Aortic arch interruption in the neonate, with emphasis on early diagnosis and management

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Summary

Interruption of the aortic arch (IAA) is a rare but lethal congenital cardiovascular anomaly. Over a 3-month period, we encountered 5 neonates with IAA, all of whom presented in the 1st week of life with congestive cardiac failure and diminished or absent pulses. All 5 underwent surgery, and 3 died from causes unrelated to the surgical repair. The diagnosis of this condition is difficult to make, although the absence of pulses may provide an important clinical clue (particularly if the left brachial and femoral pulses are involved).

Using prostaglandins to re-establish ductal patency may be life-saving in these patients, since this restores blood flow to the lower trunk and kidneys. Administration of oral prostaglandin E₂ to infants in whom this anomaly is suspected before they are referred to a tertiary care centre is therefore vital.

Complete interruption of the aortic arch (IAA) is a rare anomaly, occurring in only 19 per million live births in the New England Regional Infant Cardiac Program. It is one of the most rapidly lethal forms of neonatal congenital heart disease. The classification of Celoria and Patton remains in use and refers to one of three possible sites of interruption: distal to the left subclavian artery (type A), between the left carotid and left subclavian artery (type B), and between the innominate and left carotid artery (type C).

We present data on 5 cases of IAA seen during a 3-month period, discuss the clinical diagnosis and management, and review the surgical options available.

Patients and investigations

From July to October 1987, 5 white neonates with IAA were treated at Johannesburg Hospital. Four of them (cases 2-5) were born in the Johannesburg area and the other in Durban. Age at onset of symptoms, sex, birth weight, type of interruption and associated defects, as well as time of operation, are summarised in Table I. All 5 patients presented with congestive heart failure with respiratory distress at 2-6 days of age.

Physical examination revealed cardiomegaly, right ventricular lift, a soft pulmonary ejection systolic murmur, a palpable pulmonary artery segment and marked liver enlargement (4-7 cm). Femoral pulses were absent in all, while left upper limb pulses were not detectable in patients 3-5 (type B). Chest radiography demonstrated cardiomegaly with increased pulmonary vascular markings. Electrocardiograms showed biventricular hypertrophy with right axis deviation in all but 1 (case 3), where the mean QRS axis was -20°.

Two-dimensional echocardiographic examination demonstrated the presence of a large perimembranous ventricular septal defect (VSD) in 4 patients and a large muscular VSD in 1 (case 2). In none of them could the anatomy of the aortic arch be demonstrated satisfactorily on two-dimensional echocardiography. A retrograde left radial angiogram in case 3 showed filling of the descending aorta and the pulmonary arteries through a patent ductus arteriosus, but no filling of the arch. This was interpreted as evidence of coarctation of the aorta.

Cases 4 and 5 were evaluated by cardiac catheterisation and biplane cine-angiography. The structure of the aortic arch was reliably determined by injection of contrast into the left ventricle.

Ductal patency was maintained before surgery by continuous intravenous infusion of prostaglandin E₂. All patients received positive-pressure ventilation through a nasotracheal tube and inotropic support with either dopamine infusion or oral digoxin, and their acidosis was corrected by intravenous administration of sodium bicarbonate and assisted ventilation.

Operative management

In cases 1 and 2 the type A interruption was repaired through a left posterolateral thoracotomy. In each case there was a large patent ductus arteriosus which was transected and oversewn. In case 1 a direct anastomosis was performed between the distal end of the aortic arch and the proximal descending aorta. In case 2 the subclavian artery was utilised for the repair. It was incised longitudinally to form a flap which was anastomosed to the posterior half of the distal aortic stump. The repair was completed anteriorly with a patch of bovine pericardium.

The initial approach in case 3 was through a left posterolateral thoracotomy, since this patient was thought to have a coarctation of the aorta. Once a type B interruption was identified, the descending aorta was mobilised and the repair performed through a median sternotomy. This patient and patients 4 and 5 underwent complete repair of all their malformations, via a median sternotomy, as a single operative procedure under cardiopulmonary bypass, deep systemic hypothermia (16-18°C) and circulatory arrest. The ductus arteriosus was transected and the remaining ductal tissue trimmed from the descending aorta. The pulmonary artery side was oversewn. In each case a direct anastomosis was performed between the descending and ascending aorta. The VSD was closed through the tricuspid valve using a Teflon patch. The period of circulatory arrest averaged 31 minutes (range 13-40 minutes).

