Acute management of paediatric respiratory failure

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Respiratory failure may be defined as failure of the lungs to maintain adequate gas exchange. It may occur as a result of alveolar hypoventilation, diffusion impairment, intrapulmonary shunting or ventilation–perfusion mismatch. Respiratory failure may be associated with hypoxaemia, hypercarbia or both. Hypoxaemia can cause tissue hypoxia and progressive hypercarbia can cause carbon dioxide narcosis. Both of these complications are potentially life threatening. Thus treatment of respiratory failure must aim both to maintain gas exchange and to treat the underlying cause.

ASSESSMENT

Assessment of the child with respiratory failure follows the standard ABC (airway, breathing, circulation) approach, with an emphasis on the work of breathing and on the effects of hypoxaemia on other organ systems, particularly the heart and brain. Once an initial assessment of severity has been made and supportive measures instituted, appropriate investigations should be arranged to determine the underlying cause so that specific therapy may be commenced (*Table 1*).

SUPPORTIVE THERAPY

Supportive therapy ranges from oxygen by face mask, to non-invasive ventilation, endotracheal intubation and mechanical ventilation, nitric oxide (NO) and extracorporeal membrane oxygenation (ECMO).

Oxygen

Children with oxygen saturation (SpO_2) <92% should receive high-flow oxygen via a tight-fitting mask to achieve normal saturations. The amount can be titrated according to pulse oximetry. A fixed performance, high-flow mask provides a fractional inspired oxygen concentration (FiO_2) in the range 0.24–0.60. The FiO_2 is not known with the more common variable performance masks or with nasal cannulae. The maximum concentration of oxygen via face mask is 0.60 unless a reservoir bag is used.

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Non-invasive ventilation

Non-invasive ventilation refers to ventilatory support without endotracheal intubation. This includes continuous positive airways pressure or biphasic positive airways pressure via face mask or nasal mask. These techniques are particularly helpful in neuromuscular disorders, in obstructive sleep apnoea and in immunocompromised patients (in whom avoiding intubation reduces the risk of nosocomial pneumonia).

Ventilatory support

Endotracheal intubation and mechanical ventilation should be considered in any child who is tiring as a result of excessive work of breathing, or who has cardiovascular compromise or a reduced conscious level as a result of respiratory failure. While worsening hypoxaemia or worsening hypercarbia may confirm the imminent need for ventilation, blood gas analysis is not a substitute for clinical assessment (*Table 2*).

TABLE 1. Indicators of respiratory distress

Moderate	Tachycardia
	Respiratory rate > 50
	Flaring
	Use of accessory muscles
	Recession
	Head retraction
	Unable to feed
Severe	Cyanosis
	Getting tired
	Reduced conscious level
	Saturation <92% in spite of oxygen therapy
	Worsening hypercapnia

Inhaled nitric oxide

Inhaled NO is a pulmonary vasodilator. Although there are no randomized controlled trials to support the use of inhaled NO outside the neonatal period, older children with severe refractory hypoxaemic respiratory failure may benefit from inhaled NO. NO may also protect patients whose oxygenation might otherwise depend upon a potentially damaging ventilatory strategy.

Extracorporeal membrane oxygenation

The benefit of ECMO for acute hypoxaemic respiratory failure outside the neonatal period is also unproven. However, ECMO may be considered as a supportive therapy in the unventilatable child with respiratory failure if the duration of ventilation is under 7 days before initiation of ECMO. This time limitation is to avoid offering ECMO to children who have irreversible lung injury as a result of high pressure ventilation and who are therefore unlikely to recover.

TABLE 2. Indications for intubation and ventilation in respiratory failure

Severe respiratory distress
Tiring as a result of excessive work of breathing (may be indicated by progressive hypercarbia)
Progressive hypoxaemia
Reduced conscious level
Progressive neuromuscular weakness, e.g. Guillain–Barré syndrome

SPECIFIC CAUSES OF RESPIRATORY FAILURE

Parenchymal lung disease

Acute lung injury/acute respiratory distress syndrome: Acute lung injury (ALI) is the term used to describe the pulmonary response to a broad range of injuries occurring either directly to the lung or as a result of injury or inflammation at other sites in the body (*Table 3*).

