

# Role of Perioperative Echocardiography in Repair of Incomplete Shone Complex: A Case Series

## Abstract

Multilevel obstruction of left-sided heart structures was originally characterized by Shone *et al.* The formulation of an appropriate operative strategy remains challenging and needs to be individualized for this complex subset of patients. Intraoperative transesophageal echocardiography (TEE) not only helps in delineating spatial anatomy but also reveals associated anomalies that help in decision-making regarding operative strategies for these patients. Here, we discuss five such cases of Shone's anomaly presenting at varied age group with different associated anomaly in which intraoperative TEE played a pivotal role in the management.

**Keywords:** Mitral stenosis, shone complex, supramitral ring, transesophageal echocardiography

## Introduction

Shone's complex is an uncommon congenital heart disease consisting of multiple level left-sided obstructive lesion.<sup>[1]</sup> A typical or complete Shone's complex consists of left-sided obstruction at four levels such as supra-avalvular mitral membrane, valvular mitral stenosis (MS) due to parachute mitral valve (MV), subaortic stenosis, and coarctation of the aorta (CoA).<sup>[2]</sup> When lesion involves with two or three levels of obstruction, it is called atypical or incomplete Shone's complex.<sup>[1,2]</sup> Perioperative transesophageal echocardiography (TEE) plays a crucial role in the diagnosis and management of such cases. We present a case series of five cases of incomplete shone complex with varied presentation, features, and management with written informed consent from the patients and their families. Demographic and clinical profile of these patients is summarized in Table 1.

## Case Reports

### Case 1

A 16-year-old male patient presented with Grade II dyspnea on exertion (DOE). Clinical examination revealed feeble pulses in both lower limbs, blood pressure of 146/100 mmHg in the right upper limb and

100/66 mmHg in lower limb and a heaving apex beat located at anterior axillary line in the 6<sup>th</sup> intercostal space. Auscultation of the chest revealed loud 1<sup>st</sup> heart sound, Grade III mid-diastolic murmur at the apex, and a Grade III ejection systolic murmur at the aortic area radiating to the carotid artery. Transthoracic echocardiogram showed multilevel left-sided obstructive lesions including a supramitral ring, subaortic membrane (SAM), and CoA with associated patent ductus arteriosus (PDA) and perimembranous ventricular septal defect (VSD).

In the operating room (OR), after instituting standard monitoring and induction of anesthesia using balanced anesthesia technique, invasive arterial lines were placed in the right upper limb and right femoral artery. The pressure gradient between the upper and lower limbs was 50 mmHg. TEE after induction of anesthesia using a 5-MHz phased array transesophageal transducer (6VT-D probe) of the GE vivid E9 echocardiography system (GE Medical Systems, Horten, Norway) confirmed transthoracic echocardiographic finding of supramitral ring with mean gradient of 10 mmHg across MV, SAM with left ventricular outflow tract (LVOT) peak gradient of 26 mmHg, 8-mm perimembranous VSD with left-to-right shunt [Figure 1], PDA with bidirectional shunt, and CoA with gradient of 40 mmHg. In addition, an

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aneurysm of sinus of Valsalva and left superior vena cava draining into coronary sinus was noted on TEE.

In view of the multiple defects, decision to perform surgery in two stages was taken to reduce the circulatory arrest time and its complications thereof. The first-stage procedure constituted PDA ligation and resection of CoA with end-to-end anastomosis of the descending thoracic aorta through left posterolateral thoracotomy. Total aortic cross-clamp time was 34 min. Postrepair gradient across the coarctation decreased to 30 mmHg and over 2–3 h, pressure became equal in upper and lower limbs. Following PDA ligation and CoA repair, the gradient across the MV decreased to 4 mmHg. On the follow-up after 3 months, the gradient across the coarctation further decreased to 19 mmHg. The second-stage procedure after 3 months was performed through median sternotomy under cardiopulmonary bypass (CPB) that involved excision of

supramitral ring and SAM with polytetrafluoroethylene patch closure of VSD and plication of sinus of Valsalva aneurysm. Postoperative, patient had an uneventful course and was discharged to home.

