

Congenital Pulmonary vein stenosis - Diagnosis and treatment.

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Abstract

Congenital pulmonary vein stenosis is a rare and interesting condition. This disease was classically believed to have an exceptionally bad prognosis. This article reviews the pathophysiology of the disease and advancements in diagnosis and management.

Keywords: Congenital heart disease; Pulmonary veins, abnormalities; Pulmonary veins, physiopathology; Pulmonary veins, surgery.

Introduction

Congenital pulmonary vein stenosis is an interesting condition which is probably under diagnosed and is exceptionally difficult to manage. In spite of advances in surgical and catheter based management of this disease, prognosis remains at the best guarded in a significant proportion of the patients.

This article reviews the aetiology diagnostic methods and advancements in treatment.

Incidence

Congenital pulmonary vein stenosis is deemed to be a rare anomaly. Its reported prevalence is 1.7 per 1,00,000 children less than two years old [1]. The very fact that it has been reported in 0.5% of autopsy cases suggests that there is possibility of under diagnosis [2].

Associated cardiac defects have been reported in 30-80% of patients, most commonly septal defects [3]. It is interesting and important to note that the largest surgical series of

congenital pulmonary vein stenosis reports 23 patients, among which there was no case of isolated pulmonary vein stenosis [4]. Among the patients who succumbed to the disease in this series, 9 out of 11(81.81%) had septal defects or a patent ductus arteriosus as associated defects. This emphasises the importance of congenital pulmonary vein stenosis in the prognostication of otherwise simplecardiac defects.

Etiology and Patho-physiology

Abnormal incorporation of the common pulmonary vein into left atrium during the later stages of its development leads to congenital pulmonary vein stenosis [5]. However, there is increasing recognition of an underlying neoproliferative process caused by myofibroblastic cells leading to rapid progression of disease which might not even be evident at birth [6,7].

This has reportedly led to a preference for the term primary over congenital [3].

Figure: 1 and Figure: 2 show angiographic images of a patient with congenital pulmonary vein stenosis who developed re-stenosis post surgical repair using the suture-less technique described later.

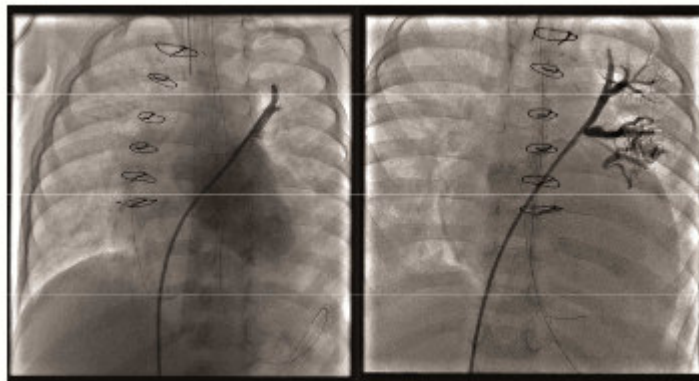


Figure 1 – Small sized, patent upper branch of left upper pulmonary vein at first catheterization study post surgery (left) and 1 month later (right) showing new areas of focal stenosis. Note that left atrium is less opacified in the right image with reflux of dye into distal and lower branches due to the proximal obstruction and 5 French catheter almost occluding the mouth of pulmonary vein completely.

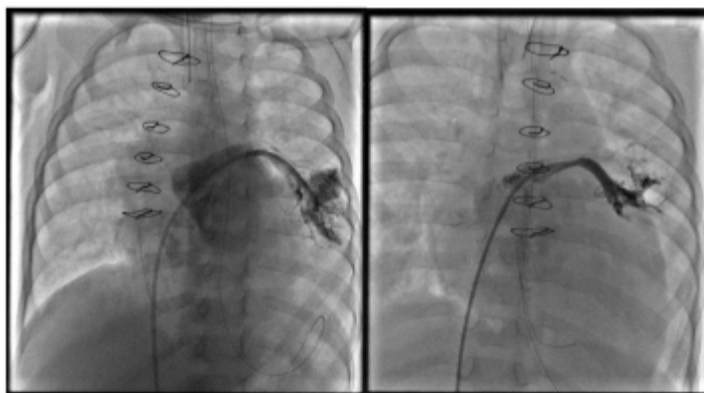


Figure 2 – Lower branch (lingular) of left upper pulmonary vein at first study post surgery (left) and 1 month later (right) showing new focal stenosis.

