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Ventral Hernia Mesh Repair

Giant omphalocele are defects larger than 10 centimeters encompassing the liver within the defect with a total loss of abdominal cavity domain. Babies born with giant omphalocele are seldom closed primarily during the neonatal due to the development of abdominal compartment syndrome, compression of the inferior vena cava and suprahepatic veins leading to multisystemic organ failure and death. The defect is covered with some bacteriostatic ointment and the amnion left to granulate creating neoskin as simple cover. Once the defect is covered with the growing skin the hernia is closed either using a prosthetic mesh, component separation or primarily. Primary repair has a 25-52% recurrence rate and is used for small < 5 cm defect. The component separation technique (CST) enlarges the abdominal wall surface by translation of the muscular layer without compromising the blood supply and innervation of the muscles. A longitudinal cut is made in the external aponeurotic fascia lateral to the rectum encompassing closure in the midline. This can be used when closing hernias between 5 and 10 cm in diameter. CST technique has a 33% of wound complications and 30% re-herniation rate. Transection of the perforating branches of the epigastric artery interfere with the blood supply of the skin of the ventral abdominal wall who will need collaterals from the intercostal artery and pudendal artery to survive. Prosthetic material can be synthetic or biologic. Prolene is a cheap synthetic mesh that creates adhesion, erosions and fistula. Biologic mesh are biodegradable unless process like cross-linking the collagen fibers take place. This extracellular material derived from human or other mammalian animal. Broad range of size helps repair larger hernia defects. Biologic mesh minimized adhesions between the mesh and viscera and incites fibrous tissue to grow and create a tough fascia with secure fixation of the mesh to the abdominal fascia. Biologic mesh are preferred to be placed underneath the peritoneal fascia (sub-lay or underlay). Biologic mesh should be biocompatible, non-toxic and nonimmunogenic. Neither antibiotic coverage nor subcutaneous drainage has an effect in the incidence of wound-related complications when placing this mesh.

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Sibson Hernia

Sibson's fascia also called the suprapleuralis membrane is the thickened portion of the endothoracic fascia extending over the cupola of the parietal pleura and reinforcing it. It is attached to the inner border of the whole length of the first rib and the first costal cartilage and transverse process of the 7th cervical vertebra. With increase in intrapleural pressure the lung apex may protrude into or through the fascia and rise a variable distance into the neck. This protrusion is referred as Sibson's hernia or apical lung hernia. The protrusion of the lung apex may be unilateral or bilateral. The right side is more frequently involved than the left. The apical segment of the lung protrudes between the scalenus anterior and sternocleidomastoid muscle. Sibson hernia is generally asymptomatic except for a swelling in the neck during coughing or Valsalva maneuver. Also, it can be associated with dysphagia. It can be diagnosed on posteroanterior and lateral chest films but CT-Scan of the chest provides a confirmatory diagnosis. It may cause problems during insertion of internal jugular or subclavian catheters and may result in inadvertent pneumothorax if not diagnosed properly. Sibson hernia has been associated with Ehlers-Danlos syndrome. Most cases of Sibson or apical lung hernia in children are identified incidentally, have a benign clinical course and resolve without surgical intervention. Surgical management is reserved for traumatic hernias and lung tissue at risk for strangulation, incarceration or punctured. The hernia resolves after discontinuation of continuous positive airway pressure breathing when physiologic tidal pressures are obtained.

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Beta Thalassemia

Sickle cell anemia and beta thalassemia are considered the most frequent inherited blood disorders around the world. Thalassemia is a genetic disease involving the genesis of hemoglobin either chains alpha or beta. Beta thalassemia refers to defective

production of the beta chain of hemoglobin. Beta thalassemia occurs when one or both Beta globin genes are either abnormal or absent. The child with thalassemia develops anemia from lower levels of hemoglobin and lacks good quality hemoglobin for oxygenation due to ineffective erythropoiesis. The gene mutation in beta thalassemia can be mild to severe, also classified as thalassemia minor or major respectively. Some patients will only need iron supplementation (non transfusion dependent thalassemia), while major cases of mutation might need regular blood transfusions for life. Hemolysis of blood is rapid, hence an overload of bilirubin is managed within the liver, reasons why some patients developed bilirubin gallstones (black pigmented stones). Patients with thalassemia experience loss of appetite, jaundice, an enlarged spleen or liver and several bone problems (osteoporosis). Iron building up in the heart leads to failure and death. A family history if thalassemia increases the chances of an individual being affected by the disease. Thalassemia affects patients with Italian, Asian, African, Middle Eastern and Greek ancestry. Whenever operating in a child with thalassemia the type of thalassemia should be ascertain as major, minor or intermedia. Also, the present of hemoglobin and iron overload must be measured. Current management of thalassemia involves red blood cell transfusions and iron chelation. Allogeneic bone marrow transplant is the only curative option limited by the availability of matching donors and graft-versus-host disease. Gene therapy using lentiviral vectors aim to correct the mutated beta-globin gene or add back a functional copy of beta or gamma-globin.

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