Great Ormond Street Hospital for Children
NHS Trust

Ventilation Strategies for Specific Conditions

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Associated GOSH clinical guidelines/protocols:
- Detailed ventilator teaching packs by Lisa Martin, Senior ventilator technician. *GOSH intranet > Guidelines & Projects > ICU Guidelines >IOL > teaching packs > Ventilator & respiratory systems support handouts.*
- Asthma
- Bronchiolitis
- BAL
- PPHN protocol

Information for Year 1 ITU Training (basic):

> **Year 1 ITU curriculum**
>
> **Basic ITU management of the following Clinical Conditions:**
> - ALI & ARDS –basic management approach. Note patholophysiology covered in "respiratory physiology & pathophysiology" module
> - Asthma & bronchiolitis
> - Pneumonia, collapse & consolidation
> - Pleural effusion, empyema and pneumothorax
>
> **Note: Congenital malformations of the upper & lower airways are covered in "congenital malformations requiring surgery" module.**

Curriculum Notes for Year 1:

The majority of children admitted to an intensive care unit are ventilated either for primary respiratory illnesses or for multi-system disorders such as sepsis, trauma and burns. With the increasing realisation that the act of ventilation may itself lead to organ failure, the adoption of an optimal ventilatory strategy for different conditions is important to achieve the best outcome with minimal risk.

Acute lung injury (ALI)/ARDS

Acute lung injury (ALI)/Acute respiratory distress syndrome (ARDS) are disorders marked by a significant inflammatory response to a local (pulmonary) or remote (systemic) insult resulting in injury to alveolar epithelial and endothelial barriers of the lung, acute inflammation and protein rich pulmonary oedema. ARDS is the more severe form of ALI.

Definition
The most used definition for acute lung injury (ALI) and ARDS is the revised one from the 1994 North American European Consensus Conference (NAECC).[1]

TABLE 1. 1994 Consensus Conference Definitions of ALI and ARDS[3]

Onset	:	Acute and persistent
Oxygenation criteria	:	$PaO_2/FiO_2 \leq 300$ for ALI; ≤ 200 for ARDS
Radiographic criteria	:	Bilateral infiltrates
Exclusion criteria	:	Pulmonary artery occlusion pressures ≥ 18 mmHg Clinical evidence of left atrial hypertension

Bernard, G. R et al Am J Respir.Crit Care Med 149: 818-824.

This is an example of a chest x-ray showing bilateral diffuse infiltrates:

From http//www.ccmtutorials.com

Incidence and aetiology
ARDS in all age groups is reported to be relatively rare (75 cases/100,000 population per year based on the NAECC definition). The incidence of paediatric ARDS has not been studied in detail – reported rates based on the NAECC criteria vary from 8-16/1000 PICU admissions. A paediatric review from 2007 suggests a 9% occurrence of ALI in ventilated children out of which 80% progress to ARDS. The commonest etiology was found to be septicaemia (Dahlem et al. Pediatric acute lung injury. *Paediatr Respir Rev. 2007 Dec;8(4):348-62*)

ALI/ARDS represent the end result of an aggressive inflammatory process in the lungs. The pulmonary response occurs to a broad range of injuries occurring either directly to the lungs or as a consequence of injury or inflammation at other sites in the body.

Local insult	*Systemic insult*
Pneumonia	Sepsis
Aspiration event	Burns
Inhalational injury	Major trauma
Fat/Air/Amniotic fluid embolism	Pancreatitis
Radiation injury	Poisoning

Pathology

Lung injury is a dynamic condition and the pathological features of ARDS are described as passing through three overlapping phases - an inflammatory or exudative phase (0-7 days), a proliferative phase (7-21 days) and lastly a fibrotic phase (from day 10).

