

IMAGES in PAEDIATRIC CARDIOLOGY

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Abstract

The mortality and morbidity of children with pulmonary atresia with intact ventricular septum (PA/IVS) is closely related with right ventricle hypoplasia and its consequent hemodynamics. Prenatal intervention for fetuses with PA/IVS has the potential to improve growth of the RV and the prospect of a biventricular outcome after birth. Successful valvulotomy of the pulmonary valve (PV) was performed in a fetus with PA/IVS at 28 weeks. Following the procedure there was an improvement in fetal hemodynamics. In utero perforation and dilation of the PV in midgestation fetuses with PA/IVS is technically feasible. Our initial results are promising and may be associated with improved right heart growth and postnatal outcome.

Keywords Pulmonary atresia with intact ventricular septum, Fetal cardiac intervention, Pulmonary valvulotomy

Introduction

Pulmonary atresia with intact ventricular septum (PA/IVS) is a disease with remarkable morphologic variability, particularly affecting the tricuspid valve (TV), right ventricle (RV) and sometimes coronary circulation. Mortality and morbidity are linked to the severity of RV and TV hypoplasia, and major abnormalities of the coronary arteries. These features are the most important determinants of univentricular versus biventricular outcomes.¹⁻³

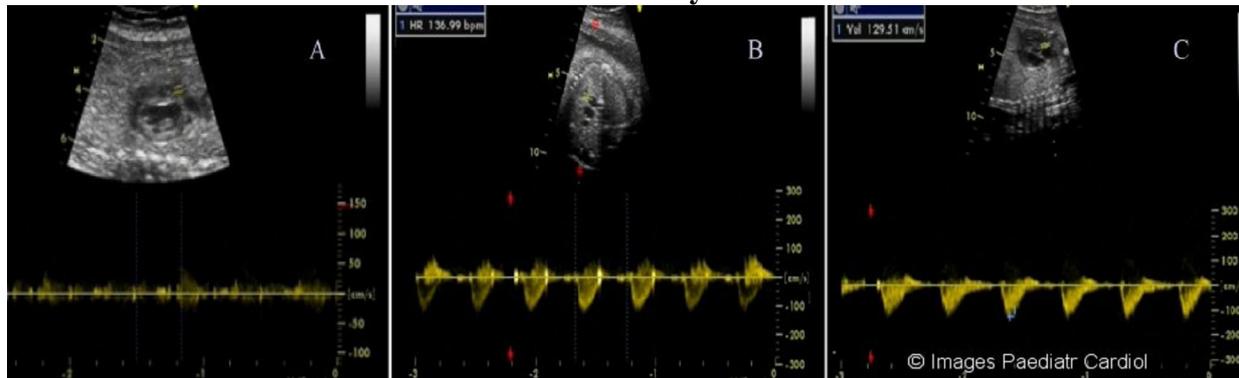
In utero valvuloplasty for PA/IVS has been performed previously in selected cases to achieve more-favorable end of the spectrum. It has been speculated that early in utero perforation and dilation of the pulmonary valve (PV) in fetuses with PA/IVS may allow antegrade blood flow through the RV, potentially stimulating right heart growth and function and improving the chances of postnatal survival with a biventricular circulation.^{4,5,6,7}

Here, we report the first case of successful prenatal pulmonary valvulotomy in a fetus with PA/IVS in Turkey. We describe our experience, with a focus on the technical details and immediate hemodynamic changes after procedure.

Case Report

A 29-year-old woman was referred at 24 weeks for fetal cardiac evaluation. Our echocardiogram displayed severe RV hypoplasia, abnormal RV filling (missing E-wave), no antegrade flow through the PV (Figure 1A) with the pulmonary flow dependent on the ductus arteriosus, holosystolic tricuspid regurgitation (TR) of 4.2 m/s leading to the diagnosis of PA/IVS. No coronary fistulae could be found and interatrial shunting was not restrictive.

Figure 1. (A) The longitudinal view of the right ventricular outflow showing no antegrade flow across the pulmonary valve before the procedure. (B) The three-vessel and (C) longitudinal view of the right ventricular outflow demonstrating antegrade flow after valvulotomy.



During weekly follow-up scans the severity of tricuspid regurgitation increased and the degree of RV hypoplasia (tricuspid valve annulus 5.2 mm, Z score -2.8) became more pronounced with the presence of reversed end-diastolic flow in the ductus venosus. After several and detailed explanations of the benefits and risks of the procedure parents consented to fetal cardiac intervention (FCI).

The procedure was performed at 28 weeks of pregnancy under ultrasound guidance. Preprocedural scan showed high velocity holosystolic TR of about 5.2 m/s with a PV annulus diameter of about 3.8 mm. The mother refused general anesthesia for the procedure. Briefly, the fetus received an intramuscular injection of rocuronium (0.6 mg/kg) to avoid fetal movements. An 18-G needle (15 cm in length) with a diamond shape stylet was introduced percutaneously through to the fetal chest wall into the RV pointing directly to the outflow tract. Care was taken to direct the needle tip to the PV, which was then perforated. A 2.7-F coronary balloon catheter (4 mm/1.2 cm, Alvi Medica-Turquoise™) was mounted on a 0.014-inch floppy tipped coronary guide wire, with 3 cm of distal wire exposed before the procedure to minimize intracardiac manipulation time.⁸ The wire/catheter assembly was then advanced through the needle until the balloon emerged. The balloon was firstly positioned in the left pulmonary artery, therefore, pulled back across the perforated valve and three consecutive inflations were performed.

