

## Case 5 / 2014 – 26-year-old Male with Moderate Stenosis of Bivalved Aortic Valve and Dilation of Ascending Aorta

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**Clinical data**: The patient is followed-up since birth with diagnosis of mild stenosis of bivalved aortic valve. Remained until two years with aortic transvalvular pressure gradient between 30 and 40 mmHg and without physical limitation. Fatigue with exertion is observed for two years, coinciding with increased aortic transvalvular pressure gradient to 60 mmHg in addition to the evidence of mild left ventricular hypertrophy on echocardiogram (interventricular septum and posterior wall = 12 mm).

**Physical examination**: Eupneic, acyanotic, normal pulse. Weight: 86 Kg, Height: 180 cm, BP: 120/80 mmHg, HR: 82 bpm, saturation  $O_2 = 96\%$ . Aorta was mildly palpable in suprasternal notch.

In precordium, *ictus cordis* was not palpable and there were no systolic impulses. Heart sounds were normophonetic and mild systolic murmur was auscultated, +/++ of intensity, rough, on aortic area, furcula, neck vessels and on the left sternal border, with thrill. There was a distinct protosystolic snap across the precordium. Liver was not palpable.

## **Complementary tests**

**Electrocardiogram:** (Figure 1) presented normal sinus rhythm and morphology of P, QRS and T waves, without signs of cavity overload. Ventricular repolarization was normal. AP:  $+40^{\circ}$ , AQRS:  $0^{\circ}$ , AT:  $20^{\circ}$ .

**Chest radiography:** Shows normal heart region and pulmonary vascular markings on expiration radiography. Mild dilation on upper right arch indicating dilation of ascending aorta (Figure 1).

**Echocardiogram Bi-Doppler:** showed heart chambers with normal size and function. There was mild left ventricular hypertrophy with 12 mm thick ventricular septum and posterior wall. Aortic annulus was 35 mm, ascending aorta was 45 mm and maximum aortic transvalvular pressure gradient was 60 mmHg, and mean, 45 mmHg. Other measurements corresponded to LA = 40, RV = 20, LV = 59, EF = 70%, delta D = 40%, RVSP = 25 mmHg (Figure 2).

## **Keywords**

Aortic Valve Stenosis, Aorta / abnormalities, Aortic Aneurysm.

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**Clinical diagnosis:** Moderate stenosis of bivalved aortic valve with dilation of ascending aorta, in adult patient in functional class II.

**Clinical reasoning:** Clinical elements were compatible with the diagnosis of aortic valvular stenosis of moderate repercussion, with systolic murmur in aortic region, fatigue with exertion, supported by mild dilation of ascending aorta clearly showed on chest radiography and confirmed on echocardiogram. The data confirmation was established by transvalvular pressure gradient and left ventricular hypertrophy showed on echocardiogram. The absence of changes of ventricular repolarization on ECG indicates that the pressure gradient does not exceed 60 mmHg.

**Differential diagnosis:** Clinical findings of complementary tests characteristic of aortic valvular stenosis must be differentiated from other obstructive areas such as supra or subvalvular aortic region. The presence of a distinct snap of valvular opening indicates obstruction at the valvular level, in addition to high-pitched systolic murmur.

**Conduct:** Indication for aortic valve replacement surgery was established with the onset of fatigue symptoms associated with signs of progressive worsening, such as increased aortic gradient, myocardial hypertrophy and dilation of ascending aorta. Percutaneous interventional treatment was not considered for being a bivalved aortic valve, with dilation of aortic annulus, and presence of the clear risk of causing aortic insufficiency. For the surgery, bivalved aortic valve was resected, thickened and Bentall and De Bono surgery with plastic tube in the ascending aorta, replacement of coronary arteries and implantation of mechanic valve n. 23 were performed, with positive evolution.

Comments: Congenital aortic valve stenosis when mild in children and teenagers, with pressure gradient below 30 mmHg, presents worsening in half the cases in adulthood. In the presence of bivalved aortic valve, there is an increased possibility for dilation of ascending aorta. It is believed that this dilation dependents on abnormal flow jets towards the vessel. However, there are those who defend genetic modification as its cause. Percutaneous interventional treatment is not recommended for these cases due to valvular failure after balloon dilation of the valve. Placement of valved stent also has difficulties, particularly when fixing it, due do aortic annulus dilation. Surgical prescription in this case was evident due to symptoms worsening, increased transvalvular gradient, onset of myocardial hypertrophy, in addition to dilation of ascending aorta. With early surgical prescription, we prevent phenomena acquired involved with valvular stenosis, which certainly place a greater risk of evolution.

## Clinicoradiological Session



Figure 1 – Normal electrocardiogram and chest radiography shows normal heart area with rounded morphology by hypertrophy and dilation of upper left arch resulting from dilation of ascending aorta.



Figure 2 – Echocardiogram enhances the stress at the aortic valve level in apical projection of 5 chambers to the left, and ascending aorta dilated in longitudinal section in left sternal border, to the right.