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CASE REPORT

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Shone's syndrome: Insights from three-dimensional echocardiography

Martín F. Vivas | María T. Politi | Laura M. Riznyk | María F. Castro | Gustavo Avegliano | Ricardo Ronderos

Section of Echocardiography, Cardiac Imaging Department, Cardiovascular Institute of Buenos Aires, Buenos Aires, Argentina

Correspondence
Ricardo Ronderos, Instituto Cardiovascular de Buenos Aires (ICBA), Buenos Aires, Argentina.
Emails: rronderos@icba.com.ar; trotare@hotmail.com

Shone's syndrome is a rare congenital anomaly defined as the presence of at least two of the following heart obstructions: a mitral supravalvular ring, a "parachute" mitral valve stenosis, subaortic stenosis, and aortic coarctation. A 58-year-old man presented with a mitral ring and a "parachute" mitral valve on two-dimensional transthoracic echocardiography, raising suspicion of Shone's syndrome. Three-dimensional transesophageal echocardiography revealed a subannular mitral ring inserted directly on the mitral leaflets, thus acting as a "valvar ring." This distinction can have therapeutic implications as a "valvar" mitral ring could require valve repair or replacement, instead of simple resection.

KEYWORDS
congenital heart defects, mitral valve obstruction, three-dimensional echocardiography

The rare congenital syndrome initially described by Shone was defined by four potentially obstructive conditions: a mitral supravalvular ring in the left atrium, mitral valve stenosis produced by a "parachute" deformity, subaortic stenosis, and aortic coarctation.[1]

We describe the case of a 58-year-old male presenting with functional class II dyspnea and supraventricular arrhythmia. He had a history of severe mitral valve stenosis thought to be rheumatic.

Three-dimensional transesophageal echocardiography (3DTEE) was planned to complement two- and three-dimensional transthoracic echocardiography (2D/3DTTE) to assess the feasibility of percutaneous mitral valvuloplasty. 2D echocardiography showed a mitral valve with a "parachute" deformity and a membranous mitral ring. Thick, short chordae converged to insert into one major apical papillary muscle. The mitral ring was subannular and seemed to be inserted distally

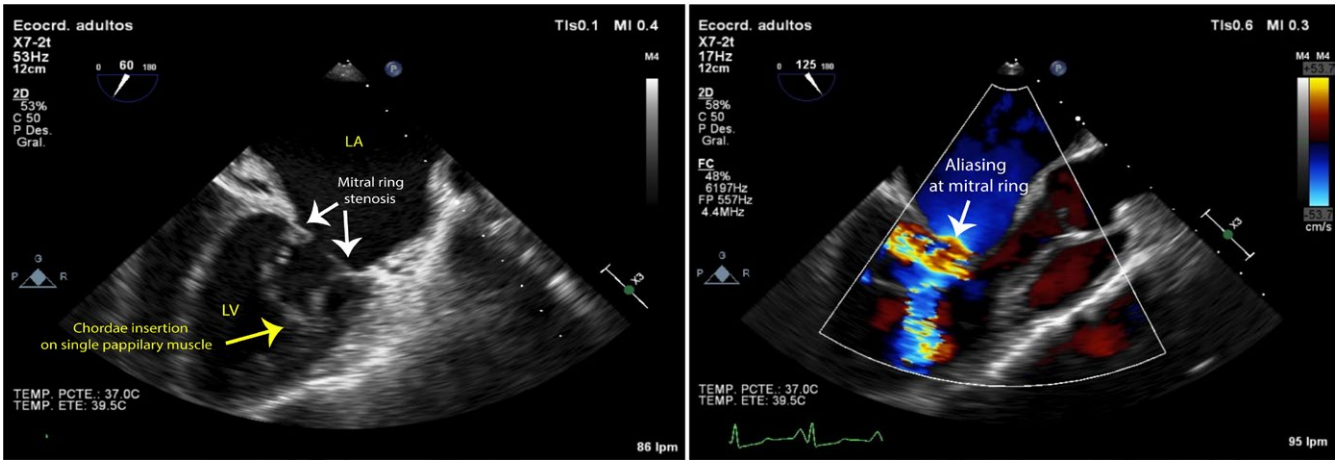

FIGURE 1 Long-axis view from two-dimensional transesophageal echocardiography (2DTEE). Left: White arrows indicate mitral ring stenosis protruding into the mitral orifice. Yellow arrow indicates thick, short chordae of the "parachute" mitral valve converging to insert on one major apical papillary muscle. Right: Turbulent transmitral flow at the mitral ring (white arrow) indicates that the membranous stenosis was obstructive. LA = left atrium; LV = left ventricle

