World Journal of Medical and Surgical Case Reports

Case Report

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Congenital Malformations of the Aortic Arch with Associated Intra-Abdominal Issues: When to Intervene?

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Abstract Congenital malformations of the aorta are rare congenital vascular defects. Severe Aortic Coartation (AoCo) and Aortic Arch Interruption (AAI) are aortic anomalies associated with left ventricular outflow tract obstruction and ductal-dependent distal systemic perfusion, requiring early surgical repair. Two neonates, one with severe AoCo and the other with AAI, presented with signs of heart failure and ductal-dependent distal systemic perfusion, and prompt surgery was required. However, other intra-abdominal issues required urgent abdominal surgery. Aortic surgery had to be deferred until the patients were recovered and optimized after the abdominal surgery. Both of the patients had successfully undergone surgical aortic surgery with good results. Timing of surgery for congenital aortic arch anomalies should be weighed according to the patient conditions to ensure a good clinical outcome and to minimize the post-operative morbidity and mortality.

Keywords Aortic Arch Interruption, Aortic Coarctation, Congenital Heart Defects, associated intra-abdominal disease, timing of surgery.

Introduction

Aortic Coarctation (AoCo) and Aortic Arch Interruption (AAI) are two different congenital heart defects involving the aortic arch and/or aortic isthmus, either isolated or associated with intra-cardiac defects [1]. Severe AoCo and AAI, generally presented during the neonatal period, require early surgical treatment. However, the ideal timing for surgery might be precluded by the presence of associated congenital non-cardiac malformations, or by the occurrence of diseases acquired in the presence of severe aortic obstruction, like necrotizing enterocolitis (NEC). In both these situations the decision making process has to consider all the concomitant variables in order to set up the appropriate time frame and sequence of abdominal and cardiac surgery approaches, for each individual patient.

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Submitted: Thursday, November 06, 2014; Accepted: Sunday, January 18, 2015; Published: Saturday, January 31, 2015

First Case presentation

A neonate was admitted immediately after birth in another hospital with the diagnosis of severe AoCo and large patent ductus arteriosus (PDA). Prostaglandins (PGE) infusion was started from the first day of life and indication was given for surgical repair of the congenital heart defect. However, he developed perforated NEC a day before the scheduled transfer to our

hospital for cardiac surgery. An emergency laparotomy with right hemicolectomy and double barrel stoma was performed. This post-operative course was complicated by multiple recurrent nosocomial infections with sepsis, and repeated episodes of lung collapse. After more than one month the clinical conditions of the patient slowly improved, even if he was still on PGE infusion with the evidence of cortical hyperostosis secondary to prolonged PGE administration. Congenital hypothyroidism was also detected. Once he was eventually cleared from active infection, he was transferred to our hospital for aortic coarctectomy and PDA closure. The first pre-operative echocardiography showed a severe AoCo with large PDA. However, a subsequent echocardiography raised the suspicion of the presence of AAI, and the complete echocardiography findings revealed the presence of aortic valve stenosis, small ventricular septal defect, and patent foramen ovale with severe left ventricular hypertrophy.

Surgery was performed through a left postero-lateral thoracotomy in the 4th inter-costal space. The dissection free of the aortic arch with the neck vessels and of the PDA with the descending thoracic aorta revealed the presence of an AAI type B, with interrupted transverse arch between the origin of the left carotid artery and left subclavian artery. The distance between the distal segment of the transverse aortic arch and the descending thoracic aorta after the PDA was quite remarkable. It was decided to perform the aortic arch reconstruction and PDA closure through the left thoracotomy approach, since the appropriate surgical treatment through median sternotomy with deep hypothermia and circulatory arrest would have required a set up at the moment not yet available in our hospital, and there was not another hospital available to accept this patient for conventional surgery. After very extensive mobilization of the aortic arch and neck vessels and of the descending thoracic aorta until the level of the diaphragm, PDA division and aortic arch reconstruction with end-to-end anastomosis of the distal aortic stump to the longitudinally incised inferior aspect of the aortic arch was performed. Immediately after aortic declamping there was good pulsatility of the distal descending thoracic aorta, with systolic pressure gradient between the invasive measurement in the right brachial artery and the non-invasive measurement on the left femoral artery of 15mmHg. The post-operative echocardiography confirmed the good reconstruction of the aortic arch, with mild residual obstruction (peak pressure gradient = 20 mmHg).

