Case Report

Congenital absence of pulmonary valve
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Introduction
Congenital absence of pulmonary valve presents a characteristic clinical, physiologic and pathologic syndrome.

Absent Pulmonary Valve (APV) is defined as total or subtotal absence of pulmonary valve leaflets.

Ruttenberg and co-workers commented on 25 cases from 1908 to 1964 and Felix Wyler and co-workers presented 15 observations from 1964 to 1969.

APV can be associated with simple or complex cardiovascular malformation. It occurs as an isolated anomaly in non-syndromic patient or as a part of a genetic syndrome in syndromic patient. The most common association is APV and Fallot's tetralogy with absence of duc tus arteriosus, Fallot type APV. APV with intact ventricular septum or muscular VSD with persistent Ductus Arteriosus is less common. Non Fallot type APV/ADA.

The main symptoms of APV are pulmonary insufficiency and bronchial obstruction. The overall frequency of APV is not known. But the Fallot type APV/ADA may be estimated to occur in 6:3000 live born infants with congenital heart disease.

William Osler said there should be no teaching without a patient for a text. This can focus attention on the condition itself as well as what we should do to help the patient.

Aneurysmal dilatation of the pulmonary artery may result either from a congenital weakness at the base of the pulmonary artery or from the hemodynamic effect of a ventricular septal defect and infundibular stenosis in the presence of pulmonary regurgitation.

Case history
An 11 year-old young lady noted exertional dyspnea from her early childhood and was treated with bronchodilators. On examination of the chest, a parasternal heave was palpable, auscultation revealed a single second heart sound, a grade 3/6 harsh systolic murmur, and heart beat at the left sternal angle. The extremities were cool and cyanosed.

A chest radiograph demonstrated cardiomegaly, a grossly enlarged left pulmonary artery, a right aortic arch and narrowing of the distal trachea (Fig 1).

Figure 1: Chest X-Ray showing grossly enlarged left pulmonary artery (long arrow). Narrowing of the trachea is evident just above the carina (paired arrows). The aortic knob can be seen arising to the right of the trachea (short arrow).

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The ORION Medical Journal 2008 Jan;29:537-538
An electrocardiogram showed sinus tachycardia with signs of right atrial abnormality and right ventricular hypertrophy (Fig 2).

Two dimensional and Doppler Echocardiogram disclosed normal LV systolic function with a large ventricular septal defect, dilated right ventricle with thickened right ventricular wall and narrowing of right ventricular outflow tract [RVOT] (Fig 3).

Cardiac catheterization done in 2006 had documented on elevated right ventricular pressure (RV) of 135/20, a pulmonary artery pressure of 24/12 and a left ventricular pressure of 135/20. Oxymetry confirmed a R>L ventricle shunt with a pulmonary to systemic flow ratio 2.2:1. A left ventriculogram demonstrated overall normal LV function with a ventricular septal defect (VSD) in membranous septum. A right ventriculogram showed RV outflow tract (RVOT) narrowing and marked dilatation of the left pulmonary artery. Ventricular septal defect with pulmonary valve stenosis was diagnosed.

The VSD was closed with a PTFE patch and the RVOT was reconstructed with placement of a 22 mm homograft valve in the pulmonary valve position.

The patient had dramatic improvement postoperatively, with immediate resolution of symptoms and signs of RV failure and reduction of the heart rate from 120-130 to 70-80 beats per minute by postoperative day 2 (Two).

**Discussion**

Congenital APV was first described by Chever in 1847. It is defined as total or subtotal absence of the pulmonary valve leaflets. This may be the result of error of complete failure in valve development.

Mild stenosis of the pulmonary artery orifice and aneurysmal dilatation of the main pulmonary artery as well as of the right and left or both pulmonary artery branches co-exist.

Compression of the major bronchi at the hilum is a secondary phenomenon and is assumed to develop in fetal life. In addition, compression of the intrapulmonary bronchi by abnormally branching pulmonary arteries may represent a serious complication.

The definite confirmation of an absent pulmonary valve can only be obtained by surgery or necropsy. The diagnosis, however, based on the presented data can be presented to correct since full correlation between auscultatory, angiocardiographic findings was established.

In our patient, the diagnosis was made by surgery and the present postoperative good clinical condition seemed not to warrant any...
surgical intervention. The surgical outcome of APV was closely related to preoperative ventricular dependency. Martin et al (2006) named 36 patients underwent surgical correction from 1979-2004, 28% were ventricular dependent, while 72% were underwent repair electively11.

The spectrum of malformations to which APV may be associated can be categorized into two types of APV with or without VSD6.

**APV with VSD**
The most common form with VSD is the association with TOF. The similarities with TOF include:
* Anterior deviation of the infundibular septum in relation to the muscular septal crest
* Malaligned VSD
* Overriding of the Aorta

The distinguishing features are:
* Unobstructed right ventricular infundibulum
* Aneurysmal dilated pulmonary arteries.

Typically absent Ductus Arteriosus is reported. It is an uncommon condition comparing 2-6% of all cases of TOF6.

Differential diagnosis of an aneurysmal dilatation of the pulmonary trunk include APV, agenesis of the ducutus arteriosus, truncus arteriosus, ventricular aneurysm and ventricular diverticula10.

**APV without VSD**
In this less frequent form without VSD, muscular VSD may be rarely observed. The association of PDA is reported4.

**Conclusion**
Surgical correction of this severe pathology has a high mortality rate (20-35%)12. The success of corrective heart surgery is in fact determined in more severe cases by progressively resolving respiratory failure preoperatively and therefore the reliance on mechanical ventilation12.

**References**