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Case Report

A window that is lost in time which is defying the odds – "A Rare Case of Pulmonary Artery Hypertension"

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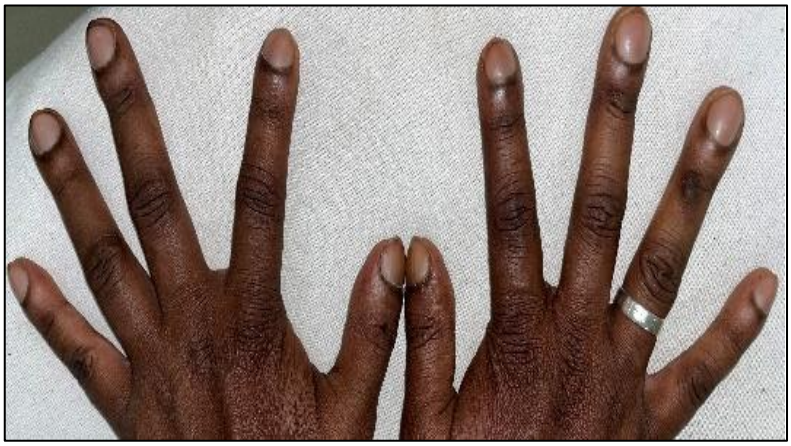
Abstract
Aortopulmonary window is an uncommon congenital heart disease that can increase pulmonary vascular resistance and result in heart failure and recurrent respiratory infections. The condition is often fatal in childhood. Here, we present a case of a 35-year-old male with a large aortopulmonary window, complicated by Eisenmenger syndrome, who has survived without surgical intervention for past 35 years.
Keywords: *Congenital heart disease, Pulmonary vascular resistance, Aortopulmonary window, Eisenmenger syndrome, Heart failure, Differential ABG, Right heart catheterization, Vasoreactivity, Endothelin receptor antagonist, Phosphodiesterase 5 inhibitor, Tunnel flap, Pan-digital clubbing, Central cyanosis, Biventricular hypertrophy.*

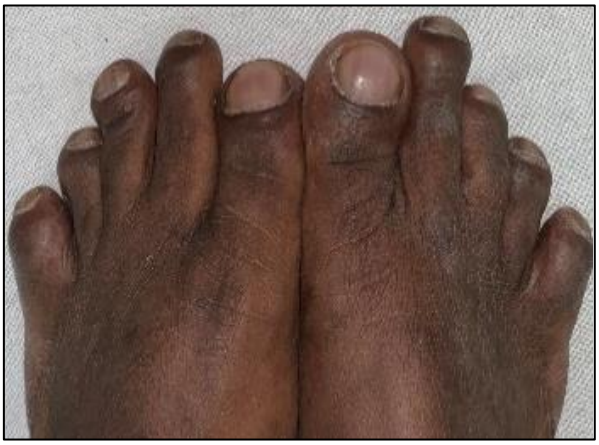
Introduction

Aortopulmonary window (APW) is an uncommon congenital heart defect, representing approximately 0.2% to 0.6% of all congenital heart conditions[1]. It is characterized by an abnormal communication between the ascending aorta and the main pulmonary artery due to incomplete embryonic development. Most infants with APW experience congestive heart failure due to a significant left-to-right shunt[2]. The prognosis for untreated large APW is poor, with a 40% mortality rate within the first year of life[3], and only a small number of cases reaching adulthood. We report a case of a 35-year-old male with a large aortopulmonary window, complicated by Eisenmenger syndrome, who has survived without surgical intervention.

