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Pulmonary Atresia with VSD and Major Aortopulmonary Collateral Arteries Presenting with Infective Endocarditis: Case Report

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Abstract: Pulmonary valve atresia with a VSD is an extreme form of tetralogy of Fallot (TOF). The majority of untreatedpatients of severe die in their first decade of life as a result of intractable congestive heart failure or respiratory distress.We report on a 18yrs old girl with, pulmonary valve atresia and ventricular septal defect (PA-VSD) with major aortopulmonary collaterals (MAPCAs) along with reformation of RPA & LPA from MAPCA's which is a complex and extremely heterogeneous anomaly. This child was diagnosed with Congenital heart disease for the first time at the age of 18yrs, when she presented with infective endocarditic.
Keywords: Pulmonary atresia; Ventricular septal defect; Infective endocarditis; major aorto-pulmonary collaterals

INTRODUCTION

Pulmonary atresia and ventricular septal defect (PA-VSD) with major aortopulmonary collaterals(MAPCAs) is a extreme form of tetralogy of Fallot (TOF) with complex pulmonary architecture with abnormal size and distribution of pulmonary arteries and Systemic collaterals that supply all or part of lung[1].It's the most severe form of TOF also known as Tetralogy of Fallot (TOF) with pulmonary atresia (PA-VSD), and is estimated to represent 5% to 10% of tetralogy of Fallot patients[2]and also accounts to 2 – 3 % of all the congenital cardiac malformations [1,3-5].

It's a duct dependent lesion of neonate with Survival in the post natal period depending on major aorto-pulmonary collaterals (MAPCAs).The survival rate withoutsurgical repair is as low as 50% at 1 year of age and 8% at10 years [6]. Adult survivors of PA-VSD are quite rare.It is more common in males than in females [7].

CASE REPORT

A 18 year old girl presented with history of moderate grade fever with chills without any other localizing symptoms since 1 month for which girl had been on treatment with multiple antibiotics. She had symptoms of progressive easy fatigability & breathlessness since 5- 6 yrs of age, presently in NYHA grade II- since last 2-3yrs. Also history of bluish discoloration of finger and lips on exposure to coldand poor weight gain since childhood for which no medical attention was seeked till date.

Clinically, she was not toxic, with stunting and wasting. Pulse-108 beats per minute in sinusrhythm, with regular good volume pulse in all four limbs. BP 110/70 mm of Hg, Respiratory rate 26/min & SPO2 80% on room air in all four limbs with JVP not raised. B/L non purulent Conjunctivalcongestion was present with peripheral cyanosis and clubbing was noted (Fig. 1). Cardiac evaluation showed tapping apex at 5th left Intercostal space (ICS) lateral to mid clavicular line with Left Parasternal heave. The girl had grade 3 ejection systolicmurmur heard maximallyin the aortic area and another grade 3-4 continuous murmur in the left clavicularregion & infra axillary region. Soft non-tender Liver was palpable 2.5 CM below costal margin with span of 9 cm and spleen 1cm below costal margin.

Laboratory examination demonstrated polycythemia (PCV-45%) with polymorphic predominant leukocytosis, increased acute phase reactants, blood cultures being sterile.Chest X-ray showed-Right sided Aortic arch and boot shaped heart with normal pulmonary blood flow (Fig. 2). ECG suggested of RVH (Fig. 3). Transthoracic 2Dand Doppler echocardiography revealed large VSD, with pulmonary atresia with large Major Aortopulmonary Collateral Arteries (MAPCA) and vegetations seen on aortic valve (Fig. 4).

Cardiac MRI- suggested TOF features- with Sub-aortic VSD and RVH with Rt sided Aortic Arch with PDA and Multiple large MAPCA's. Also there was Atresia of Main pulmonary artery (MPA) with reformation of Rt Pulmonary Artery & Lt Pulmonary Artery from MAPCA's (Fig 5&6).

DISCUSSION

PA-VSD is a cyanotic congenital heart disease characterized by underdevelopmentof the right ventricular (RV) outflow tract (i.e. sub pulmonaryinfundibulum) with atresia of the pulmonary valve, a large VSD, andoverriding of the aorta. In the past, this anomaly was termed pseudotruncusor truncusarteriosus type [3, 8-10].

