Chapter 62
Stenting of Fontan Pathway at Anastomose of Conduit to Pulmonary Artery

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Abstract A 23-year-old lady with history of Fontan surgery at 12 years ago referred to us with ascites and lower limb edema. She also declared loss of appetite and fatigue in the last 2 months ago. In physical examination and echocardiography, we suspect to stenosis at fontan pathways that this diagnosis confirmed by CT angiography, for hemodynamic evaluation and also intervention catheterization performed and stenosis at conduit to pulmonary artery confirmed. stenting of the stenotic area performed without complication. The symptoms of the patient alleviated at 2 days after procedure.

History and Clinical Presentation

A 23-year-old lady with history of Fontan surgery 12 years ago came to the ACHD clinic by fatigue, loss of appetite, abdominal swelling, and limb edema for the last 2 months. She did not return for follow-up examination from 2 years ago.

She declared that she did not any symptoms after surgery until 2 months ago.

In surgical report, she had undergone TCPC operation by an extracardiac conduit and Glenn shunt.

Physical Examination

Blood Pressure: 90/60 mmHg, **Heart Rate**: 75 beats per minute, **Oxygen Saturation**: 89% in the air room.

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General appearance: On examination, the patient was well developed. The nailbeds, lips, and mucous membranes were mildly cyanotic and digital clubbing was noted, there was not any respiratory distress at rest. The eyes and skin are icteric.

Lung/chest: No deformity of the chest wall, the lungs were clear.

Heart: Cardiac auscultation revealed a normal first and second heart sounds. No systolic and diastolic murmur or gallop was heard. The rhythm was regular.

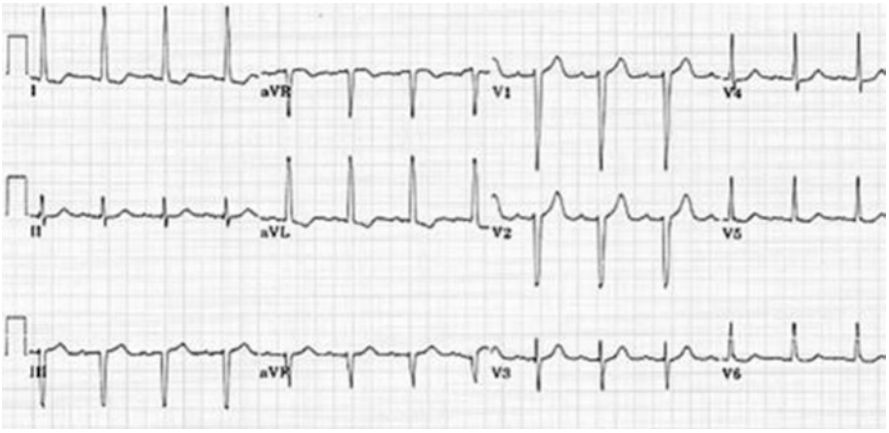
Abdomen: The abdomen was generalized distended, and also the abdominal examination showed fluctuant sensation in pulsation and also dullness in flanks.

Extremities: There was lower limb peripheral edema. The pulses were regular and symmetric but weak.

Laboratory Data

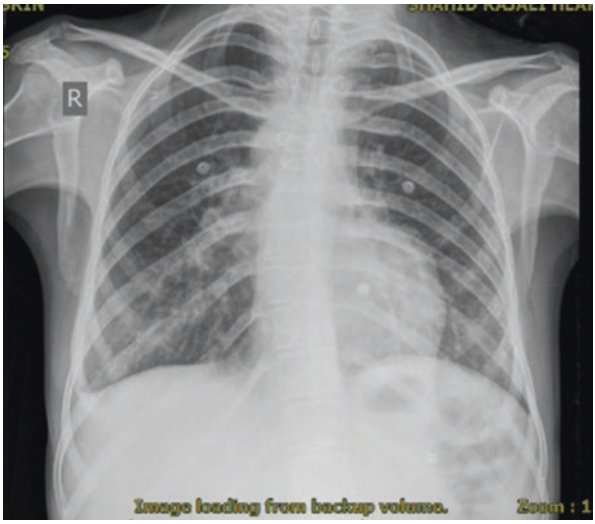
Hemoglobin	16.5 gr/dl
Hematocrit	48.5%
White Blood Count	5300 cells/mm^3
Platelet	216,000 cells/mm^3
Creatinine	0.7 mg/dl
Blood urea nitrogen	13 mg/dl
Cholesterol	115 mg/dl
Triglyceride	86 mg/dl
ALT (Alanine transaminase)	62 IU/L
AST (Aspartate transaminase)	71 IU/L
TSH (Thyroid stimulating hormone)	3.2 micro IU/ml
HIV-Ab	Negative
HBS-Ag	Negative
HCV-Ab	Negative
Bilirubin	7 mg/dl

Electrocardiogram



Twelve lead standard electrocardiogram
Normal sinus rhythm
Heart Rate: 90 beats per minute, regular
Normal QRS duration
Left axis deviation
Normal PR interval
Loss of RV forces suggested tricuspid atresia or DILV in a cyanotic patient

