

Simultaneous Bentall Procedure and Partial Atrioventricular Septal Defect Repair in a 64-Year-Old Patient with Idiopathic Thrombocytopenic Purpura: A Case Report and Literature Review

INTRODUCTION

Partial atrioventricular septal defect (PAVSD) is a congenital heart condition characterized by specific anatomical abnormalities.¹ It involves the presence of an atrial septal defect (ASD) at the ostium primum, a single annulus for the atrioventricular (AV) valves with 2 distinct orifices, and a cleft in the anterior leaflet of the left AV valve.² Approximately, 10%-15% of individuals with PAVSD experience regurgitation in the AV valves, particularly the left AV valve. There is often a left-to-right shunt at the atrial level, leading to increased volume load on the right atrium and right ventricle. Pulmonary artery pressure tends to be within normal limits in childhood, but long-lasting volume overload might lead to pulmonary hypertension in the long run. Partial atrioventricular septal defect often presents without symptoms in the infantile and pediatric populations, frequently eluding diagnosis until later in life. In the present case, an acute bronchitis episode unmasked underlying dyspnea, peripheral edema, and pulmonary hypertension, which were subsequently attributed to a previously unrecognized PAVSD.

Bicuspid aortic valve (BAV) is a congenital cardiovascular malformation characterized by 2 leaflets or cusps in the aortic valve instead of the usual 3. It is the most common heart defect with an estimated prevalence of 1%-2% in the general population.³ This condition can lead to complications such as aortic stenosis, aortic regurgitation, and infective endocarditis. Significantly, BAV is often associated with coarctation of the aorta and ascending aortic aneurysm, due to inherent weakness in the aortic wall. The reported frequency of ascending aortic aneurysm in patients with BAV varies widely but has been noted to be as high as 50% in some studies. These associated pathologies often necessitate surgical intervention, particularly when significant aortic dilation or aneurysm formation is present.

This report discusses the Bentall procedure, hemiarch replacement, and PAVSD repair as viable treatment options for an elderly patient with BAV, aortic insufficiency, ascending aortic aneurysm, PAVSD, and idiopathic thrombocytopenic purpura (ITP) who had previously been asymptomatic but recently experienced a bronchitis episode.

CASE REPORT

We describe a 64-year-old male patient, initially presenting with symptoms indicative of bronchitis, who subsequently developed clinical signs of decompensated heart failure including lower extremity edema and tachypnea during evaluation at a local hospital. Following preliminary evaluations, the patient was referred to our center for further diagnostic and therapeutic management. Computed tomography (CT) imaging demonstrated a 56 mm aneurysm located at the ascending aorta root, with additional dilatation measuring 42 mm proximal to the aortic arch (Figure 1, Figure 2A). Transthoracic echocardiography revealed a significant

CASE REPORT

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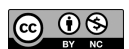
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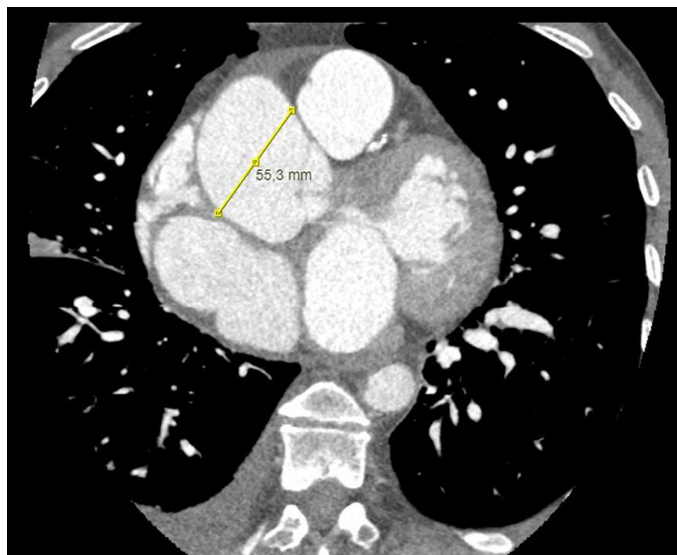


Figure 1. Preoperative contrast-enhanced computed tomography images demonstrating aortic dilation, particularly prominent at the root level.

