VOLUME - 11, ISSUE - 05, MAY - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Cardiology

INTERRUPTED AORTIC ARCH IN A ONE DAY OLD NEWBORN: A CASE REPORT

Dr. Puli Samyu	Junior resident, department of paediatrics, Prathima institute of medical sciences, Nagnur road, Karimnagar.
Dr. N Sai Jayachandra Reddy	Junior resident, department of paediatrics, Prathima institute of medical sciences, Nagnur road, Karimnagar.
Dr. SK.Khasim	Cardiology Junior resident, department of General medicine, Prathima institute of medical sciences, Nagnur road, Karimnagar.
Dr. Kasturi Ravinder Reddy	Consultant cardiologist, Professor and head of department of medicine, Prathima institute of medical sciences, Nagnur road, Karimnagar.

ABSTRACT Interrupted aortic arch (IAA) is a rare congenital heart disease (CHD) which affects approximately 1.5% of CHD patients. It is a serious congenital vascular malformation and accounts severe mortality when associated with other intracardiac defects. This report describes the case of one day old infant with atresia of aortic arch presented with rapid breathing, poor feeding, and lethargy. The diagnosis of IAA and Ventricular septal defect (VSD) was made by two-dimensional echocardiography and confirmed intraoperatively which successfully corrected by immediate surgical intervention.

KEYWORDS :

INTRODUCTION:

CHD is an abnormal formation of the heart or blood vessels next to the heart and has incidence of 8 cases of every 1000 live births worldwide. [1] In the united states, congenital heart disease affects 1% of births (40,000) per year, of which 25% have critical congenital heart disease.[2],[3] Interrupted aortic arch is a rare CHD which affects approximately 1.5% of CHD patients.[4] It is defined as complete absence of luminal continuity between ascending and descending portions of aorta. IAA has higher mortality rates when associated with other intracardiac defects.

CASE REPORT:

A full-term male, newborn first baby for a non-consanguineous parent, weighting 3.4 kg, was presented with severe tachypnea, poor suckling, irritability, poor feeding, and lethargy on the day of delivery. In the antenatal period TIFFA showed echogenic focus in the heart, for confirmation fetal echocardiography was done which showed the presence of interrupted aortic arch, patent ductus arteriosus, ventricular septal defect. The remaining antenatal period was uneventful with no maternal illness or exposures. Elective LSCS was done in view of fetal echocardiography suggesting congenital heart defect and was uneventful.

Baby presented with severe tachypnea, poor suckling, irritability, poor feeding, and lethargy on the day of delivery. On examination, the patient showed no dysmorphic features, and he was in poor general condition, feeble crying, with marked tachypnea and tachycardia. His skin was grayish appearance. On examination of pulses, there were no peripheral pulsations in lower half of his body. The cardiac auscultation disclosed a gallop rhythm and grade two systolic murmur on base of the heart, with accentuated second heart sound. He had delayed capillary refilling time of around 8 seconds. On vitals examination; arterial blood pressure was 70/30 mmHg, mean pressure 42 mmHg as measured from left arm, heart rate of 159 beats per minute. The arterial blood pressure was undetectable in his right arm.

Laboratory investigations including arterial blood gas analysis revealed lactic acidosis with high anion gap. Serum electrolytes showed hyperkalemia with normal sodium and chloride. Random capillary blood glucose level was 89 milligrams/decilitre. The electrocardiogram showed sinus rhythm with signs of left auricular hypertrophy, right ventricular hypertrophy. The chest radiography (Fig.1) showed mild cardiomegaly, increased pulmonary markings and an absent aortic knob.

Echocardiography shows situs solitus, levocardia, normal systemic and pulmonary venous drainage, dilated right heart with normal systolic function. There was moderate degree of tricuspid

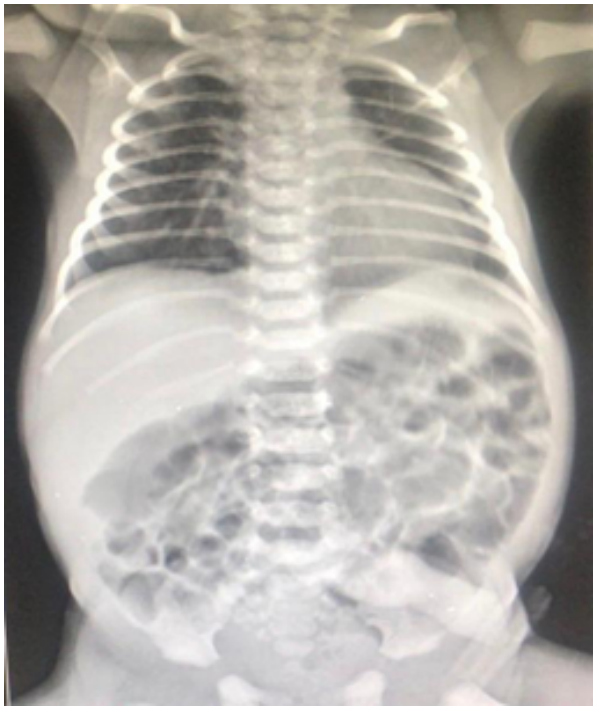

Fig.1: Chest radiography of same baby showing mild cardiomegaly, increased pulmonary markings and an absent aortic knob.

regurgitation, and the estimated pulmonary artery pressure was found to be 66 mmHg after calculation of the right atrial

pressure from the inferior vena cava size and variation with respiration. The echocardiogram showing interrupted aortic arch, large PDA, larger sub aortic VSD (bidirectional shunt) and abdominal aortic flow from the subcostal views was non-pulsatile flow (monophasic flow) with high flow across a large PDA.

