Case 6 / 2014 – Five-year Old Child with Scimitar Syndrome and Pulmonary Sequestration of Right Lower Lobe

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Clinical data: Heart murmur was auscultated in routine examination in a two-year old male child, who remained asymptomatic and without use of specific medication.

Physical examination: Eupneic, acyanotic, normal pulse. Weight: 18.6 Kg, Height: 109 cm, BP: 100/60 mmHg, HR: 90 bpm, saturation O_2= 99%. Aorta was not palpable in furcula.

In precordium, *ictus cordis* was not palpable and there were no systolic impulses. Heart sounds were normophonetic, but audible on the right sternal border, and the second heart sound was constant and with mild systolic murmur, +/++ of intensity, harsh, in the aortic area, irradiated to the right sternal border, larger than the left one. Liver was palpable at 1 cm from the right costal margin.

Complementary tests

Electrocardiogram: Showed sinus rhythm and RS morphology in right precordial and rs in V6 compatible with right ventricular overload. Ventricular repolarization was normal. AP: +20º, AQRS: +250º, AT: +30º.

Chest radiography: Shows hypoplasia of right lung with increased cardiac area positioned to the right and increased pulmonary vascular markings. There were signs of vascular imaging on the right side of the retrocardiac image with an outlining that resembled a scimitar (Figure 1).

Bi-Doppler echocardiogram: showed a clear enlargement of right cavities, dilation of the pulmonary trunk and left pulmonary artery. 5 mm *ostium secundum* interatrial communication with shunt from the left to the right. Pulmonary veins on the left normally drain in normal left atrium and right veins on the dilated inferior vena cava, next to the right atrium. Measurements: RV = 26, LV = 31, PT = 21, RPA = 7, LPA = 13, annulus T = 22, annulus M = 16, annulus P = 15 annulus Ao = 15 mm, RVSP = 37 mmHg, LVEF = 66%, (Figure 2).

Keywords

Scimitar syndrome; Heart murmurs; Bronchopulmonary sequestration.

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Cardiac catheterization confirmed the diagnosis of Scimitar syndrome with abnormal draining of right pulmonary veins in inferior vena cava and interatrial communication. It was verified with an injection of contrast agent in the descending aorta the presence of systemic-to-pulmonary collateral vessel which was being directed to the right lower lobe (pulmonary sequestration) and underwent embolization by placing four spring coils (Figure 2). Pressures: RA = 11, RV = 40/12, PT = 36/8-17, PC = 16, LA = 12, LV = 82/17, Ao = 90/60-70 mmHg.

Clinical diagnosis: Scimitar Syndrome in hypoplasia of the right lung and interatrial communication, with shunt from left to right, and pulmonary sequestration of the right lower lobe, in asymptomatic child.

Clinical reasoning: Clinical elements were compatible with the diagnosis of interatrial communication and abnormal drainage of right pulmonary veins on the lower vena cava in hypoplasia of the right lung, indicating Scimitar syndrome. Pulmonary sequestration on the right side was established by the cardiac catheterization.

Differential diagnosis: Clinical findings and complementary tests, characteristic of Scimitar syndrome, have not found differentials similar to other abnormalities. These elements must be always highlighted and remembered in the presence of hypoplasia of the right lung. Under these circumstances, the associated pulmonary sequestration must also be remembered.

Conduct: Surgery for redirection of right pulmonary veins to the left atrium and closure of interatrial communication is supported by the effects repercussion, given the right heart cavities large increase, motivated by shunt from left to right at the atrial level.

Comments: Scimitar syndrome published by Cooper[1] in 1836, and therefore designated by Neill, given the morphologic analogy with the Turkish curved in 1960[2], is systematically associated with homonymous pulmonary hypoplasia to the right and sometimes with pulmonary sequestration, congenital cardiopathies (more commonly IAC) and other defects known as "venolobar syndrome". The clinical status is similar to that of an interatrial communication, which is repercussion when pulmonary hypoplasia is not significant. Otherwise, the more hypoplastic the lung is, the more reduced the pulmonary flow is to the right and clinical manifestation is frustrated, to a point of not having a benefit from surgical correction. In association with pulmonary sequestration, it is always recommended the embolization of systemic-to-pulmonary vessel in order to avoid complications of pulmonary hemorrhage and localized pulmonary hypertension, in addition to pulmonary infection.

