Surgical and transcatheter management alternatives in refractory pulmonary hypertension: Potts shunt

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ABSTRACT

Despite advances in the medical treatment of children with pulmonary arterial hypertension that have resulted in improved health quality and life expectancy, the progression of the disease is still the main problem for some patients. Because of this undesirable condition, the search for new treatment strategies continues for pediatric cardiologists. At this point, the Eisenmenger physiology is the main target because of the long-life expectancy and more stable hemodynamics of patients with Eisenmenger syndrome. Therefore, some invasive procedures may be used for conversion to Eisenmenger physiology with the aim of decompressing the right ventricle. *(Anatol J Cardiol 2015; 15: 843-7)*

Keywords: pulmonary hypertension, Potts shunt, atrial septostomy

Introduction

Notwithstanding the innovations and gradually increasing successful results in pulmonary arterial hypertension (PAH) diagnosis, evaluation (1, 2), and treatment, clinical deterioration and even death are inevitable because of the progressive nature of the disease (3, 4). The search for a treatment to improve the clinical state and increase the survival at the end-stage, which is a non-responder to medical PAH therapy, is ongoing.

Clinical observations in patients with Eisenmenger syndrome have created opportunities in this pursuit. Because patients with Eisenmenger syndrome have a more stable clinical status and longer survival, it gives the idea that a similar physiology may become an option in idiopathic pulmonary hypertension (IPAH) treatment as well (5-8). For this purpose, atrial septostomy has been used as an option for many years. Atrial septostomy (9), which was performed on a drug-refractory PAH patient by Rich and Lam in 1983, has been conducted on many patients over time in spite of the risk for mortality and was included in the guidelines of the World Symposium of Primary Pulmonary Hypertension in 1998 (10).

Looking for new aspects

As a result of seeking an alternative because of the procedural difficulties and mortality risk of atrial septostomy, Potts shunt (11) was introduced as a new option for suprasystemic PAH in recent years (12). Potts shunt, which is created between the left pulmonary artery and descending aorta, was used for this purpose (7, 13). Potts shunt has some advantages when compared with atrial septostomy. The shunt does not cause oxygen desaturation in the upper part of the body by continuously creating a post-cardiac right-to-left shunt. Thus, coronary and brain circulations are not affected as they are in atrial septostomy (12, 13). Additional disadvantages of atrial septostomy are the high mortality rate and repetitive surgery requirement in 10% of the patients, depending on the spontaneous closure of the defect (14). A number of case series has been reported on Potts shunt as an alternative treatment option in refractory PAH since 2004 (Table 1).

Potts shunt was introduced by Blanc et al. (13) in 2004 as a new surgical method for patients with PAH. They applied Potts shunt to 2 male patients with suprasystemic PAH who were 4 and 14 years old and had an arterial switching operation before. After the procedure, the clinical status of the patients

