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Perioperative care of the vascular anomaly patient

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ABSTRACT

Patients with vascular anomalies present specific and unique challenges to providers of their postoperative care. Vascular anomalies can range from localized solitary lesions to diffuse lesions with vessel malformations and associated soft tissue, muscle, organ, and bone involvement. Perioperative issues for these patients can be complicated and include coagulopathies requiring anticoagulation, the need for postoperative surgical drains and specialized wound care, and the use of compression garments to maintain the desired postoperative result. This article will address these specific concerns for patients with vascular anomalies in order to create a framework for consistent and appropriate perioperative care.

Patients with vascular anomalies can present with unique perioperative needs related to the particular nature and characteristics of their vascular tumor or malformation. Preoperative evaluation may require assessment and management of associated coagulopathy and co-morbidities. Coordination of multiple services including the potential need for preoperative interventional radiology procedures such as caval filter placement or embolization is often essential. Of particular interest during the perioperative period is the management of anticoagulation as well as postoperative surgical drains, wound care, and use of compression garments. These issues will be discussed in order to create a framework for pertinent perioperative care specific to patients with vascular anomalies.

Perioperative anticoagulation

Patients with slow-flow vascular malformations are at an increased risk for hematologic complications, which include both bleeding and clotting. Coagulopathy affecting both hemostasis (control of bleeding) and thrombosis (regulation of blood clotting) occurs when blood stagnates in abnormal slow-flow vessels, thereby activating the coagulation system. This results in a process known as localized intravascular coagulopathy (LIC) marked by low fibrinogen, elevated D-dimer, and mild thrombocytopenia, which can progress into disseminated intravascular coagulopathy (DIC) with consumption of coagulation factors.[1]

The etiology of coagulopathy is multifactorial. Abnormal endothelial lining leads to interactions with blood products that initiate the coagulation system. Due to variations in channel size and structural abnormalities, flow abnormalities can occur resulting in local pooling of blood and stasis that may further damage the endothelium and activate the coagulation process.[2]

The risk of complications secondary to coagulopathy is increased after surgical or invasive radiological procedures involving many vascular malformations. Identification of high-risk patients prior to procedures will help the team to prepare these patients in order to decrease their risks for hematologic complications. Patients at high risk for hematologic complications include those with venous malformations, lymphatic malformations, and combined malformations, such as CLOVES and Klippel–Trenaunay (capillary lymphaticovenous malformation) syndromes. These are slow-flow lesions with abnormal venous anatomy and a lymphatic component that are prone to hemorrhagic as well as thrombotic events.

To identify which patients with these diagnoses are at risk for hematologic complications, a complete preoperative hematologic evaluation is necessary prior to considering any invasive procedure. This includes blood work, which may differ by institution but at a minimum should include CBC, PT, PTT, D-dimer, and fibrinogen. Obtaining an antiphospholipid antibody panel, thrombotic profile (protein S, protein C, antithrombin III, plasminogen, and plasminogen activator inhibitor), and thrombotic polymorphism panel (factor V Leiden, prothrombin 20210, PAI-I genotype, and MTHFR genotype) should also be considered.

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The patients identified as being at high risk for complications based on their laboratory results, extent of the lesion, or the presence of large ectatic vessels are prepared by initiation of low-molecular-weight heparin (LMWH) at prophylactic (0.5 mg/kg/dose; maximum dose of 60 mg/dose) or treatment dose (1.0 mg/kg/dose) once or twice daily. The LMWH is usually given for 14 days prior to the invasive procedure and should be stopped 12–24 h prior to the procedure. LMWH is generally restarted 12–24 h following the procedure depending on the extent of postoperative bleeding and the expected period of inactivity. It should be administered for 14 days following the procedure or until the patient returns to reasonable activity, whichever is longer. This will help to reduce the risk of clotting events even in patients with only mildly elevated D-dimers or with normal fibrinogens.[3]

Careful planning should be done prior to the procedure, including holding blood products for the procedure and communicating the risks and plan for management with the anesthesia provider. Patients with high D-dimers and/or a low fibrinogen may benefit from cryoprecipitate dripped in slowly throughout the procedure to decrease the risk of bleeding/clotting. For those highest risk patients, it is important to have other blood products on hold or ready, including packed red blood cells, FFP, and platelets. Recombinant factor 7 (NOVO seven) should be available for emergency use if the patient has extensive, uncontrolled bleeding during the procedure.

