

Sutureless Repair of Left Primary Pulmonary Vein Stenosis—A Case Report

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Abstract

Primary left pulmonary vein stenosis is a very rare condition. It is associated with variable form of Pulmonary artery hypertension. Catheter based intervention is used to treat the disease like balloon dilation and stenting but this has high recurrence rate. Here is a case in which we did a sutureless repair of left pulmonary vein thus decreasing the possibility of recurrence. Sutureless surgical management is technically superior and less challenging than direct left atrial wall anastomosis and it provides better results than catheter based intervention in terms of recurrence of stenosis.

Keywords

Pulmonary Vein Stenosis, Trans Thoracic Echocardiogram, Atrial Septal Defect

1. Introduction

Primary Pulmonary vein stenosis (PVS) occurs due to developmental aberrations in the incorporation of pulmonary veins into left atrial wall. It is a very rare anomaly and is present in 0.5% of autopsy in children. It may manifest as isolated lesion or associated with other cardiac defects (more than 50% of cases). Though the catheter based interventions like balloon dilation, stent insertion, excision of stenotic segment, patch grafting are used to correct such stenotic lesions, the re-stenosis rates are higher than the sutureless surgical management. We present a case, who underwent surgical sutureless repair for left side primary pulmonary vein (LSPV) stenosis and had an uneventful post operative recovery with no residual stenosis.

2. Case Report

A 4 year old female child, 14 kg was admitted for dyspnea, tachypnea and cya-

nosis. Her SpO₂ was 88%, heart rate 130 beats/min, systolic blood pressure 90 mmHg, and respiratory rate 45 per minute. The second heart sound was accentuated, while grade 2/6 pan-systolic murmur was audible over the lower left sternal border. Her jugular vein was distended and the liver was palpable 4 cm below the right costal margin complete blood count, electrolytes, renal, and hepatic profiles are normal. Chest X-ray showed marked pulmonary venous congestion with cardiomegaly (**Figure 1**).

Trans thoracic Echocardiogram (TTE) showed mild pulmonary hypertension (PAH) with a dilated right ventricle, bilateral shunting across the atrial septal defect (ASD) with obstructed left side pulmonary veins opening to left atrium. CT pulmonary angiography showed long segment LSPV stenosis (**Figure 2 (a)**, **Figure 2 (b)**).

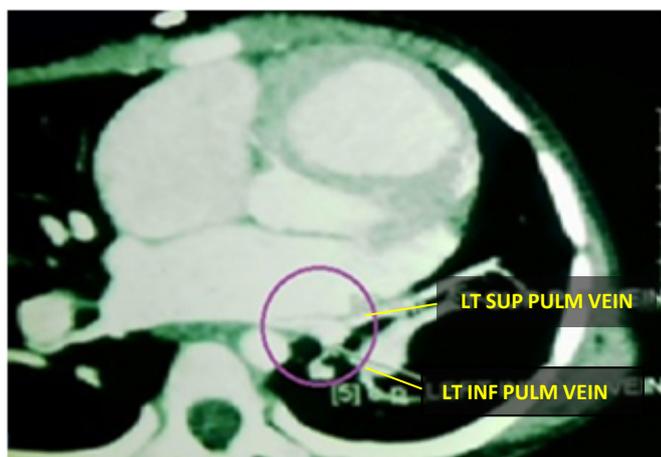
Patient was operated on cardiopulmonary bypass with cold cardioplegic arrest. Heart was retracted to right chest cavity. Incisions are made in the left atrium and extended to the base of left atrial appendage and in the left pulmonary venous confluence extended to both superior and inferior pulmonary veins across the stenosed segment (**Figure 3 (a)**, **Figure 3 (b)**).

The divided edge of the atrial wall is then sutured to the pericardium (not the pulmonary vein) in a suture line remote from the divided edge of the pulmonary veins using a running 5 - 0 non-absorbable suture. The suture line is technically easy to sew and hemostatic challenges are less as it connects the left atrial edge to the pericardium in a circle around the pulmonary veins and the suture line ignores the complex shapes of the pulmonary vein incisions. Pericardial patch closure of ASD done through right atriotomy.

