Rare Case of APW Type3 with Successful Surgical Closure-A Case Report

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Abstract

Aortopulmonary window is a rare congenital anomaly. Majority of cases present in early childhood with symptoms of heart failure, recurrent respiratory infection and failure to thrive. Very rarely they remain undiagnosed till adolescence or rarely still to adulthood. We report a case of 12 yr old Fch who presented to us with symptoms of heart failure and history of recurrent respiratory infection. Examination revealed cachectic child with indrawn lower ribs. Cardiac apex was shifted lower and lateral to its normal position. Heart sounds were prominent with a soft third heart sound. A loud systolic murmur with rough quality was audible over the precordium which was spilling over the second sound. TTE revealed dilated left heart chambers. An echo dropout of approximately 13 mm was noticed between aorta and pulmonary artery with flow from left to right was demonstrated. Associated defects were looked for and ruled out. With a diagnosis of APW-type3 patient was taken for surgical closure which was done successfully. Post-surgical echo revealed intact patch in situ with no residual shunt. Mild RPA stenosis was detected in the post op echo which is haemodynamically significant and is to be followed up. Patient was discharged after successful surgery and is being followed up.

Keywords: Aortopulmonary Window; Heart Failure; APW-type3

Introduction

Aortopulmonary window is a rare congenital anomaly. Majority of cases present in early childhood with symptoms of heart failure, recurrent respiratory infection and failure to thrive. Very rarely they remain undiagnosed till adolescence or rarely still to adulthood. We report a case of 12 yr old female who presented to us with symptoms of heart failure and history of recurrent respiratory infection, who was successfully operated at our center.

Case History

A 12 year old female child was brought by her parents with complaints of shortness of breath of class III severity. She gave history of similar symptoms on and off which gets aggravated sometimes and relieved by treatment from local physician. Her current medication included digoxin, diuretics, antibiotics and iron supplementation. On examination she looked cachectic with cachectic indrawn lower ribs. Cardiac apex was shifted lower and lateral to its normal position. Heart sounds were prominent with a soft third heart sound. A loud systolic murmur with rough quality was audible over the precordium which was spilling over the second sound. Auscultation of lungs revealed fine basal creeps. TTE revealed dilated left heart chambers. An echo dropout of approximately 13 mm was noticed between aorta and pulmonary artery with flow from left to right was demonstrated (Figure 1 and 2). Associated defects were looked for and ruled out. With a diagnosis of APW- patient was taken for surgical closure which was done successfully. Post-surgical echo revealed intact patch in situ with no residual shunt. Mild RPA stenosis was detected in the post op echo which is haemodynamically not significant and is to be followed up (Figure 3 and 4). Patient was discharged after successful surgery and is being followed up (Figure 5).
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Aortopulmonary window is an uncommon anomaly, occurring in less than 1% of all persons with a congenital heart disease and results from an incomplete development of the conotruncal septum. In half of the cases aorto-pulmonary windows are associated with other anomalies such as: tetralogy of fallot, pulmonary atresia, atrial septal defect, interrupted aortic arch, truncus arteriosus, patent ductus arteriosus. Fifty percent of patients usually have no other heart defects. It consists of a communication between the ascending aorta and the pulmonary trunk and/or the right pulmonary artery. Depending upon location of the defect it can be classified into different types, namely Type I, or proximal: the communication is usually rounded and located above the semilunar valves, between the ascending aorta and the pulmonary trunk. Type II, or distal: the defect, in a spiral curve, involves the pulmonary bifurcation at the level of the right pulmonary artery (RPA). In this type of defect, there can be hypoplasia or even aortic arch interruption due to the significant deviated flow from the aorta towards the RPA during fetal life. Type III is characterized by total absence of the aortopulmonary septum resulting from
the combination of proximal and distal defects. Babies that have a hole in between the aorta and pulmonary artery have blood from the aorta that flows into the pulmonary artery, and as a result too much blood flows to the lungs. This causes high blood pressure in the lungs (a condition called pulmonary hypertension) and congestive heart failure. Symptoms can include: delayed growth, irritability, rapid heartbeat, heart failure and recurrent respiratory tract infections. Patients with large APW usually have symptoms of pulmonary hypertension and congestive heart failure (tachypnea, diaphoresis, failure to thrive, and recurrent respiratory difficulty) in the first weeks of life. Severe pulmonary vascular hypertension can occur in the first months of life. Without surgical correction, 40 - 50% of patients will die due to congestive heart failure during the first year of life, and a large number of survivors will suffer from sequelae of congestive heart failure or pulmonary vascular disease in their lives. Very few cases of those who have survived to adulthood and been operated on successfully have been described. In a retrospective study of consecutive adult patients with aortopulmonary window treated at a tertiary charitable cardiovascular institute in South India between 1996 and 2006, Six adult patients successfully underwent aortopulmonary window closure. Although reasons for such adult survival are unknown, in our case initially undetected branch pulmonary stenosis which was detected after the patch closure in post-operative TTE, might have been protective. Majority of APW cases are dealt with surgical closure. But some select cases of type II APW can be closed by device closure technique. In our case surgical closure was done successfully. Post-operative TTE revealed successful closure of the defect without any residual shunt. LPA showed haemodynamically significant stenosis, and we postulate that the same might have remained undetected in pre-operative TTE and is the reason why the child survived till adulthood. She is in close follow up and doing reasonably well [1-5].

Conclusion

Aortopulmonary window is a rare congenital anomaly which presents in infancy and early childhood. But some rare cases may remain undetected and present later in adolescence or rarely in early adulthood. Successful closure by surgical or device closure technique is safe when feasible.

Bibliography

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