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Brief Report

Aortic arch thrombectomy in a 2.8 kilogram neonate – a case report and review of the literature

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Abstract Aortic arch thrombus is a rare occurrence in neonates. In the few described cases, this has mainly been associated with sepsis or early postnatal interventions, such as insertion of umbilical arterial line. We describe a case of occlusive aortic arch thrombus in a neonate who presented with signs of critical coarctation and successfully underwent surgical thrombectomy on deep hypothermic circulatory arrest. We also present a review of the most recently published cases of aortic arch thrombus in neonates and the treatment options employed.

Keywords: Aorta; thrombus; circulatory arrest

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History

A 2-DAY-OLD NEONATE WAS REFERRED TO US FROM the local hospital as having "critical coarctation of the aorta" with signs of left ventricular failure. The baby was born by normal vaginal delivery, with no history of maternal illness or antenatal/early postnatal interventions. There was an early rupture of maternal membrane – up to 72 hours before birth, otherwise no reported risk of sepsis. The baby was initially discharged home following a 24-hour period of postnatal observation. He, however, presented to the accident and emergency unit on day 2 of life with poor feeding, lethargy, and mild cyanosis around the lips. On physical examination, the femoral pulses were barely palpable. Initial echocardiogram at the local hospital showed "a long narrow coarctation segment" distal to the left subclavian artery, with severely impaired left ventricular function – ejection fraction 33%. The child was subsequently started on prostine and transferred to our unit.

On arrival and following another echocardiogram, a large thrombus was visualised in the aortic arch. The thrombus appeared to originate from the ampulla of the arterial duct, extending into the ostia of the left subclavian artery and the left common carotid artery (Fig 1). Other echo findings were a small patent foramen ovale, moderate mitral regurgitation, and reasonable left ventricular function. The aortic valve, ascending aorta, and arch were morphologically of good size. There was a large patent arterial duct with bidirectional flow across it.

In view of the findings and the baby's clinical condition, a decision was made for urgent surgical thrombectomy under deep hypothermic circulatory arrest.

Surgical procedure

The procedure was performed via a mid-line sternotomy. Cardiopulmonary bypass was established with an aortic cannula and a single venous cannula in the right atrium. The child was cooled in readiness for circulatory arrest, the duct was suture-ligated and transected, and the head vessels were mobilised and looped. At a core temperature of 18°C, the aorta was

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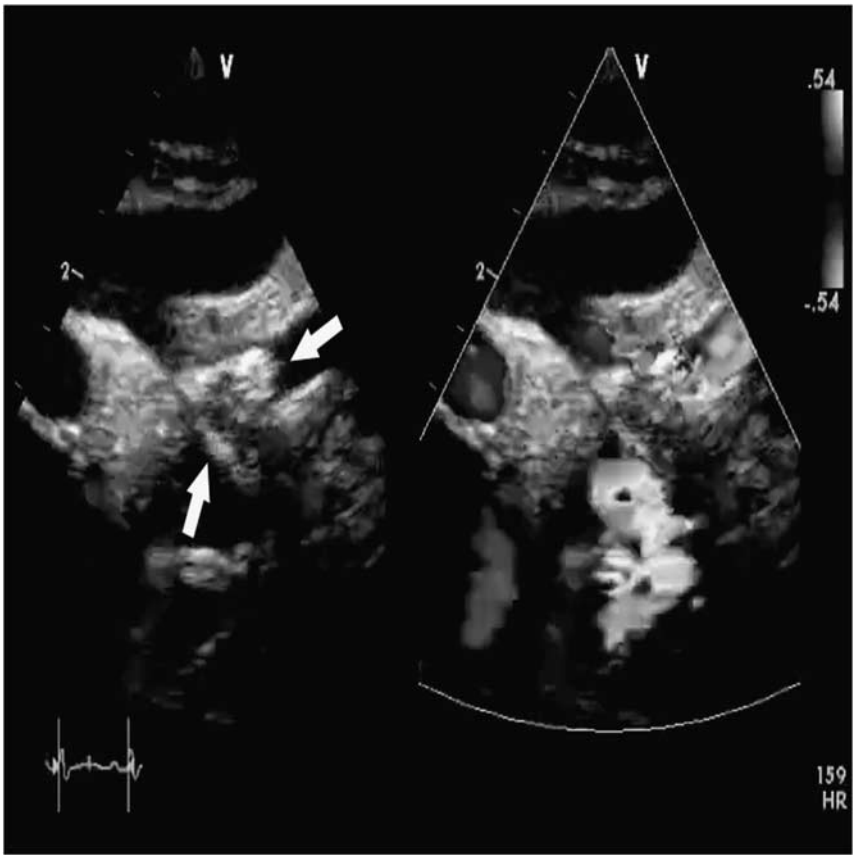

