

An Usual Case of Congenital Pulmonary Stenosis

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Introduction

Pulmonary valve stenosis as an isolated entity is not a common diagnosis, and isolated pulmonary valve stenosis is most commonly a congenital form [1]. The valvular stenosis is characterized by a trileaflet valve with fused commissures, although, rarely a dysplastic valve without commissural fusion may also be encountered [2]. Such

a dysplastic and stenotic valve is seen in Noonan's syndrome. Right ventricular hypertrophy usually results due to right ventricular pressure overload from the stenotic valve, and this may also include infundibular hypertrophy. Isolated infundibular stenosis is rare and so is isolated supraventricular stenosis. Approximately 5 out of 1000 infants are born with a congenital cardiac malformation [3]. Prevalence of pulmonary stenosis is 8-12% of all congenital heart defects [3].

Rheumatic inflammation of the pulmonary valve is very uncommon and is almost always associated with the involvement of the other valves. Also, rheumatic involvement rarely leads to a serious deformity.

As stated earlier, isolated supraventricular right ventricular outflow tract obstruction seldom occurs in isolation. It may be associated with Tetralogy of Fallot, Williams syndrome, Noonan's syndrome, Ventricular Septal Defect (VSD) or arteriohepatic dysplasia (Alagille syndrome). It progresses in severity with the passage of time and needs to be monitored.

Case History

A 15 year old male presented to us with history of cyanosis for the past 6 years associated with shortness of breath on exertion. Symptoms had been persisting even before the onset of frank cyanosis, in the form of exertional fatigue and light headedness.

On examination, his pulse rate was 100 per minute, blood pressure was 110/76 mmHg. Central cyanosis and grade 4 clubbing were present (Figures 1 and 2). JVP was 13 cm of H₂O and showed a prominent *a* wave. Right ventricular heave was present and a thrill was palpable in the second left intercostal space. On cardiac auscultation, S₁ was normal, P₂ was soft while A₂ was normal and a systolic ejection murmur was heard in the second left intercostal space. On examination there was no evidence of any connective tissue disorder or Marfan's Syndrome/Ehler's Danlos Syndrome.

Routine investigation showed a polycythemia with a hematocrit of 52%, with a normal TLC (3600 per mm³) and a normal DLC (P₆₄L₃₆). Serum electrolytes, blood urea and serum creatinine were normal. Electrocardiography showed P-pulmonale with right ventricular hypertrophy along with right ventricular strain pattern as well as tall R waves in V₁R (Figure 3).

Chest X-ray revealed oligemic lung fields with right atrial and right ventricular hypertrophy (Figure 4).

Combined two dimensional echocardiography and continuous-wave Doppler examination were carried out which revealed right atrial



Figure 1: Central cyanosis.



Figure 2: Grade 4 clubbing.

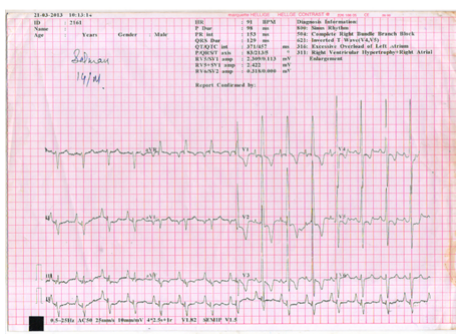


Figure 3: Electrocardiography showed P-pulmonale with right ventricular hypertrophy along with right ventricular strain pattern as well as tall R waves in V4R. Electrocardiography showed P-pulmonale with right ventricular hypertrophy along with right ventricular strain pattern as well as tall R waves in V4R.

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Figure 4: Chest X-ray revealed oligemic lung fields with right atrial and right ventricular hypertrophy.

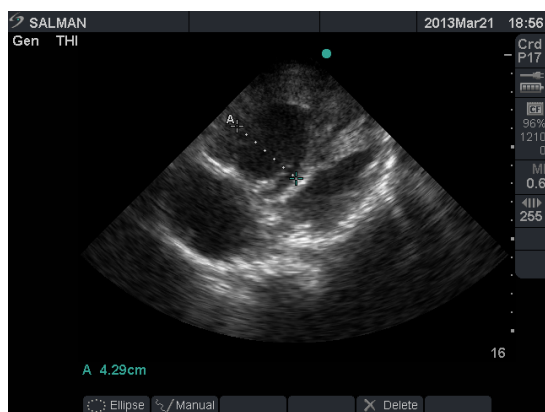


Figure 5: Echocardiography.

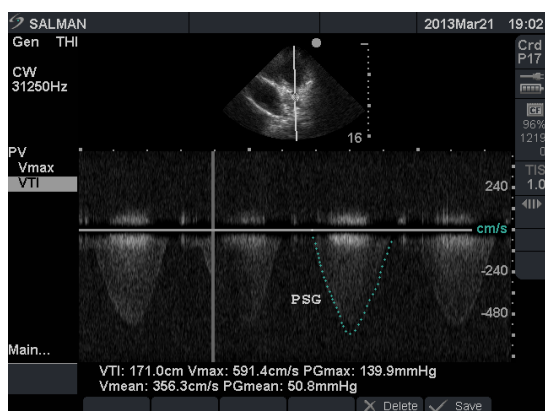


Figure 6: Continuous-wave Doppler examination.

and right ventricular hypertrophy along with a stenotic pulmonary valve, with thickened leaflets and restricted systolic excursion. Peak gradient across the pulmonary valve was 140 mmHg and mean gradient was 68 mmHg (Figures 5-7). There was no evidence of pre-existing Patent Foramen Ovale or Atrial Septal Defect to explain cyanosis in this patient. There was no evidence of any acquired infection like AIDS.