ostium primum ASD and severe insufficiency involving the mitral, aortic, and tricuspid valves. Additionally, a BAV was detected. Cardiac catheterization revealed a pulmonary-to-systemic flow ratio of 4.4. Concurrent coronary angiography demonstrated patent coronary arteries with no abnormalities. The patient's medical history was unremarkable except for a history of smoking and the presence of an emphysematous lung as revealed by CT imaging. In the preoperative echocardiography, the systolic pulmonary artery pressure was recorded at 68 mm Hg while the mean pulmonary artery pressure was measured at 43 mm Hg. Upon preoperative laboratory analysis, thrombocytopenia was observed, prompting a consultation with the hematology department. The hematologist established a diagnosis of ITP. Thrombocyte replacement therapy was administered prior to and following the operation to address the low platelet count.

We opted for surgical intervention and proceeded with the operation through a median sternotomy approach. The

patient was placed on cardiopulmonary bypass (CPB) with aorto-bicaval cannulation. The surgery was performed under CPB and aortic cross clamp. The aortic valve was calcified with fused right and non-coronary cusps (Figure 3) making valve-sparing repair impossible. As a result, a Bentall procedure was performed, involving mobilization of the right and left coronary buttons and excision of both the aortic valve and ascending aorta. Hemiarach replacement was carried out under hypothermic circulatory arrest. The composite graft, comprising a 23 mm mechanical aortic valve and a 26 mm branched Dacron conduit (Figure 2B), was securely attached to the aortic root using U sutures. The coronary buttons were reimplanted into the conduit. Subsequently, attention was directed towards the repair of the PAVSD. Interrupted prolene sutures were used to close the apposition zone of the left AV valve. Using a prolene suture and a glutaraldehyde-fixed pericardial patch, the ASD was closed. Based on the preoperative echocardiography findings, a 4 mm fenestration was created on the patch due to elevated preoperative pulmonary artery pressure. The right AV valve mural leaflet underwent plication because of chordae elongation-induced prolapse. Following the removal of the aortic cross clamp, the heart resumed spontaneous beating in sinus rhythm. After weaning from CPB, intraoperative transesophageal echocardiography revealed mild AV valve insufficiency.

To minimize the amount of bleeding in the patient with ITP, sternal closure was delayed; the open chest was packed with gauze sponges and covered by a Steri-drape dressing with a vacuum assistance for compression. After a reexploration for bleeding the sternotomy was successfully closed on the first postoperative day. Extubation took place on the same day. The patient experienced atrial fibrillation on the second day, which was successfully restored to sinus rhythm through cardioversion in the intensive care unit. The patient was transferred to the ward on the fourth day and discharged on the tenth postoperative day. Subsequent echocardiography follow-ups revealed no significant anomalies, except for mild regurgitation in the right and left AV valves. The patient has maintained a satisfactory clinical status in the 6 months following surgical intervention.

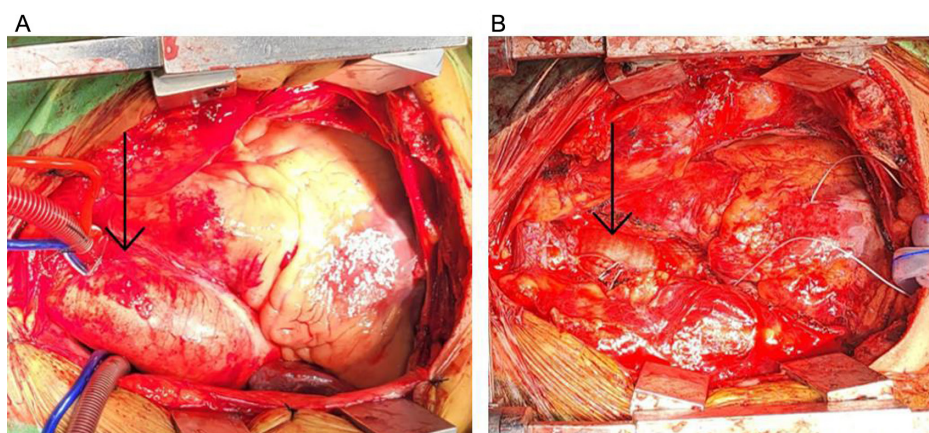


Figure 2. Paired intraoperative images showcasing the preoperative ascending aortic aneurysm (A) and the postoperative image of the Dacron graft in place of ascending aortic aneurysm (B).

