**PADIATRIC PULMONOLOGY**

Pediatric pulmonology is a medical subspecialty focused on the diagnosis, treatment, and management of respiratory disorders and diseases in children. This includes conditions such as asthma, cystic fibrosis, bronchopulmonary dysplasia, and other congenital or acquired lung diseases.

**WHO IS A PEDIATRIC PUMONOLOGIST**

A pediatric pulmonologist is a doctor who specializes in breathing and lung problems in children. They can help your child, and your family learn to manage the symptoms of breathing problems, prevent complications, and improve their quality of life. A pediatric pulmonologist will evaluate your child’s lung health, diagnose the cause of the breathing problem, and develop a treatment plan.

## **What Does a Pediatric Pulmonologist Do?**

The pediatric pulmonologist will use a wide variety of tests. These include:

* Pulmonary function test to find out how well your child’s lungs are working. This is a set of painless measurements taken while your child does simple tasks like breathing out, walking, or riding a stationary bike.
* Allergy testing, bloodwork, or a chest X-ray to look for what’s causing your child’s symptoms.

To help ease your child’s breathing symptoms, a pediatric pulmonologist can prescribe medication and/or treatments to be taken at home. These may include:

* Nebulizer treatments (your child will breathe in a mist that sends medicine directly to their lungs)
* Chest physiotherapy (a treatment that vibrates the rib cage to break up mucus in the lungs)
* CPAP and BiPAP (machines that use air to assist with breathing during sleep)
* Home oxygen or ventilator use for severe cases

## **What Conditions Does a Pediatric Pulmonologist Treat?**

Pediatric pulmonologists see children with a broad range of breathing problems, from premature newborns with chronic lung disease to children with rare disorders like interstitial lung disease.

Your child may need to see a pediatric pulmonologist because a part of their respiratory system (lungs, airway, nose, mouth, throat) developed in a way that makes it hard to breathe. Or the pulmonologist could be part of a medical team that treats your child for frequent aspiration (inhaling small amounts of food, drinks, saliva, or vomit) because of an aerodigestive disorder (a condition that affects your child’s airways and upper digestive tracts).

In asthma, your child’s lungs and airway's reaction to certain triggers by swelling and making extra mucus. This can cause wheezing, coughing, or difficulty breathing. Triggers vary but may include viruses (like colds or the flu), smoke, dust, mildew, pollen, animals, exercise, or stress. A pediatric pulmonologist can prescribe oral medications to help ease symptoms, provide inhalers and/or nebulizers that deliver medicine directly to the lungs, and order allergy testing to find out what’s triggering asthma flares.

Apnea

Short pauses in breathing can be normal for newborns. However, longer pauses are a concern. Infant apnea is a pause that lasts 20 seconds or longer (15 seconds in premature infants) or causes your baby’s heart rate to slow or their skin to turn pale or blue. Depending on the underlying cause, medication can sometimes help reduce the frequency of apnea.

Your family can learn CPR and how to make your child’s surroundings safer. The doctor may prescribe an infant apnea monitor. It can alert you if your baby stops breathing. Most babies improve over time. They only need treatment until the apnea goes away.

Sleep disorders

Obstructive sleep apnea is a pattern of brief pauses in breathing because the airway collapses during sleep. Oxygen levels drop and your child wakes up just long enough to start breathing normally again. The child may not be aware that they’re waking during the night, but they might have trouble concentrating, behavior problems, or daytime sleepiness. Central sleep apnea occurs when your child stops breathing during sleep because their brain doesn’t cue their body to breathe. Pediatric pulmonologists can diagnose sleep disorders and, when needed, prescribe a CPAP or BiPAP machine to help your child breathe normally during sleep.

Cystic Fibrosis

Cystic fibrosis (CF) is an inherited disease that leads to the production of thick mucus, clogging airways and trapping germs in the lungs. As a result, children with CF may have frequent lung infections and trouble breathing. Treatments include inhaled medications and daily at-home therapy to break up mucus and clear airways.

## **Procedures performed**

A pediatric pulmonologist performs procedures either to diagnose or treat the condition. These procedures may range from lung function test to sweat chloride test along with performing therapeutic procedures like chest physiotherapy. This is a glance at the procedures performed by them:

**Flexible fiberoptic bronchoscopy & bronchoalveolar lavage**

**Bronchoscopy** is a test to view the airways and diagnose lung disease. The flexible bronchoscope is inserted through the nose and goes into the throat and into the airways and lungs.

Usually, an anesthetic will be sprayed to reduce any pain or uncomfortableness.

**Lung Function Test**

It is a series of breathing tests designed to measure the amount of air in the lungs, and how well the lungs function in moving air in and out of the lungs. These are easy, non-invasive and painless tests which require children to follow simple directions.

Lung function tests include spirometry, body plethysmography, DLCO, exercise challenge test, methacholine challenge test, FeNO. Each exercise will be explained and demonstrated, and your child will be coached all through the way.

**Sweat Chloride test**

A sweat test measures the amount of chloride in the child’s sweat. It is the best way of checking for a health problem called cystic fibrosis (CF).

In this test, a special machine is used to release sweat from the child’s forearm. The sweat is then tested in the lab to measure the chloride level.

**Skin prick allergy test**

A skin prick allergy test is used to find which allergy-causing substances (allergens) are causing allergy symptoms in your child.

These tests may be able to confirm whether your child is allergic to a particular substance that he/she touches.

In general, these tests are most reliable for diagnosing allergies to airborne particles such as pollen, pet dander and dust mites.

**Common and Chronic Respiratory Diseases**

* Asthma
* Reactive Airway Disease (RAD)
* Bronchitis (acute and chronic)
* Bronchiolitis (commonly RSV-related)
* Pneumonia (bacterial, viral, fungal)
* Cystic Fibrosis
* Bronchopulmonary Dysplasia (BPD) / Chronic Lung Disease of Infancy
* Chronic Respiratory Failure / Insufficiency
* Wheezing disorders
* Chronic cough

## Congenital and Structural Abnormalities

* Congenital lung abnormalities (e.g., congenital cystic adenomatoid malformation)
* Congenital airway anomalies (tracheomalacia, bronchomalacia, laryngomalacia)
* Chest wall deformities
* Airway malacia and stenosis

## Rare and Diffuse Lung Diseases

* Children’s Interstitial Lung Disease (chILD)
* Bronchiolitis Obliterans
* Pulmonary fibrosis
* Neuroendocrine cell hyperplasia of infancy (NEHI)
* Alveolar hemorrhage syndromes

## Infectious Diseases

* Respiratory Syncytial Virus (RSV) infection
* Pertussis (Whooping cough)
* Tuberculosis (TB)
* Other viral and bacterial lower respiratory tract infections

## Neuromuscular and Functional Disorders

* Neuromuscular respiratory diseases (e.g., muscular dystrophy, spinal muscular atrophy)
* Muscle weakness affecting breathing and cough
* Sleep-disordered breathing (obstructive and central sleep apnea)
* Apnea of prematurity and infant apnea

## Airway and Aerodigestive Disorders

* Pulmonary aspiration syndromes (due to swallowing dysfunction or gastroesophageal reflux)
* Aerodigestive disorders involving combined airway and digestive tract issues

## Vascular and Hematologic-Related Pulmonary Diseases

* Pulmonary hypertension (including secondary to congenital heart disease or chronic lung disease)
* Hereditary hemorrhagic telangiectasia (HHT)
* Pulmonary complications of sickle cell disease

## Other Important Conditions

* Primary ciliary dyskinesia (PCD)
* Non-cystic fibrosis bronchiectasis
* Respiratory failure requiring ventilator support or tracheostomy
* Subglottic stenosis
* Acute respiratory distress syndrome
* Laryngeal web
* Esophageal atresia
* Tracheoesophageal fistula
* Alpha 1 antitrypsin deficiency
* Alveolar capillary dysplasia
* Asbestosis
* Atrioventricular septal defects
* Bronchopulmonary sequestration
* Chylothorax
* Congenital diaphragmatic hernia
* Congenital lobar emphysema
* Diaphragmatic paralysis
* Hemoptysis
* Hypoplastic left heart syndrome
* Parapneumonic effusion and empyema
* Pectus carinatum
* Pectus excavatum
* Truncus arteriosus
* Pleural effusion
* Pneumococcal pneumonia
* Pulmonary contusion
* Pulmonary stenosis
* Pulmonary stenosis
* Spontaneous pneumomediastinum
* Thoracic trauma
* Sudden infant death syndrome
* Transient tachypnea of newborn
* Total anomalous pulmonary venous return
* Sarcoidosis
* Aspirin exacerbated respiratory disease
* Community acquired pneumonia
* Bronchial atresia

**Current Procedural Terminology (CPT) codes**

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| Disease/Condition | Common CPT Codes (Evaluation & Treatment) |
| Asthma | 99202-99205 (new patient E/M), 99212-99215 (established patient E/M) |
| Reactive Airway Disease (RAD) | Same as asthma: 99202-99205, 99212-99215 |
| Bronchitis (acute and chronic) | 99202-99205, 99212-99215 |
| Bronchiolitis (commonly RSV-related) | 99202-99205, 99212-99215, 90380 (RSV monoclonal antibody administration) |
| Pneumonia (bacterial, viral, fungal) | 99202-99205, 99212-99215, 71045-71048 (Chest X-ray) |
| Cystic Fibrosis | 99202-99205, 99212-99215, 94010 (pulmonary function test), 94640 (inhalation therapy) |
| Bronchopulmonary Dysplasia (BPD) / Chronic Lung Disease of Infancy | 99202-99205, 99212-99215, 94010, 94640 |
| Chronic Respiratory Failure / Insufficiency | 99202-99205, 99212-99215, 94010, 94640, 94760 (CPAP ventilation management) |
| Wheezing disorders | 99202-99205, 99212-99215 |
| Chronic cough | 99202-99205, 99212-99215 |

## Congenital and Structural Abnormalities

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| Condition | Relevant CPT Codes |
| Congenital lung abnormalities (e.g., CCAM) | 99202-99215 (E/M), 71250 (Chest CT) |
| Congenital airway anomalies (tracheomalacia, bronchomalacia, laryngomalacia) | 31622 (Flexible bronchoscopy), 31575 (Laryngoscopy) |
| Chest wall deformities | 99202-99215 (E/M), 20610 (Muscle injection) |
| Airway malacia and stenosis | 31622 (Bronchoscopy), 31628 (Bronchoscopy with dilation) |

## 2. Rare and Diffuse Lung Diseases

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| Condition | Relevant CPT Codes |
| Children’s Interstitial Lung Disease (chILD) | 94010 (Spirometry), 71250 (Chest CT) |
| Bronchiolitis Obliterans | 94010, 31622 (Bronchoscopy) |
| Pulmonary fibrosis | 94010, 71250 |
| Surfactant metabolism disorders | 94010, genetic testing (no specific CPT) |
| Neuroendocrine cell hyperplasia of infancy (NEHI) | 94010, 71250 |
| Alveolar hemorrhage syndromes | 31622 (Bronchoscopy with lavage) |

## 3. Infectious Diseases

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| Condition | Relevant CPT Codes |
| Respiratory Syncytial Virus (RSV) infection | 90380 (RSV monoclonal antibody administration) |
| Pertussis (Whooping cough) | 99202-99215 (E/M), 94640 (Inhalation therapy) |
| Tuberculosis (TB) | 99202-99215 (E/M), 86580 (TB skin test) |
| Other viral and bacterial lower respiratory infections | 99202-99215 (E/M) |

## 4. Neuromuscular and Functional Disorders

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| Condition | Relevant CPT Codes |
| Neuromuscular respiratory diseases | 99202-99215 (E/M), 94660 (CPAP management) |
| Muscle weakness affecting breathing and cough | 99202-99215 (E/M) |
| Sleep-disordered breathing (obstructive and central sleep apnea) | 95807-95811 (Polysomnography) |
| Apnea of prematurity and infant apnea | 99202-99215 (E/M), 94762 (CO2 monitoring) |

## 5. Airway and Aerodigestive Disorders

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| Condition | Relevant CPT Codes |
| Pulmonary aspiration syndromes | 99202-99215 (E/M), 31622 (Bronchoscopy) |
| Aerodigestive disorders | 31622 (Bronchoscopy), 31575 (Laryngoscopy) |

## 6. Vascular and Hematologic-Related Pulmonary Diseases

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| Condition | Relevant CPT Codes |
| Pulmonary hypertension | 99202-99215 (E/M), 93306 (Echocardiography) |
| Hereditary hemorrhagic telangiectasia (HHT) | 99202-99215 (E/M) |
| Pulmonary complications of sickle cell disease | 99202-99215 (E/M), 71250 (Chest CT) |

## Other Important Conditions

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| Condition | Relevant CPT Codes |
| Primary ciliary dyskinesia (PCD) | 94010 (Spirometry), 31622 (Bronchoscopy) |
| Non-cystic fibrosis bronchiectasis | 94010, 71250 |
| Respiratory failure requiring ventilator support or tracheostomy | 94660 (CPAP/ventilation management), 31600 (Tracheostomy) |
| Pulmonary complications of childhood cancer and stem cell transplantation | 99202-99215 (E/M), 71250 (Chest CT) |

## Additional Common CPT Codes for Pediatric Pulmonary Care

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| CPT Code | Description |
| 94010 | Spirometry pulmonary function test |
| 94640 | Inhalation treatment for acute airway obstruction |
| 31622 | Flexible bronchoscopy, diagnostic |
| 31575 | Laryngoscopy, flexible or rigid |
| 90380 | RSV monoclonal antibody administration |
| 94660 | CPAP ventilation management |
| 95810-95811 | Polysomnography (sleep study) |
| 71250 | Chest CT without contrast |
| 86580 | Tuberculosis skin test |

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| Condition | ICD-10 Code(s) | Notes / Additional Info |
| Subglottic stenosis | J38.6 | Stenosis of subglottic larynx |
| Acute respiratory distress syndrome (ARDS) | J80 | Acute respiratory distress syndrome |
| Laryngeal web | Q31.1 | Congenital laryngeal web |
| Esophageal atresia | Q39.0 | Esophageal atresia without fistula |
| Tracheoesophageal fistula | Q39.1 | Tracheoesophageal fistula |
| Alpha-1 antitrypsin deficiency | E88.01 | Alpha-1-antitrypsin deficiency |
| Alveolar capillary dysplasia | P28.89 | Other specified respiratory conditions of newborn |
| Asbestosis | J61 | Pneumoconiosis due to asbestos and other mineral fibers |
| Atrioventricular septal defects | Q21.2 | Atrioventricular septal defect |
| Bronchopulmonary sequestration | Q33.2 | Congenital bronchopulmonary sequestration |
| Chylothorax | I31.3 | Chylothorax |
| Congenital diaphragmatic hernia | Q79.0 | Congenital diaphragmatic hernia |
| Congenital lobar emphysema | Q33.4 | Congenital lobar emphysema |
| Diaphragmatic paralysis | J98.81 | Diaphragmatic paralysis |
| Hemoptysis | R04.2 | Hemoptysis |
| Hypoplastic left heart syndrome | Q23.4 | Hypoplastic left heart syndrome |
| Parapneumonic effusion and empyema | J90 (pleural effusion), J86 (empyema) | J90 for pleural effusion, J86 for empyema |
| Pectus carinatum | Q67.6 | Pectus carinatum |
| Pectus excavatum | Q67.5 | Pectus excavatum |
| Truncus arteriosus | Q20.0 | Truncus arteriosus |
| Pleural effusion | J90 | Pleural effusion, not elsewhere classified |
| Pneumococcal pneumonia | J13 | Pneumonia due to Streptococcus pneumoniae |
| Pulmonary contusion | S27.2 | Pulmonary contusion |
| Pulmonary stenosis | Q22.1 | Pulmonary valve stenosis |
| Spontaneous pneumomediastinum | J98.2 | Mediastinal emphysema (pneumomediastinum) |
| Thoracic trauma | S27.9 | Injury of thorax, unspecified |
| Sudden infant death syndrome (SIDS) | R95 | Sudden infant death syndrome |
| Transient tachypnea of newborn | P22.1 | Transient tachypnea of newborn |
| Total anomalous pulmonary venous return | Q26.4 | Total anomalous pulmonary venous connection |
| Sarcoidosis | D86 | Sarcoidosis |
| Aspirin exacerbated respiratory disease | J45.50 | Asthma with aspirin sensitivity |
| Community acquired pneumonia | J18.9 | Pneumonia, unspecified organism |
| Bronchial atresia | Q33.3 | Bronchial atresia |

**ICD-10 CODES**

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| Disease/Condition | ICD-10 Code(s) | Notes |
| Acute Bronchitis | J20.0–J20.9 | Includes specific organisms like RSV, Mycoplasma, etc. |
| Acute Bronchiolitis | J21.0–J21.9 | Often viral, e.g., RSV; unspecified acute bronchiolitis included |
| Pneumonia | J12–J18 | J18.9 = Pneumonia, unspecified organism |
| Whooping Cough (Pertussis) | A37.0–A37.9 | Includes with/without pneumonia variants |
| Congenital Malformations of Respiratory System | Q30–Q34 | Includes congenital laryngeal, tracheal, bronchial anomalies |
| Interstitial Lung Diseases of Childhood | J84.848 | Other interstitial lung diseases of childhood |
| Chronic Lung Disease (e.g., Bronchopulmonary Dysplasia) | P27.0–P27.9 | Neonatal chronic respiratory diseases (not in search but standard) |
| Cystic Fibrosis | E84 | Common pediatric chronic pulmonary disease (not in search but standard) |
| Tuberculosis of Lung | A15.0–A16.9 | Pulmonary tuberculosis codes |
| Other Disorders of Lung | J98.4 | Includes acute interstitial pneumonitis, pulmonary insufficiency |
| Respiratory Symptoms | R05 (cough), R06.2 (wheezing), R06.00–R06.09 (shortness of breath), R07.1 (chest pain on breathing) | Symptom codes used in pediatrics |
| Allergic Rhinitis | J30.0–J30.9 | Includes seasonal and animal dander allergies |
| Viral and Bacterial Pharyngitis | J02.0–J02.9 | Streptococcal and other acute pharyngitis |

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| **ICD 10 Code** | **Classification** |
| A0103 | Typhoid pneumonia |
| A0222 | Salmonella pneumonia |
| A065 | Amebic lung abscess |
| A15 | Respiratory tuberculosis |
| A150 | Tuberculosis of lung |
| A151 | Tuberculosis of lung, confirmed by culture only |
| A152 | Tuberculosis of lung, confirmed histologically |
| A153 | Tuberculosis of lung, confirmed by unspecified means |
| A155 | Tuberculosis of larynx, trachea and bronchus |
| A157 | Primary respiratory tuberculosis |
| A160 | Tuberculosis of lung, bacteriologically and histologically negative |
| A161 | Tuberculosis of lung, bacteriological and histological examination not done |
| A162 | Tuberculosis of lung, without mention of bacteriological or histological confirmation |
| A164 | Tuberculosis of larynx, trachea and bronchus, without mention of bacteriological or histological confirmation |
| A3700 | Whooping cough due to Bordetella pertussis without pneumonia |
| A3701 | Whooping cough due to Bordetella pertussis with pneumonia |
| A3710 | Whooping cough due to Bordetella parapertussis without pneumonia |
| A3711 | Whooping cough due to Bordetella parapertussis with pneumonia |
| A3780 | Whooping cough due to other Bordetella species without pneumonia |
| A3781 | Whooping cough due to other Bordetella species with pneumonia |
| A3790 | Whooping cough, unspecified species without pneumonia |
| A3791 | Whooping cough, unspecified species with pneumonia |
| A403 | Sepsis due to Streptococcus pneumoniae |
| A5004 | Early congenital syphilitic pneumonia |
| A5272 | Syphilis of lung and bronchus |
| A5484 | Gonococcal pneumonia |
| B012 | Varicella pneumonia |
| B052 | Measles complicated by pneumonia |
| B0681 | Rubella pneumonia |
| B206 | HIV disease resulting in Pneumocystis jirovecii pneumonia |
| B671 | Echinococcus granulosus infection of lung |
| B7781 | Ascariasis pneumonia |
| B953 | Streptococcus pneumoniae as the cause of diseases classified elsewhere |
| B960 | Mycoplasma pneumoniae [M. pneumoniae] as the cause of diseases classified elsewhere |
| B961 | Klebsiella pneumoniae [K. pneumoniae] as the cause of diseases classified elsewhere |
| C33 | Malignant neoplasm of trachea |
| C34 | Malignant neoplasm of bronchus and lung |
| C340 | Malignant neoplasm of main bronchus |
| C3400 | Malignant neoplasm of unspecified main bronchus |
| C3401 | Malignant neoplasm of right main bronchus |
| C3402 | Malignant neoplasm of left main bronchus |
| C341 | Malignant neoplasm of upper lobe, bronchus or lung |
| C3410 | Malignant neoplasm of upper lobe, unspecified bronchus or lung |
| C3411 | Malignant neoplasm of upper lobe, right bronchus or lung |
| C3412 | Malignant neoplasm of upper lobe, left bronchus or lung |
| C342 | Malignant neoplasm of middle lobe, bronchus or lung |
| C343 | Malignant neoplasm of lower lobe, bronchus or lung |
| C3430 | Malignant neoplasm of lower lobe, unspecified bronchus or lung |
| C3431 | Malignant neoplasm of lower lobe, right bronchus or lung |
| C3432 | Malignant neoplasm of lower lobe, left bronchus or lung |
| C348 | Malignant neoplasm of overlapping sites of bronchus and lung |
| C3480 | Malignant neoplasm of overlapping sites of unspecified bronchus and lung |
| C3481 | Malignant neoplasm of overlapping sites of right bronchus and lung |
| C3482 | Malignant neoplasm of overlapping sites of left bronchus and lung |
| C349 | Malignant neoplasm of unspecified part of bronchus or lung |
| C3490 | Malignant neoplasm of unspecified part of unspecified bronchus or lung |
| C3491 | Malignant neoplasm of unspecified part of right bronchus or lung |
| C3492 | Malignant neoplasm of unspecified part of left bronchus or lung |
| C390 | Malignant neoplasm of upper respiratory tract, part unspecified |
| C399 | Malignant neoplasm of lower respiratory tract, part unspecified |
| C465 | Kaposi’s sarcoma of lung |
| C4650 | Kaposi’s sarcoma of unspecified lung |
| C4651 | Kaposi’s sarcoma of right lung |
| C4652 | Kaposi’s sarcoma of left lung |
| C780 | Secondary malignant neoplasm of lung |
| C7800 | Secondary malignant neoplasm of unspecified lung |
| C7801 | Secondary malignant neoplasm of right lung |
| C7802 | Secondary malignant neoplasm of left lung |
| C7A090 | Malignant carcinoid tumor of the bronchus and lung |
| D021 | Carcinoma in situ of trachea |
| D022 | Carcinoma in situ of bronchus and lung |
| D0220 | Carcinoma in situ of unspecified bronchus and lung |
| D0221 | Carcinoma in situ of right bronchus and lung |
| D0222 | Carcinoma in situ of left bronchus and lung |
| D142 | Benign neoplasm of trachea |
| D143 | Benign neoplasm of bronchus and lung |
| D1430 | Benign neoplasm of unspecified bronchus and lung |
| D1431 | Benign neoplasm of right bronchus and lung |
| D1432 | Benign neoplasm of left bronchus and lung |
| D381 | Neoplasm of uncertain behavior of trachea, bronchus and lung |
| D3A090 | Benign carcinoid tumor of the bronchus and lung |
| D860 | Sarcoidosis of lung |
| D862 | Sarcoidosis of lung with sarcoidosis of lymph nodes |
| F18 | Inhalant related disorders |
| F181 | Inhalant abuse |
| F1810 | Inhalant abuse, uncomplicated |
| F1811 | Inhalant abuse, in remission |
| F1812 | Inhalant abuse with intoxication |
| F18120 | Inhalant abuse with intoxication, uncomplicated |
| F18121 | Inhalant abuse with intoxication delirium |
| F18129 | Inhalant abuse with intoxication, unspecified |
| F1814 | Inhalant abuse with inhalant-induced mood disorder |
| F1815 | Inhalant abuse with inhalant-induced psychotic disorder |
| F18150 | Inhalant abuse with inhalant-induced psychotic disorder with delusions |
| F18151 | Inhalant abuse with inhalant-induced psychotic disorder with hallucinations |
| F18159 | Inhalant abuse with inhalant-induced psychotic disorder, unspecified |
| F1817 | Inhalant abuse with inhalant-induced dementia |
| F1818 | Inhalant abuse with other inhalant-induced disorders |
| F18180 | Inhalant abuse with inhalant-induced anxiety disorder |
| F18188 | Inhalant abuse with other inhalant-induced disorder |
| F1819 | Inhalant abuse with unspecified inhalant-induced disorder |
| F182 | Inhalant dependence |
| F1820 | Inhalant dependence, uncomplicated |
| F1821 | Inhalant dependence, in remission |
| F1822 | Inhalant dependence with intoxication |
| F18220 | Inhalant dependence with intoxication, uncomplicated |
| F18221 | Inhalant dependence with intoxication delirium |
| F18229 | Inhalant dependence with intoxication, unspecified |
| F1824 | Inhalant dependence with inhalant-induced mood disorder |
| F1825 | Inhalant dependence with inhalant-induced psychotic disorder |
| F18250 | Inhalant dependence with inhalant-induced psychotic disorder with delusions |
| F18251 | Inhalant dependence with inhalant-induced psychotic disorder with hallucinations |
| F18259 | Inhalant dependence with inhalant-induced psychotic disorder, unspecified |
| F1827 | Inhalant dependence with inhalant-induced dementia |
| F1828 | Inhalant dependence with other inhalant-induced disorders |
| F18280 | Inhalant dependence with inhalant-induced anxiety disorder |
| F18288 | Inhalant dependence with other inhalant-induced disorder |
| F1829 | Inhalant dependence with unspecified inhalant-induced disorder |
| F189 | Inhalant use, unspecified |
| F1890 | Inhalant use, unspecified, uncomplicated |
| F1891 | Inhalant use, unspecified, in remission |
| F1892 | Inhalant use, unspecified with intoxication |
| F18920 | Inhalant use, unspecified with intoxication, uncomplicated |
| F18921 | Inhalant use, unspecified with intoxication with delirium |
| F18929 | Inhalant use, unspecified with intoxication, unspecified |
| F1894 | Inhalant use, unspecified with inhalant-induced mood disorder |
| F1895 | Inhalant use, unspecified with inhalant-induced psychotic disorder |
| F18950 | Inhalant use, unspecified with inhalant-induced psychotic disorder with delusions |
| F18951 | Inhalant use, unspecified with inhalant-induced psychotic disorder with hallucinations |
| F18959 | Inhalant use, unspecified with inhalant-induced psychotic disorder, unspecified |
| F1897 | Inhalant use, unspecified with inhalant-induced persisting dementia |
| F1898 | Inhalant use, unspecified with other inhalant-induced disorders |
| F18980 | Inhalant use, unspecified with inhalant-induced anxiety disorder |
| F18988 | Inhalant use, unspecified with other inhalant-induced disorder |
| F1899 | Inhalant use, unspecified with unspecified inhalant-induced disorder |
| G001 | Pneumococcal meningitis |
| G4732 | High altitude periodic breathing |
| I2723 | Pulmonary hypertension due to lung diseases and hypoxia |
| J09010 | Influenza due to identified avian influenza virus with identified avian influenza pneumonia |
| J09018 | Influenza due to identified avian influenza virus with other specified type of pneumonia |
| J09019 | Influenza due to identified avian influenza virus with unspecified type of pneumonia |
| J09110 | Influenza due to identified novel H1N1 influenza virus with identified novel H1N1 influenza pneumonia |
| J09118 | Influenza due to identified novel H1N1 influenza virus with other specified type of pneumonia |

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| J09119 | Influenza due to identified novel H1N1 influenza virus with unspecified type of pneumonia |
| J09-J18 | Influenza and pneumonia |
| J09X1 | Influenza due to identified novel influenza A virus with pneumonia |
| J100 | Influenza due to other identified influenza virus with pneumonia |
| J1000 | Influenza due to other identified influenza virus with unspecified type of pneumonia |
| J1001 | Influenza due to other identified influenza virus with the same other identified influenza virus pneumonia |
| J1008 | Influenza due to other identified influenza virus with other specified pneumonia |
| J110 | Influenza due to unidentified influenza virus with pneumonia |
| J1100 | Influenza due to unidentified influenza virus with unspecified type of pneumonia |
| J1108 | Influenza due to unidentified influenza virus with specified pneumonia |
| J12 | Viral pneumonia, not elsewhere classified |
| J120 | Adenoviral pneumonia |
| J121 | Respiratory syncytial virus pneumonia |
| J122 | Parainfluenza virus pneumonia |
| J123 | Human metapneumovirus pneumonia |
| J128 | Other viral pneumonia |
| J1281 | Pneumonia due to SARS-associated coronavirus |
| J1282 | Pneumonia due to coronavirus disease 2019 |
| J1289 | Other viral pneumonia |
| J129 | Viral pneumonia, unspecified |
| J13 | Pneumonia due to Streptococcus pneumoniae |
| J14 | Pneumonia due to Hemophilus influenzae |
| J15 | Bacterial pneumonia, not elsewhere classified |
| J150 | Pneumonia due to Klebsiella pneumoniae |
| J151 | Pneumonia due to Pseudomonas |
| J152 | Pneumonia due to staphylococcus |
| J1520 | Pneumonia due to staphylococcus, unspecified |
| J1521 | Pneumonia due to staphylococcus aureus |
| J15211 | Pneumonia due to Methicillin susceptible Staphylococcus aureus |
| J15212 | Pneumonia due to Methicillin resistant Staphylococcus aureus |
| J1529 | Pneumonia due to other staphylococcus |
| J153 | Pneumonia due to streptococcus, group B |
| J154 | Pneumonia due to other streptococci |
| J155 | Pneumonia due to Escherichia coli |
| J156 | Pneumonia due to other Gram-negative bacteria |
| J1561 | Pneumonia due to Acinetobacter baumannii |
| J1569 | Pneumonia due to other Gram-negative bacteria |
| J157 | Pneumonia due to Mycoplasma pneumoniae |
| J158 | Pneumonia due to other specified bacteria |
| J159 | Unspecified bacterial pneumonia |
| J16 | Pneumonia due to other infectious organisms, not elsewhere classified |
| J160 | Chlamydial pneumonia |
| J168 | Pneumonia due to other specified infectious organisms |
| J17 | Pneumonia in diseases classified elsewhere |
| J170 | Pneumonia in bacterial diseases classified elsewhere |
| J171 | Pneumonia in viral diseases classified elsewhere |
| J172 | Pneumonia in mycoses |
| J173 | Pneumonia in parasitic diseases |
| J178 | Pneumonia in other diseases classified elsewhere |
| J18 | Pneumonia, unspecified organism |
| J181 | Lobar pneumonia, unspecified organism |
| J182 | Hypostatic pneumonia, unspecified organism |
| J188 | Other pneumonia, unspecified organism |
| J189 | Pneumonia, unspecified organism |
| J200 | Acute bronchitis due to Mycoplasma pneumoniae |
| J30-J39 | Other diseases of upper respiratory tract |
| J39 | Other diseases of upper respiratory tract |
| J393 | Upper respiratory tract hypersensitivity reaction, site unspecified |
| J398 | Other specified diseases of upper respiratory tract |
| J399 | Disease of upper respiratory tract, unspecified |
| J44 | Other chronic obstructive pulmonary disease |
| J440 | Chronic obstructive pulmonary disease with (acute) lower respiratory infection |
| J441 | Chronic obstructive pulmonary disease with (acute) exacerbation |
| J448 | Other specified chronic obstructive pulmonary disease |
| J4489 | Other specified chronic obstructive pulmonary disease |
| J449 | Chronic obstructive pulmonary disease, unspecified |
| J45 | Asthma |
| J450 | Predominantly allergic asthma |
| J451 | Nonallergic asthma |
| J452 | Mild intermittent asthma |
| J4520 | Mild intermittent asthma, uncomplicated |
| J4521 | Mild intermittent asthma with (acute) exacerbation |
| J4522 | Mild intermittent asthma with status asthmaticus |
| J453 | Mild persistent asthma |
| J4530 | Mild persistent asthma, uncomplicated |
| J4531 | Mild persistent asthma with (acute) exacerbation |
| J4532 | Mild persistent asthma with status asthmaticus |
| J454 | Moderate persistent asthma |
| J4540 | Moderate persistent asthma, uncomplicated |
| J4541 | Moderate persistent asthma with (acute) exacerbation |
| J4542 | Moderate persistent asthma with status asthmaticus |
| J455 | Severe persistent asthma |
| J4550 | Severe persistent asthma, uncomplicated |
| J4551 | Severe persistent asthma with (acute) exacerbation |
| J4552 | Severe persistent asthma with status asthmaticus |
| J458 | Mixed asthma |
| J459 | Other and unspecified asthma |
| J4590 | Unspecified asthma |
| J45901 | Unspecified asthma with (acute) exacerbation |
| J45902 | Unspecified asthma with status asthmaticus |
| J45909 | Unspecified asthma, uncomplicated |
| J4599 | Other asthma |
| J45991 | Cough variant asthma |
| J45998 | Other asthma |
| J46 | Status asthmaticus |
| J4A | Chronic lung allograft dysfunction |
| J4A8 | Other chronic lung allograft dysfunction |
| J4A9 | Chronic lung allograft dysfunction, unspecified |
| J60-J70 | Lung diseases due to external agents |
| J630 | Aluminosis (of lung) |
| J631 | Bauxite fibrosis (of lung) |
| J633 | Graphite fibrosis (of lung) |
| J66 | Airway disease due to specific organic dust |
| J668 | Airway disease due to other specific organic dusts |
| J670 | Farmer’s lung |
| J672 | Bird fancier’s lung |
| J674 | Maltworker’s lung |
| J675 | Mushroom-worker’s lung |
| J676 | Maple-bark-stripper’s lung |
| J677 | Air conditioner and humidifier lung |
| J68 | Respiratory conditions due to inhalation of chemicals, gases, fumes and vapors |
| J690 | [Pneumonitis](https://www.pulmonologyadvisor.com/ddi/pneumonitis/) due to inhalation of food and vomit |
| J691 | Pneumonitis due to inhalation of oils and essences |
| J698 | Pneumonitis due to inhalation of other solids and liquids |
| J702 | Acute drug-induced interstitial lung disorders |
| J703 | Chronic drug-induced interstitial lung disorders |
| J704 | Drug-induced interstitial lung disorders, unspecified |
| J705 | Respiratory conditions due to smoke inhalation |
| J8281 | Chronic eosinophilic pneumonia |
| J8282 | Acute eosinophilic pneumonia |
| J8283 | [Eosinophilic asthma](https://www.pulmonologyadvisor.com/ddi/eosinophilic-asthma/) |
| J8411 | Idiopathic interstitial pneumonia |
| J84111 | Idiopathic interstitial pneumonia, not otherwise specified |
| J84115 | Respiratory bronchiolitis interstitial lung disease |
| J84116 | Cryptogenic organizing pneumonia |
| J84117 | Desquamative interstitial pneumonia |
| J84170 | Interstitial lung disease with progressive fibrotic phenotype in diseases classified elsewhere |
| J842 | Lymphoid interstitial pneumonia |
| J8483 | Surfactant mutations of the lung |
| J8484 | Other interstitial lung diseases of childhood |
| J84848 | Other interstitial lung diseases of childhood |
| J85 | Abscess of lung and mediastinum |
| J850 | Gangrene and necrosis of lung |
| J851 | Abscess of lung with pneumonia |
| J852 | Abscess of lung without pneumonia |
| J85-J86 | Suppurative and necrotic conditions of lower respiratory tract |
| J9584 | Transfusion-related acute lung injury (TRALI) |
| J9585 | Complication of respirator [ventilator] |
| J95850 | Mechanical complication of respirator |
| J95851 | Ventilator associated pneumonia |
| J95859 | Other complication of respirator [ventilator] |
| J980 | Diseases of bronchus, not elsewhere classified |
| J9809 | Other diseases of bronchus, not elsewhere classified |
| J984 | Other disorders of lung |
| J990 | Rheumatoid lung disease |
| M051 | Rheumatoid lung disease with rheumatoid arthritis |
| M0510 | Rheumatoid lung disease with rheumatoid arthritis of unspecified site |
| M0511 | Rheumatoid lung disease with rheumatoid arthritis of shoulder |
| M05111 | Rheumatoid lung disease with rheumatoid arthritis of right shoulder |
| M05112 | Rheumatoid lung disease with rheumatoid arthritis of left shoulder |
| M05119 | Rheumatoid lung disease with rheumatoid arthritis of unspecified shoulder |
| M0512 | Rheumatoid lung disease with rheumatoid arthritis of elbow |
| M05121 | Rheumatoid lung disease with rheumatoid arthritis of right elbow |
| M05122 | Rheumatoid lung disease with rheumatoid arthritis of left elbow |
| M05129 | Rheumatoid lung disease with rheumatoid arthritis of unspecified elbow |
| M0513 | Rheumatoid lung disease with rheumatoid arthritis of wrist |
| M05131 | Rheumatoid lung disease with rheumatoid arthritis of right wrist |
| M05132 | Rheumatoid lung disease with rheumatoid arthritis of left wrist |
| M05139 | Rheumatoid lung disease with rheumatoid arthritis of unspecified wrist |
| M0514 | Rheumatoid lung disease with rheumatoid arthritis of hand |
| M05141 | Rheumatoid lung disease with rheumatoid arthritis of right hand |
| M05142 | Rheumatoid lung disease with rheumatoid arthritis of left hand |
| M05149 | Rheumatoid lung disease with rheumatoid arthritis of unspecified hand |
| M0515 | Rheumatoid lung disease with rheumatoid arthritis of hip |
| M05151 | Rheumatoid lung disease with rheumatoid arthritis of right hip |
| M05152 | Rheumatoid lung disease with rheumatoid arthritis of left hip |
| M05159 | Rheumatoid lung disease with rheumatoid arthritis of unspecified hip |
| M0516 | Rheumatoid lung disease with rheumatoid arthritis of knee |
| M05161 | Rheumatoid lung disease with rheumatoid arthritis of right knee |
| M05162 | Rheumatoid lung disease with rheumatoid arthritis of left knee |
| M05169 | Rheumatoid lung disease with rheumatoid arthritis of unspecified knee |
| M0517 | Rheumatoid lung disease with rheumatoid arthritis of ankle and foot |
| M05171 | Rheumatoid lung disease with rheumatoid arthritis of right ankle and foot |
| M05172 | Rheumatoid lung disease with rheumatoid arthritis of left ankle and foot |
| M05179 | Rheumatoid lung disease with rheumatoid arthritis of unspecified ankle and foot |
| M0518 | Rheumatoid lung disease, other |
| M0519 | Rheumatoid lung disease with rheumatoid arthritis of multiple sites |
| M301 | Polyarteritis with lung involvement [Churg-Strauss] |
| M3213 | Lung involvement in systemic lupus erythematosus |
| M3481 | Systemic sclerosis with lung involvement |
| M3502 | Sjogren syndrome with lung involvement |
| N80B2 | Endometriosis of lung |
| O2902 | Pressure collapse of lung due to anesthesia during pregnancy |
| O29021 | Pressure collapse of lung due to anesthesia during pregnancy, first trimester |
| O29022 | Pressure collapse of lung due to anesthesia during pregnancy, second trimester |
| O29023 | Pressure collapse of lung due to anesthesia during pregnancy, third trimester |
| O29029 | Pressure collapse of lung due to anesthesia during pregnancy, unspecified trimester |
| P23 | Congenital pneumonia |
| P230 | Congenital pneumonia due to viral agent |
| P231 | Congenital pneumonia due to Chlamydia |
| P232 | Congenital pneumonia due to staphylococcus |
| P233 | Congenital pneumonia due to streptococcus, group B |
| P234 | Congenital pneumonia due to Escherichia coli |
| P235 | Congenital pneumonia due to Pseudomonas |
| P236 | Congenital pneumonia due to other bacterial agents |
| P238 | Congenital pneumonia due to other organisms |
| P239 | Congenital pneumonia, unspecified |
| Q32 | Congenital malformations of trachea and bronchus |
| Q321 | Other congenital malformations of trachea |
| Q323 | Congenital stenosis of bronchus |
| Q324 | Other congenital malformations of bronchus |
| Q33 | Congenital malformations of lung |
| Q330 | Congenital cystic lung |
| Q331 | Accessory lobe of lung |
| Q332 | Sequestration of lung |
| Q333 | Agenesis of lung |
| Q335 | Ectopic tissue in lung |
| Q336 | Congenital hypoplasia and dysplasia of lung |
| Q338 | Other congenital malformations of lung |
| Q339 | Congenital malformation of lung, unspecified |
| R06 | Abnormalities of breathing |
| R0602 | Shortness of breath |
| R063 | Periodic breathing |
| R065 | Mouth breathing |
| R068 | Other abnormalities of breathing |
| R0689 | Other abnormalities of breathing |

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| R069 | Unspecified abnormalities of breathing |
| R071 | Chest pain on breathing |
| R91 | Abnormal findings on diagnostic imaging of lung |
| R918 | Other nonspecific abnormal finding of lung field |
| S110 | Open wound of larynx and trachea |
| S1102 | Open wound of trachea |
| S11021 | Laceration without foreign body of trachea |
| S11021A | Laceration without foreign body of trachea, initial encounter |
| S11021D | Laceration without foreign body of trachea, subsequent encounter |
| S11021S | Laceration without foreign body of trachea, sequela |
| S11022 | Laceration with foreign body of trachea |
| S11022A | Laceration with foreign body of trachea, initial encounter |
| S11022D | Laceration with foreign body of trachea, subsequent encounter |
| S11022S | Laceration with foreign body of trachea, sequela |
| S11023 | Puncture wound without foreign body of trachea |
| S11023A | Puncture wound without foreign body of trachea, initial encounter |
| S11023D | Puncture wound without foreign body of trachea, subsequent encounter |
| S11023S | Puncture wound without foreign body of trachea, sequela |
| S11024 | Puncture wound with foreign body of trachea |
| S11024A | Puncture wound with foreign body of trachea, initial encounter |
| S11024D | Puncture wound with foreign body of trachea, subsequent encounter |
| S11024S | Puncture wound with foreign body of trachea, sequela |
| S11025 | Open bite of trachea |
| S11025A | Open bite of trachea, initial encounter |
| S11025D | Open bite of trachea, subsequent encounter |
| S11025S | Open bite of trachea, sequela |
| S11029 | Unspecified open wound of trachea |
| S11029A | Unspecified open wound of trachea, initial encounter |
| S11029D | Unspecified open wound of trachea, subsequent encounter |
| S11029S | Unspecified open wound of trachea, sequela |
| S170 | Crushing injury of larynx and trachea |
| S170XXA | Crushing injury of larynx and trachea, initial encounter |
| S170XXD | Crushing injury of larynx and trachea, subsequent encounter |
| S170XXS | Crushing injury of larynx and trachea, sequela |
| S1982 | Other specified injuries of cervical trachea |
| S1982XA | Other specified injuries of cervical trachea, initial encounter |
| S1982XD | Other specified injuries of cervical trachea, subsequent encounter |
| S1982XS | Other specified injuries of cervical trachea, sequela |
| S273 | Other and unspecified injuries of lung |
| S2730 | Unspecified injury of lung |
| S27301 | Unspecified injury of lung, unilateral |
| S27301A | Unspecified injury of lung, unilateral, initial encounter |
| S27301D | Unspecified injury of lung, unilateral, subsequent encounter |
| S27301S | Unspecified injury of lung, unilateral, sequela |
| S27302 | Unspecified injury of lung, bilateral |
| S27302A | Unspecified injury of lung, bilateral, initial encounter |
| S27302D | Unspecified injury of lung, bilateral, subsequent encounter |
| S27302S | Unspecified injury of lung, bilateral, sequela |
| S27309 | Unspecified injury of lung, unspecified |
| S27309A | Unspecified injury of lung, unspecified, initial encounter |
| S27309D | Unspecified injury of lung, unspecified, subsequent encounter |
| S27309S | Unspecified injury of lung, unspecified, sequela |
| S2731 | Primary blast injury of lung |
| S27311 | Primary blast injury of lung, unilateral |
| S27311A | Primary blast injury of lung, unilateral, initial encounter |
| S27311D | Primary blast injury of lung, unilateral, subsequent encounter |
| S27311S | Primary blast injury of lung, unilateral, sequela |
| S27312 | Primary blast injury of lung, bilateral |
| S27312A | Primary blast injury of lung, bilateral, initial encounter |
| S27312D | Primary blast injury of lung, bilateral, subsequent encounter |
| S27312S | Primary blast injury of lung, bilateral, sequela |
| S27319 | Primary blast injury of lung, unspecified |
| S27319A | Primary blast injury of lung, unspecified, initial encounter |
| S27319D | Primary blast injury of lung, unspecified, subsequent encounter |
| S27319S | Primary blast injury of lung, unspecified, sequela |
| S2732 | Contusion of lung |
| S27321 | Contusion of lung, unilateral |
| S27321A | Contusion of lung, unilateral, initial encounter |
| S27321D | Contusion of lung, unilateral, subsequent encounter |
| S27321S | Contusion of lung, unilateral, sequela |
| S27322 | Contusion of lung, bilateral |
| S27322A | Contusion of lung, bilateral, initial encounter |
| S27322D | Contusion of lung, bilateral, subsequent encounter |
| S27322S | Contusion of lung, bilateral, sequela |
| S27329 | Contusion of lung, unspecified |
| S27329A | Contusion of lung, unspecified, initial encounter |
| S27329D | Contusion of lung, unspecified, subsequent encounter |
| S27329S | Contusion of lung, unspecified, sequela |
| S2733 | Laceration of lung |
| S27331 | Laceration of lung, unilateral |
| S27331A | Laceration of lung, unilateral, initial encounter |
| S27331D | Laceration of lung, unilateral, subsequent encounter |
| S27331S | Laceration of lung, unilateral, sequela |
| S27332 | Laceration of lung, bilateral |
| S27332A | Laceration of lung, bilateral, initial encounter |
| S27332D | Laceration of lung, bilateral, subsequent encounter |
| S27332S | Laceration of lung, bilateral, sequela |
| S27339 | Laceration of lung, unspecified |
| S27339A | Laceration of lung, unspecified, initial encounter |
| S27339D | Laceration of lung, unspecified, subsequent encounter |
| S27339S | Laceration of lung, unspecified, sequela |
| S2739 | Other injuries of lung |
| S27391 | Other injuries of lung, unilateral |
| S27391A | Other injuries of lung, unilateral, initial encounter |
| S27391D | Other injuries of lung, unilateral, subsequent encounter |
| S27391S | Other injuries of lung, unilateral, sequela |
| S27392 | Other injuries of lung, bilateral |
| S27392A | Other injuries of lung, bilateral, initial encounter |
| S27392D | Other injuries of lung, bilateral, subsequent encounter |
| S27392S | Other injuries of lung, bilateral, sequela |
| S27399 | Other injuries of lung, unspecified |
| S27399A | Other injuries of lung, unspecified, initial encounter |
| S27399D | Other injuries of lung, unspecified, subsequent encounter |
| S27399S | Other injuries of lung, unspecified, sequela |
| S274 | Injury of bronchus |
| S2740 | Unspecified injury of bronchus |
| S27401 | Unspecified injury of bronchus, unilateral |
| S27401A | Unspecified injury of bronchus, unilateral, initial encounter |
| S27401D | Unspecified injury of bronchus, unilateral, subsequent encounter |
| S27401S | Unspecified injury of bronchus, unilateral, sequela |
| S27402 | Unspecified injury of bronchus, bilateral |
| S27402A | Unspecified injury of bronchus, bilateral, initial encounter |
| S27402D | Unspecified injury of bronchus, bilateral, subsequent encounter |
| S27402S | Unspecified injury of bronchus, bilateral, sequela |
| S27409 | Unspecified injury of bronchus, unspecified |
| S27409A | Unspecified injury of bronchus, unspecified, initial encounter |
| S27409D | Unspecified injury of bronchus, unspecified, subsequent encounter |
| S27409S | Unspecified injury of bronchus, unspecified, sequela |
| S2741 | Primary blast injury of bronchus |
| S27411 | Primary blast injury of bronchus, unilateral |
| S27411A | Primary blast injury of bronchus, unilateral, initial encounter |
| S27411D | Primary blast injury of bronchus, unilateral, subsequent encounter |
| S27411S | Primary blast injury of bronchus, unilateral, sequela |
| S27412 | Primary blast injury of bronchus, bilateral |
| S27412A | Primary blast injury of bronchus, bilateral, initial encounter |
| S27412D | Primary blast injury of bronchus, bilateral, subsequent encounter |
| S27412S | Primary blast injury of bronchus, bilateral, sequela |
| S27419 | Primary blast injury of bronchus, unspecified |
| S27419A | Primary blast injury of bronchus, unspecified, initial encounter |
| S27419D | Primary blast injury of bronchus, unspecified, subsequent encounter |
| S27419S | Primary blast injury of bronchus, unspecified, sequela |
| S2742 | Contusion of bronchus |

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| S27421 | Contusion of bronchus, unilateral |
| S27421A | Contusion of bronchus, unilateral, initial encounter |
| S27421D | Contusion of bronchus, unilateral, subsequent encounter |
| S27421S | Contusion of bronchus, unilateral, sequela |
| S27422 | Contusion of bronchus, bilateral |
| S27422A | Contusion of bronchus, bilateral, initial encounter |
| S27422D | Contusion of bronchus, bilateral, subsequent encounter |
| S27422S | Contusion of bronchus, bilateral, sequela |
| S27429 | Contusion of bronchus, unspecified |
| S27429A | Contusion of bronchus, unspecified, initial encounter |
| S27429D | Contusion of bronchus, unspecified, subsequent encounter |
| S27429S | Contusion of bronchus, unspecified, sequela |
| S2743 | Laceration of bronchus |
| S27431 | Laceration of bronchus, unilateral |
| S27431A | Laceration of bronchus, unilateral, initial encounter |
| S27431D | Laceration of bronchus, unilateral, subsequent encounter |
| S27431S | Laceration of bronchus, unilateral, sequela |
| S27432 | Laceration of bronchus, bilateral |
| S27432A | Laceration of bronchus, bilateral, initial encounter |
| S27432D | Laceration of bronchus, bilateral, subsequent encounter |
| S27432S | Laceration of bronchus, bilateral, sequela |
| S27439 | Laceration of bronchus, unspecified |
| S27439A | Laceration of bronchus, unspecified, initial encounter |
| S27439D | Laceration of bronchus, unspecified, subsequent encounter |
| S27439S | Laceration of bronchus, unspecified, sequela |
| S2749 | Other injury of bronchus |
| S27491 | Other injury of bronchus, unilateral |
| S27491A | Other injury of bronchus, unilateral, initial encounter |
| S27491D | Other injury of bronchus, unilateral, subsequent encounter |
| S27491S | Other injury of bronchus, unilateral, sequela |
| S27492 | Other injury of bronchus, bilateral |
| S27492A | Other injury of bronchus, bilateral, initial encounter |
| S27492D | Other injury of bronchus, bilateral, subsequent encounter |
| S27492S | Other injury of bronchus, bilateral, sequela |
| S27499 | Other injury of bronchus, unspecified |
| S27499A | Other injury of bronchus, unspecified, initial encounter |
| S27499D | Other injury of bronchus, unspecified, subsequent encounter |
| S27499S | Other injury of bronchus, unspecified, sequela |
| S275 | Injury of thoracic trachea |
| S2750 | Unspecified injury of thoracic trachea |
| S2750XA | Unspecified injury of thoracic trachea, initial encounter |
| S2750XD | Unspecified injury of thoracic trachea, subsequent encounter |
| S2750XS | Unspecified injury of thoracic trachea, sequela |
| S2751 | Primary blast injury of thoracic trachea |
| S2751XA | Primary blast injury of thoracic trachea, initial encounter |
| S2751XD | Primary blast injury of thoracic trachea, subsequent encounter |
| S2751XS | Primary blast injury of thoracic trachea, sequela |
| S2752 | Contusion of thoracic trachea |
| S2752XA | Contusion of thoracic trachea, initial encounter |
| S2752XD | Contusion of thoracic trachea, subsequent encounter |
| S2752XS | Contusion of thoracic trachea, sequela |
| S2753 | Laceration of thoracic trachea |
| S2753XA | Laceration of thoracic trachea, initial encounter |
| S2753XD | Laceration of thoracic trachea, subsequent encounter |
| S2753XS | Laceration of thoracic trachea, sequela |
| S2759 | Other injury of thoracic trachea |
| S2759XA | Other injury of thoracic trachea, initial encounter |
| S2759XD | Other injury of thoracic trachea, subsequent encounter |
| S2759XS | Other injury of thoracic trachea, sequela |
| T17 | Foreign body in respiratory tract |
| T174 | Foreign body in trachea |
| T1740 | Unspecified foreign body in trachea |
| T17400 | Unspecified foreign body in trachea causing asphyxiation |
| T17400A | Unspecified foreign body in trachea causing asphyxiation, initial encounter |
| T17400D | Unspecified foreign body in trachea causing asphyxiation, subsequent encounter |
| T17400S | Unspecified foreign body in trachea causing asphyxiation, sequela |
| T17408 | Unspecified foreign body in trachea causing other injury |
| T17408A | Unspecified foreign body in trachea causing other injury, initial encounter |
| T17408D | Unspecified foreign body in trachea causing other injury, subsequent encounter |
| T17408S | Unspecified foreign body in trachea causing other injury, sequela |
| T1741 | Gastric contents in trachea |
| T17410 | Gastric contents in trachea causing asphyxiation |
| T17410A | Gastric contents in trachea causing asphyxiation, initial encounter |
| T17410D | Gastric contents in trachea causing asphyxiation, subsequent encounter |
| T17410S | Gastric contents in trachea causing asphyxiation, sequela |
| T17418 | Gastric contents in trachea causing other injury |
| T17418A | Gastric contents in trachea causing other injury, initial encounter |
| T17418D | Gastric contents in trachea causing other injury, subsequent encounter |
| T17418S | Gastric contents in trachea causing other injury, sequela |
| T1742 | Food in trachea |
| T17420 | Food in trachea causing asphyxiation |
| T17420A | Food in trachea causing asphyxiation, initial encounter |
| T17420D | Food in trachea causing asphyxiation, subsequent encounter |
| T17420S | Food in trachea causing asphyxiation, sequela |
| T17428 | Food in trachea causing other injury |
| T17428A | Food in trachea causing other injury, initial encounter |
| T17428D | Food in trachea causing other injury, subsequent encounter |
| T17428S | Food in trachea causing other injury, sequela |
| T1749 | Other foreign object in trachea |
| T17490 | Other foreign object in trachea causing asphyxiation |
| T17490A | Other foreign object in trachea causing asphyxiation, initial encounter |
| T17490D | Other foreign object in trachea causing asphyxiation, subsequent encounter |
| T17490S | Other foreign object in trachea causing asphyxiation, sequela |
| T17498 | Other foreign object in trachea causing other injury |
| T17498A | Other foreign object in trachea causing other injury, initial encounter |
| T17498D | Other foreign object in trachea causing other injury, subsequent encounter |
| T17498S | Other foreign object in trachea causing other injury, sequela |
| T175 | Foreign body in bronchus |
| T1750 | Unspecified foreign body in bronchus |
| T17500 | Unspecified foreign body in bronchus causing asphyxiation |
| T17500A | Unspecified foreign body in bronchus causing asphyxiation, initial encounter |
| T17500D | Unspecified foreign body in bronchus causing asphyxiation, subsequent encounter |
| T17500S | Unspecified foreign body in bronchus causing asphyxiation, sequela |
| T17508 | Unspecified foreign body in bronchus causing other injury |
| T17508A | Unspecified foreign body in bronchus causing other injury, initial encounter |
| T17508D | Unspecified foreign body in bronchus causing other injury, subsequent encounter |
| T17508S | Unspecified foreign body in bronchus causing other injury, sequela |
| T1751 | Gastric contents in bronchus |
| T17510 | Gastric contents in bronchus causing asphyxiation |
| T17510A | Gastric contents in bronchus causing asphyxiation, initial encounter |
| T17510D | Gastric contents in bronchus causing asphyxiation, subsequent encounter |
| T17510S | Gastric contents in bronchus causing asphyxiation, sequela |
| T17518 | Gastric contents in bronchus causing other injury |
| T17518A | Gastric contents in bronchus causing other injury, initial encounter |
| T17518D | Gastric contents in bronchus causing other injury, subsequent encounter |
| T17518S | Gastric contents in bronchus causing other injury, sequela |
| T1752 | Food in bronchus |
| T17520 | Food in bronchus causing asphyxiation |
| T17520A | Food in bronchus causing asphyxiation, initial encounter |
| T17520D | Food in bronchus causing asphyxiation, subsequent encounter |
| T17520S | Food in bronchus causing asphyxiation, sequela |
| T17528 | Food in bronchus causing other injury |
| T17528A | Food in bronchus causing other injury, initial encounter |
| T17528D | Food in bronchus causing other injury, subsequent encounter |
| T17528S | Food in bronchus causing other injury, sequela |
| T1759 | Other foreign object in bronchus |
| T17590 | Other foreign object in bronchus causing asphyxiation |
| T17590A | Other foreign object in bronchus causing asphyxiation, initial encounter |
| T17590D | Other foreign object in bronchus causing asphyxiation, subsequent encounter |
| T17590S | Other foreign object in bronchus causing asphyxiation, sequela |
| T17598 | Other foreign object in bronchus causing other injury |
| T17598A | Other foreign object in bronchus causing other injury, initial encounter |
| T17598D | Other foreign object in bronchus causing other injury, subsequent encounter |
| T17598S | Other foreign object in bronchus causing other injury, sequela |
| T178 | Foreign body in other parts of respiratory tract |
| T1780 | Unspecified foreign body in other parts of respiratory tract |
| T17800 | Unspecified foreign body in other parts of respiratory tract causing asphyxiation |
| T17800A | Unspecified foreign body in other parts of respiratory tract causing asphyxiation, initial encounter |
| T17800D | Unspecified foreign body in other parts of respiratory tract causing asphyxiation, subsequent encounter |
| T17800S | Unspecified foreign body in other parts of respiratory tract causing asphyxiation, sequela |
| T17808 | Unspecified foreign body in other parts of respiratory tract causing other injury |
| T17808A | Unspecified foreign body in other parts of respiratory tract causing other injury, initial encounter |
| T17808D | Unspecified foreign body in other parts of respiratory tract causing other injury, subsequent encounter |
| T17808S | Unspecified foreign body in other parts of respiratory tract causing other injury, sequela |
| T1781 | Gastric contents in other parts of respiratory tract |
| T17810 | Gastric contents in other parts of respiratory tract causing asphyxiation |
| T17810A | Gastric contents in other parts of respiratory tract causing asphyxiation, initial encounter |
| T17810D | Gastric contents in other parts of respiratory tract causing asphyxiation, subsequent encounter |
| T17810S | Gastric contents in other parts of respiratory tract causing asphyxiation, sequela |
| T17818 | Gastric contents in other parts of respiratory tract causing other injury |
| T17818A | Gastric contents in other parts of respiratory tract causing other injury, initial encounter |
| T17818D | Gastric contents in other parts of respiratory tract causing other injury, subsequent encounter |
| T17818S | Gastric contents in other parts of respiratory tract causing other injury, sequela |
| T1782 | Food in other parts of respiratory tract |
| T17820 | Food in other parts of respiratory tract causing asphyxiation |
| T17820A | Food in other parts of respiratory tract causing asphyxiation, initial encounter |
| T17820D | Food in other parts of respiratory tract causing asphyxiation, subsequent encounter |
| T17820S | Food in other parts of respiratory tract causing asphyxiation, sequela |
| T17828 | Food in other parts of respiratory tract causing other injury |
| T17828A | Food in other parts of respiratory tract causing other injury, initial encounter |
| T17828D | Food in other parts of respiratory tract causing other injury, subsequent encounter |
| T17828S | Food in other parts of respiratory tract causing other injury, sequela |
| T1789 | Other foreign object in other parts of respiratory tract |
| T17890 | Other foreign object in other parts of respiratory tract causing asphyxiation |
| T17890A | Other foreign object in other parts of respiratory tract causing asphyxiation, initial encounter |
| T17890D | Other foreign object in other parts of respiratory tract causing asphyxiation, subsequent encounter |
| T17890S | Other foreign object in other parts of respiratory tract causing asphyxiation, sequela |
| T17898 | Other foreign object in other parts of respiratory tract causing other injury |
| T17898A | Other foreign object in other parts of respiratory tract causing other injury, initial encounter |
| T17898D | Other foreign object in other parts of respiratory tract causing other injury, subsequent encounter |
| T17898S | Other foreign object in other parts of respiratory tract causing other injury, sequela |
| T179 | Foreign body in respiratory tract, part unspecified |
| T1790 | Unspecified foreign body in respiratory tract, part unspecified |
| T17900 | Unspecified foreign body in respiratory tract, part unspecified causing asphyxiation |
| T17900A | Unspecified foreign body in respiratory tract, part unspecified causing asphyxiation, initial encounter |
| T17900D | Unspecified foreign body in respiratory tract, part unspecified causing asphyxiation, subsequent encounter |
| T17900S | Unspecified foreign body in respiratory tract, part unspecified causing asphyxiation, sequela |
| T17908 | Unspecified foreign body in respiratory tract, part unspecified causing other injury |
| T17908A | Unspecified foreign body in respiratory tract, part unspecified causing other injury, initial encounter |
| T17908D | Unspecified foreign body in respiratory tract, part unspecified causing other injury, subsequent encounter |
| T17908S | Unspecified foreign body in respiratory tract, part unspecified causing other injury, sequela |
| T1791 | Gastric contents in respiratory tract, part unspecified |
| T17910 | Gastric contents in respiratory tract, part unspecified causing asphyxiation |
| T17910A | Gastric contents in respiratory tract, part unspecified causing asphyxiation, initial encounter |
| T17910D | Gastric contents in respiratory tract, part unspecified causing asphyxiation, subsequent encounter |
| T17910S | Gastric contents in respiratory tract, part unspecified causing asphyxiation, sequela |
| T17918 | Gastric contents in respiratory tract, part unspecified causing other injury |
| T17918A | Gastric contents in respiratory tract, part unspecified causing other injury, initial encounter |
| T17918D | Gastric contents in respiratory tract, part unspecified causing other injury, subsequent encounter |
| T17918S | Gastric contents in respiratory tract, part unspecified causing other injury, sequela |
| T1792 | Food in respiratory tract, part unspecified |
| T17920 | Food in respiratory tract, part unspecified causing asphyxiation |
| T17920A | Food in respiratory tract, part unspecified causing asphyxiation, initial encounter |
| T17920D | Food in respiratory tract, part unspecified causing asphyxiation, subsequent encounter |
| T17920S | Food in respiratory tract, part unspecified causing asphyxiation, sequela |
| T17928 | Food in respiratory tract, part unspecified causing other injury |
| T17928A | Food in respiratory tract, part unspecified causing other injury, initial encounter |
| T17928D | Food in respiratory tract, part unspecified causing other injury, subsequent encounter |
| T17928S | Food in respiratory tract, part unspecified causing other injury, sequela |
| T1799 | Other foreign object in respiratory tract, part unspecified |
| T17990 | Other foreign object in respiratory tract, part unspecified in causing asphyxiation |
| T17990A | Other foreign object in respiratory tract, part unspecified in causing asphyxiation, initial encounter |
| T17990D | Other foreign object in respiratory tract, part unspecified in causing asphyxiation, subsequent encounter |
| T17990S | Other foreign object in respiratory tract, part unspecified in causing asphyxiation, sequela |
| T17998 | Other foreign object in respiratory tract, part unspecified causing other injury |
| T17998A | Other foreign object in respiratory tract, part unspecified causing other injury, initial encounter |
| T17998D | Other foreign object in respiratory tract, part unspecified causing other injury, subsequent encounter |
| T17998S | Other foreign object in respiratory tract, part unspecified causing other injury, sequela |
| T18100 | Unspecified foreign body in esophagus causing compression of trachea |
| T18100A | Unspecified foreign body in esophagus causing compression of trachea, initial encounter |
| T18100D | Unspecified foreign body in esophagus causing compression of trachea, subsequent encounter |
| T18100S | Unspecified foreign body in esophagus causing compression of trachea, sequela |
| T18110 | Gastric contents in esophagus causing compression of trachea |
| T18110A | Gastric contents in esophagus causing compression of trachea, initial encounter |
| T18110D | Gastric contents in esophagus causing compression of trachea, subsequent encounter |
| T18110S | Gastric contents in esophagus causing compression of trachea, sequela |
| T18120 | Food in esophagus causing compression of trachea |
| T18120A | Food in esophagus causing compression of trachea, initial encounter |
| T18120D | Food in esophagus causing compression of trachea, subsequent encounter |
| T18120S | Food in esophagus causing compression of trachea, sequela |
| T18190 | Other foreign object in esophagus causing compression of trachea |
| T18190A | Other foreign object in esophagus causing compression of trachea, initial encounter |
| T18190D | Other foreign object in esophagus causing compression of trachea, subsequent encounter |
| T18190S | Other foreign object in esophagus causing compression of trachea, sequela |
| T27 | Burn and corrosion of respiratory tract |
| T270 | Burn of larynx and trachea |
| T270XXA | Burn of larynx and trachea, initial encounter |
| T270XXD | Burn of larynx and trachea, subsequent encounter |
| T270XXS | Burn of larynx and trachea, sequela |
| T271 | Burn involving larynx and trachea with lung |
| T271XXA | Burn involving larynx and trachea with lung, initial encounter |
| T271XXD | Burn involving larynx and trachea with lung, subsequent encounter |
| T271XXS | Burn involving larynx and trachea with lung, sequela |
| T272 | Burn of other parts of respiratory tract |
| T272XXA | Burn of other parts of respiratory tract, initial encounter |
| T272XXD | Burn of other parts of respiratory tract, subsequent encounter |
| T272XXS | Burn of other parts of respiratory tract, sequela |
| T273 | Burn of respiratory tract, part unspecified |
| T273XXA | Burn of respiratory tract, part unspecified, initial encounter |
| T273XXD | Burn of respiratory tract, part unspecified, subsequent encounter |
| T273XXS | Burn of respiratory tract, part unspecified, sequela |
| T274 | Corrosion of larynx and trachea |
| T274XXA | Corrosion of larynx and trachea, initial encounter |
| T274XXD | Corrosion of larynx and trachea, subsequent encounter |
| T274XXS | Corrosion of larynx and trachea, sequela |
| T275 | Corrosion involving larynx and trachea with lung |
| T275XXA | Corrosion involving larynx and trachea with lung, initial encounter |
| T275XXD | Corrosion involving larynx and trachea with lung, subsequent encounter |
| T275XXS | Corrosion involving larynx and trachea with lung, sequela |
| T276 | Corrosion of other parts of respiratory tract |
| T276XXA | Corrosion of other parts of respiratory tract, initial encounter |
| T276XXD | Corrosion of other parts of respiratory tract, subsequent encounter |
| T276XXS | Corrosion of other parts of respiratory tract, sequela |
| T277 | Corrosion of respiratory tract, part unspecified |
| T277XXA | Corrosion of respiratory tract, part unspecified, initial encounter |
| T277XXD | Corrosion of respiratory tract, part unspecified, subsequent encounter |
| T277XXS | Corrosion of respiratory tract, part unspecified, sequela |
| T410 | Poisoning by, adverse effect of and underdosing of inhaled anesthetics |
| T410X | Poisoning by, adverse effect of and underdosing of inhaled anesthetics |
| T410X1 | Poisoning by inhaled anesthetics, accidental (unintentional) |
| T410X1A | Poisoning by inhaled anesthetics, accidental (unintentional), initial encounter |
| T410X1D | Poisoning by inhaled anesthetics, accidental (unintentional), subsequent encounter |
| T410X1S | Poisoning by inhaled anesthetics, accidental (unintentional), sequela |
| T410X2 | Poisoning by inhaled anesthetics, intentional self-harm |
| T410X2A | Poisoning by inhaled anesthetics, intentional self-harm, initial encounter |
| T410X2D | Poisoning by inhaled anesthetics, intentional self-harm, subsequent encounter |
| T410X2S | Poisoning by inhaled anesthetics, intentional self-harm, sequela |
| T410X3 | Poisoning by inhaled anesthetics, assault |
| T410X5 | Adverse effect of inhaled anesthetics |
| T410X5A | Adverse effect of inhaled anesthetics, initial encounter |
| T410X5D | Adverse effect of inhaled anesthetics, subsequent encounter |
| T410X5S | Adverse effect of inhaled anesthetics, sequela |
| T410X6 | Underdosing of inhaled anesthetics |
| T410X6A | Underdosing of inhaled anesthetics, initial encounter |
| T410X6D | Underdosing of inhaled anesthetics, subsequent encounter |
| T410X6S | Underdosing of inhaled anesthetics, sequela |
| T711 | Asphyxiation due to mechanical threat to breathing |
| T71111A | Asphyxiation due to smothering under pillow, accidental, initial encounter |
| T71111D | Asphyxiation due to smothering under pillow, accidental, subsequent encounter |
| T71111S | Asphyxiation due to smothering under pillow, accidental, sequela |
| T71112S | Asphyxiation due to smothering under pillow, intentional self-harm, sequela |
| T71113S | Asphyxiation due to smothering under pillow, assault, sequela |
| T71114A | Asphyxiation due to smothering under pillow, undetermined, initial encounter |
| T71114D | Asphyxiation due to smothering under pillow, undetermined, subsequent encounter |
| T71114S | Asphyxiation due to smothering under pillow, undetermined, sequela |
| T71131A | Asphyxiation due to being trapped in bed linens, accidental, initial encounter |
| T71131D | Asphyxiation due to being trapped in bed linens, accidental, subsequent encounter |
| T71131S | Asphyxiation due to being trapped in bed linens, accidental, sequela |
| T71132A | Asphyxiation due to being trapped in bed linens, intentional self-harm, initial encounter |
| T71132D | Asphyxiation due to being trapped in bed linens, intentional self-harm, subsequent encounter |
| T71132S | Asphyxiation due to being trapped in bed linens, intentional self-harm, sequela |
| T71133A | Asphyxiation due to being trapped in bed linens, assault, initial encounter |
| T71133D | Asphyxiation due to being trapped in bed linens, assault, subsequent encounter |
| T71133S | Asphyxiation due to being trapped in bed linens, assault, sequela |
| T71134 | Asphyxiation due to being trapped in bed linens, undetermined |
| T71134A | Asphyxiation due to being trapped in bed linens, undetermined, initial encounter |
| T71134D | Asphyxiation due to being trapped in bed linens, undetermined, subsequent encounter |
| T71134S | Asphyxiation due to being trapped in bed linens, undetermined, sequela |
| T7114 | Asphyxiation due to smothering under another person’s body (in bed) |
| T71141 | Asphyxiation due to smothering under another person’s body (in bed), accidental |
| T71141A | Asphyxiation due to smothering under another person’s body (in bed), accidental, initial encounter |
| T71141D | Asphyxiation due to smothering under another person’s body (in bed), accidental, subsequent encounter |
| T71141S | Asphyxiation due to smothering under another person’s body (in bed), accidental, sequela |
| T71143 | Asphyxiation due to smothering under another person’s body (in bed), assault |
| T71143A | Asphyxiation due to smothering under another person’s body (in bed), assault, initial encounter |
| T71143D | Asphyxiation due to smothering under another person’s body (in bed), assault, subsequent encounter |
| T71143S | Asphyxiation due to smothering under another person’s body (in bed), assault, sequela |
| T71144 | Asphyxiation due to smothering under another person’s body (in bed), undetermined |
| T71144A | Asphyxiation due to smothering under another person’s body (in bed), undetermined, initial encounter |
| T71144D | Asphyxiation due to smothering under another person’s body (in bed), undetermined, subsequent encounter |
| T71144S | Asphyxiation due to smothering under another person’s body (in bed), undetermined, sequela |
| T71151A | Asphyxiation due to smothering in furniture, accidental, initial encounter |

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| T71151D | Asphyxiation due to smothering in furniture, accidental, subsequent encounter |
| T71151S | Asphyxiation due to smothering in furniture, accidental, sequela |
| T71152S | Asphyxiation due to smothering in furniture, intentional self-harm, sequela |
| T71153S | Asphyxiation due to smothering in furniture, assault, sequela |
| T71154A | Asphyxiation due to smothering in furniture, undetermined, initial encounter |
| T71154D | Asphyxiation due to smothering in furniture, undetermined, subsequent encounter |
| T71154S | Asphyxiation due to smothering in furniture, undetermined, sequela |
| T7119 | Asphyxiation due to mechanical threat to breathing due to other causes |
| T71191 | Asphyxiation due to mechanical threat to breathing due to other causes, accidental |
| T71191A | Asphyxiation due to mechanical threat to breathing due to other causes, accidental, initial encounter |
| T71191D | Asphyxiation due to mechanical threat to breathing due to other causes, accidental, subsequent encounter |
| T71191S | Asphyxiation due to mechanical threat to breathing due to other causes, accidental, sequela |
| T71192 | Asphyxiation due to mechanical threat to breathing due to other causes, intentional self-harm |
| T71192A | Asphyxiation due to mechanical threat to breathing due to other causes, intentional self-harm, initial encounter |
| T71192D | Asphyxiation due to mechanical threat to breathing due to other causes, intentional self-harm, subsequent encounter |
| T71192S | Asphyxiation due to mechanical threat to breathing due to other causes, intentional self-harm, sequela |
| T71193 | Asphyxiation due to mechanical threat to breathing due to other causes, assault |
| T71193A | Asphyxiation due to mechanical threat to breathing due to other causes, assault, initial encounter |
| T71193D | Asphyxiation due to mechanical threat to breathing due to other causes, assault, subsequent encounter |
| T71193S | Asphyxiation due to mechanical threat to breathing due to other causes, assault, sequela |
| T71194 | Asphyxiation due to mechanical threat to breathing due to other causes, undetermined |
| T71194A | Asphyxiation due to mechanical threat to breathing due to other causes, undetermined, initial encounter |
| T71194D | Asphyxiation due to mechanical threat to breathing due to other causes, undetermined, subsequent encounter |
| T71194S | Asphyxiation due to mechanical threat to breathing due to other causes, undetermined, sequela |
| T7120 | Asphyxiation due to systemic oxygen deficiency due to low oxygen content in ambient air due to unspecified cause |
| T7120XA | Asphyxiation due to systemic oxygen deficiency due to low oxygen content in ambient air due to unspecified cause, initial encounter |
| T7120XD | Asphyxiation due to systemic oxygen deficiency due to low oxygen content in ambient air due to unspecified cause, subsequent encounter |
| T7120XS | Asphyxiation due to systemic oxygen deficiency due to low oxygen content in ambient air due to unspecified cause, sequela |
| T71221A | Asphyxiation due to being trapped in a car trunk, accidental, initial encounter |
| T71221D | Asphyxiation due to being trapped in a car trunk, accidental, subsequent encounter |
| T71221S | Asphyxiation due to being trapped in a car trunk, accidental, sequela |
| T71222A | Asphyxiation due to being trapped in a car trunk, intentional self-harm, initial encounter |
| T71222D | Asphyxiation due to being trapped in a car trunk, intentional self-harm, subsequent encounter |
| T71222S | Asphyxiation due to being trapped in a car trunk, intentional self-harm, sequela |
| T71223A | Asphyxiation due to being trapped in a car trunk, assault, initial encounter |
| T71223D | Asphyxiation due to being trapped in a car trunk, assault, subsequent encounter |
| T71223S | Asphyxiation due to being trapped in a car trunk, assault, sequela |
| T71224 | Asphyxiation due to being trapped in a car trunk, undetermined |
| T71224A | Asphyxiation due to being trapped in a car trunk, undetermined, initial encounter |
| T71224D | Asphyxiation due to being trapped in a car trunk, undetermined, subsequent encounter |
| T71224S | Asphyxiation due to being trapped in a car trunk, undetermined, sequela |
| T7123 | Asphyxiation due to being trapped in a (discarded) refrigerator |
| T71231 | Asphyxiation due to being trapped in a (discarded) refrigerator, accidental |
| T71231A | Asphyxiation due to being trapped in a (discarded) refrigerator, accidental, initial encounter |
| T71231D | Asphyxiation due to being trapped in a (discarded) refrigerator, accidental, subsequent encounter |
| T71231S | Asphyxiation due to being trapped in a (discarded) refrigerator, accidental, sequela |
| T71232 | Asphyxiation due to being trapped in a (discarded) refrigerator, intentional self-harm |
| T71232A | Asphyxiation due to being trapped in a (discarded) refrigerator, intentional self-harm, initial encounter |
| T71232D | Asphyxiation due to being trapped in a (discarded) refrigerator, intentional self-harm, subsequent encounter |
| T71232S | Asphyxiation due to being trapped in a (discarded) refrigerator, intentional self-harm, sequela |
| T71233 | Asphyxiation due to being trapped in a (discarded) refrigerator, assault |
| T71233A | Asphyxiation due to being trapped in a (discarded) refrigerator, assault, initial encounter |
| T71233D | Asphyxiation due to being trapped in a (discarded) refrigerator, assault, subsequent encounter |
| T71233S | Asphyxiation due to being trapped in a (discarded) refrigerator, assault, sequela |
| T71234 | Asphyxiation due to being trapped in a (discarded) refrigerator, undetermined |
| T71234A | Asphyxiation due to being trapped in a (discarded) refrigerator, undetermined, initial encounter |
| T71234D | Asphyxiation due to being trapped in a (discarded) refrigerator, undetermined, subsequent encounter |
| T71234S | Asphyxiation due to being trapped in a (discarded) refrigerator, undetermined, sequela |
| T7129 | Asphyxiation due to being trapped in other low oxygen environment |
| T7129XA | Asphyxiation due to being trapped in other low oxygen environment, initial encounter |
| T7129XD | Asphyxiation due to being trapped in other low oxygen environment, subsequent encounter |
| T7129XS | Asphyxiation due to being trapped in other low oxygen environment, sequela |
| T8181 | Complication of inhalation therapy |
| T8181XA | Complication of inhalation therapy, initial encounter |
| T8181XD | Complication of inhalation therapy, subsequent encounter |
| T8181XS | Complication of inhalation therapy, sequela |
| T863 | Complications of heart-lung transplant |
| T8630 | Unspecified complication of heart-lung transplant |
| T8631 | Heart-lung transplant rejection |
| T8632 | Heart-lung transplant failure |
| T8633 | Heart-lung transplant infection |
| T8639 | Other complications of heart-lung transplant |
| T8681 | Complications of lung transplant |
| T86810 | Lung transplant rejection |
| T86811 | Lung transplant failure |
| T86812 | Lung transplant infection |
| T86818 | Other complications of lung transplant |
| T86819 | Unspecified complication of lung transplant |
| W75-W84 | Other accidental threats to breathing |
| W77 | Threat to breathing due to cave-in, falling earth and other substances |
| W770 | Threat to breathing due to cave-in, falling earth and other substances, home |
| W7700 | Threat to breathing due to cave-in, falling earth and other substances, home, while engaged in sports activity |
| W7701 | Threat to breathing due to cave-in, falling earth and other substances, home, while engaged in leisure activity |
| W7702 | Threat to breathing due to cave-in, falling earth and other substances, home, while working for income |
| W7703 | Threat to breathing due to cave-in, falling earth and other substances, home, while engaged in other types of work |
| W7704 | Threat to breathing due to cave-in, falling earth and other substances, home, while resting, sleeping, eating or engaging in other vital activities |
| W7708 | Threat to breathing due to cave-in, falling earth and other substances, home, while engaged in other specified activities |
| W7709 | Threat to breathing due to cave-in, falling earth and other substances, home, during unspecified activity |
| W771 | Threat to breathing due to cave-in, falling earth and other substances, residential institution |
| W7710 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while engaged in sports activity |
| W7711 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while engaged in leisure activity |
| W7712 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while working for income |
| W7713 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while engaged in other types of work |
| W7714 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W7718 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, while engaged in other specified activities |
| W7719 | Threat to breathing due to cave-in, falling earth and other substances, residential institution, during unspecified activity |
| W772 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area |
| W7720 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while engaged in sports activity |
| W7721 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while engaged in leisure activity |
| W7722 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while working for income |
| W7723 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while engaged in other types of work |
| W7724 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W7728 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, while engaged in other specified activities |
| W7729 | Threat to breathing due to cave-in, falling earth and other substances, school, other institution and public administrative area, during unspecified activity |
| W773 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area |
| W7730 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while engaged in sports activity |
| W7731 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while engaged in leisure activity |
| W7732 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while working for income |
| W7733 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while engaged in other types of work |
| W7734 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W7738 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, while engaged in other specified activities |
| W7739 | Threat to breathing due to cave-in, falling earth and other substances, sports and athletics area, during unspecified activity |
| W774 | Threat to breathing due to cave-in, falling earth and other substances, street and highway |
| W7740 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while engaged in sports activity |
| W7741 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while engaged in leisure activity |
| W7742 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while working for income |
| W7743 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while engaged in other types of work |
| W7744 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W7748 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, while engaged in other specified activities |
| W7749 | Threat to breathing due to cave-in, falling earth and other substances, street and highway, during unspecified activity |
| W775 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area |
| W7750 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while engaged in sports activity |
| W7751 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while engaged in leisure activity |
| W7752 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while working for income |
| W7753 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while engaged in other types of work |
| W7754 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W7758 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, while engaged in other specified activities |
| W7759 | Threat to breathing due to cave-in, falling earth and other substances, trade and service area, during unspecified activity |
| W776 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area |
| W7760 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while engaged in sports activity |
| W7761 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while engaged in leisure activity |
| W7762 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while working for income |
| W7763 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while engaged in other types of work |
| W7764 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W7768 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, while engaged in other specified activities |
| W7769 | Threat to breathing due to cave-in, falling earth and other substances, industrial and construction area, during unspecified activity |
| W777 | Threat to breathing due to cave-in, falling earth and other substances, farm |
| W7770 | Threat to breathing due to cave-in, falling earth and other substances, farm, while engaged in sports activity |
| W7771 | Threat to breathing due to cave-in, falling earth and other substances, farm, while engaged in leisure activity |
| W7772 | Threat to breathing due to cave-in, falling earth and other substances, farm, while working for income |
| W7773 | Threat to breathing due to cave-in, falling earth and other substances, farm, while engaged in other types of work |
| W7774 | Threat to breathing due to cave-in, falling earth and other substances, farm, while resting, sleeping, eating or engaging in other vital activities |
| W7778 | Threat to breathing due to cave-in, falling earth and other substances, farm, while engaged in other specified activities |
| W7779 | Threat to breathing due to cave-in, falling earth and other substances, farm, during unspecified activity |
| W778 | Threat to breathing due to cave-in, falling earth and other substances, other specified places |
| W7780 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while engaged in sports activity |
| W7781 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while engaged in leisure activity |
| W7782 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while working for income |
| W7783 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while engaged in other types of work |
| W7784 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W7788 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, while engaged in other specified activities |
| W7789 | Threat to breathing due to cave-in, falling earth and other substances, other specified places, during unspecified activity |
| W779 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place |
| W7790 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while engaged in sports activity |
| W7791 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while engaged in leisure activity |
| W7792 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while working for income |
| W7793 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while engaged in other types of work |
| W7794 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W7798 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, while engaged in other specified activities |
| W7799 | Threat to breathing due to cave-in, falling earth and other substances, unspecified place, during unspecified activity |
| W78 | Inhalation of gastric contents |
| W780 | Inhalation of gastric contents, home |
| W7800 | Inhalation of gastric contents, home, while engaged in sports activity |
| W7801 | Inhalation of gastric contents, home, while engaged in leisure activity |
| W7802 | Inhalation of gastric contents, home, while working for income |
| W7803 | Inhalation of gastric contents, home, while engaged in other types of work |
| W7804 | Inhalation of gastric contents, home, while resting, sleeping, eating or engaging in other vital activities |
| W7808 | Inhalation of gastric contents, home, while engaged in other specified activities |
| W7809 | Inhalation of gastric contents, home, during unspecified activity |
| W781 | Inhalation of gastric contents, residential institution |
| W7810 | Inhalation of gastric contents, residential institution, while engaged in sports activity |
| W7811 | Inhalation of gastric contents, residential institution, while engaged in leisure activity |
| W7812 | Inhalation of gastric contents, residential institution, while working for income |
| W7813 | Inhalation of gastric contents, residential institution, while engaged in other types of work |
| W7814 | Inhalation of gastric contents, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W7818 | Inhalation of gastric contents, residential institution, while engaged in other specified activities |
| W7819 | Inhalation of gastric contents, residential institution, during unspecified activity |
| W782 | Inhalation of gastric contents, school, other institution and public administrative area |
| W7820 | Inhalation of gastric contents, school, other institution and public administrative area, while engaged in sports activity |
| W7821 | Inhalation of gastric contents, school, other institution and public administrative area, while engaged in leisure activity |
| W7822 | Inhalation of gastric contents, school, other institution and public administrative area, while working for income |
| W7823 | Inhalation of gastric contents, school, other institution and public administrative area, while engaged in other types of work |
| W7824 | Inhalation of gastric contents, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W7828 | Inhalation of gastric contents, school, other institution and public administrative area, while engaged in other specified activities |
| W7829 | Inhalation of gastric contents, school, other institution and public administrative area, during unspecified activity |
| W783 | Inhalation of gastric contents, sports and athletics area |
| W7830 | Inhalation of gastric contents, sports and athletics area, while engaged in sports activity |
| W7831 | Inhalation of gastric contents, sports and athletics area, while engaged in leisure activity |
| W7832 | Inhalation of gastric contents, sports and athletics area, while working for income |
| W7833 | Inhalation of gastric contents, sports and athletics area, while engaged in other types of work |
| W7834 | Inhalation of gastric contents, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W7838 | Inhalation of gastric contents, sports and athletics area, while engaged in other specified activities |
| W7839 | Inhalation of gastric contents, sports and athletics area, during unspecified activity |
| W784 | Inhalation of gastric contents, street and highway |
| W7840 | Inhalation of gastric contents, street and highway, while engaged in sports activity |
| W7841 | Inhalation of gastric contents, street and highway, while engaged in leisure activity |
| W7842 | Inhalation of gastric contents, street and highway, while working for income |
| W7843 | Inhalation of gastric contents, street and highway, while engaged in other types of work |
| W7844 | Inhalation of gastric contents, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W7848 | Inhalation of gastric contents, street and highway, while engaged in other specified activities |
| W7849 | Inhalation of gastric contents, street and highway, during unspecified activity |
| W785 | Inhalation of gastric contents, trade and service area |
| W7850 | Inhalation of gastric contents, trade and service area, while engaged in sports activity |
| W7851 | Inhalation of gastric contents, trade and service area, while engaged in leisure activity |
| W7852 | Inhalation of gastric contents, trade and service area, while working for income |
| W7853 | Inhalation of gastric contents, trade and service area, while engaged in other types of work |
| W7854 | Inhalation of gastric contents, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W7858 | Inhalation of gastric contents, trade and service area, while engaged in other specified activities |
| W7859 | Inhalation of gastric contents, trade and service area, during unspecified activity |
| W786 | Inhalation of gastric contents, industrial and construction area |
| W7860 | Inhalation of gastric contents, industrial and construction area, while engaged in sports activity |
| W7861 | Inhalation of gastric contents, industrial and construction area, while engaged in leisure activity |
| W7862 | Inhalation of gastric contents, industrial and construction area, while working for income |
| W7863 | Inhalation of gastric contents, industrial and construction area, while engaged in other types of work |
| W7864 | Inhalation of gastric contents, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W7868 | Inhalation of gastric contents, industrial and construction area, while engaged in other specified activities |
| W7869 | Inhalation of gastric contents, industrial and construction area, during unspecified activity |
| W787 | Inhalation of gastric contents, farm |
| W7870 | Inhalation of gastric contents, farm, while engaged in sports activity |
| W7871 | Inhalation of gastric contents, farm, while engaged in leisure activity |
| W7872 | Inhalation of gastric contents, farm, while working for income |
| W7873 | Inhalation of gastric contents, farm, while engaged in other types of work |
| W7874 | Inhalation of gastric contents, farm, while resting, sleeping, eating or engaging in other vital activities |
| W7878 | Inhalation of gastric contents, farm, while engaged in other specified activities |
| W7879 | Inhalation of gastric contents, farm, during unspecified activity |
| W788 | Inhalation of gastric contents, other specified places |
| W7880 | Inhalation of gastric contents, other specified places, while engaged in sports activity |
| W7881 | Inhalation of gastric contents, other specified places, while engaged in leisure activity |
| W7882 | Inhalation of gastric contents, other specified places, while working for income |
| W7883 | Inhalation of gastric contents, other specified places, while engaged in other types of work |
| W7884 | Inhalation of gastric contents, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W7888 | Inhalation of gastric contents, other specified places, while engaged in other specified activities |
| W7889 | Inhalation of gastric contents, other specified places, during unspecified activity |
| W789 | Inhalation of gastric contents, unspecified place |
| W7890 | Inhalation of gastric contents, unspecified place, while engaged in sports activity |
| W7891 | Inhalation of gastric contents, unspecified place, while engaged in leisure activity |
| W7892 | Inhalation of gastric contents, unspecified place, while working for income |
| W7893 | Inhalation of gastric contents, unspecified place, while engaged in other types of work |
| W7894 | Inhalation of gastric contents, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W7898 | Inhalation of gastric contents, unspecified place, while engaged in other specified activities |
| W7899 | Inhalation of gastric contents, unspecified place, during unspecified activity |
| W79 | Inhalation and ingestion of food causing obstruction of respiratory tract |
| W790 | Inhalation and ingestion of food causing obstruction of respiratory tract, home |
| W7900 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while engaged in sports activity |
| W7901 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while engaged in leisure activity |
| W7902 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while working for income |
| W7903 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while engaged in other types of work |
| W7904 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while resting, sleeping, eating or engaging in other vital activities |
| W7908 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, while engaged in other specified activities |
| W7909 | Inhalation and ingestion of food causing obstruction of respiratory tract, home, during unspecified activity |
| W791 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution |
| W7910 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while engaged in sports activity |
| W7911 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while engaged in leisure activity |
| W7912 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while working for income |
| W7913 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while engaged in other types of work |
| W7914 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W7918 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, while engaged in other specified activities |
| W7919 | Inhalation and ingestion of food causing obstruction of respiratory tract, residential institution, during unspecified activity |
| W792 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area |
| W7920 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in sports activity |
| W7921 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in leisure activity |
| W7922 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while working for income |
| W7923 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in other types of work |
| W7924 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W7928 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in other specified activities |
| W7929 | Inhalation and ingestion of food causing obstruction of respiratory tract, school, other institution and public administrative area, during unspecified activity |
| W793 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area |
| W7930 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while engaged in sports activity |
| W7931 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while engaged in leisure activity |
| W7932 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while working for income |
| W7933 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while engaged in other types of work |
| W7934 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W7938 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, while engaged in other specified activities |
| W7939 | Inhalation and ingestion of food causing obstruction of respiratory tract, sports and athletics area, during unspecified activity |
| W794 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway |
| W7940 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while engaged in sports activity |
| W7941 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while engaged in leisure activity |
| W7942 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while working for income |
| W7943 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while engaged in other types of work |
| W7944 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W7948 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, while engaged in other specified activities |
| W7949 | Inhalation and ingestion of food causing obstruction of respiratory tract, street and highway, during unspecified activity |
| W795 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area |
| W7950 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while engaged in sports activity |
| W7951 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while engaged in leisure activity |
| W7952 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while working for income |
| W7953 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while engaged in other types of work |
| W7954 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W7958 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, while engaged in other specified activities |
| W7959 | Inhalation and ingestion of food causing obstruction of respiratory tract, trade and service area, during unspecified activity |
| W796 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area |
| W7960 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while engaged in sports activity |
| W7961 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while engaged in leisure activity |
| W7962 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while working for income |
| W7963 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while engaged in other types of work |
| W7964 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W7968 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, while engaged in other specified activities |
| W7969 | Inhalation and ingestion of food causing obstruction of respiratory tract, industrial and construction area, during unspecified activity |
| W797 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm |
| W7970 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while engaged in sports activity |
| W7971 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while engaged in leisure activity |
| W7972 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while working for income |
| W7973 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while engaged in other types of work |
| W7974 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while resting, sleeping, eating or engaging in other vital activities |
| W7978 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, while engaged in other specified activities |
| W7979 | Inhalation and ingestion of food causing obstruction of respiratory tract, farm, during unspecified activity |
| W798 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places |
| W7980 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while engaged in sports activity |
| W7981 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while engaged in leisure activity |
| W7982 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while working for income |
| W7983 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while engaged in other types of work |
| W7984 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W7988 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, while engaged in other specified activities |
| W7989 | Inhalation and ingestion of food causing obstruction of respiratory tract, other specified places, during unspecified activity |