Some patients with extensive lesions and chronic pain can see an improvement in their pain with the use of daily anticoagulation therapy for extended periods of time. Coumadin and vitamin K antagonists do not seem to be as effective with this group of patients, so LMWH is the therapy of choice. Dosing will be at a prophylactic level of 0.5 mg/kg/dose once or twice daily. If the patient is on long-term therapy with LMWH, it is important to check anti-factor Xa levels regularly with a goal of < 0.5 units/ml for prophylaxis. This level needs to be drawn 4–6 h following a dose of LMWH. These patients will also need regular blood work, including CBC and liver function tests. A yearly DEXA scan should be performed for chronic use of LMWH secondary to the potential risk of osteopenia.

Novel oral anticoagulants that directly target factor Xa are being used in adults with some evidence that they help with coagulopathy associated with vascular lesions similar to the effect of LMWH. Studies will need to be performed in children to prove the efficacy and elucidate the side-effect profile of oral agents prior to changing these recommendations to use oral agents in place of LMWH.[4]

Patients identified as high risk may need to be admitted overnight after undergoing sclerotherapy. Sclerosing agents can cause red cell hemolysis resulting in hemoglobinuria, which can damage the kidneys. Intravenous fluids, running at 1.5 times the maintenance rate, are recommended for adequate flushing. If the urine is red post-procedure, sodium bicarbonate is added to the intravenous fluid to alkalinize the urine. Patients are encouraged to drink larger than normal amounts of fluids for several days following the procedure to keep the urine dilute.

Patients with large, ectatic vessels may undergo preoperative embolization of the large vessels or may need extra precautions during surgery. They often will receive a single dose of LMWH during the procedure to decrease the risk of clotting or bleeding. They may also have the extremities that are not directly involved in the surgical procedure compressed with wraps throughout the procedure to decrease the risk of thrombus formation. Careful evaluation and planning for patients identified as high risk will help to make invasive procedures safer and decrease the number of hematologic complications. A team of specialists who are experts in the care of individuals with vascular anomalies is best prepared to provide knowledgeable and appropriate care for these patients. Severe and fatal pulmonary emboli have occurred even at our experienced centers, where we have learned to be very proactive about screening and prevention.

Postoperative surgical drains

Patients with vascular anomalies who undergo surgical resection of their lesions may require intraoperative placement of a surgical drain to evacuate dead space and drain blood and serous fluid from the surgical area during the postoperative period. Surgical drain systems can be open or closed and active or passive. Active, closed surgical drains, such as Jackson–Pratt® drains and Blake® drains, use negative pressure to remove accumulated blood and serous fluid and reduce dead space to promote healing.[5] Active, closed surgical drains are particularly important in both patients with primary lymphatic anomalies and those with a combined vascular anomaly with a significant lymphatic component. Both of these types of patients have a propensity for profound lymphatic flow into the surgical field postoperatively. If not well drained, this can result in fluid collections that contribute to swelling of the surgical site, which may adversely affect the integrity of a closed surgical wound. A single drain may be used if the surgical area is localized, while multiple drains may be necessary when the resection is extensive.

Initially, the fluid draining through the surgical drain will be sanguineous, but the fluid will slowly become more serosanguineous over time. The amount of drainage in the immediate postoperative period before the patient becomes more ambulatory may be minimal or reduced, but patients and families are counseled to expect an increase in drainage as the patient becomes more mobile. Drainage can range from minimal amounts in patients with non-lymphatic vascular anomalies to hundreds of milliliters drained in each 24-h period for patients with an associated lymphatic component. The amount of drainage will gradually decrease over time until it slows enough for the body to reabsorb the fluid on its own. Patients with surgically resected vascular anomalies, particularly those with a significant lymphatic component, may need to have the drain in place for weeks to months.

Patients and families are advised to empty the drainage bulb as often as it appears nearly full or at least once every 24 h and to measure and record the drainage amount. A small dressing may cover the drain tubing insertion site. This dressing may be changed either once daily or when soiled. The patient and family may opt not to cover the insertion site with a dressing if they are comfortable with this option and there is no drainage around the site.

The drain tubing should be milked (stripped) on a regular basis in order to remove any small clots or tissue debris that may prevent the fluid from draining freely. If there is a decrease in fluid draining through the tubing or an increase in leakage of fluid around the tubing at the insertion site, this may be a sign that the tubing is blocked and requires milking.

Patients may shower and infants may have a sponge bath with the surgical drain in place. Patients who like to swim may continue to do so in clean, chlorinated swimming pools when they have recovered to the point where the surgical wound is sufficiently healed and they are ready to return to more normal activities. With both bathing and swimming, a dab of antibiotic ointment should be placed around the drain insertion site to prevent water from entering the site.

Patients and families are advised to report drainage amounts calculated in 24-h increments every 2 or 3 days so that a decision can be made when the drain(s) can be removed. Generally, if the output drops to 20 cm^3 or less every 24 h for a period of 4 or