The comparative images on the left and the right are from angiograms done at an interval of one month and shows rapid progression of disease. Stenosis may appear as a discrete shelf, a segment of narrowing extending from the mouth of the pulmonary vein for a short distance or a diffuse hypoplasia extending into the hilum/intrapulmonary segments [7,8]. Differentiating focal stenosis from diffuse hypoplasia is of extreme importance as the latter has exceptionally worse prognosis [9]. Active inflammation has not been found in or around the affected veins [3].

The fixed obstruction in the pulmonary veins produces a secondary increase in pulmonary artery pressure and decreased pulmonary blood flow in the corresponding lung/lobes. In these lung/lobes, the elevated pressure causes vascular muscle to develop more peripherally than is normal and medial hypertrophy of arteries develops, as has been observed in other forms of pulmonary venous obstruction. The reduced flow leads to decreased arterial size. Any unobstructed lobe accommodates the diverted pulmonary blood flow and can lead to a functional stenosis of the anatomically unobstructed pulmonary vein. Arterial changes can be produced in the unobstructed lobes by the increased flow and the resulting increased pressure. The small- and medium-sized pulmonary veins show medial hypertrophy [9]. Insufficient drainage of blood from the affected lung leads to pulmonary congestion, recurring respiratory infections, and haemoptysis [10].

Clinical presentation

Controversy persists as to whether the number of pulmonary veins involved and the degree of obstruction affects the onset and severity of disease [3,11]. It seems plausible that progression of symptoms may be less rapid if only one or two pulmonary veins are involved [12]. Even in initially less severe cases, progression to bilateral pulmonary vascular disease and death has been described [5,7, 13-20].

The presentation is usually in the first months to years of life with history of significant respiratory symptoms. Patients may have recurrent pneumonia. They also have diffuse or localized pulmonary oedema depending on the anatomic involvement. Haemoptysis is prominent in older patients [3]. Features of pulmonary hypertension become evident later in the course as the disease progresses. The frequency of associated heart defects and reports of progression from normal pulmonary venous flow to progressive stenosis warrants evaluation of pulmonary veins during every echocardiographic study [21]. Also the pulmonary veins need to be critically looked at in any patient with unexplained pulmonary hypertension. Other diseases that can present similarly include most other causes of pulmonary venous hypertension including mitral atresia and different anatomical types of congenital mitral stenosis, cor triatriatum, tumour of the left atrium and total anomalous pulmonary venous connection with obstruction. Pulmonary venoocclusive disease, a close differential diagnosis is a poorly understood syndrome wherein there is extensive and diffuse occlusion of pulmonary venules and small veins by fibrous tissue. Larger veins are rarely involved in pulmonary venoocclusive disease [22].

Chest x-rays show a normal sized heart or cardiomegaly once the right heart gets involved. Characteristically, the pulmonary fields in pulmonary venous obstruction show a fine, reticulated vascular pattern, passive pulmonary congestion, or frank pulmonary edema with distribution determined by the vein/veins involved. Features of pulmonary artery hypertension appear later.

Electrocardiograms show right ventricular and right atrial enlargement [8]. Secondary pulmonary vein stenosis is seen in pediatric patients most commonly after surgery for total anomalous pulmonary venous connection [3]. Other causes include invasion by tumour tissue, mediastinitis, constrictive pericarditis and pulmonary vein phlebitis as in tuberculosis [8].

Primary pulmonary vein stenosis, similar to that in children has been rarely reported in adults. Of late, the commonest cause of secondary pulmonary vein stenosis in adults has been radiofrequency ablation for treatment of atrial fibrillation [23,24]. Sarcoidosis has also been described as a cause for secondary pulmonary vein stenosis in adults, in addition to the causes mentioned for children.

Diagnosis

Echocardiography is the primary modality of diagnosis and is usually adequate especially in children with good acoustic windows. 2D evaluation of the size and Doppler interrogation of individual pulmonary veins are done. Turbulent flow found during colour Doppler interrogation of pulmonary veins raises suspicion. A monophasic flow or flow velocities more than 1.6 m/s indicates significant obstruction [26]. A pre-operative pulmonary vein stenosis score incorporating 2 Dimensional echocardiographic assessment and Doppler interrogation of the pulmonary veins has been proposed. This score is found to have excellent correlation with intra-operative and autopsy findings and is helpful in predicting mortality post surgery [4]. Echocardiography also helps in ruling out most of the other causes of pulmonary venous hypertension. Multidetector CT is an excellent tool for more detailed evaluation, especially for planning therapeutic strategies. An important concern with CT is its inability to differentiate completely occluded pulmonary veins from those with a tiny residual opening still adequate for catheter intervention [27]. CT might add information to exclude compression of extrapulmonary segments of pulmonary veins and also help in differentiating pulmonary veno-occlusive disease. MRI has the advantage of avoiding ionising radiation, but has limitations of long acquisition times, sensitivity to motion artefacts and arrhythmias and limited spatial resolution [3].

