

Aortic coarctation and horseshoe kidney in a 3-year-old infant

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ABSTRACT

Aortic coarctation is a disease that consists in a deformation of the tunica media at the point of origin of the descending aorta. Although clinical manifestations usually occur in newborns and infants, some cases may be diagnosed in later stages of childhood, especially mild to moderate coarctation. Therefore, physical examination is essential when there is suspicion of its presence. The case of a female patient whose diagnosis was made at 3 years of age is reported. It was based on clinical elements (history, auscultatory characteristics, of peripheral pulses, arterial hypertension) and confirmed by imaging techniques. A horseshoe kidney was found as an associated malformation. An interventional procedure (therapeutic cardiac catheterization) was performed and the patient improved.

Key words: Aortic coarctation, Doppler echocardiography, Tomography

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RESUMEN

La coartación aórtica es una enfermedad que consiste en una deformación de la túnica media en el punto de origen de la aorta descendente. Aunque las manifestaciones clínicas se presentan generalmente en recién nacidos y lactantes, algunos casos pueden diagnosticarse en etapas más avanzadas de la infancia, sobre todo las coartaciones leves o moderadas, por ello el examen físico es esencial ante su sospecha. Se presenta el caso de una paciente cuyo diagnóstico se realiza a los 3 años de edad, basado en elementos clínicos (antecedentes, características auscultatorias, de los pulsos periféricos, hipertensión arterial) y se corrobora por técnicas de imagen. Como malformación asociada se detectó riñón en herradura. Se realizó procedimiento intervencionista (cateterismo terapéutico) y la paciente evolucionó favorablemente.

Palabras clave: Coartación aórtica, Ecocardiografía Doppler, Tomografía

INTRODUCTION

Congenital heart defects are anatomic abnormalities of the cardiovascular system at birth, which results functional alterations¹. They occur during the

early stage of fetal life and are the result of alterations in the embryonic development of a normal structure^{1,2}. Aortic coarctation (AC) is a disease that consists in a deformation of the tunica media at the point of origin of the descending aorta. It affects the wall by a fold, in the form of a curtain, which enters the vessel lumen and produces an eccentric narrowing. It is generally located below the origin of the left subclavian artery and is responsible for 8% of all congenital heart diseases. In terms of frequency, it follows ventricular septal defect and patent ductus arteriosus¹⁻⁴.

The case of a 3-year-old girl with AC and horseshoe kidney is reported.

CASE REPORT

A 3-year-old, female, white patient, from the municipality of Camajuani, Villa Clara, with a history of failure to thrive, was examined at the childcare consultation and a heart murmur was found during auscultation. She was referred to a Pediatric Cardiology consultation. Her parents said she got tired relatively easy; and her weight/height nutritional status was below the third percentile.

Physical examination revealed a normal precordium, without thrill or epigastric beat, but the apex beat was found in the fifth left intercostal space and in the midclavicular line. Heart sounds were rhythmic, S₁ and S₂ were normal, with II/VI systolic murmur in aortic area, radiating to the neck vessels and to the interscapular region. There was no S₃ and peri-pheral femoral pulses were weak. Heart rate was 115 beats per min-ute and blood pressure 130/80 mmHg.

The Doppler echocardiography (Figure 1, A and B) showed: *situs solitus*, concordance of all segments, signs of left

ventricular hypertrophy, competent valve apparatus, without pathological gradients; left aortic arch with a mild AC (peak gradient of 45 mmHg), continuous flow in abdominal aorta with diastolic component (Figure 1C), normal interventricular and interatrial septum, preserved global systolic and segmental function (ejection fraction of 70%), normal pericardium, no spills, type I pulmonary flow curve and pulmonary acceleration time of 96 ms.

