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# Double outlet right atrium with three atrioventricular valves

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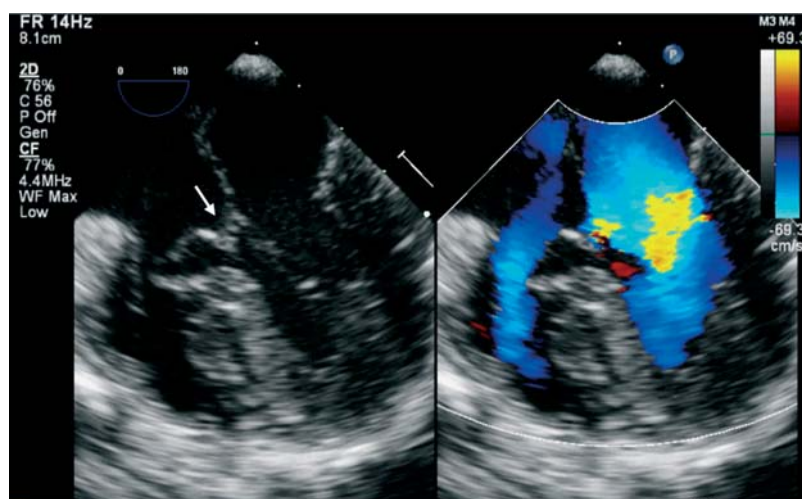
## ABSTRACT

The atrioventricular (AV) valves are complex anatomical structures which perform sophisticated functions.<sup>1</sup> A wide spectrum of malformations of these valves can occur in patients with AV septal defects. We here describe the anatomic and functional abnormalities of a rare form of the disease, where two valves connected the right atrium to both the right and left ventricles, in addition to a third valve that connected the left atrium to the left ventricle, with no evidence of regurgitation or cyanosis in spite of the relatively large communication between the right atrium and the left ventricle. In addition, the patient had severe subaortic stenosis. The pathophysiology, hemodynamics and method of repair of the condition are discussed.

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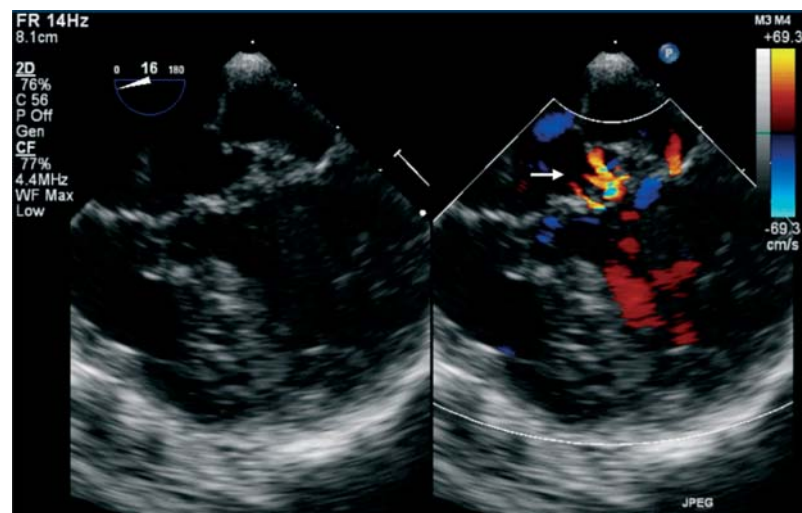


**Figure 1.** Pre-operative trans-oesophageal echocardiography, showing the diastolic flow across the atrioventricular valves, with almost no diastolic flow across the smaller right atrioventricular valve orifice (arrow).

