

Case Reports

A Modified EXIT-to-ECMO with Optional Reservoir Circuit for Use during an EXIT Procedure Requiring Thoracic Surgery

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Abstract: A 34 year old mother with a history of polyhydramnios and premature rupture of membranes presented for an ex utero intrapartum treatment (EXIT) procedure to deliver her 34 week gestation fetus. The fetus had been diagnosed with a large cervical mass which significantly extended into the right chest. The mass compressed and deviated the airway and major neck vessels posteriorly. Imaging also revealed possible tumor involvement with the superior vena cava and right atrium. The plan was for potential extracorporeal membrane oxygenation (ECMO) during the EXIT procedure (EXIT-to-ECMO) and the potential for traditional cardiopulmonary bypass (CPB) for mediastinal tumor resection. A Modified EXIT-To-ECMO with Optional Reservoir (METEOR) circuit was devised to satisfy both therapies. A fetal airway could not be established during the EXIT procedure and so the EXIT-to-ECMO strat-

egy was utilized. The fetus was then delivered and transferred to an adjoining operating room (OR). Traditional cardiopulmonary bypass with a cardiotomy venous reservoir (CVR) was utilized during the establishment of an airway, tumor biopsy and partial resection. The patient was eventually transitioned to our institution's standard ECMO circuit and then transferred to the intensive care unit. The patient was weaned from ECMO on day of life (DOL) eight and had a successful tumor resection on DOL 11. The patient required hospitalization for numerous interventions including cardiac surgery at 4 months of age. She was discharged to home at 5 months of age. **Keywords:** ex utero intrapartum treatment (EXIT) procedure, extracorporeal membrane oxygenation, ECMO, cardiopulmonary bypass, CPB, hybrid bypass circuit, cervical teratoma, METEOR circuit. *JECT. 2016;48:35–38*

OVERVIEW

Fetal neck masses can result in significant morbidity and mortality if not identified prenatally (1,2). Although fetal ultrasound and magnetic resonance imaging (MRI) allow for excellent assessment of neck masses, these masses result in potentially life-threatening airway compromise for the neonate upon delivery (1,3). The ex utero intrapartum treatment (EXIT) procedure was originally developed as a therapy for congenital diaphragmatic hernia patients in whom in utero tracheal occlusion was used to promote

pulmonary development (4,5). The EXIT procedure is performed with a partial cesarean delivery, which exposes the fetal head and neck. The fetus remains on intrapartum uteroplacental support while the airway is assessed and established. Uterine relaxation using high dose halogenated agents along with maintenance of stable maternal hemodynamics is crucial during uteroplacental support (6). Extracorporeal membrane oxygenation (ECMO) can be used if an airway cannot be established, if the fetus and its placental support become unstable, or if the lungs cannot support the patient during the initial neonatal period. This has been termed the EXIT-to-ECMO strategy (7).

We describe a case where the fetus presented with a significant cervical teratoma, which significantly extended into the right chest. There was potential for ECMO during the EXIT procedure (EXIT-to-ECMO) using central cannulation as cervical cannulation was not an option because of the size and location of the mass and the potential need for traditional cardiopulmonary bypass (CPB) for mediastinal

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tumor resection. A Modified EXIT-To-ECMO with Optional Reservoir (METEOR) circuit was devised to satisfy both therapies.

DESCRIPTION

Patient History

A 34-year-old African-American woman, with a history of polyhydramnios and premature rupture of membranes, presented with a female fetus in whom a large cervical mass was identified via fetal ultrasound and MRI. The cervical mass extended from the oropharynx into the thoracic outlet and was positioned slightly to the right of midline. Imaging was suggestive of the mass to be a teratoma with possible involvement of the superior vena cava and right atrium. The fetus was estimated to be 2.5 kg via ultrasound using gestational age, sex, and fetal measurements. A large multidisciplinary team from two institutions was assembled for a planned EXIT procedure and the potential for immediate thoracic surgery.

The Extracorporeal Support Plan

ECMO has been described as a useful therapy for providing pulmonary support during the EXIT procedure if an airway cannot be established while on placental support (1,3). ECMO has the advantage of being a closed system. This may be particularly important for a fetus with limited hemodynamic monitoring. The closed ECMO system maintains the patient in a euvoletic state and allows for normal cardiac ejection as long as the circuit prime is warm and properly balanced for electrolyte values. Maintaining cardiac ejection is essential during the EXIT procedure since there is a period of time when there is no dynamic blood pressure monitoring of the neonate. A closed extracorporeal system allows for oxygenation and ventilation with little, if any, negative effect on cardiovascular dynamics.

