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A Rare Case of Shone Complex, Successfully Managed with Percutaneous Transcatheter Coarctoplasty and Balloon Aortic Valvuloplasty.

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Abstract:

Shone complex is an exceptionally rare and severe congenital heart condition characterized by left heart obstruction at various levels, including the supravalvular mitral ring, parachute mitral valve, subaortic stenosis, and coarctation of the aorta. In this case, we present a patient diagnosed with Shone complex who successfully underwent percutaneous transcatheter coarctoplasty and balloon aortic valvotomy. The interventional cardiologist's role is to accurately diagnose the condition and determine the appropriate treatment strategy for each individual patient, whether that involves surgery, balloon dilation, or stenting.

Keywords: Shone complex; percutaneous transcatheter coarctoplasty; balloon aortic valvotomy.

Introduction:

Shone syndrome, also known as Shone complex, is a rare congenital condition characterized by the presence of four left-sided obstructive or potentially obstructive defects. (Fig 1) These include a supravalvular mitral ring, parachute deformity of the mitral valve, subaortic stenosis, and coarctation of the aorta. The complete form of Shone syndrome is diagnosed when all four lesions are present. [1] Nonetheless, when there are fewer than four lesions, it is classified as the incomplete form. [2-4] It is described by Shone in 1963 [5].

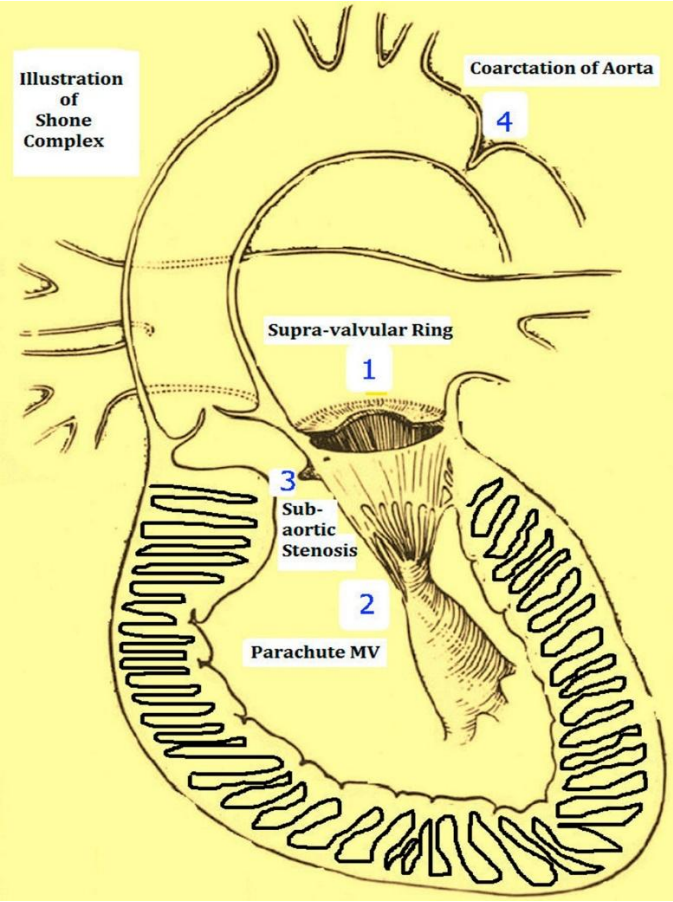

Figure 1: An illustration of Shone Complex showing the 4 defects with the direction of blood flow.

Case Report:

3-year-old developmentally normal child was admitted to the Cardiology Department as a case of Shone's complex. She had history of recurrent episodes of respiratory tract infection requiring hospitalisation. Upon examination, the primary complaints included shortness of breath and peri-oral cyanosis. The pulse rate in the upper limb was recorded at 130 beats per minute, while the blood pressure measured was 80/40 mm Hg in the right upper limb. A long ejection systolic murmur (graded III/VI) was detected in the aortic region and was conducted to the carotid arteries. Given these observations, a provisional diagnosis of coarctation of the aorta accompanied by left-sided obstructive valvular lesions in a case of Shone's complex was established.

From the laboratory examination, complete blood count was in normal limits. Echocardiography revealed normal ejection fraction (EF) of 65%, bicuspid aortic valve with severe stenosis, maximum velocity (Vmax) of 4.25 m/s, mean pressure gradient (P-mean) of 42 mm Hg, and left ventricular outflow tract area was 1.1 cm2 (Figures 2 and 3).