The chief pathological changes and clinical features are summarised below:

• Diffuse alveolar damage • Proteinaceous and haemorrhagic interstitial oedema • Hyaline membrane formation • Surfactant dysfunction • Small airway collapse
• Reduced lung compliance • V/Q mismatching • Hypoxaemia • 'Wet lungs'

Therapeutic strategies

ARDS has no specific pharmacological treatment. Various support therapies have been studied; few of them have shown consistent benefit, perhaps due to the extremely heterogeneous nature of the disease.

Summary of the various therapeutic strategies

TABLE 5. Therapeutic Strategies in ARDS[38]

Control of causative factors (sepsis, shock, etc)
Mechanical ventilation
 • Controlled oxygen exposure (FiO_2)
 • Avoidance of volutrauma (low V_T)
 • Avoidance of atelectrauma (appropriate PEEP)
Non-conventional ventilation
 • High frequency ventilation
 • Liquid ventilation
Careful fluid administration
Drug-based therapies
 • Nitric oxide
 • Surfactant
 • Corticosteroids and other anti-inflammatory agents
Positioning (Prone ventilation)
Supportive therapy
 • Analgesia and sedation
 • Nutrition/Immunonutrition
 • Psychosocial support

Vasudevan A et al. Indian J Pediatr. 2004 Aug;71(8):743-50.

Conventional mechanical ventilation

The goal of ventilation in ARDS is to maintain adequate gas exchange with minimal ventilator induced lung injury (VILI).

Mechanisms implicated in VILI
• Oxygen toxicity from use of high FiO2 • Over distension of alveoli leading to volutrauma and barotrauma • Repetitive opening and closing of alveoli causing shear stress and triggering further inflammation (atelectrauma)

The most revolutionary change in the management of children with ARDS has been adoption of the concept of "lung protective" ventilation where lower tidal volume (VT) and appropriate higher positive end-expiratory pressure (PEEP) are used.[2]

Key features of "lung protective" ventilation
- Controlled oxygen exposure
- Permissive hypercapnia
- Low tidal volumes 4-6 ml/kg
- Adequate PEEP
- Peak pressure < 30 cm H_2O

Controlled Oxygen Exposure
High FiO2 should be avoided to minimise the risk of direct cellular toxicity and avoid re-absorption atelectasis. Even though there is no clinical evidence to suggest a threshold value, it is preferable to decrease FiO2 below 0.60 as soon as possible.

Permissive hypercapnia
Using the 'lung protective' strategy may cause hypercapnia. It is increasingly being accepted that a mild elevation of PaCO2 (permissive hypercapnia) and maintaining limits on VT and airway pressure is associated with a significantly lower mortality from ARDS. There are no data to confirm the degree of acidosis that is safe. It is probably safe to maintain a pH above 7.25 with a PaCO2 of less than 10 kPa.

Minimise Tidal Volume (VT)
A couple of large studies have evaluated the role of lung-protective (low VT) strategy for ARDS, which has shown an improved survival rate in patients with ARDS, although two other studies on patients with ARDS failed to show any benefit using low VT. Children with ARDS should be ventilated with a low VT of 6 ml/kg. If a child is ventilated on a pressure-controlled mode, tidal volumes should be accurately monitored.
Note: Use of large tidal volumes even in patients without ARDS/ALI has shown to induce VILI and as a default, one should try and avoid tidal volumes above 8ml/kg in any pathology.

Positive End-expiratory Pressure (PEEP)
The application of adequate levels of PEEP improves oxygenation and is associated with favourable physiological outcomes. PEEP improves oxygenation by providing movement of fluid from the alveolar to interstitial space, recruitment of small airways and collapsed alveoli and an increase in functional residual capacity. In clinical practice, PEEP is adjusted between 8 cm H_2O and 20 cm H_2O; PEEP is progressively increased by 2-3 cm H2O increments to maintain saturation between 90 and 95% with FiO2 < 0.5. The child should be monitored for any evidence of cardiovascular compromise and hyperinflation.

