Cardiology in the Young

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Percutaneous rescue therapy in a child with blocked cavo-pulmonary shunt

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Abstract

A 7-month-old infant presented with bilateral blocked cavo-pulmonary anastomosis within 2 months of surgery. Due to extreme haemodynamic instability, surgical options were abandoned and rescue intervention from left jugular line was planned. Acute thrombosis of the left-sided Glenn was noted with significant anastomotic narrowing. Successful rescue thrombolysis was done using recombinant tissue plasminogen activator (Alteplase) along with balloon dilatation of the attenuated segments.

Brief Report

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Case report

Over the years, bidirectional cavo-pulmonary anastomosis or Glenn procedure has been the standard of care primary palliation for the univentricular pathway. Most patients remain fairly well for the initial few years after the surgery.[1] We hereby report a child who presented to us with extreme cyanosis and severe respiratory distress within 2 months of bilateral cavo-pulmonary anastomosis.

This 7-month-old male infant with single ventricle and pulmonary atresia underwent bilateral cavo-pulmonary shunt surgery at 5 months of age. The child had borderline branch pulmonary arteries on pre-operative evaluation [RPA – 3.8 mm (−2.18 Z), LPA – 4.1 mm (−1.15 Z)]. However, the above procedure was done in preference to aorto-pulmonary shunt due to its potential advantages. The procedure was prolonged and challenging due to smallish branch pulmonary arteries. Post-operatively, echocardiography showed turbulent flow across both anastomosis (mean gradients of 3 mmHg on either side). As the child was relatively stable with decent saturation in room air (80−83%) and no congestive features suggestive of high venous pressures, it was decided to keep him on close follow-up. Unfortunately, he was lost to follow-up after discharge. He then turned up at our emergency with extreme desaturation and gasping respirations requiring urgent intubation and initiation of mechanical ventilation. It was brought to our notice that the child was suffering from fever and diarrhoea for the previous 3 days subsequent to which his general condition deteriorated to the present state.

After initial stabilisation in the cardiac ICU relevant investigations were done. The child had significant facial puffiness with prominent veins all over the chest wall. Chest X-ray revealed bilateral massive pleural effusion. Inter-costal tubes were urgently inserted to expand the lungs and chylous fluid was drained from either side. Alarmingly, echocardiography failed to show any flow across the Glenn anastomosis on either side, even after expansion of the lungs. The child continued to deteriorate with intermittent episodes of bradycardia requiring cardiopulmonary resuscitation. Emergency surgical options were contemplated but abandoned due to extreme risks and the child was urgently shifted to the cardiac catheterisation laboratory for rescue intervention.

Initial venograms were done from both sided ante-cubital veins. The right injection failed to visualise the right-sided superior vena cava. The azygous system filled up through multiple collaterals and drained retrogradely into the inferior vena cava (Fig 1a). The left superior vena cava also failed to show up and there was reversal of flow into the left internal jugular vein with a large filling defect at the lower end suggesting a thrombus (Fig 1b). In the hope of re-establishment of flows, urgent cannulation of left internal jugular vein was done and a 5 Fr introducer sheath was placed. A 0.018 × 150 cm angled Terumo wire was then passed into the sheath, carefully threaded through the thrombus, across the anastomosis into the distal left pulmonary artery, under fluoroscopic and angiographic guidance (Fig 1c). Injection Heparin 50 units/kg was given locally and another 100 units/kg via the systemic route. Systemic thrombolysis was initiated with injection Alteplase (recombinant tissue plasminogen activator) at a high dose of 0.3 mg/kg/dose.

The child showed marginal improvements in saturation over the next few minutes. Sheath angiogram highlighted partial restoration of flows but persistence of large thrombi at the lower end of the sheath as well in the lower branches of the left pulmonary artery (Fig 2a). There was also significant narrowing at the left superior vena cava and the left pulmonary artery

