Transthoracic Balloon Pulmonary Valvuloplasty for Treatment of Congenital Pulmonary Atresia Patients with Intact Ventricular Septum

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Background: To summarize our clinical experience in performing transthoracic balloon pulmonary valvuloplasty for the treatment of patients suffering from congenital pulmonary atresia with intact ventricular septum (PA/IVS).

Material/Methods: Between April 2009 and April 2016, 38 patients with PA/IVS underwent transthoracic balloon pulmonary valvuloplasty in our hospital. All of them were combined with patent ductus arteriosus, tricuspid insufficiency, and atrial septal defect or patent foramen ovale. The valvuloplasty was performed from the right ventricular outflow tract through a median sternotomy incision under TEE guidance for all cases.

Result: Thirty-five patients were successfully discharged, and 3 patients died after the operation. The 35 surviving patients were followed up. SpO2 in the 35 patients was 88–96% after the operation. The transpulmonary valvular gradient pressure was less than or equal to 30 mmHg in 31 patients and between 36 and 52 mmHg in the other 4 patients. After the surgery, tricuspid regurgitation was significantly reduced. We found only 4 patients with moderate regurgitation, 5 patients with mild to moderate regurgitation, and mild regurgitation in the remaining 26 patients. Five patients underwent a second-stage operation, including biventricular repair in 4 patients and ligation of ductus arteriosus in 1 patient.

Conclusions: The application of transthoracic balloon pulmonary valvuloplasty for the treatment of PA/IVS is minimally invasive and safe, which has great significance for improving the curative effect for this condition and reducing operation mortality.

MeSH Keywords: Cardiac Catheterization • Heart Defects, Congenital • Pulmonary Atresia

Abbreviations: PA/IVS – pulmonary atresia/intact ventricular septum; PDA – patent ductus arteriosus; ASD – atrial septal defect; BT – shunt Blalock-Taussig shunt; RVOT – right ventricular outflow tract

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Background

Pulmonary atresia with intact ventricular septum (PA/IVS), a rare, complex congenital heart disease with extensive morphological variability, continues to be a difficult clinical problem. Although techniques for surgery and extracorporeal circulation have improved in recent years, the operative mortality is still as high as 5.2–16% [1–3]. As methods for minimally invasive surgery and the associated technology have progressed, transthoracic balloon pulmonary valvuloplasty, a hybrid procedure that combines catheter-based therapy and traditional surgical intervention, has become an attractive alternative to the standard surgical or catheter-based techniques [4–7]. From April 2009 to April 2016, this hybrid procedure was performed on 38 patients with PA/IVS in our hospital. The clinical effect was satisfactory, and a report of our experiences in performing the procedure follows.

Material and Methods

The present study was approved by the Ethics Committee of Fujian Medical University, China and adhered to the Declaration of Helsinki. Written informed consent was acquired from the patients or the patient’s relatives.

Thirty-eight patients with congenital pulmonary atresia with intact ventricular septum (PA/IVS), including 26 males and 12 females, were enrolled at our institution from April 2009 to April 2016 (Table 1). The patients’ ages ranged from 11 days to 1.5 years (0.8±0.37 years). Their weight ranged from 2.8 to 11.7 kg with the average weight being 5.6±1.2 kg. The hospitalization days were from 7 days to 1 month, with a median of 18 days. There were clinical symptoms of PA/IVS in 36 cases. Preoperative SPO2 in the resting state was 65–86% with a mean of 71.5±6.1%. Preoperative diagnosis was based primarily on color Doppler echocardiography. All 38 patients presented with membranous pulmonary atresia, and all of them were combined with patent ductus arteriosus, tricuspid insufficiency, and atrial septal defect or patent foramen ovale. Prostaglandin E1 was administered to maintain arterial duct patency for pulmonary oxygen exchange. Nine patients underwent tracheal intubation and ventilator support because of obvious polypnea and low saturation of blood oxygen. Preoperative echocardiography showed that the z value of the tricuspid valve ranged from 0 to –2.0, and there were 5 patients in which the z value of the tricuspid valve ranged from –1 to –2.0. The results showed that the 3 components (inlet portion, trabecular portion, and outlet portion) of the right ventricle were well developed in 36 patients who underwent cardiac CT angiography before the operation. Individuals with right ventricular dysplasia or other cardiac malformations were excluded from this study.

