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Split Cord Malformations

Split cord malformations (SCM) refers to a group of malformations where the spinal cord is split or clefted over a portion of its length. Most affected children are females with a mean age of seven years. The malformations have been recognized into two types. Type I consist of double dural sac malformations (both spinal cord and dural sac are split) associated with an extradural bony cartilaginous spur interposed between the two thecal sacs. Type II are single dural sac malformations with a clefted spinal cord (both hemicord are contained within a common dural sac). Both types contain distal aberrant nerve roots that exit from the cord and can tethered the spinal cord. Tethering the cord can produce neurological deterioration such as sensorimotor deficit, change in bowel or bladder habit and orthopedic deformities. Associated enteric malformations (posterior mediastinal foregut cyst) are frequently found with SCM. The enteric cyst can contain stomach, small or large bowel and even bronchial tissue. The child born with a SCM usually has a cutaneous stigmata (focal hypertrichosis) under the malformation. Evaluation of SCM includes MRI with intravenous contrast and/or CT-Scan myelography. Since neurological deficit increases with age, prophylactic repair of the SCM with untethering of the spinal cord is recommended before neurological signs develop.

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Segmental Bowel Dilatation

Dilatation of a segment of bowel in newborns and infants is a rare congenital malformation associated with partial or complete bowel obstruction. The dilatation is limited to a segment of the small or large intestine with a three to fourfold increase size and abrupt transition between normal and dilated bowel. Histology demonstrates that the neuronal enteric plexus

is normal in the affected segment, proximally and distally. The origin of the dilatation is unknown. Theory states intrauterine extrinsic compression of the bowel or a primitive neuromuscular dysfunction can explain the dilatation. Most cases are found in newborn explore for another surgical malformation. Other will present a bowel complication such as obstruction, perforation or bleeding. Bleeding due to ulceration of the dilated segment is found more commonly in infants. Most frequent associated malformation are omphalocele, bowel atresias and imperforate anus. Contrast studies of the distal and proximal bowel will uncover the segmental dilatation. The first diagnostic possibility to consider is aganglionosis (Hirschsprung's disease) and a suction rectal biopsy is in order. If the symptomatic child has normal rectal ganglion cells, the next step in management consists of surgical exploration with histological confirmation. Most segmental dilatation affects the ileum. Resection of the affected segment with end to end anastomosis is curative.

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Sialoblastoma

Sialoblastoma, originally called embryoma, is the most common type of congenital epithelial tumor of the salivary gland in children. Is an extremely rare salivary gland neoplasm encountered after the age of five. Sialoblastoma is locally aggressive with a high recurrence rate. The parotid gland is most commonly affected. Diagnosis can be suggested by fine needle aspiration biopsy. Prenatal diagnosis has been reported. The tumor is characterized by solid nests of epithelial cells intermingled with proliferating ductal structures lined by a double layer of cells. Sialoblastomas are mitotically active primitive cell masses with formative ducts and pseudoductular spaces without acinar differentiation. MRI can help establish the surrounding anatomy and relationship of the tumor to other vital structures. Sialoblastomas should be managed with early conservative surgery provided that free margins are obtained. The patient's prognosis is likely to be determined by the tumor grade as well as the stage at presentation and the extent of resection. Adjuvant chemotherapy or radiotherapy has not been effective in control of recurrence.

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