

## **A Rare Case of Shone Complex, Where Coarctation was Treated Successfully with Covered Cheatham Platinum Stent**

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### **Authors' contributions**

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

### **Article Information**

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**Case Study**

### **ABSTRACT**

Shone complex is an extremely rare and severe congenital heart disease characterized by left - heart obstruction at multiple levels namely supra-avalvular mitral ring, parachute mitral valve, subaortic stenosis and coarctation of the aorta. We in the present case describe a patient of shone complex who underwent successful coarctoplasty with the help of BIB (balloon in balloon) dilatation catheter and covered CP (cheatham platinum) stent. The role of interventional cardiologist is to diagnose and choose the right approach for the specific patient, whether surgery, balloon or stent.

**Keywords:** *Shone complex; BIB (Balloon in Balloon) dilatation catheter and covered CP (Cheatham Platinum) stent.*

### **1. INTRODUCTION**

Shone's complex is an extremely rare and severe congenital heart disease with about only

fifty cases reported in the literature. It involves congenital left-heart obstructions at multiple levels. First described in 1963, it is a rare congenital heart disease consisting of a

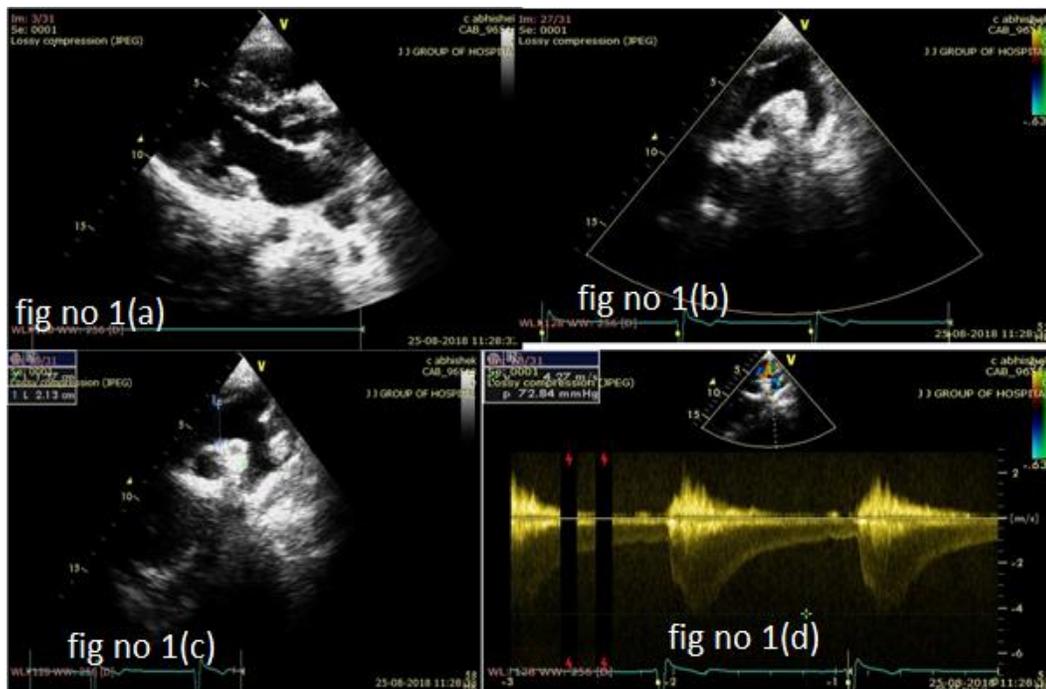
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supravalvular mitral ring, parachute mitral valve, subaortic stenosis, and coarctation of the aorta. When only two or three of these components are present, it is labelled as the incomplete form of Shone's complex. In the present case, we describe a patient of incomplete shone complex who had parachute mitral valve, bicuspid aortic valve, and coarctation of the aorta and underwent successful aortic stenting using BIB Balloon and covered CP stent.

## 2. CASE REPORT

A 15-year adolescent boy referred from primary physician to us as young hypertensive with a complaint of an intermittent headache. On examination, the patient had feeble lower limb pulses (femoral, popliteal and tibial) as compared to upper limb with significantly appreciable radio-femoral delay. Blood pressure of 200/100 mm of Hg was recorded in upper limb and 100/70 mm of Hg was recorded in lower limbs. Systolic murmur of grade 3/6 that was best heard in the aortic area with no associated thrill but there was radiation to carotid and back. ECG (electrocardiogram) was suggestive of left ventricular hypertrophy by voltage criteria. X-ray chest P/A view was suggestive of rib notching in