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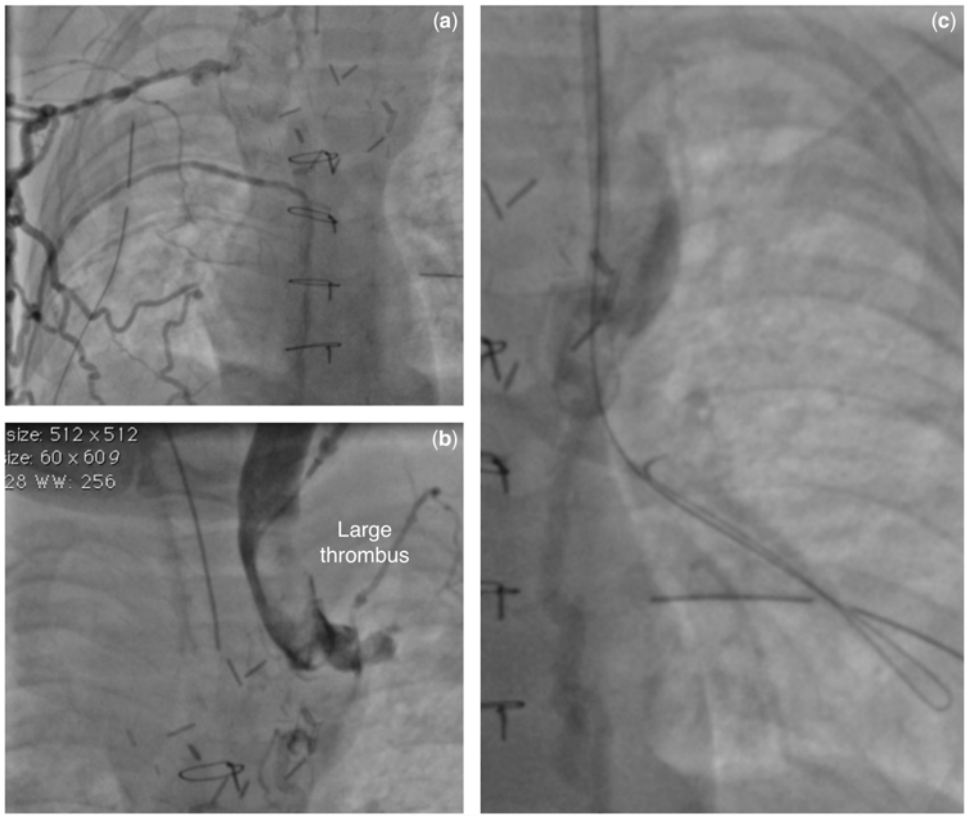

Figure 1. (**a**) Right venogram showing retrograde drainage to IVC. (**b**) Large thrombus at the lower end of left internal jugular vein, causing complete blockage. (**c**) Wire in distal left pulmonary artery.