PA-VSD demonstrates a wide spectrum of severity, depending onthe degree of pulmonary artery development. Pathologically, PA-VSD isfrequently considered the most severe end of the spectrum of TOF but controversy exists as to whether PA-VSD and TOF should be treated as two distinct entities.Unlike PA-VSD, patients with the standard type of TOF with pulmonary atresia have pulmonary arteries that are usually normal in size with normal peripheral pulmonary arborisation[4]. In addition, systemic-to-pulmonary collateral vessels are not as well developedin patients with TOF with pulmonary atresia as they are in patients with PA-VSD[4].

Our patient has a complex congenital heart defect which is characterized by theSub-aortic VSD and pulmonary atresia with RVH withRt sided Aortic Arch with PDA and large Major Aortopulmonary Collateral Arteries (MAPCA) and vegetations seen on aortic valve.

Also there was Atresia of Main pulmonary artery (MPA) with reformation of Rt Pulmonary Artery & Lt Pulmonary Artery from MAPCA's.Survival of PA-VSD patients is dependent on the adequacyof pulmonary blood flow derived from direct orindirect aortopulmonary collateral vessels. The well-developed MAPCAs might have enabled our patient to survive withoutmuch symptoms till the time she landed with infective endocarditis. Becauseof the increased survival rate of children with congenital heart disease (CHD) and the overall decrease in rheumatic valvular heart disease in developed countries, CHD now constitutes the predominant underlying condition for Infective Endocarditis in children [11].

Like in our patient infective endocarditis features may be the first presentation of the patient stressing on the importance of detailed evaluation of patients with pyrexia of unknown origin. In our patient blood cultures were sterile may be because of multiple antibiotics the girl had been. The management of infective endocarditis is similar to those in adultendocarditis. Antimicrobial prophylaxis is particularly important in these children.

This case has been reported as the rare complex congenital heart disease went undetected till 18 years age and presented as Infective endocarditis, without any invasive procedure.

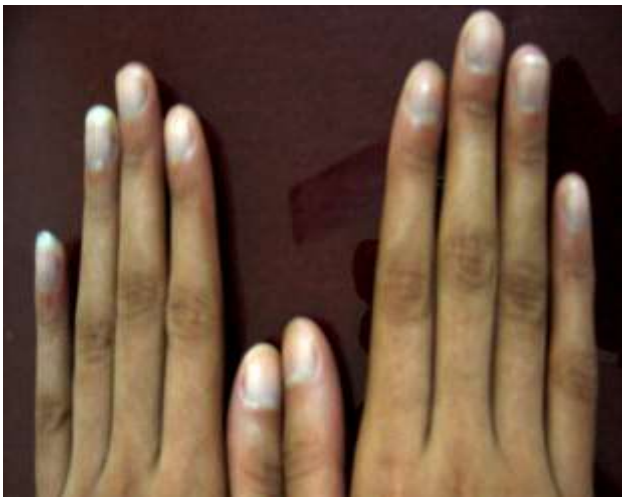

Fig. 1: Clubbing and cyanosis

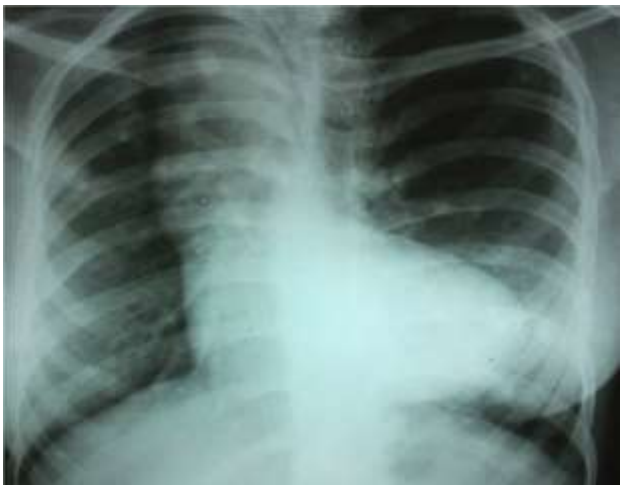

Fig. 2:Chest X-ray showing –Rt sided Aortic arch and boot shaped heart with pulmonary blood flow

