



## Acute Pulmonary Edema in a Young Patient with Shone's Complex

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### Abstract

Shone's complex is characterized by multiple left-sided heart anomalies leading to blood flow blockages. Misdiagnoses are common, making treatment decisions challenging. Transthoracic echocardiogram with Doppler can be implemented for the diagnosis. The treatment is based on surgical correction, and the prognosis depends on the severity of the condition and the timing of intervention. We present a case of a patient with Shone's complex who experienced acute pulmonary edema as a complication. Surgical correction was performed, but the patient was lost in follow-up, he later presented to the emergency service with acute dyspnea and palpitations. Further surgical intervention was undertaken.

### Introduction

Congenital Aortic Valve Stenosis (AVS) is characterized by blood flow obstruction from the left ventricle to the aorta and is dependent on the morphological characteristics of the valve [1]. The prevalence of bicuspid aortic valves ranges from 0.5% to 2%, making it the most common congenital heart disease; it is often related to other left-sided congenital lesions [2].

Congenital Mitral Stenosis (MS) is a condition where the left ventricular inflow is obstructed due to a constriction in the mitral valve orifice [3]. There are four types: typical, hypoplastic, supramitral ring, and parachute mitral valve. Typical MS is the most common, characterized by thickened leaflets, chordae tendineae, and underdeveloped papillary muscles. Parachute mitral valve is the least common, with thickened and shortened chordae tendineae inserted in a single papillary muscle [4].

Supravalvular mitral membranes are abnormal connective tissue ridges that cause mitral inflow obstruction. Two types of supravalvular rings have been identified: supramitral (above the mitral annulus) and intramitral (within the mitral tunnel) [5]. This results in increased left atrial and pulmonary venous pressure, leading to pulmonary edema. Although difficult to detect through routine echocardiography, Doppler flow acceleration before the mitral valve annulus can be observed [6].

Coarctation of the Aorta (CoA) is a localized narrowing in the aortic arch, typically distal to the left subclavian artery. This results in an increased pressure load on the left ventricle during systole, producing left ventricular hypertrophy; it also leads to decreased blood flow distal to the obstruction, resulting in reduced perfusion of the lower extremities. Complications include: Headache, epistaxis, brachiocephalic hypertension; shock and multiorgan failure can present with ductus arteriosus closure. The increased workload can eventually lead to heart failure [7]. There is significant disparity in systolic and diastolic blood pressures, with measurements being increased in upper extremities and decreased in lower extremities [8].

Shone's complex comprises a series of obstructive anomalies of the left heart and aorta. It involves a supraventricular ring, parachute mitral valve, subaortic stenosis, and CoA (Figure 1). Patients typically present with congestive heart failure, failure to thrive, diaphoresis, dyspnea, fatigue, and chest pain. Physical examination reveals a harsh pansystolic murmur. Additional defects like bicuspid aortic or pulmonary valves and endocardial sclerosis can be found. Patients typically have lower median arterial pressure in the lower limbs and may exhibit left precordial bulging, tachycardia, and cyanosis. Chest X-rays reveal accentuated pulmonary vascular markings

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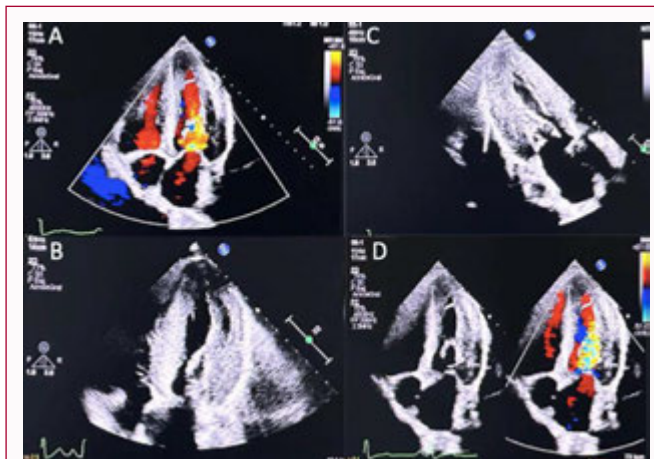
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and electrocardiograms biventricular growth and right axis deviation [9,10]. Diagnosis is through Doppler transthoracic echocardiogram. Patients have better outcomes if correction of obstructive anomalies is achieved before the development of pulmonary hypertension [9].