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Accepted Date: 03.08.2015

DOI:10.5152/AnatolJCardiol.2015.6447

Table 1. Surgical Potts shunt experiences in recent literature

	Number of patients	Age	Diagnosis	Technique of Potts	Result	Pre BNP /Post BNP (pg/mL)	Pre 6MWT/ Post 6MWT (m)	Complications	Follow up
Blanc et al. (13)	2	4 and 14 (years)	Suprasystemic PAH	Surgical	Improvement in clinical signs and NYHA class, no syncope episodes	NA	NA	None	18 and 6 months
Labombarda et al. (15)	2	3 and 6(years)	Suprasystemic IPAH	Surgical	Improvement in clinical signs and NYHA class (IV to II)	457/104 657/21	NA/539 NA/504	None	2 years
Baruteau et al. (12)	8	4 to 180 months	Suprasystemic IPAH	Surgical	Six patients discharged. Functional status markedly improves with WHO functional class I (n:4) or II (n:2)	608±109 /76±45	302±95 /456±91	Two patients died of acute pulmonary hypertensive crisis	63.7± 16.1 months
Petersen et al. (17)	1	10 (years)	End-stage PAH after late repair of VSD	Surgical	Clinical condition markedly improved	461*/10*	NA	None	13 months
Keogh et al. (18)	1	19 (years)	Residual suprasystemic PAH after completion Atrioventricular canal defect repair	Surgical modified Potts	Discharged on the postoperative 11th day. Heart failure regressed to class I	NA	328/368	None	3 years
Latus et al. (19)	1	20 (years)	Shone's complex+ Suprasystemic postcapillary PAH	Atrial septostomy +Potts shunt	Heart failure decreased from class IV to class II according to the 3 months	340/135	75/375	None	3 months
Bui et al. (20)	14 pigs (animal model)	NA	Suprasystemic PAH	Surgical unidirectional valved Potts	Mean partial pressure of carbon dioxide increased and mean peripheral oxygen decreased	NA	NA	One1 Inferior paraplegia in 24 hours, 1 died of mesenteric microembolism after 36 hours	NA
Baruteau et al. (23)	24	2.3 to 9.7 (years)	Drug refractory IPAH (23 suprasystemic and 1 infrasystemic)	19 surgical (1 unidirectional valved Potts shunt with infrasystemic PAH), 5 transcatheter	Twenty-one patients discharged. One patient died during a severe RSV infection 2 years after PDA stenting. All patients had a dramatic improvement in their functional and clinical status. One patient underwent double lung transplantation 6 years later	71.4% (>500) /0% (>500**)	260.2 ±85.1 /522.6 ±93.2	Surgical group: 3 early dead, 1 transient paraplegia at day 3, 1 medically treated chylothorax 1 significant tracheal stenosis, 1 profound upper limp arteria oxygen desaturation	3 months to 14.3 years

*N-terminal pro-brain natriuretic peptide (pmol/L**71.4 % of the patients >500 / non of the patients > 500.)

IPAH - idiopathic pulmonary arterial hypertension; NA - not available; NYHA – New York Heart Association; PAH - pulmonary arterial hypertension; PDA - patent ductus arteriosus; Pre 6MWT - preoperative six-minute walking test; Pre BNP - preoperative brain natriuretic peptide; Post 6MWT - postoperative six-minute walking test; Post BNP - postoperative brain natriuretic peptide; RSV - respiratory syncytial virus; VSD - ventricular septal defect; WHO - World Health Organization

rapidly improved with class II heart failure according to the NYHA criteria and no syncope episode in the follow-up.

In 2009, Labombarda et al. (15) performed Potts shunt to 2 patients with IPAH. They successfully applied Potts anastomosis to their patients. Two years later, the physical development of the patients was normal with class II heart failure (15). Connecting the atria in patients with severe right heart failure and significantly high right atrial pressure may threaten life because a massive right-to-left shunt may lead to insufficient pulmonary blood flow and severe hypoxemia (16). For this reason, atrial septostomy was not preferred in both patients, and Potts shunt was applied with similar success. Therefore, they suggested Potts shunt as an alternative treatment method in patients with severe PAH and right ventricular failure after receiving appropriate medical treatment.

In 2012, Baruteau et al. (12) retrospectively examined 8 patients with IPAH and reported long-term results. In their study, they applied Potts shunt to 8 children with suprasystemic IPAH and class IV heart failure according to the WHO criteria after receiving medical treatment between 2003 and 2008. Surgical Potts shunt operation was performed with a mean delay of 41.9±54.3 months after IPAH diagnosis. Two patients for whom PAH treatment was ceased after the surgery died due to acute pulmonary hypertensive crisis on the postoperative 11th and 13th days. Transient paraplegia was seen in 1 patient on the postoperative 3rd day. At the last follow-up, while 4 patients had class I heart failure, 2 had class II heart failure according to the WHO criteria. The authors suggested palliative Potts shunt as a new alternative method in patients with IPAH because it significantly prolongs the survival period and improves the functional capacity (12).

