Restrictive Lung Disease: An Educational Guide for Hospital Clinicians

Overview of Restrictive Lung Disease

Restrictive lung disease (RLD) is characterized by reduced lung volumes and decreased lung compliance, leading to difficulty in fully expanding the lungs during inspiration. This results in a reduced total lung capacity (TLC), forced vital capacity (FVC), and forced expiratory volume in 1 second (FEV1), with a preserved or increased FEV1/FVC ratio (>0.7). RLD can be caused by intrinsic lung diseases (e.g., interstitial lung disease) or extrinsic factors (e.g., pleural disease, neuromuscular disorders). In the hospital setting, RLD often presents as an acute exacerbation of a chronic condition or as a new diagnosis requiring urgent management. This guide provides a comprehensive overview of RLD, including causes, clinical presentation, diagnostic workup, and hospital management, with tables and clinical scenarios for practical application.

Causes of Restrictive Lung Disease

Intrinsic Lung Diseases (Parenchymal Causes):

- Interstitial Lung Diseases (ILD):
  - **Idiopathic Pulmonary Fibrosis (IPF):** Most common ILD, characterized by progressive fibrosis.
  - **Sarcoidosis:** Granulomatous inflammation, often with hilar lymphadenopathy.
  - **Hypersensitivity Pneumonitis (HP):** Immune response to inhaled antigens (e.g., mold, bird dander).
  - **Connective Tissue Disease-Associated ILD (CTD-ILD):** E.g., rheumatoid arthritis, systemic sclerosis.
  - **Drug-Induced ILD:** E.g., amiodarone, nitrofurantoin, chemotherapy (bleomycin).
  - **Pneumoconioses:** Occupational exposures (e.g., asbestosis, silicosis).
  - **Radiation Fibrosis:** Post-radiation therapy (e.g., breast cancer, lymphoma).
  - **Acute Interstitial Pneumonia (AIP):** Rapid-onset ILD, often mimicking ARDS.
- Extrinsic Causes (Non-Parenchymal Causes):
  - Pleural Diseases:
    - **Pleural Effusion:** Fluid accumulation (e.g., CHF, malignancy, infection).

        - **Pleural Thickening/Fibrosis:** Post-infection, asbestos exposure.
        - **Pneumothorax:** Air in pleural space, restricts lung expansion.
    - Chest Wall Disorders:
        - **Kyphoscoliosis:** Severe spinal deformity restricts chest expansion.
        - **Obesity Hypoventilation Syndrome (OHS):** Excess weight limits chest wall movement.
        - **Ankylosing Spondylitis:** Spinal and costovertebral joint rigidity.
    - Neuromuscular Disorders:
        - **Amyotrophic Lateral Sclerosis (ALS):** Progressive respiratory muscle weakness.
        - **Myasthenia Gravis (MG):** Autoimmune neuromuscular junction disorder.
        - **Guillain-Barré Syndrome (GBS):** Acute demyelinating neuropathy.
        - **Diaphragmatic Paralysis:** Unilateral/bilateral diaphragm dysfunction.
    - Other:
        - **Ascites:** Abdominal distension compresses diaphragm (e.g., cirrhosis).
        - **Pregnancy:** Uterine enlargement restricts diaphragm movement in late stages.

Clinical Presentation

Symptoms:

- **Dyspnea:** Exertional initially, progressing to rest (due to reduced lung volumes).
- **Dry Cough:** Common in ILD (e.g., IPF, sarcoidosis).
- **Fatigue:** Due to chronic hypoxemia and increased WOB.
- **Chest Pain:** If pleural involvement (e.g., pleuritis in CTD-ILD).
- Systemic Symptoms:
    - Weight loss, fever (sarcoidosis, CTD-ILD).
    - Joint pain, rash (CTD-ILD, e.g., RA, SLE).
    - Raynaud’s phenomenon (systemic sclerosis-ILD).

Physical Exam:

- Respiratory:
    - Tachypnea (RR >20/min), shallow breathing.
    - **Fine inspiratory crackles (ILD, e.g., IPF:** "Velcro" crackles at bases).
    - Decreased breath sounds (pleural effusion, pneumothorax).
- Chest Wall:
    - Spinal deformity (kyphoscoliosis).

    - Obesity (OHS).
    - Reduced chest expansion (ankylosing spondylitis).
- Neuromuscular:
    - Muscle weakness (ALS, MG, GBS).
    - **Paradoxical breathing (diaphragmatic paralysis:** abdomen moves inward on inspiration).
- Systemic:
    - Clubbing (IPF, asbestosis).
    - Lymphadenopathy, erythema nodosum (sarcoidosis).
    - Joint swelling, rash (CTD-ILD).
    - Ascites, jaundice (cirrhosis-related restriction).

