Case 4/2015 A 48-year-old Male Patient with Coarctation of the Aorta, Bicuspid Aortic Valve and Normal Ascending Aorta

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Clinical data: The patient reported that, at the age of 23 years, he had been diagnosed with hypertension, which was controlled with five antihypertensive medications. An ultrasound study performed one month ago detected low blood flow through the abdominal aorta. Coarctation of the aorta was suspected, and further confirmed by angiographic CT scan. He had no cardiovascular complaints, but had a sedentary lifestyle.

Physical examination: normal breathing; acyanotic; brisk pulses in upper limbs, absent in lower limbs. Weight: 77 kg; height: 168 cm; Blood pressure – BP = right arm = left arm = 125/75 mmHg; BP legs = 75 mmHg; heart rate – HR: 86 bpm. Aorta clearly palpable on the suprasternal notch. Mild grade 1/4 systolic murmur on the suprasternal notch.

Apical impulse not palpable in the precordium; absence of systolic impulses on the left sternal border. Normal heart sounds; grade 1/4 coarse systolic murmur in the aortic area, with no irradiation. No murmurs in the back. Liver not palpable.

Laboratory tests

Electrocardiogram: Normal sinus rhythm, no chamber overload. Sokoloff index = 20 mm; mild ventricular repolarization abnormality. Left anterior hemiblock and septal fibrosis. PA: +60º; QRSA: -45º; TA: +100º (Figure 1).

Chest radiograph: Normal cardiac silhouette (cardiothoracic ratio: 0.48). Normal pulmonary vascular network and concave arch of pulmonary artery. The posterior aortic arch was not bulged; hyperdensity in some rib borders (Figure 1).

Echocardiogram: Bicuspid aortic valve with maximum and mean pressure gradient of 14 and 7 mmHg, respectively. The ascending aorta (AscAo) was not dilated (35 mm); aortic arch = 37 mm; narrowing in the isthmic region (3 mm), with pressure gradient of 51 mmHg. Cardiac chambers of normal size. No myocardial hypertrophy; septum and posterior wall with 9-mm thickness. Normal left ventricular function (69%).

CT angiography of the aorta: AscAo = 40 mm; aortic crossing and descending aorta (DescAo) = 32 mm; narrowing at isthmus = 3 mm and at diaphragm = 25 mm. Marked collateral circulation, corresponding to the intercostal, bronchial and mammary arteries.

Cardiac catheterization: (Figure 2). Pressure levels were as follows: left ventricle – LV = 160/12, AscAo = 130/80 and DescAo = 90/80 mmHg. Angiography showed clear narrowing at the aortic isthmus of approximately 2.5 cm past the left subclavian artery, with no AscAo or DescAo dilatation distal to the obstruction.

Clinical diagnosis: Severe coarctation of the aorta at the isthmic region and mild aortic valve stenosis in bicuspid valve with no AscAo dilatation or hypertrophy, in natural course.

Clinical reasoning: The clinical elements of coarctation of the aorta are clear and easy to identify, and are mainly represented by contrasting pulses and BP between the upper and lower limbs, as occurred in this case, although detected late by indirect means of abdominal ultrasound. Noteworthy, despite the long-standing heart disease with pressure overload, no myocardial hypertrophy, signs of electric overload or symptoms are observed. This results from the fact that, in this obstructive anomaly, there is formation of collateral vessels that deviate blood in order to nourish the general circulation. Chest radiograph with normal cardiac silhouette expresses preserved ventricular function, and surprisingly there is no AscAo dilatation despite the presence of bicuspid aortic valve and coarctation of the aorta.

Differential diagnosis: Aortic obstructions in adulthood usually result from previous aortitis, as occurs in Takayasu and Kawasaki disease, as well as in connective tissue and infectious diseases.

Management: In view of the long-standing systolic impact, even without myocardial hypertrophy and with increased pressures in the proximal aorta circuit, operation is mandatory aiming at relieving the arterial obstruction, which would imply a greater possibility of development of myocardial fibrosis, heart failure, arrhythmias and earlier death. Dilatation of the isthmic region was chosen using therapeutic cardiac catheterization from the right femoral artery with placement of a 40 x 18-mm stent just past the origin of the left subclavian artery. Systolic BP immediately decreased to 100 mmHg even without the use of antihypertensive drugs. Adequate enlargement of the region was achieved (Figure 2) with equalization of pressures between limbs. The mild systolic murmur of aortic valve stenosis remained unchanged.