Table 1. Clinical data of patients undergoing transthoracic balloon pulmonary valvuloplasty of the 38 patients with PA/IVS.

| Item                              | Value   |
|----------------------------------|---------|
| Sex (M: F)                       | 26: 12  |
| Age (years)                      | 0.8±0.37|
| Weight (kg)                      | 5.6±1.2 |
| SPO2 (%)                         | 71.5±6.1|
| Z value of TVA (–2—1)            | 5       |
| Z value of TVA (–1—0)            | 33      |
| Operative time (minutes)         | 56.2±11.5|
| ICU stay (hours)                 | 5.3±13.5|
| Hospital stay (days)             | 8.5±7.2 |
| Follow-up (months)               | 24.5±12.3|
the patent ductus arteriosus and BT shunting. The atrial septal defect and patent foramen ovale were not addressed. All the patients underwent echocardiography to assess the transpulmonary valvular gradient pressure and the degree of pulmonary insufficiency and tricuspid insufficiency while they were in the intensive unit (ICU) and 3 months after surgery. SPO2 in the resting state was recorded 3 months after surgery. Five patients had to undergo a second-stage operation, including ligation of the ductus arteriosus in 1 patient at 1 year after surgery. The other 4 patients underwent biventricular repair, which consisted of ligation of the BT shunt followed by a longitudinal incision on the right ventricular outflow tract, passing through the pulmonary annular, towards the proximal part of the right and left pulmonary artery. Transannular patching to the pulmonary bifurcation was performed. Simultaneous surgical procedures included repair of the atrial septal defect in 4 patients and tricuspid valvuloplasty in 2 patients.

### Results

Thirty-five patients were satisfactorily discharged; all were followed up for 3–83 months, with an average of 28.2 months. SPO2 in the resting state at 3 months after surgery in the 35 surviving patients was 88–96%, with a mean of 92.3±3.8%. The transpulmonary valvular gradient pressure was less than or equal to 30 mmHg in 31 patients and between 36 and 52 mmHg (38.2±6.2 years) in the other 4 patients. Nine patients in whom the PDA had not been addressed underwent echocardiography at 6 months after the operation. The ductus arteriosus had closed spontaneously in 4 patients. The ASD had closed spontaneously in 6 patients. Twenty-six patients were associated with tricuspid insufficiency. Severe tricuspid insufficiency was observed in 4 patients, moderate tricuspid insufficiency in 15 patients, and mild tricuspid insufficiency in 16 patients. There were 22 patients with mild pulmonary regurgitation, 12 patients with moderate pulmonary regurgitation, and no severe pulmonary regurgitation observed in any patient. Five patients underwent a second procedure without operative death, and the complications in these 5 patients were as follows: pulmonary infection in 1 patient and right heart failure in 1 patient.

### Table 2. The surgical methods used in treating PA/IVS.

| Age       | Type of surgical methods                                      | No. of patients |
|-----------|----------------------------------------------------------------|-----------------|
| <1 month  | BPV + Ligation of PDA + B-T shunt                             | 13              |
|           | BPV                                                            | 2               |
| >1 months | BPV + Ligation of PDA + B-T shunt                             | 5               |
|           | BPV                                                            | 9               |
|           | BPV                                                            | 9               |

BPV – balloon pulmonary valvuloplasty; PDA – patent ductus arteriosus.

