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Case Report

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Complete Transcatheter Repair of Fallot's Trilogy: A Case Report

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Abstract

Fallot's trilogy is a relatively uncommon form of cyanotic congenital heart disease characterized by severe valvular pulmonary stenosis (PS), right ventricular hypertrophy and right to left interatrial shunt via an atrial septal defect (ASD) or patent foramen ovale (PFO). Historically, patients with the combination of these lesions were routinely treated by surgical correction. In recent times, transcatheter repair has become an increasingly attractive modality of treating such patients. There is limited experience with such combined transcatheter interventions, and from the scarce data available in published literature it seems that such approach has promising safety and efficacy. Herein, we describe a case of an 18 year old cyanotic male patient who was diagnosed of having severe valvular PS with supra systemic right ventricular pressures and reversed interatrial shunt via a large secundum ASD. We successfully treated him percutaneously with serial balloon pulmonary valvuloplasty, using single balloon and Inoue balloon techniques, followed by Amplatzer device closure of the ASD. The outcome was excellent with marked reduction of trans-pulmonary valvular gradient, complete cessation of shunting across the ASD, dramatic improvement of symptoms and completes disappearance of cyanosis. There were no procedure related complications and the benefits were sustained at follow up of three years. Our case demonstrates that transcatheter repair is a feasible, safe and effective alternative to surgery in selected patients with Fallot's trilogy. With further experience, it may well become the standard of care in the management of such patients.

Keywords

Fallot's trilogy; Transcatheter repair; Valvular pulmonary stenosis; Atrial septal defect; Balloon pulmonary valvuloplasty

Introduction

Fallot's trilogy describes a combination of severe pulmonary stenosis (PS), intact ventricular septum, right ventricular hypertrophy (RVH) and right to left interatrial shunt across an atrial septal defect (ASD) or patent foramen ovale (PFO) [1]. Traditionally, surgical correction has been the standard treatment of this condition. Recently, transcatheter repair has become an attractive alternative to surgery in managing such patients. Only a few cases have been reported wherein balloon pulmonary valvuloplasty (BPV) and ASD device closure were performed in cyanotic adults with severe PS and secundum ASD [2,3].

Case Report

An 18 year old male patient presented with a history of progressive exercise intolerance and cyanosis since 3 years. Examination revealed central cyanosis, clubbing and a harsh grade IV/VI ejection systolic murmur at the pulmonary area. ECG demonstrated right axis deviation and right ventricular hypertrophy. Chest X-ray showed cardiomegaly, dilated central pulmonary arteries and oligemic lung fields. Transthoracic echocardiogram (TTE) revealed a dilated right atrium (RA), hypertrophied right ventricle (RV) and moderate tricuspid regurgitation. Pulmonary valve was thin, mobile and showed systolic doming. Peak gradient across pulmonary valve was 124 mmHg. There was a suggestion of an associated ASD. Transesophageal echocardiography (TEE) confirmed a 22 mm ostium secundum ASD with adequate rims and right to left interatrial shunt. Cardiac catheterization demonstrated systemic arterial saturation of 80%, Q_p:Q_s ratio of 0.73:1 (right to left shunt) and peak gradient of 118 mmHg across pulmonary valve. There was a 40 mmHg subvalvular gradient across the right ventricular outflow tract (RVOT) and pulmonary annulus diameter was 21 mm. Right ventricular angiography demonstrated a hypertrophied RV and RVOT, mobile pulmonary valve with doming and central jet of contrast during systole (Figure 1). BPV was performed serially with 18 mm × 4 cm Tyshak XTM (NuMed, Hopkinton, NY) balloon followed by 23 mm Inoue balloon catheter using standard techniques (Figure 2). Following valvuloplasty, peak gradient across pulmonary valve decreased to 10 mmHg and subvalvular RVOT gradient decreased to 30 mmHg. Systemic arterial saturation increased to 96% and Qp:Qs ratio increased to 2.14:1 (left to right shunt). Under general

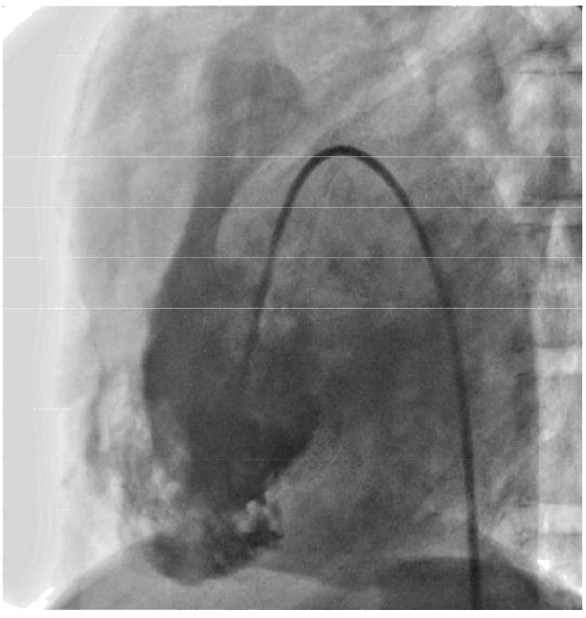

Figure 1: Right ventricular angiography demonstrating a hypertrophied right ventricle and doming stenotic pulmonary valve.