The immediate post-operative course was complicated by massive atelectasis of the left lung, requiring mechanical ventilation until the 3rd post-operative day. Tracheal extubation was then followed by later consolidation of the right lung, requiring prolonged antibiotics treatment. The general conditions slowly improved and the patient was discharged on the 32nd post-operative day, with the program to monitor the potential growth of the reconstructed aortic arch. Five months after surgery the patient remains asymptomatic, without any increase of the non-invasive pressure gradient through the aortic arch on echocardiography.

Second Case presentation

A 2.2kg girl was born premature after 34-week gestation to a 40 year-old woman with diabetes mellitus on insulin therapy, was admitted immediately after birth to the neonatal intensive care unit (NICU) because of duodenal atresia. She was incidentally found to have a large PDA evidenced by clinical findings of a continuous heart murmur best heard over the left infra-clavicular region and a wide pulse pressure with bounding peripheral pulses. Her apex beat was felt at the 5th inter-costal space

over the mid-clavicular line. The lungs auscultation was clear, but the liver was palpable 3cm below the sub-costal margin. Non-invasive blood pressure at right upper limb, left upper limb, right lower limb and left lower limb were respectively 90/52, 85/46, 81/55, 79/44 mmHg. Echocardiography confirmed the presence of a large PDA but showed also the presence of "Shone complex": mitral stenosis, aortic stenosis with bicuspid valve, aortic coarctation with long segment tubular hypoplasia. Anti-failure treatment with frusemide and spironolactone was started.

On the 4th day of life she underwent laparotomy and duodeno-jenunostomy. Intra-operative findings were a duodenal atresia with apple-peel small bowel (Christmas tree syndrome) and situs abdominal inversus. Post-operatively the patient had prolonged feeding intolerance due to her underlying intra-abdominal anomalies, with slow improvement after two weeks.

The medical treatment failed to control the heart failure, and the child remained oxygen dependent and continued to lose weight. Decision was made to proceed with cardiac surgery, performed when the patient was 34 days old and 2.0kg. Through a left postero-lateral thoracotomy in the 4th intercostal space, the PDA was closed with double transfixed 6-0 mono-filament suture. Aortic coarctectomy was performed by resection and end-to-end anastomosis with enlargement of the aortic arch. Good pulsatility of the distal descending thoracic aorta was palpable immediately after aortic declamping. The invasive blood pressure measured in the right brachial artery was 67/34 mean 42 mmHg while the non-invasive left femoral artery pressure was 63/33 mean 41 mmHg.

The immediate post-operative course was uneventful, with progressive weaning from inotropic support and mechanical ventilation until tracheal extubation on 3rd post-operative day. The post-operative echocardiography confirmed the disappearance of the PDA and the absence of any significant obstruction at the level of the aortic isthmus. On the 5th post-operative day after episodes of bradycardia the electrocardiographic diagnosis of Wolf-Parkinson-White syndrome was made and β-blocking treatment was started, clinically well tolerated. Eight months after surgery she is doing well with good weight gain.

Discussion

The aortic arch has three segments: proximal, distal and isthmus. The proximal component extends from the takeoff of the innominate artery to the left common carotid artery. The distal part extends from the left common carotid artery to the left subclavian artery. The isthmus is the segment of aorta connecting the distal aortic arch with the juxtaductal region of the descending thoracic aorta [1]. This complex composite of segments presents the possibility of developmental anomalies in the form of interruption, hypoplasia or coarctation [1, 2].

AAI is a rare congenital anomaly, with an incidence of 0.7% among congenital heart defects. It is defined as a lack of luminal continuity between the ascending and descending thoracic aorta. This discontinuity may be complete or with a present of an atretic fibrous band. AAI is classified into three types according to the site of discontinuity of the aortic arch. Type A: the discontinuity is distal to the left subclavian artery; type B: between the left carotid and left subclavian arteries; type C: between the innominate and left carotid arteries [3]. More than 98% of AAI presents with associated intra-cardiac defects [1].

AoCo is a congenital narrowing of the aortic lumen, most commonly located between the origin of the left subclavian artery and the

aortic isthmus. It is more frequent than the AAI and accounts for approximately 4.6% of all congenital heart defects [1]. Also the AoCo frequently presents with associated intra-cardiac defects.

