Ex-utero intrapartum treatment as a novel bridging strategy to surgery in hypoplastic left heart syndrome with intact atrial septum—cross-circulation revisited

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Hypoplastic left heart syndrome with an intact or nearly intact septum (HLHS-IS/NIS) constitutes 6%-20% of hypoplastic left heart syndrome cases and represents the most severe form of the disease.1-3 Unless there is a pathway for pulmonary venous decompression, hemodynamic instability and death ensue within minutes to hours after birth. Survival depends upon emergent relief of this obstruction, either by operative or interventional techniques. The optimal strategy to bridge a newborn to a stable circulation remains undefined.1,2,4 We describe a novel EXIT (ex-utero intrapartum treatment) strategy using maternal-placental support to bridge a fetus with HLHS-IS/NIS to palliative intervention.

A 26-year-old mother was referred for a fetal echocardiogram (at 30 weeks gestation), and the diagnosis of HLHS-IS/NIS (with aortic atresia, mitral stenosis) was confirmed (Figure 1, A). The family elected to proceed with fetal intervention, which was performed (at 31 weeks) in another institution. This intervention was complicated by interatrial stent malposition with no demonstrable flow across the septum, and pleural effusion (Figure 1, B). As delivery neared, the parents requested aggressive management. Although our usual approach at birth is immediate catheter intervention, this strategy would be complicated by the oblique stent position in the right atrium. An EXIT to achieve cardiopulmonary bypass (CPB) was proposed to allow the fetus to remain on placental support while undergoing median sternotomy and initiation of CPB. The strategy was the consensus from a multidisciplinary fetal care conference as the safest means of maintaining hemodynamic stability before establishing interatrial communication.

After the mother suffered premature rupture of membranes, a multidisciplinary team was mobilized for an emergent EXIT procedure. General anesthesia was induced using total intravenous anesthesia to minimize transplacental passage of inhalational agent and fetal myocardial depression. A Pfannenstiel incision was made to expose the lower uterine segment, and the fetal head and upper extremities were delivered through the hysterotomy. Once the uterus was exposed, the total intravenous anesthesia was switched to a high-dose desflurane inhalational agent to provide maximal uterine relaxation and preserve utero-placental gas exchange. The amniotic cavity was infused with warm fluid (37°C) via a level-1 rapid infusion device to prevent cord compression. A cocktail of fentanyl, atropine, and rocuronium was injected into the fetal deltoid muscle to supplement the transplacental inhalational anesthetics. A 24-gauge angiocatheter and a pulse oximeter were placed on the right hand. The fetus was intubated and tube position was confirmed by flexible bronchoscopy.

A median sternotomy was performed while maintaining position of the fetus partially within the womb (Figure 1, C). This approach avoided an empty uterus, which otherwise could have stimulated strong uterine contraction and placental separation. After heparinization through a purse string in the right atrium, a 1.6-French right-atrium cannula and an 8-French arterial cannula (distal pulmonary artery into ductus arteriosus) were placed to initiate CPB. In coordination with the anesthesiologists, the inhalational agent was reduced to allow uterine tone to return, and when adequate, CPB was initiated. An immediate umbilical cord constriction occurred due to high oxygen tension. The umbilical cord was then divided (Figure 1, D). The newborn was transferred to the adjacent operating table (Figure 1, E). Intravenous prostaglandin infusion was initiated. Following bicaval cannulation and right atriotomy, the stent was found abutting the septum, partly adherent to the right atrium free wall without evidence of interatrial communication (Figure 1, F). The stent was removed, and atrial septectomy was performed without cardiopлегic arrest, taking care to avoid systemic air embolism. The bypass was then weaned off on inotropes.

The postoperative course was challenging, with hemodynamic instability and highly variable pulmonary blood flow. The systemic oxygen delivery was marginal with pulmonary overcirculation. Therefore, the patient underwent bilateral branch pulmonary artery–banding (3 mm; day 4 of life). The lungs had the appearance of significant pulmonary lymphangiectasia intraoperatively.
Despite aggressive efforts, the patient developed profound systemic inflammatory response with diffuse capillary leak and was allowed to die (day 10 of life).