Discussion

In patients of congenital pulmonary stenosis survival into adulthood is common [4]. Clinical features in the neonatal period include central cyanosis due to right to left shunting at the atrial level and depend on the prostaglandin infusion to maintain the patency of the ductus arteriosus. In infants and children, the disease is largely indolent and patients are mostly referred for a murmur detected on routine examination. Adults may present with exertional fatigue, dyspnea, light-headedness and chest discomfort (referred to as right ventricular angina).

Physical examination reveals a prominent jugular *a* wave, a right ventricular lift and a thrill in the second left intercostals space. Auscultation reveals a normal S_1 , a single or split S_2 with a diminished P_2 , unless the obstruction is supravalvular, in which the intensity of P_2 is normal or increased. A systolic ejection murmur with maximal intensity at the left upper sternal border. The murmur duration increases with increasing degrees of stenosis, and its peak intensity occurs later in systole. An ejection click occurs with dysplastic pulmonary stenosis. Cyanosis may be present when a PFO or ASD permits right to left shunting.

Diagnostic tests used in the evaluation for pulmonic stenosis include the electrocardiogram, echocardiography, and cardiac catheterization.

Electrocardiography

In the newborn period may reveal left axis deviation with left ventricular dominance in cases with significant right ventricular hypoplasia. Other patients may have a normal QRS axis. As the stenosis progresses, evidence of right ventricular hypertrophy appears. Severe stenosis is seen in the form of tall R waves in right sided precordial leads with a deep S wave in V_6 . A tall QR wave in the right precordial leads with T wave inversion and ST segment depression (RV strain) reflects severe stenosis.

Chest X-ray

Chest X-ray in the newborn period may show pulmonary oligemia. In infants, children and adults, if there is mild to moderate stenosis, the chest radiograph will reveal a normal sized heart shadow with normal pulmonary vascularity.

Two-dimensional echocardiography

Two-dimensional echocardiography is an excellent modality for

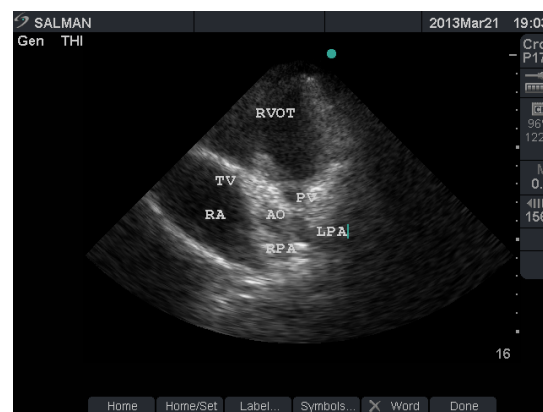


Figure 7: Peak gradient across the pulmonary valve was 140 mmHg and mean gradient was 68 mmHg.

assessment of pulmonary valvular anatomy, localization of the stenosis, and evaluation of right ventricular size and function. Typical valvular stenosis is characterized by mildly thickened leaflets with restricted systolic excursion, leading to a domed appearance. Presystolic doming of the valve may be seen during atrial contraction in patients with more severe stenosis who have a noncompliant right ventricle [5]; doming correlates with the progressive decrease in the systolic ejection click [6]. Truly dysplastic valves are characterized by markedly thickened and seemingly immobile leaflets, hypoplasia of the valve annulus, and often superannular narrowing [7].

Continuous wave Doppler measurement

Continuous wave Doppler measurement of peak systolic velocities provides estimates of the transpulmonic gradient that are remarkably accurate when compared with simultaneous values obtained at cardiac catheterization [8,9]

Cardiac catheterization

Diagnostic cardiac catheterization is rarely required. Invasive hemodynamic measurements and ventriculography may be useful when the severity of stenosis is unclear or a significant secondary infundibular stenosis is suspected in addition to the valvular stenosis [10].

Management and outcomes

Mild and moderate degrees of pulmonary stenosis (peak gradient <40 mmHg) are well tolerated and generally do not require intervention [11]. In one study, 94% patients of mild pulmonary stenosis were asymptomatic, without cyanosis or congestive heart failure [12]. Valvulotomy has been shown to improve morbidity and mortality and is indicated with these gradients. The newer modality nowadays is the transcatheter balloon valvuloplasty [13].

Conclusion

Thus, pulmonary stenosis, although a rare entity, has to be detected early because outcomes are good with early intervention and the natural history of the disease is quite conducive with interventional therapy. We present this case here because our patient had presented with main symptom of cyanosis at 15 years of age which is an unusual

presentation. It may be because of the severity of pulmonary stenosis and RV dysfunction, as critical PS will not have survival upto 15 years of age and patient would not have been so asymptomatic throughout the first decade of life.

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