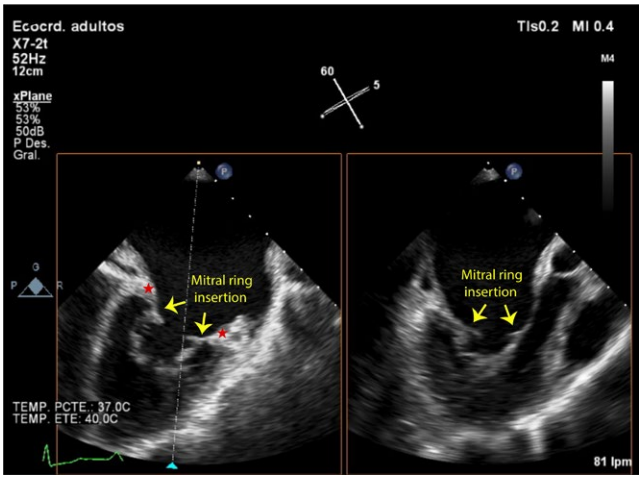

FIGURE 2 X-plane image from two-dimensional transesophageal echocardiography (2DTEE). Yellow arrows point out the mitral ring insertion just below the annulus (red stars) and suggest distal membranous insertion directly on the mitral leaflets

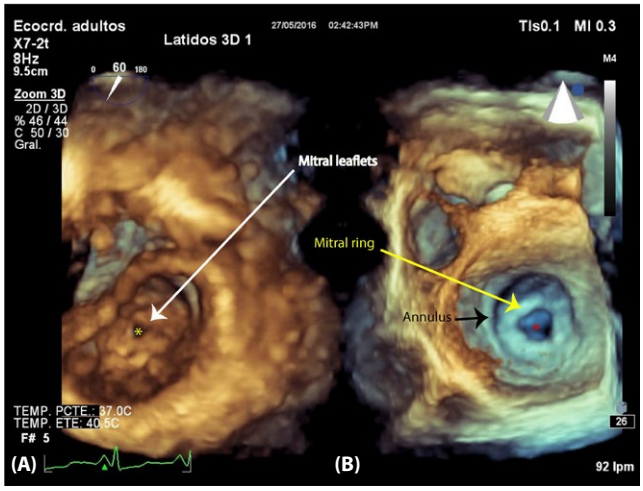

FIGURE 4 Three-dimensional transesophageal echocardiography (3DTEE) view of mitral valve from the left ventricle (A) showing the mitral leaflets (white arrow) and from the left atrium (B) showing the mitral ring (yellow arrow) and annulus (black arrow). The minimum orifice area of the mitral ring viewed from the left ventricle (yellow asterisk) was much smaller than from the left atrium (red asterisk)

directly on the leaflets, contributing to mitral stenosis (Figures 1A and 2, Movies S1 and S2). Color Doppler showed a transmitral flow with a mean gradient of 11 mm Hg (Figure 1B). Pulmonary artery systolic pressure was 39 mm Hg. Left atrium volumes were severely enlarged (57 mL/m^2). 3DTEE confirmed that the mitral ring had a proximal insertion beneath the mitral annulus and a distal insertion directly on the body of both valves (Figures 3 and 4, Movie S3). The area of the minimum orifice of the mitral ring was measured by 3D-zoom acquisition using multiplanar reconstruction. The proximal orifice area was 1.43 cm^2, while the distal orifice area was 0.81 cm^2, thus confirming a severe mitral stenosis (Figure 5). Neither subaortic stenosis nor aortic coarctation was present.

We consider that this could be an incomplete form of Shone's syndrome, in which only two of the four obstructive anomalies coexist. In Shone's original description, incomplete forms were the most common presentation, and the obstruction of the mitral valve was considered the main pathological feature.[1] In this patient, the membranous ring was located below the mitral annulus. A literature review found images very similar to ours, but in these cases, the mitral ring was described as a "supravalvular ring." Examples are Figure 5 in the original Shone et al. publication in 1963 and Figures 1 and 5 in the Popescu et al. article in 2008.[1,2] Particularly, Figure 5 in this last article shows the anatomical specimen of the heart of a patient with Shone's syndrome, in which the "supravalvular" mitral membranous ring is clearly below the annulus. With 3D echocardiography, this fact is evident without having to resort to anatomical specimens.

We consider that the term "valvar ring" proposed by Chauvaud et al.[3] is more appropriate to describe this structural anomaly for two reasons: First, because the annulus is the classic reference to establish which structures are part of the mitral valve[4]—and at least in this case, the ring is clearly below the annulus. Second, and more importantly, because the term "valvar ring" entertains the possibility that the ring may be directly attached to the valve. A similar case was presented by Cassano who described with great detail the anatomical specimen of a "so-called supravalvular ring" that was inserted on the anterior mitral valve.[5] This distinction has crucial therapeutic implications, as true supravalvular mitral rings can be corrected with a simple surgical resection, whereas the subannular mitral ring found in this case