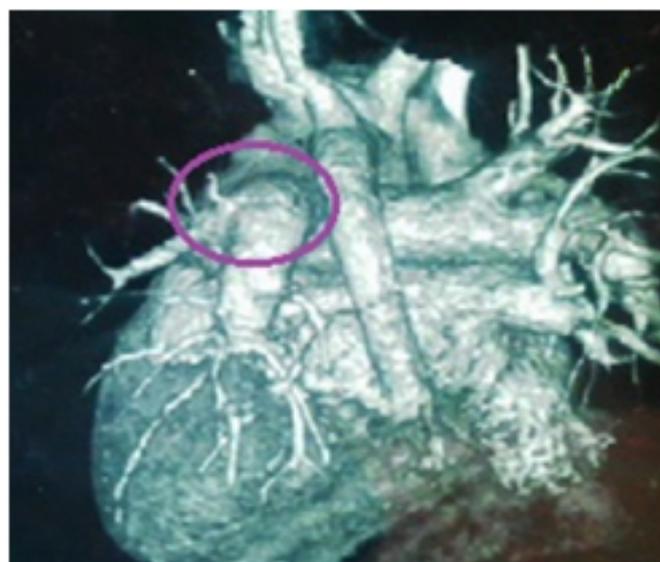
Postoperatively, patient recovered well, extubated on 2nd Post operative day with SpO₂-100% and discharged on 7th postoperative day. Post operative Cardiac CT scan revealed pulmonary vein confluence opening into left atrium with no residual stenosis with an intact neoatrial septum (**Figure 4**). Post operative TTE revealed mild Pulmonary artery hypertension (PAH) with non obstructive flow across the anastomosis.



Figure 1. Chest X-ray showing marked pulmonary venous congestion with cardiomegaly.



(a)



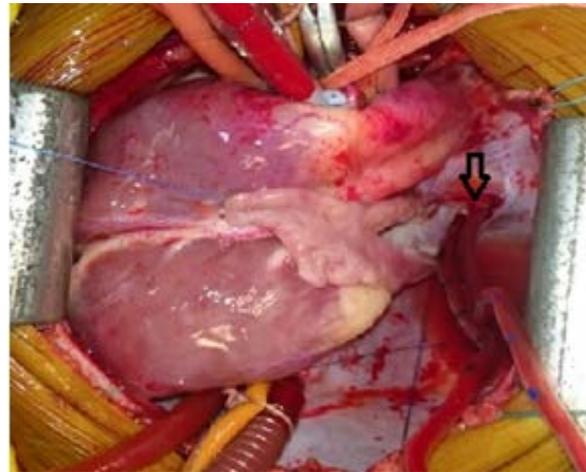
(b)

Figure 2. (a) (b) CT pulmonary angiography showing long segment LSPV stenosis.

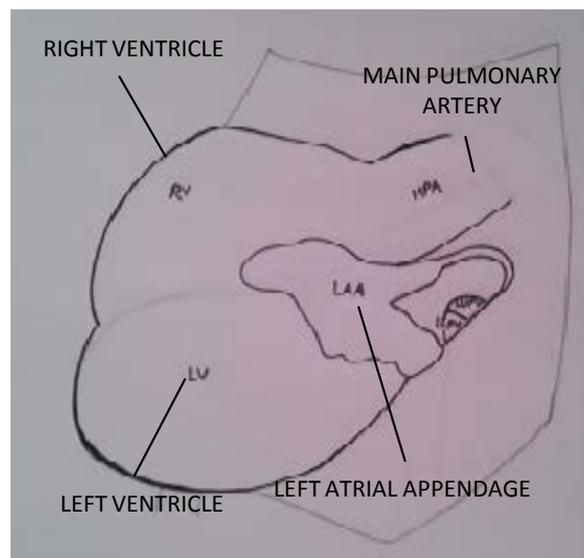
In our patient we did this procedure and postoperative result were good. Patient did not develop restenosis in 1 year follow up.

