Balloon valvotomy for rheumatic valvar stenosis in a patient with complex congenital heart disease

Sir,

The occurrence of rheumatic heart disease in patients with complex congenital heart disease is documented, but rare.[1,2] The present report draws attention to rheumatic affection in the setting of situs inversus and congenitally corrected transposition of great vessels. Balloon valvotomy was technically challenging in view of the complex anatomy.

A 17-year-old male presented with dyspnea on exertion since 2 years, worsening to NYHA class III since 3 months. He had a history of rheumatic fever at the age of 10 years. On examination, he had a regular pulse of 70 per minute, blood pressure of 110/70 mmHg in the right upper limb, and a normal jugular venous pulse. Cardiovascular examination revealed a loud first heart sound, a loud second heart sound, a long mid-diastolic murmur at the right lower sternal border, and an early diastolic murmur at the left second intercostal space. Gastric tympany was appreciated in the right hypochondrium. Investigations were consistent with situs inversus, mesocardia, congenitally corrected transposition of great vessels, and associated rheumatic systemic atrioventricular valve stenosis [Figure 1, Video 1]. In view of symptomatic severe systemic atrioventricular valve stenosis with pulmonary hypertension, balloon valvotomy was planned [Figure 2, Video 2]. The systemic and pulmonary venous drainage were normal.

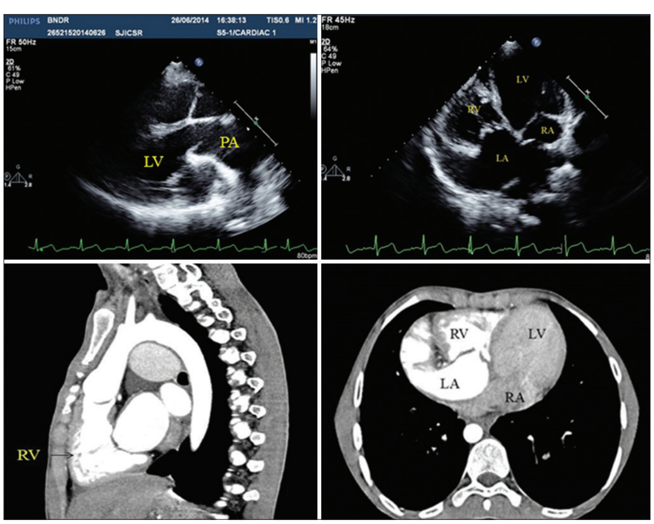

Figure 1: Pre-procedure imaging depicting the cardiac anatomy: Two-dimensional echocardiography and cardiac computed tomography highlighting atrioventricular and ventriculo-arterial discordance. In addition, thickening of systemic atrioventricular valve is appreciated. (RA: Right atrium, RV: Right ventricle, LA: Left atrium, LV: Left ventricle, PA: Pulmonary artery)

Left femoral vein and left femoral artery access were obtained. Cardiac catheterization was performed and showed severe pulmonary arterial hypertension [Figure 3, Video 3]. Subsequently, fluoroscopic imaging was inverted left to right. Pulmonary angiography was performed, and the interatrial septum was delineated in levophase [Figure 4, Video 4]. The septal descent was performed in the antero-posterior view, with the needle kept in the 7 o'clock position. Septal puncture was done in steep RAO view [Figure 5, Video 5], under transthoracic echocardiographic guidance. Transesophageal echocardiography was kept as a standby. The interatrial septum was dilated with a septal dilator and a 26-mm Accura balloon was tracked over the coiled guide wire to the left atrium. It was difficult to maneuver the Accura balloon across the systemic atrioventricular valve, and repeated attempts failed [Figure 6]. From the left femoral vein access, a 5F Swan-Ganz catheter was used to engage the stenotic valve through Mullin's sheath. A 0.025″ J-tip Terumo wire was introduced into the systemic ventricle and aorta [Figure 7, Video 6] and snared from the left femoral artery. A venoarterial loop was established. A 22 × 40 mm Tyshak® (NuMED Inc.; Hopkinton, New York, USA) peripheral balloon was introduced from the femoral artery through an 8F sheath, across the stenotic valve. Successful dilatation was done under echocardiographic and fluoroscopic guidance [Figure 8, Video 7]. Post-procedure echocardiography showed a significant reduction in gradient across the systemic atrioventricular valve, an increase in valve area, and split of one of the commissures [Figure 9, Video 8]. The hemodynamic data is summarized in Table 1. There were no complications and in-hospital stay was uneventful. Patient is asymptomatic at 1-year follow-up.