3rd and 4th ribs with a three sign. Echocardiography (Fig. 1) showed left ventricular concentric hypertrophy and a gradient of 64 mm of hg gradient across descending thoracic aorta with constriction of aorta distal to left subclavian artery (post ductal type) with associated findings (Fig. 2) of a bicuspid aortic valve, mild aortic stenosis and parachute mitral valve which helped us to reach the diagnosis of a shone complex. CT Aortogram further confirmed the diagnosis which showed the reduced calibre of descending thoracic aorta measuring 4.8 mm at about 14 mm distal to left subclavian artery with prominent collaterals from bilateral subclavian artery, internal mammary artery and intercostal arteries. Catheterization done via right femoral and radial route was suggestive of thin segment aortic narrowing (Fig. 3) with the gradient of 120 mm of Hg. A JR (Judkins right) catheter 6f (French) along with 0.035 wire (straight tip) was passed across the coarct segment followed by replacement with Amptzer super stiff wire and finally stenting with covered CP stent 4.5 cm which was deployed with 14x10 mm BIB balloon. Final result thus obtained is shown in Fig. 3 (f) and both pre and post BP procedure pressures in Table 1.



**Fig. 1(a).** Left upper panel is showing left ventricular hypertrophy, Fig. 1(b) and 1(c) upper right and lower left panel shows coarctation and Fig. 1(d) lower right panel shows gradient of about 64 mm of hg across the coarctation segment with continuous flow

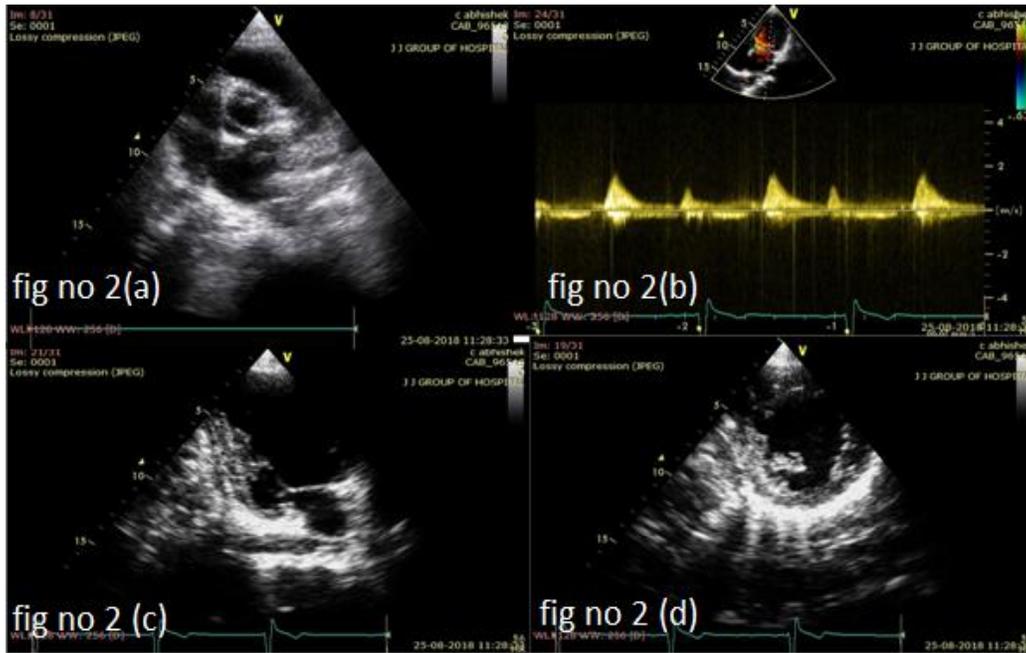


Fig. 2(a). Upper left panel shows short axis view showing bicuspid aortic valve, Fig. 2(b) upper right panel shows mild mitral stenosis and lower panels Fig. 2(c) and (d) are showing single papillary muscle i.e. parachute mitral valve in PLAX and short axis views

