Case report

Coarctation of the aorta with persistent pulmonary hypertension

Mahmuda Hassan[1], Kona Chowdhury[2], Taslim Uddin Ahmed[3], Marium Begum[3], Ashraf Uddin Ahmed[3], Asif Zaman Tushar[4], Hamidur Rahman[5]

Abstract:

Coarctation of the aorta (CoAo) is the fifth most common congenital heart defect, accounting for 6–8% of live births with congenital heart disease, with an estimated incidence of 1 in 2,500 births1. The age at which people with CoAo are diagnosed depends on the severity of the condition. If the aortic coarctation is severe, it is usually diagnosed during infancy. A 7 month male baby was admitted at Ad-din Medical College and Hospital with the complaints of difficulty in breathing, lethargy, vomiting, and cyanosis with history of poor feeding. He had past history of transient cyanotic attack 3 to 4 times in his life which was resolved automatically at home. At the age of 3 months of his age he was admitted in other hospital for cough and cold and respiratory distress. X-ray chest and colour Doppler cofirmedthe case as CoAo.

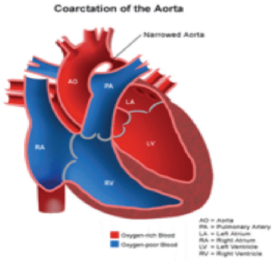
Key Words: Coarctation of the aorta (CoAo), Persistent Pulmonary Hypertension (PPHN).

Introduction

Coarctation of the aorta (CoAo) is the fifth most common congenital heart defect, accounting for 6–8% of live births with congenital heart disease, with an estimated incidence of 1 in 2,500 births[1]. It affects more male babies than female, with a reported male: female ratio between 1.27:1 and 1.74:1.[2,3] Older infants and children remain asymptomatic resulting in delayed diagnosis. In young children, CoAo may present with hypertension, and/or murmurs. The age at which people with CoAo are diagnosed depends on the severity of the condition. If the aortic coarctation is severe, it is usually diagnosed during infancy.

Etiology

The etiology of the discrete isthmic constriction of the aorta in patients with CoAo remains very much in dispute. Although familial cases have been reported[4,5] and association with various gene deletions described[6]. of blood flow.[7] Abnormal migration patterns of the developing aortic arch, and excessive distribution of Developmental theories have focused on abnormalities arterial duct-like tissue around the aortic isthmus[8] has also been proposed. But this mechanistic view do not reflect the widespread changes seen both in left heart structures (mitral valve abnormalities, bicuspid aortic valve) and upper body vascular structure (cerebral aneurysms) which commonly associated with CoAo. Changes induced by a gridlock mutationin the hey2 gene in the zebra fish lead to changes mimicking CoAo in this species.[6] Interestingly inducing up-regulation of vascular endothelial growth factor (VEGF) early in development is sufficient to suppress the gridlock phenotype and aortic abnormality in this model. VEGF plays a vital role in aortic development, acting as a chemo-attractant, stimulating angioblast migration toward the midline before formation of the aorta[9]. Indeed, targeted disruption of VEGF in mice leads to significant disruption of the developing aorta[10]. VEGF is also involved in stimulating generalized arterial differentiation through its effect on angioblast migration.



1. Associate Professor, Pediatrics, Ad-din Women's Medical College Hospital. Dhaka.
2. Assistant Professor, Pediatrics, Ad-din Women's Medical College Hospital. Dhaka.
3. Associate Professor, Pediatrics, Ad-din Women's Medical College Hospital. Dhaka.
4. Assistant Registrar. NICVD. Dhaka.
5. Professor and Head of the department, Pediatrics, Ad-din Medical College Hospital. Dhaka.

Correspondence : Mahmuda Hassan
e-mail : mahmudahasn@yahoo.com

Case Presentation and management

Presentation in infancy may vary from irritability, pale skin, sweating, respiratory distress, poor feeding, poor weight gain, cold feet and/or legs, diminished or absent pulses in the feet, blood pressure in the arms significantly greater than the blood pressure in the legs.

Present case was a 7 month baby boy admitted at Ad-din Medical College and Hospital with

Complaints of

1. difficulty in breathing
2. lethargy
3. vomiting,
4. cyanosis with history of poor feeding.

Past history: The child was delivered at term following an unremarkable pregnancy, uneventful neonatal period. At the age of 3 months of his age he was admitted in other hospital for cough and cold and respiratory distress. He had past history of transient cyanotic attack 3 to 4 times in his life which was resolved automatically at home.

On examination

1. temperature was 101 degree Fahrenheit
2. respiratory rate was 67/min with chest in drawing
3. heart rate was 110/ min and had a soft systolic murmur in the left sternal border
4. also had few rhonchi and crepitation in the lung fields.
5. His admission weight was 5kg. Weight for age was below the 3rd centile line, length was 61 cm. which is also below the 3rd centile line.
6. Blood pressure in upper limb was 110/80mm of Hg, in lower limb was 60/35 mm of Hg. with weak femoral pulse.

