Extracorporeal Life Support Organization (ELSO)

Guidelines for Pediatric Respiratory Failure

Introduction

This pediatric respiratory failure guideline is a supplement to ELSO's "General Guidelines for all ECLS Cases" which describes prolonged extracorporeal life support (ECLS, ECMO). This supplement addresses specific discussion for pediatric respiratory failure.

This guideline describes prolonged extracorporeal life support (ECLS, ECMO). This guideline describes useful and safe practice, but these are not necessarily consensus recommendations. These guidelines are not intended as a standard of care, and are revised at regular intervals as new information, devices, medications, and techniques become available.

The background, rationale, and references for these guidelines are found in "ECMO: Extracorporeal Cardiopulmonary Support in Intensive Care (The Red Book)" published by ELSO. These guidelines address technology and patient management during ECLS. Equally important issues such as personnel, training, credentialing, resources, follow up, reporting, and quality assurance are addressed in other ELSO documents or are center-specific.

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Pediatric Respiratory Cases

I Patient Condition

A. Indications

While no absolute indicators are known, consideration for ECMO is best within the first 7 days of mechanical ventilation at high levels of support

B. Contraindications

1. Recent neurosurgical procedures or intracranial bleeding (within 10 days). Grade II or III intracranial hemorrhage is a general contraindication.

2. Recent surgery or trauma: increased risk of bleeding. While ECMO has been performed successfully in these patients, use of heparinized circuits and/or oxygenators may limit bleeding initially. Care to maintain adequate coagulation factors, platelet counts and use of low ACT's (160-180) may be helpful.

3. Age and size: pediatric patients are >30 days of age and <18 years of age.

No weight limit although obese patients (especially >100kgs) may require special beds, have high risk of decubiti. May also be more difficult to cannulate.

4. Patients with severe neurologic compromise, genetic abnormalities (not including Trisomy 21).

5. Relative: endstage hepatic failure, renal failure, primary pulmonary hypertension.

C. Specific patient considerations

1. Patients with chronic respiratory failure (BPD, emphysema, cystic fibrosis) may be candidates based on pre-ICU status. Need for bronchodilators, diuretics, oxygen therapy and prognosis for long-term survival should be considered prior to offering ECMO. Cystic fibrosis patients should be eligible for lung transplantation

II Vascular Access

A. Venovenous access with a double lumen catheter placed into the internal right atrium via the internal jugular vein is preferred for pts <10-15 kgs. Catheter placement is best done by direct visualization, using direct cannulation for the semi-Seldinger technique described in the general protocol.

B. Venovenous access may also be performed via 2 separate sites. Usually the right internal jugular vein and the right or left femoral veins are used for access. Percutaneous or the semi-Seldinger technique can be performed. Recirculation is limited if drainage from the femoral site and reinfusion via the R IJ site can obtain enough flow to support the patient.

C. Modified venoarterial approach: Venous access may be obtained by femoral or internal jugular route. A low-lying arterial cannula is placed in the femoral artery (usually 18 cm). Saturation of upper body blood flow (head,heart) can be monitored via venous saturation monitor, pulse oximeter (best placed on right hand or ear), NIRS monitor on forehead, or via arterial blood gas line in right radial. Flow from arterial cannula retrograde up aorta will depend on native cardiac function. Low lying cannula can also be rewired to long (50cm) cannula if cardiac output poor and oxygenation to upper body poor.

D. Other: for additional drainage, other venous cannulas can be added into unused femoral or internal jugular sites (while the left IJ has been used, the angle of entry into the heart makes it difficult to get cannula properly placed).

III Patient and disease-specific protocols

A. Selective CO_2 removal.

For status asthmaticus and other conditions in which blood pCO_2 is very high, reducing the blood pCO_2 gradually to avoid acid base imbalance or cerebral complications. A suggested rate of decreasing arterial pCO_2 is 20 mm/Hg/hr.

When selective CO_2 removal is used to treat permissive hypercarbia and to achieve rest lung settings in ARDS, CO_2 can be normalized at acceptable rest lung settings with low blood flow (20% of cardiac output). If the lung failure is severe this can result in major hypoxemia. If the cardiac output and hemoglobin concentration are normal, arterial saturation as low as 75% is safe and well tolerated. However, increasing extracorporeal blood flow to improve oxygenation is preferable to increasing ventilator pressure or FiO_2 when selective CO_2 removal is used.

B. Support of the tracheobronchial tree:

ECMO may be extremely useful in providing airway support during or following surgical repair of the tracheobronchial tree. It allows adequate carbon dioxide removal and oxygenation at low levels of mechanical ventilator support. Use of ECLS may also eliminate the need for endotracheal intubation and use of mechanical ventilator support altogether. This may enhance the ease of surgical repair and subsequently facilitate healing without concern for rupturing suture lines from applied positive pressure.

C. Mediastinal masses:

ECLS may be applied in conditions where anterior mediastinal masses cause airway compression and high risk of death during endotracheal intubation. Application of ECLS under local anesthesia or light sedation with the patient in the upright position may avoid acute death in situations where loss of negative pressure from spontaneous breathing results in collapse of compressed airways which cannot be reexpanded with routine tracheal intubation.