**Case 2**

A 2-year-old male child presented with recurrent lower respiratory tract infection. Preoperative TEE (9T-RS probe) of the GE vivid E9 echocardiography system revealed severe LVOT obstruction (peak systolic gradient = 86 mmHg and mean gradient = 50 mmHg) due to SAM 2 mm below aortic valve, no aortic regurgitation [Figure 2], normal MV leaflet movement with no commissural fusion, severe MS (peak gradient 15 mmHg and mean gradient 7 mmHg) due to supramitral ring, 7 mm PDA with left-to-right shunt, periductal CoA, enlarged left atrium (LA), and left ventricle (LV) hypertrophy with normal ejection fraction (EF = 60%–65%). Clear delineation of anatomy on TEE helped in deciding for a single-stage repair. Ligation of PDA and resection of CoA with end-to-end anastomosis of descending thoracic aorta through lateral thoracotomy followed by resection of SAM and supramitral ring under CPB through median sternotomy was done in single setting. Postoperative course was uneventful with no MS/mitral regurgitation (MR) and no LVOT obstruction or gradient across coarcted aortic segment.

**Case 3**

A 5-year-old girl diagnosed during infancy with incomplete shone was scheduled for elective intracardiac repair. She presented with Grade III DOE. Preoperative TEE

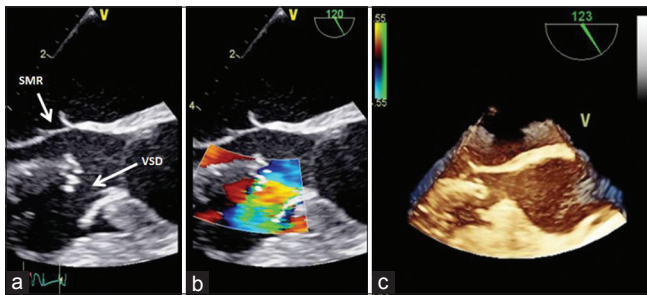


Figure 1: Transesophageal echocardiography: Mid-esophageal aortic valve long-axis view (a) two dimensional (b) color (c) three-dimensional showing supramitral ring and ventricular septal defect with left-to-right shunt

**Table 1: Demographic and clinical profile of the patients**

Variable	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age (years)	16	2	5	4	9
Gender	Male	Male	Female	Female	Male
Weight (kg)	42	10	13	10	24
BSA	1.24	1.58	0.6	0.5	1.3
Surgery time (min)	187 and 330*	480	450	160	430
Aortic cross clamp time (min)	147*	70	60	44	72
CPB time (min)	180*	105	87	68	115
PAH	Severe	Moderate	Severe	Moderate	Moderate
CoA	+ (Preductal)	+ (Preductal)	+ (Preductal)	+ (Postductal)	+ (Postductal)
SAM	+	+	–	–	
SMR	+	+	+	+	+
Mitral valve	Normal	Commissural fusion, moderate MR	Normal	Normal	Mild MR, single papillary muscle
Associated cardiac anomalies	PDA VSD SOVA LSVC	PDA	PDA Moderate TR Severe LV dysfunction	Mild TR	PDA VSD
Surgery	Staged	Single setting	Single setting	Single setting (CoA not repaired)	Single setting

\*Second-stage surgery. BSA: Body surface area, CoA: Coarctation of aorta, LSVC: Left superior vena cava, LV: Left ventricle, MR: Mitral regurgitation, PAH: Pulmonary arterial hypertension, PDA: Patent ductus arteriosus, SAM: Subaortic membrane, SMR: Supramitral ring, SOVA: Sinus of Valsalva aneurysm, TR: Tricuspid regurgitation, VSD: Ventricular septal defect

revealed severe MS due to supramitral ring, moderate MR, severe periductal CoA (peak gradient = 60 mmHg), large PDA (6–7 mm L→R shunt), severe pulmonary arterial hypertension (PAH), moderate tricuspid regurgitation (TR), and severe LV dysfunction (EF = 25%). Ligation of PDA and resection of coarctation with end-to-end anastomosis of descending thoracic aorta was done through lateral thoracotomy approach followed by resection supramitral ring under CPB was carried out through sternotomy in single setting. The postoperative patient had mild MR and mild TR with LVEF of 30%. The patient had no other complication and had an uneventful course postoperatively.