Rheumatic heart disease continues to be a major cause of cardiovascular morbidity and mortality. The recent

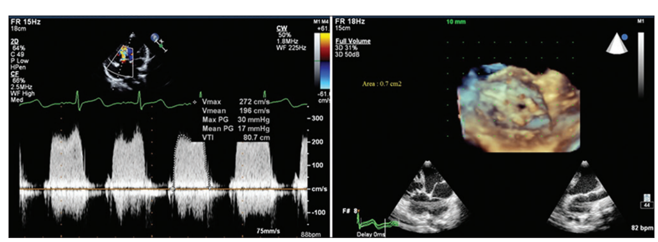

Figure 2: Echocardiography highlighting the severity of stenosis: Continuous wave Doppler across the systemic atrioventricular valve highlighting severe stenosis (peak gradient of 30 mmHg and mean gradient of 17mmHg). Valve area is 0.7 cm² on three-dimensional echocardiography, with fused commissures

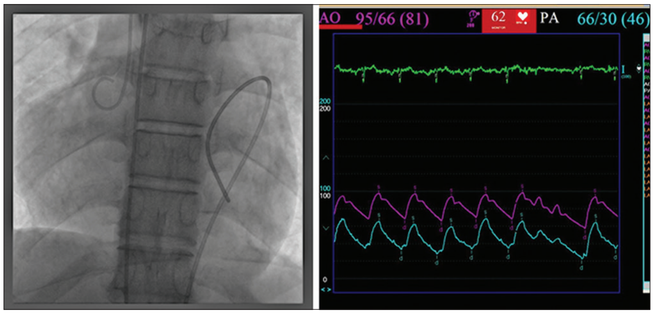

Figure 3: Pre-procedure cardiac catheterization: Catheter course: 6F Pigtail: LFA > LIA > AA > DTA > AORTIC ARCH > AORTIC ROOT. NIH: LFV > LIV > Left-sided IVC > RA > LV > PA. At the level of diaphragm, aorta is on the right and inferior venacava on the left, suggestive of situs inversus. There is evidence of severe pulmonary arterial hypertension > 66/30 (46) mmHg. (LFA: Left femoral artery, LIA: Left iliac artery {external and common iliac}, AA: Abdominal aorta, DTA: Descending thoracic aorta, NIH: National Institute of Health, LFV: Left femoral vein, LIV: Left iliac vein {external and common iliac}, IVC: Inferior venacava, RA: Right atrium, LV: Left ventricle, PA: Pulmonary artery)

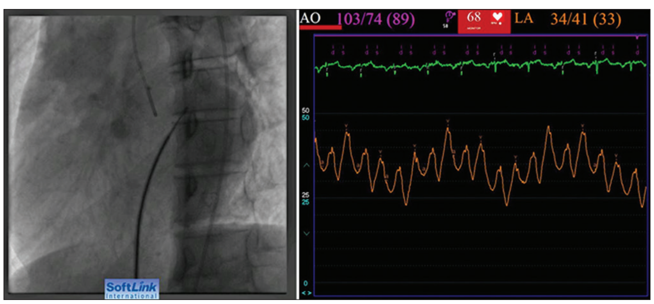

Figure 5: Transseptal puncture: Septal puncture in steep RAO (pseudo-LAO) view. The mean left atrial pressure is 33 mmHg

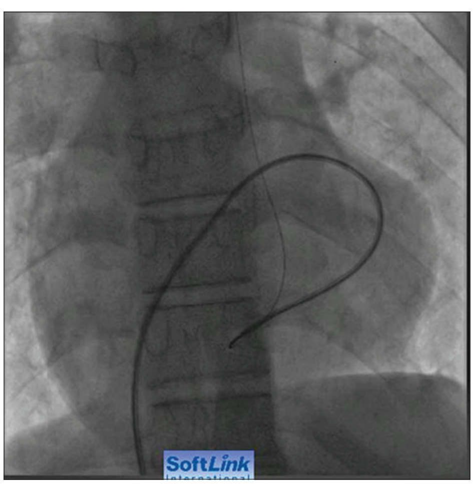

Figure 7: Systemic atrioventricular valve crossing with Terumo wire: Through Mullin's sheath, a 5F Swan-Ganz catheter is used to engage the stenotic valve. A 0.025" J-tip Terumo wire is introduced into the systemic ventricle and ascending aorta

