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Esophageal Atresia and Tracheo-Esophageal Fistula

Esophageal atresia (EA) with distal tracheo-esophageal fistula (TEF) is the most common congenital anomaly of the esophagus, followed by EA without TEF also known as pure esophageal atresia and pure TEF. Incidence is one in every 2500 live births. Polyhydramnios is most commonly seen in pure EA. EA causes excessive salivation, choking, coughing, regurgitation with first feed and inability to pass a feeding tube into the stomach. Contrast studies are rarely needed and of potential disaster (aspiration). Correct dehydration, acid-base disturbances, respiratory distress and decompress proximal esophageal pouch (Reploge tube). Evaluate for associated conditions such as VACTERL association. Early surgical repair (transpleural or extrapleural) is undertaken for those babies with adequate arterial blood gases, adequate weight (>1200 gm) and no significant associated anomalies. Delayed repair (gastrostomy first) for all other patients. Repair consists of muscle-sparing thoracotomy, closure of TEF and primary anastomosis. Esophagogram is done 7-10 days after repair. Most important predictors of outcome: birth weight, severity of pulmonary dysfuntion, and presence of major congenital cardiac disease. Complications after surgery: anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia and recurrent TEF. Increase survival is associated with improvements in perioperative care, meticulous surgical technique and aggressive treatment of associated anomalies.

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Insulinoma

Insulinoma, a rare islet beta-cell adenoma, is the most common pancreatic endocrine tumor in children. The vast majority are sporadic in nature (90%) consisting of a benign solitary small intra-pancreatic nodule. The rest of the time insulinomas are associated to familial syndromes (MEN type 1) with multiple localization within the gland. Insulinomas cause hypoglycemia, diaphoresis, anxiety, dizziness, seizures and even coma. Aggressive

management is mandatory to avoid permanent sequelae. Whipple's triad: symptoms of hypoglycemia while fasting, serum glucose level less than 40 mg% and resolution of the symptoms after sugar administration suggest the diagnosis. Measurement of insulin level while fasting produced by a plasma insulin to glucose ratio greater than 1.0 is diagnostic. Preop localization of the tumor is generally difficult to achieve (CT-Scan, selective arteriography and MRI) and believed to be neither indicated nor cost-effective. At surgery the tumor is pink, firm, discrete and well-encapsulated being amenable to enucleation. Intra-operative US localization or palpation of the tumor is essential to resect the tumor or perform a distal pancreatectomy. Once the tumor is eradicated, the child is cured and the prognosis is excellent. Familial syndromes may need 95% gland resection for cure (multiple microadenomatosis).

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Peutz-Jeghers Syndrome

Peutz-Jeghers represent the association of benign hamartomatous polyps of the gastrointestinal tract with abnormal pigmentation of the mouth and skin transmitted as a familial autosomal dominant syndrome. Mucocutaneous melanin deposits include small blacks or brown dark spots around the lips, buccal mucosa, fingers and toes usually identified during infancy. Polyps are large, pedunculated with high malignant potential (48%) later in adult life (cancer development is 18 times greater than general population). Clinical presentation may include recurrent abdominal colicky pain, gastrointestinal bleeding, obstruction, rectal prolapse, anemia and intussusception. Most polyps affect the jejunum and are multiple, although they are also found in the stomach, small bowel, and colon. Management is dictated by symptoms. A cautious approach is advised to preserve as much bowel length as possible. If surgical intervention is necessary, intraoperative endoscopy with polypectomy may prevent the development of short bowel syndrome.

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