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

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCE Zhi-qin Lin*
ABCE Qiang Chen*
CDF Hua Cao
CFG Liang-wan Chen
DF Gui-can Zhang
DEF Dao-zhong Chen
DEF Qin-min Wang
DEF Han-fan Qiu
DEF Dong-shan Liao
ABCDEFG Feng Lin

Department of Cardiovascular Surgery, Union Hospital, Fujian Medical University, Fuzhou, Fujian, P.R. China

* These authors contributed equally to this study and share first authorship

Corresponding Author: Feng Lin, e-mail: linfeng0781@163.com

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Background: To summarize our clinical experience in performing transthoracic balloon pulmonary valvuloplasty for the treatment of patients suffering from congenial 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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2591 3 2 21

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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 congenial 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	
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 operation was performed under general anesthesia with endotracheal intubation. We cut open the pericardium to expose the heart through a median sternotomy in the supine position. The entire process was performed on a beating heart (off-pump method). After systemic heparinization, a purse-string suture was positioned in the right ventricular outflow tract 1–1.5 cm away from the pulmonary trunk. Then, a fine needle and a guide wire were punctured through the pulmonary valve to the pulmonary artery (Figure 1). A balloon catheter was inserted under the guidance of a guide wire. Sequential dilations were performed (the size of the balloon diameter was selected as 120–140% larger than the diameter of the pulmonary valve annulus) until forward diastolic flow through the pulmonary valve was observed with the guidance of epicardial echocardiography (Figure 2). Finally, the guide wire and the balloon catheter were withdrawn, and the incision was closed. According to intraoperative exploration, simultaneous surgical procedures, including BT shunting and/or ligation of the ductus arteriosus, were performed on some patients (Table 2). Among 15 infants aged less than 1 month, 13 underwent simultaneous surgical procedures, including ligation of the ductus arteriosus and BT shunting, after the balloon pulmonary valvuloplasty. The surgical procedure depended on arterial oxygen saturation in patients aged more than 1 month. Nine patients in whom the oxygen saturation increased to more than 85% after balloon pulmonary valvuloplasty underwent ligation of the patent ductus arteriosus. The oxygen saturation decreased significantly in 9 patients when we attempted to clamp the ductus arteriosus after balloon pulmonary valvuloplasty; thus, we did not perform ligation of the patent ductus arteriosus in those cases. Five patients in whom the oxygen saturation showed no obvious changes after balloon pulmonary valvuloplasty underwent simultaneous surgical procedures, including ligation of

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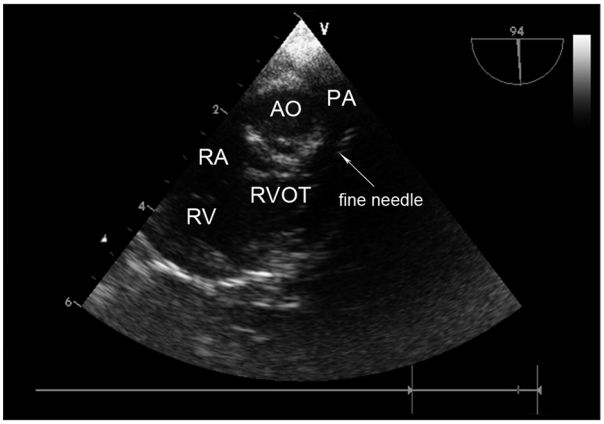

Figure 1. The fine needle was punctured through the pulmonary valve to the pulmonary artery through the right ventricular outflow tract.

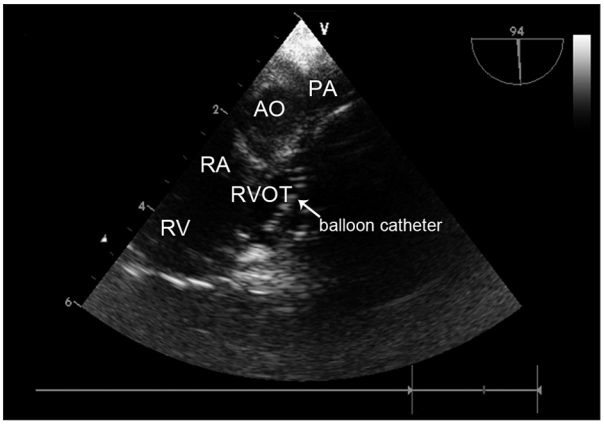

Figure 2. The balloon catheter was inserted under the guidance of a guide wire, after which sequential dilations were performed.

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.

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

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

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

Table 3. Early postoperative complications.

Item		
Pulmonary infections	8	(21.1%)
Right heart failure	3	(7.9%)
Renal failure	2	(5.3%)
Arrhythmia	3	(7.9%)

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.

The major early complications after the hybrid procedure were as follows: pulmonary infection in 8 patients, right heart failure in 3 patients, renal failure in 2 patients, and arrhythmia in 3 patients (Table 3).