Mitral valve showed colour flow turbulence in mitral region with severe stenosis, P-mean was 12, and mitral valve area of 1.0 cm2 (Figures 4-5).

There was also a 3 mm PDA (Patent ductus arteriosus) with left to right shunt.

Further investigations, including cardiac catheterization, revealed the following.

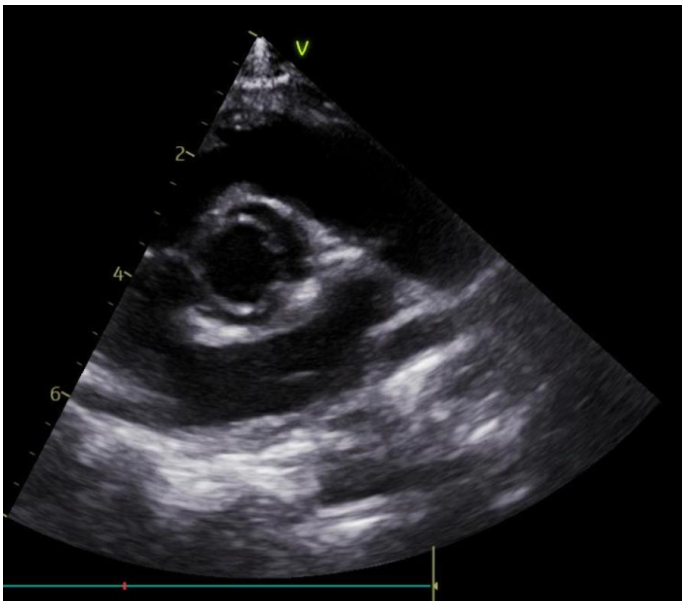

Figure 2: Two-dimensional transthoracic echocardiography demonstrates an anterior-posterior bicuspid aortic valve. (White arrow).

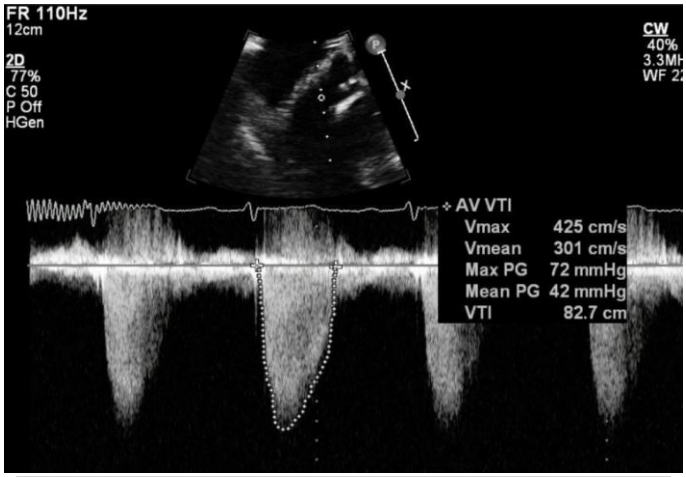

Figure 3: Doppler tracings from the patient, the aortic continuous wave (CW) recording shows a peak gradient of 72 mm Hg and a mean gradient of 42 mm Hg. These findings are consistent with severe aortic stenosis.

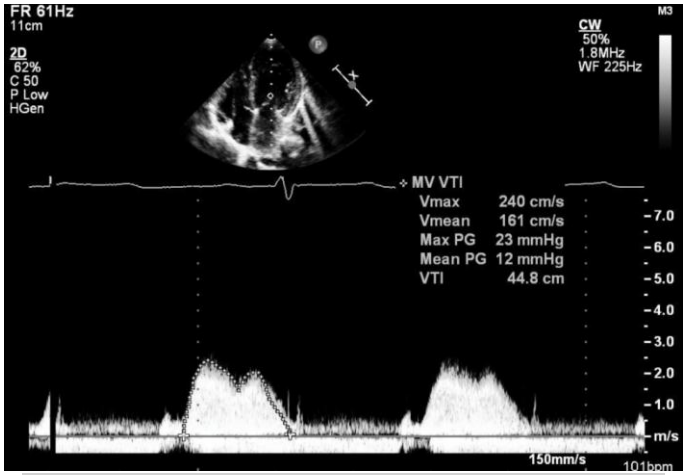

Figure 4: Continuous-wave Doppler echocardiogram across the stenotic mitral valve showed Vmax of 240 cm/s and mean Pressure Gradient of 12 mm Hg.