Treatment was started immediately with prostaglandin infusion (alprostadil) at the rate of 0.05-0.4 microgram/kilogram/minute to maintain the patency of the ductus arteriosus until taken up for surgery. After stabilization, on day 3 surgical intervention was done.

Thoracotomy confirmed the interruption of aortic arch; it was felt that the distance separating the proximal and distal aortic segments was sufficiently great that direct anastomosis would not be possible without undue tension. Therefore, a partial occluding clamp was placed on the ascending aorta, and a 5mm expanded polytetrafluoroethylene (PTFE) graft was sewn end to side to the aorta. When the clamps were removed, good pulsatile flow in the descending aorta was observed. The ductus arteriosus was ligated, and VSD closure was done with polytetrafluoroethylene synthetic patch. The operative period was uneventful. Following surgical reconstruction, echocardiographic evaluation was done, which showed adequacy of repair.

DISCUSSION:

Aortic interruption is a relatively uncommon congenital heart disease accounting for 1.5% of all congenital heart disease, with type B being the most common form accounting for approximately 84 percent of the cases. Interrupted aortic arch is an anomaly that can be considered the most severe form of aortic coarctation. More than 97% of the cases have associated cardiac anomalies and complicating their treatment. The median age at death in untreated cases is 10 days and this condition usually occurs as a complicated neonatal surgical emergency.

IAA is a ductus dependent lesion since this is the only way the blood flow can travel to places distal to the disruption. Nearly all patients with IAA present to hospital within first two weeks of life when ductus arteriosus closes. Most patients present in first day of life. According to the Celoria and Patton classification, interrupted aortic arch can be grouped into three types, depending on the site of the disruption. [5],[6],[7]

Type A: The disruption is located distal to the left subclavian artery; this is the 2nd most common disruption represents approximately 13 percent of the cases.

Type B: The disruption is located between the left carotid artery and the left subclavian artery; this is the most common anomaly, representing approximately 84 percent of the cases.

Type C: The disruption is located between the innominate artery and the left carotid artery; this is a rare type represents approximately 3 percent of all cases.

Our case has fetal echocardiography which shows interrupted aortic arch in antenatal period itself, so surgical correction was done successfully immediately after birth. In our case we corrected interrupted aortic arch and VSD in single-stage procedure. There are different surgical techniques that are used for treatment of interrupted aortic arch, and some patients require multi-staged operations through their lifetime. [8],[9],[10]

CONCLUSION:

Interrupted aortic arch is a complex and highly lethal lesion, and an aggressive approach to early operation is warranted. Prostaglandin E1 is necessary to start early to avoid sudden cardiac collapse and death.[1] The prognosis of the infant with interrupted aortic arch is dependent on birth weight, associated heart defects and time of surgery. Restoration of aortic continuity and VSD closure with expanded PTFE grafts is a good single-stage procedure for the management of interrupted aortic arch with associated VSD.

REFERENCES:

1. Hu XJ, Ma XJ, Zhao QM, Yan WL, Ge XL, Jia B, Liu F, Wu L, Ye M, Liang XC, Zhang J, Gao Y, Zhai XW, Huang GY. Pulse Oximetry and Auscultation for Congenital Heart Disease Detection. Pediatrics. 2017 Oct;140(4) https://pubmed.ncbi.nlm.nih.gov/28939700/
2. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002 Jun 19; 39(12):1890-900. https://pubmed.ncbi.nlm.nih.gov/12084585/
3. Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005. J Pediatr. 2008 Dec; 153(6):807-13. https://pubmed.ncbi.nlm.nih.gov/18657826/
4. Varghese R, Saheed SB, Omoregbee B, Ninan B, Pavithran S, Kothandam S. Surgical Repair of Interrupted Aortic Arch and Interrupted Pulmonary Artery. Ann Thorac Surg. 2015 Dec; 100(6):e139-40. https://pubmed.ncbi.nlm.nih.gov/26652572/
5. Schreiber C, Mazzitelli D, Haehnel JC, Lorenz HP, Meisner H. The interrupted aortic arch: an overview after 20 years of surgical treatment. Eur J Cardiothorac Surg. 1997 Sep;12(3):466-9; discussion 469-70. https://pubmed.ncbi.nlm.nih.gov/9332928/
6. Sato S, Akagi N, Uka M, Kato K, Okumura Y, Kanazawa S. Interruption of the aortic arch: diagnosis with multidetector computed tomography. Jpn J Radiol. 2011 Jan;29(1):46-50. https://pubmed.ncbi.nlm.nih.gov/21264661/
7. Sato S, Akagi N, Uka M, Kato K, Okumura Y, Kanazawa S. Interruption of the aortic arch: diagnosis with multidetector computed tomography. Jpn J Radiol. 2011 Jan;29(1):46-50. https://pubmed.ncbi.nlm.nih.gov/21264661/
8. Luciani G.B., Ackerman R.J., Chang A.C., Wells W.J., Starnes V.A: One-stage repair of interrupted aortic arch, ventricular septal defect, and subaortic obstruction in the neonate: a novel approach, J Thorac Cardiovasc Surg , 1996, vol. 111 (pg. 48-358)
9. Tláskal T., Hucin B., Kucera V., Vojtovic P., Gebauer R., Chaloupecky V., Skovranek J.: Repair of persistent truncus arteriosus with interrupted aortic arch, Eur J Cardiothorac Surg , 2005, vol. 28 November (5)(pg. 736-741)
10. Pankaj Kumar Mishra, Management strategies for interrupted aortic arch with associated anomalies, Eu J Cardio-Thoracic Surgery, Volume 35, Issue 4, April 2009, Pages 569–576, https://doi.org/10.1016/j.ejcts.2008.12.044