

Invited Lecture

Invited Lecture 1

座長:須田 憲治 (久留米大学医学部小児科学講座)

Thu. Jul 11, 2024 10:40 AM - 11:25 AM ROOM 1 (3F Main Hall)

[I-IL1-1]Fetal cardiac interventions for congenital heart defects

○Wayne Tworetzky (Boston Children's Hospital, Boston, MA / Harvard University, Cambridge, MA)

Fetal cardiac interventions performed for the following defects.

- 1.severe aortic valve stenosis with features of evolving hypoplastic left heart syndrome.
2. Established hypoplastic left heart syndrome with intact or highly restrictive atrial septum.
- 3.pulmonary atresia with intact ventricular septum and hypoplastic right ventricle

Aortic stenosis with evolving hypoplastic left heart syndrome

This is the most frequent indication for fetal cardiac intervention. When evaluating a fetus for this procedure, the fetus has to have anatomic and physiologic features that would predict evolution to hypoplastic left heart syndrome at birth. These features are left ventricular dysfunction, aortic stenosis, left-to-right flow at the PFO and retrograde flow in the aortic arch. The fetus also has to have features that would predict that the left ventricle is salvageable. The most important features are an apex forming left ventricle, not more than mild or moderate eFE, high left ventricular pressure as estimated by mitral regurgitation jet velocity and a normal volume left ventricle. The procedure is performed by a team that consists of ultrasound imaging, interventional catheterization, anesthesia and maternal-fetal medicine. The mother receives an epidural for analgesia. Fetal positioning is very important for technical success. The fetus receives intramuscular analgesia and paralysis. The procedure is performed by ultrasound guidance with a 19-gauge cannula inserted into the left ventricle. The aortic valve is dilated with a coronary artery balloon dilation catheter that is oversized to ensure opening of the aortic valve. Bradycardia and hemopericardium are common during the procedure and easily treatable. In the neonatal period the child has to be managed by the usual criteria and deciding on a univentricular or biventricular circulation. Novel creative surgeries have been developed to manage this complex group of patients. In some cases a borderline or hypoplastic left heart can be converted to a biventricular circulation with staged surgeries.

Hypoplastic left heart syndrome with intact or highly restrictive atrial septum

This is one of the most complicated heart defects to treat, with very high mortality and significant morbidity. Balloon dilation of the atrial septum in utero as well as placement of stents has been performed. The overall outcome is still unsatisfactory. Both procedural success and overall survival has been quite modest. In theory, creating a large atrial septal defect in early gestation might be the way to improve outcome in this complex group of patients.

Pulmonary atresia with intact ventricular septum, hypoplastic right heart syndrome

This is a rare congenital heart defect. Most fetuses are not candidates for fetal cardiac intervention. Those with fibromuscular atresia have no valve and therefore cannot undergo procedure. Patients with a large tricuspid valve and ventricle did not need a fetal intervention and can be managed successfully with postnatal therapy. Patients with hypoplastic right ventricles with an identifiable pulmonary valve can undergo in utero perforation and balloon dilation of the valve. However this is a very difficult procedure from a technical standpoint because the right ventricle has complex geometry and is small. In summary, fetal cardiac intervention for aortic stenosis is the most frequently performed procedure.

Success rates have improved and longer-term results are encouraging