Transventricular valvotomy for pulmonary atresia with intact ventricular septum in neonates: a single-centre experience in mid-term follow-up

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Abstract

OBJECTIVES: Transventricular valvotomy is a kind of hybrid therapy that is widely used in developing countries where cardiac catheter intervention is limited in neonates. The purpose of this study was to evaluate the surgical outcomes and effectiveness of transventricular valvotomy in neonates with pulmonary atresia and an intact ventricular septum.

METHODS: Between August 2008 and July 2013, 22 neonates with pulmonary atresia and an intact ventricular septum underwent transventricular valvotomy at our institution. The mean age and weight at initial palliation were 14.3 ± 5.2 days (range 1–24 days) and 3.3 ± 0.6 kg (range 2.5–3.8 kg), respectively. The mean tricuspid valve Z-score was −1.7 ± 1.09 (range −3.5 to −0.2). All patients presented with membranous atresia and no evidence of Ebstein’s malformation or right ventricle-dependent coronary circulation.

RESULTS: There were two hospital deaths and no late deaths. Postoperative echocardiography showed that the gradient over the open pulmonary valve was 26.7 ± 5.4 mmHg (range 20–32 mmHg). Patients who received patent ductus arteriosus ligation and were treated with a modified Blalock–Taussig shunt experienced a shorter intensive care unit stay than those without patent ductus arteriosus ligation ($P = 0.004$). The median follow-up time was 32.5 months (range 6–60 months). The freedom from reoperation was 85, 78.9, 56.5% at 6 months, 1 and 5 years, respectively. Fourteen patients underwent definitive repair, 10 underwent biventricular repair, and four received one and one-half ventricle repair.

CONCLUSIONS: Transventricular valvotomy can be used successfully as an easy and safe alternative to right ventricular decompression. Furthermore, we note that oxygen saturation is a valuable indicator in determining whether a modified Blalock–Taussig shunt should be performed or not.

Keywords: Congenital heart disease • Pulmonary arteries • Surgery

INTRODUCTION

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare congenital heart disease characterized by an atretic pulmonary valve (PV) and results in limited communication between the right ventricle (RV) and the pulmonary trunk. Consequently, most children suffer from severe cyanosis soon after birth. Surgical correction to provide a reliable source of pulsatile flow is required early in order to increase arterial oxygen saturation ($\text{SaO}_2$) and promote growth of the RV and pulmonary vascular beds.

Pulmonary atresia with intact ventricular septum manifests a wide spectrum of anatomical heterogeneity, including a well-developed or hypoplastic RV, varying sizes of tricuspid valve (TV), and the potential for presence of RV-dependent coronary circulation. In fact, these differences are so distinct that assessment of RV morphology is necessary in order to determine whether to proceed with bi- or univentricular repair.

Previously, there were only two kinds of RV decompression, namely conventional surgical therapy, including surgical valvotomy or right ventricular outflow tract (RVOT) reconstruction, and transcatheter-based intervention, including balloon valvotomy. As one might imagine, both techniques have noteworthy advantages and risk factors. However, transcatheter-based intervention is limited in neonates in developing countries and, as a result, transventricular valvotomy has become a more popular alternative, with several reports indicating positive results in patients with PA/IVS. [1–3].

The present study summarizes the results from our recent use of transventricular valvotomy in 22 neonates with PA/IVS in order to evaluate the effectiveness of this method and better define future guidelines for more successful patient selection.
PATIENTS AND METHODS

With approval of the Ethics Committee, 22 neonates with PA/IVS underwent transventricular valvotomy at Shanghai Children’s Medical Center from August 2008 to July 2013. All neonates were admitted to our hospital as emergency patients and were administered prostaglandin prior to surgery. Cardiac anomalies were diagnosed by two-dimensional and Doppler echocardiography. Great attention was paid to the RV cavity, the type of pulmonary atresia (membranous versus muscular), TV size and function, and coronary artery abnormalities. These details, in addition to demographic data, are shown in Table 1. Patients with severely hypoplastic RV who underwent univentricular repair or those who presented with Ebstein’s malformation, RV-to-coronary artery fistulas or right ventricle-dependent coronary circulation were excluded from this study.

Surgical technique

The initial transventricular valvotomy included pulmonary valvuloplasty and balloon dilatation with simultaneous patent ductus arteriosus (PDA) ligation or a modified Blalock-Taussig (mBT) shunt. Following a median sternotomy, a purse-string suture was made in the RVOT ~1.5–2.0 cm away from the pulmonary trunk. With the continuous guidance of transoesophageal echocardiography, a 16 gauge intravenous catheter was used to puncture the RVOT wall through the middle of the purse string and then perforate the atretic PV. A guide wire was passed through the needle, and a sheath was passed over the guide wire into the RVOT (Fig. 1). The balloon diameter was set at 100% of the PV annulus and was followed by consecutive balloon dilatations of the PV until transoesophageal echocardiography confirmed adequate pulmonary pulsatile blood flow (Fig. 2). At the end of this procedure, the change in intraoperative SaO2 was used to evaluate whether a PDA ligation or mBT shunt should be performed.