Commentaries: Cardiac compensation with maintained antegrade flow, in coarctation of the aorta, is achieved by the myocardial hypertrophy and/or development of collateral circulation, which alleviates the aortic obstruction and satisfactorily nourishes the circulation. In this case, the marked aortic obstruction detected in adulthood and manifested

Keywords

Coarctation of Aorta, Bicuspid Aortic Stenosis; Percutaneous; Aortic Treatment.

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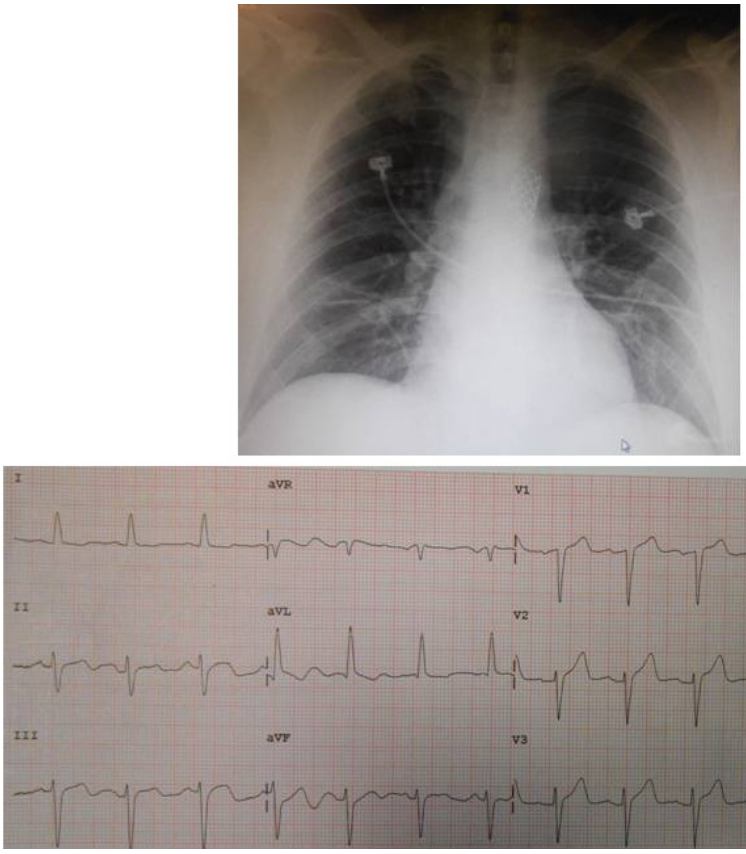

Figure 1 – *Chest radiography showing normal cardiac silhouette. 40 x 18-mm stent positioned in the beginning of the descending aorta, corresponding to the arch of the pulmonary artery. Hyperdensity can be observed in the lower border of some ribs. On electrocardiogram, left anterior hemiblock, septal fibrosis and upper lateral ischemia.*

by hypertension, was compensated by exuberant collateral circulation. Hence the favorable outcome until the fifth decade of life. Left anterior hemiblock and septal fibrosis on the electrocardiogram are difficult to explain in the absence of myocardial hypertrophy. Even when hemodynamically compensated, this obstructive anomaly should be treated early so as to prevent an unfavorable outcome in relation to the development of myocardial fibrosis, arrhythmias and heart failure. In the course of bicuspid aortic valve, unfavorable elements such as AscAo dilatation arise, and these invariably should be treated surgically in a timely manner. However, in this patient, the expected aortic dilatation was not observed, and this makes a good outcome possible even for a longer term. Aortic dilatation seems to be more related to inherent factors of the arterial wall (adequate morphology of the parietal muscle) than to hemodynamic aspects[1].