**Case 4**

A 4-year-old girl diagnosed with incomplete shone scheduled for elective intracardiac repair presented with Grade II DOE and history of recurrent respiratory tract infection. Preoperative TEE revealed severe MS (mean gradient of 30 mmHg) due to supramitral ring [Figure 2], normal mitral leaflet, periductal CoA (peak gradient = 50 mmHg), mild TR, moderate PAH (right ventricle systolic pressure = 45 + right atrial pressure), slightly enlarged LA and LV with normal LV function (EF 65%), and no PDA or SAM. Computerized tomography angiogram revealed long segment attenuation of aorta distal to left subclavian artery, dilated pulmonary arteries with distal pruning [Figure 3]. Preinduction TTE in OR showed low (12 mmHg) gradient across narrowest portion of the coarcted segment that

was subsequently confirmed on TEE after induction of anesthesia. He underwent excision of supramitral ring through median sternotomy through left atrial approach under CPB. Due to low-gradient across narrowed segment and young age of patient, coarctation was not addressed at this stage and patient was put on regular follow-up. On serial postoperative follow-up, TTE confirmed the low-gradient across the coarcted segment, no MS/MR, mild TR, and good biventricular function.

**Case 5**

A 9-year-old male child presented with Grade III DOE, Grade IV pansystolic murmur over parasternal area and holodiastolic murmur over mitral area. Preoperative TTE revealed severe MS (peak gradient 27 mmHg and mean gradient 11 mmHg) owing to supramitral ring, mild MR, single papillary muscle, PDA (3 mm L→R shunt), periductal CoA (peak gradient = 30 mmHg), 8 mm perimembranous VSD, mildly dilated LA/LV, and normal LV function (EF 55%). Intraoperative TEE revealed funnel-shaped MV with all chordae tendineae attached to only one of the two heads of the posteromedial papillary muscle. Three dimensional (3D) TEE revealed that supramitral membrane was encroaching on the leaflets of MVs [Figures 4 and 5]. The patient underwent resection and end-to-end anastomosis of coarctation segment of descending thoracic aorta along with the closure of PDA through lateral thoracotomy approach followed by excision

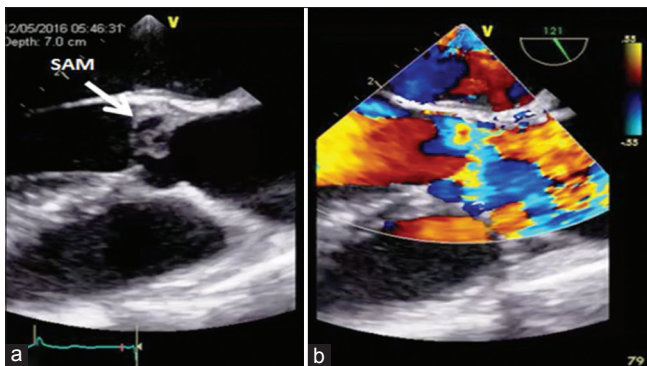


Figure 2: Transesophageal echocardiography: Mid-esophageal aortic valve long-axis view showing (a) subaortic membrane and (b) turbulent flow across left ventricular outflow tract due to subaortic membrane

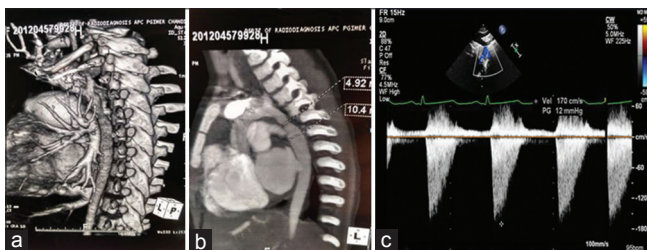


Figure 4: (a) 3D reconstruction of Computed tomography image showing (b) long attenuated segment of descending thoracic aorta with (c) maximum gradient of 12 mmHg across coarctation of aorta on continuous wave doppler in transthoracic echocardiography