Statistical analysis

Data were analysed with the statistical computing package SPSS 20.0. Preoperative and postoperative data were collected from medical records. The continuous data were expressed as a median value for non-normally distributed variables and as a mean ± SD for normally distributed variables. The categorical data were listed as frequencies. Two-group comparisons were assessed with Student’s t-tests for continuous variables that were normally distributed and Fisher’s exact tests for categorical data. Rates for survival and reoperation were estimated with a Kaplan–Meier curve. Values of P < 0.05 were considered statistically significant.

RESULTS

The median duration of intensive care unit stay was 5.5 days (range 2–24 days) and the median duration of mechanical ventilation was 46.5 h (range 14–336 h). There were two hospital deaths and no late deaths. One hospital death occurred on postoperative day 2 due to severe low cardiac output syndrome. The other early
death was attributed to ventricular tachyarrhythmia. Overall survival after initial palliation was 90.9% at 6 months and 1 year.

Postoperative transthoracic echocardiography showed that the gradient over the open PV was 26.7 ± 5.4 mmHg (range 20–37 mmHg) and no patients suffered from more than mild pulmonary insufficiency. Tricuspid regurgitation was improved postsurgery. Postoperative mild tricuspid regurgitation was present in 15 patients, while moderate tricuspid regurgitation was detected in only five patients.

In this study, 16 patients received PDA ligation and an additional mBT shunt, while five patients remained with the PDA left open. The remaining patient underwent PDA banding. The patent duct was banded to approximate 3–4 mm to avoid pulmonary overcirculation. Patients who received PDA ligation and mBT shunt experienced shorter intensive care unit stays than those with the PDA left open (P = 0.004).

The duration of postoperative prostaglandin infusion was 5.7 ± 2.3 days. Two patients experienced persistent cyanosis after weaning from prostaglandin. Transthoracic echocardiography indicated an unsuccessful transventricular valvotomy in one patient with a 75 mmHg gradient over the open PV. This patient underwent RVOT reconstruction with pulmonary valvotomy and a transannular patch. The other patient received a PDA stent 6 days after initial operation.

Follow-up data were available for all surviving patients. The median follow-up time was 32.5 months (range 6–60 months). Patients were required to return to hospital every 3 months after discharge. Transthoracic echocardiography indicated moderate pulmonary insufficiency in four patients and moderate tricuspid regurgitation in six patients.

At follow-up examination, the mean gradient over the open PV for patients before reintervention or definitive repair increased to 52.7 ± 16.4 mmHg (range 26–84 mmHg). Reintervention (transcatheter balloon dilatation) was performed in two patients at 6 and 9 months following the initial transventricular valvotomy as a result of unsatisfactory RV decompression and progressive cyanosis.

Definitive repair was performed in 14 patients. The median interval between initial palliation and definitive repair was 14.5 months (range 12–20 months). Biventricular repair was performed in 10 patients, and the remaining four patients received one and a half ventricle repair (Fig. 3). Reintervention was performed in two patients for postoperative severe TV regurgitation and in one patient for pulmonary branch stenosis.
DISCUSSION

The present study presents one of the largest published data sets for transventricular valvotomy in patients with PA/IVS thus far and demonstrates that transventricular valvotomy can be achieved successfully (as indicated by decreases in mortality and morbidity) in neonates with PA/IVS. To a large extent, this procedure simulates aspects of RV decompression by providing adequate pulmonary circulation for RV development.

A previous study reported the 5-year survival rate in treated neonates with PA/IVS to be <50% [4]. However, more recent studies indicated an increase in the survival rate, ranging from 60 to ∼98% at 5 years [5–8]. Despite the improved survival rates, controversy still exists regarding the most effective surgical method to employ based on specific characteristics related to the disparate presentation of morphologies.

Based on the degree of RV hypoplasia, the size and function of the TV and the presence or absence of RV-dependent coronary circulation, the surgical strategy for PA/IVS usually includes biventricular repair, one and a half ventricle repair and univentricular repair [9–11]. Patients with a tripartite or even bipartite RV and TV Z-scores larger than ∼3.5 may be candidates for further biventricular repair, demanding RV decompression during the neonatal period [1, 12]. Right ventricular decompression is used to reconstruct the connection between the RV and the pulmonary circulation in order to increase SaO2 and promote RV growth. This method can also result in improvement of impaired left ventricular function [13–15].

Transcatheter-based intervention as an alternative to RV decompression has been widely used in developed countries. However, the necessary technology to enable this kind of procedure to be performed on neonates is lacking in many developing countries. As an alternative, transventricular valvotomy is a minimally invasive procedure that is considered safe and relatively easy to perform. Furthermore, it permits surgeons to use an mBT shunt or PDA ligation during the initial palliation.

