

TWO DIMENSIONAL AND DOPPLER ECHOCARDIOGRAPHIC DIAGNOSIS OF BICUSPID PULMONARY VALVE

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The diagnosis of bicuspid pulmonary valve is usually established postmortem, occurring in 7% of patients with pulmonary stenosis and is usually associated to tetralogy of Fallot or ventricular septal defect. The authors report the case of a 14-year-old girl with patent ductus arteriosus in whom 2-D echocardiography disclosed a pulmonary valve formed of two slightly thickened leaflets that opened with the shape of a dome in systole, and moved under the valver ring plane during diastole. A Doppler examination showed a 21 mmHg transvalvar gradient. These characteristic features suggest that 2-D and Doppler echocardiography are the techniques of choice in the diagnosis of bicuspid pulmonary valve.

DIAGNÓSTICO ECODOPPLERCARDIOGRÁFICO DE VALVA PULMONAR BICÚSPIDE

O diagnóstico de valva pulmonar bicúspide é habitualmente feito à necrópsia, ocorrendo em 7% dos pacientes com estenose pulmonar e associando-se em geral a tetralogia de Fallot ou a comunicação interventricular. É relatado caso e menina de 14 anos com persistência do canal arterial, em que se detectou à ecocardiografia bidimensional valva pulmonar formada por dois folhetos espessados, abrindo-se em cúpula durante a sístole, e projetando-se abaixo do plano de anel valvar na diástole. Complementação do exame com Doppler demonstrou gradiente transvalvar em 21 mmHg. Estes aspectos característicos sugerem a ecocardiografia bidimensional, associada ao Doppler, como método de escolha para o diagnóstico de valva pulmonar bicúspide.

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While stenosis of the pulmonary valve, either isolated or associated with other defects, is a common anomaly which occurs in almost 10% of all congenital cardiopathies, bicuspid pulmonary valve is only found in a small subgroup (7%) of patients with stenosis. It is generally associated with tetralogy of Fallot or ventricular septal defect (1). The diagnosis of bicuspid pulmonary valve is usually established postmortem. Phono or echocardiographic identification has required confirmation with the anatomic specimen or has been a fortuitous finding following an attempted balloon catheter pulmonary valvulotomy (2).

The purpose of this report is to demonstrate how two dimensional (2-D) and color codified Doppler images can be used to identify bicuspid pulmonary valve in a patient with associated patent ductus arteriosus.

Case Report.

A 14 year old normally developed female was referred for study because of a heart murmur. She had been asymptomatic until 4 months before, when she presented exertional dyspnea and occasional palpitations. Physical exploration revealed absence of cyanosis, broad arterial pulses and a suprasternal thrill. On auscultation of the second left intercostal space S₁ was duplicated by a protosystolic click; a continuous murmur was evident. The EKG was normal, and a chest roentgenogram showed absence of cardiomegaly, a prominent

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pulmonary artery and increased pulmonary vasculature.

A 2-D echocardiographic examination from a parasternal position on the second left intercostal space allowed transverse exploration of the pulmonary valve. Only two slightly thickened leaflets were identified, a large one in a left anterior position and a smaller right posterior one, and two commissures (Fig. 1). In a conventional parasternal short axis image of great vessels the pulmonary leaflets were observed to have a domed opening (Fig. 2) in systole. In diastole they moved the right ventricular outlet, passed the plane of the valve ring and formed an inverted dome (Fig. 3). A continuous wave Doppler registry oriented by color Doppler images demonstrated a transvalvular pressure gradient of 21 mmHg. Patent ductus arteriosus was confirmed both by visualization in two dimensional registries and by the presence of retrograde diastolic flow in the pulmonary artery. On the basis of the findings described the patient has been programmed for surgical closure of the ductus arteriosus without the necessity of cardiac catheterization.

