PEDIATRIC SURGERY *Update**
Vol. 55 No. 02 AUGUST 2020

Thoracoscopic Repair Diaphragmatic Eventration

Diaphragmatic eventration occurs due to a congenital structural defect in the diaphragm or after injury to the phrenic nerve. Phrenic nerve in jury occurs due to traction on the nerve during birth or directly after an open cardiac procedure in the child. When the child develops diaphragmatic eventration, he can develop mild gastrointestinal symptoms up to life-threatening respiratory distress requiring mechanical ventilatory support. In infants, the diaphragmatic eventration causes progressive dyspnea on exertion or respiratory infection. When the child develops symptoms (dyspnea, shortness of breath, labored respiration, chest retraction) or need of mechanical ventilation plication repair of the diaphragm is needed. Diaphragmatic plication is usually performed using a standard posterolateral open thoracotomy through the 5th or 6th intercostal space. During the past years thoracoscopy has been utilized to accomplish plication of the diaphragm. The technique uses general anesthesia with single lung ventilation most commonly. Three to four trocars are placed depending on the need to reduce the elevated diaphragm. Carbon dioxide is insufflated in low pressure levels (4-6 mm Hg) to reduce the risk of developing hypercapnia, hemodynamic instability, acidosis and hypoxia. The single lung ventilation can be accomplished using a Fogarty catheter to block the main bronchus ipsilateral to the side of the eventration. Plication is the accomplished posterolateral to anteromedial with interrupted nonabsorbable sutures to avoid phrenic nerve injury and suture breakdown respectively. A chest tube drainage is utilized for a short period of time until the pneumothorax is evacuated. Thoracoscopy offers the advantage of smaller wounds, better cosmetic results, faster recovery, less thoracic pain, less incidence of late thoracotomy sequela (scoliosis), immediate pulmonary function improvement with less impaired ventilatory function postoperatively. Plication can be performed using a double purse-string technique. Plicating the diaphragm using a thoracoscopic approach is feasible, safe, easy to perform and efficient. Early thoracoscopic plication is a good treatment option for pediatric patients with symptomatic diaphragmatic eventration after surgery for congenital heart disease.

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Intestinal Pseudo-obstruction

Pediatric intestinal pseudo-obstruction (PIPO) is a rare disorder characterized by the chronic inability of the bowel to propel its content in the absence of any mechanical lesion occluding the gut. Diagnosis of PIPO needs at least two parameters: objective measure of small intestinal neuromuscular involvement (manometry, histopathology, transit), recurrent or persistently dilated bowel loops, associated genetic or metabolic abnormalities, and inability to maintain nutrition or growth with oral feedings. PIPO patients develop bile vomiting, failure to pass gas and stool, and progressive abdominal distension. PIPO could be associated with bladder dysmotility or after a Ladd's procedure for malrotation. Hirschsprung's disease and hypothyroidism should be excluded. Megacystis is a prenatal sign of PIPO. Most cases present signs within the first month of life. Immaturity of the intestinal motility in premature infants can mimic PIPO. Late-onset or infant PIPO presents with recurrent and intermittent episodes of gastric, intestinal or colonic obstruction and is triggered by infections, fever, general anesthesia or emotional stress. Associated abdominal pain could lead to feeding difficulties and malnutrition. Megacystis with hypocontractile detrusor, increase bladder capacity and compliance occurs in more than 50% of PIPO cases. This usually associated with uretero-hydronephrosis and minimal vesico-ureteral reflux. Abdominal films show bowel dilatation with air-fluid levels. Entero CT-Scan, MRI and/or small bowel follow-through studies using water soluble contrast is needed to exclude mechanical obstruction. The most accurate and sensitive measure of GI transit is obtained with nuclear studies such as gastric emptying scans. Antroduodenal manometry studies are indicated for diagnosis and classification of the pseudo-obstruction present. Esophageal, colonic and anorectal manometry is used to determine extend of disease. Unnecessary surgery should be avoided as these patients develop dense adhesions. Should surgery be performed full-thickness biopsy of the affected bowel for nerve, muscle and interstitial cell of Cajal should be performed. Laparoscopic biopsy are indicated for diagnostic purpose obtaining a minimum tissue specimen of 0.5 x 0.5 cm. Full work-up include labs tests, genetic testing, endoscopy, neurologic evaluation and imaging. Nutrition should be optimized. Drug therapy aims to promote GI motility, limit intestinal inflammation and suppress bacterial overgrowth. Erythromycin, pyridostigmine and octreotide are effective drug therapy in some children. Venting or feeding gastrostomy and/or jejunostomy should be considered in patients with PIPO, and decompressive ileostomy in those on parenteral nutrition. Bowel resection should be avoided to avoid short bowel syndrome or liver failure. Definitive cure is bowel transplantation. Stomal prolapse, recurrent pancreatitis, diversion colitis, and electrolytes imbalance are the most common complications.

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Diffuse Hyperplastic Perilobar Nephroblastomatosis

Nephroblastomatosis refers to a premalignant condition associated with Wilms tumor characterized by multiple residual embryonal cells known as nephrogenic rests. It is considered an intermediate preneoplastic stage in the sequence of Wilms tumorigenesis. Nephrogenic rests are clusters of embryonic metanephric that can be found incidentally in less than 1% of infants. They can be single or multiple, focal or diffuse lesions identified in the renal parenchyma. Panlobar nephroblastomatosis denotes complete replacement of the renal lobe by nephrogenic tissue. The fate of nephrogenic rests and nephroblastomatosis varies and includes obsolescence, sclerosis, dormancy, hyperplasia, or neoplasia. Evidence strongly suggests that neoplastic transformation of nephrogenic rests results in Wilms' tumor (nephroblastoma). Depending on where in the renal parenchyma they are located they are either classified as perilobar or intralobar. Perilobar rests show a strong association with synchronous bilateral Wilms' tumors, whereas intralobar rests are more strongly associated with metachronous tumors. Perilobar nephrogenic rests are found in the renal peripheral cortex associated with fetal overgrowth disorders. Intralobar rests occur as focal lesion inside the central renal parenchyma. Diffuse hyperplastic perilobar nephroblastomatosis (DHPLNB) is associated with an increased risk of developing into a malignant nephroblastoma. DHPLNB can be seen in US or CT Scan as enlarged diffusely hypoechogenic kidneys or enhancing peripheral nodularity with scattered patches of adjacent normal renal parenchyma respectively. MRI demonstrates peripheral nodules with low signal intensity on T1 and T2 images. When DHPLNB is associated with Wilms tumor management is multimodal including surgery, chemotherapy and radiotherapy. Poor prognostic factors include an anaplastic tumor, high stage, unfavorable molecular and genetic marker and age greater than two years of age. The diagnosis of a Wilms tumor should be favored over a nephrogenic rest when a renal mass is spherical, exophytic, or larger than 1.75 cm. Most DHPLNB cases occur with bilateral renal involvement and as such management strategies should consider nephron-sparing procedures to avoid leaving the child anephric hence renal insufficiency, dialysis dependent and in need for renal transplantation. In such cases 18 weeks of vincristine and actinomycin D should be used as chemotherapy for an extended period until nephrogenic rest disappear. In case of bilateral DHPLNB laparoscopic nephro-sparing resection can be performed so long as the capsule

of the nephrogenic rest stays intact and there is no spillage of the lesion as they upstage the tumor.

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**** PSU 1993-2020***

ISSN 1089-7739