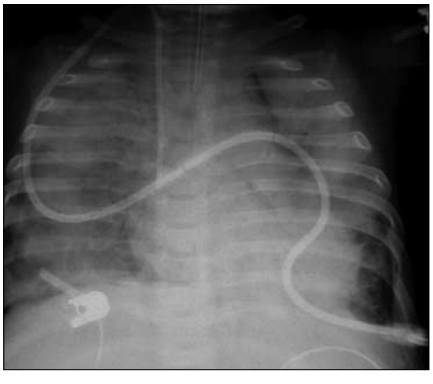
Acute respiratory distress syndrome (ARDS) represents the severe end of this condition. Diagnostic criteria were developed for the syndrome in 1994 by an American–European consensus conference (Bernard et al, 1994). These are:

- Bilateral pulmonary infiltrates on chest X-ray (*Figure 1*)
- Pulmonary capillary wedge pressure <18 mmHg or no clinical suspicion of raised left atrial pressure
- Partial pressure of arterial oxygen (PaO_2)/FiO_2 <300 = ALI
- PaO_2/FiO_2 <200 = ARDS

TABLE 3.
Common causes of acute lung injury and acute respiratory distress syndrome

Sepsis
Pneumonia
Trauma
Aspiration/near drowning
Burns/inhalational injury
Massive blood transfusion
Transfusion-related acute lung injury

Figure 1. Bilateral diffuse pulmonary infiltrates as a result of acute respiratory distress syndrome in a leukaemic child with febrile neutropenia.



Pathogenesis: ALI is characterized by widespread airway collapse, surfactant deficiency and reduced lung compliance. It is an inflammatory disorder with three phases: an exudative phase, a proliferative phase and a fibrotic phase.

The exudative phase lasts for the first week and is marked by alveolar oedema with hyaline membranes. In the proliferative phase, in the second 2 weeks, organization of the inflammatory exudates occurs. In the fibrotic phase, which may occur from day 10, pulmonary fibrosis is seen. However, many studies show that survivors return to normal lung function, with complete resolution of pulmonary fibrosis.

Therapeutic strategies: The goals of treating patients with ALI or ARDS are to maintain adequate gas exchange while avoiding ventilator-induced lung injury, to treat the underlying cause of the condition and to attenuate the inflammatory response.

Oxygenation: High concentration inspired oxygen should be avoided to limit the risk of direct cellular toxicity and to avoid reabsorption atelectasis. SaO_2 values of around 88–92% are commonly accepted. Positive end expiratory pressure (PEEP) may improve oxygenation by encouraging movement of fluid from the alveolar to the interstitial space, recruiting collapsed alveoli, increasing functional residual capacity and preventing cyclical alveolar collapse. A long inspiratory time may also improve lung recruitment.

Lung protective ventilation: Traditional mechanical ventilation, using high tidal volumes and low PEEP, is likely to induce lung injury in patients with ARDS. However, in ARDS, a 'lung protective strategy', optimizing PEEP, using a tidal volume of <6 ml/kg, permissive hypercapnia, and pressure limited ventilation with peak inspiratory pressure limited to <40 cmH_2O, has been shown to improve outcome (Anon, 2000).

High frequency oscillatory ventilation: High frequency oscillatory ventilation (HFOV) may be of benefit for patients with ARDS because it delivers small tidal volumes (typically 2 ml/kg), thus preventing 'atelectotrauma', it keeps the lungs open and improves alveolar recruitment, and it may improve ventilation–perfusion matching. Such data as exist in paediatric ARDS suggest that a trial of HFOV may be beneficial when the mean airway pressure is >16 cmH_2O and FiO_2 >0.6 (Arnold et al, 1994).

Corticosteroids: In two small trials, steroid therapy in ARDS was associated with improved oxygenation and successful extubation (Meduri et al, 1998; Varpula et al, 2000). Many centres now use steroids for ARDS which has not resolved after 7 days supportive therapy. The current ARDS Network Late Steroid Rescue Study may provide further evidence for prescribing low-dose steroids in ARDS.

Other therapies: Other therapies occasionally of benefit include prone positioning, inhaled NO, exogenous surfactant and ECMO.

Other causes of parenchymal lung disease: The principles of ventilating patients with ARDS applies to any cause of parenchymal lung disease, such as pulmonary oedema, pulmonary haemorrhage, pneumonia and aspiration syndromes. As in any cause of ARDS, however, the underlying cause of the problem must also be addressed, for example, pneumonia should be investigated and treated with appropriate antibiotics (Anon, 2002).

Small airway disease

Asthma: Management of acute severe asthma is almost always possible without mechanical ventilation. Children should receive oxygen therapy to achieve normal saturations and nebulized salbutamol every 20–30 minutes. If symptoms are refractory to initial treatment, ipratropium bromide should be added to each nebulizer. The dose frequency may be reduced as clinical improvement occurs.