Figure 1.
Pre-operative echocardiogram. Arrows show a large thrombus in the distal arch and the left subclavian artery.

cross-clamped, cardioplegia instilled into the aortic root, and circulation was subsequently arrested. The head vessels were snared and the aorta transected at the level of the isthmus. The incision was extended proximally into the transverse arch. Distally, remnant ductal tissue with part of the aortic isthmus was excised. There was a large thrombus in the transverse arch, extending all the way into the descending aorta and partially occluding the left subclavian artery. To the naked eye, the thrombus appeared old and organised. There was no evidence of aortic coarctation.

The thrombus was completely removed and the two ends of the aorta were anastomosed using a running 6-0 surgilene suture. The small patent foramen ovale was also directly closed with 6-0 surgilene suture and cardiopulmonary bypass was re-established. The patient was re-warmed and subsequently weaned from bypass. Owing to supra-systemic pressures in the right ventricle, the child was started on nitric oxide and the chest was uneventfully closed.

Post-operative course

The child made a good recovery after initial difficulties associated with high pulmonary pressures. He was extubated on the 6th post-operative day and weaned off inotropic support. His intravenous heparin, initiated after surgery, was substituted for sub-cutaneous low-molecular-weight heparin before discharge from hospital. The child was discharged home 2 weeks after surgery and would remain on low-molecular-weight heparin for the next 3 weeks.

Post-operative echocardiogram showed a widely patent aortic arch with laminar flow in the descending aorta (Fig 2). It also showed significantly improved left ventricular function.

The results of septic screening carried out at the time of admission were all negative. Coagulation screening for possible thrombophilia, including activated protein C resistance, showed that all parameters were within the normal range. Factor V Leiden assay was not indicated as the modified activated protein C ratio was normal.

Discussion

Spontaneous aortic arch thrombus is a rare finding in neonates. Most of the reported cases have been associated with sepsis, perinatal asphyxia, inherited thrombophilia, or the insertion of umbilical

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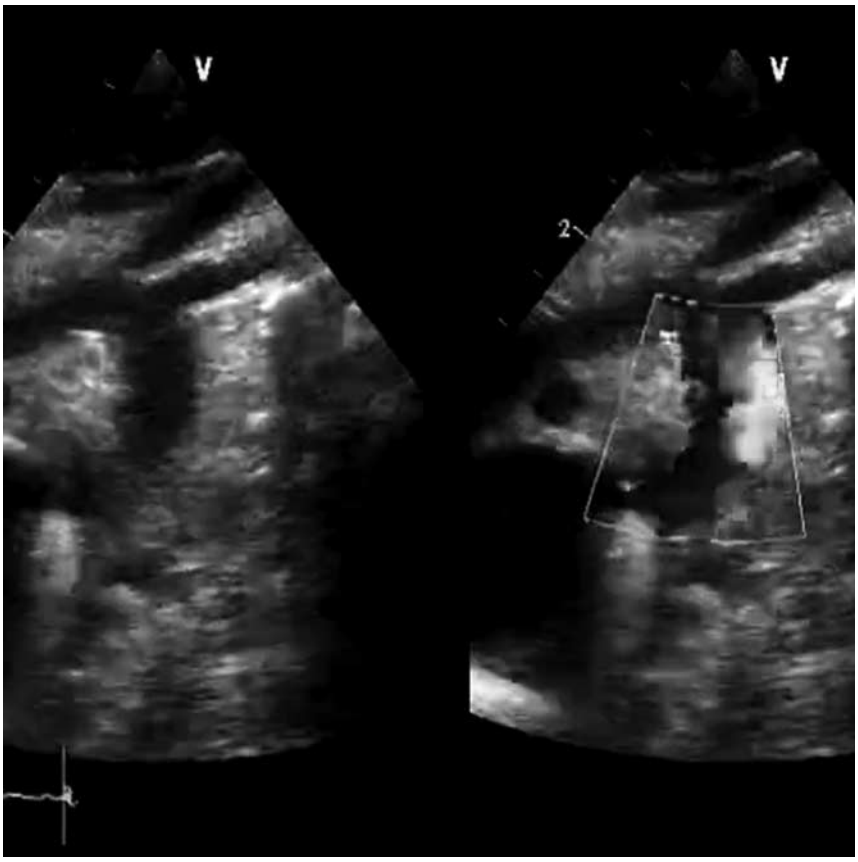