Outcome

Of the 5 patients, 2 have survived (cases 3 and 5) and at the time of writing had been followed up for periods of 24 weeks and 10 weeks respectively.
Patient 1 was referred to this centre in the 3rd week of life, in established chronic renal failure requiring peritoneal dialysis. Renal biopsy revealed cortical necrosis with involvement of at least 50% of the nephrons. He required mechanical ventilation, because the volume of dialysate interfered with normal diaphragmatic function. An ultrasound scan of the head showed moderate periventricular haemorrhage. He died in chronic renal failure without having undergone closure of the VSD.

Patient 2 underwent successful repair of the aortic interruption but could not be weaned from the ventilator. A large muscular VSD had been demonstrated on two-dimensional echocardiography and this was thought to be contributing to the respiratory problem. Multiple muscular VSDs were found at surgery, performed on day 17 of life. Despite this she continued to be ventilator-dependent. Mildly dysmorphic facial features prompted chromosomal studies, which demonstrated a partial deletion of one of the X chromosomes. She went on to develop Serratia septicaemia before death. At autopsy cytomegalovirus was isolated from the lung.

Patient 4 underwent successful repair of the aortic interruption and VSD, but remained ventilator-dependent. He became grossly oedematous and this was thought to be caused by a combination of factors, viz. hypo-albuminaemia, sepsis and inappropriate secretion of antidiuretic hormone. Staphylococcus epidermidis was cultured from the blood and Candida albicans from the urine. He was treated with vancomycin, ceftriaxone and amphotericin B and received daily infusions of albumin because the volume of dialysate interfered with normal diaphragmatic function. An ultrasound scan of the head showed moderate periventricular haemorrhage. He died in chronic renal failure without having undergone closure of the VSD.

Patient 5 is well and thriving but has been left with a 50 mmHg systolic gradient between the upper and lower limbs. Her systemic hypertension is well controlled with propranolol and prazosin. Patient 5 is also thriving and has a 10 mmHg systolic gradient between the upper and lower limbs. Her early neonatal course was complicated by hypocalcaemia, which was controlled with calcium supplementation. At surgery no thymic tissue was found. Tests for lymphocyte subsets showed almost complete absence of T lymphocytes, which, together with the hypocalcaemia, suggested the diagnosis of DiGeorge syndrome. On day 29 of life she underwent thymic transplantation with VSD closure. However, the aortic arch continued to dilate, and by day 39 he was still dependent on mechanical ventilation. 

**Discussion**

IAA is a rare congenital malformation that usually occurs together with severe intracardiac anomalies. Celoria and Patton divided aortic interruption into three types, depending on the location of the interruption. Type A interruption was first reported by Steidel in 1778 and is the second most common type. The commonest form, type B, was first described by Seidel in 1818, and it was only in 1948 that type C, the rarest form, was first documented.

Embryologically, blood flow through the aortic arch is of critical importance to its enlargement or involution (interruption). The great majority of cases of IAA appear to be related to reduced antegrade blood flow in the ascending aorta as a result of the associated intracardiac shunts. Infrequently, the patient with type C interruption may have no intracardiac malformations. In such cases morphogenic factors other than reduced antegrade aortic blood flow may be operative.

Because of the rarity of this condition we were surprised by the unusually high number of 5 cases over a 3-month period. From the medical histories obtained, we were unable to identify a common aetiological factor. However, the incidence of coarctation of the aorta, which is embryologically related to IAA, appears to be higher in the Transvaal than in the rest of RSA.

The common presentation in our patients was rapid onset of congestive heart failure, with significant hepatomegaly and associated respiratory distress in the 1st week of life. The absence of pulses provides a very important clinical clue to the diagnosis. When both lower limb pulses weaken or disappear, it is extremely difficult to differentiate between coarctation of the aorta and type A aortic interruption. Statistically, coarctation of the aorta is by far the more common lesion. When there is a deficit in both lower limb pulses as well as the left upper limb pulse, the presence of type B interruption is probable. In such instances, cardiac catheterisation is essential to outline the structure of the aortic arch. In expert hands echocardiography may also provide the required anatomical information. If untreated, up to 80% of infants with IAA will not survive the 1st month of life.

