Center Experience and Step-by-Step Approach for Fetal Pulmonary Valvuloplasty

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12.1 Center Experience

In our center (the Children's Heart Center Linz), the fetal cardiac intervention program was started in the year 2000. In fact, the very first procedure, which we performed, was in a fetus with pulmonary atresia with intact ventricular septum (PAIVS) at 28 weeks [1]. Since then a total of 91 interventions have been here conducted, the majority in fetuses with critical aortic stenosis, few in fetuses with hypoplastic left heart syndrome and restrictive foramen ovale, and altogether 20 procedures in 14 fetuses with PAIVS or critical pulmonary stenosis with intact ventricular septum. We reported our first case together with a case that was done in the UK in a publication in the year 2002. This fetus was thought to develop restriction at the atrial level with increased central venous pressures and imminent hydrops as indicated by severely abnormal venous Doppler waveforms – a complication that may result in fetal hydrops and intrauterine death. The fetus had a hypoplastic RV with growth arrest and suprasystemic right ventricular pressures, a significant tricuspid regurgitation (TR), and a favorable anatomy with a membranous atresia of the pulmonary valve and well-developed confluent pulmonary arteries. For this first procedure, we used a 16-gauge needle and managed to perforate the membrane between the RV and main pulmonary artery. Clear antegrade as well as retrograde flow across the pulmonary valve annulus was observed immediately after the procedure by color-flow and CW Doppler. This fetus was followed longitudinally in our center, and over the following weeks, significant changes in hemodynamics were documented: growth of RV long axis, growth of

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G. Butera et al. (eds.), *Fetal and Hybrid Procedures in Congenital Heart Diseases*, DOI 10.1007/978-3-319-40088-4_12

tricuspid valve annulus, change from a short monophasic to a longer biphasic RV inflow, as well as complete disappearance of the TR jet. Six weeks after the procedure, TR reappeared again with high velocities due to an increasing restenosis of the perforated pulmonary valve. This baby was delivered at 38 + 2 weeks; postnatally it received a pulmonary valvuloplasty, and because of a still borderline RV output, a 3.5 mm modified right BT shunt was placed. This shunt was successfully removed at the age of 8 months. Now 15 years later, this girl still has had no other interventions; she has mild to moderate pulmonary stenosis and mild pulmonary regurgitation with some dilation of the main pulmonary artery. She is on no medication, is very sportive, and has no exercise intolerance at all.

This encouraging preliminary experience motivated us to continue and to further develop in utero treatment of PAIVS. Our experience with the first series of five and six cases, respectively, was published in 2006 and 2011 [2, 3]. At that time we had a technical success rate – which means that we were able to perforate and to dilate the pulmonary valve – in four out of six cases. Of the successful cases, three became biventricular and one ended up with a one-and-a-half ventricle circulation. In 2015 we presented our experience of 12 procedures in ten fetuses. We were able to perforate and to dilate the valve in 80 % of these fetuses. Again we saw RV and TV growth and longer RV inflow durations but with remaining high PV gradients in most of them. Six of them became biventricular, one with a one-and-a-half ventricle and one with a univentricular circulation. Currently our experience consists of 20 procedures in 14 fetuses; three of them are still in utero.

12.2 Step-by-Step Approach

12.2.1 Assessing Fetal Position

The single most important factor for a successful procedure is the fetal position. Once the fetus is in a dorso-posterior position with the RV facing toward the maternal abdomen and the fetal spine is positioned between 5 and 7 o'clock, the procedure has a very high chance of being successful. There are "borderline" positions, where an intervention theoretically is still possible but certainly more difficult and a challenge for the whole team: amount of amniotic fluid, position of the placenta, distance between maternal skin and fetal heart, maternal obesity, and obvious obstacles in the way of the intended needle insertion (umbilical cord, fetal extremities, etc.). Additionally there are fetal positions, where a successful intervention is impossible (e.g., spine anterior), and the procedure has to be postponed until a better position is present. This initial evaluation is usually done by both perinatologist and cardiologist together and certainly needs a lot of experience.

12.2.2 Setting Up the OR

All procedures were performed in the operating theater with enough space for the anesthesia team (we performed all procedures under general anesthesia); on one side of the table, there is the ultrasound machine positioned and on the other side the

catheter table with the necessary equipment. Our interventional team consists of an anesthetist with one or two nurses, one perinatologist for the needling, two fetal cardiologists for scanning and handling of the catheter, and one physician or nurse handling the balloon inflation.