### Table 3. Early postoperative complications.

| Item                  | No. (%) |
|-----------------------|---------|
| Pulmonary infections   | 8 (21.1%) |
| Right heart failure    | 3 (7.9%)  |
| Renal failure          | 2 (5.3%)  |
| Arrhythmia             | 3 (7.9%)  |

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Three patients died after the operation. The z value of the tricuspid valve in these patients ranged from −1 to −2.0. One patient (aged 20 days) with PA/IVS with PDA who underwent balloon pulmonary valvuloplasty and simultaneous surgical procedures, including BT shunting and ligation of the ductus arteriosus, died of pulmonary infection and right heart failure. Pulmonary insufficiency and tricuspid insufficiency were moderate to severe after surgery in 1 patient (aged 42 days) with PA/IVS with PDA and ASD, who underwent balloon pulmonary valvuloplasty and ligation of the ductus arteriosus, and the patient then died of pulmonary infection and renal failure. Severe hypoxia occurred after surgery in 1 patient with PA/IVS with PDA who underwent balloon pulmonary, and the patient then died due to multiple organ failure.

**Discussion**

With the development of imaging technology and interventional devices, the strategy for the treatment of congenital heart disease has changed from surgery alone to a hybrid procedure combining catheter-based therapy and traditional surgical intervention. Both surgical and transcatheter approaches have drawbacks, but the hybrid procedure can maximize the potentials and minimize the limitations of both to reduce patient risk. It may thus be a new therapeutic option for complex congenital heart disease [8]. As medical technology has advanced, the hybrid procedure has attracted increasing attention due to its outstanding advantages.

PA/IVS, a type of cyanotic congenital heart disease that involves duct-dependence and has a high surgical mortality rate, has always posed a challenging treatment problem. With the development of new medical technologies, the mortality rate of PA/IVS was gradually reduced from 56% to 16% [9]. At present, improving hypoxia, promoting development of the right ventricle, and providing adequate blood flow to the pulmonary artery are the main problems facing the physician. Medical treatment of PA/IVS has the main goal of maintaining the patency of the ductus arteriosus to prevent severe hypoxia and metabolic acidosis. Pulmonary valvotomy is the traditional surgical treatment for critical pulmonary stenosis and PA/IVS in neonates and infants [10]; however, recent advances have led percutaneous balloon pulmonary valvuloplasty (PBPV) to become the preferred treatment method for pulmonary valve stenosis. PBPV has also been applied for the treatment of PA/IVS [11], but it is generally appropriate only for patients aged 2–4 years. Indeed, PBPV has rarely been applied to treat infants with severe cyanosis, and percutaneous perforation and ballooning have been associated with higher rates of procedural failure and serious complications. Unfortunately, even after successful percutaneous balloon valvuloplasty, 43–51% of neonates require urgent procedures, including Blalock-Taussig shunt placement, RVOT reconstruction, or both [12]. Complications of percutaneous intervention, such as cardiac rupture, cardiac perforation, damage to papillary muscles and chordae tendineae, occur easily in neonates and infants. It has also been reported that performing PBPV in infants is relatively intratable, with a high failure rate [13].

Typically, the operation method is selected according to the development of the tricuspid valve and the right ventricle and the existence of right ventricular-dependent coronary circulation [14,15]. Depending on the report, the z value of the tricuspid valve in patients with PA/IVS with severe right ventricular dysplasia is <−4 [16]. Wu Song [17] reported that the z value for the tricuspid valve in neonates and infants with a well-developed right ventricle and without muscular stenosis of the right ventricle outflow tract ranged from 0 to −1. Thirty-eight patients in our study presented with membranous pulmonary atresia, and we obtained the z value of the tricuspid valve for all of them. For all patients, the 3 components (inlet portion, trabecular portion, and outlet portion) of the right ventricle were well developed and right ventricular-dependent coronary circulation was absent. All 38 patients underwent the hybrid procedure successfully. However, 3 patients later died, and the z value of the tricuspid valve for these patients ranged from −1 to −2.0. Two patients died due to severe pulmonary infection, recurrent heart failure, or renal failure. One patient died of severe hypoxia and multiple organ failure. Despite these setbacks, the overall results were satisfactory. Postoperative SPO2 in the resting state in the surviving patients was 88–96%, and postoperative echocardiography showed that the transpulmonary valvular gradient pressure was less than or equal to 30 mmHg in 88.6% of the survivors, revealing that transthoracic balloon pulmonary valvuloplasty had advantages for the treatment of infants or neonates with PA/IVS. Balloon pulmonary valvuloplasty can cause pulmonary regurgitation; therefore, surgeons must obtain color Doppler ultrasound imaging of the pulmonary valve fusion and accurately deploy the balloon to the central coaptation point. The final deployed diameter is smaller than the diameter of the pulmonary artery ring [18]; thus, severe pulmonary regurgitation is relatively rare. Moderate to severe postoperative pulmonary insufficiency and tricuspid insufficiency occurred in only 1 patient, causing high pressure in the right atrium. The patient then died of renal failure. Normal or near-normal pulmonary valve function was beneficial to the maintenance of right ventricular function. After balloon pulmonary valvuloplasty, the pressure in the right ventricle was reduced, and the forward flow through the right ventricle could promote right ventricle development, potentially improving the size and function of the right ventricle [19]. The procedure also significantly reduced tricuspid regurgitation in the patients in this group. PA/IVS is characterized by varying degrees of dysplasia of the right ventricle; thus, the prevention of right heart...
failure after the operation is very important. Alprostadil can be applied to reduce pulmonary arterial resistance, milrinone to enhance the heart’s contractile ability, and diuretics to enhance the diuretic effect.