The use of prostaglandins to achieve ductal patency has now enabled many such infants to survive and reach tertiary medical centres where surgical treatment may be undertaken. Any neonate suspected of having coarctation of the aorta or IAA should therefore be given oral prostaglandin E2 30 μg/kg hourly through a nasogastric tube before referral to a tertiary care centre. In addition, stabilisation of the patient with regard to acid-base status, renal function and control of congestive heart failure is essential before surgery.

Type B interruption is frequently associated with the DiGeorge syndrome, both of which are thought to result from neural crest cell abnormalities. Characteristically, the thymus and parathyroid glands are absent in this syndrome. It may pose a problem in the neonatal period and causes hypocalcaemia and/or infection because cellular immunity is defective. Thus this association should always be borne in mind when a diagnosis of type B interruption is made. Partial forms of the

<table>
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<th>Patient</th>
<th>Birth weight (g)</th>
<th>Sex</th>
<th>Age at onset of symptoms (d)</th>
<th>Type of interruption</th>
<th>Associated lesions</th>
<th>Age at surgery (d)</th>
<th>Outcome</th>
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<tr>
<td>1</td>
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<td>A</td>
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<tr>
<td>3</td>
<td>2400</td>
<td>F</td>
<td>6</td>
<td>B</td>
<td>VSD</td>
<td>9*</td>
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<tr>
<td>4</td>
<td>2900</td>
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<td>2</td>
<td>B</td>
<td>VSD</td>
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<tr>
<td>5</td>
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<td>B</td>
<td>VSD</td>
<td>9*</td>
<td>Alive</td>
</tr>
</tbody>
</table>

*Closure of VSD.

Complete repair.
DiGeorge syndrome have been reported, and we believe that our case 5 falls into this category because she was producing some endogenous parathyroid hormone.

Because of the invariable association of IAA with intracardiac lesions causing increased blood flow, selection of the ideal operative approach is difficult. The options are complete primary repair or palliative surgery. The latter involves repair of the IAA and banding of the pulmonary artery. Closure of the VSD would then be carried out at a later stage. In 1975 Trusler and Izukawa\(^1\) reported complete repair of a type B interruption and VSD in a 13-day-old patient. Since then several groups\(^2\)\(^-\)\(^6\) have presented series of patients treated in this manner. Furthermore, Hammon et al.\(^7\) and Schumacher et al.\(^8\) had good results with deep hypothermic circulatory arrest, and this has now become the approach of choice in many centres.

We consider primary total correction using deep hypothermia with or without circulatory arrest as the procedure of choice for cases of type B interruption. On the other hand, our policy for neonates with type A interruption and VSD has been to repair the interruption through a posterolateral thoracotomy and review the need to close the VSD at a later stage. This is similar to our approach in symptomatic infants with coarctation of the aorta and a VSD.\(^9\) It is true that both of our patients with type A interruption died. However despite severe renal failure patient 1 survived surgery for the IAA. Had he not progressed to chronic renal impairment surgical closure of the VSD would have been undertaken. Patient 2 survived surgery for both the IAA and the muscular VSDs. It was a combination of cytomegalovirus infection of the lung and septicaemia that contributed to her death. Because of the multiple VSDs, palliation (i.e. pulmonary artery banding) at the time of correction of the IAA might have been the better option in this case. Another situation in which a palliative approach might be preferable is the rare occurrence of type B interruption with an aberrant right subclavian artery. Banding of the pulmonary artery at the time of repair of the aortic interruption would divert flow up the aorta and stimulate growth of that vessel. Palliation is also indicated for those infants with complicated intracardiac defects, e.g. transposition of the great vessels, truncus arteriosus and single ventricle. In our 3 patients who died, the repair of the interruption was found to be adequate at autopsy. Patient 2, who had multiple VSDs, had a single small residual VSD. In the other case closure was complete. (Patient 1 did not have his VSD closed (Table 1).

In summary, the clinical picture in IAA is typically that of an initially well neonate who becomes acutely ill within the first week of life, when the ductus constricts. The femoral pulses weaken or disappear. When arch interruption is proximal to the left subclavian artery (i.e. in type B) the left brachial pulse also disappears. In the rare situation of type B interrup-

**REFERENCES**

4. Steidley RJ, Summ Chr u Med Both 177: 2: 114 (quoted in 7).