

# Hypoplastic left heart syndrome with coronary-cameral fistulas: Echocardiographic demonstration of coronary artery steal and successful interventional treatment

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## ABSTRACT

**We report a case of a patient with hypoplastic left heart syndrome with significant coronary-cameral fistulas and exertional symptoms from coronary steal. Symptoms resolved following successful coil occlusion of his left ventricle.**

**Keywords:** Coronary artery fistulas, coronary steal, hypoplastic left heart syndrome

## CLINICAL SUMMARY

A 9-year-old male patient with hypoplastic left heart syndrome (HLHS), (mitral stenosis and aortic atresia) who had successfully completed three staged palliation presented 6 years following Fontan palliation with new-onset exertional symptoms characterized by chest pain and syncope during exercise. An exercise stress test demonstrated ST-segment depression in the inferolateral leads [Figure 1]. These findings corresponded clinically with arterial desaturation related to low cardiac output, hypotension, chest pain, and dizziness.

To evaluate his coronary arterial circulation, cardiac catheterization with coronary angiography and transesophageal echocardiogram (TEE) was performed. By angiography, there were multiple and prominent fistulous communications between his left coronary artery system and the hypoplastic left ventricle (LV) [Figure 2]. His right coronary artery (RCA) was normal with no evidence of fistulous communications to the LV or left coronary system. His LV systolic pressure was 258 mmHg with an end-diastolic pressure (EDP) of 15 mmHg, with mild mitral regurgitation. The simultaneous right

ventricular pressure was 74 (systolic)/10 (EDP) mmHg. His systemic blood pressure was 80/51 mmHg. His baseline TEE demonstrated antegrade diastolic flow across a dilated RCA during diastole into his systemic right ventricle [Figure 3]. Isoprenaline was then infused to simulate exercise. With drug-induced chronotropic and inotropic response, there was retrograde flow from the RCA by Doppler interrogation during systole [Figure 4]. These findings suggested that diastolic runoff into the fistulas created a steal phenomenon from the RCA and was responsible for both his clinical symptoms as well as the electrocardiographic ST segment changes in the distribution of the RCA.

Given the hemodynamic significance of these fistulas, transcatheter coil embolization of the LV was performed using several coils to completely obliterate the LV cavity. Access to the LV was obtained through the Fontan baffle. A renal double curve catheter was advanced into the LV across the mitral valve to perform a left ventricular angiogram. A 2.7-Fr Progreat catheter (Terumo Interventional systems, New Jersey, USA) was used

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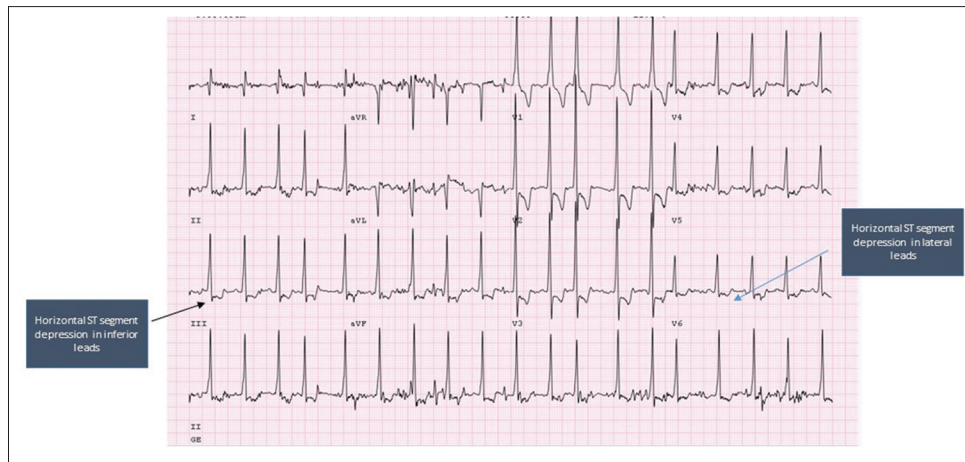


Figure 1: 12-lead electrocardiogram during exercise stress test showing evidence of horizontal ST-segment depression in inferior leads and lateral leads during peak exercise



Figure 2: An angiogram in the native ascending aorta shows dilated left coronary system with several small fistulous communications (arrowheads) from the circumflex coronary artery into the hypoplastic left ventricle which fills because of the fistulas communications

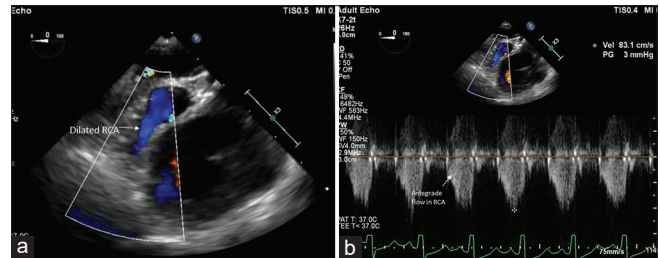


Figure 3: (a and b) Transesophageal echocardiogram still frame shows dilated right coronary artery system with antegrade blood flow during diastole (blue), spectral Doppler imaging to confirm antegrade blood flow in diastole



Figure 5: A repeat angiogram in the neo-aorta shows minimal flow into the hypoplastic left ventricle after coil occlusion of these fistulas

