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Congenital Esophageal Stenosis

Esophageal stenosis in children can be of congenital (5%) or most commonly acquired nature (95%). Acquired stenosis is the result of repaired esophageal atresia, caustic injury, penetrating injury or reflux esophagitis. Congenital esophageal stenosis (CES) can be the result of a membranous diaphragm, segmental hypertrophy of the muscularis and submucosal layer (submucosal fibrosis), or presence of ectopic tracheobronchial rest. CES most commonly affect the middle and distal third of the esophagus and rarely cause symptoms in the neonatal period. Symptoms can be vomiting of undigested food, regurgitation, food impaction, difficulty swallowing solid and failure to thrive. CES affecting the upper third of the esophagus is very rare and usually produce respiratory symptoms such as stridor and repeated respiratory infections. Esophageal atresia is associated with one-third of cases of CES. To establish a diagnosis investigation has to include esophagogram (relatively long, smooth circumferential narrowing), esophagoscopy with biopsy, pH monitoring and in selected cases manometry. Recognition of the correct etiologic factor that caused the stricture will pave the way for adequate management. CES is managed with forceful dilatation or hydrostatic balloon dilatation, while resection with anastomosis will be needed for intractable (fibromuscular hypertrophy) cases and those harboring tracheobronchial rests. Most intractable cases are due to the presence of tracheobronchial rest.

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Limb Body Wall Complex

Limb-body wall complex (LBWC) is a rare, lethal, sporadic congenital anomaly pattern in body-wall defects. Diagnosis of this entity is based on finding at least two of the three following characteristics: (1) neural tube defect (exencephaly/encephalocele and facial clefts), (2) body-wall disruption (thoraco- and/or abdominoschisis, diaphragmatic hernia, eventration, cloacal exstrophy, absent external genitalia or abnormal internal genitalia) and, (3) limb abnormalities (ectrodactyly, phocomelia). Three pathogenic mechanisms have been proposed: early amnion rupture, vascular disruption and embryonic dysgenesis. The internal defects are secondarily to vascular disruption. Cocaine abuse has been associated with LBWC by impairing uteroplacental fetal blood flow during critical periods of development. The limb defects are due to mechanical rupture through the amnion in the presence of a persistent extraembryonic coelom or from adhesions of the amnion to necrotic embryonic tissue. Diagnosis of LBWC is established with prenatal sonography. LBWC is usually incompatible with life.

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Ingrown Toe Nails

Onychocryptosis is the medical term use to describe ingrown toe nails (ITN). Congenital hyperplasia of the nail bed lead to this condition in infants and children. Adolescent and athletes are susceptible to develop ITN. Other conditions associate with ingrown nails are hallux valgus, claw toes, gout, diabetes, arthritis, or fungus infections of the nails. Initial management should be conservative inserting cotton wool pledgets moistened with an antiseptic under the ingrowing nail edge along with oral antibiotics if there are signs of infection. Insertion of the pledget under anesthesia improves the results and chronicity does not adversely affect the outcome though the procedure is time consuming. Other alternatives are segmental total nail avulsion, nail edge excision, nail matrix phenol cauterization or wedge resections. Total nail avulsion and nail edge excision carries the highest recurrence rate. Nail matrix phenolization is an effective short procedure that can be done under the presence of an infection with a low recurrence rate. Radical wedge resection is simple, has a low recurrence rate, leaves patients with an intact, pain-free, cosmetically acceptable nail-bearing toe, and permits the wearing of normal shoes within a short period. In children conservative treatment is more promising than in adults. If unsuccessful the next reliable surgical approach is wedge resection. Attentions to hygiene

and always cutting nails transversely are important preventing factors.

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