



PEDIATRIC SURGERY Update*

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Vanishing Testicular Syndrome

Testicular regression syndrome, also known as Vanishing testicular syndrome (VTS), refers to the disappearance of a normally developed testis during fetal life. The spermatic cord ends blindly as evidence of a testis that was present but disappeared. VTS occur usually unilateral the consequence of late antenatal or perinatal vascular thrombosis, torsion or infarction seen in less than 5% of undescended testes. The testis is non-palpable in almost 20% of cases of undescended testes in children and of these 50% are directly caused by VTS. VTS is more common than testicular agenesis. Due to earlier normal descent of the left testis anatomically this site is affected more commonly than the right testis. Most VTS are sporadic events while a few are associated with severe mental retardation or members of the same family. The clinical presentation is an empty scrotum with non-palpable testes in the inguinal canal or elsewhere. Once a child is found with a non-palpable undescended testis a diagnostic laparoscopy is in order to find an intraabdominal testis. When vas deferens and spermatic vessels are visualized exiting the internal inguinal ring on laparoscopy a groin exploration is usually carried out to identify and remove the testicular tissue remnant (nubbin) associated with the vanishing testis. Fibrosis, dystrophic calcification, giant cell reaction and hemosiderin deposition in association with identifiable testicular structure are features of VTS nubbin. Histopathological examination has confirmed the presence of germ cells in 10% of VTS testicular nubbin remnants. The pathological diagnosis of VTS can be established by the presence of the following diagnostic criteria: 1) a vascularized fibrous nodule with calcification and/or hemosiderin, and 2) a minimum of a vascularized fibrous nodule with cord elements in proximity. Intraabdominal testes are six times more likely to develop a testis malignant tumor than the palpable undescended testis in the inguinal canal. There is controversy whether to remove the nubbin from the VTS due to the low rate of viable germ cell tissue and almost negligible incidence of malignant transformation.

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Leiomyoma

Leiomyoma is a benign tumor usually encountered in the genitourinary (uterus) and gastrointestinal tract (small bowel and esophagus) in adults. Leiomyomas are slow growing tumors that can occur in any body part where smooth muscle is present. It is a very rare tumor in children. The skin is the second most common location for leiomyomas after the uterus. Depending on the site of origin leiomyomas can be classified into three types: piloleiomyoma (cutaneous), angioleiomyomas (vascular) and dartoic leiomyomas (deep soft tissue). Piloleiomyomas originate from the arrector pili muscles of the hair follicles. Angioleiomyomas come from the vascular smooth muscle, while dartoic leiomyomas originate from the smooth muscle of genital skin. Leiomyomas may present clinically as either solitary or multiple lesions. In the skin they can have a red-surface and most commonly located in the extremity. Piloleiomyomas are the most common type of leiomyoma in the skin ranging in diameter from two to 20 mm and they are typically painful. The pain may be spontaneous or the result of exposure to cold, pressure, emotional stress or growth of the tumor compressing surrounding nerve tissues. Piloleiomyoma are non-encapsulated circumscribed dermal tumors composed of multiple fascicles of smooth muscle in an interlacing whorled arrangement. Calcifications have been reported in isolated deep tissue leiomyomas. Differential diagnosis includes osteochondroma, desmoid tumor, spindle cell carcinoma and other soft tissue tumors from the adjacent region. Although leiomyomas are benign they have a low but definite malignant potential and should be removed. Atypical cells, necrosis and mitotic activity must be interpreted as a warning sign in terms of malignant transformation and post=surgical recurrence. Definitive diagnosis is established after surgical removal. The management of leiomyomas is surgical excision whenever the anatomic location permit. The prognosis is good for patients after complete surgical resection.

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Fracture CVC

As medicine advance and more invasive treatment options are available the number of children needing long-term central venous access increases. Long term venous access can

be achieved using central venous catheters (CVC) such as Hickman, Broviac, percutaneous inserted central catheter (PICC), subcutaneous implantable ports or umbilical catheters. Most CVC are safe but complications do occur. Most complications can be categorized as infectious, thrombotic or mechanical. Fracture central venous catheter is a serious mechanical complication that occurs rarely. CVC fracture can be defined as a break in the sterility of the line that allows leakage as a result of a tear, hole, or shearing injury in the external line tubing. These fractures occur more frequently in peripherally inserted catheters. Factors that predispose CVC to break are related to the characteristic of the catheter, the insertion or removal technique, and the clinical problem for which they were used. Silicone catheters are prone to fracture at or near the entrance site where the caliber of the line narrows. Catheter damage may be caused by the introducer needle in PICC lines. Some catheters might rupture in the axillary area due to repeated stress caused by flexion and extension of the arm. Repeated compression of the catheter between the clavicle and first rib in the costoclavicular ligament area can lead to fracture of the CVC (Pinch off syndrome). If the catheter is occluded and high pressure infusion is utilized the catheter can rupture. Two primary methods are utilized to manage CVC fracture: line replacement or line repair. CVC comes with repair kit. The incidence of infection is not increased after repairing the fracture catheter. Line repair can be performed safely at the bedside. The major drawback of CVC repair over replacement is decreased durability of the catheter. Risk of leaving catheter fragments in the child body can include pulmonary embolism, sepsis, endocarditis, arrhythmias and cardiac perforation. Due to the risks associated with fragmented catheters they should be removed. This is performed using a percutaneous endovascular approach.

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