Diagnostic Workup

Pulmonary Function Tests (PFTs):

- **Hallmark:** Reduced TLC (<80% predicted), reduced FVC, preserved FEV1/FVC ratio (>0.7).
- Patterns:
    - **ILD:** Reduced FVC, TLC, DLCO (diffusing capacity for carbon monoxide).
    - **Neuromuscular:** Reduced FVC, normal DLCO (unless coexisting lung disease).
    - **Pleural Disease:** Reduced FVC, TLC, normal DLCO.

Labs:

- **CBC:** Anemia (chronic disease in CTD-ILD), eosinophilia (HP, drug-induced).
- **CMP:** Hypoalbuminemia (cirrhosis), elevated Cr (scleroderma renal crisis).
- Inflammatory Markers:
    - ESR, CRP (elevated in CTD-ILD, sarcoidosis).
- Autoimmune Serologies:
    - ANA, RF, anti-CCP (RA-ILD).
    - Anti-Scl-70, anti-centromere (systemic sclerosis-ILD).
    - Anti-Ro/La (Sjögren's-ILD).
    - ACE levels (sarcoidosis, supportive but non-specific).
- Hypersensitivity Panel:
    - Specific IgG to antigens (e.g., avian proteins in bird fancier's lung).

Imaging:

- Chest X-Ray (CXR):
  - **ILD:** Reticular opacities, honeycombing (IPF), hilar lymphadenopathy (sarcoidosis).
  - **Pleural Disease:** Blunting of costophrenic angle (effusion), pleural thickening.
  - **Chest Wall:** Kyphoscoliosis, rib abnormalities.
- High-Resolution CT (HRCT):
  - **IPF:** Usual interstitial pneumonia (UIP) pattern—subpleural, basal-predominant reticulation, honeycombing.
  - **Sarcoidosis:** Perilymphatic nodules, upper lobe fibrosis, hilar lymphadenopathy.
  - **HP:** Ground-glass opacities, centrilobular nodules, mosaic attenuation (air trapping).
- **CTD-ILD:** Non-specific interstitial pneumonia (NSIP) pattern—ground-glass, reticulation.
- Echocardiogram:
  - Assess pulmonary hypertension (PH) secondary to chronic hypoxemia (e.g., RVSP >35 mmHg).

Other Tests:

Bronchoalveolar Lavage (BAL): Lymphocytosis in sarcoidosis/HP, eosinophilia in drug-induced ILD.

Lung Biopsy: Surgical (VATS) for UIP in IPF; transbronchial for sarcoidosis (non-caseating granulomas).

Pulmonary Artery Catheterization: If PH suspected (PA pressure >25 mmHg at rest).

Neuromuscular Assessment: EMG/NCS for ALS, GBS; anti-AChR antibodies for MG.

Hospital Management

General Principles:

- Supportive Care:
  - **Oxygen Therapy:** Titrate to SpO2 88-92% (avoid hyperoxia, e.g., SpO2 >95%).
  - **Pulmonary Rehabilitation:** If stable, to improve exercise tolerance.
  - **Nutrition Support:** Address malnutrition (albumin <3.0 g/dL), especially in OHS, cirrhosis.

- Monitoring:
  - **ABG:** Assess hypoxemia (PaO_2 <60 mmHg), hypercapnia ($PaCO_2$ >45 mmHg).
  - **SpO_2:** Continuous monitoring, target 88-92%.
  - **PFTs:** If feasible, to quantify restriction (TLC, FVC).
  - **6-Minute Walk Test (6MWT):** Assess functional capacity, desaturation (<88%).

Specific Management by Cause:

- Interstitial Lung Diseases (ILD):
  - IPF:
    - **Antifibrotics:** Pirfenidone 801 mg PO TID or nintedanib 150 mg PO BID (slows FVC decline).
    - **Oxygen:** Long-term oxygen therapy (LTOT) if PaO_2 <55 mmHg.
    - **Steroids:** Not routinely used (no benefit in IPF); consider in acute exacerbation (methylprednisolone 1 g IV daily x 3 days).
    - **Transplant:** Refer for lung transplant evaluation (age <70, no major comorbidities).
  - Sarcoidosis:
    - **Steroids:** Prednisone 20-40 mg PO daily if symptomatic (e.g., dyspnea, organ involvement).
    - **Immunosuppressants:** Methotrexate 15 mg PO weekly if steroid-resistant.
    - **Anti-inflammatories:** Hydroxychloroquine 200 mg PO BID for skin involvement.
  - CTD-ILD:
    - **Immunosuppression:** Prednisone 1 mg/kg/day PO + cyclophosphamide 2 mg/kg/day (Scleroderma-ILD).
    - **Biologics:** Rituximab 1 g IV q2 weeks (RA-ILD, refractory cases).
    - **Supportive:** Oxygen, infection prophylaxis (e.g., TMP-SMX for PCP).
  - HP:
    - **Antigen Avoidance:** Remove exposure (e.g., birds, mold).
    - **Steroids:** Prednisone 0.5-1 mg/kg/day PO, taper over 3-6 months.
  - Pleural Diseases:
    - Pleural Effusion:
      - **Thoracentesis:** Diagnostic (exudate vs. transudate), therapeutic if symptomatic.
      - **Underlying Cause:** Treat CHF (diuretics), infection (antibiotics), malignancy (chemotherapy).
      - **Pleurodesis:** If recurrent (e.g., talc slurry for malignant effusion).