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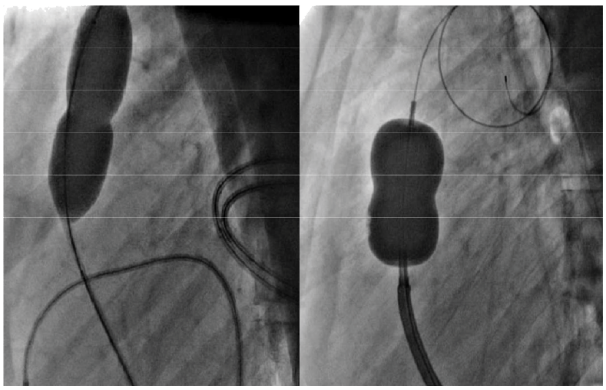

Figure 2: Serial Pulmonary valve balloon dilatation with 18 mm × 4 cm Tyshak balloon and 23 mm Inoue balloon catheter.

anaesthesia, TEE and fluoroscopic guidance, a 28mm Amplatzer atrial septal occluder (AGA Med, MN, USA) was deployed across the ASD as per standard protocol (Figure 3). Total procedure time was 100 min and fluoroscopy time was 30 min. There were no procedure related complications. TTE on the next day revealed complete occlusion of ASD, peak instantaneous gradient of 15 mmHg across pulmonary valve, and an infundibular gradient of 32 mmHg (Figure 4). Patient was discharged on aspirin 150 mg daily. Patient did well on follow up and aspirin was stopped after 6 months. He remained asymptomatic up to 3 years of follow up. An echocardiogram after 3 years revealed stable device position, no residual shunt, peak gradient of 10 mmHg across pulmonary valve and 20 mmHg across the infundibulum. There was only mild TR with estimated RVSP of 26 mmHg. RA size and RV hypertrophy had considerably regressed.

Discussion

In Fallot's trilogy, supra-systemic RV pressures and high RA pressure (secondary to severe pulmonary stenosis) cause right to left shunting across the ASD leading to cyanosis and low pulmonary blood flow [1]. The history of cyanosis may date from birth, but typically, such patients present with late onset of increasing cyanosis and disability in the second and third decades of life [4].

BPV is the standard treatment for severe pulmonary stenosis [5]. In adults with pulmonary annulus size of more than 20 mm (as in our case), the traditional single balloon technique may not be adequate to relieve PS. In such situations, a double balloon technique, or an Inoue balloon technique may be used [6,7]. We prefer Inoue balloon over the double balloon technique as it offers several advantages including: minimal risk of RVOT or MPA injury due to its short and self-positioning nature; ability to give serial incremental dilatations, thus reducing the risk of over-dilatation and pulmonary annulus injury; short inflation-deflation cycle (approximately 5 s) which limits any hemodynamic compromise resulting from complete obstruction of RV outflow during balloon inflation; ability to be introduced through the femoral vein without a sheath due to its stiffer profile; and eliminating the need of an additional venous access required to introduce an additional balloon for the double balloon technique [7]. Cyanotic patients with severe PS and right to left interatrial shunt are at increased risk of developing complications like bradycardia, hypotension, cerebral infarction, and even death during BPV [8]. Rapid catheter manipulation and serial dilations using smaller balloons initially followed by larger ones are advisable to minimize such complications [8]. We first dilated the pulmonary valve with a smaller (18 mm × 4 cm) Tyshak XTM (NuMed, Hopkinton, NY) balloon, followed by a larger (23 mm) Inoue balloon, without encountering any complications.

The decision regarding device closure of secundum ASD following BPV depends on the size of the ASD, presence of adequate rims, residual gradient across RVOT, and direction of interatrial shunt [2,9]. When the RVOT obstruction is adequately relieved, shunting across ASD is left to right, and the defect is suitable for device closure, ASD may be closed in the same sitting [9-11]. Persistent right to left shunt after adequate relief of PS may be a consequence of high residual infundibular gradient, poor RV compliance or RV dysfunction [2]. In such situations, ASD may prevent systemic congestion in the face of high right heart pressures, and thus immediate closure of ASD may be detrimental. It may thus be prudent to wait for few months and close the ASD after the shunt becomes left to right [3]. In our case, the shunt direction reversed immediately and residual gradient was insignificant; hence we closed the ASD concurrently. Transcatheter approach offers several advantages over surgery including: avoidance of sternotomy and cardio-pulmonary bypass; shorter recovery time; and lower overall complication rate [12]. The main disadvantage of transcatheter approach is its higher cost compared to surgery, which may be a limiting factor in developing countries. In our case, the benefits of this procedure were apparent immediately and were sustained up to a follow up period of three years.