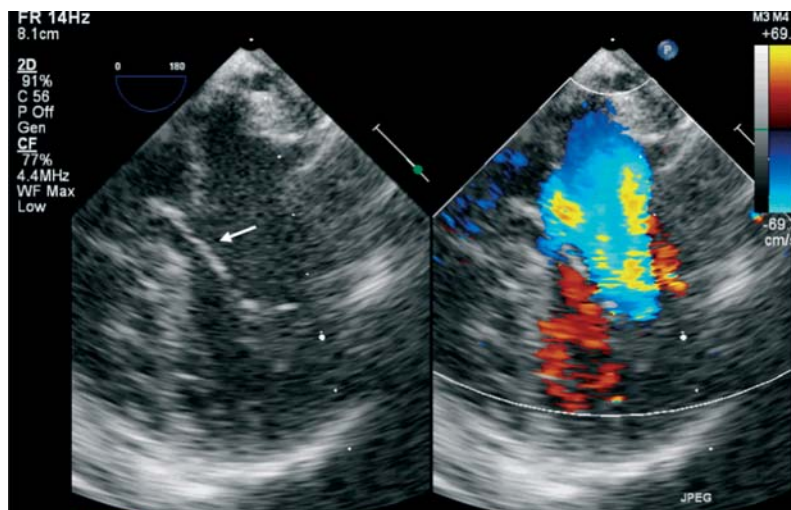
#### PATIENT AND METHODS

A 12-year-old female patient, presented with dyspnea on moderate effort. She had no history of cyanosis. Her resting oxygen saturation was 98%. Echocardiography revealed the diagnosis of atrioventricular septal defect (AVSD) with two adequate ventricles, closed atrial component and almost closed ventricular component, with what seemed to be aneurysmal tissue that had a tiny leak (Figures 1, 2). A cord could be seen attached to that aneurysmal tissue (Figure 3). There was mild regurgitation through “cleft” left atrioventricular (AV) valve. The patient also had a subaortic membrane with a peak gradient of 70 mmHg across the left ventricular outflow tract, mild aortic regurgitation and left ventricular hypertrophy. There was no right or left ventricular dilatation.

The patient was referred to surgery for resection of the subaortic membrane and repair of the left AV valve. In surgery, the right atrium was opened for trans-septal access of the left AV valve. On opening the right atrium, two AV valves were found: a bigger AV valve opening to the right ventricle, directly attached to the muscular interventricular septum with no ventricular septal defect or aneurysmal tissue; and another small orifice opening to the left ventricle (Figure 4). There was no atrial septal defect.



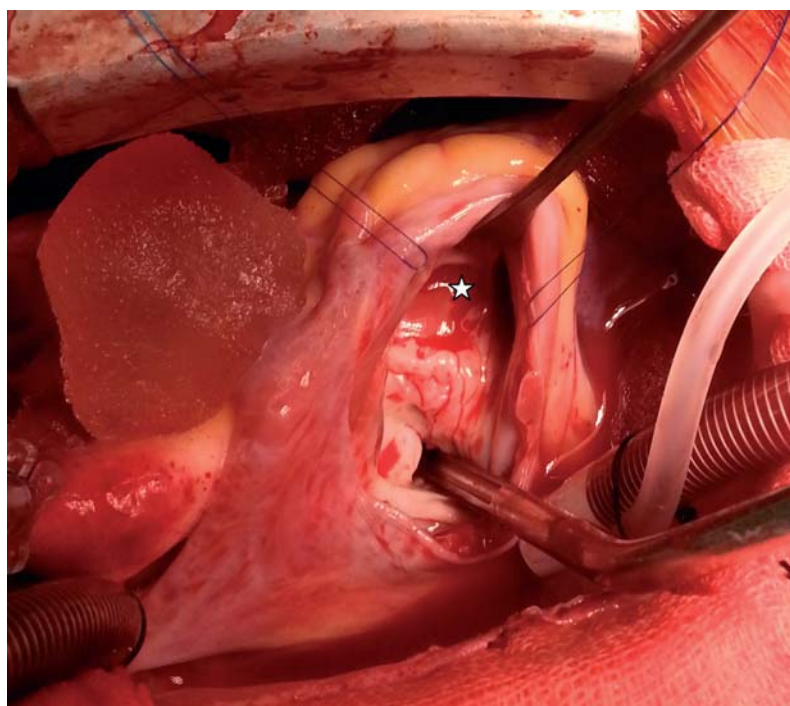
**Figure 2.** Pre-operative trans-oesophageal echocardiography, showing the systolic “leak” through the accessory orifice (arrow).



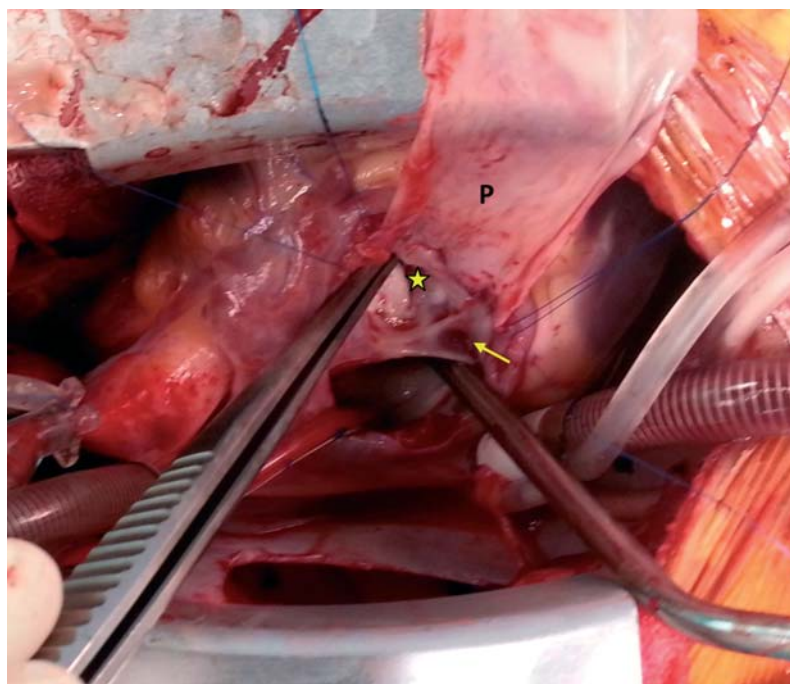
**Figure 3.** Pre-operative trans-oesophageal echocardiography, showing a chord attached to the accessory orifice (arrow)