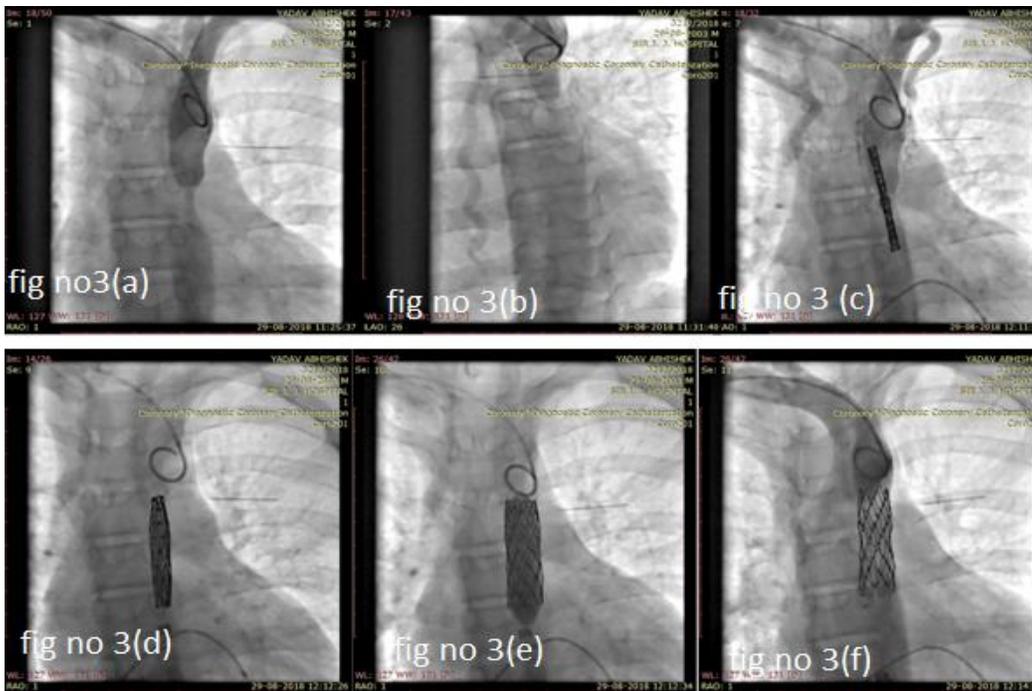


Fig. 3(a). Pigtail shoot (6f) in arch shows long segment narrowing suggestive of coarctation of aorta. Fig. 3(b) shows large collaterals due to coarctation. Fig. 3(c) Shows placement of CP stent across coarctated segment. Fig. 3(d) and (e) showing Expansion of BIB balloon inner followed by outer balloon Fig. 3(f). Final result

**Table 1. Showing pre and post coarctoplasty radial artery and femoral artery pressures**

	<b>Pre stenting</b>	<b>Post stenting</b>
Radial artery	224/86	150/80
Femoral artery	98/78	120/76
Gradient between above and below the coarctation segment	120(Peak)	10

### 3. DISCUSSION

Shone's complex is an extremely rare and severe congenital heart disease. It typically consists of four obstructive lesions of the left side of the heart and circulation namely parachute-like mitral valve, supravulvar mitral ring, subaortic stenosis, and coarctation of the aorta. Complete as well as incomplete forms of shones complex have been described wherein all four lesions or two to three lesions are present respectively [1]. In the present case, we described a case of Incomplete shone complex with coarctation of the aorta, a bicuspid aortic valve with mild aortic stenosis and parachute mitral valve. The LVOT obstruction in shone complex features may include subaortic stenosis, valvar aortic stenosis, bicuspid aortic valve, and coarctation of aorta [2]. A supravulvar mitral ring is a circumferential ridge or membrane, may be thin or thick which may arise from left atrial wall overlying the mitral valve besides its adhesion to the valve may impair opening of the leaflets causing mitral-valve inflow obstruction in some patients [3]. Parachute mitral valve is defined as a unifocal attachment of mitral valve chordae independent of the number of papillary muscles. A true parachute mitral valve (PMV) is characterized by attachment of the chordae to a single or fused papillary muscle The unifocal attachment of the chordae results in a restricted valve opening and sub valvular obstruction and rarely valvular regurgitation [4].

There are few case reports from earlier literature [5,6] regarding the Shone complex but the present case also highlights successful coarctoplasty using Covered CP stent and BIB balloon. BIB balloon dilatation catheter (NuMed, Hopkinton, NY) having two balloons one shorter (inner) and one longer (outer). The inner balloon inflates the centre of the stent and outer one is for proper positioning of the stent. CP stent (developed by NuMed Hopkinton, New York) is made from heat-tempered 90% platinum and 10% Iridium .01 inch wire arranged in a zig pattern [7].

Complications during the plain balloon angioplasty may include residual gradient exceeding 20 mm of Hg in 14% of cases, recurrent stenosis after initial successful angioplasty is rare and aneurysm formation in about 5% of the patients [8] however Balloon expandable stents provide effective therapy in the form of endovascular buttress to support the dilated segment. Complications such as residual gradient, recoarctation and aneurysms are much less as compared to plain balloon angioplasty but may require redilatation in growing children [8,9,10].

### 4. CONCLUSION

Shone complex is rare congenital heart disease comprising of obstructive lesions of left side of the heart. With the help of proper clinical examination, echocardiography, radiological investigation like CT scans and MRI scans and catheterization diagnosis can be established and quantified for the severity. Coarctation of aorta which is one the component of shones complex may present as asymptomatic hypertension with radio femoral delay. It can be effectively treated with percutaneous transcatheter coarctoplasty with the BIB balloon and covered CP stent.

### CONSENT

It is not applicable.

### ETHICAL APPROVAL

It is not applicable.

### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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