        - Pneumothorax:
            - **Chest Tube:** For large (>2 cm) or symptomatic pneumothorax.
            - **Oxygen:** High-flow O2 to promote reabsorption.
            - **Surgery:** VATS pleurodesis if recurrent.
    - Chest Wall Disorders:
        - Kyphoscoliosis:
            - **Non-Invasive Ventilation (NIV):** BiPAP (e.g., IPAP 12 cmH2O, EPAP 4 cmH2O) for nocturnal hypoventilation.
            - **Surgical Correction:** If severe (Cobb angle >50°), consult orthopedics.
    - OHS:
        - **Weight Loss:** Nutrition consult, bariatric surgery if BMI >40 kg/m².
        - **NIV:** BiPAP for sleep-disordered breathing (e.g., IPAP 14 cmH2O, EPAP 6 cmH2O).
    - Neuromuscular Disorders:
        - ALS/GBS/MG:
            - **Monitor FVC:** Intubate if FVC <15 mL/kg or <1 L, or if rapid decline (>30% in 24h).
            - **NIV:** BiPAP as bridge (e.g., IPAP 12 cmH2O, EPAP 4 cmH2O) if FVC 15-30 mL/kg.
            - **Mechanical Ventilation:** If FVC <15 mL/kg, severe hypercapnia (PaCO2 >60 mmHg, pH <7.30).
        - MG-Specific: Plasmapheresis or IVIG for crisis, pyridostigmine 60 mg PO q4h.
        - GBS-Specific: IVIG 0.4 g/kg/day x 5 days, monitor respiratory status.

Management of Acute Exacerbations:

- Oxygen Therapy:
    - High-flow nasal cannula (HFNC) or NIV if PaO2 <60 mmHg, SpO2 <88%.
- Steroids:
    - Methylprednisolone 1-2 mg/kg/day IV for ILD exacerbation (e.g., IPF, HP).
- Antibiotics:
    - If infection suspected (e.g., pneumonia in sarcoidosis; ceftriaxone 1 g IV daily + azithromycin 500 mg IV daily).
- Ventilatory Support:
    - **NIV:** BiPAP for hypercapnic respiratory failure (e.g., OHS, neuromuscular disease).
    - **Intubation:** If NIV fails (e.g., PaCO2 >60 mmHg, pH <7.25), or severe hypoxemia (PaO2/FiO2 <150).

- Supportive Care:
  - Fluids, nutrition, DVT prophylaxis (enoxaparin 40 mg SC daily).

Table: Causes of Restrictive Lung Disease

Category	Cause	Key Features	Diagnostic Findings
Intrinsic (Parenchymal)	Idiopathic Pulmonary Fibrosis	Progressive dyspnea, dry cough, clubbing	HRCT: UIP pattern (honeycombing), PFTs: ↓ FVC, ↓ DLCO
	Sarcoidosis	Dyspnea, cough, erythema nodosum	CXR: Hilar lymphadenopathy, HRCT: Perilymphatic nodules
	Hypersensitivity Pneumonitis	Dyspnea, exposure history (e.g., birds)	HRCT: Ground-glass, centrilobular nodules
	CTD-ILD (e.g., RA, Scleroderma)	Dyspnea, joint pain, rash	HRCT: NSIP pattern, serologies: ANA+, anti-Scl-70+
Extrinsic (Pleural)	Pleural Effusion	Dyspnea, chest pain	CXR: Blunting costophrenic angle, thoracentesis
	Pneumothorax	Sudden dyspnea, chest pain	CXR: Pleural line, lung collapse
Extrinsic (Chest Wall)	Kyphoscoliosis	Dyspnea, spinal deformity	CXR: Cobb angle >50°, PFTs: ↓ TLC
	Obesity Hypoventilation	Dyspnea, obesity (BMI >40)	PFTs: ↓ FVC, ABG: Hypercapnia
Extrinsic (Neuromuscular)	ALS	Dyspnea, muscle weakness	EMG: Denervation, PFTs: ↓ FVC, normal DLCO
	Myasthenia Gravis	Dyspnea, ptosis, diplopia	Anti-AChR+, PFTs: ↓ FVC

