

## CASE REPORT

# Percutaneous endovascular catheter aspiration thrombectomy of severe superior vena cava syndrome

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We report a combined percutaneous endovascular approach, including thrombus aspiration and catheter directed local thrombolysis, followed by systemic thrombolytic therapy to treat severe superior vena cava syndrome in a 2½ week old infant. This procedure was performed on the fifth postoperative day after major surgery. No treatment complications were observed. The only predisposing condition for thrombosis was a central venous line. No other acquired or genetic risk factors for thrombosis could be found.

The widespread use of central venous lines (CVL) in neonatal intensive care units has led to better management of neonates requiring major surgery and parenteral nutrition. However, this invasive practice has increased infectious and thrombotic complications. Treatment options include supportive care, anticoagulant therapy, thrombolytic therapy, and surgery.<sup>1,2</sup>

Alternatively, mechanical thrombus disruption using soft wires and balloon angioplasty in conjunction with continuous site directed thrombolytic infusion into the clot (local pharmacomechanical thrombolysis),<sup>3</sup> as well as transcatheter recanalisation and subsequent balloon dilatation of the occluded vessel (without local thrombolysis),<sup>4</sup> have proved to be effective in infants and children with major thromboses.

Management problems arise when heparin resistant central venous thrombosis with severe superior vena cava syndrome develops shortly after major surgery, precluding aggressive management with fibrinolytic agents.

We report a 2½ week old neonate carrying a CVL, who developed a catheter related thrombosis with severe superior vena cava syndrome on the second postoperative day after major abdominal surgery.

### CASE REPORT

A 3130 g male infant was born at 40 weeks gestation. Prenatal ultrasonography showed polyhydramnios and dilated, fluid filled bowel loops. The boy was born by normal spontaneous delivery. The Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. No resuscitation measures were required. Initial treatment included nasogastric decompression. The infant's physical examination was unremarkable apart from a tense, distended abdomen. Exploratory laparotomy at the first day of life revealed seven jejunal atresias. Resection of all atretic segments with a proximal and distal stoma was performed. Two weeks later, a CVL was inserted via the left internal jugular vein; an end to end anastomosis combined with a tapering enteroplasty of the proximal distended bowel segment and a Bishop–Koop stoma was performed. On the second postoperative day, the patient developed swelling and discoloration of the head, neck, and upper limbs, as well as bilateral pleural effusions. Thrombosis of the superior vena cava and innominate vein was diagnosed by ultrasound and confirmed by

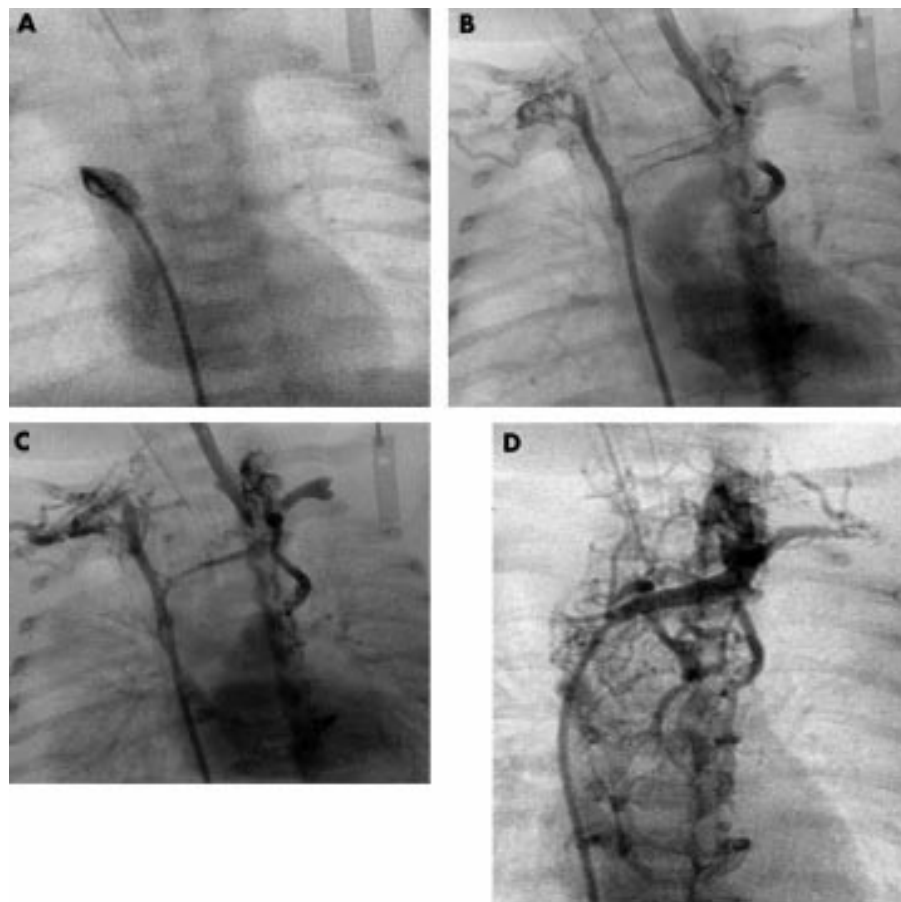
venography. Doppler investigations showed no blood flow through the thrombus.