Our experiences performing transthoracic balloon pulmonary valvuloplasty can be summarized as follows: (1) Patients with membranous pulmonary atresia should be strictly selected. The \( z \) value of the tricuspid valve should range from 0 to –1, and the 3 components (inlet portion, trabecular portion, and outlet portion) of the right ventricle should be well developed. (2) The piercing process should be gentle to avoid pulmonary artery perforation. (3) In principle, the balloon diameter should be 120–140% larger than the diameter of the pulmonary valve annulus. (4) The puncture point should be located in the right ventricular outflow tract 1–1.5 cm away from the pulmonary trunk. (5) Esophagus echocardiography can be used for guidance during the operation. (6) The occurrence of atrial septal defect together with PA/IVS was not addressed in our group. Not all patients should undergo ligation of the ductus arteriosus, as the PDA can provide adequate pulmonary blood flow [20]. During the follow-up of our cases, the ductus arteriosus had closed spontaneously in approximately 45% of the patients in whom the PDA had not been addressed during the operation. (7) According to our experience, the indications for balloon pulmonary valvuloplasty concomitant with BT shunting are as follows: in the neonatal period; infants aged more than 1 month, although there was no significant change in \( \text{SatO}_2 \) after balloon pulmonary valvuloplasty; and patients with a small PDA.

Compared to the traditional surgical method and catheter interventional treatment, utilizing thoracotomy for balloon pulmonary valvuloplasty for the treatment of PA/IVS has the following advantages: (1) It has little negative effect on cardiac function because the operative route is short, and the operation is performed without myocardial incision and without cardiopulmonary bypass. (2) It can avoid the complications of extracorporeal circulation and reduce the pulmonary inflammatory response, thereby reducing the operative mortality rate. (3) The hybrid approach can result in decreased risk, less invasive procedures, and improved outcome [21].

Conclusions

In short, the hybrid procedure may be an effective treatment method for infants with PA/IVS. Importantly, it can improve the efficacy of surgical treatment for PA/IVS, although careful assessment of the surgical indications is necessary. Only patients with membranous pulmonary atresia with a \( z \) value of the tricuspid valve ranging from 0 to –1 should be selected, and the 3 components (inlet portion, trabecular portion, and outlet portion) of the right ventricle should be well developed. Selected patients should also be without right ventricular-dependent coronary circulation and should have no muscular stenosis of the right ventricle outflow tract. A systematic postoperative follow-up plan should also be conducted, and second-stage operative schemes executed as soon as possible. It should be noted that the application of the hybrid procedure for the treatment of PA/IVS is still in its infancy, and the best methods for hybrid operating room construction, improvement of interventional devices, and other aspects require additional clinical exploration and research [21].

Competing interests

The authors declare that they have no competing interests.

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