



## ASYMPTOMATIC PRIMARY PULMONARY VEIN STENOSIS: A CASE REPORT

**Dr Thota Chiranjeevi**

MBBS, MS General Surgery

**Dr Hemachandra Tokala**

MBBS, MS General Surgery

**Dr Rama Krishna Dev T**

MBBS, MS, Mch CT Surgery

**Dr R. V. Kumar\***

MBBS, MS, Mch CT Surgery \*Corresponding Author

**ABSTRACT** Pulmonary vein stenosis is a rare congenital disease that affects children, and the prognosis is poor in most of the patients. Stenosis of the individual pulmonary veins, first described by Reve[1] is uncommon. It is characterized by congenital extrapulmonary obstruction and leads to significant alteration of pulmonary structure and function. An angiography will typically be able to confirm the diagnosis. When there is a history of recurrent lung infections, Dyspnea, Failure to thrive, Hemoptysis, or Pulmonary Hypertension that cannot be explained, stenosis of particular veins should be examined.

**KEYWORDS :** Pulmonary veins, congenital , primary , stenosis

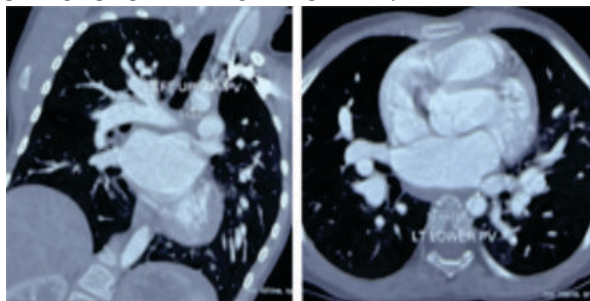
### INTRODUCTION

Pulmonary vein stenosis is an uncommon congenital disease that almost exclusively affects in young children with or without various forms of congenital heart disease[2]. It is a disease with a bad prognosis, and it is extremely rare for a patient to make it to adulthood without receiving therapy. This is due to the fact that the symptom first appears in infants, and it is accompanied by progressively worsening pulmonary Hypertension[3].

Herein , we present a patient who diagnosed to have sub-Aortic VSD with Pulmonary vein stenosis with mild pulmonary hypertension.

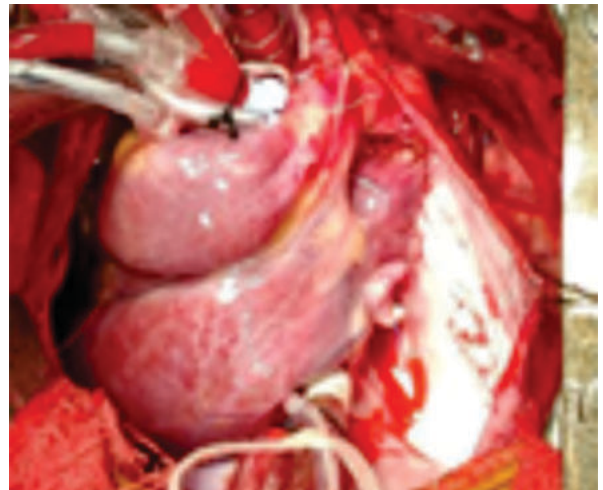
### CASE REPORT

A 7-year-old female child who is known case of ACHD, Sub Aortic VSD + Left Pulmonary vein stenosis with no regular follow up now came with complain of shortness of Birth on exertion, Frequent URTI's and decreased appetite. On examination vital signs were stable, No pallor and Cyanosis, Respiratory system Examination- Symmetrical chest expansion with no abnormal lung sounds. Cardio vascular examination- S1 and S2 present, Pansystolic murmur+. Arterial blood gas analysis at room air with in normal range. Electrocardiogram shows right ventricular Hypertrophy. On Transthoracic Echocardiography a 1cm Subaortic VSD with left to Right shunt, with normal systemic and pulmonary veins drainage, Good Biventricular function. X ray no obvious defect is seen. On CT Thoracic Angiogram showed Subaortic VSD (1.3mm), Right upper and lower Pulmonary veins draining into left atrium, moderate stenosis of left upper and lower pulmonary veins drain into left atrium ( diameter of left upper pulmonary vein 2.8mm, lower pulmonary vein 3.6mm) [Fig.1], with pruning of peripheral sub segmental pulmonary arteries.



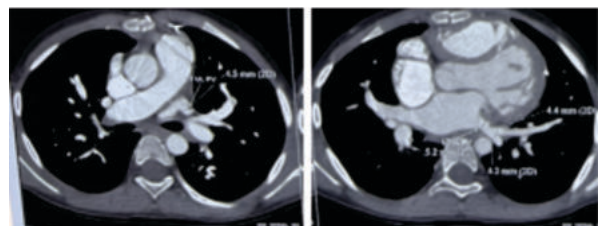
**Fig.1 – Left Upper And Lower Pulmonary Vein Stenosis.**

Patient was taken up for surgery, VSD closure done with pericardial patch and left superior Pulmonary vein augmentation done with left atrial appendage [ Fig.2] ( intraoperatively left lower pulmonary vein no significant stenosis seen).



**Fig.2 Left Upper Pulmonary Vein Augmentation Done With Left Atrial Appendage.**

Postop recovery was uneventful, and patient discharged on postop day 5. Patient is on regular follow up with no fresh complaints, follow up CT Thoracic Angiogram was done, no Stenosis at operated site[Fig.3]



	Preop	Postop
Left upper pulmonary vein	2.8mm	4.5mm
Left lower pulmonary vein	3.6mm	5.2mm

### DISCUSSION

Pulmonary vein stenosis is a rare congenital disease accounts for 0.4% seen in neonates and children[2]. It has been postulated that it results from abnormal incorporation of the common pulmonary vein into left atrium in the later stages of the cardiac development.

Patients become symptomatic in the first few months to years of life, and frequently associated with other congenital cardiac anomalies[4].

Congenital pulmonary vein stenosis refers to pulmonary stenosis in children and even adults without a known antecedent or concurrent cause of stenosis. Evaluation for pulmonary vein stenosis to be done in any young patients with severe pulmonary hypertension. Patients with congenital pulmonary vein stenosis have a poor prognosis. Prognosis depends upon the severity of the associated cardiac disease and number of pulmonary veins stenosed[5]. Patients with only 1 or 2 pulmonary veins affected have a substantially more benign course, although the exact natural history of milder forms of congenital pulmonary vein stenosis is unclear. The cause of death usually a pulmonary hypertensive crisis, recurrent pulmonary infection, or massive hemoptysis.

Given in the rarity and severity of this condition, treatment options, and long-term management are necessary to improve the prognosis and quality of life for individuals affected by pulmonary vein stenosis.

#### REFERENCES

1. Reye RDK. Congenital stenosis of the pulmonary veins in their extrapulmonary course. *Med J Aust* 1951; 1:801-802.
2. Edward J. Congenital stenosis of the pulmonary veins: pathologic and developmental considerations. *Lab Invest* 1960; 9:46-66.
3. Van Son JA, Danielson GK, Puga FJ, Edwards WD, Driscoll DJ. Repair of congenital and acquired pulmonary vein stenosis. *Ann Thorac Surg* 1995; 60:144-150
4. Latson LA, Prieto LR. Congenital and acquired pulmonary vein stenosis. *Circulation* 2007; 115:103-108
5. Breinholt JP, Hawkins JA, Minich L, Tani LY, Orsmond GS, RitterS, et al. Pulmonary vein stenosis with normal connection: associatedcardiac abnormalities and variable outcome. *Ann Thorac Surg* 1999; 68:164-168