Cardiac catheterisation study and angiography was the modality of choice for confirmatory diagnosis of congenital pulmonary vein stenosis before the era of advancement in noninvasive diagnostic techniques [5]. In the current era, invasive study is done mainly with the intention of percutaneous dilatation. The presence of pulmonary artery hypertension along with an elevated pulmonary capillary wedge pressure and a normal left atrial pressure suggests congenital obstruction of the extra pulmonary segments of the pulmonary veins [9].

Selective angiograms can be done from arterial segments that drain to each of the pulmonary veins. Pulmonary artery segments draining into severely stenosed pulmonary veins may have little antegrade flow and the contrast may even flow backwards into arteries that drain less stenotic veins. Balloon wedge angiography from the segmental pulmonary artery is helpful in such situations and may even show patency missed by non invasive methods [3,27]. Alternately, direct visualization of non occluded pulmonary veins can be made entering the left atrium through an atrial septal defect/patent foramen ovale or via transeptal puncture if the atrial septum is intact. In pulmonary veno-occlusive disease, a close differential diagnosis, the characteristic finding described has been inability to obtain a pulmonary artery wedge tracing. If at all the catheter is wedged, the wedge pressure recording would be normal [22]. Radionuclide quantitative pulmonary flow imaging identifies redistribution of flow in case of asymmetric pulmonary vein stenosis and is recommended before any intervention and for follow up.

Treatment

Untreated, long term survival is rare and the mode of death is a pulmonary hypertensive crisis, inter current infection or hemoptysis [3]. Early surgical techniques and catheter interventions were reported to have dismal results . This was attributed to relentless re stenosis. A variety of techniques have been proposed for surgical repair ever since the first attempt by Kawashima and colleagues [28]. It was speculated that the patch material used in surgical venoplasty techniques acted as a substrate for turbulent blood flow triggering intimal hyperplasia. Trauma caused by direct suturing of the pulmonary vein intima was also believed to stimulate further injury response, accelerating restenosis. Lacour-Gayet et al

The ostium of the left lower pulmonary vein is also narrow as seen from within the left atrium. The left atrial appendage can also be used for marsupialization of the left sided veins.

first described a technique to avoid both these issues during surgical repair² . This technique, called the suture-less pulmonary vein repair (Figure 3) avoids stitches at the cut edges of the pulmonary veins and uses pedicled autologous pericardium for marsupialisation around the openings of pulmonary veins forming a neo atrium. This has better results in both congenital and acquired forms and is increasingly used in primary repair for pulmonary vein anomalies at high risk of post repair restenosis [3,30]. Mid to long term results of this technique are encouraging (Table - 1).The technique best suits anatomically localized disease, diffuse disease has been recognized as a technical challenge [31].

Figure 3 – The “suture-less” technique of repair for pulmonary vein stenosis. The figure shows the approach to right sided veins. The dotted line illustrates the line of excision of the narrow pulmonary veins along with left atrial wall if required. The dashed line illustrates the final suture line which would approximate the atrial wall with pericardium. The pulmonary veins will drain into the neo atrium thus created and then into the left atrium.

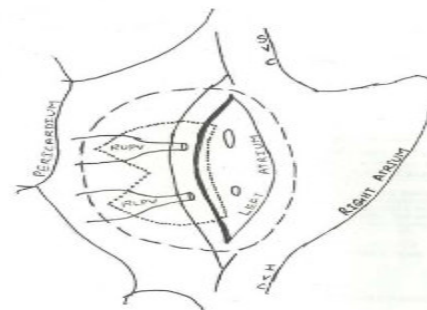


Figure 3

Table-1 Results of the suture-less technique of pulmonary vein repair for congenital pulmonary vein stenosis