So the atrial septum was incised at the fossa ovalis, through which the left AV valve was seen opening to the left ventricle with “a cleft” (zone of apposition between the bridging leaflets).

The fossa ovalis was excised, creating good communication and the “cleft” in the left AV valve was closed. Then a fresh autologous pericardial patch was used to separate the two right AV valve orifices (Figure 5). The patch was then used to separate the right and left atria, leaving the small orifice connected to the left atrium (Figures 6, 7). The coronary sinus was kept in the left atrium to avoid making a “waist” between the left AV valve and the small orifice. The subaortic membrane was resected with a limited myectomy.



**Figure 4.** Intra-operative surgeon view, through the opened right atrium, showing the normal sized AV valve (star) opening to the right ventricle, in addition to a smaller orifice through which the suction tip is passing to the left ventricle.

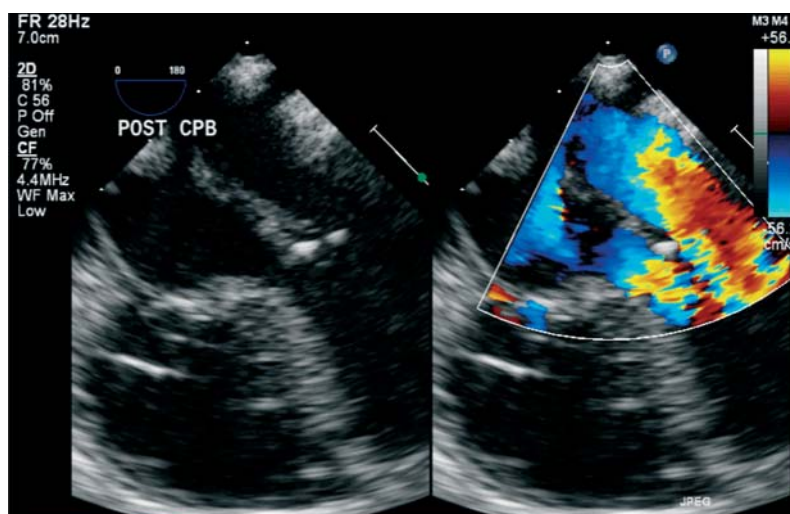


**Figure 5.** Intra-operative surgeon view, through the opened right atrium, after excision of the fossa ovalis, showing the small right atrioventricular (AV) valve orifice (star), separated from the bigger right AV valve orifice (not shown) by the pericardial patch (P). The suction tip is passing through the created atrial septal defect to the left AV valve orifice. The arrow points to the coronary sinus ostium.

The patient had a smooth post-operative course and was discharged from hospital after 5 days. The post-operative echo showed trivial right and left AV valve regurgitation. The mean diastolic gradient across the left AV valve was 4 mmHg. There was no significant gradient across the left ventricular outflow tract. The patient remained symptom-free and with the same echo findings in the routine follow-up after 2 months.



