# Literature on Fetal Therapy After Pulmonary Valvuloplasty

# 14

Roland Gitter and Gerald Tulzer

Due to the relative rarity of the lesion and the complexity of patient selection and the intervention itself, current numbers of reported cases are limited to approximately two dozens. The first publication worldwide was issued in 2002 by Tulzer et al. [1] from Children's Heart Center Linz, describing a successful valvulotomy of the pulmonary valve, performed in a fetus with heart failure at 28 weeks of gestation. Following the procedure significant growth of the tricuspid valve and the right ventricle was observed. The neonate was delivered at 38 weeks with a RV suitable for biventricular repair leading the authors to the conclusion that in utero pulmonary valvulotomy is feasible and may change the natural history of the condition in affected fetuses with PA/IVS [1]. In 2006 another successful intervention was reported by Galindo et al. [2] performed in a fetus with critical PS-IVS and heart failure at 25 weeks of gestation. After the procedure a significant improvement in fetal hemodynamics was seen up to 34 weeks, when a significant restenosis with signs of circulatory failure leads to premature delivery of the baby and an immediate postnatal valvuloplasty provided a biventricular repair for the baby [2].

Another 3 years later in 2009, the first consecutive series was published by Tworetzky W. et al. from the Boston group [3]. During a 6-year period, ten fetuses at a median gestational age of 24 weeks (range from 21–28 weeks) underwent the attempt of balloon dilation of the PV in utero. Whereas the first four procedures were technically unsuccessful, the most recent six were technically successful. Success was defined by the inflation of a balloon within the pulmonary valve followed by unequivocal antegrade blood flow across the valve detected by color Doppler imaging. Compared to 15 control fetuses with PA/IVS who did not undergo prenatal intervention and out of whom nine had univentricular outcomes after birth, the tricuspid

R. Gitter (✉) • G. Tulzer
Department of Pediatric Cardiology, Children's Heart Center Linz,
Kepler University Hospital Linz, Linz, Austria
e-mail: Roland.gitter@gespag.at



G. Butera et al. (eds.), *Fetal and Hybrid Procedures in Congenital Heart Diseases*, DOI 10.1007/978-3-319-40088-4_14

valve annulus, right ventricle length, and PV annulus grew significantly more from mid- to late gestation in the six fetuses who underwent successful interventions. Five of these successfully treated patients had maintained antegrade pulmonary blood flow postnatally and underwent neonatal augmentation of the right ventricular outflow tract combined with a systemic to pulmonary shunt, which could be closed later on in four patients – the fifth patient in the intermediate period at the time of the paper's publication. Biventricular circulation – in which per definition the right ventricle is the only source of pulmonary blood flow, with systemic arterial saturation of more than 90 % in room air, with or without interatrial communication – was the primary goal of the intervention and could be achieved in five out of six successfully treated valves. The authors conclude that the initial results are promising, but fetal intervention for PA/IVS remains an experimental therapy, and it is necessary to compare the outcomes of this approach with those of postnatal transcatheter and surgical interventions that are currently the standard of care. The risk/benefit balance must be clear before prenatal intervention becomes an accepted therapy.

In 2012 Polat et al. [4] yielded a case of PA/IVS which they followed during mid-gestation from 24–28 weeks, observing increased tricuspid regurgitation and a lack of growth of the right ventricle ending up with a tricuspid valve annulus Z-score of −2.8. Intervention was successfully performed at 28 weeks. Immediately after the procedure, there was an improvement in fetal hemodynamics, but no further follow-up is described in this case report. Another small series of four interventions was published by Gomez-Montes et al. [5] in 2012. Fetal valvuloplasty was offered to three fetuses with signs of heart failure and one fetus with a predicted univentricular course. All interventions were considered successful; two babies died postnatally, but the other two were fine with a biventricular course at follow-up period of 8 years and one-and-a-half ventricle repair after 21 months, respectively [5].

In 2014 the group of Children's Heart Center Linz reported their experience of 12 attempted fetal valvuloplasties performed in 10 fetuses with suprasystemic RV pressures. Eight cases were considered successful and the intervention resulted in better RV filling and continuous, but slower than normal growth of the tricuspid valve and RV. Out of the first five newborns, four were biventricular at the end of the first year; one child received a Glenn shunt. One 6-month-old patient still has a Blalock-Taussig shunt; two fetuses were still in utero at the time of publication. Currently there is no literature available concerning the definitive treatment of fetuses, who underwent intrauterine pulmonary valvuloplasty. This field is predominantly still unexplored resulting in a lack of publications.

## References

1. Tulzer G, Arzt W, Franklin RCG, Loughna PV, Mair R, Gar- diner HM. Fetal pulmonary valvuloplasty for critical pulmonary stenosis/atresia with intact septum. Lancet. 2002;360(9345):1567–8.
2. Galindo A, Gutierrez-Larraya F, Velasco JM, de la Fuente P. Pulmonary balloon valvuloplasty in a fetus with critical pulmonary stenosis/atresia with intact ventricular septum and heart failure. Fetal Diagn Ther. 2006;21:100–4.

3. Tworetzky W, McElhinney DB, Marx GR, et al. In utero valvuloplasty for pulmonary atresia with hypoplastic right ventricle: techniques and outcomes. Pediatrics. 2009;124(3):e510–8.
4. Polat T, Danısman N. Pulmonary valvulotomy in a fetus with pulmonary atresia with intact ventricular septum: first experience in Turkey. Images Paediatr Cardiol. 2012;14(3):6–11.
5. Gómez Montes E, Herraiz I, Mendoza A, Galindo A. Fetal intervention in right outflow tract obstructive disease: selection of candidates and results. Cardiol Res Pract. 2012;2012:592403.