Chest X-Ray



Posteroanterior projection, full inspiration
Levocardia (apex orientation to the left), situs solitus (the gastric bubble is seen in the left)
Left aortic arch
Straight border of the right silhouette in favor of tricuspid atresia
Decreased pulmonary vascularity
Narrow mediastinum in favor of TGA

Echocardiography

- Enlarged systemic left ventricle with preserved systolic function (left ventricle ejection fraction = 50%)
- Hypoplastic RV (Video 62.1)

- Large interatrial septal defect
- Normal mitral valve leaflets, no mitral stenosis, mild mitral regurgitation
- Normal tricuspid aortic valve, no aortic stenosis, no aortic insufficiency
- Atretic tricuspid valve
- Pulmonary artery closed surgically
- Extracardiac conduit was seen with an increased gradient of fenestration(10 mmHg) and turbulent flow with 8 mmHg gradient was seen between the conduit and right pulmonary artery (Video 62.2)
- -Normal laminar flow through Glenn shunt

Congenital CT Angiography

Normal epicardial coronary arteries. Tricuspid atresia and hypoplastic RV was seen. Severe stenosis at conduit insertion to the pulmonary artery was noted (Fig. 62.1).

Catheterization

- Hemodynamic and pressure study showed evidence of elevated pressure in IVC and conduit (20 mmHg), and normal pressure in PA and SVC (12 mmHg).
- Conduit injection showed severe stenosis at conduit anastomose to right pulmonary artery (Fig. 62.2).

Management

- After conduit injection, we decided to stent the stenosis area. Cefazolin IV 1000 mg was administered and guidewire was advanced through catheter from the femoral vein to IVC, conduit, and RPA. The stiff wire then passed through

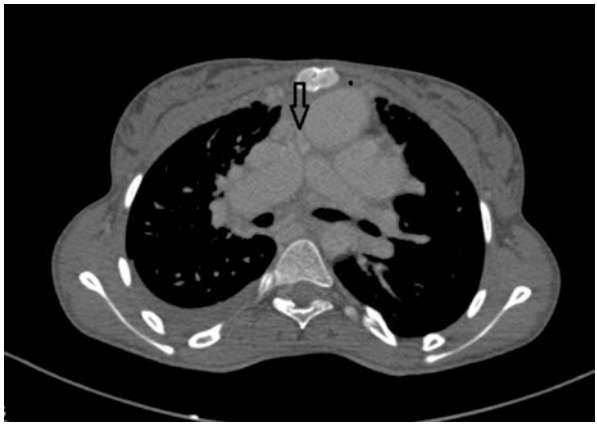


Fig. 62.1 Ct Angio showed Severe stenosis at conduit insertion to the right pulmonary artery

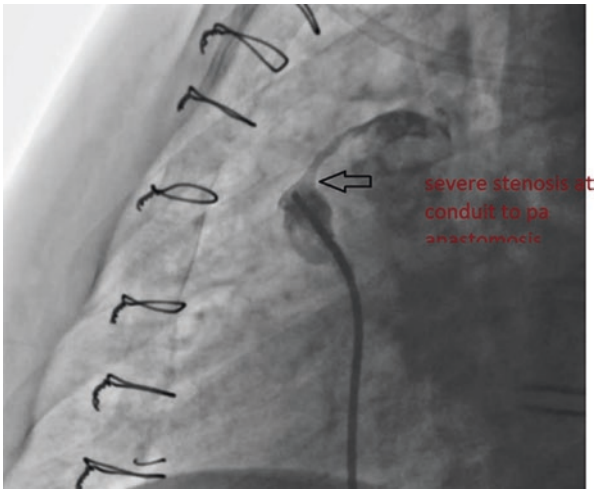


Fig. 62.2 Severe stenosis at conduit insertion to RPA

the catheter. The venous sheath and catheter were then removed and an introducer sheath was advanced over the stiff guidewire to the RPA, the guidewire was removed. Next, a Palmaz Genesis stent 28 mm mounted on Balloon 30 × 10 mm, advanced through the introducer sheath and inflated at the anastomosis site. Then post-dilation performed by balloon 16 × 30 mm and gradient eliminated completely.

- During some days after the procedure the ascites and icterus alleviated and after 1 month patient feels completely healthy condition. She had under treatment with warfarin and clopidogrel. After 3 months clopidogrel discontinued and warfarin continued.

Discussion The Fontan procedure is an operation that bypasses a right ventricle and diverts a systemic venous blood to the pulmonary arteries [1]. It is a palliative treatment for many complex congenital heart diseases that biventricular repair is impossible or more challenging. There are different types of Fontan circulation. (1) Atriopulmonary connection that is historical and not performed nowadays; (2) intracardiac total Cavo pulmonary connection (lateral tunnel or intracardiac conduit); (3) extracardiac total Cavo pulmonary connection [2].

The patients who undergone Fontan procedure may have many complications during follow-up. One of the complications is obstruction at Fontan circuit and it may be focal or diffuse and can happen at IVC anastomosis to conduit or conduit anastomosis to the pulmonary artery or at through the conduit length [3]. This obstruction may be due to technical surgical problems with less probability, multiple thrombosis formation at the circuit, or more likely stretching of the conduit during child growth. Most of this stenosis amenable for catheter intervention [4] and symptoms of the patient and NYHA functional class improved after the intervention.

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