Provisionally diagnosed as a case of severe pneumonia with congenital heart disease.

Investigations

a) In complete blood picture, hemoglobin was 11.6 gm/dl, total white blood cell was 21000/ cu.mm with increased neutrophil count.

b) X ray chest shadowed cardiomegaly and multiple patchy opacity over both the lung fields.

c) Color Doppler ECHO done on 4th hospital day which showed left sided arch with coarctation of the aorta, intact intra atrial and intra-ventricular septum with normal coronary arteries. Left Ventricular Ejection Fraction (LVEF) 77%, Fractional Shortening (FS) 43%. Severe coarctation of the aorta noticed. Peak Pressure Gradient (PPG) 55 mm of Hg. Diameter of the aorta at the level of CoAo a was 3.5mm, mild TR noticed, PPR 44 mmHg, calculated Pulmonary Artery Systolic Pressure (PASP) 55 mm of Hg, severe left ventricular hypertrophy with hypertrophied IVS, moderate RVH ,

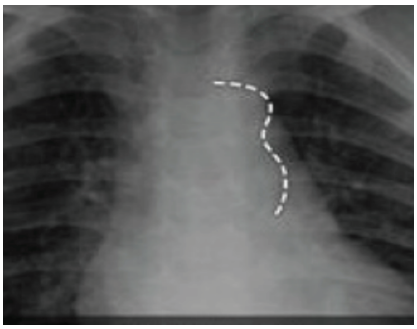
d) No patent ductus arteriosus (PDA) and evidence of severe pulmonary hypertension.

e) On admission SpO2 was 88% and maintained SpO2 > 95% with FiO2 1L/minute. Antibiotic was started as per protocol. Sildenafil has both pulmonary and systemic vascular relaxation effects was prescribed.

Paediatric cardiologist advised for urgent Balloon angioplasty for CoAo. His fever subsided and respiratory distress was resolved after treatment. Unfortunately the baby died at home while waiting for his surgical treatment 1 month after discharged from the hospital.

Discussion

CoAo is a congenital malformation that usually presents early in life and is often associated with congenital abnormal aortic valve. The mean survival for untreated patients is 35 years with a 25% survival rate beyond 50 years. The natural history of unrepaired coarctation of the aorta includes the development of systemic hypertension and subsequent morbidity and death from cardiovascular disease[11]. The age of diagnosis and correction is the most important factor for relief of hypertension and long-term survival[12]. Despite the fact that the coarctation of aorta appears more often in young males may present with hypertension, and/or murmurs resulting from collaterals or associated heart defects[11]. The age at which people with coarctation of the aorta are diagnosed depends on the severity of the condition. If the aortic coarctation is severe, it is usually diagnosed during infancy. Diagnosis is usually based on clinical suspicion and physical findings. It includes blood pressure difference between the upper and lower extremities, pulse delay and systolic murmur over the thoracic spines due to collateral circulation. Other manifestations can include systolic ejection sound and/or murmur and neurological complaints, as well as persistent pulmonary hypertension.



Plain X ray chest shows figure of 3 sign contour abnormality of the aorta, inferior rib notching also called

Roesler sign is due to secondary to dilated inter-costal collateral vessels which form as a way to bypass the coarctation and supply the descending aorta, the dilated and tortuous vessels erode the inferior margins of the ribs, resulting in notching but our case did not show any such signs on x ray because it is seen only in long standing cases, and therefore not seen in infancy (unusual in patients <5 years of age)[13] seen in 70% of cases presenting at older children or adults.

A reported 7 month female infant was presented with aortic stenosis, preductal coarctation, and pulmonary hypertension underwent operation. Intra-operative lung biopsy revealed marked medial hypertrophy of the pulmonary arterioles. This histopathology is compatible with persistent pulmonary hypertension in the newborn. She was alive and about 5 years after the operation pulmonary hypertension remained.[14]

Preoperative treatment consists of treatment of hypertension can be effectively treated using beta-blockers.15 The goal should be to reduce upper extremity hypertension, but remember that vigorous attempts to achieve normal upper extremity blood pressure (BP) may result in inadequate lower-body perfusion. Beta-blocker therapy prior to surgery may reduce the severity of postoperative hypertension, although most patients with preoperative hypertension require at least transient postoperative therapy. Relieving the aortic obstruction promptly rather that attempting to treat hypertension with antihypertensive medications is better. Sildenafil is a PDE5 inhibitor, its mechanism of action is to augment nitric oxide-cGMP signaling by inhibiting the degradation of cGMP. Increased cGMP results in pulmonary vascular relaxation.[16]