Several successful cases with Potts shunt are reported. In 2013, Petersen et al. (17) performed Potts shunt in a patient with lately repaired VSD and end-stage PAH and obtained successful results. Postoperative sildenafil, treprostinil, and bosentan treatments were continued, and the patient was discharged 2 weeks later. Thirteen months from the operation, the patient's clinical condition significantly improved and NT-proBNP level regressed from 461 pmol/L to 10 pmol/L. Atrial septostomy was not considered because it could have led to severe hypoxemia; thus Potts shunt, which is an extracardiac shunt providing successful results, was performed (17).

The first Potts shunt performed in adulthood was reported by Keogh et al. (18). An 18-year-old patient, who had been diagnosed with Down syndrome, underwent repair of a complete atrioventricular canal defect at the age of 4 months and presented with postoperative PAH and was admitted more than 10 times for syncope during the preceding 12 months. A modified Potts shunt was performed at the age of 19 months by inserting a 10-mm Dacron graft between the left pulmonary artery and the descending aorta. They mentioned that an advantage of preferring modified shunt over classic Potts shunt is that the pulmonary artery is too distant from the aorta in adult patients. In the 3-year follow-up period, the patient who was discharged on the postoperative 11th day after bosentan and aspirin treatment had no hospitalization requirement or syncope complaint. Heart failure regressed to class I (18).

Latus et al. (19) reported a case of the combined application of atrial septostomy and modified Potts shunt (13-mm graft from the left pulmonary artery to the descending aorta). They performed atrial septostomy and Potts shunt together in a 20-year-old patient with Shone's complex, severe diastolic dysfunction, and severe pulmonary hypertension, while waiting for heart–lung transplantation and obtained successful results. The patient was evaluated with right and left catheterization while receiving combined treatment with bosentan and sildenafil. Suprasystemic post-capillary pulmonary hypertension [pulmonary arterial pressure (PAP): 125/75 mm Hg, systemic arterial pressure: 105/55 mm Hg) was determined. Atrial septostomy and Potts shunt were applied to the patient through a hybrid procedure. At hospital discharge, the BNP level regressed from 340 pg/mL to 135 pg/mL, and heart failure decreased from class IV to class III according to the WHO criteria. After 3 months, the patient's clinical status improved to class II and the 6MWT distance increased from 75 m to 375 m (19).

Bui et al. (20) performed a unidirectional valved Potts anastomosis in an animal model. In this study performed in 14 pigs, PAP of the animals was brought to the suprasystemic level by glue injection, and they performed a unidirectional valved Potts anastomosis. The unidirectional valve was closing in the case of infrasystemic and isosystemic PAPs and prevented the right-to-left shunt. The first unidirectional valved Potts shunt performed in an animal model was reported with this study (20).

Transcatheter creation of Potts

Potts shunt can also be performed with the transcatheter technique. Boudjemline et al. (21) reported a case series with transcatheter Potts shunt. In the series, cardiac catheterization was performed in 28 patients with IPAH, and residual or probe patent ductus arteriosus (PDA) was detected in 4 patients. Because one of these patients had infrasystemic PAP, PDA stenting was not performed. During the procedure, vasovagal collapse occurred in 1 patient, and cardiac resuscitation was required. No complication was observed in 2 other patients. All 3 patients were discharged 1 day after the procedure, and their right ventricular functions significantly improved. After a mean follow-up of 14±9 months, all patients showed an improved functional capacity and a patent PDA stent on echocardiography (21). A detailed cardiac catheterization should be performed in patients with PAH, and transcatheter Potts shunt creation should be kept in mind as an alternative treatment method in patients with suprasystemic PAP.

Esch et al. (22) reported a case series on transcatheter Potts shunt and its short-term results. They described a technique for transcatheter Potts shunt creation by fluoroscopically-guided retrograde needle perforation of the descending aorta at the side of apposition to the left pulmonary artery. They considered the creation of transcatheter Potts shunt in 7 patients, but performed it in 4 of them. All patients were adults with severe PAH. The procedure was successful in 3 patients, but 1 patient died during the procedure because of fatal hemothorax. One survivor died on the postoperative 5th day because ventilator-associated pneumonia with multiorgan dysfunction unrelated to the shunt. The other 2 survivors were followed-up for 10 and 4 months, and no late complications were seen. Additionally, their clinical symptoms were significantly improved. The death of the patient during the procedure highlighted the risks of this procedure, and this risk should be kept in mind in patient selection.

