NEONATE FOR REPAIR OF DIAPHRAGMATIC HERNIA ON ECMO

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OBJECTIVES

1. Review common treatment strategies in patients with diaphragmatic hernia
2. Discuss indications and contraindications for initiation of ECMO support
3. Review issues related to timing and location of surgical repair of diaphragmatic hernia in patients requiring support with extracorporeal membrane oxygenation (ECMO)
4. Review anesthetic options for patients on veno-arterial and veno-venous ECMO
5. Discuss management of anticoagulation for patients requiring surgical repair while on ECMO
6. Review risks and benefits of antifibrinolytic therapy for patients requiring surgical repair while on ECMO

STEM CASE – KEY QUESTIONS

A 3.5-kg, 51-cm girl with a prenatally diagnosed diaphragmatic hernia was born at 39-2/7 weeks via induced vaginal delivery to a healthy 25-year-old G1P0 mother. The neonate had strong spontaneous crying immediately after birth, with APGAR scores of 9 at both 1 and 5 minutes. Because of the known diaphragmatic hernia, she was intubated in the delivery room and remained adequately oxygenated on modest mechanical ventilation settings. However, on the second day-of-life her oxygen requirements increased requiring initiation of high-frequency oscillation ventilation (HFOV) and inhaled nitric oxide. Chest radiograph is shown in Figure 1. Severe pulmonary hypertension and right ventricular dysfunction were seen with transthoracic echocardiography. With increasing doses of inotrope (dopamine 15 mcg/kg/min) needed to support her hemodynamics, the decision was made to place the neonate on veno-arterial ECMO.

Cannulation of the right carotid artery (8-Fr) and right internal jugular vein (14-Fr) via cut-down was performed in the intensive care unit. ECMO flow was maintained between 1.5 and 2.0 liters/min/m². A vasopressin infusion was used initially to maintain a mean blood pressure >40 mmHg while the dopamine was weaned off. On ECMO-day 2 she underwent patent ductus arteriosus ligation via left thoracotomy to correct the persistent large left-to-right shunt.

On ECMO she was anticoagulated with heparin (4-16 units/kg/hour) to maintain an activated clotting time (ACT) (I-stat® with kaolin cartridge) of >160 seconds. On ECMO-days 3-4 she exhibited a mild coagulopathy requiring transfusion of fresh frozen
plasma, cryoprecipitate and platelets to maintain adequate coagulation parameters. Sedation was achieved with infusions of midazolam and morphine, and low concentrations (0.2-0.4% inspired) isoflurane. With little improvement of aeration due to persistent bilateral lung consolidation seen on multiple chest radiographs, she remained on HFOV (mean 14 cmH\textsubscript{2}O, amplitude 36 cmH\textsubscript{2}O, 8 hZ, FiO\textsubscript{2} 21%).

Figure 1. Chest radiograph from day-of-life 2 prior to initiation of ECMO

On ECMO-day 5 she was hemodynamically stable, and did not require any inotrope or vasopressor infusions. Her hemoglobin was 10.7 g/dL. Coagulation tests showed a prothrombin time (PT) of 11.6 sec, partial thromboplastin time (PTT) between 52-63 sec, fibrinogen of 147 mg/dL and a platelet count of 86,000/mm\textsuperscript{3}. Heparinase thromboelastogram (kaolin) showed R=16.7 mm, angle (\(\alpha\))= 28.7\(^{\circ}\), mean amplitude (MA)=50.1 mm and no fibrinolysis. On her morning chest radiograph, a large pneumothorax with pneumoperitoneum was visualized, presumably the result of barotrauma. Because of the pneumothorax, the HFOV mean pressure was reduced to 10 cmH\textsubscript{2}O and amplitude reduced to 28 cmH\textsubscript{2}O. Rather than simply placing a chest tube, the pediatric surgical team requested to repair the diaphragmatic hernia that day.
DISCUSSION

Treatment of CDH and Indications for ECMO Therapy
What are the different ventilation strategies/modes used in these patients? What is the role of nitric oxide, and surfactant? What cardiovascular and/or respiratory parameters determine the need for ECMO support? What measures of oxygenation are used as indications/contraindications for ECMO?

Once ECMO support has been initiated, there are several issues that need to be addressed prior to proceeding with diaphragmatic hernia repair.

Timing
We must ask if this is the optimal time for surgical repair. As some centers suggest, should repair be delayed until the patient can be weaned from ECMO, or until they are off ECMO completely? Will the repair at this time help lung aeration or hemodynamics improve? If proceeding with repair, is the patient in optimal condition for the procedure? Is repair in the intensive care unit appropriate, or would transporting the patient to the operating room provide better operating conditions?

Anesthetic Delivery
Does the fact that the patient is currently on ECMO affect your anesthetic technique? Does the ECMO cannulation (veno-arterial vs. veno-venous) affect your technique? Should the patient be maintained on HFOV, or converted to traditional mechanical ventilation for the procedure?

**Anticoagulation**
The anticoagulation required for ECMO remains a challenge, potentially leading to increased bleeding and transfusion requirement related to the surgical repair. Prior to proceeding with repair, what values for the various coagulation parameters will you consider adequate? How will you achieve these values in this patient? Will you adjust the rate of heparin infusion for the procedure and/or the postoperative period? What are your target ACT and PTT values for this time period? Will these be different if the entire circuit, including cannulas, are heparin-coated?

**Antifibrinolytic Therapy**
Numerous studies suggest there are less ECMO-related bleeding and hemorrhagic complications with administration of the lysine analogues aminocaproic acid or tranexamic acid. However there are also reports of thrombotic events occurring when these agents are used. Will you administer aminocaproic acid or tranexamic acid during or after the procedure? What are potential circuit-related consequences of using these agents?

**SELECT REFERENCES**