A brief and self-limited episode of bradycardia was observed immediately after puncture of the RV; the fetal heart rate remained stable for the rest of the procedure. The balloon was retrieved leaving the wire in position and a careful scan showed antegrade pulmonary flow. The needle was then withdrawn into the amniotic space, still leaving the wire in position, and only a 2 mm pericardial effusion located anterior to the RV was observed afterwards. Finally, the wire and needle were withdrawn respectively. During this last attempt, the proximal spiral part of the floppy guidewire was stripped and broke off when passing through the sharp edge of the needle, and wedged into the fetal chest wall. The free portion of the wire extended into the lumen of the main pulmonary artery (Figure 2). There was no further evidence of bleeding, arrhythmic events and no maternal complication.

Figure 2. The floppy guidewire fragment (white arrow) extended into the lumen of the main pulmonary artery.



Echocardiography performed a few hours later showed the patency of the anterograde flow through the pulmonary valve with a velocity of 1.29 m/s (Figure 1B, 1C), and slightly decrease in the severity of TR with a maximum velocity of 3.4 m/s. Slight improvement in right ventricular compliance was documented as barely visible E-wave Doppler pattern after procedure (Figure 3). However, there was still reversed flow in the ductus arteriosus and reversed end-diastolic flow in the ductus venosus (Figure 4). The pericardial effusion disappeared next day and mother was discharged.

Figure 3. Pulsed Doppler recordings of right ventricular inflow velocities registering visible E-wave Doppler pattern after procedure.

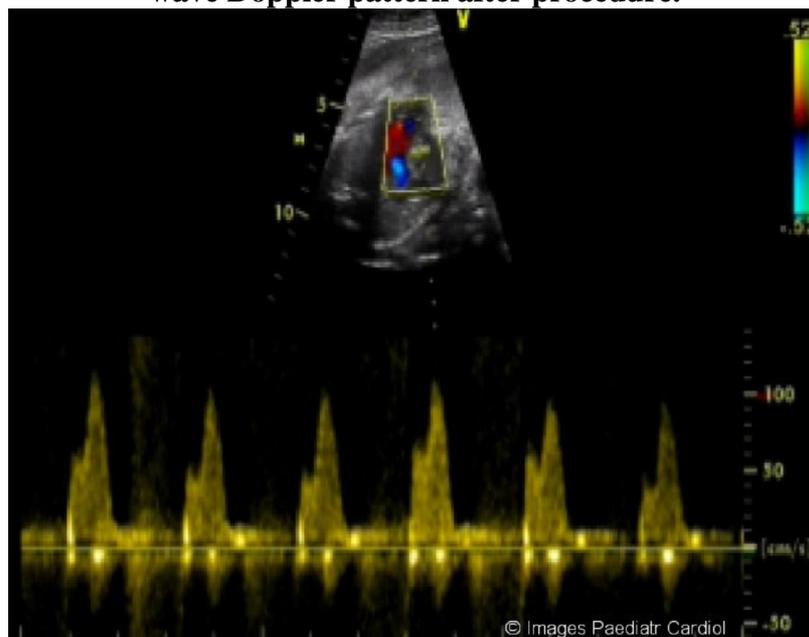
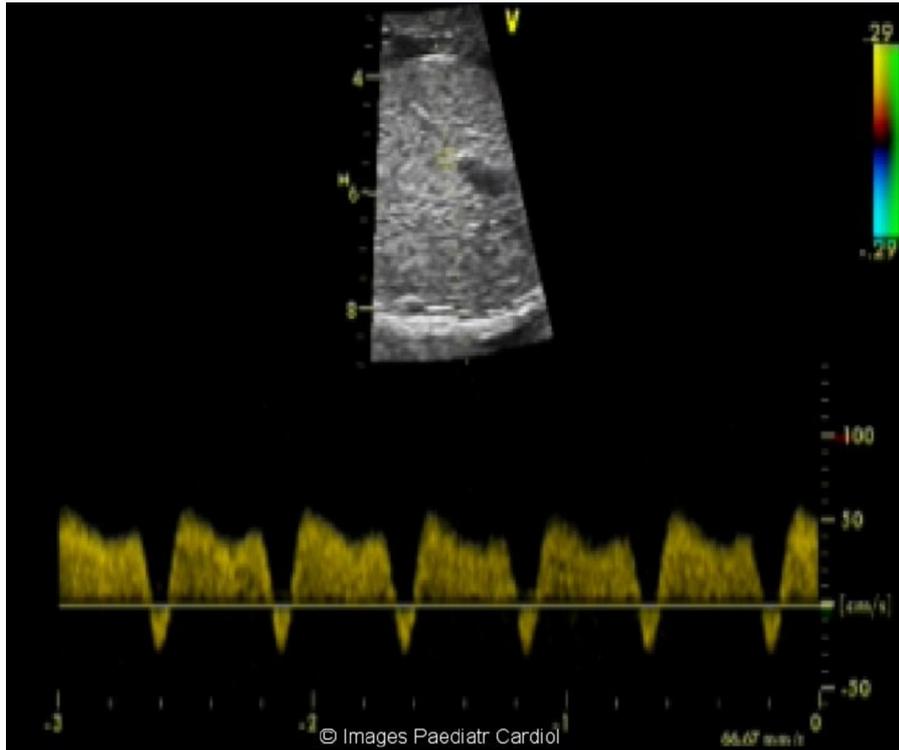


