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Case Report

Double-Orifice Mitral Valve Associated with Atrioventricular Canal Defects: One Case Report

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Abstract

A 6-month-old male child with exertional dyspnea, was diagnosed on echocardiography as having an atrioventricular canal. This was confirmed during open heart surgery with the identification of a congenital double orifice mitral valve.

The defect was fixed but the double orifice mitral valve was preserved due to the absence of hemodynamic disturbances and in front of satisfying tests.

We report this case along with a review of the literature.

Keywords: *Mitral Valve; Double-Orifice; Atrioventricular Canal Defects*

Introduction

Double orifice mitral valve (DOMV) is a rare congenital defect and was first reported by Greenfield in 1876 [1].

It is usually associated with other cardiac malformations and often with abnormalities of the atrioventricular canal, as it can be isolated [2].

Its existence can compromise the functioning of the valve by causing stenosis or regurgitation [2].

We report here a rare case of DOMV associated with an atrioventricular canal.

Case Report

A 6-month-old male child with Down syndrome, a product of non-consanguineous marriage, presented exertional dyspnea. Clinical examination revealed a left parasternal systolic murmur.

The diagnosis of the atrioventricular canal was detected by electrocardiography.

Per-operatively, the complete atrioventricular canal defect with a Mitral cleft was visualized and two mitral valve orifices were discovered (Figure 1).

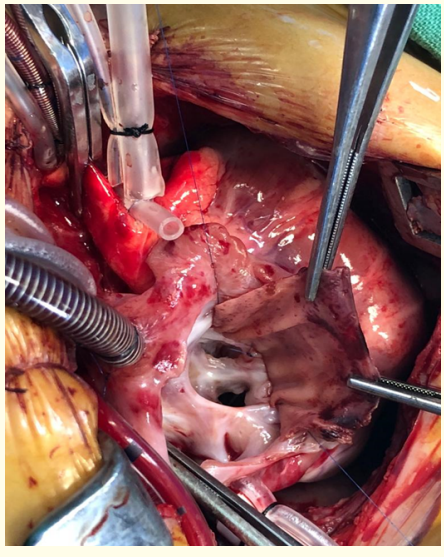

Figure 1: *Preoperative view of DOMV.*

The septal defect was closed by the Nunn technique with a single patch, including partial closure of the cleft.

The double orifice mitral valve was preserved due to satisfying leakage tests and in front of the absence of hemodynamic disturbances.

The postoperative electrocardiography control showed a well-balanced double orifice mitral valve without leakage or stenosis (Figure 2).

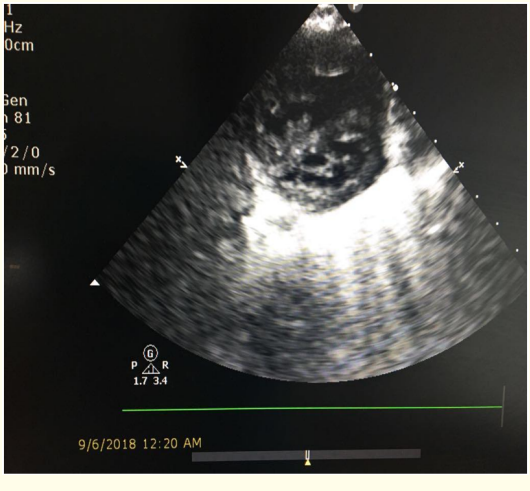

Figure 2: *Postoperative echocardiographic control.*

The postoperative follow-up was good.

Discussion

In a DOMV, an abnormal tissue subdivides the mitral orifice into two portions [3].

The mitral orifices are unequal in size, the smaller one being directed toward the anterolateral commissure (41%) or the posteromedial commissure (44%).

Rosenberg., *et al*. reported that 25% of patients with DOMV had a partial persistent AV defect and about 5% of patients with a partial AV defect had a DOMV [4], in our case, it was a Complete atrioventricular canal (CAVC) A type.

Bano-Rodrigo., *et al*. [5] reported that both orifices are equalized in 15% of cases.

Trowitsch., *et al*. [6] classified the DOMV into three types according to the echocardiography results: the hole type (accessory orifices surrounded by leaflets tissue), complete bridging (fibrous bridge) and incomplete bridging.

In our case, it is the complete bridging type.

DOMV could be the result of fetal endocarditis or a developmental abnormality, the accessory orifice represents retention of the left side of the atrioventricular canal [4].

De Dominic., *et al*. [7], report two cases of DOMV associated with complete AVC defect A type, for which the decision to repair the mitral valve by resection of the bridge led to the death of the patient by massive mitral regurgitation. Hence the interest of preoperative echocardiographic evaluation along with the surgical difficulty to have a balance between the residual leak and the mitral stenosis.

Bibhuti., *et al*. [3] reported that the mitral valve was functionally normal in 9 patients (50%), slightly impaired in 7 (38%) and that only 2 patients (11%) had regurgitation or stenosis.

In our case the mitral valve did not present either stenosis or regurgitation.

Conclusion

This case demonstrates the value of careful preoperative imaging and the benefit of preserving the integrity of the mitral valve if there is no evidence of mitral leakage.

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