Recurrent Hemoptysis in 11-year-old girl- Pulmonary Atresia with Ventricular Septal defect and Patent Ductus Arteriosus

MP Ranjith MD, KF Rajesh MD, Nair Anishkumar MD, G Rajesh MD DM, MN Krishnan MD DM

Introduction: Pulmonary atresia and Ventricular septal defect (PA-VSD) often present as cyanotic newborn. Natural history of PA-VSD patients depends on the adequacy of pulmonary blood supply. Here we report an 11 yr-old girl with PA-VSD who presented with recurrent hemoptysis.

Case Report description: An 11 yr old girl presented to us with recurrent hemoptysis since 1 yr. She was a known case of ventricular septal defect with pulmonary atresia but not on any treatment. On examination, her pulse rate was 70 bpm and blood pressure 90/40 mm Hg. There was central cyanosis (spO2 83%), polycythemia (hematocrit of 71%), and digital clubbing. Cardiovascular system examination revealed right ventricular type apex, loud single S2, ejection click and a continuous murmur over infracavicular area. Chest X-ray showed hypoplastic main pulmonary artery. ECG showed north west axis, right atrial enlargement and right ventricular hypertrophy with strain pattern (Figure 1). Echo showed large perimembranous VSD extending to the inlet region with aortic override. Shunt was predominantly right to left. Pulmonary valve atretic with confluent left and right pulmonary arteries supplied by a large patent ductus arteriosus (PDA) from undersurface of aorta (Figure 2). She was managed symptomatically and planned for surgery after detailed imaging studies.

Discussion: In PA-VSD there is lack of continuity between the right ventricle and the pulmonary artery. The pulmonary blood supply is variable may be from systemic pulmonary collaterals or PDA. Most of the patients who survive to adolescence or adulthood have had palliative or corrective surgery, and have developed an alternative blood supply to the lungs through systemic-to-pulmonary collateral vessels (APCs). These collateral vessels are prone to rupture and may result in serious and even fatal hemoptysis. PDA, if present, is an important source of pulmonary blood supply. When ductus arteriosus is patent, generally the native pulmonary arteries will be of adequate size. When a PDA connects with a central pulmonary artery, the peripheral distribution is normal, and there are no MAPCAs in the hemithorax. Thus, a PDA does not coexist with MAPCAs in the same lung. Our patient had confluent pulmonary artery which was supplied by long PDA arising from the inferior aspect of aortic arch.

Implications to clinical practice: It is critical to

*Senior Resident, †Additional Professor, ‡Professor & Head, Department of Cardiology, Government Medical College, Kozhikode

Corresponding Author: MP Ranjith, drranjithmp@gmail.com
map the pulmonary artery architecture and source of blood supply in patients with PA-VSD preoperatively. Color Doppler flow studies are reliable in making the distinction between PDA and APCs based on the direction of blood flow in the proximal pulmonary artery. For complete delineation of pulmonary arterial tree cardiac catheterization is frequently necessary.

Figure 1

Figure 1: A- Chest X-ray PA view showing hypoplastic main pulmonary artery. B- ECG showing north west axis, right atrial enlargement and right ventricular hypertrophy with strain pattern.

Reference


Figure 2

Figure 2: A-Parasternal long axis view showing malaligned VSD. B- Basal short axis view showing atretic pulmonary valve. C- Suprasternal view showing PDA arising from the inferior aspect of aortic arch