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Component Separation Technique

The component separation technique (CST) was initial developed in 1990 to repair primary large ventral hernias defects without using mesh or prothesis. CST was originally described as incising the external oblique aponeurosis and releasing the external oblique from the internal oblique. This release leads to improved tissue mobility and decreases tension allowing fascial approximation closure either to be done primarily or intercalating a mesh when the defect is very large. The CST does not compromise the innervation and blood supply of the muscles. The procedure is performed in adults to repair large giant ventral hernias with lost of abdominal domain. The procedure is rarely used in children. In short the technique encompass identifying the borders of the hernial defect, creating bilateral tissue flaps (skin and subcutaneous tissue) until the external oblique aponeurosis is exposed, bilateral longitudinal incision of the external oblique fascia one centimeter lateral to the rectus muscle dividing the external oblique muscle from the internal oblique muscle until the midline can be approximated with minimal tension. At this moment of closure the surgeon might use onlay or underlay biologic mesh to support the midline closure. The mesh can alleviate further the tension that exists with the hernia repair and provide a scaffold for cellular growth. In adults with large ventral defects the risk of recurrence is decreased when the CST closure is supported by either mesh onlay or underlay. In children, CST can be utilized for large ventral hernia defects after initial management of abdominal wall defects such as omphalocele and gastroschisis. The CST offers the potential for definitive repair as a newborn as well as avoidance of the morbidity associated with recurrent hernias and multiple operations. Complications associated with the CST include wound infection, seroma, hematoma or skin necrosis.

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Immune Thrombocytopenic Purpura

Immune thrombocytopenia purpura (ITP) is an acquired autoimmune disease characterized by platelet destruction due to antiplatelet autoantibodies. The reticuloendothelial system engulfs the affected platelets causing early destruction hence thrombocytopenia. In children is self-limited with a 6-month remission rate in more than 70% of cases. After six months the condition is termed chronic ITP and it occurs in 20% of all cases. Clinically mean age at onset is six years, males and females are equally affected, and the disease usually follows an infection. Most children present with simple petechiae and bruising symptoms. Most children with ITP experience spontaneous resolution without medication. Severe bleeding is more likely in children with severe thrombocytopenia from ITP. First-line therapy for symptomatic and/or severely thrombocytopenic patients include steroids, anti-D antibody, or intravenous gamma globulin. Second-line therapy is reserved for those who have severe bleed or are refractory to first-line medical therapy and include high-dose steroids, rituximab, and other chemotherapeutics. Laparoscopic splenectomy is a safe, efficacious, and cost-effective strategy option for children with uncontrolled chronic ITP or life-threatening hemorrhage complicating acute ITP. Unfortunately there is no clear prediction which patient will benefit from splenectomy. Almost 70% of cases respond favorably and persistently to splenectomy. Steroid-resistant children are most likely to have complete respond to splenectomy. Older age, longer duration of ITP, and male gender correlates with complete response. Splenectomy improves quality of life of children with ITP. Intracranial hemorrhage is a devastating complication of ITP preceded by headache and mucosal bleeding which can be precipitate in the event of aspirin intake, extreme exercise, head trauma or a congenital vascular cerebral lesion.

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Lipofibromatosis

Lipofibromatosis (LF) is a rare benign fibrofatty tumor of childhood largely composed of adipose tissue traversed by bundles of spindle fibroblastic-like cells. Most lipofibromatosis are found in the distal extremity. Clinically, LF presents as a slow growing painless mass with a propensity to affect the hands and feet. The lesion is subcutaneous in location or in the deep soft tissue with poorly demarcated margins. The lesion usually measures one to 3 cm, with a median size of 2 cm. LF can infiltrate adjacent structures like vessels, nerves and muscle. Functional impairment is very rare. The findings at MRI are nonspecific, but it can evaluate the extension of the lesion and involvement of adjacent structures. Hyperintense signal on T1W and T2W images with loss of signal on fat-saturated images confirm the lipomatous content of the lesion. Diagnosis can be made easily with distinctive histopathologic features without an indication of immunohistochemistry. Mitotic rate is low and there is usually no cellular atypia. Management of LF is complete surgical excision if at all feasible. Due to its infiltrative nature, LF has a high rate of recurrence. The following events were more common in the group with recurrent or persistent disease: congenital onset, male sex, hand and foot location, incomplete excision, and mitotic activity in the fibroblastic element.

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