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63.1 Tongue-Tie (Ankyloglossia)

This may be defined as a congenital anomaly in which a short lingual frenulum or a tight genioglossus muscle restricts tongue movement. It may interfere with breastfeeding and tooth development and (disputed) speech development.

- Male > female (2:1)
- Usually *sporadic*, but some genetic link has been mentioned in the literature (X-linked mutation of T box transcription factor TBX22) with or without the association of cleft lip/palate.
- *Posterior tongue-tie*
 - Thickened submucosal base of perhaps normal-looking frenulum from posterior to middle part of the tongue. In some cases, it can restrict the tongue movement.
- *Upper lip tie*
 - Rarely a cause of breastfeeding problems from ineffective latching due to inadequate eversion of the upper lip and release of these upper lip tie may be helpful in these cases.

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63.1.1 Clinical Features

Tongue-tie is mostly asymptomatic but the best-defined consequence is difficulty with breastfeeding causing poor latch, insufficient and pronged feeding, and maternal nipple pain. It may lead to failure to thrive. Most infants with tongue-tie actually can breastfeed without difficulty.

Articulation problem of speech due to restricted tongue movement has been seen in some toddlers and young children. Speech sounds that may be affected include “t,” “d,” “z,” “s,” “th,” “n,” “l” (sibilants and lingual sounds). Involvement of a speech therapist is also important in the management process along with the release of tongue-tie. An actual delay in acquisition of speech is not caused by tongue-tie and other possible diagnoses should be sought.

Protrusion of the tongue causes its tip to become notched and not extend past the incisors or touch the roof of the mouth. In severe cases, the tongue is completely immobile and fixed.

Difficulty to maintain oral hygiene may cause periodontal disease due to lack of sweeping food debris from the teeth, and licking of lips.

Cosmetic and social problems like difficulty in kissing, playing a wind instrument, or licking an ice cream in public could be an embarrassing problem for these patients.

63.1.2 Management

The natural history of tongue-tie is unknown. Spontaneous elongation or natural release with time is postulated.

Assessment and management by breastfeeding counsellor/midwife are imperative before surgical referral. Frenotomy can be considered (without anesthetic) as curative by surgeon or the aforementioned. After infancy, it is judicious to wait for a reason (e.g., speech therapy defined problem) rather than preempt matters.

Division of frenulum is straightforward but should avoid the submandibular ducts on either side.

63.2 Umbilical Issues

The umbilicus is the last link with fetal life and is the site of many peculiar embryonic vestiges.

63.2.1 Embryology and Anatomy

Vascular connection to the placenta is maintained by a single large umbilical vein (to the porta hepatis) at 12 o'clock and two inferior umbilical arteries from the internal

iliac arteries at 5 and 7 o'clock. At birth, these structures close and obliterate but leave a potential space through the umbilical ring, which also has to close by a cicatrizing process. Failure to achieve this leads to the development of *umbilical herniation*.

Two further structures may lead to vestigial-related problems:

- *Vitello-intestinal duct*
 - Communication from the apex of the midgut to the yolk sac of embryonic life. It should vanish, but remnants may include a *Meckel's diverticulum*, a fibrous connection to the underside of umbilicus, or a completely patent duct.
- *Urachus*
 - Connection between the apex of bladder and allantois.¹

63.2.2 Umbilical Hernia

- Prevalence is greatly increased in Afro-Caribbean ethnicity (particularly of West African origin) than those of Caucasian/Asian background.
- Low birth weight.
- M = F.
- Spontaneous closure is less likely if the defect is >1.5 cm.
- 13% (vs. 2%) at 1 year of age (USA study).

63.2.2.1 Associations

- Congenital hypothyroidism (defective cicatrization)
 - May be associated with constipation
- ↑ Intra-abdominal fluid
 - For example, ascites and VP shunts
- Beckwith-Wiedemann syndrome, mucopolysaccharidoses
- Trisomy 21, Trisomy 18

63.2.2.2 Clinical Features

These are mostly asymptomatic but in very rare cases it can cause feeding problems in young infants and seems to be related to the intestinal protrusion through the hernial ring.

Umbilical herniation is unique among hernias in that it may spontaneously reduce and close over time. There is a low risk (<3%) of irreducibility and intestinal obstruction. Incarceration if happens is more common in smaller defects.

63.2.2.3 Management

As most hernias will close spontaneously, surgical closure should be considered in:

- Large defects (usually above 1.5 cm) at age 3–4 years.

¹Allantois (Greek—sausage)—is a diverticulum of cloaca within connecting stalk to placenta—main function in reptiles and birds (both lay eggs) is as a repository of nitrogenous waste.