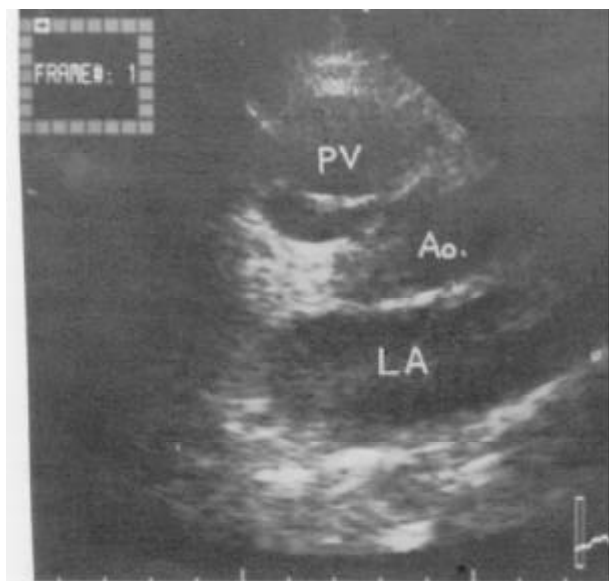


Fig. 1 Parasternal image on second left intercostal space for evaluating the pulmonary valve at the level of implantation. Two leaflets can be identified, a large left anterior one and a smaller right posterior one as well as two commissures. PV | Pulmonary valve; Ao | Aorta; LA | Left atrium.

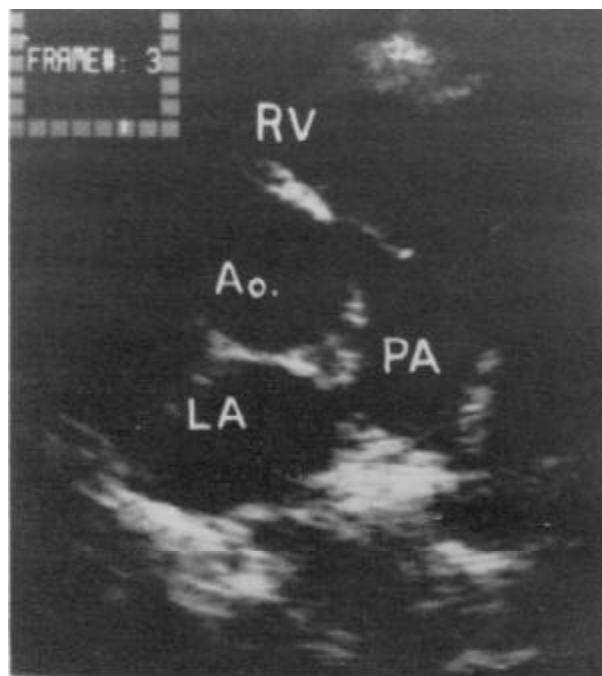


Fig. 2 Conventional parasternal short axis image. Systolic doming of the pulmonary valve is apparent. RV | Right ventricle; PA | Pulmonary artery. Other abbreviations as before.

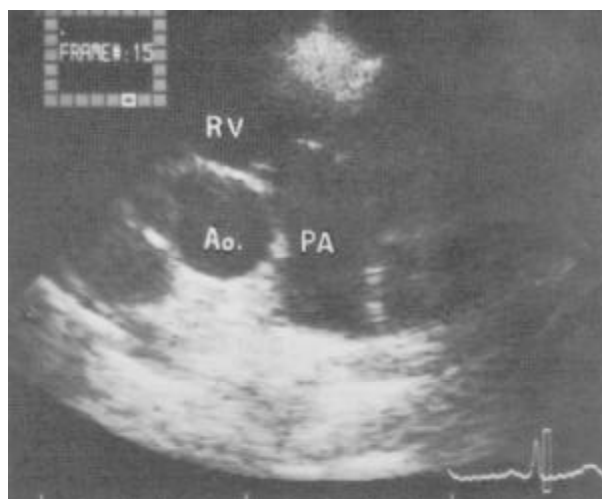


Fig. 3 Conventional parasternal short axis image. The end-diastolic displacement of the pulmonary leaflets toward the right ventricular outlet can be observed. Abbreviations as before.

DISCUSSION

Unlike bicuspid aorta, bicuspid pulmonary valve is rarely diagnosed^{1,2}. Perhaps for this reason little attention has been paid to looking for it and attempting by non-invasive methods. We believe that in any patient with a basal protosystolic click and a tranvalvular gradien-

te detected by Doppler bicuspid pulmonary valve should be investigated. Two dimensional parasternal short axis views complemented by transverse images of the pulmonary valve in parasternal position on the second left intercostal space are of particular utility in establishing the diagnosis. In the former the dome morphology of the valve oriented toward the pulmonary artery in systole and the right ventricle in diastole is characteristic. Diagnosis is confirmed with the latter images, which demonstrate the existence of only two pulmonary valve leaflets. With these data and the information derived from doppler registries

it is possible to quantify the transvalvular pulmonary pressure gradient and investigate valvular regurgitation as well as other associated defects. These observations indicate that 2-D and Doppler echocardiography are the technique of choice in the diagnosis of bicuspid pulmonary valve.

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