A traditional CPB system with an open reservoir would be less than ideal in this setting. An open reservoir system is designed to efficiently empty the heart (8). This would not be preferred during the EXIT procedure since maintenance of ventricular ejection is desired to maintain effective circulation. An empty heart on traditional CPB in the setting of an uncontrolled patent ductus arteriosus (PDA) could result in distention of the left heart. Inadequate cerebral blood flow may also result because of circulatory steal to the pulmonary circulation and ultimately the left side of the heart. Normally, the perfusionist can judge and effect the ventricular status with integration of left atrial, central venous, and dynamic arterial blood pressure monitoring values. Those values are not available during the EXIT procedure. Furthermore, before placing a neonate on traditional CPB, there is normally time to dissect out the PDA for control immediately before or once

bypass is initiated. That is also not feasible during the EXIT procedure.

The METEOR circuit allows for ECMO support with an option for traditional CPB with an open reservoir and cardiomy suction. Figure 1 provides a simplified schematic of the METEOR circuit. Switching between the two therapies only requires moving paired clamps and the patient remains on extracorporeal support at all times. This system allows use of an institution's standard bypass circuit without significant changes to the heart-lung machine settings. The addition of the parallel ECMO limb to the circuit only required adding servo-regulated pressure monitoring for the ECMO bladder. A Terumo CDI (Terumo Cardiovascular, Inc., Ann Arbor, MI) in-line blood gas monitor was used throughout extracorporeal support. The origin of flow for arterial monitoring was a post oxygenator port for both therapies. The return for the CDI shunt sensor flow was to the cardiomy venous reservoir during traditional bypass and to the ECMO bladder during ECMO support. A cuvette in the venous limb of the circuit was used as well for venous saturation and hematocrit monitoring during both therapies.

The extracorporeal support plan was to use EXIT-to-ECMO with the option of traditional CPB with an open reservoir once the patient was delivered and transferred to a separate operating room (OR). The CPB circuit consisted of a Terumo CAPIOX FX05 oxygenator with hard-shell reservoir (Terumo Cardiovascular, Inc., Minneapolis, MN),

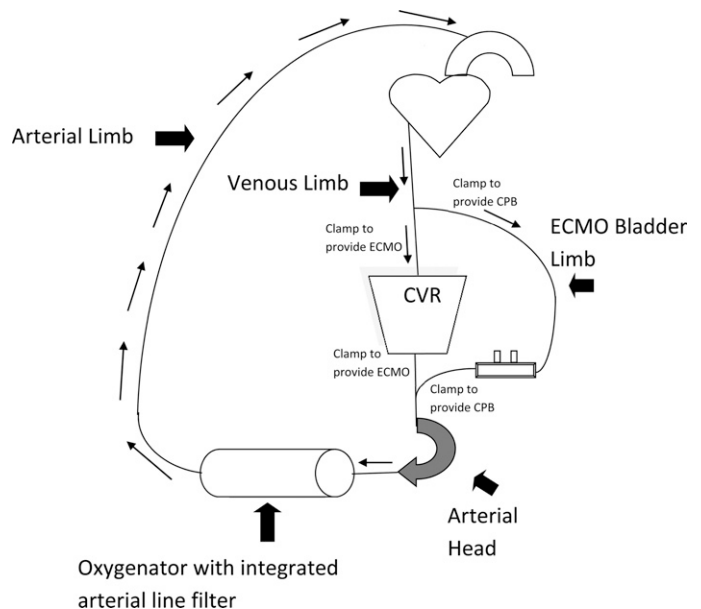


Figure 1. Simplified schematic of a hybrid ECMO-CPB circuit. Paired tubing clamps are placed on the “ECMO” label areas to provide ECMO support. These clamps may be moved to the “CPB” label areas to allow for traditional cardiopulmonary bypass with cardiomy suction. Adapted from *Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population* (9), with permission of John Wiley & Sons (ISBN 978-1-118-90079-6).

and a custom tubing pack with 3/16" arterial and boot lines and a 1/4" venous line (Sorin Group USA, Arvada, CO). A 30-mL ECMO bladder (Medtronic Inc., Minneapolis, MN) was spliced into the circuit for creation of the METEOR circuit. The circuit was CO₂ flushed before being crystalloid primed and circulated through a 5 µm prebypass filter. The final 300 mL circuit prime consisted of 50 mL Plasma Lyte-A 7.4 (Baxter Healthcare, Deerfield, IL), 250 mL of red blood cells with plasma added, 900 U heparin (3 U heparin/mL of prime), 9 mEq sodium bicarbonate, and 1000 mg calcium gluconate. Antibiotic coverage with 63 mg cefazolin (25 mg/kg) and steroids consisting of 75 mg methylprednisolone (30 mg/kg) were also added before instituting support. A lab sample of the circuit prime revealed normal pH, blood gas, and electrolyte values. The circuit prime was maintained at 35°C.