Case Report

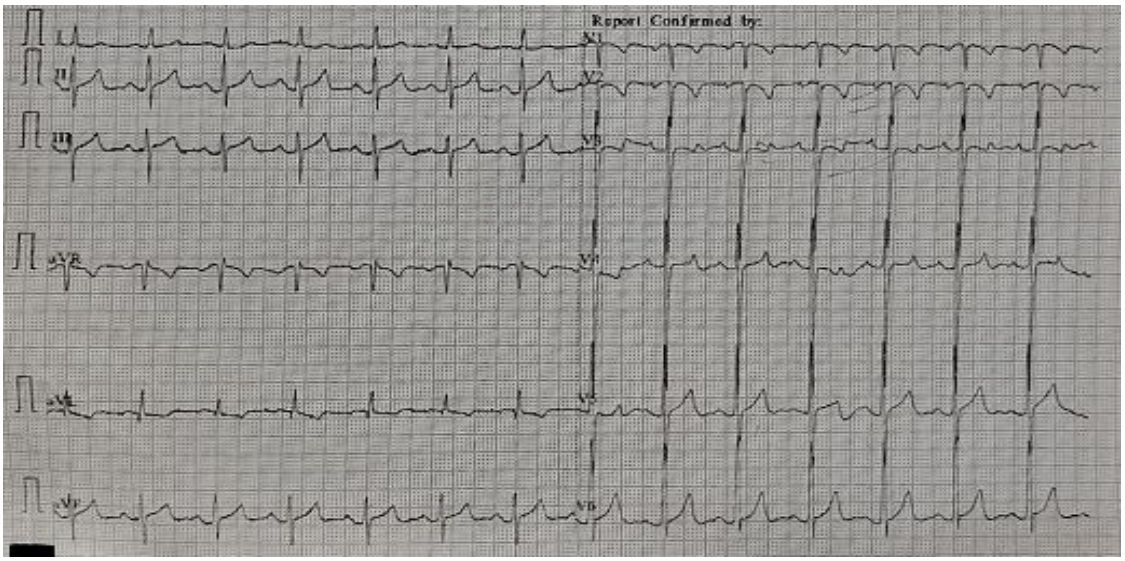
A 35-Year-old male came with the chief complaints of dyspnea on exertion -NYHA class II for past 6 months and class IV for past 15 days associated with retro-sternal chest pain even at rest and cough with expectoration. Clinical examination revealed Pan-digital clubbing, central cyanosis, elevated JVP with a prominent 'a' wave. CVS examination showed loud P2, pansystolic murmur over left lower sternal border.





Evaluation & management

ECG: Sinus rhythm, Left atrial enlargement present, Biventricular hypertrophy present,



CBC showed Polycythemia, Hemoglobin: 20.8 g/dl, PCV: 53%, MCV: 81 fl, WBC: 11,000 cells/mm, Platelets: 4,32,000,