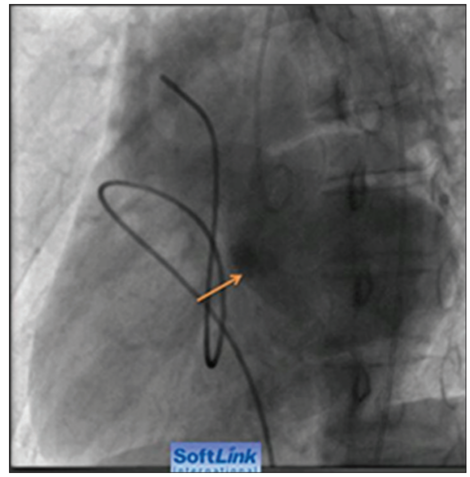

Figure 4: Pulmonary angiogram: Levophase pulmonary artery angiogram delineating the interatrial septum (arrowhead)

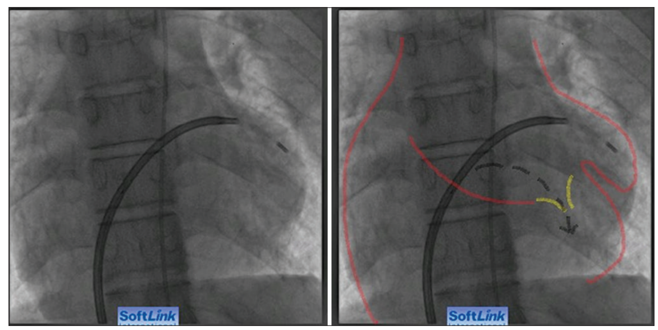

Figure 6: Attempted systemic atrioventricular valve crossing with Accura balloon: The Accura balloon could not be negotiated across the stenotic valve, despite repeated attempts. (Dotted line illustrates the intended direction of balloon passage. The balloon is entering the left atrial appendage)

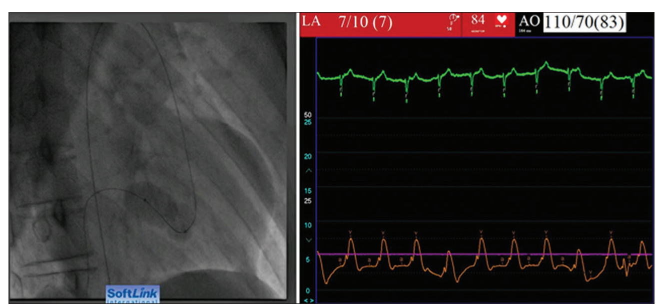

Figure 8: Systemic atrioventricular valve dilatation with peripheral balloon: A 22 × 40 mm Tyshak® balloon is introduced from the femoral artery across the stenotic valve and inflated under echocardiographic and fluoroscopic guidance. The mean left atrial pressure is decreased to 7 mmHg

prevalence in India is estimated to be 5-6.7 per 10,000.[3] The occurrence of rheumatic mitral stenosis in congenital heart disease is uncommon. The pathogenesis of the rheumatic fever is because of the molecular mimicry, and the most common valve to be affected depends on the hemodynamic stress. The most important long-term sequelae are mitral valve disease, as it is subject to the maximal hemodynamic stress (having to withstand a pressure of 120 mmHg during systole). In our report, we reinforce Paul Wood's hypothesis highlighting rheumatic affection in the valve exposed to maximum hemodynamic burden. In congenitally corrected transposition of great