12.2.3 Preparing the Procedure

Mother: After having given informed consent on the day before the procedure, an ultrasonic assessment of the fetal position is done. If the fetus is found to be in a favorable position, the mother is brought into the theater and scanned again, and if the fetus has not changed position, anesthesia is started instantly. Only after effective anesthesia of the mother and fetus has been confirmed, all other necessary preparations continue: positioning of the mother, skin sterilization, and preparing the operation field.

Fetus Fetal weight is estimated by ultrasound and emergency medication is prepared (epinephrine, atropine).

Catheter and needling equipment (Fig. 12.1): The following items were prepared: 18G or 19G needles (Cook Medical®) depending on gestational age and distance to the fetal heart, floppy 0.0014 in. guidewires, and 2.5–4 mm semi-compliant, monorail coronary artery balloon catheters (Maverick®, Boston Scientific) with a manometer-equipped syringe and sterile strips to mark catheters and wires. The guidewire is inserted into the catheter and then both together through the needle after the trocar was removed. Markers on the catheter and the wires are placed to make sure the depth of insertion can be controlled. Then catheter and wire are removed from the needle; the trocar is inserted back again and handed over to the perinatologist to perform the puncture. The wire remains in the catheter and both will be inserted together, once the needle will be in place.

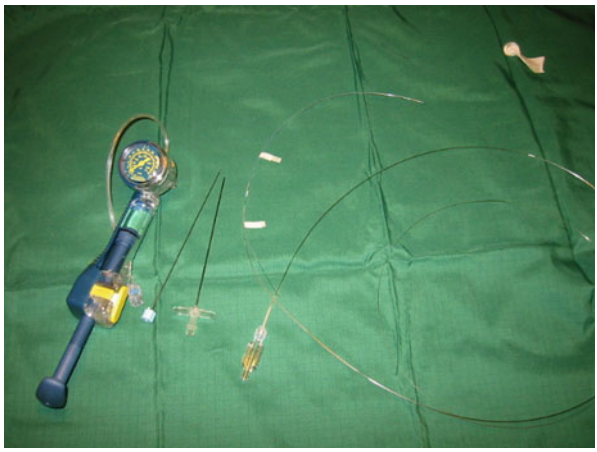

Fig. 12.1 Equipment used for fetal valvuloplasty: *left*, syringe with manometer for balloon inflation; *middle*, 18G needle with the trocar removed; and *right*, monorail balloon catheter with a 0.0014 in. floppy wire inside. Note the two markers on the catheter and on the guidewire, respectively

Ultrasound We use a Voluson E8 (GE Healthcare) ultrasound machine for imaging with standard curve array scanners equipped with Doppler and color Doppler. Keyboard and probe get a sterile cover. All procedures are recorded continuously on DVD.

12.2.4 Procedure

Puncture + imaging: This is the most critical part. The direction of the needle has to be perfect from the beginning, because larger manipulations or angle corrections are not possible once inside the fetal heart. So the synchronization between the scanning and needling person is crucial; sometimes both have to be done simultaneously by the same person. This procedure is significantly more challenging than aortic valve dilation, because the target, the hypoplastic RV, is usually very small. It has to be entered in the region of the RVOT about 1 cm proximal to the atretic valve and almost perpendicular to the fetal chest.

Testing the right needle position: Before perforating the atretic pulmonary valve or advancing a guidewire across a critical pulmonary stenosis, one has to be sure to be inside the cavity of the RV, which by ultrasound alone is very difficult to judge. So at this time, the trocar is slowly removed, and if blood is coming out through the needle (usually with quiet a high pressure), one can be sure to have an intracardiac position.

Perforation of the atretic valve: In the case of pulmonary atresia, the trocar has to be inserted again – be careful to do it very slowly; otherwise, one will push air bubbles into the RV and imaging will be a dramatic worse afterward. Under ultrasound guidance the needle is then advanced through the valve into the main PA. If there is critical pulmonary stenosis with still a tiny opening, the trocar should stay outside, and the whole ensemble (catheter with guidewire) will be slowly inserted. The ultrasound picture as well as the markers on the catheter will show you when the catheter tip has reached the tip of the needle. Then only the guidewire is advanced forward to cross the tiny opening, which is successful in most of the cases after a few attempts. Once the guidewire is clearly seen in the main PA or in the ductus arteriosus, the catheter is manipulated over the wire into an optimal position across the valve annulus (Fig. 12.2).