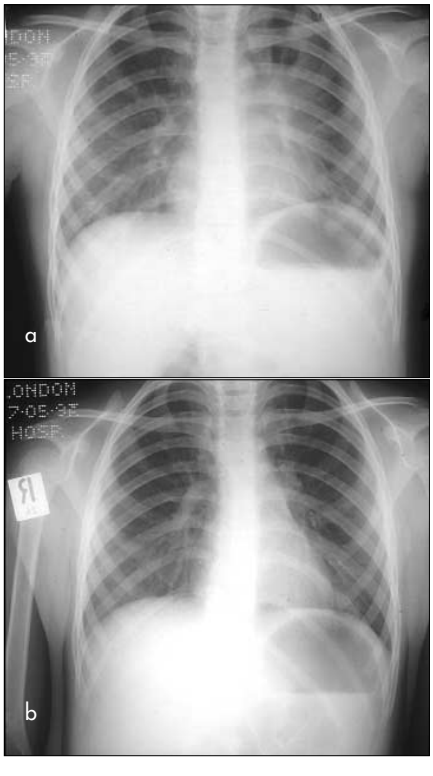
Steroids should be given early in acute asthma. In children unable to take oral medication, intravenous hydrocortisone should be given 4-hourly.

The early addition of a bolus dose of intravenous salbutamol can be an

effective adjunct in severe cases. A continuous infusion should be considered in refractory asthma. Patients on intravenous salbutamol should have continuous cardiac monitoring and regular monitoring of electrolytes. Aminophylline can be used in children with refractory bronchospasm. A loading dose should be given over 20 minutes with cardiac monitoring followed by a continuous infusion. A loading dose should not be given to patients on oral theophylline treatment. Magnesium sulphate is also effective and should be given by slow infusion over 30 minutes (Anon, 2003).

If mechanical ventilation is required, a low tidal volume, slow rate strategy with a long expiratory time is recommended to avoid the problem of progressive hyperinflation as a result of inadequate expiration. Permissive hypercapnia is recommended for the same reason. Humidification, regular chest physiotherapy and suctioning are essential to prevent and treat mucus plugging (*Figure 2*). Sedation and muscle relaxation may be required to facilitate mechanical ventilation.

Figure 2. a. Left upper lobe and lingula collapse as a result of mucus plugging in a child with acute severe asthma. b. Complete resolution after bronchodilator therapy and expectoration of a mucus plug.

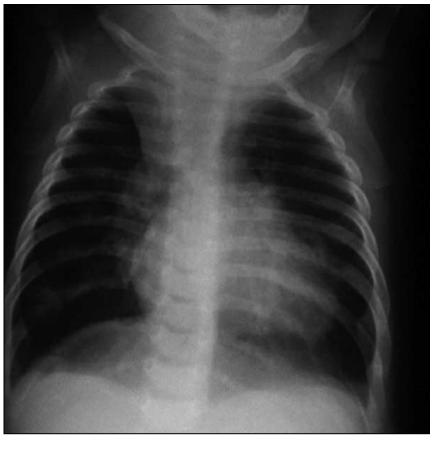


Bronchiolitis: Bronchiolitis is a clinical syndrome of infancy characterized by respiratory distress and crepitations and wheezes on auscultation. It is often preceded by coryzal illness and usually has a viral aetiology such as respiratory syncytial virus (RSV), influenza, parainfluenza, adenovirus or metapneumovirus. Co-infection with *Mycoplasma pneumoniae* has been described but secondary bacterial infection is very uncommon. However, co-infection with pertussis should be considered in very severely affected infants, particularly in the context of echocardiographic evidence of pulmonary hypertension.

The pathophysiology is caused by airway obstruction, which may be partial or complete, leading to areas of hyperinflation and atelectasis, and a resultant ventilation–perfusion mismatch (*Figure 3*). Treatment is largely supportive. Other than nebulized epinephrine, which has been shown to be of benefit in some randomized controlled studies (Hartling et al, 2004), there is no effective specific treatment. Ribavirin, an antiviral medication with specific activity against RSV, has not been shown to be of any benefit (Randolph and Wang, 2000).

Some infants also develop an ARDS picture with four quadrant shadowing (Tasker et al, 2000). Typically, this latter group are much sicker and have a much longer time course on the intensive care unit. Thus it is possible for both airway obstruction and an ARDS-type picture to be present in the same infant with RSV. This has implications for ventilation. Infants with predominant airway obstruction should be ventilated as per 'asthma' above. However, infants with a predominantly ARDS-type picture may need to be ventilated as per 'ARDS' above. Very severely affected infants in either group may respond to HFOV, inhaled NO and/or surfactant (Tibby et al, 2000), or require ECMO (Khan et al, 1995).

Figure 3. Gross hyperinflation and right upper lobe collapse in a 7-month-old boy with respiratory syncytial virus bronchiolitis. The child was subsequently found to have cystic fibrosis.



Large airway disease

Tracheobronchomalacia and stenosis: Tracheobronchomalacia is a condition of dynamic airway collapse during expiration. In tracheal stenosis, fixed airway narrowing causes airway obstruction in inspiration and in expiration. These conditions may be primary, when cartilaginous rings are congenitally malformed, or secondary, as a result of degeneration of previously normal cartilage.

