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Management of coarctation of the aorta in a neonate

by *H. S. Saw* *and* *K. T. Singham*
A. W. Grieve *A. E. Delilkan*

Department of Surgery, University Hospital, Kuala Lumpur.

Introduction

Coarctation of the aorta which is a constriction that may vary from slight to complete occlusion and which can be located at any site, was first noted by Morgagni (Keith, Rowe and Vlad, 1966) in the 18th century. Since the first successfully treated cases by Gross (1945) and later by Crafoord (1945) resection in older children and adults have become the standard treatment. It's recognition in early infancy and the reluctance to operate on these sick infants, in non-cardiac and non-paediatric oriented centres still remain a significant contributor to the high mortality rate of this serious condition.

We wish here to present a typical case presenting in the neonatal period.

Case Report

M.S. is a 32-day old male child who presented with complaints of difficulty in respiration of 1 day's duration. 3 days previously he was noticed to have a mild cough. There was no other history of note.

At initial evaluation, he was found to be mildly febrile with a temperature of 37.4°C. There was marked tachypnoea: the respiratory rate was about 80-100/min. This was associated with intercostal and subcostal recession and nasal flaring. Examination of the chest revealed fine crepitations in both lung bases.

He had a pulse rate of 180/min. No differential cyanosis was noted. The heart was enlarged and there was an obvious heave. No thrills were palpable. Although his radial pulses were readily palpable, the femoral pulses were either absent or very weak. Blood pressure measurement using the flush technique showed an upper limb systolic pressure of 110 mm Hg. while that in the lower limb was only 20 Hg. There was no obvious murmur heard in the precordium or back. The liver was about 1 cm. below the subcostal margin.

With these findings, a clinical diagnosis of coarctation of the thoracic aorta with associated congestive heart failure and pneumonia was made.

Arterial blood gas analysis showed the patient to be markedly acidotic. The chest x-ray done confirmed the cardiomegaly and pneumonia; there was no pulmonary oedema. Electrocardiograms showed marked right axis deviation and right ventricular hypertrophy.

The patient was started on crystalline penicillin, Digoxin and Lasix. He showed slight improvement initially but about 36 hours after admission, his condition deteriorated. More intensive therapy was instituted and when his vital signs stabilised, cardiac catheterization and angiography were performed. This revealed a right ventricular and pulmonary artery pressure of 56/8 and 55/28 mm of Hg. respectively. A size 5 NIH catheter was then passed from the right atrium through a patent foramen ovale into the left atrium and a left ventricular injection was done. This excluded the possibility of a VSD, PDA or hypoplastic left heart syndrome and confirmed the presence of the coarctation (Figs. 1 & 2).

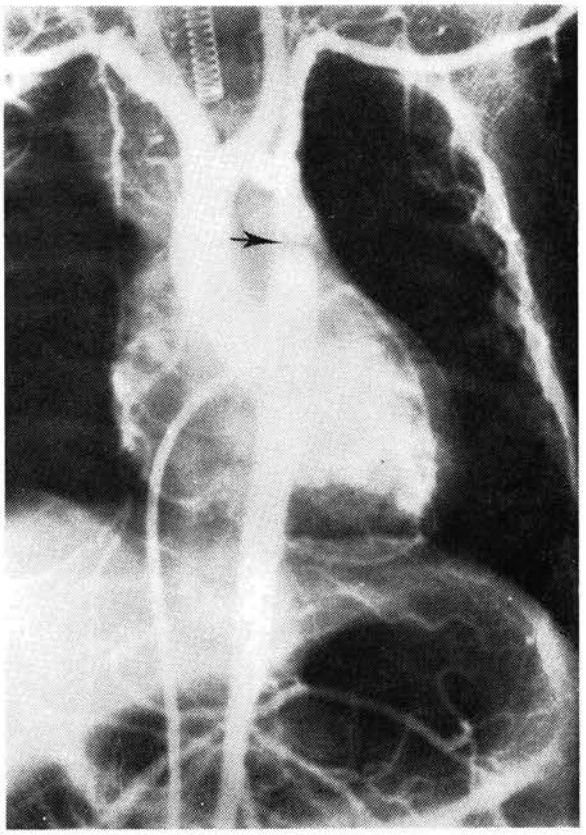

Figure 1: Aortogram AP view showing coarcted segment.

