

## Anaesthesia for massive venous malformation with DIC and strategies for minimising blood loss. Case report

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### Key points

Venous Malformations are the most commonly occurring vascular malformation. The life-threatening manifestations of DIC are rarely reported. The anaesthetic considerations are multifactorial and requires particular investigation into respiratory, cardiovascular and coagulation complications. Surgery may be prolonged and difficult with major blood loss anticipated. Positive outcomes of endovascular balloons have been reported; reducing surgical duration by providing a clear surgical field and reducing intra-operative blood loss. Evidence for cell-salvage is limited within paediatric surgery but benefits include improved oxygen-carrying capacity and reduction of allogeneic transfusion. It is both cost-saving and cost-effective.

### Abstract

We present our experience of anaesthetising a 7 year-old girl with massive venous malformation causing rarely associated Disseminated Intravascular Coagulation (DIC). The malformation extended across her neck, anterior chest wall and upper limb. The exceptional size of this lesion meant sclerotherapy was not viable and high-risk surgical resection was the only treatment option. As part of a multi-modal strategy to minimise peri-operative haemorrhage, we employed intermittent endovascular occlusive balloons. Commencing our anaesthesia in the angiography suite, an intra-arterial device was placed in the left subclavian artery, which when inflated also occluded of the left internal mammary artery and left vertebral artery. A left venous balloon was inserted in the left brachiocephalic/subclavian vein. The patient was then transferred to the surgical theatres where a subtotal resection was performed over a total of 18

hours. In addition to the balloons, we utilised a CAT cell salvage device to conserve transfusion demand.

This case is of particular interest due to lesions size, site and directly resulting consumptive-coagulopathy. The successful employment of endovascular balloons is rarely reported in the literature to have been used in this type of paediatric surgery, but has been cited as a useful adjunct in reducing blood flow during surgery. The prolonged, complex surgery and management of potentially life-threatening haemorrhage were considered in advance by a multi-disciplinary team. This was essential and responsible for the positive outcome of this case.

**Keywords:** Congenital syndromes, PICU, General Anaesthesia, Massive transfusion, Interventional radiology, Endovascular occlusion device.

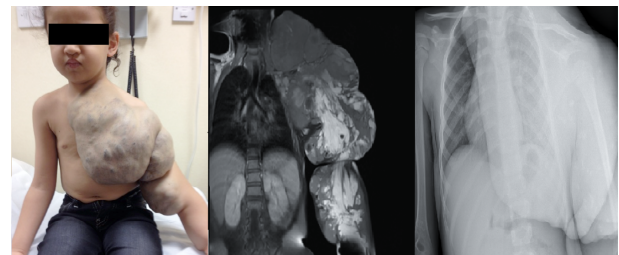
## Introduction

Venous malformations are the most commonly occurring vascular malformations (1:5-10,000). 40% are found in the head and neck, characterised by low-flow, compressibility, rapid-filling and growth from birth. Treatment options include laser, sclerotherapy or surgery. Combinations may be required to treat complex lesions with complete treatment not always achievable.

## Case report

The patient was a 30 kg, 7-year-old girl from Libya. She presented to our institution with a massive venous malformation extending across the anterior chest-wall and upper limb [figure1]. From 8-months of age, the malformation developed to such a size that her parents were required to take precautions to prevent trauma and potential exsanguination. She had no respiratory or airway symptoms. Echocardiography showed normal cardiac function. Haematological investigations were grossly abnormal, showing consumptive coagulopathy, resulting directly from the malformation. A test-dose of fibrinogen increased the fibrinogen levels to 0.3 and reduced the APTT to 45. Sclerotherapy was excluded because the malformation had a large stromal component, likely to be unresponsive and the potential adverse effects from the high-dose of sclerosant required for the size of lesion. The procedure began in the angiography suite with induction of Fentanyl (1mcg/kg), Propofol (2.5mg/kg) and Atracurium (0.5mg/kg). Both mask ventilation and intubation were uncomplicated. Via femoral access, a left subclavian artery balloon was inserted, which when inflated also occluded the left internal mammary artery. A left brachiocephalic/subclavian venous balloon was inserted. A double-lumen femoral vas cath and a basilic double-lumen PICC line were inserted. Intravenous heparin was commenced. Following transfer to theatre, standard monitoring continued plus invasive blood pressure (BP), central venous pressures (CVP), nasopharyngeal temperature and urine output. Active warming was maintained with a fluid warmer and bairhugger®. Maintenance anaesthesia was provi-

