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Ondine's Curse

Congenital Central hypoventilation syndrome (CCHS), also known as Ondine curse, is a rare disorder of ventilatory control characterized by a lack of response to normal respiratory stimulants, especially hypercapnia. The child develops prolonged apnea, cyanosis and hypoventilation in the absence of cardiac, pulmonary or neuromuscular disease. Low respiratory rates during sleep are characteristic of Ondine's curse. Initial mechanical ventilation dependency is followed by ability to sustain respiration during awake periods. Ondine's curse is believed to be caused by aberrant development of neural crest tissue (neurocristopathy), and as such is associate with Hirschsprung's disease mostly of the long segment variety. It can also be associated with multifocal congenital neuroblastoma, ganglioneuroma, neuroma and hypothalamic dysfunction. Abnormalities of the eye and autonomic nervous system are also common. CCHS can potentially be managed with long-term assisted mechanical ventilation, or electrodes surgically implanted on the phrenic nerve to pace the diaphragm. Electrodes can be placed thoracoscopically. Diaphragmatic pacing can result in successful weaning from mechanical ventilation, appears to be effective in reducing pulmonary vascular resistance and pulmonary hypertension, and improve the quality of life in children with CCHS. Diaphragmatic fatigue and loss of phrenic nerve conductivity seem to be causes of failure of electrical pacing. The prognosis for patients with CCHS is poor with death resulting from pulmonary infections or cardiac failure due to pulmonary hypertension from hypoxemia.

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Splenic Autotransplantation

Splenic autotransplantation (SAT) as a means of preserving some immunologic function after emergent (trauma), or elective (Schistosomiasis and Gaucher's disease) splenectomy in children has been reported in several studies. Thinly slice splenic segments (around 20

to 30 gm tissue) are deposited into a greater omentum pouch. Studies have shown that these implants grow with time, taking around five years. More than 30% of normal splenic tissue is needed for adequate filtration and immunologic clearance. After implantation Technetium scanning is use to show viability of the graft. Fc-receptor scintigram with IgG coated and Tc-labeled RBC are use to verify mononuclear phagocyte function. Filtration capacity is follow-up with disappearance of Howell-Jolly bodies and platelet numbers. Phagocytosis and Immunologic capacity is done measuring tuftsin and IgM respectively, and NBT test for evaluating phagocytic function of granulocytes. Complications associated with SAT are adhesive small bowel obstruction, torsion and aseptic necrosis of the transplant. A better antibiotic response to pneumococcal vaccine is found in patients after splenectomy and autotransplantation. SAT is simple and appears to preserve splenic function.

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Laparoscopic Varicocelectomy

Varicoceles develop during early adolescence. Varicocelectomy represents the treatment of choice for scrotal varicocele associated with scrotal pain or discomfort, testicular atrophy or infertility (low sperm count). Prepubertal varicocelectomy is controversial. Traditionally, the surgical procedure entailed an inguinal or high retroperitoneal open approach to ligate the internal spermatic veins associated significant postop morbidity (recurrence and postop hydrocele formation) and prolonged return to normal activity. During the past ten years the technique has take advantage of the laparoscopic approach including minimal surgical trauma, lower morbidity, cost and time sparing, faster recovery, better microscopic dissection with preservation of the spermatic artery along amenable bilateral ligation without a second incision. The Palomo mass high retroperitoneal ligation of the internal spermatic vessels results in a significant decrease in failure rate as compared with artery-sparing procedures. The Palomo approach is also safe after previous inguinal hernia repair. The procedure entails mass clipping and division of the spermatic veins. Collateral veins should be cauterized. Scrotal subdermal injection of methylene blue help contrast delineates and preserve lymphatics vessels to reduce the incidence of postop hydrocele formation.

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