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| W799 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place |
| W7990 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while engaged in sports activity |
| W7991 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while engaged in leisure activity |
| W7992 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while working for income |
| W7993 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while engaged in other types of work |
| W7994 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W7998 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, while engaged in other specified activities |
| W7999 | Inhalation and ingestion of food causing obstruction of respiratory tract, unspecified place, during unspecified activity |
| W80 | Inhalation and ingestion of other objects causing obstruction of respiratory tract |
| W800 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home |
| W8000 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while engaged in sports activity |
| W8001 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while engaged in leisure activity |
| W8002 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while working for income |
| W8003 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while engaged in other types of work |
| W8004 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while resting, sleeping, eating or engaging in other vital activities |
| W8008 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, while engaged in other specified activities |
| W8009 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, home, during unspecified activity |
| W801 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution |
| W8010 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while engaged in sports activity |
| W8011 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while engaged in leisure activity |
| W8012 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while working for income |
| W8013 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while engaged in other types of work |
| W8014 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W8018 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, while engaged in other specified activities |
| W8019 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, residential institution, during unspecified activity |
| W802 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area |
| W8020 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in sports activity |
| W8021 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in leisure activity |
| W8022 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while working for income |
| W8023 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in other types of work |
| W8024 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W8028 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, while engaged in other specified activities |
| W8029 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, school, other institution and public administrative area, during unspecified activity |
| W803 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area |
| W8030 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while engaged in sports activity |
| W8031 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while engaged in leisure activity |
| W8032 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while working for income |
| W8033 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while engaged in other types of work |
| W8034 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W8038 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, while engaged in other specified activities |
| W8039 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, sports and athletics area, during unspecified activity |
| W804 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway |
| W8040 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while engaged in sports activity |
| W8041 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while engaged in leisure activity |
| W8042 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while working for income |
| W8043 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while engaged in other types of work |
| W8044 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W8048 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, while engaged in other specified activities |
| W8049 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, street and highway, during unspecified activity |
| W805 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area |
| W8050 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while engaged in sports activity |
| W8051 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while engaged in leisure activity |
| W8052 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while working for income |
| W8053 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while engaged in other types of work |
| W8054 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W8058 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, while engaged in other specified activities |
| W8059 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, trade and service area, during unspecified activity |
| W806 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area |
| W8060 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while engaged in sports activity |
| W8061 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while engaged in leisure activity |
| W8062 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while working for income |
| W8063 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while engaged in other types of work |
| W8064 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W8068 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, while engaged in other specified activities |
| W8069 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, industrial and construction area, during unspecified activity |
| W807 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm |
| W8070 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while engaged in sports activity |
| W8071 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while engaged in leisure activity |
| W8072 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while working for income |
| W8073 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while engaged in other types of work |
| W8074 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while resting, sleeping, eating or engaging in other vital activities |
| W8078 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, while engaged in other specified activities |
| W8079 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, farm, during unspecified activity |
| W808 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places |
| W8080 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while engaged in sports activity |
| W8081 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while engaged in leisure activity |
| W8082 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while working for income |
| W8083 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while engaged in other types of work |
| W8084 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W8088 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, while engaged in other specified activities |
| W8089 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, other specified places, during unspecified activity |
| W809 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place |
| W8090 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while engaged in sports activity |
| W8091 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while engaged in leisure activity |
| W8092 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while working for income |
| W8093 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while engaged in other types of work |
| W8094 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W8098 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, while engaged in other specified activities |
| W8099 | Inhalation and ingestion of other objects causing obstruction of respiratory tract, unspecified place, during unspecified activity |
| W82 | Other accidental threats to breathing |
| W83 | Other specified threats to breathing |
| W830 | Other specified threats to breathing, home |
| W8300 | Other specified threats to breathing, home, while engaged in sports activity |
| W8301 | Other specified threats to breathing, home, while engaged in leisure activity |
| W8302 | Other specified threats to breathing, home, while working for income |
| W8303 | Other specified threats to breathing, home, while engaged in other types of work |
| W8304 | Other specified threats to breathing, home, while resting, sleeping, eating or engaging in other vital activities |
| W8308 | Other specified threats to breathing, home, while engaged in other specified activities |
| W8309 | Other specified threats to breathing, home, during unspecified activity |
| W831 | Other specified threats to breathing, residential institution |
| W8310 | Other specified threats to breathing, residential institution, while engaged in sports activity |
| W8311 | Other specified threats to breathing, residential institution, while engaged in leisure activity |
| W8312 | Other specified threats to breathing, residential institution, while working for income |
| W8313 | Other specified threats to breathing, residential institution, while engaged in other types of work |
| W8314 | Other specified threats to breathing, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W8318 | Other specified threats to breathing, residential institution, while engaged in other specified activities |
| W8319 | Other specified threats to breathing, residential institution, during unspecified activity |
| W832 | Other specified threats to breathing, school, other institution and public administrative area |
| W8320 | Other specified threats to breathing, school, other institution and public administrative area, while engaged in sports activity |
| W8321 | Other specified threats to breathing, school, other institution and public administrative area, while engaged in leisure activity |
| W8322 | Other specified threats to breathing, school, other institution and public administrative area, while working for income |
| W8323 | Other specified threats to breathing, school, other institution and public administrative area, while engaged in other types of work |
| W8324 | Other specified threats to breathing, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W8328 | Other specified threats to breathing, school, other institution and public administrative area, while engaged in other specified activities |
| W8329 | Other specified threats to breathing, school, other institution and public administrative area, during unspecified activity |
| W833 | Other specified threats to breathing, sports and athletics area |
| W8330 | Other specified threats to breathing, sports and athletics area, while engaged in sports activity |
| W8331 | Other specified threats to breathing, sports and athletics area, while engaged in leisure activity |
| W8332 | Other specified threats to breathing, sports and athletics area, while working for income |
| W8333 | Other specified threats to breathing, sports and athletics area, while engaged in other types of work |
| W8334 | Other specified threats to breathing, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W8338 | Other specified threats to breathing, sports and athletics area, while engaged in other specified activities |
| W8339 | Other specified threats to breathing, sports and athletics area, during unspecified activity |
| W834 | Other specified threats to breathing, street and highway |
| W8340 | Other specified threats to breathing, street and highway, while engaged in sports activity |
| W8341 | Other specified threats to breathing, street and highway, while engaged in leisure activity |
| W8342 | Other specified threats to breathing, street and highway, while working for income |
| W8343 | Other specified threats to breathing, street and highway, while engaged in other types of work |
| W8344 | Other specified threats to breathing, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W8348 | Other specified threats to breathing, street and highway, while engaged in other specified activities |
| W8349 | Other specified threats to breathing, street and highway, during unspecified activity |
| W835 | Other specified threats to breathing, trade and service area |
| W8350 | Other specified threats to breathing, trade and service area, while engaged in sports activity |
| W8351 | Other specified threats to breathing, trade and service area, while engaged in leisure activity |
| W8352 | Other specified threats to breathing, trade and service area, while working for income |
| W8353 | Other specified threats to breathing, trade and service area, while engaged in other types of work |
| W8354 | Other specified threats to breathing, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W8358 | Other specified threats to breathing, trade and service area, while engaged in other specified activities |
| W8359 | Other specified threats to breathing, trade and service area, during unspecified activity |
| W836 | Other specified threats to breathing, industrial and construction area |
| W8360 | Other specified threats to breathing, industrial and construction area, while engaged in sports activity |
| W8361 | Other specified threats to breathing, industrial and construction area, while engaged in leisure activity |
| W8362 | Other specified threats to breathing, industrial and construction area, while working for income |
| W8363 | Other specified threats to breathing, industrial and construction area, while engaged in other types of work |
| W8364 | Other specified threats to breathing, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W8368 | Other specified threats to breathing, industrial and construction area, while engaged in other specified activities |
| W8369 | Other specified threats to breathing, industrial and construction area, during unspecified activity |
| W837 | Other specified threats to breathing, farm |
| W8370 | Other specified threats to breathing, farm, while engaged in sports activity |
| W8371 | Other specified threats to breathing, farm, while engaged in leisure activity |
| W8372 | Other specified threats to breathing, farm, while working for income |
| W8373 | Other specified threats to breathing, farm, while engaged in other types of work |
| W8374 | Other specified threats to breathing, farm, while resting, sleeping, eating or engaging in other vital activities |
| W8378 | Other specified threats to breathing, farm, while engaged in other specified activities |
| W8379 | Other specified threats to breathing, farm, during unspecified activity |
| W838 | Other specified threats to breathing, other specified places |
| W8380 | Other specified threats to breathing, other specified places, while engaged in sports activity |
| W8381 | Other specified threats to breathing, other specified places, while engaged in leisure activity |
| W8382 | Other specified threats to breathing, other specified places, while working for income |
| W8383 | Other specified threats to breathing, other specified places, while engaged in other types of work |
| W8384 | Other specified threats to breathing, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W8388 | Other specified threats to breathing, other specified places, while engaged in other specified activities |
| W8389 | Other specified threats to breathing, other specified places, during unspecified activity |
| W839 | Other specified threats to breathing, unspecified place |
| W8390 | Other specified threats to breathing, unspecified place, while engaged in sports activity |
| W8391 | Other specified threats to breathing, unspecified place, while engaged in leisure activity |
| W8392 | Other specified threats to breathing, unspecified place, while working for income |
| W8393 | Other specified threats to breathing, unspecified place, while engaged in other types of work |
| W8394 | Other specified threats to breathing, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W8398 | Other specified threats to breathing, unspecified place, while engaged in other specified activities |
| W8399 | Other specified threats to breathing, unspecified place, during unspecified activity |
| W84 | Unspecified threat to breathing |
| W840 | Unspecified threat to breathing, home |
| W8400 | Unspecified threat to breathing, home, while engaged in sports activity |
| W8401 | Unspecified threat to breathing, home, while engaged in leisure activity |
| W8402 | Unspecified threat to breathing, home, while working for income |
| W8403 | Unspecified threat to breathing, home, while engaged in other types of work |
| W8404 | Unspecified threat to breathing, home, while resting, sleeping, eating or engaging in other vital activities |
| W8408 | Unspecified threat to breathing, home, while engaged in other specified activities |
| W8409 | Unspecified threat to breathing, home, during unspecified activity |
| W841 | Unspecified threat to breathing, residential institution |
| W8410 | Unspecified threat to breathing, residential institution, while engaged in sports activity |
| W8411 | Unspecified threat to breathing, residential institution, while engaged in leisure activity |
| W8412 | Unspecified threat to breathing, residential institution, while working for income |
| W8413 | Unspecified threat to breathing, residential institution, while engaged in other types of work |
| W8414 | Unspecified threat to breathing, residential institution, while resting, sleeping, eating or engaging in other vital activities |
| W8418 | Unspecified threat to breathing, residential institution, while engaged in other specified activities |
| W8419 | Unspecified threat to breathing, residential institution, during unspecified activity |
| W842 | Unspecified threat to breathing, school, other institution and public administrative area |
| W8420 | Unspecified threat to breathing, school, other institution and public administrative area, while engaged in sports activity |
| W8421 | Unspecified threat to breathing, school, other institution and public administrative area, while engaged in leisure activity |
| W8422 | Unspecified threat to breathing, school, other institution and public administrative area, while working for income |
| W8423 | Unspecified threat to breathing, school, other institution and public administrative area, while engaged in other types of work |
| W8424 | Unspecified threat to breathing, school, other institution and public administrative area, while resting, sleeping, eating or engaging in other vital activities |
| W8428 | Unspecified threat to breathing, school, other institution and public administrative area, while engaged in other specified activities |
| W8429 | Unspecified threat to breathing, school, other institution and public administrative area, during unspecified activity |
| W843 | Unspecified threat to breathing, sports and athletics area |
| W8430 | Unspecified threat to breathing, sports and athletics area, while engaged in sports activity |
| W8431 | Unspecified threat to breathing, sports and athletics area, while engaged in leisure activity |
| W8432 | Unspecified threat to breathing, sports and athletics area, while working for income |
| W8433 | Unspecified threat to breathing, sports and athletics area, while engaged in other types of work |
| W8434 | Unspecified threat to breathing, sports and athletics area, while resting, sleeping, eating or engaging in other vital activities |
| W8438 | Unspecified threat to breathing, sports and athletics area, while engaged in other specified activities |
| W8439 | Unspecified threat to breathing, sports and athletics area, during unspecified activity |
| W844 | Unspecified threat to breathing, street and highway |
| W8440 | Unspecified threat to breathing, street and highway, while engaged in sports activity |
| W8441 | Unspecified threat to breathing, street and highway, while engaged in leisure activity |
| W8442 | Unspecified threat to breathing, street and highway, while working for income |
| W8443 | Unspecified threat to breathing, street and highway, while engaged in other types of work |
| W8444 | Unspecified threat to breathing, street and highway, while resting, sleeping, eating or engaging in other vital activities |
| W8448 | Unspecified threat to breathing, street and highway, while engaged in other specified activities |
| W8449 | Unspecified threat to breathing, street and highway, during unspecified activity |
| W845 | Unspecified threat to breathing, trade and service area |
| W8450 | Unspecified threat to breathing, trade and service area, while engaged in sports activity |
| W8451 | Unspecified threat to breathing, trade and service area, while engaged in leisure activity |
| W8452 | Unspecified threat to breathing, trade and service area, while working for income |
| W8453 | Unspecified threat to breathing, trade and service area, while engaged in other types of work |
| W8454 | Unspecified threat to breathing, trade and service area, while resting, sleeping, eating or engaging in other vital activities |
| W8458 | Unspecified threat to breathing, trade and service area, while engaged in other specified activities |
| W8459 | Unspecified threat to breathing, trade and service area, during unspecified activity |
| W846 | Unspecified threat to breathing, industrial and construction area |
| W8460 | Unspecified threat to breathing, industrial and construction area, while engaged in sports activity |
| W8461 | Unspecified threat to breathing, industrial and construction area, while engaged in leisure activity |
| W8462 | Unspecified threat to breathing, industrial and construction area, while working for income |
| W8463 | Unspecified threat to breathing, industrial and construction area, while engaged in other types of work |
| W8464 | Unspecified threat to breathing, industrial and construction area, while resting, sleeping, eating or engaging in other vital activities |
| W8468 | Unspecified threat to breathing, industrial and construction area, while engaged in other specified activities |
| W8469 | Unspecified threat to breathing, industrial and construction area, during unspecified activity |
| W847 | Unspecified threat to breathing, farm |
| W8470 | Unspecified threat to breathing, farm, while engaged in sports activity |
| W8471 | Unspecified threat to breathing, farm, while engaged in leisure activity |
| W8472 | Unspecified threat to breathing, farm, while working for income |
| W8473 | Unspecified threat to breathing, farm, while engaged in other types of work |
| W8474 | Unspecified threat to breathing, farm, while resting, sleeping, eating or engaging in other vital activities |
| W8478 | Unspecified threat to breathing, farm, while engaged in other specified activities |
| W8479 | Unspecified threat to breathing, farm, during unspecified activity |
| W848 | Unspecified threat to breathing, other specified places |
| W8480 | Unspecified threat to breathing, other specified places, while engaged in sports activity |
| W8481 | Unspecified threat to breathing, other specified places, while engaged in leisure activity |
| W8482 | Unspecified threat to breathing, other specified places, while working for income |
| W8483 | Unspecified threat to breathing, other specified places, while engaged in other types of work |
| W8484 | Unspecified threat to breathing, other specified places, while resting, sleeping, eating or engaging in other vital activities |
| W8488 | Unspecified threat to breathing, other specified places, while engaged in other specified activities |
| W8489 | Unspecified threat to breathing, other specified places, during unspecified activity |
| W849 | Unspecified threat to breathing, unspecified place |
| W8490 | Unspecified threat to breathing, unspecified place, while engaged in sports activity |
| W8491 | Unspecified threat to breathing, unspecified place, while engaged in leisure activity |
| W8492 | Unspecified threat to breathing, unspecified place, while working for income |
| W8493 | Unspecified threat to breathing, unspecified place, while engaged in other types of work |
| W8494 | Unspecified threat to breathing, unspecified place, while resting, sleeping, eating or engaging in other vital activities |
| W8498 | Unspecified threat to breathing, unspecified place, while engaged in other specified activities |
| W8499 | Unspecified threat to breathing, unspecified place, during unspecified activity |
| W930 | Contact with or inhalation of dry ice |
| W9302 | Inhalation of dry ice |
| W9302XA | Inhalation of dry ice, initial encounter |
| W9302XD | Inhalation of dry ice, subsequent encounter |
| W9302XS | Inhalation of dry ice, sequela |
| W931 | Contact with or inhalation of liquid air |
| W9312 | Inhalation of liquid air |
| W9312XA | Inhalation of liquid air, initial encounter |
| W9312XD | Inhalation of liquid air, subsequent encounter |
| W9312XS | Inhalation of liquid air, sequela |
| X130 | Inhalation of steam and other hot vapors |
| X130XXA | Inhalation of steam and other hot vapors, initial encounter |
| X130XXD | Inhalation of steam and other hot vapors, subsequent encounter |
| X130XXS | Inhalation of steam and other hot vapors, sequela |
| X140 | Inhalation of hot air and gases |
| X140XXA | Inhalation of hot air and gases, initial encounter |
| X140XXD | Inhalation of hot air and gases, subsequent encounter |
| X140XXS | Inhalation of hot air and gases, sequela |
| Y3646 | War operations involving intentional restriction of air and airway |
| Y36460 | War operations involving intentional restriction of air and airway, military personnel |
| Y36460A | War operations involving intentional restriction of air and airway, military personnel, initial encounter |
| Y36460D | War operations involving intentional restriction of air and airway, military personnel, subsequent encounter |
| Y36460S | War operations involving intentional restriction of air and airway, military personnel, sequela |
| Y36461 | War operations involving intentional restriction of air and airway, civilian |
| Y36461A | War operations involving intentional restriction of air and airway, civilian, initial encounter |
| Y36461D | War operations involving intentional restriction of air and airway, civilian, subsequent encounter |
| Y36461S | War operations involving intentional restriction of air and airway, civilian, sequela |
| Y3647 | War operations involving unintentional restriction of air and airway |
| Y36470 | War operations involving unintentional restriction of air and airway, military personnel |
| Y36470A | War operations involving unintentional restriction of air and airway, military personnel, initial encounter |
| Y36470D | War operations involving unintentional restriction of air and airway, military personnel, subsequent encounter |
| Y36470S | War operations involving unintentional restriction of air and airway, military personnel, sequela |
| Y36471 | War operations involving unintentional restriction of air and airway, civilian |
| Y36471A | War operations involving unintentional restriction of air and airway, civilian, initial encounter |
| Y36471D | War operations involving unintentional restriction of air and airway, civilian, subsequent encounter |
| Y36471S | War operations involving unintentional restriction of air and airway, civilian, sequela |
| Y3746 | Military operations involving intentional restriction of air and airway |
| Y37460 | Military operations involving intentional restriction of air and airway, military personnel |
| Y37460A | Military operations involving intentional restriction of air and airway, military personnel, initial encounter |
| Y37460D | Military operations involving intentional restriction of air and airway, military personnel, subsequent encounter |
| Y37460S | Military operations involving intentional restriction of air and airway, military personnel, sequela |
| Y37461 | Military operations involving intentional restriction of air and airway, civilian |
| Y37461A | Military operations involving intentional restriction of air and airway, civilian, initial encounter |
| Y37461D | Military operations involving intentional restriction of air and airway, civilian, subsequent encounter |
| Y37461S | Military operations involving intentional restriction of air and airway, civilian, sequela |
| Y3747 | Military operations involving unintentional restriction of air and airway |
| Y37470 | Military operations involving unintentional restriction of air and airway, military personnel |
| Y37470A | Military operations involving unintentional restriction of air and airway, military personnel, initial encounter |
| Y37470D | Military operations involving unintentional restriction of air and airway, military personnel, subsequent encounter |
| Y37470S | Military operations involving unintentional restriction of air and airway, military personnel, sequela |
| Y37471 | Military operations involving unintentional restriction of air and airway, civilian |
| Y37471A | Military operations involving unintentional restriction of air and airway, civilian, initial encounter |
| Y37471D | Military operations involving unintentional restriction of air and airway, civilian, subsequent encounter |
| Y37471S | Military operations involving unintentional restriction of air and airway, civilian, sequela |
| Y480 | Adverse effects in the therapeutic use of inhaled anaesthetics |
| Z03822 | Encounter for observation for suspected aspirated (inhaled) foreign body ruled out |
| Z2911 | Encounter for prophylactic immunotherapy for respiratory syncytial virus (RSV) |
| Z3684 | Encounter for antenatal screening for fetal lung maturity |
| Z4824 | Encounter for aftercare following lung transplant |
| Z48280 | Encounter for aftercare following heart-lung transplant |
| Z7951 | Long term (current) use of inhaled steroids |
| Z801 | Family history of malignant neoplasm of trachea, bronchus and lung |
| Z825 | Family history of asthma and other chronic lower respiratory diseases |
| Z851 | Personal history of malignant neoplasm of trachea, bronchus and lung |
| Z8511 | Personal history of malignant neoplasm of bronchus and lung |
| Z85110 | Personal history of malignant carcinoid tumor of bronchus and lung |
| Z85118 | Personal history of other malignant neoplasm of bronchus and lung |
| Z8512 | Personal history of malignant neoplasm of trachea |
| Z85860 | Personal history of secondary malignant neoplasms of lung |
| Z8701 | Personal history of pneumonia (recurrent) |
| Z902 | Acquired absence of lung [part of] |
| Z92240 | Personal history of inhaled steroid therapy |
| Z942 | Lung transplant status |
| Z943 | Heart and lungs transplant status |
| Z983 | Post therapeutic collapse of lung status |
| Z991 | Dependence on respirator |
| Z9911 | Dependence on respirator [ventilator] status |
| Z9912 | Encounter for respirator [ventilator] dependence during power failure |

**ASTHMA**

## **Alternative Names for Asthma**

* Reactive Airway Disease (RAD)  
  Often used in children or when a definitive asthma diagnosis is not yet confirmed.
* Bronchial Asthma  
  A formal term emphasizing the involvement of the bronchi (airways).
* Allergic Asthma  
  Asthma triggered primarily by allergens such as pollen, dust mites, or pet dander.
* Non-Allergic Asthma  
  Asthma triggered by factors other than allergens, such as cold air, exercise, or irritants.
* Exercise-Induced Asthma (EIA) or Exercise-Induced Bronchoconstriction (EIB)  
  Asthma symptoms specifically triggered by physical activity.
* Cough-Variant Asthma  
  A type of asthma where the main symptom is a persistent cough rather than wheezing or breathlessness.
* Occupational Asthma  
  Asthma caused or worsened by workplace exposures to irritants or allergens.
* Intrinsic Asthma  
  Asthma not linked to allergic reactions, often developing in adulthood.
* Extrinsic Asthma  
  Asthma caused by external allergens, typically starting in childhood.

**DEFINITION AND DESCRIPTION**

Asthma is a condition in which your airways narrow and swell and may produce extra mucus. This can make breathing difficult and trigger coughing, a whistling sound (wheezing) when you breathe out and shortness of breath.

For some people, asthma is a minor nuisance. For others, it can be a major problem that interferes with daily activities and may lead to a life-threatening asthma attack.

Asthma can't be cured, but its symptoms can be controlled. Because asthma often changes over time, it's important that you work with your doctor to track your signs and symptoms and adjust your treatment as needed.

### **Types of asthma**

Types of asthma include:

* **Allergic asthma**: when allergies trigger asthma symptoms
* **Cough-variant asthma**: when your only asthma symptom is a cough
* **Exercise-induced asthma**: when exercise triggers asthma symptoms
* **Occupational asthma**: when substances you breathe in at work cause you to develop asthma or trigger asthma attacks
* **Asthma-COPD overlap syndrome (ACOS)**: when you have both asthma and COPD (chronic obstructive pulmonary disease)

## **Causes**

It isn't clear why some people get asthma and others don't, but it's probably due to a combination of environmental and inherited (genetic) factors.

### **Asthma triggers**

Exposure to various irritants and substances that trigger allergies (allergens) can trigger signs and symptoms of asthma. Asthma triggers are different from person to person and can include:

* Airborne allergens, such as pollen, dust mites, mold spores, pet dander or particles of cockroach waste
* Respiratory infections, such as the common cold
* Physical activity
* Cold air
* Air pollutants and irritants, such as smoke
* Certain medications, including beta blockers, aspirin, and nonsteroidal anti-inflammatory drugs, such as ibuprofen (Advil, Motrin IB, others) and naproxen sodium (Aleve)
* Strong emotions and stress
* Sulfites and preservatives added to some types of foods and beverages, including shrimp, dried fruit, processed potatoes, beer and wine
* Gastroesophageal reflux disease (GERD), a condition in which stomach acids back up into your throat

**Risk factors**

Several factors are thought to increase your chances of developing asthma. They include:

* Having a blood relative with asthma, such as a parent or sibling
* Having another allergic condition, such as atopic dermatitis — which causes red, itchy skin — or hay fever — which causes a runny nose, congestion and itchy eyes
* Being overweight
* Being a smoker
* Exposure to secondhand smoke
* Exposure to exhaust fumes or other types of pollution
* Exposure to occupational triggers, such as chemicals used in farming, hairdressing and manufacturing

**Symptoms**

Asthma symptoms vary from person to person. You may have infrequent asthma attacks, have symptoms only at certain times — such as when exercising — or have symptoms all the time.

Asthma signs and symptoms include:

* Shortness of breath
* Chest tightness or pain
* Wheezing when exhaling, which is a common sign of asthma in children
* Trouble sleeping caused by shortness of breath, coughing or wheezing
* Coughing or wheezing attacks that are worsened by a respiratory virus, such as a cold or the flu

Signs that your asthma is probably worsening include:

* Asthma signs and symptoms that are more frequent and bothersome
* Increasing difficulty breathing, as measured with a device used to check how well your lungs are working (peak flow meter)
* The need to use a quick-relief inhaler more often

For some people, asthma signs and symptoms flare up in certain situations:

* **Exercise-induced asthma,** which may be worse when the air is cold and dry
* **Occupational asthma,** triggered by workplace irritants such as chemical fumes, gases or dust
* **Allergy-induced asthma,** triggered by airborne substances, such as pollen, mold spores, cockroach waste, or particles of skin and dried saliva shed by pets (pet dander)

### **When to see a doctor**

Severe asthma attacks can be life-threatening. Work with your doctor to determine what to do when your signs and symptoms worsen — and when you need emergency treatment. Signs of an asthma emergency include:

* Rapid worsening of shortness of breath or wheezing
* No improvement even after using a quick-relief inhaler
* Shortness of breath when you are doing minimal physical activity

See your doctor:

* **If you think you have asthma.** If you have frequent coughing or wheezing that lasts more than a few days or any other signs or symptoms of asthma, see your doctor. Treating asthma early may prevent long-term lung damage and help keep the condition from getting worse over time.
* **To monitor your asthma after diagnosis.** If you know you have asthma, work with your doctor to keep it under control. Good long-term control helps you feel better from day to day and can prevent a life-threatening asthma attack.
* **If your asthma symptoms get worse.** Contact your doctor right away if your medication doesn't seem to ease your symptoms or if you need to use your quick-relief inhaler more often.

Don't take more medication than prescribed without consulting your doctor first. Overusing asthma medication can cause side effects and may make your asthma worse.

* **To review your treatment.** Asthma often changes over time. Meet with your doctor regularly to discuss your symptoms and make any needed treatment adjustments.

## **Diagnosis and test**

### **Physical exam**

Your doctor will perform a physical exam to rule out other possible conditions, such as a respiratory infection or chronic obstructive pulmonary disease (COPD). Your doctor will also ask you questions about your signs and symptoms and about any other health problems.

You may be given lung function tests to determine how much air moves in and out as you breathe. These tests may include:

* **Spirometry.** This test estimates the narrowing of your bronchial tubes by checking how much air you can exhale after a deep breath and how fast you can breathe out.
* **Peak flow.** A peak flow meter is a simple device that measures how hard you can breathe out. Lower than usual peak flow readings are a sign that your lungs may not be working as well and that your asthma may be getting worse. Your doctor will give you instructions on how to track and deal with low peak flow readings.

Lung function tests often are done before and after taking a medication to open your airways called a bronchodilator (brong-koh-DIE-lay-tur), such as albuterol. If your lung function improves with use of a bronchodilator, it's likely you have asthma.

Other tests to diagnose asthma include:

* **Methacholine challenge.** Methacholine is a known asthma trigger. When inhaled, it will cause your airways to narrow slightly. If you react to the methacholine, you likely have asthma. This test may be used even if your initial lung function test is normal.
* **Imaging tests.** A chest X-ray can help identify any structural abnormalities or diseases (such as infection) that can cause or aggravate breathing problems.
* **Allergy testing.** Allergy tests can be performed by a skin test or blood test. They tell you if you're allergic to pets, dust, mold or pollen. If allergy triggers are identified, your doctor may recommend allergy shots.
* **Nitric oxide test.** This test measures the amount of the gas nitric oxide in your breath. When your airways are inflamed — a sign of asthma — you may have higher than normal nitric oxide levels. This test isn't widely available.
* **Sputum eosinophils.** This test looks for certain white blood cells (eosinophils) in the mixture of saliva and mucus (sputum) you discharge during coughing. Eosinophils are present when symptoms develop and become visible when stained with a rose-colored dye.
* **Provocative testing for exercise and cold-induced asthma.** In these tests, your doctor measures your airway obstruction before and after you perform vigorous physical activity or take several breaths of cold air.

### **How asthma is classified**

To classify your asthma severity, your doctor will consider how often you have signs and symptoms and how severe they are. Your doctor will also consider the results of your physical exam and diagnostic tests.

Determining your asthma severity helps your doctor choose the best treatment. Asthma severity often changes over time, requiring treatment adjustments.

Asthma is classified into four general categories:

|  |  |
| --- | --- |
| **Asthma classification** | **Signs and symptoms** |
| Mild intermittent | Mild symptoms up to two days a week and up to two nights a month |
| Mild persistent | Symptoms more than twice a week, but no more than once in a single day |
| Moderate persistent | Symptoms once a day and more than one night a week |
| Severe persistent | Symptoms throughout the day on most days and frequently at night |

## **Treatment**

Prevention and long-term control are key to stopping asthma attacks before they start. Treatment usually involves learning to recognize your triggers, taking steps to avoid triggers and tracking your breathing to make sure your medications are keeping symptoms under control. In case of an asthma flare-up, you may need to use a quick-relief inhaler.

### **Medications**

The right medications for you depend on a number of things — your age, symptoms, asthma triggers and what works best to keep your asthma under control.

Preventive, long-term control medications reduce the swelling (inflammation) in your airways that leads to symptoms. Quick-relief inhalers (bronchodilators) quickly open swollen airways that are limiting breathing. In some cases, allergy medications are necessary.

**Long-term asthma control medications,** generally taken daily, are the cornerstone of asthma treatment. These medications keep asthma under control on a day-to-day basis and make it less likely you'll have an asthma attack. Types of long-term control medications include:

* **Inhaled corticosteroids.** These medications include fluticasone propionate (Flovent HFA, Flovent Diskus, Xhance), budesonide (Pulmicort Flexhaler, Pulmicort Respules, Rhinocort), ciclesonide (Alvesco), beclomethasone (Qvar Redihaler), mometasone (Asmanex HFA, Asmanex Twisthaler) and fluticasone furoate (Arnuity Ellipta).

You may need to use these medications for several days to weeks before they reach their maximum benefit. Unlike oral corticosteroids, inhaled corticosteroids have a relatively low risk of serious side effects.

* **Leukotriene modifiers.** These oral medications — including montelukast (Singulair), zafirlukast (Accolate) and zileuton (Zyflo) — help relieve asthma symptoms.

Montelukast has been linked to psychological reactions, such as agitation, aggression, hallucinations, depression and suicidal thinking. Seek medical advice right away if you experience any of these reactions.

* **Combination inhalers.** These medications — such as fluticasone-salmeterol (Advair HFA, Airduo Digihaler, others), budesonide-formoterol (Symbicort), formoterol-mometasone (Dulera) and fluticasone furoate-vilanterol (Breo Ellipta) — contain a long-acting beta agonist along with a corticosteroid.
* **Theophylline.** Theophylline (Theo-24, Elixophyllin, Theochron) is a daily pill that helps keep the airways open by relaxing the muscles around the airways. It's not used as often as other asthma medications and requires regular blood tests.

**Quick-relief (rescue) medications** are used as needed for rapid, short-term symptom relief during an asthma attack. They may also be used before exercise if your doctor recommends it. Types of quick-relief medications include:

* **Short-acting beta agonists.** These inhaled, quick-relief bronchodilators act within minutes to rapidly ease symptoms during an asthma attack. They include albuterol (ProAir HFA, Ventolin HFA, others) and levalbuterol (Xopenex, Xopenex HFA).

Short-acting beta agonists can be taken using a portable, hand-held inhaler or a nebulizer, a machine that converts asthma medications to a fine mist. They're inhaled through a face mask or mouthpiece.

* **Anticholinergic agents.** Like other bronchodilators, ipratropium (Atrovent HFA) and tiotropium (Spiriva, Spiriva Respimat) act quickly to immediately relax your airways, making it easier to breathe. They're mostly used for emphysema and chronic bronchitis but can be used to treat asthma.
* **Oral and intravenous corticosteroids.** These medications — which include prednisone (Prednisone Inten sol, Rayos) and methylprednisolone (Medrol, Depo-Medrol, Solu-Medrol) — relieve airway inflammation caused by severe asthma. They can cause serious side effects when used long term, so these drugs are used only on a short-term basis to treat severe asthma symptoms.

If you have an asthma flare-up, a quick-relief inhaler can ease your symptoms right away. But you shouldn't need to use your quick-relief inhaler very often if your long-term control medications are working properly.

Keep a record of how many puffs you use each week. If you need to use your quick-relief inhaler more often than your doctor recommends, see your doctor. You probably need to adjust your long-term control medication.

**Allergy medications** may help if your asthma is triggered or worsened by allergies. These include:

* **Allergy shots (immunotherapy).** Over time, allergy shots gradually reduce your immune system reaction to specific allergens. You generally receive shots once a week for a few months, then once a month for a period of three to five years.
* **Biologics.** These medications — which include omalizumab (Xolair), mepolizumab (Nucala), dupilumab (Dupixent), reslizumab (Cinqair), benralizumab (Fasenra) and tezepelumab (Tezspire) — are specifically for people who have severe asthma.

### **Bronchial thermoplasty**

This treatment is used for severe asthma that doesn't improve with inhaled corticosteroids or other long-term asthma medications. It isn't widely available nor right for everyone.

During bronchial thermoplasty, your doctor heats the insides of the airways in the lungs with an electrode. The heat reduces the smooth muscle inside the airways. This limits the ability of the airways to tighten, making breathing easier and possibly reducing asthma attacks. The therapy is generally done over three outpatient visits.

### **Treat by severity for better control: A stepwise approach**

Your treatment should be flexible and based on changes in your symptoms. Your doctor should ask about your symptoms at each visit. Based on your signs and symptoms, your doctor can adjust your treatment accordingly.

For example, if your asthma is well controlled, your doctor may prescribe less medication. If your asthma isn't well controlled or is getting worse, your doctor may increase your medication and recommend more-frequent visits.

### **Asthma action plan**

Work with your doctor to create an asthma action plan that outlines in writing when to take certain medications or when to increase or decrease the dose of your medications based on your symptoms. Also include a list of your triggers and the steps you need to take to avoid them.

Your doctor may also recommend tracking your asthma symptoms or using a peak flow meter on a regular basis to monitor how well your treatment is controlling your asthma.

## A**sthma Treatment Side Effects**

## 1. Inhaled Corticosteroids (Preventers)

* Examples: Beclomethasone, Budesonide, Fluticasone
* Common Side Effects:
  + Hoarse voice
  + Sore mouth or throat
  + Oral thrush (fungal infection)
* Prevention:
  + Use proper inhaler technique
  + Rinse and gargle mouth after use
  + Use spacer devices to reduce oropharyngeal deposition
* Rare/Long-term Risks:
  + Possible risk of osteoporosis with high-dose long-term use
  + Increased risk of pneumonia or nontuberculous mycobacterial infections in older adults

## 2. Short-Acting Beta-2 Agonists (Relievers)

* Examples: Albuterol (Salbutamol), Levalbuterol
* Common Side Effects:
  + Tremor or shakiness
  + Rapid or irregular heartbeat (tachycardia, palpitations)
  + Headache
  + Throat or nasal irritation
  + Muscle cramps
* Notes:
  + Side effects often resolve quickly
  + Overuse can worsen asthma control and increase sensitivity to triggers

## 3. Long-Acting Beta-2 Agonists (LABAs)

* Used in combination with inhaled corticosteroids for maintenance
* Side effects similar to short-acting beta-2 agonists but less frequent

## 4. Leukotriene Receptor Antagonists

* Example: Montelukast
* Possible Side Effects:
  + Mood changes (anxiety, depression)
  + Nightmares, sleep disturbances
* Note:
  + Rare but serious neuropsychiatric effects reported, especially in children
  + Report any behavioral changes to your doctor immediately

## 5. Anticholinergic Bronchodilators

* Examples: Ipratropium, Tiotropium
* Side Effects:
  + Dry mouth
  + Difficulty urinating (especially in patients with prostate issues)

## 6. Theophylline

* Side Effects:
  + Nausea, vomiting
  + Headache
  + Insomnia
  + Cardiac arrhythmias (at high doses)
  + Seizures (rare)

## 7. Cromolyn Sodium (Mast Cell Stabilizer)

* Side Effects:
  + Cough
  + Throat irritation
  + Rarely, headache or nausea

**Lifestyle and home remedies**

Although many people with asthma rely on medications to prevent and relieve symptoms, you can do several things on your own to maintain your health and lessen the possibility of asthma attacks.

### **Avoid your triggers**

Taking steps to reduce your exposure to asthma triggers is a key part of asthma control. To reduce your exposure, you should:

* **Use your air conditioner.** Air conditioning reduces the amount of airborne pollen from trees, grasses and weeds that finds its way indoors. Air conditioning also lowers indoor humidity and can reduce your exposure to dust mites. If you don't have air conditioning, try to keep your windows closed during pollen season.
* **Decontaminate your decor.** Minimize dust that may worsen nighttime symptoms by replacing certain items in your bedroom. For example, encase pillows, mattresses and box springs in dustproof covers. Avoid using down-filled pillows and blankets. Throughout the house, remove carpeting and install hardwood or linoleum flooring. Use washable curtains and blinds.
* **Maintain optimal humidity.** If you live in a damp climate, talk to your doctor about using a dehumidifier.
* **Prevent mold spores.** Clean damp areas in the bathroom, kitchen and around the house to keep mold spores from developing. Get rid of moldy leaves or damp firewood in the yard.
* **Reduce pet dander.** If you're allergic to dander, avoid pets with fur or feathers. Having pets regularly bathed or groomed may also reduce the amount of dander in your surroundings.
* **Clean regularly.** Clean your home at least once a week. If you're likely to stir up dust, wear a mask or have someone else do the cleaning. Wash your bedding regularly.
* **Cover your nose and mouth if it's cold out.** If your asthma is worsened by cold or dry air, wearing a face mask can help.

### **Stay healthy**

Taking care of yourself can help keep your symptoms under control, including:

* **Get regular exercise.** Having asthma doesn't mean you have to be less active. Treatment can prevent asthma attacks and control symptoms during activity.

Regular exercise can strengthen your heart and lungs, which helps relieve asthma symptoms. If you exercise in cold temperatures, wear a face mask to warm the air you breathe.

* **Maintain a healthy weight.** Being overweight can worsen asthma symptoms, and it puts you at higher risk of other health problems.
* **Control heartburn and gastroesophageal reflux disease (GERD).** It's possible that the acid reflux that causes heartburn may damage lung airways and worsen asthma symptoms. If you have frequent or constant heartburn, talk to your doctor about treatment options. You may need treatment for GERD before your asthma symptoms improve.

## **Alternative medicine**

Certain alternative treatments may help with asthma symptoms. However, keep in mind that these treatments are not a replacement for medical treatment, especially if you have severe asthma. Talk to your doctor before taking any herbs or supplements, as some may interact with the medications you take.

In most cases, more research is needed to see how well alternative remedies work and to measure the extent of possible side effects. Alternative asthma treatments include:

* **Breathing exercises.** These exercises may reduce the amount of medication you need to keep your asthma symptoms under control.
* **Herbal and natural remedies.** A few herbal and natural remedies that may help improve asthma symptoms include black seed, caffeine, choline and pycnogenol.

## **Outlook / Prognosis**

Most people with asthma can manage their symptoms. Asthma management means you:

* Can do the things you want to do at work and home
* Have no (or minimal) asthma symptoms
* Rarely need to use your rescue inhaler
* Can sleep without asthma symptoms waking you up
* Don’t need oral steroids for flare-ups more than twice a year

Some people can avoid triggers and have no symptoms most of the time. Others need to use a maintenance inhaler or other medications in addition to avoiding triggers. Kids may have fewer or no symptoms as they get older and their airways get bigger.

You might be able to reduce or avoid asthma symptoms with a few everyday habits. These include:

* Avoid triggers whenever possible. It might be helpful to keep a symptoms journal to figure out what makes your symptoms worse.
* Be physically active to a level that’s right for you. Ask your provider what they recommend. A pulmonary rehabilitation program might help.
* Don’t smoke or vape.
* Let your provider know if you’re unable to use inhalers or take medication as prescribed.

## **Diagnostic Considerations**

## Vocal cord dysfunction or inducible laryngeal obstruction (ILO)

Vocal cord dysfunction may exist alone or with asthma, it is caused by paradoxical adduction of the vocal cords during inspiration, and may disappear with panting, speech, or laughing.Patients with chronic symptoms suggestive of asthma, normal spirometry, poor response to asthma medications, and frequent evaluations should be evaluated for vocal cord dysfunction.Usually, the diagnosis can be made using direct laryngoscopy, but only during symptomatic periods or after exercise. The presence of flattening of the inspiratory limb of the flow-volume loop may also suggest vocal cord dysfunction, but this is only seen in 28% of patients at baseline.

## Tracheal and bronchial lesions

A variety of airway tumors are reported to manifest with symptoms like those of asthma. These tumors include endobronchial carcinoid and mucoepidermoid tumors, as shown in the images below. In one case, a 14-year-old boy with hyperlucency in the left lung was ultimately found to have a bronchial carcinoid in the left mainstem bronchus.

Other tracheal lesions can include bronchocentric granulomatosis, subglottic stenosis, subglottic web, tracheal hamartoma, bronchogenic cysts, leiomyoma, and tracheobronchopathia osteoplastica. All these types of tracheal lesions have been reported with symptoms similar to asthma.

## **Foreign bodies**

Foreign body aspiration may cause not only localized wheezing but also generalized wheezing. Wheezing occurs in toddlers as well as in adults. As described in one patient, foreign body aspiration may necessitate bronchoscopic retrieval before the patient even recalls the inciting event, and as many as 25% of patients may never recall the event.Furthermore, aspirated foreign bodies may be radiolucent and therefore not be visible on a chest radiograph. Radiography may show unilateral hyperinflation (from air trapping), infiltrate (from occlusion of a bronchus), or may be normal.

## **Pulmonary migraine**

Pulmonary migraine consists of combined recurrent asthma; cough with thick mucoid sputum; lower back pain radiating to the shoulder; subtotal or total atelectasis of a segment or lobe; and, occasionally, nausea with vomiting.The symptoms are often accompanied closely in time by focal headache. Spastic narrowing of the bronchi is postulated—along with retained mucous secretions, smooth muscle hypertrophy, and thickened bronchial walls—to cause expiratory collapse of selected airways. Cerebral and abdominal vascular migraine episodes are believed to accompany pulmonary migraine.

## **Congestive heart failure**

Congestive heart failure causes engorged pulmonary vessels and interstitial pulmonary edema, which reduce lung compliance and contribute to the sensation of dyspnea and wheezing. Cardiac asthma is characterized by wheezing secondary to bronchospasm in congestive heart failure, and it is related to paroxysmal nocturnal dyspnea and nocturnal coughing.

## **Diffuse pan bronchiolitis**

Diffuse pan bronchiolitis is prevalent in Japan and the Far East, and it may mimic bronchial asthma with wheezing, coughing, dyspnea on exertion, and sinusitis.High-resolution CT (HRCT) findings include centrilobular nodules and linear markings that usually are more profuse than the multifocal bronchiolar impaction sometimes observed with asthma.

## **Aortic arch anomalies**

Aortic arch anomalies may occur later in adulthood. In one case, the anomalies, which simulated exercise-induced asthma, were noticed first in a young woman only after a vigorous exercise program.On testing, the flow-volume display of this patient suggested an intrathoracic obstruction. The patient had a right aortic arch with ligamentum arteriosum that extended anterior to the trachea. This condition caused constriction when increased pulmonary blood flow, oxygen demand, and tracheal airflow and decreased intratracheal pressure from downstream turbulence distal to the tracheal ring occurred with exercise; combined, these factors produced wheezing and dyspnea.

## **Sinus disease**

Sinus disease, especially in children, is associated with bronchial asthma and wheezing. Although the association is not strong in patients with CT evidence of mild sinus mucosal thickening, a scoring system developed by Newman et al showed that extensive sinus disease was correlated with a substantially higher extent of wheezing than that in patients with only mild thickening. Of 104 adults, 39% had extensive disease, as visualized on CT scans, which was correlated with asthma and peripheral eosinophilia.

## **Gastroesophageal reflux**

Cough, recurrent bronchitis, pneumonia, wheezing, and asthma are associated with gastroesophageal reflux (GER).The incidence of GER in patients with asthma ranges from 38% in patients with only asthma symptoms to 48% in patients with recurrent pneumonia. Scintigraphic studies performed after technetium-99m sulfur-colloid ingestion have shown radionuclide activity in the lungs the next day, but no causal relationship between reflux and asthma has been established. Nevertheless, evidence suggests that increased pulmonary resistance occurs with symptoms of reflux during acid provocation testing; as some have suggested, the changes may be sufficiently significant to produce clinically evident bronchoconstriction.

## **Other conditions and factors**

Other extrinsic conditions, such as lymphadenopathy from sarcoidosis or Hodgkin lymphoma of the upper mediastinum, can contribute to asthma. In addition, aspirin or NSAID hypersensitivity and reactive airways dysfunction syndrome may be mistaken for asthma. Misdiagnoses as refractory bronchial asthma has resulted in inappropriate long-term treatment with corticosteroids.

A significant history of smoking greater than 20-pack years should make the diagnosis of chronic obstructive pulmonary disease (COPD) a stronger consideration than asthma.

Consideration for alternative diagnoses should be given in all patients, and in particular in those older than 30 years and younger than 2 years with new symptoms suggestive of asthma. An absence of airway obstruction on initial spirometry findings should prompt consideration for alternative diagnoses and additional testing.

## **Differential Diagnoses**

* Allergic and Environmental Asthma
* Alpha1-Antitrypsin (AAT) Deficiency
* Aspergillosis
* Bronchiectasis
* Bronchiolitis
* Chronic Obstructive Pulmonary Disease (COPD)
* Chronic Sinusitis
* Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss Syndrome)
* Cystic Fibrosis
* Exercise-Induced Anaphylaxis
* Food-Dependent Exercise-Induced Anaphylaxis (FDEIA)
* Foreign Body Aspiration
* Gastroesophageal Reflux Disease
* Heart Failure
* Pediatric Airway Foreign Body
* Pediatric Tracheomalacia
* Pulmonary Embolism (PE)
* Pulmonary Eosinophilia
* Sarcoidosis
* Upper Respiratory Tract Infection
* Vocal Cord Dysfunction
* Wheat-Dependent Exercise-Induced Anaphylaxis (WDEIA)

## **Epidemiology**

Asthma affects 5-10% of the US population or an estimated 25 million persons, including 4.7 million children.The overall prevalence rate of exercise-induced bronchospasm is 3-10% of the general population if persons who do not have asthma or allergy are excluded, but the rate increases to 12-15% of the general population if patients with underlying asthma are included. Asthma affects an estimated 300 million individuals worldwide. Annually, the World Health Organization (WHO) has estimated that 15 million disability-adjusted life-years are lost, and 250,000 asthma deaths are reported worldwide.

In the United States, asthma prevalence, especially morbidity and mortality, is higher in blacks than in whites. Although genetic factors are of major importance in determining a predisposition to the development of asthma, environmental factors play a greater role than racial factors in asthma onset. A national concern is that some of the increased morbidity is due to differences in asthma treatment afforded certain minority groups. Larger asthma-associated lung function deficits are reported in Hispanics, especially females.

Asthma is common in industrialized nations such as Canada, England, Australia, Germany, and New Zealand, where much of the asthma data have been collected. The prevalence rate of severe asthma in industrialized countries ranges from 2-10%. Trends suggest an increase in both the prevalence and morbidity of asthma, especially in children younger than 6 years. Factors that have been implicated include urbanization, air pollution, passive smoking, and change in exposure to environmental allergens.

Asthma predominantly occurs in boys in childhood, with a male-to-female ratio of 2:1 until puberty, when the male-to-female ratio becomes 1:1. Asthma prevalence is greater in females after puberty, and the majority of adult-onset cases diagnosed in persons older than 40 years occur in females. Boys are more likely than girls to experience a decrease in symptoms by late adolescence.

Asthma prevalence is increased in very young persons and very old persons because of airway responsiveness and lower levels of lung function.Two thirds of all asthma cases are diagnosed before the patient is aged 18 years. Approximately half of all children diagnosed with asthma have a decrease or disappearance of symptoms by early adulthood

**RECENT GUIDELINE**

The 2024 Global Initiative for Asthma (GINA) guidelines categorize asthma severity as mild, moderate, or severe. Severity is assessed retrospectively from the level of treatment required to control symptoms and exacerbations, as follows:

* Mild asthma: Well-controlled with low-intensity treatment such as as-needed reliever medication alone or with low-intensity controller treatment such as low-dose inhaled corticosteroids (ICSs)
* Moderate asthma: Well-controlled with low- or medium-dose ICS/long-acting beta2-agonists (LABA)
* Severe asthma: Requires high-dose ICS/LABA to prevent it from becoming uncontrolled, or asthma that remains uncontrolled despite this treatment

European Respiratory Society/American Thoracic Society (ERS/ATS) guidelines on evaluation and treatment of severe asthma reserves the definition of severe asthma for patients with refractory asthma and those in whom response to treatment of comorbidities is incomplete.

The 2022 GAN guidelines stress the importance of distinguishing between severe asthma and uncontrolled asthma, as the latter is a much more common reason for persistent symptoms and exacerbations, and it may be more easily improved. The most common problems that need to be excluded before a diagnosis of severe asthma can be made are the following:

* Poor inhaler technique
* Poor medication adherence
* Incorrect diagnosis of asthma, with symptoms due to alternative conditions such as upper airway dysfunction, cardiac failure, or lack of fitness
* Comorbidities and complicating conditions such as rhinosinusitis, gastroesophageal reflux, obesity, and obstructive sleep apnea
* Ongoing exposure to sensitizing or irritant agents in the home or work environment.

## **Management Guidelines**

Management and Prevention" include the following:

* Achieve and maintain control of asthma symptoms
* Maintain normal activity levels, including exercise
* Maintain pulmonary function as close to normal as possible
* Prevent asthma exacerbations
* Avoid adverse effects from asthma medications
* Prevent asthma mortality

### **Stepwise pharmacologic therapy**

The pharmacologic treatment of asthma is based on stepwise therapy. Asthma medications should be added or deleted as the frequency and severity of the patient's symptoms change.

Step 1 for intermittent asthma is as follows:

* Controller medication not indicated
* Reliever medication is a short-acting beta-agonist (SABA) as needed for symptoms

Step 2 for mild persistent asthma is as follows:

* Preferred controller medication is a low-dose inhaled corticosteroid
* Alternatives include cromolyn, leukotriene receptor antagonist (LTRA),or theophylline

Step 3 for moderate persistent asthma is as follows:

* Preferred controller medication is either a low-dose inhaled corticosteroid (ICS) plus a long-acting beta-agonist (LABA) (combination medication is the preferred choice to improve compliance) or an inhaled medium-dose corticosteroid
* Alternatives include a low-dose ICS plus either an LTRA or theophylline

Step 4 for moderate-to-severe persistent asthma is as follows:

* Preferred controller medication is an inhaled medium-dose corticosteroid plus a LABA (combination therapy)
* Alternatives include an inhaled medium-dose corticosteroid plus either an LTRA or theophylline

Step 5 for severe persistent asthma is as follows:

* Preferred controller medication is an inhaled high-dose corticosteroid plus LABA

Step 6 for severe persistent asthma is as follows:

* Preferred controller medication is an inhaled high-dose corticosteroid plus LABA plus oral corticosteroid

The 2024 GINA guidelines employ a stepwise, two track system depending on the choice of medications. For safety reasons, treatment with short-acting beta2-agonists (SABA) only is no longer recommended. To reduce risk of serious exacerbations and to control symptoms, all adults and adolescents with asthma should receive controller treatment containing inhaled corticosteroids (ICS).

For mild asthma, as-needed low-dose ICS and low-dose formoterol are recommended. If formoterol is not available, the patient should take low-dose ICS whenever SABA is taken. ICS-containing treatment should be initiated as soon as possible after asthma diagnosis. Asthma medications should be added or deleted as the frequency and severity of the patient's symptoms change. Track 1 is the preferred management approach with low dose ICS-formoterol taken at any step when the patient is symptomatic. The steps are as follows:

* Step 1-2: As-needed low-dose ICS-formoterol
* Step 3: Daily low-dose ICS-formoterol as maintenance and reliever
* Step 4: Daily medium-dose ICS-formoterol as maintenance and reliever
* Step 5: Refer for expert investigation and add-on LAMA treatment

The Track 2 approach is the alternative if Track 1 is not possible, or if a patient is stable, with good adherence and no exacerbations in the past year on their current therapy. The steps are as follows:

* Step 1: As-needed SABA and a low dose ICS are taken together (in combination, or with the ICS taken right after the SABA)
* Step 2: Daily low-dose ICS as maintenance plus as-needed SABA as reliever
* Step 3: Daily low-dose ICS-LABA as maintenance plus as-needed SABA as reliever
* Step 4: Medium- to high-dose ICS-LABA as maintenance plus as-needed SABA as reliever
* Step 5: Refer for expert investigation and add-on LAMA treatment
* For severe allergic asthma, a therapeutic trial of omalizumab
* Do not use methotrexate or macrolide antibiotics to treat severe asthma
* For severe asthma and recurrent exacerbations of allergic bronchopulmonary aspergillosis (ABPA), antifungal agents should be given
* Do not use antifungal agents for severe asthma without ABPA irrespective of sensitization to fungi (ie, positive skin prick test or fungus-specific immunoglobulin E in serum)

***Diagnosis***

The JSA recommends spirometry for assessing the extent of airflow limitation or airway reversibility.

The JSA recommends daily measurement of peak expiratory flow for unstable asthma and patients lacking obvious dyspnea during attack.

Although useful for diagnosing asthma, the JSA does not recommend assessing bronchial hyperresponsiveness in patients with low FEV1 (≤1 L) or low %FEV1 (≤50%) since excess airway narrowing may occur due to irritant inhalation.

*Treatment of long-term adult asthma*

The JSA recommends using a jet nebulizer for budesonide (BUD) inhalation suspension.

The JSA recommends adding one or more agents other than inhaled corticosteroids (ICSs), as opposed to increasing the dose of an ICS, to control asthma.

The JSA recommends long-acting β2-agonists (LABAs), leukotriene receptor antagonists (LTRAs), sustained-release theophylline, and long-acting muscarinic antagonists as add-on drugs.

The JSA recommends that anti-immunoglobulin E antibodies and other biologics as well as oral steroids be reserved for very severe and persistent asthma related to allergic reactions.

The JSA recommends inhaled β2-agonists, aminophylline, corticosteroids, adrenaline, oxygen therapy, and other approaches be used as needed during acute exacerbations.

***Treatment during pregnancy***

The JSA recommends ICSs as first-line treatment for long-term management of pregnant women with asthma.

The JSA recommends a short-acting beta-agonist (SABA) as needed for pregnant women with mild intermittent asthma.

The JSA recommends low-dose ICS; LTRA, controlled-release theophylline, and/or disodium cromoglycate (DSCG) as needed in pregnant women with mild persistent asthma.

The JSA recommends low-dose ICS and LABA or moderate-dose ICS and LABA in combination with LTRA or controlled-release theophylline as needed in pregnant women with moderate persistent asthma.

The JSA recommends high-dose ICS and LABA; oral steroids as needed for pregnant women with severe persistent asthma.

## **Exercise-Induced Asthma Guidelines**

**Guidelines for the management of exercise-induced bronchoconstriction** (EIB), which included the following recommendations:

* Administration of an inhaled SABA before exercise (strong recommendation); the SABA is typically administered 15 minutes before exercise
* A controller agent is added whenever SABA therapy is used daily or more frequently
* Interval or combination warm-up exercise before planned exercise (strong recommendation)
* Recommend against daily use of an inhaled long-acting beta2-agonist as single therapy (strong recommendation)
* For patients who continue to have symptoms despite using an inhaled SABA before exercise or who require an inhaled SABA daily or more frequently: (1) Daily ICS (strong recommendation), (2) Daily administration of an LTRA (strong recommendation), (3) Administration of a mast cell‒stabilizing agent before exercise (strong recommendation), and (4) Inhaled anticholinergic agent before exercise (weak recommendation)
* For patients with EIB and allergies who continue to have symptoms despite using an inhaled SABA before exercise or who require an inhaled SABA daily or more frequently consider administration of an antihistamine (weak recommendation)
* For exercise in cold weather, routine use of a device (eg, mask) that warms and humidifies the air during exercise (weak recommendation)

**GENOMIC DATA**

Key genomic findings include variants in genes such as:

* IL1RL1, IL33, HLA-DQA1, GSDMB, FLG, TSLP, STAT6, and SMAD3, among others, which are associated with asthma risk, age of onset, and severity.
* These genes often relate to immune system regulation, airway epithelium function, and inflammatory pathways, highlighting the immune system's critical role in asthma pathogenesis.

Recent whole-genome sequencing studies focusing on moderate-to-severe asthma have shown that genetic risk correlates with lung function measures and that different genetic components contribute independently to asthma susceptibility.

Despite these advances, the known genetic variants explain only a portion of asthma heritability, prompting ongoing research integrating multiomics data to better understand asthma's genetic architecture and improve personalized diagnosis and treatment

**PREDEFINED Q AND A**

For asthma, some basic questions to ask your doctor include:

* Is asthma the most likely cause of my breathing problems?

If you are experiencing breathing problems, asthma is a common and likely cause, but it is important to confirm this with proper evaluation because other conditions can cause similar symptoms

* Other than the most likely cause, what are other possible causes for my symptoms?

## Other Possible Causes of Breathing Problems

* Respiratory infections (e.g., bronchitis, pneumonia)
* Chronic obstructive pulmonary disease (COPD)
* Allergies or allergic reactions
* Gastroesophageal reflux disease (GERD)
* Heart conditions
* Anxiety or panic disorders
* Structural abnormalities in the lungs or airways
* What kinds of tests do I need?

To diagnose asthma and rule out other causes, your doctor will likely recommend:

* Physical exam and medical history review to assess symptoms and possible triggers.
* Lung function tests (spirometry): Measures airflow and how well your lungs work before and after using a bronchodilator (e.g., albuterol). Improvement after bronchodilator use supports asthma diagnosis.
* Peak flow measurement: A simple test to measure how hard you can breathe out, useful for monitoring asthma.
* Methacholine challenge test: If initial tests are inconclusive, this test uses a known asthma trigger to assess airway sensitivity.
* Exhaled nitric oxide (FeNO) test: Measures airway inflammation linked to asthma.
* Allergy testing: To identify potential triggers.
* Imaging tests (chest X-ray or CT scan): To rule out infections or other lung diseases.
* Blood tests: To check for markers of inflammation or allergic responses
* Is my condition likely temporary or chronic?

Asthma is generally a chronic condition that can be managed but not cured. Symptoms may vary over time, with periods of control and flare-ups. Some breathing problems may be temporary if caused by infections or allergies, but asthma tends to be persistent

* What's the best treatment?
* Inhaled corticosteroids to reduce airway inflammation (mainstay of asthma control).
* Bronchodilators (short acting like albuterol) for quick relief of symptoms.
* Long-acting bronchodilators or combination inhalers for persistent asthma.
* Avoidance of known triggers such as allergens, smoke, or irritants.
* Your doctor will develop an asthma action plan tailored to your symptoms and severity
* What are the alternatives to the primary approach that you're suggesting?
* Leukotriene modifiers (oral medications)
* Biologic therapies for severe asthma (e.g., monoclonal antibodies)
* Allergy immunotherapy if allergies are a significant trigger
* Lifestyle modifications including breathing exercises and avoiding triggers
* I have these other health conditions. How can I best manage them together?

If you have other health issues, coordinate care with your healthcare provider to avoid drug interactions and overlapping symptoms. For example, GERD can worsen asthma symptoms and may need simultaneous treatment

* Are there any restrictions that I need to follow?
* Avoid known asthma triggers (smoke, allergens, pollution)
* Follow your medication regimen strictly
* Avoid strenuous exercise if it triggers symptoms unless managed with pre-exercise medication
* Smoking cessation if applicable
* Should I see a specialist?

Seeing a pulmonologist or allergist is recommended if diagnosis is uncertain, symptoms are severe, or asthma is difficult to control

* Is there a generic alternative to the medicine you're prescribing me?

Many asthma medications, including inhaled corticosteroids and bronchodilators, have generic versions available. Ask your doctor or pharmacist about generic options to reduce cost

**Doctor patient conversation**

Doctor: Good morning! What brings you in today?

Patient: Hi, I’ve been having trouble breathing lately, especially at night and when I exercise. I also cough a lot sometimes.

Doctor: I see. How long have you been experiencing these symptoms?

Patient: It’s been a few months now, but it’s getting worse.

Doctor: Do you notice if anything triggers your symptoms? Like dust, cold air, or exercise?

Patient: Yes, cold weather and running seem to make it worse.

Doctor: Have you used any inhalers or medications for this?

Patient: I tried using an over-the-counter inhaler a couple of times, but it didn’t help much.

Doctor: Okay. Based on what you’re describing, asthma is a possibility. It’s a condition where your airways become inflamed and narrow, making it harder to breathe. We’ll do some tests to confirm.

Patient: What kind of tests?

Doctor: We’ll start with spirometry, which measures how well your lungs are working. It’s a simple breathing test. Sometimes we also do a test to see how your lungs respond to medication.

Patient: Is asthma permanent? Will I have this forever?

Doctor: Asthma is usually a chronic condition, meaning it tends to last a long time, but with proper treatment, most people can control their symptoms and live normal lives.

Patient: What treatments are available?

Doctor: The main treatments are inhalers. There are two types: one for quick relief when you have symptoms, and another you use daily to reduce inflammation and prevent attacks. We’ll find the right combination for you.

Patient: Are there side effects?

Doctor: Inhaled medications are generally safe. Some people notice mild throat irritation or hoarseness, but serious side effects are rare. We’ll monitor you closely.

Patient: Should I avoid anything?

Doctor: Yes, try to avoid your triggers like cold air and strenuous exercise without proper warm-up or medication. Also, avoid smoke and allergens if possible.

Patient: I also have allergies and acid reflux. Will that affect my asthma?

Doctor: Yes, allergies can worsen asthma, and acid reflux can sometimes trigger symptoms. Managing those conditions alongside asthma is important.

Patient: Do I need to see a specialist?

Doctor: For now, we can start treatment here. If your symptoms don’t improve or are severe, I may refer you to a pulmonologist or allergist.

Patient: Are there generic options for inhalers? I’m worried about the cost.

Doctor: Yes, many inhalers have generic versions. We’ll choose medications that fit your budget.

Doctor: I’ll give you some brochures about asthma and websites you can visit for more information. Do you have any other questions?

Patient: Not right now. Thank you!

Doctor: You’re welcome! Let’s schedule your lung function test and a follow-up appointment in a few weeks to see how you’re doing.

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**REACTIVE AIRWAY DISEASE**

## **Alternative Names for Reactive Airway Disease (RAD)**

* Wheezing Bronchitis  
  Sometimes used interchangeably with RAD, particularly in pediatric cases.
* Reactive Airways Dysfunction Syndrome (RADS)  
  A condition similar to RAD but typically refers to asthma-like symptoms following a single, high-level exposure to irritants.
* Bronchial Hyperreactivity  
  Describes increased sensitivity of the airways, a hallmark of RAD and asthma.
* Asthma-like Syndrome  
  A descriptive term used when symptoms resemble asthma, but a formal diagnosis is pending.
* Transient Wheezing  
  Refers to temporary episodes of wheezing that may be labeled as RAD in infants or toddlers.
* Presumptive Asthma  
  Sometimes used when RAD is considered a preliminary diagnosis before confirming asthma.

**DEFINITION AND DESCRIPTION**

“Reactive airway disease” (RAD) is a term that healthcare providers use to describe breathing symptoms that are similar to asthma, but they’re not sure of the exact cause. Your symptoms develop when the tubes that carry air to and from your lungs (bronchial tubes) swell, which causes narrowing of them. This makes it difficult for air to move into and out of your lungs, resulting in difficulty breathing.

Reactive airway disease isn’t the same as reactive airways dysfunction syndrome (RADS). The names, acronyms and symptoms are similar, but it’s important to keep them separate.

RAD isn’t an official clinical diagnosis, and it doesn’t have a precise definition. There’s controversy in the medical community over its use because some providers use the terms RAD and asthma interchangeably, but they don’t have the same meaning. Its use should be limited to being a placeholder term until providers can make an official diagnosis.

**Symptoms**

Reactive airway disease symptoms include:

* Chronic cough that clears mucus (sputum) from your airways.
* Shortness of breath (dyspnea).
* Difficulty breathing.
* Wheezing.
* Chest tightness.

If a provider describes your condition as RAD, your chest may feel tight and it may be difficult to breathe.

### **CAUSES**

The following may trigger symptoms that healthcare providers label as RAD:

* Allergies.
* Bacterial infections.
* Viral infections.
* Chemical gases, perfumes or fumes.
* Smoke.
* Exercise.
* Cold air, hot air, humidity or changes in the weather.

### **How long can reactive airway disease last?**

It depends on the underlying cause of your symptoms. Once a healthcare provider makes an official diagnosis, they can prescribe medicine or recommend treatment to relieve your symptoms. They’ll detail when you can expect to feel better.

### **Does reactive airway disease go away?**

It depends on the cause. Talk to a healthcare provider about your symptoms. They can give you an idea of what to expect after making an official diagnosis.

#### **Who does reactive airway disease affect?**

A healthcare provider may use the term reactive airway disease for anyone who has breathing problems without a clear cause. However, providers are most likely to describe breathing and airway symptoms as RAD in infants and children who are too young to take a lung function test.

**Diagnosis and Tests**

Talk to a healthcare provider if you or your child have symptoms that affect your breathing or if you have a cough that won’t go away. They’ll ask you about your symptoms and medical history. They’ll also conduct a physical examination. During the physical exam, they’ll listen to your lungs with a stethoscope (auscultation). A stethoscope is a medical device with a small, metal disc (diaphragm) that connects to earpieces with rubber tubing. They’ll also order tests to help them confirm their diagnosis.

To help determine the cause of your breathing problems, a healthcare provider may order the following tests:

* **Spirometry:** Spirometry is a common lung function test that measures how much air goes into and out of your lungs when you breathe.
* **Imaging tests:** Imaging tests are painless, noninvasive tests that help a provider take a closer look at your lungs, heart and bones. They may order a chest X-ray, echocardiogram (echo) or CT (computed tomography) scan.
* **Blood tests:** During a blood test, a provider will use a small needle (about the size of a standard earring post) to withdraw a small amount of blood. They’ll look at your blood under a microscope to see if you have any signs of an infection, allergies or inflammation.
* **Skin (scratch) prick test:** During a skin prick test, a provider will scratch small areas of your skin with different allergens to test for allergies.
* **Electrocardiogram (ECG or EKG):** An electrocardiogram checks how well your heart works. A provider may order this test to rule out heart disease as a cause of shortness of breath.
* **Pulse oximetry:** Pulse oximetry measures how much oxygen is in your blood.
* **Exercise testing:** Exercise testing measures whether your blood oxygen levels decrease when you exercise.

**Management and Treatment**

Reactive airway disease treatment depends on an official diagnosis. In an emergency setting, providers may use:

* **Bronchodilators:** Bronchodilators (inhalers) help relax your airways. Providers commonly prescribe bronchodilators to treat asthma and chronic obstructive pulmonary disease (COPD).
* **Oxygen therapy:** Oxygen therapy helps give your body oxygen when you’re having a hard time breathing.
* **Epinephrine injection:** Providers may give an epinephrine injection if you have a severe allergic reaction (anaphylaxis) or asthma attack.
* **Corticosteroids (steroids):** Steroids help reduce inflammation in your lungs. Providers commonly provide steroids to treat asthma, COPD or allergies.

#### **Does albuterol help reactive airway disease?**

It depends. Albuterol (Accuneb®) is a type of bronchodilator that helps open up your airways if you have asthma, COPD and exercise-induced bronchospasm. It may not treat other RAD causes.

## **Common Medications for Reactive Airway Disease**

|  |  |  |  |
| --- | --- | --- | --- |
| Medication Type | Examples | Purpose | Common Side Effects |
| Short-acting bronchodilators (SABAs) | Albuterol (Ventolin, ProAir), Levalbuterol | Provide quick relief by relaxing airway muscles during acute symptoms or attacks | Tremors, nervousness, increased heart rate, headache |
| Inhaled corticosteroids (ICS) | Fluticasone (Flovent), Budesonide (Pulmicort) | Reduce airway inflammation to prevent symptoms long-term | Hoarseness, throat irritation, oral thrush (fungal infection) |
| Oral corticosteroids | Prednisone, Methylprednisolone (Medrol) | Used short-term in severe exacerbations to reduce inflammation | Weight gain, mood changes, increased blood sugar, osteoporosis (long-term use) |
| Leukotriene modifiers | Montelukast (Singulair) | Block inflammatory chemicals to reduce symptoms | Headache, abdominal pain, rare mood changes |
| Long-acting bronchodilators (LABAs) | Salmeterol (Serevent) | Used with corticosteroids for long-term control (not alone) | Muscle cramps, palpitations, headache |
| Combination inhalers | Fluticasone/salmeterol (Advair), Budesonide/formoterol (Symbicort) | Combine anti-inflammatory and bronchodilator effects | Side effects of both ICS and LABA |
| Anticholinergics | Ipratropium (Atrovent) | Alternative bronchodilator, sometimes added for symptom relief | Dry mouth, cough, headache |

* Oxygen therapy: In severe cases with low oxygen levels.
* Epinephrine injection: Used in emergency situations such as severe airway constriction or anaphylaxis.
* Immunotherapy: For allergic triggers contributing to RAD.

## Side Effect Considerations

* Inhaled corticosteroids are generally safe but require rinsing the mouth after use to prevent fungal infections.
* Oral steroids are effective but should be used for short periods due to systemic side effects.
* Bronchodilators can cause jitteriness or increased heart rate, especially if overused.
* Montelukast has rare reports of mood or behavioral changes; monitor accordingly.

## Non-Pharmacological Management

* Avoidance of triggers (allergens, smoke, irritants)
* Breathing exercises and pulmonary rehabilitation
* Lifestyle modifications including diet and exercise

## **Outlook / Prognosis**

If a healthcare provider says you have RAD, it means you have breathing problems, but they aren’t sure of the cause. They’ll conduct a physical exam and tests to help determine the cause of your breathing issues so you can get proper treatment.

They may also refer you to a healthcare provider who specializes in conditions that affect your lungs (pulmonologist).

If a healthcare provider describes your symptoms as RAD, your treatment, recovery and management depend on an official diagnosis. Some conditions are treatable, while others may progressively get worse.

**Prevention**

The best way to lower your risk of RAD is to:

* Avoid known allergens or other triggers.
* Take medicines that prevent your airways from swelling.
* Quit smoking.
* Wear a mask or other respiratory protection while handling chemicals.

**When should I see my healthcare provider?**

See your healthcare provider if you have symptoms of reactive airway disease or if your symptoms don’t improve with treatment.

Call your local emergency number or get to an emergency room right away if you’re using a lot of energy to breathe (severe respiratory distress), aren’t responding to breathing treatments, have low oxygen levels or notice anaphylaxis symptoms, including:

* Difficulty breathing.
* Drop in blood pressure.
* Swollen lips.
* Sudden weakness.
* Fainting (syncope).
* Confusion.

## **Diagnostic Considerations**

The clinical diagnosis of asthma in children aged 5 years or younger is often based on symptoms during and between viral respiratory tract infections. The symptom pattern is as follows:

* Cough, wheeze, and heavy breathing for more than 10 days during upper respiratory tract infections
* More than three episodes per year or severe episodes and/or night worsening
* Cough, wheeze, or heavy breathing during play or when laughing, between episodes
* Allergic sensitization, atopic dermatitis, food allergy, or family history of asthma

## **Differential Diagnoses**

* Anaphylaxis
* Aspiration Pneumonitis and Pneumonia
* Emergent Management of Croup (Laryngotracheobronchitis)
* Pediatric Acute Respiratory Distress Syndrome
* Pediatric Anaphylaxis
* Pediatric Epiglottitis
* Pediatric Foreign Body Ingestion
* Pediatric Pneumonia

## **Epidemiology**

### United States statistics

Pediatric asthma is a chronic, multifactorial, lower airway disease that affects 5-15% of children (2.7 million children in the United States alone). In the United States, approximately one half of all emergency department (ED) and clinic visits for asthma are children younger than 18 years. ED visits peak in the fall, whereas school holidays disrupt the spread of infections, resulting in a subsequent decrease in ED visits and hospitalizations. Status asthmaticus appears to be on the rise; several retrospective studies reflect an increase in hospital admissions, particularly in those younger than 4 years. Fewer hospital and ED visits occur in children using inhaled corticosteroid therapy.

Asthma prevalence appears to be increasing worldwide. Air pollutants may play a role in the prevalence increase. Higher prevalence occurs in poverty stricken urban areas where children are less likely to have routine doctor visits and readily available access to medications.

A correlation may exist between high levels of exposure to cockroach allergen and the frequency of asthma-related health problems in inner-city children. Homes in poverty areas were more likely to have high cockroach allergen levels. Asthma may develop in children from early exposure to cockroach allergen.An association may exist between obesity and childhood asthma. Increased resisting, an adipokine produced by adipose tissue, may play a negative predictive role in asthma.

An algorithm has been developed to determine the risk factors for developing persistent asthma symptoms among children younger than 3 years of age who had 4 or more episodes of wheezing during the previous year.The Asthma Predictive Index included either (1) one of the following: parental history of asthma, a physician diagnosis of atopic dermatitis, or evidence of sensitization to aeroallergens; or (2) two of the following: evidence of sensitization to foods, ≥4% peripheral blood eosinophilia, or wheezing apart from colds.

### International statistics

Worldwide, the prevalence of asthma is increasing. Asthma is more common in Western countries than in developing countries. Asthma is more prevalent in English-speaking countries. Prevalence increases as a developing country becomes more Westernized and urbanized.

### Race-, sex-, and age-related demographics

Reactive airway disease is more common in Black and Hispanic children; hospitalization rates in African Americans are 4 times greater than in the White population. A correlation may exist between high levels of exposure to cockroach allergen and the frequency of asthma-related health problems in inner-city children. No correlation exists between education levels from a retrospective review.

The male-to-female ratio is 1.5:1. The peak prevalence of asthma is in those aged 6-11 years.

### Impact of COVID-19 on pediatric asthma exacerbation

Pediatric asthma attacks decreased during the COVID-19 pandemic, probably because of reduced environmental allergen exposure and decreased risk of other viral respiratory tract infections. Asthma is not considered to be a risk factor for COVID-19 severity.

## **Procedures**

Procedures include the following:

* Spirometry (decreased forced expiratory volume in one second [FEV1])
  + Bedside spirometry is the primary procedure for children with RAD who are older than 5 years.
  + Patients with decreased FEV1 require further evaluation and treatment.
* A barium swallow may be indicated to determine any esophageal, pulmonary, or vascular pathology, particularly a tracheoesophageal fistula.
* Bronchoscopy (rarely indicated) (see Table 1 below)
* Peak expiratory flow (PEF) is the most common form of pulmonary function test monitoring. Record the best of 3 attempts. Possible life-threatening asthma exacerbation with PEF predicted of less than 30%; severe exacerbation, with less than 50%; and moderate exacerbation, with less than 80%.

Peak flow rates are described in the table below.

Table 1. Peak Flow Rates in Liters per Minute

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| Height in  Inches | Average  Rate | Range\* | Height in  Inches | Average  Rate | Range\* |
| 40 | 150 | 110-190 | 56 | 330 | 240-420 |
| 41 | 160 | 115-205 | 57 | 340 | 240-420 |
| 42 | 170 | 120-220 | 58 | 360 | 260-460 |
| 43 | 180 | 130-220 | 59 | 375 | 270-480 |
| 44 | 190 | 135-245 | 60 | 390 | 280-500 |
| 45 | 200 | 145-255 | 61 | 400 | 290-510 |
| 46 | 210 | 150-270 | 62 | 415 | 300-530 |
| 47 | 220 | 160-280 | 63 | 430 | 310-550 |
| 48 | 230 | 165-295 | 64 | 445 | 320-570 |
| 49 | 240 | 175-305 | 65 | 460 | 330-590 |
| 50 | 250 | 180-320 | 66 | 480 | 345-615 |
| 51 | 260 | 190-330 | 67 | 500 | 360-640 |
| 52 | 270 | 195-345 | 68 | 515 | 370-660 |
| 53 | 280 | 200-360 | 69 | 530 | 380-680 |
| 54 | 300 | 215-385 | 70 | 550 | 395-705 |
| 55 | 315 | 225-405 | 71 | 570 | 410-730 |
| \*Includes 95% of white males aged 7-20 years. | | | | | |

**RECENT GUIDELINES AND RECOMMENDATION**

Recommendations include the following:

* The ERS recommends spirometry as first-line diagnosis in children aged 5-16 years with suspected asthma.
* ERS recommends bronchodilator reversibility testing as first-line diagnosis in all children with FEV1 < LLN or < 80% predicted and/or FEV1/FVC < LLN or < 80% predicted.
* ERS recommends FeNO as first-line diagnosis in children aged 5-16 years with suspected asthma.
* ERS recommends against PEFR variability testing as the primary objective test on its own to diagnose asthma in children aged 5-16 years.
* ERS recommends against diagnosing asthma in children aged 5-16 years based on clinical history alone or following a single abnormal objective test.
* ERS recommends against using an improvement in symptoms after a trial of preventive medication alone to diagnose asthma in children aged 5-16 years.
* ERS recommends against using skin prick tests as diagnostic tests for asthma in children aged 5-16 years.
* ERS recommends a direct bronchial challenge test using methacholine in children aged 5-16 years with suspected asthma where asthma diagnosis could not be confirmed with first-line tests.
* ERS recommends an indirect bronchial challenge test using a treadmill or a bicycle in children aged 5-16 years with suspected asthma with exercise-related symptoms where asthma diagnosis could not be confirmed with first-line tests.

## **PREDEFINED Questions and answers**

### **What is the difference between reactive airway disease and asthma?**

People sometimes use the terms “reactive airway disease/RAD” and “asthma” interchangeably, but they don’t have the same meaning. Asthma is a chronic condition that irritates and narrows your airways. It may also cause extra mucus production. Reactive airway disease is a placeholder term providers use to indicate that something is affecting your airways, but they aren’t sure of the exact cause.

### **Is reactive airway disease the same as COPD?**

Healthcare providers may sometimes describe COPD symptoms as reactive airway disease. But they’re not the same. COPD is an umbrella term for chronic (long-lasting) lung conditions that affect your ability to breathe and progressively get worse. Providers may describe a condition — including COPD — as RAD until they can make an official diagnosis.

## 1. What’s causing my reactive airway disease?

RAD is caused by overreaction of your bronchial tubes to various irritants or triggers, leading to swelling and narrowing of the airways. Common causes include:

* Allergens such as pet hair, pollen, mold, dust
* Respiratory infections, especially viral infections like RSV
* Environmental irritants like smoke, strong odors, perfumes, chemical fumes
* Exercise, stress, and changes in weather or temperature  
  Children with a family history of asthma or prior lung infections are at higher risk.

## 2. Is there a cure for my condition?

Reactive Airway Disease is generally considered a transient or temporary condition, especially in young children, and not a formal diagnosis like asthma. It may improve or resolve over time, particularly if triggers are avoided and infections are managed. However, RAD can sometimes progress to chronic asthma, which currently has no cure but can be effectively controlled with treatment.

## 3. How can I improve my breathing?

* Avoid known triggers such as smoke, allergens, strong odors, and cold air.
* Use prescribed medications regularly to reduce airway inflammation and prevent attacks.
* Manage respiratory infections promptly.
* Practice breathing exercises and maintain good overall lung health.
* Follow your doctor’s asthma or RAD action plan closely.

## 4. What medications do you recommend?

Typical medications used include:

* Short-acting bronchodilators (e.g., albuterol) for quick relief of symptoms.
* Inhaled corticosteroids to reduce airway inflammation and prevent flare-ups.
* In some cases, leukotriene modifiers or combination inhalers may be prescribed.
* Oral corticosteroids may be used short-term for severe episodes.

## 5. What’s the correct way to use these medications?

* Inhalers: Shake well before use, exhale fully, place the mouthpiece in your mouth, inhale deeply while pressing the inhaler, then hold your breath for 10 seconds before exhaling. Use a spacer if recommended. Rinse your mouth after corticosteroid inhalers to prevent irritation.
* Follow dosage instructions carefully and do not overuse quick-relief inhalers.
* Take long-term control medications daily, even when you feel well, to prevent symptoms.

## 6. Should I see a pulmonologist?

* If your symptoms are severe, persistent, or difficult to control, or if your primary care provider cannot confirm a diagnosis, seeing a pulmonologist (lung specialist) is recommended.
* A specialist can perform detailed lung function tests and tailor treatment to your needs

**GENOMIC DATA**

* Asthma-related genetic loci: Many genes identified in asthma research are relevant to RAD because of overlapping airway hyperreactivity. Genome-wide association studies (GWAS) have pinpointed several key genes linked to airway disease susceptibility, including:
  + ORMDL3 and GSDMB on chromosome 17q21, strongly associated with childhood-onset asthma and airway hyperreactivity.
  + IL1RL1/IL18R1, IL33, HLA-DQ, SMAD3, IL2RB and others, which regulate immune responses and airway inflammation.
* Genetic variants in CFTR, alpha-1 antitrypsin, and MBL genes: Studies have explored these genes for their roles in obstructive lung diseases. For example, the F508del mutation in CFTR is marginally associated with increased asthma risk but is not a major marker for COPD or RAD.
* Gene-environment interactions: Genetic predisposition combined with environmental triggers (viruses, allergens, pollutants) influences RAD development and severity. For instance, defects in barrier proteins (e.g., FLG, SPINK5) can predispose to airway sensitivity and atopic conditions.
* Mouse model studies: Research using mouse models has identified genes like mClca3 and mClca5 that regulate mucous cell metaplasia and airway hyperreactivity, illustrating genetic segregation of airway disease traits and pointing to potential therapeutic targets

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand you or your child have been experiencing breathing difficulties. Can you tell me more about the symptoms?

Patient: Yes, there’s been a lot of coughing and wheezing, especially at night, and sometimes after playing or running around.

Doctor: That sounds like reactive airway disease, or RAD, which is a term we use when someone has asthma-like symptoms but we haven’t confirmed asthma with lung tests yet. It’s common in young children who can’t do lung function tests.

Patient: What causes this? Is it serious?

Doctor: RAD is usually caused by your airways reacting strongly to triggers like viral infections, allergens, cold air, or smoke. It can make your chest feel tight and breathing difficult. The good news is that with proper treatment and avoiding triggers, most people improve.

Patient: How do you diagnose it?

Doctor: I’ll listen to your lungs with a stethoscope and ask about your symptoms and history. Sometimes we do lung function tests if possible. For young children, diagnosis is mostly clinical. We may also check for allergies or infections.

Patient: What treatment do you recommend?

Doctor: We usually start with inhalers that help open your airways quickly when you have symptoms, like albuterol. If symptoms happen often, we add inhaled corticosteroids to reduce inflammation. I’ll also advise you on avoiding triggers and what to do if symptoms worsen.

Patient: How long will it take to get better?

Doctor: It depends on the cause and how well you avoid triggers and use medications. Many children outgrow RAD, but some may develop asthma later. We’ll monitor your progress and adjust treatment as needed.

Patient: Should I see a specialist?

Doctor: If symptoms are severe, frequent, or not improving with treatment, I may refer you to a pulmonologist or allergist for further evaluation.

Patient: Thank you. Is there anything else I should know?

Doctor: Yes, it’s important to follow your asthma action plan, use medications correctly, and come back for follow-ups. I’ll give you some brochures and reliable websites for more information.

REFERENCES

[Reactive Airway Disease: Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24661-reactive-airway-disease#overview)

<https://emedicine.medscape.com/article/800119-guidelines>

**BRONCHITIS**

## **Alternative Names for Bronchitis**

* Acute Bronchitis  
  Refers to short-term inflammation of the bronchial tubes, usually caused by infections.
* Chronic Bronchitis  
  A long-term condition characterized by persistent cough and mucus production, often part of Chronic Obstructive Pulmonary Disease (COPD).
* Tracheobronchitis  
  Inflammation involving both the trachea (windpipe) and bronchi.
* Bronchial Inflammation  
  A descriptive term emphasizing the inflamed state of the airways.
* Smoker’s Cough  
  Informal term often used to describe chronic bronchitis symptoms in smokers.
* Bronchopneumonia  
  Sometimes used when bronchitis is accompanied by pneumonia, indicating infection of both bronchi and lung tissue.
* Lower Respiratory Tract Infection (LRTI)  
  A broader term that includes bronchitis along with other infections like pneumonia.

**DEFINITION AND DESCRIPTION**

Bronchitis is an inflammation of the lining of your bronchial tubes. These tubes carry air to and from your lungs. People who have bronchitis often cough up thickened mucus, which can be discolored. Bronchitis may start suddenly and be short term (acute) or start gradually and become long term (chronic).

Acute bronchitis, which often develops from a cold or other respiratory infection, is very common. Also called a chest cold, acute bronchitis usually improves within a week to 10 days without lasting effects, although the cough may linger for weeks.

Chronic bronchitis, a more serious condition, is a constant irritation or inflammation of the lining of the bronchial tubes, often due to smoking. If you have repeated bouts of bronchitis, you may have chronic bronchitis, which requires medical attention. Chronic bronchitis is one of the conditions included in chronic obstructive pulmonary disease (COPD).

**Causes**

Acute bronchitis is usually caused by viruses, typically the same viruses that cause colds and flu (influenza). Many different viruses — all of which are very contagious — can cause acute bronchitis. Antibiotics don't kill viruses, so this type of medication isn't useful in most cases of bronchitis.

Viruses spread mainly from person to person by droplets produced when an ill person coughs, sneezes or talks and you inhale the droplets. Viruses may also spread through contact with an infected object. This happens when you touch something with the virus on it and then touch your mouth, eyes or nose.

The most common cause of chronic bronchitis is cigarette smoking. Air pollution and dust or toxic gases in the environment or workplace also can contribute to the condition.

**Risk factors**

Factors that increase your risk of bronchitis include:

* **Cigarette smoke.** People who smoke or who live with a smoker are at higher risk of both acute bronchitis and chronic bronchitis.
* **Low resistance.** This may result from another acute illness, such as a cold, or from a chronic condition that compromises your immune system. Older adults, infants and young children have greater vulnerability to infection.
* **Exposure to irritants on the job.** Your risk of developing bronchitis is greater if you work around certain lung irritants, such as grains or textiles, or are exposed to chemical fumes.
* **Gastric reflux.** Repeated bouts of severe heartburn can irritate your throat and make you more prone to developing bronchitis.

**Symptoms**

If you have acute bronchitis, you may have cold symptoms, such as:

* Cough
* Production of mucus (sputum), which can be clear, white, yellowish-gray or green in color — rarely, it may be streaked with blood
* Sore throat
* Mild headache and body aches
* Slight fever and chills
* Fatigue
* Chest discomfort
* Shortness of breath and wheezing

While these symptoms usually improve in about a week, you may have a nagging cough that lingers for several weeks.

For chronic bronchitis, signs and symptoms may include:

* Cough
* Production of mucus
* Fatigue
* Chest discomfort
* Shortness of breath

Chronic bronchitis is typically defined as a productive cough that lasts at least three months, with bouts that recur for at least two consecutive years. If you have chronic bronchitis, you're likely to have periods when your cough or other symptoms worsen. It's also possible to have an acute infection on top of chronic bronchitis.

### **When to see a doctor**

Contact your doctor or clinic for advice if your cough:

* Is accompanied by a fever higher than 100.4 F (38 C).
* Produces blood.
* Is associated with serious or worsening shortness of breath or wheezing.
* Includes other serious signs and symptoms, for example, you appear pale and lethargic, have a bluish tinge to your lips and nail beds, or have trouble thinking clearly or concentrating.
* Lasts more than three weeks.

Before you go in, your doctor or clinic can give you guidance on how to prepare for your appointment.

## **DIAGNOSIS**

During the first few days of illness, it can be difficult to distinguish the signs and symptoms of acute bronchitis from those of a common cold. During the physical exam, your doctor will use a stethoscope to listen closely to your lungs as you breathe.

In some cases, your doctor may suggest the following tests:

* **Chest X-ray.** A chest X-ray can help determine if you have pneumonia or another condition that may explain your cough. This is especially important if you smoke or have ever smoked.
* **Sputum tests.** Sputum is the mucus that you cough up from your lungs. It can be tested to see if you have illnesses that could be helped by antibiotics. Sputum can also be tested for signs of allergies.
* **Pulmonary function test.** During a pulmonary function test, you blow into a device called a spirometer, which measures how much air your lungs can hold and how quickly you can get air out of your lungs. This test checks for signs of asthma, chronic bronchitis or emphysema.

**Treatment**

Most cases of acute bronchitis get better without treatment, usually within a couple of weeks.

### **Medications**

In some circumstances, your doctor may recommend other medications, including:

* **Cough medicine.** If your cough keeps you from sleeping, you might try cough suppressants at bedtime.
* **Other medications.** If you have allergies, asthma or chronic obstructive pulmonary disease (COPD), your doctor may recommend an inhaler and other medications to reduce inflammation and open narrowed passages in your lungs.
* **Antibiotics.** Because most cases of acute bronchitis are caused by viral infections, antibiotics aren't effective. However, if your doctor suspects that you have a bacterial infection, he or she may prescribe an antibiotic.

### **Therapies**

If you have chronic bronchitis, you may benefit from:

* **Pulmonary rehabilitation.** This is a breathing exercise program in which a respiratory therapist teaches you how to breathe more easily and increase your ability to be physically active.
* **Oxygen therapy.** This delivers extra oxygen to help you breathe.

## **Common Medications Used to Treat Bronchitis**

|  |  |  |  |
| --- | --- | --- | --- |
| Medication Type | Examples | Purpose | Common Side Effects |
| Over-the-Counter Pain Relievers | Acetaminophen (Tylenol), Ibuprofen (Advil), Naproxen | Reduce fever, body aches, and inflammation | Stomach upset, liver toxicity (acetaminophen overdose), kidney issues (NSAIDs) |
| Cough Suppressants | Dextromethorphan (Robitussin DM) | Suppress dry cough when mucus is minimal | Drowsiness, dizziness |
| Expectorants / Mucolytics | Guaifenesin (Mucinex) | Loosen mucus to help clear airways | Nausea, vomiting, dizziness |
| Bronchodilators | Albuterol (inhaler/nebulizer), Ipratropium | Relax airway muscles to ease breathing | Tremors, nervousness, increased heart rate, dry mouth |
| Inhaled Corticosteroids | Beclomethasone, Fluticasone | Reduce airway inflammation (for chronic or severe cases) | Hoarseness, throat irritation, oral thrush |
| Antibiotics | Azithromycin, Amoxicillin, Doxycycline, Levofloxacin (only if bacterial infection suspected) | Treat bacterial bronchitis or secondary infections | Diarrhea, allergic reactions, antibiotic resistance |
| Antiviral Medications | Oseltamivir (Tamiflu) (if influenza-related) | Reduce severity and duration of flu-related bronchitis | Nausea, vomiting, headache |

* Acute bronchitis is mostly viral, so antibiotics are generally not recommended unless a bacterial infection is confirmed or strongly suspected.
* Symptomatic treatment with rest, fluids, and OTC medications is often sufficient.
* Bronchodilators may be prescribed if wheezing or airway obstruction occurs.
* Corticosteroids are usually reserved for chronic bronchitis or severe cases.

## Side Effects

* Pain relievers: Can cause stomach upset or liver/kidney issues if overused.
* Cough suppressants: May cause drowsiness or dizziness.
* Expectorants: Generally well-tolerated but may cause mild nausea.
* Bronchodilators: Can cause tremors, increased heart rate, or nervousness.
* Inhaled corticosteroids: Risk of oral thrush; rinsing mouth after use reduces this.
* Antibiotics: Risk of diarrhea, allergic reactions, and antibiotic resistance if overused.
* Antivirals: Possible nausea and headache.

## **Lifestyle and home remedies**

If you have bronchitis, to help you feel better, you may want to try the following self-care measures:

* **Get enough rest.** Rest and sleep help your body heal.
* **Drink plenty of fluids.** Staying hydrated can help to thin mucus.
* **Avoid lung irritants.** Don't smoke. Wear a mask when the air is polluted or if you're exposed to irritants, such as paint or household cleaners with strong fumes.
* **Use a humidifier.** Warm, moist air helps relieve coughs and loosens mucus in your airways. Be sure to clean the humidifier according to the manufacturer's recommendations to avoid the growth of bacteria and fungi in the water container.
* **Consider a face covering in cold air.** If cold air makes your cough worse and causes shortness of breath, put on a face mask or cover your mouth and nose with a scarf before you go outside.

## **Complications**

Although a single episode of bronchitis usually isn't cause for concern, it can lead to pneumonia in some people. Repeated bouts of bronchitis, however, may mean that you have chronic obstructive pulmonary disease (COPD).

**Prevention**

To reduce your risk of bronchitis, follow these tips:

* **Get an annual flu shot.** Many cases of acute bronchitis result from influenza, a virus. Getting a yearly flu vaccine can help protect you from getting the flu. Also ask your doctor or clinic if you need a vaccination that protects against certain types of pneumonia.
* **Wash your hands.** To reduce your risk of catching a viral infection, wash your hands frequently and get in the habit of using alcohol-based hand sanitizers. Also, avoid touching your eyes, nose and mouth.
* **Avoid close contact with people who have a viral infection.** Stay away from people who have the flu or another respiratory illness.
* **Avoid cigarette smoke.** Cigarette smoke increases your risk of chronic bronchitis.
* **Wear appropriate face covering.** If you have COPD, consider wearing a face mask at work if you're exposed to dust or fumes. Talk to your employer about the appropriate protection. Wearing a face mask when you're going to be among crowds helps reduce exposure to infections.

## **Outlook / Prognosis**

Acute bronchitis usually isn’t serious. While frustrating, you have to wait out the symptoms for a few weeks. If you’re living with a heart condition or another breathing condition, like asthma, it could make your symptoms worse or last longer.

Chronic bronchitis can be a serious condition and might mean you have lung damage. While the damage can’t be reversed, your provider can help you manage your symptoms and have fewer flare-ups.

If you have an ongoing condition like asthma, diabetes, chronic obstructive pulmonary disease or heart failure, bronchitis might make it worse (exacerbation). Tell your healthcare provider if you have any ongoing conditions.

### **Can bronchitis go away on its own?**

Yes, acute bronchitis usually goes away on its own. It’s almost always caused by a virus, and you can’t get rid of most viruses with medicine. You can treat the symptoms at home while you wait for the inflammation to go down.

Bronchitis caused by something else may need treatment to help it go away. Chronic bronchitis usually doesn’t go away completely but can get better with treatment.

Most people get over bronchitis in about two weeks, but it might take as long as three to six weeks. You can manage your symptoms at home with over-the-counter medicines while you get better. If you don’t feel better after three weeks, see your healthcare provider.

## **Diagnostic Considerations**

Streptococcal pharyngitis is most caused by group A streptococci (45%) and anaerobes (18%), which often occur as a co-infection.

Much of the concern about diagnosing streptococcal pharyngitis is related to the complications of infection, particularly acute rheumatic fever and poststreptococcal glomerulonephritis as a late complication. Therefore, maintaining a high level of suspicion for streptococci group A in the presence of pharyngitis is advisable.

Other medical issues/problems to consider include the following:

* Exercise-induced asthma
* Bacterial tracheitis
* Cough
* Cystic fibrosis
* Influenza
* Hyperreactive airway disease
* Retained foreign body
* Tonsillitis
* Occupational exposures

## **Differential Diagnoses**

* Acute Sinusitis
* Alpha1-Antitrypsin (AAT) Deficiency
* Asthma
* Bacterial Pharyngitis
* Bronchiectasis
* Bronchiolitis
* Bronchitis
* Chronic Obstructive Pulmonary Disease (COPD)
* Chronic Sinusitis
* Gastroesophageal Reflux Disease
* Group A Streptococcal (GAS) Infections
* Influenza
* Viral Pharyngitis

## **Epidemiology**

According to estimates from national interviews taken by the National Center for Health Statistics in 2006, approximately 9.5 million people, or 4% of the population, were diagnosed with chronic bronchitis. These statistics may underestimate the prevalence of chronic obstructive pulmonary disease by as much as 50%, because many patients underreport their symptoms, and their conditions remain undiagnosed.

An overdiagnosis of chronic bronchitis by patients and clinicians has also been suggested, however. The term bronchitis is often used as a common descriptor for a nonspecific and self-limited cough, thereby falsely increasing its incidence even though the patient does not meet the criteria for diagnosis.

In one study, acute bronchitis affected 44 of 1000 adults annually, and 82% of episodes occurred in fall or winter.By way of comparison, 91 million cases of influenza, 66 million cases of the common cold, and 31 million cases of other acute upper respiratory tract infections occurred that year.

Acute bronchitis is common throughout the world and is one of the top 5 reasons for seeking medical care in countries that collect such data. No difference in racial distribution is reported, though bronchitis occurs more frequently in populations with a low socioeconomic status and in people who live in urban and highly industrialized areas.

In terms of gender-specific incidence, bronchitis affects males more than females. In the United States, up to two thirds of men and one fourth of women have emphysema at death. Although found in all age groups, acute bronchitis is most frequently diagnosed in children younger than 5 years, whereas chronic bronchitis is more prevalent in people older than 50 years.

**GENOMIC DATA**

* Acute Viral Bronchiolitis:  
  A genome-wide association study (GWAS) of infants hospitalized for bronchiolitis found significant associations with variants in the GSDMB and CDHR3 genes. These genes modulate susceptibility to bronchiolitis, especially when caused by viruses other than respiratory syncytial virus (RSV). Increased expression of GSDMB and ORMDL3 in immune cells correlates with higher bronchiolitis risk. Children with bronchiolitis in infancy linked to these genetic variants are more likely to develop asthma later in life, suggesting shared genetic pathways between bronchiolitis and asthma.
* Chronic Bronchitis and CFTR Mutations:  
  Studies have shown that mutations in the CFTR gene (notably F508del, W1282X, N1303K) contribute to chronic bronchitis and bronchial asthma in children. These mutations affect ion transport and mucus clearance in the airways, increasing susceptibility to chronic respiratory conditions. A hereditary predisposition was found in about one-third of affected children’s parents, highlighting a genetic basis for chronic bronchitis.
* Other Genetic Markers:  
  Research suggests associations between chronic bronchitis and certain blood group systems (MNS, Rhesus), and complement component alleles (e.g., absence of C3F allele linked to higher chronic bronchitis prevalence), indicating a multifactorial genetic influence

**PREDEFINED Questions and answer**

## 1. What’s the best way to treat my symptoms at home?

* Drink plenty of fluids (8-12 glasses a day) to thin mucus and make coughing easier.
* Get plenty of rest to help your body heal.
* Use a humidifier or inhale steam to loosen mucus and soothe airways.
* Gargle saltwater several times a day to relieve throat irritation.
* Avoid lung irritants like smoke, strong fumes, and cold air (wear a scarf or mask if needed).
* Use over-the-counter pain relievers such as ibuprofen or acetaminophen for fever and aches.
* Try honey (for adults and children over 1 year) to soothe cough.
* Avoid cough suppressants unless the cough is dry and disturbing sleep; productive coughs help clear mucus.
* Keep your environment clean and dust-free.

## 2. How long should I expect bronchitis to last?

* Acute bronchitis typically lasts about 7 to 10 days, but the cough can persist for up to 3 to 4 weeks or longer.
* Most people recover fully within 2 to 3 weeks, but some symptoms, especially cough, may linger.
* Chronic bronchitis lasts at least 3 months and requires medical management.

## 3. What new or worsening symptoms should I look out for?

* High or persistent fever (above 101.3°F or 38.5°C)
* Shortness of breath or difficulty breathing that worsens
* Chest pain or tightness
* Cough producing green, yellow, or bloody mucus
* Symptoms lasting longer than 3 weeks without improvement
* Signs of pneumonia such as chills, rapid breathing, or fatigue

## 4. When should I see you again if symptoms haven’t improved?

* If symptoms do not improve within 2 to 3 weeks or worsen at any time
* If you develop any of the worsening symptoms listed above
* If you have underlying lung diseases (e.g., asthma, COPD) and symptoms worsen
* If you experience difficulty breathing or chest pain

Bronchitis vs. Pneumonia: How are they Different?

### **What’s the difference between bronchitis and pneumonia?**

Bronchitis is an inflammation of the airways leading to the lungs. [Pneumonia](https://my.clevelandclinic.org/health/diseases/4471-pneumonia) is an inflammation of the lungs themselves.

Bronchitis causes inflammation and mucus in your trachea and bronchi that make you cough a lot. Pneumonia causes inflammation and fluid in the small sacs in your lungs (alveoli) that makes it hard to breathe. You also usually have a cough and a fever. Pneumonia is more serious than bronchitis.

While you could have an infection that causes both, bronchitis doesn’t usually turn into pneumonia.

### **What’s the difference between bronchitis and bronchiolitis?**

Bronchitis is inflammation in the larger airways (trachea and bronchi) coming into the lungs. Bronchiolitis is an inflammation of the next smaller airways (bronchioles) that come off of the bronchi. Children usually get bronchiolitis while adults get bronchitis.

### **Is menthol vapor rub good for bronchitis?**

You might use vapor rubs, like Vicks VapoRub® or Mentholatum® ointment, for anything that ails you and wonder if they work for bronchitis. Vapor rubs have ingredients in them intended to calm down coughs, so they may help your bronchitis symptoms. Don’t use vapor rubs on children under two without asking your pediatrician first.

**DOCTOR PATIENT CONVERSATION**

Doctor: Can you tell me what’s bothering you today?

Patient: I’ve had a cough for the past few days. It started after I had a cold.

Doctor: How long has the cough lasted, and is it dry or productive?

Patient: It’s been about a week now, and I’m coughing up some mucus.

Doctor: Do you have any other symptoms like fever, chest pain, or shortness of breath?

Patient: I had a mild fever a few days ago, and sometimes my chest feels tight, especially when I cough a lot.

Doctor: Based on your symptoms, it sounds like acute bronchitis, which is inflammation of the airways usually caused by a viral infection. It’s common after a cold or flu.

Patient: Is it serious? How long will it last?

Doctor: Most people recover in 2 to 3 weeks, but the cough can linger a bit longer. It’s usually not serious in healthy adults.

Patient: What can I do to feel better?

Doctor: Rest, drink plenty of fluids, and use over-the-counter pain relievers like ibuprofen or acetaminophen for fever and aches. Using a humidifier or inhaling steam can help loosen mucus. Avoid smoking and irritants.

Patient: Do I need antibiotics?

Doctor: Since bronchitis is usually viral, antibiotics aren’t needed unless there’s a bacterial infection. If your symptoms worsen or don’t improve after a few weeks, let me know.

Patient: When should I come back or see a specialist?

Doctor: If you develop high fever, difficulty breathing, chest pain, or if your cough lasts more than 3 weeks, you should come back. If you have underlying lung conditions like asthma or COPD, or if symptoms worsen, a referral to a pulmonologist might be necessary.

Patient: Thank you. Is there anything else I should watch for?

Doctor: Yes, watch for worsening shortness of breath, coughing up blood, or severe fatigue. If any of these happen, seek medical attention promptly.