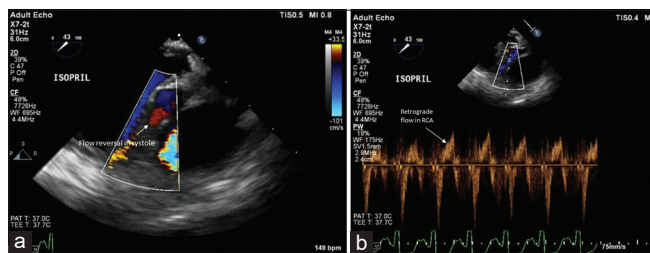
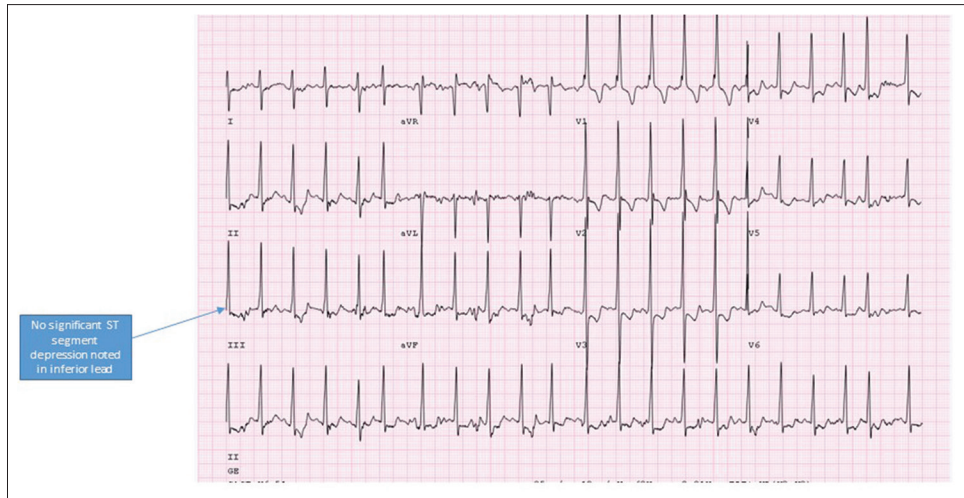


Figure 4: (a and b) Transesophageal echocardiogram still frame at elevated heart rate of around 150 bpm now shows that there is evidence of retrograde flow in systole in proximal right coronary artery by color flow mapping and with spectral Doppler imaging. Also note that there is evidence of decreased antegrade blood flow across right coronary artery during diastole using pulse wave Doppler, electrocardiogram tracing showed increased ST depression at heart rate of 154

to perform coil embolization of the LV using 7 AZUR coils (Terumo Interventional systems, New Jersey,

USA). A repeat angiogram after coil embolization demonstrated minimal flow into the LV through these fistulous communications [Figure 5]. Three months later, repeat exercise stress testing showed marked improvement in exercise tolerance with no chest pain and minimal ST-segment depression in the inferior and lateral leads [Figure 6]. The patient is currently doing



**Figure 6: A repeat electrocardiogram exercise stress test shows no significant ST-segment depression in inferior and lateral leads at peak exertion**

well with no exertional symptoms under follow-up at 6 monthly intervals.

## DISCUSSION

Coronary artery fistulous communications with the ventricular cavity, also called coronary-cameral communications, have been described in HLHS with mitral stenosis and aortic atresia. O'Connor *et al.* initially described these communications in an autopsy study of nine patients.<sup>[1]</sup> These fistulas are congenital and have been known to enlarge with age. The pathophysiology is that there is increased flow across these fistulas due to lower resistance than normal arterioles and capillaries, which traverse the myocardium. With larger fistulas, runoff occurs which draws blood way from the normal coronary pathway leading to coronary steal. Our patient had prominent fistulas from his left coronary system and during simulated exercise with isoprenaline, had retrograde flow in his RCA system with ST-segment depression proving that these fistulas were significant and explaining his symptoms. During elevated heart rate, the flow in the RCA is both during systole and diastole.<sup>[2]</sup> We hypothesize that during exercise, there is elevated systolic and diastolic right ventricular pressure causing an increased pressure gradient across the normal right coronary vascular bed. This leads to reversal of flow in systole and decreased diastolic flow in his normal right coronary arterial system with preferential shunting of flow through the lower resistance fistulas into the LV during diastole.

HLHS with coronary artery fistulas has been noted to have variable morbidity and mortality outcomes, with several reports identifying their presence as a risk factor for increased mortality.<sup>[3,4]</sup> A recent paper reported an operative mortality of approximately 29%

in these patients with autopsies suggesting myocardial ischemia as the cause of death.<sup>[3]</sup> At present, there is little data to guide assessment and risk stratification of these fistulas to determine which fistulas will become clinically and hemodynamically significant<sup>[4,5]</sup> and interventions to interrupt coronary cameral fistulas are not routinely performed at the time of initial surgery in most centers.

In conclusion, we present a case of HLHS s/p Fontan palliation with significant coronary artery fistulas into the hypoplastic LV and clinical symptoms of syncope and exertional intolerance. By coil embolization, the LV was occluded and the drainage site of these fistulas was eliminated, resulting in resolution of clinical symptoms in the short term.

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### Conflicts of interest

There are no conflicts of interest.

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