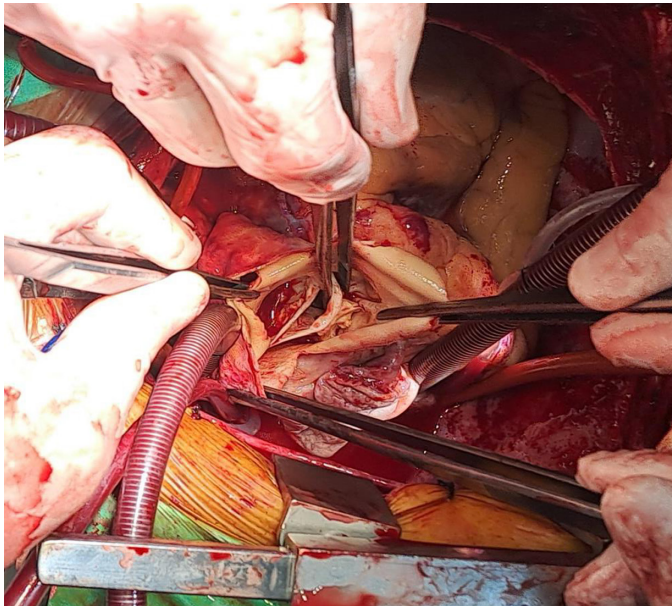


Figure 3. Detailed view of the extensively calcified bicuspid aortic valve, illustrating fused right and non-coronary cusps.

DISCUSSION

We reviewed the literature on adult PAVSD repair and found that there have been limited reports on this specific condition. While there are reports on the Bentall procedure and valve repair, to the best of our knowledge, our case represents the first instance of a Bentall procedure combined with PAVSD repair in the literature. In our literature review, we observed that in 499 cases of adult PAVSD repair with mean age of 37.3; the presence of a BAV was documented only in 3 patients (Table 1). However, our case presented with a prominently calcified BAV and an aortic aneurysm measuring 56 mm at the root level. This finding is significant as it adds to the complexity of the surgical intervention and highlights the uniqueness of our case. Additionally, it underscores the potential for ongoing complications related to BAV, such as aortic aneurysm or valve dysfunction, even when the primary condition (PAVSD) is addressed early. Here are the results of our literature review:

- Hynes et al⁴ reported multiple accompanying lesions in their cohort of 52 adult PAVSD patients, including

cleft anterior mitral leaflet, tricuspid leaflet abnormality, secundum ASD, persistent left superior vena cava, left atrial orifice to coronary sinus, double orifice mitral valve, and prolapsed aortic cusp with severe aortic insufficiency. However, none of the patients had a BAV. The study reported 3 fatalities during the operative period and 2 later deaths, with no cases requiring reoperation.⁴

- Barnett et al⁵ studied a smaller group of 8 patients with adult PAVSD, observing the presence of mitral regurgitation, left ventricular hypertrophy, and accompanying lesions, but no BAV. Two mortalities occurred at a later stage, and 2 cases required reoperation.
- Bergin et al⁶ reported on 31 adult PAVSD patients with various accompanying lesions, including secondary ASD, membranous ventricular septal aneurysm, patent foramen ovale, tricuspid valve abnormalities, subaortic stenosis, persistent left superior vena cava, coronary sinus ASD, and double orifice mitral valve. Only 1 patient had a BAV without an aortic aneurysm. The series included 2 early deaths, nine late deaths, and reoperation in 5 cases.
- Gatzoulis et al⁷ described a series of 50 adult PAVSD patients, with additional diagnoses including Down syndrome, secondary ASD, unroofed coronary sinus, small ventricular septal defect, right AV valve abnormalities, subaortic stenosis, pulmonary valve stenosis, and coronary artery disease. None of the patients had a BAV. Eight late deaths and 2 cases requiring reoperation were recorded in this series.
- In the report of Zhou et al⁸ there are 133 adult patients with PAVSD. Apart from PAVSD, conduction block, atrial fibrillation, left AV valve regurgitation are the accompanying lesions in this series. It is worth mentioning that none of the patients exhibited a BAV. One early death, 2 late deaths were reported, and no reoperation were required.⁸
- According to Song et al's⁹ report, a total of 46 adult individuals were included in the study, all of whom were diagnosed with PAVSD. The researchers noted associated conditions such as mitral regurgitation, tricuspid regurgitation, atrial fibrillation, AV block type 3, left bundle branch block. Nonetheless, no BAV was reported. One early death and 4 cases requiring reoperation were noted.