Although AAI and AoCo are different in term of anatomical and morphological definition, both cause left ventricular outflow tract obstruction, followed by subsequent left ventricle hypertrophy.

Neonates with severe AoCo and AAI have ductal-dependent perfusion of the distal systemic circulation, with the blood flow to the distal part of the body maintained by pulmonary hypertension and patency of the ductus arteriosus. Upon closure of the ductus arteriosus, the neonates present sudden hemodynamic collapse with profound metabolic acidosis, due to the inadequate distal systemic perfusion. Hence, the initial management after the diagnosis of severe AoCo and AAI includes PGE infusion to maintain the ductal patency, followed by optimization of the clinical conditions: correction of the metabolic acidosis, stabilization of hemodynamic status with mechanical ventilation and inotropic support are the essential elements to provide the neonates with the best conditions before proceeding to surgery [1, 4, 5].

Because of the ductal-dependency of the distal systemic circulation, the surgical treatment of severe AoCo and AAI requires to be performed in the first few weeks of life. However, as in our reported experience, the neonates might present with other issues needing urgent abdominal surgery before the cardiac surgery procedure.

In both of our cases the aortic surgery had to be postponed for about a month, in order to optimize the clinical conditions after the abdominal surgeries. Perforated NEC carries a high mortality rate with reported survival of 35% pertaining to the development of Gram negative sepsis [6]. It needs a prompt surgical treatment to remove the source of infection. Unfortunately, this patient developed multiple complications after the abdominal surgery for perforated NEC which delayed the surgery for AAI.

The standard surgical treatment of AAI consists of complete repair performed through a median sternotomy, with cardio-pulmonary bypass on deep hypothermia and circulatory arrest or selective cerebral perfusion [1, 7]. However, this would have required a set up not yet available in our center. The diagnosis of AAI was mistaken as AoCo before the surgery. Ideally, an additional pre-operative imaging such as cardiac catheterization with angiography, magnetic resonance angiography or computed tomography angiography, should be considered especially if there is an equivocal finding from echocardiography [8-10]. An accurate pre-operative imaging should provide correct characterization of the aortic morphology, and therefore allow a proper surgical planning.

With regard to the abdominal issues of case repot number 2, duodenal atresia, apple peel deformity or type IIIB jejunal atresia is a rare association. Apple peel atresia is due to intrauterine vascular accident of superior mesenteric artery. Heterotaxy is a congenital transposition of internal organs. Situs inversus abdominis, also called abdominal heterotaxia, is a very rare condition with frequency of 1 in 4000 to 20000 live birth [11]. Congenital heart disease is present in 5-10% of patients with situs inversus abdominis. Duodenal atresia, apple peel deformity with situs inversus abdominis is definitely a very rare association.

Patient described in case report 2 had complex congenital anomalies involving intra-abdominal organs (duodenal atresia, apple-peel small bowel in the presence of

situs inversus abdominis). Patient with duodenal atresia need an early surgical intervention to restore the continuity of alimentary tract to allow optimization of nutrition via enteral feeding, to avoid bowel mucosa atrophy secondary to prolonged fasting and subsequently bacteria translocation. Moreover, duodenal atresia are associated with risk of aspiration pneumonia and electrolytes imbalance due to loss of gastric and bile secretion. Surgery for the repair of AoCo was deferred in order to allow the correction of duodenal atresia.

Conclusion

The timing of surgery for severe congenital aortic malformations in neonates with associated abdominal issues should be balanced between the urgency of the treatment of the abdominal problems and the risk of delaying the treatment of the aortic defect. Nevertheless, optimization of the patients before surgery is a crucial step before proceeding with any major aortic surgery, especially when dealing with neonates and young infants.

Conflicts of Interests:

The authors declare that there are no conflicts of interests.

Authors contributions:

CYW: Performed the literature search and prepared the draft manuscript

NR: Participated in design of study and edited the manuscript

SA: Participated in design of study and edited the manuscript

HASSAN: Participated in design of study and edited the manuscript

ARW: Participated in design of study and edited the manuscript

AFC: Conceived the study, participated in design and edited the final manuscript

All authors read and approved the final manuscript for submission

Ethical Consideration

The written informed consents were obtained from the parents/legal guardians of the patients for publication of this case report

Acknowledgement

We would like to thank Dr. A. M. Shamsuddin for the technical assistance in the preparation of the manuscript.

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