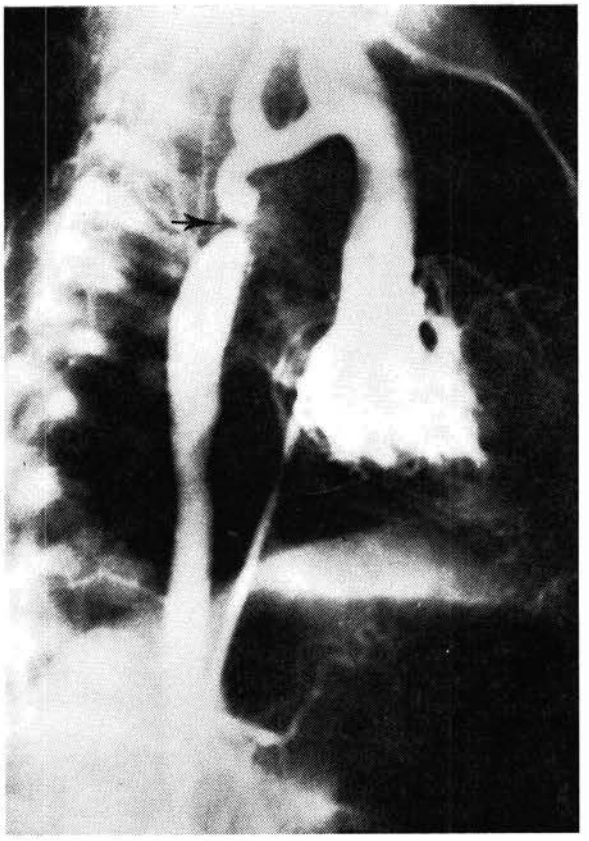

Figure 2: Aortogram: Lateral view.

Under the same general anaesthetic, the patient was wheeled into the Operating Theatre where a left thoracotomy and a resection of the coarcted segment was carried out. The coarctation was found to be a preductal (infantile) type.

Post-operatively, he was electively ventilated for 36 hours, after which he was extubated and nursed in a head box. His post-operative course was uneventful and he was discharged well on the 14th post-operative day with no medications.

Discussion

The classical clinical features are easily elucidated if a high level of suspicion is present.

Congestive heart failure is the commonest feature in coarctation. Interestingly, coarctation is also the commonest congenital heart lesion to present with congestive heart failure; other conditions that may present with heart failure in infancy include large patent ductus arteriosus, ventricular septal defects, closed septum pulmonary stenosis or atresia, total anomalous pulmonary venous drainage and endocardial fibroelastosis.

In 75% of patients there is a blood pressure difference between the upper and lower limbs (Waldhausen, *et al.,* 1964). This can often be elicited by the "flush technique". A difference of 20 mm of Hg. has been accepted as suggestive of coarctation. This difference may not be evident when the child is in overt heart failure but as congestive heart failure improves, the pressure difference will become more obvious, hence the importance of a high level of suspicion and the value of repeated blood pressure measurements.

Apart from the blood pressure difference in the upper and lower limbs, examination of the cardiovascular system in these young infants is very unrewarding in lending support to the diagnosis of coarctation. A murmur across the coarctation and murmurs of collaterals in the back are often absent. In addition, the so-called classical differential cyanosis, is invariably non-existent.

Cardiomegaly as confirmed by electrocardiograms and chest x-rays is frequently present but is not specific to coarctation alone.

Infections, especially of the respiratory system is a common feature and one should be very wary of making the diagnosis of congestive heart failure secondary to pneumonia.

