CASE FIVE

Short case number: 3_20_5
Category: Children & Young People / Respiratory & ENT Systems
Discipline: Paediatrics Medicine
Setting: Emergency Department
Topic: The child with stridor

Case

It is 3 am; Lily Griffen, who is 4 years old presents with difficulty breathing. She has been unwell for 2 days with an upper respiratory tract infection and fever to 38^0C. Last night she developed a barking cough. She has awoken with noisy difficult breathing.
Vital signs: Temp 38^0C, HR 100 bpm, RR 20min, BP 75/50, SaO_2 – 95% [room air]
Observation: Stridorous breathing with use of accessory muscles at rest.

Questions

1. Outline your immediate management of Lily. Summarise the clinical indications for use of nebulised adrenalin and/or corticosteroids in acute laryngotracheobronchitis. [croup]
2. What is stridor? Outline the underlying pathophysiology.
3. Lily has now settled following acute treatment. What are the key features of history that you would assess to determine the cause of Lily's problem?
4. What are the key features of physical examination that you would assess and why?
5. Summarise the key pathological and clinical features of the following causes of stridor; acute larynogtracheobronchitis, laryngomalacia, sub-glottic stenosis, and sub-glottic haemangioma.
6. Once Lily has been stabilised you decide to revise your respiratory physiology, summarise the pressure changes that occur in the lungs and airways during inspiration and expiration and how affected by airway obstruction.
7. Lily's mother asks *"if Lily gets worse again should I sit with her in a steamy bathroom?"* What would you explain to her?
8. Occasionally children experience recurrent episodes of croup. Outline the clinical features that differentiate recurrent [spasmodic] croup from viral croup.

Suggested reading:

- South M, Isaacs D editors. Practical Paediatrics. 7th edition. Edinburgh: Churchill Livingstone; 2012.
- Thomson K, Tey D, Marks M, editors. Paediatric Handbook. Staff of the Royal Children's Hospital Melbourne, Australia. 8th edition. Chichester: Wiley-Blackwell; 2009.

ANSWERS

1. Outline your immediate management of Lily. Summarise the clinical indications for use of nebulised adrenalin and/or corticosteroids in acute laryngotracheobronchitis. [croup]

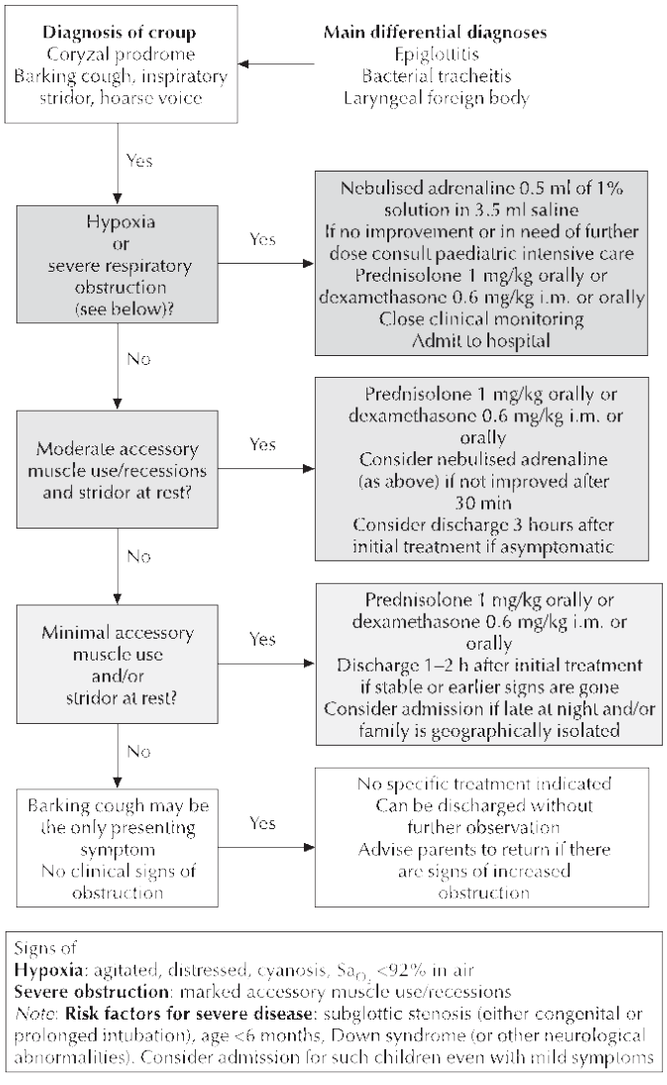

Fig. 36.3 Management of croup

2. What is stridor? Outline the underlying pathophysiology.

Stridor is a harsh, high pitched noise heard predominantly during inspiration. The presence of an added respiratory sound implies an obstruction to the free flow of gas through the airway tree. Flow limitation in a compliant tube, such as the airways, is accompanied by fluttering of the walls, which occurs to conserve energy when driving pressure exceeds the pressure required to produce the maximal flow. The fluttering of the walls produces a respiratory noise. When this phenomenon occurs during inspiration, the resultant noise is known as stridor, and when it occurs during expiration, the noise is known as wheeze

3. Lily has now settled following acute treatment. What are the key features of history that you would assess to determine the cause of Lily's problem?