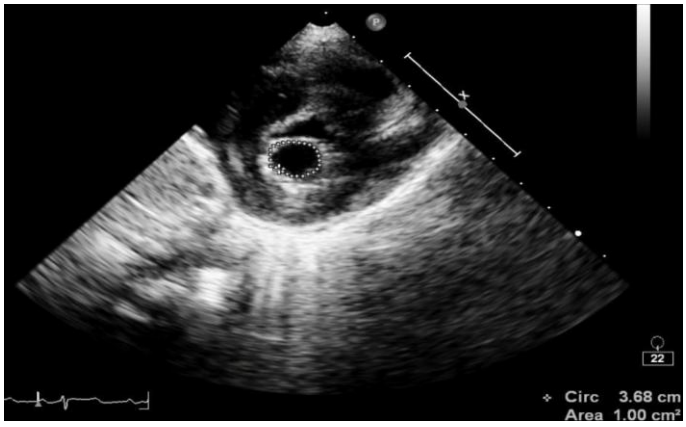

Figure 5: Parasternal Short axis (PSAX) view shows mitral valve area of 1.0 cm2.

Aortic root angiogram:

Doming aortic valve was noted in the LAO view. The arch was left sided with normal origin of arch vessels. Severe coarctation was noted just distal to the left subclavian artery origin. (Fig 6)

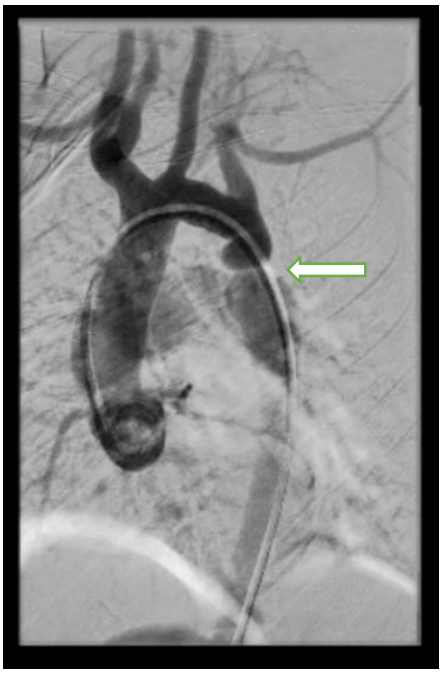

Figure 6: Aortic root angiogram showing severe coarctation just distal to the left subclavian artery origin. (White arrow).

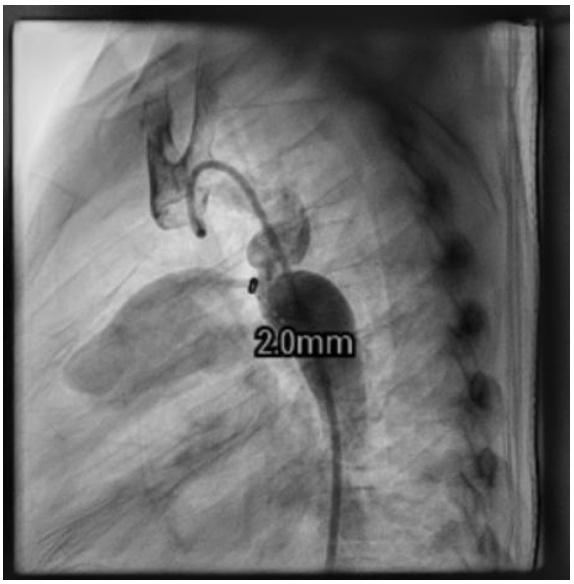

Figure 7: Arch angiogram showing a PDA measuring 2 mm at the coarct segment with filling of branch Pas (Pulmonary artery).