Operative Course

The mother was anesthetized and the EXIT procedure commenced. A uterine incision was made as a continuous amnioinfusion of warm crystalloid solution was infused into the uterus. This infusion helps maintain uterine volume and temperature while additionally helping prevent umbilical cord spasm (10). The infant's head, neck, and upper chest were delivered and the airway was examined by the otorhinolaryngology team. The large neck mass was externally visible, and on direct laryngoscopic examination of the upper airway, no tracheal lumen was identified as the fetus became bradycardic. The infant's chest and upper abdomen were delivered. An emergent sternotomy was performed and 750 U of heparin (300 U/kg) were administered through the right atrial appendage. The aorta was cannulated with an 8 French Biomedicus arterial cannulae (Medtronic Inc., Minneapolis, MN), whereas the right atrium was cannulated with single 12 French angled metal tip venous cannulae (Medtronic Inc.). Within three minutes, ECMO support was initiated at 250 mL/min (100 mL/kg), the umbilical cord was clamped and the baby was fully delivered. The cardiac surgeon moved her to a small instrument table, which was prepared as the transport bed. ECMO flow was maintained at 250 mL/min as measured by an arterial limb flow probe and ECMO bladder pressure was closely monitored to ensure adequate venous drainage. The venous blood oxygen saturation was 85–90% with ECMO flows at 100 mL/kg.

The patient was transferred to an adjacent OR and an umbilical artery line was used to monitor the arterial blood pressure. Target parameters included an arterial blood pressure of 45–50 mmHg, hematocrit 35–40%, normothermia, venous saturation ≥85%, and a normal electrocardiogram and near-infrared spectroscopy values at or above their baseline of 75%. Once in the new OR while still on ECMO support, the infant's airway was more carefully examined by the otorhinolaryngology team, who

were still unable to visualize or establish an antegrade airway. The trachea was visualized in the mediastinum and a tracheotomy was performed in the mediastinal trachea. Through this opening, a suction catheter was passed retrograde, over which an endotracheal tube was then passed. The suction catheter and endotracheal tube were pulled through the mouth into the airway antegrade from the mediastinum. Once the baby was orotracheally intubated, the tracheotomy was repaired. With the airway established, attempts were made to expand the lungs, which could only accommodate tidal volumes of less than 5mL/kg. With the airway established, the decision was made to transition the patient to traditional CPB from ECMO. The paired bypass clamps for transitioning therapies were moved. The hardshell reservoir venous inlet and venous reservoir outlet clamps were moved to the parallel ECMO limb inflow and outflow limbs. Once on traditional CPB, tumor biopsy was performed for diagnosis. Gas exchange was thought to be insufficient without extracorporeal support. The plan was to maintain the patient on ECMO in the intensive care unit (ICU) until signs of lung recovery were evident. Complete resection of the tumor was deferred until circulatory support was no longer necessary. The patient was not transitioned to bicaval cannulation since it was decided to delay full tumor resection. It was determined at this time that tumor resection would best be performed off of extracorporeal support and without systemic anticoagulation. The patient was transitioned from traditional bypass to ECMO via the METEOR circuit while our institution's traditional ECMO circuit was prepared. Volume supplementation during both ECMO periods was achieved by briefly unclamping the venous reservoir outlet while simultaneously clamping the ECMO bladder outlet tubing. The patient was transitioned to the traditional ECMO circuit and then transported to the cardiac ICU.

The patient was weaned from ECMO support in the ICU on DOL 8. The teratoma was resected in the OR without CPB on DOL 11. The patient remained in the hospital and required surgery with CPB for repair of supravalvar aortic stenosis and removal of a transverse aortic arch thrombus at 4 months of age. The patient was discharged to home at 5 months of age.

COMMENTS

ECMO has been successfully used during EXIT procedures in which an airway could not be established (3,5,7). This patient presented with a rare and unique cervical teratoma. It was surmised that there was a high likelihood that cardiothoracic surgery would be needed shortly after delivery since the tumor potentially involved the superior vena cava and right atrium. A perfusion circuit was devised

which allowed ECMO during the EXIT procedure with a simple transition to traditional cardiopulmonary bypass if the need arose for tumor resection. The METEOR circuit allowed for seamless transitions between the two extracorporeal therapies.

It is essential to provide a properly balanced bypass circuit blood prime for the EXIT procedure. If the circuit prime were to be cold or if it had abnormal electrolyte values, the baby's heart could develop an arrhythmia or asystole once placed on extracorporeal support (with either venoarterial ECMO or traditional CPB). If this occurred with an uncontrolled PDA, the heart could distend and/or there could be inadequate cerebral blood flow due to circulatory steal to the pulmonary vasculature and ultimately the left side of the heart. This could also happen if the heart were to be emptied out with traditional CPB with an open system. Peng et al. published a case report including an EXIT to CPB but this was in the setting of hypoplastic left heart syndrome presenting for atrial septostomy (8). His team cannulated centrally and they were able to avoid the concerns we had in devising the METEOR circuit since the arterial cannula was inserted into the pulmonary artery and advanced into the ductus arteriosus. Their venous cannulation strategy required bicaval placement. Our patient did not have complex congenital cardiac disease requiring this type of cannulation. We believe that the METEOR circuit is a desirable option when traditional CPB with cardiotomy field suction

and an open reservoir may be needed after an EXIT-to-ECMO procedure.

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