High frequency oscillatory ventilation (HFOV)

There has been a resurgence of interest in HFOV over the last few years but the role in paediatric respiratory failure and ARDS remains a source of debate. The advantages of HFOV are (a) use of low VT and avoidance of barotrauma and (b) maintenance of near normal $PaCO_2$ with improved minute ventilation. Some studies in paediatric population have shown that early initiation of HFOV is associated with better oxygenation but none have demonstrated clearly improved outcome.[3]

Drugs

- Inhaled nitric oxide
- Surfactant therapy
- Corticosteroids

Nitric Oxide (NO)
NO causes pulmonary vasodilation and decrease in pulmonary hypertension. Maximal improvement in oxygenation is usually achieved with <10 ppm in most patients. The effect can be frequently seen in less than ten minutes or may take several hours. Data from various

paediatric studies suggest that iNO improves short-term oxygenation in children with ARDS but little change is seen in long-term oxygenation indices.[4] Cochrane Metaanalysis from 2007 doesnot show any added advantage of adding iNO in ARDS.

Surfactant Therapy
Few studies have shown improvement in oxygenation after using surfactant but there are no good randomized trials in paediatric ARDS to draw any definite conclusions. Surfactant may be a useful adjuvant in special situations when oxygenation cannot be achieved with conventional methods. The patients who would most benefit from this therapy remains to be elucidated.

Corticosteroids
The use of corticosteroid does not prevent the development of ARDS, nor is it beneficial when employed during the initial phase of the clinical course. However, in a prospective, randomized controlled trial prolonged administration of methylprednisolone in adult patients with unresolving ARDS (ARDS>7 days) was associated with improvement in lung injury and MODS scores and reduced mortality.[5] These findings have not been replicated in larger more recent RCTs, which have shown improved ventilatory parameters but increased late mortality in the group given steroids (see below (Year 2) for these papers).

Prone positioning
The mechanism of improvement in oxygenation with prone positioning is complex and varied. Changes in regional lung perfusion and in regional pleural pressures and recruitment of dorsal lung have been postulated to improve oxygenation during prone positioning. In a large study in adults, prone position was not associated with any improvement in clinical outcome. Prone positioning may be used in selected children with ARDS as it has few risks or cost involved.

Asthma

Asthma is characterised by reversible airway obstruction and inflammation. Intubation and ventilation are life saving in children with life-threatening features of severe asthma. Intubation can be associated with serious decompensation. Once intubated, the main strategies applicable to the ventilation of patients with asthma are:

- Minimise minute ventilation
- Maximise time for expiration
- Slow rate, adequate inspiratory time (lower than physiologal rates)
- Avoid hyperinflation (air trapping)
- Accept lower pH (>7.2)
- Permissive hypercapnia
- Either Volume/pressure ventilation- maybe a marginal advantage of volume targeted with constant flow.

In general, pressure controlled ventilation (keep PIP <30) or volume controlled ventilation (Tv 5-8 ml/kg) may be used. A long expiratory time (with a optimum inspiratory time) with an I/E ratio of >1:2 and a slow rate allow emptying of the lungs and avoid 'air trapping' and progressive hyperinflation. Sometimes a manual decompression of chest helps to deflate overinflated lungs and improves ventilation.

Sedation and neuromuscular paralysis are important to avoid the complications of air-leak. Preferred drugs for these patients are ketamine (has a bronchodilator effect) & fentanyl (as morphine causes histamine release which might aggravate bronchospasm). Suction and physiotherapy are important to clear mucus plugging and prevent atelectasis. Other specific treatments for asthma in the form of nebulised and intravenous salbutamol, intravenous aminophylline, systemic steroids and magnesium sulphate are important in the management of the intubated asthmatic.

Bronchiolitis

Bronchiolitis is a clinical syndrome of infancy characterised by respiratory distress and crepitations and wheezes on auscultation, due to inflammation of bronchiole mostly due to a viral infection. The main features are partial or complete airway obstruction, resultant atelectasis and a resultant V/Q mismatch.