Relevant laboratory evaluation included: platelets  $469 \times 10^9/l$ , INR 1.0, aPTT 41.2 s, fibrinogen 2.8 g/l, and D-dimer 2.4 mg/l (normal range  $<0.3$  mg/l).

The CVL was removed and treatment with unfractionated heparin (UFH) started with a continuous infusion rate of 30 IU/kg/h to maintain the aPTT at 50% above normal. Because the size of the thrombus had remained unchanged and progression of superior vena cava syndrome was noted during the next three days, we decided to perform a percutaneous endovascular catheter aspiration thrombectomy in combination with catheter directed local thrombolysis.

A venous sheath was placed into the right femoral vein. The catheter was inserted through the sheath into the distal end of the superior vena cava. Angiography confirmed complete thrombotic occlusion (fig 1A). A guidewire was advanced through the catheter into the thrombus. Once the catheter had been advanced, the guidewire was removed and multiple thrombi were aspirated, while the catheter was pressed gently into the thrombus. This was followed by fibrinolytic treatment with two local applications of 0.2 mg/kg recombinant tissue type plasminogen activator (rt-PA) into the superior vena cava as slow bolus injections over 10 minutes. Partial reperfusion was noted (fig 1B). The guidewire was then advanced into the innominate vein and again multiple thrombi were aspirated, leading to partial recanalisation (fig 1C). Flow through the innominate vein could be further improved by two doses of 0.2 mg/kg rt-PA, given into the innominate vein as slow bolus injections over 10 minutes (fig 1D). The sheath was left in place and we started systemic treatment with low dose continuous infusion of rt-PA at 1.0 mg/kg/day and low dose UFH (5 IU/kg/h). However, reocclusion occurred during the following 48 hours. We therefore repeated the percutaneous endovascular catheter aspiration thrombectomy procedure (without local application of rt-PA) and started treatment with UFH at a higher dosage (which maintained the aPTT at 50% above normal), continuous infusion of rt-PA at 2 mg/kg/day, and multiple bolus infusions of rt-PA (given as slow bolus injections over 30 minutes into a peripheral scalp vein). Dosage of bolus injections of rt-PA were started at 0.2 mg/kg and subsequently raised to 0.9 mg/kg until Doppler investigations of blood flow improved markedly. Prior to each bolus administration of rt-PA, cranial ultrasound was performed to assess for the presence of intraventricular haemorrhage. Three days later, thrombolytic therapy was stopped. With this regimen, the clot dissolved completely, blood flow in the superior vena cava normalised, and the infant's condition improved, with regression of superior vena cava syndrome.

