Immediate and short-time outcomes of pulmonary valvuloplasty in a fetus with pulmonary atresia

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To the Editor: The incidence of pulmonary atresia with intact ventricular septum (PA/IVS) is 1.9% in congential heart disease. The most serious manifestation is hypoplastic right heart syndrome (HRHS). HRHS and low birth weight have been determined as the major risk factors of fetal death after birth. Fetal pulmonary valvuloplasty (FPV) could relieve obstruction of the right ventricular outflow tract, restore growth potential of right ventricle, and increase the chances of biventricular circulation. However, so far fewer than 70 HRHS cases have been reported at the International Fetal Cardiac Intervention Registry (http://www.ifcir.org/) and there is a lack of post-natal treatment experience.

In 2018, our center reported the minimum gestational week of fetal cardiac intervention (FCI) in China.[1] FPV was performed on PA/IVS with HRHS of fetus at the 26th week of gestation. The procedure was smooth without serious complications and the pregnant woman was discharged from hospital smoothly [Figure 1A–F]. Conventional echocardiograph after FPV showed right heart size with a qualitative improvement in comparison with pre-FPV and tricuspid regurgitation velocity consistent with 2.8 to 3.3 m/s. The pregnant woman gave birth to a female newborn weighing 3050 g by cesarean section at the 38th week of gestation. The baby was transferred to cardiac intensive care unit (CICU) with a pulse oxygen saturation of 90%. Prostaglandin E_1 (PGE_1) was infused immediately to keep the ductus arteriosus open. Neonatal echocardiographic examination showed critical pulmonary stenosis, moderate HRHS. The inner diameter of ductus arteriosus was about 3 mm, with no RV-dependent coronary circulation.

The percutaneous balloon pulmonary valvuloplasty (PBPV) was performed at the age of 24 h under general anesthesia. Intraoperative balloon dilatation of 1.5 to 4.5 mm (Boston Scientific, USA) was used respectively to dilate the valves gradually [Supplementary Figure 1A, http://links.lww.com/CM9/A57]. PGE_1 was continuously infused to keep the ductus arteriosus open, the pulse oxygen saturation was 95% to 100%. After 1 week, pulse oxygen saturation (SpO_2) dropped gradually to 85%, which may probably indicating restenosis of pulmonary valves. A second PBPV was performed by adopting an 8 mm × 20 mm balloon (Numed, Germany) to dilate the pulmonary valve ultimately. Ductus stent (3.5 mm × 18.0 mm; Lepu Medical Technology, China) was implanted [Supplementary Figure 1B, http://links.lww.com/CM9/A57], and the SpO_2 rose to 95%. After 4 months of follow-up, the SpO_2 was maintained above 99%, the RV was able to support the pulmonary circulation. Biventricular circulation was obtained in this child, suggesting a good long-term prognosis.

The success rate of FPV was 70%, biventricular repair rate after birth was 40%. The incidence of bradycardia was 31%, which were the major causes for intra-operative death of the fetuses. Case of FPV selection still remains controversial. Gómez Montes *et al*[2] put forward to evaluate single ventricle results by using a scoring system that comprehensively combines three morphologic indices and one functional index of echocardiography of fetuses during mid-term pregnancy. Three indicators were consistent with the scoring system, which predicted a single-ventricular result with a sensitivity of 100% and a post-natal specificity of 98%. Before operation, we strictly consulted the scoring system to indicate that the indications of operation were clear. Post-operative related indicators were significantly improved, and the operation achieved satisfactory results. Consistent with the study of foreign scholars, we found that fetal tricuspid regurgitation speed increased again, suggesting that post-operative pulmonary valve adhesion occurred gradually.[3]

Considering the advantages of less trauma, higher safety and faster recovery in PBPV, it has become the preferred method for patients with central pain syndrome. Because