Infants can present predominantly with apneas, air trapping and wheeze, with atelectasis and parenchymal lung disease (ARDS) or a mixture of the above. The latter group are quite sick in the first 24 hours (AadO2 >400) and have a long ICU stay. Ventilatory strategies for each of the clinical presentations is different:

- Apnoeas (relatively normal lungs) – Minimise VILI with low Tv
- Air trapping – Manage like asthma
- ARDS – Manage like ARDS (including HFOV, iNO, ECMO)

Co-infection with pertussis is common in infants who are partially or fully unimmunised (<4 months); this may result in severe lung disease and ARDS.

Pneumonia/Lung collapse

The principles of ventilating a child with pneumonia or lung collapse/consolidation, aspiration syndromes, pulmonary oedema or pulmonary haemorrhage are similar to ARDS management. The predominant pathology in all the above conditions is air space disease.

Summary of ventilation strategy:

- Minimise oxygen toxicity (FiO2 <0.60)
- Minimise atelectrauma (adequate PEEP)
- Minimise volutrauma (low Tv 5-8 ml/k)
- Permissive hypercapnia

Pleural effusion, empyema and pneumothorax

There are no specific ventilatory strategies for the management of pleural effusions or empyema other than removing the cause of the respiratory failure (antibiotics, drainage of the collection etc). Instillation of urokinase or streptokinase has been used in aiding drainage of empyema; pig tail drains are no worse than ordinary chest drains; surgical decortication may be necessary in the management of complicated empyema. Cardiothoracic and Respiratory input is essential to manage these children.

Draining a pneumothorax is usually sufficient. Multiple air-leaks may be caused by Staphylococcal infection or necrotising pneumonias from Gram-negative organisms such as Klebsiella. In multiple air-leaks (pneumothoraces, pneumomediastinum etc), the current ventilatory strategies are:

- High frequency oscillatory ventilation (HFOV)
- ECMO for refractory air leaks

Note: Non invasive ventilatory techniques can be used in carefully selected patients with any of the diseases discussed above to either prevent intubation or assist extubation.

Other sources of information:
1. Pulmonary Physiology and Pathophysiology: An Integrated, Case-based Approach. John B. West. Lippincott, Williams and Wilkins. December 2000
Websites:
1. www.ardsnet.org - Excellent site that lists all the studies (completed and ongoing) in ARDS; list of relevant publications; and future research paths

2. www.ccmtutorials.com - Excellent summary of adult ICU problems; Respiratory failure is covered well
3. http://pedsccm.wustl.edu/FILE-CABINET/File_cab_index.html - Resources on the PedsCCM website dealing with Pulmonary topics

References:

Key articles attached

1. Bernard, G. R., Artigas, A., Brigham, K. L., Carlet, J., Falke, K., Hudson, L., Lamy, M., LeGall, J. R., Morris, A. and Spragg, R. (1994) The American-European Consensus Conference on ARDS. Definitions, mechanisms, relevant outcomes, and clinical trial coordination, Am J Respir.Crit Care Med 149: 818-824.

2. Anon. (2000) Ventilation with lower tidal volumes as compared with traditional tidal volumes for acute lung injury and the acute respiratory distress syndrome. The Acute Respiratory Distress Syndrome Network, N.Engl.J Med 342: 1301-1308.

3. Arnold JH, Hanson JH, Toro-Figuero LO, Gutierrez J, Berens RJ, Anglin DL. Prospective, randomized comparison of high-frequency oscillatory ventilation and conventional mechanical ventilation in pediatric respiratory failure. Crit Care Med. 1994;22:1530-9

4. Taylor RW, Zimmerman JL, Dellinger RP, et al. Low-dose inhaled nitric oxide in patients with acute lung injury: a randomized controlled trial. JAMA. 2004 Apr 7;291(13):1603-9.

