

Unrecognized incomplete Shone's complex. Awareness is key in congenital heart disease

Katarzyna Elżbieta Gil¹, Christine Jellis², Brian Griffin²

¹The Ohio State University Division of Cardiovascular Medicine, Columbus, OH, United States

²Section of Cardiovascular Imaging, Heart and Vascular Institute, Cleveland Clinic, Cleveland, OH, United States

Correspondence to:

Katarzyna Elżbieta Gil, MD, PhD,
The Ohio State University Division
of Cardiovascular Medicine,
452 W 10th Ave, Columbus, 43210
OH, United States,
phone: +61 49 075 600,
e-mail:
katarzyna77.gil@gmail.com

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A 30-year-old woman presented with dyspnea with a background of remote aortic valve replacement for bicuspid aortic valve stenosis at an outside institution. A redo surgery with aortic root enlargement had been performed 8 years later at the same institution for reported patient-prosthetic mismatch. The intra-operative transesophageal echocardiography (TEE) showed restriction of the posterior mitral valve (MV) leaflet (mean gradient 8 mm Hg) with mild to moderate mitral regurgitation. The redo surgery had been complicated by complete heart block and treated with dual-chamber pacemaker implantation. The results of the follow-up transthoracic echocardiograms (TTE) were conflicting regarding the assessment of the MV pathology.

Our TTE performed a year after the redo surgery, demonstrated subvalvular narrowing of the left ventricular outflow tract (LVOT) with increased gradients on continuous wave Doppler, possibly related to LVOT obstruction or the small prosthesis size, and severe mitral stenosis with moderate mitral regurgitation. TEE showed a normally functioning size 21 mm aortic valve prosthesis. The MV was morphologically abnormal with a funnel shape and parachute-like anatomy in the context of a dominant anterolateral papillary muscle, to which the majority of chordae were attached (Figure 1A–C, arrows). An intra-mitral valve ring resulted in severe mitral stenosis (mean gradient, 19 mm Hg; Figure 1D, arrows). Left heart catheterization demonstrated a left ventricular intracavitary peak gradient of 50 mm Hg and a transmitral gradient of 22 mm Hg, but no significant gradient across the aortic valve prosthesis. There was no evidence of mitral or aortic valve calcifications on the

cardiac computed tomography. The patient was diagnosed with Shone's syndrome, based upon this constellation of findings, and underwent the Konno procedure to augment the size of the LVOT and aortic annulus along with mechanical aortic valve and ascending aorta replacements. Since the TEE in the operating room confirmed the pre-operative pathology of the MV with significant tethering of the subvalvular apparatus, the MV was deemed unreparable and was replaced with a mechanical prosthesis.

Shone's complex is a rare congenital disease consisting of a spectrum of left-sided obstructive lesions [1]. Complete Shone's complex can be diagnosed given the presence of four lesions of which the parachute MV, supramitral ring, subaortic membrane, and aortic coarctation are most common [1]. The more frequent incomplete Shone's complex warrants presence of at least two lesions [1]. Identification of one left-sided obstructive lesion should prompt a search for other anomalies [2]. Mitral valve obstruction is considered the earliest and most common pathological event in Shone's syndrome, causing underdevelopment of the left ventricle and thus leading to LVOT obstruction and aortic coarctation [1]. The severity of MV obstruction is the main predictor of long-term outcomes [1, 2]. Two types of mitral rings might be the cause of progressive mitral stenosis: the supramitral ring, originating just above the mitral annulus with no adherence to the leaflets and normal subvalvular apparatus, and the intra-mitral ring adherent to the leaflets with abnormal subvalvular apparatus [3]. The Doppler flow acceleration at the level of the ring might be the only finding on TTE [3]. Mitral stenosis

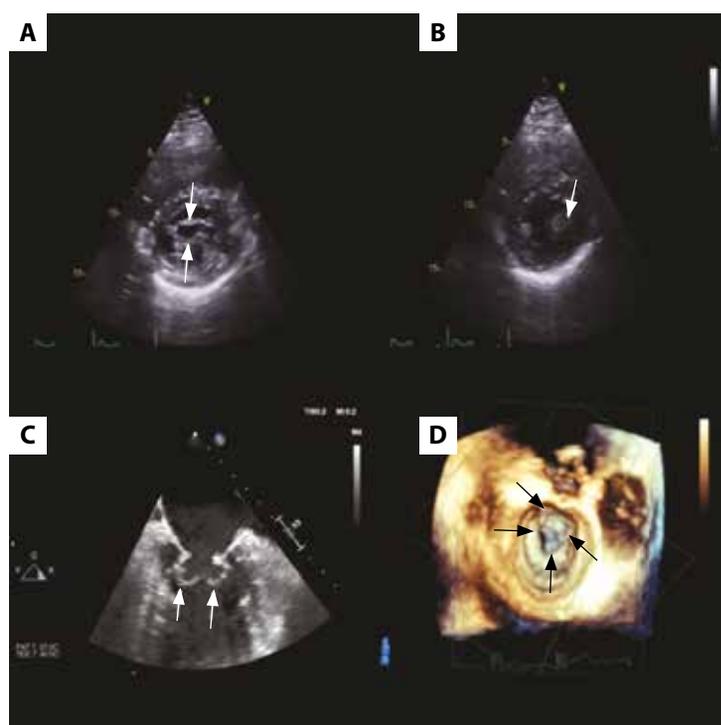


Figure 1. **A.** Mitral valve TTE; **B.** Papillary muscles TTE; **C.** Mitral valve TEE; **D.** Intra-mitral ring 3D TEE

Abbreviations: TEE, transesophageal echocardiography; TTE, transthoracic echocardiography

might also be caused by unequal distribution of chordal attachments in the parachute MV and its variant, parachute-like asymmetric MV [4].

Determination of appropriate management strategies in Shone's complex is challenging due to the variability in presentation and severity of individual lesions [2]. Repair, rather than replacement, of the mitral valve should be the first-line strategy when feasible [2]. Stenosis at the subvalvular, valvular, and supra-annular level can be relieved with the anterior aortoventriculoplasty, known as the Konno procedure [5].

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