



# Double orifice left AV valve with partial atrio-ventricular septal defect

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## SUMMARY

Partial atrioventricular septal defects (PAVSD) account for three percent of all major congenital cardiac anomalies. It is surgically managed by patch closure of the primum atrial septal defect (ASD) and repair of the cleft left atrioventricular (AV) valve. Double orifice left AV valve, however, occurs in five percent of PAVSD and constitutes a management problem. We report here the echocardiographic presentation, illustrate the surgical findings and suggest technical tips for management of this rare condition.

## BACKGROUND

Atrioventricular (AV) septal defects account for approximately three percent of all major congenital cardiac defects. They are caused by faulty development of the endocardial cushions and are classically divided into partial, transitional, and complete AV canal defects.

We report here a case of a 14-year-old boy who was accidentally discovered to have a partial AV septal defect. The patient was asymptomatic and with no family history of cardiac disease. Echocardiography showed a large primum atrial septal defect (2 cm in diameter), a smaller secundum defect (1 cm), and a cleft left atrioventricular valve (LAVV) with moderate regurgitation. Fig. 1(a) shows the intraoperative three-dimensional trans-oesophageal echocardiography (TOE) with a surgeon's view of the LAVV, showing a double orifice valve. There was a larger anterolateral orifice with a cleft leaflet (arrow), a smaller posteromedial orifice (star), and a bridge of valve tissue in between. The posteromedial orifice was found competent. The cleft in the anterolateral orifice was closed by interrupted sutures. Fig. 1(b) shows the LAVV after closure of the cleft. The atrial septum was closed by a generous pericardial patch, enlarging the left atrium. Fig. 2(a) is a three-dimensional trans-thoracic echocardiography showing a post-operative view of the LAVV after closure of the cleft. Postoperative echocardiography showed mild LAVV regurgitation (Fig. 2(b)). The patient had a smooth postoperative course.

## DISCUSSION

In the partial AV septal defect, a tongue of tissue extends between the two bridging leaflets so that there are dual orifices within the common junction. This creates a trifoliate left AV valve formed by the mural leaflet and the left ventricular (LV) components of the two bridging leaflets. The zone of apposition between the two bridging leaflets, commonly called the cleft, is the site at which left AV valve regurgitation occurs [1].

If an additional tongue of tissue extends between the mural leaflet and one of the LV components of the bridging leaflets, a double orifice may result in the left AV valve [2]. This occurs in about five percent of cases with partial AV septal defects [3]. The smaller accessory orifice is usually present at the posteromedial commissure, while the bigger main orifice is usually anterolateral. Each orifice is supported by a distinct subvalvular apparatus. The double orifice left AV valve shows a variable degree of regurgitation, but is unlikely to have stenosis [4].

The diagnosis of a double orifice left AV valve defect is not easily made and can often be missed [5]. The echocardiographic features include finding two different valve orifices at slightly