ded by isoflurane with remifentanyl (0.1-0.2 mcg/kg/min). To minimise intraoperative blood loss, the vascular-occlusive-balloons were inflated for 1hour periods and deflated for 20minutes on continuous cycling. We utilised a Continuous Auto-Transfusion (CAT) cell-salvage device. Fibrinogen (2 g), Tranexamic acid (10mg/kg) and calcium were administered. Regular blood samples were reviewed by a consultant haematologist and allogenic blood products administered accordingly. In total: 5 administrations of fibrinogen, 19 units of blood, 5 units of FFP, 5 units of platelets and 2445mls of autologous blood were given. Continuous assessment of fluid balance with replacement resulted in 10,000mls of crystalloid or colloid being infused. Total anaesthetic time was 18 hours with a surgical subtotal resection of 14 hours.



**Figure1.** Pre-operative photograph of the venous malformation with MRI and chest radiograph imaging

## Discussion

Anaesthetic considerations for venous malformation surgery are multifactorial, dependant on site, extent, age and clinical manifestations. Particular inquiry into respiratory, cardiovascular and coagulation complications is required. Surgery may be prolonged, difficult with major blood loss. Respiratory considerations include airway compression, sleep apnoea, difficult intubation, dyspnoea with postural features. Cardiovascular complications include high-output failure, venous hypertension and tachycardia. A spectrum of coagulopathy and electrolyte disturbance may be seen. Intraoperative management should include a strategy for a cardiovascular stable induction, anticipating ventilation or intubation difficulties. Monitoring should be continuous using in-

vasive-pressure devices, both to optimise fluid balance and facilitate regular blood sampling. Emphasis on patient warming, positioning and pressure areas are important. For blood loss, adequate products should be available with sufficient anaesthetic and portering staff dedicated to the case. Controlled phlebotomy, in cases where compression of the lesion is employed, may prevent congestive failure associated with auto-massive transfusion [1]. Post-operatively, high-level care is required for such lesions and staged surgical procedures often necessary. Ongoing bleeding is compounded by pre-existing coagulopathy. Cardiac-output monitoring should be available. Tracheomalacia may complicate extubation. Common manifestations of slow-flow malformations are phleboliths and Local-Intravascular-Coagulation (LIC). Factors such as stress, infection, trauma or surgery may trigger progression to DIC. Development of DIC due to venous malformations is rarely seen, with a cohort of only 6 children between 1980 and 2005 reported by our tertiary-referral centre [2]. The aims for managing DIC, are replacement of consumed clotting-factors and maintaining adequate platelet count and fibrinogen level, ultimately increasing oxygen delivery to the tissues. By binding to Antithrombin-III, Heparin inhibits thrombin, decreasing the amount of circulating clot. It can prevent pain, thrombosis and decompensation of LIC to DIC. The use of endovascular balloons are rarely reported to have been used in this type of paediatric surgery, but have been cited as a useful adjunct in reducing intra-operative blood flow[3]. Common surgical applications include high-risk obstetric haemorrhage, resection of sacral tumours and managing major blood loss in pelvic fractures. Provision of a clear surgical field reduces surgical duration and intra-operative blood loss, with a subsequent reduction in transfusion. Distal thrombosis has been reported and where arterial occlusive devices have been used alone, an increase in venous bleeding has been described. Organ dysfunction from ischaemia and reperfusion can be reduced by shortening inflation times. Pre-operative

vessel embolization has alternatively been performed, but collateral vessels may develop[4]. Traditionally little evidence existed for use of cell-salvage in paediatric practice, mainly due to the 300ml minimum processing requirement. Today, non-centrifugal devices, such as the CATS, can process any volume. Indications for use in paediatric cardiac, craniofacial and orthopaedic surgery, is supported by evidence showing a cost-effective reduction in allogenic blood transfusion[5]. Further advantages include increased erythrocyte 2,3-DPG and ATP, mean erythrocyte viability of 88% and maintained bioconcavity, which improves oxygen-carrying capacity.

#### **Key learning points**

1. Venous malformations are associated with numerous multisystem complications requiring particular investigation.
2. DIC is a rare, life threatening complication of venous malformations, greatly increasing the risk of peri-operative morbidity.
3. Endovascular occlusive-balloons were used effectively as part of a multi-modal blood conservation strategy.

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#### **Conflict of interest**

The authors declare no conflict of interest

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