Baruteau et al. (23) reported short- and long-term results with Potts shunt (19 surgical and 5 transcatheter) performed on 24 patients with drug-refractory PAH in a retrospective study. The age range was 2.3–9.7 years, and the median weight was 22 kg. They applied unidirectional valved Potts shunt, which was performed in an animal model before by Micheleti et al. (14), to a 6-year-old patient with infrasystemic PAP in right cardiac catheterization. Because of rapid clinical deterioration and recurring catheter infections, Potts shunt was performed although the patient had infrasystemic PAP. This is the first case of unidirectional valved Potts shunt performed in a human. When the short-term results of these patients were examined, major postoperative complications occurred in 6 patients (25%) in whom surgical shunt was performed. Twenty-one survivors were discharged, but 1 child died due to severe respiratory syncytial virus (RSV) infection 2 years after PDA stenting. During the follow-up, all survivors experienced a dramatic improvement in their functional condition. Syncope and right heart failure signs disappeared, and all children had normal growth curves. Performing lung transplantation in patients in whom surgical Potts shunt had been performed showed that lung transplantation as the last option in the treatment may still remain along with Potts anastomosis. The number of PAH treatments significantly decreased in the final check-up. In this study, in spite of complications that occurred in 25% of the patients in the early period, long-term results were significantly better. Baruteau et al. (23) suggested Potts shunt, which does not lead to cerebral and coronary oxygen desaturation, as the first treatment option instead of atrial septostomy in patients with drug-refractory PAH.

Newer challenges

Potts shunt is not the only surgical option for drug-refractory pulmonary hypertension. New ideas have also come up for the holistic evaluation of the problem and a better understanding of communication between the ventricles. Although the right and left ventricles have different functions, they have a common septum and pericardium. Fifty percent of the right ventricle functions are provided by the contractions of the left ventricle (24). The relationship between the two ventricles suggested that the working capacity of the right ventricle increases with aortic banding (25). The aim is to increase the working capacity of the right ventricle by pushing the interventricular septum, which deviated to the left by increased pulmonary pressure, to the right. Along with this hypothesis, researchers showed that such a relation between the ventricles was also based on the interactions at histological and molecular levels (4). Even though clinical studies are scarce, the study by Schranz et al. (26) regarding the possible recovery of existing left ventricular failure by pulmonary artery banding in children with dilated cardiomyopathy was promising. In this study, significant decreases in left ventricle end-diastolic diameter and plasma BNP values were shown in the 3–6 month short-term follow-up of 12 children in whom pulmonary artery banding was performed.

Conclusion

In spite of improved PAH-specific medication options and effective treatment protocols, the clinical status of patients with IPAH and residual PAH rapidly worsens and their lifetimes are too short. This resulted in cardiologists investigating a new approach in patients with suprasystemic PAH. Although it is not a new idea to transform such patients into Eisenmenger physiology for a better prognosis, having alternative surgical options in addition to atrial septostomy would provide diversity in terms of treating such cases. Potts shunt should be considered in cases that do not respond to medical PAH therapy with suprasystemic PAP. Patients should be carefully selected to prevent disappointment. It is difficult to predict which patient will benefit or which will deteriorate after atrial septostomy and Potts shunt. We can obtain accurate information indicating which invasive procedure would be a better surgical option with more randomized controlled studies. Factors such best surgical time, optimum age, hemodynamic parameters and clinical/anatomic status for Potts shunt and aortic banding procedures should be meticulously determined. Because of its high risk, Potts shunt should be reserved for patients in whom atrial septostomy or transplantation is contraindicated.

Finally, we should not forget the suggestion of Potts and his colleagues in their original report in 1946: "… In attempting this new procedure, it seemed only fair to choose those patients whose condition was such that without aid, the future was hopeless …" (11).

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