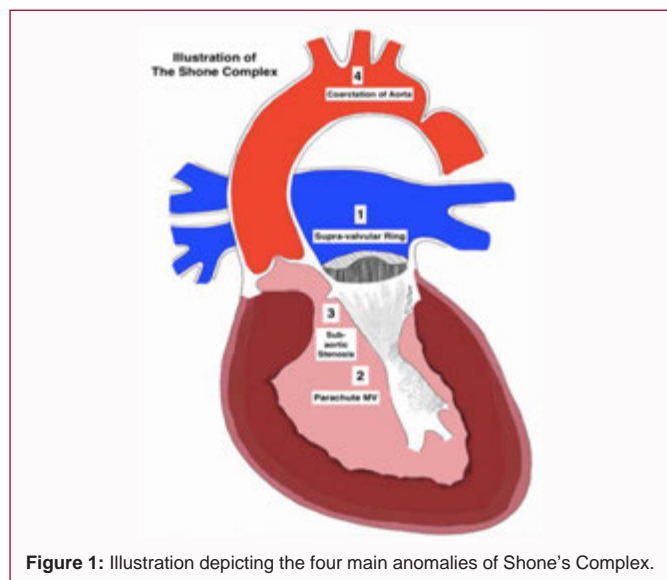
**Case Presentation**

The patient initially presented to the National Institute of Cardiology (NIC) in 2013 for follow-up of an unspecified heart murmur diagnosed at 7 years old. After initial workup, the following diagnosis were established: MS (Parachute Mitral Valve [PMV], supra-valvular mitral membrane, thickened anterior valve leaflet), aortic stenosis (sub-valvular aortic stenosis, Bicuspid Aortic Valve [BAV], hypoplastic ring), CoA, hypoplastic transverse arch, Patent Ductus Arteriosus (PDA) and concentric hypertrophy of both ventricles.

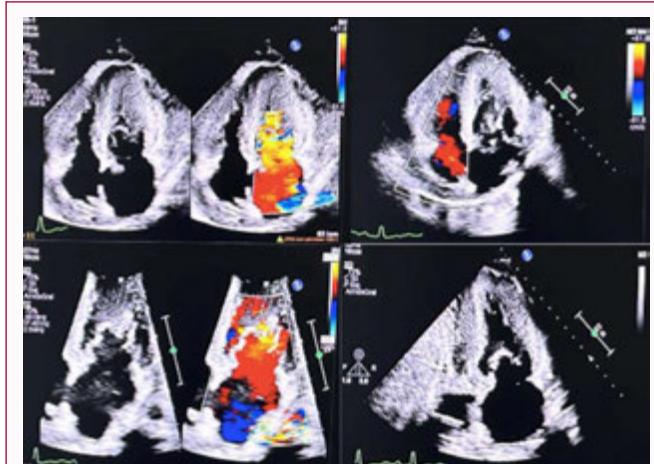
Surgical correction was undertaken with correction of the CoA with end-to-end anastomosis and surgical closure of PDA; the patient would be shortly re-evaluated to determine a plan to correct the remaining heart defects. Out-patient follow-up was considered; however, the patient’s last check-up was performed in 2014.