The management of HLHS-IS/NIS remains most challenging in the current era, with high mortality regardless of intervention. Attrition continues, even with initial successful stage-1 or stage-2 palliation.\textsuperscript{2,4} In-utero left atrial and pulmonary hypertension leads to pathologic lymphatic changes and pulmonary venous “arterialization.”\textsuperscript{2,4} Our strategy is reminiscent the earliest era of congenital heart surgery in which maternal-child cross-circulation was used very successfully. This concept was then utilized for fetuses with upper-airway obstruction, to allow time for the airway to be secured before separation from maternal-placental circulation.\textsuperscript{5}

In our case, alternative EXIT strategies would include: (1) open atrial septectomy with inflow occlusion, (2) neck cannulation for extracorporeal membrane oxygenation with immediate balloon atrial septostomy, (3) sternotomy with per-atrial balloon septostomy. We concluded that the best chance for survival without profound cardiac instability or neurologic injury would be rapid left atrial decompression under the most controlled circumstances. In this patient, an initial stabilization with peripheral extracorporeal membrane bypass was utilized.

\begin{figure}
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\caption{A, Fetal echocardiogram preintervention: 4-chamber view showed fetal hypoplastic left heart syndrome with a thickened atrial septum bowing into the right atrium (denoted by \textit{asterisk} $[\ast]$). B, Preintervention fetal echocardiogram showed a severely abnormal to-fro pulmonary venous Doppler pattern (forward-to-reverse velocity-time integral flow ratio of 1.7) without evidence of a decompressing vessel. Postintervention, there was a persistent abnormal to-fro flow pattern (forward-to-reverse velocity-time integral flow ratio of 1.6), with no demonstrable flow across the atrial septum. C, With the fetus partially delivered (pelvis and lower limbs remained in-utero), and the umbilical cord still intact, an incision was made in the midline for sternotomy. D, After cannulation of the ductus arteriosus and right atrium, CPB was initiated. The umbilical cord was then clamped and divided as shown. E, The diagram shows the layout of the fetal surgery operating room for the EXIT to CPB procedure (\textit{green}: cardiac surgeons; \textit{red}: fetal surgeons; \textit{yellow}: anesthesiologist; \textit{orange}: scrub nurses; \textit{gray}: cardiologist; \textit{blue}: operating table). F, Intraoperative photo showing interatrial stent in the right atrium (\textit{white arrow}). \textit{LV}, Left ventricle; \textit{LA}, left atrium; \textit{RV}, right ventricle; \textit{RA}, right atrium; \textit{CPB}, cardiopulmonary bypass.}
\end{figure}
Oxygenation would not have achieved effective cardiac decompression in the absence of an interatrial communication, and the need to remove an intracardiac foreign body and perform atrial septectomy required immediate open heart surgery. The EXIT strategy described here could conceivably be used when immediate open heart surgery is required, as an approach to secure a fetus hemodynamically before separation from maternal-placental circulation.

References

Complete thoracoscopic S9 or S10 segmentectomy through a pulmonary ligament approach

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Video clip is available online.

The pulmonary ligament (PL) forms the intersegmental septum and enters the lung parenchyma. Consequently, the PL can be easily separated from the lung parenchyma at surgery. We report thoracoscopic segmentectomy of the lateral basal segment (S9), the posterior basal segment (S10), or both by a new approach that does not require interlobar separation of the lung.

SURGICAL TECHNIQUE

Since March 2009, we have performed thoracoscopic segmentectomy of S9, S10, or both through a PL approach in 23 patients in whom a nodular shadow or ground-glass opacity measuring 2 cm or less was found in S9 or S10 on chest computed tomography. All patients were evaluated for the absence of extrapulmonary metastatic disease by fluorodeoxyglucose F 18 (INN fluorodeoxyglucose [18F]) positron emission tomographic scan before resection. Reconstruction of 3-dimensional models by preoperative computed tomography has been previously described (Figure 1, A). Involved pulmonary vessels and bronchi were identified, and the surgical procedure was determined on the basis of such factors as tumor location and margin status. Surgery was performed with the patient under general anesthesia with single-lung ventilation and positioned in the lateral decubitus position. A small skin incision, 2.5 to 4 cm in length, was made along the anterior axillary line at the fifth intercostal space. A camera port was placed at the midaxillary line of the seventh or eighth intercostal space, and an assist port was placed at the posterior axillary line of the seventh or eighth intercostal space. First, the PL was incised up to the inferior pulmonary vein, and the basal pulmonary vein was exposed (Figure 1, B and C, Figure 2, A, and Video 1). Branches of the basal pulmonary vein that had been preoperatively identified on a 3-dimensional image and branches of the segmental pulmonary vein were transected if necessary (Figure 2, B and E). With right-angle forceps, dissection proceeded along the intersegmental septum to expose the pulmonary arteries running alongside the bronchi, which were then transected (Figure 2, C and F). When the bronchi...