Intrauterine Intervention of Pulmonary Atresia at 26th Gestational Week

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To the Editor: Congenital heart diseases (CHDs) are the most common congenital structural anomalies in China. Most of them are curable after birth. However, some defects progress during fetal period, thus resulting in high neonatal mortality and morbidity. Pulmonary atresia with intact ventricular septum (PA/IVS) during fetal period is partly associated with right ventricle (RV) hypoplasia and its consequent hemodynamic deterioration, which limits the possibility of biventricular (BV) repair after birth. The 1-year survival of PA/IVS live births is around 70%, but the rate of BV outcome is around 30%.[1]

Such cases may benefit from fetal cardiac intervention (FCI), which achieves a promising BV circulation by relieving outflow tract obstruction. Early ventricular decompression may stop pathological progression, alter the natural history, and improve postnatal outcomes either by preserving a BV circulation or by improving the outlook for single-ventricle candidates. In 2002, Tulzer *et al*.[2] reported their first successful attempts to perform pulmonary valvuloplasty in second trimester fetuses via a needle perforated through the RV to induce the development of the right heart. Up to now, 35 FCI cases were subsequently reported using similar procedures. In this case, we described a family in which the fetus was reported RV hypoplasia due to PA/IVS. We performed the FCI at 26 weeks of gestation and gained technically success. As far as we know, this is the smallest gestational age of prenatal FCI of PA/IVS in Asia.

A 33-year-old woman was referred to our center at 25 weeks of gestation because of severe fetal pulmonary stenosis. Both parents had no family history of CHDs and genetic disorders. Their first child, a 5-year-old girl, was born after an uneventful pregnancy and did not suffer from any neonatal complications. The present pregnancy was the second pregnancy of this couple, following to a postterm cesarean delivery. The parents refrained from amniocentesis as diagnostic test (due to the risk of miscarriage).

Repeated echocardiography showed PA/IVS with hypoplastic right heart. The multidisciplinary team including the maternal-fetal medicine specialist, pediatric cardiologist, ultrasonologist, and anesthetist provided detailed counseling and a FCI was planned. The procedure was approved by the hospital ethics committee. The pregnant woman and her families were informed about the maternal and fetal benefits and risks of the intervention. Written informed consent was obtained.

The FCI was performed at 26 weeks of gestation. After maternal general anesthesia with inhaled and intravenous agents, the external obstetrical version was performed in order to adjust the fetal position to clearly show the RV outflow tract (RVOT). An 18G trocar needle was introduced toward the RVOT by direct puncture. Then after the guide wire (0.014", Conquest Pro, Asahi) was accessed into the pulmonary artery through the pulmonary valve, the coronary angioplasty balloon (3.5 mm × 12 mm, Boston Scientific) was advanced across the pulmonary valve, inflated to dilate the valve. The intervention took around 30 min. A small amount of fetal hemopericardium was observed immediately after the intervention and spontaneously absorbed within 24 h.

After the intervention, the transplacental therapy of maternal oral digoxin (0.25 mg/d) was administrated for 2 weeks in order to improve RV function. The postprocedure complications including uterine contraction, fetal arrhythmia and hemopericardium were monitored. Moreover, the fetal growth of right heart structure was followed up weekly by echocardiography. The RV function and hemodynamic parameters were significantly improved after 2 weeks, and the ratio of the tricuspid valve inflow duration and cardiac cycle significantly improved [Table 1].

PA/IVS is a rare cyanotic CHD, accounting for 5% of fetal CHD. It will eventually lead to fetal hypoplastic right heart to some extent, resulting in failure to achieve BV circulation after birth.

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Table 1: Right ventricular function and hemodynamic parameters before and after FCI

Timing	GA (weeks)	TV/MV	RV/LV	PV/AV	TVI/CC	TR (m/s)	PE (mm)
1 day before FCI	26 + 2	0.80	0.80	0.70	0.26	3.3	3.5
1 week after FCI	27 + 2	1.01	0.84	0.98	0.34	2.0	1.5
2 weeks after FCI	28 + 2	1.08	0.86	0.99	0.37	3.8	1.0

FCI: Fetal cardiac intervention; GA: Gestational age; TV/MV: Ratio of the tricuspid valve annulus and mitral valve annulus; RV/LV: Longitudinal ratio of the right ventricle and left ventricle; PV/AV: Ratio of pulmonary valve annulus and aortic valve annulus; TVI/CC: Ratio of the tricuspid valve inflow duration and cardiac cycle; TR: Tricuspid regurgitation; PE: Pericardial effusion.

PA/IVS may result in fetal hydrops and even intrauterine death. Fetal pulmonary valvuloplasty can restore blood flow from RV into pulmonary artery, which can theoretically promote RV development to achieve BV circulation and better long-term prognosis after birth.

As the limited literature of FCI for PA/IVS, no accurate predictors of successful BV circulation after birth were documented. Gómez Montes *et al*.[3] proposed a scoring system according to three size-based parameters and one functional marker in the second trimester to predict the single ventricular outcome after birth: (1) tricuspid valve/mitral valve <0.83, (2) RV/left ventricular length <0.64, (3) pulmonary valve/aortic valve <0.75, and (4) tricuspid inflow duration/cardiac cycle length <0.36. This predicts a non-BV outcome with sensitivity of 100% and specificity of 92% when three criteria are fulfilled. The preoperative echocardiographic evaluation of this case shows three markers which are in accordance with this scoring system.

The timing of FCI is controversial. The earlier intervention might provide more possibility for ventricular growth. Moon-Grady *et al*. illustrated that the average gestational age of FCI was 26 + 4 weeks (20 + 3 ~ 33 + 1 weeks).[4] Tworetzky *et al*.[5] performed 10 cases of *in utero* valvuloplasty for PA with hypoplastic RV, pointing out the larger gestational age was associated with higher rate of technical success. This case is the FCI of PA/IVS with smallest gestational age in Asia. Velocity of tricuspid regurgitation decreased 1 week after FCI and increased significantly 2 weeks later because of a better RV function from the mother's oral usage of digoxin. Moreover, the blood flow passing through the pulmonary valve was detected continuously. From our practice, the most significant determinant of a successful intervention is the favorable fetal position.

Most studies reported that FCI was performed under both maternal and fetal anesthesia to enable the relaxation of uterus and the manipulation of fetal position. In the present case, the transverse lie of fetal head and FCI were successfully performed only via maternal anesthesia.

In conclusion, PA/IVS can be severe and life threatening during fetal life. The strategy of FCI in the second trimester, as described above, is technically feasible and may be associated with improved right heart growth, thus improving the chances for a BV outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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