**Abbreviations:** CVL, central venous line; LMWH, low molecular weight heparin; rt-PA, recombinant tissue type plasminogen activator; UFH, unfractionated heparin



**Figure 1** Angiography into the distal end of the superior vena cava confirms complete thrombotic occlusion (A). Angiography after endovascular thrombus aspiration and two local applications of 0.2 mg/kg rt-PA into the superior vena cava reveals partial recanalisation (B). Partial recanalisation of the innominate vein after percutaneous endovascular thrombus aspiration (C). Improved flow through the innominate vein after two doses of 0.2 mg/kg rt-PA, given as slow bolus injections (D).

Pleural effusions disappeared gradually. All cranial ultrasound examinations were negative for signs of intraventricular haemorrhage.

During thrombolytic therapy, prothrombin time, aPTT, fibrinogen, and D-dimers were measured twice daily. Fibrinogen was maintained at levels between 1.8 and 3.2 g/l. Evaluation for thrombophilic disorders revealed no abnormalities: protein C, protein S, antithrombin III, plasminogen, fibrinogen, lipoprotein (a), and homocystein were in the normal range. Assays for the C677T mutation in the methylenetetrahydrofolate reductase (MTHFR) gene, the G20210A mutation in the prothrombin gene, and the G1691A mutation in the factor V gene, as well as assays for anticardiolipin antibodies and lupus anticoagulants were negative.

To avoid reocclusion after thrombolysis, infusion of UFH at a dosage to maintain the aPTT at 50% above normal was given for 45 days, followed by anticoagulation with low molecular weight heparin (LMWH). The further course was complicated by jaundice (direct reacting hyperbilirubinaemia with a maximal value of 11.5 mg/dl) which resolved spontaneously, and by gastrointestinal disturbances requiring reoperation. Enterostomy closure was performed at 55 days of life and the patient was discharged home in good clinical condition at the age of 2½ months. LMWH treatment was continued for a further period of six months.

## DISCUSSION

The use of CVLs to optimise parenteral nutrition and monitoring of critically ill neonates has become common practice in

neonatal intensive care units. However, the use of CVLs is associated with increased infectious and thrombotic complications.

It is important to prevent immediate and potential long term complications in catheter related deep vein thrombosis. Short term sequelae of catheter related occlusion include organ and limb dysfunction, chylothorax (as part of superior vena cava syndrome), as well as thromboembolic events. Long term sequelae after thrombotic events are not well defined but may be considerable.<sup>5</sup>

There is no consensus in the literature about the treatment of choice for neonates with central venous thrombosis. The degree of thrombus extension, the patient's clinical symptoms, the underlying cause of thrombosis, and the increased rate of complications encountered with more aggressive treatment should guide the choice of therapy. Treatment modalities are supportive care, anticoagulation, thrombolysis, transcatheter recanalisation, and subsequent balloon dilatation of the occluded vessel, local pharmacomechanical thrombolysis, and surgery.<sup>1,3,4</sup>

Our patient presented a management dilemma because thrombosis was diagnosed on the second postoperative day after major abdominal surgery, precluding aggressive medical management with fibrinolytic agents. Therefore we elected to start UFH therapy at a dosage that maintained the aPTT at 50% above normal. Unfortunately, this resulted in progression of superior vena cava syndrome. As surgical thrombectomy as well as thrombolytic therapy were considered too risky, we opted for percutaneous endovascular catheter aspiration thrombectomy<sup>6</sup> with catheter directed thrombolysis, which

resulted in partial reperfusion. However, reocclusion was noted despite systemic low dose continuous infusion of rt-PA and low dose UFH, which may reflect marked activation of coagulation caused by damage of the intima as a result of the thrombus aspiration procedure. Reocclusion after the second percutaneous endovascular catheter aspiration thrombectomy could be avoided by increasing the doses of UFH and rt-PA. Dosage and duration of thrombolytic therapy after the second aspiration thrombectomy were determined by the response to treatment and by exclusion of bleeding complications prior to each rt-PA bolus infusion.

We suggest that percutaneous endovascular catheter aspiration of central vein thrombi (either with or without catheter directed local application of thrombolytic agents) may represent a potential therapeutic alternative in neonates with severe superior vena cava syndrome, particularly in patients where medical management fails or where thrombolytic therapy and surgical thrombectomy are contraindicated or considered too risky.

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