REFERENCES

[Bronchitis: Causes, Symptoms, Diagnosis & Treatment](https://my.clevelandclinic.org/health/diseases/3993-bronchitis#outlook-prognosis)

[Bronchitis - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/bronchitis/diagnosis-treatment/drc-20355572)

<https://emedicine.medscape.com/article/297108-workup#c8>

**BRONCHIOLITIS**

## **Alternative Names for Bronchiolitis**

* Acute Bronchiolitis  
  Refers to the sudden onset inflammation of the small airways (bronchioles), usually in infants and young children.
* Viral Bronchiolitis  
  Emphasizes that the condition is typically caused by viral infections, most commonly respiratory syncytial virus (RSV).
* Infantile Bronchiolitis  
  Highlights the age group most affected infants under 2 years old.
* Obstructive Bronchiolitis  
  Describes the airway obstruction caused by inflammation and mucus.
* Bronchiolitis Obliterans (Constrictive Bronchiolitis)  
  A rare, chronic form of bronchiolitis characterized by permanent scarring and narrowing of the bronchioles, often due to injury or infection.
* Small Airway Disease  
  A broader term sometimes used to describe conditions affecting the bronchioles, including bronchiolitis.

**DEFINITION AND DESCRIPTION**

Bronchiolitis is a common lung infection in young children and infants. It causes swelling and irritation and a buildup of mucus in the small airways of the lung. These small airways are called bronchioles. Bronchiolitis is almost always caused by a virus.

Bronchiolitis starts out with symptoms much like a common cold. But then it gets worse, causing coughing and a high-pitched whistling sound when breathing out called wheezing. Sometimes children have trouble breathing. Symptoms of bronchiolitis can last for 1 to 2 weeks but occasionally can last longer.

Most children get better with care at home. A small number of children need a stay in the hospital.

Bronchiolitis is the most common lower respiratory tract infection among children younger than 2 years old.

The viruses that cause bronchiolitis are contagious. These viruses spread through respiratory droplets (saliva or mucus) from someone’s mouth or nose. When someone sneezes or coughs, these droplets become airborne and spread from person to person or stick to frequently touched surfaces or objects.

### **Bronchiolitis and bronchitis**

Bronchiolitis and bronchitis are two conditions that both sound similar and have similar symptoms. A virus causes both conditions, which target the airways in your lungs. Bronchitis affects the bronchi, or the larger airways. Bronchiolitis affects your smaller airways (bronchioles). Bronchitis usually affects older children and adults, while bronchiolitis is more common in younger children.

**Causes**

Bronchiolitis happens when a virus infects the bronchioles, which are the smallest airways in the lungs. The infection makes the bronchioles swollen and irritated. Mucus collects in these airways, which makes it difficult for air to flow freely in and out of the lungs.

Bronchiolitis is usually caused by the respiratory syncytial virus (RSV). RSV is a common virus that infects just about every child by 2 years of age. Outbreaks of RSV infection often happen during the colder months of the year in some locations or the rainy season in others. A person can get it more than once. Bronchiolitis also can be caused by other viruses, including those that cause the flu or the common cold.

The viruses that cause bronchiolitis are easily spread. You can get them through droplets in the air when someone who is sick coughs, sneezes or talks. You also can get them by touching shared items — such as dishes, doorknobs, towels or toys — and then touching your eyes, nose or mouth.

**Risk factors**

Bronchiolitis usually affects children under the age of 2 years. Infants younger than 3 months have the highest risk of getting bronchiolitis because their lungs and their ability to fight infections aren't yet fully developed. Rarely, adults can get bronchiolitis.

Other factors that increase the risk of bronchiolitis in infants and young children include:

* Being born too early.
* Having a heart or lung condition.
* Having a weakened immune system. This makes it hard to fight infections.
* Being around tobacco smoke.
* Contact with lots of other children, such as in a childcare setting.
* Spending time in crowded places.
* Having siblings who go to school or get childcare services and bring home the infection.

### **signs and symptoms of bronchiolitis**

Early signs and symptoms of bronchiolitis resemble those of the common cold, including:

* A runny nose.
* A slight fever (under 101 degrees Fahrenheit or 38 degrees Celsius).
* A cough.
* Fatigue.
* Fussiness or irritability (infants).

A bronchiolitis infection targets your child’s airways and can cause the following symptoms that affect their breathing:

* Rapid or shallow breathing.
* Wheezing.
* Grunting noises when they breathe.
* Flaring of the nostrils.

**If your child has trouble breathing or you notice the following symptoms of severe bronchiolitis, call their healthcare provider or visit the emergency room immediately**:

* Difficulty sucking and/or swallowing (unable to feed).
* Flaring (widening) nostrils when breathing.
* Chest retracts during breathing (their skin pulls down tightly against their rib cage and makes their chest look like it’s pulling inward).
* Blue, gray or pale skin tone on their lips, fingers or toes (cyanosis).
* Dry mouth, not urinating (peeing) or crying without producing tears (dehydration).

### **When to see a doctor**

If symptoms become serious, call your child's health care provider. This is especially important if your child is younger than 12 weeks old or has other risk factors for bronchiolitis — for example, being born too early, also called premature, or having a heart condition.

Get medical attention right away if your child has any of these symptoms:

* Has blue or gray skin, lips and fingernails due to low oxygen levels.
* Struggles to breathe and can't speak or cry.
* Refuses to drink enough or breathes too fast to eat or drink.
* Breathes very fast — in infants this can be more than 60 breaths a minute — with short, shallow breaths.
* Can't breathe easily and the ribs seem to suck inward when breathing in.
* Makes wheezing sounds when breathing.
* Makes grunting noises with each breath.
* Appears slow moving, weak or very tired.

## **Diagnosis**

Your child's health care provider can usually diagnose bronchiolitis by the symptoms and listening to your child's lungs with a stethoscope.

Tests and X-rays are not usually needed to diagnose bronchiolitis. But your child's provider may recommend tests if your child is at risk of severe bronchiolitis, if symptoms are getting worse or if the provider thinks there may be another problem.

Tests may include:

* **Chest X-ray.** A chest X-ray can show if there are signs of pneumonia.
* **Viral testing.** A sample of mucus from your child's nose can be used to test for the virus causing bronchiolitis. This is done using a swab that's gently inserted into the nose.
* **Blood tests.** Occasionally, blood tests might be used to check your child's white blood cell count. An increase in white blood cells is usually a sign that the body is fighting an infection. A blood test also can show if the level of oxygen in your child's bloodstream is low.

Your child's provider may look for symptoms of dehydration, especially if your child has been refusing to drink or eat or has been vomiting. Signs of dehydration include dry mouth and skin, extreme tiredness, and making little or no urine.

**Treatment**

Bronchiolitis usually lasts for 1 to 2 weeks but symptoms occasionally last longer. Most children with bronchiolitis can be cared for at home with comfort measures. It's important to be alert for problems with breathing that are getting worse. For example, struggling for each breath, not being able to speak or cry because of struggling to breathe, or making grunting noises with each breath.

Because viruses cause bronchiolitis, antibiotics — which are used to treat infections caused by bacteria — don't work against viruses. Bacterial infections such as pneumonia or an ear infection can happen along with bronchiolitis. In this case, your child's health care provider may give an antibiotic for the bacterial infection.

Medicines called bronchodilators that open the airways don't seem to help bronchiolitis, so they usually aren't given. In severe cases, your child's health care provider may try a nebulized albuterol treatment to see if it helps. During this treatment, a machine creates a fine mist of medicine that your child breathes into the lungs.

Oral corticosteroid medicines and pounding on the chest to loosen mucus, a treatment called chest physiotherapy, have not been shown to be effective for bronchiolitis and are not recommended.

### **Hospital care**

A small number of children may need a stay in the hospital. Your child may receive oxygen through a face mask to get enough oxygen into the blood. Your child also may get fluids through a vein to prevent dehydration. In severe cases, a tube may be guided into the windpipe to help breathing.

**bronchiolitis treatment drugs and their side effects**

1. Supportive Care (Mainstay of Treatment)

* Oxygen therapy: To maintain adequate oxygen saturation (above 92%).
* Hydration and nutrition support: Oral or intravenous fluids as needed.
* Nasal suctioning: To clear mucus and ease breathing.

## 2. Bronchodilators (e.g., Albuterol/Salbutamol)

* Sometimes used to relieve wheezing, but evidence of benefit is inconsistent.
* If used, treatment should continue only if objective improvement in breathing or oxygenation is observed.
* Side effects: Tremors, increased heart rate, nervousness.

## 3. Corticosteroids (Systemic or Inhaled)

* Corticosteroids like dexamethasone or prednisolone have been studied extensively.
* Routine use is not recommended as most studies show no significant benefit in acute bronchiolitis.
* May be considered in specific cases with a history of reactive airway disease or atopy.
* Side effects: Potential for immune suppression, increased risk of infections, mood changes, and in infants, unknown long-term effects on lung development.

## 4. Epinephrine (Nebulized)

* Some evidence supports use in infants aged 6 weeks to 12 months as cost-effective treatment combined with corticosteroids.
* Side effects: Increased heart rate, anxiety, tremors.

## 5. Antibiotics

* Not indicated unless there is confirmed bacterial infection.
* Side effects: Allergic reactions, antibiotic resistance, gastrointestinal upset.

## 6. Ribavirin

* Antiviral agent rarely used; reserved for immunocompromised patients.
* Side effects: Anemia, rash, fatigue.

## 7. Other Therapies

* Hypertonic saline nebulization (3% saline): May improve mucus clearance and symptoms in mild to moderate cases.
* Chest physiotherapy: Not recommended routinely as it has not shown benefit.

**Lifestyle and home remedies**

Though it may not be possible to shorten the length of your child's illness, you may be able to make your child more comfortable. Here are some tips:

* **Humidify the air.** If the air in your child's room is dry, a cool-mist humidifier or vaporizer can moisten the air. This may help loosen mucus and lessen coughing. Be sure to keep the humidifier clean so that bacteria and molds don't grow in the machine.
* **Give your child liquids to stay hydrated.** Infants should have formula or breast milk only. Your child's health care provider may add oral rehydration therapy. Older kids can drink whatever they want, such as water, juice or milk, as long as they're drinking. Your child may drink more slowly than usual because of swelling and mucus in the nose. Offer small amounts of liquid often.
* **Try saline nose drops to ease stuffiness.** You can buy these drops without a prescription. They are effective, safe and won't irritate the nose, even for children. Put several drops into the opening on one side of the nose, called the nostril, then bulb suction that nostril right away. Be careful not to push the bulb too far into the nose. Repeat the same steps in the other nostril.
* **Consider pain relievers that you can buy without a prescription.** For treatment of fever or pain, ask your child's health care provider about giving your child infants' or children's over-the-counter fever and pain medicines such as acetaminophen (Tylenol, others) or ibuprofen (Advil, Motrin, others). Those are safer than aspirin. Aspirin is not recommended in children due to the risk of Reye's syndrome, a rare but potentially life-threatening condition. Children and teenagers recovering from chickenpox or flu-like symptoms should never take aspirin, as they have a higher risk of Reye's syndrome.
* **Avoid secondhand smoke.** Smoke can worsen symptoms of respiratory infections. If a family member smokes, ask them to smoke outside of the house and outside of the car.

Don't use other over-the-counter medicines, except for fever reducers and pain relievers, to treat coughs and colds in children under 6 years old. Also, consider avoiding the use of these medicines for children younger than 12 years old. The risks to children outweigh the benefits.

## **Complications**

Complications of severe bronchiolitis may include:

* Low oxygen in the body.
* Pauses in breathing, which is most likely to happen in babies born too early and in babies under 2 months old.
* Not being able to drink enough liquids. This can cause dehydration, when too much body fluid is lost.
* Not being able to get the amount of oxygen needed. This is called respiratory failure.

If any of these happen, your child may need to be in the hospital. Severe respiratory failure may require that a tube be guided into the windpipe. This helps your child breathe until the infection improves.

**Prevention**

Because the viruses that cause bronchiolitis spread from person to person, one of the best ways to prevent infection is to wash your hands often. This is especially important before touching your baby when you have a cold, flu or other illness that can be spread. If you have any of these illnesses, wear a face mask.

If your child has bronchiolitis, keep your child at home until the illness is past to avoid spreading it to others.

To help prevent infection:

* **Limit contact with people who have a fever or cold.** If your child is a newborn, especially a premature newborn, avoid being around people with colds. This is especially important in the first two months of life.
* **Clean and disinfect surfaces.** Clean and disinfect surfaces and items that people often touch, such as toys and doorknobs. This is especially important if a family member is sick.
* **Wash hands often.** Frequently wash your own hands and those of your child. Wash with soap and water for at least 20 seconds. Keep an alcohol-based hand sanitizer handy to use when you're away from home. Make sure it contains at least 60% alcohol.
* **Cover coughs and sneezes.** Cover your mouth and nose with a tissue. Throw away the tissue. Then wash your hands. If soap and water aren't available, use a hand sanitizer. If you don't have a tissue, cough or sneeze into your elbow, not your hands.
* **Use your own drinking glass.** Don't share glasses with others, especially if someone in your family is ill.
* **Breastfeed, when possible.** Respiratory infections are less common in breastfed babies.

### **Immunizations and medicines**

In the U.S., respiratory syncytial virus (RSV) is the most common cause of bronchiolitis and pneumonia in children who are less than a year old. Two options for immunization can help prevent young infants from getting severe RSV. Both are recommended by the American Academy of Pediatrics, the American Academy of Family Physicians, the American College of Obstetricians and Gynecologists, and others.

You and your healthcare professional should discuss which option is best to protect your child:

* **Antibody product called nirsevimab (Beyfortus).** This antibody product is a single dose shot given in the month before or during RSV season. It's for newborn babies and those younger than 8 months born during or entering their first RSV season. In the U.S., the RSV season typically is November through March, but it varies in Florida, Alaska, Hawaii, Puerto Rico, Guam and other U.S. Pacific Island territories.
* Nirsevimab also should be given to children 8 months through 19 months old who are at higher risk of severe RSV disease through their second RSV season. Higher risk conditions include:
  + Children with active chronic lung disease from being born too soon (prematurely).
  + Children with a severely weakened immune system.
  + Children with severe cystic fibrosis.
  + American Indian or Alaska Native children.

In rare situations, when nirsevimab is not available or a child is not eligible for it, another antibody product called palivizumab may be given. But palivizumab requires monthly shots given during the RSV season, while nirsevimab is only one shot. Palivizumab is not recommended for healthy children or adults.

* **Vaccine for pregnant people.** The FDA approved an RSV vaccine called Abrysvo for pregnant people to prevent RSV in infants from birth through 6 months of age. A single dose shot of Abrysvo can be given sometime from 32 weeks through 36 weeks of pregnancy during September through January in the U.S. Abrysvo is not recommended for infants or children.

Other viruses can cause bronchiolitis too. These include COVID-19 and influenza or flu. Getting seasonal COVID-19 and flu shots every year is recommended for everyone older than 6 months.

**Outlook / Prognosis**

Your child may have symptoms for up to a week if they have bronchiolitis. During their illness, they may have trouble eating full meals or lose their appetite. To help your child eat when they don’t want to, try feeding them multiple small meals throughout the day instead of larger meals less often. It’s important to keep your child hydrated since they’re at a high risk of dehydration during their illness.

To alleviate your child’s symptoms, talk to their healthcare provider to see what’s safe for your child to take, like over the counter (OTC) medications to reduce a fever. Don’t give your child aspirin, as it can lead to Reye’s syndrome.

If your child has symptoms that don’t improve after one week or get worse, contact their healthcare provider. If your child has trouble breathing, contact emergency services or visit the emergency room immediately.

Some children develop asthma as they grow if they had bronchiolitis when they were infants. While less common, some children may develop pneumonia after bronchiolitis.

#### **Can you have bronchiolitis more than once?**

Yes. Since there are many viruses that can cause bronchiolitis, your child can develop the infection more than once.

## **Diagnostic Considerations**

Bronchiolitis and asthma have similar symptoms and signs, and some concern exists that patients with asthma could be misdiagnosed with bronchiolitis. The pathology of bronchiolitis involves edema of the airway wall rather than bronchoconstriction (as in asthma).

In addition to the conditions listed in the differential diagnosis, other problems to be considered include the following:

* Branchial cleft cyst
* Bronchiolitis obliterans-organizing pneumonia (BOOP)
* Bronchomalacia
* Cardiac disease
* Congenital heart disease
* Congenital lobar emphysema
* Congenital structural airway anomaly
* Constrictive bronchiolitis
* Gastroesophageal reflux
* Tracheal ring
* Vascular ring

## **Differential Diagnoses**

* Altitude-Related Disorders
* Aspiration Pneumonitis and Pneumonia
* Aspiration Syndromes
* Asthma
* Bacterial Pneumonia
* Chlamydial Pneumonias
* Chronic Obstructive Pulmonary Disease (COPD)
* Croup
* Cystic Fibrosis
* Heart Failure
* Mycoplasmal Pneumonia
* Pediatric Apnea
* Pediatric Foreign Body Ingestion
* Pediatric Pneumonia
* Pediatric Sepsis
* Pertussis
* Pneumothorax Imaging
* Viral Pneumonia

## **Epidemiology**

### United States statistics

Respiratory infection is observed in 25% of children younger than 12 months and 13% of children aged 1-2 years.Of these 25%, one half have wheezing-associated respiratory disease.RSV can be cultured from one third of these outpatients and from 80% of hospitalized children younger than 6 months of age.

Nearly 100% of children experience an RSV infection within 2 RSV seasons, and 1% are hospitalized.Among healthy full-term infants, 80% of hospitalizations due to bronchiolitis occur in the first year, and 50% of hospitalizations occur in children aged 1-3 months.Fewer than 5% of hospitalizations occur in the first 30 days of life, presumably because of transplacental transfer of maternal antibody.

Descriptive analysis of the US National Hospital Discharge Survey data from 1980 through 1996 showed that admissions associated with bronchiolitis totaled 1.65 million.In a retrospective analysis of data from the same source for 1997-2006, RSV-coded hospitalizations accounted for 24% of an estimated 5.5 million lower respiratory tract infection hospitalizations among children younger than 5 years of age.Between 2-3% of all children younger than 12 months of age are hospitalized with a diagnosis of bronchiolitis, which accounts for between 57,000 and 172,000 hospitalizations annually.The cost of hospitalization for bronchiolitis in children younger than 2 years is estimated to be more than $1.7 billion in 2009. While bronchiolitis remains a cause of significant mortality among children in the developing world, fewer than 100 annual deaths in the United States among young children are attributable to RSV infection.

In most regions of the United States, the highest RSV activity usually occurs in winter with peaks from October to February and a relative subsidence only from March to July. An exception is the subtropical regions of the southeastern United States (eg, Florida) where RSV is endemic throughout the year.

Secondary RSV infections occur in 46% of family members, 98% of other children attending a childcare center, 42% of hospital staff, and 45% of previously uninfected hospitalized infants.Infection is spread through self-inoculation of nasopharyngeal or ocular mucous membranes after direct contact with respiratory fomites and contaminated environmental surfaces. RSV can survive for several hours on hands and surfaces; therefore, handwashing and using disposable gloves and gowns may reduce nosocomial spread.

### International statistics

Bronchiolitis is a significant cause of respiratory disease worldwide. According to the World Health Organization bulletin, an estimated 150 million new cases occur annually; 11-20 million (7-13%) of these cases are severe enough to require hospital admission. Worldwide, 95% of all cases occur in developing countries.

The frequency of bronchiolitis in developed countries appears to be similar to that in the United States. Epidemiologic data for underdeveloped countries are incomplete. Epidemiologic data from underdeveloped countries show that RSV is a predominant viral cause of acute lower respiratory tract infections and accounts for about 65% of hospitalizations attributed to viruses.

Despite incomplete data about RSV-associated mortality from developing countries, in 2005, RSV alone was estimated to cause 66,000 to 199,000 deaths among children younger than 5 years of age.Morbidity and mortality is higher in less-developed countries likely because of poor nutrition and lack of resources for supportive medical care.

In the northern hemisphere, RSV epidemics generally occur annually in winter and late spring, whereas parainfluenza outbreaks usually occur in the fall. Conversely, in the southern hemisphere, wintertime epidemics occur from May to September.

Descriptive epidemiologic data from a population-based cohort (Georgia Air Basin, Canada) reported by Koehoorn et al indicated that from 1999 through 2002, bronchiolitis was associated with 12,474 inpatient and outpatient physician contacts during the first year of life.This equates to 134.2 cases per 1000 person-years. In total, 1588 bronchiolitis cases resulted in hospitalization (17.1 cases per 1000 person-years).

### Age-related demographics

Although infection with the agents that cause bronchiolitis may occur at any age, the clinical entity of bronchiolitis includes only infants and young children. About 75% of cases of bronchiolitis occur in children younger than 1 year and 95% in children younger than 2 years. Incidence peaks in those aged 2-8 months.

Age is a significant factor in the severity of infection: The younger the patient is, the more severe the infection tends to be, as measured by the lowest oxygen saturation. Infants younger than 6 months are most severely affected, owing to their smaller, more easily obstructed airways and their decreased ability to clear secretions.

Intrauterine cigarette-smoke exposure may impair in utero airway development or alter the elastic properties of the lung tissue. Exposure to second-hand cigarette smoke (e.g., by a parent or family member) in the postnatal period compounds the severity of RSV bronchiolitis in infants.

Although RSV bronchiolitis is clearly a significant disease of the young child, immunity has been shown to wane over time; susceptible adults may be asymptomatic or mildly symptomatic and act as carriers. With the increasing use of treatment modalities that compromise cellular immunity, RSV infection may be life-threatening to older children and adults undergoing organ and bone marrow transplantation, as well as to the elderly.

### Sex-related demographics

Severe bronchiolitis occurs more frequently in males than in females; a pattern like other respiratory viral infections. The exact reason for this difference is unknown.Death is 1.5 times more likely in males.

### Race-related demographics

Race and low socioeconomic status may adversely affect outcome in patients with acute bronchiolitis. Multiple population-based reports sponsored by the Centers for Disease Control and Prevention (CDC) indicate no disparity in the rates of hospitalization for RSV infection between black and white children. A study by La Via et al demonstrated that although more minority children than white children were hospitalized with RSV infection, nothing indicated that the infections in minority children were more or less severe than those in white children.

Lower socioeconomic status may increase the likelihood of hospitalization. Hospitalization rates are higher in Native American, Alaskan, and Hispanic populations, but it is not clear if this is due to more severe infection or to a lower threshold for admission.

## **Guidelines**

### **AAP guidelines for pediatric bronchiolitis**

The American Academy of Pediatrics released updated guidelines on the diagnosis, treatment, and prevention of bronchiolitis in children aged 1 to 23 months. The guidelines emphasize the use of supportive care, including hydration and oxygen.Other recommendations include the following:

* As multiple viruses may cause bronchiolitis, testing for specific viruses is not necessary.
* Routine radiographic or laboratory studies are also not necessary. Diagnosis and assessment of bronchiolitis severity should be based on patient history and physical examination.
* There is no need for a trial dose of a bronchodilator.
* Otherwise, healthy infants with gestational age of 29 weeks or more should not receive palivizumab to prevent respiratory syncytial virus infections. Infants under one year of age with hemodynamically significant heart disease or chronic lung disease of prematurity should be treated with palivizumab, up to a maximum of 5 monthly doses, during the respiratory syncytial virus season.
* Risk factors for severe disease include age less than 12 weeks, prematurity, underlying cardiopulmonary disease, and immunodeficiency.
* Epinephrine and chest physiotherapy should not be administered to infants and children with bronchiolitis

**GENOMIC DATA**

* TLR4 Gene Variants:  
  Mutations in the Toll-like receptor 4 (TLR4) gene, specifically the Asp299Gly and Thr399Ile polymorphisms, are strongly associated with increased severity of RSV bronchiolitis in infants under 12 months. These variants impair TLR4 receptor function on bronchial epithelial cells, leading to reduced immune signaling and a blunted response to RSV infection, which increases the risk of severe disease requiring hospitalization.
* DNAH9 Gene Mutations:  
  Variants in the DNAH9 gene have been linked to post-infectious bronchiolitis obliterans (PIBO), a chronic obstructive lung disease following severe bronchiolitis. Whole exome sequencing in pediatric patients identified multiple DNAH9 mutations associated with airway obstruction after infections such as Mycoplasma pneumoniae and adenovirus.
* ABCA3 and Surfactant Protein Mutations:  
  Mutations in surfactant-related genes like ABCA3 may contribute to the pathogenesis of PIBO by affecting lung surfactant function and airway repair mechanisms.
* Interleukin-8 (IL-8) Polymorphisms:  
  Certain IL-8 gene polymorphisms have been studied in infants with bronchiolitis, suggesting a role in modulating inflammatory responses during infection.
* RSV Viral Genetic Variations:  
  Studies of the RSV virus itself shows multiple mutations, especially in the fusion (F) and attachment (G) proteins, which may affect viral infectivity and immune evasion, potentially impacting disease severity and vaccine development

**PREDEFINED Q AND A**

## 1. Should I give my child medication? If so, for how long and at what times of the day?

* Most bronchiolitis cases are managed with supportive care rather than medications.
* Routine use of bronchodilators (like albuterol) or corticosteroids is not recommended as they have not shown consistent benefit.
* In some cases, a single trial of a bronchodilator may be considered if there is a family history of asthma or atopy, but treatment should be stopped if no improvement is seen.
* Antibiotics are only needed if there is a confirmed bacterial infection.
* Always follow your doctor’s specific instructions if medications are prescribed.

## 2. When will my child start to feel better?

* Symptoms usually improve within 7 to 10 days, but the cough and mild breathing difficulties can last for 2 to 3 weeks.
* Most infants recover fully without complications.

## 3. Will I need to bring my child back for a follow-up visit?

* Follow-up is recommended if symptoms worsen or do not improve within 2 to 3 weeks.
* Also bring your child back if you notice increased difficulty breathing, poor feeding, dehydration, or persistent high fever.
* Some children with risk factors or severe illness may require hospital observation.

## 4. Should I keep my child home from school or daycare?

* Yes, keep your child home while they have symptoms, especially if they have a cough, runny nose, or fever, to prevent spreading the infection.
* Your child can return once they are fever-free and feeling better, but consult your healthcare provider for specific advice.

## 5. Which over-the-counter pain relievers do you recommend?

* Use acetaminophen (paracetamol) or ibuprofen to reduce fever and relieve discomfort.
* Avoid aspirin in children due to the risk of Reye’s syndrome.
* Follow dosing instructions carefully based on your child’s age and weight.

## 6. Which symptoms should I look out for?

Seek medical attention if your child develops:

* Difficulty breathing or rapid breathing
* Persistent high fever (above 38.5°C or 101.3°F)
* Poor feeding or signs of dehydration (few wet diapers, dry mouth)
* Bluish color around lips or face
* Severe lethargy or unresponsiveness
* Wheezing or chest retractions (pulling in of skin between ribs or neck)

## **Admission Criteria**

A decision must be made as to whether the patient should be treated in an inpatient or an outpatient setting. For hospitalized patients, the length of stay averages 2-3 days, with a readmission rate of 1-4%. Considerations for hospital admission may include the following:

* Persistent resting oxygen saturation below 90% in room air
* Markedly elevated respiratory rate (>70-80 breaths/min)
* Dyspnea, intercostal retractions and cyanosis (indicating respiratory distress)
* Chronic lung disease, especially if the patient is already receiving supplemental oxygen
* Congenital heart disease, especially if hemodynamically significant (associated with cyanosis or pulmonary hypertension)
* Prematurity
* Age younger than 3 months, when severe disease is most common
* Inability to maintain oral hydration in patients younger than 6 months and difficulty feeding as a consequence of respiratory distress
* Parent unable to care for child at home

A decision must also be made regarding admission to an intensive care unit (ICU). Criteria for ICU admission vary greatly. In general, ICU admission is uncommon for previously healthy infants who present with bronchiolitis. Severely ill children should be admitted to an adequately equipped intensive care unit (ICU). If this requires transfer to another hospital, transport personnel and vehicles specifically intended for pediatric transport are desirable.

Patients with the following conditions should be evaluated for ICU admission:

* Worsening hypoxemia or hypercapnia
* Worsening respiratory distress
* Persistent oxygen desaturation and/or severe cyanosis in spite of adequate oxygen delivery
* Apnea
* Acidosis
* Extrapulmonary symptoms
* Worsening mental status
* Unclear etiology of symptoms

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I see your child has been coughing and having some breathing difficulties. Can you tell me more about the symptoms?

Parent: Yes, my baby has had a runny nose and cough for a few days, and now they seem to be breathing faster and wheezing a bit.

Doctor: That sounds like bronchiolitis, which is a common viral infection in young children that causes inflammation in the small airways. It usually gets better on its own with supportive care.

Parent: Is this serious? What can I do to help my child?

Doctor: Most cases are mild and can be managed at home. The best treatment is supportive care: keeping your child well hydrated, using saline nose drops and gentle suctioning to clear nasal mucus, and making sure they get plenty of rest. If your child has trouble breathing or low oxygen levels, they may need hospital care.

Parent: Should my child be given any medications?

Doctor: Routine use of bronchodilators like albuterol or steroids is not recommended because studies show they don’t help most children with bronchiolitis. Antibiotics are only needed if there’s a bacterial infection, which is uncommon. You can give acetaminophen or ibuprofen if your child has a fever or seems uncomfortable.

Parent: How long will it take for my child to get better?

Doctor: Symptoms usually peak around 3 to 5 days and improve over 1 to 2 weeks. The cough may linger for a few more weeks.

Parent: When should I bring my child back or seek emergency care?

Doctor: If your child has difficulty breathing, is feeding poorly, has persistent high fever, becomes very lethargic, or if you notice blue lips or face, you should seek medical attention immediately. Also, if symptoms don’t improve after 2 to 3 weeks, a follow-up visit is advisable.

Parent: Should my child stay home from daycare?

Doctor: Yes, it’s best to keep your child home while they have symptoms to prevent spreading the infection to others.

Doctor: I’ll give you some written information about bronchiolitis and what signs to watch for. If you have any concerns, don’t hesitate to contact us.

REFERENCES

[Bronchiolitis Causes & Symptoms](https://my.clevelandclinic.org/health/diseases/8272-bronchiolitis#outlook-prognosis)

[Bronchiolitis - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/bronchiolitis/diagnosis-treatment/drc-20351571)

<https://www.ncbi.nlm.nih.gov/books/NBK573086/>

<https://emedicine.medscape.com/article/961963-treatment>

### **Pneumonia**

## **Alternative Names for Pneumonia**

* Lung Infection  
  A general term describing infection of lung tissue.
* Pulmonary Infection  
  Another broad term emphasizing infection within the lungs.
* Lobar Pneumonia  
  Pneumonia affecting a large, continuous area (lobe) of the lung.
* Bronchopneumonia (Lobular Pneumonia)  
  Pneumonia involving patches of infection around the bronchi and bronchioles.
* Community-Acquired Pneumonia (CAP)  
  Pneumonia acquired outside of healthcare settings.
* Hospital-Acquired Pneumonia (HAP) / Nosocomial Pneumonia  
  Pneumonia acquired during or after hospitalization.
* Aspiration Pneumonia  
  Pneumonia caused by inhalation of foreign material such as food, liquids, or vomit into the lungs.
* Atypical Pneumonia  
  Pneumonia caused by atypical bacteria like Mycoplasma pneumoniae or Chlamydophila pneumoniae, often with milder symptoms.
* Viral Pneumonia  
  Pneumonia caused by viruses such as influenza or respiratory syncytial virus (RSV).
* Bacterial Pneumonia  
  Pneumonia caused by bacteria like Streptococcus pneumoniae.
* Walking Pneumonia  
  A mild form of pneumonia, often caused by Mycoplasma pneumoniae, where patients remain ambulatory.

**DEFINITION AND DESCRIPTION**

Pneumonia is an infection in your lungs caused by bacteria, viruses or fungi. Pneumonia causes your lung tissue to swell (inflammation) and can cause fluid or pus in your lungs. Bacterial pneumonia is usually more severe than viral pneumonia, which often resolves on its own.

Pneumonia can affect one or both lungs. Pneumonia in both of your lungs is called bilateral or double pneumonia.

### **Viral and bacterial pneumonia**

While all pneumonia is inflammation caused by an infection in your lungs, you may have different symptoms depending on whether the root cause is a virus, bacteria or fungi.

Bacterial pneumonia tends to be more common and more severe than viral pneumonia. It’s more likely to require a hospital stay. Providers treat bacterial pneumonia with antibiotics. Viral pneumonia causes flu-like symptoms and is more likely to resolve on its own. You usually don’t need specific treatment for viral pneumonia.

### **Types of pneumonia**

We categorize pneumonia by which pathogen (virus, bacteria or fungi) caused it and how you got it — community-acquired, hospital-acquired or ventilator-associated pneumonia.

#### **Community-acquired pneumonia (CAP)**

When you get pneumonia outside of a healthcare facility, it’s called community-acquired pneumonia. Causes include:

* **Bacteria:** Infection with *Streptococcus pneumoniae* bacteria, also called pneumococcal disease, is the most common cause of CAP. Pneumococcal disease can also cause ear infections, sinus infections and meningitis. Mycoplasma pneumoniae bacteria causes atypical pneumonia, which usually has milder symptoms. Other bacteria that cause CAP include *Haemophilus influenza*, *Chlamydia pneumoniae* and *Legionella* (Legionnaires’ disease).
* **Viruses:** Viruses that cause the common cold, the flu (influenza), COVID-19 and respiratory syncytial virus (RSV) can sometimes lead to pneumonia.
* **Fungi (molds):** Fungi, like *Cryptococcus*, *Pneumocystis jirovecii* and *Coccidioides*, are uncommon causes of pneumonia. People with compromised immune systems are most at risk of getting pneumonia from a fungus.
* **Protozoa:** Rarely, protozoa like *Toxoplasma* cause pneumonia.

#### **Hospital-acquired pneumonia (HAP)**

You can get hospital-acquired pneumonia (HAP) while in a hospital or healthcare facility for another illness or procedure. HAP is usually more serious than community-acquired pneumonia because it’s often caused by antibiotic-resistant bacteria, like methicillin-resistant *Staphylococcus aureus* (MRSA). This means HAP can make you sicker and be harder to treat.

#### **Healthcare-associated pneumonia (HCAP)**

You can get HCAP while in a long-term care facility (such as a nursing home) or outpatient, extended-stay clinics. Like hospital-acquired pneumonia, it’s usually caused by antibiotic-resistant bacteria.

**Ventilator-associated pneumonia (VAP)**

If you need to be on a respirator or breathing machine to help you breathe in the hospital (usually in the ICU), you’re at risk for ventilator-associated pneumonia (VAP). The same types of bacteria as community-acquired pneumonia, as well as the drug-resistant kinds that cause hospital-acquired pneumonia, cause VAP.

#### **Aspiration pneumonia**

Aspiration is when solid food, liquids, spit or vomit go down your trachea (windpipe) and into your lungs. If you can’t cough these up, your lungs can get infected.

Learn common symptoms of pneumonia and when you should seek medical attention.

### **Pneumonia versus the common cold or the flu**

It can be difficult to tell the difference between the symptoms of a cold, the flu and pneumonia, and only a healthcare provider can diagnose you. As pneumonia can be life-threatening, it’s important to seek medical attention for serious symptoms that could be signs of pneumonia, such as:

* Congestion or chest pain.
* Difficulty breathing.
* A fever of 102 degrees Fahrenheit (38.88 degrees Celsius) or higher.
* Coughing up yellow, green or bloody mucus or spit.

### **Who is most at risk of getting pneumonia?**

You’re at an increased risk of pneumonia if you:

* **Are over the age of 65 and or under the age of 2.**
* **Are living with a lung or heart condition.** Examples include cystic fibrosis, asthma, chronic obstructive pulmonary disease, emphysema, pulmonary fibrosis or sarcoidosis.
* **Are living with a neurological condition that makes swallowing difficult.** Conditions like dementia, Parkinson’s disease and stroke increase your risk of aspiration pneumonia.
* **Are in the hospital or at a long-term care facility.**
* **Smoke.**
* **Are pregnant.**
* **Have a weakened immune system.** You might have a weakened immune system if you’re on chemotherapy, are an organ transplant recipient, are living with HIV/AIDS or are taking medications that suppress your immune system.

Anyone can get pneumonia. It’s a common illness, with millions of people diagnosed each year in the United States. About 55,000 people die each year of pneumonia in the U.S. It’s the most common cause of death in developing countries.

### **Causes**

Pneumonia can develop when your immune system attacks an infection in the small sacs of your lung (alveoli). This causes your lungs to swell and leak fluids.

Many bacteria, viruses and fungi can cause the infections that lead to pneumonia. Bacteria are the most common cause in adults and viruses are the most common cause in school-aged children. Common illnesses that can lead to pneumonia include:

* Common cold (rhinovirus).
* COVID-19 (SARS-COV-2).
* The flu (influenza virus).
* Human metapneumovirus (HMPV).
* Human parainfluenza virus (HPIV).
* Legionnaires’ [disease](https://my.clevelandclinic.org/health/diseases/17750-legionnaires-disease).
* Mycoplasma pneumonia bacteria.
* Pneumococcal disease.
* Pneumocystis pneumonia.
* Respiratory syncytial virus (RSV).

### **Is pneumonia contagious?**

Pneumonia itself isn’t contagious, but the bacteria and viruses that cause it are. For instance, the flu is contagious and can lead to pneumonia, but most people who get the flu won’t get pneumonia.

The bacteria that most commonly causes pneumonia, *Streptococcus pneumoniae,* can be spread from person to person by touching infected surfaces or through coughing and sneezing.

Pneumonia caused by fungi isn’t contagious. Fungal infections aren’t spread from person to person like viruses and bacteria.

### **signs and symptoms of pneumonia**

Symptoms of pneumonia depend on the cause. Symptoms can range from mild to severe. Babies, young children and older adults may have different symptoms.

#### **Symptoms of bacterial pneumonia**

Symptoms of bacterial pneumonia can develop gradually or suddenly. Symptoms include:

* High fever (up to 105 F or 40.55 C).
* Cough with yellow, green or bloody mucus.
* Tiredness (fatigue).
* Rapid breathing.
* Shortness of breath.
* Rapid heart rate.
* Sweating or chills.
* Chest pain and/or abdominal pain, especially with coughing or deep breathing.
* Loss of appetite.
* Bluish skin, lips or nails (cyanosis).
* Confusion or altered mental state.

**Symptoms of viral pneumonia**

Symptoms of viral pneumonia usually develop over several days. You might have symptoms similar to bacterial pneumonia, or you might additionally have:

* Dry cough.
* Headache.
* Muscle pain.
* Extreme tiredness or weakness.

#### **Symptoms of pneumonia in young children**

Babies and newborns may not show any symptoms of pneumonia, or their symptoms may be different from adults, including:

* Fever, chills, general discomfort, sweating/flushed skin.
* Cough.
* Difficulty breathing or rapid breathing (tachypnea).
* Loss of appetite.
* Vomiting.
* Lack of energy.
* Restlessness or fussiness.

Signs you can look for in babies and young children include:

* Grunting sound with breathing or noisy breathing.
* A decreased amount of pee or diapers that are less wet.
* Pale skin.
* Limpness.
* Crying more than usual.
* Difficulty feeding.

#### **Symptoms of pneumonia in adults over 65**

Adults over 65 or those with weakened immune systems may have mild or less noticeable symptoms of pneumonia (like cough and shortness of breath). Symptoms of ongoing health conditions may worsen. Older adults may experience:

* A sudden change in mental state.
* Low appetite.
* Fatigue.

#### **When to see a doctor**

See your doctor if you have difficulty breathing, chest pain, persistent fever of 102 F (39 C) or higher, or persistent cough, especially if you're coughing up pus.

It's especially important that people in these high-risk groups see a doctor:

* Adults older than age 65
* Children younger than age 2 with signs and symptoms
* People with an underlying health condition or weakened immune system
* People receiving chemotherapy or taking medication that suppresses the immune system

For some older adults and people with heart failure or chronic lung problems, pneumonia can quickly become a life-threatening condition.

## **Diagnosis and Tests**

To diagnose pneumonia, a healthcare provider will ask about your health history and conduct a physical exam. They’ll listen to your lungs with a stethoscope and may perform or order additional tests. These include imaging (like chest X-rays), pulse oximetry (checking oxygen levels in your blood), blood tests or sputum (spit) tests.

Even if your healthcare provider confirms that you have pneumonia, sometimes, they can’t find the exact cause.

Your provider may perform tests that look at your lungs for signs of infection, measure how well your lungs are working and examine blood or other body fluids to try to determine the cause of your pneumonia. These include:

* **Imaging:** Your provider can use chest X-ray or CT scan to take pictures of your lungs to look for signs of infection.
* **Blood tests:** Your provider can use a blood test to help determine what kind of infection is causing your pneumonia.
* **Sputum test:** You’re asked to cough and then spit into a container to collect a sample for a lab to examine. The lab will look for signs of an infection and try to determine what’s causing it.
* **Pulse oximetry:** A sensor measures the amount of oxygen in your blood to give your provider an idea of how well your lungs are working.
* **Pleural fluid culture:** Your provider uses a thin needle to take a sample of fluid from around your lungs. The sample is sent to a lab to help determine what’s causing the infection.
* **Arterial blood gas test:** Your provider takes a blood sample from your wrist, arm or groin to measure oxygen levels in your blood to know how well your lungs are working.
* **Bronchoscopy:** In some cases, your provider may use a thin, lighted tube called a bronchoscope to look at the inside of your lungs. They may also take tissue or fluid samples to be tested in a lab.

## **Management and Treatment**

Treatment for pneumonia depends on the cause — bacterial, viral or fungal — and how serious your case is. In many cases, the cause can’t be determined, and treatment is focused on managing symptoms and making sure your condition doesn’t get worse.

Some treatments may include:

* **Antibiotics:** Antibiotics treat bacterial pneumonia. They can’t treat a virus, but a provider may prescribe them if you have a bacterial infection at the same time as a virus.
* **Antifungal medications:** Antifungals can treat pneumonia caused by a fungal infection.
* **Antiviral medications:** Viral pneumonia usually isn’t treated with medication and can go away on its own. A provider may prescribe antivirals such as oseltamivir (Tamiflu®), zanamivir (Relenza®) or peramivir (Rapivab®) to reduce how long you’re sick and how sick you get from a virus.
* **Oxygen therapy:** If you’re not getting enough oxygen, a provider may give you extra oxygen through a tube in your nose or a mask on your face.
* **IV fluids:** Fluids delivered directly to your vein (IV) treat or prevent dehydration.
* **Draining of fluids:** If you have a lot of fluid between your lungs and chest wall (pleural effusion), a provider may drain it. This is done with a catheter or surgery.

### **Can pneumonia go away on its own?**

Viral pneumonia often goes away on its own, but you should always follow your healthcare provider’s recommendations to treat symptoms and reduce your risk of serious complications.

### **How do I manage the symptoms of pneumonia?**

Over-the-counter medications and other at-home treatments can help you feel better and manage the symptoms of pneumonia, including:

* **Pain relievers and fever reducers:** Your provider may recommend medicines like ibuprofen (Advil®) and acetaminophen (Tylenol®) to help with body aches and fever.
* **Cough suppressants:** Check with your healthcare provider before taking cough suppressants for pneumonia. Coughing is important to help clear your lungs.
* **Breathing treatments and exercises:** Your provider may prescribe these treatments to help loosen mucus and help you to breathe.
* **Using a humidifier:** Your provider may recommend keeping a small humidifier running by your bed or taking a steamy shower or bath to make it easier to breathe.
* **Drinking plenty of fluids.**

### **How soon after treatment for pneumonia will I begin to feel better?**

How soon you’ll feel better depends on:

* Your age.
* The cause of your pneumonia.
* The severity of your pneumonia.
* If you have other health conditions or complications.

If you’re otherwise healthy, most symptoms of bacterial pneumonia usually begin to improve within 24 to 48 hours after starting treatment. You might start to feel better after a few days of treatment for viral pneumonia. Some symptoms, like cough and fatigue, may linger for several weeks.

### **How long am I contagious if I have pneumonia?**

If you have bacterial pneumonia, you’re no longer considered contagious when your fever is gone and you’ve been on antibiotics for at least two days. If you have viral pneumonia, you’re still considered contagious until you feel better and have been free of fever for several days.

## **Common Medications Used to Treat Pneumonia**

|  |  |  |  |
| --- | --- | --- | --- |
| Drug Class | Examples | Purpose | Common Side Effects |
| Macrolide Antibiotics | Azithromycin (Zithromax), Clarithromycin (Biaxin), Erythromycin | First-line for community-acquired pneumonia (CAP), effective against atypical bacteria | Gastrointestinal upset (nausea, diarrhea), liver enzyme elevation, QT prolongation (rare) |
| Tetracyclines | Doxycycline | Alternative for CAP, especially in mild cases or penicillin allergy | Photosensitivity, gastrointestinal upset, tooth discoloration in children |
| Fluoroquinolones | Levofloxacin (Levaquin), Moxifloxacin (Avelox) | Used for more severe cases or resistant bacteria | Tendonitis/tendon rupture, QT prolongation, CNS effects (dizziness) |
| Beta-lactams (Penicillins and Cephalosporins) | Amoxicillin, Amoxicillin/clavulanate (Augmentin), Ceftriaxone, Cefuroxime | Effective against common bacterial pathogens | Allergic reactions, gastrointestinal upset, rash |
| Clindamycin | Clindamycin | Used for suspected anaerobic infections or MRSA coverage | Diarrhea, risk of Clostridioides difficile infection |
| Vancomycin | Vancomycin | Reserved for MRSA or resistant infections | Nephrotoxicity, infusion reactions (red man syndrome) |
| Antiviral Medications | Oseltamivir (Tamiflu), Zanamivir (Relenza) | For viral pneumonia caused by influenza viruses | Nausea, vomiting, headache |

* Antibiotics are used only for bacterial pneumonia. Viral pneumonia typically does not require antibiotics but may be treated with antivirals if caused by influenza.
* The choice of antibiotic depends on factors such as pneumonia type, severity, patient allergies, and local resistance patterns.
* Intravenous antibiotics may be necessary for severe pneumonia or hospitalized patients.
* Always follow your healthcare provider’s instructions regarding dosage and duration.

**Outlook / Prognosis**

If you’re otherwise healthy, you can recover quickly from pneumonia when you get prompt care. However, pneumonia can be life-threatening if left untreated, especially if you have an underlying health condition.

Even people who’ve been successfully treated and have fully recovered may face long-term health issues. After recovering from pneumonia, you may experience:

* Decreased ability to exercise.
* Worsening of cardiovascular disease.
* General decline in quality of life.

Children who’ve recovered from pneumonia have an increased risk of chronic lung diseases.

Follow up with your healthcare provider if you have ongoing health concerns after recovering from pneumonia.

#### **Possible complications of pneumonia**

Pneumonia can lead to serious complications that can require hospitalization, including:

* **Breathing difficulties**. Pneumonia can lead to respiratory failure or acute respiratory distress syndrome (ARDS).
* **Fluid around your lungs (pleural effusion).**
* **Bacteria in your bloodstream (bacteremia), or sepsis.** The bacteria that cause pneumonia can enter your bloodstream, spreading the infection to other organs and leading to sepsis or organ failure.
* **Lung abscess.** Pneumonia can lead to pus-filled holes in your lungs.

#### **When would I need to be hospitalized for pneumonia?**

If you have a severe case of pneumonia or complications, you may need to stay in the hospital for treatment. You’re more likely to be hospitalized for pneumonia if you’re:

* Under age 2 or over age 65.
* Have a weakened immune system.
* Have health conditions that affect your heart and lungs.

It may take six to eight weeks to feel back to normal if you’ve been hospitalized with pneumonia.

**Prevention**

The best way to prevent pneumonia is to get vaccinated against bacteria and viruses that commonly cause it. There are also everyday precautions you can take to help reduce your risk of pneumonia.

#### **Vaccines for pneumonia**

There are two types of vaccines (shots) that prevent pneumonia caused by pneumococcal bacteria. Similar to a flu shot, these vaccines won’t protect against all types of pneumonia, but if you do get sick, it’s less likely to be severe.

* **Pneumococcal vaccines:** Pneumovax23® and Prevnar13® protect against pneumonia bacteria. They’re each recommended for certain age groups or those with increased risk for pneumonia. Ask your healthcare provider which vaccine would be appropriate for you or your loved ones.
* **Vaccinations against viruses:** As certain viruses can lead to pneumonia, getting vaccinated against COVID-19 and the flu can help reduce your risk of getting pneumonia.
* **Childhood vaccinations:** If you have children, ask their healthcare provider about other vaccines they should get. Several childhood vaccines help prevent infections caused by the bacteria and viruses that can lead to pneumonia.

#### **Other ways to reduce your risk of pneumonia**

In addition to getting vaccinated, you can reduce your risk of getting and spreading pneumonia with some healthy habits:

* Quit smoking and avoid secondhand smoke. Smoking damages your lungs and makes you more likely to get an infection.
* Wash your hands with soap and water before eating, before handling food and after using the restroom. If soap isn’t available, use an alcohol-based hand sanitizer.
* Avoid close contact and sharing items with other people if either of you has an infectious disease such as the flu, a cold or COVID-19.
* If you have to stay in a hospital or other healthcare facility, don’t be afraid to ask your providers about how to reduce your risk of getting an infection during your stay.
* Eat a healthy diet, exercise and get enough rest.
* Get treated for any other infections or health conditions you may have. These conditions could weaken your immune system, which could increase your chance of pneumonia.
* Avoid excessive alcohol consumption.

## **Living With**

You can help yourself feel better while you have pneumonia by:

* Managing your symptoms as recommended by your healthcare provider.
* Finishing all medications and therapies prescribed by your provider. Don’t stop taking antibiotics when you start feeling better. Continue taking them until no pills remain. If you don’t take all of your antibiotics, your pneumonia may come back.
* If your provider has recommended over-the-counter medicines to reduce fever (aspirin, acetaminophen, ibuprofen, naproxen), take them as directed on the label. Never give aspirin to children.
* Getting lots of rest.

If at any time you start to feel worse, call your doctor right away.

### **Signs that pneumonia is improving**

As you begin to recover from pneumonia, your temperature will probably return to normal first. After that, you may notice that you’re coughing up less mucus. Feeling like you’re up to returning to some of your normal activities is a good sign that you’re improving.

### **When can I return to work, school and regular activities if I have pneumonia?**

You can typically resume your normal activities if your symptoms are gone, mild or improving and you don’t have new or worsening:

* Shortness of breath or tiredness (less energy).
* Chest pain.
* Mucus, fever or cough.

If you’re generally healthy, most people feel well enough to return to previous activities in about a week. However, it may take about a month to feel totally back to normal.

## **Diagnostic Considerations**

Pneumonia can occur at any age, although it is more common in younger children. Different age groups tend to be infected by different pathogens, which affects diagnostic and therapeutic decisions.

Many patients referred for evaluation for recurrent pneumonia are diagnosed with asthma. In emergency department studies, 35% of children with an asthma exacerbation have abnormalities visible on chest radiographs. In a child not yet diagnosed with asthma, these abnormalities are frequently interpreted simply as pneumonia. Inflammation, often triggered by viral infection, is part of the asthmatic response. Wheezing responsive to bronchodilators, a history of atopy, a family history of asthma, and a history of cough or wheeze with exercise may be helpful in identifying such patients.

Consider any other diseases that may present with respiratory dysfunction in the first 24 hours of life. Keep in mind that any of the conditions listed below may also be superimposed by pneumonia:

* Alveolar-capillary dysplasia
* Arrhythmia
* Asphyxia
* Bronchial duplication
* Chest wall injury or anomaly
* Choanal atresia
* Chylothorax
* Diaphragmatic eventration
* Heart block
* Intracranial hemorrhage
* Laryngeal cleft
* Laryngeal nerve injury
* Mutation of *ABCA3* gene (for surfactant phospholipid transport)
* Neuromuscular disorders
* Phrenic nerve injury
* Pulmonary hemorrhage
* Pulmonary hypoplasia
* Pulmonary lymphangiectasia
* Spinal injury
* Surfactant-related protein B deficiency
* Tachycardia syndromes
* Tracheoesophageal fistula
* Transplacental medications
* Vascular catheter accident
* Other causes of airway obstruction
* Other congenital heart diseases
* Other inborn errors of metabolism
* Other neuromuscular diseases

A careful history and examination in patients with recurrent pneumonia are both very helpful to further narrow the differential diagnosis. However, additional testing is often needed to confirm most of these diagnoses and is generally outside the scope of a primary care provider.

## **Differential Diagnoses**

* Acidosis, Metabolic
* Acute Anemia
* Acute Hypoglycemia
* Afebrile Pneumonia Syndrome
* Agammaglobulinemia
* Alveolar Proteinosis
* Aortic Stenosis
* Aortic Stenosis, Subaortic
* Aortic Stenosis, Valvar
* Aseptic Meningitis
* Asphyxiating Thoracic Dystrophy (Jeune Syndrome)
* Aspiration Syndromes
* Asthma
* Atelectasis, Pulmonary
* Atrioventricular Septal Defect, Complete
* Atrioventricular Septal Defect, Unbalanced
* Bacteremia
* Birth Trauma
* Bowel Obstruction in the Newborn
* Bronchiectasis
* Bronchiolitis
* Bronchitis
* Bronchitis, Acute and Chronic
* Bronchogenic Cyst
* Cardiomyopathy, Hypertrophic
* Chronic Anemia
* Chronic Granulomatous Disease
* Coarctation of the Aorta
* Coccidioidomycosis and Valley Fever
* Severe Combined Immunodeficiency (SCID)
* Common Variable Immunodeficiency
* Complement Deficiency
* Complement Receptor Deficiency
* Congenital Diaphragmatic Hernia
* Congenital Pneumonia
* Congenital Stridor
* Cystic Adenomatoid Malformation
* Cystic Fibrosis
* Double Outlet Right Ventricle, Normally Related Great Arteries
* Double Outlet Right Ventricle, With Transposition
* Ebstein Anomaly
* Emergent Management of Atrial Flutter
* Empyema
* Esophageal Atresia With or Without Tracheoesophageal Fistula
* Foreign Body Aspiration
* Gastroesophageal Reflux
* Goodpasture Syndrome
* Head Trauma
* Hemosiderosis
* Hemothorax
* Human Immunodeficiency Virus Infection
* Hypersensitivity Pneumonitis
* Hypocalcemia
* Hypoplastic Left Heart Syndrome
* IgA and IgG Subclass Deficiencies
* Inhalation Injury
* Interrupted Aortic Arch
* Legionella Infection
* Meningitis, Bacterial
* Neural Tube Defects
* Patent Ductus Arteriosus (PDA)
* Pediatric Acute Respiratory Distress Syndrome
* Pediatric Airway Foreign Body
* Pediatric Histoplasmosis
* Pediatric Pleural Effusion
* Pediatric Pneumococcal Infections
* Pediatric Pulmonary Hypoplasia
* Pertussis
* Pneumococcal Infections
* Pneumonia, Aspiration
* Pneumonia, Bacterial
* Pneumonia, Empyema and Abscess
* Pneumonia, Immunocompromised
* Pneumonia, Mycoplasma
* Pneumothorax
* Pulmonary Atresia with Intact Ventricular Septum
* Pulmonary Atresia with Ventricular Septal Defect
* Pulmonary Hypertension, Persistent-Newborn
* Pulmonary Sequestration
* Q Fever
* Respiratory Distress Syndrome
* Respiratory Distress Syndrome
* Smoke Inhalation Injury
* Total Anomalous Pulmonary Venous Connection
* Transient Tachypnea of the Newborn
* Transposition of the Great Arteries
* Tricuspid Atresia
* Truncus Arteriosus
* Vascular Ring, Double Aortic Arch
* Vascular Ring, Right Aortic Arch

## **Epidemiology**

### United States statistics

Pneumonia can occur at any age, although it is more common in younger children. Pneumonia accounts for 13% of all infectious illnesses in infants younger than 2 years of age. In a large community-based study conducted by Denny and Clyde, the annual incidence rate of pneumonia was 4 cases per 100 children in the preschool-aged group, 2 cases per 100 children aged 5-9 years, and 1 case per 100 children aged 9-15 years.

Thompson et al reported that, after elderly persons, the second highest rates of influenza-associated hospitalizations in the United States were in children younger than 5 years of age.These investigators evaluated annual influenza-associated hospitalizations by hospital discharge category, discharge type, and age group.

In a randomized double-blind trial, the heptavalent pneumococcal vaccine reduced the incidence of clinically diagnosed and radiographically diagnosed pneumonia among children younger than 5 years of age by 4% and 20%, respectively.Although the overall rate of pneumonia has decreased in the United States with the use of the 7-valent vaccine, the rate of empyema and complicated pneumonia has increased. [[24](javascript:void(0);)] Following the use of the 13-valent conjugated pneumococcal polysaccharide vaccine, the overall rates of pneumonia are anticipated to drop further. The new vaccine includes serotypes that have become associated with complicated or antibiotic-resistant disease (19A and 6A, for example).

Among school-aged children and adolescents, bronchopneumonia occurs in 0.8-2% of all pertussis cases and 16-20% of hospitalized cases. *M pneumoniae* accounts for 14-35% of pneumonia hospitalizations in this age group, and mycobacterial pneumonia has recently been noted with increasing frequency in some inner-city areas, particularly among children in homeless shelters and group homes and those with household contacts.

### International statistics

Pneumonia and other lower respiratory tract infections are the leading cause of death worldwide. The WHO Child Health Epidemiology Reference Group estimated the median global incidence of clinical pneumonia to be 0.28 episodes per child-year.This corresponds to an annual incidence of 150.7 million new cases, of which 11-20 million (7-13%) are severe enough to require hospital admission. Ninety-five percent of all episodes of clinical pneumonia in young children worldwide occur in developing countries.

Approximately 150 million new cases of pneumonia occur annually among children younger than 5 years of age worldwide. This accounts for approximately 10-20 million hospitalizations.A WHO Child Health Epidemiology Reference Group publication cited the incidence of community-acquired pneumonia among children younger than 5 years of age in developed countries as approximately 0.026 episodes per child-year. In addition, a study conducted in the United Kingdom showed that 59% of deaths from pertussis are associated with pneumonia.

**GENOMIC DATA**

* Chlamydia psittaci:  
  Metagenomic analysis reconstructed two near-complete genomes of *C. psittaci* strains from lung samples of pneumonia patients, showing high similarity (>97%) to known isolates. This helps understand strain variation and potential virulence genes involved in pneumonia caused by this pathogen.
* Pseudomonas aeruginosa:  
  Real-time metagenomic sequencing has been used to rapidly identify *P. aeruginosa* in pneumonia patients. Whole-genome sequencing revealed numerous antibiotic resistance genes, such as the Mex efflux pump complex, which correlate with clinical resistance patterns.
* Klebsiella pneumoniae:  
  Genome sequencing of *K. pneumoniae* isolates from pneumonia cases worldwide shows a wide genetic diversity including multiple antimicrobial resistance (AMR) genes and virulence factors like yersiniabactin, colibactin, and aerobactin. This genomic framework aids in tracking pandemic multidrug-resistant clones and understanding their pathogenicity.
* Streptococcus pneumoniae:  
  Functional genomic approaches using CRISPR interference have identified key virulence genes driving pneumococcal pneumonia progression. This research sheds light on how *S. pneumoniae* interacts with viral infections (e.g., influenza) to worsen pneumonia severity.
* Metagenomic and 16S rRNA sequencing:  
  These techniques enable culture-independent identification of bacterial communities in pneumonia, detecting pathogens in lung tissue or bronchoalveolar lavage samples, improving diagnostic accuracy and understanding of polymicrobial infections.
* Antimicrobial resistance genes:  
  Genomic studies reveal widespread distribution of resistance genes across pneumonia pathogens, often localized on plasmids or chromosomes, highlighting the challenge of treating multidrug-resistant infections

## **PREDEFINED Questions and answers**

### **Is it possible to have pneumonia without having a fever?**

Yes, while fever is common in pneumonia, it’s possible to have pneumonia with a low fever or no fever. This is more likely if you:

* Are older than 65 or younger than 2 (especially newborns and infants).
* Have a weakened immune system.

### **Is pneumonia treated any differently in children?**

Pneumonia isn’t usually treated any differently in children. However, young children can be at higher risk for severe illness from pneumonia. They’re more likely to be hospitalized for treatment than adults.

**Doctor patient conversation**

Doctor: Hello, I understand you’ve been feeling unwell. Can you tell me about your symptoms?

Patient: Yes, I’ve had a cough for several days, and it’s been getting worse. I also have a fever and feel short of breath sometimes.

Doctor: I see. Have you noticed any chest pain, especially when you breathe deeply or cough?

Patient: Yes, there is some sharp pain on the right side of my chest.

Doctor: Based on your symptoms and your physical exam, it looks like you have pneumonia, which is an infection in your lungs.

Patient: Is this serious? What causes pneumonia?

Doctor: Pneumonia can be caused by bacteria, viruses, or sometimes fungi. It can range from mild to severe. Since you have fever, cough, chest pain, and difficulty breathing, it’s important we treat it promptly.

Patient: What treatment will I need?

Doctor: I’m going to prescribe antibiotics to target the most common bacteria that cause pneumonia. You should take them exactly as directed, even if you start feeling better before finishing the course. Also, rest, drink plenty of fluids, and use over-the-counter pain relievers like acetaminophen or ibuprofen to reduce fever and discomfort.

Patient: How long will it take for me to get better?

Doctor: Most people start feeling better within 3 to 5 days, but the cough and fatigue can last several weeks. If your symptoms worsen or you develop new symptoms like difficulty breathing, high fever, or chest pain, please come back immediately.

Patient: Should I avoid work or other activities?

Doctor: Yes, it’s best to rest and avoid strenuous activities until you feel well. Also, avoid smoking or exposure to smoke, which can irritate your lungs.

Patient: Do I need any follow-up?

Doctor: We’ll schedule a follow-up in about a week to make sure you’re improving. If you have any concerns before then, don’t hesitate to contact us.

Patient: Thank you, doctor. I appreciate the information.

Doctor: You’re welcome. Take care and get well soon.

References

[Pneumonia - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/pneumonia/diagnosis-treatment/drc-20354210)

[Pneumonia: Causes, Symptoms, Diagnosis & Treatment](https://my.clevelandclinic.org/health/diseases/4471-pneumonia#overview)

<https://www.ncbi.nlm.nih.gov/books/NBK526116/#article-27364.s10>

<https://emedicine.medscape.com/article/967822-treatment>

**CYSTIC FIBROSIS**

## Alternative Names for Cystic Fibrosis

* Mucoviscidosis  
  An older term referring to the thick, sticky mucus characteristic of the disease.
* Fibrocystic Disease of the Pancreas  
  Highlights the pancreatic involvement with fibrosis and cyst formation.
* Cystic Fibrosis of the Pancreas  
  Emphasizes the pancreatic pathology, often used historically.
* Congenital Mucoviscidosis  
  Refers to the hereditary nature of the disease and mucus abnormalities.
* CFTR-related Disorder  
  A broader term encompassing cystic fibrosis and related conditions caused by mutations in the CFTR gene with variable symptoms

**DEFINITION AND DESCRIPTION**

Cystic fibrosis (CF) is a condition passed down in families that causes damage to the lungs, digestive system and other organs in the body.

CF affects the cells that make mucus, sweat and digestive juices. These fluids, also called secretions, are usually thin and slippery to protect the body's internal tubes and ducts and make them smooth pathways. But in people with CF, a changed gene causes the secretions to become sticky and thick. The secretions plug up pathways, especially in the lungs and pancreas.

CF gets worse over time and needs daily care, but people with CF usually can attend school and work. They often have a better quality of life than people with CF had in past decades. Better screening and treatments mean that people with CF now may live into their mid- to late 50s or longer, and some are being diagnosed later in life.

**CAUSES**

In cystic fibrosis, a change in a gene causes problems with the protein that controls the movement of salt and water in and out of cells. This gene is the cystic fibrosis transmembrane conductance regulator (CFTR) gene. It affects the cells that make mucus, sweat and digestive juices. When the CFTR protein doesn't work as it should, the result is thick, sticky mucus in the respiratory, digestive and reproductive systems, as well as extra salt in sweat.

Changes in the CFTR gene that cause CF are divided into several different groups based on the problems they cause. Different groups of gene changes affect how much CFTR protein is made and how well it works.

To have cystic fibrosis, children must get one copy of the changed CFTR gene from each parent. If children get only one copy, they won't develop CF. But they will be carriers and could pass the changed gene to their own children. People who are carriers may have no symptoms of CF or a few mild symptoms.

**Risk factors**

Because cystic fibrosis is a condition passed down in families, family history is a risk factor.

CF occurs in all races, but it's most common in white people of Northern European ancestry. Because it's less common in people who are Black, Hispanic, Middle Eastern, Native American or Asian, this might lead to a much later diagnosis.

A late diagnosis may cause worse health issues. Early and effective treatment can improve your quality of life, prevent complications and help you live longer. If you're a person of color and have symptoms that could be CF, talk to your healthcare professional so that you can get tested for CF.

**Symptoms**

In the U.S., because of newborn screening, cystic fibrosis can be diagnosed within the first month of life, before symptoms develop. But people born before newborn screening became available may not be diagnosed until the symptoms of CF show up.

CF symptoms vary, depending on which organs are affected and how severe the condition is. Even in the same person, symptoms may worsen or get better at different times. Some people may not have symptoms until their teenage years or adulthood.

People who are not diagnosed until adulthood usually have milder symptoms and are more likely to have symptoms that aren't typical. These may include repeated bouts of an inflamed pancreas called pancreatitis, infertility and repeated bouts of pneumonia.

People with CF have a higher than usual level of salt in their sweat. Parents often can taste the salt when they kiss their children. Most of the other symptoms of CF affect the respiratory system and digestive system.

### **Respiratory symptoms**

In cystic fibrosis, the lungs are most commonly affected. The thick and sticky mucus that happens with CF clogs the tubes that carry air in and out of the lungs. This can cause symptoms such as:

* A cough that won't go away and brings up thick mucus.
* A squeaking sound when breathing called wheezing.
* Limited ability to do physical activity before tiring.
* Repeated lung infections.
* Irritated and swollen nasal passages or a stuffy nose.
* Repeated sinus infections.

### **Digestive symptoms**

The thick mucus caused by cystic fibrosis can block tubes that carry digestive enzymes from the pancreas to the small intestine. Without these digestive enzymes, the intestines can't completely take in and use the nutrients in food. The result is often:

* Foul-smelling, greasy stools.
* Poor weight gain and growth.
* Blocked intestines, which is more likely to happen in newborns.
* Ongoing or severe constipation. Straining often while trying to pass stool can cause part of the rectum to stick out of the anus. This is called a rectal prolapse.

### **When to see a doctor**

If you or your child has symptoms of cystic fibrosis — or if someone in your family has CF — talk with your healthcare professional about testing for the condition. Make an appointment with a doctor who has skills and experience in treating CF.

CF requires regular follow-up with your healthcare professional, at least every three months. Call your healthcare professional if you have new or worsening symptoms, such as more mucus than usual or a change in the mucus color, lack of energy, weight loss, or severe constipation.

Get medical care right away if you're coughing up blood, have chest pain or trouble breathing, or have severe stomach pain and bloating.

Call 911 or your local emergency number or go to the emergency department at a hospital if:

* You're having a hard time catching your breath or talking.
* Your lips or fingernails turn blue or gray.
* Others notice that you're not mentally alert.

**DIAGNOSIS**

To diagnose cystic fibrosis, healthcare professionals typically do a physical exam, review your symptoms and do tests.

### **Newborn screening and diagnosis**

Every state in the U.S. now routinely screens newborns for cystic fibrosis. Early diagnosis means that treatment can begin right away. Testing can include:

* **Newborn screening.** In this screening test, a healthcare professional takes a few drops of blood from the baby's heel. A lab checks the blood sample for higher levels than expected of a chemical called immunoreactive trypsinogen (IRT). IRT is released by the pancreas and may suggest CF. A newborn's IRT levels also may be high because of premature birth or a stressful delivery. For that reason, other tests may be needed to confirm a diagnosis of cystic fibrosis.
* **Sweat test.** To check if a baby has CF, a sweat test is done once the baby is at least 2 weeks old. A chemical that causes the skin to sweat is put on a small area of skin. Then the sweat is collected to test it and see if it's saltier than typical. Testing done at a care center accredited by the Cystic Fibrosis Foundation helps ensure results that can be trusted.
* **Genetic testing.** Healthcare professionals also may recommend genetic testing to look for specific changes on the gene responsible for CF. Genetic testing may be used along with IRT levels to confirm the diagnosis.

### **Testing of older children and adults**

Cystic fibrosis tests may be recommended for older children and adults who weren't screened at birth. Your healthcare professional may suggest genetic and sweat tests for CF if you have repeated bouts of an inflamed pancreas, nasal polyps, chronic sinus infections, lung infections, bronchiectasis or male infertility.

**Treatment**

There is no cure for cystic fibrosis, but treatment can ease symptoms, lessen complications and improve quality of life. Close monitoring and early, aggressive intervention is recommended to slow the worsening of CF over time. This can lead to a longer life.

Managing CF is complicated, so it's best to get treatment at a center with a multispecialty team of doctors and other healthcare professionals trained in CF. They can evaluate and treat your condition.

The goals of treatment include:

* Preventing and controlling infections that occur in the lungs.
* Removing and loosening mucus from the lungs.
* Treating and preventing intestinal blockage.
* Getting enough nutrition.

### **Medicines**

Options include:

* Medicines that target gene changes and improve how the CFTR protein works. These are called cystic fibrosis transmembrane conductance regulator (CTFR) modulators.
* Antibiotics to treat and prevent lung infections.
* Anti-inflammatory medicines to lessen swelling in the airways in the lungs.
* Mucus-thinning medicines, such as hypertonic saline, to help cough up mucus. This can improve lung function.
* Medicines breathed into the lungs called bronchodilators. These can help keep airways open by relaxing the muscles around the bronchial tubes.
* Pancreatic enzyme capsules taken by mouth to help the digestive tract take in and use nutrients.
* Stool softeners to prevent constipation or bowel obstruction.
* Acid-reducing medicines to help pancreatic enzymes work better.
* Specific medicines for diabetes or liver disease, when needed.

### **Medicines that target genes**

For those with cystic fibrosis who have certain gene changes, cystic fibrosis transmembrane conductance regulator (CFTR) modulators may help. About 90% of people with CF may be helped by using these medicines. Gene testing is needed to find out which specific gene change you have and if a CFTR modulator may work for you.

CFTR modulators are newer medicines that many experts think are a breakthrough in the treatment of CF. The medicines help the CFTR protein work better. This can make lung function better, help digestion and weight, and lessen the amount of salt in sweat.

The U.S. Food and Drug Administration (FDA) has approved these CFTR modulators for treating CF in people with specific changes in the CFTR gene:

* The newest combination medicine with elexacaftor, ivacaftor and tezacaftor (Trikafta) is approved for people age 2 years and older. Trikafta has been shown to be the most effective CFTR modulator.
* The combination medicine with ivacaftor and tezacaftor (Symdeko) is approved for people age 6 years and older.
* The combination medicine with ivacaftor and lumacaftor (Orkambi) is approved for people who are age 1 year and older.
* Ivacaftor (Kalydeco) is approved for people who are 1 month and older.

Your healthcare professional may do liver function tests and eye exams before prescribing these medicines. While taking these medicines, you'll likely need testing on a regular basis to check for side effects such as liver function changes and clouding of the eye lenses called cataracts. Ask your healthcare professional and pharmacist for information on possible side effects and what to watch for.

Keep regular follow-up appointments so your healthcare professional can monitor you while taking these medicines. Tell your healthcare professional about any side effects that you have.

### **Airway clearance techniques**

Airway clearance techniques, also called chest physical therapy, can help get rid of mucus blocking the airways. It also can help to lessen infection and inflammation in the airways. Airway clearance techniques loosen the thick mucus in the lungs, making it easier to cough up.

Airway clearing techniques are usually done several times a day. Different techniques, and often more than one method, can be used to loosen and remove mucus.

* Clapping with cupped hands on the front and back of the chest. This is a common technique.
* Special breathing and coughing activities.
* Mechanical devices, such as a tube that you blow into, and a machine that pulses air into the lungs called a vibrating vest.
* Vigorous exercise.

Your healthcare professional can give you instructions on the airway clearance techniques that are best for you and how often you should do them.

### **Pulmonary rehabilitation**

Your healthcare professional may recommend a long-term program called pulmonary rehabilitation. The program may improve your lung function and your overall well-being. Pulmonary rehabilitation is usually done on an outpatient basis and may include:

* Physical exercise that may improve your condition.
* Breathing techniques that may help loosen mucus and make breathing easier.
* Dietary counseling.
* Mental health counseling and support.
* Education about your condition.

### **Surgery and other treatments**

Options for certain conditions caused by cystic fibrosis include:

* **Nasal and sinus surgery.** Surgery can remove nasal polyps that get in the way of breathing. Sinus surgery may be done to treat repeated or long-term sinusitis.
* **Oxygen therapy.** If there isn't enough oxygen in your blood, you may need supplemental oxygen. You can get this extra oxygen to your lungs through a mask or through plastic tubing with tips that fit into your nose. These attach to an oxygen tank. Lightweight, portable units that you take with you can help you be more mobile. Oxygen therapy may prevent high blood pressure in the lungs, a condition called pulmonary hypertension.
* **Noninvasive ventilation.** Typically used while sleeping, noninvasive ventilation uses a nose or mouth mask to give positive pressure in the airway and lungs when breathing in. It's often used along with oxygen therapy. Noninvasive ventilation can increase air exchange in the lungs and lessen the work of breathing. The treatment also may help with airway clearance.
* **Feeding tube.** CF interferes with digestion, so you can't take in and use nutrients from food very well. A feeding tube delivers extra nutrition. This may be a short-term tube placed through your nose and guided to your stomach. Or the tube may be surgically placed in the stomach through a small cut in the skin on your belly. A feeding tube gives extra calories during the day or night and does not keep you from eating by mouth.
* **Bowel surgery.** If a blockage happens in the intestines, you may need surgery to remove it. If part of an intestine folds inside a nearby section of intestine, you may need surgery.
* **Lung transplant.** If you have severe breathing problems or life-threatening lung complications, or if antibiotics no longer work to treat lung infections, a lung transplant may be an option. Because bacteria line the airways in diseases such as CF that cause permanent widening of the large airways, both lungs need to be replaced.

Cystic fibrosis does not recur in transplanted lungs. But other complications linked with CF, such as sinus infections, diabetes, pancreas conditions and osteoporosis, can still happen after a lung transplant.

* **Liver transplant.** For severe CF-related liver disease, such as cirrhosis, liver transplant may be an option. In some people, a liver transplant may be done together with lung or pancreas transplants.

## **Lifestyle and home remedies**

Here are some ways you can manage cystic fibrosis and lessen complications.

### **Pay attention to nutrition and fluid intake**

Cystic fibrosis can cause poor nutrition because the enzymes needed for digestion can't reach the small intestine. This prevents food from being taken in and used by the body. People with CF may need a much higher number of calories daily than do people without the condition.

A healthy diet is important to growth and development and to support good lung function. It's also important to drink lots of fluids to help thin the mucus in your lungs. You may work with a dietitian to create a nutrition plan.

Your healthcare professional may recommend:

* Pancreatic enzyme capsules with every meal and snack.
* Medicines to lessen acid made in the stomach and help pancreatic enzymes work.
* High-calorie nutrition supplements.
* Special fat-soluble vitamins.
* Extra fiber to prevent intestinal blockage.
* Extra salt, especially during hot weather or before exercising.
* Drinking enough water, especially during hot weather.

### **Keep vaccinations up to date**

In addition to the other usual childhood vaccines, the annual flu vaccine is important if you have cystic fibrosis. So are any other vaccines your healthcare professionals recommend, such as the vaccine to prevent pneumonia and COVID-19. CF doesn't affect the immune system, but people with CF are more likely to develop complications when they get sick.

### **Exercise**

Regular exercise helps loosen mucus in your airways and makes your heart stronger. Because people with cystic fibrosis are living longer, it's important to keep your heart and blood vessels in good shape for a healthier life. Anything that gets you moving, including walking and biking, can help.

### **Stay away from smoke**

Don't smoke, and don't allow other people to smoke around you or your child. Secondhand smoke and air pollution are harmful for everyone, but especially if you have cystic fibrosis. Using electronic cigarettes, also called vaping, can worsen CF too.

### **Wash your hands**

Teach all the members of your family to wash their hands thoroughly before eating, after using the bathroom, when coming home from work or school, and after being around a sick person. If possible, stay away from people who have colds or flu. Washing your hands is the best way to protect against infection.

### **Keep medical appointments**

Along with ongoing care from your medical team:

* Keep your regular follow-up appointments.
* Take your medicines as prescribed and follow therapies as instructed.
* Talk with your healthcare professional about how to manage symptoms.
* Learn the warning signs of serious complications.

**Complications**

Complications of cystic fibrosis can affect the respiratory, digestive and reproductive systems, as well as other organs.

### **Respiratory system complications**

* **Damaged airways.** Cystic fibrosis is one of the leading causes of damaged airways, a long-term lung condition called bronchiectasis. Bronchiectasis results in widening and scarring of the airways. This makes it harder to move air in and out of the lungs and clear mucus from the airways.
* **Ongoing infections.** Thick mucus in the lungs and sinuses makes a place for bacteria and fungi to live and grow. Sinus infections, bronchitis or pneumonia are common and may happen repeatedly. Infections with bacteria that don't respond to antibiotics and are difficult to treat is common too.
* **Growths in the nose.** Because the lining inside the nose is irritated and swollen, it can develop soft, fleshy growths called nasal polyps.
* **Coughing up blood.** Bronchiectasis can occur next to blood vessels in the lungs. The combination of airway damage and infection can result in coughing up blood. Often this is only a small amount of blood, but rarely it can be life-threatening.
* **Collapsed lung.** Also called pneumothorax, this condition happens when air leaks into the space that separates the lungs from the chest wall. This causes part or all of a lung to collapse. Collapsed lung is more common in adults with CF. Collapsed lung can cause sudden chest pain and trouble breathing. People often have a bubbling feeling in the chest.
* **Respiratory failure.** Over time, CF can damage lung tissue so badly that it no longer works. Lung function usually worsens slowly over time and can become life-threatening. Respiratory failure is the most common cause of death with CF.
* **Bouts of worsening symptoms.** People with CF may experience times when respiratory symptoms are worse than usual. These are called exacerbations (eg-zas-er-bay-shuns). Symptoms may include coughing with more mucus than usual and trouble breathing. Low energy and weight loss also are common during exacerbations. Exacerbations are treated with antibiotics. Sometimes treatment can be given at home, but a stay in the hospital may be needed.

### **Digestive system complications**

* **Poor nutrition.** Thick mucus can block the tubes that carry digestive enzymes from the pancreas to the intestines. Without these enzymes, the body can't take in and use protein, fats or fat-soluble vitamins and can't get enough nutrients. This can result in delayed growth and weight loss. An inflamed pancreas, a condition called pancreatitis, is common.
* **Diabetes.** The pancreas makes insulin, which the body needs to use sugar. Cystic fibrosis raises the risk of diabetes. About 20% of teenagers and up to 50% of adults with CF develop diabetes.
* **Liver disease.** The tube that carries bile from the liver and gallbladder to the small intestine may become blocked and inflamed. This can lead to liver problems, such as jaundice, fatty liver disease and cirrhosis, and sometimes gallstones.
* **Intestinal obstruction.** Intestinal blockage can happen to people with CF at all ages. Sometimes, a condition in which a section of the intestine slides inside another nearby section of the intestine, like a collapsible telescope, also can happen.
* **Distal intestinal obstruction syndrome (DIOS).** DIOS is partial or complete blockage where the small intestine meets the large intestine. DIOS requires treatment right away.

### **Reproductive system complications**

* **Infertility in men.** Almost all men with cystic fibrosis are not fertile. The tube that connects the testicles and prostate gland, called the vas deferens, is either blocked with mucus or missing entirely. Sperm is still made in the testicles even though it can't pass into the semen made by the prostate gland. Certain fertility treatments and surgical procedures sometimes make it possible for men with CF to become biological parents.
* **Lower fertility in women.** Although women with CF may be less fertile than other women, it's possible for them to conceive and to have successful pregnancies. Still, pregnancy can worsen the symptoms of CF. Talk with your healthcare professional about the risks.

### **Other complications**

* **Thinning of the bones.** Cystic fibrosis raises the risk of developing a dangerous thinning of bones called osteoporosis. Joint pain, arthritis and muscle pain also may occur.
* **Out of balance electrolytes and dehydration.** CF causes saltier sweat, so the balance of minerals in the blood may be upset. This raises the risk for dehydration, especially with exercise or in hot weather. Symptoms of dehydration include a fast heartbeat, extreme tiredness, weakness and low blood pressure.
* **Gastroesophageal reflux disease (GERD).** Stomach acid repeatedly flows back up into the tube connecting the mouth and stomach, called the esophagus. This backwash is known as acid reflux, and it can irritate the lining of the esophagus.
* **Mental health conditions.** Having an ongoing medical condition that has no cure may cause fear, depression and anxiety.
* **Higher risk of digestive tract cancer.** The risk of cancer of the esophagus, stomach, small and large bowel, liver, and pancreas is higher in people with cystic fibrosis. Regular colorectal cancer screening should begin at age 40.

## **Outlook / Prognosis**

Yes, cystic fibrosis can be life-threatening. Lung damage — from thick mucus and frequent lung infections — is the most common cause of death.

#### **What is the life expectancy of cystic fibrosis?**

Experts predict the life expectancy of someone born with cystic fibrosis in the past few years is around 50 years old. Improvements in treatment in recent years have increased this from a few years ago, when life expectancy was between 30 and 40 years old.

People with atypical cystic fibrosis tend to have longer life expectancies than those with classic CF.

### **What can I expect if I have cystic fibrosis?**

There’s no cure for CF. You or your child will need lifelong treatments to manage it. This includes treating infections, maintaining nutrition and seeing a CF specialist frequently. But new treatment methods help children who have CF live well into adulthood and have a better quality of life.

Treatments work best when CF is diagnosed early, which is why newborn screening is so important. The addition of CFTR modulators at a young age may improve long-term health and increase life expectancy even more in the future.

**Prevention**

If you or your partner have close relatives with cystic fibrosis, you both may choose to have genetic testing before having children. Testing done in a lab on a sample of blood can help find out your risk of having a child with CF.

If you're already pregnant and the genetic test shows that your baby may be at risk of CF, your healthcare professional can do other tests on your unborn child.

Genetic testing isn't for everyone. Before you decide to be tested, talk with a genetic counselor about the mental health impact the test results might have.

## **Differential Diagnoses**

* Acute Sinusitis
* Bronchiolitis
* Nutritional Considerations in Failure to Thrive
* Pediatric Aspergillosis
* Pediatric Asthma
* Pediatric Bronchiectasis
* Pediatric Celiac Disease (Sprue)
* Primary Ciliary Dyskinesia
* Short Stature

## **Epidemiology**

Cystic fibrosis is an autosomal-recessive disease. Its estimated heterozygote frequency in White people is up to 1 in 20. Each offspring of 2 heterozygote parents has a 25% chance of developing cystic fibrosis.

Cystic fibrosis is the most common lethal hereditary disease in the White population. In the United States, the prevalence is as follows:

* Whites of northern European origin - 1 case per 3200-3500 population
* Hispanics - 1 case per 9200-9500 population
* African Americans - 1 case per 15,000-17,000 population
* Asian Americans - 1 case per 31,000 population

The worldwide incidence varies from 1 per 377 live births in parts of England to 1 per 90,000 Asian live births in Hawaii. The higher frequency in Asian American or African American populations compared with native Asians or Africans reflects White admixture.

### Race demographics

The distribution of *CFTR* mutations varies according to the background of patients; for example, ΔF508 is the most common mutation found in the White population of northern European origin. Variability in clinical features between people of different races with same genotype has not been reported.

Clinical manifestations are similar in Black and White populations, except that a poorer nutritional status is observed in Black patients. Black patients with cystic fibrosis are younger at diagnosis and have poorer nutritional status and pulmonary function than White patients with cystic fibrosis. Whether this is genetic or due to socioeconomic factors is unclear; low socioeconomic status is associated with significantly worse pulmonary outcomes in patients with cystic fibrosis.

### Sex demographics

Compared with males, females with cystic fibrosis have greater deterioration of pulmonary function with increasing age and younger mean age at death. Although it has been suggested that the increase in hormone secretion with puberty in females may interfere with the defense mechanisms of the immune system, thereby promoting progressive pulmonary involvement, the immune system in patients with cystic fibrosis is fundamentally intact.

## **Genotyping**

Genotype testing is recommended for individuals with a positive family history and for couples planning a pregnancy. It is not necessarily indicated for the general population.

More than 1893 CF mutations have been identified.In the commercially available CF gene sequencing method, the entire coding region, splice junction sites, and promoter region of the *CFTR* gene are amplified from genomic DNA by polymerase chain reaction (PCR) and then subjected to nucleotide sequence analysis on an automated capillary DNA sequencer.

A finding of 2 *CFTR* mutations in association with clinical symptoms is diagnostic. This test can detect more than 98% of disease-causing mutations in Whites; the detection rate is lower in Black, Hispanic, and Asian populations. Therefore, failure to find 2 abnormal genes does not exclude the disease.

In 2013, the US Food and Drug Administration (FDA) approved 4 next-generation gene sequencing devices for clinical use in CF. Two of the devices are used to screen and diagnose CF by detecting DNA changes in the CF transmembrane conductance regulator (*CFTR*) gene: the Illumina MiSeqDx Cystic Fibrosis 139-Variant Assay, which checks specific points in the patient's *CFTR* gene sequence to detect known variants in the gene, and the Illumina MiSeqDx Cystic Fibrosis Clinical Sequencing Assay, which sequences a large portion of the *CFTR* gene to detect any difference in the *CFTR* gene compared with a reference *CFTR* gene.

The other 2 FDA-approved devices are the Illumina MiSeqDx instrument platform, which analyzes the genes, and the Illumina Universal Kit reagents, which isolate and create copies of the genes of interest from patient blood samples. These 2 devices comprise the first FDA-regulated test system that allows laboratories to develop and validate sequencing of any part of a patient’s genome.

### **PREDEFINED QUESTION AND ANSWERS**

### **Why can’t people with CF touch each other?**

Healthcare professionals usually recommend that people with cystic fibrosis aren’t in close contact with one another. This is because people with CF are more likely to get infections that other people fight off easily. They’re more likely to pass the germs on to others with CF (who also can’t fight them off easily). People with CF also should avoid anyone who’s sick.

## 1. What are my treatment options?

* Medications:
  + CFTR modulators (e.g., ivacaftor, lumacaftor/ivacaftor) target the defective CFTR protein to improve lung function.
  + Antibiotics to treat and prevent lung infections.
  + Anti-inflammatory drugs to reduce airway swelling.
  + Mucus-thinning agents like hypertonic saline and dornase alfa to help clear mucus.
  + Bronchodilators to open airways.
  + Pancreatic enzyme supplements to aid digestion and nutrient absorption.
  + Acid-reducing medications to improve enzyme effectiveness.
  + Other medicines for complications like CF-related diabetes or liver disease.
* Airway clearance therapies: Chest physiotherapy and devices to loosen and remove mucus.
* Nutritional support: High-calorie, high-fat diet with vitamin supplements; sometimes tube feeding if needed.
* Surgery: Rarely, for complications like intestinal blockage or lung transplantation in advanced disease.
* Treatment is best managed at specialized CF centers with a multidisciplinary team.

## 2. What’s a healthy eating plan I can follow?

* A high-calorie, high-protein, and high-fat diet is recommended to meet increased energy needs.
* Eat frequent meals and snacks to maintain weight and growth.
* Take pancreatic enzyme supplements with all meals and snacks to aid digestion.
* Include fat-soluble vitamins (A, D, E, K) supplements as advised by your healthcare team.
* Stay well hydrated.
* Work with a CF dietitian to tailor nutrition plans to your needs.

## 3. What can I do to manage my symptoms?

* Perform daily airway clearance therapies to loosen mucus and improve breathing.
* Use prescribed inhaled medications regularly (mucolytics, bronchodilators, antibiotics).
* Avoid lung irritants like smoke and pollution.
* Get regular exercise to improve lung function and overall health.
* Monitor symptoms closely and report any worsening promptly.
* Stay up to date with vaccinations, including flu and pneumonia vaccines.

## 4. What signs of infection should I look out for?

* Increased cough or change in cough character (more frequent, productive, or bloody).
* Increased shortness of breath or wheezing.
* Fever or chills.
* Increased fatigue or decreased exercise tolerance.
* Changes in sputum color or amount.
* Chest pain or discomfort.
* Loss of appetite or weight loss.

## 5. When should I follow up with you?

* Regular follow-up visits are essential, often every 3 months or as advised by your CF care team.
* Immediately if you notice worsening respiratory symptoms, fever, or other signs of infection.
* Before starting any new medications or therapies.
* For routine monitoring of lung function, nutrition, and complications.

## 6. What symptoms should I go to the ER for?

* Severe difficulty breathing or shortness of breath at rest.
* Persistent high fever not responding to medications.
* Chest pain or sudden worsening of cough.
* Blue or gray discoloration of lips, face, or fingertips (signs of low oxygen).
* Severe dehydration (little or no urine output, dizziness).
* Confusion or extreme lethargy.

## 7. Should other family members get tested?

* Yes, family members can be carriers of CFTR mutations even if they do not have symptoms.
* Genetic counseling and testing are recommended for siblings and reproductive partners to assess carrier status and reproductive risks.
* Early diagnosis in family members can improve management and outcomes.

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello! I see you’ve been diagnosed with cystic fibrosis. How are you feeling about this news?

Patient: It’s a lot to take in. I’m worried about what this means for my health and daily life.

Doctor: That’s completely understandable. CF is a complex condition, but with current treatments and a good care team, many people live full lives. Our goal is to manage symptoms, prevent infections, and maintain your lung and nutritional health.

Patient: What kind of treatments will I need?

Doctor: You’ll likely use several therapies daily, including airway clearance techniques to help clear mucus, inhaled medications like mucolytics and bronchodilators, and pancreatic enzyme supplements to aid digestion. There are also newer medications called CFTR modulators that target the underlying genetic defect, which can improve lung function.

Patient: That sounds like a lot. How often will I need to see you or the care team?

Doctor: Regular visits every 3 months are important to monitor lung function, nutrition, and infections. We’ll do breathing tests and sometimes imaging to track your lungs. You’ll also have access to a multidisciplinary team including respiratory therapists, dietitians, and social workers.

Patient: What can I do day-to-day to manage my symptoms?

Doctor: Daily airway clearance, taking your medications as prescribed, staying active with exercise, and maintaining a high-calorie, nutritious diet are key. Also, avoid lung irritants like smoke and stay up to date with vaccinations.

Patient: What signs should I watch for that mean I need to come in sooner?

Doctor: If you notice increased cough, more mucus or a change in its color, shortness of breath, fever, chest pain, or fatigue, contact us promptly. These may indicate an infection or exacerbation.

Patient: Should my family members get tested?

Doctor: Yes, CF is genetic. Family members can be carriers even if they don’t have symptoms. Genetic counseling and testing can help assess risks for relatives and future children.

Patient: How do you usually communicate with patients about CF? It’s a lot to understand.

Doctor: We tailor communication to your needs, provide written materials, and encourage questions. We want you to feel supported and involved in your care decisions. You can always reach out between visits if you have concerns.

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## **Bronchopulmonary Dysplasia (BPD)**

## **Alternative Names for Bronchopulmonary Dysplasia**

* Chronic Lung Disease of Prematurity  
  Emphasizes that BPD primarily affects premature infants with underdeveloped lungs.
* Neonatal Chronic Lung Disease  
  Highlights the chronic nature of lung injury starting in the neonatal period.
* Infantile Chronic Lung Disease  
  Another term referring to chronic lung issues in infants, often synonymous with BPD.
* Oxygen-Dependent Lung Disease of Prematurity  
  Describes the need for prolonged oxygen therapy in affected infants.
* Pulmonary Dysplasia of Prematurity  
  A less common variant term focusing on lung development abnormalities.

**DEFINITION AND DESCRIPTION**

Bronchopulmonary dysplasia (BPD) is a lung disorder that can affect preterm babies. When a baby is born early, their lungs are underdeveloped. Because of this, they may need long-term oxygen therapy or mechanical ventilation to help them breathe.

While these therapies can save your infant’s life, they can also overstretch the tiny air sacs (alveoli) in their lung tissue. This can damage your infant’s lungs and airways (bronchi) over time, causing tissue destruction (dysplasia).

The earlier your baby is born, the greater their risk of developing bronchopulmonary dysplasia. The severity of the condition can vary. While most babies recover from BPD, some may have a lifetime of breathing difficulties and other complications.

### **Bronchopulmonary dysplasia causes**

When your baby is born early, they sometimes need help breathing because their lungs are underdeveloped. This means your baby may need to be on a ventilator. The ventilator provides oxygen and pressure to help your baby’s lungs expand and support their breathing.

Your baby’s lungs are vulnerable right after birth, and the delivery of oxygen and pressure can overstretch their fragile air sacs (alveoli). This can lead to inflammation and damage to their lung tissue over time.

#### **Risk factors**

Newborns who are especially at risk of developing bronchopulmonary dysplasia include:

* Babies born more than 10 weeks early
* Babies who weigh less than 2 pounds at birth
* Babies with underdeveloped lungs or breathing problems

It’s rare for babies born after 32 weeks to develop BPD.

### **Signs and symptoms of bronchopulmonary dysplasia**

Signs and symptoms of bronchopulmonary dysplasia (BPD) vary widely but can include:

* A blue tone of a white baby’s skin and lips, or a yellow-gray, gray or white tone in newborns of color (cyanosis)
* Difficulty breathing (respiratory distress)
* Low oxygen levels in your baby’s blood
* Pauses in breathing (apnea)
* Rapid breathing (tachypnea)
* Wheezing

### **When should my baby see their healthcare provider?**

After treatment, your baby should see their healthcare provider if they have:

* Chronic cough
* Chronic snoring
* Difficulty eating
* Flaring nostrils during each breath
* Heavy, quick breathing
* Symptoms of a viral illness, like fever, sneezing or coughing
* Wheezing

Visit the emergency room or call 911 immediately if your baby stops breathing, has trouble breathing or if their skin or lips become discolored.

## **Diagnosis and Tests**

If your baby is born preterm and needs help breathing for the first 28 days of their life, their healthcare provider will likely diagnose them with BPD. There are no specific tests to diagnose the condition. But some tests that can suggest and help manage the diagnosis include:

* Blood tests to identify how much oxygen is in your baby’s blood
* Imaging tests, like a chest X-ray, to look at your baby’s lungs

**Management and Treatment**

There’s no cure for bronchopulmonary dysplasia. But treatment reduces the risk of further lung damage and helps your baby’s lungs grow and heal.

The goal of bronchopulmonary dysplasia treatment is to wean your baby off supplemental oxygen as soon as possible. Treatment improves your baby’s lung function and their ability to breathe on their own. Treatment may include:

* **Nutrition.** Increasing the amount of calories your baby takes in can help their lungs grow.
* **Diuretics.** Diuretics help reduce the amount of fluid in and around your baby’s lungs.
* **Bronchodilators.** Bronchodilators help relax the muscles around your baby’s airways, making breathing easier.
* **Corticosteroids.** Corticosteroids help reduce or prevent inflammation in and around your baby’s lungs.
* **Nirsevimab.** Nirsevimab (or palivizumab) helps prevent viral infections like RSV in babies.
* **Nasal continuous positive airway pressure (nCPAP).** An nCPAP machine gently pushes air into your baby’s lungs through special prongs placed in their nose.
* **Tracheostomy.** If your baby has a severe case of BPD, their provider may surgically insert this tube into your baby’s windpipe to help them breathe.

After treatment, your baby’s health will gradually improve over several months. During this time, their lungs will continue to heal and grow, with the goal of breathing on their own.

## **TREATMENT DRUGS AND THEIR SIDE EFFECTS**

## 1. Diuretics

* Purpose: Reduce fluid accumulation in and around the alveoli to improve lung function.
* Common drugs: Furosemide (loop diuretic), Chlorothiazide, Hydrochlorothiazide, Spironolactone.
* Administration: Usually given orally 1-4 times daily.
* Side effects: Electrolyte imbalances (low potassium, sodium), dehydration, kidney dysfunction, increased urination.

## 2. Bronchodilators

* Purpose: Relax airway muscles to widen air passages and ease breathing.
* Common drugs: Albuterol (salbutamol), Levalbuterol, Ipratropium bromide.
* Administration: Usually inhaled via nebulizer or inhaler with spacer.
* Side effects: Tremors, increased heart rate, nervousness, potential worsening of tracheomalacia.

## 3. Corticosteroids

* Purpose: Reduce lung inflammation, swelling, and mucus production.
* Common drugs: Dexamethasone, Hydrocortisone (systemic); Budesonide, Fluticasone (inhaled).
* Administration: Systemic (IV or oral) or inhaled aerosols.
* Side effects: Growth suppression, immune suppression, increased infection risk, hypertension, hyperglycemia, potential neurodevelopmental effects (especially with systemic steroids).

## 4. Surfactant Therapy

* Purpose: Improve lung compliance and reduce surface tension in alveoli.
* Common drugs: Poractant alfa (Curosurf), Calfactant (Infasurf), Beractant (Survanta).
* Administration: Intratracheal instillation, mainly in preterm infants with respiratory distress syndrome.
* Side effects: Rare but can include transient bradycardia or oxygen desaturation during administration.

## 5. Caffeine Citrate

* Purpose: Stimulates respiratory drive, prevents apnea of prematurity, reduces inflammation.
* Side effects: Irritability, tachycardia, feeding intolerance.

## 6. Pulmonary Vasodilators (for BPD-associated pulmonary hypertension)

* Common drugs: Sildenafil, Inhaled nitric oxide (iNO), Bosentan.
* Side effects: Hypotension, headache, liver enzyme abnormalities (bosentan), bleeding risk (iNO).

## 7. Macrolide Antibiotics (Research Use)

* Purpose: Anti-inflammatory effects and activity against Ureaplasma species.
* Common drugs: Azithromycin, Clarithromycin.
* Side effects: Gastrointestinal upset, potential antibiotic resistance concerns.

### **Complications of bronchopulmonary dysplasia**

Most infants recover from BPD by the time they’re 5 years old. But they’re at risk of developing other health conditions, including:

* Feeding difficulties
* Gastroesophageal reflux disease (GERD)
* Learning disabilities and disorders
* Neurological disorders
* Problems with hearing or vision
* Pulmonary hypertension

Some children and adults who had the condition as newborns are at risk of developing certain lung complications. These include:

* Asthma
* Bronchitis
* Obstructive sleep apnea
* Pneumonia
* Reactive airway disease
* Severe respiratory syncytial virus (RSV) infections

## **Outlook / Prognosis**

Your baby’s lungs will continue to develop after they leave the hospital. Their lungs will still be vulnerable, and it’s important to keep them as healthy as possible. You can do this by:

* Avoiding people who have respiratory tract infections, including RSV
* Getting all family members vaccinated against the flu
* Keeping your baby away from secondhand smoke and fumes (especially from cigarettes)

Your baby may experience feeding difficulties, which could lead to growth faltering. Taking your baby to all scheduled appointments is very important to address these issues sooner.