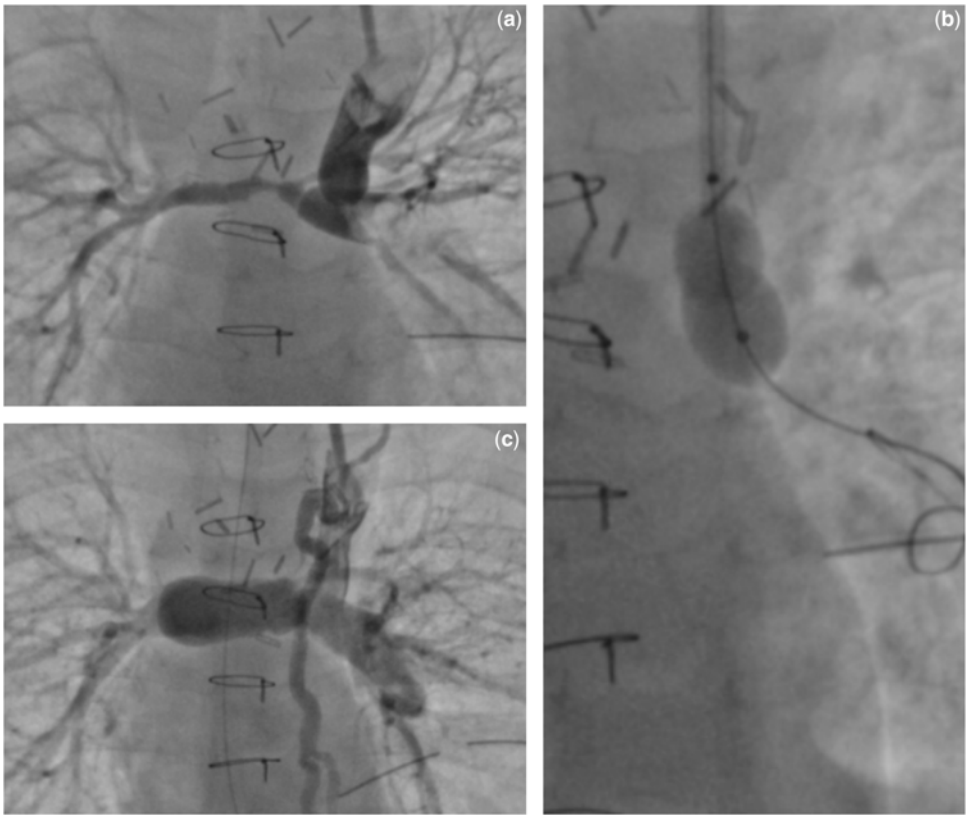

Figure 2. (**a**) Partial restoration of flows but significant thrombus load persisting. (**b**) Balloon dilation of left Glenn anastomotic site. (**c**) Complete restoration of flows after 10 hours of Alteplase infusion.

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anastomotic site (Fig 2a). The branch pulmonary artery confluence too appeared diminutive. A 8 × 30 mm Tyshak II balloon was used to adequately dilate the anastomotic site (Fig 2b). Another 0.018 × 150 cm Terumo wire was then placed in distal right pulmonary artery. The same balloon was used to dilate the pulmonary artery confluence to prevent complete blockage of the right pulmonary artery in case of thrombus breakdown and embolisation. The child was already on stiff dose of thrombolytic, but repeat sheath angiogram confirmed the persistence of large thrombi in the circuit, albeit with improved flow into both lungs. Frank bleeding was noted in the right inter-costal drain and the Alteplase infusion was reduced to 0.1 mg/kg/hour and connected to the neck sheath for better local action. The sheath was kept in situ and the child was shifted to the ICU with better haemodynamics.

Infusion Alteplase was continued for the next 10 hours. Repeat sheath angiogram done subsequently showed complete restoration of flows to both lungs with no thrombi anywhere in the circuit (Fig 2c). Right Glenn flow was not re-established but was not intervened at that point. Alteplase was stopped and Heparin infusion was continued for the next 24 hours. The child improved gradually and was successfully discharged after 7 days on oral Aspirin and Warfarin.

Discussion

Thrombotic complications are relatively rare after bidirectional cavo-pulmonary anastomosis.[2] Incidence in various studies ranges from 0 to 3%.[3,4] Most chronic thrombotic events happen when there is sluggish flow in the setting of poor compliance or inadequate drug prophylaxis. Aspirin is the usual prophylaxis of choice but some centres use oral anticoagulants alone or in combination.[5] In our case, the surgical anastomosis was difficult due to underlying pulmonary artery anatomy and the child had post-operative mild bilateral Glenn flow obstruction. He was discharged on Aspirin but then later presented to us in decompensated state with bilateral Glenn thrombosis. Perhaps the episode of diarrhoea and subsequent dehydration in the presence of the underlying anatomical narrowing led to the acute event.

After the initial venograms, it was clear that there was a relatively long-standing occlusion on the right side. However, on the left side, organised clot was visible within the vessel suggesting a more acute or sub-acute course. Once the thrombi could be crossed successfully with a wire, it was imperative to re-establish flows using thrombolytic therapy. Alteplase (recombinant tissue plasminogen activator) has been highly successful in re-establishment of flows in acute thrombosis of various aetiologies in children with a reasonable safety profile.[6] We used the high dose initially via systemic route and shifted to low dose infusion locally for better direct action.[7] While thrombolysis was life-saving, it was equally important to balloon-dilate the anastomotic site as well as the PA confluence and proximal right pulmonary artery to prevent further development and propagation thrombi.

There have been sparse reports on thrombolysis using recombinant tissue plasminogen activator in children who have undergone single ventricle palliation. In one such report, Nomura et al.[8] state the utility of catheter-mediated direct thrombolysis in a patient with thrombosed sub-segmental branches of right pulmonary artery after one and half ventricle repair. Another report mentions the successful use in a child on extra corporeal support with blocked Glenn shunt.[9] Our report adds to this select cohort suggesting that Alteplase (recombinant tissue plasminogen activator) may be quite useful as rescue thrombolytic therapy in such critical and challenging scenarios.

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Conflicts of interest. None.

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