Figure 2.
Post-operative echocardiogram depicting laminar flow in the aortic arch.

catheters in these patients. Congenital cytomegalovirus infection has also been mentioned as a likely cause of neonatal aortic arch thrombus.

An online search of PubMed for articles relating to "aortic arch thrombus in neonates" returned a total of eight publications over the last decade. These were all case reports highlighting the often "misleading" clinical presentation of neonatal aortic arch thrombosis and the associated therapeutic challenges. On the whole, 12 patients were involved, of which 10 survived.

In one of the most recent reports, Francis et al[1] presented the case of an extremely preterm baby with occlusive aortic arch thrombus following the insertion of umbilical catheters. The large thrombus – located distal to the innominate artery and extending into the carotid artery – was successfully treated with intravenous heparin and subsequent low-molecular-weight heparin.

Low-molecular-weight heparin was also employed in the three cases reported by Sharathkumar et al.[2] The three neonates involved had aortic arch thrombosis and complete occlusion of a unilateral internal carotid artery and multiple cerebral infarcts. Thrombus resolution was reported to be complete in two of these neonates and near complete in one, with all the infants showing normal growth and development.

In addition to anticoagulation treatment with heparin, thrombolysis using tissue plasminogen activator or streptokinase can be an effective treatment option as shown in two of the four cases reported by Kenny and Tsai-Goodman.[3] Of the neonates, one had low levels of anti-thrombin III and was also shown to be heterozygotic for factor V Leiden mutation. Another case of aortic arch thrombosis in a neonate with heterozygous carrier status for factor V Leiden mutation was reported by Metsvaht et al.[4] Treatment with tissue plasminogen activator and streptokinase was, however, unsuccessful in this case.

Surgical thrombectomy has its place in the treatment of aortic arch thrombosis in neonates, as shown by our report and that of Das et al[5] who reported two neonates with thrombosis of the aortic arch and isthmus masquerading as coarctation of the aorta. Of the two neonates, one had neonatal transient protein C deficiency, whereas the other patient had severe perinatal asphyxia. The former had the arch thrombus successfully removed by

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surgery, whereas the latter died of complications associated with perinatal asphyxia. Aortic thrombectomy was also successful in one of the cases reported by Kenny and Tsai-Goodman.[3]

The choice of treatment for neonatal arch thrombosis varies from case to case and generally depends on the associated risk factors for bleeding, the extent of organ involvement or ischaemia, and, to a large degree, on the availability of surgical expertise.

Our decision to carry out surgical thrombectomy in our patient was informed by the rather acute clinical presentation. With the benefit of hindsight, the decision proved to be most appropriate as the thrombus was organised and, most probably, would have made any thrombolytic treatment unsuccessful. The use of deep hypothermic circulatory arrest afforded us the leeway of extending the arch incision well into the ascending aorta if the need did arise. In addition, a short period of deep hypothermic circulatory arrest – in this case 19 minutes – carries a very low risk of brain injury.

There was no clear pointer to the likely cause of arch thrombus in our patient. The perinatal history stated an early rupture of maternal membrane but made no mention of asphyxia or sepsis, and all clinical cum laboratory investigations were negative. We can only speculate about some undetected viral infection, but that is mere speculation.

In conclusion, neonatal aortic arch thrombosis, more often than not, remains a diagnostic surprise, with serious consequences if not appropriately treated. The choice of treatment should be on a case-by-case basis, taking into consideration the patient's clinical state, the inherent risks, and the availability of needed expertise.

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