A rare case of associate surprising aortopulmonary window

Alessandra Narciso Garcia Department of Surgery, University of California-Los Angeles, Los Angeles, California, USA acarvalhob@yahoo.com

Keywords

Rare disease; Aortopulmonary window; Tetralogy of Fallot

Abstract

Aorto-pulmonary window is a rare heart disease occurring in 0.1-0.2% of the patients with the congenital cardiac disease and it results from an incomplete development of the conotruncal septum. Half of the cases aorto-pulmonary windows are associated with the other anomalies. This condition can occur on its own or with other heart defects such as: tetralogy of fallot, pulmonary atresia, atrial septal defect, interrupted aortic arch, truncus arteriosus and patent ductus arteriosus. 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, infections of the lungs. We describe a case of aortopulmonary window incidentally found during surgery for a Type an interrupted aortic arch and repaired uneventfully. In our case the anomaly was not diagnosed until the thoracotomy but anatomy allowed surgeon to perform the closure of the window.

Introduction

Aortopulmonary window may be a rare internal organ anomaly occurring in zero.1-0.2% of patients with inherent internal organ illness Associate in nursing results from an incomplete development of the conotruncal septum. Aortopulmonary window is outlined because the presence of a communication between 2 nice vessels, the aorta and also the main arterial blood vessel or the proper arterial blood vessel.

Case Description

A three weeks previous boy was admitted to the inherent viscus Surgery Unit with the diagnosing of sort AN interrupted aorta, patent blood vessel and bodily cavity congenital heart defect. Surgical indications were confirmed. In theatre, when the median sternotomy and therefore the longitudinal pericardiotomy, a little sort one aortopulmonary window (APW) according the Richardson et al. classification was incidentally detected. Each the arteria and pulmonic ends were ligated (Figure 1) and therefore the defect transacted.

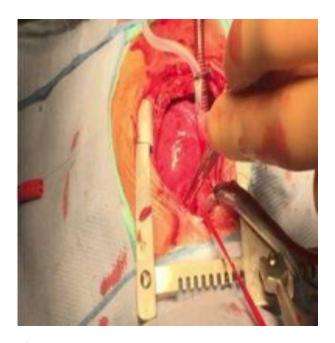


Figure 1: Aortic and pulmonary ends ligation.

Discussion

These anomalies area unit terribly rare and might be associated, as in our case, with different anomalies just like the interrupted aorta, bodily cavity congenital heart defect and series of Etienne-Louis Arthur Fallot. In literature there are a unit many classifications of APW and one among the foremost used is created by Richardson et al. that describe 3 sorts of APW: Type I: typical aortopulmonary congenital heart defect or window; Type II: distal aortopulmonary septate defects within which the aorta communicates with the origin of the proper arteria; Type III: abnormal right respiratory organ artery origin from aorta, or hemitruncus arteriosus. Diagnostic technique is rare. It is often done solely in patients while not internal organ connections abnormalities prenatally on the quality three-vessel read. The aorto-pulmonary window could be a rare malformation that consists of associate alteration within the formation of the aortopulmonary septum and is designed with a communication between aorta and arterial trunk within the presence of adequate crescent valves. Its lesion could also be isolated or together with different advanced internal organ abnormalities. The defect is mostly settled between the lateral left facet of the arterial blood vessel and also the left facet wall of the arteria. The diameter of the defect is varied, however generally it's of such size as to not be restrictive. A selected syndrome that deserves to be remembered is that the combination of a large respiratory organ arteria window, with the proper arteria that originates severally from the lateral posterior wall of the aorta, interruption of the arteria artery and patent blood vessel duct. we tend to still recall the chance of abnormal coronary origin which will arise from the window or arteria. e aortopulmonary window from a pathophysiological purpose of read behaves sort of a major blood vessel duct that causes severe myocardial infarct from early infancy. With a precise frequency, patients with aorto-pulmonary window, World Health Organization area unit already in operative part, to administer stability to vas conditions got to be treated with resuscitation maneuvers: inotropic support, autacoid infusion and aided mechanical ventilation. Surgical correction, even during this anomaly, is silent with the instant of designation unless the defect is actually restrictive . associate incision right at the window at the front 1/2 the defect permits glorious exposure and facilitates the closure with patches of arteria and respiratory organ sinus, paying specific attention to their reconstruction to avoid obstructions or during this patient, the massive shunt that's created for the naturally wide

window size develops a respiratory organ preventive illness among the primary year of life. Due to the autumn in respiratory organ resistance, clinical conditions, shortly after the time of life, area units expected to deteriorate apace. If the respiratory organ circulation doesn't fall, heart muscle insufficiency is going to be a reason behind fast clinical deterioration, however the expectancy of those patients remains restricted over time thanks to severe respiratory organ hyper flow. Associated lesions will contribute to considerably sterilisation explanation. If there's arteria interruption, these patients now when their birth area unit space encountered during a large cardioci rculatory collapse. Specific operative issues area unit respiratory organ cardiovascular disease, residual shunt and respiratory organ hypertensive crisis. In current literature there's not enough data regarding APW, and it's potential to mention that the sole tools offered for postpartum designation area unit graph. sonogram and hearing, constant instruments used for medical diagnosis. In fact, a medical diagnosis with patent blood vessel and customary truncus arteriosus are often done by the presence of the signs like arteria cardiac murmur and with signs of left bodily cavity hypertrophy on graph. Also, the standard of the peripheral pulse, further because the loud beat murmur, are often wont to diagnose APW. e presence of right bodily cavity hypertrophy, or combined bodily cavity hypertrophy, in associate child with stenosis suggests complicating associated right-sided anomalies. In our case APW wasn't diagnosed till the thoracotomy and then the treatment was unprogrammed, however the anatomy of the anomaly allowed the doctor to perform the closure of the windows. The main operation was carried on habitually and also the course was placid. The patient was admitted to the unit with lowest inotropic support and his recovery was satisfactory while not complications. At followup twelve months when repaired the patient denied any symptoms or complication.

Conclusion

The surgery for correction of APW has evolved over years, currently an open repair on cardiopulmonary bypass (CPB) with a single patch technique yields good results. Mortality is

Extended Abstract

affected by association of pulmonary hypertension and other cardiac malformations.