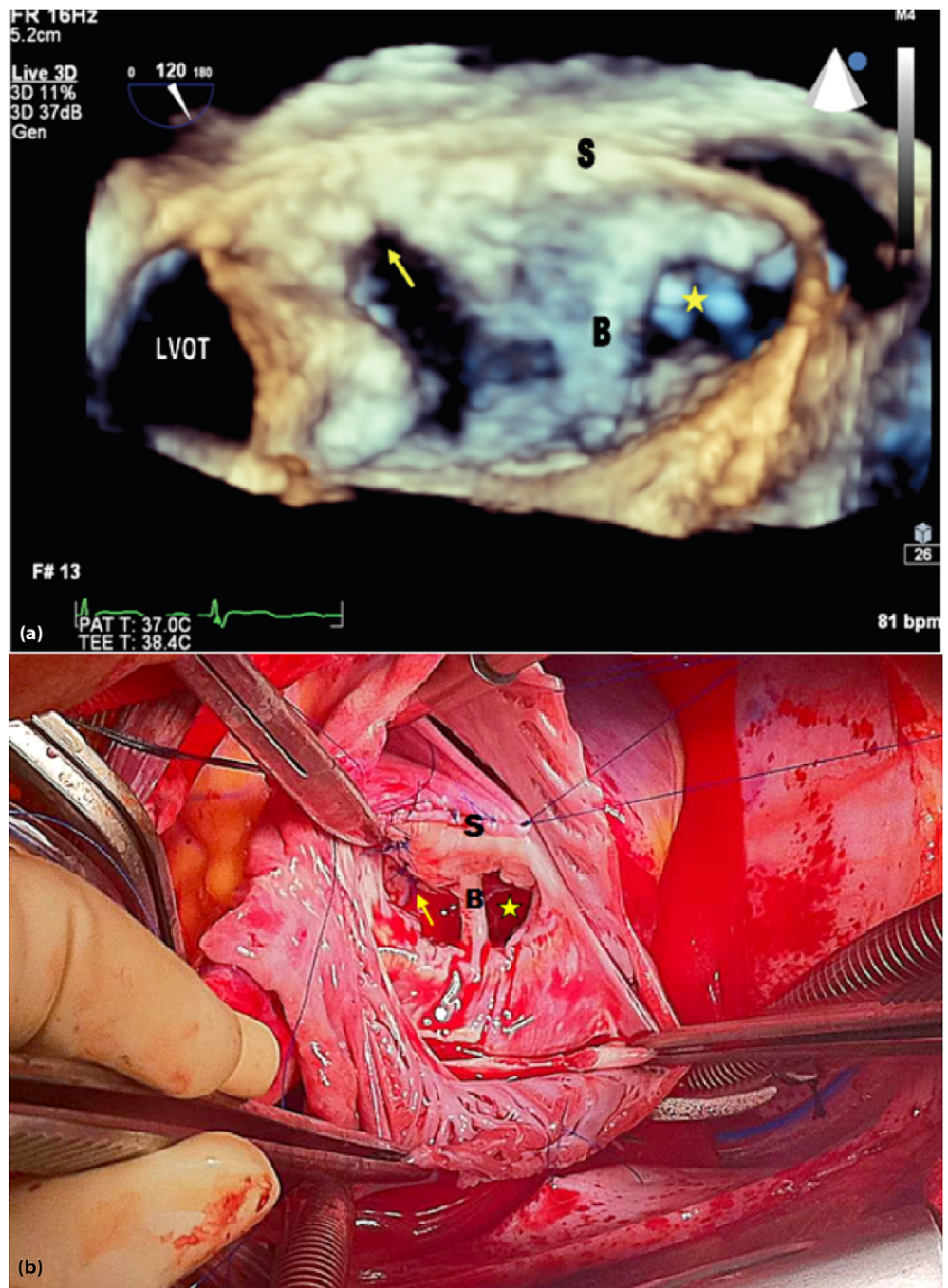
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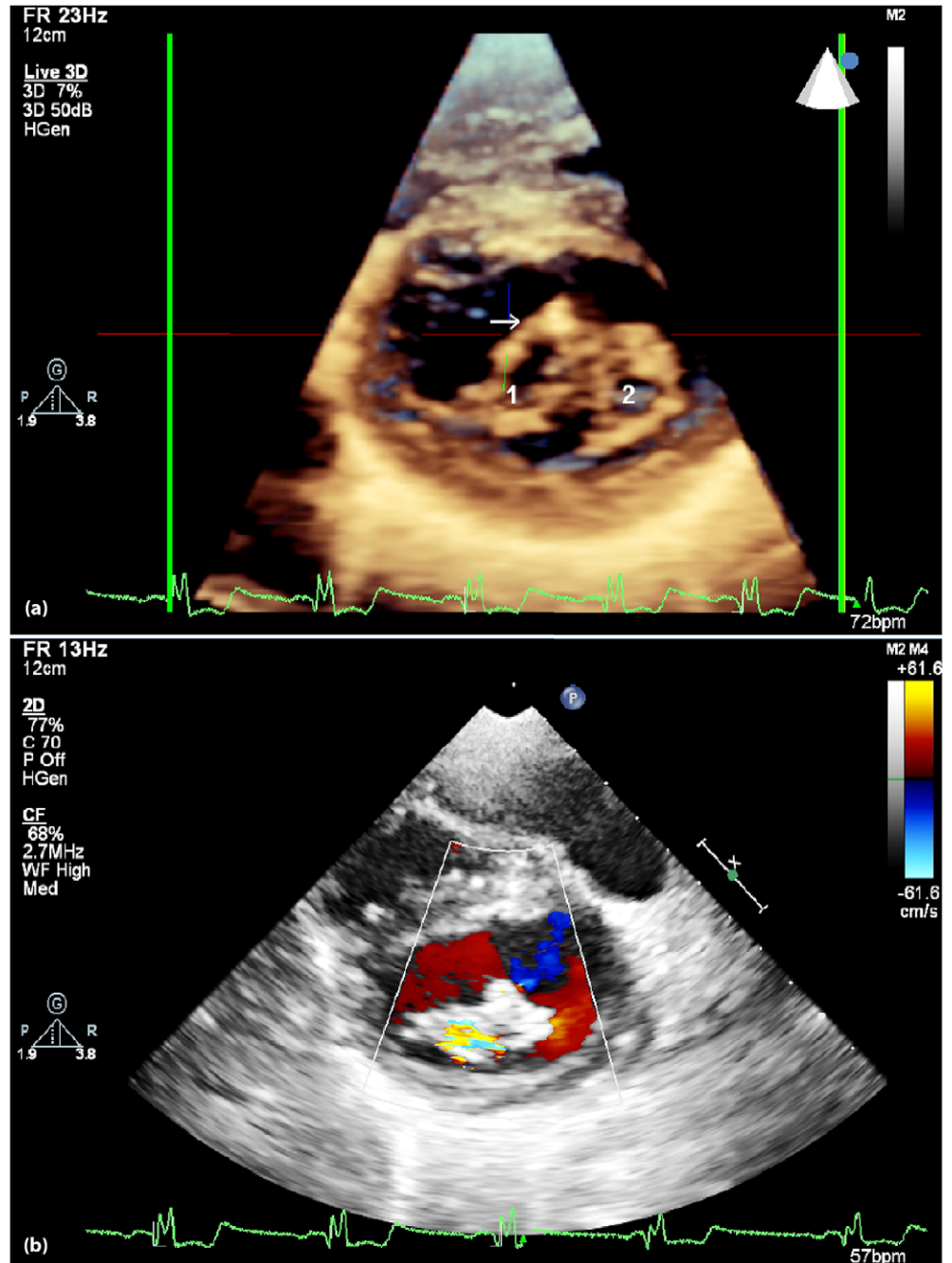
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different planes of ultrasound within the LV cavity on M-mode. In two-dimensional echocardiography, the short axis view can show the left AV valve with two separate holes in the cusps, which open in diastole. In the long axis view, a central bridge of tissue can be seen separating the two orifices [3]. Color Doppler echocardiography may detect two separate jets of left AV valve regurgitation [5].



