

Surgical Management of CDH

Background:

Neonates with Congenital Diaphragmatic Hernia of Bochdalek continue to experience high morbidity and mortality. While clinical studies are limited by small numbers, selection bias, and single center data these efforts suggest that standardization of both medical and surgical care has the potential to influence outcomes. Furthermore, the lack of standardization in many institutions, such as CHLA, precludes isolating and assessing specific variables and their impact upon clinical results.

Recognizing that CDH infants may fall into several disparate physiological categories, it is necessary to examine standardization of the “stable” infant who does not require ECMO, the unstable infant who requires ECMO within the first 48 hours of life, the unstable infant who requires ECMO after 48 hours of life, and the children who go on ECMO and are weaned off or are deemed “unweanable.” Though these are distinctions subject to the judgments of the involved clinicians, they are based on observed mortality rates and a logical stratification of risk on ECMO. Regardless, these are separations that may be revisited or modified as data is obtained to refine risk profiles.

Logistics:

The rounding Pediatric Surgeon and Neonatologists will confer at least once daily to review CDH patient status and next steps. In the event that a specific patient is being cared for by a surgeon other than the rounder, said surgeon will communicate daily with the attending neonatologist. At the point that a child has met criteria to undergo operative repair, the responsible surgeon will prioritize the scheduling of the operation. If the surgeon caring for the child will be unable to meet the time requirements, the case will be addressed by the rounding surgeon.

Equipoise:

As the current medical literature does not support selection of any one approach to children with CDH placed upon ECMO, we believe there is sufficient equipoise to support a two pathway approach to the disorder.

Group I (“Low Risk”): These infants meet the following MINIMUM criteria to undergo operative repair (type TBD by surgeon). Once infants meet the criteria below, they will undergo repair within 24 hours:

- Demographics:
 - Age > 72 hours
 - No severe CHD, lethal syndromes, severe comorbidities, or other contraindications to “early” operation that could logically improve with time.
- Physiology:
 - Blood pressure > normal minimum for gestational age with normal perfusion
 - FiO₂ < 0.5
 - Pre-ductal SaO₂ > 96% on average (episodic, non-prolonged drops are acceptable)
 - Pre – Post-ductal gradient < 10%
 - Conventional ventilation (must be off oscillator)
 - MAP < 18cm H₂O
 - No significant infections (pneumonia, blood stream, urinary)
- Medications:
 - No greater than 5 mcg/kg/min dopamine [preferably none]

- NO < 20ppm [though this may present problems for going to the OR]
- Sildenafil, allowable
- Special Studies:
 - ECHO demonstrating PA pressures < 80% Systemic and no structural cardiac disease other than PDA, ASD, VSD.

While individual surgeons may continue to exercise their clinical judgment as circumstances may require, the general approaches for neonates requiring ECMO will be as follows:

Group IIa (Weaned from ECMO): These infants are placed on ECMO and are able to be weaned and removed from the circuit AND meet the following criteria prior to operation:

- Meet all criteria of the “low risk” group above.
- At or near predicted “dry” weight

These infants will undergo operation within 24 hours of meeting criteria so long as resources and operating room time are appropriately available. For example, the Surgeon, Neonatologist, and Anesthesiologist may elect to forego corrective procedures at night or on weekends when there are fewer resources available.

Group IIb (“Unweanable” from ECMO): These infants have undergone all reasonable measures to support liberation from ECMO but by consensus opinion of Attending Neonatologist and Attending Surgeon are deemed to be unweanable at approximately the 2 week point from ECMO initiation. This may include, but is not limited to, the use of multiple pharmaceutical pulmonary dilators, lung recruitment maneuvers, trial of different ventilators and modes, diuresis to dry weight, and inotropic support. Because institutional data suggests there may be a small percentage of these children that will survive, operation will be performed unless there are clear contraindications.

These children will undergo open operation in the NICU (thoracotomy or laparotomy at attending surgeon’s discretion) at the point that they are physiologically optimized but generally not to exceed 14 days on ECMO. In recognition of the mounting risk of severe post-operative complications on ECMO and the rapidly diminishing chances of successful liberation from ECMO with time, all parties agree that the child must be removed from ECMO within 12 hours of completion of operation. Furthermore, team members understand that the infants liberated from ECMO at this juncture may not survive. Important conditions include:

Family engagement: The family must be informed that operation on ECMO carries a variety of potential risks including significant hemorrhage, transfusion, patch use, recurrence, and death. Moreover, families will be informed of the intent to liberate the infant from ECMO within 12 hours post-operatively regardless of physiologic state. Operation will only proceed with explicit family approval.

Patient status: The child should not have any comorbidities beyond those reasonably anticipated for a CDH on ECMO (intentional coagulopathy, pulmonary hypertension, depressed cardiac function, PDA, low urine output) the mitigation of which would significantly impact surgical risk or is a contraindication to further support (severe hepatic dysfunction, severe coagulopathy not related to heparin use, severe neurologic injury, severe sepsis).

Operative Preparation on ECMO:

Given the possible risks of operative intervention on ECMO, specific measures are required to best care for these children including at a minimum:

-Informed consent

-40mL/kg PRBCs and appropriate ratio of FFP, cryoprecipitate, and platelets in the room at the time of operation.

-Presence of the attending Neonatologist for the duration of the operation to monitor and manage the ECMO circuit.

-Presence of the full Anesthesia and OR team for the procedure.

-Anticoagulation High Risk Bleeding protocol: Heparin gtt 10units/kg/hr with goal Anti-Xa level less than 0.3

-Amicar 100 mg/kg bolus 2 hrs before incision and continuous gtt at 30mg/kg/hr for 48hr after the operation. May be longer if ongoing postoperative bleeding but not longer than 72hrs. Shorter continuous gtt if postop risk is low (depending on the operation) and clots are noted on the cannulas.

Group III (Early Repair on ECMO):

After appropriate stabilization, parental consent, and within 24 hours of initiation of ECMO, the responsible surgeon may opt to undertake open operative repair in the NICU (thoracotomy or laparotomy at the surgeon's discretion). All attempts will be made to use higher flows and low or no anticoagulation for at least 24-72 hours after the initiation of operative repair to mitigate bleeding risk. Contraindications to proceeding may include the following:

Genetics and Syndromes: If a child is placed on ECMO but there is concern that there is a lethal complex, syndrome, or genetic disorder that may reasonably be determined in the first week and, thus, the team may elect to defer operation.

Family Engagement: The family must be informed that operation on ECMO carries a variety of potential risks including significant hemorrhage, transfusion, patch use, recurrence, and death. Operation will only proceed with explicit family approval.

Patient Status: The child should not have any comorbidities beyond those reasonably anticipated for a CDH on ECMO (intentional coagulopathy, pulmonary hypertension, depressed cardiac function, PDA, low urine output) the mitigation of which would significantly impact surgical risk or is a contraindication to further support (severe hepatic dysfunction, severe coagulopathy not related to heparin use, other site severe bleeding, severe neurologic injury, severe sepsis).

Operative Preparation: Given the possible risks of operative intervention on ECMO, specific measures are required to best care for these children including at a minimum:

-Informed consent

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Key References:

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