Case Report

A Neonate with Berry Syndrome
(AP Window with Interrupted Aortic Arch)

Rasoul Abdulrahman MD1*, Mahmod Samadi MD2, Sohrab Negargar MD3

1. Dept. of Cardiac Surgery, Madani Heart Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.
2. Dept. of Pediatric Cardiology, Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.
3. Dept. of Anesthesiology and Intensive Care, Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.

(Received 25 Jul 2009; Accepted 22 Aug 2009)

Abstract

Full term neonate patient, about 3 kg weighs, admitted in child hospital because of Respiratory distress syndrome (RDS). Investigation by echocardiography revealed large aorto-pulmonary window (AP window) with severe coarctation of aorta. Findings at the time of operation were large AP window and interrupted aortic arch with patent ductus arteriosus (Berry syndrome). The anomalies corrected at the same operation. Unfortunately severe acidosis; brain and pulmonary edema, not responding to medical therapy contribute to patient’s death.

*Corresponding Author: Rasoul Abdulrahman MD, Madani Heart Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.
Tel: 0 9359542672  E-mail: dr_rasol@yahoo.com

Keywords: Aorto Pulmonary Window • Berry Syndrome • Interrupted Aortic Arch
Introduction

AP window is a rare malformation and the complex Berry syndrome variant is more so. At the Hospital for sick children in Toronto, only 23 (0.15%) of the 15,104 patients with congenital heart disease seen over a 20-year period had AP window. There is no known tendency for AP window to close spontaneously. An AP window is usually a large, oval defect between the aorta and pulmonary trunk, although in about 10% of patients the defect is small. The pulmonary arteries are normally related to the pulmonary trunk. Rarely, there is a complex syndrome of the AP window, usually in the downstream portion of the ascending aorta, with aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus, and interrupted aortic arch or severe coarctation (Berry syndrome). This is a particularly lethal combination; most affected infants die shortly after birth. In all, from one third to half of patients with AP window have a major associated cardiac anomaly, including the above and ventricular septal defect (VSD), tetralogy of fallot, transposition of the great arteries, anomalous origin of a coronary artery, aortic isthmic hypoplasia and sub aortic stenosis.

Case report

A full term mal neonate; about 3kg weighs; admitted in NICU department of child hospital with RDS diagnosis, in which Examination reveled symptoms and signs of heart failure with a continuous murmur; so the patient underwent echocardiography, in that subject findings include: Sever coarctation of aorta, small patent ductus arteriosus (PDA), hypoplastic isthmus, Large AP window. Left ventricular ejection fraction was nearly 60%. The patient is then referred to our center for further management and consideration for urgent cardiac surgery. During admission the patient's hemodynamic was unstable, in which case impending to shock; so intubation was done immediately. Electrocardiogram showed normal rhythm, Right axes deviation, and mild right ventricular hypertrophy. Chest radiography findings include: increased cardiothoracic ratio, congestion of the lungs, and pulmonary edema. There were no changes in hematological and biochemical tests, with normal electrolytes. After median sternotomy and pericardiectomy, mediastinum explored, aorta and pulmonary arteries were obvious and intact at their origin. Right pulmonary artery originated partially, from the downstream portion of the ascending aorta as well as connection with common pulmonary artery. The ascending aorta is then come to end by giving three branches including, brachiocephalic artery, left common carotid artery, and left subclavian artery. There was not evidence of descending aorta. Proximal part of patent ductus arteriosus with left pulmonary artery origin also detected at the same time. Based on relay to the echocardiography report of sever coarctation of aorta; we stopped the exploration; aorta canulated just before (proximal) to the brachiocephalic artery and cardiopulmonary by pass initiated. The repair was done through the aorta, opened transversely at the level of the AP window. Partially right pulmonary artery connected to the aorta released, and end of the right pulmonary artery anastomosed to the distal and side of the common pulmonary artery. The aortotomy incision is closed with two rows of continuous polypropylene sutures. Then patent ductus arteriosus followed, surprisingly its distal end was the beginning of the descending aorta, and at this stage of operation interrupted aortic arch confirmed (then coarctation of aorta excluded). We cut proximal end of PDA, beveled it and sutured to the side of ascending aorta just proximal to the brachiocephalic branching at the same operation. Finally layers were closed anatomically. The patient labored badly post operation in the ICU; because of intractable sever acidosis as well as brain and pulmonary edema the first day after operation; lead to cardio respiratory arrest, without response to advanced cardio pulmonary resuscitation. (Fig)

Fig - Sever Post Operation Face, Head and Neck Edema of Full Term Neonate with Berry Syndrome.
Discussion

Because of coexisting cardiac anomalies, the basic technique of repair must be modified and adapted to the individual situation. However, every effort should be made to accomplish a one-stage repair as it done in our case. Based on surgical literature, time-related survival is excellent when the operation is performed in infancy. Mortality is low after repair of AP window unless unusual circumstances are present. Even with coexisting major anomalies, risk of total repair may also be low. Inappropriate preservation of our case; because of echocardiography faults and/or disorientation to the anomaly after incomplete exploration at the operation time, lead to unfavorable results with high flow rate to the brain and low flow rate to other parts of the body. Patient died from post operative sever acidosis, not responding to the medical therapy; as well as brain and pulmonary edema.

References

4. Coleman En, Barcloy Rs, Reid JM, Stevenson JG, Congenital aorto pulmonary fistula combined with PDA, Br Heart J 1965; 29: 571.