The report of the computed tomography angiography (Siemens Definition of dual source), with the use of contrast, showed, after performing axial cuts and multiplanar and volumetric reconstructions, a coarctation of the aorta below the left subclavian artery and the probable existence of a small muscular ring (Figure 2A). Pre-coarctation diameter was 9 mm, post-coarctation diameter was 13 mm and the length of the constricted segment was 12 mm. At the level of the abdomen, a fusion of the lower renal poles was observed, before the aorta, which was consistent with horseshoe kidney (Figure 2B).

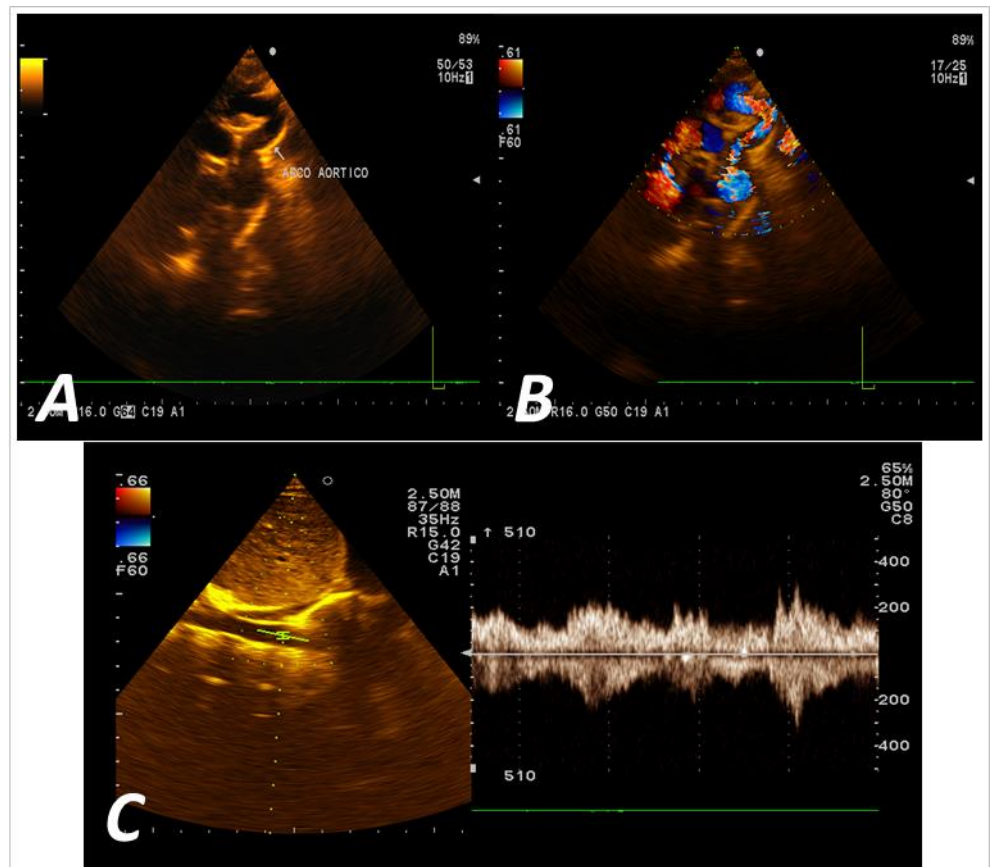


Figure 1. A. Aortic arch (B-mode). B. Color Doppler. C. Continuous Flow Chart in abdominal aorta.