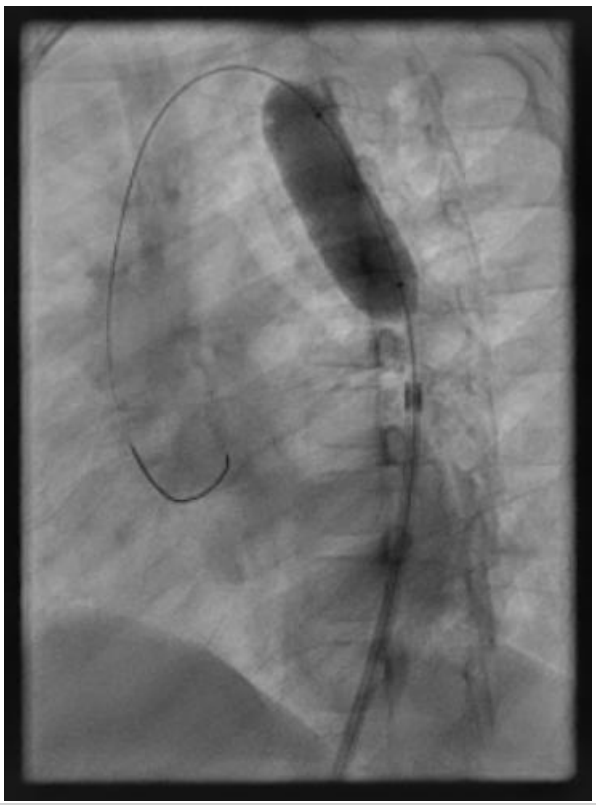

Figure 8: Dilatation of the coarct segment with Tyshak II 12x30mm balloon.

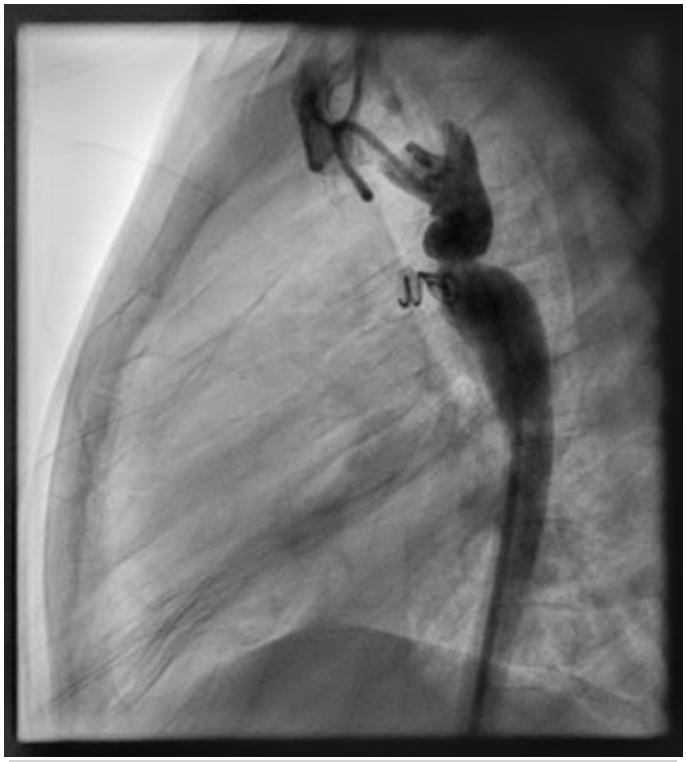

Figure 9: PDA Coil Closure (white arrow) with complete cessation of flow across the ductus.

Arch angiogram:

A PDA was noted at the coarct segment with filling of branch PAs (Pulmonary artery). PDA was measured to be 2 mm.

She underwent Balloon Aortic Valvotomy of bicuspid AV with (Aortic stenosis) with Coarctoplasty and PDA Coil Closure under general anaesthesia.

Procedure details:

Access:

RFA- 6F sheath

RFV- 6F sheath

Pressures	Pre procedure	Post procedure
Right Femoral Artery	75/42(58) mm Hg	
Descending Aorta	53/40 (47) mm Hg	69/49(63) mm Hg
Ascending Aorta	100/48(72) mm Hg	90/58(75) mm Hg
Coarctation Gradient	51 mm Hg	21 mm Hg
Left Ventricle-Aorta Gradient	16 mm Hg	11 mm Hg

Procedure:

The right femoral artery and venous sheath were placed. 1000 units of Heparin was administered intravenously. The aortic valve was crossed with 6F AL diagnostic catheter and 0.035" straight tipped Terumo glide wire (260 cm) and the AL catheter was placed in the LV. Then the Glide wire was replaced with a 0.018 Road runner wire. A 6 F long sheath was taken over the wire and placed in the ascending aorta. Dilatation of the aortic valve was carried out twice with a Tyshak II 12x30mm balloon over the 0.018 wire. The balloon was dilated until no waist was noted. Hence further dilatation was not carried out. The final residual gradient was reduced from 16 to 11 mm of Hg.

The Sheath was then pulled back to just distal to the coarct segment. The Tyshak II 12x30mm balloon was placed across the coarct segment and dilated up to 3 ATMs. The final residual gradient was reduced from 51 to 21 mm of Hg.