Because babies with bronchopulmonary dysplasia are born early, they can experience a delay in their developmental milestones. Most babies will catch up with various therapies, including physical therapy, speech therapy and occupational therapy. Severe cases of bronchopulmonary dysplasia can be life-threatening. But most babies survive, with their health gradually improving as they get older and gain weight.

**Prevention**

If you’re pregnant, it’s important to keep yourself healthy and take steps to avoid preterm labor. The risk of having a baby with BPD significantly reduces if the fetus’s lungs have enough time to develop. You can reduce your risk of having your baby preterm by:

* Avoiding tobacco, recreational drugs and alcohol while you’re pregnant
* Eating healthy and nutritious foods
* Getting regular prenatal check-ups throughout your pregnancy
* Minimizing stress

## **Diagnostic Considerations**

Consider airway injury, nosocomial infection, and subglottic stenosis in the diagnosis of bronchopulmonary dysplasia (BPD).

Associated confounding problems in infants with BPD can be severe, and delayed diagnosis can be catastrophic. For example, if an infant with bronchopulmonary dysplasia and superimposed sepsis is treated with systemic corticosteroids, the infant may have serious complications or death. When steroids (hydrocortisone, dexamethasone) are administered with indomethacin, the risk of spontaneous intestinal perforation is significantly increased.

Careful discussions between parents and caregivers should be undertaken before corticosteroids are given to high-risk infants.

Brain growth in preterm infants may also be adversely affected by severe BPD. In a study that used magnetic resonance imaging to assess the segmented brain volumes of 63 preterm infants (42 with severe BPD; 21 without severe BPD) at term equivalent age and at 18 months of corrected age and compared their brain volume and brain growth velocity, Iwanaga et al found no significant difference in brain volumes at term equivalent age between the severe-BPD and non-severe BPD infants. However, at 18 months of corrected age, the infants with severe BPD had significantly smaller brain volumes of the total brain and cerebral white matter, as well as lower brain growth velocities from term equivalent age to 18 months of corrected age in the total brain, cerebral cortex, and cerebral white matter.

## **Differential Diagnoses**

* Patent Ductus Arteriosus (PDA)
* Pediatric Hypertension
* Pediatric Pneumonia
* Pulmonary Atelectasis
* Tracheomalacia

## **Epidemiology**

### United States data

Infants with severe bronchopulmonary dysplasia are often extremely immature and have very low birth weight, although term infants with severe respiratory failure are also at increased risk. Bronchopulmonary dysplasia is uncommon in infants with a birth weight of more than 1250 g and in infants who were born at more than 30 weeks' gestation. Overall, about one fourth of infants who weigh less than 1500 g are diagnosed with bronchopulmonary dysplasia. About half of infants born at less than 30 weeks' gestation have bronchopulmonary dysplasia.

Antenatal gluco corticosteroids, early surfactant therapy, and gentle modalities of ventilation have minimized the severity of lung injury, particularly in relatively mature infants. However, improved survival has increased the prevalence of bronchopulmonary dysplasia, especially in small infants who may have been exposed to in utero infection (e.g., chorioamnionitis).

Several trials of surfactants revealed that incidences of bronchopulmonary dysplasia widely vary, from 17% to 57%. No substantial difference between placebo-treated and surfactant-treated survivors has been reported. Kresch and Clive performed a meta-analysis of surfactant-replacement therapy for infants weighing less than 2 kg.Infants receiving modified natural surfactant had improved survival without bronchopulmonary dysplasia. Van Marter and associates described the wide variation in the prevalence of bronchopulmonary dysplasia in different NICUs using various ventilatory strategies. This variation has also been noted among sites in the Vermont Oxford Network (VON) and in the NICHD research network, suggesting that differences in patient populations and clinical practices may directly affect outcomes.

### International data

Studies like those in the United States have been conducted to compare rates of bronchopulmonary dysplasia in different NICUs in Europe. Results have been similar despite the relatively homogeneous population.

### Race-, sex-, and age-related demographics

Compared with white infants, African American infants generally have a lower incidence of severe bronchopulmonary dysplasia, although the combined rate of bronchopulmonary dysplasia and death is often similar in persons of different races.

Male infants with bronchopulmonary dysplasia tend to have more severe disease and worse neurodevelopmental outcome.

Bronchopulmonary dysplasia is most common in the most immature neonates born at 22-30 weeks' gestational age. These patients frequently weigh less than 1000 g at birth.

**PREDEFINED Q AND A**

## 1. What is Bronchopulmonary Dysplasia (BPD)?

BPD is a chronic lung condition that mostly affects premature infants who needed prolonged oxygen therapy or mechanical ventilation. It causes scarring and inflammation in the lungs, leading to breathing difficulties that can persist beyond infancy.

## 2. How long does BPD last?

Symptoms of BPD are usually most severe in infancy and early childhood. Many children see improvement by age 2 to 3 years, and treatment often ends by age 5. However, lung development may remain abnormal, and some may experience lung problems or asthma-like symptoms into adulthood.

## 3. What treatments are available for BPD?

There is no cure, but treatment focuses on supporting lung function and minimizing further damage. This includes:

* Oxygen therapy at home as needed
* Diuretics to reduce lung fluid
* Bronchodilators and inhaled steroids to open airways and reduce inflammation
* Nutritional support and feeding assistance
* Regular monitoring by pediatric lung specialists
* Preventive vaccinations to reduce infections.

## 4. What care is needed after hospital discharge?

Babies with BPD often require ongoing oxygen therapy and medications at home. Caregivers should monitor breathing, avoid exposure to smoke and infections, and follow up closely with healthcare providers. Feeding difficulties and growth delays may also need management.

## 5. What complications can occur with BPD?

Complications may include:

* Apnea (pauses in breathing)
* Feeding difficulties requiring tube feeding
* Gastroesophageal reflux disease (GERD)
* Pulmonary hypertension (high blood pressure in lung vessels)
* Neurologic, vision, or hearing problems in severe cases
* Increased susceptibility to respiratory infections and hospitalizations.

## 6. How can I support my child’s lung health?

* Ensure timely vaccinations, including flu and RSV prevention
* Practice good hand hygiene to reduce infections
* Avoid exposure to tobacco smoke and other lung irritants
* Follow prescribed therapies and attend regular check-ups with specialists.

## 7. When should I seek medical help for my child?

Seek prompt care if your child has:

* Increased difficulty breathing or rapid breathing
* Persistent or worsening wheezing or cough
* High fever or signs of infection
* Poor feeding or dehydration
* Blue discoloration of lips or face.

## 8. Can BPD be prevented?

While not all cases can be prevented, risks are reduced by:

* Preventing premature birth through healthy pregnancy care
* Administering antenatal steroids (like betamethasone) to help lung maturity if preterm birth is expected
* Minimizing lung injury from ventilation and oxygen therapy in the NICU.

## 9. What support is available for families?

Many hospitals offer specialized BPD programs with multidisciplinary teams to support families. Support groups and resources like the American Lung Association’s Lung Help Line can provide guidance and emotional support

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I want to talk with you about your baby’s lung condition called bronchopulmonary dysplasia, or BPD. This is a chronic lung disease that affects many premature infants who require oxygen or ventilation support.

Parent: What exactly is BPD? Is it serious?

Doctor: BPD happens because the lungs are still developing when babies are born too early. The oxygen and ventilation they need to survive can sometimes cause inflammation and scarring in the lungs. It can make breathing harder and increase the risk of infections. It is serious, but with good care, many babies improve over time.

Parent: What kind of treatments will my baby need?

Doctor: Treatment focuses on supporting your baby’s breathing and nutrition. This may include oxygen therapy, medications like diuretics and bronchodilators to help the lungs work better, and careful nutritional support to promote growth. We also monitor closely to prevent and treat infections. Sometimes babies need feeding tubes if they have trouble eating.

Parent: Will my baby need to stay in the hospital a long time?

Doctor: It depends on the severity. Some babies need prolonged hospital stays, but many go home with oxygen support and close outpatient follow-up. We have specialized clinics where experts monitor lung health and development after discharge.

Parent: What signs should I watch for that mean my baby is having trouble?

Doctor: Watch for increased work of breathing like fast breathing, chest retractions, grunting, or if your baby becomes more tired or feeds poorly. Also, any fever or change in color, such as bluish lips, should prompt immediate medical attention.

Parent: Can my baby fully recover from BPD?

Doctor: Many infants improve significantly as their lungs grow and mature, especially in the first few years. However, some may have ongoing lung issues like asthma or increased risk of respiratory infections. That’s why regular follow-up is important.

Parent: Is there anything I can do to help at home?

Doctor: Yes, avoid exposure to smoke or sick contacts, keep up with vaccinations including flu and RSV prevention, and follow the treatment plan closely. We’re here to support you every step of the way.

Parent: Thank you. It’s reassuring to know there’s a plan.

Doctor: You’re doing a great job caring for your baby. Please call us anytime if you have concerns or questions.

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**CHRONIC RESPIRATORY FAILURE**

## **Alternative Names for Chronic Respiratory Failure**

* Chronic Respiratory Insufficiency  
  Emphasizes the long-term inability of the respiratory system to maintain adequate gas exchange.
* Chronic Hypoxemic Respiratory Failure  
  Refers specifically to chronic respiratory failure characterized by low oxygen levels (hypoxemia).
* Chronic Hypercapnic Respiratory Failure  
  Describes chronic respiratory failure with elevated carbon dioxide levels (hypercapnia).
* Type 1 Respiratory Failure (Chronic)  
  Primarily hypoxemic respiratory failure without hypercapnia.
* Type 2 Respiratory Failure (Chronic)  
  Respiratory failure with both hypoxemia and hypercapnia.
* Chronic Ventilatory Failure  
  Highlights failure of the ventilatory pump function over time.
* End-Stage Respiratory Failure  
  Used in advanced disease when respiratory failure is irreversible and severe.

**DEFINITION AND DESCRIPTION**

As chronic respiratory failure progresses, you may have symptoms such as difficulty breathing. It’s serious, and the underlying cause doesn’t always have a cure. However, treatments are available to help you manage the symptoms.

**Respiratory failure**

Respiratory failure is a condition where you don’t have enough oxygen in the tissues in your body (hypoxia) or when you have too much carbon dioxide in your blood (hypercapnia). You might also hear people use the term “acute hypoxemic respiratory failure (AHRF)” to describe it.

**Respiratory failure is often a medical emergency. Call 911 or seek medical attention right away if you think you’re experiencing respiratory failure.**

#### **How respiration works**

You can think of respiration as passengers traveling from the air to your tissues. When you breathe in, oxygen molecules travel to your lungs — the passengers arriving at the airport. The oxygen passengers arrive at the “airport gates” — small air sacs around your lungs called alveoli — and are picked up by your blood. They travel through your blood to their destination in your tissues, like your organs and muscles. You need oxygen to reach its destination to stay alive.

After your blood cells drop off oxygen in your tissues, they have room to pick up carbon dioxide. Your body doesn’t need carbon dioxide (it’s a waste product). If too much of it builds up, there isn’t room in your blood’s transportation system to deliver oxygen. Your blood circulates through your body, back to your lungs, where it drops off carbon dioxide. When you breathe out, you get rid of the unnecessary waste to make room for more oxygen.

If any parts of this system fail, you won’t have enough oxygen to keep your tissues healthy.

### **Types of respiratory failure**

Respiratory failure can come on suddenly (acute) or over time (chronic). There are two common types: hypoxemic respiratory failure (type 1) and hypercapnic respiratory failure (type 2). Other types include perioperative (related to surgery) respiratory failure (type 3) and respiratory failure due to shock (type 4).

#### **Hypoxemic respiratory failure**

Hypoxemic respiratory failure happens when you don’t have enough oxygen in your blood (hypoxemia). Heart and lung conditions are the most common causes. Hypoxemic respiratory failure is also called hypoxic respiratory failure.

**Hypercapnic respiratory failure**

Hypercapnic respiratory failure happens when you have too much carbon dioxide (CO2) in your blood. If your body can’t get rid of carbon dioxide, a waste product, there isn’t room for your blood cells to carry oxygen.

The most common causes of hypercapnic respiratory failure include heart, lung, muscle and neurological (brain and spinal cord) conditions. Certain medications can also cause it. Hypercapnic respiratory failure is also called hypercarbic respiratory failure.

#### **Perioperative respiratory failure**

Perioperative respiratory failure can happen when you have surgery. Anesthesia (medication that keeps you asleep) can keep you from breathing properly. Sometimes, air sacs in your lungs can collapse (atelectasis) and keep oxygen from getting into your blood.

##### **Respiratory failure due to shock**

Shock is a condition that causes low blood pressure, fluid in your lungs (pulmonary edema) and other issues that can lead to respiratory failure. [Sepsis](https://my.clevelandclinic.org/health/diseases/12361-sepsis), cardiac events (like a heart attack) and blood loss can cause shock.

### **causes respiratory failure**

Respiratory failure happens when something keeps your body from getting oxygen into your blood or getting carbon dioxide out of your blood. This can be due to:

* Too little airflow or blood flow to your lungs.
* Blockages, scarring or fluid in your lungs.
* Inability to breathe properly or deeply enough. Conditions that affect your lungs, issues with the nerves or muscles you use to breathe, or injuries to your chest can cause this.
* Abnormalities in the way blood flows through your heart.

#### **Risk factors for respiratory failure**

**Risk factors for respiratory failure include:**

* Lung conditions and diseases. This includes acute respiratory distress syndrome (ARDS), chronic obstructive pulmonary disease (COPD), pneumonia, asthma, cystic fibrosis, pulmonary edema, pulmonary embolism and pulmonary fibrosis.
* Heart or circulatory (blood flow) conditions and diseases. This includes heart attack, congenital heart disease, heart failure and shock.
* Conditions that affect the nerves and muscles that help you breathe. This includes muscular dystrophy, amyotrophic lateral sclerosis (ALS), severe scoliosis and Guillain-Barré syndrome.
* Chest, spinal cord or brain injuries (including stroke).
* Smoking or exposure to other lung irritants. This includes chemical fumes, dust and asbestos.
* Surgery that requires sedation or anesthesia.
* Drug use or excessive alcohol consumption.
* Age. Newborn babies (especially premature infants) and adults over 65 are at higher risk for respiratory failure.

### **symptoms of respiratory failure**

Symptoms of respiratory failure depend on the cause. Symptoms may include:

* Shortness of breath or feeling like you can’t get enough air (dyspnea).
* Rapid breathing (tachypnea).
* Extreme tiredness (fatigue).
* Fast heart rate (feeling like your heart’s racing) or heart palpitations.
* Spitting or coughing blood or bloody mucus (hemoptysis).
* Excessive sweating.
* Restlessness.
* Pale skin.
* Bluish skin, lips or nails (cyanosis).
* Headaches.
* Blurred vision.
* Agitation, confusion or being unable to think straight.
* Behavioral changes, not acting like yourself.

### **When should I see a healthcare provider?**

Talk to a healthcare provider if you have a chronic condition that puts you at risk for respiratory failure. They can tell you what signs and symptoms of respiratory failure to look out for and how to manage your condition.

Go to the nearest emergency room or call 911 if you have symptoms of respiratory failure. It can be fatal if not treated quickly.

## **causes chronic respiratory failure**

Certain lung diseases can cause chronic respiratory failure. Conditions that affect how the brain, muscles, bones, or surrounding tissues support breathing can also cause chronic respiratory failure.

Diseases and conditions that commonly lead to chronic respiratory failure include:

* chronic obstructive pulmonary disease (COPD)
* emphysema
* complicated pneumonia
* obesity hypoventilation syndrome, a type of breathing disorder that’s associated with low blood oxygen levels
* cystic fibrosis
* injury to the chest
* spinal cord injuries
* stroke
* muscular dystrophy
* amyotrophic lateral sclerosis (ALS), better known as Lou Gehrig’s disease
* drug or alcohol misuse

Smoking is another possible cause.

## **Diagnosis and Tests**

A doctor will be able to diagnose chronic respiratory failure by performing a physical exam and asking about your symptoms and medical history. They may also run certain tests to confirm the diagnosis.

Often an ongoing illness or significant injury has occurred prior to the development of chronic respiratory failure.

### **Physical examination**

During a physical exam, the doctor will use a stethoscope to listen for unusual sounds in your lungs and heart.

### **Medical history**

The doctor will ask you about lung diseases or conditions you currently have or have had in the past.

### **Pulse oximetry test**

Pulse oximetry is a simple and painless test that evaluates how well oxygen is being sent to various parts of the body.

The doctor will place a small sensor on the tip of your finger or ear lobe to determine whether you’re getting enough oxygen.

In healthy people, the normal oxygen saturation range will be between 95% and 100%, according to Stat Pearls. The American Thoracic Society states that any percentage under 89% indicates an abnormally low oxygen level.

**Arterial blood gas test**

An arterial blood gas test is a safe and easy procedure that measures the amount of carbon dioxide and oxygen in your blood. It also measures the pH, or acidity level, of your blood.

The doctor will draw blood from an artery in your wrist. They’ll then send the blood to a lab for analysis. The results of this test indicate carbon dioxide and oxygen levels in your blood as well as its overall chemistry.

### **Imaging tests**

The doctor can use a chest X-ray or CT scan to obtain a better view of your lungs.

### **Bronchoscopy**

A bronchoscope is a thin, flexible lighted instrument that can be inserted into your lungs and airways. Doctors can use bronchoscopy to get a closer look at the lung passages as well as to take samples of airway and lung tissue.

## **Management and Treatment**

Although acute respiratory failure is a medical emergency that must be treated in a hospital, chronic respiratory failure can be managed at home, depending on its cause.

In severe cases, medical professionals can help you manage the condition in a long-term healthcare center.

Treatment options typically include:

* addressing the underlying cause of your respiratory failure (perhaps with medications)
* removing excess carbon dioxide from the blood
* increasing oxygen levels in the blood

Some treatments are detailed below.

### **Oxygen therapy**

You may receive oxygen therapy if you don’t have enough oxygen in your blood. Oxygen therapy raises your oxygen levels by increasing the amount of oxygen you inhale.

Oxygen is distributed from a tank through a tube. The gas enters your lungs through a face mask, nasal tubes, or one larger tube directly inserted into your windpipe. There are small, portable oxygen machines that you can carry in a shoulder bag.

### **Tracheostomy**

In severe cases of chronic respiratory failure, you may need a tracheostomy. During this procedure, a surgeon places a tube in your windpipe so you can breathe more easily.

The tube is inserted through a cut in the front of your neck where your windpipe is located. This tube can be temporary or permanent.

### **Mechanical ventilation**

If chronic respiratory failure doesn’t improve with other treatments, the doctor may put you on a ventilator or breathing machine.

This machine pumps oxygen through a tube that’s inserted into your mouth or nose and extends down into your windpipe. Since the ventilator blows air directly into your lungs, you don’t have to work as hard to breathe oxygen in on your own.

Depending on the severity of your condition, you may only need the ventilator to help you with breathing, or you may need the ventilator to do all of the breathing for you.

Other forms of breathing assistance, known as noninvasive ventilation (NIV), include bilevel positive airway pressure (BiPAP) and continuous positive airway pressure (CPAP) machines. These may be appropriate long-term options for people with certain conditions, like COPD.

### **How is respiratory failure treated?**

How providers treat respiratory failure depends on how severe it is and what’s causing it. Treatments focus on managing the underlying cause, giving you more oxygen, or using mechanical ventilation to breathe for you until you’re able to on your own again.

Acute respiratory failure is an emergency and needs to be treated right away. Mild chronic respiratory failure can often be treated at home by managing the condition that’s causing it.

#### **Specific treatments for respiratory failure**

Providers may use medications or procedures to treat respiratory failure, including:

* **Mechanical ventilation**. Providers use a breathing machine and a tube that goes into your airways to move air in and out of your lungs.
* **Extracorporeal membrane oxygenation (ECMO).** Providers use a bypass machine to add oxygen to your blood and remove carbon dioxide.
* **Oxygen therapy.** A machine delivers extra oxygen through a breathing mask or small tube (cannula). You may get oxygen at home or in the hospital.
* **Fluids.** Your provider can give you fluids through an IV (directly to a vein). This can improve the blood flow through your body, bringing more oxygen to your tissues.
* **Managing underlying conditions.** You provider may treat you with other medications or procedures, depending on what’s causing respiratory failure.

## **Outlook / Prognosis**

There often isn’t any cure for chronic respiratory failure, but you can manage your symptoms with treatment.

Your specific outlook depends on the exact cause of your respiratory failure, your overall health, and how quickly you receive treatment. If you have a chronic lung disease, you may need continuous help with your breathing.

Respiratory failure is a very serious condition. Many people survive it, depending on what’s causing it, the severity and how quickly they’re treated.

While many causes of acute respiratory failure are treatable, it can be fatal if not treated quickly. Up to 1 in 3 people who are hospitalized for acute respiratory failure don’t survive. Chronic respiratory failure is usually caused by an ongoing condition that gets worse over time.

## **Prevention**

You can’t always prevent respiratory failure. You can lower your risk of chronic respiratory failure by managing ongoing heart, lung and neurological conditions. Talk to your healthcare provider about ways to reduce your risk if you have a condition that can cause respiratory failure.

## **Diagnostic Considerations**

Respiratory failure is a common and a life-threatening condition that demands prompt diagnosis and assessment and appropriate management.

Failure to visualize an obvious abnormality on chest radiographs in hypoxemic respiratory failure suggests the possibility of right-to-left shunting.

The vast majority of patients in acute respiratory failure due to cardiogenic pulmonary edema respond to measures to reduce preload and afterload. Those with acute respiratory distress syndrome (ARDS) require early elective intubation because the duration of respiratory failure is longer.

Hypercapnic respiratory failure occurs secondary to a variety of causes, including an increased respiratory muscle load, impaired neuromuscular function, and decreased respiratory drive caused by central nervous system (CNS) depression.

## **Differential Diagnoses**

* Acute Respiratory Distress Syndrome (ARDS)
* Angina Pectoris
* Aspiration Pneumonitis and Pneumonia
* Asthma
* Atelectasis
* Bacterial Pneumonia
* Cardiogenic Pulmonary Edema
* Cardiogenic Shock
* Community-Acquired Pneumonia (CAP)
* Cor Pulmonale
* Cyanosis
* Diaphragmatic Paralysis
* Dilated Cardiomyopathy (DCM)
* Distributive Shock
* Emphysema
* Hypertrophic Cardiomyopathy
* Idiopathic Pulmonary Fibrosis (IPF)
* Interstitial (Nonidiopathic) Pulmonary Fibrosis
* Mechanical Ventilation
* Myocardial Infarction
* Neurogenic Pulmonary Edema
* Noninvasive Ventilation
* Obstructive Sleep Apnea (OSA)
* Pneumothorax Imaging
* Idiopathic Pulmonary Arterial Hypertension
* Pulmonary Embolism (PE)
* Respiratory Acidosis
* Restrictive Lung Disease
* Pulmonary Arterial Hypertension
* Viral Pneumonia

## **Epidemiology**

Respiratory failure is a syndrome rather than a single disease process, and the overall frequency of respiratory failure is not well known. The estimates for individual diseases mentioned in this article can be found in the Medscape Reference articles specific to each disease.

The relationship between acute respiratory failure and race is still debated. A study by Khan et al suggested that no differences in mortality exist in patients of Asian and Native Indian descent with acute critical illness after adjusting for differences in case mix. [[1](javascript:void(0);)] Moss and Mannino reported worse outcome for African Americans with ARDS than for whites after adjustment for case mix.Future prospective association studies should yield a better knowledge of the impact of race on the outcome of respiratory failure.

## **Guidelines Summary**

Guidelines on acute respiratory distress syndrome published by the European Society of Intensive Care Medicine (ESICM) contain the following key recommendations regarding the management of acute hypoxemic respiratory failure (AHRF):

* Patients with AHRF not due to cardiogenic pulmonary edema or acute exacerbation of COPD should receive high-flow nasal cannula oxygen (HFNO) as compared to conventional oxygen therapy (COT) to reduce the risk of intubation
* Continuous positive airway pressure (CPAP)/noninvasive mechanical ventilation (NIV) can be considered instead of HFNO for the treatment of AHRF due to COVID-19 to reduce the risk of intubation

No recommendations could be made for or against the following treatments:

* Use of HFNO compared to CPAP/NIV to treat AHRF in unselected patients with acute hypoxemic respiratory failure not due to cardiogenic pulmonary edema or acute exacerbation of COPD
* Use of NIV compared to CPAP for the treatment of AHRF

### European Respiratory Society (ERS)

Guidelines on the use of HFNO in acute respiratory failure with the following conditional recommendations:

* Use HFNO over COT in adults with AHRF
* Use HFNO over NIV in patients with AHRF
* Use HFNO over COT when short breaks of NIV are needed due to feeding, speaking, relief from mask pressure, etc.
* Use either COT or HFNO in post-operative patients at low risk of respiratory complications
* Use either HFNO or NIV in post-operative patients at high risk of respiratory complications
* Use HFNO over COT in nonsurgical patients after extubation at low or moderate risk of extubation failure
* Use NIV over HFNO after extubation for patients with no contraindications to NIV and at high risk of extubation failure

### European Respiratory Society/American Thoracic Society (ERS/ATS)

European Respiratory Society/Amerian Thoracic Society (ERS/ATS) joint guidelines on the use of noninvasive ventilation in ARF are summarized below.

Bilevel noninvasive mechanical ventilation (NIV) may be considered in chronic obstructive pulmonary disease (COPD) patients with an acute exacerbation in the following three clinical settings:

* To prevent acute respiratory acidosis (ie, when the arterial carbon dioxide tension [PaCO2] is normal or elevated but pH is normal)
* To prevent endotracheal intubation and invasive mechanical ventilation in patients with mild-to-moderate acidosis and respiratory distress, with the aim of preventing deterioration to a point when invasive ventilation would be considered
* As an alternative to invasive ventilation in patients with severe acidosis and more severe respiratory distress

Bilevel NIV also may be used as the only method for providing ventilatory support in patients who are not candidates for or decline invasive mechanical ventilation.

Bilevel NIV is recommended as follows:

* Patients with ARF leading to acute or acute-on-chronic respiratory acidosis (pH ≤7.35) due to COPD exacerbation
* Patients considered to require endotracheal intubation and mechanical ventilation, unless the patient is immediately deteriorating

Either bilevel NIV or continuous positive airway pressure (CPAP) is recommended for patients with ARF due to cardiogenic pulmonary edema.

CPAP or bilevel NIV is suggested for patients with ARF due to cardiogenic pulmonary edema in the prehospital setting.

Early NIV is suggested for immunocompromised patients with ARF.

NIV use is suggested as follows

* For patients with postoperative ARF
* Can be offered to dyspneic patients for palliation in the setting of terminal cancer or other terminal conditions
* For the prevention of post extubation respiratory failure in high-risk patients; not suggested to prevent post extubation respiratory failure in non–high-risk patients
* To facilitate weaning from mechanical ventilation in patients with hypercapnic respiratory failure

**GENOMIC DATA**

* Surfactant Protein B (SP-B) Deficiency:  
  Mutations in the *SFTPB* gene, such as the 121ins2 mutation, cause hereditary surfactant protein B deficiency leading to severe neonatal respiratory distress and chronic respiratory failure. These loss-of-function mutations impair surfactant production, essential for lung function, resulting in progressive respiratory failure in infants.
* Hereditary Myopathy with Early Respiratory Failure (HMERF):  
  Caused by mutations in the *TTN* gene (titin), this autosomal dominant disorder affects skeletal muscles including respiratory muscles, leading to early-onset respiratory failure. Some mutations cause milder or later-onset symptoms, but homozygous mutations can cause severe respiratory muscle weakness and chronic respiratory failure.
* Chronic Obstructive Pulmonary Disease (COPD) Genetics:  
  COPD, a common cause of chronic respiratory failure, has a genetic predisposition. The most well-known genetic risk factor is alpha-1 antitrypsin deficiency (mutations in *SERPINA1*), which leads to early-onset emphysema and respiratory failure. Other candidate genes (e.g., *CHRNA3/5*, *HHIP*, *FAM13A*) have been implicated in susceptibility and progression of COPD, though data are complex and sometimes conflicting.
* Gene Mutations in Lung Cancer with COPD:  
  Studies of non-small cell lung cancer (NSCLC) patients with coexisting COPD show distinct mutation profiles, including genes like *EGFR*, *TP53*, *KRAS*, and others (e.g., *LRP1B*, *MLH1*), which may correlate with more severe lung disease and respiratory failure.
* Other Genetic Mutations Linked to Chronic Lung Disease:  
  Rare mutations in genes such as *DNAL1* have been identified in indigenous populations causing chronic lung disease with respiratory failure

**PREDEFINED Q AND A**

## 1. What are my treatment options?

* Oxygen therapy: Supplemental oxygen delivered via nasal cannula or mask to increase blood oxygen levels and reduce shortness of breath. It may be used continuously or during sleep and exercise.
* Inhaled medications: Bronchodilators and other inhaled drugs help open airways and improve lung function.
* Oral medications: Depending on the underlying cause, medications may prevent worsening lung function or treat infections and inflammation.
* Non-invasive ventilation (NIV): Devices like CPAP or BiPAP provide positive airway pressure to keep airways open, often used during sleep.
* Mechanical ventilation: In severe cases, a ventilator may assist or fully support breathing, sometimes via a tracheostomy.
* Tracheostomy: Surgical opening in the neck to place a breathing tube for long-term ventilation support if needed.
* Respiratory rehabilitation: Exercises and education to improve breathing endurance and muscle strength.
* Fluid management: Proper hydration supports circulation without causing lung fluid overload.

## 2. What caused this?

Chronic respiratory failure results from long-standing lung or respiratory muscle diseases that impair oxygenation and/or carbon dioxide removal. Common causes include:

* Chronic Obstructive Pulmonary Disease (COPD)
* Interstitial lung diseases (pulmonary fibrosis)
* Neuromuscular disorders affecting breathing muscles
* Chest wall deformities
* Severe obesity with hypoventilation
* Advanced heart or lung disease

## 3. How do I take my medications?

* Follow your healthcare provider’s instructions carefully for each medication.
* Inhaled medications are typically administered via inhalers or nebulizers; use proper technique to ensure effective delivery.
* Oral medications should be taken with or without food as directed, at prescribed times.
* Never stop or change doses without consulting your provider.

## 4. How do I use this device? Can you demonstrate?

* Devices like inhalers, nebulizers, CPAP, or BiPAP machines require specific techniques.
* Your healthcare provider or respiratory therapist can demonstrate proper use, including:
  + How to assemble and clean the device
  + How to position the mask or mouthpiece
  + How to breathe during treatment for maximum benefit
* Ask for a hands-on demonstration and practice under supervision to ensure you are comfortable using the device.

## 5. When can I expect to feel better?

* Improvement depends on the underlying cause and severity.
* Oxygen therapy and ventilation support can relieve symptoms quickly, but chronic lung conditions often require ongoing management.
* Pulmonary rehabilitation may improve exercise tolerance and quality of life over weeks to months.
* Some patients experience symptom stabilization rather than full recovery.

## 6. When should I follow up with you?

* Regular follow-up visits are essential, often every 3 to 6 months or as advised.
* Contact your provider sooner if you notice worsening symptoms such as increased breathlessness, cough, sputum changes, or fatigue.
* Follow-up includes monitoring lung function, oxygen needs, and medication effectiveness

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I want to discuss your recent diagnosis of chronic respiratory failure. How have you been feeling lately?

Patient: I’ve been feeling more short of breath, especially when I walk or do simple activities. Sometimes I feel very tired.

Doctor: That’s understandable. Chronic respiratory failure means your lungs aren’t able to get enough oxygen into your blood or remove carbon dioxide efficiently over a long period. It often results from underlying lung or muscle problems.

Patient: What caused this condition in my case?

Doctor: In your situation, it’s mainly due to your chronic lung disease, which has gradually impaired your lung function. Other causes can include neuromuscular weakness or chest wall problems, but your tests point toward lung-related causes.

Patient: What treatments are available to help me?

Doctor: We will focus on supporting your breathing and managing symptoms. This includes supplemental oxygen therapy to raise your oxygen levels, inhaled medications like bronchodilators to open your airways, and possibly non-invasive ventilation like CPAP or BiPAP to assist your breathing, especially at night. Pulmonary rehabilitation exercises can also improve your lung strength and endurance.

Patient: How do I take my medications and use these devices?

Doctor: I will show you how to use your inhalers and oxygen equipment properly. For devices like CPAP or BiPAP, a respiratory therapist will provide hands-on training to ensure you are comfortable and using them correctly. It’s important to follow the instructions carefully to get the best benefit.

Patient: When can I expect to feel better?

Doctor: Some symptoms like breathlessness can improve quickly with oxygen and ventilation support. However, chronic respiratory failure is a long-term condition, so ongoing management is needed. Pulmonary rehab and medication adherence can improve your quality of life over weeks to months.

Patient: When should I come back for follow-up?

Doctor: We’ll schedule regular visits every 3 to 6 months to monitor your lung function and adjust treatment as needed. If you notice worsening breathlessness, increased coughing, changes in sputum, or any new symptoms like confusion or chest pain, please seek medical attention promptly.

Patient: Thank you, doctor. This helps me understand what to expect and how to manage my condition.

Doctor: You’re welcome. We’re here to support you, so don’t hesitate to contact us with any concerns.

REFERENCES

https://www.ncbi.nlm.nih.gov/books/NBK526127/

[Respiratory Failure: Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24835-respiratory-failure#diagnosis-and-tests)

<https://www.nhlbi.nih.gov/health/respiratory-failure/treatment>

### **Wheezing**

## **Alternative Names for Wheezing**

* Expiratory Wheeze  
  Refers specifically to wheezing heard during exhalation.
* Inspiratory Wheeze (Stridor)  
  Though stridor is a distinct high-pitched sound during inhalation, it is sometimes confused with inspiratory wheezing.
* Bronchospasm  
  Describes the underlying mechanism causing wheezing—constriction of the bronchial muscles.
* Sibilant Wheeze  
  A medical term emphasizing the high-pitched, musical quality of wheezing.
* Airway Obstruction Sounds  
  A broader term that includes wheezing as a sign of narrowed airways.

**DEFINITION AND DESCRIPTION**

Wheezing is the shrill, coarse whistling or rattling sound your breath makes when your airway is partially blocked or narrowed. It’s usually most apparent when you breathe out (exhale). Sometimes, it can be a sign that you’re having breathing problems due to an underlying condition. But, other times, wheezing can be a reaction to dust in the air or because you have a cold.

Many treatments are available for wheezing, depending on what’s causing it. Your healthcare provider may recommend seeing a pulmonologist or allergist if your symptoms persist or if wheezing is due to a chronic health condition like asthma.

### **What does wheezing sound like?**

Some wheezes can only be heard with a stethoscope, but often, you can hear them with your own ears. Wheezing is more obvious when you breathe out but can also be heard when you breathe in. The tone of the wheeze can vary depending on which part of your respiratory system is blocked or narrowed. Narrowing in the upper respiratory system may make for a hoarser wheeze. Lower obstructions may have a more musical tone, like how a wind instrument like a clarinet might sound.

Anyone — from infants to older adults — can develop wheezing. In adults, people who smoke and people with emphysema or heart failure are most prone to wheezing.

Wheezing is also quite common in infants. Up to 25% to 30% of infants develop wheezing in their first year. This may happen because babies have smaller airways. Children under 2 are also more susceptible to wheezing because they tend to get viral upper respiratory infections.

Adults and children with asthma and allergies may also be more likely to experience wheezing.

### **causes of wheezing**

An obstruction (blockage) or narrowing of the small bronchial tubes in your chest usually causes wheezing. An obstruction in your larger airways or vocal cords can also cause it. The causes range from chronic but manageable conditions like asthma to serious conditions like heart failure.

Many different things can cause wheezing, such as medical conditions, infections or viruses, and lifestyle factors.

#### **Lung issues**

* **Asthma:** A chronic condition that causes spasms and swelling in your bronchial tubes. Exposure to airborne allergens such as pollen, mold or dust can trigger wheezing in asthma. Viral illnesses can also make asthma symptoms worse. Asthma is one of the most common causes of wheezing.
* **Aspirating:** Breathing a foreign object or substance into your lungs.
* **Bronchitis:** Inflammation of the lining inside your bronchial tubes.
* **Bronchiolitis**: A virus most common in young children that causes inflammation and irritation in their airways.
* **Bronchiectasis:** Damage to the large airways in your lungs.
* **Chronic obstructive pulmonary disease (COPD):**  Inflammation and damage of the lining of your bronchial tube, most commonly from smoking cigarettes.
* **Cystic fibrosis (CF):** A condition that causes thick mucus to clog your airways and make breathing difficult.
* **Emphysema:** A lung condition that makes you short of breath.
* **Pneumonia:** Lung inflammation caused by a virus or bacteria. Other viral infections can cause wheezing, especially in infants and toddlers.
* **Respiratory syncytial virus (RSV)**: A seasonal lung infection that’s common in children.

#### **Vocal cord issues**

* **Vocal cord dysfunction** VCD causes your vocal cords to close instead of open when you breathe in and out, making it harder to get air into or out of your lungs.

#### **Issues with your digestive tract**

* **GERD:** Chronic acid reflux can relax the lower esophageal valve, causing wheezing.

#### **Allergies**

* **Allergies**: Allergen triggers like dust mites, pollens, pets, mold spores and foods can cause wheezing.
* **Anaphylaxis**: A severe allergic reaction typically caused by food allergies or insect stings can make you wheeze.

#### **Heart conditions:**

* **Heart failure:** Fluid in your lungs due to heart failure can make you wheeze.

#### **Lifestyle factors**

* **Smoking**: Smoking tobacco increases your risk of developing COPD and emphysema and makes it harder to manage conditions like asthma.
* **Medications:** Certain medications (like aspirin) may contribute to wheezing.
* **Sleep apnea:** Sleep disorders can cause wheezing.

## **Wheezing Risk Factors**

## **Wheezing in Infants**

Wheezing is common in infants. About 50% of children have a wheezing episode in their first year of life.

Young children are more likely to wheeze because their airways are small, and they’re also more likely to get upper respiratory infections than adults are. When this happens, the infants’ small passages swell, and they often fill with mucus, which can cause wheezing and coughing.

If your infant is wheezing, that doesn’t necessarily mean they have asthma. Even if it happens repeatedly, they may outgrow it when they’re older and not have asthma. Sometimes, doctors will wait until the child is 4 or 5 before confirming a diagnosis of asthma.

Wheezing can also mean that an object is lodged in the airway and needs to be removed. This is a concern especially with young children who may have put an object in their mouth. If a child starts wheezing out of the blue, this may be the reason, Moss says.

**Diagnosing the Cause of Wheezing**

To diagnose the cause of your wheezing, your doctor will perform an exam. They will also ask you some questions, such as:

* How long have you been wheezing?
* Does it happen when you exercise?
* Do you wheeze all the time?
* Do you wheeze more during the day or at night?
* Does rest help control your wheezing?
* Do you wheeze when you breathe in, out, or both in and out?
* Do you smoke?
* Do certain foods seem to cause your wheezing?

They’ll listen to your breathing and the sounds your lungs make. They might do certain tests, such as:

* X-rays to get a picture of your lungs
* Breathing tests to see how well your lungs are functioning
* [A](https://www.webmd.com/heart/anatomy-picture-of-blood) blood test to check your oxygen levels (too low levels could signal a lung problem)

If your child is wheezing, their doctor might check to see if they’ve swallowed or inhaled something small.

**TREATMENT**

Your treatment for wheezing depends on its underlying cause. If you go to the ER or see a healthcare provider, they may begin with oxygen therapy to help you breathe. If wheezing is severe or doesn’t improve with supplemental oxygen, you may need to be hospitalized until your breathing improves.

#### **What medicines can you take for wheezing?**

Most of the time, taking medication to treat the cause of the wheezing improves your symptoms. For example, using an inhaler for asthma or taking antibiotics for an infection usually helps.

#### **Asthma medications**

If asthma is causing you to wheeze, your healthcare provider will likely prescribe an inhaler to reduce inflammation and open your airways (a bronchodilator). Inhaled corticosteroids and pills such as montelukast (Singulair®) are anti-inflammatory medicines to treat asthma.

**Bronchitis medications**

If your provider determines bronchitis is causing your wheezing, they may prescribe a bronchodilator such as albuterol (Proair® HFA, Proventil® HFA, Ventolin® HFA) or an antibiotic to heal a bacterial infection. This should help you breathe better as you recover.

Other causes of wheezing may require specific treatments like oxygen therapy. Your provider will prescribe a plan to treat the underlying cause of your condition and soothe symptoms to help you feel better faster.

### **OTC (over the counter) treatments for wheezing**

There are many ways you can improve wheezing at home without a prescription. Some of those treatments include:

* **Breathing exercises.** Taking slow, deep breaths helps expand your lung capacity and relaxes your airways (diaphragmatic breathing). Deep breathing in a moist, humid environment (like a steam room) can also help.
* **Drink hot herbal tea.** The warmth and moisture of tea will help relax your bronchial tubes. Some studies show green tea has antibacterial properties, as well.
* **Don’t smoke.** Smoking irritates your lungs and inflames your airways. Take precautions to avoid secondhand smoke.
* **Use an air purifier with a HEPA filter.** High-quality filters help remove allergens in your home.
* **Vaporize your air:** Humidifiers or vaporizers moisten the air to help you breathe better.
* **Stay away from known allergy triggers.** Avoid things that trigger your allergies.

## **When To Call the Doctor**

See your healthcare provider if your wheezing is new, if it keeps coming back or if any of the following symptoms accompany it:

* Shortness of breath or rapid breathing.
* Confusion or altered mental state.
* Chest tightness or chest pain.
* Swelling of your lips or tongue.
* A bluish tinge around your skin, mouth or nails.

Your healthcare provider will perform a physical exam, listen to your lungs and breath and ask about your symptoms. Questions could include things like:

* When did the wheezing start?
* Is the wheezing getting worse or staying the same?
* Is the wheezing constant or all day, or does it come and go?
* Do certain factors make the wheezing worse, like exercise, lying down or being outside?

Your provider may also order tests like:

* Pulse oximetry (pulse ox, a finger sensor to measure oxygen levels).
* Chest X-rays.
* Pulmonary function tests.
* Blood tests.

Whatever the cause, there are things you can do to get relief. Follow your healthcare provider’s instructions on taking medication, choosing not to smoke (or quitting) and running a vaporizer or humidifier to moisten the air. Doing all these things will help you breathe easier.

If your skin, mouth or nails are turning blue or you’re gasping for air, it’s a sign that your lungs don’t have enough air. This is a medical emergency and you should have a family member or friend take you to the nearest emergency room. If you’re alone, call 911 (or your emergency services number) and describe your breathing.

If you suddenly start wheezing after a bee sting, after you take a new medication or eat a new food, that could indicate an allergic reaction, and you should seek medical attention right away.

## **Breathing Exercises for Wheezing**

Breathing exercises can reduce your wheezing and help your lungs work more efficiently. “Breathing techniques can also help the airway relax — help to change how a person is breathing and moving air in their airway,” Moss says.

Try these breathing techniques:

**Pursed-lip breathing.** Breathe in through your nose. Breathe out for twice as long, with your lips pursed like you’re going to whistle. Breathing out through pursed lips increases the airway pressure and helps open your airways a bit, Moss says.

**Belly breathing or diaphragmatic breathing.** Breathe in through your nose. Put your hands on your belly and pay attention to how it expands. Breathe out through your mouth for at least two to three times as long as you breathe in. If you’re wheezing, your chest may feel tight. When you focus on your belly or your diaphragm while breathing, it helps relax your chest, Moss says.

**Buteyko breathing method.** Breathe in and out through your nose for a few minutes. After breathing out, plug your nose and hold your breath, inhaling when you feel the urge to breathe. Breathe in and out through your nose a few times and repeat. This helps normalize breathing patterns.

**Nasal breathing.** Breathe in and out through your nose, relaxing your jaw and throat. Breathe slow and steady. This helps warm and humidify the air as you breathe it in.

Other breathing methods used in yoga or meditation can be helpful, too, Moss says. With these techniques, you’re “focusing your breathing deeper in your core, down to your belly, so that you’re getting a much deeper breath and you’re not breathing as shallowly,” he says.

Many breathing techniques are used in yoga and meditation, and they may help improve your breathing. They include:

**Alternate nostril breathing.** While sitting up, use your right thumb to close your right nostril, and inhale deeply through your left nostril. Use your ring finger to close your left nostril, and breathe out through your right nostril. Then switch, breathing in through your right, and repeat the process.

**Bumblebee breath.** While sitting, close your eyes and put your index fingers on your eyelids, thumbs in your ears, and rest of the fingers on your face. Take a deep breath through your nose, and as you breathe out, close the back of your throat to make a humming or buzzing sound.

## **Differential Diagnoses for Wheezing**

## Common Causes

* Asthma (most common cause of recurrent wheezing)
* Chronic Obstructive Pulmonary Disease (COPD)
* Acute Bronchitis
* Allergic reactions / Anaphylaxis
* Viral respiratory infections (e.g., bronchiolitis in infants)
* Foreign body aspiration (especially in children)
* Heart failure (cardiac asthma)

## Other Causes

* Bronchiectasis
* Upper airway obstruction (e.g., vocal cord dysfunction, tumors, tracheal stenosis)
* Gastroesophageal reflux disease (GERD) causing airway irritation
* Pulmonary edema
* Cystic fibrosis
* Interstitial lung diseases (rarely cause wheezing but possible)
* Tumors or masses compressing airways
* Inhalation injury or exposure to irritants
* Vocal cord dysfunction or paradoxical vocal fold motion

**Epidemiology of Wheezing (mainly related to asthma):**

* The prevalence of current wheeze varies globally, ranging from about 5.6% in low- and middle-income countries (Asia and Africa) to 11.6% in high-income countries (Europe and North America).
* Wheezing is a common symptom of asthma, which affected an estimated 262 million people worldwide in 2019, causing approximately 455,000 deaths.
* Asthma and wheezing are more prevalent in children, with higher incidence in boys during childhood; however, adults tend to have higher morbidity and mortality from asthma.
* Severe asthma, often accompanied by persistent wheezing, affects a smaller proportion of patients—around 2-8% depending on the population studied.
* Risk factors influencing wheezing prevalence include family history of asthma, other allergic conditions (eczema, rhinitis), urban living, exposure to tobacco smoke and air pollution, viral respiratory infections early in life, and obesity.
* Wheezing prevalence and asthma rates tend to be higher in high-income countries, likely due to lifestyle and environmental factors.
* In the US, asthma prevalence among young adults is around 8-9%, with wheezing being a common symptom among these patients

**GENOMIC DATA**

* Different wheezing phenotypes (e.g., virus-induced wheezing vs. atopic asthma) are associated with distinct genetic polymorphisms. For example, virus-induced wheezing is frequently linked to IL-8 gene polymorphisms, while atopic asthma and atopy are associated with Th2 cytokine genes such as CD14 and IL-13 on chromosome 5.
* The gene ADAM33 (on chromosome 20p13) is strongly linked to asthma and bronchial hyperresponsiveness, contributing to airway remodeling and disease progression, which can manifest as wheezing.
* Filaggrin gene mutations (chromosome 1q21), which impair skin barrier function, are associated with atopic dermatitis and increase the risk of allergic sensitization and asthma with wheezing, especially in patients with atopic dermatitis.
* A significant genetic locus on chromosome 17q21 (including genes like ORMDL3 and GSDMB) is strongly associated with early-onset wheezing and childhood asthma. Children with risk variants in this region who experience early-life wheezing illnesses caused by human rhinovirus have a dramatically increased risk of developing asthma.
* Genetic susceptibility to wheezing and asthma is complex, involving multiple genes and gene-environment interactions. For example, the combination of genetic risk variants and early viral wheezing illnesses can synergistically increase asthma risk.
* Other genes implicated in wheezing and asthma include IL33, ANXA1, and genes involved in immune regulation and airway epithelial response to infections

**PREDEFINED Q AND A**

**How can I stop wheezing immediately?**

If your wheezing is severe and making it difficult to breathe, you may need albuterol or oxygen treatment. If your wheezing is not severe, you can try breathing exercises, such as belly breathing, to help relax your airways.

**Can GERD cause wheezing?**

Yes. Gastroesophageal reflux disease (GERD) is a condition that can cause wheezing.

**How can I stop wheezing without inhalers?**

You may or may not need medication for your wheezing. Breathing exercises also may help you stop wheezing. Sometimes wheezing will go away on its own. “For example, someone who’s allergic to a cat and has asthma, and goes to a house where there's a cat, may notice that they start wheezing right away and leave the house, breathe some fresh air, and feel improvement within minutes because they’ve removed that trigger,” Moss says.

**Can wheezing be cured?**

Wheezing isn’t exactly a disease that can be cured, but it can be stopped. For example, if you’re wheezing because you have asthma, taking preventative medication daily can help stop your wheezing. “A lot of people wouldn’t necessarily call that being cured because they’re still taking medication preventatively. So, a lot of times we’ll describe it as wheezing or asthma that goes into remission,

**Doctor-patient conversation about wheezing**

Doctor: Hello, how can I help you today?

Patient: Hi doctor, I’ve been having some trouble breathing lately. I noticed a whistling sound when I breathe out.

Doctor: I see. That sound is called wheezing. Have you experienced wheezing before, or do you have any history of asthma or allergies?

Patient: No, this is the first time. I also have a cough and feel a bit short of breath.

Doctor: Okay, let me listen to your lungs. Please take a deep breath and hold it for a moment.

*(Doctor listens with stethoscope)*

Doctor: I do hear some wheezing in your lungs, which suggests your airways are narrowed. This could be due to asthma, an infection, or allergies. Have you had any recent colds or exposure to irritants like smoke?

Patient: I had a cold last week, and I live with someone who smokes.

Doctor: That could be contributing. I’m going to prescribe an inhaler that will help open your airways and reduce inflammation. It’s important to use it exactly as directed. Also, try to avoid smoke and other triggers.

Patient: How soon will I feel better?

Doctor: Many patients notice improvement within a few days of starting treatment. If your symptoms worsen or you have difficulty breathing, chest pain, or high fever, please come back immediately.

Patient: Should I come back for a follow-up?

Doctor: Yes, I’d like to see you in about two to three weeks to check your progress and adjust treatment if needed.

Patient: Thank you, doctor.

Doctor: You’re welcome. Don’t hesitate to call if you have questions or concerns before your appointment.

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**CHRONIC COUGH**

**DEFINITION AND DESCRIPTION**

A chronic cough is a cough that lasts eight weeks or longer in adults, or four weeks in children. A chronic cough is more than just annoying. It can interrupt your sleep and leave you feeling very tired. Severe cases of chronic cough can cause vomiting and lightheadedness and even break a rib.

The most common causes are tobacco use and asthma. Other common causes include fluid that drips from the nose down the back of the throat, called postnasal drip, and the backward flow of stomach acid into the tube that connects the throat to the stomach, called acid reflux. Fortunately, chronic cough usually goes away once the underlying issue is treated.

**Causes**

A cough that happens once in a while is common. It helps clear irritants and mucus from your lungs and prevents infection. But a cough that lasts for weeks is usually due to a health concern. Many times, more than one health concern causes the cough.

Most cases of chronic cough are due to these causes, which can occur alone or together:

* **Postnasal drip.** When your nose or sinuses produce extra mucus, it can drip down the back of your throat and cause you to cough. This condition also is called upper airway cough syndrome.
* **Asthma.** An asthma-related cough may come and go with the seasons. It may appear after an upper respiratory tract infection. Or it can get worse when you're exposed to cold air or certain chemicals or fragrances. In one type of asthma known as cough-variant asthma, a cough is the main symptom.
* **Gastroesophageal reflux disease.** In this common condition, also called GERD, stomach acid flows back into the tube that connects your stomach and throat. This tube is also known as your esophagus. The constant irritation can lead to chronic coughing. Then the coughing can make GERD worse, creating a vicious cycle.
* **Infections.** A cough can last long after other symptoms of pneumonia, flu, a cold or another infection of the upper respiratory tract have gone away. A common cause of a chronic cough in adults — but one that often isn't recognized — is whooping cough, also known as pertussis. Chronic cough also can occur with fungal infections of the lung, as well as tuberculosis infection, also called TB, or lung infection with nontuberculous mycobacteria, also called NTM. NTM is found in soil, water and dust.
* **Chronic obstructive pulmonary disease (COPD).** Also called COPD, this is a lifelong inflammatory lung disease that limits airflow from the lungs. COPD includes chronic bronchitis and emphysema. Chronic bronchitis can cause a cough that brings up colored sputum. Emphysema causes shortness of breath and damages the air sacs in the lungs, also known as alveoli. Most people with COPD are current or former smokers.
* **Blood pressure drugs.** Angiotensin-converting enzyme inhibitors, also called ACE inhibitors, which are commonly prescribed for high blood pressure and heart failure, are known to cause chronic cough in some people.

Less commonly, chronic cough may be caused by:

* Aspiration — when food or other items are swallowed or inhaled and go into the lungs.
* Bronchiectasis — widened and damaged airways that slowly lose the ability to clear out mucus.
* Bronchiolitis — an infection that causes swelling, irritation and buildup of mucus in the small airways of the lung.
* Cystic fibrosis — a genetic disorder that affects the lungs, digestive system and other organs.
* Idiopathic pulmonary fibrosis — gradual damage and scarring of the lungs due to a cause that isn't known.
* Lung cancer — cancer that starts in the lungs, including non-small cell lung cancer and small cell lung cancer.
* Non asthmatic eosinophilic bronchitis — when airways are inflamed but asthma is not the cause.
* Sarcoidosis — groups of inflamed cells that form lumps or nodules in different parts of the body but most often in the lungs.

**Risk factors**

Being a current or former smoker is one of the leading risk factors for chronic cough. Exposure to a lot of secondhand smoke also can lead to coughing and lung damage.

**Symptoms**

A chronic cough can occur with other symptoms, including:

* A runny or stuffy nose.
* A feeling of liquid running down the back of your throat, also known as postnasal drip.
* Clearing your throat a lot.
* Sore throat.
* Hoarseness.
* Wheezing and shortness of breath.
* Heartburn or a sour taste in your mouth.
* In rare cases, coughing up blood.

### **When to see a doctor**

See your healthcare professional if you have a cough that lasts for weeks, especially one that brings up sputum or blood, disturbs your sleep, or affects school or work.

## **Diagnosis**

Your healthcare professional asks about your medical history and does a physical exam. A thorough medical history and physical exam can give important clues about a chronic cough. Your health professional also may order tests to look for the cause of your chronic cough.

But many health professionals start treatment for one of the common causes of chronic cough rather than ordering expensive tests. If the treatment doesn't work, you may be tested for less common causes.

### **Imaging tests**

* **X-rays.** Although a routine chest X-ray won't reveal the most common reasons for a cough — postnasal drip, acid reflux, tobacco use or asthma — it may be used to check for lung cancer, pneumonia and other lung diseases. An X-ray of your sinuses may reveal evidence of a sinus infection.
* **Computerized tomography scans.** These scans also are called CT scans. They may be used to check your lungs for conditions that may produce chronic cough or your sinus cavities for pockets of infection.

### **Lung function tests**

These simple, noninvasive tests, such as spirometry, are used to diagnose asthma and COPD. They measure how much air your lungs can hold and how fast you can exhale.

Your healthcare professional may request an asthma challenge test. This test checks how well you can breathe before and after inhaling the drug methacholine (Provocholine).

### **Lab tests**

If the mucus that you cough up is colored, your healthcare professional may want to test a sample of it for bacteria.

### **Scope tests**

If your healthcare professional can't find the cause of your cough, special scope tests may be used to look for possible causes. These tests may include:

* **Bronchoscopy.** A bronchoscope is a thin, flexible tube that has a light and camera attached to it. Your health professional can look at your lungs and air passages. A biopsy also can be taken from the inside lining of your airway, also known as the mucosa, to look for anything unusual. A biopsy is a procedure to remove a sample of tissue for testing in a lab.
* **Rhinoscopy.** Using a fiberoptic scope, also known as a rhinoscope, your health professional can view your nasal passageways, sinuses and upper airway.

### **Children**

A chest X-ray and spirometry, at a minimum, are usually ordered to find the cause of a chronic cough in children.

**Treatment**

Finding out what's causing a chronic cough is very important to effective treatment. In many cases, more than one underlying condition may be causing your chronic cough.

If you smoke, your healthcare professional likely will talk with you about your readiness to quit and give you advice on how to achieve this goal. If you're taking an ACE inhibitor medicine, your health professional may switch you to another medicine that doesn't have cough as a side effect.

Medicines used to treat chronic cough may include:

* **Antihistamines, corticosteroids and decongestants.** These medicines are standard treatment for allergies and postnasal drip.
* **Inhaled asthma medicines.** The most effective treatments for asthma-related cough are corticosteroids and bronchodilators. They reduce inflammation and open up your airways.
* **Antibiotics.** If a bacterial, fungal or mycobacterial infection is causing your chronic cough, your healthcare professional may prescribe antibiotic medicines for the infection.
* **Acid blockers.** When lifestyle changes don't take care of acid reflux, you may be treated with medicines that block acid production. Some people need surgery to resolve the problem.

### **Medicine to reduce coughing**

Your healthcare professional works to find the cause of your cough and the best treatment for you. During that time, your healthcare professional also may prescribe a medicine to reduce coughing, called a cough suppressant. Cough suppressants are not recommended for children.

Cough and cold medicines available without a prescription treat the symptoms of coughs and colds — not the underlying disease. Research suggests that these medicines don't work any better than no medicine at all. These medicines are not recommended for children because of potentially serious side effects, including fatal overdoses in children younger than 2 years old.

Don't use over-the-counter cough and cold medicines, except for fever reducers and pain relievers, to treat coughs and colds in children younger than 6 years old. Also, avoid use of these medicines for children younger than 12 years old. Check with your healthcare professional for guidance.

**Treatment options for chronic cough, including common drugs and their side effects**

## 1. Cough Suppressants (Antitussives)

* Purpose: Reduce the cough reflex, mainly for dry, irritating coughs.
* Common drugs:
  + *Dextromethorphan* (found in Robitussin DM, Delsym)
  + *Codeine* (prescription only)
  + *Benzonatate* (Tessalon)
* Side effects:
  + Drowsiness, dizziness
  + Nausea
  + Potential for dependence (especially codeine)
  + Not recommended for children under 6 years due to risk of serious side effects.

## 2. Expectorants

* Purpose: Loosen mucus in the airways to help clear productive coughs.
* Common drugs:
  + *Guaifenesin* (Mucinex)
* Side effects:
  + Nausea, vomiting
  + Dizziness
  + Rare allergic reactions.

## 3. Mucolytics

* Purpose: Thin mucus to make it easier to cough up.
* Common drugs:
  + *Bromhexine* (not widely available in all countries)
* Side effects:
  + Gastrointestinal discomfort
  + Rare allergic reactions.

## 4. Inhaled Bronchodilators and Corticosteroids

* Purpose: Reduce airway inflammation and open airways, especially if asthma or COPD is the cause.
* Common drugs:
  + *Albuterol* (bronchodilator)
  + *Fluticasone* or *Budesonide* (inhaled steroids)
* Side effects:
  + Tremors, nervousness (bronchodilators)
  + Oral thrush, hoarseness (inhaled steroids)
  + Long-term steroid use may have systemic effects.

## 5. Acid-Reducing Medications (for GERD-related cough)

* Purpose: Reduce stomach acid to prevent reflux-induced cough.
* Common drugs:
  + *Proton pump inhibitors* (e.g., omeprazole)
  + *H2 receptor blockers* (e.g., ranitidine)
* Side effects:
  + Headache, diarrhea
  + Long-term use may increase risk of fractures or infections.

## 6. Antihistamines and Decongestants (for postnasal drip)

* Purpose: Reduce nasal congestion and allergic inflammation causing cough.
* Common drugs:
  + *Diphenhydramine*, *chlorpheniramine* (antihistamines)
  + *Pseudoephedrine*, *oxymetazoline* (decongestants)
* Side effects:
  + Drowsiness (antihistamines)
  + Increased heart rate, insomnia (decongestants)
  + Not recommended for people with hypertension or certain heart conditions.

## 7. Antibiotics

* Purpose: Treat bacterial infections causing chronic cough (e.g., pneumonia, whooping cough).
* Side effects:
  + Gastrointestinal upset
  + Allergic reactions
  + Antibiotic resistance with misuse.

**Lifestyle and home remedies**

Follow the plan your healthcare professional gives you for treating the cause of your cough. In the meantime, you can try these tips to ease your cough:

* **Drink fluids.** Liquid helps thin the mucus in your throat. Warm liquids, such as broth, tea or juice, can soothe your throat.
* **Suck on cough drops or hard candies.** They may ease a dry cough and soothe an irritated throat.
* **Consider taking honey.** A teaspoon of honey may help loosen a cough. Don't give honey to children younger than 1 year old. Honey can contain bacteria harmful to infants.
* **Moisturize the air.** Use a cool-mist humidifier or take a steamy shower.
* **Avoid tobacco smoke.** Smoking or breathing secondhand smoke irritates your lungs and can worsen coughs. If you smoke, talk with your healthcare professional about programs and products that can help you quit.

#### **What happens if chronic cough goes untreated?**

Chronic coughing can keep you up at night and rob you of precious sleep (insomnia). The effects can snowball into fatigue and stress that impact every part of your life. The frequent coughing can make you self-conscious — so much so that you avoid being around others.

In severe cases of chronic cough, you may have related medical issues like:

* Bleeding in your eye
* Broken ribs
* Dizziness
* Headaches
* Hernia
* Loss of bladder control
* Muscle pain in your coughing muscles

### **Prevention**

You can’t always prevent chronic cough. But you can do things to reduce your risk. You can:

* **Quit smoking**. If you don’t smoke, don’t start.
* **Protect yourself from germs that can make you sick**. This may mean staying up to date on flu shots, masking or washing your hands often. There are lots of ways to protect yourself.
* **See your primary care provider and discuss the issue with them**. They’ll help you decide if you need testing or treatment based on your symptoms.

**Complications**

Having a cough that doesn't stop can be very tiring. Coughing can cause various concerns, including:

* Sleep disruption.
* Headache.
* Dizziness.
* Vomiting.
* Sweating a lot.
* Unintended bladder loss, also known as urinary incontinence.
* Broken ribs.
* Passing out, also known as syncope.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of chronic cough includes the following conditions:

* Bronchiolitis
* Bronchogenic carcinoma
* Chronic aspiration
* Congestive heart failure
* Foreign body aspiration
* Neuromuscular disorders
* Psychogenic cough

**EPIDEMIOLOGY**

Chronic cough has a global prevalence of 3% to 18% in the general adult population and is affected by several factors, such as smoking and age Eighteen percent of adults in the United States who smoke have chronic coughs. The prevalence is variable, however, based on the location of the population studied. Chronic cough was significantly more frequent in Europe and America than in Asia and Africa. However, the geographic variation is not genetic or ethnically related. The regional variation in chronic cough prevalence may be attributed to environmental factors, particularly urbanization in Western countries, which can lead to increased inhalational exposure to irritants.

The findings from the KNHANES 2010–2012 study indicate that the prevalence of chronic cough increases significantly with age. The odds ratio (OR) of 2.20 and 95% confidence interval of 1.53 to 3.16 suggests a substantial increase in the likelihood of chronic cough among individuals 65 or older compared to those aged 18 to 39. This finding aligns with the general trend observed in various studies, where chronic cough becomes more prevalent in older age groups. The cumulative effects of environmental exposures over time may also play a role. One study by Dicpinigaitis et al assessed ethnic and sex differences in cough reflex sensitivity and found no significant ethnic differences in cough reflex sensitivity among 3 distinct ethnic groups: White, Indian, and Chinese. This suggests that the variation in the prevalence of chronic cough among different races may not be directly attributed to differences in cough reflex sensitivity.

In addition to age and smoking, the factors that contribute to the increased prevalence of chronic cough include obesity, atopy, asthma, COPD, GERD, ACE inhibitors, and sleep-disordered breathing. Other factors, such as air pollution and air quality, do not significantly affect the prevalence of chronic cough, and study results have been inconclusive, except for metal exposure.

**PROGNOSIS**

Cough is a distinct and independent factor that can serve as an indicator of disease progression. The prognostic implications of cough in patients with idiopathic pulmonary fibrosis (IPF) are significant. A study of 242 IPF patients revealed that the presence of cough was an independent predictor of disease progression, regardless of the severity of the disease. Furthermore, cough was more prevalent in patients with advanced pulmonary fibrosis. Nevertheless, it is crucial to differentiate whether the cough results from the underlying inflammation or fibrosis or is due to a comorbidity when assessing cough in patients with interstitial lung disease (ILD).

A recent study revealed that many individuals diagnosed with chronic cough continued to experience persistent coughing for at least 5 years. This finding was somewhat more optimistic than a prior study on the same subject, which reported that 60% of patients experienced worsening or unchanged cough symptoms at a 7-year follow-up. The disparity in the results can be attributed to differences in the patient populations studied. The earlier investigation was limited to patients with unexplained chronic cough, whereas the recent study enrolled individuals with various cough-related disorders and idiopathic cough. Thus, the latter study may provide a more accurate representation of the general chronic cough patient population. Notably, the prevalence of autoimmune diseases was high in the more recent group, a characteristic commonly observed in patients with idiopathic chronic cough. Among the study participants, most asthma and chronic rhinitis patients were effectively managed with local corticosteroid preparations or antihistamines. However, most reflux disease patients did not receive treatment with proton pump inhibitors, an ineffective regimen for reflux-associated cough. Consequently, the study population likely reflects the real-life medication regimens of patients. The study further identified obesity as a significant predictor of continued cough-related quality of life impairment. Indeed, subjects who met the current criteria for obesity were found to be at a higher risk for ongoing cough-related distress.

**PREDEFINED Q AND A**

## 1. How long have you had the cough?

* *A cough lasting more than 8 weeks is considered chronic and warrants further evaluation.*

## 2. Is the cough dry or productive?

* *Knowing if you bring up mucus (and its color) helps identify causes like infection or bronchitis.*

## 3. Do you cough up blood (hemoptysis)?

* *Coughing up blood requires urgent investigation to rule out serious conditions.*

## 4. Are there any associated symptoms?

* *Such as fever, night sweats, weight loss, shortness of breath, or chest pain.*

## 5. What triggers or worsens your cough?

* *Exposure to dust, smoke, allergens, cold air, exercise, or certain positions.*

## 6. Have you been diagnosed with respiratory conditions like asthma, COPD, or allergies?

* *This helps identify underlying chronic diseases contributing to cough.*

## 7. Are you taking any medications, including ACE inhibitors?

* *Some drugs can cause chronic cough as a side effect.*

## 8. Do you smoke or vape?

* *Smoking is a common cause of chronic cough and worsens lung health.*

## 9. Have you had recent respiratory infections?

* *Post-infectious cough can persist for weeks after a cold or bronchitis.*

## 10. Do you have symptoms of acid reflux or heartburn?

* *GERD is a frequent cause of chronic cough.*

## 11. Have you noticed any changes in your voice or difficulty swallowing?

* *These may indicate upper airway or neurological causes.*

## 12. Have you had any recent imaging or lung function tests?

* *Helps assess for structural lung disease or airway obstruction.*

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand you’ve been experiencing a cough that’s lasted for quite some time. Can you tell me how long you’ve had this cough?

Patient: It’s been going on for about three months now. It just won’t go away.

Doctor: Thank you. Is your cough dry, or do you bring up any mucus?

Patient: It’s mostly dry, but sometimes I do cough up a little phlegm.

Doctor: Have you noticed any other symptoms like shortness of breath, wheezing, chest pain, or coughing up blood?

Patient: No blood, but I do feel a bit short of breath sometimes, especially when I’m active.

Doctor: Do you have any history of asthma, allergies, or acid reflux? Also, are you currently taking any medications, like blood pressure medicines?

Patient: I have mild seasonal allergies, and I started an ACE inhibitor for my blood pressure about six months ago.

Doctor: That’s helpful to know. ACE inhibitors can sometimes cause a persistent dry cough. Have you noticed if the cough started after you began that medication?

Patient: Yes, it started a couple of months after I began the medication.

Doctor: That could be the cause. We can consider stopping or switching your blood pressure medicine to see if your cough improves. In the meantime, I’d like to examine your chest and listen to your lungs.

*(Doctor performs examination)*

Doctor: Your lungs sound clear, which is a good sign. I’d also like to order a chest X-ray and possibly lung function tests to rule out other causes.

Patient: What else could be causing this cough?

Doctor: Common causes include postnasal drip from allergies, asthma, or acid reflux. Sometimes, infections or other lung conditions can cause a chronic cough. We’ll investigate and treat accordingly.

Patient: When should I come back to see you?

Doctor: Let’s schedule a follow-up in about four weeks to review your test results and see how you’re doing after any medication changes. If your cough worsens, you develop fever, or start coughing up blood, please come in sooner.

Patient: Thank you, doctor. That helps me understand what’s going on.

Doctor: You’re welcome. Feel free to call if you have any questions before your appointment.

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## **congenital lung abnormalities**

## **Alternative Names for Congenital Lung Abnormalities**

* Congenital Pulmonary Malformations (CPM)  
  A broad term encompassing various structural lung defects present at birth.
* Congenital Pulmonary Airway Malformation (CPAM)  
  Formerly known as Congenital Cystic Adenomatoid Malformation (CCAM), a common cystic lung lesion.
* Congenital Lung Malformations  
  General term for developmental anomalies of the lung.
* Congenital Lung Lesions  
  Refers to any abnormal mass or cystic lesion detected in the lung from birth.
* Congenital Pulmonary Sequestration  
  A specific type of congenital lung anomaly involving non-functioning lung tissue with aberrant blood supply.
* Bronchopulmonary Foregut Malformations  
  Includes abnormalities involving both lung and foregut structures.
* Pulmonary Hypoplasia  
  Underdevelopment of lung tissue present at birth.
* Congenital Lobar Emphysema  
  Overinflation of a lung lobe causing respiratory distress.
* Congenital Cystic Lung Disease  
  Describes cystic abnormalities in the lung present from birth.

**DEFINITION AND DESCRIPTION**

Congenital lung abnormalities are lung problems from birth. They happen when tissues in the lungs or airways haven’t developed properly during pregnancy. What causes them isn’t known.

There are several different types of congenital lung abnormality. For example, there could be a problem with:

the lungs

the airways

the blood supply to the lungs or the airways.

The most common type of congenital lung abnormality is congenital pulmonary airway malformations or CPAM. CPAM affects the lungs and airways.

## **congenital pulmonary airway malformations (CPAM)**

Congenital pulmonary airway malformations (CPAM) are abnormal areas of tissue (lesions) on the lung. They happen when the airway and surrounding lung tissue does not develop properly.

**Congenital** = something the baby is born with

**Pulmonary** = affects the lungs

CPAM are the most common type of congenital lung abnormality. Most babies with CPAM can be delivered normally and will not have any obvious symptoms after they are born. CPAM are usually detected before your baby is born, at the 20-week prenatal scan. They often get larger during the last trimester of pregnancy, before shrinking again before birth.

There are different types of CPAM. A single CPAM can be ‘hybrid’, meaning having multiple different tissues in it. Doctors will not know for certain exactly which type of CPAM your baby is affected by unless the decision is to remove it by an operation after birth.

At some time after birth (timing will vary between different hospitals), your child will have a scan to find out more about the CPAM. A chest CT scan with an injection of dye will show the abnormality. A type of dye called contrast is injected to help the blood vessels show up on the scan. The injection will only last a few seconds, and the scan itself is painless.

Different types of CPAM that may show up on a chest CT scan include:

**bronchogenic cysts**: this is when pockets of tissue called cysts develop on the bronchial tree (the system of airways called bronchi and bronchioles)

**bronchial atresia (absent bronchus):** this is when one or more airways at the very end of the bronchial tree (peripheral bronchi) are missing and the airway beyond the blockage swells by ongoing production of lung fluid

**bronchopulmonary sequestration:** this is when a mass of abnormal lung tissue develops next to the lung, or inside one lung. It’s usually solid rather than air filled and doesn’t function or connect with the rest of the breathing system

**congenital cystic adenomatous malformation (CCAM):** this is when pockets of tissue called cysts develop in one or more sections (lobes) of the lung

**congenital lobar overinflation (CLO):** this is when one of the lobes of the lung gets overinflated. This squashes and moves the lung tissues next to it and causes breathing difficulties. It’s sometimes known as congenital lobar emphysema (CLE) or congenital large hyperlucent lobe (CLHL).

**foregut duplication cyst:** this is when during pregnancy, some parts of the unborn baby’s lung develop abnormally. At first, in the very early stages of development, the lung bud (which goes on to form the lungs) grows out of what will become the gut. This means that some lung cystic abnormalities look like parts of the intestine when removed and examined under a microscope.

### **CPAM or cancer**

A rare cancer of young children known as pleuropulmonary blastoma (PPB) can sometimes look like a CPAM. If a CPAM has been diagnosed at your prenatal scan, it’s very unlikely to be PPB.

Once your baby is born, if doctors find a CPAM with a pneumothorax (collapsed lung), doctors will consider this very rare diagnosis and carry out tests to rule out or confirm PPB. Other cancers can happen in CPAMs, but these are very rare, and you will need to discuss this with your pediatrician.

## **CPAM and other congenital lung abnormalities**

Congenital lung abnormalities are rare. Researchers estimate they may affect about **one in every 10,000** babies born.

There are many different types of congenital lung abnormality. Each type is very rare. We don’t have accurate figures for how common each abnormality is, but CPAM is the most common.

## **causes congenital lung abnormalities**

It’s not known what causes congenital lung problems.

Some congenital lung abnormalities happen together with other congenital abnormalities, such as heart problems.

Researchers are working to understand more about how congenital lung abnormalities happen. In some cases, they have identified faulty genes and processes that cause the lungs to develop abnormally.

## **symptoms of congenital lung abnormalities**

Children’s symptoms will vary depending on the condition. Congenital lung abnormalities may be found before birth, in a newborn baby or later in life.

### **Signs before birth**

In some babies, congenital lung abnormalities may be picked up on antenatal scans before they are born. This is usually the case with CPAM.

### **Signs in new-born babies**

Most babies with CPAM have no obvious signs of chest problems.

Some babies with other congenital lung abnormalities will have breathing problems as soon as they are born. Symptoms can include:

noisy breathing, known as stridor

a blue colour on your skin, lips, tongue, or gums (cyanosis).

Sometimes a baby may have significant breathing difficulties and may need help with their breathing. This might include ventilation (a breathing machine) or taking oxygen through a face mask or tubes in their nose.

### **Signs in older children and adults**

Most children with CPAM will not experience symptoms.

In older children and adults, symptoms of congenital lung abnormalities can include:

infections that keep coming back in the affected area of the lung

bronchiectasis

asthma-like symptoms (coughing, wheezing, breathlessness) that do not respond to asthma treatment.

## **congenital lung abnormalities diagnosed**

Lung problems can sometimes show up on an ultrasound scan when the baby is still in the womb. This is usually the case with CPAM.

For children, doctors might use a bronchoscopy (a camera test to look inside the airways) if they think your child may have an upper airway abnormality.

## **Treatment**

Babies and children with CPAM and other congenital lung abnormalities will usually go to specialist centers to investigate and manage their condition. If the abnormality is found on an antenatal scan, the pregnancy will be monitored, and arrangements may be made to deliver the baby in a specialist center.

### **Treatment before birth (antenatal treatment)**

Very rarely babies need treatment before they’re born - this is called antenatal treatment. For example, in unborn babies with a bronchopulmonary sequestration, doctors might block the abnormal vessels while they are still in the womb.

### **Treatment after birth**

Treatment for CPAM after your child is born will depend on their condition and if they have symptoms.

A few babies with congenital lung abnormalities may need emergency surgery when they are born. Others may have an operation because of features shown on a CT scan or following any complications that may develop after they’re born.

If your child has symptoms, the CPAM should be removed surgically.

As your child grows up, if they do not have any symptoms, they may not need immediate treatment and will have regular check-ups to monitor their condition.

Some children may be offered surgery before any symptoms show to remove the CPAM. This may be to try and reduce infections that keep coming back. You should be involved in making the decision with the health care professional. Policies on surgery before any symptoms show differ between treatment centers, and there is not enough evidence to help decide which CPAMs are at risk of complications. You should ask your pediatrician to explain the risks and benefits of surgery.

Other than surgery, your child will need treatment for any complications that happen related to their congenital lung abnormality. For example, they’ll need antibiotics if there is an infection in the affected area. However, once a CPAM gets infected, it likely needs to be removed surgically to stop repeat infections.

## **Long-term outlook and complications of CPAM**

Some people live with congenital lung abnormalities for a long time without having any symptoms at all. Their condition might be discovered by chance when they have a chest X-ray (or especially in adults, a CT scan) for a different reason.

The long-term outlook and any long-term complications for CPAM and other congenital lung abnormalities will depend on the type of condition your child has. Generally, when CPAM is diagnosed prenatally (before your child is born), the long-term outlook is good. However, a few types of CPAM (around 5%) can develop complications and it is not known which. **You should speak to your doctor or nurse for advice specific to your child’s condition.**

**The 5 Stocker classifications of CCAM (CPAM) are as follows:**

* Type 0: the rarest form, 1-3% of cases, arises from the trachea or bronchus, presentation is severe and usually lethal, birth presentation, cysts of 0.5 cm with ciliated pseudostratified epithelial lining and goblet cells, bronchiolar cartilage present, all lobes involved.
* Type I: 50–70% of cases, arising from the distal bronchus or proximal bronchiole, a small number of large echolucent cysts measuring 3–10 cm or a single dominant, cyst walls are thin and lined with a ciliated pseudostratified epithelium with bronchiolar differentiation, mucinous cells in 33% and cartilage in 10% of cases, only one lobe involved.
* Type II: 15–30% of cases, arise from terminal bronchioles, first month of life presentation, multiple spaced cysts of 0.5-2.5 cm (sponge-like appearance) as well as solid area, ciliated cuboidal or columnar epithelial lining, absence of mucinous cells and cartilage, highest incidence of associated anomalies at up to 60%, involvement of usually only one lobe.
* Type III: 5–10% of cases, in uterus or birth respiratory distress, bulky firm mass with adenomatoid appearance and cysts of 1.5 cm, ciliated cuboidal epithelial lining, absence of mucous cells and cartilage, involvement of usually only one lobe or one lung, associated with a poorer prognosis, microcystic.
* Type IV: 5–15% of cases, newborn respiratory distress, pneumothorax, pneumonia or incidental finding, peripheral cysts with acinar-alveolar epithelial differentiation, cysts as large as 10 cm and associated with malignancy, specifically pleuropulmonary blastoma, alveolar in origin, antenatally classified as microcystic (< 5 mm).

## **CPAM Differential Diagnosis List**

* Bronchopulmonary Sequestration (BPS)  
  Non-functioning lung tissue with systemic arterial supply (from aorta), no connection to bronchial tree; may coexist as hybrid lesions with CPAM features.
* Congenital Diaphragmatic Hernia (CDH)  
  Herniation of abdominal contents (bowel loops) into the thoracic cavity; identified by presence of bowel in chest on imaging.
* Congenital Lobar Emphysema (CLE) (Congenital Lobar Overinflation)  
  Overinflation of a lung lobe causing hyperlucency without cystic or solid masses.
* Bronchogenic Cyst  
  Usually unilocular, fluid-filled cysts with no communication to the bronchial tree; lined by respiratory epithelium and containing cartilage.
* Localized Congenital Cystic Bronchiectasis  
  Cystic dilation of bronchi due to mucus retention or infection; may mimic CPAM on imaging.
* Pulmonary Hypoplasia  
  Underdevelopment of lung tissue, often associated with other anomalies.
* Pleuropulmonary Blastoma (PPB)  
  Rare malignant cystic lung tumor in children; may resemble CPAM but has malignant histology.
* Other Cystic Lung Lesions
  + Cystic teratoma
  + Enteric duplication cyst
  + Pulmonary cystic lymphangioma

**EPIDEMIOLOGY**

Though uncommon overall, about 95% of congenital cystic lung disease is accounted for by congenital pulmonary airway malformation. The incidence of CPAM is reported as 1 in 10000 to 1 in 35000 births. There have been some studies that show male predominance in lesions that present in early infancy. Overall, these malformations occur sporadically with no genetic predisposition (except Type 4), and no association with maternal factors

**GENOMIC DATA**

* Oligogenic and Heterogeneous Mutations:  
  CPAM appears to be caused by mutations in multiple genes involved in lung development and cancer pathways. Studies found an excess of damaging variants in genes linked to lung carcinoma and developmental lung abnormalities, suggesting CPAM is an oligogenic disorder with variable phenotypes depending on gene interactions.
* Cancer-Associated Genes:  
  Several CPAM patients carry damaging mutations in genes commonly mutated in lung adenocarcinomas and other cancers, such as *KRAS*, *TP53*, *FGFR2*, and *LRP2*. This genetic overlap may explain the reported association between CPAM and later development of malignancies like mucinous adenocarcinoma or pleuropulmonary blastoma.
* Somatic KRAS Mutations:  
  Somatic activating mutations in *KRAS* have been identified particularly in CPAM Type 1 lesions, primarily in mucinous and nonmucinous epithelial cells. These mutations may drive abnormal airway epithelial proliferation and are considered markers of potential malignant transformation.
* DICER1 and Related Syndromes:  
  While *DICER1* mutations are linked to pleuropulmonary blastoma (a rare lung tumor sometimes confused with CPAM), recent studies found no pathogenic *DICER1* variants in many CPAM tissue samples, suggesting distinct genetic mechanisms.
* Developmental Pathways and Immune Response:  
  Transcriptomic analyses show upregulation of genes involved in primary cilium development and ciliopathies, and downregulation of immune pathways (e.g., antigen presentation, interferon gamma response), indicating disrupted airway epithelial differentiation and suppressed innate immunity in CPAM tissue.
* Mosaic RASopathies Concept:  
  CPAM may represent a form of mosaic RASopathy, where somatic mutations in the RAS-MAPK pathway (including *KRAS*) cause localized developmental lung anomalies with potential for neoplastic progression

**PROGNOSIS**

The overall prognosis for congenital pulmonary airway malformation, when diagnosed prenatally, is excellent. There have been several cases that report prenatal regression of the lesion. If fetal hydrops is present, the survival rate drops. Surgical resection has been shown to increase survival rates and has proved to be curative in the neonatal period. There have been studies, however, that show conservative management, in selected cases, to be equally effective. Out of the different types per the Stocker classification, Type 1 is shown to have the best prognosis. Type 2’s prognosis depends on the severity of the associated anomalies. Type 3 often has hypoplasia of an entire lobe leading to complications such as pulmonary hypertension. Type 4 has an excellent prognosis with surgical resection but does have a strong correlation with pleuropulmonary blastoma

**Doctor-patient conversation about Congenital Pulmonary Airway Malformation (CPAM)**

Doctor: Hello, I’d like to talk with you about your baby’s lung condition called Congenital Pulmonary Airway Malformation, or CPAM. This is a rare lung abnormality that develops before birth.

Parent: What exactly is CPAM? Is it serious?

Doctor: CPAM is a benign mass or cystic lesion made up of abnormal lung tissue that doesn’t work properly. It usually affects one lobe of the lung. Many babies with CPAM do well, especially if the lesion is small, but some may have breathing difficulties or infections depending on the size and location of the lesion.

Parent: What causes CPAM?

Doctor: We don’t know the exact cause. It happens during lung development in the womb and is not inherited or related to anything the mother did during pregnancy. It occurs in about 1 in 25,000 pregnancies.

Parent: How is CPAM diagnosed?

Doctor: Often, CPAM is detected during routine prenatal ultrasounds. Sometimes we use fetal MRI to get more detail. After birth, we confirm the diagnosis with imaging like chest X-rays and CT scans.

Parent: What treatments are available?

Doctor: If the lesion is small and your baby has no symptoms, we may monitor closely. Surgery to remove the affected lung tissue is usually recommended when the baby is around 1 to 3 months old. Early surgery helps the remaining lung grow and reduces risks of infections or rare complications like cancer.

Parent: What happens during surgery? Is it risky?

Doctor: Surgery involves removing the affected lobe of the lung. It’s typically done through a minimally invasive approach and most babies stay in the hospital for 2 to 5 days afterward. The risks are low, and most babies recover well with normal lung function as they grow.

Parent: Will my baby need long-term follow-up?

Doctor: Most babies do well after surgery and only need follow-up for about a year to ensure normal lung growth. Some babies with larger or more complex lesions may need longer monitoring. We have specialized programs to support children with lung growth issues.

Parent: What should I watch for after birth?

Doctor: Watch for signs of breathing difficulty, persistent cough, or infections. If your baby has trouble feeding or seems unusually tired, please seek medical attention promptly.

Parent: Thank you, doctor. This helps me understand what to expect.

Doctor: You’re welcome. We’ll work closely with you to provide the best care for your baby. Please feel free to ask any questions at any time.

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### **Tracheomalacia**

## **Alternative Names for Tracheomalacia**

* Congenital Tracheomalacia  
  When the condition is present from birth.
* Acquired Tracheomalacia  
  When tracheomalacia develops later due to injury, inflammation, or external compression.
* Tracheal Cartilage Softening  
  Describes the underlying pathology of weakened tracheal cartilage.
* Dynamic Tracheal Collapse  
  Refers to the airway collapse during breathing due to tracheal wall weakness.
* Tracheal Collapse Syndrome  
  Emphasizes the functional consequence of the malacia.
* Airway Malacia  
  A broader term that includes tracheomalacia and bronchomalacia (softening of the bronchi).
* Tracheobronchomalacia (TBM)  
  When both the trachea and bronchi are involved.

**DEFINITION AND DESCRIPTION**

Tracheomalacia (*TRAY-kee-oh-muh-LAY-shia*) is when you have weak or floppy cartilage in your trachea (windpipe). The walls of your windpipe can collapse or fall in, causing symptoms like high-pitched breathing. It can also trap mucus in your lungs, making it difficult to clear them out. In severe cases, tracheomalacia may be life-threatening, but it’s curable with treatment.

Tracheomalacia usually affects newborns, but anyone can develop it. Babies born with the condition often develop symptoms when they’re 1 to 2 months old. Then, symptoms improve over the first three years of life. That’s because cartilage strengthens as your baby’s windpipe grows. But severe cases may need surgery.

#### **Types of this condition**

There are two types of tracheomalacia:

* **Congenital tracheomalacia**: “Congenital” means you’re born with the condition. It happens when a baby’s windpipe didn’t form properly during fetal development.
* **Acquired tracheomalacia**: “Acquired” means the condition developed after you were born. It happens when your windpipe breaks down or gets damaged. Injuries, surgeries or prolonged mechanical ventilation can cause this. Although it’s uncommon, acquired tracheomalacia can occur at any age.

Some people with tracheomalacia also have weak bronchi (the tubes that run from your windpipe to your lungs). Healthcare providers call this condition tracheobronchomalacia.

Congenital tracheomalacia is somewhat rare. Even so, it’s the most common birth defect affecting the windpipe. Approximately 1 in 2,100 children are born with the condition.

Acquired tracheomalacia (which can occur at any age) is also very uncommon.

### **causes tracheomalacia**

Congenital tracheomalacia happens when the cartilage in your baby’s windpipe doesn’t develop the way it should. The walls of their windpipe are floppy instead of rigid.

Acquired tracheomalacia causes include:

* Chronic acid reflux (GERD).
* Damage from surgery or other medical procedures.
* Emphysema.
* Polychondritis (inflammation of the cartilage) in your windpipe.
* Tracheoesophageal fistula repair (fixing an abnormal connection between your esophagus and trachea).
* Tracheostomy, or long-term use of a breathing tube.
* Upper respiratory infections, like bronchitis.

Tracheomalacia may be associated with other conditions like:

* Developmental delays.
* Ehlers-Danlos syndrome.
* Heart defects.

### **Risk Factors**

* **Age:** Tracheomalacia is more common in infants and young children, particularly those born prematurely.
* **Gender:** Some studies suggest a higher prevalence in males.
* **Geographic Location:** Exposure to environmental pollutants may vary by location, influencing the risk.
* **Underlying Conditions:** Individuals with chronic respiratory diseases, such as asthma or chronic obstructive pulmonary disease (COPD), may be at higher risk.

### **symptoms of tracheomalacia**

The most common tracheomalacia symptom is high-pitched or noisy breathing (stridor). Other symptoms include:

* Breathing issues that get worse when feeding, crying or coughing.
* Choking.
* Chronic coughing.
* Cyanosis (a condition where your skin, lips or nails turn blue from a lack of oxygen).
* Difficulty swallowing.
* Hoarseness.
* Recurring (returning) airway infections like pneumonia or bronchitis.
* Shortness of breath.
* Wheezing or rattling sounds.

## **Diagnosis and Tests**

A healthcare provider will do a physical exam and ask about your symptoms. They’ll also use laryngoscopy or bronchoscopy to look at your windpipe. To do this, they’ll guide a lighted scope with a camera down your throat.

Your healthcare provider may need to run more tests to diagnose tracheomalacia, like:

* Airway fluoroscopy.
* Barium swallow (esophagram).
* Chest X-ray.
* CT (computed tomography) scan.
* Lung function tests.
* MRI (magnetic resonance imaging).

## **Management and Treatment**

Healthcare providers can treat tracheomalacia with nonsurgical therapies, medications or surgery. What’s right for you depends on the extent of the condition.

#### **Nonsurgical therapies**

The following treatments help keep your airways open and your lungs clear:

* **Breathing humidified air**. Using a humidifier can thin out mucus so you can breathe comfortably.
* **Chest physical therapy**. A physical therapist can teach you breathing exercises to clear mucus from your lungs.
* **Continuous positive airway pressure (CPAP)**. Using a CPAP machine can keep your windpipe from collapsing too much during sleep.

#### **Medications**

Healthcare providers use several different medications to treat tracheomalacia, including:

* **Antibiotics** to treat bacterial infections.
* **Bronchodilators** to relax the muscles that help you breathe.
* **Corticosteroids** to reduce inflammation.
* **Mucolytics** to thin out mucus.

#### **Surgery**

Severe tracheomalacia cases may need surgery. Some options include:

* **Aortopexy**. A surgeon moves your aorta up and away from your airway and attaches it to your breastbone. This keeps your windpipe from collapsing.
* **Stenting**. This involves placing a stent (hollow tube) in your airway to keep it open.
* **Tracheopexy**. A surgeon connects part of your windpipe to a nearby structure (like your breastbone or a ligament at the top of your spine). This keeps your windpipe open and prevents collapse.

### **complications of tracheomalacia**

Without treatment, tracheomalacia can cause:

* Airway obstruction.
* Aspiration pneumonia.
* Growth faltering.
* Upper respiratory infections that linger or come back often

## **Outlook / Prognosis**

After treatment, most people with tracheomalacia go on to live healthy lives with no complications.

Treatment isn’t always necessary. Congenital tracheomalacia usually improves on its own by age 3. As your baby’s tracheal cartilage grows stronger, their symptoms will likely improve. But in some cases, your baby may need medication and/or surgery.

People with tracheomalacia need close monitoring if they develop respiratory infections. Even minor colds can cause complications. Your healthcare provider may recommend treatments or medications to manage your symptoms.

**Prevention**

There’s nothing you can do to reduce your risk for tracheomalacia. But you can manage it with the help of your healthcare provider.

**When should I see my healthcare provider?**

Schedule an appointment with your healthcare provider if your baby shows tracheomalacia symptoms. This includes things like frequent cough, noisy breathing or prolonged respiratory infections.

Tracheomalacia in adults can cause exercise intolerance and frequent respiratory infections. Tell your provider if you notice these things. They can confirm the diagnosis and recommend appropriate treatment.

## **Diagnostic Considerations**

The differential diagnosis of tracheomalacia includes laryngomalacia, subglottic stenosis, congenital cysts, vocal cord paralysis, and hypocalcemic tetany. Complications include problems with acute airway obstruction and perioperative morbidity and mortality.

According to a 2005 study by Boogaard, when pediatric pulmonologists diagnosed airway malacia (on the basis of symptoms, history, and lung function) prior to bronchoscopy, a correct diagnosis was made in 74% of the cases.However, in 52% of the diagnoses of airway malacia, the diagnosis was not suspected prior to bronchoscopy. Children with tracheomalacia present with atypical and variable clinical features; considerable overlap occurs with features of allergic asthma

## **Differential Diagnosis for Tracheomalacia**

## Structural Airway Abnormalities

* Laryngomalacia  
  Softening and collapse of supraglottic structures causing inspiratory stridor, often coexisting with tracheomalacia in infants.
* Bronchomalacia  
  Weakness and collapse of bronchial walls, causing wheezing and recurrent infections.
* Vascular Rings and Slings  
  Congenital vascular anomalies (e.g., double aortic arch, pulmonary sling) compressing the trachea externally.
* Tracheal Stenosis  
  Fixed narrowing of the trachea due to congenital or acquired causes (e.g., prolonged intubation, trauma).
* Tracheal Web or Atresia  
  Congenital membrane or absence of tracheal segments causing obstruction.

## Infectious and Inflammatory Causes

* Tracheitis (Bacterial or Viral)  
  Infection causing airway inflammation and edema mimicking airway collapse.
* Granulomatous Diseases  
  Such as tuberculosis or sarcoidosis causing airway narrowing.

## Neuromuscular Disorders

* Conditions causing poor airway muscle tone leading to dynamic airway collapse (e.g., muscular dystrophies).

## Foreign Body Aspiration

* Partial airway obstruction causing wheezing and respiratory distress.

## Tumors or Masses

* Intratracheal or extrinsic masses compressing or invading the airway.

## Chronic Obstructive Pulmonary Disease (COPD) and Emphysema

* In adults, airway collapse due to loss of elastic recoil and cartilage weakening.

## **Epidemiology**

All types of tracheomalacia are extremely rare; no definite incidence rates are available.

In a total of 512 bronchoscopies, airway malacia was diagnosed in 160 children (94 males) at a median age of 4.0 years (range, 0-17 y). Airway malacia was classified as primary in 136 children and as secondary in 24 children. The incidence of primary airway malacia was estimated to be at least 1 in 2100.

**medications used, their purposes, and potential side effects**

Medications Used in Tracheomalacia Treatment

|  |  |  |  |
| --- | --- | --- | --- |
| Medication Type | Examples | Purpose / Mechanism | Common Side Effects |
| Nebulized Medications | Saline solution | Helps humidify airways, loosen mucus, ease cough | Minimal; possible mild irritation |
| Anticholinergic Bronchodilators | Ipratropium bromide (Atrovent) | Opens airways by relaxing bronchial muscles, reduces cough and airway collapse | Dry mouth, throat irritation, cough, dizziness |
| Bronchodilators | Albuterol (short-acting beta-agonist) | Relaxes airway smooth muscle to improve airflow | Tremor, nervousness, palpitations |
| Corticosteroids | Inhaled or systemic steroids (e.g., prednisone) | Reduce airway inflammation that may worsen symptoms | Oral thrush (inhaled), hoarseness, immune suppression (systemic) |
| Antibiotics | Various (if bacterial infection present) | Treat respiratory infections that worsen airway obstruction | GI upset, allergic reactions |

## Additional Non-Drug Therapies

* Humidified air and chest physiotherapy to clear secretions and reduce airway irritation.
* Noninvasive positive pressure ventilation (CPAP or BiPAP) to stent open airways during breathing, especially at night.
* Surgical interventions (for severe cases) such as aortopexy, tracheopexy, tracheostomy, airway stenting, or tracheal reconstruction.

## Important Notes on Medication Use

* Medications like ipratropium bromide and bronchodilators help improve airway tone but do not cure tracheomalacia; they manage symptoms.
* Corticosteroids are used cautiously due to side effects and are typically reserved for cases with significant airway inflammation.
* Treating respiratory infections promptly with antibiotics is critical, as infections can worsen airway collapse.
* Many infants improve as their tracheal cartilage strengthens with growth, reducing the need for long-term medication.

#### **PREDEFINED Q AND A**

## 1. How severe is the condition?

Severity is usually assessed by the degree of airway collapse during breathing, especially exhalation:

* Mild: 25–50% airway lumen narrowing
* Moderate: 50–75% narrowing
* Severe: >75% narrowing, sometimes near complete collapse  
  Severity also depends on symptoms and whether collapse occurs during quiet breathing or only with coughing or agitation. Flexible bronchoscopy is the gold standard for diagnosis and severity assessment, but interpretation can vary between clinicians.

## 2. What caused the condition?

Tracheomalacia can be:

* Congenital: due to underdeveloped or soft tracheal cartilage present from birth
* Acquired: caused by prolonged intubation, trauma, inflammation, external compression (e.g., vascular rings), chronic infections, or chronic obstructive pulmonary disease (COPD) in adults  
  Risk factors include obesity and gastroesophageal reflux disease (GERD).

## 3. What treatment do you recommend?

Treatment depends on severity and symptoms:

* Mild cases often improve with conservative management (humidified air, airway clearance, treating infections)
* Medical therapies may include bronchodilators, corticosteroids, and antibiotics if infections occur
* Noninvasive ventilation (CPAP/BiPAP) can help stent open airways during breathing
* Severe cases may require surgical interventions like aortopexy, tracheopexy, or airway stenting.

## 4. Will surgery be necessary?

Surgery is usually reserved for:

* Severe tracheomalacia causing significant airway obstruction or respiratory distress
* Failure of conservative and medical management
* Associated anatomical abnormalities causing external airway compression  
  The decision depends on clinical symptoms, airway collapse severity, and response to other treatments.

## 5. What can I do to ease symptoms?

* Use humidified air to keep airways moist
* Avoid respiratory irritants like smoke and pollutants
* Promptly treat respiratory infections with antibiotics if needed
* Practice airway clearance techniques as advised
* Use prescribed inhalers or medications properly
* Monitor for worsening symptoms and seek medical attention if breathing difficulty increases.

## 6. How soon do I need treatment?

* Mild cases may only require monitoring and supportive care.
* Moderate to severe cases with symptoms like stridor, recurrent infections, or respiratory distress need prompt evaluation and treatment.
* If symptoms worsen rapidly (e.g., severe breathing difficulty, cyanosis), emergency care is necessary

#### **What conditions might be confused for tracheomalacia?**

The following conditions may be confused for tracheomalacia:

* **Laryngomalacia**: This condition refers to floppy tissue above your voice box. Your voice box sits above your windpipe.
* **Tracheobronchomalacia**: This describes weak bronchi (the tubes that run from your windpipe to your lungs). Some people with tracheomalacia also have tracheobronchomalacia.

