Superior vena cava thrombosis causing respiratory obstruction successfully resolved by stenting in a small bowel transplant candidate

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Abstract

A 4 year old child was referred for small bowel transplantation. He had superior vena cava obstruction secondary to numerous central venous line placements; alternative routes for long term central venous access were compromised by extensive venous occlusive disease. Patency for the superior vena cava was reestablished with stenting, which allowed for radiological placement of another central venous line.

Long term survival in infants and young children with intestinal failure is dependent on adequate central venous access for the administration of parenteral nutrition. Line sepsis and physical damage to the catheter often necessitates multiple central venous catheter placements during their early life and these children are at risk of catheter related veno-occlusive disease. Recurrent sepsis and the loss of satisfactory venous access for the administration of parenteral nutrition is life threatening and is an indication for intestinal transplantation in up to 41% of patients reported by the small bowel registry.

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Keywords: bowel transplant; vena cava thrombosis

Case report

A child of consanguineous Pakistani parents developed profuse watery diarrhoea within weeks of birth and was extensively investigated at five months of age (including full thickness biopsy of the bowel, immunological work-up, faecal electrolytes and infectious disease screening). No specific cause was found. He was not thriving and started long term parenteral nutrition. By the time he was four years old he had under gone eight surgically tunnelled venous catheter placements, complicated by episodes of line sepsis. Haematological investigations excluded a thrombotic tendency and he was treated with Warfarin.

The child was referred for assessment for suitability for small bowel transplantation because it was feared that he would soon lose all viable venous access for parenteral nutrition. His superior vena cava (SVC) obstruction progressed with marked facial and neck swelling and respiratory symptoms necessitating the use of a continuous positive airway pressure device and an emergency tracheotomy. Ultrasound and venography confirmed extensive veno-occlusive disease involving the iliac veins, the right femoral vein, the lower

inferior vena cava, the left internal jugular vein, and both subclavian veins. A short complete occlusion was present in the superior vena cava and an extensive venous collateral network was present around the lower neck and upper mediastinum (fig 1).

Emergency treatment to re-establish superior vena caval patency was performed. A transjugular liver biopsy needle was passed through a vascular sheath in the right internal jugular vein and advanced into the right atrium by pushing through the SVC obstruction. A guide wire was passed through the lumen of the needle and pulled from the right atrium with a vascular snare out through a left femoral vein sheath. The transjugular needle was removed and the SVC obstruction was dilated using an 8 mm diameter balloon catheter passed over



Figure 1 Bilateral simultaneous arm venogram showing occlusion of both subclavian veins, with numerous collaterals, and upper thorax.

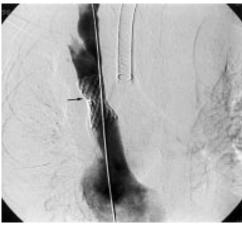


Figure 2 Post deployment venogram via the right internal jugular vein shows SVC stent satisfactorily placed with good flow into the right atrium.

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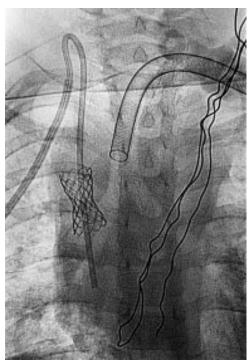


Figure 3 Chest x ray showing the Jostent (JOMED) through which dual 7French Hickman line is positioned with tip in the right atrium.

the guide wire from the left femoral vein. The balloon catheter was removed and a covered metallic 6–12 mm long Jostent (JOMED) was deployed in the SVC to cover the site of the obstruction. The stent was further balloon dilated with a good result and satisfactory flow as shown on venography (figs 2 and 3).

The child's neck and facial swelling improved dramatically after stenting and anticoagulation was continued with Warfarin. He





Figure 4 Clinical photographs of patient (A) immediatly after placement of SVC stent showing residual facial congestion and; (B) 12 months later, showing resolution of facial congestion and successful decannulation of tracheostomy.

remained well and it was possible to place a 7 French dual lumen Hickman line radiologically via the right internal jugular vein through the SVC stent into the right atrium (fig 4), when the existing catheter in the azygos system fractured. Four months after placement of the SVC stent the upper airway obstruction improved sufficiently for decannulation of the tracheotomy.

Discussion

This case describes the efficacy of radiologically re-establishing patency of the superior vena cava in a young child awaiting small bowel transplantation. Most published work and experience in the radiological management of superior vena caval obstruction has been adult based. ¹⁻³ Semi-urgent recannalisation of the SVC in a young child with severely compromised venous access and upper airway obstruction whilst awaiting small bowel transplantation has not previously been described.

We elected to cut through the complete SVC occlusion with a transcatheter method before stenting because the alternative, thrombolysis, may have taken up to 48 hours of intravenous treatment to clear thrombus and would have mandated intensive patient monitoring and repeated venography.⁴

The long term effects of stent in the growing child need to be taken into consideration and selection of the correct stent size is vital to match that of the normal SVC in the child. Stents used should allow for possible future expansion by balloon dilatation without losing radial strength to accommodate the growth of the child. Strategies for stent management in the growing child have previously been addressed, although there is limited experience in the venous system.⁵

In conclusion, recannalisation of the superior vena cava by the method described can immediately provide a route for central venous access and alleviate symptoms including upper airway obstruction. Creating a patent SVC allows access into the right atrium and can be a life saving procedure in infants and children dependent on total parenteral nutrition and can avoid the need for early intestinal transplantation.

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