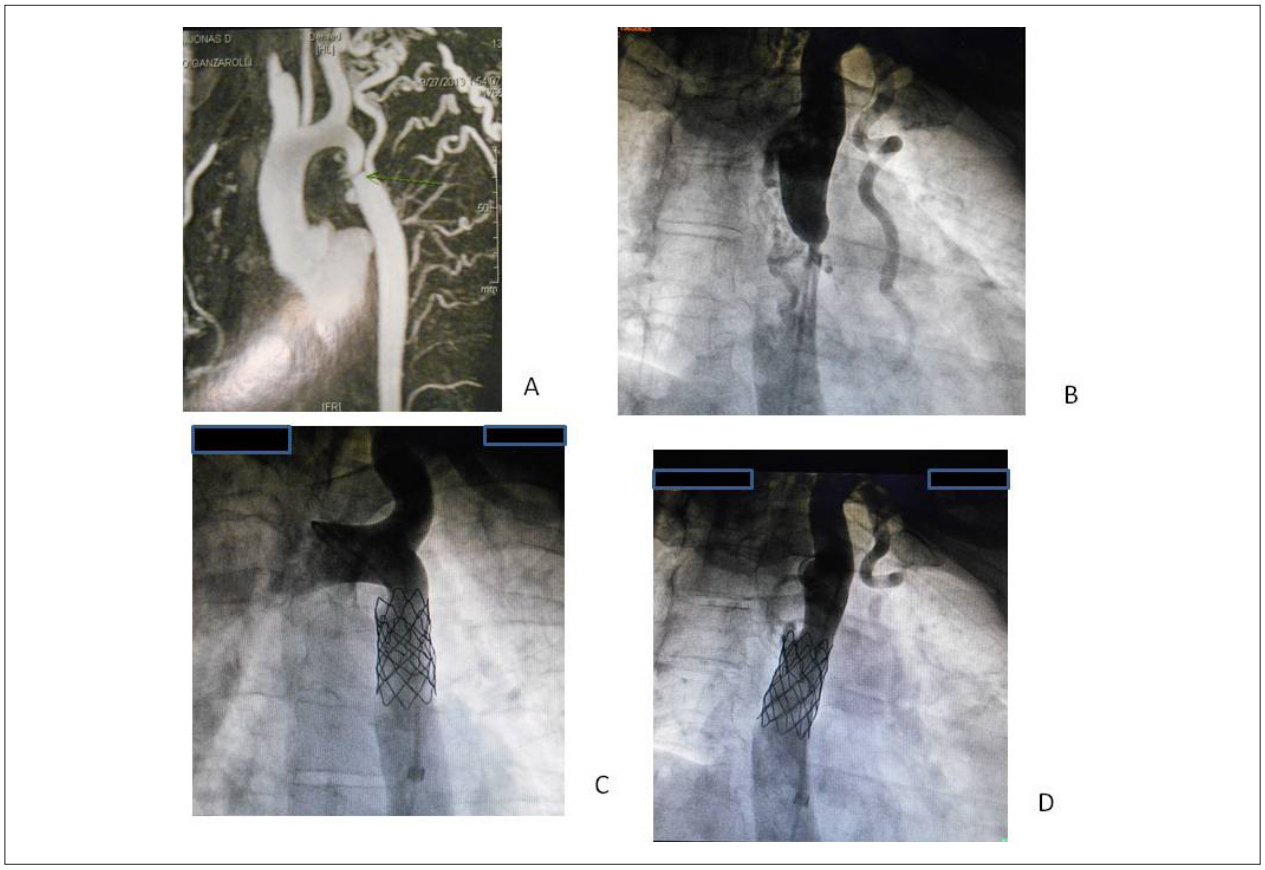

Figure 2 – *Upper angiographic images (A and B) show marked isthmic coarctation of the aorta with normal ascending aorta, and exuberant collateral circulation; C and D show images of the descending aorta after placement of a 40 x 18-mm stent, with uniform diameters and no post-stenotic dilatation.*

Reference

1. Jackson V, Petrini J, Caidahl K, Eriksson MJ, Liska J, Eriksson P, et al. Bicuspid aortic valve leaflet morphology in relation to aortic root morphology: a study of 300 patients undergoing open-heart surgery. Eur J Cardiothorac Surg. 2011;40(3):e118-24. Erratum in: Eur J Cardiothorac Surg. 2012;41(2):471.