Figure 4. Transverse view of the fetal abdomen showing persistent end-diastolic reversed flow in the ductus venosus.



A video shows the balloon firstly positioned in the left pulmonary artery, pulled back across the perforated valve and three consecutive inflations were performed (figure 5).

Figure 5. Video showing balloon positioning in the left pulmonary artery and inflations.



Discussion

The potential role of FCI in fetuses with PA/IVS is to promote right heart growth and functional development and increase the chance of a biventricular circulation after birth. Several small patient series and case reports of intrauterine valvuloplasties have been recently communicated and published.^{4,5,6,7} On the basis of this limited experience, it appears that prenatal pulmonary valve perforation and dilation may be performed successfully in midgestation fetuses, with maintenance of valvar patency throughout gestation and apparently improved growth of right heart structures.

There is limited information about prenatal predictors of postnatal outcomes in fetuses with PA/IVS. Both anatomic and hemodynamic characteristics are likely to be important. TV Z score less than -3 at birth is an important predictor of failure to achieve a biventricular circulation. Along with the RV volume, TR is an important and common feature, which should also be considered.^{2,9}

In our case, there was a progressive impairment of RV volume with a tricuspid Z score almost -3 , and of hemodynamic profiles with altered TR and existing abnormal Doppler patterns during follow-up. Thus, we decided to perform an invasive procedure in order to change the anticipated unfavorable history and to maintain tripartite RV morphology. With the opening of the PV and by establishing antegrade flow through the stiff and hypoplastic ventricle into the pulmonary artery, we provided an alternative pathway for the blood leaving the right atrium. Although a decrease of tricuspid regurgitation was achieved immediately after procedure, reversed flow in the ductus arteriosus and venosus did not change. Slight improvement in right ventricular compliance was documented as barely visible E-wave Doppler pattern after procedure. Furthermore, antegrade flow reached a velocity of 1.2 m/s, while the amount of TR and maximum TR velocity slightly decreased. We concluded that the combination of significant tricuspid regurgitation and inadequate compliance of RV can virtually decrease forward flow from the right ventricle. Therefore, persistence of flow reversal in the DV and DA immediate after the procedure may be a reflection of inadequate forward flow from the noncompliant RV with significant TR.

Small pericardial fluid collections were common during FCI with both atrial and ventricular access. In a large series, significant hemopericardium occurred during FCI in nine of 83 fetuses, all of which underwent FCI with attempted transventricular access, and seven of which developed major fetal hemodynamic instability.¹⁰ Given the potential hemodynamic importance of hemopericardium during FCI, we followed a step by step approach to withdraw the assembly after procedure. In this approach, by leaving the wire in position, we aimed to keep the previous access point for pericardial drainage, particularly after withdrawing the needle. However, the guidewire was severed during its retrieval. We consider that this complication was closely related to the sharp edge of the needle, and has never been reported in the literature. Nevertheless, the missing wire fragment did not cause any harmful events such as arrhythmia or obstruction during procedure and follow-up. In order to avoid this complication, we concluded that wire should be the first to withdrawn into the balloon catheter or wire along with the needle should be withdrawn simultaneously.

In all previous reports, general anesthesia was administered to avoid maternal movements and to provide fetal anesthesia. Mostly, fetuses also received neuromuscular blocking agents, intramuscularly.^{4,5,6,7} Under general anesthesia, if an ideal fetal position could not be achieved or imaging was inadequate, a maternal laparotomy was undertaken.^{7,11} However, the mother refused general anesthesia in our case and gave informed consent for a minimally invasive approach. Therefore, we decided to paralyze the fetus solely with intramuscular injections of a neuromuscular blocking agent. We preferred rocuronium, a steroidal, non-depolarizing neuromuscular blocking drug chemically related to vecuronium. Rocuronium has been studied in pregnant women, but has never been used during fetal interventions. Compared with vecuronium, rocuronium is six times less potent but has a much faster onset of action with comparable intermediate duration of action.^{12,13} We considered that the shorter onset time might help to preserve an ideal fetal position for scanning scanned prior to intervention, and if the first attempt failed due to poor fetal position, the shorter duration of action might allow rapid fetal self-repositioning for optimal further attempts.

Although our initial results are promising, it is difficult to prove some affirmations. A longer follow-up is required to answer all the questions particularly that this procedure might be able to

alter the prenatal natural history, and enable sufficient RV. However, published data supports the concept that further RV growth in this fetus would not have occurred without this procedure.

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