Table 1. Summary of Studies Reporting Adult Partial Atrioventricular Septal Defect Repairs, Detailing the Number of Bicuspid Aortic Valves, Mean Age, Mortality Rates, and Reoperation Frequencies

Reports	Number of Patients	Number of Patients with Bicuspid Aortic Valve	Mean Age	Mortality		Reoperation
				Early	Late	
Hynes et al (1982) [4]	52	0	37	3	2	0
Barnett et al (1988) [5]	8	0	48	0	2	2
Bergin et al (1995) [6]	31	1	51	2	9	5
Gatzoulis et al (1999) [7]	50	0	36.6	0	8	2
Zhou et al (2017) [8]	133	0	38	1	2	0
Song et al (2019) [9]	46	0	37.6	1	0	4
Patlolla et al (2020) [10]	179	2	34.1	4	61	34
Total	499	3	37.3	11	84	47

- Patlolla et al¹⁰ reported 179 adult patients with PAVSD. Numerous conditions such as left AV valve and right AV valve regurgitation, atrial fibrillation and flutter, supra ventricular tachycardia, ventricular ectopy, ostium secundum ASD, VSD, left ventricular outflow tract obstruction (LVOTO), right ventricular outflow tract obstruction (RVOTO), bundle branch block (BBB), ventricular septal aneurysm, double orifice left AV valve were observed as accompanying lesions. Furthermore, 2 patients had BAV with aortic valve stenosis. There are 4 early and 61 late deaths, 34 patients requiring reoperation.

For repairing aortic root, several procedures can be used including prosthetic replacement of valve such as Bentall and Cabrol procedures; valve sparing aortic root repair (V-SARR) such as valve reimplantation (David) and valve remodeling (Yacoub); and placement of biological graft such as homograft, xenograft, and autograft.^{3,11} Over the past 2 decades, V-SARR is more preferred than composite valve graft replacement.¹² The primary reasons for this preference are that V-SARR obviates the need for lifelong anticoagulant treatment, reduces the risk of postoperative complications, and minimizes the need for reoperation.¹⁰ Although controversial, valve-sparing options are not recommended in patients with BAV since these patients have a high likelihood of requiring another surgery on the aortic valve in the follow-up. In our case, we preferred the Bentall procedure instead of a valve sparing approach because the highly calcified BAV was not amenable to repair.

In addition to the Bentall procedure, we performed repair procedures on the right and left AV valves, as well as addressing the presence of PAVSD in our case. In the presented case, a simple cleft closure technique was employed to repair the left AV valve, while the anterior leaflet of the right AV valve was pliated due to chordal elongation. The ostium primum ASD was closed using an autologous pericardial patch. It is worth noting that alternative techniques have been described in the literature for the repair of AV valves. These include suture annuloplasty, ring annuloplasty, direct suture, and valve replacement for the left AV valve repair.⁴⁻¹⁰ Suture annuloplasty and ring annuloplasty are commonly employed for right AV valve repair.⁴⁻¹⁰ For ASD closure, both synthetic patch and direct suture methods are utilized.⁴⁻¹⁰ Ring annuloplasty was not preferred in our case because the diameters of the AV valves were not wide, insufficiency disappeared after repair, and we did not want to prolong the already long CPB in such a complex surgery.

We took extra precautions at the end of the operation because of the diagnosis of ITP. After a meticulous bleeding control, the chest was left open, it was packed with gauze sponges and covered by a Steri-drape dressing. A vacuum assistance was added to the gauze sponges for compression. This technique minimized the bleeding in our patient.

CONCLUSIONS

In conclusion, our case report presents a unique coexistence of a PAVSD and a bicuspid aorta, aortic insufficiency, aneurysm of the ascending aorta in an adult patient with ITP. The surgery describes the combination of 2 complex surgical techniques, PAVSD repair and Bentall procedure, hemiarch replacement in the same adult patient with ITP.

Informed Consent: Written informed consent was obtained from the patient and patient's family for publication of this case report and accompanying images.

Declaration of Interests: The authors have no conflict of interest to declare.

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