5. Meduri, G. U., Headley, A. S., Golden, E., Carson, S. J., Umberger, R. A., Kelso, T. and Tolley, E. A. (1998) Effect of prolonged methylprednisolone therapy in unresolving acute respiratory distress syndrome: a randomized controlled trial, JAMA 280: 159-165

Information for Year 2 ITU Training (advanced):

> **Year 2 ITU curriculum**
> - More in-depth understanding of pathogenesis and aetiology of ALI and ARDS. Knowledge of current research.
> - Pulmonary oedema, pulmonary haemorrhage
> - Sickle chest syndrome
> - Aspiration syndromes
> - Cystic fibrosis
> - Thoracic cage abnormalities.
>
> **(Note: PPHN & meconium aspiration is covered in the Medical Neonates module; Heliox is covered in respiratory physiology & pathophysiology module)**

Curriculum Notes for Year 2:

Further detailed information on each of the conditions mentioned in the Year 1 curriculum notes is summarised below:

Acute lung injury (ALI)/ARDS

Epidemiology
The largest proportion of ARDS cases are made up of children with sepsis, severe trauma, pneumonia and multiple transfusions of blood products. Overall, approximately 20-40% of patients with well-established risk factors and respiratory failure develop ARDS, and the more risk factors in any individual, the greater the likelihood of developing ARDS. There is evidence that certain genetic polymorphisms also influence the likelihood of developing and dying from ARDS (TNFA, surfactant protein B and ACE).

Pathogenesis
Lung injury is a dynamic process and passes through a series of three phases (day 0-7: exudative phase; day 7-21: proliferative phase; day 10 onwards: fibrotic phase).

The key points in ARDS pathogenesis are:

- There is much greater overlap of the inflammatory and fibroproliferative phases of ARDS than previously imagined, with the fibroproliferative response beginning within 24 hours and its severity being directly related to the outcome.

    - Markers for collagen turnover are detectable in BAL fluid from 24 hours after ventilation for ARDS. Within 48 hours of diagnosis, myofibroblasts are detected in BAL fluid indicating early fibrotic mediators.

- An exaggerated inflammatory response underlies the pathogenesis of ARDS – the neutrophil is the dominant leucocyte, while many proinflammatory agents including endotoxin, proinflammatory and chemotactic cytokines, vascular endothelial growth factor, high mobility group-1 protein and thrombin, together with oxidant stress, are all implicated in vascular leakage and lung damage.

    - Neutrophils cause cell damage though the production of free radicals, inflammatory mediators, and proteases. Neutrophil elastase may be an important mediator of damage to the alveolar epithelium and the progression to fibrosis.
    - The inflammatory process is driven in part by cytokines including TNF- and IL-1ß, IL-6, and IL-8.

- Surfactant deficiency is not a primary causal event in ARDS; rather, the inflammatory processes lead to surfactant dysfunction as a secondary factor.

- Alveolar type II cell proliferation and enhanced fibroproliferation are key steps in attempts at repair in the lung; both offer potential targets for future therapies.

Pathology and clinical implications

Lung mechanics

- Lung involvement in ARDS is not homogenous
- Posterior, dependent portions of the lung are more severely affected, a distribution determined largely by gravity
- CT scans of ARDS survivors have shown greatest abnormality in the anterior parts of the lung even though the posterior areas had been most severely affected in the acute phase

This is a CT chest in a patient with ARDS, showing the typical heterogeneous distribution of opacification within the lungs. The increased density of lung tissue in dorsal regions (A) is caused by consolidation and atelectasis. The aerated, ventral regions (B) have the highest compliance and tend to become overdistended (volutrauma). The interface between the two areas (C) is prone to cyclic recruitment–derecruitment (atelectrauma).
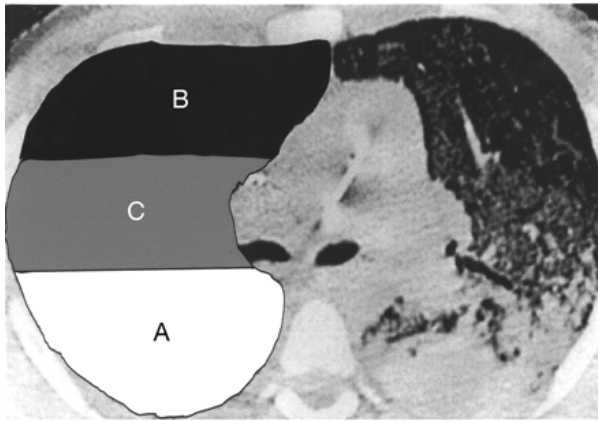
Therapeutic strategies and ongoing research