**GENOMIC DATA**

* Tracheomalacia can be associated with genetic syndromes and congenital anomalies. It often occurs alongside conditions such as Ehlers-Danlos syndrome, Marfan syndrome, Mounier-Kuhn syndrome, and other connective tissue disorders that affect cartilage strength and elasticity.
* Some cases of congenital tracheomalacia are linked to mutations affecting cartilage development and extracellular matrix proteins, though specific causative genes for isolated tracheomalacia are not well defined.
* A rare autosomal recessive condition involving mutations in the DCHS1 gene (encoding protocadherin-16) has been associated with tracheomalacia among other systemic features like intellectual disability. However, this is not a common cause of isolated tracheomalacia.
* Tracheomalacia frequently coexists with other congenital malformations such as tracheoesophageal fistula, vascular rings, and esophageal atresia, which have complex embryological and genetic backgrounds.
* Most cases of tracheomalacia are considered multifactorial or developmental anomalies rather than single-gene disorders. There is no routine genetic testing for isolated tracheomalacia, and it is often sporadic without clear inheritance patterns

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand your child has been having noisy breathing and some coughing. After reviewing the tests, it looks like your child has a condition called tracheomalacia. Have you heard of this before?

Parent: No, I haven’t. What exactly is tracheomalacia?

Doctor: Tracheomalacia means that the cartilage in the windpipe, or trachea, is softer than usual. This softness can cause the airway to partially collapse when your child breathes out, leading to noisy breathing, coughing, or sometimes difficulty breathing.

Parent: Is this serious? Will it get worse?

Doctor: In many cases, especially in infants and young children, tracheomalacia improves as the cartilage strengthens with growth. Mild to moderate cases often get better over time without surgery. However, if the airway collapse is severe, it can cause breathing difficulties that need closer monitoring or treatment.

Parent: What causes tracheomalacia?

Doctor: It can be present from birth due to how the trachea developed, or it can develop later from injury, infections, or pressure from nearby blood vessels. Sometimes it occurs along with other conditions like vascular rings or certain genetic syndromes.

Parent: What treatments are available?

Doctor: Treatment depends on how severe the symptoms are. For mild cases, we usually recommend supportive care like humidified air, avoiding irritants, and monitoring. If infections occur, they are treated promptly. In more severe cases, medications such as inhalers or breathing support like CPAP might be needed. Surgery is rarely necessary but can be considered if symptoms are severe or don’t improve.

Parent: What should I watch for at home?

Doctor: Watch for signs of increased breathing difficulty, bluish color around lips or face, poor feeding, or frequent respiratory infections. If any of these happen, seek medical care promptly.

Parent: How long will this last?

Doctor: Many children improve by the time they are 1 to 2 years old as their airway cartilage becomes stronger. We will follow your child closely and adjust treatment as needed.

Parent: Thank you, doctor. This helps me understand what’s going on.

Doctor: You’re welcome. Please feel free to ask any questions anytime, and we’ll work together to support your child’s breathing health.

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[Tracheomalacia: Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/24504-tracheomalacia#overview)

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### **Bronchomalacia**

## **Alternative Names for Bronchomalacia**

* Congenital Bronchomalacia  
  When present from birth due to underdeveloped bronchial cartilage.
* Acquired Bronchomalacia  
  Develops later due to inflammation, trauma, or external compression.
* Bronchial Cartilage Softening  
  Describes the underlying pathology of weakened bronchial walls.
* Dynamic Bronchial Collapse  
  Refers to airway collapse during breathing caused by bronchial wall weakness.
* Airway Malacia (Bronchomalacia subtype)  
  A broader term including tracheomalacia and bronchomalacia.
* Bronchial Collapse Syndrome  
  Emphasizes the functional airway obstruction caused by bronchial wall weakness.

**DEFINITION AND DESCRIPTION**

Bronchomalacia is a condition where the cartilage in the bronchi is weak. Your bronchi are flexible but firm airways in your lungs. Normally, c-shaped cartilage structures surround your bronchi like protective shells. These firm structures allow your bronchi to open as your lungs expand and contract. Without their support, your bronchi become narrow, especially during exhalation (when you breathe out).

Bronchomalacia usually affects newborns, especially those who were born prematurely (early) and those with Down syndrome. In rare cases, it affects older children and adults.

### **How does bronchomalacia affect my baby?**

Bronchomalacia can block or narrow the bronchi. When the bronchi are blocked or too narrow, your baby may have breathing problems and difficulty getting enough oxygen. In very severe cases, the condition can be life-threatening.

### **Tracheobronchomalacia (TBM)**

In many cases, people with bronchomalacia also have tracheomalacia, which is a weakness in the tracheal walls. These two conditions together are called tracheobronchomalacia (TBM).

Like bronchomalacia, tracheomalacia can interfere with breathing and is more common in newborns. Tracheomalacia is often linked to a rare birth defect known as esophageal atresia

### **causes bronchomalacia**

Bronchomalacia in newborns, known as primary bronchomalacia, is usually a result of:

* Congenital heart disease.
* Underdeveloped lung cartilage due to being born early.
* Williams-Campbell syndrome: Congenital (inherited) condition that causes weakened cartilage in the bronchi.

Bronchomalacia that isn’t present at birth is known as secondary bronchomalacia, and causes include:

* Chest injury.
* Chronic bronchitis.
* Emphysema.
* Long-term ventilator therapy.
* Tuberculosis (TB).
* Tumors or benign (noncancerous) growths in the lungs.

### **symptoms of bronchomalacia**

If you or your child has bronchomalacia, you may notice:

* Barking cough without phlegm.
* Cyanosis (blue-tinted skin).
* Fatigue.
* Frequent infections like colds and respiratory illnesses.
* Shortness of breath (dyspnea).
* Stridor (noisy, vibrating sound when breathing).
* Wheezing.

### **Is bronchomalacia contagious?**

You cannot catch bronchomalacia from someone else or spread it to others. Bronchomalacia is not caused by upper respiratory infections, influenza (flu) or other contagious illnesses.

## **Diagnosis and Tests**

Your provider will perform a physical examination and listen to your or your child’s lungs. If appropriate, they’ll order additional testing or place additional referrals.

Imaging tests allow your provider to see inside the lungs. You or your baby may need:

* Bronchoscopy.
* CT scan.
* MRI.
* X-ray.

## **Management and Treatment**

Newborns with mild bronchomalacia may only need regular check-ins with their healthcare provider. The condition often resolves on its own by 6 months of age as their lungs mature.

If bronchomalacia is interfering with your baby’s oxygen levels, providers may recommend a continuous positive airway pressure (CPAP) device. A CPAP device is a mask that goes over your baby’s nose and mouth and connects to a machine. The machine delivers gentle, continuous air pressure to the mask. The air pressure can help keep the bronchi open. Your baby may need this device for a few weeks or until their lungs have fully developed.

In very severe cases of bronchomalacia and respiratory failure, your baby may need a ventilator to keep their bronchi open and help them breathe.

Bronchomalacia treatment in adults depends on the cause. If you have emphysema or chronic bronchitis, you may need medications and regular checkups to help manage your symptoms. A CPAP device may also be helpful, especially when sleeping. In severe cases, patients may need ventilator support in a hospital.

**Outlook / Prognosis**

Many babies with bronchomalacia recover and thrive once their lung cartilage has matured and hardened. If the condition is caused by a blockage like a tumor, removing the blockage usually cures bronchomalacia.

In adults with emphysema or chronic bronchitis, bronchomalacia usually requires ongoing care and can’t be cured. However, your healthcare provider can help you manage symptoms and feel better.

## **Prevention**

There’s no known way to prevent bronchomalacia in newborns. It is not a result of something you did during pregnancy. If you’re pregnant, learn the signs of premature labor so you can seek medical care right away. Prompt care may prevent your baby from being born too early.

Because bronchomalacia in adults is often a result of lung disease, good lung care may help prevent it. Adults can take these steps to keep their lungs as healthy as possible:

* **Don’t smoke or vape.** Smoking and vaping raise the risk of chronic bronchitis, emphysema and lung cancer. If you do smoke and need help quitting, talk to your provider.
* **Get your home tested for radon.** You can’t see or smell radon gas in your home, but it can cause lung cancer.
* **Wear personal protective equipment (PPE)**. A respirator should be worn when you’re working with chemicals or particles like gases, vapors, fumes, dust and soot.

### **When should I see my healthcare provider?**

See your provider regularly for well checkups and to discuss any changes to your health. Seek emergency care if you or your child has:

* Bluish lips or skin.
* Chest retractions (pulling inward with breathing).
* Fainting or inability to wake.
* Shortness of breath or trouble breathing.
* Wheezing or stridor.

## **Differential Diagnosis for Bronchomalacia**

* Primary Bronchomalacia (Congenital)  
  Due to deficiency or weakness of cartilaginous rings in the bronchi, typically presenting in infants with wheezing and recurrent respiratory infections.
* Secondary Bronchomalacia (Acquired)  
  Caused by extrinsic compression from vascular anomalies (e.g., vascular rings), bronchogenic cysts, inflammation, trauma, or chronic infections.
* Tracheomalacia / Tracheobronchomalacia (TBM)  
  Involvement of the trachea along with bronchi; symptoms and airway collapse overlap with bronchomalacia.
* Asthma  
  Reversible airway obstruction causing wheezing and cough, often confused with bronchomalacia in infants.
* Chronic Obstructive Pulmonary Disease (COPD)  
  In adults, airway collapse and obstruction can mimic bronchomalacia symptoms.
* Bronchogenic Cyst  
  Congenital cystic lesion causing airway compression and symptoms similar to bronchomalacia.
* Vascular Rings and Slings  
  Congenital vascular anomalies compressing the airway externally, causing wheezing and respiratory distress.
* Foreign Body Aspiration  
  Partial airway obstruction causing wheezing and cough.
* Tracheal or Bronchial Stenosis  
  Fixed airway narrowing due to congenital or acquired causes.
* Laryngomalacia  
  Softening of the laryngeal structures causing stridor, often coexisting with bronchomalacia.
* Recurrent Respiratory Infections  
  May cause airway inflammation and mimic symptoms of bronchomalacia.

**Epidemiology of Bronchomalacia:**

* In a large study of 459 children with airway malacia, bronchomalacia (BM) was the most common type, present in 94.8% of cases. It affected the right lung in 53.3% of patients, the left lung in 11%, and both lungs in 35.6%.
* The condition is more frequently diagnosed in male children aged 2 years or younger, consistent with other studies showing a male predominance and early childhood onset.
* The incidence of primary airway malacia (including bronchomalacia) is estimated to be at least 1 in 2,100 children in the general population.
* Among children undergoing bronchoscopy for respiratory symptoms, airway malacia (including bronchomalacia) is diagnosed in approximately 1% to 4.5% of cases.
* Bronchomalacia often coexists with tracheomalacia or tracheobronchomalacia, and its clinical presentation can overlap with other respiratory conditions such as asthma or recurrent pneumonia.
* Severity varies, with most patients showing mild to moderate malacia; severe cases are less common but associated with more frequent and severe respiratory infections

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand you or your child has been experiencing wheezing and difficulty breathing. After reviewing the tests, it appears there is a condition called bronchomalacia. Are you familiar with this term?

Patient/Parent: No, I haven’t heard of bronchomalacia before. What does it mean?

Doctor: Bronchomalacia means that the cartilage in the bronchi—the airways inside the lungs—is softer and weaker than normal. This causes the airways to narrow or collapse, especially when breathing out, which can lead to wheezing, coughing, and sometimes trouble breathing.

Patient/Parent: Is this a serious condition? Will it get worse?

Doctor: It depends on the severity. Many infants, especially those born prematurely, have mild bronchomalacia that improves as they grow and their airways strengthen. In adults or severe cases, it might cause more persistent symptoms and require treatment.

Patient/Parent: What causes bronchomalacia?

Doctor: It can be congenital, meaning present from birth due to how the airways developed, or acquired later from infections, inflammation, or pressure from nearby blood vessels or masses.

Patient/Parent: How do you treat it?

Doctor: Treatment focuses on managing symptoms and preventing infections. Mild cases may only need supportive care like humidified air and monitoring. In some cases, inhalers or breathing support devices like CPAP can help. Surgery is rarely needed but may be considered for severe cases.

Patient/Parent: What should I watch for?

Doctor: Watch for worsening wheezing, difficulty breathing, frequent respiratory infections, or if you notice any bluish color around the lips or face. If these occur, seek medical attention promptly.

Patient/Parent: How long will this last?

Doctor: Many children improve by age 2 as their airways mature. Adults may have persistent symptoms depending on the cause. We will monitor and adjust treatment as needed.

Patient/Parent: Thank you for explaining. It helps me understand what’s going on.

Doctor: You’re welcome. Please feel free to ask any questions anytime.

REFERENCES

[Bronchomalacia: Definition, Treatment & Causes](https://my.clevelandclinic.org/health/diseases/22771-bronchomalacia#overview)

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### **Laryngomalacia**

## **Alternative Names:**

* Congenital laryngomalacia
* Floppy larynx
* Laryngeal flaccidity
* Infantile laryngomalacia
* Supraglottic laryngomalacia

**DEFINITION AND DESCRIPTION**

Laryngomalacia (*lah-ring-oh-ma-LAY-shia*) is a larynx (voice box) abnormality that can happen in newborn babies. It occurs when weak, floppy tissues above the voice box temporarily fall back over the airway. It’s the most common cause of noisy breathing in babies.

Laryngomalacia sounds like a high-pitched squeak (stridor) when your baby breathes in. It usually isn’t serious. But in severe cases, it can cause breathing and feeding issues, among other complications.

Congenital laryngomalacia (meaning you’re born with it) is common in infants. Over half of all newborn babies have it during the first week of life, and even more develop it when they’re 2 to 4 weeks old.

Laryngomalacia can also occur in adults, but it’s rare. (Healthcare providers refer to this as acquired laryngomalacia.)

### **causes laryngomalacia**

Experts aren’t exactly sure why some babies develop laryngomalacia and others don’t. But they have a few theories about why it happens, in general:

* **Structural abnormalities**: The cartilage or muscles around the voice box form atypically (differently) during fetal development.
* **Neuromuscular disorders**: These include disorders that affect the vocal cord nerves and muscles.
* **GERD**: If acid reflux reaches the voice box, it can cause swelling in the area. This can worsen any structural abnormalities causing laryngomalacia.

#### **Types of laryngomalacia**

Healthcare providers categorize laryngomalacia into three types according to cause:

1. **Type 1**: The mucous membranes of the voice box are too tight or too short.
2. **Type 2**: The upper part of the voice box has excess soft tissue.
3. **Type 3**: An underlying disorder (like GERD or neuromuscular disorder) causes laryngomalacia.

### **symptoms of laryngomalacia**

Laryngomalacia symptoms can range from mild to severe. Loud, noisy or squeaky breathing is the main thing to watch for. This often worsens over the first several months but resolves within a year or two.

Most babies with laryngomalacia have no trouble breathing or feeding, even when their breathing sounds concerning. Breathing usually gets louder when lying down, sleeping, crying or feeding.

Babies with severe laryngomalacia may have these symptoms:

* Apnea (long pauses in breathing).
* Aspiration (pulling food into the lungs).
* Cyanosis (a condition that causes the skin to develop a bluish hue).
* Difficulty swallowing (dysphagia).
* Inability to gain weight.
* “Tugging” or “pulling in” at the neck or chest when breathing.

If your baby shows any of the symptoms listed above, call their pediatrician right away

## **Diagnosis and Tests**

Nasopharyngolaryngoscopy (NPL) is the main test healthcare providers use to diagnose laryngomalacia. An otolaryngologist (ENT) uses a scope with a tiny camera (endoscopy) to view your baby’s voice box. They’ll gently guide the scope into your baby’s nostril and down their throat. Providers can do this routine test in about two to five minutes.

#### **Other laryngomalacia tests**

If your baby has laryngomalacia, their provider may need to run other tests to determine the extent of the condition. These tests may include:

* **Airway fluoroscopy:** This procedure combines X-rays and a contrast agent (like dye) that illuminates affected areas within your baby’s body. Your provider may do this as a swallow study to see how laryngomalacia affects your baby’s swallowing function.
* **Impedance probe:** A healthcare provider inserts a small tube through your baby’s nose and into their esophagus. Then, they use a measuring device to see how much stomach acid reaches your baby’s voice box. Babies who have this procedure usually stay for at least one night in the hospital.
* **Microlaryngoscopy and bronchoscopy (ML&B):** A healthcare provider uses a lighted scope to examine your baby’s trachea (windpipe) and voice box to see what’s causing loud breathing. They’ll do this procedure under general anesthesia.
* **Neck or chest X-rays:** These imaging tests can tell your healthcare provider if your baby has any structural abnormalities that may cause noisy breathing.

## **Management and Treatment**

Most of the time, laryngomalacia goes away on its own within a year or two and the noisy breathing improves over time. In mild cases, you can manage your baby’s symptoms at home. But if your baby has severe laryngomalacia, they might need medication or surgery.

#### **Treatment at home**

If your baby has mild symptoms, you can usually keep an eye on things at home. Laryngomalacia management depends on your baby’s unique situation:

* **If your baby has trouble with feeding**, you may need to feed them more often to make up for lost calories and nutrition. You can also try thickening their formula. (You can do this with infant cereal or over-the-counter thickeners.) This increases the “stickiness” of their food so it’s less likely to come back up into their esophagus.
* **If your baby has breathing difficulties**, your provider might recommend elevating the head of their mattress. This may help open their airway.

Ask your provider about specific ways to manage your baby’s laryngomalacia symptoms.

#### **Medication**

When GERD occurs with laryngomalacia, your baby’s provider may prescribe an anti-reflux medication like a proton pump inhibitor (PPI) or H2 blocker. GERD can worsen swelling associated with laryngomalacia, so it’s important to keep reflux in check if it’s a contributing factor.

#### **Surgery**

Laryngomalacia surgery involves trimming the weak, floppy tissue above your baby’s voice box. This procedure is a supraglottoplasty. An ENT surgeon will do a supraglottoplasty in an operating room while your child is under general anesthesia. Your baby will typically stay overnight in the hospital for observation.

### **How long will it take for my baby to feel better after treatment?**

Anti-reflux medication usually improves symptoms within two weeks. But your baby will probably need to stay on the medication for several weeks or months.

If your baby had laryngomalacia surgery, their breathing may sound worse for a few days. This is normal. It’s due to post-op inflammation (swelling) around their vocal cords. The noisy breathing should gradually improve, with full recovery taking about two weeks.

**Outlook / Prognosis**

Despite the noisy breathing, laryngomalacia is usually not dangerous. While most babies outgrow laryngomalacia, a few will need surgery to correct the issue, especially if they’re having trouble gaining weight or are having severe breathing difficulties. Your healthcare provider can tell you what to expect if your baby receives a diagnosis.

Laryngomalacia usually goes away on its own by age 1 or 2. But you should keep an eye out for severe symptoms like apnea and a bluish color around their lips. These things can cause serious complications.

**Prevention**

You can’t prevent laryngomalacia. But you can manage your baby’s symptoms with treatment.

As a parent, you want to shield your baby from all harm. But laryngomalacia is just something that happens. It doesn’t mean you’ve done something wrong. Although the sounds your child makes may be scary at first, treatment may not be necessary.

**When should I call my baby’s healthcare provider?**

If your baby shows symptoms of laryngomalacia, like noisy breathing, consider scheduling an appointment with your healthcare provider. They can examine your baby and make recommendations for referral to ENT, home care and management.

Call your provider right away if your baby develops sudden symptoms, or if they have GERD.

Head to the nearest emergency room if your baby:

* Stops breathing for more than 10 seconds at a time.
* Has a “tugging” or “pulling in” at the chest or neck when breathing.
* Turns blue around the lips.

## **Differential Diagnoses**

* Congenital Stridor
* Croup
* Hypocalcemia
* Pediatric Airway Foreign Body
* Pediatric Gastroesophageal Reflux
* Pediatric Subglottic Stenosis Surgery
* Respiratory Papillomatosis

Unilateral vocal fold paralysis typically presents after a surgical procedure in the thoracic cavity or the neck, though it can be congenital as well. A hoarse cry is common, and these infants may have difficulties feeding. When bilateral vocal fold paralysis is present, infants typically have biphasic stridor and may require a tracheostomy if there is significant respiratory distress. Flexible fiberoptic laryngoscopy is used to diagnose these conditions.

Laryngeal papillomatosis may cause a hoarse cry with upper airway obstruction, which may present early in infancy, and diagnosis is by either flexible fiberoptic laryngoscopy or direct laryngoscopy and biopsy of the lesions.

Subglottic hemangiomas are a rare cause of stridor, typically expiratory. Hemangiomas in a beard-like distribution are clinically suggestive of subglottic extension or occult subglottic hemangioma. Confirmation is achievable with direct laryngoscopy and bronchoscopy.

Subglottic stenosis is usually congenital in patients in this age group, though it may also result from scarring of the subglottic region following prolonged intubation. Stridor may be heard but does not change with the infant's position.

Tracheomalacia and bronchomalacia may be present along with laryngomalacia. Expiratory airway sounds are generally present. Diagnosis is via bronchoscopy.

A vascular ring is a rare cause of airway obstruction. Feeding difficulties and stridor may be present. Diagnostic confirmation is with a contrasted computed tomography scan of the chest. This condition should be a suspected diagnosis with tracheomalacia seen on bronchoscopy or compression of the esophagus seen on an esophagram.

Foreign body aspiration is a possibility after finding an infant in respiratory distress after being unaccompanied or after ingesting food, causing a choking or coughing event. Diagnosis is suggested with chest x-ray findings and decreased unilateral breath sounds. Bronchoscopy should be performed to diagnose and retrieve the foreign body.

## **Epidemiology**

### United States statistics

Frequency is unknown. Often, the diagnosis is presumed.

### Race-, sex-, and age-related demographics

*Race*

No known race predilection has been reported.

*Sex*

Although previous reports in predominately White populations have reported a male predominance (58-76% of cases), a more recent study of a more ethnically diverse population demonstrated no significant difference between males and females.

*Age*

Although this is a congenital lesion, airway sounds typically begin at age 4-6 weeks. Until that age, inspiratory flow rates may not be high enough to generate the sounds. Symptoms typically peak at age 6-8 months and remit by age 2 years.

Late-onset laryngomalacia may be a distinct entity, which can present after age 2 years.

## **Procedures**

### Laryngoscopy and bronchoscopy

These studies are the best studies used to confirm the diagnosis. However, in an infant with typical inspiratory noises (worse when supine) who has a normal cry and normal growth and development, clinical diagnosis is not unreasonable.

A pediatric pulmonologist or pediatric otorhinolaryngologist may perform flexible laryngoscopy or bronchoscopy. Bronchoscopy under anesthesia has been shown to be more sensitive and specific than bronchoscopy in infants who are awake.

Direct visualization of the airway reveals an omega-shaped epiglottis that prolapses over the larynx during inspiration. Enlarged arytenoid cartilages that prolapse over the larynx during inspiration may also be present.

The International Pediatric Otolaryngology Group recommends that microlaryngotracheobronchoscopy to detect synchronous airway lesions be reserved for patients with severe, progressive, or atypical disease.

## **Drug Treatment**

* Anti-reflux medications are commonly prescribed when GERD coexists with laryngomalacia, as reflux can worsen laryngeal swelling and noisy breathing.
  + Proton pump inhibitors (PPIs) (e.g., omeprazole)
  + H2 receptor blockers (e.g., ranitidine)

These medications help reduce acid reflux, thereby decreasing inflammation and improving symptoms indirectly.

## Side Effects of Anti-reflux Medications:

* PPIs:
  + Headache
  + Diarrhea or constipation
  + Increased risk of respiratory and gastrointestinal infections with prolonged use
  + Possible nutrient malabsorption (e.g., magnesium, calcium, vitamin B12) with long-term use
* H2 blockers:
  + Headache
  + Dizziness
  + Diarrhea or constipation
  + Rarely, confusion or arrhythmias in sensitive individuals

## Surgical Treatment

* Supraglottoplasty is the preferred surgical procedure for severe laryngomalacia. It involves trimming or releasing the floppy supraglottic tissues to open the airway.
* Tracheotomy is rarely performed and reserved for very severe cases or when other treatments fail.

## **Genetic and Genomic Insights:**

* Genetic Mutations and Hereditary Patterns:  
  Congenital laryngomalacia can be caused by genetic mutations (pathogenic variants), some of which may be hereditary. There is evidence supporting autosomal dominant transmission in certain families, indicating a genetic predisposition in some cases.
* Associated Genetic Syndromes:  
  About 8–20% of infants with laryngomalacia have congenital anomalies or genetic syndromes, rising to 40% in severe cases requiring surgery. The most commonly reported genetic disorder associated with laryngomalacia is Down syndrome, where about 50% of affected children have respiratory symptoms including laryngomalacia.
* Genes Implicated:  
  Several genes have been linked to laryngomalacia or related phenotypes, often through studies of mouse models and ClinVar genetic variation databases. Notable genes include:
  + WFS1, NFIX, MECP2, CHD7, ARID1B, KIF1A, PHOX2B, COL2A1, ATP4A, ATP12A, UBE3B, SHOC2 among others.
  + These genes are involved in nervous system development, craniofacial formation, gene regulation, and other biological processes relevant to airway structure and function.

## **PREDEFINED Questions and answers**

### **Laryngomalacia vs. tracheomalacia: What’s the difference?**

Both laryngomalacia and tracheomalacia are conditions affecting the airway. While laryngomalacia refers to floppy tissues above the voice box, the characteristics of tracheomalacia include floppy or weak cartilage of the windpipe, which is below the voice box. Tracheomalacia is far less common — and usually more serious — than laryngomalacia.

### **Can laryngomalacia cause weight gain?**

Not usually. In fact, babies with severe laryngomalacia may struggle to gain weight.

### **What worsens laryngomalacia?**

Lying on their back could make your baby’s laryngomalacia symptoms worse. If you notice that your baby is having difficulty breathing when sleeping on their back, please see your healthcare provider.

Additionally, GERD — which is common in babies with laryngomalacia — may make their symptoms worse.

**DOCTOR PATIENT CONVERSATION**

Doctor: "Good morning. We're here to discuss your child's noisy breathing. Based on the examination and what you've described, I suspect it might be laryngomalacia. Have you heard of that before?"

Parent: "I've heard the term, but I'm not entirely sure what it means. What exactly is it?"

Doctor: "Laryngomalacia is a common condition in infants where the tissues above the voice box are soft and floppy. When your child breathes in, these tissues can collapse slightly into the airway, causing that noisy sound, which we call stridor."

Parent: "So, the noisy breathing is the main thing we should look out for? We first noticed it a couple of weeks after birth."

Doctor: "Yes, inspiratory stridor is the main symptom, and it often appears shortly after birth, typically around two weeks. It can sometimes sound worse when your child is eating, crying, sleeping, or lying down. We often see it worsen between 6 to 8 months of age before gradually improving."

Parent: "Is it serious? Should I be worried?"

Doctor: "In about 90% of cases, laryngomalacia is mild and resolves on its own as the child grows, usually by 18 to 20 months of age. Many children with mild cases thrive despite the noisy breathing. However, we do categorize it as mild, moderate, or severe based on associated symptoms, not just how loud the stridor is."

Parent: "What are those other symptoms that would make it more serious?"

Doctor: "We look for signs like significant breathing difficulties, such as stopping breathing (apneas), episodes where the skin turns blue, or if they need extra oxygen. We also watch for feeding difficulties, such as choking or poor weight gain, as these can indicate a more moderate or severe case. If your child is growing well and doesn't have these other issues, it's likely a mild case."

Parent: "How do you confirm the diagnosis?"

Doctor: "To confirm the diagnosis, we'll refer you to a pediatric ENT specialist. They'll perform a procedure called a flexible laryngoscopy. It involves passing a thin, flexible tube through your child's nose to look at their airway and vocal cords directly. This allows us to see how the laryngeal structures move during breathing and rule out other causes of stridor, like vocal cord paralysis or a laryngeal cyst."

Parent: "And what about treatment? Is there anything we need to do?"

Doctor: "For most mild cases, we'll monitor your child's growth and development closely. Many infants with laryngomalacia also experience gastroesophageal reflux (GERD), which can worsen the noisy breathing. If reflux is a problem, we might prescribe anti-reflux medication to help manage it. In rare, severe cases where there are significant breathing problems or poor weight gain, a surgical procedure called a supraglottoplasty might be considered to trim the floppy tissue and open the airway."

Parent: "So, for now, it's mostly observation and keeping an eye on things?"

Doctor: "Exactly. We'll focus on supportive care, monitoring their feeding, breathing, and growth. We'll schedule follow-up appointments to track their progress. If you notice any of those more severe symptoms we discussed, or have any other concerns, please don't hesitate to contact us immediately

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#### **Chest Wall Deformities**

## Common Alternative Names and Terms:

* Thoracic deformities
* Thoracic cage abnormalities
* Thoracic wall deformities
* Chest cage deformities
* Pectus deformities (when referring specifically to pectus excavatum or pectus carinatum)
* Rib cage deformities
* Chest wall anomalies
* Thoracic skeletal abnormalities

**DEFINITION AND DESCRIPTION**

Chest wall deformities are conditions that cause physical abnormalities of the chest. The two most common types of chest wall deformities are:

Pectus Excavatum

Also known as funnel chest or sunken chest, pectus excavatum causes the chest to be depressed (turn inwards) at the sternum (breastbone). It is the most common form of chest wall deformity. Patients may be born with the condition, or it can develop later in adolescence.

Pectus Carinatum

Also known as pigeon chest or protruding chest, this condition causes the sternum and ribs to protrude. It can be caused by an excessive growth of cartilage. Pectus Carinatum can be present from birth or appear during adolescence, triggered by a growth spurt at puberty. It can also appear following surgery on the heart during which the sternum splits.

* Poland syndrome: Poland syndrome is a rare condition characterized by underdeveloped or absent chest muscles on one side, often accompanied by hand abnormalities.
* Sternal cleft: Sternal cleft is a rare congenital condition where the sternum does not fuse properly during fetal development, resulting in a visible split or gap.
* Rib abnormalities: Abnormalities of the ribs, such as fused ribs, missing ribs, or extra ribs, can contribute to chest wall deformities.

#### **Causes Of Chest Wall Deformities**

The causes of chest wall deformities can vary, including:

* Genetic factors: Certain chest wall deformities, such as pectus excavatum and pectus carinatum, can have a genetic component and may run in families.
* Fetal development: In some cases, chest wall deformities result from disruptions in fetal development, leading to abnormalities in the formation of the sternum, ribs, or chest muscles.
* Connective tissue disorders: Some connective tissue disorders, such as Marfan syndrome or Ehlers-Danlos syndrome, can predispose individuals to chest wall deformities.
* Environmental factors: External factors, such as intrauterine compression or mechanical forces on the chest during fetal development, may contribute to chest wall deformities.

Causes of Pectus Excavatum include:

* Hereditary
* Poland Syndrome
* Marfan syndrome
* Scoliosis
* Rickets

Causes of Pectus Carinatum include:

* Present at birth
* Homocystinuria
* Trisomy 21
* Trisomy 18
* Marfan syndrome
* Multiple lentigines syndrome
* Morquio syndrome
* Osteogenesis imperfecta

#### **How are Chest Wall Deformities Diagnosed?**

Pectus Excavatum

Doctors may use computed tomography (CT) scans to see the chest’s internal structures and the degree of depression. Something called the Haller index is used to measure how severe the depression is. Pulmonary function tests and cardiac evaluations may be performed to understand how the deformity is impacting the patient.

Pectus Carinatum

The condition is usually diagnosed in young people, with a doctor performing a thorough physical examination. They may also take photographs of the chest and take measurements to monitor the condition.

#### **Treatment**

Pectus Excavatum

Surgery is required to treat a sunken chest. The preferred option is the minimally invasive Nuss procedure. It involves making small incisions under the arm pits and another to insert a very small camera. A surgeon implants metal bars underneath the sternum to elevate it. The results of the surgery are immediately felt by the patient. The bars can be removed one or two years later, when the desired bone structure is established.

The Ravitch procedure is an open surgery which involves opening the chest and removing sections of rib cartilage and then raising the sternum. The surgeon may implant struts, which are metal bars, which hold the chest wall in a normal position and reshape the chest. The struts can be removed around 6 months later.

Pectus Carinatum

A non-surgical option is a chest brace, which is custom built for each patient. The brace is worn over a period of time and reduces chest protrusion. In combination with exercise program, the therapy can completely correct the protrusion. Results are usually seen after around 6 months of treatment.

For patients with more severe pectus carinatum, surgery may be required. This involves open surgery during which sections of rib cartilage are removed and the sternum flattened. Patients notice a correction in the deformity immediately after surgery.

**DIFFERENTIAL DIAGNOSIS**

**Pectus deformities**

Marfan syndrome is the hallmark entity of differential diagnosis; however, all related syndromes to pectus deformities are part of differential diagnosis constellation.

**Poland Syndrome**

Differential diagnoses in Poland syndrome highlights include normal breast asymmetry in females, and Swayer-James syndrome and giant bulla in hyper-lucent hemithorax RX setting.

**Sternal Clefts**

The differential diagnosis depends on syndromic association constellation. Highlights are limb body wall complex, amniotic band syndrome, and Cantrell pentalogy.

**Ectopia Cordis**

Differential diagnosis highlights are; trisomy 18 and Turner syndrome.

**Jeunes Syndrome**

Differential diagnosis highlights are; Ellis-Van Creveld syndrome, short rib polydactyly syndrome (I-IV types), Barnes syndrome, and Shwachman-Diamond syndrome.

**Jarcho-Levin syndrome**

Differential diagnosis highlights include spondyloepiphyseal dysplasia, Morquio syndrome, chondrodysplasia, Klippel-Feil syndrome, and short rib polydactyly syndrome

**EPIDEMIOLOGY**

**Pectus Excavatum**

This anomaly represents 90% of chest wall deformities; it is more common in the White race with an incidence of 1 per 400 live births and 5 to 1 male predominance. Pectus excavatum could present as isolated non-familiar deformity; however, familiar non-syndromic positive history is present in up to 40%, highlighting an autosomal dominant inheritance patron. Pectus excavatum deformity can be part of genetic syndromes. Noonan and Marfan are the most important associated syndromes identified by POSSUM (Pictures Of Standard Syndromes and Undiagnosed Malformations version 5.7) and WBDD (Winter-Baraitser Dysmorphology Database version 1.0.14). It is of paramount importance that Poland syndrome rib hypoplasia causes secondary pectus excavatum, which could need modified Ravitch procedure repair. Less than 1% of pectus excavatum cases have an underlying connective tissue disorder. Data shows that 80% of cases get diagnosed at two years of life. Mitral valve prolapse (17%), arrhythmias (15%), and congenital heart disease (2%) are the most important cardiac anomalies related.MASS phenotype is present in two-thirds of pectus excavatum patients.

**Pectus Carinatum**

Is the second most common chest-wall deformity (5 to 6 times less common than pectus excavatum), has an incidence of 0.06% of live births with a 4 to 1 male predominance. Pectus carinatum could present as an isolated disease with a 25% to 30% positive family history, or can be part of genetic syndromes. As pectus excavatum, Marfan and Noonan are the most prevalent associated syndromes. Cardiac anomalies association could exist. Thoracolumbar scoliosis is linked to an underlying connective disorder in 12% to 14% of cases. Chondrogladiolar type is more common (95%). Pectus carinatum may present at any age; however, patients generally request medical attention at puberty.

**Poland Syndrome**

The average incidence is 1 in 20000 to 1 in 30000 live births with male predominance between 2 to 1 and 3 to 1. Unilateral presentation (over 90%) is considered a non-genetic disease. The deformity is most commonly right-sided in men (60% to 75%), females have no side tendency. Familial cases span less than 1% of cases (rare), highlighting bilateral or non-right sided with an equal male-to-female ratio as specific features. The left side anomaly is associated with dextrocardia. Poland syndrome has correlated with; Moebius anomaly, Klippel-Feil syndrome, renal anomalies, leukemia, non-Hodgkin lymphoma, breast cancer, cervical cancer, leiosarcoma, and lung cancer.

**Sternal Clefts**

Sternal clefts are the most common type of sternal defects and represent 0.15% of chest wall deformities. Of these, 67% are superior clefts,19.5% complete clefts,11% inferior clefts, and 2.5% sternal foramen. Sternal clefts incidence is difficult to estimate; however, there are reports of a female predominance of 2 to 1. Sternal clefts can be part of PHACES syndrome (90% female), midline fusion defects, and cavernous hemangiomata (female predominance), chromosome 22 syndromes, and 1 chromosome arrangement problems have been linked to sternal clefts by the POSSUM system. The inferior sternal cleft could be considered a pathognomonic sign of Cantrell pentalogy. The superior sternal cleft is commonly an isolated deformity.

**Ectopia Cordis**

It is an extremely rare chest-wall deformity with an estimated prevalence of 1 of 5.5 to 7.9 per million live births. Ectopia cordis types are thoracic (65% to 90%), thoracoabdominal (7% to 20%), and cervical (2.8 %). Ectopia cordis may occur as an isolated condition; however, it has a strong association with congenital heart diseases or other midline defects. Fallot tetralogy is the most common congenital cardiac defect. Thoracoabdominal ectopia cordis highlights include high indices of left ventricle diverticulum and Cantrell pentalogy association. Omphalocele is the most common abdominal defect associated with ectopia cordis.

**Jeunes Syndrome**

The incidence is 1 per 120000 live births. Most deaths occur in half to three-fourths of cases in the first two years of life. Skeletal chest wall growth arrest is the most serious complication core. Clavicle, limbs, pelvis, renal, hepatic, pancreatic are the main malformations that correlate with Jeune syndrome.

**Jarcho-Levin syndrome**

Epidemiological data are scarce. Spondylothoracic dysostosis incidence is unknown. It has an autosomal recessive inheritance patron linked to chromosome 2q32.1, without DLL3 gene mutation, it does not demonstrate gender predilection. Puerto Rican ancestry history is commonly positive. Most deaths (80 to 100%) occur in infancy; however, reports exist of mild phenotypes cases with a good prognosis.

Spondylocostal dysostosis has a reported prevalence of 0.25 per 10000 births and demonstrates a better prognosis. This phenotype has both dominant and recessive autosomal inheritance, with the DLL3 (chromosome 19) and MESP2 (chromosome 15) gene mutation. Also, MESP2 mutation has links to the ethnic Puerto Rican group. Other anomalies associated with Jarcho Levin syndrome are a short trunk, dwarfism, craniofacial anomalies, short and rigid neck, inguinal and abdominal hernias, urinary tract anomalies, and talipes equinovarus

**PROGNOSIS**

**Pectus Excavatum**

Surgical correction is achieved with open and MARPE surgery with satisfactory results (86% to 98.1%), from a functional standpoint, cardio-pulmonary fitness performance improve at 6 to 12 months after surgery.Mitral valve prolapse related to pectus excavatum resolution occurs in 50 % of cases after surgical repair.

**Pectus Carinatum**

Highlighting that minimally invasive surgery still needs more evidence support and acceptance, pectus carinatum surgical repair results are based on the modified Ravitch repair principles, therefore in several series, outcomes, and prognosis (esthetic satisfaction and symptomatology improvement) homogeneity results between both pectus deformities have been demonstrated. For bracing treatment, anatomical changes can be observed within 2 to 3 months of its use, bracing treatment duration is from 2 to 2.5 years; however, there are reports that the bracing device is poorly tolerated.

**Poland Syndrome**

Prognosis will depend on age, gender, phenotype severity, associated cancer if coexisting, and type of surgical reconstruction if indicated.

**Sternal Clefts**

Prognosis and outcomes will depend on defect phenotype, age, and associated anomalies. Superior sternal clefts commonly present as an isolated feature with an orthotropic hearth without significant intra-cardiac abnormalities, so they have a good prognosis. In general, a better prognosis is achievable with an appropriate surgical repair timing (neonatal period); however, survival will depend more on the associated anomalies than sternal cleft.

**Ectopia Cordis**

Ectopia cordis general prognosis is poor. Thoracic ectopia cordis, complex congenital cardiac defects, and Cantrell pentalogy have the poorest prognosis. Thoracoabdominal ectopia cordis has been reported with a better prognosis due to the non-malrotated ectopic heart.

**Jeunes Syndrome**

Generally, Jeune's syndrome prognosis is poor, and it has a reported mortality rate of 60 to 80%; however, some centers have reported up to a 50% chance of survival with surgery in mild cases. Survival prognosis may improve with age progression.

**Jarcho-Levin syndrome**

Prognosis will depend on phenotype. Spondylothoracic dysostosis generally is considered a lethal condition and surgery has no benefit; however, in some cases series, mild forms have been reported with up to 56% survival with aggressive medical treatment focused on respiratory function.Spondylocostal dysostosis has a better prognosis. VEPTR surgery has a more probable better prognosis. Both phenotypes have normal intellectual development, and the overall prognosis improves if patients reach six months of age.

## **Treatment of Chest Wall Deformities: Drug Information and Side Effects**

## 1. Medical (Drug) Treatment

* Anti-inflammatory or pain medications:  
  Used postoperatively or to manage discomfort related to chest wall deformities or surgery.
  + *Common drugs:* NSAIDs (ibuprofen), acetaminophen
  + *Side effects:* GI upset, kidney effects (NSAIDs), liver toxicity (acetaminophen in overdose)
* Medications for Gastroesophageal Reflux Disease (GERD):  
  GERD can exacerbate symptoms in some patients with chest wall deformities, especially pectus excavatum.
  + *Drugs:* Proton pump inhibitors (PPIs) like omeprazole, H2 blockers
  + *Side effects:* Headache, diarrhea, nutrient malabsorption with long-term use
* Physical therapy and exercise:  
  No drugs involved but important non-pharmacologic adjunct to improve posture and respiratory function.

## 2. Non-Drug Treatments

* Bracing (mainly for pectus carinatum):  
  Custom chest wall braces apply pressure to remodel the protruding sternum over time. No drug side effects but requires compliance.

## 3. Surgical Treatment

Surgery is the mainstay for moderate to severe chest wall deformities, especially pectus excavatum and some cases of pectus carinatum.

* Nuss Procedure (Minimally invasive repair):  
  A curved metal bar is inserted behind the sternum to elevate it. The bar remains for 2-3 years before removal.
  + *Risks/Side effects:*
    - Pain (managed with analgesics)
    - Pneumothorax (collapsed lung)
    - Infection
    - Bar displacement or allergic reaction to metal
    - Rare cardiac or lung injury
* Ravitch Procedure (Open surgery):  
  Removal of abnormal cartilage and repositioning of the sternum, sometimes with metal struts.
  + *Risks/Side effects:*
    - Longer recovery and scarring
    - Infection
    - Bleeding
    - Pain

**PREDEFINED Q AND A**

## 1. What are chest wall deformities?

Answer:  
Chest wall deformities are structural abnormalities affecting the shape and structure of the chest. The most common types are pectus excavatum (sunken or funnel chest) and pectus carinatum (pigeon chest), which involve either a depression or protrusion of the breastbone and ribs.

## 2. What causes chest wall deformities?

Answer:  
The exact cause is often unknown but is thought to involve abnormal cartilage growth connecting the ribs to the sternum. Genetic factors may play a role, as these deformities often run in families. They can be congenital or develop during childhood, especially during growth spurts.

## 3. What symptoms do chest wall deformities cause?

Answer:  
Symptoms vary by severity. Mild cases may have no symptoms, while more severe deformities can cause chest pain, shortness of breath, reduced lung capacity, and sometimes cardiac compression. Cosmetic concerns and low self-esteem are common.

## 4. How are chest wall deformities diagnosed?

Answer:  
Diagnosis involves a physical exam and imaging studies such as chest X-rays, CT scans, or MRI. The Haller index from CT scans measures severity in pectus excavatum. Additional tests may include echocardiograms and pulmonary function tests to assess heart and lung impact.

## 5. What treatment options are available?

Answer:  
Treatment depends on severity and type:

* Observation for mild cases.
* Bracing for pectus carinatum to remodel the chest wall.
* Surgery (e.g., Nuss or Ravitch procedures) for moderate to severe pectus excavatum or carinatum, usually after the child reaches adolescence.

## 6. When is surgery recommended?

Answer:  
Surgery is typically recommended for moderate to severe deformities that cause symptoms such as breathing difficulties, chest pain, or significant cosmetic concerns. It is usually performed after the child’s bones have matured, often in adolescence.

## 7. Can chest wall deformities affect breathing or heart function?

Answer:  
Yes, severe deformities can compress the lungs and heart, leading to reduced lung capacity, shortness of breath, and cardiac symptoms like palpitations. This is why evaluation with pulmonary and cardiac tests is important.

## 8. Are chest wall deformities hereditary?

Answer:  
They often run in families, suggesting a genetic component. Certain connective tissue disorders like Marfan syndrome are also associated with chest wall deformities.

## 9. What is the prognosis for children with chest wall deformities?

Answer:  
Many children with mild deformities live normal lives without treatment. Those with severe deformities who undergo surgery generally have good outcomes with improved chest appearance and function.

## 10. Are there any non-surgical treatments?

Answer:  
Yes, bracing is effective for pectus carinatum in many cases, especially if started early. Physical therapy and exercises may help improve posture and respiratory function but do not correct the deformity itself

## **Genomic Data:**

* + Inheritance Patterns:
    - Non-syndromic isolated cases of PE and PC often show autosomal dominant inheritance with variable penetrance.
    - Familial cases suggest a multifactorial inheritance pattern with low recurrence risk.
    - Syndromic cases are linked to well-characterized genetic mutations.
  + Genetic Syndromes Associated:
    - Marfan Syndrome: Caused by mutations in the FBN1 gene, affecting connective tissue and frequently associated with PE or PC.
    - Noonan Syndrome: Often caused by mutations in genes of the RAS/MAPK pathway, including PTPN11 and SOS1. These mutations are linked to thoracic deformities, pulmonary stenosis, and short stature.
    - Turner Syndrome: Chromosomal disorder frequently associated with chest wall deformities.
    - Other syndromes include DiGeorge syndrome and various chromosomal abnormalities.
  + Molecular Findings:
    - Mutations in PTPN11 (a non-receptor tyrosine phosphatase) are common in Noonan syndrome cases with chest wall deformities.
    - SOS1 gene variants are also implicated, affecting signal transduction pathways.
    - Incidental findings in genes like CACNA1C (associated with cardiac syndromes) have been reported in patients with chest wall deformities.
  + Prevalence in Syndromic vs. Isolated Cases:
    - Approximately 40% of patients with chest wall deformities may have identifiable genetic abnormalities when syndromic features are present.
    - Over 32 syndromes have been described with PE or PC as common features.

**DOCTOR PATIENT CONVERSATION**

Doctor: "Hello! I understand you have some concerns about your child's chest shape. Can you tell me what you’ve noticed?"

Parent: "Yes, I’ve noticed that my child’s chest looks sunken in the middle, and sometimes they seem to have trouble breathing during exercise. I’m worried it might be something serious."

Doctor: "Thank you for sharing that. Based on what you describe, your child may have a condition called pectus excavatum, which is the most common chest wall deformity where the breastbone is sunken inward."

Parent: "Is this dangerous? Will it get worse?"

Doctor: "In many cases, pectus excavatum is primarily a cosmetic issue and doesn’t cause serious health problems. However, in some children, especially if the deformity is moderate or severe, it can affect lung and heart function, leading to symptoms like shortness of breath or chest pain."

Parent: "How do you confirm the diagnosis?"

Doctor: "We usually start with a physical exam and then order imaging, like a chest X-ray or CT scan, to measure the severity. We also might do lung function tests and an echocardiogram to see if the heart is affected."

Parent: "What treatments are available?"

Doctor: "Treatment depends on severity. For mild cases, we often monitor without intervention. For pectus carinatum, which is a protruding chest, we sometimes use bracing to reshape the chest wall. For moderate to severe pectus excavatum, surgery is an option. The most common surgery now is the Nuss procedure, which is minimally invasive and involves placing a metal bar to lift the sternum."

Parent: "What about risks from surgery?"

Doctor: "Like any surgery, there are risks such as pain, infection, or complications like pneumothorax (air in the chest cavity). But these are relatively uncommon, and most children recover well with improved chest appearance and function."

Parent: "Is this condition hereditary? Should I be worried about other family members?"

Doctor: "Chest wall deformities can run in families, so it’s worth checking siblings or other children for similar signs. We can offer screening if you’re concerned."

Parent: "Thank you. What should I watch for at home?"

Doctor: "Keep an eye on your child’s breathing, exercise tolerance, and any chest pain. If symptoms worsen or if you notice difficulty feeding or blue discoloration, seek immediate care. Otherwise, we’ll schedule regular follow-ups to monitor progress."

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**AIRWAY MALACIA**

## **Alternative Names:**

* Laryngomalacia (when involving the larynx)
* Tracheomalacia (when involving the trachea)
* Bronchomalacia (when involving the bronchi)
* Congenital airway malacia
* Floppy airway syndrome
* Dynamic airway collapse
* Soft airway syndrome
* Airway cartilage weakness
* Supraglottic collapse (specific to laryngomalacia)

**DEFINITION AND DESCRIPTION**

### **tracheobronchomalacia (TBM)**

Tracheobronchomalacia (TRAY-key-oh-bronco-mă-LAY-cia) is a condition where your trachea (windpipe) and your bronchi (the tubes that lead from your trachea to your lungs) are so weak that they collapse and close down when you take a breath or cough. If you have TBM, you may have issues breathing. You may wheeze or cough a lot.

Some people are born with tracheobronchomalacia. This is primary or congenital TBM. But you can also develop it during your lifetime. Healthcare providers may call this secondary or acquired tracheobronchomalacia.

Healthcare providers have treatments to help you manage tracheobronchomalacia symptoms. In some cases, they can do surgery to support your trachea and bronchi.

Based on one study, experts estimate 1 in 2,100 babies are born with tracheobronchomalacia. In other research, experts estimate between 4% and 13% of people with airway problems have TBM. But that’s just an estimate, as providers don’t always make the connection between common respiratory problems and collapsing airways.

### **causes tracheobronchomalacia**

Tracheobronchomalacia happens when the walls of your trachea and bronchi are weak and collapse when you take a breath. Your trachea is a stiff, flexible tube made of cartilage that carries air in and out of your lungs. Your bronchi move air into your lungs and are lined with tiny hair cells that help move mucus and particles out of your lungs.

#### **Causes of primary/congenital TBM**

Primary/congenital tracheobronchomalacia in babies may happen for no known reason (idiopathic TBM). But it may happen if they’re born with conditions, including:

* **Vascular rings**: Your baby may have a vascular ring because their aorta didn’t form normally during fetal development. The aorta is the main artery that carries oxygenated blood away from your baby’s heart to the rest of their body. If your baby has a vascular ring, their aorta wraps around their trachea to cause tracheobronchomalacia.
* **Certain inherited disorders**: Hunter syndrome, Hurler syndrome and Ehlers-Danlos syndrome are inherited disorders that may cause tracheobronchomalacia.
* **Premature birth**: Babies born before 37 weeks of pregnancy may have an increased risk of TBM if their trachea isn’t fully formed.

#### **Causes of secondary/acquired TBM**

Several things may cause secondary/acquired tracheo bronchomalacia including:

* Certain medical conditions.
* Certain treatments.
* Exposure to certain toxins.

##### **Medical conditions**

Adults with tracheo bronchomalacia often have the following conditions that can lead to TBM:

* Asthma.
* Bronchitis.
* Obesity.
* Chronic obstructive pulmonary disease (COPD).
* Gastroesophageal reflux disease (GERD). Studies show nearly half of people treated for GERD have TBM.
* Goiters. This is an enlargement on your thyroid, which wraps around your trachea. Goiters can put pressure on your trachea.
* Relapsing polychondritis. This is a rare degenerative disease that causes your cartilage to deteriorate.

##### **Medical treatments**

Some medical treatments may cause tracheobronchomalacia:

* **Prolonged tracheostomy use**. A tracheostomy is a medical treatment to help you breathe by inserting a tube into your trachea.
* **Prolonged use of inhaled corticosteroids**. Inhaled corticosteroids can treat asthma.
* **Prolonged use of intubation and ventilation** that injures your airway.

##### **Exposure to toxins**

Exposure to the following toxins may increase your risk of TBM:

* Toxic gases such as mustard gas.
* Secondhand smoke.

### **symptoms of tracheobronchomalacia**

Symptoms may be different depending on whether you’re born with the condition or develop it over time.

Babies born with TBM may have symptoms that start when they’re 2 to 4 months old. Symptoms may include:

* Stridor.
* Having a hard time breathing when you breastfeed or bottle feed them.
* Persistent cough.
* Frequent colds.
* Frequent respiratory tract infections.

Tracheomalacia symptoms in adults may develop over time and get progressively worse. Symptoms include:

* Difficulty breathing after everyday activities like climbing stairs or walking.
* “Barking” or dry, harsh cough.
* Temporarily losing consciousness during coughing.
* Wheezing.
* Difficulty coughing up mucus.
* Frequent colds.
* Obstructive sleep apnea (OSA).

## **Diagnosis and Tests**

A healthcare provider will do a physical examination. If they’re treating your child, they’ll ask you about their symptoms, their overall health and any medical conditions that may be the reason why your child is coughing, wheezing or has trouble breathing.

If you’re having symptoms, your provider will do a physical examination and ask the same sorts of questions. They may ask if you have frequent respiratory infections or other conditions that affect how you breathe.

They may do the following tests:

* **Pulmonary function tests**: These are tests to see how well air moves in and out of your lungs.
* **Bronchoscopy**: In this test, your provider checks how narrow your trachea becomes when you take a deep breath or cough.
* **Computed tomography (CT) scan**: Your provider may order this test to make sure other conditions aren’t causing your symptoms.

## **Management and Treatment**

## Treatments vary depending on how tracheobronchomalacia affects your or your baby’s ability to breathe. For example:

* Your baby’s healthcare provider may prescribe antibiotics to treat any underlying infections. They may show you ways to breastfeed or bottle feed your baby that don’t affect their ability to breathe.
* They may prescribe medications to help manage your baby’s symptoms.
* They may prescribe pulmonary rehabilitation programs that teach you or your child ways to manage symptoms.
* You may use supportive devices that help you breathe.
* You or your child may need surgery to support weakened tracheas.

#### **Supportive devices for TBM treatment**

Several kinds of supportive devices or durable medical equipment treat TBM symptoms. These devices can help you or your child manage symptoms and limit how much tracheobronchomalacia affects your quality of life. Treatments include:

* **CPAP or BiPAP® machines**: These machines help to keep your trachea open.
* **External percussion vests**: TBM can make it hard to cough up mucus. External percussion vests connect to a machine that vibrates very fast, shaking up mucus in your bronchi so you can cough up mucus more easily.
* **Nebulizer**: Nebulizers change liquid medication into fine droplets that you or your child inhale through a mask or mouthpiece.

#### **Surgery for tracheobronchomalacia**

There are several surgical treatments for TBM. They include:

* **Bronchoscopy**: This is the same procedure that healthcare providers use to diagnose issues with your lungs or airway. In treatment, healthcare providers use a rigid bronchoscope to place a temporary stent in your airway. A stent is a small plastic, rubber or metal tube that holds your airway open. Stents help healthcare providers decide if you need additional surgery.
* **Tracheobronchoplasty:** This procedure involves sewing mesh to the outside of your trachea. The mesh gives your trachea more structure so it’s less likely to collapse.
* **Tracheopexy**: This is surgery that attaches the back of your trachea to ligaments in your spine to keep your trachea from collapsing.

##### **Complications of TBM surgery**

Tracheobronchoplasty and tracheopexy are major surgeries. Like most surgeries, there may be complications, including:

* Excessive bleeding.
* Blood clots.
* Infection of the surgical site.

If your healthcare provider recommends surgery to treat TBM, they’ll explain the risk of complications.

## **Outlook / Prognosis**

Your child’s trachea cartilage will become stiffer as they grow up. That means there’s less chance that their trachea and bronchi will collapse and affect their breathing. But often, children with TBM need ongoing medical treatment and support to help them breathe.

Sometimes, losing weight or treatment may help with TBM symptoms. But TBM in adults is often a chronic (continuing) condition. If you have tracheobronchomalacia, you’ll probably work with a healthcare provider who specializes in airway treatment for the rest of your life.

Studies suggest that surgery helps ease tracheobronchomalacia symptoms right away and for several years after surgery. If you’re like many people with tracheobronchomalacia, you have other medical conditions that affect your overall health. Those conditions can make it more difficult for you to respond to treatment or recover from surgery.

### **What is the life expectancy for people with tracheobronchomalacia?**

People who develop TBM often have other serious medical conditions that may affect their life expectancy. Your healthcare provider is your best source if you want to know if having tracheobronchomalacia could affect how long you’ll live.

#### **Does tracheobronchomalacia go away?**

TBM in adults can be life-threatening because it doesn’t go away and gets worse over time. TBM in babies may cause less serious symptoms as they grow up.

## **Prevention**

There’s no way to prevent congenital tracheobronchomalacia in children. Adults can develop TBM over time if they have certain medical conditions or have exposure to secondhand smoke or toxic gases.

Tracheobronchomalacia gets worse over time, so talk to a healthcare provider if you know you have exposure to secondhand smoke, toxic gases or conditions that may increase your risk. You may not be able to keep TBM from happening, but there may be ways to keep it from getting worse.

## **Living With**

Most children with tracheobronchomalacia will need medical support throughout childhood. For example, you or your child will need treatment like taking medications or using devices that keep your airway clear.

Your child’s pediatrician may schedule regular follow-up appointments to check on your child’s overall health. You may want to ask your child’s pediatrician about ways to protect your child’s overall health, like ways to avoid respiratory infections.

If you have tracheobronchomalacia, here are some suggestions that may be helpful:

* **Take care of your health**: Many people with TBM also have asthma, COPD or bronchitis. If you do, be sure to follow your treatment plan and your provider’s advice for managing your underlying illness.
* **Eat well**: Having obesity can make it hard for you to breathe. Getting to or maintaining a weight that’s right for you may help ease breathing issues that TBM can cause.
* **Get some regular physical activity:** Regular activity may help you manage your weight.
* **Avoid secondhand smoke**: Studies suggest secondhand smoke exposure increases your risk of developing TBM.

### **When should I see my healthcare provider?**

If you have TBM, you’ll have ongoing medical care and regular medical checkups. Contact your provider if you notice your symptoms like coughing or wheezing get worse.

## **Differential Diagnoses for Airway Malacia**

* Laryngomalacia
* Tracheomalacia
* Bronchomalacia
* Subglottic stenosis (congenital or acquired narrowing below vocal cords)
* Vocal cord paralysis (unilateral or bilateral)
* Congenital cysts or laryngeal masses (e.g., laryngeal cysts)
* Tracheal or bronchial stenosis (fixed airway narrowing due to scarring or congenital anomaly)
* Vascular rings or slings (vascular anomalies compressing the airway externally)
* Foreign body aspiration (acute airway obstruction)
* Hypocalcemic tetany (laryngospasm due to low calcium)
* Infectious causes (e.g., croup, bacterial tracheitis causing airway inflammation)
* Asthma or reactive airway disease (wheezing without dynamic airway collapse)
* Excessive dynamic airway collapse (EDAC) (posterior membrane bowing without cartilage weakness)

## **Epidemiology of Airway Malacia**

* Prevalence:  
  Airway malacia, including tracheomalacia and bronchomalacia, is relatively rare but not uncommon in pediatric populations undergoing airway evaluation. In a study of 512 bronchoscopies, airway malacia was diagnosed in approximately 31% (160 children), with a median age of 4 years (range 0–17 years).
* Incidence:  
  The estimated incidence of primary airway malacia is at least 1 in 2,100 children.
* Sex Distribution:  
  Males appear to be more frequently affected; in one cohort, 94 out of 160 diagnosed children were males.
* Age at Diagnosis:  
  Median age at diagnosis is around 4 years, but airway malacia can present from infancy through childhood.
* Associated Conditions:  
  Up to 30% of children with airway malacia have congenital syndromes or anomalies such as esophageal atresia/tracheoesophageal fistula (EA/TEF), cardiovascular anomalies, or primary ciliary dyskinesia.
* Types:  
  Airway malacia can be congenital (primary) or acquired (secondary), with congenital forms often improving spontaneously by 18–24 months of age.
* Clinical Impact:  
  Symptoms include stridor, recurrent respiratory infections, cough, and exercise intolerance. Severity and symptom persistence vary depending on the type and extent of Malacia

## **PREDEFINED Q AND A**

## 1. What is airway Malacia?

Answer:  
Airway Malacia refers to abnormal softness or floppiness of the airway walls, leading to dynamic collapse during breathing. It includes laryngomalacia (upper airway), tracheomalacia (trachea), and bronchomalacia (bronchi).

## 2. What are the common symptoms of airway Malacia in children?

Answer:  
Symptoms often include noisy breathing (stridor or wheezing), persistent cough, recurrent respiratory infections such as pneumonia or bronchitis, episodes of respiratory distress, and sometimes cyanosis or difficulty feeding. Wheezing and cough are common, with cough present in over 98% of cases and wheezing in about 67%.

## 3. How is airway malacia diagnosed?

Answer:  
Diagnosis is primarily by flexible bronchoscopy, which allows direct visualization of airway collapse during breathing. Imaging such as dynamic CT scans can assist. Clinical suspicion arises when children have persistent noisy breathing or recurrent respiratory infections not explained by other causes.

## 4. What infections are commonly associated with airway malacia?

Answer:  
Children with airway malacia complicated by pneumonia often have infections caused by *Mycoplasma pneumoniae*, *Streptococcus pneumoniae*, and respiratory syncytial virus (RSV).

## 5. What are the risk factors or associated conditions for airway malacia?

Answer:  
Premature birth, history of mechanical ventilation, and congenital anomalies such as esophageal atresia or cardiovascular malformations increase the risk. Children with airway malacia often have longer hospital stays and more severe respiratory symptoms.

## 6. How is airway malacia managed?

Answer:  
Management is mostly supportive, including treatment of infections, respiratory support if needed, and monitoring. Severe cases may require surgical intervention or airway stabilization. Avoidance of irritants and treatment of associated conditions like gastroesophageal reflux may help.

## 7. What is Excessive Dynamic Airway Collapse (EDAC) and how is it related to airway malacia?

Answer:  
EDAC is a related condition characterized by excessive bulging of the posterior airway membrane during exhalation without cartilage weakness. It can coexist with or mimic airway malacia and may require specialized evaluation and management.

## 8. Can airway malacia improve over time?

Answer:  
Yes, many cases of congenital airway malacia improve spontaneously as the airway cartilage strengthens, often by 18-24 months of age, though some cases may persist or require intervention

**DOCTOR PATIENT CONVERSATION**

Doctor: "Hello, I understand you've been experiencing some breathing difficulties. Can you tell me more about your symptoms?"

Patient: "Yes, I often feel like I’m short of breath, especially when I exercise or cough. Sometimes I even have episodes where I gasp for air. It’s been going on for years, and I was told it might be asthma, but the medications don’t seem to help much."

Doctor: "Based on what you’re describing, one possibility is a condition called airway malacia. This means that parts of your airway, like your windpipe or the tubes leading to your lungs, are softer than normal and tend to collapse when you breathe out or cough."

Patient: "I’ve never heard of that before. How does it happen?"

Doctor: "It can be something you’re born with, called congenital airway malacia, or it can develop later due to injury, inflammation, or other conditions. The cartilage that normally keeps your airway open is weak or floppy, so the airway narrows or collapses, making it harder to breathe."

Patient: "How do you diagnose this?"

Doctor: "We usually start with imaging tests like a CT scan, especially a dynamic CT that takes pictures while you breathe in and out. The most definitive test is a bronchoscopy, where we use a thin camera to look directly inside your airway while you breathe. This helps us see if and where your airway is collapsing."

Patient: "Is there treatment for this? Can it be fixed?"

Doctor: "Yes, treatment depends on the severity. Mild cases can be managed with supportive care and physiotherapy to help clear mucus and improve breathing. For more severe cases, there is a surgical option called tracheobronchoplasty, where a mesh is placed to support the airway and prevent collapse. This surgery has helped many patients return to normal activities."

Patient: "Are there risks with surgery?"

Doctor: "Like any surgery, there are risks such as infection or complications from anesthesia, but it’s generally safe when done by experienced surgeons. We carefully evaluate each patient to decide the best approach."

Patient: "What should I do now?"

Doctor: "We’ll arrange the necessary tests to confirm the diagnosis and assess severity. Meanwhile, if you experience worsening breathing difficulty, persistent cough, or infections, please seek medical attention promptly. We’ll work together to develop a treatment plan tailored for you."

REFERENCES

[Tracheobronchomalacia: Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/22061-tracheobronchomalacia#diagnosis-and-tests)

### **Tracheal stenosis**

## Common Alternative Names:

* Tracheal narrowing
* Tracheal stricture
* Subglottic stenosis (when narrowing is just below the vocal cords)
* Laryngotracheal stenosis (when both larynx and trachea are involved)
* Tracheal obstruction
* Congenital tracheal stenosis (if present from birth)
* Acquired tracheal stenosis (if due to injury, infection, or intubation)
* Tracheal web or membrane (if caused by thin tissue bands causing narrowing)

**DEFINITION AND DESCRIPTION**

Tracheal stenosis involves narrowing of your trachea (windpipe) that makes it harder to breathe. Your trachea is a tube made of cartilage and soft tissue. Air travels through your trachea on the way from your nose and mouth to your lungs. When you have tracheal stenosis, inflammation, injury or scar tissue in your trachea makes it harder for air to flow through.

“Stenosis” is the medical term for atypical narrowing in a body passage. It can develop in different parts of your throat. For example, a related condition called laryngotracheal stenosis involves narrowing in both your trachea and larynx (voice box). Subglottic stenosis is narrowing in the area above your trachea and below your vocal cords.

Regardless of the part of your throat that’s affected, you should contact a healthcare provider if you’re having difficulty breathing.

#### **Types**

There are two types of tracheal stenosis:

* **Acquired tracheal stenosis** is acquired (developed) during your lifetime because of an injury or illness. The most common type, it affects both adults and children.
* **Congenital tracheal stenosis (CTS)** is present at birth. A rare and potentially fatal condition, it affects 1 out of every 64,500 [babies](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10964263/). Most are diagnosed at birth or within the first few months of being born.

Tracheal stenosis is life-threatening in infants. **Seek emergency care if your newborn is showing signs of tracheal stenosis, like struggling for air**.

That said, both forms of tracheal stenosis make it harder to breathe, which can affect your quality of life. That’s why seeing a healthcare provider is so important if you or your child has this condition.

### **causes tracheal stenosis**

With congenital tracheal stenosis (CTS), the cartilage that makes up an infant’s trachea doesn’t form correctly, causing their windpipe to be too narrow.

Long-term intubation is the most common cause of acquired tracheal stenosis. Intubation is a lifesaving treatment that involves inserting a tube into your trachea so you can breathe. Sometimes, intubation causes damage that leads to stenosis. Children may develop tracheal stenosis if they were born with premature lungs and their provider used breathing tubes to help them get air.

You might also develop tracheal stenosis if you:

* Have an autoimmune disorder, like granulomatosis with polyangiitis (GPA)
* Have an inflammatory condition, like pulmonary sarcoidosis
* Have an infectious disease, like tuberculosis, or other bacterial and viral infections that affect your respiratory system
* Have a benign or malignant tumor pressing on your trachea
* Inhaled a substance that damaged your trachea, including chemicals or poisonous gases
* Had radiation therapy directed toward your neck or chest
* Had a tracheostomy (emergency surgery that creates a hole in your trachea so that you can breathe)

### **symptoms of tracheal stenosis**

Many tracheal stenosis symptoms are the same for children and adults. Common symptoms include:

* Difficulty breathing after everyday activities like climbing stairs or walking
* Stridor (sounds like a wheeze or whistle when you breathe both in and out)
* Persistent cough
* Difficulty coughing up phlegm
* Frequent colds, bouts of pneumonia or other respiratory infections
* Persistent asthma that isn’t better after treatment
* Chest congestion
* Gaps in breathing (apnea) and sleep apnea

Children are more likely to have additional symptoms:

* Infants might have difficulty breastfeeding or bottle feeding. They might also seem unusually tired after feeding.
* Older children might choke or have difficulty breathing while they eat.
* Older children’s skin around their noses and their gums might appear blue (cyanosis)

## **Diagnosis and Tests**

Otolaryngologists (ENTs) use several tests to diagnose tracheal stenosis and decide how to treat it. Tests may include:

* **Endoscopic procedures**. Bronchoscopy is the primary procedure for diagnosing tracheal stenosis. Your healthcare provider may also perform a laryngoscopy.
* **Imaging procedures**. A computed tomography (CT) scan of your chest and neck is the most common imaging procedure that shows tracheal stenosis. Sometimes, healthcare providers recommend magnetic resonance imaging (MRI) to help plan treatment.
* **Pulmonary function test**. Providers will ask you to complete several breathing tests. This helps them check things like how your trachea size and lung function affect your breathing.

You may need other tests to check for what’s causing your condition. Tests may include a blood test to check for inflammation or infection or a biopsy to see if unusual growths in your airway are cancerous.

## **Management and Treatment**

Surgeries and procedures that widen your trachea are the most common treatments for tracheal stenosis. Treatment options include:

* **Tracheal dilation**. Healthcare providers place a balloon or tracheal dilator in your trachea. The balloon or dilator stretches your trachea so that you can breathe.
* **Laser bronchoscopy**. Providers direct a laser beam at scar tissue in your trachea. The laser burns away the tissue, opening up your airways.
* **Trachea airway stent**. A provider places a small, plastic or metal tube called a stent that holds your trachea open. (Stenting may or may not be an option, depending on the location of the stenosis.)
* **Tracheal resection and reconstruction**. A provider cuts away (resects) the tissue that’s causing the narrowing. Then, they join the two remaining ends of your trachea together. This procedure reconstructs your trachea to create an unobstructed airway.

The best treatment for tracheal stenosis depends on lots of things, including where the narrowing is and how severe it is. Your provider will explain how these factors inform which procedures will work best for you.

**Outlook / Prognosis**

You’ll likely need surgery or a nonsurgical procedure if you have tracheal stenosis. Each treatment option has different recovery times and outcomes.

For example, tracheal resection and reconstruction surgeries are invasive. But they’re more likely to eliminate the narrowing in your trachea in the long term. Nonsurgical procedures, like tracheal dilation, are less invasive. They may be the only treatment you need, or you may repeat procedures.

Regardless of the procedure, your healthcare provider will monitor you to check for recurrence. Tracheal stenosis sometimes comes back because treatment can cause new scar tissue to form. Your provider will explain how likely it is that your condition will return.

## **Living With**

You might start by asking your healthcare provider how your surgery will affect you. Every procedure to treat tracheal stenosis will require different at-home care. Your healthcare provider will have information about your next steps. They may advise you on:

* **What you can eat.** For the first 24 hours, you may need to stick with soft foods that are easy to swallow. It may be a good idea to limit yourself to bland foods that won’t upset your stomach.
* **How you should sleep**. For the first few days, you may need to keep your upper body elevated as you try to sleep.
* **How to manage pain**. Your provider can recommend over-the-counter (OTC) medications or prescribe pain medicines as needed.
* **How active you should be**. You may have activity restrictions for the first week or so following surgery.

### **When should I see my healthcare provider?**

Your healthcare provider will schedule follow-up appointments to check on your recovery after treatment. At first, you may need to see your provider every few weeks or so. If you’re healing well, your provider may extend follow-up visits to every few months until they’re confident the stenosis won’t return.

**Contact your provider anytime you’re experiencing shortness of breath. While next steps vary depending on your condition, it can be dangerous to put off getting help when you’re having difficulty breathing. It’s essential to seek care.**

You should go to the emergency room if you can’t breathe or have other tracheal stenosis symptoms. The symptoms might be a sign your tracheal stenosis has come back.

**If you’re caring for a newborn or infant who’s having difficulty breathing, get them to an emergency room immediately.**

## **Differential Diagnosis of Tracheal Stenosis**

* Tracheomalacia (softening and collapse of the tracheal walls)
* Laryngomalacia (supraglottic airway collapse)
* Subglottic stenosis (narrowing just below the vocal cords, congenital or acquired)
* Vocal cord paralysis (unilateral or bilateral causing airway obstruction)
* Congenital cysts or laryngeal masses (e.g., laryngeal cysts causing narrowing)
* Foreign body aspiration (causing acute airway obstruction)
* Hypocalcemic tetany (laryngospasm due to low calcium)
* Infectious causes (e.g., bacterial tracheitis, epiglottitis causing airway swelling)
* Vascular rings or slings (external vascular compression of the trachea)
* Granulomatous diseases (e.g., sarcoidosis, Wegener’s granulomatosis causing airway inflammation and stenosis)
* Trauma-related stenosis (post-intubation injury, surgery, or blunt trauma causing scarring)
* Neoplastic lesions (benign or malignant tumors causing airway narrowing)
* Autoimmune disorders (e.g., relapsing polychondritis affecting airway cartilage)
* Asthma or reactive airway disease (may mimic symptoms but no fixed airway narrowing)

## **Epidemiology of Tracheal Stenosis**

* Incidence:
  + Overall incidence of tracheal stenosis following tracheostomy or intubation ranges widely from 0.6% to 31% depending on the population and diagnostic methods used.
  + Symptomatic tracheal stenosis occurs in about 6% of patients after tracheostomy.
  + Post-tracheotomy or post-intubation stenosis incidence is reported between 0.6% and 21%.
  + In ICU patients, tracheal stenosis after prolonged intubation or tracheostomy is a significant complication.
* Risk Factors:
  + Prolonged intubation or delayed tracheostomy increases the risk.
  + Airway trauma from endotracheal tubes, poor cuff pressure control, infection, and surgical technique influence development.
  + Systemic factors like cardiovascular disease and metabolic conditions may contribute.
  + COVID-19 patients, especially those with prolonged ventilation, have increased tracheal stenosis incidence.
* Demographics:
  + Can affect all ages but often seen in adults after prolonged mechanical ventilation.
  + Some studies report a higher prevalence in males.
  + Median age varies but often includes middle-aged adults.
* Clinical Burden:
  + Tracheal stenosis leads to respiratory distress, impaired quality of life, and may be life-threatening if severe.
  + It accounts for a significant proportion of tracheostomy-related complications and hospital readmissions.
  + Mortality rates vary; some studies report in-hospital mortality around 3-23% in severe cases.
* Common Causes:
  + The most common cause is prolonged endotracheal intubation or tracheostomy (up to 85% in some cohorts).
  + Other causes include tumors, infections (e.g., tuberculosis), trauma, autoimmune diseases, and congenital abnormalities.

## **Key Genetic Findings:**

* TBX5 gene mutations:
  + A de novo pathogenic variant in the TBX5 gene (e.g., p.Ile227Thr) has been identified in patients with congenital tracheal stenosis combined with congenital heart defects.
  + TBX5 is a transcription factor critical for embryonic development, including tracheal cartilage formation. Mutations can reduce protein stability and disrupt normal tracheal development.
* Fibroblast Growth Factor (FGF) Pathway:
  + Mutations or dysregulation in FGFR2 and FGF10 genes are implicated in tracheal stenosis.
  + Overexpression of FGF10 and its regulators (e.g., TBX4, TBX5) leads to abnormal proliferation of tracheal mesenchyme and excessive cartilage formation, contributing to stenosis.
* Sonic Hedgehog (SHH) Signaling:
  + SHH pathway is essential for tracheal chondrogenesis. Loss of SHH function disrupts expression of downstream genes like BMP4 and SOX9, impairing cartilage development and causing tracheal malformations.
* GLI3 mutations:
  + Associated with syndromes such as Pallister-Hall syndrome, GLI3 mutations can cause tracheal stenosis as part of broader congenital malformations.
* Genetic predisposition to acquired tracheal stenosis:
  + Recent research suggests that genetic variants affecting wound healing and scarring pathways may predispose certain ethnic groups (e.g., African Americans) to acquired laryngotracheal stenosis after intubation or tracheostomy.
* Congenital tracheal stenosis (CTS):
  + Characterized by complete tracheal rings without a membranous portion, CTS is a rare but life-threatening condition with a strong genetic basis involving the genes mentioned above

## **Treatment of Tracheal Stenosis: Drug Information and Side Effects**

## 1. Medical (Drug) Treatments

* Corticosteroids (Anti-inflammatory drugs):
  + Used to reduce airway inflammation and prevent progression of stenosis, either systemically (oral or intravenous) or by inhalation.
  + Side effects:
    - Oral/IV steroids: Immunosuppression, hyperglycemia, weight gain, osteoporosis, adrenal suppression, mood changes.
    - Inhaled steroids: Oral thrush, hoarseness, cough.
* Other supportive medications:
  + Antibiotics if infection is present.
  + Bronchodilators may be used symptomatically but do not treat stenosis itself.

## 2. Endoscopic (Non-Surgical) Procedures

* Tracheal dilation:
  + Balloon or mechanical dilators widen the narrowed tracheal segment. Provides temporary relief.
  + May require repeated procedures due to restenosis.
* Laser bronchoscopy:
  + Laser ablation removes scar tissue causing narrowing.
  + Usually temporary relief; can sometimes worsen stenosis if not carefully applied.
* Cryotherapy:
  + Freezing abnormal tissue to reduce size and improve airway patency. Minimally invasive.
* Photodynamic therapy:
  + Uses light-sensitive drugs and laser to destroy abnormal cells. Experimental but promising.
* Tracheobronchial stenting:
  + Placement of silicone or metal stents to keep airway open, used in inoperable cases or as a bridge to surgery.
  + Side effects: Granulation tissue formation, stent migration, infection, mucous plugging.

## 3. Surgical Treatment

* Tracheal resection and reconstruction:
  + Removal of stenotic segment and rejoining healthy ends.
  + Considered the definitive treatment with the highest cure rates (~95%).
  + Requires good overall patient health and suitable stenosis characteristics.

### **PREDEFINED QUESTIONS AND ANSWERS**

## 1. Why did I develop tracheal stenosis?

Tracheal stenosis often develops due to injury or irritation to the tracheal lining, most commonly from prolonged intubation or tracheostomy. Factors such as pressure from the breathing tube cuff, airway trauma during insertion, infections, poor cuff pressure management, and underlying conditions like diabetes or cardiovascular disease can contribute to damage and scarring that narrows the airway. Other causes include autoimmune diseases, tumors, radiation therapy, or congenital abnormalities, but the majority of acquired cases relate to airway instrumentation and prolonged mechanical ventilation.

## 2. Will I need surgery?

Whether you need surgery depends on the severity, location, and cause of your tracheal stenosis, as well as your overall health. Mild cases may be managed conservatively or with endoscopic procedures like dilation or laser therapy. However, surgical resection and reconstruction of the narrowed tracheal segment is often the definitive treatment for significant or symptomatic stenosis that impairs breathing.

## 3. What are the typical side effects of surgery to treat tracheal stenosis?

Surgery carries risks such as:

* Bleeding
* Infection
* Airway swelling or restenosis (re-narrowing)
* Anastomotic complications (problems at the site where the trachea is reconnected)
* General risks of anesthesia

Most patients recover well with careful surgical technique and postoperative care, but close follow-up is essential to monitor for complications.

## 4. How likely is it that my tracheal stenosis will come back after surgery?

The chance of recurrence varies but is generally low with successful surgical resection, reported cure rates can be as high as 95%. However, restenosis can occur, especially if underlying risk factors persist or if the surgery is incomplete. Factors like ongoing inflammation, infection, or poor healing increase the risk of recurrence.

## 5. How can I care for myself during recovery?

* Follow all postoperative instructions carefully, including wound care and activity restrictions.
* Attend all follow-up appointments for airway evaluation and monitoring.
* Avoid respiratory infections by practicing good hygiene and getting vaccinations.
* Manage underlying conditions such as diabetes or reflux that may affect healing.
* Report any new or worsening symptoms such as difficulty breathing, coughing, or noisy breathing promptly.
* Avoid smoking and irritants that can worsen airway inflammation

**Doctor-patient conversation about tracheal stenosis**

Doctor: "Hello, I want to discuss the results of your recent evaluation. You have a condition called tracheal stenosis, which means there is a narrowing in your windpipe that’s making it harder for you to breathe."

Patient: "What causes this narrowing? Did I do something wrong?"

Doctor: "Not at all. The most common cause is injury or irritation from previous intubation or a tracheostomy tube, especially if it was in place for a long time. Other causes include infections, inflammation, or sometimes congenital issues. It’s not your fault, and we have ways to manage it."

Patient: "Will I need surgery to fix this?"

Doctor: "That depends on how severe the narrowing is and how much it’s affecting your breathing. Mild cases might be managed with less invasive procedures like balloon dilation or laser treatment. But if the narrowing is significant, surgery to remove the narrowed segment and reconnect the healthy parts of your trachea is usually the best option."

Patient: "What are the risks if I have surgery?"

Doctor: "Like any surgery, there are risks such as bleeding, infection, or the airway narrowing again after surgery. We monitor patients closely after surgery to catch any issues early. Most people recover well and have significant improvement in breathing."

Patient: "How will I communicate if I have a tracheostomy or after surgery?"

Doctor: "If you have a tracheostomy tube, we can use special speaking valves that allow air to pass through your vocal cords so you can talk. Speech therapists will work with you to help optimize your voice and breathing. We also ensure you have alternative communication methods if needed."

Patient: "What should I expect during recovery?"

Doctor: "Recovery involves careful monitoring of your airway, managing any infections, and attending follow-up appointments. You’ll need to avoid irritants like smoking and practice good hygiene. Our team, including speech therapists and physiotherapists, will support you throughout the process."

Patient: "Is there anything I can do to prevent this from happening again?"

Doctor: "We recommend avoiding prolonged intubation if possible, ensuring proper care of any airway tubes, and managing any underlying health issues. If you have symptoms like difficulty breathing or noisy breathing in the future, seek medical attention early."

REFERENCES

[Tracheal Stenosis: Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/21866-tracheal-stenosis#symptoms-and-causes)

## [**https://rarediseases.info.nih.gov/diseases/12008/congenital-tracheal-stenosis**](https://rarediseases.info.nih.gov/diseases/12008/congenital-tracheal-stenosis)

## **Interstitial lung disease**

**DEFINITION AND DESCRIPTION**

The term ‘interstitial lung disease’ covers a large group of lung conditions that affect the interstitium, the tissue and space around the alveoli (air sacs) in the lungs. This is where oxygen is taken into the body and carbon dioxide is taken out of the body. These conditions can affect the lungs in different ways. Most of the time, the walls of the interstitium get thicker, which makes it harder for the lungs to expand (when breathing in) or contract (when breathing out). This can lead to lower levels of oxygen in the blood and may make a child short of breath. To make up for this they will often breathe faster, which uses up more energy and makes them very tired. ChILD also includes some conditions whose effects are more widespread in the lungs and are not just focused on the interstitium. These are called diffuse lung diseases. The symptoms and effects on the body are usually the same, which is why they are known as ChILD conditions. ChILD conditions are linked to a range of potential causes, including the genes a child inherits from their parents, having problems with the immune system, being exposed to air pollution, and going through chemotherapy or radiotherapy.

## **Causes**

ChILD can crop up without a known cause. On the other hand, certain genes, toxins, or other diseases may be culprits.

Some possible causes are:

**Inherited conditions**: Disorders that cause problems with surfactant -- a fluid in the lungs that helps your child breathe -- can be passed on through genes.

**Immune system disorders**: Certain immune system problems make it harder for kids to fight off illnesses.

**Autoimmune diseases**: These happen when your child’s immune system mistakenly attacks healthy tissues. Inflammatory bowel disease and collagen vascular disease are two autoimmune conditions commonly linked to chILD.

**Infection**: Some children get chILD after a cold or virus.

**Birth defects**: Babies can be born with a birth defect that causes problems with their lungs.

**Aspiration**: When you inhale food, liquid, or vomit into your lungs, damage can happen. Aspiration often affects kids with swallowing problems or a condition called gastroesophageal reflux disease (GERD).

**Cancer treatments**: Therapies, such as radiation and chemo, may lead to chILD.

**Environmental triggers**: Chemicals and molds can irritate your child’s lungs.

**Surgeries**: A lung transplant or bone marrow transplant may be to blame for certain cases of chILD.

**Types**

There are different kinds of chILD. Many have long, hard-to-pronounce names. Although they’re all considered rare diseases, some forms are more common in specific age groups.

ChILD diseases that usually affect babies are:

* Surfactant dysfunction mutations
* Developmental disorders, such as alveolar capillary dysplasia
* Lung growth abnormalities
* Neuroendocrine cell hyperplasia of infancy (NEHI)
* Pulmonary interstitial glycogenosis (PIG)

Types of chILD that are more common in children and teens are:

**Idiopathic interstitial pneumonias**: This category includes cryptogenic organizing pneumonia, acute interstitial pneumonia, nonspecific interstitial pneumonia, desquamative interstitial pneumonia, and lymphocytic interstitial pneumonia.

**Other primary disorders:** These disorders may be alveolar hemorrhage syndromes, aspiration syndromes, hypersensitivity pneumonitis, bronchiolitis obliterans, eosinophilic pneumonia, pulmonary alveolar proteinosis, pulmonary infiltrates with eosinophilia, pulmonary lymphatic disorders (lymphangiomatosis, lymphangiectasis), or pulmonary vascular disorders (haemangiomatosis).

**ILD-associated with systemic disease processes:** Examples are connective tissue diseases, histiocytosis, malignancy-related lung disease, sarcoidosis, and storage diseases.

**Disorders of the compromised immune system:** This group includes opportunistic infection, disorders related to therapeutic intervention, lung and bone marrow transplant-associated lung diseases, and diffuse alveolar damage of unknown cause.

**Symptoms**

Signs and symptoms of chILD often depend on the type of disease and how severe it is. They may include:

* Difficulty breathing or shortness of breath
* Fast or noisy breathing
* Wheezing
* Coughing or chest congestion
* Repeated bouts of pneumonia or bronchitis
* Low oxygen levels
* Failure to gain weight or grow in height

## **Adults vs. Child Interstitial Disease**

Some kids who get chILD will have the condition throughout their lives, so it can technically happen in both children and adults.

But when an adult is diagnosed with an interstitial lung disease, doctors usually consider it a completely different condition than chILD.

Kids with chILD should see a pediatric pulmonologist, rather than a doctor who specializes in adults.

## **Diagnosis**

It’s often hard to diagnose chILD. Each type is different, so the methods your doctor uses will vary.

Tests that help diagnose chILD include:

**Chest X-ray or CT scan:** These imaging procedures use X-rays to take pictures of your child’s lungs.

**Lung function tests**: Doctors measure how kids breathe in and out to examine how well their lungs work.

**Blood tests**: Blood draws are sometimes used to check for abnormal genes.

**Bronchoalveolar lavage**: With this procedure, a doctor injects salt water through a tube in your child’s lungs to see specific types of cells. It can help spot a lung injury, aspiration, infection, or an airway problem.

**Lung biopsy**: A surgeon takes out a small piece of lung tissue to test in a lab.

## **Treatment**

Very little research has been done on how to treat chILD. But some therapies can help children’s lungs work better, ease symptoms, or simply make them feel better.

Your doctor may recommend:

**Medicines**: Steroids lower lung inflammation, antimicrobial drugs treat infections, and bronchodilators help relax muscles around the airways.

**Oxygen**: More oxygen can help children breathe better and give their hearts a rest.

**Nutrition**: An eating plan that focuses on weight gain may benefit some kids with chILD.

**Pulmonary rehab and exercises**: Special therapies aim to ease congestion and improve how well the lungs work (your doctor may call this “lung function”).

**Breathing machines**: Devices called ventilators can help children breathe easier.

**Lung transplant**: This may be an option for kids with serious or life-threatening cases of chILD. So far, chILD doesn’t seem to come back in children who have the surgery.

## **Outlook**

With no cure for chILD, the condition comes about and advances differently in each child.

Some cases are severe and tend to be life-threatening at an early age. Other types stay the same or worsen slowly. But certain forms of the disease, such as neuroendocrine cell hyperplasia of infancy, can even improve over time.

Kids with chILD may have special needs. It’s a good idea to talk with teachers, family members, and other parents about ways to support your child and the rest of your family. Take good care of your own health, too. Caregivers often put themselves last, but you need to be well to help your family.

## **Epidemiology**

Overall, ILD is rare in children, and individual ILDs are extremely rare. Because of the different approaches to case ascertainment and definition, determining the incidence and prevalence of ILDs is difficult. In a systematic review of the literature, the incidence of chILD was estimated at 0.13-16.2 cases per 100,000 children per year.

### United States data

Most of the literature is composed of case reports and small series. One of the first relatively large series was a combined retrospective and prospective study by Fan et al performed over a 15-year period at a leading referral center for ILD.The study included 99 patients, in whom the case definition included respiratory symptoms lasting longer than 1 month, diffuse infiltrates depicted on chest radiography, and the absence of known bronchopulmonary dysplasia (BPD), heart disease, malignancy, immunodeficiency, autoimmunity, cystic fibrosis (CF), aspiration, or acquired immunodeficiency syndrome (AIDS).

A retrospective study that attempted a relatively complete case ascertainment of children undergoing biopsy for ILD in 11 referral centers in the United States and Canada over a 5-year period reported 187 cases in children younger than 2 years old. Fan et al expanded this case ascertainment in a study of children aged 2-18 years undergoing lung biopsies over a 4-year period at 12 centers across North America; the completed study reported 191 cases of chILD.In a single-center study, chILD cases were retrospectively reviewed and classified according to the classification system used by Fan et al; 93 cases were identified and 91% were classifiable.

The National Registry for Childhood Interstitial and Diffuse Lung Diseases was created in the United States to improve the understanding of chILDs. The prospective registry enrolled 683 patients with various diagnoses from centers across the United States; the most common diagnosis was NEHI (23% of patients).

### International data

A national survey of cases of chronic ILD in immunocompetent children aged 0-16 years in the United Kingdom and Ireland over a 3-year period yielded an estimated prevalence of 3.6 per million. Griese et al used data from the Surveillance Unit for Rare Paediatric Disorders to determine that the incidence of chILD in Germany is 1.32 new cases per 1 million children per year.

In Europe, an international registry enrolled 575 patients over a 3-year period; the distribution of some of the diagnostic categories was as follows:

* Diffuse developmental disorders, 2.6%
* Growth abnormalities, 6.4%
* Other diagnoses in infancy (eg, NEHI, PIG), 18.5%
* Surfactant dysfunction, 22.3%
* Respiratory distress syndrome in a mature or almost mature neonate, 4%
* Diffuse parenchymal lung disease (DPLD) related to systemic disease, 15.6%
* Exposure-related DPLD in an immune intact host, 13.3%
* DPLD in an immunocompromised host, 4.3%
* DPLD related to lung vessels, 4.6%; reactive lymphoid lesions, 1.2%; and airway disorders, 3.5%

A study from Australia and New Zealand of patients aged 0-18 years with a diagnosis of chILD gathered questionnaire data from clinical providers and data from reference genetics laboratories; the investigators calculated the prevalence at 1.5 cases per million.

### Sex- and age-related demographics

There appears to be a slight male predominance (53-60%) in reported cases of chILD. This male predominance is found primarily in cohorts of children younger than 2 years; the male: female ratio is close to 50:50 in the diagnoses that cluster in the older ages.

ILD can present at any age from birth to adulthood. Some diagnoses cluster in infancy, such as NEHI and PIG, while other forms of chILD present throughout childhood and adolescence. A lung biopsy review of 378 chILD cases in children aged 0-2 years versus those aged 2-18 years demonstrated significant differences in the distribution and spectrum of diseases based on age. A European task force described 185 cases of ILDs in immunocompetent children; they demonstrated a significant clustering of cases among children younger than 2 years. In an 18-year retrospective review of 93 cases identified at a single institution, the median age at diagnosis was 90 months

## **Differential Diagnoses**

* Afebrile Pneumonia Syndrome
* Anti-GBM Antibody Disease
* Aspiration Syndromes
* Bone Marrow Transplantation
* Bronchopulmonary Dysplasia
* Cystic Fibrosis
* Granulomatosis with Polyangiitis (GPA, formerly Wegener Granulomatosis)
* Hemosiderosis
* Histiocytosis
* Histoplasmosis
* Hypersensitivity Pneumonitis
* Inhalation Injury
* Juvenile Systemic Sclerosis
* Long-Term Effects of Bone Marrow Transplantation
* Lymphoproliferative Disorders
* Partial Anomalous Pulmonary Venous Connection
* Pediatric Anti-GBM Disease (Goodpasture Syndrome)
* Pediatric Sarcoidosis
* Pediatric Severe Combined Immunodeficiency
* Posttransplant Lymphoproliferative Disease (PTLD)
* Idiopathic Pulmonary Arterial Hypertension
* Systemic Lupus Erythematosus (SLE)
* Total Anomalous Pulmonary Venous Connection

## **Staging**

No widely used staging system is available for chILD, which is appropriate because the spectrum of possible diagnoses is large.

In adults, a scoring system is available for IPF, based on clinical, radiographic, and pathologic findings (ie, CRP scoring system).

Fan devised a simple scoring system for chILD. A score of 5 indicates the worst outcome, with a 38% survival rate at 60 months. A score of 2, 3, or 4 indicates a survival rate of 76%. Data from Cox proportional hazards modeling suggests a 140% increase in risk of death with each unit increase in score. The score can be applied to most ILD diagnoses with the likely exception of NEHI, which has a good eventual prognosis in the majority of cases.

The Fan severity scoring system is as follows:

1. Asymptomatic
2. Symptomatic with normal oxyhemoglobin saturation
3. Symptomatic with nocturnal or exercise-induced desaturation
4. Desaturation at rest
5. Pulmonary hypertension

## **Procedures**

### Bronchoalveolar lavage

Bronchoscopy with bronchoalveolar lavage (BAL) is useful in diagnosing certain conditions in the differential diagnosis of ILD, including alveolar proteinosis, aspiration syndromes, pulmonary hemosiderosis, eosinophilic syndromes, and various infections. Occasionally, results of cytologic analysis may be diagnostic—for example, when Langerhans cells are present, indicating histiocytosis. Most authorities believe BAL should precede biopsy.

Problems with the use of BAL include the lack of a standardized methodology in children, the paucity of reference values for differential cell counts, the variability of BAL findings at different times in a disease course, and the lack of correlation between BAL and histologic findings.

Fluid should be sent for differential cell counts, cultures and special staining for bacteria (including mycobacteria) and fungi, cytologic analysis (including oil red O staining for lipid-laden macrophages and staining for hemosiderin), pepsin levels, and viral diagnostic studies.

BAL findings can be diagnostic of pulmonary alveolar proteinosis (PAP), demonstrating a cloudy or milky appearance of the fluid with periodic acid-Schiff (PAS)–positive amorphous debris. Electron microscopy may reveal lamellar membranous structures in a fibrous and granular matrix with degenerative cells.Increased eosinophils (>30% of total) are consistent with eosinophilic pneumonia syndromes, whereas predominant lymphocytosis can be associated with hypersensitivity pneumonitis. CD1a-positive cells are diagnostic for Langerhans cell histiocytosis.

A study evaluated quantitation of levels of surfactant proteins B (SP-B) and C (SP-C) in BAL fluid for the diagnosis of chILD and reported that low SP-C levels may suggest diseases caused by mutations in *NKX2.1*, *SFTPC*, *ABCA3*, and other genes involved in surfactant metabolism. SP-B levels may be used to screen for SP-B deficiency; however, given the availability of genetic testing, this approach is not the standard of care for diagnosis.

### Lung biopsy

Analysis of tissue obtained during lung biopsy can be the best route to a diagnosis if it cannot be established by noninvasive means. For NEHI, an expert clinician can be confident of a presumptive diagnosis based on the clinical history and characteristic HRCT findings, but this is not true of most other chILD entities. An increasing number of chILDs can be diagnosed by genetic testing.

Much of the classification of ILD, especially in disorders of unknown cause, is based on histopathology. However, it is important to note that a histopathologic pattern is not synonymous with a diagnosis in many cases. To maximize the diagnostic yield, a pediatric lung biopsy protocol has been developed and supported by the ChILD Research Network.However, a diagnosis is not reached in all patients, even after biopsy is performed.

The number of biopsy procedures performed and the method used (eg, open vs thoracoscopic) have little influence on diagnostic yield. Diagnostic yield may be enhanced if HRCT is used to direct the biopsy sites. The biopsy sample should be taken from a region of involvement. If diffuse involvement is found, any site (except the tip of the right middle lobe or lingua) is appropriate.

Communication between the clinician, surgeon, pathologist, and radiologist before the biopsy is useful and appropriate for determining biopsy sites and prioritizing use of the tissue.

Compared with open lung biopsy, thoracoscopic biopsy shortens the surgical time, duration of chest tube placement, and hospital stay without substantially altering the diagnostic yield. The choice between thoracoscopic and open approaches should be left to the consulting surgeon. Transbronchial biopsies are not recommended for chILDs.

Regardless of the method used, biopsy samples should be processed for bacterial, fungal, and mycobacterial cultures and staining, including special staining, light microscopy, immunofluorescence, and electron microscopy. Immunostains, such as bombesin for NEHI and vimentin for pulmonary interstitial glycogenesis (PIG), may aid in the diagnosis of specific forms of ILD.

### Cardiac catheterization

This procedure should be considered in any child with noninvasive evidence of pulmonary hypertension but especially in children with a history of hemoptysis or absence of crackles on examination. These findings have been correlated with pulmonary veno-occlusive disease.

**PREDEFINED QUESTIONS AND ANSWERS**

## 1. What is Interstitial Lung Disease (ILD)?

Answer:  
ILD is a group of lung diseases that cause inflammation and scarring (fibrosis) of the lung tissue around the air sacs (alveoli). This thickening makes it harder for oxygen to pass into the blood, leading to breathing difficulties.

## 2. What is Childhood Interstitial Lung Disease (chILD)?

Answer:  
chILD refers to a diverse group of rare lung diseases affecting infants, children, and teenagers. These diseases cause similar symptoms like rapid breathing, cough, and difficulty getting enough oxygen due to changes in the lung tissue.

## 3. What causes ILD in children?

Answer:  
Causes vary widely and include genetic abnormalities, developmental lung disorders, immune system problems, infections, and exposure to harmful substances. Sometimes the exact cause is unknown.

## 4. What are the common symptoms of ILD or chILD?

Answer:  
Children with ILD often have rapid or difficult breathing, a dry cough, poor growth or weight gain, low oxygen levels, and may tire easily during physical activity.

## 5. How is ILD diagnosed?

Answer:  
Diagnosis involves ruling out more common lung diseases first. Doctors use chest imaging (X-rays, CT scans), lung function tests, blood tests, and sometimes lung biopsy or genetic testing to confirm ILD.

## 6. Can ILD be cured?

Answer:  
Many forms of ILD cannot be cured, but treatments can slow disease progression, reduce symptoms, and improve quality of life. In severe cases, lung transplantation may be considered.

## 7. What treatments are available for ILD?

Answer:  
Treatment depends on the type and cause but may include anti-inflammatory medications (like corticosteroids), oxygen therapy, medications to suppress the immune system, and supportive care such as nutrition and physical therapy.

## 8. How does ILD affect daily life?

Answer:  
ILD can cause fatigue and breathing difficulties that limit physical activities. Children may need ongoing medical care and support at school and home to manage symptoms and maintain growth.

## 9. Is ILD hereditary?

Answer:  
Some types of ILD have a genetic basis, especially in children. Genetic testing and family history can help identify inherited forms.

## 10. Where can I find support and more information?

Answer:  
Specialist centers, patient support groups, and organizations like Lung Foundation Australia, Boston Children’s Hospital, and national lung disease associations provide resources and support for families affected by ILD.