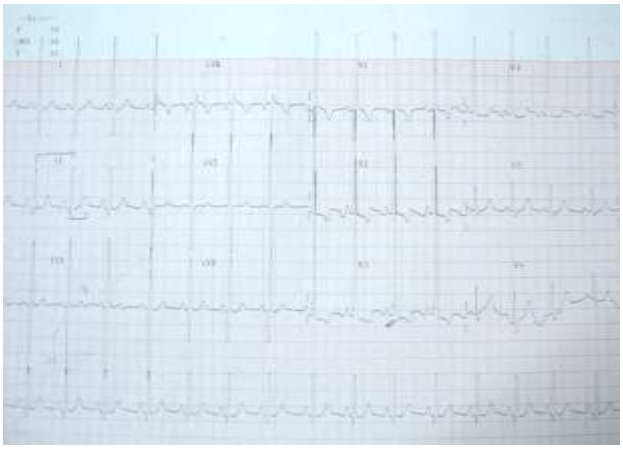

Fig. 3: ECG showing features of Right ventricular hypertrophy

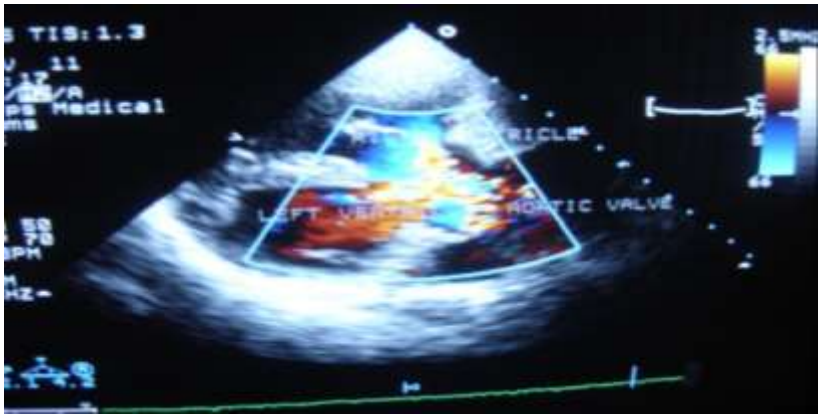

Fig. 4: Transthoracic 2D and Doppler echocardiography revealed large VSD,with pulmonary atresia with large Major Aortopulmonary Collateral Arteries (MAPCA) and vegetations seen on aortic valve

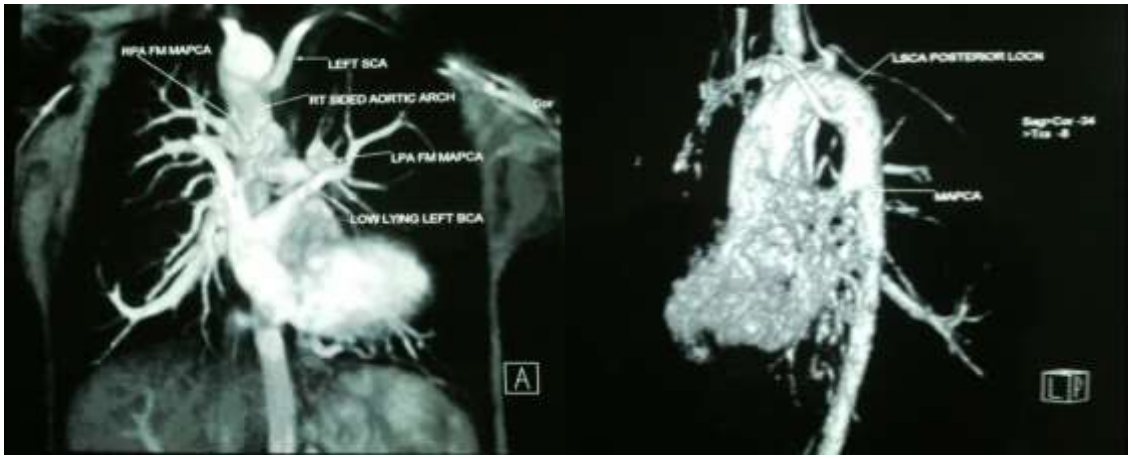

Fig. 5 & 6: Cardiac MRI- showing TOF features- with Sub-aortic VSD and RVH with Right sided Aortic Arch with PDA and Multiple large MAPCA's. Also there was Atresia of Main pulmonary artery (MPA) with reformation of Right Pulmonary Artery & Left Pulmonary Artery from MAPCA's.

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