Surfactant
Although the use of surfactant has been tried on and off in ARDS, so far there has been no definite evidence of its benefit. However, a recent large multi-centre RCT showed that tracheal instillation of Calfactant resulted in a significant acute improvement in oxygenation and mortality, but did not suggest any significant decrease in the number of ventilator days, ICU stay or hospital length of stay.[1]

Prone positioning
A number of uncontrolled studies showed improvement in oxygenation following prone positioning in children with ARDS. However, a recent large multicentre RCT in children did not suggest any differences at 28 days between the group that was positioned prone for 20 hours/day for 7 days compared to the supine group.[2]

Ketoconazole
Small studies suggested that ketoconazole had immunomodulatory properties that might be useful in ARDS. However, a large RCT co-ordinated by the ARDS Network did not show nay differences between the groups.

Lisophylline
Lisophylline was postulated as a drug that would dampen the pro-inflammatory mediators in ARDS; however, an RCT co-ordinated by the ARDS Network was stopped mid-way for futility.

Steroids in late ARDS
A recent adult RCT co-ordinated by the ARDS Network has just concluded and results have now been published [3]. Patients with ARDS of over 7 days in duration were randomised to receive 2mg/kg/d methylprednisolone (or placebo) for 14 days followed by a tapering dose over 11 days. Methylprednisolone increased the number of ventilator-free and shock-free days during the first 28 days in association with an improvement in oxygenation, respiratory-system compliance, and blood pressure with fewer days of vasopressor therapy. As compared with placebo, methylprednisolone did not increase the rate of infectious complications but was associated with a higher rate of neuromuscular weakness. However, Methylprednisolone was associated with significantly increased 60- and 180-day mortality rates among patients enrolled at least 14 days after the onset of ARDS. These results do not support the routine use of steroids in ARDS.

Fluid management
A recent ARDSNet study involved randomising patients to a "fluid liberal" or "fluid restricted" strategy based on the use of central venous lines or pulmonary artery catheters. The results have now been published [4]. Although there was no significant difference in the primary outcome of 60-day mortality, conservative strategy of fluid management improved lung function and shortened the duration of mechanical ventilation and intensive care without increasing non pulmonary-organ failures.

Immunonutrition
Addition of glutamine, arginine and omega-3-fatty acids to diet has been suggested to promote immunomodulation. This therapy is unproven.

Extra corporeal membrane oxygenation is reserved as a last resort.

Pulmonary oedema and pulmonary haemorrhage

Pulmonary oedema in children usually falls into one of three categories:

- Cardiogenic
- Negative pressure (post-obstructive)
- Neurogenic
- Non-cardiogenic

Pulmonary haemorrhage is uncommon in children; severe coagulopathy and fluid overload are common causes. Unusual causes include tumours, Wegener's granulomatosis, idiopathic haemosiderosis and aspergillosis (in the immunocompromised host – usually fatal).

In both conditions, the main ventilatory strategies are:

Conventional ventilation
- Minimise oxygen toxicity (FiO2 <0.60)
- Minimise atelectrauma (adequate PEEP)
- Minimise volutrauma (low Tv 5-8 ml/k)
- Permissive hypercapnia

High frequency ventilation
- Constant MAP
- Recruitment of lung

Adjunctive therapies such as bronchoscopy may be helpful in identifying focal areas of bleeding.