**Doctor-patient conversation about interstitial lung disease (ILD)**,

Doctor: "Hello, I want to talk with you about your recent diagnosis of interstitial lung disease, or ILD. This means that the tissue in your lungs has become inflamed and scarred, which can make it harder for your lungs to work properly."

Patient: "What causes this disease? Did I do something to cause it?"

Doctor: "ILD can have many causes. Sometimes it’s related to autoimmune diseases, exposure to harmful substances, infections, or medications. In some cases, we don’t know the exact cause. It’s important to understand that it’s not your fault — it’s a condition that happens due to changes in your lung tissue."

Patient: "What symptoms should I expect, and how will this affect my life?"

Doctor: "Common symptoms include shortness of breath, a dry cough, and feeling tired more easily. The disease can affect your ability to do physical activities, but with treatment and support, many people maintain a good quality of life. We’ll work together to manage your symptoms and slow the progression."

Patient: "What treatments are available?"

Doctor: "There are medications that can help slow down the scarring process, like antifibrotic drugs and immunosuppressants. We also focus on managing symptoms, such as providing oxygen therapy if needed and helping with cough and breathlessness. In some cases, lung transplantation may be considered."

Patient: "How will we know if the treatment is working?"

Doctor: "We’ll monitor your lung function regularly with tests and imaging. It’s important to have ongoing follow-up visits to adjust treatment as needed. Also, please let me know if your symptoms change or worsen."

Patient: "What about my prognosis? Will I get worse?"

Doctor: "ILD is a chronic condition, and it can progress over time, but the rate varies widely between individuals. Some people remain stable for years, especially with treatment. We’ll focus on maximizing your quality of life and addressing any concerns you have."

Patient: "Is there anything I can do to help myself?"

Doctor: "Yes, staying active within your limits, avoiding smoking and lung irritants, getting vaccinated against respiratory infections, and joining support groups can all help. It’s also important to communicate openly about how you’re feeling."

Patient: "This sounds overwhelming. How can I cope?"

Doctor: "It’s normal to feel overwhelmed. We have resources including patient education, counseling, and peer support groups. You’re not alone, and we’ll support you every step of the way.

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### **https://my.clevelandclinic.org/health/diseases/17809-interstitial-lung-disease**

### [**https://www.nhlbi.nih.gov/health/childhood-interstitial-lung-diseases**](https://www.nhlbi.nih.gov/health/childhood-interstitial-lung-diseases)

### **Popcorn lung**

## Common Alternative Names:

* Bronchiolitis obliterans
* Obliterative bronchiolitis
* Constrictive bronchiolitis
* Bronchiolitis obliterans syndrome (BOS) (especially in transplant patients)
* Chemical bronchiolitis
* Toxic bronchiolitis

**DEFINITION AND DESCRIPTION**

Bronchiolitis obliterans, also called popcorn lung, is a respiratory condition that affects the bronchioles of your lungs. The bronchioles are the smallest airways in your lungs. If you have this condition, these airways become inflamed, damaged and then scarred because of inhaling toxic substances or from infections. Other terms for this condition include obliterative bronchiolitis or constrictive bronchiolitis.

### **Why is bronchiolitis obliterans called popcorn lung?**

The name probably comes from when researchers first identified the disease among workers in a microwave popcorn factory. The workers had breathed in diacetyl, a flavoring chemical used to make the popcorn taste buttery.

Other industries used diacetyl for flavoring. Providers diagnosed workers in those other industries who had breathed in diacetyl with bronchiolitis obliterans. The liquid in electronic cigarettes (e-cigarettes) or vapes also contains diacetyl. There were also cases of the disease found in workers at a coffee roasting plant.

### **Who is more likely to get popcorn lung?**

Certain people have a higher risk of developing bronchiolitis obliterans because they come in close contact with toxic substances in the air. These chemicals are used in some types of manufacturing and can also be found in vapes and e-cigarettes.

#### **Toxic substances associated with developing popcorn lung**

* Acetaldehyde.
* Ammonia.
* Chlorine.
* Diacetyl.
* Formaldehyde.
* Fumes from metal oxides.
* Hydrochloric acid.
* Mustard gas or sulfur mustard.
* Nitrogen oxides.
* Sulfur dioxide.

**Medical conditions associated with developing popcorn lung**

* Infections, like respiratory syncytial virus, some types of pneumonia and some types of bronchitis.
* Stevens-Johnson syndrome.
* Rheumatic conditions like rheumatoid arthritis.
* Organ transplant recipients.

Popcorn lung is a rare disorder, but it can happen to anyone since it can result from an infection or exposure to certain substances.

Bronchiolitis obliterans can also occur without a specific exposure in people who've had a lung transplant. About 50% of people who have lung transplants will develop bronchiolitis obliterans syndrome within five years of their transplant procedure. About 10% of recipients of donor marrow also develop bronchiolitis obliterans syndrome within five years.

### **Bronchiolitis obliterans syndrome**

This condition, which results in reduced lung function because of the scarring to small airways in the lungs, is the most common type of lung transplant rejection among lung recipients. It can look like an infection at first.

The disease progresses in stages, but not the same way for everyone who has it. One person might stay in an early stage for quite some time, while another goes quickly from one stage to another more advanced stage. A lung function test called spirometry can determine how severe the disease is.

### **causes popcorn lung**

Popcorn lung isn’t contagious. You can’t get it from other people or give it to other people.

Popcorn lung happens from inhaling toxic chemicals, such as diacetyl, formerly used to flavor microwave popcorn. Exposure to these chemicals can happen at work or by vaping. E-cigarettes have many different types of chemicals in them that may be dangerous to your lung tissue. It’s thought that the vapor may affect not only the person vaping but also the people around them.

In people who've had a lung transplant, the disease can occur without exposure to a chemical or infection.

### **signs and symptoms of popcorn lung**

Signs and symptoms of popcorn lung include:

* Coughing, especially during and after exercise. Coughs may sometimes bring up mucus.
* Shortness of breath (dyspnea), especially during and after exercise.
* Wheezing.
* Tiredness.
* Fever.
* Night sweats.
* Skin rash.

Sometimes people who have popcorn lung don’t have symptoms initially.

**Diagnosis and Tests**

Your provider will ask you questions about your medical history and how you’re feeling. Because symptoms like difficulty breathing or being fatigued are common in other conditions, your doctor may recommend certain tests to clarify the diagnosis. These include:

* Imaging tests like a chest X-ray or [computed tomography (CT) scan](https://my.clevelandclinic.org/health/diagnostics/4808-ct-computed-tomography-scan).
* Lung function tests.
* Bronchoscopy.
* Lung biopsy.

## **Management and Treatment**

The damage from popcorn lung can be severe and reversal isn’t always possible. Management is likely to be more effective if your provider catches the disease early.

The first thing you should do is avoid exposure to the chemicals that cause popcorn lung. If you’re at a job that exposes you to these chemicals, you need to use the recommended protective equipment. If you’re smoking or vaping, you should quit.

Treatments for popcorn lung may include:

* Corticosteroids to fight inflammation, such as prednisone.
* Inhalers to help with breathing, such as those with albuterol.
* Oxygen therapy.
* Lung transplant, but this is recommended only in the most severe and extreme circumstances.

#### **Complications/side effects of corticosteroids**

Corticosteroids work very well to reduce inflammation, but they may cause a host of side effects. These side effects may include:

* Weight gain.
* Changes in mood.
* Nervousness or restlessness.
* Diabetes or causing existing diabetes to get worse.
* Trouble sleeping.

## **Outlook / Prognosis**

There’s no cure for popcorn lung. You’ll need to have life-long care to manage the symptoms, which may not always respond well to treatment.

**Prevention**

Start early by taking care of your lungs as best you can. You can do this in the following ways:

* Avoid using tobacco and e-cigarettes and avoid secondhand smoke and polluted places.
* Avoid infections when possible. Certain infections can damage your lungs.
* Follow your healthcare provider's suggestions on maintaining vaccine protection.
* If you work around dangerous substances, always wear personal protective equipment.

## **Living With**

Make sure that you tell your provider about any gastroesophageal reflux disease symptoms so they can be treated and not cause any additional harm to your lungs.

If you have a chronic illness, you might find it useful to join a support group. Sometimes sharing with others who have similar problems gives you answers and perspectives you might not find elsewhere. The support system may also be useful to caregivers, family and friends.

### **When should I see my healthcare provider?**

Contact your healthcare provider if you have breathing difficulties that aren’t relieved by your rescue inhaler. Call if you have any symptoms that are new or get worse.

**DIFFERENTIAL DIAGNOSIS**

* Asthma
* Bronchogenic carcinoma
* Bronchiectasis
* Cystic fibrosis
* Interstitial lung disease
* Pleural effusion
* Pulmonary edema
* Recurrent aspiration
* Recurrent pulmonary emboli
* Tracheobronchomalacia

**EPIDEMIOLOGY**

Bronchiolitis obliterans syndrome is considered a form of chronic allograft rejection after lung transplantation. The majority of lung transplant recipients who are long term survivors develop bronchiolitis obliterans syndrome. More than 50% of recipients will develop some degree of BO by 5 years post-transplant. The average time to diagnose BO is 16 to 20 months after lung transplant but has been reported as early as 3 months after transplantation. About 5% to 14% of Hematopoietic stem cell transplantation (HSCT) recipients also develop bronchiolitis obliterans syndrome, which is pulmonary graft vs. host disease and can present several months to years later after transplantation.

**Doctor-patient conversation about bronchiolitis obliterans (BO)**

Doctor: "I want to talk to you about a condition called bronchiolitis obliterans, or BO. It’s a rare lung disease that causes inflammation and scarring in the small airways of the lungs, which can make it harder to breathe."

Patient: "How did I get this? Is it contagious?"

Doctor: "BO is not contagious. It can happen after a severe lung infection, exposure to certain chemicals, or sometimes after a lung or stem cell transplant. The inflammation causes permanent narrowing of the small airways."

Patient: "What symptoms should I expect?"

Doctor: "Common symptoms include a persistent dry cough, shortness of breath, wheezing, and feeling tired easily. These symptoms can vary depending on how much your airways are affected."

Patient: "How do you diagnose it?"

Doctor: "We start by reviewing your symptoms and medical history. Tests like lung function tests and high-resolution CT scans help us see the extent of airway narrowing. Sometimes, we may use bronchoscopy or biopsy, but that’s less common."

Patient: "Is there a cure?"

Doctor: "There’s no cure, but we have treatments that can slow the disease and help with symptoms. These include medications like steroids to reduce inflammation and antibiotics like azithromycin. Oxygen therapy may be needed if your oxygen levels are low."

Patient: "Will I need surgery?"

Doctor: "Surgery is rarely needed, but in very severe cases, a lung transplant might be considered."

Patient: "What can I do to help myself?"

Doctor: "Avoid lung irritants like smoke, follow your treatment plan, attend all follow-ups, and report any worsening symptoms promptly. Support from a team of specialists will help manage your condition."

Patient: "What is the outlook?"

Doctor: "The outlook varies. Some people improve with treatment, others may have stable disease, and some may worsen. We will monitor you closely and adjust treatment as needed."

**PREDEFINED Questions and answers**

### **What is the difference between bronchiolitis obliterans and bronchiolitis obliterans organizing pneumonia?**

Bronchiolitis obliterans and bronchiolitis obliterans organizing pneumonia (BOOP) are two different diseases. Both diseases affect the bronchioles, but the cause of BOOP is infections, drugs or other diseases. The new name of BOOP is cryptogenic organizing pneumonia (COP) if there is no known cause, and most cases of BOOP have no known cause. The most common symptom is a cough that lasts for some time and doesn’t bring up secretions.

### **Can popcorn lung fix itself?**

The short answer to this is no. Bronchiolitis obliterans is irreversible. Once the damage happens, you can’t fix it. You can only try to stop making it worse. Without treatment, it could be fatal.

## Commonly Used BOS Staging System

|  |  |  |
| --- | --- | --- |
| Stage | FEV₁ (% of baseline) | Description |
| 0-p (Potential BOS) | 81–90% of baseline and/or FEF25–75 ≤ 75% of baseline | Early or potential BOS; subtle decline in lung function, may predict progression |
| 1 | 66–80% of baseline | Mild BOS; moderate decline in FEV₁ indicating airway obstruction |
| 2 | 51–65% of baseline | Moderate BOS; further decline in lung function |
| 3 | ≤50% of baseline | Severe BOS; significant airway obstruction and impaired lung function |

* FEF25–75% (forced expiratory flow at 25–75% of pulmonary volume) may also be used as an early marker but is less specific than FEV₁

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[Popcorn Lung: Causes, Symptoms, Treatment & Is It Real](https://my.clevelandclinic.org/health/diseases/22590-popcorn-lung-bronchiolitis-obliterans#overview)

[Learn About Bronchiolitis Obliterans (Popcorn Lung) | American Lung Association](https://www.lung.org/lung-health-diseases/lung-disease-lookup/popcorn-lung/learn-about-popcorn-lung)

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**PULMONARY FIBROSIS**

## Common Alternative Names:

* Idiopathic Pulmonary Fibrosis (IPF) — when the cause is unknown
* Interstitial Pulmonary Fibrosis
* Fibrotic Interstitial Lung Disease
* Usual Interstitial Pneumonia (UIP) — a specific histopathologic pattern often seen in IPF
* Diffuse Pulmonary Fibrosis
* Fibrosing Alveolitis (older term)
* Pulmonary Interstitial Fibrosis
* Chronic Interstitial Pneumonitis

**DEFINITION AND DESCRIPTION**

Pulmonary fibrosis is a lung disease that occurs when lung tissue becomes damaged and scarred. This thickened, stiff tissue makes it harder for the lungs to work properly. Pulmonary fibrosis worsens over time. Some people can stay stable for a long time, but the condition gets worse faster in others. As it gets worse, people become more and more short of breath.

The scarring that happens in pulmonary fibrosis can be caused by many things. Often, doctors and other healthcare professionals cannot pinpoint what's causing the problem. When a cause cannot be found, the condition is called idiopathic pulmonary fibrosis.

Idiopathic pulmonary fibrosis usually occurs in middle-aged and older adults. Sometimes pulmonary fibrosis is diagnosed in children and infants, but this is not common.

The lung damage caused by pulmonary fibrosis cannot be repaired. Medicines and therapies can sometimes help slow down the rate of fibrosis, ease symptoms and improve quality of life. For some people, a lung transplant might be an option.

**Causes**

Pulmonary fibrosis is scarring and thickening of the tissue around and between the air sacs called alveoli in the lungs. These changes make it harder for oxygen to pass into the bloodstream.

Damage to the lungs that results in pulmonary fibrosis may be caused by many different things. Examples include long-term exposure to certain toxins, radiation therapy, some medicines and certain medical conditions. In some cases, the cause of pulmonary fibrosis is not known.

### **Your work and surroundings**

The type of work you do and where you work or live could be the cause or part of the cause for pulmonary fibrosis. Having continuous or repeated contact with toxins or pollutants — substances that harm the quality of water, air or land — can damage your lungs, especially if you do not wear protective gear. Examples include:

* Silica dust.
* Asbestos fibers.
* Hard metal dusts.
* Wood, coal and grain dusts.
* Mold.
* Bird and animal droppings.

### **Radiation treatments**

Some people who receive radiation therapy to the chest, such as for lung or breast cancer, show signs of lung damage months or sometimes years after treatment. How severe the damage is may depend on:

* How much of the lung was exposed to radiation.
* The total amount of radiation given.
* Whether chemotherapy also was used.
* Whether there is underlying lung disease.

### **Medicines**

Many medicines can damage the lungs. Some examples include:

* **Chemotherapy.** Medicines designed to kill cancer cells, such as methotrexate (Trexall, Otrexup, others), bleomycin and cyclophosphamide (Cytoxan), can damage lung tissue.
* **Heart medicines.** Some medicines used to treat irregular heartbeats, such as amiodarone (Nexterone, Pacerone), may harm lung tissue.
* **Some antibiotics.** Antibiotics such as nitrofurantoin (Macrobid, Macrodantin) or ethambutol (Myambutol) can cause lung damage.
* **Anti-inflammatory medicines.** Certain anti-inflammatory medicines such as rituximab (Rituxan) or sulfasalazine (Azulfidine) can cause lung damage.

### **Medical conditions**

Lung damage can also result from a number of conditions, including:

* **Dermatomyositis,** an inflammatory disease marked by muscle weakness and a skin rash.
* **Lupus,** a disease that occurs when the body's immune system attacks its own tissues and organs.
* **Mixed connective tissue disease,** which has a mix of symptoms of different disorders, such as lupus, scleroderma and polymyositis.
* **Pneumonia,** an infection that inflames the air sacs in one or both lungs.
* **Polymyositis,** an inflammatory disease that causes muscle weakness on both sides of the body.
* **Rheumatoid arthritis,** an inflammatory disease that affects joints and other body systems.
* **Sarcoidosis,** an inflammatory disease that most often affects the lungs and lymph nodes.
* **Scleroderma,** a group of rare diseases that involve hardening and tightening of the skin as well as problems inside the body.

### **Idiopathic pulmonary fibrosis**

Many substances and conditions can lead to pulmonary fibrosis. Even so, in many people, the cause is never found. But risk factors such as smoking or exposure to air pollution could be related to the condition, even if the cause cannot be confirmed. Pulmonary fibrosis with no known cause is called idiopathic pulmonary fibrosis.

Many people with idiopathic pulmonary fibrosis also may have gastroesophageal reflux disease, also called GERD. This condition occurs when acid from the stomach flows back into the esophagus. GERD may be a risk factor for idiopathic pulmonary fibrosis or cause the condition to worsen faster. But more studies are needed.

**Risk factors**

Pulmonary fibrosis has been found in children and infants, but this is not common. Idiopathic pulmonary fibrosis is much more likely to affect middle-aged and older adults. Other types of pulmonary fibrosis, such as that caused by connective tissue disease, can occur in younger people.

Factors that can raise your risk of pulmonary fibrosis include:

* **Smoking.** If you smoke now or used to smoke, you're at a higher risk of pulmonary fibrosis than people who never smoked. People with emphysema are at higher risk, too.
* **Certain types of work.** You have a higher risk of developing pulmonary fibrosis if you work in mining, farming or construction. The risk also is higher if you have continuous or repeated contact with pollutants known to damage the lungs.
* **Cancer treatments.** Having radiation treatments to your chest or using certain chemotherapy medicines can raise your risk of pulmonary fibrosis.
* **Genetics.** Some types of pulmonary fibrosis run in families, so genes may play a role.

**Symptoms**

Symptoms of pulmonary fibrosis may include:

* Shortness of breath.
* Dry cough.
* Extreme tiredness.
* Weight loss that's not intended.
* Aching muscles and joints.
* Widening and rounding of the tips of the fingers or toes, called clubbing.

How fast pulmonary fibrosis worsens over time and how severe the symptoms are can vary greatly from person to person. Some people become ill very quickly with severe disease. Others have moderate symptoms that worsen more slowly, over months or years.

### **When symptoms suddenly get worse**

In people with pulmonary fibrosis, especially idiopathic pulmonary fibrosis, shortness of breath can suddenly get worse over a few weeks or days. This is called an acute exacerbation. It can be life-threatening. The cause of an acute exacerbation may be another condition or an illness, such as a lung infection. But usually the cause is not known.

### **When to see a doctor**

If you have symptoms of pulmonary fibrosis, contact your doctor or other healthcare professional as soon as possible. If your symptoms get worse, especially if they get worse fast, contact your healthcare team right away.

## **Diagnosis**

To diagnose pulmonary fibrosis, your doctor or other healthcare professional reviews your medical and family history and does a physical exam. You can talk about your symptoms and review any medicines you take. You also will likely be asked about any continuous or repeated contact with dusts, gases, chemicals or similar substances, especially through work.

During the physical exam, your healthcare professional listens carefully to your lungs while you breathe. Pulmonary fibrosis often occurs along with a crackling sound at the base of the lungs.

You may have one or more of these tests.

### **Imaging tests**

* **Chest X-ray.** Images of the chest may show the scar tissue that is usually part of pulmonary fibrosis. Sometimes the chest X-ray may not show any changes. More tests may be needed to find out why you are short of breath.
* **Computerized tomography (CT) scan.** A CT scan combines X-ray images taken from many different angles to create images of structures inside the body. A high-resolution CT scan can be helpful in diagnosing pulmonary fibrosis and in finding out how much lung damage has occurred. Some kinds of fibrosis have certain patterns.
* **Echocardiogram.** An echocardiogram uses sound waves to look at the heart. The test can create pictures of the heart's structures. It also can create videos that show how the heart is working. This test can tell the amount of pressure in the arteries of the lungs and in the right side of the heart.

### **Lung function tests**

Also called pulmonary function tests, these are done to find out how well your lungs are working:

* **Spirometry.** In this test, you breathe out quickly and forcefully through a tube connected to a machine. The machine measures how much air the lungs can hold and how quickly air moves in and out of the lungs.
* **Lung volume test.** This test measures the amount of air the lungs hold at different times when breathing in and out.
* **Lung diffusion test.** This test shows how well the body moves oxygen and carbon dioxide between the lungs and the blood.
* **Pulse oximetry.** This simple test uses a small device placed on one of the fingers to measure how much oxygen is in the blood. The percentage of oxygen in the blood is called oxygen saturation. Your healthcare professional may recommend a six-minute walking test with a check of your oxygen saturation.
* **Exercise stress test.** An exercise test on a treadmill or stationary bike may be used to monitor heart and lung function during activity.
* **Arterial blood gas test.** In this test, a sample of blood, usually taken from an artery in the wrist, is tested. The oxygen and carbon dioxide levels in the sample are measured.

In addition to showing whether you have pulmonary fibrosis, imaging and lung function tests can be used to check your condition over time and see how treatments are working.

### **Tissue sample**

If other tests cannot find the cause of your condition, a small amount of lung tissue may need to be removed. This is called a biopsy. The biopsy sample is then examined in a laboratory to diagnose pulmonary fibrosis or rule out other conditions. One of these methods can be used to get a tissue sample:

* **Surgical biopsy.** Although a surgical biopsy is invasive and has potential complications, it may be the only way to make the right diagnosis. This procedure may be done as a minimally invasive surgery called video-assisted thoracoscopic surgery (VATS). The biopsy also may be done as an open surgery called a thoracotomy.

During VATS, a surgeon inserts surgical instruments and a small camera through two or three small cuts between the ribs. The surgeon looks at the lungs on a video monitor while removing tissue samples from the lungs. During the surgery, a combination of medicines put you in a sleep-like state called general anesthesia.

During a thoracotomy, a surgeon removes a lung tissue sample through a cut that opens the chest between the ribs. This open surgery also is done using general anesthesia.

* **Bronchoscopy.** In this procedure, very small tissue samples are removed — usually no larger than the head of a pin. A small, flexible tube called a bronchoscope is passed through the mouth or nose into the lungs to remove the samples. The tissue samples are sometimes too small to make the right diagnosis. But this form of biopsy also may be used to rule out other conditions.

### **Blood tests**

You may have blood tests to look at your liver and kidney function. Blood tests also can check for and rule out other conditions.

**Treatment**

The lung scarring and thickening that occurs in pulmonary fibrosis cannot be repaired. And no current treatment has proved effective in stopping the disease from getting worse over time. Some treatments may improve symptoms for a time or slow how fast the disease worsens. Others may help improve quality of life.

Treatment depends on the cause of your pulmonary fibrosis. Doctors and other healthcare professionals evaluate how severe your condition is. Then together you can decide on the best treatment plan.

### **Medicines**

If you have idiopathic pulmonary fibrosis, your healthcare professional may recommend the medicine pirfenidone (Esbriet) or nintedanib (Ofev). Both are approved by the U.S. Food and Drug Administration (FDA) for idiopathic pulmonary fibrosis. Nintedanib also is approved for other types of pulmonary fibrosis that get worse quickly. These medicines may help slow the worsening of pulmonary fibrosis and may prevent bouts when symptoms suddenly get worse.

Nintedanib can cause side effects such as diarrhea and nausea. Side effects of pirfenidone include nausea, loss of appetite and skin rash from sunlight. With either medicine, your healthcare professional uses regular blood tests to check how well the liver is working.

New medicines and therapies are being developed or tested in clinical trials but are not yet approved by the Food and Drug Administration (FDA). Researchers continue to study medicines to treat pulmonary fibrosis.

Doctors may recommend anti-acid medicines if you have symptoms of gastroesophageal reflux disease (GERD). GERD is a digestive condition that commonly occurs in people with idiopathic pulmonary fibrosis.

### **Oxygen therapy**

Using extra oxygen, called supplemental oxygen, cannot stop lung damage, but it can:

* Make breathing and exercise easier.
* Prevent or lessen complications from low blood oxygen levels.
* Possibly lessen strain on the right side of the heart.
* Improve sleep and sense of well-being.

You may use oxygen when you sleep or exercise. But some people need oxygen all the time. Carrying a small tank of oxygen or using a portable oxygen concentrator can help you be more mobile.

### **Pulmonary rehabilitation**

Pulmonary rehabilitation can help manage your symptoms and improve your ability to do daily tasks. Pulmonary rehabilitation programs focus on:

* Physical exercise to improve how much you can do.
* Breathing techniques that may improve how well your lungs use oxygen.
* Nutritional counseling.
* Emotional counseling and support.
* Education about your condition.

### **When symptoms suddenly get worse**

When symptoms suddenly get worse, called an acute exacerbation, you may need more supplemental oxygen. In some cases, you may need mechanical ventilation in the hospital. In this treatment, a tube is guided into the lungs and attached to a machine that helps with breathing. Your healthcare professional may recommend antibiotics, corticosteroid medicines or other medicines when symptoms suddenly get worse.

### **Lung transplant**

A lung transplant may be an option for some people with pulmonary fibrosis. Having a lung transplant can improve your quality of life and allow you to live a longer life. But a lung transplant can involve complications such as rejection and infection. After a lung transplant, you take medicines for the rest of your life. You and your healthcare team may discuss a lung transplant if it's thought to be the right treatment option for your condition.

**Lifestyle and home remedies**

Being actively involved in your treatment and staying as healthy as possible are essential to living with pulmonary fibrosis. It's important to:

* **Stop smoking.** If you have lung disease, it is important to stop smoking. Talk with your healthcare team about options for quitting, including smoking cessation programs. These use proven techniques to help people quit. Because secondhand smoke can be harmful to your lungs, avoid being around people who are smoking.
* **Avoid other things that can irritate your lungs.** Breathing indoor pollutants, such as fumes from heating fuel or chemicals, can irritate your lungs. So can outdoor pollutants, such as dust or car exhaust.
* **Eat well.** People with lung disease may lose weight both because eating is not comfortable and because of the extra energy it takes to breathe. A healthy diet that contains enough calories is needed. Try to eat smaller meals more often during the day. A dietitian can give you more information on healthy eating for your condition.
* **Get moving.** Regular exercise can help you keep your lung function and manage your stress. Aim to include physical activity, such as walking or biking, into your daily routine. Talk to your healthcare team about what activities may be best for you. If over time you need help getting around, such as using a wheelchair, look for active movements you can do that do not require walking. One example is tai chi.
* **Take time to rest.** Make sure to get enough rest. Taking time to rest can help you have more energy and cope with the stress of your condition. If you have problems sleeping, talk with your healthcare team.
* **Get vaccinated.** Respiratory infections, such as colds and flu, can worsen symptoms of pulmonary fibrosis. Make sure that you get the pneumonia vaccine, an annual flu shot and COVID-19 vaccines. It's important that your family members also be vaccinated. Try to stay out of crowds when possible.
* **Follow your treatment plan.** You usually need ongoing treatment from your healthcare team. Follow the care team's instructions. Take your medicines as prescribed. Adjust your diet and exercise as needed. Attend pulmonary rehabilitation sessions. Go to all of your appointments and contact your care team if symptoms worsen.

**Complications**

Complications of pulmonary fibrosis may include:

* **High blood pressure in the lungs.** Called pulmonary hypertension, this type of high blood pressure affects the arteries in the lungs. These are the pulmonary arteries. Stiff and thick arteries may slow down or block blood flow through the lungs. This raises the pressure inside the pulmonary arteries and the lower right heart chamber, called the right ventricle.
* **Right-sided heart failure.** This serious condition occurs when your heart's right chamber has to pump harder than usual to move blood through partly blocked pulmonary arteries.
* **Respiratory failure.** This is often the last stage of long-term lung disease. It occurs when blood oxygen levels fall dangerously low.
* **Lung cancer.** Long-standing pulmonary fibrosis increases your risk of developing lung cancer.
* **Other lung problems.** As pulmonary fibrosis gets worse over time, it may lead to serious problems such as blood clots in the lungs, a collapsed lung or lung infections.

**Outlook / Prognosis**

Lung scarring is nearly always permanent (unless caused by a medication and caught early). If you have an underlying disease, management of it might help prevent further damage. If the cause is unknown, your healthcare provider will treat your symptoms and try to prevent more damage.

Healthcare providers can’t easily predict how pulmonary fibrosis will progress. Your symptoms may get worse very slowly, over years. In some cases, the disease may lead to severe symptoms quickly (over months).

#### **How long can a person live with pulmonary fibrosis?**

The life expectancy of someone with the most common form, idiopathic pulmonary fibrosis, is three to five years. But life expectancies for people with PF have been getting longer in recent years.

**Prevention**

Many causes of pulmonary fibrosis aren’t preventable. You can reduce your risk of lung scarring from environmental exposures by:

* Avoiding substances that can harm your lungs, such as asbestos, metal dust or chemicals, and wearing a respirator (a mask that filters particles from the air) if you have to work with them
* Avoiding things that can cause chronic allergic reactions, like hay, grain, bird droppings or feathers, and heating and cooling systems — and wearing a respirator mask if you have to work with them
* Not smoking or quitting smoking

## **Differential Diagnoses**

* Acute Respiratory Distress Syndrome (ARDS)
* Amyloidosis
* Aspiration Pneumonitis and Pneumonia
* Cardiogenic Pulmonary Edema
* Drug-Induced Pulmonary Toxicity
* Malignancy
* Nitrogen Dioxide Toxicity
* Pneumocystis jiroveci Pneumonia (PJP)
* Pulmonary Alveolar Proteinosis
* Pulmonary Embolism (PE)
* Pulmonary Eosinophilia
* Restrictive Lung Disease
* Viral Pneumonia

## **Epidemiology**

### Frequency

As a group, diffuse interstitial diseases of the lung are uncommon. Of patients referred to a pulmonary disease specialist, an estimated 10-15% have a DPLD.

In the United States, the 2021 Global Burden of Disease (GBD) study estimated over 650,000 cases, with an age-standardized prevalence varying from 101 to 156 cases per 100,000 people among states.

### Race

One multicenter study that included 5275 ILD patients in the United States showed a racial and ethnic group breakdown of 83.2% White, 10.2% Black, and 6.7% Hispanic patients. The etiology of ILD varied among racial and ethnic groups, with IPF having the highest prevalence in White patients, HP having the highest prevalence in Hispanic patients, and CTD-ILD having the highest prevalence in Black patients. Black patients were more likely than Hispanic and White patients to be hospitalized, undergo lung transplant, and die at a younger age.

### Sex

Evaluation of data from the GBD study showed a higher prevalence of ILD among females, with age-adjusted prevalence of 131.4 per 100,000, compared to 121.3 per 100,000 in males in the United States.

Several DPLDs show sex-related differences in frequency. In general, IPF affects men more than women (at a ratio of 1.5:1), while LAM and pulmonary tuberous sclerosis almost exclusively affect women. Women are much more likely to develop CTD-ILD than men and thus are more likely to experience pulmonary manifestations of those diseases. However, when affected, men with certain rheumatologic diseases (e.g. RA) are more likely to develop pulmonary manifestations than women. The pneumoconiosis (e.g., silicosis) are much more common in men than in women, which may be due to higher rates of occupational exposure.

### Age

Many of the DPLDs develop over many years and therefore are more prevalent in older adults. For example, most patients with IPF present in the sixth or greater decade of life. Others form of interstitial lung disease, such as sarcoidosis, LAM, connective-tissue disease–associated lung disease, and inherited forms of lung disease primarily present in younger adults.

## **Procedures**

The role of lung biopsy remains controversial, with expert opinion weighing in on both sides. Consensus appears to be building on the side of forgoing biopsy when the typical clinical and high-resolution CT scan features of a specific disease process are present.

A multidisciplinary discussion is an essential part of diagnosis of ILD and deciding whether biopsy is necessary. This discussion often includes pulmonologists, radiologists, pathologists, thoracic surgeons, and rheumatologists. Multidisciplinary discussions can improve diagnosis by involving more specialties in the discussion, minimizing the effects of interobserver variability, and potentially avoiding invasive procedures.

Transbronchial and endobronchial lung biopsies may be diagnostic, particularly for sarcoidosis, HP, or lymphangitic spread of carcinoma but frequently are not useful for other diagnoses. This is due to the patchy and often peripheral, rather than airway-centric, distribution of the majority of these diseases.

Surgical lung biopsy is regarded as the gold standard diagnostic test, however the most recent guidelines suggest that transbronchial cryobiopsy may be an acceptable alternative, in centers with sufficient experience performing the procedure. A balance of risks of these procedures must be weighed with the likelihood of obtaining a diagnostic result.

Bronchiolar lavage (BAL) may be helpful in differentiating some types of DPLD from others. Bronchiolar lavage is useful in evaluating the possibility of infection or malignancy. Cell count and differential analysis can differentiate DPLDs that have particular associations with lymphocyte-, neutrophilic-, eosinophilic predominance. For example, IPF is associated with neutrophil predominance, while HP is associated with lymphocyte predominance. Special stains can provide diagnostic value, as in the case of Periodic acid Schiff stain positivity for pulmonary alveolar proteinosis (PAP).

**Predisposition to Pulmonary Fibrosis: Genes and Distinguishing Clinical Features**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|  | **Gene 1** | **% of all FPF** | **MOI** | **Age of Onset of Pulmonary Fibrosis** | **Distinguishing Features** |
| **Genes associated w/telomere maintenance 2** | *DKC1* | ~1% 3 | XL 4 | Adult | Short telomere syndrome features |
| *NAF1* | ~1% 5, 6 | AD | Adult | Combined PF & emphysema; short telomere syndrome features |
| *PARN* | 4%-5% 5, 7 | AD AR | Mean: 64 yrs |  |
| *RTEL1* | 4%-9% 5, 8 | AD | Mean: 60 yrs |  |
| *TERC* | 2%-5% 9 | AD | Mean: 51 yrs | Short telomere syndrome features |
| *TERT* | 15%-20% 5, 10, 11 | AD | >40 yrs (mean: 58 yrs; PF in >60% of those age >60 yrs) | Some persons have combined PF & emphysema |
| *TINF2* | ~1% 12 | AD | Adult | Short telomere syndrome features |
| *ZCCHC8* | <1% | AD | Adult | ± bone marrow failure |
| **Genes associated w/surfactant metabolism** | *ABCA3* | <1% 13 | AR | Rare in adults, more common in childhood-onset PF | Lung radiographs show ground glass opacities, reticulations, & cysts of variable size, predominantly involving upper lobes |
| *SFTPA1* | <1% 14, 15 | AD | Adult | Lung adenocarcinoma |
| *SFTPA2* | <1% 14, 16 | AD | Adult | Lung adenocarcinoma |
| *SFTPC* | 1%-5% 14, 17 | AD | Infancy to late adulthood | Wide array of radiographic abnormalities, incl combined PF & emphysema, & atypical upper lobe involvement 18 |

## **PREDEFINED Questions and answers**

### **What are the stages of pulmonary fibrosis?**

While there’s no official staging system for pulmonary fibrosis, some providers may describe pulmonary fibrosis as mild, moderate, severe or very severe. They base this diagnosis on your symptoms, lung function tests and imaging.

#### **At what age does pulmonary fibrosis start?**

Most cases of pulmonary fibrosis are diagnosed in people 65 and older

**DOCTOR PATIENT CONVERSATION**

Doctor: "Hello, I want to discuss the results of your tests. You have a condition called pulmonary fibrosis, which means there is scarring in your lungs that makes it harder for oxygen to pass into your bloodstream."

Patient: "What causes this? Is it something I did?"

Doctor: "Pulmonary fibrosis can have many causes, including environmental exposures, certain medications, infections, or autoimmune diseases. Sometimes, we don’t find a specific cause, which we call idiopathic pulmonary fibrosis. It’s not your fault, and it’s not contagious."

Patient: "What symptoms should I expect?"

Doctor: "Common symptoms include shortness of breath, especially when active, a dry cough, and fatigue. These symptoms can slowly get worse over time."

Patient: "Is there a cure?"

Doctor: "Currently, there is no cure, but we have treatments that can slow the progression of the disease and help you manage symptoms. We’ll also focus on keeping you as comfortable as possible and maintaining your quality of life."

Patient: "What treatments are available?"

Doctor: "There are medications called antifibrotics that can slow lung scarring. We may also recommend oxygen therapy if your oxygen levels are low, pulmonary rehabilitation to help with breathing and exercise, and support services."

Patient: "How will this affect my life?"

Doctor: "Pulmonary fibrosis is a chronic condition, and it can impact your daily activities. We’ll work together to monitor your lung function regularly and adjust treatments as needed. It’s important to avoid smoking and lung irritants, stay active within your limits, and get vaccinated against respiratory infections."

Patient: "What should I do if my symptoms get worse?"

Doctor: "If you notice increased shortness of breath, coughing, or fatigue, please contact us promptly. Early intervention can help manage flare-ups and complications."

Patient: "This sounds overwhelming. How can I cope?"

Doctor: "It’s normal to feel overwhelmed. We have resources including patient education, counseling, and support groups. Our team, including nurses and therapists, will support you throughout your journey. Please feel free to ask any questions at any time."

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**NEUROENDOCRINE HYPERPLASIA OF INFANCY**

**ALTERNATIVE NAMES**

* chronic tachypnea of infancy (CTI)
* persistent tachypnea of infancy (PTI)
* NEHI

**DEFINITION AND DESCRIPTION**

Neuroendocrine hyperplasia of infancy (NEHI), is a rare lung disease that affects children, typically presenting in the first two years of life. NEHI typically presents in otherwise healthy infants during the first months to the first year of life. It is characterized by persistent rapid and labored breathing (tachypnea), crackles and low oxygen levels (hypoxemia).

The cause of NEHI is currently poorly understood.The prevalence is not well known, but it is considered a rare disease.

The hallmark diagnostic features of NEHI on chest computed tomography (CT) include hyperinflation and ground-glass opacities in characteristic distributions, without other parenchymal abnormalities.2,7 While symptoms such as breathing difficulties and low oxygen levels tend to improve with time, they can persist for years, underscoring the importance of early diagnosis and long-term management.

Some patients undergo a lung biopsy which shows an increased number of pulmonary neuroendocrine cells (PNEC). The role of PNECs in NEHI is not well understood. Treatment is largely supportive including supplemental oxygen and ensuring the patient has adequate growth

### **Causes**

The cause of NEHI remains unknown, but there is evidence suggesting genetic factors may play a role. A change (variant) in the *NKX2-1* gene was identified in a patient with NEHI, along with family members who had childhood lung disease, indicating a possible genetic link. While increased numbers of pulmonary neuroendocrine cells (PNECs) are found in NEHI patients, it is not clear whether these cells are directly involved in the development of the disease or serve as markers of the disorder.These cells play a role in oxygen sensing and lung epithelial regeneration, potentially contributing to disease symptoms. However, more research is needed to understand the full range of influences on NEHI.

### **Signs & Symptoms**

Children with NEHI often present in the first year of life with:1,2,4,6,7

– Rapid, shallow breathing (tachypnea)

– Increased work of breathing, including retractions

– Low oxygen levels in the blood (hypoxemia)

– Crackling sounds (crackles) heard with a stethoscope

– Failure to gain weight (failure to thrive)

These symptoms can often be mistaken for more common conditions, leading to initial misdiagnoses of asthma or recurrent respiratory infections. Unlike asthma, wheezing is not a common symptom, and children with NEHI do not typically respond to asthma medications such as inhaled corticosteroids.

### **Disorders with Similar Symptoms**

Asthma/reactive airway disease

Bronchiolitis

### **Diagnosis**

Doctors typically diagnose NEHI by looking at symptoms and using specialized tests. A high-resolution CT scan of the chest is a key tool. It can show characteristic patterns like ground-glass opacities (which look hazy on the scan) and air trapping, both of which are common in NEHI. Additionally, infant pulmonary function tests (PFTs) may help confirm the diagnosis by detecting signs of air trapping in the lungs.

In most people, these noninvasive imaging and testing methods are enough to make a diagnosis of NEHI without needing more invasive procedures. However, if the imaging results are unclear or the symptoms are not typical of NEHI, doctors might need to do a lung biopsy. During this procedure, a small piece of lung tissue is removed and analyzed. The biopsy is stained with a special marker (called bombesin) to identify the increased presence of pulmonary neuroendocrine cells (PNECs). Since biopsies are more invasive, they are usually only done in cases where the noninvasive image tests are not conclusive. In most people, NEHI can now be confidently diagnosed through clinical symptoms and imaging tests alone.

### **Standard Therapies**

There is no cure or specific treatment for NEHI, and management is primarily supportive.

Most of the affected children require supplemental oxygen to address low oxygen levels in the blood (hypoxemia) and nutritional support is often necessary to manage growth delays.

The use of medications such as inhaled corticosteroids typically used for asthma, has not been found to be effective in NEHI.

Immunizations, including the influenza and pneumococcal vaccines are crucial in preventing further respiratory complications.

Genetic counseling may be recommended, especially in cases where familial lung disease is present. Long-term supportive care is recommended for families coping with a diagnosis of NEH

**DIFFERENTIAL DIAGNOSIS**

* Other Childhood Interstitial Lung Diseases (chILD):
  + Surfactant dysfunction disorders (e.g., SFTPB, SFTPC, ABCA3 mutations)
  + Pulmonary interstitial glycogenosis (PIG)
  + Diffuse developmental lung disorders (e.g., acinar dysplasia, alveolar capillary dysplasia)
  + Alveolar growth abnormalities (e.g., chronic neonatal lung disease, pulmonary hypoplasia)
* Hypersensitivity Pneumonitis (HP)
* Infectious Pneumonitis (viral or bacterial infections causing chronic symptoms)
* Bronchiolitis Obliterans
* Congenital Lung Malformations (e.g., congenital pulmonary airway malformation, bronchopulmonary sequestration)
* Primary Ciliary Dyskinesia
* Cystic Fibrosis
* Asthma or Reactive Airway Disease (usually older children, reversible airway obstruction)
* Pulmonary Vascular Disease (rare in infants but can cause respiratory distress)

## **Epidemiology of Neuroendocrine Cell Hyperplasia of Infancy (NEHI)**

* Prevalence:  
  The exact prevalence of NEHI is unknown. It is considered a rare pediatric interstitial lung disease, but likely underdiagnosed due to overlapping symptoms with other lung diseases and the limited need for lung biopsy in mild cases.
* Age of Onset:  
  NEHI primarily affects infants, with the mean age of presentation around 3 to 4 months. Most cases occur within the first year of life, but older children can also be affected.
* Gender Distribution:  
  Some studies report a male predominance (about 66% male in one large series).
* Clinical Presentation:  
  Infants typically present with persistent tachypnea (rapid breathing), chest retractions, hypoxemia (low oxygen levels), crackles on lung auscultation, and sometimes failure to thrive.
* Prognosis:  
  The prognosis is generally good, with many infants improving over time. However, some patients may have persistent respiratory symptoms, oxygen requirement, or recurrent infections for months or years.
* Etiology:  
  The cause of NEHI remains unknown, but genetic factors are suspected given rare familial cases reported. It is not inherited in a typical Mendelian pattern, and no clear environmental cause has been identified.
* Diagnostic Imaging:  
  High-resolution CT typically shows patchy ground-glass opacities, especially in the right middle lobe and lingula, along with air trapping

## **Predefined Questions and Answers for NEHI**

1. What is Neuroendocrine Cell Hyperplasia of Infancy (NEHI)?  
NEHI is a rare lung disease in infants where there is an increased number of neuroendocrine cells in the small airways. This causes inflammation and narrowing of the airways, leading to breathing difficulties.

2. What causes NEHI?  
The exact cause of NEHI is unknown. It is not inherited or contagious. It may be related to abnormal lung development or repair after injury, but research is ongoing.

3. What are the common symptoms of NEHI?  
Infants with NEHI usually have persistent rapid breathing (tachypnea), low oxygen levels (hypoxemia), chest retractions, crackling sounds in the lungs, and sometimes poor weight gain.

4. How is NEHI diagnosed?  
Diagnosis is mainly based on clinical symptoms and a high-resolution CT scan showing characteristic ground-glass opacities in the right middle lobe and lingula. Lung biopsy is rarely needed but can confirm diagnosis by showing increased neuroendocrine cells.

5. Is NEHI curable?  
There is no cure, but the condition often improves over time. Many children gradually get better with supportive care.

6. What treatments are available for NEHI?  
Treatment is supportive and includes supplemental oxygen to maintain adequate oxygen levels, nutritional support, and monitoring. There are no specific medications to cure NEHI.

7. How long does NEHI last?  
Symptoms often improve over months to years, but some children may have persistent mild symptoms or oxygen needs for a longer time.

8. Can NEHI be prevented?  
Since the cause is unknown, there are no known prevention strategies.

9. Will NEHI affect my child’s growth and development?  
Most children with NEHI grow and develop normally with proper medical care, although close monitoring is important.

10. Where can I get support and more information?  
Support groups, specialist pediatric pulmonologists, and organizations like the chILD Foundation provide resources and guidance for families affected by NEHI.

## **Neuroendocrine Cell Hyperplasia of Infancy (NEHI) — Treatment, Drugs, and Side Effects**

## 1. Main Treatments for NEHI

* Oxygen Therapy:
  + Most infants with NEHI require supplemental oxygen to maintain adequate oxygen levels, especially during sleep or illness.
  + Oxygen may be needed continuously or intermittently and is gradually weaned as the child improves.
  + Side effects are generally minimal but may include nasal irritation or dryness.
* Nutritional Support:
  + Infants with NEHI often have increased energy needs due to labored breathing and may experience poor weight gain.
  + High-calorie feeds, nutritional supplements, or feeding tubes (e.g., nasogastric tube) may be necessary.
  + Side effects depend on feeding method but can include discomfort or risk of aspiration with tube feeding.

## 2. Medications Used (Off-Label / Empirical)

* Corticosteroids:
  + Sometimes used to reduce airway inflammation, but evidence suggests they are less effective in NEHI than in other interstitial lung diseases.
  + Side effects include immunosuppression, growth retardation, weight gain, hypertension, and mood changes.
  + Use is individualized and discussed case-by-case.
* Azithromycin:
  + An antibiotic with anti-inflammatory properties occasionally used to help reduce airway inflammation.
  + Side effects may include gastrointestinal upset and potential antibiotic resistance.
* Hydroxychloroquine:
  + An antimalarial drug with anti-inflammatory effects used in some interstitial lung diseases.
  + Side effects include retinal toxicity (rare), gastrointestinal symptoms, and skin reactions.

## 3. Other Management Considerations

* Vaccinations:
  + Ensuring all routine childhood vaccines, including influenza, to reduce risk of respiratory infections which can worsen NEHI.
* Monitoring and Follow-Up:
  + Regular clinical and imaging follow-up to monitor lung function and oxygen needs.
* Supportive Care:
  + Avoidance of respiratory irritants and infections.
  + Physical therapy and developmental support as needed.

## 4. Prognosis and Treatment Outcomes

* Most children with NEHI improve over months to years, often weaning off oxygen and achieving normal growth and development.
* No specific curative drug treatment exists; supportive care remains the cornerstone.
* Clinical improvement often occurs regardless of initial treatment intensity.

**Doctor-patient conversation about Neuroendocrine Cell Hyperplasia of Infancy (NEHI)**

Doctor: "I want to talk with you about your baby's breathing difficulties. The diagnosis is called Neuroendocrine Cell Hyperplasia of Infancy, or NEHI for short. It’s a rare lung condition that affects infants, causing fast and heavy breathing."

Parent: "What exactly is NEHI? How does it affect my baby?"

Doctor: "In NEHI, there are extra neuroendocrine cells in the small airways of the lungs. These cells are normally present but in NEHI they are increased, which can cause inflammation and narrowing of the airways. This makes it harder for your baby to get enough oxygen and can cause symptoms like rapid breathing, low oxygen levels, and sometimes trouble gaining weight."

Parent: "What causes NEHI? Is it something I did or something contagious?"

Doctor: "We don’t know the exact cause yet. It’s not contagious and not caused by anything you did. Researchers think it might be related to both genetics and environmental factors, but more studies are needed to understand it fully."

Parent: "How do you know my baby has NEHI? What tests did you do?"

Doctor: "We rely mainly on your baby’s symptoms and a special type of lung scan called a high-resolution CT. This scan shows a typical pattern of changes in certain parts of the lungs, especially the right middle lobe and the lingula. Sometimes, if needed, a lung biopsy can confirm the diagnosis, but usually the clinical picture and imaging are enough."

Parent: "Is there a cure? What treatments are available?"

Doctor: "There’s no cure at the moment, but the good news is most babies with NEHI improve over time. Treatment focuses on supporting your baby's breathing and nutrition. Many babies need supplemental oxygen to keep their oxygen levels normal, and some may need extra help with feeding to ensure they grow well."

Parent: "How long will my baby need oxygen? Will this get better?"

Doctor: "Most children gradually get stronger as they grow, and many can stop needing oxygen after some time. Older children might only need oxygen during sleep or when they have infections. We will monitor your baby closely and adjust care as needed."

Parent: "What should I watch for? When should I call you?"

Doctor: "If your baby is working very hard to breathe, has increased breathing difficulty, or if you notice any changes like poor feeding or unusual tiredness, please contact us right away. Also, protecting your baby from infections is very important, so make sure vaccinations are up to date and avoid exposure to sick people."

Parent: "Is there support available for families like ours?"

Doctor: "Yes, there are support groups and resources through organizations like the chILD Foundation. We can connect you with specialists and other families who understand what you’re going through."

REFERENCES

[Neuroendocrine Cell Hyperplasia of Infancy - Symptoms, Causes, Treatment | NORD](https://rarediseases.org/rare-diseases/neuroendocrine-cell-hyperplasia-of-infancy/#causes)

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**Respiratory syncytial virus (RSV)**

## Alternative Names for Respiratory Syncytial Virus (RSV)

* RSV (common abbreviation)
* Human Respiratory Syncytial Virus
* Human Orthopneumovirus (official taxonomic name by ICTV)
* Pneumovirus (historical classification)
* Respiratory Syncytial Pneumovirus (less common)

**DEFINITION AND DESCRIPTION**

RSV is a respiratory illness that can cause cold-like symptoms or, sometimes, serious illness. RSV stands for respiratory syncytial (sin-SISH-uhl) virus, the germ that makes you sick with RSV. Almost everyone gets it for the first time before the age of 2. But the protection (immunity) you get from being infected doesn’t last, so you can get it more than once.

RSV is more common than you might think. We most often hear about babies getting severely ill from RSV, but adults and kids of all ages get it. Infants are more likely to get very sick from RSV because their immune systems are still developing. Their smaller airways are also more likely to get inflamed.

You can also get RSV many times throughout your life — sometimes even twice in one year. For adults, it usually causes mild, cold-like symptoms. But adults over 65 and people with compromised immune systems are at a higher risk for serious illness.

### **RSV causes**

A virus — respiratory syncytial virus — causes RSV. It’s not caused by bacteria. RSV spreads through:

* Close contact with someone who has it
* Coughing and sneezing (respiratory droplets)
* Contaminated objects or surfaces (think toys, countertops and phones)

#### **Is RSV contagious?**

Yes, RSV is contagious while you have symptoms — usually three to eight days. RSV spreads easily from person to person. It can live on hard surfaces, like tables, for several hours.

You may be able to spread RSV a day or two before you develop symptoms. Babies and people with compromised immune systems may be contagious even after symptoms go away, for up to four weeks.

## **Risk factors**

By age 2, most children will have been infected with respiratory syncytial virus, but they can get infected by RSV more than once. Children who attend child care centers or who have siblings who attend school are at a higher risk of exposure and reinfection. RSV season — when outbreaks tend to occur — is the fall to the end of spring.

People at increased risk of severe or sometimes life-threatening RSV infections include:

* Infants, especially premature infants or babies who are 6 months or younger
* Children who have heart disease that's present from birth (congenital heart disease) or chronic lung disease
* Children or adults with weakened immune systems from diseases such as cancer or treatment such as chemotherapy
* Children who have neuromuscular disorders, such as muscular dystrophy
* Adults with heart disease or lung disease
* Older adults, especially those age 65 and older

## **Symptoms**

Signs and symptoms of respiratory syncytial virus (RSV) infection most commonly appear about four to six days after exposure to the virus. In adults and older children, RSV usually causes mild cold-like signs and symptoms. These may include:

* Congested or runny nose
* Dry cough
* Low-grade fever
* Sore throat
* Sneezing
* Headache

### **In severe cases**

RSV infection can spread to the lower respiratory tract, causing pneumonia or bronchiolitis — inflammation of the small airway passages entering the lungs. Signs and symptoms may include:

* Fever
* Severe cough
* Wheezing — a high-pitched noise that's usually heard on breathing out (exhaling)
* Rapid breathing or difficulty breathing — the person may prefer to sit up rather than lie down
* Bluish color of the skin due to lack of oxygen (cyanosis)

Infants are most severely affected by RSV. Signs and symptoms of severe RSV infection in infants include:

* Short, shallow and rapid breathing
* Struggling to breathe — chest muscles and skin pull inward with each breath
* Cough
* Poor feeding
* Unusual tiredness (lethargy)
* Irritability

Most children and adults recover in one to two weeks, although some might have repeated wheezing. Severe or life-threatening infection requiring a hospital stay may occur in premature infants or in anyone who has chronic heart or lung problems.

### **RSV and COVID-19**

Because RSV and coronavirus disease 2019 (COVID-19) are both types of respiratory viruses, some symptoms of RSV and COVID-19 can be similar. In children, COVID-19 often results in mild symptoms such as fever, runny nose and cough. For adults with COVID-19, symptoms may be more severe and may include trouble breathing.

Having RSV may lower immunity and increase the risk of getting COVID-19 — for kids and adults. And these infections may occur together, which can worsen the severity of COVID-19 illness.

If you have symptoms of a respiratory illness, your doctor may recommend testing for COVID-19 .

### **When to see a doctor**

Seek immediate medical attention if your child — or anyone at risk of severe RSV infection — has difficulty breathing, a high fever, or a blue color to the skin, particularly on the lips and in the nail beds.

## **Diagnosis**

Your doctor may suspect respiratory syncytial virus based on the findings of a physical exam and the time of year the symptoms occur. During the exam, the doctor will listen to the lungs with a stethoscope to check for wheezing or other abnormal sounds.

Laboratory and imaging tests aren't usually needed. However, they can help diagnose RSV complications or rule out other conditions that may cause similar symptoms. Tests may include:

* Blood tests to check white cell counts or to look for viruses, bacteria and other germs
* Chest X-rays to check for lung inflammation
* Swab of secretions from inside the mouth or nose to check for signs of the virus
* Pulse oximetry, a painless skin monitor, to detect lower than normal levels of oxygen in the blood

## **Treatment**

Treatment for respiratory syncytial virus generally involves self-care measures to make your child more comfortable (supportive care). But hospital care may be needed if severe symptoms occur.

### **Supportive care**

Your doctor may recommend an over-the-counter medication such as acetaminophen (Tylenol, others) to reduce fever. (Never give aspirin to a child.) Use of nasal saline drops and suctioning may help clear a stuffy nose. Your doctor may prescribe antibiotics if there's a bacterial complication, such as bacterial pneumonia.

Keep your child as comfortable as possible. Offer plenty of fluids and watch for signs of loss of body fluids (dehydration), such as dry mouth, little to no urine output, sunken eyes, and extreme fussiness or sleepiness.

### **Hospital care**

If the respiratory syncytial virus (RSV) infection is severe, a hospital stay may be necessary. Treatments at the hospital may include:

* Intravenous (IV) fluids
* Humidified oxygen
* A breathing machine (mechanical ventilation), in rare cases

An inhaler (bronchodilator) or steroids are not proved to be helpful in treating RSV infection.

## **Self-care**

You may not be able to shorten the length of a respiratory syncytial virus (RSV) infection, but you can try to relieve some signs and symptoms.

If your child has RSV, do your best to comfort or distract him or her — cuddle, read a book or play a quiet game. Other tips for relieving symptoms are:

* **Create moist air to breathe.** Keep the room warm but not overheated. If the air is dry, a cool-mist humidifier or vaporizer can moisten the air and help ease congestion and coughing. Be sure to keep the humidifier clean to prevent the growth of bacteria and molds.
* **Drink fluids.** Continue breastfeeding or bottle-feeding your infant as you would normally. For older children and adults, keep a steady supply of cool water at the bedside. Offer warm fluids, such as soup, which may help loosen thickened secretions. Ice pops may be soothing as well.
* **Try saline nasal drops.** Over-the-counter (OTC) drops are a safe, effective way to ease congestion, even for young children. Follow your doctor's recommendations and the instructions on the product.
* **Use over-the-counter pain relievers.** OTC pain relievers such as acetaminophen (Tylenol, others) may help reduce fever and relieve a sore throat. Ask a doctor for the correct dose for your child's age.
* **Stay away from cigarette smoke.** Secondhand smoke can aggravate symptoms.

## **Complications**

Complications of respiratory syncytial virus include:

* **Hospitalization.** A severe respiratory syncytial virus (RSV) infection may require a hospital stay so that doctors can monitor and treat breathing problems and give intravenous (IV) fluids.
* **Pneumonia.** RSV is the most common cause of inflammation of the lungs (pneumonia) or the lungs' airways (bronchiolitis) in infants. These complications can occur when the virus spreads to the lower respiratory tract. Lung inflammation can be quite serious in infants, young children, older adults, immunocompromised individuals, or people with chronic heart or lung disease.
* **Middle ear infection.** If germs enter the space behind the eardrum, you can get a middle ear infection (otitis media). This happens most frequently in babies and young children.
* **Asthma.** There may be a link between severe RSV in children and the chance of developing asthma later in life.
* **Repeated infections.** Once you've had RSV, you could get infected again. It's even possible for it to happen during the same RSV season. However, symptoms usually aren't as severe — typically it's in the form of a common cold. But they can be serious in older adults or in people with chronic heart or lung disease.

## **Prevention**

Respiratory syncytial virus can infect anyone. But premature babies and young infants, as well as older adults, with heart or lung disease or a weakened immune system are at higher risk of severe infection.

### **Protection for babies and high-risk young children**

Two main options exist to help prevent young infants from getting severe Respiratory syncytial virus (RSV). One is an antibody product given to the infant. The other is an RSV vaccine for pregnant people to help protect their baby from birth through 6 months of age. Both are approved by the U.S. Food and Drug Administration (FDA). You and your healthcare professional can discuss which option is best to protect your child.

* **Antibody product called nirsevimab (Beyfortus).** This antibody product is a single dose shot given in the month before or during RSV season. It's for babies younger than 8 months born during or entering their first RSV season. Nirsevimab also can be given to children 8 months through 19 months old who are at higher risk of severe RSV disease through their second RSV season. In the U.S., the RSV season typically is November through March, but it varies in Florida, Alaska, Hawaii, Puerto Rico, Guam and other U.S. Pacific Island territories.

In rare situations, when nirsevimab is not available or a child is not eligible for it, another antibody product called palivizumab may be given. But palivizumab requires monthly shots given during the RSV season, while nirsevimab is only one shot. Palivizumab is not recommended for healthy children or adults.

* **Vaccine for pregnant people.** The FDA approved an RSV vaccine called Abrysvo for pregnant people to prevent RSV in infants from birth through 6 months of age. A single dose shot of Abrysvo can be given sometime from 32 weeks through 36 weeks of pregnancy during September through January in the U.S.

### **Vaccine for older adults**

Older adults have weaker immune systems, especially those with ongoing conditions, such as heart or lung disease. To help prevent RSV infection, the FDA approved RSV vaccines for adults age 60 and older.

The CDC recommends that adults age 60 and older talk with their healthcare professional about getting an RSV vaccine, especially if they're at higher risk of getting severe RSV. Two vaccines are available for this age group: Abrysvo and Arexvy. The CDC does not recommend one over the other. Each is a single dose shot.

Talk with your healthcare team about the benefits and risks of RSV vaccines for your situation.

### **Lifestyle habits**

These lifestyle habits can help prevent the spread of this infection:

* **Wash your hands often.** Teach your children the importance of handwashing.
* **Avoid exposure.** Cover your mouth and nose when you cough or sneeze. Limit your baby's contact with people who have fevers or colds.
* **Keep things clean.** Make sure kitchen and bathroom countertops, doorknobs, and handles are clean. Put used tissues in the trash right away.
* **Don't share drinking glasses with others.** Use your own glass or disposable cups when you or someone else is sick. Label each person's cup.
* **Don't smoke.** Babies who are exposed to tobacco smoke have a higher risk of getting RSV and potentially more-severe symptoms. If you do smoke, never do so inside the house or car.
* **Wash toys regularly.** Do this especially when your child or a playmate is sick.

## **Outlook / Prognosis**

RSV can last a week or two. You might have a lingering cough for a while. Severe cases of RSV may last longer.

### **Is there anything I can do to feel better?**

If you have mild symptoms, you can take care of yourself at home with:

* A cool-mist humidifier to help with breathing
* Nasal saline spray to help relieve cough and congestion
* Suctioning your child’s nose to remove mucus
* Plenty of fluids to avoid dehydration
* Over the counter (OTC) medications (like acetaminophen or ibuprofen)

Always check with your provider or your child’s pediatrician before using any medications or giving them to kids.

## **Diagnostic Considerations**

High-risk groups for severe respiratory syncytial virus (RSV) infection include the following:

* Premature infants in their first year of life (the younger the child is [in gestational and chronologic age] at the start of RSV season, the greater the risk)
* Infants with chronic lung disease (eg, bronchopulmonary dysplasia or cystic fibrosis) during their first 2 years of life
* Children with hemodynamically significant congenital heart disease, especially with increased pulmonary blood flow
* Patients with immunodeficient states
* Children with metabolic disorders, structural airway abnormalities, and neuromuscular disorders
* Children of multiple births (triplets or greater)

## **Differential Diagnoses**

* Adenovirus
* Asthma
* Bronchiolitis
* Croup
* Human Metapneumovirus
* Influenza
* Neonatal Sepsis
* Other respiratory viruses

Upper respiratory tract infection, croup, and bronchiolitis caused by RSV is clinically indistinguishable from other respiratory infections

* Human Parainfluenza Viruses (HPIV) and Other Parainfluenza Viruses
* Pediatric Bronchitis
* Pediatric Pneumonia

## **Epidemiology**

RSV LRT infection develops annually in 4-5 million children, and more than 125,000 children are admitted per year for RSV-related illness. The burden of RSV infection is not limited to only young children. In United States, it is responsible for 177,000 hospitalizations and 14,000 deaths in elderly ≥ 65 years of age.

Seasonal variations in incidence are observed (see the image below). Reinfection occurs throughout life, with the disease generally limited to the upper respiratory tract in persons older than 3 years. RSV infection is primarily seen in the winter months throughout United States except in the state of Florida where it extends throughout much of the year. Nationally, the onset of RSV season ranges from mid-September to mid-November, peaks from mid-December to mid-February, and the off-season occurs mid-April to mid-May. In tropical climates peak RSV activity correlates with the rainy season.

The COVID-19 pandemic disrupted the typical patterns of circulation for RSV. Starting in the southern United States, RSV circulation increased during the spring of 2021 and peaked in the summer.

Severe RSV disease has been reported in older children and adults with SCID (e.g., bone marrow transplantation), and RSV disease of the lower respiratory tract has been reported in elderly persons. RSV infection can also be severe in adults with COPD, those with immunodeficiency, and those ≥ 65 years of age.

Worldwide, RSV infection is prevalent, with clinical manifestations and early occurrence of RSV LRTI comparable to those seen in the United States.Each year RSV infection causes more than 100,000 deaths among children younger than 5 years throughout the world. Nearly half of those deaths occur in infants younger than 6 months old. In addition, RSV infection accounts for an estimated 3.6 million hospital admissions globally each year.

Severe RSV disease is primarily a disease of young infants and children, with a peak occurrence at the age of 2-8 months. Reinfection with RSV occurs throughout life, with disease becoming increasingly limited to the upper respiratory tract with advancing age. Although boys and girls are equally affected by milder RSV disease, males are approximately twice as likely to be hospitalized for RSV disease. All races appear to be susceptible to RSV, showing similar disease patterns.

## **Predefined Questions and Answers for Respiratory Syncytial Virus (RSV)**

1. What is RSV?  
RSV (Respiratory Syncytial Virus) is a common respiratory virus that causes cold-like symptoms in most people. It can cause mild upper respiratory illness but may lead to serious lung infections like bronchiolitis and pneumonia, especially in infants and older adults.

2. What are the symptoms of RSV?  
Symptoms usually start with a runny nose, decreased appetite, coughing, sneezing, fever, and wheezing. In very young infants, symptoms may be subtle, such as irritability, decreased activity, and breathing difficulties.

3. Who is at risk for severe RSV disease?  
Severe RSV is most common in infants under 6 months, premature babies, children with chronic lung or heart disease, immunodeficient individuals, and older adults with underlying health conditions.

4. How is RSV transmitted?  
RSV spreads through respiratory droplets when an infected person coughs or sneezes, and by direct contact with contaminated surfaces or people.

5. How long is the incubation period?  
Symptoms typically appear 4 to 7 days after exposure to the virus.

6. How is RSV diagnosed?  
Diagnosis is usually clinical but can be confirmed by laboratory tests such as PCR or antigen detection from nasal swabs.

7. Is there a treatment for RSV?  
There is no specific antiviral treatment for RSV. Care focuses on relieving symptoms, maintaining hydration, and oxygen therapy if needed. Fever and pain can be managed with acetaminophen or ibuprofen (never aspirin in children).

8. When should I seek medical care for RSV?  
Seek care if there is difficulty breathing, persistent high fever, dehydration, or if the infant is very young or has underlying health conditions.

9. Can RSV be prevented?  
Preventive measures include good hand hygiene, avoiding close contact with sick individuals, and cleaning surfaces. There are now vaccines and monoclonal antibodies available for high-risk infants and older adults to prevent severe RSV disease.

10. What is the prognosis for RSV infection?  
Most people recover within 1 to 2 weeks. However, severe cases can lead to hospitalization and, rarely, death, especially in high-risk groups. Early RSV infection may increase the risk of wheezing or asthma later in childhood

## **Genomic Data of Respiratory Syncytial Virus (RSV)**

* Genome Type:  
  RSV has a single-stranded, negative-sense RNA genome approximately 15,191 to 15,226 nucleotides in length, typically around 15.2 kb.
* Genome Organization:  
  The genome is non segmented and linear, containing 10 genes arranged in the order:  
  3′ NS1 - NS2 - N - P - M - SH - G - F - M2 - L 5′  
  These genes encode 11 proteins, including structural and nonstructural proteins.
* Proteins Encoded:
  + NS1 and NS2: Nonstructural proteins that inhibit host interferon responses.
  + N (nucleoprotein): Encapsidates the RNA genome.
  + P (phosphoprotein): Cofactor for RNA polymerase.
  + M (matrix protein): Involved in virus assembly.
  + SH (small hydrophobic protein): Forms ion channels; function not fully understood.
  + G (attachment glycoprotein): Mediates viral attachment; highly glycosylated and variable; modulates immune response.
  + F (fusion protein): Facilitates membrane fusion and viral entry; activates TLR-4 signaling.
  + M2: Encodes two proteins (M2-1 and M2-2) involved in transcription regulation.
  + L (large polymerase protein): RNA-dependent RNA polymerase.
* Replication:  
  RSV replication involves a complementary antigenome RNA that serves as a template for genome synthesis. Both genome and antigenome lack 5′ caps and 3′ polyA tails but are encapsidated by N protein to protect from degradation and immune detection.
* Genetic Variability:  
  RSV exists as two major subgroups, RSV A and RSV B, which differ in genetic sequences, especially in the G (attachment) and F (fusion) proteins.
  + RSV A generally has a higher mutation density and genetic diversity than RSV B.
  + Both subgroups show numerous synonymous and missense mutations, with the attachment glycoprotein gene exhibiting the highest variability.
  + These mutations contribute to viral evolution, epidemic potential, and challenges in vaccine development.
* Structural Insights:  
  RSV particles can be spherical, filamentous, or asymmetric, with a consistent organization of surface glycoproteins and internal proteins such as M and M2-1, which influence virus assembly and infectivity

**Doctor-patient conversation about Respiratory Syncytial Virus (RSV)**

Doctor: "Hello, I understand you’re concerned about your symptoms. Based on your exam and the season, it looks like you have an infection caused by Respiratory Syncytial Virus, or RSV."

Patient: "What exactly is RSV? Is it serious?"

Doctor: "RSV is a common virus that causes cold-like symptoms in most people. For healthy adults, it usually causes mild illness. However, in infants, older adults, or people with lung or heart conditions, it can cause more serious breathing problems like bronchiolitis or pneumonia."

Patient: "How did I get it? Is it contagious?"

Doctor: "Yes, RSV spreads easily through coughs, sneezes, or touching contaminated surfaces. It’s very common in the fall and winter months when respiratory viruses circulate more."

Patient: "Do I need antibiotics?"

Doctor: "No, RSV is caused by a virus, so antibiotics won’t help. Treatment focuses on relieving symptoms—rest, fluids, and if needed, medications for fever or pain. If your breathing becomes difficult or you feel worse, please come back."

Patient: "How long will it last?"

Doctor: "Most people recover in one to two weeks. Some symptoms like cough may linger longer, but you should gradually feel better."

Patient: "Is there a vaccine or prevention?"

Doctor: "There are vaccines and monoclonal antibodies available for high-risk groups like infants and older adults to prevent severe RSV. For most people, good hand hygiene and avoiding close contact with sick individuals help reduce risk."

Patient: "Thank you, doctor. I’ll take care and watch my symptoms."

Doctor: "That’s good. If you have any concerns or your symptoms worsen, don’t hesitate to contact us."

REFERENCES

[RSV Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/rsv-respiratory-syncytial-virus#what-is-rsv)

[Respiratory syncytial virus (RSV) - Symptoms & causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/respiratory-syncytial-virus/symptoms-causes/syc-20353098)

[Symptoms and Care of RSV | RSV | CDC](https://www.cdc.gov/rsv/symptoms/index.html)

<https://www.ncbi.nlm.nih.gov/books/NBK459215/#article-28424.s10>

## **Whooping Cough (Pertussis)**

## **Alternative Names for Whooping Cough (Pertussis)**

* Pertussis (medical term)
* Whooping Cough (common name)
* 100-day Cough (historical term, reflecting prolonged cough duration)
* Bordetella pertussis infection (refers to the causative bacterium)
* Paroxysmal Cough (describes the characteristic coughing fits)

**DEFINITION AND DESCRIPTION**

Whooping cough is an illness that can spread easily. It's also called pertussis. An infection with bacteria causes it. Many people with the illness get a serious hacking cough. Breathing in after coughing often causes a high-pitched noise that sounds like a "whoop."

Before the vaccine for pertussis came out, whooping cough was thought of as a childhood disease. Today, whooping cough mainly affects children too young to have gotten all their shots of the vaccine. The illness also tends to affect teenagers and adults whose protection from the vaccine has faded.

Deaths linked with whooping cough are rare. Most often, they occur in infants. But pregnant people can help protect their babies by getting a booster shot of the vaccine during pregnancy. Vaccination also is recommended for other people who will have close contact with an infant.

## **Causes**

A type of bacteria called Bordetella pertussis causes whooping cough. When an infected person coughs or sneezes, tiny germ-filled droplets spray into the air. Anyone who happens to be nearby can breathe in the droplets. The bacteria that cause whooping cough also can spread when people are together for a long time. Or these germs can spread when people share breathing space, such as while holding a newborn on your chest.

## **Risk factors**

The whooping cough vaccine you receive as a child wears off over time. This leaves many teenagers and adults prone to the infection during an outbreak. And regular outbreaks still happen.

Certain infants are most at risk for serious health problems and death from whooping cough. These include babies younger than 12 months old who are not vaccinated or haven't received all their vaccine doses.

## **Symptoms**

Once you become infected with whooping cough, it takes about 5 to 10 days for symptoms to start. Sometimes it takes up to three weeks. The symptoms often are mild at first. They may seem like those of a common cold. They can include:

* Runny or stuffy nose.
* Red, watery eyes.
* Fever.
* Cough.

After a week or two, the symptoms become worse. Thick mucus builds up inside the airways. This causes rapid coughing that can't be controlled. The cough can last for weeks or months, and it may be worse at night. Intense coughing attacks may cause:

* Vomiting.
* A red or blue face.
* Extreme tiredness.
* A high-pitched "whoop" sound during the next breath of air.

People with mild illnesses often don't make the whooping sound. Sometimes, an ongoing hacking cough is the only symptom of whooping cough in teens and adults.

Many babies with the illness don't cough at all. Some babies and young children might.

* Gag or struggle to breathe.
* Have skin, lips or nails that turn blue or purple.
* Have life-threatening pauses in breathing called apnea.

#### **Stages of whooping cough**

The three stages of whooping cough are:

* Common cold symptoms
* Coughing fits
* Gradual recovery

##### **Stage 1: Common cold symptoms**

This stage lasts about one to two weeks. You or your child is most contagious during this time, but you might think it’s just a cold. Symptoms include:

* Stuffy nose
* Runny nose
* Sore throat
* Watery eyes
* Mild or occasional cough that gradually gets worse
* Low-grade [fever](https://my.clevelandclinic.org/health/symptoms/10880-fever) (under 100.4 degrees Fahrenheit/38 degrees Celsius) or normal temperature
* Feeling generally unwell or “off” (malaise)

Infants may struggle to breathe or have pauses in their breathing (apnea). You might notice:

* Your baby is working hard to breathe.
* Their stomach seems to be caving in or breathing looks like they’re panting.
* Their skin or area around their mouth is turning blue or gray from lack of oxygen (cyanosis)

##### **Stage 2: Coughing fits**

The second stage usually lasts anywhere from one to six weeks. But it sometimes lasts as long as 10 weeks. During this stage, you have thick mucus in your airways and severe coughing fits (paroxysms). Here’s what to expect:

* A coughing fit means you cough many times in a row for several minutes.
* When you try to catch your breath between coughs, you may make a high-pitched “whoop” sound.
* You may vomit and/or feel exhausted after a coughing fit.
* Crying, eating or laughing may trigger a coughing fit.
* Coughing fits occur during the day and night but may get worse at night.

Coughing fits get more frequent before leveling off and then gradually going down in number.

##### **Stage 3: Gradual recovery**

This stage lasts up to six weeks. During this time, you may have a mild cough that comes and goes, but you won’t have severe coughing fits like before. You’ll gradually cough less and less.

Even as you start to feel better, you’re more vulnerable to other respiratory infections during this time. Your body is still healing. Try to avoid exposure to germs. If you do get sick with something else, the coughing fits may return.

#### **Whooping cough sound**

Whooping cough gets its name from the “whoop” sound that some people make after coughing. If you’re coughing over and over, it can be difficult to catch your breath. When you finally take in some air, your efforts may come across as a high-pitched “whoop” or gasp. But not everyone with pertussis develops a “whoop sound.”

## **When to see a doctor**

Call your healthcare professional if ongoing coughing spells cause you or your child to:

* Vomit.
* Turn red, purple or blue.
* Breathe in with a whooping sound.
* Not drink enough fluid.

Call 911 or your local emergency number right away if you or your child seems to struggle to breathe. Also call for emergency care if you notice pauses in breathing.

## **Diagnosis**

It may take time to find out if you have whooping cough. The symptoms can seem like those of other common illnesses, such as a common cold or the flu.

Sometimes, healthcare professionals can determine that you have whooping cough by asking about your symptoms and doing a physical exam.

But you may need medical tests such as:

* **A nose or throat culture and test.** Your healthcare professional takes a mucus sample from the back of the throat, where the nose and throat meet. The sample is checked for signs of whooping cough bacteria.
* **Blood tests.** A blood sample may be taken and sent to a lab. The lab checks for certain proteins called antibodies that the body makes to fight infections. This is a general test and not specific for whooping cough.
* **A chest X-ray.** This test may be recommended to check for signs of pneumonia due to whooping cough.

## **Treatment**

Most often, infants need treatment in the hospital for whooping cough. That's because the illness is more dangerous for babies. If your child can't keep down liquids or food, fluids given through a vein may be needed. Your child is cared for away from others. This prevents the infection from spreading.

Treatment for older children and adults often can be given at home, since the illness tends to be milder.

### **Medications**

Antibiotics kill the bacteria that causes whooping cough. When you take them early, they might make your illness less serious. They also may shorten the amount of time you're at risk of spreading the illness. If you live with other people, they may be given antibiotics to help prevent them from getting sick.

Not much is available to relieve the cough itself. Cough medicines that are sold without a prescription, for instance, do not help treat whooping cough. Do not take them unless your healthcare professional tells you to.

## **Self-care**

The following tips can help you deal with coughing spells while you recover from whooping cough at home:

* **Get plenty of rest.** A cool, quiet and dark bedroom may help you relax and rest better.
* **Drink plenty of fluids.** Water, juice and soups are good choices. Be aware of dehydration symptoms, especially if your child is sick. The symptoms include dry lips, crying without tears and urinating much less often.
* **Eat smaller meals.** To help prevent vomiting after coughing, eat smaller, more-frequent meals rather than large ones.
* **Clean the air.** Keep your home free of irritants that can trigger coughing spells. These include dust, tobacco smoke and fumes from fireplaces.
* **Prevent the spread of whooping cough.** Cover your cough or sneeze with a tissue or the inside of your elbow. Throw away used tissues right away. Wash your hands often and for at least 20 seconds. If you must be around others, wear a mask.

Stay home from work or school until you've completed at least five days of treatment. If a young child is sick, keep the child home from daycare until finishing five days of treatment. If you don't get treatment, it's safest to stay away from others for three weeks after your symptoms start. Your body gets rid of the bacteria by then, even though you may still have symptoms.

## **Whooping Cough (Pertussis) — Treatment Drugs and Their Side Effects**

## 1. Preferred Antibiotics for Treatment and Prophylaxis

* Macrolides:
  + Azithromycin
    - Dosing:
      * Infants >6 months & adults: 5-day course (500 mg day 1, then 250 mg daily days 2–5)
      * Children: weight-based dosing (10 mg/kg day 1, then 5 mg/kg days 2–5)
    - Side Effects: Gastrointestinal upset (nausea, diarrhea, abdominal pain), rare allergic reactions, possible QT prolongation.
  + Clarithromycin
    - Dosing:
      * Children >6 months: 15 mg/kg/day divided twice daily for 7 days
      * Adults: 500 mg twice daily for 7 days
    - Side Effects: GI upset, taste disturbances, liver enzyme elevation, drug interactions. Not recommended in pregnancy.
  + Erythromycin
    - Dosing:
      * Infants: 40–50 mg/kg/day divided into 4 doses for 14 days
      * Adults: 2 g/day divided into 4 doses for 14 days
    - Side Effects: GI upset (especially abdominal cramping and diarrhea), risk of infantile hypertrophic pyloric stenosis in neonates, liver toxicity, drug interactions. Estolate formulation preferred in children but avoided in adults and pregnant women.
* Trimethoprim-Sulfamethoxazole (TMP-SMX):
  + Alternative for those intolerants to macrolides.
  + Dosing:
    - Children >2 months: 8 mg TMP/40 mg SMX/kg/day divided twice daily for 14 days
    - Adults: 320 mg TMP/1600 mg SMX daily divided twice daily for 14 days
  + Side Effects: Allergic reactions, rash, photosensitivity, hematologic effects (rare), not recommended in pregnancy or infants <2 months.

## 2. Treatment Timing and Considerations

* Antibiotics are most effective when started within the first 1–2 weeks of cough onset to reduce severity and transmission.
* After 3 weeks, antibiotics do not change the course but may still be given to infants and pregnant women up to 6 weeks after onset.
* Early treatment reduces contagiousness after 5 days of antibiotics.

## 3. Supportive Care

* Rest, hydration, and avoiding cough irritants (smoke, dust).
* Cough medicines are generally ineffective and not recommended.
* Hospitalization may be required for severe cases, especially infants.

## **Complications**

Teens and adults often recover from whooping cough with no problems. When other health conditions happen, they tend to be side effects of intense coughing, such as:

* Bruised or cracked ribs.
* Tissue that bulges through a weak spot in the muscles of the stomach area, called an abdominal hernia.
* Broken blood vessels in the skin or the whites of the eyes.

Teens and adults also can have:

* Loss of bladder control.
* Weight loss.
* Fainting.

### **Infants**

Health conditions that can happen with whooping cough are more serious in infants, especially those under 6 months of age. They can include:

* The lung infection pneumonia.
* Ear infection.
* Slowed or stopped breathing.
* Dehydration or weight loss due to trouble feeding.
* Seizures.
* Brain damage.

Infants and toddlers have the highest risk of other health conditions due to whooping cough. So, they're more likely to need treatment for these in a hospital. Health conditions due to whooping cough can be life-threatening for infants younger than 6 months old.

## **Prevention**

The best way to prevent whooping cough is with the pertussis vaccine. It's often given together with vaccines against two other serious diseases: diphtheria and tetanus. Healthcare professionals recommend starting vaccination during infancy.

The vaccine is a series of five shots. You might hear it called DTaP. It's most often given to children at these ages:

* 2 months.
* 4 months.
* 6 months.
* 15 to 18 months.
* 4 to 6 years.

### **Vaccine side effects**

Most often, side effects of the vaccine are mild. They may include a fever, crankiness, headache, fatigue or soreness at the site of the shot.

### **Booster shots**

Booster shots strengthen the body's defenses against whooping cough. They help protect against whooping cough, diphtheria and tetanus. You might hear your healthcare professional call your booster shot Tdap. It's recommended for the following people.

* **Pre-teens.** Protection from the pertussis vaccine tends to fade by age 11. So, healthcare professionals recommend a shot of Tdap at 11 or 12 years old.
* **Pregnant women.** Health experts now recommend getting a shot of Tdap between 27 and 36 weeks of each pregnancy. This also may give some protection to an infant during the first few months of life.
* **Adults.** Sometimes, Tdap is given instead of a booster shot for tetanus and diphtheria. The tetanus and diphtheria booster shot is recommended for adults every 10 years. If you're due for that shot and you've never received Tdap before, your healthcare professional likely will give you Tdap instead. That's because Tdap protects against all three diseases. In general, adults who never received the Tdap booster shot can get one at any time. It's key to make sure you're up to date on your shots at least two weeks before you get close to a baby. The vaccine lowers your risk of spreading whooping cough to infants.

### **Preventive medications**

If you or your infant has been exposed to someone with whooping cough, talk with your healthcare professional. Medicines called antibiotics may be recommended to protect against infection if you:

* Are a healthcare professional.
* Are pregnant.
* Are younger than 12 months old.
* Have a health condition that could put you at risk of serious illness or complications. This includes conditions such as a weakened immune system or asthma.
* Live with someone who has whooping cough.
* Live with someone who is at high risk of serious illness or complications from a whooping cough infection. This includes babies and pregnant women.

## **Outlook / Prognosis**

Whooping cough can make you feel miserable for weeks or months. And it’s possible to develop severe complications.

The good news is that your body will likely build up some immunity against *Bordetella pertussis* bacteria. So, you won’t get whooping cough again for a while — anywhere from four to 20 years, according to some experts. But you can get whooping cough again in the future once immunity wanes.

Your child might need to stay in the hospital, especially if they’re very young.

#### **When is it safe to return to daycare, school or work?**

It depends. If you or your child is taking antibiotics, it’s generally safe to return after five full days of treatment. Otherwise, you’re still contagious for three weeks after the start of the coughing fits.

### **Is there anything I can do to feel better?**

Things you can do to feel a little better — and possibly ease coughing at night — include:

* **Take some honey**. This may help calm your cough. But it’s only safe for adults and kids over age 1. Never give honey to an infant (because of the risk of botulism).
* **Rest**. Getting lots of rest helps your body heal.
* **Keep up the fluids**. Drinking plenty of fluids can help thin the mucus that’s making you cough.
* **Use a cool-mist humidifier**. This helps break up mucus.
* **Eat small meals**. Eating just a small amount of food at a time may keep you from vomiting.
* **Avoid irritants**. Things like dust, smoke and chemical fumes (for example, from some cleaning supplies) might make you cough.

## **Diagnostic Considerations**

Illnesses that mimic clinical pertussis include the following:

* Adenoviral respiratory infection - Children present with fever, sore throat, and conjunctivitis
* Mycoplasmal pneumonia - Patients with mycoplasmal infections have more pronounced systemic symptoms, fever and headache may occur, and rales may be appreciated on chest auscultation
* Chlamydial pneumonia - young infants with chlamydial infections present with staccato cough, purulent conjunctival discharge, tachypnea, rales, and wheezing
* Respiratory syncytial virus infection - Patients present with predominantly lower respiratory tract signs (eg, wheezing, rales)

Other conditions to consider in the differential diagnosis of pertussis include the following:

* Common cold
* Influenza
* Cystic fibrosis
* Interstitial pneumonitis
* Bronchiolitis
* Croup (Laryngotracheobronchitis)
* Dehydration
* Febrile Seizures
* Fever
* Gastroenteritis
* Intussusception
* Tachycardia
* Aspiration pneumonia
* Bacterial pneumonia
* Viral pneumonia
* Tuberculosis

## **Differential Diagnoses**

* Afebrile Pneumonia Syndrome
* Asthma
* Bronchiolitis
* Chlamydia (Chlamydial Genitourinary Infections)
* Chronic Obstructive Pulmonary Disease (COPD) and Emphysema in Emergency Medicine
* Emergent Treatment of Gastroenteritis
* Encephalitis
* Pediatric Mycoplasma Infections
* Respiratory Syncytial Virus Infection
* Trachea Foreign Bodies

## **Epidemiology**

### Occurrence in the United States

Since the early 1980s, pertussis incidence has cyclically increased, with peaks occurring every 2-5 years.Most cases occur between June and September. Neither acquisition of the disease nor vaccination provides complete or lifelong immunity. Protection against typical disease wanes 3-5 years after vaccination and is not measurable after 12 years.

The rate of pertussis peaked in the 1930s, with 265,269 cases and 7518 deaths reported in the United States. This rate decreased to a low of 1010 cases in the United States, with 4 deaths, in 1976. Starting in the 1980s, however, the reported incidence of US pertussis cases dramatically increased across all age groups. Although the largest increase in pertussis cases has been among adolescents and adults, the annual reported incidence has been highest among infants younger than 1 year.

In 2010, according to the Centers for Disease Control and Prevention (CDC), the US pertussis rate reached 27,550 cases (the highest number since 1959), with 27 related deaths.

In 2011, according to preliminary statistics from the CDC, adolescents (ages 11-19 years) and adults together accounted for 47% of pertussis cases, whereas children aged 7-10 years accounted for 18% of cases.

According to the CDC, during the first half of 2012, most states had reported either increased pertussis activity or outbreaks of the disease. By July 5 of that year, 37 states had reported increases in pertussis cases over those reported during the same period in 2011.

For example, as listed by the CDC and the states’ health departments, the number of reported cases in Washington State (where a pertussis epidemic was declared), Minnesota, and Wisconsin in 2012 were as follows:

* Washington State - 2012: 3400 cases reported through Aug 4; 2011: 287 cases reported through Aug 4
* Minnesota - 2012: 2039 cases reported as of Aug 2; 2011 (entire year): 661 cases reported
* Wisconsin - 2012: 3496 confirmed or probable cases reported through July 31, 2012; 2011 (entire year): 1192 confirmed or probable cases reported

The CDC listed a provisional national figure of 17,000 pertussis cases between Jan 1 and July 21, 2012, including 9 pertussis-related deaths. The reasons that pertussis cases peak in some years is not completely understood, according to the CDC.

The CDC has estimated that 5-10% of all cases of pertussis are recognized and reported. Pertussis remains the most commonly reported vaccine-preventable disease in the United States in children younger than 5 years. In studies, 12-32% of adults with prolonged (1-4 wks.) cough have been found to have pertussis.

Between January 1, 2014, and June 10, 2014, California's public health department reported 3,458 cases of pertussis. The department declared the outbreak to have reached epidemic proportions, with 800 cases reported in the span of just 2 weeks.A study that examined a similar outbreak in California in 2010 determined that nonmedical vaccine exemptions played a role.

Nationally, the CDC stated that the 4,838 cases of pertussis reported from January 1, 2014, to April 14, 2014, represented a 24% increase over the same period in 2013.

The CDC reports that during January 1–November 26, 2014, a total of 9,935 cases of pertussis with onset in 2014 were reported in California. Severe and fatal disease occurs almost exclusively in infants who are too young to be vaccinated against pertussis. Therefore, pregnant women are encouraged to receive tetanus, diphtheria, and acellular pertussis vaccine (Tdap) during the third trimester of each pregnancy to provide placental transfer of maternal antibodies to the infant.

### International occurrence

The annual worldwide incidence of pertussis is estimated to be 48.5 million cases, with a mortality rate of nearly 295,000 deaths per year.The case-fatality rate among infants in low-income countries may be as high as 4%.

In England, the percentage of people vaccinated for pertussis over the last 4 decades has decreased to less than 30%. This decline has resulted in thousands of recently reported cases of the disease, with the incidence rate approaching that of the pre vaccination era. Similar epidemic outbreaks recently have occurred in Sweden, Canada, and Germany. Nearly 300,000 deaths from pertussis are thought to have occurred in Africa over the last decade.

### Race- and sex-related demographics

With regard to race, the CDC reported that among individuals with pertussis between 2001 and 2003, 90% were white, 7% were black, 1% were Asian/Pacific Islander, and 1% were American Indian/Alaska Native, and 1% were identified as “other race”. From 2001-2003, females accounted for 54% of pertussis cases in the United States.

### Age-related demographics

From 2001-2003 of patients with pertussis, 23% were younger than 1 year, 12% were aged 1-4 years, 9% were aged 5-9 years, 33% were aged 10-19 years, and 23% were older than 20 years.

Because of the lack of maternal immunity transfer, 10-15% of all cases of pertussis occur in infants younger than 6 months; more than 90% of all deaths occur in this same age group. However, the growing majority of cases now are in persons aged 10 years and older, which has led to increased booster recommendations.

## **Genomic Data of Whooping Cough (Pertussis) — *Bordetella pertussis***

* Genome Structure:  
  *Bordetella pertussis* has a circular chromosome approximately 4.1 million base pairs (bp) in length (e.g., 4,124,236 bp in strain CS) with a high G+C content of about 67.3%.
* Gene Content:  
  The genome encodes roughly 3,400 to 3,900 protein-coding sequences (CDSs), depending on the strain, with an average protein size of around 327 amino acids. It also contains multiple copies of insertion sequence IS481, a mobile genetic element presents in high numbers (over 200 copies), which plays a role in genome rearrangements and deletions.
* Genomic Variation and Evolution:  
  Despite vaccination, pertussis has resurged in many countries, partly due to pathogen adaptation and genome evolution. Comparative genomics of hundreds of isolates reveal:
  + Low nucleotide diversity: *B. pertussis* is considered a highly monomorphic pathogen with few single nucleotide polymorphisms (SNPs) between strains.
  + Structural rearrangements: Large-scale chromosomal rearrangements mediated by IS481 elements alter gene order and may affect gene expression, potentially contributing to vaccine escape and epidemiological changes.
  + Virulence gene variation: Mutations and deletions in key antigen genes such as pertactin (prn) and pertussis toxin promoter (ptxP) have been observed, with some circulating strains being pertactin-deficient, which may provide a fitness advantage

## **Whooping Cough (Pertussis) — Predefined Questions and Answers**

1. What is whooping cough?  
Whooping cough, also called pertussis, is a highly contagious respiratory infection caused by the bacterium *Bordetella pertussis*. It causes severe coughing fits that can make it hard to breathe.

2. How does whooping cough spread?  
It spreads through droplets when an infected person coughs or sneezes. Close contact with someone who has pertussis increases the risk of catching it.

3. What are the symptoms of whooping cough?  
Early symptoms resemble a common cold: runny nose, sneezing, mild cough, and low-grade fever. After 1–2 weeks, severe coughing fits develop, often followed by a “whooping” sound when inhaling.

4. Who is most at risk for severe disease?  
Infants under 1 year old, especially those not fully vaccinated, are at highest risk. Older adults and people with weakened immune systems can also have severe illness.

5. How is whooping cough diagnosed?  
Diagnosis is made based on symptoms and confirmed by laboratory tests such as PCR or culture from a nasal swab.

6. What treatments are available?  
Antibiotics such as azithromycin, clarithromycin, or erythromycin are used to treat pertussis and reduce transmission. Supportive care includes rest, fluids, and monitoring for breathing difficulties.

7. Can whooping cough be prevented?  
Yes. Vaccination is the best prevention. The DTaP vaccine is given to children, and Tdap boosters are recommended for adolescents and adults, especially pregnant women.

8. How long is someone contagious?  
People are contagious from the start of cold-like symptoms until about 5 days after starting appropriate antibiotics. Without treatment, they can be contagious for up to 3 weeks after coughing begins.

9. When should I seek medical care?  
Seek care if coughing spells cause difficulty breathing, vomiting, or if an infant shows signs of distress, poor feeding, or apnea (pauses in breathing).

10. What complications can occur?  
Complications include pneumonia, seizures, brain damage, and in severe cases, death—especially in infants.

**Doctor-patient conversation about Whooping Cough (Pertussis)**

Doctor: "Hello, I understand your child has been having a persistent cough. Based on the symptoms and examination, it looks like your child may have whooping cough, also called pertussis."

Parent: "What exactly is whooping cough? Why is it called that?"

Doctor: "Whooping cough is a contagious respiratory infection caused by bacteria called *Bordetella pertussis*. It gets its name from the characteristic 'whoop' sound children make when they gasp for air after a severe coughing fit. The cough can be very intense and last for weeks."

Parent: "How does it start and how long does it last?"

Doctor: "It usually begins like a common cold with a runny nose, sneezing, mild cough, and sometimes a low fever. After one to two weeks, the cough worsens and comes in violent fits. These coughing spells can cause vomiting, redness in the face, and sometimes a whooping sound. This stage can last from two to eight weeks, and the cough may persist for up to 100 days, which is why it's sometimes called the '100-day cough.'"

Parent: "Is it dangerous? What should I watch for?"

Doctor: "Infants and young children are at highest risk for complications like pneumonia, difficulty breathing, or even seizures. Watch for signs like difficulty breathing, turning blue around the lips, poor feeding, or extreme tiredness. If you see any of these, seek medical care immediately."

Parent: "How is it treated?"

Doctor: "We treat whooping cough with antibiotics, which help reduce the contagiousness and can be most effective if started early. Supportive care like rest, fluids, and monitoring breathing is important. Sometimes hospitalization is needed for severe cases."

Parent: "Can it be prevented?"

Doctor: "Yes, vaccination is the best prevention. Children receive the DTaP vaccine, and adults and pregnant women get booster shots called Tdap to protect themselves and their babies."

Parent: "How contagious is it? How can I protect others?"

Doctor: "Whooping cough spreads easily through coughing or sneezing. Your child is contagious from the start of symptoms until about five days after starting antibiotics. Good hand hygiene, avoiding close contact with others, and completing the full course of antibiotics help prevent spread."

Parent: "Thank you, doctor. What should I do if the cough gets worse?"

Doctor: "If your child's coughing spells become more severe, if they have trouble breathing, or if they stop feeding well, please come back immediately. We will monitor closely and adjust care as needed."

REFERENCES

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[Pertussis (Whooping Cough) | Whooping Cough | CDC](https://www.cdc.gov/pertussis/index.html)

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**TUBERCULOSIS**

**DEFINITION AND DESCRIPTION**

Tuberculosis, also called TB, is a serious illness that mainly affects the lungs. The germs that cause tuberculosis are a type of bacteria.

Tuberculosis can spread when a person with the illness coughs, sneezes or sings. This can put tiny droplets with the germs into the air. Another person can then breathe in the droplets, and the germs enter the lungs.

Tuberculosis spreads easily where people gather in crowds or where people live in crowded conditions. People with HIV/AIDS and other people with weakened immune systems have a higher risk of catching tuberculosis than people with typical immune systems.

Medicines called antibiotics can treat tuberculosis. But some forms of the bacteria no longer respond well to treatments.

## **Causes**

Tuberculosis is caused by a bacterium called Mycobacterium tuberculosis.

People with active TB disease in the lungs or voice box can spread the disease. They release tiny droplets that carry the bacteria through the air. This can happen when they're speaking, singing, laughing, coughing or sneezing. A person can get an infection after inhaling the droplets.

The disease is more likely to spread when people spend a lot of time together in an indoor space. So the disease spreads easily in places where people live or work together for long periods. Also, the disease spreads more easily in crowded gatherings.

A person with a latent TB infection cannot pass the disease to other people. A person taking medicine to treat active TB disease usually can't pass the disease after 2 to 3 weeks of treatment.

### **Drug-resistant TB**

Some forms of the TB bacteria have become drug resistant. This means that medicines that once cured the disease no longer work.

This happens, in part, because of naturally occurring genetic changes in bacteria. A random genetic change in a bacterium might give it some quality that makes it more likely to survive the attack of an antibiotic. If it does survive, then it can multiply.

When antibiotic medicines aren't used correctly — or medicines fail to clear out all the bacteria for another reason — the conditions are ideal for more-resistant versions of the bacteria to take hold and multiply. If these bacteria are passed on to other people, a new drug-resistant strain can grow over time.

Problems that can lead to such drug-resistant strains of bacteria include the following:

* People didn't follow directions for taking the medicines or stopped taking the medicines.
* They weren't prescribed the right treatment plan.
* Medicines were not available.
* The medicines were of poor quality.
* The body didn’t absorb the medicines as expected.

## **Risk factors**

Anyone can get tuberculosis, but certain factors increase the risk of getting an infection. Other factors increase the risk of an infection becoming active TB disease.

The Centers for Disease Control and Prevention recommends a TB test for people who have an increased risk of TB infection or active TB disease. Talk to your healthcare professional if you have one or more of the following risk factors.

### **Risk of TB infection**

Certain living or working conditions make it easier for the disease to pass from one person to another. These conditions increase the risk of getting a TB infection:

* Living with someone with active TB disease.
* Living or traveling in a country where TB is common, including several countries in Latin America, Africa, Asia and the Pacific Islands.
* Living or working in places where people live close together, such as prisons, nursing homes and shelters for homeless people.
* Living in a community identified as being at high risk of tuberculosis.
* Working in healthcare and treating people with a high risk of TB.