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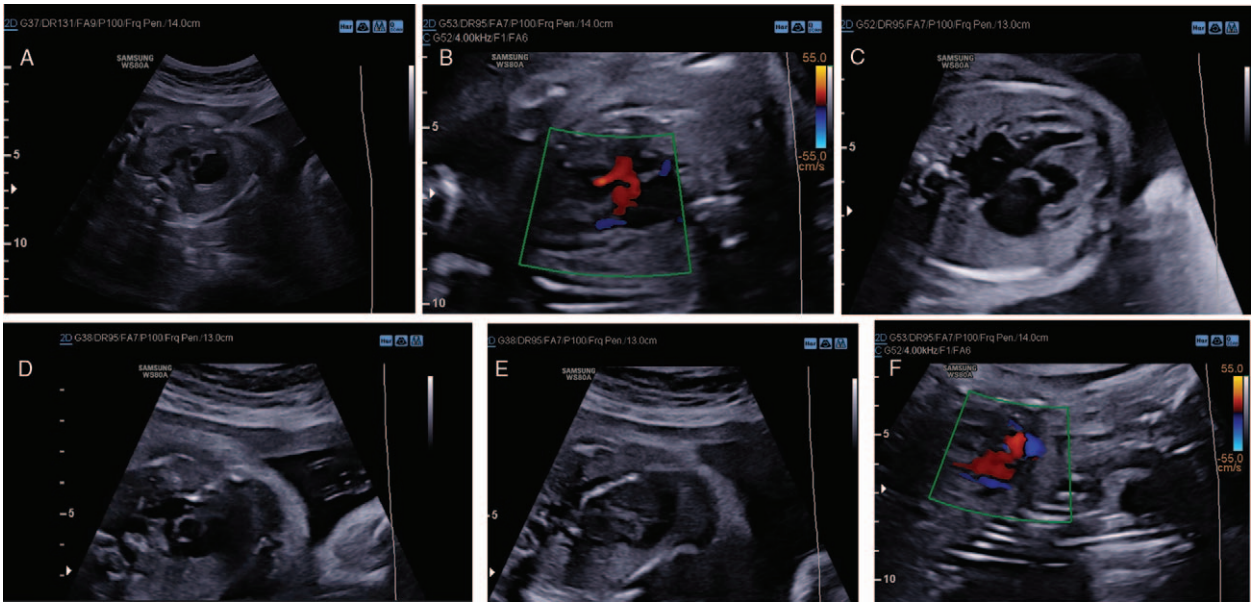

Figure 1: Fetal pulmonary valvuloplasty in the fetus at the 26th week of gestation. (A) Echocardiography showed pulmonary atresia. (B) Doppler ultrasound showed no forward flow of pulmonary valve. (C) Echocardiography showed small right ventricular cavity, myocardial hypertrophy, and right ventricular dysplasia. (D) Echocardiography showed the needle tip was pushed through the pulmonary valve in the valve middle for perforation. (E) The coronary balloon (3.5 mm × 12.0 mm; Boston Scientific, USA) was sent in along the guidewire to dilate the pulmonary value. (F) Doppler ultrasound showed pulmonary valve forward flow recovery after operation.

of right ventricular hypoplasia and poor adaptability, normal volume load of right ventricle could not be performed immediately after PBPV. If the situation continues to deteriorate, it is necessary to implement a systemic to pulmonary artery shunt or a ductal stent during the neonatal period.

The success rate of arterial duct stenting was about 90%, the fatality rate was lower than 10%, and there was no operative mortality. The ductal stent has become an alternative to the modified Blalock Taussig shunt surgery. Based on the experience of this case, we believe that PBPV and arterial duct stenting should be performed simultaneously in fetus with moderate to severe right ventricular dysplasia after birth. This method could reduce repeated procedures and could shorten hospital stay, without increasing fatality and disability rate.

This study had currently the lowest gestational age in Asia receiving FCI treatment as far as we know. PBPV and arterial duct stenting were performed after birth, that not only avoid surgical thoracotomy, but also help to achieve biventricular circulation.

Declaration of patient consent

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

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Conflicts of interest

None.

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