PDA was crossed from Aortic side using 4F JR diagnostic catheter and 018 Road runner wire. The JR diagnostic Catheter was taken over the road runner wire and placed distally in the MPA. A 5-5 PDA Coil was loaded on a TDS 110cm coil delivery system and was taken across the ductus through the JR catheter.2 Coils were deployed on the PA side,2 coils deployed in the ductus and remaining 1 coil deployed in Aortic side. Angio done later showed complete cessation of flow across the ductus.

Patient remained hemodynamically stable. Echocardiogram showed a well opened bicuspid aortic valve, with Mild AR, the mean gradient across mitral valve was noted to be 4 mmHg. Coarctation segment was well open and no residual flow through the ductus was noted. There was no pericardial effusion. Sheath was removed in lab and manual compression given and then the patient was shifted to the ward in a hemodynamically stable condition.

On follow-up, the patient reported great improvement in her symptoms, and she had no other complaints. Her follow up echo showed mild residual aortic stenosis, mean gradient of 12mmHg with no Aortic regurgitation with improvement of coarctation gradient and PDA coil seen in situ with tiny residual ductal flows.

Discussion:

Initially identified in 1963, Shone's anomaly is recognized as a rare congenital condition characterized by left-sided obstructive lesions. Key features include a parachute mitral valve and a muscular or membranous supravalvular ring in the left atrium. Together, these two characteristics encapsulate the classic presentation of the condition. [6]

A parachute mitral valve is characterized by having two leaflets and two commissures, where all chordae attach to a single papillary muscle rather than two. The supra-valvular ring of the left atrium refers to a connective tissue structure that originates at the base of the mitral valve leaflets on the atrial surface and extends into the valve's opening. Additionally, the third and fourth observations are subaortic stenosis and aortic coarctation. [6]

Shone's complex typically manifests early on. A retrospective study revealed that the diagnosis of this condition was made at a median age of 14 days. [7]

A different retrospective cohort study found that synchronous connections (SC) is frequently not recognized as a diagnostic condition, with a prevalence rate of 0.7% in a sample of 4,000 patients diagnosed

with congenital heart disease (CHD). The incomplete form of SC was identified more often than the complete form. Complications were primarily associated with arrhythmias, heart failure, and surgical interventions, resulting in elevated rates of cardiac hospitalizations. Among adult patients, there was a notable incidence of reinterventions for mitral valve, aortic valve, and left ventricular outflow tract issues. Nevertheless, the mortality rate among patients who reached adulthood remained relatively low.

SC poses a diagnostic dilemma because of its diverse clinical presentations. Transthoracic echocardiography (TTE) is usually the first imaging technique used, offering an extensive evaluation of left ventricular function and notable valvular defects. Additionally, TTE facilitates the identification of related conditions such as subaortic stenosis, supramitral ring, and coarctation of the aorta (CoA). In instances of suspected or confirmed coarctation of the aorta (CoA), cross-sectional imaging using cardiac magnetic resonance (CMR) or cardiac computed tomography (CCT) is often advised. This imaging technique facilitates the accurate determination of the coarctation's location, length, and diameter, which helps in deciding on the most suitable interventions. [8]

Handling this group of patients presents difficulties owing to the diverse ways in which they present and the varying severity of each individual heart defect. Recent research has underscored the significance of assessing the severity of left ventricular inflow obstruction when strategizing surgical interventions and forecasting patient outcomes.

While earlier literature has limited case reports on the Shone complex, the current case emphasizes the success of coarctoplasty. Potential complications from simple balloon angioplasty may involve a residual gradient that surpasses 20 mm Hg in 14% of instances, while recurrent stenosis after an initially successful angioplasty is uncommon. Additionally, aneurysm formation occurs in approximately 5% of patients. [9] Although complications like residual gradient, recoarctation, and aneurysms are less frequent, they may necessitate redilatation in growing children. [10]

Conclusion:

Shone's complex is an uncommon congenital heart condition characterized by obstructive lesions on the left side of the heart. A thorough clinical examination, along with echocardiography, radiological tests such as CT and MRI scans, and catheterization, can accurately diagnose and assess the severity of the condition. The components of Shone's complex include aortic coarctation and subaortic stenosis. It can be effectively treated with percutaneous transcatheter coarctoplasty and balloon aortic valvotomy.

Ethics approval: The patient's father provided written informed consent to be published as a case report.

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Competing interests: No conflict of interest is to be declared by the authors.

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