Conclusion

There is limited experience with the dual transcatheter interventional approach of concurrent balloon pulmonary

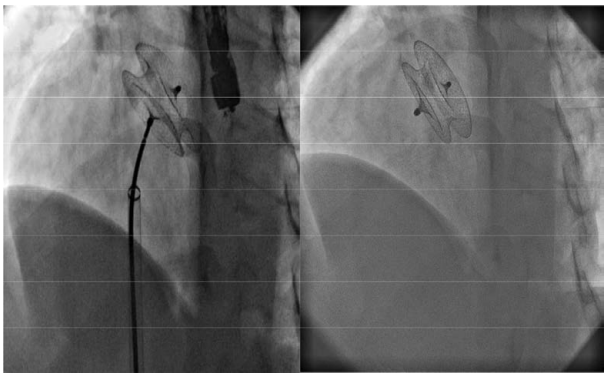

Figure 3: 28 mm Amplatzer atrial septal occluder deployed across the atrial septal defect.

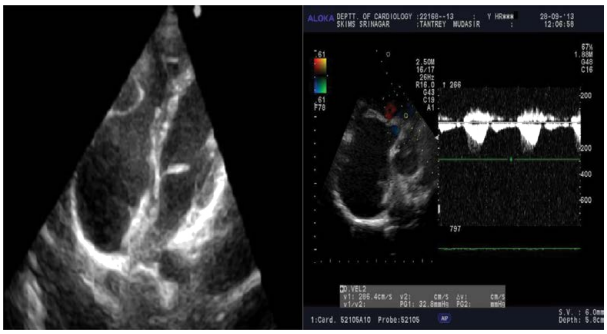

Figure 4: Echocardiographic images revealing stable device position and a residual gradient of 32 mmHg across right ventricular outflow tract.

valvuloplasty and ASD device closure in cyanotic adults with Fallot's trilogy. Our case demonstrates the feasibility, safety and efficacy of this minimally invasive approach with excellent immediate and long term results. With growing expertize and further experience from larger studies, it may well become the standard of care in the management of such patients.

References

1. Joly F, Carlotti J, Sicot JR, Piton A (1950) Congenital heart disease. II. Fallot's trilogies. Arch Mal Coeur Vaiss 43: 687-704.
2. Behjati-Ardakani M, Sadr-Bafghi SM, Andishmand A (2012) Combined transcatheter closure of atrial septal defect and pulmonary valve balloon dilatation. Acta Medica Iranica 50: 710-712.
3. Huang Y, Li Y, Huang T (2000) Transcatheter therapy of Fallot's trilogy. Zhonghua Zhongliu Zazhi 28: 458-459.
4. Abrahams DG, Wood P (1951) Pulmonary stenosis with normal aortic root. Br Heart J 13: 519-548.
5. Feltes TF, Bacha E, Beekman RH, Cheatham JP, Feinstein JA, et al. (2011) Indications for cardiac catheterization and intervention in paediatric cardiac disease a scientific statement from the American Heart Association. Circulation 123: 2607-2652.
6. Al Kasab S, Ribeiro PA, Al Zaibag M, Halim M, Moh'D AH, et al. (1988) Percutaneous double balloon pulmonary valvotomy in adults: One-to two-year follow-up. Am J Cardiol 62: 822-824.
7. Lau KW, Hung JS, Wu JJ, Chern MS, Yeh KH, et al. (1993) Pulmonary valvuloplasty in adults using the Inoue balloon catheter. Cathet Cardiovasc Diagn 29: 99-104.
8. Mahmood ZI, Farhan HA (2006) Pulmonary balloon valvuloplasty in high risk patients. Iraq Postgrad Med J 5: 15-17.
9. Shrivastava S, Radhakrishnan S (2001) Concurrent percutaneous atrial septal defect closure and pulmonary valvuloplasty. Indian Heart J 53: 773-735.
10. Yip WC, Chan KY, Godman MJ (1998) Simultaneous transcatheter valvuloplasty and amplatzer Septal occlusion for pulmonary valvar stenosis and secundum atrial septal defect. Ann Acad Med Singapore 27: 578-581.
11. Medina A, de Lezo JS, Delgado A, Caballero E, Segura J, et al. (2000) Combined percutaneous atrial septal defect occlusion and pulmonary balloon valvuloplasty in adult patients. Tex heart Inst J 27: 216-217.
12. Suchon E, Peiculewicz M, Tracz W, Przewlocki T, Sadowski J, et al. (2009) Transcatheter closure as an alternative and equivalent method to the surgical treatment of atrial septal defect in adults: Comparison of early and late results. Med Sci Monit 15: 612-617.

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