

Anesthesia recommendations for **Klippel-Trénaunay syndrome**

Disease name: Klippel-Trénaunay syndrome

ICD 10: Q87.2

ORPHAcode: 90308

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.

Differential diagnostics: Parkes-Weber syndrome, a congenital vascular disease that consists of capillary, venous, lymphatic, and arteriovenous malformations, can be misdiagnosed as Klippel-Trenaunay syndrome. The presence of arteriovenous fistulas is the differentiating feature in Parkes-Weber syndrome. Doppler ultrasound and magnetic resonance angiography can help to diagnose and differentiate these syndromes [1,2,3]. Klippel-Trenaunay syndrome should be differentiated by the other overgrowth syndromes presenting as hemihypertrophy and other vascular syndromes with cutaneous lesions such as CLOVES (congenital lipomatous overgrowth vascular malformation with epidermal nevus and skeletal abnormalities); MCAP syndrome (megalencephaly-capillary malformation); DCMO (diffuse capillary malformation with overgrowth); FAO (fibro adipose overgrowth); Proteus syndrome, post-natal skeletal and connective tissue overgrowth with epidermal nevi and vascular malformation; Beckwith-Wiedemann syndrome characterized by hemihypertrophy, neonatal hypoglycemia, omphalocele, ear abnormalities; Maffucci syndrome-multiple enchondromas; plexiform neurofibromatosis in NF-1 presenting as localized hypertrophy [4].

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this guideline Perhaps new knowledge.



Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.



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

Emergency information

A	AIRWAY / ANESTHETIC TECHNIQUE	<p>GA is preferred, but intubation could be difficult as patients may have facial anomalies, upper airway angiomas and soft tissue hypertrophy within the airway. Careful and gentle intubation, with hemodynamic response blunted by opioids/betablockers, may be needed if cerebral angiomas are present, to prevent intracranial bleeds.</p> <p>RA: Central neuraxial block has high risk for epidural hematomas, as there could be AV malformations in the spine. Peripheral nerve blocks should also be done carefully with ultrasound guidance to prevent inadvertent vascular puncture.</p>
B	BLOOD PRODUCTS (COAGULATION)	<p>Local intravascular coagulation occurs within the malformation and distal flow may have depletion of coagulation factors, resulting in Disseminated intravascular coagulation, consumptive coagulopathy and thrombocytopenia. Massive blood loss can occur.</p> <p>Hence, full coagulation profile should be checked preoperatively, and adequate blood and blood products should be arranged. Autologous transfusion can be considered. Preoperative embolization should be done whenever possible to prevent massive blood loss.</p> <p>Some patients may be on DVT prophylaxis in view of their high incidence of thromboembolism.</p>
C	CIRCULATION	<p>Preoperative cardiology evaluation must be ensured as many patients have venous thrombophlebitis and pulmonary thromboembolism which may lead to pulmonary hypertension and right ventricular failure. Arteriovenous malformations can produce high output congestive heart failure.</p>
D	DRUGS	None reported.
E	EQUIPMENT	<p>To avoid direct sustained pressure on bony prominences or areas with significant vascular malformations to prevent skin breakdown, injury to vessels, and potential bleeding; use pillows or specialized gel devices to cushion vulnerable areas.</p>

Typical surgery and procedures

Orthopedic 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 [5], neurosurgical procedures such as coiling or clipping of intracranial aneurysms [6], and surgical resection of the bowel in case of gastrointestinal hemorrhage [7].

Type of anesthesia

General anesthesia (GA) appears safer.

Central neuraxial block is fraught with risk due to hemangiomas and spinal arteriovenous and neurovascular malformations in the surrounding structure of the spine, tendency for coagulation disorders, and venous dilation, all of which can cause epidural hematomas [7]. Epidural hemangioma rupture can occur spontaneously or due to trauma caused by the needle, leading to progressive paraplegia [8].

Peripheral nerve blocks have a high risk of vascular puncture due to prominent superficial vessels, so should be done carefully and only with ultrasound guidance, specifically with high resolution ultrasound or superficial echography [9].

Necessary additional preoperative testing (beside standard care)

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 [10]. Preoperatively, deep vein thrombosis prophylaxis must be considered. If neuraxial block 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 hemorrhage [5]. In certain cases, preoperative embolization 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 [5]. Intubation should be gentle and carefully performed to avoid trauma and bleeding in the airway. If vascular malformations are suspected in the airway, it will be prudent to use videolaryngoscopy for optimum visualization of the larynx during intubation [11].

Hemodynamic responses to laryngoscopy and tracheal intubation maneuvers must be minimized or avoided with opioids and beta-blockers. The sudden increase in blood pressure may result in increased intracranial pressure and rupture of cerebral hemangiomas [12].

Particular preparation for transfusion or administration of blood products

Potential for massive intraoperative hemorrhage 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) [13]. Anesthesiologists 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. Autologous transfusion can be considered preoperatively. The use of an intraoperative autologous blood collection device may help reduce the amount of blood needed for transfusion [9].

Particular preparation for anticoagulation

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

Particular precautions for positioning, transportation and mobilization

To avoid direct sustained pressure on bony prominences or areas with significant vascular malformations to prevent skin breakdown, injury to vessels, and potential bleeding; use pillows or specialized gel devices to cushion vulnerable areas, such as the heels, elbows, hips, and knees, while positioning for surgery.

Disproportionally enlarged and asymmetric limbs may pose a challenge to positioning, hence careful preoperative planning for extra extensions may be needed to accommodate oversized limbs on the operating table.

Interactions of chronic disease and anesthesia medications

Not reported.

Anesthetic procedure

GA is commonly used. If central neuraxial block 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 [5]. If central venous pressure (CVP) monitoring is planned, ultrasound guidance can help to rule out vascular malformations of the jugular / subclavian veins [7]. Femoral cannulation is better avoided in view of lower limb thrombophlebitis with venous anomalies. The possible presence of brain hemangiomas pose a risk for intraoperative intracranial hemorrhage. Hence, measures must be taken to minimize surges in blood pressure, especially during intubation, extubation and at the time of surgical incision [9].

Particular or additional monitoring

If massive blood loss and consequent hemodynamic instability is anticipated, it will be beneficial to have invasive monitoring with intra-arterial blood pressure monitoring and CVP

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 cardiopulmonary 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 anesthetic 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. Moreover, 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 hemodynamic 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.

Disease-related acute problems and effect on anesthesia and recovery

Not reported.

Ambulatory anesthesia

Not reported.

Obstetrical anesthesia

Thromboembolic, cardiovascular and hemorrhagic complications are exacerbated in pregnancy, so multidisciplinary team involving obstetricians, anesthesiologist, radiologists, cardiologists, hematologists and pediatricians must individualize each case to decide on management. It is recommended to evaluate, with periodic imaging and coagulation studies, vascular malformations in the pelvis, uterus, and vagina and to identify neuraxial alterations, to guide the safest method of delivery and anesthetic technique for these patients [14,15]. Prophylactic anticoagulation is advised in the postpartum period.

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