- *Age of onset.* A stridor present from the first few days of life suggests a congenital or structural cause
- *Speed of onset of symptoms.* Infective causes such as croup tend to come on quickly; however, most cases of congenital or structural stridor commonly first present following a viral upper respiratory illness

- *Progression of stridor.* Stridor increasing in severity over weeks to months suggests a progressive lesion, such as subglottic haemangioma
- *Effect of body position.* Stridor that is worse when lying supine is seen commonly with laryngomalacia
- *Presence of an expiratory component.* This suggests a more severe obstruction that limits flow during expiration as well as during inspiration
- *Quality of voice.* While the voice is frequently normal, a hoarse voice would suggest a vocal cord lesion
- *Other medical conditions that could contribute to the pathogenesis or presentation:* febrile illness, ex-premature infant, gastro-oesophageal reflux, cutaneous haemangiomas, Möbius syndrome (a very rare syndrome characterized by congenital palsy of the external rectus and facial muscles, usually bilateral, associated with paralysis of the sixth and seventh nerves)

4. What are the key features of physical examination that you would assess and why?
Avoid any examination that may distress the child.

5. Summarise the key pathological and clinical features of the following causes of stridor; acute larynogtracheobronchitis, laryngomalacia, sub-glottic stenosis, and sub-glottic haemangioma.

ACUTE LARYNGOTRACHEOBRONCHITIS (CROUP)
Typically a disease of toddlers. Usually begins with symptoms of URTI and progresses to typical croup over 1-2 days. The typical barking cough usually begins at night or in the early hours of the morning. As the disease progresses, stridor may be heard on exertion initially. If the subglottic obstruction progresses further, stridor may be heard at rest and an expiratory component may be heard (the typical cough continues).

If the degree of obstruction continues to worsen, the stridor may become more difficult to hear and the child may become distressed and restless. Cough may be absent at this stage. The lack of stridor comes about because the amount of air moving through the obstructed airway is not sufficient to generate the noise (see above). The distress and restlessness are most likely to be due to hypoxia and signal impending complete respiratory obstruction.

The viral illness generally lasts 7-10 days, but the typical croupy cough usually only occurs on the first 2-3 nights.

LARYNGOMALACIA
Laryngomalacia, which is sometimes known as infantile larynx, is the most common cause of persistent stridor. It is not well named, as the larynx and vocal cords are actually normal. The supraglottic tissues appear as if they are too large for the size of the glottis and narrow the glottic aperture during inspiration instead of the more normal widening during inspiration. This can occur in a number of ways, the most common being:

- a long, curled (sometimes called omega-shaped) epiglottis collapsing during inspiration so that the lateral walls touch, restricting the free passage of air
- floppy arytenoid processes prolapsing into the glottic aperture during inspiration
- a long epiglottis collapsing against the posterior pharyngeal wall during inspiration.

In more severe cases combinations of these mechanisms may be responsible for the inspiratory obstruction.

Laryngomalacia classically produces a cog-wheel stridor, with no expiratory component. The cog-wheel nature to the stridor is likely to come from vibrations of the supraglottic tissues as the degree of obstruction varies during the inspiratory effort. The stridor may be worse when the infant is lying supine, although this feature is not always seen. More severe obstruction may be associated with suprasternal and sternal retraction during inspiration.

Laryngomalacia is usually a benign condition that does not require any treatment, except to reassure the parents that this is the case. Severe laryngomalacia may be associated with failure to thrive and gastro-oesophageal reflux

SUB-GLOTTIC STENOSIS

Subglottic stenosis refers to a narrowing in the upper part of the trachea, immediately below the glottis. This narrowing may be congenital or acquired. Congenital subglottic stenosis occurs typically at the level of, and involves, the cricoid cartilage. The tracheal epithelium typically appears normal but the cross-sectional area of the lumen is reduced and typically does not vary with respiration. Acquired subglottic stenosis usually results from trauma and is most commonly seen in premature infants who required intubation. Older infants and children who require prolonged intubation are also at risk. Here the tracheal epithelium is more likely to be replaced by scar tissue

Subglottic stenosis may present soon after birth or the presentation may be delayed. The stenosis, either congenital or acquired, is usually not progressive but the degree of obstruction may increase, e.g. as the child's activity levels increase or at times of respiratory infection. The typical presentation is with stridor, particularly at times of respiratory infection. If the obstruction is severe enough, the stridor may have an expiratory component and be associated with suprasternal and sternal retractions

Many cases of subglottic stenosis do not require treatment and most will improve with growth.