3. Discussion

Primary PVS is a rare disease. In such cases, patient becomes symptomatic because of pulmonary edema and hypoxemia resulting from increased hydrostatic pressure in the pulmonary venous system, which in turn triggers an increase in pulmonary vascular resistance and further worsen the symptoms. This vicious cycle is remarkable in first few years of life as the pulmonary vascular recruitment is very limited even in patients of unilateral disease. Patients present initially with tachypnea and recurrent pneumonia and with the progression of disease patient may develop a new murmur, right ventricular failure and pulmonary edema. Patient may develop failure to thrive and hemoptysis [1]. Our patient had unilateral



(a)



(b)

Figure 3 (a) Showing Incisions made in the left atrium and extended to the base of left atrial appendage and in the left pulmonary venous confluence extended to both superior and inferior pulmonary veins across the stenosed segment; (b) Pictorial presentation of incisions made in the left atrium and extended to the base of left atrial appendage and in the left pulmonary venous confluence extended to both superior and inferior pulmonary veins across the stenosed segment.

PVS but with radiographic evidence of bilateral reticular markings consistent with pulmonary edema and it explains the clinical presentation of tachypnea and hypoxemia.

TTE with doppler ultrasound detecting the turbulent flow gives rise to a suspicion of PVS. CT angiography and cardiac MRI though provide three dimensional view and noninvasive diagnostic tools for PVS, but are limited by spatial

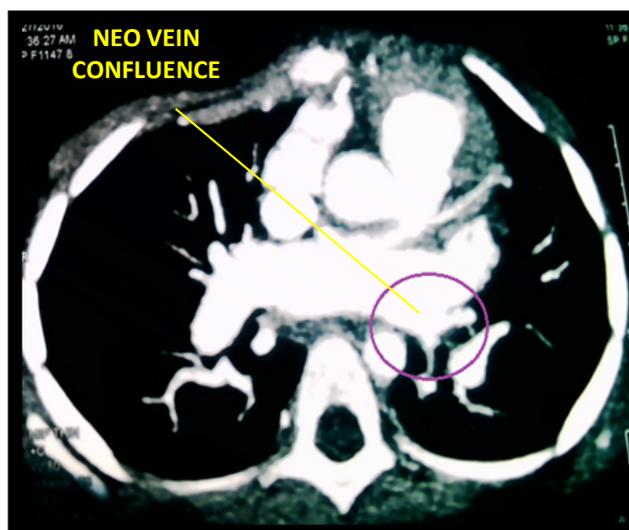


Figure 4. Post operative Cardiac CT scan revealing pulmonary vein confluence opening into left atrium with no residual stenosis with an intact neoatrial septum.

resolution. Radionuclide quantitative pulmonary flow imaging is another excellent tool to evaluate the distribution of pulmonary blood flow. Cardiac catheterization (or angiography) remains the gold standard for the diagnosis of PVS. A catheter is advanced into pulmonary artery and the contrast flow is observed till the pulmonary segments; if PVS is present, flow will be reduced or absent [1] [2].

Though role of catheter based interventions like balloon dilation and stent insertion for the management of resistant PVS lesions have been argued by authors in the literature, the restenosis rates are higher than the surgical methods [3] [4]. The Coles procedure (a “sutureless” technique) is the preferred surgical repair method [1] [5] [6]. The advantage of this technique is that there is no suture line on free pulmonary vein edges; hence stimulus for fibrotic tissue growth in pulmonary vein is less [1]. Lacour-Gayet F *et al.*, Najm HK *et al.* and Yun T-J *et al.* have also used the same procedure and supported its use in the patients of PVS. Left atrial wall anastomosis with pericardium is technically less challenging in a retracted heart with minimal exposure on posterior aspect of heart than the direct anastomosis with the pulmonary vein confluence. Pneumonectomy and lung transplantation are reserved for the patients having refractory hemoptysis and severe PAH respectively.

4. Conclusion

A high index of clinical suspicion should be made for PVS in patients presenting with symptoms of PAH. Once the diagnosis is made, management is primarily surgical. Sutureless surgical management is technically superior and less challenging than direct left atrial wall anastomosis and it provides better results than catheter based intervention in terms of recurrence of stenosis.

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Consent

Consent for the publication has been given by his parents as the patient is minor.

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