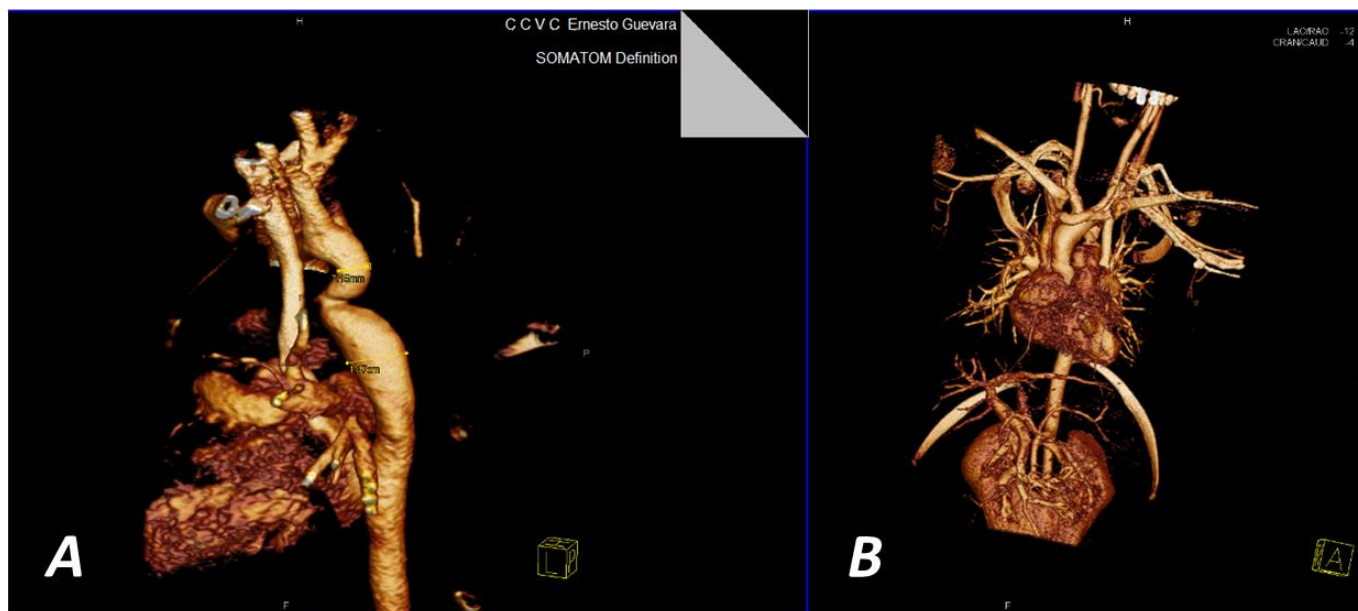


Figura 2. A. Aortic coarctation and its pre and post-coarctation diameters. **B.** Association with horseshoe kidney, fused at their lower poles.

The patient was sent to the national reference center where a hemodynamic study was conducted showing a gradient which was sufficient to perform an interventional procedure. The outcome has been satisfactory after the intervention.

COMMENT

AC is a congenital heart disease which usually appears in the early stages of life⁵. However, some cases, as the one reported here, may be diagnosed at later stages of childhood, especially mild to moderate coarctation^{5,6}. The clinical approach remains the cornerstone for diagnosis. The medical history and physical examination were essential in this case. Failure to thrive, the auscultation of a systolic murmur with interscapular radiation, the difference in the weak peripheral pulses in the lower limbs and hypertension, are all characteristic elements of great interest⁷⁻¹⁰.

AC increases the resistance to blood flow, which is greater the narrower the lumen of the vessel¹¹. The level of obstruction caused by aortic coarctation progresses slowly after birth. The systolic and diastolic blood pressures above AC are often abnormally high, while in the femoral artery, below AC, the systolic pressure is lower^{11,12}. As a consequence of hypertension in the upper body, an abundant collateral circulation develops, which often maintains asymptomatic the patient⁵. A quarter of the patients with this

condition presents heart failure in the first week of life, and another quarter in the next three weeks. It is possible that the closure of the ductus arteriosus contributes to the obstruction and the sudden onset of symptoms in some patients. The clinical symptoms of these children include: dyspnea, tachypnea, trouble feeding and failure to thrive¹⁻³. The precordial beats, from both ventricles, are hyperdynamic and there may be gallop rhythm; moreover, a low intensity systolic murmur may be heard in the interscapular region. Clinical diagnosis is made by comparing the pulses of the upper and lower limbs: femoral pulse is weak, delayed or not palpable, and systolic blood pressure in the upper arm is generally 30 mmHg higher than in the leg of the same side of the body^{1,4-6}.