An increasing number of studies have reported limitations and negative results regarding transcatheter-based intervention [16, 17]. However, it should be noted that the majority of the negative reports are likely to result from the high rate of reintervention or conversion to open surgery due to inadequate pulmonary pulsatile blood flow and prolonged prostaglandin administration after the initial palliation. Cheung et al. [18] reported that nine of the original 15 patients required a reinterventional catheterization. Likewise, McLean et al. [17] noted that only five of their 14 patients were uneventful at discharge. Even after a successful transcatheter-based intervention, more than half of the patients required an mBT shunt or PDA stent and RVOT reconstruction within 1 month of intervention [14, 19].

Previously, the criteria for patient selection for transventricular valvotomy have not been unanimous. At our institute, we are confident that transventricular valvotomy can be achieved successfully in most patients who need RV decompression and who do not present with RV-dependent coronary circulation, muscular atresia, severe RV and TV hypoplasia or Ebstein’s anomaly. However, until now, these determinations were made based on our experience and not based on published data. Although several studies report biventricular repair in patients with Ebstein’s anomaly [20], we still exclude these patients because of the likelihood of RV volume overload due to severe TV regurgitation and decreased forward flow. Transventricular valvotomy is also unsuitable for patients with a hypoplastic PV annulus. Consecutive balloon dilatation will not enlarge the PV annulus but instead will result in severe pulmonary regurgitation. The goal of transventricular valvotomy is to help neonates survive early in life; thus, for patients who are old enough to undergo surgical treatment, RVOT reconstruction is preferable to transventricular valvotomy.

There are some noteworthy differences specific to transcatheter-based intervention. When perforating the atretic PV, it is possible for the needle not to be positioned in the middle of the valve plate. To avoid such a situation, a purse-string suture should be made 1.5–2.0 cm away from the pulmonary trunk and the needle should not be at too great an angle to the RVOT wall. Transoesophageal echocardiography should be used to monitor this procedure carefully. We typically inflate the balloon to 100% of the size of the PV annulus (instead of 120–125%) to avoid potential damage to the PV before the balloon is positioned in the middle of the valve.

Some authors have suggested a more reliable source of pulmonary blood flow to the pulmonary artery shunt (i.e., systemic) to relieve cyanosis [17]. While we agree that the pulmonary circulation will be reliant on a systemic-to-pulmonary artery shunt, it is difficult to decide whether an mBT shunt should be used or whether the PDA should be left open. Li et al. [1] recommended using an mBT shunt in all patients <1 month old. However, we focus on the SaO2 to evaluate which route to take. If SaO2 levels are 80–85% after transventricular valvotomy, an mBT shunt is used. If SaO2 increases to 85–90% after transventricular valvotomy, we try to occlude the PDA during the procedure in order to monitor the changes in SaO2 before the PDA is left open. If the patient’s saturation decreases to <75–80% after the tentative PDA occlusion, an mBT shunt is required. If SaO2 remains close to 80%, the PDA is left open. In this study, five patients were left with their PDA open. In two of them, we detected spontaneous closure of the PDA but with acceptable oxygen saturation levels. One patient required conversion to RVOT reconstruction, one patient received a PDA stent and in another patient the PDA remained open. Although leaving the PDA open will increase the risk for postoperative reintervention and intensive care unit stay, it will also circumvent the need to perform and undo the mBT shunt during definitive repair.
A transoesophageal echocardiographic examination during the follow-up period is essential for patients with PA/IVS to evaluate the effectiveness of transventricular valvotomy, in order to assess the growth of the RV and TV and to determine the feasibility of biventricular repair. Once the patient is a candidate for definitive repair, we feel the procedure should be scheduled immediately. It is impossible for transventricular valvotomy to relieve pulmonary stenosis and infundibular thickening. Moreover, the surgical success of transventricular valvotomy is less favourable than that of RVOT reconstruction. It is therefore necessary for patients to undergo an early definitive repair, especially when progressive TV regurgitation and restrictive atrial septal defect are present. Right ventricular volume overload can be devastating in these patients.

While the results presented in the present study demonstrate that transventricular valvotomy can be used successfully as an easy and safe alternative to RV decompression, there are several limitations. This is a retrospective and non-randomized clinical trial, and some results may be affected by selection bias. It should also be noted that this is a single-centre report that includes a small patient population with follow-up times not exceeding 5 years. Furthermore, our facility lacks the magnetic resonance imaging capabilities to be able to evaluate right and left ventricular function properly before and after transventricular valvotomy. It is best to use the results of this study for comparison of the different kinds of RV decompression in order to provide the best representation of the advantages of this procedure.

In conclusion, transventricular valvotomy can be achieved successfully in neonates with PA/IVS in terms of mortality and morbidity. The hybrid procedure is an easy and safe alternative to RV decompression in developing countries. Furthermore, oxygen saturation is a valuable indicator of whether an mBT shunt is necessary. Follow-up investigation is required to evaluate the effectiveness of this procedure and to determine when to perform the definitive repair.

Conflict of interest: none declared.

REFERENCES