Author	Number of patients	Follow up duration	Freedom from re-stenosis /death	Deaths	Comments
Devany et al [20]	Suture-less in 8/14	25 months (median)	50%(suture-less)	2 early; 1 late (all associated with re-stenosis)	Bilaterality predicts diffuse disease & poor outcome
Azsakie et al [30]	7	34months (median)	57%(suture-less)	1 late death (during a gen surgical procedure)	Effective for discrete stenosis; diffuse disease – technical challenge
Nicola Viola et al [4]	Suture-less in 19/23	40.2+/-42.2 months (mean +/- SD)	64% -1yr 47%-5yr 31% -10yr (overall) 42% mortality excluding non suture-less repairs	11 – total deaths	Preoperative pulmonary vein stenosis score predicts mortality

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Among the available treatment options for diffuse disease, bilateral sequential lung transplantation has good short term results. Mendeloff et al reported three patients who underwent bilateral sequential lung transplantation. Average hospital stay was 42 days after transplantation [32]. One of these patients was on ECMO (extra corporeal membrane oxygenation) for 24 days before transplantation. There was no echo evidence of PAH on 0.5 to 1.8 years follow up. An analysis of the pediatric cardiac care consortium database identified younger age at diagnosis, higher initial mean pulmonary artery pressure and bilateral vessel involvement as predictors of lung death and recommended early consideration of lung transplantation for these subsets [11]. Breinholt et al described two patients who underwent cardiac transplantation for other indications. Stenosed recipient pulmonary veins were not addressed during transplantation. Both patients were fine at six and 11 months post transplant [21]. As pulmonary pressures were in the range acceptable for heart transplant, these patients possibly had a milder form of disease. As diffuse disease is attributed to neoproliferative process, the possibility of use of antiproliferative drugs for prevention of re-stenosis after surgery or as treatment seems attractive. There have been anecdotal reports on use of oral steroids to prevent post operative pulmonary vein stenosis [33]. Antiproliferative drugs are another option. A prospective phase 2 trial of weekly vinblastine and methotrexate in children had only 38% survival at 1 yr for patients with associated congenital heart disease. There were two patients with isolated disease, both of them died from progression [34]. Oral sirolimus has been successfully used for recurrent stenosis in adults [35]. Catheter interventions have acceptable immediate results with the disadvantage of recurrence; but can be used multiple times for temporary relief of focal stenoses. Repeat procedures can slow down the disease process [3]. High pressure balloon dilatations and cutting balloons give better results [3]. Overall; results of catheter intervention for pulmonary vein stenosis are better in adults compared to children as the larger lumen achieved decreases restenosis rates; lesser number of veins are involved and few patients have associated heart disease. Stenting has better results in adults compared to balloon angioplasty; literature is conflicting regarding the use of drug eluting stents [3,36]. Although initial reports were discouraging [12,37,38]; a variety of stents - bare-metal, drug eluting and covered were recently reported to have acceptable acute results in children for focal obstruction. Recurrence was more with smaller stents. Survival rate was 50±8% at 5 years after pulmonary vein stent implantation. Freedom from reintervention was 62±7% at 6 months and 42±7% at 1 year [38]. Cryo-balloon [39] and drug eluting balloon have acceptable acute results [40]. Intrastent sonotherapy reportedly has good result for re stenosis after stent implantation [41]. Challenges in children include diffuse disease, small size of diffusely narrowed veins and need for trans-septal puncture if there is no PFO.

Summary

Abnormal embryologic events, a neoproliferative process has been identified, which lends a progressive Congenital pulmonary vein stenosis is a rare disease. Although its origin is attributed to nature to the disease. Echocardiography is currently the primary modality of diagnosis and a high index of suspicion is required for identification of the disease process. Other non invasive modalities provide detailed information and are used mainly for planning catheter interventions. Surgical relief using the suture-less technique is the preferred treatment modality. Catheter interventions can be used repeatedly for re-stenosis. Encouraging results are being recently reported, repeat procedures can also slow down the disease process. A variety of investigational modalities have been applied to manage diffuse and repeated restenosis which can be tried before a decision is made for lung transplantation. The prognosis in certain subsets of patients remains grim in spite of advances in treatment. Larger studies using newer modalities of treatment would help to improve prognosis in the difficult to treat subsets.

Disclosure

The images shown in Figure 1 and 2 had been included in the paper "Our experience with the suture-less technique of repair for primary pulmonary vein stenosis" presented at the Annual Conference of the Pediatric Cardiac Society of India, held at Chennai, India in October 2012.

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