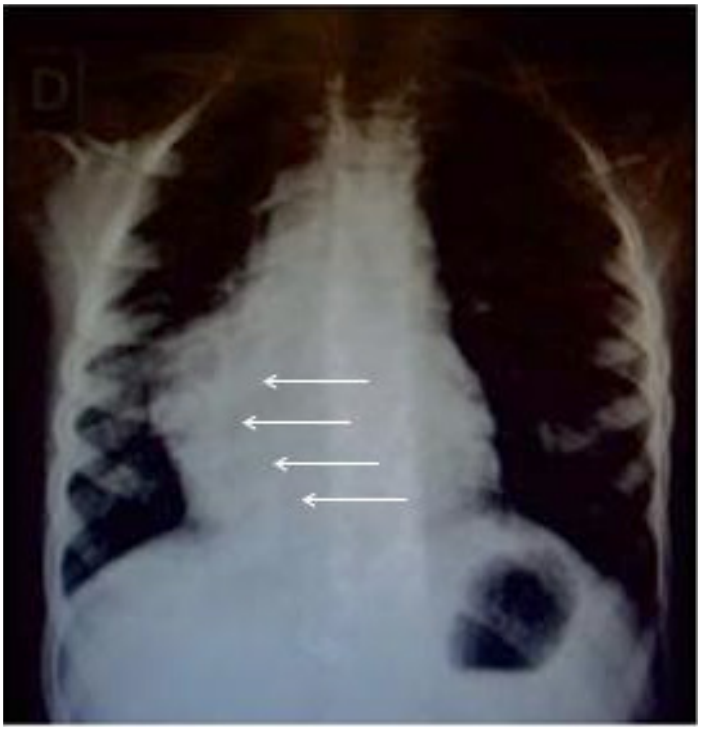

Figure 1 – *Chest radiography shows the increased heart area to the right with increased pulmonary vascular markings and venous vessel in the hypoplastic right lung, simulating the shape of scimitar, in the retrocardiac image (arrows).*

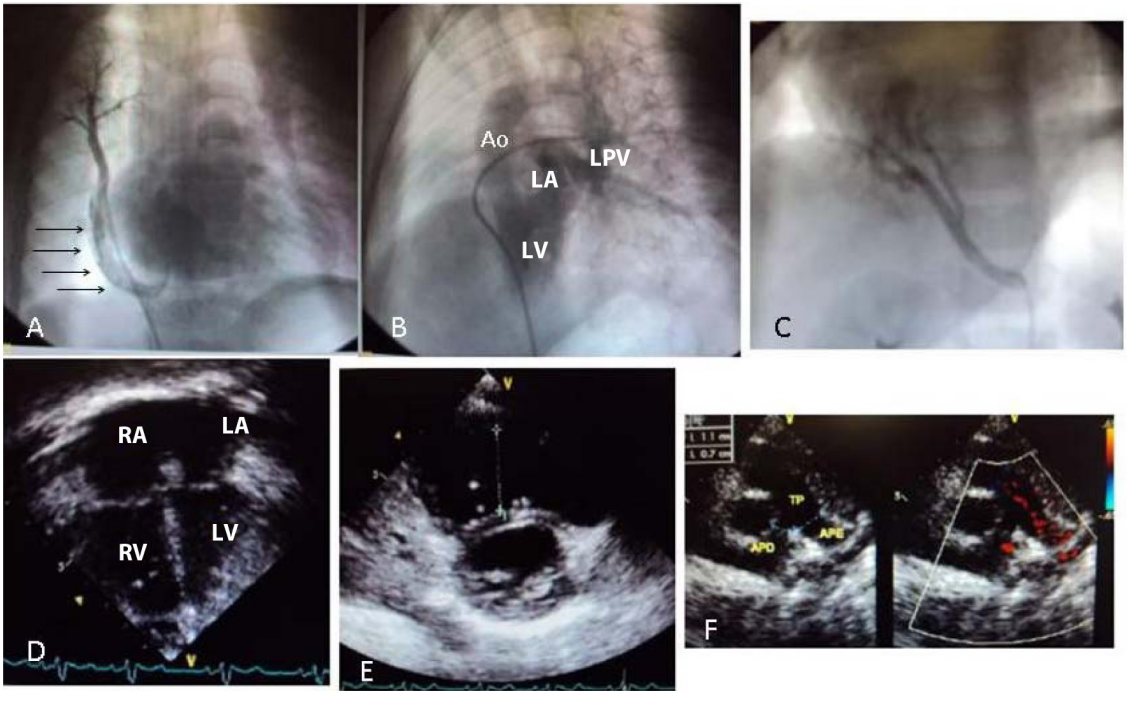

Figure 2 – *Angiography in right upper pulmonary vein enhances the Scimitar shape of right common pulmonary vein (arrows) towards lower vena cava in A, Left Pulmonary Vein (LPV) in the Left Atrium (LA) in continuity of the Left Ventricle (LV) and Aorta (Ao) in B and systemic-to-pulmonary vessel of descending aorta to right lower lobe in C. Echocardiographic images show the increased right cavities in apical view of 4 chambers in D, the large increase of right ventricle in cross-sectional view in E and dilated pulmonary trunk and left pulmonary artery in cross-sectional view in F.*

References

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