Intrauterine Cardiac Intervention for Fetal Pulmonary Valve Obstruction Lesion

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Most of the congenital heart diseases can be diagnosed *in utero* using fetal echocardiography. With the development of ultrasonography and intrauterine cardiac intervention technique, fetal cardiac intervention (FCI) for critical aortic stenosis, highly restrictive or intact atrial septum, pulmonary atresia with an intact ventricular septum (PA/IVS), and critical pulmonary stenosis (CPS) are available in more countries.

In September 2016 and March 2017, experts from Guangdong Cardiovascular Institute performed the first and second cases of FCI in China. Both patients were referred to our center because an abnormal fetal right heart structure was observed during routine obstetric ultrasonographic screening at mid-gestation age. In the first case, the mother was 39 years, G2P1A0. This time was a nature conception. She delivered a baby in 2013 using *in vitro* fertilization and received partial thyroidectomy in 2002 due to a thyroid cyst, but she maintained euthyroidism during pregnancy. In the second case, the mother was 37 years, G3P1A1. She had a baby in 2010 but underwent induced abortion in 2013 due to fetal cervical lymphatic hygroma. Other obstetric examinations were unremarkable in both patients.

Detailed fetal echocardiography was performed. The first fetus was diagnosed as PA/IVS, and the second fetus was CPS. Hypoplasia of both right ventricles (RVs) was found but without signs of the RV-dependent coronary circulation. FCIs were performed at 28 weeks' gestation under complete ultrasound visualization. Maternal-fetal cardiologists, pediatric cardiologists, and anesthetists were involved, with cardiac surgeons, obstetrics, and neonatologists on standby. The fetal position was identified first to clearly view the right ventricular outflow tract (RVOT), and then, maternal general anesthesia was given. An 18-gauge trocar needle was used to form procedural trajectory by direct puncture through mother's abdomen, uterine wall, placenta, fetal chest wall, fetal RVOT, and finally, to the pulmonary valve. Then, a 4.0 mm × 9.0 mm coronary angioplasty balloon was guided across the pulmonary valve for dilation. The intervention took about 30 min.

Monitoring of the growth and development of the right heart structure, the presence of fetal arrhythmia, fetal hydrops, uterine contraction, and other complications will be performed closely. Follow-up visits on the fetal cardiac status and obstetric examination should be performed every 2–4 weeks until delivery after discharge.

Born at term, the baby in the first case was diagnosed as CPS and RV hypoplasia. The RV was still underdeveloped, especially the trabecular portion. Modified Blalock-Taussig shunting, pulmonary valve commissurotomy, ligation of the patent ductus arteriosus, and enlargement of the atrial septal defect were performed at day 8. A recent examination showed pulse oximetry results were 82–85% saturation, and the orifice of the pulmonary valve was 3.0 mm. In the second case, the baby was premature at 35 weeks' gestation, and the echocardiogram demonstrated severe pulmonary stenosis and severe tricuspid regurgitation. Percutaneous pulmonary balloon

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Table 1: Before and after FCI and postnatal characteristics of the two patients undergoing intrauterine cardiac intervention for fetal pulmonary valve obstruction lesion

Case number	Diagnosis	GA at diagnosis (weeks)	BPD (cm)	FL (cm)	HR (beats per minute)	Before FCI			After FCI		
						TV/MV	RV/LV	TVI/CC	TV/MV	RV/LV	TVI/CC
1	PA/IVS	24^{+6}	7.13	5.11	143	0.56	0.42	0.26	0.59	0.51	0.32
2	CPS/IVS	26^{+1}	7.33	5.38	139	0.70	0.63	0.35	0.80	0.80	0.42

Case number	Postpartum							Latest examination		
	Wt (g)	L (cm)	PO (%)	RV (mm)	LV (mm)	PV (mm)	*Z*-score	PO (%)	PV (mm)	PG (mmHg)
1	2970	50	80–90	14.1×9.5	27.5×21.8	2.8	1.12	82–85	3.0	50
2	3390	45	88–92	22.7×11.0	25.1×22.7	3.5	1.00	92–96	6.3	36

1 mmHg = 0.133 kPa. FCI: Fetal cardiac intervention; GA: Gestational age; BPD: Biparietal diameter; FL: Femur length; HR: Fetal heart rate; TV/MV: Ratio of the tricuspid valve annulus and mitral valve annulus; RV/LV: Longitudinal ratio of the right ventricle and left ventricle; TVI/CC: Ratio of the tricuspid valve inflow duration and cardiac cycle; Wt: Weight of the neonate; L: Length of the neonate; PO: Pulse oximetry; RV: Right ventricle; LV: Left ventricle; PV: Orifice of the pulmonary valve; *Z*-score: *Z*-score for the pulmonary valve annulus; PG: Pressure gradient across pulmonary valves; PA/IVS: Pulmonary atresia with an intact ventricular septum; CPS: Critical pulmonary stenosis.

valvuloplasty was performed on day 18. A 4.0 mm × 12.0 mm coronary balloon and an 8.0 mm × 10.0 mm balloon were used for sequential dilation. At the latest visit, pulse oximetry results were 92–96% saturation and the opening of the pulmonary valve was 6.3 mm with a pressure gradient of 36 mmHg (1 mmHg = 0.133 kPa). Both babies require close follow-up after initial treatment, and whether further treatment is needed depends on the follow-up results. Profiles of these two babies are demonstrated in Table 1.

PA/IVS or CPS is a vital situation for a fetus, and univentricular repair may be necessary for survival due to an unfavorable right ventricular anatomy. RV hypertrophies with an elevating pressure instead of dilation, which makes it hard to adapt to the nonphysiologically hemodynamic changes; hence, the risks of irreversible myocardial changes significantly increases. FCI offers a feasible treatment choice.

Operative steps and equipment are essentially the same, however, institutional criteria for pulmonary FCI vary. Boston and New York centers[1,2] emphasized the Z-score of right heart structures, but Toronto and Madrid centers focused on ratios between the right heart structures and counterparts in the left heart.[3,4] Due to the lack of effective evaluation of Z-score in our country, we adopted the Madrid scoring system to select eligible candidates. Compared with the four cases reported by Gómez Montes *et al*.,[4] the different outcomes may result from the lighter degree of compromise of right heart structure of our cases. There was no decrease in the RV/LV ratio in our fetus with PA/IVS at late gestation, and for the fetus with CPS, no restenosis occurred, and she was stable to undergo balloon pulmonary valvuloplasty. No prenatal of postnatal death occurred at our center.

Based on 33 cases of pulmonary FCI published worldwide, the technical success rate is about 70%, and the BV rate is approximately 40%; however, with the small series of FCI patients and limited experience, there is no accurate prediction for a successful BV surgery after birth. Numerous efforts should be made to improve indications and selective criteria, as well as long-term outcomes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials 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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