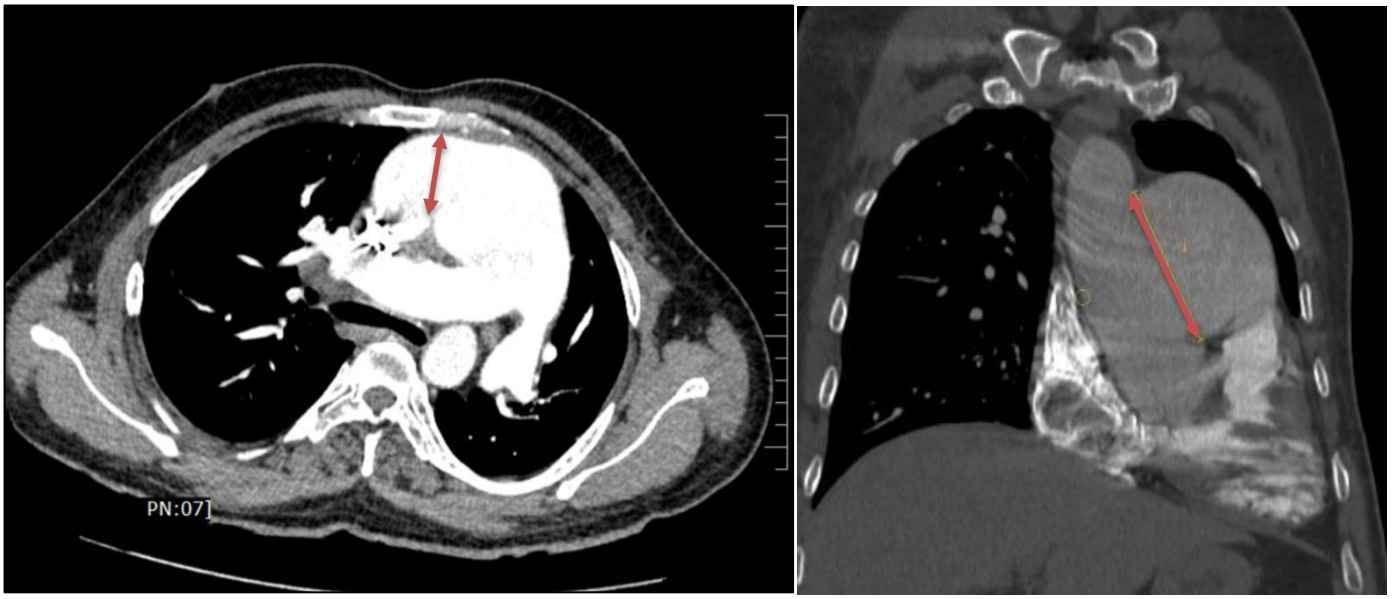
ABG showed Hypoxemia (PO2:59mmHg), Differential ABG: no difference,

Chest Xray showed Apparent cardiomegaly

Transthoracic ECHO showed RA, RV dilated Severe pulmonary hypertension (RVSP-80mmHg), trivial TR, LVEF-60%,

NT-pro BNP was 1400pg/ml

CT Aortogram showed Aorto-Pulmonary window Type III of size 7cms

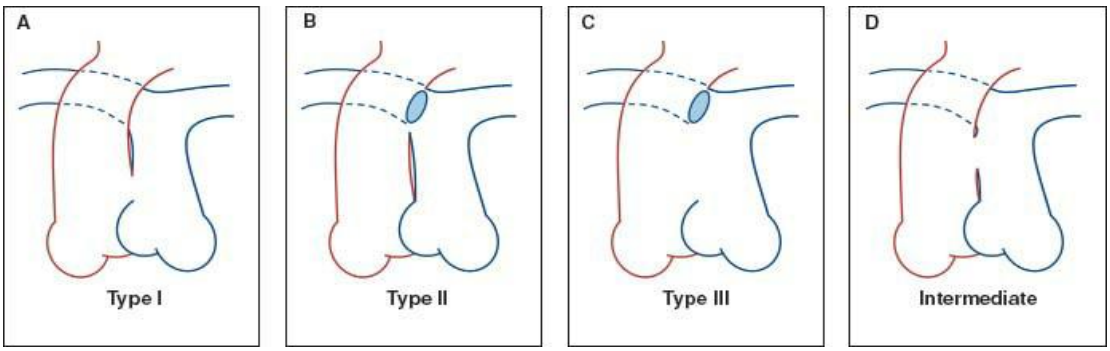


Right Heart catheterization showed Acute vasoreactivity was negative, mPAP50.8mmHg.

Patient was treated with phosphodiesterase 5 inhibitor with endothelin receptor antagonist and after 3 months of follow up patient was symptomatically better (RVSP: 68mmHg).

Discussion

Aorto pulmonary window is a direct communication between ascending aorta and pulmonary artery due to deficiency of aortopulmonary septum. It is a rare form of congenital heart disease, accounting for 0.15% of all congenital cardiac defects and the incidence is estimated at approximately 1 in 300,000 live births, with most cases diagnosed in infancy due to significant left-to-right shunting and early heart failure. However, untreated cases surviving into adulthood, especially with Eisenmengerization, are exceedingly rare[4]. There are four types of AP window Type 1- consists of a proximal defect with very little inferior aortopulmonary septum above the semilunar valves, Type 2- distal defect with absence of the superior septum, Type 3- large defect that spans from the semilunar valves to the pulmonary artery bifurcation, Intermediate aortopulmonary window is a central defect with proximal and distal septal rims[5].