Performing the dilatation: Now the balloon is inflated with the respective diameter. Balloon size should be at least 10–20 % larger than the valve diameter. With the current available equipment, a 4 mm balloon will be the largest size that would fit through an 18G needle. Using larger needles will allow larger balloons, but will increase the likelihood for complications.

Withdrawal of all equipment: A deflated balloon sometimes cannot be retracted back into the needle again. Instead of applying increased force, we prefer to withdraw the needle together with the deflated balloon, which is still outside. After a successful dilatation, one should see good antegrade flow as well as some pulmonary regurgitation (Fig. 12.3a, b).

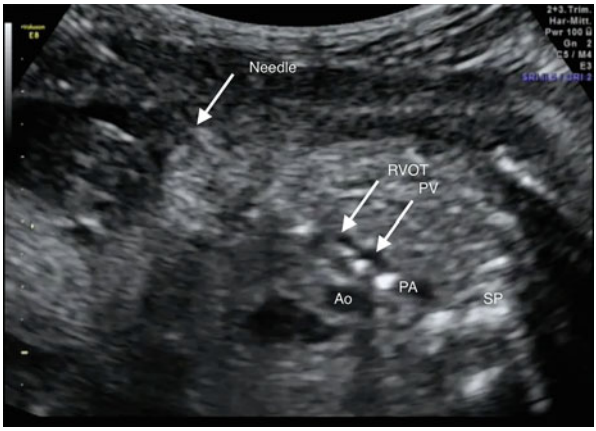

Fig. 12.2 Fetal pulmonary valvulotomy procedure: the needle is already passed from the right ventricular outflow tract through the perforated pulmonary valve in the main pulmonary artery. *Ao* aorta, *PA* pulmonary artery, *PV* pulmonary valve, *RVOT* right ventricular outflow tract

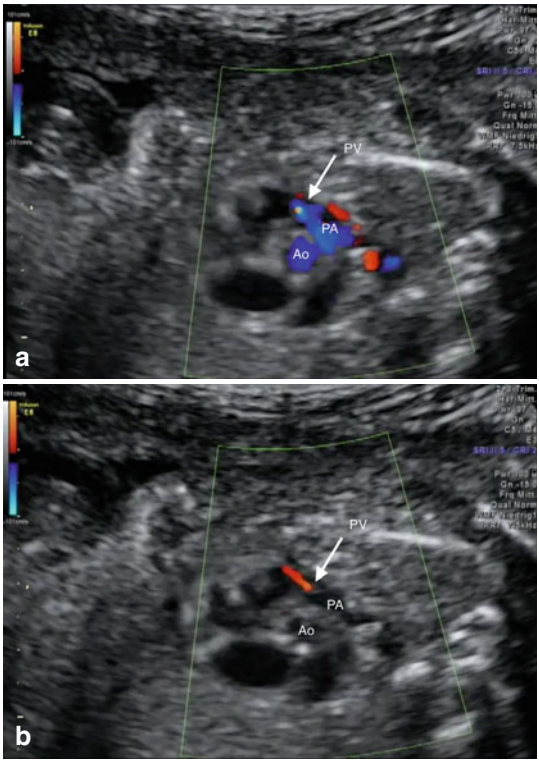

Fig. 12.3 Color Doppler after a successful pulmonary valvuloplasty: (**a**) picture in systole (note the blue color across the pulmonary valve indicating now antegrade flow from the right ventricle into the pulmonary artery) and (**b**) picture in diastole (note the red pulmonary regurgitation jet back into the right ventricle indicating some damage of the pulmonary valve after valvuloplasty). *Ao* aorta, *PA* pulmonary artery, *PV* pulmonary valve

12.2.5 Post-procedural Assessment

Immediate: Watch for pericardial effusion and drain instantly if it gets too large with signs of hemodynamic compromise. If persistent bradycardia develops, epinephrine is administered (according to estimated fetal weight) into the left ventricle (now using a 22G needle).

First 24–48 h: Nonstress test to watch for premature labor or fetal arrhythmias; observe if there are signs of premature rupture of the membranes or amnioinfection.

Long term: The patients were discharged 2 days after the procedure is stable, and follow-up visits were recommended every 2 or 4 weeks to assess hemodynamic changes.

So far we did not see any major maternal complication from this procedure [4].

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