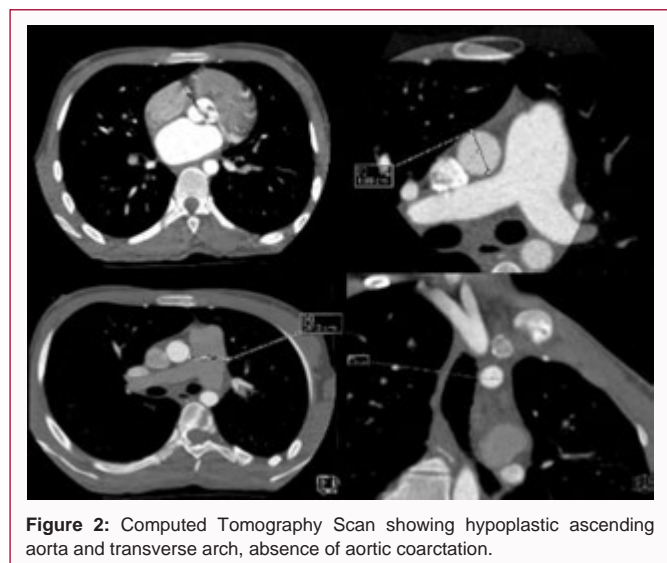
**Figure 3:** Preoperative Transthoracic Echocardiogram (TTE). A) TTE showing mild to moderate mitral regurgitation with a small central jet. B) TTE showing thickened bicuspid aortic valve, with presence of sub-valvular aortic membrane. C) TTE showing concentric hypertrophy of the left ventricle. D) TTE showing dysplastic parachute mitral valve, with supra-valvular mitral membrane.



**Figure 1:** Illustration depicting the four main anomalies of Shone's Complex.



**Figure 4:** Postoperative Transthoracic Echocardiogram (TTE) showing left ventricle with concentric hypertrophy, normally functioning mechanical prosthesis with adequate mobility of its discs without paravalvular leaks, right ventricle with dynamic obstruction in the outflow tract, and biventricular diastolic dysfunction.



**Figure 2:** Computed Tomography Scan showing hypoplastic ascending aorta and transverse arch, absence of aortic coarctation.

The patient presented to the emergency service of the NIC in November 2022 due to acute dyspnea, oxygen saturation of 65% which partially improved with the administration of oxygen, cough with salmon-colored sputum and palpitations. A chest X-ray was performed, observing bilateral opacities in the upper lobes. Subsequently, a CT scan was performed, where an absence of coarctation and hypoplastic arch was observed, and a control echocardiogram was also performed (Figure 2, 3). The patient was admitted and treated for heart failure while surgical correction was considered.

During his stay, the following procedures were performed: Transarterial and transaortic approach, mitral valve repair, supra-valvular mitral membrane resection, papillary muscles fenestrations, aortic ring enlargement (Manougian technique) and aortic valve replacement (prosthetic valve). The patient presented atrial fibrillation during surgery. Control echocardiogram was performed (Figure 4).

The first postsurgical hours presented with hemodynamic instability despite the use of vasopressors, the patient required multiple blood transfusions and the diagnosis of mixed shock (hemorrhagic, cardiogenic) was established. Greater than expected bleeding prompted re-exploration of the mediastinum finding hemopericardium, drainage was realized as well as hemostasis of multiple bleeding points. Following hemopericardium drainage the patient presented adequate hemodynamic stability to allow discharge from the hospital, with plans for posterior surgical intervention.

## Discussion

This patient was diagnosed in childhood with findings compatible with Shone's complex, including MS, aortic stenosis, CoA, hypoplastic transverse arch, PDA, and concentric hypertrophy of both ventricles. However, most patients present only 2 to 3 of these anomalies alongside other heart defects such as PDA, bicuspid aortic valve, atrial and ventricular septal defects [11]. Mitral valve obstruction is considered the earliest malformation in the heart, thus leading to other consequent left-side obstructive events [12]. MYH6 mutations have been found in 11% of patients with Shone's complex and are associated with other conditions such as atrial septal defects, ventricular septal defects, and cardiomyopathies [13].

The condition is often misdiagnosed or underdiagnosed due to its wide range of clinical presentation and unspecific symptoms. This issues a significant challenge when it comes to determining the appropriate approach and management of these patients [9]. Surgical correction is necessary, and prognosis is dependent on the patient's severity and moment of intervention. Shone's complex is associated with multiple morbidities including atrial flutter, aortic dissection, pulmonary edema and presents an important perioperative mortality rate [11].

Surgical corrections such as biventricular repair are associated with good postoperative results and long-term survival in patients with Shone's complex [14]. This case highlights the importance of preventing complications through an early diagnosis and surgical treatment. The responsibility of patient education is of critical importance in contributing to adequate patient follow-up and falls upon the physician's shoulders.

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