SUB-GLOTTIC HAEMANGIOMA
Sub-glottic haemangiomas are typical haemangiomas occurring in the submucosal layer of the tracheal wall. As with other haemangiomas, they enlarge during the first year of life and typically present with increasing stridor and inspiratory obstruction. The stridor is rarely present at birth and most come to attention around 4-6 months of age. As the obstruction becomes worse, the stridor develops an expiratory component and is associated with sternal and suprasternal retractions. Approximately 50% of subglottic haemangiomas are associated with cutaneous haemangiomas, although the converse association is much less frequent.

6. **Once Lily has been stabilised you decide to revise your respiratory physiology, summarise the pressure changes that occur in the lungs and airways during inspiration and expiration and how affected by airway obstruction.**

During breathing, there are pressure gradients between the airway opening and the alveoli. Inspiration occurs when alveolar pressure is lowered below atmospheric pressure and air flows in to equalize the pressures. At the onset of expiration, alveolar pressure exceeds atmospheric pressure and air flows out. There are also pressure gradients across the airway wall and these tend to alter airway calibre. The pressure around the extrathoracic airways, that is, those above the thoracic inlet, is atmospheric, while the pressure around the intrathoracic airways essentially is equal to the pleural pressure. The pressure gradients across the airway wall during inspiration means that there is a net force tending to narrow the extrathoracic airways and to dilate the intrathoracic airways. During expiration, the direction of the forces is opposite, resulting in a tendency to narrow intrathoracic airways and dilate extrathoracic airways.

As stridor is an inspiratory noise, the predominant site of obstruction (the site responsible for the flow limitation) is generally in the extrathoracic airways. Stridor with an expiratory component, that is, where the noise can also be heard at the beginning of expiration, can result from either a severe obstruction producing flow limitation during expiration as well, or from a lesion that extends into the intrathoracic airways.

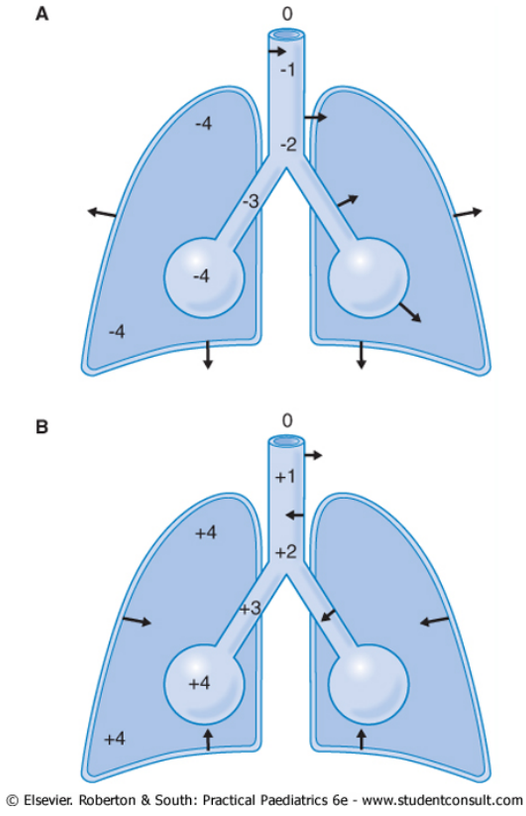

Figure: The distribution of pressures throughout the respiratory system during (**A**) inspiration and (**B**) expiration. Atmospheric pressure is shown as zero. During inspiration, the expansion of the thorax results in pleural pressure falling below atmospheric. This relatively negative pressure is transmitted to the alveoli and a pressure gradient is established between the airway opening and the alveoli. Gas flows into the lungs along this pressure gradient. The pressure outside the airways is essentially pleural pressure and results in net forces that tend to expand intrathoracic airways and to collapse the extrathoracic trachea. As shown in B, the pressure gradients are opposite during expiration

7. Lily's mother asks *"if Lily gets worse again should I sit with her in a steamy bathroom?"* What would you explain to her?

There is a widespread belief that exposing these children to steam, especially by steaming up the home bathroom, helps relieve stridor. There is no evidence to support this treatment. The only benefit that is likely to come from sitting with the child in a steamy bathroom is from sitting quietly with the child and not from the steam

8. Occasionally children experience recurrent episodes of croup. Outline the clinical features that differentiate recurrent [spasmodic croup] from viral croup.

Some children suffer recurrent episodes of croup, frequently without the preceding viral prodrome usually seen in acute viral croup. Typically these children are well when they go to bed and wake in the early hours of the morning with a barking cough and stridor. Fever is unusual in this form of croup. The same viruses as found in acute viral croup may be found in the upper airways of children with spasmodic croup, although the relationship between the viruses and the symptoms is less clear. Frequently children with recurrent croup have a family history of atopy and asthma or have asthma themselves. This, together with the uncertain relationship between the clinical symptoms and the presence of a virus, have led to the concept that spasmodic croup maybe a manifestation of upper airway hyper responsiveness. There are no direct data to support or refute this hypothesis