Interrupted aortic arch detected 4 days after surgical ventricular septal defect and patent ductus arteriosus closure: a case report

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ABSTRACT

Background: Interrupted aortic arch is a loss of luminal continuity between ascending and descending aorta. In a patient with an interrupted aortic arch, systemic perfusion is supplied by ductus arteriosus. Therefore, ductal closure will lead to systemic hypoperfusion. We present a case of undiagnosed type A interrupted aortic arch on initial examination.

Case Presentation: A 4-month-old patient had signs of systemic hypoperfusion after a successful surgical ventricular septal defect and patent ductus arteriosus closure. Blood pressure between the upper and lower extremities showed a wide discrepancy. Echocardiography showed no flow from the aortic arch to the descending aorta. Emergency re-operation was performed, and type A interrupted aortic arch was found with descending aorta connected to the pulmonary artery. The interrupted aortic arch repair was performed using end to side technique. The patient was discharged on a post-operative day 26 without any complication, and no abnormalities were found on a 3-month follow-up.

Conclusion: Careful physical examination, including BP measurement of all extremities, is warranted. To avoid misdiagnosis, preoperative echocardiography must be performed thoroughly to assess cardiac and great vessel abnormalities, including color flow Doppler.

Keywords: Interrupted Aortic Arch, Echocardiography, Systemic Hypoperfusion.

INTRODUCTION

Interrupted aortic arch (IAA) is a cardiovascular malformation accounting for ~1% of infants with congenital heart disease, with an incidence of 3 in 1 million live birth.¹ The anatomical abnormality in IAA is loss of luminal continuity between ascending and descending aorta.¹,² Interrupted aortic arch (IAA) is rarely isolated.³ Other associated cardiovascular anomalies most commonly seen in IAA patients are ventricular septal defect (VSD) (90%) and patent ductus arteriosus (PDA) (97%), which provide blood flow to the descending aorta.⁴,⁵ Symptoms usually occur during the early neonatal period, and clinical deterioration is often rapid after ductal closure, which leads to systemic hypoperfusion.⁶ Diagnosis of IAA can be provided by cross-sectional echocardiography combined with color flow Doppler.⁷ Even though rarely reported, misdiagnosis of IAA could occur even in adults.⁸,⁹ In this case report, we present an interesting case of undiagnosed type A IAA in early diagnostic examination and known 4 days after surgical VSD closure and PDA closure.

CASE REPORT

A 4-month-old boy presented with shortness of breath. The complaint worsened 3 months before admission. Signs of failure to thrive also exist. The chest radiograph showed cardiomegaly and increased pulmonary vascular markings on diagnostic examination. Echocardiography showed subaortic VSD 6-7 mm in diameter, dilatation of LA and LV, suspected AS valvar-subvalvular, and no PDA. The aortic arch appeared normal 6-7 mm in diameter, good functioning aortic valve. VSD closure was performed using IMPRA patch 0.4 mm. TEE showed no residual VSD and no residual PDA post-operatively.

On post-operative day 1, the patient was anuric despite adequate blood pressure with arterial blood pressure (ABP) 89/42 (56) mmHg and a heart rate of 137 bpm. Blood gas analysis showed mild metabolic acidosis. Echocardiography showed good
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There are three types of IAA based on the position of interruption. Type A is interruption distal to the left subclavian artery; type B is between the left common carotid artery and left subclavian artery; type C is between the brachiocephalic artery and the left common carotid artery. Isolated IAA is very uncommon and

LV and RV contractility, no residual VSD and PDA, with mean pulmonary artery pressure of 42 mmHg. Blood creatinine was doubled, raised from 0.37 to 0.75 mg/dL. Therefore, the patient was diagnosed with low cardiac output syndrome and pre-renal acute kidney injury, complicated with pulmonary hypertension and treated accordingly. The patient was given a furosemide drip, and urine output started to become adequate. During the following 3 days, we observed that patient would have anuria or oliguria when the blood pressure was within normal range, and it would be resolved when the blood pressure achieved more than 100 mmHg.

On post-operative day 4, the possibility of coarctation of the aorta was evaluated. Non-invasive blood pressure of all extremities was measured. A wide discrepancy of blood pressure (>30 mmHg) was found during measurement, as follows: ABP of the left radial artery was 122/52 (79) mmHg, NBP of the right arm was 126/50 mmHg, NBP of the right foot was 33/23 mmHg, left foot was 42/24 mmHg. Echocardiography revealed no flow from the aortic arch to the descending aorta. It was suspected of IAA or iatrogenic stenosis of the aorta during ostium PDA closure. Emergency re-operation was performed. During exploration, IAA type A was found with descending aorta connected to the pulmonary artery. IAA repair was performed using end to side technique.