D. Pulmonary embolism

Many patients with primary or secondary ARDS will have small (segmental) pulmonary emboli on contrast CT or angiography. Such emboli do not require any specific treatment aside from the heparinization which accompanies ECLS. When **major or massive pulmonary embolism** is the cause of respiratory/cardiac failure, venoarterial ECLS is very successful management if cannulation and extracorporeal support can be instituted before brain injury occurs. After VA access and successful ECLS is established, document the extent of pulmonary embolism by appropriate imaging studies. Massive pulmonary emboli will usually resolve or move into segmental branches within 48-72 hours of ECLS support. The patient can be weaned from ECLS then from ventilation and managed by pulmonary embolism prophylaxis. Almost all such patients are managed with placement of an inferior vena caval filter. If heart/lung function has not recovered within two days, or if there is a secondary reason to get the patient off ECLS (GI bleeding for example), the patient should undergo pulmonary thrombectomy with cardiopulmonary bypass support. When thrombectomy is done it is usually necessary to continue ECLS for days until lung function is normal.

E. ARDS with secondary lung injury (following shock, trauma, sepsis, etc.)

Once the patient is on ECLS support for stability, adequate repair of secondary organ damage must be performed. If surgical repair of organ injury is required (for example pancreatic resection and drainage for necrotizing pancreatitis, fasciotomy and/or amputation for compartment syndromes and gangrene, excision and drainage of abscesses, etc), these procedures may be adequately performed during ECMO support.

F. Fluid overload

ECLS offers the opportunity to treat massive fluid overload easily. Adequate renal perfusion through native cardiac output or through VA perfusion can be assured minute to minute with appropriate management. As long as renal perfusion is adequate, pharmacologic diuresis can be instituted and maintained even in septic patients with active capillary leak. Continuous hemofiltration may be added to the circuit if pharmacologic diuresis is inadequate. The hourly fluid balance goal should be set and maintained until normal extracellular fluid volume is reached (no systemic edema, within 5% of “dry” weight). Use of renal replacement therapy to enhance fluid removal and allow adequate nutritional support is often performed. Despite the literature surrounding fluid overload (>10%) as a risk factor for death, review of the ELSO registry also finds that use of renal replacement therapy is also a risk factor for poor outcome. Even if acute renal failure occurs with ECLS, resolution in survivors occurs in >90% of patients without need for long-term dialysis.

G. Post ECLS recovery and management

A patient is weaned off ECLS on non-damaging ventilator settings as described in V. If respiratory function is tenuous the vascular access catheters can be left in place as described in V. Once the patient is off ECLS ventilator weaning continues per unit protocol. There is a tendency to drift into positive fluid balance, more sedation, increasing ventilator settings which should be carefully avoided. Tracheostomy is not often performed in pediatric ECLS but may be indicated if respiratory improvement to allow cessation of mechanical ventilation is prolonged.

Patients who experience severe lung injury from necrotizing pneumonia, or from very high plateau pressures prior to ECLS will have the physiologic syndrome of very high alveolar level dead space. This is characterized by adequate oxygenation on low FiO_2 but CO_2 retention, respiratory acidosis, the need for hyperventilation (either spontaneous or via the ventilator) to maintain $PaCO_2$ under 60, and an emphysematous (honeycomb) appearance on chest x-ray or CT scan. Although this condition has the characteristics of chronic irreversible obstructive lung disease, it may reverse to normal within 1-6 weeks. These conditions heal by contracture eliminating the alveolar level dead space.

H. Lung biopsy

The cause of severe respiratory failure may be unknown when the patient is started on ECLS. Although lung biopsy is the next step in diagnosis, it is potentially dangerous in patients on ECLS with anticoagulation. If pulmonary function rapidly improves during ECLS (the first few days) lung biopsy may be delayed until the patient is off anticoagulation. However, if pulmonary function is not improving after several weeks, the primary diagnosis has not been established by bronchoscopy or other means and the status of lung recovery uncertain, lung biopsy can and should be performed. Lung biopsy is best done by thoracotomy (or thoracoscopy) rather than transbronchially because of the risk of major hemorrhage into the airway with transbronchial biopsy.

I. Rare conditions

ECLS has been used for rare causes of pulmonary failure with variable success. When considering ECLS for a specific diagnosis for the first time in any given center it may be helpful to consult the ELSO registry for the worldwide experience with that condition. Examples are vasculitis, autoimmune lung disease, bronchiolitis, obliterans, Goodpasture syndrome, rare bacterial, fungal or viral infections.

IV Weaning and follow-up

Patients should have a thorough neurologic evaluation, including imaging with a CT or MRI scan, done prior to hospital discharge. A follow-up plan that involves a minimum of yearly patient assessment should also be developed. A minimum level of progress in normal developmental milestones, school performance, required medications and subsequent hospitalizations post-ECMO should be obtained.