Associated Lesions are frequently occurs alongside ventricular septal defect, tetralogy of Fallot, subaortic stenosis, atrial septal defect, and patent ductus arteriosus. In rare cases, the right and left coronary arteries may be abnormally positioned, originating from the pulmonary trunk.

Early presentation – continuous murmur and signs of left sided heart dilation, infants with large AP window shows features of heart failure, timely surgical repair is required, unless pulmonary hypertension is deemed irreversible. Late presentation–when AP window is large–

Pulmonary artery hypertension, secondary polycythemia, cyanosis, Eisenmenger syndrome and heart failure. When the AP window is small - variable degrees of left ventricular dilation or heart failure without fixed pulmonary hypertension

Evaluation: Electrocardiogram, ABG, Transthoracic/Trans esophageal echocardiogram, CT Angiogram / Contrast enhanced MRI, NT-pro BNP, Right heart catheterization. Adults with unoperated aortopulmonary windows warrant a full diagnostic workup: Confirm the presence of the aortopulmonary window and identify associated lesions, assess the magnitude of left-to-right shunting, assess the degree of pulmonary hypertension, if present, transthoracic echocardiography provides most of this information[6].

If evidence of pulmonary hypertension is present, patients should undergo cardiac catheterization for assessment of pulmonary vascular resistance with reversibility studies before any consideration is given to closure.

Surgical closure of an aortopulmonary window in the adult is a low-risk procedure under cardiopulmonary bypass. Selected patients may be suitable for transcatheter closure.

For adults with repaired aortopulmonary windows, the following points need to be addressed:

1. Confirm the absence of residual communication. 2. Assess associated lesions, if present. 3. Assess left ventricular size and function. 4. Exclude pulmonary hypertension. 5. Exclude important supravalvar pulmonary stenosis, if the tunnel type of repair was employed. 6. Exclude ischemia due to coronary artery ostial obstruction.

Management: Heart failure management, Pulmonary artery hypertension management (Calcium channel blocker, Phosphodiesterase 5 inhibitor and endothelin receptor antagonist), surgical (direct suture/patch/tunnel type flap), palliative care and follow up.

Conclusion

Adults with Aortopulmonary -Window repaired early should have a normal life expectancy. Management depends on the pulmonary artery vasoreactivity –irreversible (Medical) / reversible (surgical). The prognosis of surgically uncorrected AP window is poor and adult survival rate is also very less.

Reference

1. Kutsche LM, Van Mierop LH. Anatomy and pathogenesis of aorticopulmonary septal defect. Am J Cardiol 1987; 59: 443–447.
2. Aggarwal SK, Mishra J, Sai V, et al. Aortopulmonary window in adults: diagnosis and treatment of late-presenting patients. Congenit Heart Dis 2008; 3: 341–346.
3. El Dick J, El-Rassi I, Tayeh C, et al. Aortopulmonary window in adults: a rare entity leading to Eisenmenger syndrome. Echocardiography 2019; 36: 1173–1178.
4. Michael A. Gatzoulis, Gary D. Webb and Piers E.F. Daubeney –Patent ductus arteriosus and Aortopulmonary window (Pg:406) - Diagnosis and management of adult congenital heart disease –3rd edition; 2018
5. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital heart surgery nomenclature and database project: Aortopulmonary window. *Ann Thorac Surg*.2000;69:S44–S49
6. Cui M, Xia B, Wang H, Liu H, Yin X. A Rare Case of Adult Aortopulmonary Window Combined with Anomalous Origin of the Right Pulmonary Artery from the Aorta Leading to Eisenmenger Syndrome. J Int Med Res. 2021 Jan;49(1):300060520984656. doi: 10.1177/0300060520984656.