

Double Aortic Arch with Double Coarctation: Presentation and Management

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Received: November 17, 2017; **Published:** November 30, 2017

Newborns presenting with acute respiratory distress, especially in a week or two following discharge home should be evaluated for congenital cardiac anomalies [1]. One of the best ways to screen them would be to perform upper and lower extremity blood pressure measurements. If there is a gradient, then it is likely a coarctation of the aorta. The presentation of respiratory distress and failure is secondary to the closure of PDA in the first one or two weeks following birth and the resultant lactic acidosis from the decreased perfusion to the lower body. Chest X-ray (CXR) shows cardiomegaly and pulmonary edema and electrocardiography (ECG) may show extreme right axis deviation and absent left ventricular forces in the left precordial leads.

Emergent management includes performing a blood gas and simultaneously securing the airway with endotracheal intubation and hemodynamic resuscitation with an infusion of PGE and fluid boluses to restore perfusion. An echo would help determine the degree of myocardial dysfunction resulting from the sudden increase in afterload on the heart. Milrinone infusion should be slowly introduced to help the heart muscle relax. Correction of the metabolic acidosis with sodium bicarbonate infusions will help improve cardiac contractility.

A complete echocardiogram in most cases helps determine that the patient has coarctation of the aorta and whether or not the left ventricle and aortic arch are hypoplastic. The presence of abnormal aortic valve, a carotid subclavian index < 1.1, Isthmus/Ascending aorta ratio < 0.53, and Distal thoracic aorta/Ascending Aorta ratio < 0.6 suggest the presence of coarctation of the aorta [2-4]. If there is a variable presentation, then a cardiac CT angiogram is recommended to better define the anatomy and degree and levels of obstruction in the aortic arch. Very rarely, some patients present with a double aortic arch and both arches appearing to be with coarctation [5]. There is usually a patent ductus arteriosus providing blood flow to the distal aorta and its closure would be catastrophic to the patient.

Management of double aortic arch with double coarctation involves a median sternotomy and aortic clamping, followed by ligation and division of the right aortic arch which is actually on the left side of the patient and resection and end to end anastomosis of the left aortic arch which is on the right side of the patient and is in the anatomically correct position. The PDA obviously is ligated and divided. Patients usually respond well to this surgery unless there was an episode of cardiorespiratory arrest before a diagnosis is made. In which case they may end up needing a feeding tube and, or a tracheostomy with or without ventilator support. The presence of a double aortic arch sometimes forms a complete vascular ring like anomaly which can present as stridor or trachea bronchomalacia in some cases.

Evaluation for genetic abnormalities like DiGeorge syndrome should be made with a karyotype, FISH analysis and microarray. An abdominal ultrasound would reveal any evidence of heterotaxy.

In the long term, hypertension and recoarctation of the repaired or nearby segments will need follow up and interventions. A balloon dilation in the cardiac catheterization lab is usually first attempted before committing to another thoracotomy and patch augmentation [6].

For early recognition of these cases, performance of upper and lower extremity oxygen saturations helps determine the presence of any congenital cyanotic heart disease. This can easily be performed in the nursery. If there is any abnormality noted, then we should proceed to check four extremity blood pressure check and potentially an echocardiogram to screen for any congenital cardiac conditions.

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Volume 6 Issue 4 November 2017

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