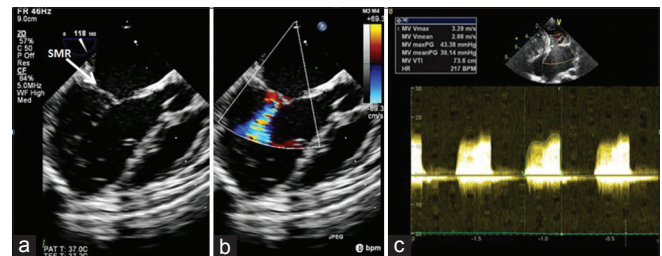


Figure 3: Transesophageal echocardiography: Mid-esophageal long-axis view showing (a) supramitral ring(SMR); (b) mitral stenosis in color flow and (c) severe mitral stenosis on continuous wave doppler

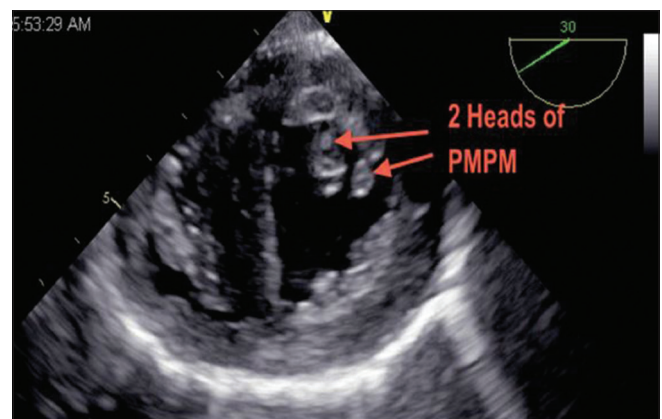


Figure 5: Transesophageal echocardiography: Transgastric midpapillary short-axis view showing single papillary muscle with two heads

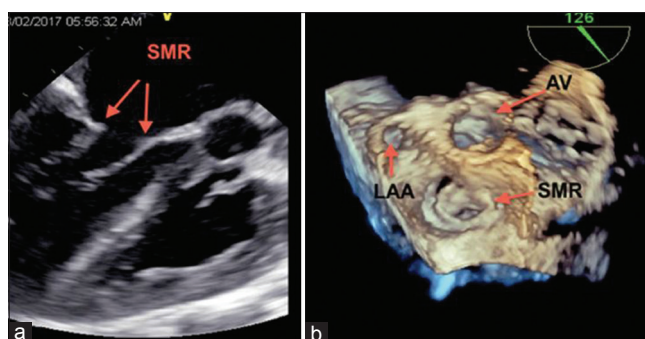


of supramitral ring, peeling off of the membrane from mitral leaflets, and closure of VSD under CPB. Postoperative echocardiography revealed no residual gradient across MV or residual flow through VSD.

## Discussion

The Shone's complex is a very rare entity, diagnosed most frequently in its incomplete form.<sup>[1]</sup> It encompasses broad anatomic and physiologic spectrum, ranging from mild lesions that may not require intervention to severe form that inevitably require surgery. Patients with Shone's syndrome have a poor long-term prognosis, with a perioperative mortality rate of 24%–27%.<sup>[3]</sup> A good outcome is possible when surgical intervention is undertaken early before the onset of pulmonary hypertension.<sup>[3]</sup> Shone *et al.* noted that MV obstruction appeared to be the most critical lesion. Patients with the most severe forms of mitral obstruction presented with severely elevated pulmonary artery pressure and had the poorest prognosis.<sup>[3–5]</sup>

MV morphologic assessment in Shone's anomaly is critical to determine the reconstructive surgical approach. Supravulvar mitral ring and parachute MV are the most prevalent variants of MS in this disease and are rarely amenable to balloon angioplasty.<sup>[2,4]</sup> Imaging of this lesion is challenging as one requires a high index of suspicion to diagnose it. The abnormalities of the area of the ring and MV are recognized from the mid-esophageal (ME) four-chamber (Ch) and ME two-Ch views. Alternate cross sections of the MV as provided by the ME MV commissural and ME long-axis (LAX) views [Figures 1, 3, and 6] adequately display the pathology. The supravulvar ring may be difficult to detect because it lies within the atrial surface of the valve leaflets, just above the MV annulus. 2D TEE LAX view shows small circumferential web attached to the atrial surface of the mitral leaflets [arrows in Figures 1, 3 and 6]. Color flow imaging may show the level of obstruction caused by the supravulvar mitral ring. 3D TEE en face view reveals the supravulvar mitral ring to be a circular membrane [Red arrow, Figure 6]. Parachute MVs can cause varying degrees of hemodynamic compromise depending on chordae development and annular size. In case of typical congenital, MS annular size is small, with thickened and rolled leaflet margins and there is decreased chordae tendineae interpapillary distance. In this variant, commissurotomy with separation of the chordae may postpone or eliminate the need for valve replacement.<sup>[6]</sup> Transgastric (TG) short-axis views (anteflexed, 0°) and TG two-Ch view (anteflexed, 90°) can delineate the subvalvular apparatus clearly showing the papillary muscles and the chordae attachments. Rarely, both papillary muscles are present with chordal attachments to one. Oosthoek *et al.* called this malformation “parachute-like asymmetric MV.”<sup>[7]</sup> Unlike others, we found supramitral ring as the most common cause of severe MS and not “typical” congenital MS of Shone's anomaly. In one of our cases,