Asthma and bronchiolitis

The key to ventilate severe asthmatic is to achieve adequate oxygenation (sats 88-92%) and minimize hyperinflation by hypoventilating the patient (pH >/= 7.2). The main ventilatory strategies in these conditions are covered in Year 1 material. Novel adjunctive therapies are:

- Heliox (helium-O2 mixture) either before or during mechanical ventilation
- Aerosol delivery using heliox
- Inhaled or nebulised magnesium sulphate

Use of heliox in the pre-ICU setting has received much attention – although there is lack of clear evidence based on the previous studies, it is likely to be useful in the sick asthmatic child in the first 24 hours. Use of heliox during mechanical ventilation has also resulted in dramatic improvement in respiratory acidosis in some case series.[5]

The use of extrinsic PEEP in the management is controversial and though some patients respond to extrinsic PEEP which matches the auto PEEP, others donot. The auto PEEP however could be monitored by measuring the end expiratory pressure. *(Ref: Respir Care 2008; 53: 740-748.)*

In very severe cases not responding to any of the above treatment strategies, buffer therapy with bicarbonate or THAM, inhaled anaesthetic gases, airway clearance with bronchoscopy & mucolytics, ECMO need to considered very very cautiously.

Sickle chest syndrome

Acute chest syndrome (ACS) in sickle cell disease is characterised by fever, infiltrates on chest radiograph, pleuritic chest pain, and hypoxaemia. ACS is the leading cause of hospitalisation in children with sickle cell disease; it is also associated with a 10-12% mortality rate and is the single most common cause of death in the >12 year old child with Hb SS.

Clinical features
- Fever
- New (bilateral) infiltrates
- Pleuritic chest pain
- Hypoxaemia
- Leucocytosis

Hypoxaemia may result from a number of pathological changes:

Pathophysiology
- Microvascular occlusion (sickled RBCs and increased adhesion to vascular endothelium)
- Endothelial vasoactive mediator disturbance
- Release of inflammatory cytokines
- Activation of coagulation cascade system
- Fat embolism from bone marrow infarction
- Exaggerated regional hypoxic pulmonary vasoconstriction

Ventilatory modalities:

Conventional ventilation
- ARDS type ventilation (low tidal volume, high PEEP, minimal O2)
- Suction and physiotherapy
- Use of N-acetyl cysteine, Dornase alpha
- Bronchoscopy and lavage
- Inhaled nitric oxide*

High frequency oscillation
- With or without nitric oxide*

*As many as 60% adult patients with Hb SS are affected by pulmonary hypertension. Oral agents such as L arginine have been used. During ACS leading to ARDS, the successful use of iNO has been reported in case reports.

Exchange transfusion is invaluable (usually double volume), although even a simple transfusion may increase the haemoglobin A2 sufficiently to dilute the level of HbSS. Antibiotics are normally prescribed to cover for atypical organisms. Data from a large cohort of ACS (adult and paediatric) suggests that infection is a common precipitating factor in ACS, although in the majority, the cause remains unknown.[6]

TABLE 4. CAUSES OF THE ACUTE CHEST SYNDROME. *

CAUSE	ALL EPISODES (N=670)	AGE AT EPISODE OF ACUTE CHEST SYNDROME		
		0–9 YR (N=329)	10–19 YR (N=188)	≥20 YR (N=153)
		no. of episodes (%)		
Fat embolism, with or without infection†	59 (8.8)	24	16	19
Chlamydia‡	48 (7.2)	19	15	14
Mycoplasma§	44 (6.6)	29	7	8
Virus	43 (6.4)	36	5	2
Bacteria	30 (4.5)	13	5	12
Mixed infections	25 (3.7)	16	6	3
Legionella	4 (0.6)	3	0	1
Miscellaneous infections¶	3 (0.4)	0	3	0
Infarction‖	108 (16.1)	50	43	15
Unknown**	306 (45.7)	139	88	79

