

Anaesthesia recommendations for patients suffering from

Klippel-Trénaunay syndrome

Disease name: Klippel-Trénaunay syndrome

ICD 10: Q 87.2

Synonyms: Angio osteohypertrophy, naevus vasculosus osteohypertrophicus, capillary venous lymphatic malformation (CLVM)

Disease summary: Klippel-Trénaunay syndrome is a rare congenital malformation with an incidence of 1 out of 27,500 live births, characterized by a triad of venous malformations or varicose veins, cutaneous capillary malformations and bony or soft tissue hypertrophy in affected limbs. The lung, trunk, gastrointestinal tract, neurovascular structures and the bladder may be involved with the presence of vascular malformations.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong



Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Orthopaedic surgeries to correct limb discrepancies, debulking surgeries and amputations; interventions like sclerotherapy and laser therapy for vascular malformations, vascular surgeries like surgical stripping and endoscopic ligation of perforating veins, and surgical resection of the bowel in case of gastrointestinal haemorrhage.

Type of anaesthesia

General anaesthesia is safer. Central neuraxial blockade is fraught with risk due to haemangiomas and spinal arteriovenous malformations and neurovascular malformation in the surrounding structure of the spine, tendency for coagulation disorders, and venous dilation, all of which can cause epidural hematomas.

Necessary additional diagnostic procedures (preoperative)

Preoperative cardiology evaluation must be ensured as many patients have venous thrombophlebitis (50%) and pulmonary thromboembolism (22%) which may lead to pulmonary hypertension and right ventricular failure. Arteriovenous malformations can produce high output congestive heart failure. Preoperatively, deep vein thrombosis prophylaxis must be considered. If neuraxial blockade is planned, it is mandatory to do preoperative CT/MRI to rule out vascular malformations in the CNS and ensure absence of cutaneous lesions overlying site of needle insertion. Preoperative coagulation profile must be done in these patients. Enough packed RBC should be ready before the operation for appropriate fluid resuscitation in case of haemorrhage.

In certain cases, preoperative embolisation can be carried out to reduce intraoperative bleeding. This will need close collaboration with interventional radiologists.

Particular preparation for airway management

Difficult intubation must be anticipated as patients may have facial anomalies, upper airway angiomias and soft tissue hypertrophy in the airway.

Particular preparation for transfusion or administration of blood products

Potential for massive intraoperative haemorrhage must be considered. In this syndrome, local intravascular coagulation occurs within the malformation and distal flow has depletion of coagulation factors. This is also often associated with disseminated intravascular coagulation and Kasabach-Merritt syndrome (consumptive coagulopathy and thrombocytopenia). The anaesthesiologist must therefore be vigilant with sufficient intravenous access (although limb involvement can limit peripheral venous access), adequate blood and blood product reserve and appropriate monitoring intraoperatively.

Particular preparation anticoagulation

Deep vein thrombosis prophylaxis must be considered as many patients have venous thrombophlebitis (50%) and pulmonary thromboembolism (22%).

Particular precautions for positioning, transport or mobilisation

Not reported.

Probable interaction between anaesthetic agents and patient's long-term medication

Not reported.

Anaesthesiologic procedure

General anaesthesia is commonly used. If central neuraxial blockade is considered, CT/ MRI imaging of the spine is carried out to rule out vascular malformations and examination of the back done to ensure there are no cutaneous lesions at the planned site of needle insertion. If central venous pressure monitoring is planned, ultrasound guidance can help to rule out vascular malformations of the jugular/ subclavian veins.

Femoral cannulation is better avoided in view of lower limb thrombophlebitis with venous anomalies. The possible presence of brain haemangiomas pose a risk for intraoperative intracranial haemorrhage. Hence measures must be taken to minimize surges in blood pressure, especially during intubation, extubation and at the time of surgical incision.

Particular or additional monitoring

If massive blood loss and consequent haemodynamic instability is anticipated, it will be beneficial to have invasive monitoring with intra-arterial blood pressure monitoring and central venous pressure monitoring. It is important that the patient stays normotensive during induction and throughout the operation. Fluctuation of blood pressure might lead to a hypertensive state that could potentially cause rupture of multiple intracranial and peripheral arteriovenous shunts, aneurysms and capillary malformations. Other complications such as internal bleeding from vascular abnormalities and fistulas, might be exacerbated by elevated blood pressure.

Maintaining a normotensive state is important if the patient is positioned prone during the surgical operation. Prone positioning is associated with predictable changes in cardio-pulmonary physiology. In the prone posture, pressure on the abdomen compresses the inferior vena cava and femoral veins, diverting blood from the distal parts of the body into perivertebral venous plexuses.

Essential part of the anaesthetic plan is the preparation for unexpected vascular complication such as hypertensive and hypotensive states. Nitroprusside sodium, dopamine hydrochloride and phenylephrine infusions may have to be kept ready before the surgical operation.

More, It is important to bear in mind that excessive venous pulsation can result in inaccurately low pulse oximetry reading if the probe is placed on an affected limb.

Possible complications

Massive blood transfusion might be necessary as haemodynamic instability is a possibility even in minor surgeries, due to intraoperative blood loss, as a result of presence of widespread varicosities and venous malformations. Thromboembolic complications are likely.

Postoperative care

The level of monitoring needed for each individual depends on the surgical procedure done, the preoperative status, and the intraoperative complications.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.: not common

Not reported.

Ambulatory anaesthesia

Not reported.

Obstetrical anaesthesia

Thromboembolic, cardiovascular and haemorrhagic complications are exacerbated in pregnancy , so multidisciplinary team involving obstetricians, anaesthesiologists, radiologists, cardiologists, haematologists and paediatricians must individualise each case to decide on management. Prophylactic anticoagulation is advised in the post partum period.

Literature and internet links

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