The patient was hemodynamically stable after IAA repair during monitoring in the ICU. Urine output was adequate, creatine level was improved, and metabolic acidosis was resolved. The patient was intubated for 9 days post-re-operation and on NIV for the following 5 days. Difficulty in weaning from the ventilator was caused by pulmonary hypertension and was complicated by recurrent lung atelectasis and pneumonia. The patient also had paralytic ileus, which contributed to a prolonged hospital stay. The patient was discharged on the 26th day after surgery without significant complications. On a 3-month follow-up, the patient had gained 2 kg in body weight, no breathing or feeding difficulty was reported and observed, and no issue with urine output. Echocardiography showed good LV and RV contractility, no residual VSD and PDA and a wide aortic arch with a gradient of 6 mmHg.

DISCUSSION

The interrupted aortic arch is defined as lacking luminal continuity between the ascending and descending aorta. It is a rare congenital heart disease with an incidence of 3 in 1 million live birth. IAA can usually be detected in intrauterine using fetal echocardiography. By 1 month, 76% of cases died due to heart failure if left untreated. Less than 10% survived until 1 year of age without operation. The classification system of IAA introduced by Celoria and Patton is still universally used. There are three types of IAA based on the position of interruption. Type A is interruption distal to the left subclavian artery; type B is between the left common carotid artery and left subclavian artery; type C is between the brachiocephalic artery and the left common carotid artery. Isolated IAA is very uncommon and

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incompatible with life. Association of the interrupted aortic arch, ventricular septal defect and patent ductus arteriosus are constant. It has been considered the triad of congenital vascular defects by Everts-Suarez and Carson. Other cardiac anomalies usually present with IAA are truncus arteriosus, transposition of great arteries, bicuspid aortic valve, aberrant innominate arteries and aortopulmonary window.

Clinical presentations of IAA are respiratory distress, variable degrees of cyanosis, poor peripheral pulses and signs of congestive heart failure during the first days of life. Differential cyanosis is uncommon because of the frequent association of VSD. In the most common form of IAA with associated PDA and VSD, clinical deterioration is abruptly seen after ductal closure. The patient will become acidic and anuric as systemic blood flow to the inferior body part is disrupted. Signs of systemic hypoperfusion can be observed in the elevation of serum creatinine, hepatic enzymes and lactic acid dehydrogenase and evidence of necrotizing enterocolitis.

Cross sectional echocardiography combined with color flow Doppler can provide an accurate preoperative diagnosis of IAA in infants and children with congenital heart disease. Assessment of the aorta must be done routinely and thoroughly by evaluating the branching pattern of the great artery and its flow from ascending to descending aorta to avoid misdiagnosis of aortic abnormalities. If IAA is suspected, the length of discontinuity, the narrowest dimension of the left ventricular outflow tract, the diameter of the aortic annulus, and the size of the ascending and descending aorta should be measured. Careful echocardiography examination often eliminates the need for preoperative catheterization or angiography, even in a patient with complex cardiac anomalies. In some cases where the further examination is needed to evaluate the aorta and great vessels, MRI and CT angiography are useful to clarify anatomy before surgery. These diagnostic tools can be used to evaluate cardiac and congenital aortic abnormalities and the origins of great and thoracic vessels.

In this case report, preoperative diagnostic examination failed to detect IAA type A. Surgical closure of VSD and PDA caused no blood flow to the lower part of the body, which subsequently caused systemic hypoperfusion. The patient survived this state for 4 days.

The presence of interrupted aortic arch or coarctation of the aorta should be suspected in a patient with persistent acidosis and signs of low perfusion of a lower body part (anuria or oliguria, paralytic ileus) despite successful cardiac operation. The discrepancy between the upper and lower extremities’ blood pressure arise the suspicion of aortic arch disruption. Color flow Doppler echocardiography is performed to establish the diagnosis. We suspect there is numerous collateral circulation between the two separate aortic systems, which become the key to survival in this patient.

CONCLUSION
Patient with VSD who presents with severe symptoms of heart failure since neonate should raise suspicion of associated congenital cardiovascular anomalies. Interrupted aortic arch is most commonly seen with VSD and PDA. Careful physical examination, including BP measurement of all extremities, is warranted. Preoperative echocardiography must be performed thoroughly to assess cardiac and great vessel abnormalities, including color flow Doppler, to avoid misdiagnosis. However, evaluation of the aorta and great vessels can occasionally be limited. MRI and CT angiography has recently been adjunct to echocardiography in evaluating this entity. Failure to detect this abnormality in time is the main cause of morbidity and mortality in infants with severe cases.

CONFLICT OF INTEREST
The authors declared explicitly that there are no conflicts of interest concerning this article’s authorship and/or publication.

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REFERENCES