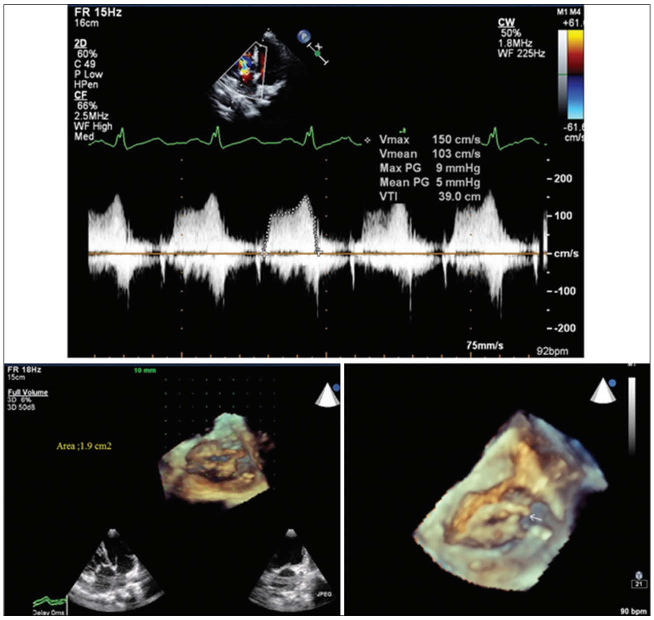

Figure 9: Post-procedure echocardiography: Two-dimensional echocardiography showing a gradient of 9/5 mmHg across the systemic atrioventricular valve. Three-dimensional echocardiography depicting a valve area of 1.9 cm^2 with split of one of the commissures (arrowhead)

Table 1: Echocardiographic and catheterization data of balloon valvotomy

	Pre-procedure	Post-procedure
Valve orifice area (cm^2)	0.7	1.9
Gradient (mmHg)	30/17	9/5
Mitral regurgitation	Nil	Nil
Left atrial pressure (mmHg)	34/41 (33)	7/10 (7)
Pulmonary artery pressure (mmHg)	66/30 (46)	35/21 (26)
Aortic pressure (mmHg)	95/66 (81)	110/70 (83)

arteries, the valve exposed to maximum hemodynamic burden is a tricuspid valve, which is the systemic atrioventricular valve.

There are only a few reports in literature on balloon mitral valvotomy in the setting of situs inversus / dextrocardia and rheumatic mitral stenosis.[4-6] However, our case is unique and challenging in view of associated congenitally corrected transposition of great arteries. Balloon valvotomy was performed with modifications of the standard Inoue technique, previously described in similar settings.[7] The important modifications include the following:

a. In situs inversus, a left femoral vein access is preferred to reduce the septal puncture needle angulation at the confluence of iliac veins to the left-sided inferior vena cava.
b. The fluoroscopic images are to be acquired in inverted position to simulate normal anatomy. This facilitates manipulation of the septal puncture needle and maneuvring of the balloon in the left atrium.
c. During septal puncture needle descent, the external indicator of the needle should point at 7 o' clock position.
d. Delineation of the interatrial septum with levophase pulmonary angiography guides transseptal puncture in complex anatomy. In addition, it is appropriate to utilize transesophageal echocardiography and/or intracardiac echocardiography in such a setting. Cardiac computed tomography can be useful to profile the interatrial septum and define the cardiac anatomy, as illustrated [Figure 1].

In our report, interatrial septum was profiled with transthoracic echocardiography, cardiac computed tomography, and levophase pulmonary angiogram. Septal puncture was done under transthoracic echocardiographic guidance, with transesophageal echocardiography as a backup. The Accura balloon could not be maneuvred across the systemic atrioventricular valve, despite repeated attempts. Hence, it was decided to establish a venoarterial loop as described. Subsequently, the valve was successfully dilated with a peripheral balloon, introduced from the femoral artery. The balloon could be used either transseptally or via the femoral artery. Tyshak® balloon is a low-profile balloon, compatible with an 8F introducer sheath. The position of the balloon should be confirmed on echocardiography, before inflation. This approach is a reasonable alternative in cases of complex cardiac anatomy and malpositions, as highlighted in this report.

To conclude, balloon valvotomy is technically feasible, even in complex anatomy. In our case, balloon valvotomy was unique and challenging, in view of associated situs inversus and congenitally corrected transposition of great arteries. In situs inversus, a left femoral vein access is preferred for balloon valvotomy of systemic atrioventricular valve. Multimodality imaging for interatrial septum delineation is helpful, as fluoroscopic landmarks of septal puncture are unreliable in complicated anatomy. In cases of difficulty in negotiating a stenotic atrioventricular valve, it is preferable to establish a venoarterial loop. Peripheral balloon can be utilized to successfully dilate stenotic atrioventricular valves.

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Conflicts of interest

There are no conflicts of interest.

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