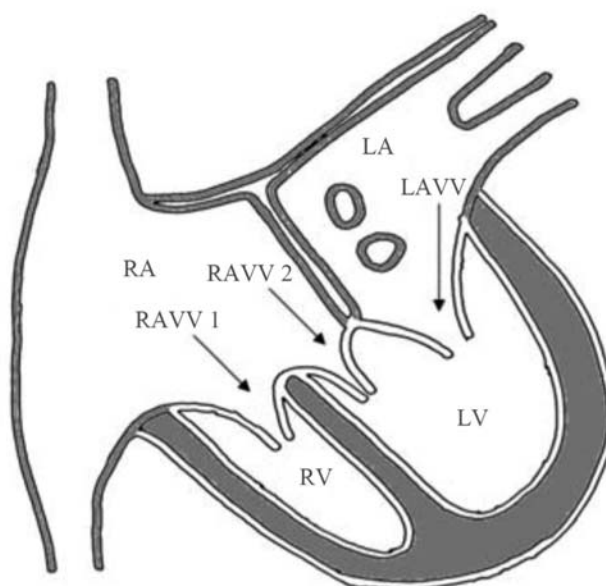
**Figure 6.** Post-operative trans-oesophageal echocardiography, showing the unobstructed communication of the left atrium to the two atrioventricular valve orifices (white arrows) on the left side of the pericardial patch. The star shows the created atrial septal defect and the yellow arrow points to the pericardial patch.



**Figure 7.** Post-operative trans-oesophageal echocardiography, showing the diastolic flow across the two atrioventricular valve orifices on the left side of the pericardial patch.

## DISCUSSION

This report illustrates several features of a rare variant of AV septal defect, with three AV valve orifices. The patient had AVSD with totally closed atrial component dividing the common AV valve into a right and left components. Misalignment between the atrial septum and muscular ventricular septum caused overriding and straddling of the right component of the common AV valve. The ventricular component of the AVSD has also totally closed causing the crest of the ventricular septum to divide the right AV valve into a valve connecting the right atrium to the right ventricle and another smaller valve connecting the right atrium to the left ventricle. Thus although the atrioventricular connections were seemingly concordant, at the same time there was double outlet right atrium and double inlet left ventricle with two balanced ventricles (Figure 8).



**Figure 8.** Diagram showing the atrioventricular (AV) connections. The malalignment of the atrial and ventricular septae caused overriding and straddling of the right AV valve, resulting in double outlet right atrium and double inlet left ventricle with two adequate ventricles. RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle, RAVV: right atrioventricular valve, LAVV: left atrioventricular valve. (Modified from reference 8)

The tiny systolic flow, detected pre-operatively, that was thought to be a leak in the aneurysmal tissue closing the ventricular septal defect, was actually trivial regurgitation in the small orifice of the right AV valve connecting the right atrium to the left ventricle (Figure 2).

Although there was a communication between the right atrium and the left ventricle, the patient did not give history of cyanosis and her resting oxygen saturation was 98%. This can be explained by the fact that the blood preferentially flowed from the right atrium to the right ventricle because the AV valve connecting them was significantly bigger than the other small orifice opening to the left ventricle, in addition the left ventricular compliance was much less than that of the right ventricle, which is secondary to the coexisting left ventricular Hypertrophy due to the subaortic membrane. Due to the difference in compliance between the right and left ventricles, which is increased further in our patient by the presence of left ventricular hypertrophy, due to the co existing subaortic stenosis. This is clearly evident by color Doppler in the pre-operative echo that shows almost no forward flow (from the right atrium to the left ventricle) across the small orifice (Figure 1), in comparison to the post-operative echo, that shows good flow across both orifices (Figure 7).

We chose to connect the small orifice to the left atrium rather than just closing it, because the left AV valve alone would have been small for the patient's body size, especially after closing the "cleft".

AVSD can rarely occur without inter-atrial or inter-ventricular communications.<sup>2</sup> The hallmark of diagnosis would then be the presence of common AV junction with trileaflet left AV valve.

Double orifice left AV valve occurs in AVSD when a tongue of tissue extends between the mural leaflet and one of the LV components of the bridging leaflets.<sup>3</sup> It occurs in about five percent of patients with partial AVSD.<sup>4</sup> This can also rarely occur with the right AV valve. Surgical repair of the left AV valve involves closure of the cleft in the main orifice leaving the accessory orifice intact, and the bridging tissue should not be divided as it is crucial for valve function.<sup>5</sup>

Double orifice left AV valve occurs when the two left valve orifices drain to the same ventricle. But if each orifice drains to a different ventricle, this is called double outlet atrium.<sup>6,7</sup> Double outlet atrium is a quite rare condition. It can be double outlet right atrium or double outlet left atrium, and is generally caused by misaligned atrial or ventricular septae.<sup>8</sup> In some situations, as in our case, this can result in the presence of three AV valves.<sup>8-10</sup> If one AV connection is absent with straddling of the solitary AV valve, the condition will represent uni-atrial but bi-ventricular connection.<sup>11</sup>

In conclusion, this was a rare case of AVSD with intact and misaligned atrial and ventricular septae and overriding and straddling of the right AV valve resulting in double outlet right atrium and double inlet left ventricle; in addition to subaortic membrane.

## Acknowledgements

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