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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 intractable, 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

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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 SatO2 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]. The residence time in the operating room is short; therefore, the postoperative recovery and prognosis are excellent. (4) Compared with catheter interventional treatment, the hybrid procedure can provide an open surgical visual field that facilitates the operation procedure in the infant and avoids radiation damage to the patient. According to the intraoperative exploration, the BT shunt can be placed immediately in some cases, thereby avoiding the occurrence of hypoxia. If rupture or hemorrhage of the right ventricular wall or the pulmonary artery wall occurs, it can be addressed immediately under direct vision.

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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References:

1. Petrucci O, Obrien SM, Jacobs ML et al: Risk factors for mortality and morbidity after the neonatal Blalock-Taussing shunt procedure. Ann Thorac Surg, 2011, 92: 642–51
2. Zheng JH, Gao BT, Zhu ZQ et al: Surgical results for pulmonary atresia with intact ventricular septum: A single-centre 15-year experience and medium-term follow-up. Eur J Cardiothorac Surg, 2016; 50(6): 1083–88
3. Moller JH: Operative and interventional procedures in 1039 neonates with pulmonary valvular atresia and intact ventricular septum: A multi-institutional study. Progress in Pediatric Cardiology, 2010; 29: 15–18
4. Li SJ, Zhang H, Sheng XD et al: Intraoperative hybrid cardiac surgery for neonates and young children with congenital heart disease: 5 years of experience. Ann Thorac Cardiovasc Surg, 2010; 16: 406–9
5. Leacche M, Umakanthan R, Zhao DX et al: Surgical update hybrid procedures, do they have a role? Circ Cardiovasc Interv, 2010, 3: 511–18
6. Zampi JD, Hirsch-Romano JC, Goldstein BH et al: Hybrid approach for pulmonary atresia with intact ventricular septum: Early single center resultsand comparison to the standard surgical approach. Catheter Cardiovasc Interv, 2014; 83(5): 753–61

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7. Li S, Chen W, Zhang Y et al: Hybrid therapy for pulmonary atresia with intact ventricular septum. Ann Thorac Surg, 2011; 91(5): 1467–71
8. Schmitz C, Esmailzadeh B, Herberg U et al: Hybrid procedures can reduce the risk of congenital cardiovascular surgery. Eur J Cardiothorac Surg, 2008; 34: 718–25
9. Viana FF, Fukushima S, Clarke AJB et al: Long-term results of the treatment of pulmonary atresia with intact ventricular septum. Heart, Lung and Circulation, 2011; 20: 54–55
10. Zhang H, Luo Y, Li L Hou J: Surgical treatment of pulmonary atresia with intact ventricular septum and critical pulmonary stenosis in neonates and infants. Journal of Cardiovascular and Pulmonary Diseases, 2008; 27(3): 138–43
11. Humpl T, Söderberg B, McCrindle BW et al: Percutaneous balloon valvotomy in pulmonary atresia with intact ventricular septum: Impact on patien. Circulation, 2003; 108(7): 826–32
12. Park JY, Seo DM, Shin HJ et al: Hybrid procedure for pulmonary atresia with ventricular septal defect in a low birth weight neonate. Korean J Thorac Cardiovasc Surg, 2013; 46(1): 56–59
13. Asnes JD, Fahey JT: Novel catheter positioning technique for atretic pulmonary valve perforation. Catheterization and Cardiovascular Interventions, 2008; 71: 850–52
14. Burkholder H, Balaguru D: Pulmonary atresia with intact ventricular septum: Management options and decision-making. Pediat Therapeut, 2012; 01(S5): 1–7
15. Schneider AW, Blom NA, Bruggemans EF, Hazekamp MG: More than 25 years of experience in managing pulmonary atresia with intact ventricular septum. Ann Thorac Surg, 2014; 98(5): 1680–86
16. Amin P, Levi DS, Likes M et al: Pulmonary atresia with intact ventricular septum causing severe left ventricular outflow tract obstruction. Pediatr Cardiol, 2009; 30: 851–54
17. Wu S, Liu Y, Wan F et al: Surgical treatment of the pulmonary artery atresia with the intact ventricular septum. Journal of Clinical Cardiology, 2012; 22(11): 76–78
18. Zhang H, Li SJ, Li YQ et al: Hybrid procedure for the neonatal management of pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg, 2007; 133: 1654–56
19. Kim SJ, Huh J, Song JY et al: The hybrid perventricular closure of apical muscular ventricular septal defect with Amplatzer duct occluder. Korean J Pediatr, 2013; 56(4): 176–81
20. Foker JE, Berry JM, Pyles LA: Treatment algorithm for pulmonary atresia with intact ventricular septum. Progress in Pediatric Cardiology, 2010; 29: 61–63
21. Hudorović N, Rogan SA, Lovričević I et al: The vascular hybrid room – operating room of the future. Acta Clin Croat, 2010; 49: 289–98

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