The most useful investigation is cardiac catheterization and angiography. Although the clinical diagnosis may be obvious, the surgeon must convince himself that he is not dealing with a hypoplastic left heart syndrome, which to this day is considered inoperable in many centres. He should also know if there is any other intracardiac anomaly, especially a ventricular septal defect which may require an initial temporary banding of the pulmonary artery before the aorta is cross-clamped for resection of the coarctation.

Anaesthesia is a tricky proposition in coarctation of the aorta in infancy. The management involves two stages – first for the emergency cardiac catheterization and angiography and then for the emergency surgical correction.

Pre-operative assessment of the patient is vital to assess the left heart failure and the respiratory status. Digitalization and diuretic therapy are usually necessary (Keith, Rowe and Vlad, 1966) and pre-anaesthetic IPPV might be necessary depending on the respiratory state. With the exception of Hyoscine, no other pre-anaesthetic medication is given. Conscious intubation is the safest procedure and non-depolarizing neuro-muscular blocking drugs e.g. curare can be administered intravenously.

The neonate is known to be sensitive to non-depolarizing agents and titrating, incremental doses of 0.25 mgm d-tubocurarine should be given; it is our impression that after two such intravenous doses (0.5 mgm total dose) 50-70% of all neonates are paralysed while all neonates should be paralysed after a third dose (0.75 mgm total dose). The N_2O/O_2/ Relaxant controlled ventilation technique is the technique of choice.

During angiography, 5 mEq of Sodium Bicarbonate is given when the contrast is injected to cater for the metabolic acidosis which arises.

Operation is performed via a left postero-lateral thoracotomy through the 4th intercostal space. Rib resection is often unnecessary. The lung is displaced anteriorly and an incision is made in the mediastinal pleura over the upper descending extended superiorly over the left subclavian artery. The superior intercostal vein is ligated and transected in the process.

The aorta is mobilised circumferentially, proximally and distally for as long a distance as is possible taking care not to injure the intercostal arteries, which are usually large and delicate.

Traction on umbilical tapes passed about the coarctation, elevates the aorta and allows placement of vascular clamps proximal and distal to the coarctation. The ligamentum arteriosus or ductus arteriosus is then ligated and the coarcted segment excised.

Primary anastomosis is then carried out using 5-0 prolene as a continuous posterior layer. A few interrupted sutures are used for the anterior surface. This combination of continuous and interrupted sutures facilitates rapid anastomosis with some reduction of the risks of re-coarctation as the child grows. Very infrequently, the coarctation involves a very long segment and primary anastomosis without tension may not be feasible. Under such circumstances, the use of a short segment of woven dacron graft, the use of the left subclavian artery or patch aortoplasty (or isthmoplasty) may have to be resorted to.

During thoracotomy the problems which may be encountered are as for any thoracotomy in a neonate, namely, haemorrhage (the incision tends to be more bloody because of collaterals) and hypothermia. Deliberate hypothermia can lead to problems in the neonate unless the team is tuned to, and experienced in, its use. If collaterals are not well developed clamping of the aorta can endanger thoracic aorta posterior to the vagus nerve and the spinal cord and kidneys, necessitating the use of hypothermia or even left atrio-femoral bypass. The use of deliberate hypotension to lessen haemorrhage can be a problem in the neonate.

Monitoring the neonate during anaesthesia is always a technical problem because of the small size of the patient. An oesophageal stethoscope is invaluable as evidenced in this case. At one stage after the resection of the coarcted segment and end-to-end anastomosis of the aorta, pressure was put on the suture line for haemostasis. A marked bradycardia was picked up by the oesophageal stethoscope indicating interference with the vagus nerve. On removing the gauze, the bradycardia promptly disappeared confirming the vagal problem.

The results of larger series (Waldhausen *et al.*, 1964, Hallman and Cooley, 1975) show that apart from the complications of a standard thoracotomy