### **Risk of active TB disease**

A weakened immune system increases the risk of a TB infection becoming active TB disease. Conditions or treatments that weaken the immune system include:

* HIV/AIDS.
* Diabetes.
* Severe kidney disease.
* Cancers of the head, neck and blood.
* Malnutrition or low body weight.
* Cancer treatment, such as chemotherapy.
* Medicine to prevent rejection of transplanted organs.
* Long-term use of prescription steroids.
* Use of unlawful injected drugs.
* Misuse of alcohol.
* Smoking and using other tobacco products.

### **Age and active TB disease**

The risk of a TB infection becoming active TB disease changes with age.

* **Under 5 years of age.** Until children reach age 5, they have high risk of a TB infection becoming active TB disease. The risk is greater for children under age 2. Tuberculosis in this age group often leads to serious disease in the fluid surrounding the brain and spinal column, called meningitis.
* **Age 15 to 25.** People in this age group have an increased risk of developing more-severe active TB disease in the lungs.
* **Age 65 and older.** The immune system weakens during older age. Older adults have a greater risk of active TB disease. Also, the disease may be more difficult to treat.

## **Symptoms**

When TB germs survive and multiply in the lungs, it is called a TB infection. A TB infection may be in one of three stages. Symptoms are different in each stage.

**Primary TB infection.** The first stage is called the primary infection. Immune system cells find and capture the germs. The immune system may completely destroy the germs. But some captured germs may still survive and multiply.

Most people don't have symptoms during a primary infection. Some people may get flu-like symptoms, such as:

* Low fever.
* Tiredness.
* Cough.

**Latent TB infection.** Primary infection is usually followed by the stage called latent TB infection. Immune system cells build a wall around lung tissue with TB germs. The germs can't do any more harm if the immune system keeps them under control. But the germs survive. There are no symptoms during latent TB infection.

**Active TB disease.** Active TB disease happens when the immune system can't control an infection. Germs cause disease throughout the lungs or other parts of the body. Active TB disease may happen right after primary infection. But it usually happens after months or years of latent TB infection.

Symptoms of active TB disease in the lungs usually begin gradually and worsen over a few weeks. They may include:

* Cough.
* Coughing up blood or mucus.
* Chest pain.
* Pain with breathing or coughing.
* Fever.
* Chills.
* Night sweats.
* Weight loss.
* Not wanting to eat.
* Tiredness.
* Not feeling well in general.

**Active TB disease outside the lungs.** TB infection can spread from the lungs to other parts of the body. This is called extrapulmonary tuberculosis. Symptoms vary depending on what part of the body is infected. Common symptoms may include:

* Fever.
* Chills.
* Night sweats.
* Weight loss.
* Not wanting to eat.
* Tiredness.
* Not feeling well in general.
* Pain near the site of infection.

Active TB disease in the voice box is outside the lungs, but it has symptoms more like disease in the lungs.

Common sites of active TB disease outside the lungs include:

* Kidneys.
* Liver.
* Fluid surrounding the brain and spinal cord.
* Heart muscles.
* Genitals.
* Lymph nodes.
* Bones and joints.
* Skin.
* Walls of blood vessels.
* Voice box, also called larynx.

**Active TB disease in children.** Symptoms of active TB disease in children vary. Typically, symptoms by age may include the following:

* **Teenagers.** Symptoms are similar to adult symptoms.
* **1- to 12-year-olds.** Younger children may have a fever that won't go away and weight loss.
* **Infants.**The baby doesn't grow or gain weight as expected. Also, a baby may have symptoms from swelling in the fluid around the brain or spinal cord, including:
  + Being sluggish or not active.
  + Unusually fussy.
  + Vomiting.
  + Poor feeding.
  + Bulging soft spot on the head.
  + Poor reflexes.

## **When to see a doctor**

The symptoms of tuberculosis are similar to symptoms of many different illnesses. See your healthcare professional if you have symptoms that don't improve with a few days of rest.

Get emergency care if you have:

* Chest pain.
* Sudden, severe headache.
* Confusion.
* Seizures.
* Difficulty breathing.

Get immediate or urgent care if you:

* Cough up blood.
* Have blood in your urine or stool.

## **Diagnosis**

To diagnosis a tuberculosis (TB) infection, your healthcare professional will do an exam that includes:

* Listening to you breathe with a stethoscope.
* Checking for swollen lymph nodes.
* Asking you questions about your symptoms.

### **TB tests**

Your healthcare professional will order tests if:

* Tuberculosis is suspected.
* You were likely exposed to a person with active TB disease.
* You have health risks for active TB disease.

Your healthcare team will determine whether a skin test or blood test is the best option.

### **Skin test**

A tiny amount of a substance called tuberculin is injected just below the skin on the inside of one forearm. Within 48 to 72 hours, a healthcare worker will check your arm for swelling at the injection site. The size of the raised skin is used to determine a positive or negative test.

This test is seeing if your immune system reacts, or has made an antibody, to tuberculosis. A positive test indicates you likely have either a latent TB infection or active TB disease. People who had a TB vaccination might get a positive test even if they have no infection.

A negative test means that your body didn't react to the test. It doesn't necessarily mean you don't have an infection.

### **Blood tests**

A sample of blood is sent to a lab. One lab test finds out whether certain immune system cells can "recognize" tuberculosis. A positive test shows that you have either a latent TB infection or active TB disease. Other tests of the blood sample can help determine if you have active disease.

A negative result means you likely do not have a TB infection.

### **X-ray**

A chest X-ray can show irregular patches in the lungs that are typical of active TB disease.

### **Sputum tests**

Your healthcare professional may take a sample of the mucus that comes up when you cough, also called sputum. If you have active TB disease in your lungs or voice box, lab tests can detect the bacteria.

A relatively quick laboratory test can tell if the sputum likely has the TB bacteria. But it may be showing bacteria with similar features.

Another lab test can confirm the presence of TB bacteria. The results often take several weeks. A lab test also can tell if it's a drug-resistant form of the bacteria. This information helps your healthcare professional choose the best treatment.

### **Other lab tests**

Other lab tests that may be ordered include:

* Breath test.
* Procedure to remove sputum from your lungs with a special tube.
* Urine test.
* Test of the fluid around the spine and brain, called cerebrospinal fluid.

## **Treatment**

If you have a latent TB infection, your healthcare professional may begin treatments. This is especially true for people with HIV/AIDS or other factors that increase the risk of active TB disease. Most latent TB infections are treated for three or four months.

Active TB disease may be treated for four, six or nine months. Specialists in TB treatment will determine which medicines are best for you.

You will have regular appointments to see if you're improving and to watch for side effects.

### **Take all of the medicines**

It is important to take every dose as instructed. And you must complete the full course of treatment. This is important for killing the bacteria in your body and preventing new drug-resistant bacteria.

Your public health department may use a program called directly observed therapy (DOT). With DOT, a healthcare worker visits you at home to watch you take your dose of medicines.

Some healthcare departments have programs that let you take your medicines on your own. The Centers for Disease Control and Prevention has printable forms you can use to keep track of your daily doses.

### **Most common TB medicines**

If you have a latent TB infection, you might need to take only one or two types of medicines. Active TB disease requires taking several medicines. Common ones used to treat tuberculosis include:

* Isoniazid.
* Rifampin (Rimactane).
* Rifabutin (Mycobutin).
* Rifapentine (Priftin).
* Pyrazinamide.
* Ethambutol (Myambutol).

You may be prescribed other medicines if you have drug-resistant tuberculosis or other complications from your illness.

### **Medication side effects**

Most people can take TB medicines without serious side effects. If you have serious side effects, your healthcare professional may ask you to stop taking a medicine. You may have to change the dose of a medicine.

Talk to your healthcare professional if you experience any of the following:

* Upset stomach.
* Vomiting.
* Loss of appetite.
* Severe diarrhea.
* Light-colored stool.
* Dark urine.
* Yellowish skin or eye color.
* Changes in vision.
* Dizziness or trouble with balance.
* Tingling in hands or feet.
* Easy bruising or bleeding.
* Unexplained weight loss.
* Unexplained tiredness.
* Sadness or depression.
* Rash.
* Joint pain.

It is important for you to list all medicines, dietary supplements or herbal remedies you take. You may need to stop taking some of these during your treatment.

## **Prevention**

If you test positive for latent TB infection, you may need to take medicines to prevent active TB disease.

### **Preventing the spread of disease**

If you have active TB disease, you'll need to take steps to prevent other people from getting an infection. You will take medicines for four, six or nine months. Take all of the medicines as directed during the entire time.

During the first 2 to 3 weeks, you will be able to pass TB bacteria to others. Protect others with these steps:

* **Stay home.** Don't go to work or school.
* **Isolate at home.** Spend as little time as possible among members of your household. Sleep in a separate room.
* **Ventilate the room.** Tuberculosis germs spread more easily in small, closed spaces. If it's not too cold outdoors, open the windows. Use a fan to blow air out. If you have more than one window, use one fan to blow air out and another to blow air in.
* **Wear face masks.** Wear a mask when you have to be around other people. Ask other members of the household to wear masks to protect themselves.
* **Cover your mouth.** Use a tissue to cover your mouth anytime you sneeze or cough. Put the dirty tissue in a bag, seal it and throw it away.

### **Vaccinations**

In countries where tuberculosis is common, infants often are vaccinated with the bacille Calmette-Guerin (BCG) vaccine. This protects infants and toddlers who are more likely to have active TB disease in the fluid surrounding the brain and spinal cord.

The vaccine may not protect against disease in the lungs, which is more likely in the United States. Dozens of new TB vaccines are in various stages of development and testing.

**PROGNOSIS**

The cure rate for patients with drug-susceptible TB who can complete a therapeutic regimen can exceed 95%. Variables such as disease extent, delay in treatment, comorbidities, age, the need for mechanical ventilation, drug resistance, and adverse drug reactions influence the therapy outcome. Novel regimens will likely improve outcomes in people treated for drug-resistant TB.

The WHO 2018 global estimates of successful treatment outcomes are as follows:

* 85% for people with new and relapsed TB
* 76% of HIV-coinfected people
* 57% for people with MDR-TB

More than 80% of TB-associated mortality occurs in LMICs. In 2022, the WHO estimated 1.13 million deaths among HIV-negative people and 167,000 deaths among PLHIV. TB is the leading cause of death in PLHIV. The WHO estimates that 15% of patients with MDR-TB die of disease, and 26% of those deaths are due to XDR-TB. The current estimate of the prognosis of untreated TB is difficult to calculate; it would have to account for regional differences in healthcare resources, those who have either failed or never initiated treatment, those with drug-resistant strains, and people with different underlying comorbidities. Estimates based on pre-chemotherapy era data may be unreliable due to case definition, patient selection, and reporting heterogeneity. The study by Tiemersma, et al estimated a 70% lifetime case fatality among untreated HIV-negative individuals

## **Diagnostic Considerations**

Tuberculosis (TB) is well known for its ability to masquerade as other infectious and disease processes. For example, congenital TB can mimic congenital syphilis or cytomegalovirus (CMV) infection. Along with the differentials listed in the next section, conditions with a presentation that may resemble pulmonary TB include the following:

* Blastomycosis
* Tularemia
* Actinomycosis
* *Mycobacterium avium-intracellulare* infection
* *M chelonae* infection
* *M fortuitum* infection
* *M gordonae* infection
* *M kansasii* infection
* *M marinum* infection
* *M xenopi* infection
* Squamous cell carcinoma

Conditions to be included in the differential diagnosis of extrapulmonary TB include the following:

* Blastomycosis
* Tularemia
* Actinomycosis
* Hidradenitis suppurativa
* Eosinophilic granuloma
* *M avium-intracellulare* infection
* *M chelonae* infection
* *M fortuitum* infection
* *M gordonae* infection
* *M kansasii* infection
* *M marinum* infection
* *M xenopi* infection
* Endemic syphilis
* Erythema induratum (nodular vasculitis)
* Erythema nodosum
* Leishmaniasis
* Leprosy
* Cat scratch disease
* Syphilis
* Syringoma
* Rheumatoid arthritis

Dermatologic differential diagnosis:

Diagnosis of skin infection with *M tuberculosis* involves the following:

* Differentiate primary-inoculation TB from ulceroglandular complexes and mycobacteriosis
* Differentiate TB verrucosa cutis from diseases such as North American blastomycosis, chromoblastomycosis, iododerma and bromoderma, chronic vegetative pyoderma, verruca vulgaris, verrucous carcinoma, verrucous atypical mycobacterial infection, and verrucous lupus vulgaris
* Differentiate miliary TB of the skin (which appears as small, non characteristic, erythematous, papular, or purpuric lesions) from drug reactions.
* Differentiate scrofuloderma from suppurative lymphadenitis with sinus-tract formation, such as blastomycosis or coccidioidomycosis.
* Differentiate TB cutis orificialis from glossitis, apotheosis, and deep fungal infections.
* Differentiate lupus vulgaris from lupoid rosacea, deep fungal or atypical mycobacterial infection, chronic granulomatous disease, granulomatous rosacea, and Wegener granulomatosis.
* Differentiate erythema induratum from nodular panniculitides (eg, Weber-Christian disease) and nodular vasculitides (eg, syphilitic gumma, nodular pernio)
* Differentiate papulonecrotic tuberculid from other papulonecrotic entities, such as leukocytoclastic vasculitis, lymphomatoid papulosis, papular eczema, and prurigo simplex with neurotic excoriation.
* Differentiate lichen scrofulosorum from keratosis spinulosa, lichenoid sarcoid, and lichenoid secondary syphilis

## **Differential Diagnoses**

* Actinomycosis
* Aspergillosis
* Bronchiectasis
* Constrictive Pericarditis
* Fungal Pneumonia
* Histoplasmosis
* Lung Abscess
* Nocardiosis
* Non-Small Cell Lung Cancer (NSCLC)
* Pott Disease (Tuberculous [TB] Spondylitis)

## **Epidemiology**

### United States

With the improvement of living conditions and the introduction of effective treatment (streptomycin) in the late 1940s, the number of patients in the United States with reported TB began to steadily decline (126,000 TB patients in 1944; 84,000 in 1953; 22,000 in 1984; 14,000 in 2004), despite explosive growth in the total population (140 million people in 1946, 185 million in 1960, 226 million in 1980).

In 2022, 8,300 TB cases were reported in the USA, compared with 7,874 cases in 2021. TB incidence increased slightly in 2022 (2.5 cases per 100,000 persons) after an artificially generated substantial decline in 2020. This drop was associated with the COVID-19 pandemic-induced social distancing, masking, and missed or delayed diagnoses. Now, after the social distancing restrictions are lifted, and people are seeking health care increasingly, reported TB cases and TB incidence in the United States are returning to pre-pandemic levels. According to the 2023 CDC report, an estimated 13 million people are living with latent TB in the USA.

### International

In 2023, tuberculosis (TB) was responsible for approximately 1.25 million fatalities, including 161,000 individuals co-infected with HIV. TB likely reclaimed its status as the predominant global cause of mortality from a single infectious agent; a position it held prior to being overtaken by COVID-19 for three years. It continued to be the foremost cause of death among HIV-positive individuals and played a significant role in deaths linked to antimicrobial resistance.

The same year saw an estimated 10.8 million new TB cases worldwide, comprising 6.0 million men, 3.6 million women, and 1.3 million children. TB is ubiquitous, affecting all countries and age demographics, yet it remains both preventable and curable.

### Demographics

In terms of demographics, the majority of reported TB cases occurred among non-US–born persons (71.4%), with the incidence rate being 15.8 times higher among non-US–born persons (12.5 cases per 100,000 persons) than US-born persons (0.8 cases per 100,000 persons). Among non-US–born persons with TB disease in 2021, the most common countries of birth included Mexico, the Philippines, India, Vietnam, and China. Among non-US–born persons, the countries of birth with the highest US incidence rates (cases per 100,000 persons from the country population living in the United States) of TB disease were the Republic of the Marshall Islands, the Republic of Congo, Mongolia, Bhutan, Myanmar, and Somalia. In 2021, among the persons with TB disease in the United States, 36% identified as non-Hispanic Asian persons. The remainder included Hispanic or Latino persons (30.6%), non-Hispanic, African Americans (18.0%), and non-Hispanic White persons (11.2%). TB incidence rates are higher among adults than among children. Among individuals 15 years and older, the incidence rates increase with age, with adults 65 years or older having the highest TB incidence rate in 2021 (4.0 cases per 100,000 persons). In 2021, males accounted for 61.3% of TB cases in the United States, including 62.6% of cases among US-born persons and 60.7% among non-US–born persons

## **Latent Tuberculosis Infection (LTBI) Treatment Guidelines**

The National Tuberculosis Controllers Association (NTCA) and Centers for Disease Control and Prevention (CDC) have issued updated treatment guidelines for latent tuberculosis infection (LTBI) among persons who live in the United States.

The recommended 2020 LTBI treatment guidelines include three preferred rifamycin-based regimens and two alternative daily-isoniazid monotherapy regimens. These recommendations are intended for *Mycobacterium tuberculosis* infections with presumed susceptibility to isoniazid or rifampin. *M tuberculosis* strains resistant to isoniazid and rifampin are exempt from these recommendations.

Generally, rifamycin-based treatment regimens administered in short courses are preferred over isoniazid monotherapy administered in longer courses for treating LTBI.

Preferred treatment regimens for LTBI

The rifamycin-based preferred regimens for LTBI are as follows:

* Once-weekly isoniazid plus rifapentine for 3 months (strongly recommended in adults and children >2 years, including those with HIV infection) OR
* Daily rifampin for 4 months (strongly recommended in HIV-negative adults and children of all ages) OR
* Daily isoniazid plus rifampin for 3 months (conditionally recommended in adults and children of all ages and HIV-positive persons)

Prescribing providers or pharmacists should note that rifampin and rifapentine are not interchangeable, and care should be taken to administer the correct medication for the intended regimen.

Alternative treatment regimens for LTBI

The alternative treatment regimens are as follows:

* Daily isoniazid for 6 months (strongly recommended in HIV-negative adults and children of all ages and conditionally in HIV-positive adults and children of all ages) OR
* Daily isoniazid for 9 months (conditionally recommended in adults and children of all ages regardless of HIV infection status)

The following is a list of commonly occurring adverse events and their associated anti-TB drugs:

* Hepatitis (malaise, fatigue, fever, anorexia, nausea, dark urine)
  + INH
  + Bedaquiline
  + Rifampin
  + Pyrazinamide
* Peripheral neuropathy
  + INH
  + Linezolid
* Ocular toxicity
  + Ethambutol
* Rash
  + Pyrazinamide
  + Ethambutol
  + Fluoroquinolones
  + Amikacin
  + Beta-lactams
  + INH
  + Streptomycin
  + Para-aminosalicylic acid
* Cranial nerve VIII dysfunction and renal dysfunction
  + Amikacin
  + Streptomycin
  + Capreomycin
  + Kanamycin
* Gastrointestinal upset
  + All anti-TB drugs
* Myalgias-arthralgias
  + Bedaquiline
  + Pyrazinamide
* Anxiety, confusion, psychosis
  + Cycloserine
  + Fluoroquinolones
* Hypoglycemia
  + Fluoroquinolones
* Tendonitis
  + Fluoroquinolones

People with TB and HIV coinfection undergoing antiretroviral therapy present additional challenges. Coadministration of anti-TB and antiretroviral agents can pose significant risks due to adverse reactions and drug-drug interactions. For example, rifamycins cause decreased plasma concentrations of protease inhibitors and nonnucleoside reverse-transcriptase inhibitors. In coinfected individuals, the restoration of immunity may result in clinical deterioration due to immune reconstitution inflammatory syndrome. Excellent references provide discussions of the management of these pharmacological challenges

### **PREDEFINED QUESTIONS AND ANSWERS**

### **How common is tuberculosis?**

In 2020, about 10 million people became ill with TB throughout the world, and about 1.5 million people died from it. TB was once the leading cause of death in the U.S., but with new treatments, the number of cases fell rapidly in the 1940s and 1950s. There were 7,860 tuberculosis cases reported in the U.S. in 2021.

## 1. What's the most likely cause of my symptoms?

Your symptoms of chronic cough, fever, night sweats, and weight loss could be caused by tuberculosis (TB), a bacterial infection caused by *Mycobacterium tuberculosis*. However, other conditions can cause similar symptoms, so further evaluation is needed to confirm the diagnosis.

## 2. Do I need tests?

Yes. To diagnose TB, your healthcare provider will perform:

* A medical history and physical exam
* A TB skin test (Mantoux test) or a TB blood test (Interferon Gamma Release Assay, IGRA) to check for immune response to TB bacteria
* A chest X-ray to look for lung changes
* Sputum tests including smear microscopy, molecular tests (PCR), and culture to detect TB bacteria and test for drug resistance  
  These tests help determine if you have latent TB infection or active TB disease.

## 3. What treatments are available? Which do you recommend?

TB is treated with a combination of antibiotics over several months. The most common regimen for drug-sensitive TB includes:

* Isoniazid
* Rifampin
* Ethambutol
* Pyrazinamide  
  Treatment usually lasts 6 months, with the first 2 months including all four drugs (intensive phase), followed by 4 months of isoniazid and rifampin (continuation phase).  
  Your doctor will tailor treatment based on drug susceptibility testing and your overall health.

## 4. What if the treatment doesn't work?

If symptoms persist or worsen, or if drug resistance is detected, your healthcare provider may:

* Perform further testing for drug-resistant TB
* Adjust your treatment regimen with second-line drugs, which may be longer and more complex
* Monitor closely for side effects and adherence to therapy  
  Early detection of treatment failure is important to prevent complications and transmission.

## 5. How long do I have to stay on the treatment?

Standard treatment for drug-sensitive TB is 6 months. For drug-resistant TB, treatment may last 9 months to 2 years depending on the resistance pattern. It is crucial to complete the full course to ensure cure and prevent relapse or resistance.

## 6. How often do I need to follow up with you?

You will have regular follow-ups, especially during the intensive phase (first 2 months), to:

* Monitor your response to treatment
* Check for side effects
* Review sputum test results  
  Follow-up frequency may be weekly or biweekly initially, then monthly during continuation phase. Your doctor will advise based on your progress.

## 7. I have other health problems. How can I best manage these conditions together?

Managing TB alongside other health conditions (like diabetes, HIV, or liver disease) requires:

* Coordinated care between your TB specialist and other healthcare providers
* Close monitoring for drug interactions and side effects
* Adjusting TB treatment if needed based on your other conditions
* Maintaining good control of your other illnesses to support your immune system  
  Be sure to inform all your doctors about your TB diagnosis and medications

## **Genomic Data of *Mycobacterium tuberculosis* (Mtb)**

* Genome Size and Structure:  
  The *M. tuberculosis* genome is a circular chromosome of approximately 4.4 million base pairs (Mb), containing around 4,000 genes (e.g., 4,411,529 bp in the H37Rv reference strain). The genome has a high GC content (~64-65%).
* Gene Content and Families:
  + About 40% of genes have experimentally confirmed functions, with another 44% having putative functions assigned.
  + The genome encodes many genes involved in fatty acid and lipid metabolism (around 250 genes), essential for the bacterium’s waxy, lipid-rich cell envelope and survival in host macrophages.
  + A notable feature is the PE/PPE gene families, which constitute about 10% of the coding capacity. These genes encode glycine-rich proteins involved in antigenic variation and immune evasion.
  + The genome also contains genes related to virulence factors, immune modulation, dormancy, and intracellular survival.
* Lineages and Genetic Diversity:
  + *M. tuberculosis* is classified into multiple lineages (L1–L10), with Lineage 2.2 (Beijing family) and Lineage 4 being prevalent in many regions.
  + Genetic variation among strains is relatively low (monomorphic), but important mutations and gene copy number variations contribute to differences in virulence, drug resistance, and host adaptation.
  + Drug resistance mutations are often found in genes encoding targets of first-line drugs (e.g., isoniazid, rifampicin).
* Virulence and Adaptation:
  + The genome encodes multiple secretion systems, including the ESX-5 type VII secretion system, which exports many PE/PPE proteins important for virulence.
  + Regions of difference (RDs) in the genome distinguish *M. tuberculosis* from related species like *M. bovis* and contribute to host specificity and pathogenicity.
  + Genes involved in host cell entry (e.g., mce family), stress response, and immune modulation are critical for persistence and disease progression

**Doctor-patient conversation about Tuberculosis (TB)**

Doctor: Good morning, Mrs. Rani. I understand you’ve been coughing for several weeks and recently noticed some blood in your sputum. Let’s talk about what might be causing your symptoms.

Patient: Yes, doctor. I’ve been feeling weak and coughing a lot. What could this be?

Doctor: Based on your symptoms and examination, it’s possible you have tuberculosis, or TB. It’s a bacterial infection that mainly affects the lungs and causes a persistent cough, sometimes with blood, along with weight loss, night sweats, and fever.

Patient: How do people get TB? Is it contagious?

Doctor: TB spreads through the air when someone with active TB coughs or sneezes. It’s important to avoid close contact with others until we confirm your diagnosis and start treatment.

Patient: What tests will I need?

Doctor: We’ll do a chest X-ray and collect sputum samples to look for the bacteria. We may also do a skin or blood test to see if your body has been exposed to TB.

Patient: Is TB curable?

Doctor: Yes, TB can be cured with the right antibiotics taken regularly for at least six months. It’s very important to complete the full course to fully clear the infection and prevent resistance.

Patient: What if the treatment doesn’t work?

Doctor: If your symptoms don’t improve or if the bacteria are resistant to standard drugs, we’ll adjust your treatment and may use different medications. We’ll monitor you closely throughout.

Patient: How often will I need to come back?

Doctor: Initially, we’ll see you every few weeks to check your progress and side effects. Once stable, follow-ups will be less frequent but regular until treatment is complete.

Patient: I also have diabetes. Will that affect my TB treatment?

Doctor: It can make TB harder to treat, so we’ll coordinate your care carefully to manage both conditions. Controlling your blood sugar is very important during TB treatment.

Patient’s Husband: What can we do to prevent TB in our family?

Doctor: Good ventilation, covering coughs, and avoiding sharing utensils help reduce spread. Also, make sure everyone completes their vaccinations and gets tested if they have symptoms.

Patient: Thank you, doctor. I feel better knowing what to expect.

Doctor: You’re welcome. Remember, adherence to treatment and regular follow-up are key to your recovery. Don’t hesitate to contact us if you have any concerns.

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**LOWER RESPIRATORY TRACT INFECTIONS**

**DEFINITION AND DESCRIPTION**

**Lower Respiratory Tract Infections (LRTIs)** are severe infections of the lungs that can affect anyone. They are a significant reason for morbidity and mortality in children (below five years of age) and people (above 65) globally.

Lower respiratory tract infections cause difficulty breathing and may threaten the patient's life if left untreated. Keep reading to learn more about the causes of Lower respiratory tract infections, their definition, symptoms, types, diagnosis, prevention, medicine, treatment, and other important detail in this article.

## **Lower Respiratory Tract Infection**

Lower Respiratory Tract Infection refers to infection of the body's airways. These infections develop below the larynx (voice box), including the trachea and the alveolar sacs in the lungs.

Different viruses and bacteria usually cause lower respiratory tract infections. LRTI is a contagious infection which means it can easily spread from one person to another through close contact.

Lower respiratory tract infection generally lasts from 7 to 21 days. However, if lower respiratory infections last for more than 21 days, it could be a sign of a severe lower respiratory tract infection. Hence, it's always best to seek medical care as soon as the patient feels the symptoms.

## **Lower Respiratory Tract Infection Types**

There are various types of lower respiratory tract infections. It depends on the kind of virus or bacteria that causes them. Many of them are treatable. However, if they remain untreated for a long time, they can cause severe health complications.

The important types of lower respiratory infections are as follows:

1. **Pneumonia**: It is a lower respiratory tract infection in which the air sacs in the lungs fill with pus or fluids. This condition could inflame one or both lungs. People with weakened immune systems and infants are more prone to pneumonia than others.
2. **Bronchitis**: It is the swelling in the lining of the bronchial tubes, chiefly the large airways. Bronchial tubes bring in and take out the air from the lungs. Bronchitis is most common in older adults and infants under five years.
3. **Bronchiolitis**: It refers to lung inflammation, especially of the small airways. Bronchiolitis is most common in infants and toddlers under two years of age.
4. **Tuberculosis**: It is an infectious disease that affects the lungs. Tuberculosis can happen to people of any age group.

Other LRTIs include acute exacerbations of chronic obstructive pulmonary disease/chronic bronchitis (AECB) and acute bronchiectasis.

## **Lower Respiratory Tract Infections Causes**

The major causes of lower respiratory tract infections are **viruses and bacteria**. However, the type of bacteria/ virus depends on the specific lower respiratory tract disease.

The following are the major causes of lower respiratory infections:

1. Bacteria such as Mycoplasma pneumonia and Streptococcus pneumonia (pneumococcus), Haemophilus influenzae, Pseudomonas aeruginosa, and so on.
2. Viruses, such as influenza and the respiratory syncytial virus (RSV).
3. Fungal infection

Apart from these, environmental and artificial factors also cause lower respiratory tract infections. These are as follows:

1. Air pollution
2. Tobacco smoke
3. Vapours and fumes
4. Allergens
5. Dust
6. Chemicals

## **Lower Respiratory Tract Infection Risk Factors**

There are several risk factors that make a person more likely to develop lower respiratory infections. Some of the lower respiratory tract infection risk factors include:

1. Weakened immune system
2. Being more than 65 years old
3. Being under five years old
4. Recent surgery
5. A recent cold or flu
6. Smoking

## **Lower Respiratory Tract Infection Symptoms**

The most common lower respiratory infection symptoms are:

1. Breathlessness
2. Dry cough
3. Stuffy nose
4. Sore throat
5. Low fever
6. Dull headache

The major lower respiratory tract infection symptoms are as follows:

1. Sneezing
2. Stuffy nose
3. Runny nose
4. Sore throat
5. Difficulty breathing
6. Headache
7. High fever (above 100 F)
8. Wheezing
9. Cough
10. Muscle pain

## **Lower Respiratory Tract Infections diagnosis**

A primary care physician generally diagnoses lower respiratory tract infections based on the symptoms and physical examination. For physical examination, the doctor will listen to the patient's breathing sound and pattern and check the patient’s history to track any past respiratory disease.

However, the doctor might also conduct numerous medical examinations to determine the exact causative agent of the patient's lower respiratory tract infection. Some of them are as follows:

1. **Blood tests:** To check for the vital measures and presence of bacteria or viruses in the blood.
2. **Sputum test:** To help detect the type of bacteria or virus causing lower respiratory tract infection.
3. **Pulse oximetry:** To check the oxygen level in the patient's bloodstream as lower respiratory tract infection causes difficulty breathing.
4. **Swab test:** To confirm the possibility of a lower respiratory tract infection.
5. **CT scans and chest X-rays:** To diagnose diseases like pneumonia and bronchitis.

Heart tests like Echocardiogram and Electrocardiogram can also be performed to rule out any chances of cardiac diseases.

## **Lower Respiratory Tract Infection Treatment**

### **Home Remedies for Lower Respiratory Tract Infection**

1. **Steam inhalation:** The patient should take inhaling steams as this helps to clear the respiratory tract of any mucus. It relieves nasal congestion as well.
2. **Saline water gargle:** Gargling with saltwater helps relieve pain in the throat. It aids in thinning the mucus lining along the respiratory tract, too.
3. **Ginger:** Ginger is antiviral and antimicrobial. Thus, patients can eat ginger every day to help treat the infection.
4. **Saline nasal drops:** These drops help clear the nasal passageways of any mucus.

### **Medications for Lower Respiratory Tract Infection**

1. **Allopathic Medicines**
   1. Lower respiratory tract infections are often caused by viruses. Hence, antibiotics do not work against them. However, over-the-counter medicines for cough or fever can help with lower respiratory tract infections. Apart from that, patients can also take medication such as a bronchodilator inhaler.
   2. Consult the doctor before taking any medicines for lower respiratory tract infections.
2. **Ayurvedic Medicines:**
   1. **Tulsi:** Holy basil or tulsi is an effective remedy to remove cough from the body. It also strengthens the immune system.
   2. **Neem:** It consists of anti-inflammatory and antimicrobial characteristics. Thereby helping in lower respiratory tract infections.
   3. **Giloy:** It helps to remove toxins from the lungs. Giloy is beneficial in tuberculosis and asthma too.
3. **Homeopathic Medicines:**  
   Homoeopathic medicines are a great treatment for LRTI too. A few helpful homoeopathic medicines are as follows:
   1. Belladonna
   2. Adrenaline
   3. Eriodictyon californicum
   4. Phosphorus
   5. Aconitum napellus

Always consult a homoeopathic doctor before taking any of the above medicines.

### **Other treatment methods for Lower Respiratory Tract Infection**

Intravenous fluid is essential for the body but in a limited amount.

1. **To treat low levels of IV fluid in the body:** Patients can receive oral medications
2. **To treat high levels of IV fluid in the body:** Patients can receive diuretics

To treat oxygen levels in the body, the following treatments are given to the patients:

1. Mechanical ventilation
2. Supplemental oxygen

Note: Please consult a doctor before giving the patient any of the above-mentioned treatments.

## **Risks and Complications of Lower Respiratory Tract Infections**

If lower respiratory tract infections are not treated on time, they might lead to:

1. Pus buildup (in lungs)
2. Inflammation in the lungs
3. Lung abscess
4. Bronchiectasis
5. Deafness

Can also lead to life-threatening conditions such as:

1. Respiratory failure
2. Respiratory arrest
3. Heart failure
4. Sepsis

### **When to see a doctor**

Consult a doctor if the patient feels the following symptoms:

1. Excessive cough
2. Unable to take liquid for 4-5 hours
3. Wheezing
4. High fever
5. Patient Appears pale
6. Feels difficulty breathing
7. Grunts or wheezes

## **Diet for Lower Respiratory Tract Infections**

Staying hydrated and eating a diet rich in minerals and vitamins will help reduce the risk of developing any disease condition. Below are some dietary measures for people with lower respiratory tract infections.

1. Take diets rich in green leafy vegetables, cauliflower, cabbage, peas, broccoli, and spinach.
2. Eat fruits rich in Vitamin C such, as Kiwi, orange, and sweet lime.
3. Avoid refined and processed foods
4. Avoid dairy and high-fat products
5. Avoid sugar and high-sugar products
6. Avoid cold drinks, alcohol and caffeine

### **Lifestyle changes for Lower Respiratory Tract Infections**

1. Start exercising daily. Do yoga and pranayama as it helps improve the breathing process and relieve mucus and nasal congestion.
2. Get 8 hours of good sleep.
3. Wear a face mask when going or doing outdoor activities.
4. Wash or sanitize the hands frequently.

## **Prevention of Lower Respiratory Tract Infection**

Lower respiratory tract infections are highly contagious, especially in small children and older adults. Nevertheless, there are a variety of factors that, if taken into action, can prevent lower respiratory tract infections from occurring. To prevent lower respiratory tract infection, one should follow the crucial guidelines listed below:

1. Wash hands frequently, especially before eating food.
2. Maintain distance from an infected person.
3. Cover the mouth while sneezing or coughing.
4. Get vaccine shots, such as MMR vaccines and pneumococcal vaccines.
5. Stay away from allergens, smoke, chemicals, and other harmful substances that can put one at risk of developing LRTI.
6. Do not visit an LRTI endemic area.
7. Avoid touching any objects, as they might be contaminated with bacteria or viruses.
8. Avoid chewing tobacco.
9. Avoid smoking or drinking alcohol.

## **Differential Diagnosis (DDx) List for Lower Respiratory Tract Infection (LRTI)**

## Viral Causes

* Respiratory Syncytial Virus (RSV)
* Influenza A and B viruses
* Adenoviruses
* Parainfluenza viruses
* Human Metapneumovirus
* SARS-CoV-2 (COVID-19)
* Other respiratory viruses

## Bacterial Causes

* *Streptococcus pneumoniae* (typical pneumonia)
* *Haemophilus influenzae*
* *Mycoplasma pneumoniae* (atypical pneumonia)
* *Chlamydophila pneumoniae*
* *Legionella* species
* *Staphylococcus aureus*
* Gram-negative bacteria (e.g., *Klebsiella*, *Pseudomonas*) especially in hospital-acquired pneumonia
* Anaerobic bacteria (aspiration pneumonia)

## Other Infectious Causes

* *Chlamydia trachomatis* (especially in infants)
* *Chlamydophila psittaci* (psittacosis)
* Mycobacterium tuberculosis (chronic cough, cavitary lesions)
* Fungal infections (e.g., *Histoplasma*, *Aspergillus*) in immunocompromised patients

## Non-Infectious Conditions Mimicking LRTI

* Idiopathic chronic eosinophilic pneumonia
* Organizing pneumonia (cryptogenic or secondary)
* Pulmonary embolism with infarction
* Bronchogenic carcinoma or lung malignancy
* Interstitial lung diseases
* Heart failure with pulmonary edema

## **Epidemiology of Lower Respiratory Tract Infections (LRTIs)**

* Global Burden (2019 Data):  
  There were approximately 489 million new cases of LRTIs worldwide, causing about 2.4 million deaths in 2019. This represents a significant cause of morbidity and mortality globally.
* Incidence and Mortality Rates:  
  The global age-standardized incidence rate was around 6,295 cases per 100,000 population, and the death rate was approximately 34.3 deaths per 100,000 population in 2019.  
  Since 1990, incidence has decreased by about 24%, and mortality has declined by nearly 49% worldwide.
* High-Burden Countries:  
  Countries with the highest incidence rates include Guinea, Chad, and India.  
  Regions with the greatest mortality and disability-adjusted life years (DALYs) include South Asia, Sub-Saharan Africa, and parts of Latin America.
* Age Distribution:  
  The highest burden is seen in children under 5 years of age and adults over 70 years. In 2021, about 502,000 deaths occurred in children under 5, mostly in low socio-demographic index (SDI) countries.
* Risk Factors:  
  Major contributors to LRTI burden include:
  + Child wasting (malnutrition): 33.1%
  + Household air pollution from solid fuels: 24.9%
  + Lack of access to handwashing facilities: 14.4%  
    Additionally, non-optimal temperatures (cold exposure) contribute significantly to LRTI mortality and morbidity.
* Impact of COVID-19 Pandemic:  
  Non-pharmaceutical interventions during the COVID-19 pandemic reduced transmission of many respiratory pathogens, temporarily decreasing LRTI incidence and mortality in 2020–21.
* Sex Differences:  
  Males generally have a higher burden of LRTI deaths compared to females, particularly in older age groups

## **Predefined Questions and Answers for Lower Respiratory Tract Infection (LRTI)**

1. What is a lower respiratory tract infection?  
A lower respiratory tract infection is an infection in the lungs or airways below the voice box, including conditions like bronchitis, bronchiolitis, pneumonia, and tuberculosis.

2. What are the common symptoms of LRTI?  
Common symptoms include cough (which may be severe), fever, chest discomfort, difficulty breathing, wheezing, and sometimes chest pain. In severe cases, there may be rapid breathing, blue discoloration of the skin, and difficulty maintaining oxygen levels.

3. What causes lower respiratory tract infections?  
Most LRTIs are caused by viruses such as respiratory syncytial virus (RSV), influenza, adenovirus, and coronaviruses. Bacteria like *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Mycoplasma pneumoniae* can also cause LRTIs, especially pneumonia.

4. How is LRTI diagnosed?  
Diagnosis is usually based on clinical symptoms and physical examination, supported by chest X-rays and laboratory tests including sputum cultures and viral PCR tests when needed.

5. How are lower respiratory tract infections treated?  
Treatment depends on the cause:

* Viral infections often require supportive care (rest, fluids, oxygen if needed).
* Bacterial infections are treated with appropriate antibiotics.
* Severe cases may require hospitalization and respiratory support.

6. When should I see a doctor for LRTI?  
Seek medical care if you have difficulty breathing, persistent high fever, chest pain, coughing up blood, or if symptoms worsen or do not improve after a few days.

7. Can LRTIs be prevented?  
Prevention includes good hand hygiene, avoiding close contact with sick individuals, vaccination against influenza and pneumococcus, and reducing exposure to air pollution and smoking.

8. What is the difference between upper and lower respiratory tract infections?  
Upper respiratory tract infections affect areas above the larynx (nose, throat), causing symptoms like runny nose and sore throat. Lower respiratory tract infections affect the airways and lungs below the larynx, often causing cough, fever, and breathing difficulties

**Doctor-patient conversation about a lower respiratory tract infection (LRTI)**

Doctor: Hello, what brings you in today?

Patient: I’ve had a cough for about a week now, and I’m feeling quite tired. I had a cold a few days before the cough started.

Doctor: I see. Have you noticed any other symptoms like fever, shortness of breath, or chest pain?

Patient: I had a mild fever a couple of days ago, and sometimes it’s hard to catch my breath, especially when I’m active.

Doctor: Thanks for sharing. Based on your symptoms and the exam, it looks like you have a viral lower respiratory tract infection, similar to acute bronchitis. This is common, especially during this season.

Patient: Do I need antibiotics? I’m worried it might get worse.

Doctor: In most cases like yours, antibiotics aren’t needed because viruses cause the infection. Antibiotics won’t help and can cause side effects. The best treatment is rest, plenty of fluids, and medications to relieve symptoms like fever or cough if needed.

Patient: What if I don’t get better or if it gets worse?

Doctor: If your symptoms worsen—like high fever, difficulty breathing, or coughing up blood—or if you don’t improve after 10 days, please come back. We can then do further tests, like chest X-rays or sputum tests, to check for bacterial infections or other causes.

Patient: How long will this last?

Doctor: Most people start feeling better within 1 to 2 weeks, but sometimes the cough can linger for a few more weeks.

Patient: Is there anything I can do to prevent this in the future?

Doctor: Good question. Regular handwashing, avoiding close contact with sick people, and staying up to date with vaccinations like the flu shot help reduce your risk.

Patient: Okay, thank you. I’ll follow your advice.

Doctor: You’re welcome. Feel free to call or come in if you have any concerns or if your symptoms change.

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**MUSCULAR DYSTROPHY**

**DEFINITION AND DESCRIPTION**

Muscular dystrophy is a group of diseases that causes muscles to become weaker and lose mass over time. The condition is caused by changes in the genes that make proteins needed to form healthy muscles.

There are many types of muscular dystrophy. Symptoms of the most common type start in childhood, mostly in boys. Other types sometimes don't start until adulthood.

Medicines and other treatments can help manage the symptoms and slow the course of muscular dystrophy.

#### **Types of muscular dystrophy**

There are more than 30 types of muscular dystrophy. Some of the more common forms include:

* **Duchenne muscular dystrophy (DMD)**: This is the most common form of muscular dystrophy. It mainly affects boys, but girls can also have a milder version of it. As DMD progresses, it affects your heart and lungs.
* **Becker muscular dystrophy (BMD)**: BMD is the second most common type of muscular dystrophy. It mainly affects boys, but girls can have milder symptoms. Symptoms of BMD can appear any time between the ages of 5 and 60, but they typically start by your teenage years. The severity of BMD varies from person to person.
* **Myotonic dystrophy**: This is the most common type of muscular dystrophy that’s diagnosed in adulthood. It affects men and women equally. People with myotonic dystrophy have difficulty relaxing their muscles after using them. The condition can also affect your heart and lungs and may cause endocrine issues, such as thyroid disease and diabetes.
* **Congenital muscular dystrophies (CMD)**: CMD refers to a group of muscular dystrophies that become apparent at or near birth (“congenital” means “present from birth”). CMD causes overall muscle weakness with possible joint stiffness or looseness. Depending on the type, CMD may also involve spinal curvature (scoliosis), breathing issues, intellectual disabilities, learning disabilities, eye issues or seizures.
* **Distal muscular dystrophy**: This type affects the muscles of your hands, feet, lower arms and lower legs. It tends to affect people in their 40s and 60s.
* **Emery-Dreifuss muscular dystrophy (EDMD)**: EDMD mainly affects male children and young adults. It tends to cause muscle weakness in your shoulders, upper arms and shins. EDMD also affects your heart. The condition usually progresses slowly.
* **Facioscapulohumeral muscular dystrophy (FSHD)**: FSHD most commonly affects muscles in your face, shoulders and upper arms. Symptoms tend to appear before age 20. About 4 out of 100,000 people in the U.S. have this form.
* **Limb-girdle muscular dystrophy (LGMD)**: LGMD affects the muscles in your upper arms, upper legs, shoulders and hips. It affects people of all ages. Approximately 2 out of 100,000 people in the U.S. have LGMD.
* **Oculopharyngeal muscular dystrophy (OPMD)**: OPMD weakens muscles in your eyelids and throat. Symptoms, such as droopy eyelids (ptosis) and difficulty swallowing (dysphagia), often appear in your 40s or 50s. About 1 in 100,000 people have OPMD.

Muscular dystrophy is relatively rare. All of the different types combined affect about 16 to 25 per 100,000 people in the U.S.

The most common childhood form is Duchenne muscular dystrophy. The most common adulthood form is myotonic dystrophy.

### **causes muscular dystrophy**

Mutations (changes) in the genes that are responsible for healthy muscle structure and function cause muscular dystrophy. The mutations mean that the cells that would normally maintain your muscles can no longer fulfill this role, leading to progressive muscle weakness.

There are several genes — and possible genetic mutations — that play a role in muscle function. This is why there are so many different forms of muscular dystrophy.

In the majority of muscular dystrophy cases, you inherit the genetic mutation from one or both of your biological parents.

There are three ways you can inherit muscular dystrophy, depending on the specific type:

* **Recessive inheritance**: This means you’ve inherited a genetic mutation that causes the condition from both of your biological parents. Some forms of limb-girdle muscular dystrophy have this inheritance.
* **Dominant inheritance**: This means you only need to inherit the mutated gene from one of your biological parents to develop the condition. Myotonic, facioscapulohumeral and oculopharyngeal muscular dystrophies have this type of inheritance.
* **Sex-linked (X-linked) inheritance**: A genetically male person has one X and one Y chromosome, and a genetically female person has two X chromosomes. A genetic mutation on the X chromosome causes a sex-linked condition. As genetically male people only have one copy of each gene on the X chromosome, they’ll develop the condition if one of those genes is mutated. A genetically female person can have X-linked disorders, but the symptoms are usually less severe. Duchenne and Becker muscular dystrophies have this type of inheritance.

In rare cases, a person may develop muscular dystrophy spontaneously, meaning the mutation happened randomly and wasn’t inherited. This is called a *de novo* mutation.

## **Risk factors**

Muscular dystrophy occurs in people of all ages and ethnic groups. But the most common type, Duchenne, usually affects young boys. People with a family history of muscular dystrophy have a higher risk of getting the disease or passing it on to their children.

## **Symptoms**

The main symptom of muscular dystrophy is muscle weakness that becomes worse over time. This makes everyday tasks harder to do. The type of muscular dystrophy that a person has determines the:

* Exact symptoms.
* Age range at which the symptoms start.
* Muscle groups that are affected.

There are over 30 types of muscular dystrophy. Here are some of the main types along with key symptoms.

### **Duchenne type muscular dystrophy**

This is the most common form. Girls can carry the gene change that causes the disease, and some have symptoms. But this type of muscular dystrophy is much more common in boys.

Most often, the symptoms of Duchenne muscular dystrophy start in early childhood. The symptoms can include challenges with movement, such as:

* Late walking.
* Frequent falls.
* Trouble rising from the floor or a lying or seated position.
* Trouble running, jumping or climbing stairs.
* Waddling gait.
* Walking on the toes or the balls of the feet.

Other symptoms can include:

* Large calf muscles.
* Muscle pain and stiffness.
* Learning or behavior-related challenges.
* Delayed growth.

### **Becker muscular dystrophy**

The symptoms of this type are like those of Duchenne muscular dystrophy, but Becker muscular dystrophy tends to be milder, and it becomes worse more slowly. In general, symptoms start in the teens or early adulthood. They might not occur until the mid-20s or later.

### **Other types of muscular dystrophy**

Some types of muscular dystrophy are defined by a certain feature. Or they're defined by where in the body the symptoms start. Examples include:

* **Emery-Dreifuss.** The symptoms of this type often start by age 10. Emery-Dreifuss muscular dystrophy causes certain joints to become stiff. Early on, some children may walk on their toes due to stiff tendons in the heels. They also may have trouble bending the elbows. This type of muscular dystrophy also causes muscles in the shoulders, upper arms and calves to slowly waste and weaken. Some people with the disease develop heart conditions that can cause fainting. Heart conditions need to be watched closely by a healthcare team.
* **Myotonic.** With this type of muscular dystrophy, the muscles can't relax at will. For example, it might be hard to let go of someone's hand after shaking it. Facial and neck muscles often are the first to be affected. Symptoms often start between the ages of 20 and 30, but some have symptoms shortly after birth to childhood. As the disease becomes worse, the heart might beat out of rhythm and the heart muscle can grow weaker. Heart rhythm issues can be the first complication for some people. Muscles involved in breathing also can become weaker. This can lead to poor breathing, especially during sleep. Other names for myotonic dystrophy are Steinert's disease or dystrophia myotonica.
* **Facioscapulohumeral (**FSHD)**.** With this type, muscle weakness usually starts in the face, shoulders and upper arms. The weakness often affects one side of the body more than the other. When muscles around the eyes are affected, that can cause trouble fully closing the eyelids and lead to dryness of the eye. When the shoulders are affected, the shoulder blades might stick out like wings when the arms are raised. Sometimes, mild hearing loss also may occur. The symptoms of FSHD tend to start in the teenage years.
* **Congenital.** There are many types of congenital MD. These types cause symptoms at birth or before age 2. Most children with congenital muscular dystrophy have muscle weakness that becomes worse. But some forms of congenital muscular dystrophy progress slowly and cause mild disability. Others become worse quickly and cause serious health challenges. Babies with muscle weakness or lack of muscle tone can seem "floppy." Later on, babies and toddlers might take longer than usual to roll over, sit up or walk. Or they might not meet certain milestones of development at all.
* **Limb-girdle.** This type often affects muscles around the shoulders and hips. Some people with limb-girdle dystrophy develop minor disabilities over time. Others develop serious trouble using their arms or legs to do everyday activities, such as walking or carrying things. The age at which symptoms start, how serious they become and how quickly they get worse varies.
* **Oculopharyngeal.** This type tends to weaken the eyelids and the throat muscles. It's also been linked with muscle weakness in the limbs and near the center of the body from top to bottom. It can cause trouble swallowing, weakness of the tongue and drooping eyelids. In time, some people also have trouble with movement. Symptoms often start in the 40s and 50s, and they become worse slowly.

## **When to see a doctor**

Get medical advice if you notice symptoms of muscle weakness in yourself or your child. Symptoms could include delays in development, more clumsiness than usual and falling.

## **Diagnosis**

Your healthcare team asks about your or your child's medical history, including general health and past illnesses. A physical exam is done.

After that, the healthcare team might recommend tests. These tests are based on the type of muscular dystrophy suspected. They may include the following:

* **Enzyme tests.** Damaged muscles release proteins called enzymes into the blood. These enzymes include creatine kinase. In a person who hasn't had a serious injury, high blood levels of creatine kinase suggest a muscle disease.
* **Genetic testing.** Blood samples can be checked for changes in some of the genes that cause types of muscular dystrophy.
* **Muscle biopsy.** A small piece of muscle can be removed through an incision or with a hollow needle. Then a lab checks this tissue sample. The lab can tell muscular dystrophies apart from other muscle diseases.
* **Tests to monitor the heart, such as electrocardiography and echocardiogram.** These tests are used to check how well the heart works, especially in people who have myotonic muscular dystrophy.
* **Tests to monitor the lungs.** These tests are used to check lung function.
* **Electromyography.** A special needle is placed into the muscle to be tested. Electrical activity is measured as the muscle is relaxed and as it is gently tightened. Changes in the pattern of electrical activity can confirm a muscle disease. This test rarely is used to find out if Duchenne or Becker muscular dystrophy is the cause of someone's symptoms.

## **Treatment**

No cure exists for any type of muscular dystrophy. But treatment for some types of the disease can help people:

* Maintain muscle strength.
* Prevent complications.
* Stay independent and mobile longer.
* Improve heart and lung health.

Treatments for some types of Duchenne muscular dystrophy, in particular, are quickly expanding based on research.

People with muscular dystrophy often need to be monitored throughout life by a team of healthcare professionals. A primary care doctor often helps oversee your overall medical care. Most often, the care team includes:

* A doctor called a neurologist, who treats brain and nervous system conditions. It's key to find a neurologist who has experience treating diseases that affect how muscles work due to nerves and muscles in the body. These are called neuromuscular diseases.
* A doctor called a physical medicine and rehabilitation physician or physiatrist, who helps people with disabilities function better, be more independent and have less pain.
* A physical therapist, who teaches exercises to improve movement and keep muscles stronger and flexible for longer.
* An occupational therapist, who teaches various ways to make everyday tasks easier.

Many people with muscular dystrophy also might need other doctors, including a:

* Lung doctor called a pulmonologist, as well as a respiratory therapist who helps with breathing.
* Heart doctor called a cardiologist.
* Doctor whose speciality is treating sleep conditions.
* Hormone expert called an endocrinologist to help with bone density and growth concerns.
* Orthopedic surgeon, who treats problems with bones, joints, ligaments, tendons and muscles.
* Doctor called a gastroenterologist, who treats problems with the stomach and intestinal tract.
* A palliative care doctor, who cares for people living with serious illnesses.
* A genetics doctor or counselor to help guide you about the course your muscular dystrophy likely will take.

Many people with muscular dystrophy also might need to see other specialists, such as a:

* Speech therapist for swallowing and communication.
* Dietitian or nutritionist, who recommends special diets when needed.
* Social worker for support and community resources.
* Psychologist or psychiatrist to assess and treats mood or behavior-related conditions.

Muscular dystrophy treatment includes medicines, physical and occupational therapy, equipment, surgery, and other procedures. Ongoing tests of walking, swallowing, breathing and hand function help the treatment team change treatments as needed over time.

### **Medicines**

Your healthcare team might recommend medicines such as:

* **Corticosteroids.** These medicines can help with muscle strength and slow some types of muscular dystrophy from becoming worse. Examples of corticosteroids include prednisone and deflazacort (Emflaza). Long-term use of this type of medicine can cause weight gain and weaker bones. That raises the risk of breaking a bone, also called a fracture.
* **Targeted medicines and gene therapies.** These are tailored to treat some people with Duchenne muscular dystrophy who have certain confirmed gene changes. More and more treatments are becoming available based on research. These include eteplirsen (Exondys 51), golodirsen (Vyondys 53), viltolarsen (Viltepso), casimersen (Amondys 45), and delandistrogene moxeparvovec-rokl (Elevidys).

Ask your healthcare professional what your treatment choices are. Approved targeted treatments may vary by country.

* **Heart medicines.** If muscular dystrophy damages the heart muscle and causes heart symptoms, these medicines may be prescribed. They include angiotensin-converting enzyme (ACE) inhibitors and beta blockers.

### **Therapy**

Various therapy and assistive devices can improve the quality of life in people with muscular dystrophy. They may help some people live longer too. Examples include:

* **Range-of-motion and stretching exercises.** Muscular dystrophy can limit how flexible and mobile affected joints are. Range-of-motion exercises can help to keep joints as flexible as possible.
* **Exercise.** Low-impact aerobic exercise can help with strength, movement and general health. Examples include walking and swimming. Some types of strengthening exercises also might be helpful. But talk to your healthcare professional before you start exercising. Some types of workouts might be harmful or unsafe to do.
* **Braces.** These devices can help keep muscles and tendons stretched and flexible. This slows contractures from becoming worse. Braces also can aid movement and function by supporting weaker muscles.
* **Mobility aids.** Canes, walkers and wheelchairs can help people with muscular dystrophy move around and stay independent.
* **Breathing assistance.** As muscles involved in breathing become weaker, deep breathing and coughing exercises taught by a healthcare professional can help. Some people also need help breathing during sleep. A device that delivers air through a face mask may be prescribed. Other people who have serious breathing problems need to use a machine that forces air in and out of the lungs, called a ventilator.

### **Surgery**

Surgery might be needed to fix contractures or a curve in the spine that could make breathing harder over time. Heart health may be improved with a pacemaker or other heart device.

### **Preventing respiratory infections**

Illnesses that affect the muscles involved in breathing can become a problem with muscular dystrophy. So, it's key to be vaccinated for pneumonia. Also, keep up to date with shots that protect against the flu and COVID-19. Try to stay away from people who are sick.

## **Complications**

Muscular dystrophy can lead to serious health issues, such as the following:

* **Trouble walking.** In time, some people with muscular dystrophy will use a wheelchair for mobility.
* **Trouble using arms.** Daily activities can become harder if the muscles of the arms and shoulders are affected.
* **Shortening of muscles or tendons around joints, called contractures.** Along with the weakness caused by the muscular dystrophy, less range of motion in joints also can limit walking and arm use.
* **Trouble breathing or coughing.** Over time, muscular dystrophy can affect the muscles involved in breathing. Some people who develop serious trouble breathing might need to use devices that help keep the airways open or assist with breathing. Muscular dystrophy also can affect muscles that are used to cough. That can make it hard for the body to remove mucus from the lungs and windpipe. If your child has trouble coughing, ask your healthcare professional to recommend techniques or treatments that can help.
* **Curved spine, also called scoliosis.** This is most likely to happen in a growing child who can't walk. The curving may play a role in breathing troubles and challenges with getting into certain positions. Sometimes, surgery is needed to correct the curving.
* **Heart problems.** Muscular dystrophy can make it harder for the heart to pump well or beat with a regular rhythm.
* **Swallowing problems.** If the muscles involved with swallowing are affected, that can lead to trouble getting enough nutrition. Trouble swallowing also raises the risk of a lung infection caused by breathing food or liquid into the airways. This is called aspiration pneumonia. To lower these risks, nutrition may need to be given through a flexible device called a feeding tube.
* **Brain-related differences.** Some children with certain types of muscular dystrophy have challenges with learning, intelligence or social skills. For example, those with Duchenne muscular dystrophy may have higher rates of autism spectrum disorder, ADHD, obsessive compulsive disorder and anxiety compared with people who don't have muscular dystrophy. If your child has a learning, intellectual or mental health condition, talk with your child's school. Ask for changes that can help your child learn more easily, called accommodations.
* **Weakening bones.** Children may have fewer minerals in their bones than usual. This is called low bone density. It can lead to weaker bones over time, especially in children who use wheelchairs. Low bone density raises the risk of broken bones, also called fractures. A healthcare professional might prescribe medicines to help strengthen weak bones.

## **Outlook / Prognosis**

The prognosis (outlook) for muscular dystrophy varies depending on the type. Your healthcare provider will be able to give you a better idea of what to expect based on the type of muscular dystrophy you have and your unique situation.

The life expectancy for muscular dystrophy varies significantly depending on the type.

For example, people with Duchenne muscular dystrophy (DMD) often die from the condition by the age of 25. But other forms of muscular dystrophy, such as oculopharyngeal muscular dystrophy, don’t typically affect life expectancy.

**Living With**

If you have muscular dystrophy or you’re taking care of someone with it, it’s important to advocate for yourself/them to ensure you/they get the best medical care and as much access to therapy as possible. Advocating for care can help you/them have the best possible quality of life.

You and your family may also want to consider joining a support group to meet others who can relate to your experiences.

**DIFFERENTIAL DIAGNOSIS**

Muscular dystrophy is not always a straightforward diagnosis and can be mistaken for several other diseases with overlapping etiologies. Clinicians must carefully consider and exclude the following conditions in patients presenting with a muscular disorder, as these alternative diagnoses can lead to significant morbidity and mortality:

* Adrenal insufficiency
* Electrolyte imbalance: sodium, potassium, and magnesium
* Hypercalcemia
* Porphyrias
* Rabies
* Complicated migraine
* Postictal (Todd) paralysis
* Hypoglycemia
* Acute spastic paraparesis (a medical emergency)
* Myasthenia gravis
* Pancoast tumor
* Paraneoplastic syndromes

An acute onset gait disorder is often indicative of acute systemic decompensation. A thorough and systematic evaluation is essential to exclude catastrophic presentations. It is not recommended to attribute a gait disorder to a single disease. Gait abnormalities can indicate a severe medical emergency, especially when the problem is associated with any of these additional symptoms:

* Headache (raised intracranial pressure)
* Nausea or vomiting
* Decreased alertness
* Impaired coordination presenting on only one side of the body
* Recent viral illness/immunization (Guillain-Barré syndrome)
* Trauma.

## **Epidemiology**

### United States statistics

The incidence of MD varies, depending on the specific type of MD under consideration. Duchenne MD is the most common MD and is sex-linked, with an inheritance pattern of 1 case per 3500 live male births.One third of cases occur as a result of spontaneous new mutations. Becker MD is the second most common form, with an incidence of 1 case per 30,000 live male births.Other types of MD are rare. For example, limb-girdle dystrophy occurs in only 1.3% of patients with MDs.

### International statistics

The incidence internationally is similar to that of the US for most of the dystrophies, except for the oculopharyngeal type, which is more common in French Canadians than in other groups.Distal MD tends to occur in Sweden.

**GUIDELINES**

## Screening

In 2013, the American Academy of Pediatrics (AAP) published a clinical report from a multidisciplinary expert panel that developed an algorithm for the screening of children for motor delays with guidance for the initial workup and referral. Identification of motor delays includes ongoing surveillance of the following milestones:

* Sitting
* Standing
* Walking
* Running
* Going up stairs

The report recommends that developmental screenings take place at well-child visits at 9, 18, 30, and 48 months of age. The following motor skills are typically acquired at earlier ages, and their absence at these ages signifies delay:

* 9-month visit - Roll to both sides, sit well without support, and demonstrate motor symmetry without established handedness; ability to grasp and transfer objects hand to hand
* 18-month visit - Sitting, standing, and walking independently; ability to grasp and manipulate small objects; mild motor delays undetected at the 9-month screening visit may now be apparent
* 30-month visit - Most motor delays will have already been identified and more subtle gross motor, fine motor, speech, and oral motor impairments may emerge; progressive neuromuscular disorders may manifest as a loss of previously attained gross or fine motor skills
* 48-month visit - Elementary school skills, with emerging fine motor, handwriting, gross motor, communication, and feeding abilities that promote participation with peers in group activities; loss of skills should alert to the possibility of a progressive disorder

The Centers for Disease Control and Prevention (CDC) also supports early identification and evaluation of motor delays to enable a quicker referral to a specialist for diagnosis. In collaboration with the National Task Force for Early Identification of Childhood Neuromuscular Disorders, the CDC developed a Web-based diagnostic tool, to assist providers in primary care, rehabilitation medicine, and physical and occupational therapy in the evaluation of children with motor delay and early manifestations of neuromuscular disorders. The website content was endorsed by the AAP.

ChildMuscleWeakness.org provides guidance on motor surveillance and screening and includes an aid to the assessment of motor development milestones and recommendations for evaluating the following milestones :

* Infant+: Head lag on pull to sit
* Age 6+ months: Achieving and maintaining sitting
* Age 12+ months: Rising to stand from the floor and gait (walking and running)

If a delay is found by using the surveillance aid, a motor delay algorithm provides guidance on testing and referral. The following findings are red flags that indicate the need for an urgent referral to a neurologist:

* Tongue fasciculations
* Loss of motor milestones
* Creatine phosphokinase (CK) level higher than three times normal (however, children with some neuromuscular disorders have normal CK levels)

Many neuromuscular conditions increase the risk for malignant hyperthermia with anesthesia use, and anticipated surgery should increase the urgency of a diagnostic evaluation.

Questions to consider when evaluating a patient:

**Are reflexes hypoactive?** Hypoactive reflexes may suggest myotonic dystrophy, tabes dorsalis, and progressive muscular atrophy. However, diminished reflexes can also be a result of weakness.

**Is there any pain present?** Muscle weakness and, occasionally, myalgia are typical features of muscle disease. Conversely, a short history of painful weakness in adulthood would suggest an inflammatory myositis.

**Is the problem acute or chronic?** The age and rate of progression can help determine the type of muscle disease. Progressive slow weakness without pain from childhood may indicate degenerative muscular dystrophy. Diseases like Dreifuss muscular dystrophy and Bethlem myopathy cause early fixed contractures, representing distinctive features.

**Is weakness primarily distal or proximal?** The distribution of weakness helps define the likely type of muscle disease. Proximal arm and leg weakness may be present in limb-girdle muscular dystrophy.

**Is the weakness localized or diffuse?** Muscle weakness could be attributed to various causes, including myopathies (e.g., dermatomyositis), inflammatory diseases (e.g., rheumatoid arthritis), neurologic disorders (e.g., Guillain-Barré syndrome), or infections (e.g., Lyme disease or trichinosis)

**Are there any focal neurological lesions?** Conditions like myotonic dystrophy, myasthenia gravis, and progressive muscular atrophy may present with partial ptosis.

**Duchenne muscular dystrophy**

* **Genetics:** X-linked recessive disorder caused by mutations in the DMD gene.
* **Onset:** Typically, early childhood (around 3 to 5).
* **Clinical features:** Progressive muscle weakness, particularly in the pelvic and shoulder girdle muscles.
* **Diagnostic tests:** Elevated CK levels, genetic testing for DMD gene mutations.
* **Prognosis:** Rapid progression, wheelchair dependence by adolescence, cardiopulmonary complications.

**Becker muscular dystrophy**

* **Genetics:** X-linked recessive disorder caused by mutations in the DMD gene.
* **Onset:** Later than DMD, often in adolescence or adulthood.
* **Clinical features:** Similar to DMD but with milder symptoms and slower progression.
* **Diagnostic tests:** Elevated CK levels, genetic testing for DMD gene mutations.
* **Prognosis:** Variable, some individuals may remain ambulatory into adulthood.

**Myotonic dystrophy**

* **Genetics:** Autosomal dominant disorder, involving DMPK gene (DM1) or CNBP gene (DM2).
* **Clinical features:** Myotonia, muscle wasting, cataracts, cardiac conduction abnormalities.
* **Diagnostic tests:** Genetic testing for CTG repeats (DM1) or CCTG repeats (DM2).
* **Prognosis:** Variable, with a range of severity; may involve multiple systems.

**Facioscapulohumeral muscular dystrophy**

* **Genetics:** Autosomal dominant disorder, involving DUX4 gene.
* **Clinical features:** Progressive weakness in face, shoulders, and upper arms.
* **Diagnostic tests:** Genetic testing for DUX4 gene mutations.
* **Prognosis:** Variable, may involve asymmetric muscle weakness.

**Limb-Girdle muscular dystrophy**

* **Genetics:** Heterogeneous group, both autosomal dominant and recessive forms.
* **Clinical features:** Proximal muscle weakness in the pelvic and shoulder girdles.
* **Diagnostic tests:** Genetic testing for various associated genes associated.
* **Prognosis:** Variable, depending on the specific subtype.

**Emery-Dreifuss muscular dystrophy**

* **Genetics:** X-linked or autosomal dominant, involving EMD and LMNA genes.
* **Clinical features:** Early contractures, muscle wasting, cardiac conduction abnormalities.
* **Diagnostic tests:** Genetic testing for EMD and LMNA gene mutations.
* **Prognosis:** Cardiac complications can be life-threatening

## **Predefined Questions and Answers for Muscular Dystrophy**

1. What is muscular dystrophy?  
Muscular dystrophy is a group of genetic disorders that cause progressive weakening and wasting of the muscles due to mutations affecting muscle structure and function.

2. What causes muscular dystrophy?  
MD is caused by mutations in genes responsible for healthy muscle proteins. Most forms are inherited, but some cases result from spontaneous gene mutations with no family history.

3. What are the common symptoms?  
Symptoms include muscle weakness, difficulty walking, frequent falls, trouble climbing stairs, muscle pain or stiffness, and sometimes respiratory or cardiac problems depending on the type.

4. How is muscular dystrophy diagnosed?  
Diagnosis involves clinical examination, family history, blood tests for muscle enzymes (like creatine kinase), genetic testing, muscle biopsy, and sometimes MRI or electromyography.

5. Is there a cure for muscular dystrophy?  
Currently, there is no cure. Treatment focuses on managing symptoms, slowing progression, and improving quality of life.

6. What treatments are available?

* Physical therapy and regular low-intensity exercise to maintain muscle strength.
* Corticosteroids like prednisone or deflazacort to slow muscle degeneration, though they have side effects.
* Newer drugs such as eteplirsen target specific genetic defects in Duchenne muscular dystrophy (DMD).
* Supportive care for respiratory and cardiac complications.

7. How does muscular dystrophy affect life expectancy?  
Life expectancy varies by type. For example, Duchenne muscular dystrophy often reduces life expectancy to early adulthood, while other types like oculopharyngeal MD may have normal life expectancy.

8. Can muscular dystrophy be prevented?  
Since MD is genetic, it cannot be prevented. However, genetic counseling and prenatal testing are options for families with a history of MD.

9. How can I manage muscular dystrophy daily?  
Management includes physical therapy, assistive devices, respiratory care, cardiac monitoring, and joining support groups for emotional and practical help.

10. Are there ongoing research and clinical trials?  
Yes, research is ongoing to develop gene therapies, exon-skipping drugs, and other treatments to alter disease progression, especially for Duchenne MD

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand you or your child have been experiencing muscle weakness and difficulty with movement. Let’s talk about what might be going on.

Patient/Parent: Yes, we’ve noticed increasing weakness, especially in the legs, and trouble walking. What could be causing this?

Doctor: Based on your symptoms and examination, it’s possible you have a type of muscular dystrophy. Muscular dystrophy is a genetic condition that causes progressive muscle weakness over time.

Patient/Parent: Is this something inherited? How serious is it?

Doctor: Most types of muscular dystrophy are inherited, often passed down through families, though sometimes a new mutation can occur. The severity and progression vary depending on the type. For example, Duchenne muscular dystrophy usually starts in early childhood and progresses faster, while other types may have a milder course.

Patient/Parent: How do you confirm the diagnosis?

Doctor: We’ll do some blood tests to check muscle enzymes like creatine kinase, which are often elevated. Genetic testing can identify specific mutations. Sometimes a muscle biopsy or imaging like MRI helps. These tests help us confirm the diagnosis and determine the exact type.

Patient/Parent: Is there a cure?

Doctor: Currently, there is no cure for muscular dystrophy. However, treatments like physical therapy, medications such as corticosteroids, and newer gene-targeted therapies can help slow progression, improve muscle strength, and maintain quality of life.

Patient/Parent: What kind of care will be needed long-term?

Doctor: Managing muscular dystrophy requires a team approach, including neurologists, physical and occupational therapists, cardiologists, and respiratory specialists. Regular monitoring helps manage complications like heart and lung issues.

Patient/Parent: What can we do at home to help?

Doctor: Maintaining gentle exercise and stretching can help keep muscles flexible. Using assistive devices when needed improves mobility and independence. It’s also important to watch for breathing or swallowing difficulties and report them promptly.

Patient/Parent: How will this affect life expectancy?

Doctor: It depends on the type. Duchenne muscular dystrophy can reduce life expectancy, but with advances in care, people are living longer. Other types may have little or no impact on lifespan.

Patient/Parent: Are there any new treatments or research?

Doctor: Yes, research is ongoing, including gene therapies and drugs that target specific genetic defects. Clinical trials may be available, and we can discuss if you’re interested.

Patient/Parent: Thank you, doctor. This helps us understand what to expect and how to manage the condition.

Doctor: You’re welcome. We’ll work together to provide the best care and support for you or your child. Please feel free to ask any questions anytime.

REFERENCES

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### **Spinal muscular atrophy**

**DEFINITION AND DESCRIPTION**

Spinal muscular atrophy (SMA) represents a group of genetic (inherited) neuromuscular disorders that cause certain muscles to become weak and waste away (atrophy).

SMA involves the loss of a specific type of nerve cell in your spinal cord called lower motor neurons, or anterior horn cells. These cells control muscle movement. Without these motor neurons, muscles don’t receive the nerve signals that make them move.

The weakness in SMA tends to be more severe in the muscles that are close to the center of your body (proximal muscles) than in the muscles farther away from your body’s center (distal muscles). Muscle weakness tends to worsen with time.

#### **Types of SMA**

There are five subtypes of SMA. Healthcare providers classify them based on the age of onset, as well as the severity and life expectancy. The subtypes include:

* **SMA type 0 (congenital SMA)**: This is a rare subtype that affects a fetus before birth. Pre-birth, there are typically decreased fetal movements. At birth, infants with type 0 have severe muscle weakness and typically go into respiratory failure. Death usually happens at birth or within the first month of life.
* **SMA type 1 (severe SMA)**: About 60% of SMA cases are type 1 — also called Werdnig-Hoffman disease. Symptoms arise within the first six months of life and include limited head control and decreased muscle tone (hypotonia). Infants with type 1 SMA also have difficulty swallowing and breathing. Without breathing support, children with type 1 SMA die before their second birthday.
* **SMA type 2 (intermediate SMA)**: Symptoms of type 2 SMA (also called Dubowitz disease) appear between six months and 18 months of life. Symptoms include hypotonia and worsening muscle weakness, which tends to affect their legs more than their arms. Children with type 2 SMA may be able to sit up but can’t walk. Around 70% of people with type 2 will survive until 25, with some surviving into their 30s. Respiratory issues are the major cause of death.
* **SMA type 3 (mild)**: Symptoms of type 3 SMA (also called Kugelbert-Welander disease) appear after a child’s first 18 months of life. Type 3 symptoms include lower limb muscle weakness, leading to difficulty walking. People with type 3 MSA don’t tend to develop breathing issues, and it typically doesn’t affect life expectancy.
* **SMA type 4 (adult):** This is the mildest form of SMA. It doesn’t typically appear until after the age of 21. Muscle weakness symptoms progress slowly, so most people with type 4 remain mobile. It typically doesn’t affect life expectancy.

Although SMA is thought of as uncommon, it’s the second most common severe hereditary disease of infancy and childhood after cystic fibrosis. Researchers estimate that it affects between 1 in 6,000 to 1 in 11,000 live births. SMA is about twice as common in white and Asian people as it is in Black and Hispanic people.

### **causes spinal muscular atrophy (SMA)**

SMA is a genetic condition, which means you inherit genes from your biological parents that cause the condition.

Mutations (changes) in the *SMN1* (survivor motor neuron 1) gene cause all types of spinal muscular atrophy. The number of copies that you have of the *SMN2* gene alters the severity of the condition.

A healthy *SMN1* gene produces SMN protein. Motor neurons need this protein to survive and function properly. If you have SMA, your body doesn’t make enough SMN protein, so your motor neurons shrink and die. As a result, your brain can’t control voluntary movements, especially motion in your head, neck, chest and legs.

The *SMN2* gene also produces a small amount of SMN protein. A person may have up to eight copies of an *SMN2* gene. Having multiple copies of the *SMN2* gene typically leads to less severe SMA symptoms because the extra genes make up for the missing *SMN1* protein.

#### **Inheritance pattern of SMA**

You inherit SMA in an autosomal recessive pattern, which means both of your biological parents pass on mutations in the *SMN1* gene. In most cases, the biological parents of someone with an autosomal recessive condition each carry one copy of the mutated gene. But these carriers typically don’t have symptoms of the condition. In the general population, mutations of the *SMN1* gene are common. Among white people, 1 in 50 may be a carrier.

In rare cases, you may inherit an *SMN1* gene mutation from one parent and acquire a new mutation in the other copy of the gene during embryonic development.

### **symptoms of SMA**

In general, the main symptom of SMA is muscle weakness — typically, in the muscles closest to the center of your body. But the symptoms vary based on the type.

Symptoms of SMA type 0 include:

* Decreased fetal movements.
* Arthrogryposis.
* Decreased muscle tone (hypotonia).
* Severe muscle weakness.
* Severe breathing issues.

Symptoms of SMA type 1 include:

* Limited head control.
* Hypotonia.
* Lack of reflexes (areflexia).
* Inability to sit without support.
* Abnormal breathing pattern and a bell-shaped chest.
* Difficulty swallowing, which can lead to growth issues (failure to thrive).
* Facial muscle weakness. This develops later on in the condition.

Symptoms of SMA type 2 include:

* Hypotonia and areflexia.
* Progressive muscle weakness that affects your child’s legs more than their arms.
* Scoliosis.
* Muscle weakness in your child’s chest, which can result in restrictive lung disease.
* Jerky movements in your child’s hands (polyminimyoclonus).
* Stiffening of your child’s jaw (mandible) joint due to bone fusion (ankylosis).
* Joint contractures.

Symptoms of SMA type 3 mainly involve progressive muscle weakness that affects your child’s legs more than their arms.

The main symptom of SMA type 4 is mild leg weakness.

## **Diagnosis and Tests**

To start the diagnosis process, a healthcare provider will ask about your or your child’s medical history and symptoms. They’ll then do physical and neurological exams.

If your provider suspects SMA, the main test that can confirm it is genetic testing. This blood test can confirm 95% of SMA cases by identifying problems with the *SMN1* gene. Each of the 50 U.S. states now routinely screens newborns for SMA.

Some SMA symptoms resemble those of other neuromuscular disorders, like muscular dystrophy. If your provider doesn’t immediately suspect SMA, they may recommend any of the following tests to find the cause:

* **Creatine kinase blood test**: Deteriorating muscles release this enzyme into your bloodstream. But the levels are typically normal with SMA, unlike with other neuromuscular disorders.
* **Electromyogram (EMG) and nerve conduction study**: These tests measure the electrical activity of your muscles and nerves.
* **Muscle biopsy**: Rarely, your provider may request a muscle biopsy. This procedure involves removing a small amount of your muscle tissue and sending it to a lab for examination.

#### **Can spinal muscular atrophy be diagnosed during pregnancy?**

If you’re pregnant and have a family history of SMA, prenatal genetic testing may be able to check if the developing fetus has the condition. Prenatal tests for SMA include:

* **Amniocentesis**: During amniocentesis, your healthcare provider inserts a thin needle into your belly to draw out a small amount of amniotic fluid. A pathologist checks the sample for SMA. This test happens after the 14th week of pregnancy.
* **Chorionic villus sampling (CVS)**: Your provider removes a small tissue sample from the placenta through your cervix or belly. A pathologist checks the sample for SMA. CVS can take place as early as the 10th week of pregnancy.

## **Management and Treatment**

Unfortunately, there isn’t a cure for SMA. Treatment for SMA mainly seeks to manage symptoms and prevent complications. Symptom management therapies may include:

* Physical therapy, which can help improve posture, prevent joint immobility and slow muscle weakness.
* Occupational therapy, which can improve your ability to perform daily tasks.
* Assistive devices, like orthopedic braces, crutches, walkers and wheelchairs.
* Therapy for speech and swallowing difficulties.
* A feeding tube if swallowing is too difficult and/or dangerous.
* Assisted ventilation for breathing issues.

#### **Medications for SMA**

Between 2016 and 2020, the U.S. Food and Drug Administration (FDA) approved treatments that can significantly improve the course of SMA. They include:

* **Disease-modifying therapy**: These medications stimulate the production of SMN2 protein. The FDA has approved Nusinersen (Spinraza®) for both children and adults. A healthcare provider injects the medication into the space around your spinal canal. A different medication, risdiplam (Evrysdi®), helps those with SMA who are 2 months and older. You take risdiplam daily by mouth (orally).
* **Gene replacement therapy**: Children younger than 2 may benefit from a one-time intravenous (IV) infusion of a medication called onasemnogene abeparvovec-xioi (Zolgensma®). This therapy replaces a missing or faulty *SMN1* gene with a functioning gene.

These newer treatments may be particularly effective if started early, even before symptoms of SMA appear. Given the availability of these treatments, there’s now routine screening of newborns for SMA in the United States.

Your child may also be able to participate in a clinical trial for SMA. Talk with your child’s healthcare team to see if this is an option.

## **Spinal Muscular Atrophy (SMA) Treatment Drugs and Their Side Effects**

## 1. Spinraza (Nusinersen)

* Type: Antisense oligonucleotide
* Administration: Intrathecal injection (into the spinal canal)
* Mechanism: Modifies splicing of SMN2 gene to increase production of functional SMN protein
* Common Side Effects:
  + Headache
  + Back pain
  + Respiratory infections
  + Constipation
  + Risk of thrombocytopenia and renal toxicity (rare)
* Notes: Requires repeated lumbar punctures; well-established safety profile.

## 2. Evrysdi (Risdiplam)

* Type: Oral SMN2 splicing modifier
* Administration: Oral liquid or tablet form (FDA approved tablet form in 2025)
* Mechanism: Increases SMN protein production by modifying SMN2 gene splicing
* Common Side Effects:
  + Fever
  + Diarrhea
  + Rash
  + Mouth ulcers
  + Upper respiratory tract infections
* Notes: Non-invasive, daily dosing; tablet form improves accessibility and convenience.

## 3. Zolgensma (Onasemnogene abeparvovec-xioi)

* Type: Gene replacement therapy
* Administration: Single intravenous infusion
* Mechanism: Delivers functional copy of SMN1 gene via viral vector
* Common Side Effects:
  + Elevated liver enzymes (hepatotoxicity risk)
  + Vomiting
  + Thrombocytopenia
  + Fever
  + Respiratory infections
* Notes: One-time treatment; requires monitoring of liver function and platelet counts.

## 4. Emerging Therapy: Apitegromab (SRK-015)

* Status: Investigational; FDA priority review expected in September 2025
* Mechanism: Myostatin inhibitor that prevents muscle wasting by blocking myostatin activation
* Potential Side Effects: Under investigation; expected to be well tolerated based on trials.

## **Genomic Data of Spinal Muscular Atrophy (SMA)**

* Causative Gene:  
  SMA is primarily caused by biallelic mutations or deletions in the *SMN1* gene located on chromosome 5q13. This gene encodes the survival motor neuron (SMN) protein essential for motor neuron health.
* Gene Structure and Homology:  
  The *SMN1* gene is highly homologous to a nearly identical paralog, *SMN2*, which differs by a few nucleotides. The number of *SMN2* copies varies between individuals and modulates disease severity because *SMN2* produces some functional SMN protein but less efficiently than *SMN1*.
* Genetic Variations:
  + Most SMA patients have homozygous deletion of *SMN1* (zero copies), which is diagnostic.
  + Some patients have compound heterozygous mutations including deletions and single-nucleotide variants (SNVs) in *SMN1* exons or intron/exon boundaries.
  + Rare cases involve deep intronic mutations or complex rearrangements that are harder to detect.
  + Carrier testing is complicated by the presence of silent carriers who have two copies of *SMN1* on one chromosome (2+0 genotype), making standard copy number tests insufficient alone.
* Copy Number Variation (CNV):
  + The copy number of *SMN1* is critical for diagnosis and carrier detection; zero copies indicate SMA, one copy indicates carrier status.
  + The copy number of *SMN2* influences disease severity; more copies generally correlate with milder phenotypes.
* Diagnostic Techniques:
  + Copy number analysis of *SMN1* and *SMN2* using PCR-based methods or genome sequencing (GS) with specialized algorithms to distinguish *SMN1* from *SMN2*.
  + Long-read sequencing and next-generation sequencing (NGS) enable detailed profiling of *SMN1* and *SMN2* including SNVs and structural variants.
  + Advanced methods combine microsatellite marker analysis, long-read sequencing, and de novo assembly to resolve complex deletions and improve carrier detection.

## **Outlook / Prognosis**

The prognosis (outlook) for someone with SMA varies based on the subtype. Your healthcare team can give you a better idea of what to expect based on your or your child’s situation.

#### **Complications of SMA**

Over time, children with SMA experience progressive muscle weakness and loss of muscle control. Potential complications include:

* Bone fractures, hip dislocation and scoliosis.
* Malnutrition and dehydration due to problems eating and swallowing.
* Chest infections, like aspiration pneumonia, due to swallowing issues.
* Weak lungs and breathing problems that may require breathing support (ventilation).

People with SMA are also prone to metabolic acidosis, especially during periods of illness or fasting. Researchers aren’t sure why this happens.

#### **What is the life expectancy of someone with SMA?**

The life expectancy of someone with SMA largely depends on the type:

* SMA type 0 results in death at birth or within one month of life.
* SMA type 1 often results in death by the age of 2 without breathing support.
* Life expectancy for SMA type 2 varies but is usually between 20 and 40 years.
* SMA types 3 and 4 typically don’t affect life expectancy.

It’s important to note that disease-modifying and gene replacement therapies have been proven to substantially improve survival in SMA type 1. Your healthcare team will be able to give you a better idea of what to expect.

## **Prevention**

SMA is an inherited condition, so it’s not typically preventable. But genetic testing can help you understand your odds of having a biological child with SMA.

Carrier testing for SMA is available using a genetic test. If you or your partner carry the mutated gene that causes SMA, a genetic counselor can explain the chances of your child having SMA or being a carrier.

You may be able to take steps before pregnancy to lower the risk of passing on SMA. A process called preimplantation genetic diagnosis (PGD) identifies embryos that don’t have the mutated gene. Your healthcare provider implants healthy embryos during in vitro fertilization (IVF).

**Living With**

If your child has SMA, it’s important to advocate for them to ensure they get the best medical care and as much access to therapy as possible. Advocating for care can help them have the best possible quality of life.

You and your family may also want to consider joining a support group to meet others who can relate to your experiences.

### **PREDEFINED Questions and answers**

## 1. What type of SMA do I or my child have?

Spinal Muscular Atrophy (SMA) is typically classified into five main types (0, 1, 2, 3, and 4) based on the age of symptom onset and severity. All these types are primarily caused by mutations in the *SMN1* gene, with the number of copies of the *SMN2* gene influencing the severity.

* SMA Type 0: This is a very rare and severe type where symptoms begin before birth. Infants have severe weakness, and significant difficulty breathing and feeding. Life expectancy is very short, often resulting in death at birth or within one month.
* SMA Type 1 (Werdnig-Hoffman disease): This is the most common and severe form, with symptoms appearing at birth or by 6 months of age. Infants experience severe muscle weakness, trouble breathing, coughing, and swallowing. Without breathing support, type 1 often results in death by age 2.
* SMA Type 2 (Intermediate SMA/Dubowitz disease): Symptoms typically begin between 6 and 18 months of age. Children with SMA Type 2 can usually sit without support but cannot stand or walk independently. Muscle weakness is mainly in the proximal muscles, affecting lower limbs more than upper limbs, while face and eye muscles are usually unaffected. Life expectancy for Type 2 varies but is often between 20 and 40 years.
* SMA Type 3 (Kugelberg-Welander disease): Symptoms appear after 18 months of age and can range from 18 months to adulthood. Individuals can walk independently but may experience difficulty running, climbing stairs, and frequent falls due to proximal weakness. Many may eventually require a wheelchair. Life expectancy is typically comparable to that of the general population.
* SMA Type 4: This is the mildest and latest-onset form, usually appearing after age 30, though sometimes as early as 18. Symptoms include mild to moderate leg muscle weakness. Patients can achieve motor milestones and maintain mobility throughout life, and their lifespan remains normal.

Your healthcare provider will determine the specific type through clinical evaluation and genetic testing.

## 2. What's the prognosis for this type of SMA?

The prognosis for SMA varies significantly by type, primarily depending on the severity and age of onset:

* SMA Type 0: Extremely poor, with death usually occurring at birth or within the first month.
* SMA Type 1: Without advanced breathing support, life expectancy is often less than two years.
* SMA Type 2: Life expectancy is variable, often ranging from 20 to 40 years.
* SMA Type 3: Typically, life expectancy is normal, although individuals may experience progressive muscle weakness and may eventually need a wheelchair.
* SMA Type 4: Life expectancy is generally normal, as it is the mildest form.

It is important to note that with recent advancements in treatment, the prognosis for all types, especially Types 1 and 2, has improved significantly.

## 3. What's the best treatment for this type of SMA?

The best treatment for SMA is disease-modifying therapy aimed at increasing the production of the SMN protein, which is deficient in all SMA types. The choice of treatment depends on the specific type of SMA, age, and other factors, and your doctor will recommend the most suitable option:

* Nusinersen (Spinraza): An antisense oligonucleotide that increases SMN protein production. It is administered via intrathecal injection (into the spinal canal).
* Onasemnogene abeparvovec (Zolgensma): A gene replacement therapy that delivers a functional copy of the *SMN1* gene. It is a one-time intravenous infusion [Search Result 9].
* Risdiplam (Evrysdi): An oral medication that also increases SMN protein levels by modifying gene splicing. It is taken daily [Search Result 9].

In addition to these medications, supportive care is crucial for all SMA types, including physical therapy, occupational therapy, respiratory support, and nutritional management to address specific symptoms and improve quality of life

## 4. What are the treatment risks and side effects?

Each treatment has specific risks and side effects:

* Nusinersen (Spinraza): Common side effects include headache, back pain, and respiratory infections. Rare but serious risks can include thrombocytopenia (low platelet count) and kidney toxicity. It also requires repeated lumbar punctures.
* Onasemnogene abeparvovec (Zolgensma): Common side effects include elevated liver enzymes (indicating liver injury), vomiting, and thrombocytopenia. Liver function must be closely monitored before and after treatment.
* Risdiplam (Evrysdi): Common side effects include fever, diarrhea, rash, and mouth ulcers. It can also cause upper respiratory tract infections.

Your healthcare provider will discuss these risks in detail and explain how they will be monitored throughout treatment.

## 5. Are other members of my family at risk for getting SMA? If so, should we get genetic tests?

Yes, SMA is a genetic condition typically inherited in an autosomal recessive pattern, meaning that an individual usually inherits two mutated copies of the *SMN1* gene (one from each parent) to develop the condition.

* If you have a child with SMA, both biological parents are typically carriers (meaning they each have one healthy copy and one mutated copy of the *SMN1* gene). Each child they have together would then have a 25% chance of having SMA, a 50% chance of being a carrier, and a 25% chance of being neither affected nor a carrier.
* Other family members, such as siblings of an affected individual, may also be carriers.

Genetic testing for carrier status is highly recommended for family members who are considering having children, including parents, siblings, and extended family members, to understand their risk. Genetic counseling can help interpret results and explain family risks.

## 6. What type of ongoing care will I or my child need?

Ongoing care for SMA is multidisciplinary and focuses on managing symptoms, preventing complications, and supporting overall well-being:

* Neurological follow-ups: Regular visits with a neurologist to monitor disease progression and adjust treatment.
* Physical and occupational therapy: To maintain muscle strength, flexibility, prevent contractures, and improve daily function.
* Respiratory care: Monitoring breathing function, and providing respiratory support (e.g., non-invasive ventilation, airway clearance techniques) as needed to prevent and manage respiratory infections.
* Nutritional support: Monitoring weight and growth and addressing swallowing difficulties (dysphagia) or feeding issues, potentially through feeding tubes.
* Orthopedic care: Monitoring for scoliosis (curvature of the spine) and joint contractures and considering interventions like bracing or surgery if necessary.
* Speech and language therapy: For those with swallowing or communication difficulties.
* Psychological and social support: For patients and families to cope with the challenges of SMA.

## 7. What signs of complications should I look out for?

You should be vigilant for signs of complications, as early detection allows for timely intervention:

* Respiratory distress: Increased difficulty breathing, rapid breathing, use of accessory muscles for breathing, blue discoloration around the lips or fingertips, or frequent respiratory infections.
* Feeding difficulties: Choking, gagging, recurrent aspiration, or significant weight loss.
* Scoliosis: Noticeable curvature of the spine, which can worsen breathing problems.
* Joint contractures: Joints becoming stiff or difficult to move through their full range of motion.
* Signs of medication side effects: Such as persistent fever, vomiting, or changes in urine color (if on Zolgensma, indicating liver issues), or severe headache/back pain (if on Nusinersen).
* Sudden worsening of weakness: Any rapid decline in muscle function or motor skills

## **Epidemiology**

### Frequency

United States

The spinal muscular atrophies are the second most common autosomal-recessive inherited disorders after cystic fibrosis. The acute infantile-onset SMA (type I) affects approximately 1 per 10,000 live births; the chronic forms (types II and III) affect 1 per 24,000 births. SMA types I and III each account for about one fourth of cases, whereas SMA type II is the largest group and accounts for one half of all cases.

International

The incidence of spinal muscular atrophy is about 1 in 10,000 live births with a carrier frequency of approximately 1 in 50.

### Mortality/Morbidity

The mortality and/or morbidity rates of spinal muscular atrophy are inversely correlated with the age at onset. High death rates are associated with early onset disease. In patients with SMA type I, the median survival is 7 months, with a mortality rate of 95% by age 18 months.

* Respiratory infections account for most deaths.
* In type II SMA, the age of death varies, but death is most often due to respiratory complications.
* See Prognosis for more information.

### Sex

Male individuals are most frequently affected, especially with the early-onset forms of spinal muscular atrophy, ie, types I and II.

### Age

According to the ISMAC system, the age of onset for spinal muscular atrophies is as follows:

* SMA type I (acute infantile or Werdnig Hoffman): Onset is from birth to 6 months.
* SMA type II (chronic infantile): Onset is between 6 and 18 months.
* SMA type III (chronic juvenile): Onset is after 18 months.
* SMA type IV (adult onset): Onset is in adulthood (mean onset, mid 30s).

## **SMA Differential Diagnosis**

* Congenital Muscular Dystrophy
* Congenital Myopathies
* Duchenne and Becker Muscular Dystrophies
* Juvenile Myasthenia Gravis
* Amyotrophic Lateral Sclerosis (ALS) (especially for adult-onset SMA type 4)
* Guillain-Barré Syndrome
* Transverse Myelitis
* Pompe Disease (Glycogen Storage Disease type II)
* Charcot-Marie-Tooth Disease
* Prader-Willi Syndrome
* Congenital Myasthenic Syndromes
* Fazio-Londe Syndrome
* Monomelic Amyotrophy
* Cerebral Palsy (hypotonic diplegia)
* Progressive Muscular Atrophy

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand you have some concerns about your child’s muscle weakness. Can you tell me more about what you’ve noticed?

Parent: Yes, my baby seems very floppy and has trouble holding up their head. They also don’t move as much as other babies their age.

Doctor: Thank you for sharing that. Based on the symptoms and initial tests, it looks like your child may have a condition called Spinal Muscular Atrophy, or SMA. It’s a genetic disorder that affects the motor neurons, which are the nerve cells that control muscle movement.

Parent: Is this something serious? What causes it?

Doctor: SMA can vary in severity, but it often causes progressive muscle weakness. It is caused by changes in a gene called *SMN1*, which leads to a shortage of a protein necessary for motor neuron survival. Because of this, muscles become weaker over time.

Parent: How do you confirm this diagnosis?

Doctor: We confirm SMA by doing a genetic test that looks for mutations or deletions in the *SMN1* gene. This test is very accurate and can tell us the exact type of SMA your child has.

Parent: What treatments are available? Can it be cured?

Doctor: While there is currently no cure, there are effective treatments that can slow the progression of the disease and improve muscle function. These include medications like Nusinersen (Spinraza), Risdiplam (Evrysdi), and gene therapy (Zolgensma). We also focus on supportive care such as physical therapy and respiratory support.

Parent: Are there any risks or side effects with these treatments?

Doctor: Yes, each treatment has potential side effects. For example, Spinraza requires injections into the spinal canal and can cause headaches or back pain. Gene therapy can affect liver function, so we monitor that closely. We will discuss these in detail and monitor your child carefully during treatment.

Parent: What about other family members? Could they have SMA?

Doctor: SMA is inherited in an autosomal recessive pattern, which means both parents usually carry one copy of the mutated gene. We recommend genetic testing for family members to understand their carrier status and discuss family planning options.

Parent: What kind of ongoing care will my child need?

Doctor: Your child will need regular follow-ups with a multidisciplinary team including neurologists, physical therapists, respiratory specialists, and nutritionists. We’ll monitor muscle strength, breathing, and nutrition closely to manage symptoms and prevent complications.

Parent: What signs should I watch for that mean my child’s condition is getting worse?

Doctor: Watch for increased difficulty breathing, swallowing problems, persistent cough, or changes in muscle strength. If you notice any of these, please contact us immediately.

Parent: Thank you, doctor. This helps me understand what to expect and how we can help our child.

Doctor: You’re very welcome. We’re here to support you every step of the way. Please don’t hesitate to reach out with any questions or concerns.

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## **Sleep disordered breathing**

**DEFINITION AND DESCRIPTION**

Sleep disordered breathing (SDB) refers to conditions with breathing disturbances during sleep. These include increased airway resistance, heavy snoring, hypopnea (impaired airflow), or apnea (complete cessation of breathing). The severity of SDB can range from mild airway resistance to severe apnea episodes. These issues affect sleep quality and overall health. Timely recognition and treatment are crucial, as untreated cases can lead to complications like cardiovascular problems and fatigue.

What is a sleep disordered breathing child?

The term breathing disorders during sleep in children refers to a group of diseases or conditions which involve a disorder in breathing during sleep, from a simple recurring noise to complex obstructive apnea. This disorder is often related to factors such as hypertrophied tonsils or adenoids, obesity, and facial structural abnormalities. Some of the symptoms of SDB include severe snoring, restless sleep, airway obstruction during daytime, mouth breathing, nocturnal enuresis, excessive daytime sleepiness and poor concentration. The assessment usually consists of polysomnography and treatment techniques can range from the removal of diseased adenoids in the throat, dietary control in overweight patients or finally continuous positive airway pressure in very severe conditions. It is important to remember that timely initiation of treatments is crucial in terms of prevention of stunted development and impaired intellectual abilities.

## **Types of sleep disordered breathing**

Among the different sleep disorders, each has its distinct characteristics and health effects.

### Snoring

* **Characteristics**: Snoring is the sound produced by airflow through the nose and throat being partially blocked. Causing vibrations in surrounding tissues.
* **Causes**: It can be caused by several things including nasal congestion, obesity, alcohol, and positions taken while sleeping.
* **Impact on Health**: Snoring is mostly not a serious threat. However, it may disrupt sleep to the great discomfort of both the snorer and the snooze partner leading to excessive daytime fatigue and irritability.

### Upper Airway Resistance Syndrome (UARS)

* **Characteristics**: UARS is one in which the upper airway collapses somewhat during sleep, increasing respiratory efforts.
* **Causes**: Anatomical factors may play a part in UARS such as jaw size and enlarged tonsils.
* **Impact on Health**: These can cause frequent awakenings at night, excessive daytime sleepiness, and if not treated, can go on to develop more severe forms of sleep disorder.

### Obstructive Sleep Apnea (OSA)

* **Characteristics**: OSA is when the airway becomes partially blocked or totally obstructed during sleep, leading to increased respiratory efforts.
* **Causes**: Causes can basically be obesity, relaxation of throat muscles, enlarged tonsils, and jaws being set in an unusual position.
* **Impact on Health**: Apnea can lead to life-threatening situations such as high blood pressure, heart disease, stroke, and diabetes because of frequent drops in blood oxygen levels and consequent frequent wakeups during sleep.

### Central Sleep Apnea (CSA)

* **Characteristics**: CSA is a rarer form of Sleep Apnea where the brain does not send the right signals with awesome rhythm.
* **Causes**: Certain medical conditions that include heart failure, stroke, and any particular medication may be taken.
* **Impact on Health**: It can cause sleep disruption, fatigue, and in extreme cases, worsen cardiovascular problems and promote decline in cognitive activities.