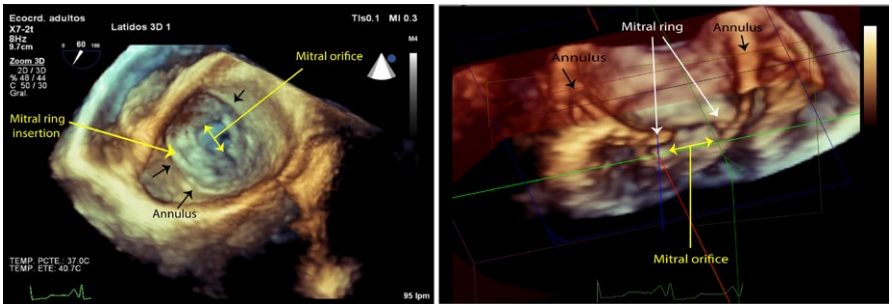

FIGURE 3 Three-dimensional transesophageal echocardiography (3DTEE). Left: Left atrial view of mitral valve. Yellow double-arrow marks the mitral orifice. Yellow single arrow indicates membranous mitral ring insertion beneath the mitral annulus (black arrows). 3DTEE confirmed that the location of the membranous ring was clearly below the annulus. Right: Sectioned view of mitral valve from the left atrium. Black arrows mark the position of the annulus. White arrows indicate mitral membrane insertion. Yellow double-arrow points out the mitral orifice

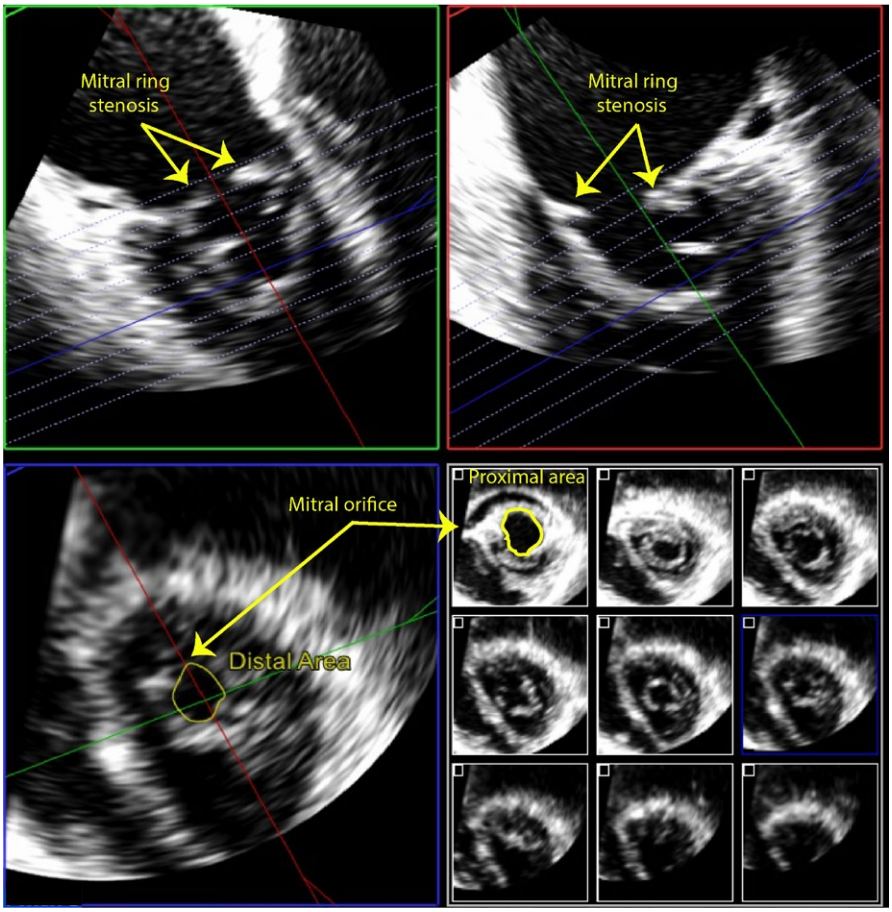

FIGURE 5 3D-zoom acquisition of the mitral valve using multiplanar reconstruction. The proximal orifice area was 1.43 cm^2, while the distal orifice area was 0.81 cm^2. Yellow arrows point out the location of the mitral ring

could require an attempt of plastic repair or, eventually, mitral valve replacement.

We conclude that at least some variants of Shone's syndrome may present with a membranous ring proximally inserted just below the mitral annulus and distally attached to the mitral valves. We believe that the term "valvar ring" is more appropriate than "supravalvular ring" and that 3D echocardiography is an excellent tool to evaluate these abnormalities, especially when considering a surgical scenario.

ORCID

María T. Politi http://orcid.org/0000-0002-4158-9080

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SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

Movie S1. Left parasternal long-axis view from two-dimensional transthoracic echocardiography (2DTTE). "Parachute" mitral valve with thick chordae inserting on a single apical papillary muscle.
Movie S2. Left atrial view of the mitral valve from three-dimensional transesophageal echocardiography (3DTEE). The membranous mitral ring was clearly below the plane of the mitral annulus.
Movie S3. Sectioned view of the mitral valve from the left atrium using three-dimensional transesophageal echocardiography (3DTEE). The membranous mitral ring was located between the mitral annulus and the valve leaflets.

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