Table: Hospital Management Strategies for Restrictive Lung Disease

Cause	Acute Management	Chronic Management	Monitoring
IPF Exacerbation	Methylprednisolone 1 g IV daily x 3 days, HFNC if SpO2 <88%	Pirfenidone 801 mg PO TID, LTOT if PaO2 <55 mmHg	ABG q6h, PFTs q3 months, 6MWT
Sarcoidosis	Prednisone 20-40 mg PO daily, oxygen if needed	Methotrexate 15 mg PO weekly if steroid-resistant	CXR q6 months, ACE levels
Pleural Effusion	Thoracentesis, treat underlying cause (e.g., diuretics for CHF)	Pleurodesis if recurrent (e.g., malignant effusion)	CXR post-thoracentesis, monitor recurrence
OHS	BiPAP (IPAP 14, EPAP 6 cmH2O), HFNC if hypoxemic	Weight loss, bariatric surgery if BMI >40	ABG (PaCO2), sleep study

Cause	Acute Management	Chronic Management	Monitoring
Neuromuscular (GBS)	Monitor FVC q4h, intubate if FVC <15 mL/kg, IVIG 0.4 g/kg/day	Long-term NIV, physical therapy	FVC trends, EMG/NCS

Clinical Scenarios

Scenario 1: Elderly Male with IPF Exacerbation

- Presentation: A 70-year-old male with known IPF presents with 3 days of worsening dyspnea and dry cough. Exam shows T 37.5°C, BP 120/80 mmHg, HR 100 bpm, RR 28/min, SpO2 86% on room air, fine basal crackles, clubbing.
- Labs/Studies: **ABG:** PaO2 50 mmHg, PaCO2 38 mmHg, pH 7.40, HRCT: Honeycombing with new ground-glass opacities, PFTs (prior): FVC 60% predicted, DLCO 45% predicted.
- Diagnosis: Acute exacerbation of IPF → Worsening dyspnea, new ground-glass on HRCT, known IPF.
- Management: Admit to ICU (hypoxemia). Start HFNC 40 L/min, FiO2 60% (SpO2 improves to 92%). Methylprednisolone 1 g IV daily x 3 days. Prophylactic antibiotics (ceftriaxone + azithromycin) to cover infection. LTOT initiated (PaO2 <55 mmHg). Consult pulmonology: Continue pirfenidone, consider transplant evaluation. After 5 days, dyspnea improves, discharged on 2 L O2 via nasal cannula.

Scenario 2: Middle-Aged Female with Sarcoidosis and Pulmonary Involvement

- Presentation: A 45-year-old female presents with 2 months of dyspnea, dry cough, and fatigue. Exam shows T 37°C, BP 130/85 mmHg, HR 90 bpm, RR 20/min, SpO2 90% on room air, erythema nodosum, hilar lymphadenopathy on exam.
- Labs/Studies:WBC 8,000/µL, Hgb 12 g/dL, ESR 40 mm/h, ACE levels elevated, HRCT: Perilymphatic nodules, hilar lymphadenopathy, PFTs: FVC 70% predicted, DLCO 65% predicted, biopsy: Non-caseating granulomas.
- Diagnosis: Sarcoidosis with pulmonary involvement → Dyspnea, hilar lymphadenopathy, granulomas on biopsy.
- Management: Admit for evaluation. Start prednisone 40 mg PO daily (dyspnea, pulmonary involvement). Supplemental O2 2 L/min via nasal cannula (SpO2 94%). Consult pulmonology: Plan to taper prednisone over 3 months, consider methotrexate if refractory. Monitor PFTs q3 months. Discharged on day 3 with improved symptoms.

Scenario 3: Young Male with GBS and Respiratory Failure

- Presentation: A 30-year-old male with GBS (diagnosed 1 week ago) presents with worsening dyspnea and weakness. Exam shows T 37°C, BP 110/70 mmHg, HR 100 bpm, RR 24/min, SpO2 88% on room air, proximal muscle weakness, paradoxical breathing.
- Labs/Studies: **ABG:** PaO2 55 mmHg, PaCO2 50 mmHg, pH 7.32, FVC 12 mL/kg (0.8 L), EMG: Demyelination, anti-ganglioside antibodies positive.
- Diagnosis: GBS with acute hypercapnic respiratory failure → FVC <15 mL/kg, hypercapnia, paradoxical breathing.
- Management: Admit to ICU (respiratory failure). Intubate (FVC <15 mL/kg, PaCO2 >50 mmHg): AC-VC mode, VT 6 mL/kg (420 mL), RR 14 breaths/min, FiO2 40%, PEEP 5 cmH2O. IVIG 0.4 g/kg/day x 5 days initiated. DVT prophylaxis (enoxaparin 40 mg SC daily). Monitor FVC q4h, ABG q6h. After 5 days, FVC improves to 20 mL/kg, weaned to BiPAP, extubated on day 7.

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