Intravenous continuous PGI2 is also effective in treating older children with primary pulmonary hypertension. PGI2works by increasing cAMP, and iNO works by increasing cGMP. Prostacyclins also causes vasodilation in the pulmonary vasculature, leading to decreased pulmonary pressures and improved cardiac output.[17]

Proper therapy for hypertension, endocarditis prophylaxis and corrective treatment for coarctation lesions with a high gradient. Indications for intervention in children include heart failure, a peak instantaneous pressure gradient across the coarctation>20mmHg, and/or radiologic detection of collateral circulation. Systemic hypertension, accelerated coronary heart disease, stroke, aortic dissection, and heart failure are common complications in adults who have not undergone correction for their coarctation or were operated later in life. Coarctation repair after early child-hood does not prevent persistence or late recurrence of systemic hypertension. As a result, correction of coarctation should be performed in infancy or early childhood to prevent the development of chronic systemichypertension13. In this case report, our patient was asymptomatic and his problem was diagnosed after hospital admission due to other clinical illness.

Death in these patients is usually due to heart failure, coronary artery disease, aortic rupture/dissection, concomitant aortic valve disease, infective endarteritis, or cerebral hemorrhage.

Conclusion

Early diagnosis and timely intervention is required for better the prognosis. If CoAo is severe and diagnosed is delayed in childhood or adulthood prognosis is guarded. To avoid late diagnosis and prevent complications, through physical examination including routine measurement of blood pressure and echocardiogram of suspected cases are advised.

References

1. Anderson R, Baker EJ, Macartney FJ, Rigby ML, Shinebourne EA, Tynan M eds. Paediatric cardiology. 2nd Ed. Churchill Livingstone, London 2002.
2. Campbell M, Polani PE. Aetiology of coarctation of aorta. Lancet,1961; 1: 463–467.
3. Fyler DC BD, Hellenbrand WC, Cohn HE. Report on the New England Regional Infant Cardiac Program. Pediatrics, 1980; 65:375–461.
4. Sehested J. Coarctation of the aorta in monozygotic twins. Br Heart J, 1982; 47: 619–620.
5. 5. Wessels MW, Berger RM, Frohn-Mulder IM et al. Autosomal dominant inheritance of left ventricular outflow tract obstruction. Part A. Am J Med Genet, 2005; 134A: 171–179.
6. 6. Peterson RT, Shaw SY, Peterson TA et al. Chemical suppression of a genetic mutation in a zebra fish model of aortic coarctation. Nature Biotechnol, 2004; 22: 595–599.
7. Rudolph AM, Spitznas U, Heymann MA. Hemodynamic considerations in development of narrowing of aorta. Am J Cardiol, 1972; 30: 514–525.
8. Russell GA, Berry PJ, Watterson K, Dhasmana JP, Wisheart JD. Patterns of ductal tissue in coarctation of the aorta in the first three months of life. J Thoracic CardiovascSurg, 1991; 102: 596–601.
9. Cleaver O, Krieg PA. VEGF mediates angioblast migration during development of the dorsal aorta in Xenopus. Development, 1998; 125: 3905–3914.
10. Carmeliet P, Ferreira V, Breier G et al. Abnormal blood vessel development and lethality in embryos lacking a single VEGF allele. Nature, 1996; 380: 435–439.
11. M. Campbell, "Natural history of coarctation of the aorta," British Heart Journal .1970;32(5): 633-640.
12. M. H. J. Brouwer, M. E. Erasmus, T. Ebels, and A. Eijgelaar,

"Influence of age on survival, late hypertension, and re-coarctation in elective aortic coarctation repair: including long-term results after elective aortic coarctation repair with a follow-up from 25 to 44 years," Journal of Thoracic and CardiovascularSurgery, 1994; 108(3): 525–531.

13. Higgins CB. Radiography of congenital heart disease. In: Webb WR, Higgins CB (eds). Thoracic imaging: pulmonary and cardiovascular radiology (pp742-767). Lippincott Williams and Wilkins. (2011).
14. Aota M1, Nomoto S, Yamaki S, Ban T Pulmonary hypertension caused by medial hypertrophy associated with aortic stenosis and preductalcoarctation.Ann Thorac Surg. 1997 ;64(1):244-7.
15. Lopez-Sendon J, Swedberg K, McMurray J, et al. Expert consensus document on beta-adrenergic receptor blockers. Eur Heart J. 2004 Aug. 25(15):1341-62.
16. Bharani A, Mathew V, Sahu A, Lunia B. The efficacy and tolerability of sildenafil in patients with moderate-to-severe pulmonary hypertension. Indian Heart J 2003;55(1):55–59.
17. Kelly LK, Porta NFM, Goodman DM, Carroll CL, Steinhorn RH. Inhaled prostacyclin for term infants with persistent pulmonary hypertension refractory to inhaled nitric oxide. J Pediatr. 2002;141:830–832.