Scientific advances make diagnosis easier with the use of innovative diagnostic imaging techniques, however, echocardiography remains the mainstay^{12,13}.

Horseshoe kidney was detected as an associated malformation, which reaffirms that birth defects often do not occur in isolation, but in association with other defects^{1,3}. Timely therapy, often by interventional procedures, is very effective and minimizes complications¹³.

REFERENCES

1. Alcívar J, García E, Gutiérrez-Larraya F, Moreno F, Pan M, Santos J. Guías de actuación clínica de la

- Sociedad Española de Cardiología. Requerimientos y equipamiento de las técnicas invasivas en cardiología pediátrica: aplicación clínica. *Rev Esp Cardiol*. 1999;52:688-707.
- Oyen N, Poulsen G, Boyd HA, Wohlfahrt J, Jensen PK, Melbye M. Recurrence of congenital heart defects in families. *Circulation*. 2009;120:295-301.
 - Vega T, González G, Llanes MC, Ley L, Garí M, García Y. Morbilidad y mortalidad por cardiopatías congénitas en lactantes. Villa Clara, años 2006-2010. *Acta Méd Centro* [Internet]. 2011 [citado 31 Ene 2015];5:[aprox. 9 p.]. Disponible en: http://www.actamedica.sld.cu/r4_11/lactantes.htm
 - Calderón-Colmenero J, Cervantes-Salazar JL, Curi-Curi PJ, Ramírez-Marroquín S. Problemática de las cardiopatías congénitas en México. Propuesta de regionalización. *Arch Cardiol Mex*. 2010;80:133-40.
 - Herranz B. Control de los niños con cardiopatía congénita en atención primaria. *Rev Pediatr Aten Primaria*. 2009;11:639-55.
 - Lee MG, d'Udekem Y. Coarctation of the aorta can no longer be considered a benign condition. *Heart Lung Circ*. 2014;23:297-8.
 - Oyen N, Poulsen G, Wohlfahrt J, Boyd HA, Jensen PK, Melbye M. Recurrence of discordant congenital heart defects in families. *Circ Cardiovasc Genet*. 2010;3:122-8.
 - Donazzan L, Crepez R, Stuefer J, Stellin G. Abnormalities of aortic arch shape, central aortic flow dynamics, and distensibility predispose to hypertension after successful repair of aortic coarctation. *World J Pediatr Congenit Heart Surg*. 2014;5:546-53.
 - Dzsinich C, Vaszily M, Vallus G, Dzsinich M, Berek P, Barta L, *et al*. Late complications and treatment options of aortic coarctation operated in childhood. *Orv Hetil*. 2014;155:1189-95.
 - Früh S, Knirsch W, Dodge-Khatami A, Dave H, Prêtre R, Kretschmar O. Comparison of surgical and interventional therapy of native and recurrent aortic coarctation regarding different age groups during childhood. *Eur J Cardiothorac Surg*. 2011;39:898-904.
 - Moore JW, Vincent RN, Beekman RH, Benson L, Bergersen L, Holzer R, *et al*. Procedural results and safety of common interventional procedures in congenital heart disease: initial report from the National Cardiovascular Data Registry. *J Am Coll Cardiol*. 2014;64:2439-51.
 - Riesenkampff E, Fernandes JF, Meier S, Goubergrits L, Kropf S, Schubert S, *et al*. Pressure fields by flow-sensitive, 4D, velocity-encoded CMR in patients with aortic coarctation. *JACC Cardiovasc Imaging*. 2014;7:920-6.
 - Ostovan MA, Kojuri J, Mokhtaryan M, Razazi V, Zolghadrasli A. Procedural Outcome and One Year Follow up of Patients Undergoing Endovascular Stenting for Coarctation of Aorta: A Single Center Study. *J Cardiovasc Thorac Res*. 2014;6:117-21.