**Figure 6:** Transesophageal echocardiography: Mid-esophageal long-axis view showing funnel-shaped mitral valve opening with supramitral ring encroaching on mitral valve leaflets (a) two-dimensional and (b) three-dimensional image

intraoperative TEE demonstrated a parachute MV with short-thickened chordae attached to the single posterior medial papillary muscle. The anterolateral papillary muscle was absent suggesting parachute-like asymmetric MV.

Identification of subaortic stenosis requires high index of suspicion. SAMs may be difficult to visualize on transthoracic echocardiogram. TEE generally offers better visualization of the subaortic anatomy and should be considered whenever there is high transvalvular velocity with normal aortic valve leaflets. The ME LAX view at 135° focused for LVOT provides adequate delineation of the subaortic obstruction [Figure 2] while the deep TG-LAX view allows evaluation of the pressure gradient. A possible subvalvular aortic ridge may not be noted in TTE as the image quality precludes definitive diagnosis. Multiplane TEE performed in such situation correctly identifies the precise location and cause of the gradient.

TEE can provide helpful 2D information about the location of the coarcted segment in the descending aorta. In the upper esophageal short-axis view at 0°, the coarctation will appear as a narrowing of the cross-sectional area with or without variable dilatation on either side. The descending aorta long-axis view at 90° could provide additional insight about the length and morphology of the coarctation. However, in either view, Doppler estimation of pressure gradient is difficult as the Doppler beam and the direction of the blood flow is not aligned.

Shone's syndrome rarely has associated perimembranous VSD and PDA.<sup>[8,9]</sup> CoA occurs in 20%–59% of cases with MV anomalies, whereas the supravulvar mitral ring is associated with other defects in almost 90% of cases.<sup>[2]</sup> Therefore, the finding of these defects should prompt for search of other cardiac anomalies. Our patients had perimembranous VSD and PDA in addition to CoA, SAM, and supramitral ring causing severe MS and severe pulmonary artery hypertension.

It is unusual for a patient to remain asymptomatic throughout childhood. Our cases had been asymptomatic in early childhood. The presence of PDA and VSD may have

contributed for the delay in presentation and worsening of the symptoms. Two-stage repair is still an effective technique for CoA associated with VSD, to diminish mortality, recoarctation rate, complications related to circulatory arrest, and allogenic blood transfusion.<sup>[10]</sup>

Recent advances in operative technique and the routine use of intraoperative TEE has significantly contributed to the success of valve reconstruction as the initial treatment for congenital MS. Reports suggest that late outcomes of valve repair are superior to replacement in patients with isolated congenital mitral anomalies.<sup>[6,7,11]</sup> Successful repair will permit continued annular growth and delay or eliminate the need for future valve replacement and lifelong anticoagulation.<sup>[6]</sup>

Operative outcome of patients diagnosed with Shone complex is excellent. Operative strategies for such patients should be individualized. Mitral interventions may generally be deferred for need of redo surgeries. Awareness of the atypical form of the complex is pivotal in appropriate management of the disease. Perioperative echocardiography plays a key role in diagnosis and management of such cases. In the index case series, TEE helped in the diagnosis of associated lesions such as sinus of valsalva aneurysm in the case 1, low-gradient across narrowest portion of the coarcted segment of aorta in the case 4, and demonstration of single papillary muscles with two heads and supramitral ring encroaching in the MV leaflet besides confirmation of preoperative TTE finding that helped in tailoring surgical treatment for the repair of this complex disease.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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