**Figure. 1 (a)** Three-dimensional trans-oesophageal echocardiography showing a view of the left AV valve from the left atrium. Arrow: cleft in the larger anterolateral orifice. Star: smaller posteromedial orifice. B: Bridge in between two orifices. S: Atrial septum. LVOT: Left ventricular outflow tract. **(b)** Surgical view of the left AV valve. Arrow: the cleft closed (in the larger anterolateral orifice). Star: smaller posteromedial orifice. B: Bridge in between two orifices. S: Atrial septum closed by a pericardial patch.

Surgical repair of the left AV valve involves closure of the cleft in the main orifice till the edge of the leaflets or total closure of the cleft. Where the mural leaflet is too small or the cleft forms a major pathway from the left atrium to the left ventricle, the cleft should be partially closed otherwise left AV valve stenosis or left ventricular outflow obstruction may result. The accessory orifice should be left intact, and the bridging tissue should not be divided as it is crucial for valve function [2,4].



**Figure. 2 (a)** Three-dimensional trans-thoracic echocardiography showing a post-operative view of the left AV valve. Arrow: the site of cleft closed. 1: the larger anterolateral orifice. 2: the smaller posteromedial orifice. **(b)** Short axis view for the left AV valve with color Doppler showing mild regurgitation from the smaller orifice.

**References**

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