### Sleep-Related Hypoventilation Disorders

* **Characteristics**: In most cases, these disorders involve decreased breathing and shallow breaths during sleep, resulting in high carbon dioxide levels in the blood.
* **Causes**: Obesity hypoventilation syndrome (OHS), chronic lung disease, and neuromuscular disorders may all cause Hypoventilation.
* **Impact on Health**: People with Hypoventilation may suffer from chronic morning headaches. They also experience excessive daytime sleepiness and eventually develop respiratory failure.

### Sleep-Related Hypoxemia Disorder

* **Characteristics**: This is a condition characterized by abnormally low blood oxygen levels during sleep.
* **Causes**: Persistent low oxygen levels during sleep occur when someone has lung conditions such as chronic obstructive pulmonary disease (COPD).
* **Impact on Health**: Long-term low oxygen levels strain the heart and lungs, leading to complications like pulmonary hypertension, hypertension, and arrhythmias.

## **Sleep Disordered Breathing vs. Sleep Apnea**

Sleep disordered breathing refers to a group of breathing problems during sleep. But sleep apnea only applies to conditions when breathing pauses entirely or partially in sleep cycles. There are two types of sleep apnea under SDB category: obstructive sleep apnea (OSA) and central sleep apnea (CSA).

## **How to Diagnose Sleep Disordered Breathing**

The diagnosis of sleep disordered breathing spectrum usually requires a sleep study (polysomnography). Where the patient’s breathing, oxygen levels, heart rate, and some other parameters are monitored during sleep. Home sleep apnea testing may also be used, especially for OSA diagnosis. For certain patients, the need for evaluation by a sleep specialist or imaging studies may be warranted to evaluate airway anatomy.

## **Treatment Options for Sleep Disordered Breathing**

Sleep disordered breathing treatment options depend on both the type and severity of SDB

* **CPAP Machines for OSA**: Continuous positive airway pressure (CPAP) treats OSA more frequently than any other remedy by being a system of delivering constant air pressure to keep the airway open.
* **Lifestyle Changes**: Moderate weight loss and smoking cessation can significantly improve SDB symptoms by lessening the chances of airway occlusion.
* **Surgical Options**: In some cases, surgery may be needed to remove or shrink airway tissues (such as tonsils and soft palate) for adequate airflow.
* **Dental Devices and Positional Therapy**: Oral appliances can draw the jaw and tongue forward. Positional therapy advises sleeping a certain way such as on one’s side to lessen airway obstruction.

## Initial Treatment

### Elimination of contributing factors

The first task in treating patients with sleep-disordered breathing (SDB) is to eliminate all possible contributing factors. This includes weight loss for patients who are obese and elimination of alcohol or sedative use, especially near bedtime. Benzodiazepines, narcotics, and barbiturates can worsen SDB, or sometimes they initiate it where it had not previously been present.

A 10% weight loss was associated with a 26% decrease in the respiratory disturbance index (RDI; also referred to as the apnea-hypopnea index [AHI]) in a population-based study. Weight loss should be recommended for all obese patients with sleep apnea; however, weight loss takes time, and only a minority of patients successfully maintain it.

### Alteration of body positioning during sleep

Body positioning during sleep can improve SDB in some patients. Because lying supine can allow gravity to assist in pulling lax tongue muscles back toward the posterior pharyngeal wall, patients should sleep on their sides, on their stomachs, or propped up 60°. These positions can improve SDB in patients whose symptoms occur primarily while supine.

Avoidance of supine sleeping can easily be accomplished with a sock, tennis ball, and safety pins. The tennis ball in a sock is pinned to the back of the pajamas, positioning the tennis ball between the scapulae.

Avoidance of supine sleeping can easily be accomplished with a sock, tennis ball, and safety pins. The tennis ball in a sock is pinned to the back of the pajamas, positioning the tennis ball between the scapulae. When the patient rolls into the supine position during sleep, this lump is uncomfortable enough that the position is immediately shifted, usually without the patient awakening.

### Thyroid hormone replacement therapy

In patients with hypothyroidism and SDB, thyroid hormone replacement therapy is usually accompanied by an improvement in the SDB.

### Use of oral appliances

In some individuals, a mouthpiece may improve the anatomy of the airway to the point where snoring or mild obstructive sleep apnea (OSA) can be corrected. Oral appliances, or mandibular advancement devices (MADs), can be an effective alternative for mild and medium-to-moderate OSA syndrome (OSAS), but they require strict monitoring because of differences in individual response to this therapy.

Many types of oral appliances have been designed for the treatment of sleep apnea. Most are custom fitted to the teeth of both dental arches to reposition the mandible and to enlarge the retropalatal and retrolingual airway space. However, consistent patient tolerance for this treatment is relatively low, and it is less effective than continuous positive airway pressure (CPAP) in reducing the frequency of apnea and hypopnea.

### Restriction or elimination of alcohol use

Alcohol significantly worsens SDB. Eliminating use of alcohol, especially near bedtime, improves SDB

## **Diagnostic Considerations**

The differential diagnosis of sleep-disordered breathing (SDB) includes the following:

* Simple snoring
* Central sleep apnea (CSA)
* Other disorders that cause daytime sleepiness (eg, insufficient sleep, a circadian-rhythm abnormality, narcolepsy, periodic limb movement disorder).

## **Epidemiology**

Epidemiologic studies indicate that sleep apnea is more common in men than in women (male-to-female ratio, 2-3:1). Sleep apnea occurs in 4% of men and 2% of women aged 30-60 years. A retrospective study on 830 patients with OSAS reported a male-to-female ratio (M:F) that increases with the gravity of the disease: 2.2:1 in mild OSAS and 7.9:1 in severe OSAS.Hypersomnolence is reported with a percentage of 16% in men and 22% in women; 24% of men and 9% of women have an apnea-hypopnea index of at least 5.

The discrepancy between the lower prevalence of OSA, the greater frequency of obesity, and the smaller airway size in women compared with men suggests that a gender difference underlies this condition.

Men tend to have a larger but more collapsible airway during mandibular movement than women, and this may play a partial role in the positional dependency and severity of OSA in men.

Another possible reason for the lower prevalence of OSAS may be reluctance on the part of many women to report symptoms mostly considered inappropriate, like snoring; this reluctance may cause a clinical underestimation of the problem in females.

The gender-related protective effect decreases in females who are postmenopausal and not on hormone replacement therapy.

The association between age and OSA is complex. Several studies have shown a higher prevalence of OSA in elderly persons than in middle-aged persons, though daytime symptoms may be less common with advancing age.

The Sleep Heart Health Study demonstrated that the influence of male sex and body mass index (BMI) on OSA tends to wane with age. For unclear reasons, the overall prevalence of OSA plateaus after age 65 years.

The prevalence of OSAS among African American persons seems to be at least equal to and possibly greater than that among white persons. The prevalence among men in urban India and men and women in Korea is similar to that observed in Western countries. Some researchers have noticed an increased incidence of OSA in persons of Asian origin.

## **Prognosis**

Excessive daytime sleepiness resulting from SDB can impact focus and concentration, causing decreased work effectiveness. Even mild-to-moderate SDB lengthens reaction time, causing performance decreases similar to alcohol intoxication. This can lead to motor vehicle accidents and other serious accidents in situations where alertness is required for safety (e.g., heavy machinery operators).

Moderate-to-severe OSA is associated with earlier death. The cardiovascular sequelae of untreated OSA include hypertension, cor pulmonale, arrhythmias, and increased risk of myocardial infarction or stroke.SDB is associated with higher levels of IL-6, a marker of myocardial infarction risk and mortality.Adiposity may mediate the increased levels of C-reactive protein (CRP), fibrinogen, intercellular adhesion molecule (ICAM)-1, and P-selectin observed in SDB.

OSA is associated with difficult-to-control hypertension.CPAP also reduces markers of hypercoagulability, and this is a potential mechanism by which it can reduce the rate of cardiovascular morbidity and mortality in OSAS patients.

In heart failure patients with sleep apnea, studies have not shown the use of PAP to reduce the risks of cardiovascular outcomes or death; however, such therapy has been associated with some improvements in OSA symptoms.

Treatment of OSA may reduce new first-time cerebrovascular events and recurrences. A study by Gupta et al suggested that in patients with stroke and OSA, CPAP treatment can yield significantly better stroke outcomes and statistically nonsignificant favorable outcomes in terms of recurrence of vascular events.

Untreated OSA has been associated with cognitive deficits and changes in brain electrophysiology, and there is evidence to suggest that CPAP may mitigate these effects.

Many of the studies examining the relation between OSA and glucose tolerance have shown a direct and independent relation between OSA and diabetes. The Wisconsin Sleep Study Cohort showed a greater prevalence of diabetes in subjects with increasing levels of OSA. Several studies have shown a beneficial effect of CPAP therapy on insulin resistance or glucose levels.

The probable mechanisms connecting OSA with glucose tolerance and type 2 diabetes mellitus include increased sympathetic activity, sympathovagal dysfunction, alterations in neuroendocrine function (especially in growth hormone [GH] and cortisol levels), and a high inflammatory state with an increase in the release of proinflammatory cytokines

## **Predefined Questions and Answers for Sleep Disordered Breathing**

1. What is sleep disordered breathing?  
Sleep disordered breathing refers to abnormal breathing patterns during sleep, including snoring, pauses in breathing (apneas), and shallow breathing (hypopneas) that can disrupt sleep quality and oxygen levels.

2. What are common symptoms of sleep disordered breathing?

* Loud and frequent snoring
* Pauses in breathing observed by a bed partner
* Gasping or choking during sleep
* Excessive daytime sleepiness or fatigue
* Morning headaches
* Difficulty concentrating or memory problems
* Restless sleep or frequent awakenings

3. How is sleep disordered breathing diagnosed?  
Diagnosis often involves:

* A detailed sleep history, including questions about snoring, breathing pauses, and daytime symptoms
* Sleep questionnaires such as the Berlin Questionnaire or STOP-Bang to assess risk
* Overnight sleep studies (polysomnography) either in a sleep lab or at home to monitor breathing, oxygen levels, and brain activity during sleep

4. What questions might my doctor ask to assess for sleep disordered breathing?

* Do you snore loudly?
* Has anyone noticed that you stop breathing or gasp during sleep?
* Do you wake up feeling short of breath or choking?
* Do you feel excessively sleepy during the day?
* Do you have high blood pressure or other cardiovascular conditions?
* What is your typical sleep schedule and sleep environment like?

5. What are the risk factors for sleep disordered breathing?

* Obesity
* Older age
* Male sex
* Anatomical factors such as enlarged tonsils, a small jaw, or nasal obstruction
* Smoking and alcohol use
* Medical conditions like hypertension, heart failure, or stroke

6. What treatments are available for sleep disordered breathing?

* Lifestyle changes: weight loss, avoiding alcohol and sedatives before bedtime, changing sleep position
* Continuous positive airway pressure (CPAP) therapy to keep airways open during sleep
* Oral appliances to reposition the jaw or tongue
* Surgery to remove or reduce airway obstruction in selected cases
* Treatment of underlying medical conditions

7. Can sleep disordered breathing cause other health problems?  
Yes, untreated sleep disordered breathing can lead to:

* High blood pressure
* Heart disease and stroke
* Diabetes
* Daytime fatigue increasing risk of accidents
* Cognitive impairment and mood disorders

**Doctor-patient conversation about sleep disordered breathing (SDB)**

Doctor: Hello, I understand you’ve been having some trouble with your sleep. Can you tell me what symptoms you’ve noticed?

Patient: Yes, I’ve been snoring loudly, and my partner says I sometimes stop breathing during the night. I also feel very tired during the day, even after a full night’s sleep.

Doctor: Thank you for sharing that. Loud snoring and pauses in breathing are common signs of a condition called obstructive sleep apnea, or OSA. This means your airway may be partially or completely blocked during sleep, causing you to stop breathing briefly.

Patient: That sounds serious. How do you confirm if I have sleep apnea?

Doctor: The best way to diagnose sleep apnea is through a sleep study, called a polysomnography. It monitors your breathing, oxygen levels, heart rate, and brain activity while you sleep. We can arrange this either at a sleep center or sometimes with a home sleep test.

Patient: What happens if I do have sleep apnea? How is it treated?

Doctor: Treatment depends on the severity, but the most common and effective treatment is a CPAP machine. It delivers continuous positive airway pressure to keep your airway open during sleep. There are also other options like oral appliances or surgery in some cases.

Patient: Are there lifestyle changes I can make to help?

Doctor: Absolutely. Losing weight if you’re overweight, avoiding alcohol and sedatives before bedtime, and sleeping on your side instead of your back can all help reduce symptoms.

Patient: What are the risks if I don’t treat sleep apnea?

Doctor: Untreated sleep apnea can increase your risk of high blood pressure, heart disease, stroke, diabetes, and daytime accidents due to sleepiness. It can also affect your mood and concentration.

Patient: I’d like to get tested. What are the next steps?

Doctor: I’ll refer you for a sleep study. Meanwhile, try to keep a sleep diary noting your sleep patterns and symptoms. After the test, we’ll review the results and discuss the best treatment plan for you.

Patient: Thank you, doctor. I feel better knowing there’s a way to find out what’s going on and get help.

Doctor: You’re welcome. Please don’t hesitate to contact me if you have any questions or concerns before your sleep study.

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**CENTRAL SLEEP APNEA**

**DEFINITION AND DESCRIPTION**

Central sleep apnea is a disorder in which breathing repeatedly stops and starts during sleep.

Central sleep apnea occurs because the brain doesn't send proper signals to the muscles that control breathing. This condition is different from obstructive sleep apnea, in which breathing stops because the throat muscles relax and block the airway. Central sleep apnea is less common than obstructive sleep apnea.

Central sleep apnea can result from other conditions, such as heart failure and stroke. Another possible cause is sleeping at a high altitude.

Treatments for central sleep apnea might involve managing existing conditions, using a device to assist breathing or using supplemental oxygen.

**Causes**

Central sleep apnea occurs when your brain doesn't transmit signals to your breathing muscles.

The brainstem links the brain to the spinal cord. It controls many functions, including heart rate and breathing. Central sleep apnea can be caused by a number of conditions that affect the ability of the brainstem to control breathing.

The cause varies with the type of central sleep apnea you have. Types include:

* **Cheyne-Stokes breathing.** This type of central sleep apnea is most commonly associated with congestive heart failure or stroke.

During Cheyne-Stokes breathing, breathing effort and airflow gradually rise and then lessen. During the weakest breathing effort, a total lack of airflow can occur.

* **Drug-induced apnea.** Taking certain medicines such as opioids can cause breathing to become irregular or stop completely for a short time. These medicines can be taken by mouth or by shot, also called injection. They include morphine (MS Contin, Mitigo, others), oxycodone (Roxicodone, Oxycontin, others) and codeine.
* **High-altitude periodic breathing.** A Cheyne-Stokes breathing pattern can occur if you're at a very high altitude. The change in oxygen at a high altitude can cause fast breathing, known as hyperventilation, followed by taking in too little air.
* **Treatment-emergent central sleep apnea.** Some people with obstructive sleep apnea develop central sleep apnea while using continuous positive airway pressure (CPAP) for treatment. This condition is known as treatment-emergent central sleep apnea. It is a combination of obstructive and central sleep apneas.
* **Medical condition-induced central sleep apnea.** Several medical conditions, including end-stage kidney disease and stroke, may lead to central sleep apnea. This type of sleep apnea doesn't involve Cheyne-Stokes breathing.
* **Primary central sleep apnea, also known as idiopathic sleep apnea.** The cause of this uncommon type of central sleep apnea is not known.

**Risk factors**

Certain factors put you at increased risk of central sleep apnea:

* **Sex.** Males are more likely to develop central sleep apnea than are females.
* **Age.** Central sleep apnea is more common among older adults, especially those older than age 60. This could be because people older than 60 are likely to have other medical conditions or sleep patterns that are linked to central sleep apnea.
* **Heart disorders.** Heart problems put people at higher risk of central sleep apnea. An irregular heartbeat, known as atrial fibrillation, can increase the risk. Having heart muscles that don't pump enough blood for the body's needs, known as congestive heart failure, also can raise the risk.
* **Stroke, brain tumor or a structural problem with the brainstem.** These brain conditions can affect the brain's ability to regulate breathing.
* **High altitude.** Sleeping at an altitude higher than you're used to may increase your risk of sleep apnea. High-altitude sleep apnea resolves a few weeks after returning to a lower altitude.
* **Opioid use.** Opioid medicines may increase the risk of central sleep apnea.
* **CPAP.** Some people with obstructive sleep apnea develop central sleep apnea while using continuous positive airway pressure (CPAP). This condition is known as treatment-emergent central sleep apnea. It is a combination of obstructive and central sleep apneas.

For some people, complex sleep apnea goes away with continued use of their CPAP device. Other people may be treated with a different kind of positive airway pressure therapy.

**Symptoms**

Common symptoms of central sleep apnea include:

* Observed episodes of not breathing during sleep.
* Sudden awakenings with shortness of breath.
* Not being able to stay asleep, known as insomnia.
* Excessive daytime sleepiness, known as hypersomnia.
* Trouble focusing.
* Mood changes.
* Morning headaches.
* Snoring.

Although snoring suggests some degree of a blocked airway, snoring also can occur in people with central sleep apnea. However, snoring may not be as prominent with central sleep apnea as it is with obstructive sleep apnea.

### **When to see a doctor**

Consult a medical professional if you have — or if your partner notices — any symptoms of central sleep apnea, particularly:

* Shortness of breath that awakens you from sleep.
* Pauses in your breathing during sleep.
* Trouble staying asleep.
* Excessive daytime drowsiness, which may cause you to fall asleep while you're working, watching television or even driving.

Ask a member of your health care team about any sleep problem that leaves you regularly fatigued, sleepy and irritable. Excessive daytime drowsiness can be due to other disorders, so it's important to get an accurate diagnosis. Sleepiness during the day may be caused by obstructive sleep apnea, by not allowing yourself time to get enough sleep at night or by sudden attacks of sleep, known as narcolepsy.

## **Diagnosis**

A primary care professional might evaluate your condition based on your symptoms. Or you may be referred to a sleep specialist in a sleep disorder center.

A sleep specialist can help you decide on your need for further evaluation. That might involve overnight monitoring of your breathing and other body functions during a sleep study called polysomnography.

During polysomnography, you're connected to equipment that monitors your heart, lung and brain activity, breathing patterns, arm and leg movements, and blood oxygen levels while you sleep. You may have a full-night or split-night sleep study.

In a split-night sleep study, you're monitored during the first half of the night. If you're diagnosed with central sleep apnea, staff might wake you to start a therapy for the second half of the night. The therapy might be positive airway pressure or supplemental oxygen.

Polysomnography can help diagnose central sleep apnea. It also can help rule out other sleep disorders, such as obstructive sleep apnea, repetitive movements during sleep or narcolepsy. These other disorders can cause excessive daytime sleepiness but require different treatment.

Doctors trained in nervous system diseases, known as neurologists, and in heart diseases, known as cardiologists, and others might be involved in evaluating your condition. You might need imaging of your head or heart to look for contributing conditions.

**TREATMENT**

Treatments for central sleep apnea might include:

* **Addressing associated medical problems.** Possible causes of central sleep apnea include other disorders. Treating those conditions might help your central sleep apnea. For example, therapy for heart failure might improve central sleep apnea.
* **Reduction of opioid medicines.** If opioid medicines are causing your central sleep apnea, your health care team might reduce your dose of those medicines over time.
* **Continuous positive airway pressure (CPAP).** This method, also used to treat obstructive sleep apnea, involves wearing a mask over the nose or over the nose and mouth while asleep.

The mask is attached to a small pump that supplies a continuous amount of pressurized air to hold open the upper airway. CPAP may prevent the airway closure that can trigger central sleep apnea.

As with obstructive sleep apnea, in central sleep apnea it's important that you use the CPAP device only as directed. If your mask is uncomfortable or the pressure feels too strong, talk with your health care team. Several types of masks are available. The air pressure also can be adjusted.

* **Adaptive servo-ventilation (ASV).** If CPAP doesn't effectively treat your condition, you might be given ASV. Like CPAP, ASV also delivers pressurized air.

Unlike CPAP, ASV adjusts the amount of pressure breath-by-breath when you take a breath. This smooths out your breathing pattern. The device also might automatically deliver a breath if you haven't taken a breath within a certain number of seconds.

ASV isn't recommended for people with symptomatic heart failure.

* **Bilevel positive airway pressure (BPAP).** Like ASV, BPAP delivers a set amount pressure when you breathe in and a different amount of pressure when you breathe out. Unlike ASV, the amount of pressure delivered when you breathe in is fixed rather than variable. BPAP also can be set to deliver a breath if you haven't taken a breath within a certain number of seconds.
* **Supplemental oxygen.** Using supplemental oxygen while you sleep might help if you have central sleep apnea. Various devices are available to deliver oxygen to your lungs.
* **Medicines.** Medicines such as acetazolamide have been used to stimulate breathing in people with central sleep apnea. These medicines might be prescribed to help your breathing as you sleep if you can't tolerate positive airway pressure.

### **Surgery or other procedures**

A newer therapy for central sleep apnea is transvenous phrenic nerve stimulation. A device approved by the U.S. Food and Drug Administration known as Remede System delivers an electrical pulse to the nerve that controls the diaphragm during sleep. This causes you to take a breath. The system includes a battery-powered pulse generator that's implanted under the skin in the upper chest.

Used for moderate to severe central sleep apnea, this system produces a steady breathing pattern. More study is needed.

## **Diagnostic Considerations**

This discussion includes the differentiation of central sleep apnea from non–central sleep apnea conditions.

## Obstructive sleep apnea

This is a sleep disorder in which recurrent complete or partial upper airway obstruction produces snoring, oxygen desaturations, and numerous arousals. The repetitive upper airway collapse occurs during sleep because negative pressure generated during inspiration is not effectively counteracted by splinting by pharyngeal dilators, especially when narrowing occurs as a result of excessive soft tissue or vulnerable craniofacial anatomy. Patients may report loud snoring, witnessed apneas, and excessive daytime sleepiness. Physical examination characteristics often include a crowded oropharynx, increased neck and waist circumferences, and increased body mass index. Further, polysomnography (PSG) demonstrates prominent snoring and obstructive respiratory events (airflow is absent but ventilatory effort persists, as opposed to absent ventilatory effort in central sleep apnea

## Pseudocentral sleep apnea

Patients with diaphragmatic paralysis and other neuromuscular diseases, who are dependent on accessory muscles of breathing to maintain ventilation, may appear to have central apneas during rapid eye movement (REM) sleep. This is due to the REM atonia of skeletal muscles. Many of these patients actually have obstructive sleep apnea but do not have enough diaphragmatic excursions to be recorded by the piezoelectric belts used during routine PSG. A history of neuromuscular disease and worsening of central apneas during REM sleep should alert to the possibility of pseudocentral apnea.

## Sleep-related hypoventilation syndrome

Sleep-related hypoventilation with central sleep apneas can be observed in many conditions, such as neuromuscular weakness or chronic obstructive pulmonary disease. These conditions are characterized by a history of a preexisting disorder of hypoventilation, elevated resting PaCO2, and severe oxygen desaturation during sleep, which is more prominent during REM sleep in contrast to primary central sleep apnea and Cheyne-Stokes breathing-central sleep apnea (CSB-CSA), which are mostly observed during NREM sleep.

## **Epidemiology**

### US frequency

Predominant central apnea is uncommon and is seen in less than 10% of patients presenting for PSG. In the general population, the prevalence of central sleep apnea is less than 1%.CSB-CSA has been reported in 25-40% of patients with heart failure and in 10% of patients who have had a stroke. CSA related to chronic kidney disease has been reported as in up to 10% of patients. Although data is insufficient to establish the prevalence, CSA occurs in neurological disorders with impaired central command and/or peripheral muscle weakness including multiple-system atrophy, amyotrophic lateral sclerosis, multiple sclerosis and neuromuscular disease.

One studyhas reported the prevalence rate of central sleep apnea at 30% in a population of patients in a stable methadone maintenance program.

### Race

No data are available on the racial distribution of central sleep apnea syndromes.

### Sex

CSB-CSA shows a striking male preponderance. Sex distribution in other types of central sleep apnea syndromes has not been studied. Central sleep apnea is uncommon in premenopausal women. One explanation for this discrepancy is the presence of a lower apneic threshold of PaCO2 in women compared with men. Thus, women require a greater reduction in their PaCO2 to initiate apnea than do men.

### Age

Primary central sleep apnea mostly affects middle-aged or elderly individuals. CSB-CSA increases in prevalence among individuals older than 60 years.Age distribution in other central sleep apnea syndromes is unknown

## **Prognosis**

The mortality and morbidity associated with primary central apnea remains unknown; however, these individuals are unlikely to develop significant hypercarbia or hypoxia to the detriment of pulmonary circulation or cor pulmonale. Patients with heart failure and CSB-CSA have a higher mortality rate than those without it. In one study, the 2-year survival rate for patients in heart failure with concomitant CSB-CSA higher than in those without CSB-CSA.A more recent study demonstrated a higher mortality rate in congestive heart failure patients with central sleep apnea than those with no sleep apnea. However, the observed difference was no longer significant after adjusting for age and New York Heart Association functional class.

A study of sleep-disordered breathing and nocturnal cardiac arrhythmias in older men documented that the likelihood of atrial fibrillation or complex ventricular ectopy increased along with the severity of sleep-disordered breathing, which included obstructive sleep apnea and CSB-CSA.Different forms of sleep-disordered breathing were associated with different types of arrhythmias, and central sleep apnea was strongly associated with atrial fibrillation/flutter. The odds of atrial fibrillation (*P* = .01) and of complex ventricular ectopy (*P*< .001) increased with increasing quartiles of the respiratory disturbance index (a major index including all apneas and hypopneas).

## **Predefined Questions and Answers for Central Sleep Apnea (CSA)**

1. What is central sleep apnea?  
Central sleep apnea is a sleep disorder where breathing repeatedly stops and starts during sleep because the brain fails to send proper signals to the muscles that control breathing. This differs from obstructive sleep apnea, which is caused by airway blockage.

2. What causes central sleep apnea?  
Common causes include heart failure, stroke, brainstem injuries, high altitude, use of opioid medications, and certain neurological conditions. Sometimes, no cause is found (idiopathic CSA).

3. What are the symptoms of central sleep apnea?  
Symptoms include pauses in breathing during sleep, sudden awakenings with shortness of breath, difficulty staying asleep, excessive daytime sleepiness, morning headaches, trouble concentrating, mood changes, and sometimes snoring (less common than in obstructive sleep apnea).

4. How is central sleep apnea diagnosed?  
Diagnosis is made through a sleep study (polysomnography) that monitors breathing patterns, oxygen levels, and brain activity during sleep. It helps distinguish CSA from other sleep disorders like obstructive sleep apnea or narcolepsy.

5. What treatments are available for central sleep apnea?  
Treatment depends on the cause and severity:

* Managing underlying conditions (e.g., heart failure)
* Positive airway pressure (PAP) therapies such as CPAP, BiPAP, or adaptive servo-ventilation (ASV) (ASV is contraindicated in some heart failure patients)
* Supplemental oxygen during sleep
* Medications like acetazolamide or theophylline in select cases
* Implantable phrenic nerve stimulation devices for moderate to severe CSA unresponsive to other treatments.

6. What are the risks or side effects of treatment?  
PAP therapies may cause discomfort, nasal congestion, or dry mouth. Implantable devices carry surgical risks such as infection. Medications can have side effects and require close monitoring.

7. Can lifestyle changes help with central sleep apnea?  
Avoiding sedatives and opioids, maintaining a healthy weight, and treating underlying medical conditions can help reduce symptoms.

8. How can I live well with central sleep apnea?  
Education about the condition, adherence to treatment, regular follow-up with your healthcare provider, and support from sleep specialists can improve quality of life

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I reviewed your sleep study results, and it appears you have a condition called *central sleep apnea*. This means that during sleep, your brain sometimes doesn’t send the right signals to your breathing muscles, causing pauses in breathing.

Patient: I see. How is that different from the sleep apnea I hear about, like obstructive sleep apnea?

Doctor: That’s a great question. In obstructive sleep apnea, the airway is physically blocked, so you try to breathe but can’t get air in. In central sleep apnea, the problem is that your brain temporarily stops telling your body to breathe. It’s less common than obstructive sleep apnea but still important to treat.

Patient: What causes central sleep apnea?

Doctor: It can be caused by several things, including heart failure, certain neurological conditions, or use of some medications like opioids. Sometimes it happens without a clear cause, which we call primary central sleep apnea.

Patient: What treatments are available?

Doctor: Treatment depends on the cause and severity. We often start with positive airway pressure therapy, like CPAP or adaptive servo-ventilation, to help regulate your breathing during sleep. We’ll also manage any underlying conditions that may contribute. In some cases, supplemental oxygen or medications may be helpful.

Patient: Are there any risks or side effects with these treatments?

Doctor: Some people find the machines uncomfortable at first, and there can be side effects like nasal dryness or congestion. We’ll work closely with you to adjust the settings and address any issues. Implantable devices are an option for some patients but involve surgery and monitoring.

Patient: How will I know if the treatment is working?

Doctor: We’ll monitor your symptoms and may do follow-up sleep studies or use remote monitoring to see how well your breathing improves. Many patients notice better sleep quality and less daytime fatigue when treatment is effective.

Patient: What should I watch out for or report?

Doctor: If you notice worsening daytime sleepiness, new breathing difficulties, or side effects from treatment, please contact us promptly. Also, any new symptoms related to heart or neurological health should be evaluated.

Patient: Thank you. It helps to understand what’s going on and what to expect.

Doctor: You’re welcome. We’ll support you throughout your treatment and are here to answer any questions along the way.

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**OBSTRUCTIVE SLEEP APNEA**

**DEFINITION AND DESCRIPTION**

Obstructive sleep apnea is the most common sleep-related breathing disorder. People with obstructive sleep apnea repeatedly stop and start breathing while they sleep.

There are several types of sleep apnea. Obstructive sleep apnea occurs when the throat muscles relax and block the airway. This happens off and on many times during sleep. A sign of obstructive sleep apnea is snoring.

Treatments for obstructive sleep apnea are available. One treatment is a device that uses positive pressure to keep the airway open during sleep. Another option is a mouthpiece to thrust the lower jaw forward during sleep. In some people, surgery might be an option too.

**Causes**

Obstructive sleep apnea occurs when the muscles in the back of the throat relax too much to allow for proper breathing. These muscles support the back of the roof of the mouth, known as the soft palate. The muscles also support the tongue and side walls of the throat.

When the muscles relax, the airway narrows or closes as you breathe in. This can lower the level of oxygen in the blood and cause a buildup of carbon dioxide.

Your brain senses this impaired breathing and briefly rouses you from sleep so that you can reopen your airway. This awakening is usually so brief that you don't remember it.

You may awaken with shortness of breath that corrects itself quickly, within one or two deep breaths. Or you might make a snorting, choking or gasping sound.

This pattern can repeat itself 5 to 30 times or more each hour, all night long. These disruptions impair your ability to reach the deep, restful phases of sleep, and you'll probably feel sleepy during your waking hours.

People with obstructive sleep apnea might not be aware of their interrupted sleep. Many people with this type of sleep apnea don't realize they haven't slept well all night.

**Risk factors**

Anyone can develop obstructive sleep apnea. However, certain factors put you at increased risk, including:

* **Excess weight.** Most but not all people with obstructive sleep apnea are overweight. Fat deposits around the upper airway can obstruct breathing. Medical conditions that are associated with obesity, such as hypothyroidism and polycystic ovary syndrome, also can cause obstructive sleep apnea.
* **Older age.** The risk of obstructive sleep apnea increases as you age but appears to level off after your 60s and 70s.
* **Narrowed airway.** A naturally narrow airway is a trait that can be passed down in your family. Or your tonsils or adenoids might become enlarged and block your airway.
* **High blood pressure, known as hypertension.** Obstructive sleep apnea is relatively common in people with hypertension.
* **Chronic nasal congestion.** Obstructive sleep apnea occurs twice as often in those who have consistent nasal congestion at night, regardless of the cause.
* **Smoking.** People who smoke are more likely to have obstructive sleep apnea.
* **Diabetes.** Obstructive sleep apnea might be more common in people with diabetes.
* **Male sex.** In general, men are 2 to 3 times more likely as premenopausal women to have obstructive sleep apnea. However, the risk of obstructive sleep apnea increases in women after menopause.
* **A family history of sleep apnea.** Having family members with obstructive sleep apnea might increase your risk.
* **Asthma.** Research has found an association between asthma and the risk of obstructive sleep apnea.

**Symptoms**

Symptoms of obstructive sleep apnea include:

* Excessive daytime sleepiness.
* Loud snoring.
* Observed episodes of stopped breathing during sleep.
* Waking during the night and gasping or choking.
* Awakening in the morning with a dry mouth or sore throat.
* Morning headaches.
* Trouble focusing during the day.
* Mood changes, such as depression or being easily upset.
* High blood pressure.
* Decreased interest in sex.

### **When to see a doctor**

Consult a health care professional if you have, or if your partner observes, the following:

* Snoring loud enough to disturb your sleep or the sleep of others.
* Waking up gasping or choking.
* Pausing in your breathing during sleep.
* Having excessive daytime drowsiness. This may cause you to fall asleep while working, watching television or even driving a vehicle.

Snoring doesn't necessarily indicate something potentially serious, and not everyone who snores has obstructive sleep apnea.

Be sure to talk to a member of your health care team if you snore loudly, especially if your snoring is interrupted by periods of silence. Snoring may be loudest — and breath pauses known as apneas may be more common — when you sleep on your back.

Ask your health care team about any sleep problem that leaves you fatigued, sleepy and irritable on a regular basis. Excessive daytime drowsiness may be due to other disorders, such as narcolepsy.

## **Diagnosis**

A member of your health care team evaluates your condition based on your symptoms, an exam, and tests. You may be referred to a sleep specialist for further evaluation.

The physical exam involves an examination of the back of your throat, mouth and nose. Your neck and waist circumference may be measured. Your blood pressure also may be checked.

A sleep specialist can further evaluate you. The specialist can diagnose and determine the extent of your condition. The specialist also can plan your treatment. The evaluation might involve staying at a sleep center overnight. At the sleep center, your breathing and other body functions are monitored as you sleep.

### **Tests**

Tests to detect obstructive sleep apnea include:

* **Polysomnography.** During this sleep study, you're hooked up to equipment that monitors your heart, lung and brain activity and breathing patterns while you sleep. The equipment also measures arm and leg movements and blood oxygen levels.

You might be monitored all night or part of the night. If you're monitored for part of the night, it's called a split-night sleep study.

In a split-night sleep study, you'll be monitored during the first half of the night. If you're diagnosed with obstructive sleep apnea, staff members may wake you and give you continuous positive airway pressure for the second half of the night

The sleep study also can help look for other sleep disorders that can cause excessive daytime sleepiness but have different treatments. The sleep study can uncover leg movements during sleep, known as periodic limb movement disorder. Or the study can help evaluate people who have sudden bouts of sleep during the day, known as narcolepsy.

* **Home sleep apnea testing.** Under certain circumstances, you may have an at-home version of polysomnography to diagnose obstructive sleep apnea. Home sleep apnea testing kits monitor a limited number of variables to detect breathing pauses during sleep.

**TREATMENT**

* **Positive airway pressure.** If you have obstructive sleep apnea, you may benefit from positive airway pressure. In this treatment, a machine delivers air pressure through a piece that fits into your nose or is placed over your nose and mouth while you sleep.

Positive airway pressure reduces the number of times you stop breathing as you sleep. The therapy also reduces daytime sleepiness and improves your quality of life.

The most common type is called continuous positive airway pressure, also known as CPAP (SEE-pap). With this treatment, the pressure of the air breathed is continuous, constant and somewhat greater than that of the surrounding air. The pressure of the air is just enough to keep your upper airway passages open. This air pressure prevents obstructive sleep apnea and snoring.

Although CPAP is the most successful and commonly used method of treating obstructive sleep apnea, some people find the mask uncomfortable or loud. However, newer machines are smaller and less noisy than older machines. And there are a variety of mask designs for individual comfort.

Also, with some practice, most people learn to adjust the mask to obtain a comfortable and secure fit. You may need to try different types to find a suitable mask. Several options are available, such as nasal masks, nasal pillows or face masks.

If you're having trouble tolerating pressure, some machines have special adaptive pressure functions to improve comfort. You might also benefit from using a humidifier along with your CPAP system.

CPAP may be given at a continuous pressure, known as fixed. Or the pressure may be varied, known as autotitrating positive airway pressure (APAP). In fixed CPAP, the pressure stays constant. In autotitrating CPAP, the levels of pressure are adjusted if the device senses increased airway resistance.

Bilevel positive airway pressure (BPAP) is another type of positive airway pressure. BPAP delivers a preset amount of pressure when you breathe in and a different amount of pressure when you breathe out.

CPAP is more commonly used because it's been well studied for obstructive sleep apnea and has been shown to effectively treat the condition. People who have difficulty tolerating fixed CPAP might want to try BPAP or APAP.

Don't stop using your positive airway pressure machine if you have problems. Check with your health care team to see what adjustments you can make to improve its comfort.

In addition, contact your health care team if you still snore despite treatment, if you begin snoring again, or if your weight goes up or down by 10% or more.

* **Mouthpiece, known as an oral device.** Though positive airway pressure is often an effective treatment, oral appliances are an alternative for some people with mild or moderate obstructive sleep apnea. They're also used for people with severe sleep apnea who can't use CPAP. The devices may reduce sleepiness and improve quality of life.

These devices are designed to keep the throat open. Some devices keep the airway open by bringing the lower jaw forward, which can sometimes relieve snoring and obstructive sleep apnea. Other devices hold the tongue in a different position.

If you decide to explore this option, you'll need to see a dentist experienced in dental sleep medicine appliances for the fitting and follow-up therapy. A number of devices are available. Close follow-up is needed to ensure successful treatment and that use of the device doesn't cause changes to your teeth.

A newer device uses electrical stimulation on the tongue. The device helps improve snoring and breathing during sleep in people with very mild sleep apnea and snoring. This device isn't meant to be used in place of CPAP when it's recommended for moderate to severe obstructive sleep apnea.

It's a removable device that you place around your tongue while you're awake. It delivers electrical impulses to improve the muscle tone of the tongue. This helps prevent the tongue from collapsing and blocking the airway during sleep. The device is used for 20 minutes a day. It takes six weeks to see improvement. A dentist makes a custom device that fits you.

Only a small number of studies has looked at how well these devices work. Larger studies are still needed. Don't use a tongue muscle stimulation device if you have a pacemaker or another implanted electrical device.

### **Surgery or other procedures**

Surgery is usually considered only if other therapies haven't been effective or haven't been appropriate options for you. Surgical options may include:

* **Surgical removal of tissue.** Uvulopalatopharyngoplasty (UPPP) is a procedure in which a surgeon removes tissue from the back of the mouth and top of the throat. The tonsils and adenoids may be removed as well. UPPP usually is performed in a hospital and requires a medicine that puts you in a sleep-like state. This medicine is called a general anesthetic.
* **Upper airway stimulation.** This new device is approved for use in people with moderate to severe obstructive sleep apnea who can't tolerate CPAP or BPAP.

A small, thin impulse generator, known as a hypoglossal nerve stimulator, is implanted under the skin in the upper chest. When you inhale, the device stimulates the nerve that controls the movement of the tongue. The tongue moves forward instead of moving backward and blocking the throat.

Studies have found that upper airway stimulation greatly improves obstructive sleep apnea symptoms and quality of life.

* **Jaw surgery, known as maxillomandibular advancement.** In this procedure, the upper and lower parts of the jaw are moved forward compared with the rest of the facial bones. This enlarges the space behind the tongue and soft palate, making obstruction less likely.
* **Surgical opening in the neck, known as a tracheostomy.** You may need this form of surgery if other treatments have failed and you have life-threatening obstructive sleep apnea.

During a tracheostomy, a surgeon makes an opening in the neck and inserts a metal or plastic tube for breathing. Air passes in and out of the lungs, bypassing the blocked air passage in your throat.

Other types of surgery may help reduce snoring and sleep apnea by clearing or enlarging air passages, including:

* Nasal surgery to remove polyps or straighten a crooked partition between the nostrils, called a deviated septum.
* Surgery to remove enlarged tonsils or adenoids.

**Lifestyle and home remedies**

In many cases, self-care may be the most appropriate way for you to deal with obstructive sleep apnea. Try these tips:

* **Lose weight.** If you're overweight or obese, even moderate weight loss may help relieve constriction of your airway. Losing weight also can improve your health and quality of life and might reduce your daytime sleepiness.
* **Exercise.** Exercising, such as aerobic exercise and strength training, can help improve your condition. Aim to exercise about 150 minutes a week, and generally try to exercise most days of the week.
* **Don't drink alcohol or use some anti-anxiety medicines or sleeping pills.** Alcohol, some anti-anxiety medicines and some sleeping pills can worsen obstructive sleep apnea and sleepiness.
* **Sleep on your side or stomach rather than on your back.** Sleeping on your back can cause your tongue and soft palate to rest against the back of your throat and block your airway.

To prevent sleeping on your back, try sewing a tennis ball in the back of your pajama top or place pillows behind you to keep you sleeping on your side.

* **Keep your nasal passages open while you sleep.** If you have congestion, use a saline nasal spray to help keep your nasal passages open. Talk to a member of your health care team about using nasal decongestants or antihistamines, because some medicines may be recommended for only short-term use.

**Complications**

Obstructive sleep apnea is considered a serious medical condition. Complications can include:

* **Daytime fatigue and sleepiness.** Because of a lack of restorative sleep at night, people with obstructive sleep apnea often have severe daytime drowsiness, fatigue and irritability. They might have difficulty concentrating and find themselves falling asleep at work, while watching TV or even when driving. This can put them at higher risk of work-related accidents.

Children and young people with obstructive sleep apnea might do poorly in school and commonly have attention or behavior problems.

* **Cardiovascular problems.** Sudden drops in blood oxygen levels that occur during obstructive sleep apnea increase blood pressure and strain the cardiovascular system. Many people with obstructive sleep apnea develop high blood pressure, which can increase the risk of heart disease.

The worse the obstructive sleep apnea, the greater the risk of coronary artery disease, heart attack, heart failure and stroke.

Obstructive sleep apnea also increases the risk of heart rhythm problems known as arrhythmias. Arrhythmias can lower blood pressure. If there's underlying heart disease, these repeated multiple episodes of arrhythmias could lead to sudden death.

* **Complications with medicines and surgery.** Obstructive sleep apnea also is a concern with certain medicines and general anesthesia. Medicines such as sedatives, some prescription painkillers and general anesthetics, relax the upper airway and can make obstructive sleep apnea worse.

If you have obstructive sleep apnea, having major surgery can worsen breathing problems. This is especially true if you have been sedated and you were lying on your back. People with obstructive sleep apnea might be more prone to complications after surgery.

Before you have surgery, tell your surgeon if you have obstructive sleep apnea or symptoms related to the condition. You may need to get tested for obstructive sleep apnea before surgery.

* **Eye problems.** Some research has found a connection between obstructive sleep apnea and certain eye conditions, such as glaucoma. Eye complications can usually be treated.
* **Sleep-deprived partners.** Loud snoring can keep those around you from getting good rest and eventually disrupt your relationships. Some partners choose to sleep in another room.

People with obstructive sleep apnea also may complain of memory problems, morning headaches, and mood swings or depression. They also may need to urinate often at night.

Obstructive sleep apnea might be a risk factor for COVID-19. People with obstructive sleep apnea have been found to be at higher risk for developing a severe form of COVID-19. They may be more likely to need hospital treatment than do those who don't have obstructive sleep apnea.

## **Outlook / Prognosis**

Untreated OSA may reduce your life expectancy and increase your risk of dangerous complications. But OSA is a treatable condition. A healthcare provider is the best person to talk to about what you can expect, as this answer is very unique to you.

### **What’s the outlook for obstructive sleep apnea?**

The outlook for OSA depends on many factors like the severity and whether you have other underlying conditions, too. However, you can expect a positive outcome if you stick to your treatment plan after you and your healthcare provider find one that works best for you.

## **Prevention**

You can’t prevent all cases of OSA. But you can take steps to reduce your risk and improve your overall sleep by:

* Eating nutritious foods and participating in regular physical activities
* Maintaining a healthy weight
* Practicing good sleep hygiene (like setting a bedtime routine and turning off electronic devices before bed)
* Managing any existing health conditions, such as high cholesterol, high blood pressure and Type 2 diabetes
* Not smoking and not drinking beverages that contain alcohol before bed
* Seeing your healthcare provider annually for a check-up

## **Predefined Questions and Answers for Obstructive Sleep Apnea (OSA)**

1. What is obstructive sleep apnea?  
Obstructive sleep apnea is a common sleep disorder where the airway becomes partially or completely blocked during sleep, causing breathing pauses, snoring, and disrupted sleep.

2. What are the common symptoms of OSA?  
Symptoms include loud snoring, gasping or choking during sleep, excessive daytime sleepiness, morning headaches, difficulty concentrating, and restless sleep.

3. How is OSA diagnosed?  
Diagnosis is made through a sleep study (polysomnography) that monitors breathing, oxygen levels, heart rate, and brain activity during sleep. Home sleep apnea tests may be used in some cases.

4. What causes obstructive sleep apnea?  
Causes include excess weight, enlarged tonsils, a small jaw or airway, nasal congestion, alcohol or sedative use, and certain medical conditions like hypothyroidism.

5. What treatments are available for OSA?  
The primary treatment is positive airway pressure (PAP) therapy, such as CPAP, which keeps the airway open during sleep. Other treatments include lifestyle changes (weight loss, avoiding alcohol), oral appliances, and surgery in severe cases.

6. How does a CPAP machine work?  
A CPAP machine delivers a steady stream of pressurized air through a mask to keep your airway open while you sleep, reducing apneas and improving sleep quality.

7. What lifestyle changes can help manage OSA?  
Weight loss, avoiding alcohol and sedatives before bedtime, quitting smoking, and sleeping on your side instead of your back can reduce symptoms.

8. What are the risks if OSA is left untreated?  
Untreated OSA increases the risk of high blood pressure, heart disease, stroke, diabetes, daytime accidents due to sleepiness, and reduced quality of life.

9. How can I adjust to using a CPAP machine?  
It may take time to get used to the mask and machine. Communicate any discomfort or issues with your healthcare provider, who can help with mask fitting and adjustments.

10. Should I see a sleep specialist?  
If you have symptoms like loud snoring, daytime sleepiness, or witnessed breathing pauses, seeing a sleep specialist for evaluation and possible sleep study is recommended

## **Obstructive Sleep Apnea (OSA) Treatment Drugs and Their Side Effects**

## 1. Modafinil (Provigil) and Armodafinil (Nuvigil)

* Use: Promote wakefulness in patients with residual daytime sleepiness despite CPAP therapy.
* Mechanism: Central nervous system stimulants that enhance alertness.
* Side Effects:
  + Headache
  + Nausea
  + Nervousness or anxiety
  + Insomnia
  + Rare serious skin reactions (e.g., Stevens-Johnson syndrome)
* Notes: Do not treat the underlying apnea; only improve wakefulness.

## 2. Solriamfetol (Sunosi)

* Use: Treat excessive daytime sleepiness in OSA patients who remain sleepy despite primary therapy.
* Mechanism: Dopamine and norepinephrine reuptake inhibitor.
* Side Effects:
  + Headache
  + Nausea
  + Anxiety
  + Insomnia
  + Increased blood pressure and heart rate
* Notes: Requires monitoring of cardiovascular status.

## 3. Tirzepatide (Zepbound)

* Use: Recently FDA-approved for moderate to severe OSA, primarily via weight loss effects as a GLP-1 receptor agonist.
* Side Effects:
  + Nausea
  + Diarrhea
  + Decreased appetite
  + Injection site reactions
* Notes: Weight loss can improve OSA severity.

## 4. Other Off-label or Investigational Drugs

* Theophylline: A bronchodilator with mild stimulant effects; limited use due to side effects.
* Acetazolamide: Carbonic anhydrase inhibitor; may improve breathing stability but limited by side effects like metabolic acidosis.
* Dronabinol: Cannabinoid receptor agonist; some evidence for reducing apnea events but not widely used.
* Antidepressants and nasal decongestants: Occasionally used but evidence is limited.

## Non-Pharmacologic Treatments (Primary Therapy)

* CPAP: Delivers continuous positive airway pressure to keep airways open. Side effects include nasal dryness, congestion, discomfort, and mask intolerance.
* Oral appliances: Mandibular advancement devices for mild to moderate OSA.
* Surgery: For anatomical causes or CPAP intolerance (e.g., hypoglossal nerve stimulation).
* Lifestyle changes: Weight loss, positional therapy, avoiding alcohol and sedatives

## **Diagnostic Considerations**

A diagnosis of narcolepsy may be delayed if obstructive sleep apnea (OSA) is considered the only condition. Patients should be routinely screened clinically for symptoms of narcolepsy. These patients do not typically have normal sleepiness when OSA has been treated; they may experience improvement in sleepiness, but it is important to question the diagnosis of sleepiness due to OSA despite ideal treatment.

## Indices for sleep-disordered breathing

The indices commonly used to assess sleep disordered breathing (SDB) are the apnea-hypopnea index (AHI) and the respiratory disturbance index (RDI).

The AHI is defined as the average number of episodes of apnea and hypopnea per hour. The RDI is defined as the average number of respiratory disturbances (obstructive apneas, hypopneas, and respiratory event–related arousals [RERAs]) per hour. If the AHI or RDI is calculated based on less than 2 hours of continuous recorded sleep, the total number of recorded events to calculate the AHI or RDI during sleep testing is at least the number of events that would have been required in a 2-hour period.

No universal consensus exists on whether the AHI or the RDI should be the standard index used to determine treatment by specialists and insurance carriers, with Medicare being the most confusing as it varies by region as to whether AHI and RDI can be used. This needs to be resolved as soon as possible. One study found that 30% of symptomatic patients would have been left untreated if the AHI were used rather the RDI.

In the authors’ view, the RDI is preferable to the AHI because it includes flow-limitation events that end with arousals. The RDI is better suited to meet the new American Academy of Sleep Medicine (AASM) diagnostic criteria for OSA (see below). One study has demonstrated that use of the

Diagnostic criteria for OSA

A positive test for OSA is established if either of the following criteria using the AHI or the RDI is met:

* AHI or RDI greater than or equal to 15 events per hour, or
* AHI or RDI greater than or equal to 5 and less than or equal to 14 events per hour with documented symptoms of excessive daytime sleepiness (EDS); impaired cognition; mood disorders; insomnia; or documented hypertension, ischemic heart disease, or history of stroke

The AASM has developed its own criteria, as listed in the *International Classification of Sleep Disorders: Diagnostic and Coding Manual, Second Edition.*At least 1 of the following criteria must apply for OSA to be diagnosed:

* The patient reports daytime sleepiness, unrefreshing sleep, fatigue, insomnia, and/or unintentional sleep episodes during wakefulness. The patient awakens with breath holding, gasping, or choking. The patient’s bed partner reports loud snoring, breathing interruptions, or both during the patient’s sleep.
* Polysomnography (PSG) shows more than 5 scoreable respiratory events (eg, apneas, hypopneas, RERAs) per hour of sleep and/or evidence of respiratory effort during all or a portion of each respiratory event.
* PSG shows more than 15 scorable respiratory events (eg, apneas, hypopneas, RERAs) per hour of sleep and/or evidence of respiratory effort during all or a portion of each respiratory event.
* Another current sleep disorder, medical or neurologic disorder, medication use, or substance use does not better account for the patient’s condition.

Accreditation of sleep centers by the AASM is critical because there are still more centers that are unaccredited than there are centers that have chosen to meet the highest standards in the field (as evidenced by achieving AASM accreditation). Whether AASM accreditation translates into insurance companies deciding to pay for studies performed at an AASM-accredited center has yet to be determined, although in the authors’ opinion, payment should depend on achieving AASM accreditation.

Diagnostic considerations include the following:

* Chronic insufficient sleep
* Dyspnea due to pulmonary edema
* Idiopathic hypersomnia
* Nocturnal panic attacks
* Nonobstructive alveolar hypoventilation
* Obesity-hypoventilation syndrome (pickwickian syndrome)
* Periodic limb movement disorder
* Simple snoring
* Approximately 25% of narcoleptic persons also have obstructive sleep apnea)

## **Differential Diagnoses**

* Asthma
* Central Sleep Apnea Syndromes
* Chronic Obstructive Pulmonary Disease (COPD)
* Depression
* Gastroesophageal Reflux Disease
* Hypothyroidism
* Narcolepsy
* Periodic Limb Movement Disorder

## **Epidemiology**

SDB is common in the United States. The National Commission on Sleep Disorders Research estimated that minimal SDB (RDI >5) affects 7-18 million people in the United States and that relatively severe cases (RDI >15) affect 1.8-4 million people. The prevalence increases with age. SDB remains undiagnosed in approximately 92% of affected women and 80% of affected men.

OSA is increasingly prevalent, in both adults and children, in modern society. The estimated prevalence has been 2% for women and 4% for men.Similar data have been found in an epidemiologic study from Pennsylvania.More recent research indicates a prevalence of 4% for women and 9% for men. Data from the Wisconsin Cohort Study indicate that the prevalence of OSA in people aged 30-60 years is 9-24% for men and 4-9% for women.

The prevalence in children is less certain, but the author’s sleep center is seeing increasing numbers of adolescent patients, who are often obese and present similarly to many of their adult counterparts, with the important exception that they may be sleepy and/or hyperactive. A 2007 study has suggested that approximately 6% of adolescents have weekly SDB.

### International statistics

The prevalence of OSA in non-American populations has only been studied in men and has been found to be as low as 0.3% (England) and as high as 20-25% (Israel and Australia). The prevalence of OSA in Australian men is estimated to be 3%.

### Age distribution for OSA

Aging is an important consideration of risk for OSA. OSA prevalence increases 2-3 times in older persons (>65 y) compared with individuals aged 30-64 years,with an estimated rate as high as 65% in a community sample of people older than 65 years.

After age 65 years, no further relative disparity is noted in the incidence of OSA. One explanation for this plateau is the relative increase in mortality in persons older than 65 years; however, data to support this contention, as attractive as it appears, are insufficient. Scant data are available to help clinicians determine if clinical management should differ between the age cohorts.

### Sex distribution for OSA

The male-to-female ratio in community-based studies is 2-3:1.Androgenic patterns of body fat distribution (deposition in the trunk, including the neck area) predispose men to OSA. In general, sex hormones may affect neurologic control of UA-dilating muscles and ventilation.

In population studies that have examined the incidence of OSA, women were not only less likely than men to have OSA but also less likely to be diagnosed early in the disease process. Survival rates are lower for women than for men, after an OSA diagnosis has been established by PSG, presumably due to the delayed OSA diagnosis.

Three large epidemiologic studies have demonstrated that the prevalence of OSA in women appears to increase after menopause.In these studies, women on hormone replacement therapy (HRT) had a prevalence similar to that of premenopausal women. Postmenopausal women are 3 times more likely to have moderate-to-severe OSA compared with premenopausal women. Women who are on HRT are half as likely to have OSA compared with postmenopausal women who are not on HRT.

Premenopausal women with OSAHS tend to be more obese than men with the same severity of disease. Thin women with symptoms of OSAHS appear to have an increased frequency of craniofacial abnormalities.

Evidence indicates that women underreport the symptoms of loud snoring and witnessed apneas, leading to underreferral to sleep centers. This may explain the marked male predominance (male-to-female ratio of approximately 8:1) in sleep center–based studies. Additionally, women have lower AHIs than men, even after correcting for other demographic factors such as BMI and neck circumference.

### Prevalence of OSA by race or ethnicity

African American individuals appear to be more predisposed to SDB than white persons. This increased predisposition varies according to age. The odds ratio is greater than 3 in children younger than 13 years and is 1.88 in persons younger than 25 years. In elderly African Americans, the risk is increased 2-fold. Examination of craniofacial morphology found that brachycephaly is associated with an increased AHI in whites but not in African Americans.

Chinese patients with OSA have a more crowded upper airway and relative retrognathia compared with their white counterparts, with statistical controls for BMI and neck circumference.Asians are known to have a shorter cranial base and a more acute cranial base flexure, increasing OSA risk, with BMI and neck circumference being roughly equal. Therefore, interestingly, obesity plays a more prominent role in OSA predisposition in whites than in Chinese persons. This may serve to underscore the role that craniofacial factors have in Chinese patients.

Other populations that may be at increased risk include Mexican Americans and Pacific Islanders.

## **Guidelines Summary**

The *AASM Scoring Manual* also provides guidance on standard montages, electrode placements and technical and digital specifications. The recommendations for the indications and performance of polysomnography include the following:

* Sleep stages are recorded via an electroencephalogram, electro-oculogram, and chin electromyogram
* Heart rhythm is monitored with a single-lead electrocardiogram
* Leg movements are recorded via an anterior tibialis electromyogram
* Breathing is monitored, including airflow at the nose and mouth (using both a thermal sensor and a nasal pressure transducer), effort (using inductance plethysmography), and oxygen saturation
* The breathing pattern is analyzed for the presence of apneas and hypopneas (as per definitions standardized by the American Academy of Sleep Medicine)

The guidelines include the following strong recommendations:

* Clinical tools, questionnaires and prediction algorithms should not be used to diagnose OSA in adults without polysomnography or home sleep apnea testing (HSAT).
* Polysomnography or home sleep apnea testing with a technically adequate device, should be used for the diagnosis of OSA in uncomplicated adult patients with clinical signs of an increased risk of moderate to severe OSA.
* When a single HSAT is negative, inconclusive, or technically inadequate, polysomnography should be performed for the diagnosis of OSA.
* Polysomnography should be used for the diagnosis of OSA in adults with significant cardiorespiratory disease, potential respiratory muscle weakness due to neuromuscular condition, awake hypoventilation or suspicion of sleep-related hypoventilation, chronic opioid medication use, history of stroke or severe insomnia.

In 2021 the AASM published guideline on referral for surgical therapy with the following recommendations:

* Referral to a sleep surgeon for alternative treatments should be discussed with patients with BMI < 40 kg/m2 if the patient's adherence and tolerance does not support adequate PAP or if the patient rejects PAP.
* Referral to a bariatric surgeon should be discussed with adults with OSA and obesity as an alternative treatment option if the patient's adherence and tolerance does not support adequate PAP or if the patient rejects PAP.
* Discussion regarding a referral to both sleep and bariatric surgeons to discuss management options may be appropriate in patients with a BMI range of 35–40 kg/m2.
* PAP should be the initial therapy for adults with OSA and a major upper airway anatomic abnormality prior to referral for upper airway surgery

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I’ve reviewed your symptoms and test results, and it looks like you have obstructive sleep apnea. This means that during sleep, your airway is partially blocked, causing pauses in breathing and disrupted sleep.

Patient: I see. What causes this blockage? Is it serious?

Doctor: The blockage can be due to several factors like excess weight, enlarged tonsils, or anatomical differences in your airway. It is serious because these breathing pauses can lower oxygen levels and disrupt your sleep, leading to daytime fatigue and increasing risks for heart disease, high blood pressure, and other health problems.

Patient: How do you confirm this diagnosis?

Doctor: The diagnosis is confirmed with a sleep study, called polysomnography, which monitors your breathing, oxygen levels, and sleep stages overnight. Sometimes a home sleep test is sufficient, depending on your situation.

Patient: What treatment options do I have?

Doctor: The most effective treatment is usually a CPAP machine, which delivers continuous air pressure to keep your airway open while you sleep. There are also oral appliances that reposition your jaw, lifestyle changes like weight loss and avoiding alcohol before bedtime, and in some cases, surgery.

Patient: I’m worried about using a CPAP machine. Is it hard to get used to?

Doctor: It can take some time to adjust, but we’ll work with you to find a mask that fits well and address any discomfort. Many patients find their sleep quality improves significantly once they get used to it. Also, involving your bed partner in this process can help with motivation and adherence.

Patient: Are there any other things I should do to improve my condition?

Doctor: Yes, losing weight if you’re overweight, sleeping on your side instead of your back, avoiding sedatives or alcohol before bed, and managing any nasal congestion can all help reduce symptoms.

Patient: What happens if I don’t treat this?

Doctor: Untreated OSA can lead to serious complications like high blood pressure, heart disease, stroke, diabetes, and increased risk of accidents due to daytime sleepiness. So, treatment is very important.

Patient: Thank you, doctor. What’s the next step?

Doctor: I’ll refer you for a sleep study if you haven’t had one yet, and once we have the results, we’ll discuss the best treatment plan tailored to you. Meanwhile, try to note your sleep patterns and any symptoms you experience.

Patient: I appreciate that. I feel better knowing there are options.

Doctor: You’re welcome. We’re here to support you every step of the way. Please reach out anytime with questions or concerns.

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### **Croup**

**DEFINITION AND DESCRIPTION**

Croup (laryngotracheobronchitis) is a respiratory infection that affects young children. Viral infections are the most common cause of the condition. Croup causes swelling of your child’s voice box (larynx) and windpipe (trachea). This swelling causes the airway below their vocal cords to narrow, which makes their breathing noisy and difficult.

Croup in babies is most common, along with children younger than 3 years old. As children get older, croup isn’t seen as often. This is because their windpipes get larger and swelling is less likely to get in the way of their breathing.

Croup causes a distinctive cough that may sound similar to the call of a seal. The condition is usually mild, but symptoms can become severe and life-threatening.

#### **RSV vs. croup — what’s the difference?**

RSV (respiratory syncytial virus) and croup are both respiratory illnesses that can affect babies and young children. RSV is a viral infection that can affect both children and adults. It causes coughing, sneezing and other cold-like symptoms.

While RSV is its own illness, the respiratory syncytial virus is also one of the viruses that can lead to croup.

#### **Whooping cough vs. croup — what’s the difference?**

Whooping cough (pertussis) and croup are both respiratory infections that can affect babies and children. Both conditions cause a distinctive cough, although the sound of whooping cough is a more high-pitched gasping or “whooping” noise.

Whooping cough is a bacterial infection whereas a viral infection usually causes croup. Therefore, no vaccines can prevent croup and antibiotics can’t treat it. (Antibiotics can’t kill viruses.) There’s a vaccine to prevent whooping cough, but it doesn’t go away quickly on its own as croup usually does.

Croup affects about 3% of U.S. children every year. The condition accounts for 7% of all hospitalizations in children younger than 5 years old. It’s more common in males. Healthcare providers define about 85% of croup cases as mild. They consider less than 1% of cases severe.

## **Symptoms and Causes**

The croup cough sounds like a harsh “barking” sound. This is the most common symptom of croup. Your child may also have stridor, which is a raspy, vibrating sound that occurs when your child is breathing in.

Croup is typically mild and lasts less than one week, but symptoms can get more severe. Symptoms normally start slowly and may begin with a runny or stuffy nose. Over the next 12 to 48 hours, symptoms can worsen, and the barking cough may start. Symptoms are usually worse at night.

Other mild croup symptoms include:

* Hoarseness.
* Fever.
* Rash.
* Eye redness (conjunctivitis).
* Swollen lymph nodes.

Symptoms of moderate to severe croup may include:

* Difficulty breathing.
* Restlessness or nervousness.
* Retractions (sucking in the skin around your child’s ribs and the top of their breastbone).
* Cyanosis (blue-tinged skin).

### **What causes croup?**

The most common cause of croup is a viral infection. Croup viruses include parainfluenza, influenza, respiratory syncytial virus (RSV), measles and adenovirus. Viral croup causes your child’s upper airways to swell, making it difficult for them to breathe. However, these viruses are common and most children with viral infections don’t develop croup. Rarely, bacteria can complicate the viral infection and make it more difficult to breathe.

#### **Is croup contagious?**

Yes, croup is highly contagious because the viruses that lead to the condition are easily spreadable.

#### **How do you get croup?**

The viruses that cause croup spread easily through the air. When someone with a viral or bacterial infection that can cause croup sneezes or coughs, they send respiratory droplets containing croup-causing germs into the air. When your child breathes in these droplets, they can catch an illness that’ll cause croup. Your child can also get croup by touching objects contaminated by germs that can cause croup.

#### **How long is croup contagious?**

Your child is contagious for three days after their symptoms first appeared or until their fever is gone. You should keep your child home from school until 24 hours have passed without a fever and without using fever-reducing medication.

### **Complications of croup**

Most cases of croup are mild and you can treat them at home. Complications of croup are rare. Less than 5% of children with croup need in-hospital care. Your child’s condition may lead to hospitalization if they:

* Need oxygen therapy to keep their oxygen levels within a safe range.
* Have severe dehydration that requires IV (intravenous, or through your vein) fluids.
* Need multiple doses of inhaled breathing treatments to provide relief.
* Have severe symptoms despite initial treatment.

## **Diagnosis and Tests**

You can usually tell if your child has croup based on their signs and symptoms. The most common symptoms are a barking cough and stridor. This condition is especially widespread in the fall and winter months. If your child’s condition is severe, a healthcare provider may order X-rays and laboratory tests, but this is rare.

## **Management and Treatment**

Croup treatment depends on the severity of your child’s condition and the risk of it rapidly worsening. If your child has a history of respiratory problems or was born prematurely, that may also affect the treatment approach.

#### **Mild croup**

You can usually treat mild croup at home. Home treatment includes using a cool mist humidifier to help soothe dry and irritated airways. You can also sit with your child in a bathroom filled with steam generated from hot water running in the shower. (Don’t sit in the shower or let your child near the hot water.) If your child’s condition doesn’t improve with mist treatment, you should contact their healthcare provider.

Other croup home remedies include:

* Letting your child breathe cool air at night by opening a door or window.
* Treating your child’s fever with an over the counter (OTC) medication such as acetaminophen (Tylenol®) or ibuprofen (Advil®).
* Treating your child’s cough with warm, clear fluids to help loosen the mucus on their vocal cords.
* Avoiding smoking in your home, as smoke can worsen your child’s cough.
* Keeping your child’s head elevated with an extra pillow. (Don’t use pillows with infants younger than 12 months old.)

You may wish to sleep in the same room as your child so you’re there if they start to have trouble breathing.

#### **Moderate to severe croup**

For moderate to severe croup, you should take your child to the nearest urgent care center or emergency room (ER). Severe croup can be life-threatening, and you shouldn’t delay taking your child in. Treatment for moderate to severe croup will vary based on your child’s symptoms. Croup treatments may include:

* Humidified air or oxygen.
* IV fluids for dehydration.
* Monitoring of [v](https://my.clevelandclinic.org/health/articles/10881-vital-signs)ital signs, including oxygen levels, breathing and heart rate.
* Croup medication, including steroids (glucocorticoids) and nebulized breathing treatments (epinephrine).
* Placement of a breathing tube (mechanical ventilation). This is rare.

#### **Specific croup medication**

If you take your child to their provider’s office or the emergency room, their provider will give them a glucocorticoid and a nebulized breathing treatment (epinephrine).

##### **Glucocorticoids**

Glucocorticoids are a type of steroid that decreases the swelling of your child’s voice box (larynx), typically within six hours of the first dose. For a child with mild croup, glucocorticoids may reduce the need for a repeat visit to their provider’s office or the emergency room.

The glucocorticoids healthcare providers use most often are dexamethasone and prednisolone. Your child will usually only need one dose taken by mouth (orally). If your child is vomiting or can’t keep the medicine down, their provider can also give dexamethasone intravenously (IV) or through an intramuscular (IM) injection.

##### **Nebulized breathing treatment (epinephrine)**

Your child will receive epinephrine as an inhaled mist (nebulizer). This also reduces the swelling in your child’s airways and usually starts working within 10 minutes. Epinephrine works for two hours or less, and your child may receive this treatment every 15 to 20 minutes for severe symptoms.

#### **Complications/side effects of the treatment**

Serious side effects of epinephrine are rare. However, side effects could include a rapid heartbeat (tachycardia). A healthcare provider will monitor your child for three to four hours after their last dose to ensure symptoms of airway blockage don’t return.

#### **How soon after treatment will my child feel better?**

Glucocorticoids usually start working within six hours of the first dose. Epinephrine typically begins working faster than glucocorticoids.

## **Outlook / Prognosis**

Croup can be mild, moderate or severe, depending on how difficult it is for your child to pull air into their lungs. The size (diameter) of their windpipe and the amount of narrowing due to the swelling determine the severity of your child’s condition. In addition, your child’s condition may become more severe if they become upset.

#### **Mild croup**

A child with mild croup may have a barking cough and stridor. Symptoms can worsen throughout your child’s illness, especially during the evening hours. So it’s important to keep an eye on their breathing, but you can usually treat their condition at home.

#### **Moderate croup**

A child with moderate croup may have stridor along with retractions (sucking in the skin around their ribs and the top of their breastbone). They may also be slightly agitated or disoriented and may have moderate trouble breathing. You should take your child to see a healthcare provider for treatment.

#### **Severe croup**

A child with severe croup has stridor and retractions. They may also be agitated, anxious or fatigued. Cyanosis (blue-tinged skin) is common. Severe croup is a life-threatening condition. Take your child to the emergency room immediately.

#### **How long does croup last?**

Symptoms of croup usually clear up in most children within two days. However, symptoms can persist for up to one week.

#### **When can my child go back to school?**

Croup is very contagious. Your child should stay home from school until after their fever is gone.

## **Prevention**

Croup can spread by physical contact or through the air. To help prevent its spread:

* Wash and dry your hands thoroughly after caring for your child.
* Wash toys between each use.
* Encourage your child to cover their mouth and nose when coughing and sneezing.
* Keep your child home from school or daycare when they’re ill or if outbreaks occur.
* Throw used tissues away.

### **When should I take my child to see their healthcare provider?**

You should call your child’s healthcare provider if:

* Your child has a fever that lasts for more than three days.
* Your child has symptoms of mild croup that last for more than one week.
* You have questions or are concerned about your child’s condition.

If your child develops symptoms of severe or worsening croup, seek immediate medical attention. These symptoms include:

* Difficulty breathing.
* Blue-tinged skin (cyanosis).
* Severe coughing spells.
* Drooling or difficulty swallowing.
* Inability to cry or speak due to trouble taking a breath.
* A noisy, high-pitched whistling sound while breathing.
* Sucking in the skin around your child’s ribs and the top of their breastbone (retractions).

## **PREDEFINED Questions and answers**

### **Why does my child keep getting croup?**

If your child keeps getting croup, it may be a sign they have a narrowing in their airway and that they’re at a higher likelihood to be affected by the infection. Your child may have been born with the narrowing, or it may have developed after birth. If croup returns (recurs) repeatedly, your child’s provider may refer them to a specialist such as an otolaryngologist (ear, nose and throat doctor) or a pulmonologist (breathing and lung disease doctor).

### **Can adults get croup?**

Adults can get croup, but it’s rare. The reason babies and young children get croup is because their windpipes (tracheas) are narrower and not fully developed. As children get older, their windpipes get larger and fully develop, so any swelling is less likely to affect breathing. Adults have larger airways, so croup doesn’t typically affect them. When adults do get croup, their symptoms are usually worse and they may need more aggressive treatment.

## Diagnostic Considerations

Although croup is considered the most common cause of stridor and respiratory distress in the pediatric population, diagnostic differentials should be considered, dependent on clinical history and presenting symptoms, and include the following:

* Spasmodic croup (recurrent croup, afebrile)
* Retropharyngeal abscess
* Subglottic stenosis
* Angioedema
* Allergic reaction
* Tracheomalacia
* Laryngeal web
* Laryngeal papillomatosis
* Laryngeal hemangioma
* Subglottic hemangioma
* Vocal cord paralysis
* Uvulitis
* Innominate artery compression
* Right aortic arch vascular ring
* Double aortic arch
* Aberrant subclavian artery
* Pulmonary artery sling
* Gastroesophageal reflux (diagnostic consideration for recurrent croup)
* Rarer etiologies in the pediatric population:
  + Laryngeal tuberculosis, neoplasm (compressing trachea), sarcoidosis, Wegener granulomatosis

## Differential Diagnoses

* [Bacterial Tracheitis](https://emedicine.medscape.com/article/961647-overview)
* Inhalation Injury
* [Laryngeal Fractures](https://emedicine.medscape.com/article/865277-overview)
* [Laryngomalacia](https://emedicine.medscape.com/article/1002527-overview)
* [Measles](https://emedicine.medscape.com/article/966220-overview)
* [Pediatric Airway Foreign Body](https://emedicine.medscape.com/article/1001253-overview)
* [Pediatric Diphtheria](https://emedicine.medscape.com/article/963334-overview)
* [Pediatric Epiglottitis](https://emedicine.medscape.com/article/963773-overview)
* [Pediatric Mononucleosis and Epstein-Barr Virus Infection](https://emedicine.medscape.com/article/963894-overview)
* [Pediatric Peritonsillar Abscess](https://emedicine.medscape.com/article/970260-overview)

EPIDEMIOLOGY

The majority of patients with croup, more than 85%, exhibit mild symptoms, and severe croup is rare, fewer than 1%. In the United States, croup accounts annually for approximately 7% of pediatric hospitalizations, with fewer than 3% of admitted cases requiring intubation, and contributes to up to 15% of emergency department visits among children younger than 5.[[5]](https://www.ncbi.nlm.nih.gov/books/NBK431070/#)

Croup typically affects children between the ages of 6 months and 3 years, but can occur as early as 3 months and up to 15 years. The annual incidence is approximately 532 cases per 100,000 individuals, affecting 3% of children younger than 5 globally. Croup rarely occurs in adults. PIV is responsible for more than two-thirds of croup infections. Croup is more common in boys than in girls, with a 1.5:1 ratio, and shows no preference for any race. Croup is more prevalent in resource-limited countries, likely due to a higher proportion of children younger than 6 and suboptimal nutritional status

## Croup Treatment Drugs and Their Side Effects

## 1. Corticosteroids (Dexamethasone and Prednisolone)

* Use:
  + Reduce airway swelling and inflammation in croup.
  + Proven to decrease hospital admissions, shorten hospital stays, and reduce severity of symptoms.
  + Usually given as a single oral dose; sometimes repeated if symptoms persist.
* Common Side Effects:
  + Vomiting
  + Upset stomach
  + Restlessness
  + Headache
  + Hiccups (rare)
* Safety:
  + Short courses (single dose or up to 5 days) have minimal to no significant side effects.
  + Well tolerated and considered very safe in children with croup.
* Administration:
  + Oral syrup or liquid form.
  + Sometimes given as an injection if oral intake is difficult.

## 2. Epinephrine (Adrenaline) Nebulized

* Use:
  + For moderate to severe croup with significant airway obstruction.
  + Rapidly reduces airway swelling with effects starting within 10 minutes but lasting less than 2 hours.
  + Typically administered in hospital settings.
* Common Side Effects:
  + Rapid heartbeat (tachycardia)
  + Nervousness or jitteriness
  + Rare serious side effects; patients are monitored closely after administration.
* Notes:
  + Multiple doses may be given every 15-20 minutes if needed.
  + Observation for several hours after last dose is standard to watch for symptom recurrence.

## 3. Supportive Treatments (Not Drugs but Important)

* Pain and Fever Relief:
  + Paracetamol (acetaminophen) or ibuprofen can be used to reduce fever and ease discomfort.
  + Aspirin should not be given to children under 16.
* Avoid:
  + Cough medicines and decongestants, as they do not help and can cause dangerous drowsiness.
  + Steam inhalation is no longer recommended.

### **Pain relief for children**

Children’s [paracetamol](https://www.nhsinform.scot/tests-and-treatments/medicines-and-medical-aids/types-of-medicine/paracetamol/) can ease pain and may help lower your child’s temperature if they have a fever.

Children under 16 years of age should not be given aspirin.

Speak to your pharmacist or GP if you are unsure about what type of pain relief is right for your child.

Do not use cough medicines or decongestants. These do not help ease the symptoms of croup. These treatments often have drowsy side effects, which can be dangerous when a child has breathing difficulties.

### **Hospital treatment**

In severe cases of croup, treatment in hospital may be needed. This might include your child being given adrenaline or oxygen.

DOCTOR PATIENT CONVERSATION

Doctor: Hello, I understand your child has been coughing and having some noisy breathing. Can you tell me more about what you’ve noticed?

Parent: Yes, my child has a harsh, barking cough and sometimes sounds like they’re struggling to breathe, especially at night. They also have a hoarse voice.

Doctor: Based on what you’re describing, it sounds like your child has croup. Croup is a common viral infection that causes swelling around the vocal cords and windpipe, which leads to that barking cough and noisy breathing.

Parent: Is it serious? Should I be worried?

Doctor: Most cases of croup are mild and get better on their own within a few days. However, sometimes the swelling can make breathing harder, which is why we monitor symptoms carefully. If your child is having trouble breathing, is very restless, or is drooling and unable to swallow, those are signs to seek emergency care immediately.

Parent: What can we do to help them feel better?

Doctor: The main treatment is a steroid medicine called dexamethasone, which reduces the swelling in the airway. It’s usually given as a single dose and works quite quickly. For more severe breathing difficulty, we might use a nebulized epinephrine treatment in the hospital to open up the airway.

Parent: Are there any side effects from the steroid?

Doctor: Steroids like dexamethasone are very safe when used in a single dose for croup. Some children might feel a bit restless or have a mild upset stomach, but serious side effects are very rare.

Parent: Is there anything I can do at home to help?

Doctor: Keeping your child calm is very important because crying can make breathing harder. Make sure they stay hydrated. Sometimes, cool mist or sitting with your child in a steamy bathroom can help soothe their airway, but avoid hot steam or smoke exposure.

Parent: When should I bring my child back or go to the emergency room?

Doctor: If your child has persistent noisy breathing even when calm, is struggling to breathe, has blue lips or face, is very sleepy or difficult to wake, or starts drooling and can’t swallow, please seek emergency care immediately.

Parent: Thank you, doctor. That helps me feel more prepared.

Doctor: You’re welcome. We’ll give you the steroid medication today and keep an eye on your child. If you notice any worrying signs, don’t hesitate to come back or call us.

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## **Apnea of Prematurity**

Apnea of prematurity is a breathing condition that affects babies born before 37 weeks gestation (preterm birth). Your baby’s body is still growing, and the parts that support breathing are still developing. So, your baby isn’t quite ready to breathe in a normal rhythm yet. Instead, they have periods where they don’t breathe (apnea). Providers call these periods apneic spells.

It can feel very scary to think your baby isn’t breathing right. But this condition is common among preterm babies. And providers know how to treat it. Your baby needs to spend some time in the neonatal intensive care unit (NICU). Providers monitor your baby’s vital signs and give them treatments to support breathing. Your baby can likely go home once they can breathe normally without treatments.

### **Apnea of prematurity causes**

Apnea of prematurity occurs because your baby’s airways and center for breathing in the brain aren’t fully developed yet. These parts of your baby’s body work together to allow them to breathe. Providers divide apnea in newborns into three main types according to what’s causing the breathing pauses:

* Obstructive
* Central
* Mixed

#### **Obstructive apnea**

This is when there’s a blockage in your baby’s airways. These passages carry oxygen-rich air into your baby’s lungs and remove carbon dioxide from their body. Your baby’s airways need to stay open wide enough to let air pass through.

But your baby’s airways (typically, the part that passes through their neck) may not be developed enough to stay open all the time. This can cause breathing pauses.

#### **Central apnea**

This is when your baby’s brainstem (the center in the brain responsible for breathing) doesn’t send out certain signals when expected. Normally, your baby’s brainstem sends signals to your baby’s respiratory muscles (like their diaphragm and intercostal muscles). These muscles help your baby’s lungs pull in and push out air so they can breathe.

But if your baby’s brainstem isn’t fully developed yet, it’s not ready to send out signals in a predictable or reliable way. This can lead to pauses in breathing.

**Mixed apnea**

This is when your baby’s airways and brainstem aren’t working as expected. It’s the most common type of apnea among preterm infants.

### **Symptoms of apnea of prematurity**

Apnea of prematurity signs and symptoms include:

* Pauses in breathing that last 15 to 20 seconds or longer
* Slow heart rate (bradycardia)
* Low blood-oxygen level (hypoxemia), which can cause your baby’s skin to look blue, white, yellow-gray or gray — especially the lips and/or tongue (cyanosis)

Your baby may have breathing pauses that are shorter than 15 seconds. Providers consider these symptoms of apnea if they occur along with bradycardia and/or hypoxemia.

It’s important to distinguish these pauses from periodic breathing. Periodic breathing involves short pauses (less than 10 seconds) with no bradycardia or low oxygen level, followed by rapid breathing, and is considered normal.

**Diagnosis and Tests**

Healthcare providers diagnose apnea of prematurity in the neonatal intensive care unit (NICU). This is an area of the hospital for babies who need extra care and medical attention. If your baby is born preterm, they may need to spend some time in the NICU before they can go home.

While your baby is in the NICU, providers monitor their:

* Heart rate
* Breathing
* Blood-oxygen level

Monitors go off if your baby’s heart rate drops or their breathing pauses for a certain amount of time. Data from these monitors — along with nurses’ observations — help providers diagnose apnea.

Apnea in newborns is sometimes a sign of other medical conditions. These include metabolic disorders, brain bleeds and infections. Providers check your baby for signs and symptoms of other medical conditions. They rule out all other causes before diagnosing your baby with apnea of prematurity.

An apnea of prematurity diagnosis means your baby’s breathing pauses are due to early birth. Their nervous system and respiratory system aren’t fully developed yet. So, your baby needs a little extra help with breathing until their body is ready to handle things on its own.

## **Management and Treatment**

Apnea of prematurity treatment typically includes one or both of the following:

* Medications (usually caffeine citrate)
* Breathing support

**Caffeine for apnea of prematurity**

Caffeine citrate is a medication providers use to treat apnea of prematurity. Just like your morning coffee gives you a jolt, this medicine stimulates your baby’s nervous system. This “wake-up call” helps your baby’s brainstem and nerve cells send signals that regulate breathing.

Caffeine helps your baby have fewer apneic spells. It also helps shorten these pauses in breathing.

#### **Breathing support**

Your baby’s upper airway (pharynx and larynx) might need a little help staying open. If this is the case, providers often use continuous positive airway pressure (CPAP). With CPAP, your baby is still breathing on their own. But tiny prongs in their nose send air through their airway. This lowers the risk of breathing pauses from airway blockages.

If your baby continues to have severe apneic spells, they may need mechanical ventilation. This means a machine (a ventilator) does the work of breathing for your baby until their body can take over

## Apnea of Prematurity (AOP) Treatment Drugs and Their Side Effects

Apnea of Prematurity is a common condition in preterm infants characterized by pauses in breathing due to immature respiratory control. Pharmacological treatment primarily involves respiratory stimulants, especially methylxanthines.

## 1. Caffeine Citrate

* Mechanism: Stimulates the central nervous system, enhancing respiratory drive by improving brainstem signaling to respiratory muscles.
* Dosage: Typical loading dose is 20 mg/kg (oral or IV), followed by maintenance doses of 5-10 mg/kg daily.
* Efficacy: Preferred treatment due to effectiveness, wide therapeutic window, and ease of administration. Reduces apnea episodes and need for mechanical ventilation.
* Side Effects:
  + Generally well tolerated.
  + Possible side effects include tachycardia (increased heart rate), feeding intolerance, jitteriness, and irritability.
  + Rarely, may cause seizures or arrhythmias at high doses.
* Monitoring: Plasma levels usually not required due to predictable pharmacokinetics.

## 2. Theophylline / Aminophylline

* Mechanism: Similar to caffeine; methylxanthines that stimulate respiratory centers.
* Dosage: Theophylline has a narrower therapeutic window; aminophylline is often used intravenously as a theophylline source.
* Efficacy: Effective but less favored due to more frequent dosing and need for plasma level monitoring.
* Side Effects:
  + Tachycardia (more pronounced than caffeine).
  + Feeding intolerance, irritability, tremors.
  + Risk of toxicity including seizures and arrhythmias.
* Monitoring: Requires frequent plasma level monitoring due to narrow therapeutic index.

## 3. Doxapram

* Mechanism: Respiratory stimulant acting on carotid body chemoreceptors and central nervous system.
* Use: Considered a third-line agent, used when caffeine and CPAP fail.
* Efficacy: Some studies show improved apnea control; used cautiously.
* Side Effects:
  + Hypertension, irritability, tachycardia.
  + Potential for serious side effects; requires close monitoring.
* Notes: Not widely used due to limited safety data.

## 4. Supportive Treatments

* Continuous Positive Airway Pressure (CPAP): Helps keep airways open and reduce obstructive apnea components.
* Mechanical Ventilation: For severe cases where spontaneous breathing is inadequate.
* Non-pharmacological: Tactile stimulation during apnea episodes, optimizing positioning, and treating underlying conditions

## Genomic Data and Genetic Basis of Apnea of Prematurity (AOP)

* Strong Genetic Component:  
  Multiple studies, including twin studies, indicate a high heritability of AOP, with genetic factors accounting for approximately 87% of the variance among same-gender twins. In particular, genetic influence is stronger in male twins (up to 99%) compared to females (around 78%).
* Twin Studies:  
  Research comparing monozygotic (identical) and dizygotic (fraternal) twins shows that if one identical twin has AOP, the other twin has an 87% chance of also having it, whereas this concordance is 62% in fraternal twins, supporting a significant genetic predisposition.
* Genetic Polymorphisms:  
  Specific gene polymorphisms, especially in adenosine receptor genes (A1, A2A, A2B), have been linked to both the risk of developing AOP and variability in response to caffeine therapy, the main pharmacologic treatment.
  + Some polymorphisms in the A2A receptor gene are associated with increased risk of apnea.
  + Variants in A1 receptor gene correlate with better response to caffeine treatment.
  + Polymorphisms in A2A and A2B receptors also influence the incidence of bronchopulmonary dysplasia (BPD), a common comorbidity

## **Outlook / Prognosis**

Apnea of prematurity is a temporary condition. Symptoms improve as your baby grows. Your baby may need to spend several weeks — or even a couple of months — in the NICU. They’ll receive care not just for their breathing but also for any other issues related to preterm birth.

Providers will tell you how long your baby needs to stay in the NICU. They’ll also explain what needs to happen for your baby to safely head home.

In general, providers identify when it’s safe to stop giving your baby treatments. Then, after stopping treatment, they monitor your baby to see how their body responds. Your baby needs to go a certain number of days without apnea symptoms in order for providers to send them home. The length of this “observation period” depends on how early your baby was born and their overall health.

Most babies don’t need continued monitoring with devices at home. But if your baby does need monitoring, providers will explain exactly what’s involved. Ask if anything is unclear or you’re concerned about caring for your baby at home. Providers will make sure you get the information you need to feel comfortable with this transition.

## Diagnostic Considerations

Apnea of prematurity (AOP) is a diagnosis of exclusion. For many diseases in preterm infants, apnea is a presenting symptom. The causes of these diseases are different when the differential diagnosis occurs shortly after birth compared with later in the patient's hospital stay. Other etiologies must be sought before drug and/or ventilatory therapies for apnea of prematurity are started.

## Conditions associated with apnea

Shortly after birth, apnea can be a manifestation of several types of conditions. Consider the following:

* [Respiratory distress syndrome](https://emedicine.medscape.com/article/976034-overview) and other pulmonary conditions
* Infections (eg, congenital pneumonia, bacteremia, meningitis, fetal or neonatal inflammatory response syndrome)
* Hypoglycemia and other metabolic diseases
* CNS pathology (eg, trauma, intracranial hemorrhage, anoxia and/or ischemia, stroke)

The aforementioned brain insults may be noticed because patients may have seizures and/or associated apnea. [[63](javascript:void(0);), [72](javascript:void(0);)] Some of the diseases cited above also occur relatively late during the hospitalization of prematurely born infants, but the signs and symptoms may or may not include apnea.

Intraventricular hemorrhage and posthemorrhagic hydrocephalus without seizures increases the frequency of apnea in preterm infants. [[62](javascript:void(0);), [73](javascript:void(0);)]

## Other conditions associated with apnea in preterm infants during their hospitalization

*Nosocomial bacterial or fungal infection*

Apnea, bradycardia, and desaturations are presenting symptoms of nosocomial infections caused by bacteria and fungi or viral agents. [[74](javascript:void(0);), [75](javascript:void(0);)]

In a study of 9 infants in an NICU who had respiratory syncytial viral infection, 8 had apnea as a manifestation of disease. [[76](javascript:void(0);)]

Apnea is also common in preterm infants with *Ureaplasma urealyticum* infection. [[77](javascript:void(0);)]

*Necrotizing enterocolitis*

In addition, apnea is one of several signs and symptoms associated with the onset of [necrotizing enterocolitis](https://emedicine.medscape.com/article/977956-overview). [[78](javascript:void(0);)]

*Systemic inflammation*

It is well known to the caregivers in the NICU that premature infants who present with a cluster of multiple episodes of apnea, bradycardia, and desaturations could be showing signs of developing sepsis. During the sepsis, the inflammatory mediators and cytokines play a major role in the development of apnea bradycardia and desaturations. Lipopolysaccharide (LPS) attenuates the sensitivity of the carotid body, which leads to the development of apnea, bradycardia, and desaturations. [[79](javascript:void(0);)]

*Exposure to magnesium*

The administration of magnesium to prevent seizures in preeclampsia and tocolysis of preterm labor has been associated with hypoventilation, apnea, and other adverse effects in preterm infants. [[80](javascript:void(0);)] Hypermagnesemia during parenteral nutrition has also been a cause of apnea. [[81](javascript:void(0);)]

*Anemia*

Disagreement exists regarding the role of clinically significant anemia in the development of apnea among convalescing preterm infants. Westkamp and colleagues reported that blood transfusions lowered heart and respiratory rates but had little effect on apnea of prematurity. [[82](javascript:void(0);)] Bell and associates conversely found that liberal versus restrictive blood transfusion significantly reduced apnea. [[83](javascript:void(0);)] The interest in the adverse neurodevelopmental outcomes observed in preterm infants with anemia and low iron status emphasizes neonatology-related awareness of the problem. [[84](javascript:void(0);)]

*Anemia, apnea of prematurity, and blood transfusion*

The etiology of apnea in a premature infant is a multifactorial; however, it is a common practice to transfuse packed red blood cells in an infant who is anemic and having multiple episodes of apnea and bradycardia. Many studies have shown conflicting results, as many variables play a role in the pathogenesis of apnea of prematurity. One retrospective study found that packed red blood cell transfusion reduces the number of apneic events in premature infants, with many limitations. [[85](javascript:void(0);)] When anemia of prematurity and apnea of prematurity occur in conjunction, clinicians are more inclined to transfuse at higher hematocrit levels in affected infants than in infants without apnea of prematurity. Zagol et al observed more cardiopulmonary events during the 72 hours period prior to the blood transfusion compared to 72 hours after the transfusion. [[86](javascript:void(0);)] Clinicians should judge the merits of transfusion.

*Surgery*

Postoperative apnea occurs in preterm infants, particularly those whose intraoperative pain relief involved [general anesthesia](https://emedicine.medscape.com/article/1271543-overview). [[87](javascript:void(0);), [88](javascript:void(0);)]

Additional research is required to determine whether spinal anesthesia, different analgesic agents, or caffeine can mitigate morbidity associated with apnea after surgery in prematurely born infants. [[89](javascript:void(0);), [90](javascript:void(0);)]

*Immunization*

Apnea transiently increases or recurs in hospitalized preterm infants after immunization. The increase in apnea has been attributed to the whole-cell pertussis component. [[91](javascript:void(0);)] Investigators have observed reduced morbidity with newer vaccines that contain acellular pertussis. [[92](javascript:void(0);), [93](javascript:void(0);)] Some reports still identified clinically significant apnea and other adverse events. [[94](javascript:void(0);), [95](javascript:void(0);)]

*Eye examination*

Premature infants routinely undergo screening eye evaluations for retinopathy of prematurity in the NICU. Note that approximately 19% to 25% of infants experience an increased number of apneic spells after these routine eye examinations. [[96](javascript:void(0);)]

*Gastroesophageal reflux*

As stated earlier, controversy exists regarding the role of gastroesophageal reflux (GER) as a causative factor in apnea of prematurity. One perspective is that the two conditions are related. [[56](javascript:void(0);), [97](javascript:void(0);)] Laryngeal edema identified during fiberoptic laryngeal endoscopy has been associated with GER, and antireflux surgery has dramatically reduced apnea in preterm infants at highest risk. [[98](javascript:void(0);), [99](javascript:void(0);), [100](javascript:void(0);)]

Past research failed to reveal a temporal relationship between GER and apnea of prematurity. [[51](javascript:void(0);), [52](javascript:void(0);), [101](javascript:void(0);), [102](javascript:void(0);), [103](javascript:void(0);)]

Therefore, the NICHD Review Group on Apnea of Prematurity has called for additional investigations with rigorous research designs.

*Skin-to-skin contact, or kangaroo care*

Skin-to-skin contact, or kangaroo care, for preterm infants has been associated with an increased occurrence of apnea, bradycardia, and desaturation; this appears to be unrelated to hyperthermia. [[104](javascript:void(0);), [105](javascript:void(0);)] The observation suggests that obstructive events may occur during skin-to-skin contact. These findings call attention to the importance of environmental hyperthermia as a cause of apnea in preterm infants. [[106](javascript:void(0);), [107](javascript:void(0);)]

No adverse events appear to occur during kangaroo care. [[108](javascript:void(0);)]

The disparity among the reported studies may be related to the specific practice of skin-to-skin care in a particular NICU or the validity of monitoring during skin-to-skin contact. [[109](javascript:void(0);)]

## Differential Diagnoses

* [Anemia of Prematurity](https://emedicine.medscape.com/article/978238-overview)
* [Neonatal Sepsis](https://emedicine.medscape.com/article/978352-overview)
* [Pediatric Respiratory Failure](https://emedicine.medscape.com/article/908172-overview)
* [Respiratory Syncytial Virus Infection](https://emedicine.medscape.com/article/971488-overview)

## Epidemiology

### United States data

Although not always apparent, apnea of prematurity is the most common problem in premature neonates. Approximately 70% of babies born before 34 weeks of gestation have clinically significant apnea, bradycardia, or O2 desaturation during their hospital stay. The more immature the infant, the higher his or her risk of apnea of prematurity. Apnea may occur during the postnatal period in 25% of neonates who weighed less than 2500 g at birth and in 84% of neonates who weigh less than 1000 g.

Carlo and Barrington showed that apnea may begin on the first day of life in neonates without respiratory distress syndrome. [[58](javascript:void(0);), [59](javascript:void(0);)] However, apnea of prematurity is always a diagnosis of exclusion. Many diseases manifest with apnea on the day of birth; examples are intrapartum magnesium exposure, systemic infections or the fetal inflammatory response syndrome, pneumonia, intracranial pathology, seizures, [hypoglycemia](https://emedicine.medscape.com/article/921936-overview), and other metabolic disturbances.

Approximately 50% or more of surviving infants who weighed less than 1500 g at birth have episodes of apnea that must be managed with pharmacologic intervention or ventilatory support. Mixed apnea accounts for about 50% of all cases of apnea in premature neonates; about 40% are central apneas, and 10% are obstructive apneas. [[60](javascript:void(0);)] These percentages vary in different reports. In 50% of all apneic episodes, an obstructive component precedes or follows central apnea, which leads to mixed apnea.

### International data

To the authors' knowledge, no investigators have compared the incidence of apnea of prematurity in the United States with those of other countries.

### Race-, sex-, and age-related demographics

The authors know of no systematic, prospective clinical study that has been conducted to evaluate the role of a person's race/ethnic background or sex on the incidence of apnea of prematurity.

A young gestational age at birth is associated with an increased incidence of apnea of prematurity. The age at which apnea of prematurity resolves depends on several factors. The mean time for severe apnea of prematurity to resolve is approximately 43 weeks after conception, but a prolonged duration of risk is not uncommon. [[14](javascript:void(0);)]

In one report, about 6-22% of babies with a very low birth weight had apnea at term. [[61](javascript:void(0);)] Approximately 91% of premature neonates had apnea of longer than 12 seconds at the time of hospital discharge. Of these babies, 31% also had bradycardia, and 6.5% required prolonged hospitalization because of the severity of their apnea and bradycardia.

These findings show that apnea of prematurity does not resolve at term in many low birth weight infants and that it may persist for some time after hospital discharge.

## Procedures

Several studies may reveal diagnostic findings in selected infants. These include fiberoptic examination of the larynx through the nose during spontaneous breathing, direct laryngoscopy, and bronchoscopy (which is usually performed with the patient under anesthesia).

Emergency or scheduled tracheostomy may be used to manage severe airway obstruction caused by a number of conditions. Tracheostomy might occur after the airway is stabilized by using endotracheal intubation. Jaw-distraction surgery has been used to avoid tracheostomy in neonatal conditions (eg, Robin sequence) that involve severe micrognathia as a component of malformation.

Doctor: Hello, I want to talk with you about your baby’s breathing. Since your baby was born prematurely, they have a condition called apnea of prematurity. This means your baby sometimes stops breathing for short periods because their respiratory system is still immature.

Parent: That sounds scary. How long do these pauses last, and how often do they happen?

Doctor: Apnea is defined as a pause in breathing lasting 20 seconds or more, or shorter pauses that cause the heart rate to slow down or oxygen levels to drop. These episodes can happen frequently, especially in babies born before 35 weeks of gestation. It usually starts a couple of days after birth and tends to improve as the baby matures, often resolving by around 37 to 44 weeks postmenstrual age.

Parent: How do you monitor these breathing pauses?

Doctor: We use cardiorespiratory monitors that track your baby’s breathing, heart rate, and oxygen levels continuously. If the baby stops breathing or their heart rate drops below certain thresholds, the monitor alerts us so we can intervene quickly.

Parent: What treatments are available? Will my baby need medicine?

Doctor: Treatment depends on how severe and frequent the apnea episodes are. For mild cases, supportive care like keeping your baby’s environment warm, positioning the head and neck properly, and ensuring clear nasal passages can help. For more significant apnea, we use respiratory stimulants like caffeine, which helps the brain regulate breathing better. If needed, nasal CPAP (continuous positive airway pressure) can keep the airway open. In rare cases, if these treatments don’t work, more intensive support like mechanical ventilation may be necessary.

Parent: Are there side effects from the caffeine or other treatments?

Doctor: Caffeine is generally safe and well tolerated. Some babies may experience mild side effects like increased heart rate or jitteriness, but serious problems are rare. We monitor your baby closely throughout treatment.

Parent: How long will my baby need treatment?

Doctor: Usually, treatment continues until your baby’s breathing control matures, which is typically around 34 to 35 weeks corrected gestational age or when apnea episodes have resolved for several days. Most babies outgrow apnea of prematurity as their nervous system develops.

Parent: What should I watch for or be concerned about?

Doctor: If you notice your baby turning blue, becoming very limp, or if the apnea episodes become more frequent or prolonged, please notify the medical team immediately. We are here to support you and your baby through this.

Parent: Thank you, doctor. It helps to understand what’s going on and what to expect.

Doctor: You’re welcome. We’ll keep monitoring your baby closely and adjust treatment as needed. Please feel free to ask any questions anytime.

REFERENCES

[Apnea of Prematurity (AOP) Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/apnea-of-prematurity#what-is-apnea-of-prematurity)

<https://emedicine.medscape.com/article/974971-workup#c7>