Mild tracheobronchial malacia or stenosis classically presents in early childhood with wheeze, cough, stridor and recurrent lower respiratory tract infections. It usually resolves spontaneously as cartilaginous development of the airway occurs. More severe lesions present in early infancy with respiratory failure, episodes of profound cyanosis, reflex apnoea and inability to wean from mechanical ventilation following preterm delivery or surgical procedures.

A diagnosis of tracheobronchomalacia requires contrast bronchography, which permits assessment of the trachea and bronchi throughout the respiratory cycle (*Figure 4*). Bronchoscopy is also useful, particularly to identify complete tracheal rings.

Infants requiring intensive care may need a variety of management approaches, including surgery and long-term ventilation (Inwald et al, 2001).

Restrictive lung disease

Pleural effusion, empyema and pneumothorax: These conditions do not require specific ventilatory man-

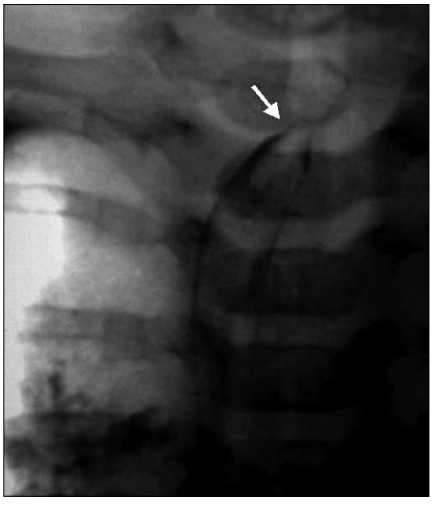
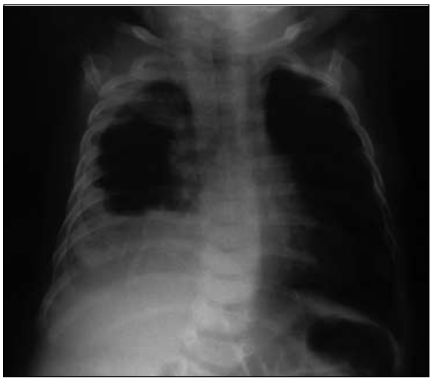


Figure 4. Contrast bronchogram demonstrating a tight mid-tracheal stenosis.

agement, but the underlying cause of respiratory failure should be removed. Effusions and pneumothoraces should be drained. Empyemas should be drained but may require more specific management including antibiotic therapy (which should cover *Pneumococcus*, *Staphylococcus* and *Mycoplasma*), instillation of fibrinolytic agents such as urokinase through the chest drain or surgical decortication (*Figure 5*). Management of empyema should be determined in conjunction with a respiratory paediatrician and cardiothoracic surgeon (Jaffe and Cohen, 2003).

Thoracic cage abnormalities and neuromuscular weakness: Both thoracic cage abnormalities such as Jeune's asphyxiating thoracic dystrophy (*Figure 6*) and progressive neuromuscular disease may cause respiratory failure as a result of alveolar hypoventilation. The principles of ventilation of these conditions are the same as those applied for ARDS, i.e. a low tidal volume/low pressure strategy with permissive hypercapnia. These conditions are often irreversible and palliative care should be considered rather than a long-term ventilation strategy (Yates et al, 2004).

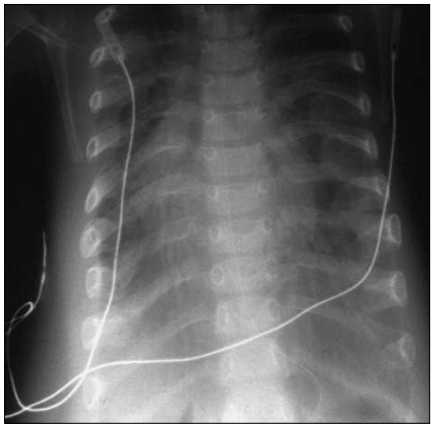
Figure 5. Right-sided staphylococcal empyema in a 9-month-old. An intercostal drain was inserted and the baby required mechanical ventilation for 12 days.



CONCLUSIONS

Previous work has demonstrated that the outcome of respiratory failure in paediatric critical care is related to the severity of associated pathology and not to the severity of respiratory failure (Peters et al, 1998). Thus early recognition, supportive treatment and appropriate specific therapy are critical to avoid any unnecessary morbidity and mortality in this eminently treatable condition. HM

Figure 6. Asphyxiating thoracic dystrophy of Jeune. The thorax is small and bell-shaped with short, horizontal ribs and has severely restricted lung growth. The child died of progressive respiratory failure.



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KEY POINTS

- Accurate assessment of severity of respiratory distress is the key to guiding management.
- Supportive treatments should be given in a timely manner.
- Intubation and mechanical ventilation should not be delayed until respiratory arrest is imminent.
- A lung protective ventilation strategy should be pursued to avoid ventilator-associated lung injury.