Vichinsky EP et al. NEJM 2000;342:1855-65

Pneumonia, lung collapse and aspiration syndromes

The principles of ventilating a child with pneumonia or lung collapse/consolidation, aspiration syndromes, pulmonary oedema or pulmonary haemorrhage are similar to ARDS management. Pulmonary aspiration occurs commonly in children with neuromuscular weakness, cerebral palsy, or obtunded airway reflexes. Common causes for aspiration are:

- Accidental (foreign body aspiration)
- Gastro-oesophageal reflux
- Comatose patient
- During intubation (emergency)
- Cerebral palsy
- Bulbar weakness from neuromuscular disease (Guillain Barre syndrome)

Summary of ventilation strategy:

- Minimise oxygen toxicity (FiO2 <0.60)
- Minimise atelectrauma (adequate PEEP)
- Minimise volutrauma (low Tv 5-8 ml/k)
- Permissive hypercapnia

Cystic fibrosis

Mechanical ventilation in children with cystic fibrosis is undertaken for a small number of reasons:

- Infective exacerbations
- Massive haemoptysis
- Post-surgical (after pleurodesis/lobar resection)

However, since most patients with CF have end-stage respiratory failure when they require ventilation, the question of how appropriate ICU care is for these patients is an important one. Although there is paucity of evidence in children, infants with reversible respiratory illnesses (with CF) may have a much better outcome than adults. Favourable results were also obtained among adults admitted post-surgery (pleurodesis) at the Royal Brompton Hospital. These results may indicate that ventilation may be appropriate in the two subgroups mentioned. Ventilation with a view to transplantation has not been advocated in the UK due to restricted organ availability and high post-transplantation mortality.

Thoracic cage abnormalities

Thoracic cage abnormalities such as kyphoscoliosis or chest wall abnormalities lead to restrictive lung disease and often result in respiratory failure from hypoventilation.

Examples
- Jeune's asphyxiating thoracic dystrophy
- Progressive neuromuscular disease
- Cerebral palsy
- Skeletal abnormalities (such as Morquio's syndrome)

Ventilation options
- Non-invasive ventilation (BiPAP, CPAP, negative pressure ventilation)
- Invasive ventilation

The principles of invasive ventilation in these conditions are the same as those applied for ARDS, ie a low tidal volume/low pressure strategy with permissive hypercapnia. Many conditions are often irreversible and palliative care should be considered early.

Other sources of information

Websites
1. http://www.informedhealthonline.org - Good reviews on common critical care conditions, and results of Cochrane reviews (in plain English...!)

References
Key articles attached

1. Willson DF, Thomas NJ, Markovitz BP, et al. Effect of exogenous surfactant (calfactant) in pediatric acute lung injury: a randomized controlled trial. JAMA. 2005 Jan 26;293(4):470-6.

2. Curley MA, Hibberd PL, Fineman LD, et al. Effect of prone positioning on clinical outcomes in children with acute lung injury: a randomized controlled trial. JAMA. 2005 Jul 13;294(2):229-37.

3. Steinberg KP, Hudson LD, Goodman RB et al. Efficacy and safety of corticosteroids for persistent acute respiratory distress syndrome. N Engl J Med. 2006;354:1671-84.

4. Wiedemann HP, Wheeler AP, Bernard GR et al. Comparison of two fluid-management strategies in acute lung injury. ARDS Clinical Trials Network. N Engl J Med 2006;354:2564-75.

5. Gupta VK, Cheifetz IM. Heliox administration in the pediatric intensive care unit: an evidence-based review. Pediatr Crit Care Med. 2005 Mar;6(2):204-11.

6. Vichinsky EP, Neumayr LD, Earles AN, et al. Causes and outcomes of the acute chest syndrome in sickle cell disease. National Acute Chest Syndrome Study Group. N Engl J Med. 2000 Jun 22;342(25):1855-65.