- No decrease in the size of the defect during a year of observation.
- Children who develop symptoms.

Large trunk-like hernias without any perceptible decrease in defect size are unlikely to close spontaneously and usually need operation.

63.2.2.4 Surgery

Sub-umbilical skin crease incision:

- Reduce contents and then dissect around the whole circumference of the neck of sac.
- Separate overlying skin from sac and then excise this back to the fascial ring.
- Repair defect (usually transverse), typically with absorbable suture.
- Umbilicoplasty.
 - Mostly the skin can be tacked back to the repair and the cavity obliterated by pressure. Sometimes, so much skin has left that excision of a triangle and “swirling” the subsequent suture line is a better form of cosmesis.

63.2.3 Umbilical swellings

- *Umbilical granuloma*
 - Commonest umbilical swelling in neonates presenting as a pink, moist swelling, usually pedunculated in nature. The size varies from 0.3 to 1 cm. It can be treated with 75% silver nitrate application (once or twice weekly for few weeks).
 - Ligation of base with a suture until it falls off. Before ligation, the possibility of umbilical polyp should be ruled out.
- *Umbilical polyp*
 - Are usually larger and less common than umbilical granulomas. It presents as a relatively firmer swelling. It comprises intestinal epithelium or uroepithelium. The treatment is excision.
- *Urachal remnants*
 - Involution is the norm but may remain as a fibrous cord between the umbilicus and dome of the urinary bladder. A persistent urachus can be complete causing urinary discharge from the umbilicus or incomplete persistence can cause cystic swelling at the umbilicus, bladder, or in the mid-duct.

Symptoms may include umbilical swelling, discharge, abdomen pain, infection, or incidental diagnosis in a US scan. US scan is usually diagnostic, and also rules out any associated renal tract problem. Treatment should be surgical excision supplemented by diagnostic. If not removed (as in asymptomatic cases), parents should be made aware of the potential (albeit low) risk of malignancy in the future.

63.3 Rectal Prolapse

Of the falling down of the Fundament
Ambroise Pare (1634)

May be defined as a protrusion of all or some of the rectal wall through the anus, though there are more complicated classifications that include internal or occult prolapse.

Types

- Type 1: Incomplete/partial/mucosal prolapse
 - Radial mucosal folds
- Type 2: Full-thickness rectal prolapse/procidentia²
 - Circumferential concentric mucosal folds

Epidemiology

- Common during infancy
- ↑ Incidence in tropical latitudes

Associations

- Increased abdominal pressure and straining
 - Chronic constipation is the usual precipitating factor.
 - Protracted coughing, excessive vomiting, toilet training.
- Acute diarrheal illness
- Cystic fibrosis
 - ~3% of children with CF, and occasionally its presenting feature. Consider CF screening for recurrent prolapse with failure to thrive.
- Pelvic floor neuromuscular weakness
 - Spina bifida, sacral agenesis
- Malnutrition
- Ehlers–Danlos syndrome

63.3.1 Clinical Features

Usually nontender, red mucosal mass protruding from the anus, initially only on straining. It may become irreducible and present all the time.

² *Procidentia ani*—(Latin) *prōcidere*—to fall forward.

63.3.2 Differential

- Intussusception—much more severe symptoms, \pm intestinal obstruction. Finger or probe can freely be passed around all circumferences between anus and intussusceptum.
- Prolapsed rectal polyp.

63.3.3 Investigations

- Exclude underlying conditions, which may be overt (spina bifida) or covert (cystic fibrosis).
- If diarrhea—stool microscopy, culture and sensitivities, and screening for ova, cysts, and parasites.
- Lower GI endoscopy—to exclude rectal polyps. There is usually little evidence of anything abnormal in prolapse.

63.3.4 Management

- Manual reduction with adequate sedation. Firm compression to reduce edema, then reduction of innermost mucosal lead-point first. Application of sucrose/sugar in very large and edematous prolapse decrease swelling and thus helps in reduction.
- Thereafter, most would advocate a simple conservative policy of treating the underlying condition (e.g., constipation/diarrhea) and avoidance of straining. Most children (<4 years) recover spontaneously.

Intervention is more likely to be needed in older children and those with neuromuscular etiology.

63.3.5 Surgery

- *Sclerotherapy*
 - For example, oily (5%) phenol, hypertonic saline, dextrose (50%) solution. May need multiple injections. Avoid anterior needle placement (prostate or vagina at risk). Success rate up to 85%.
- *Thiersch³ operation*
 - –Circumferential (typically absorbable) suture.
- *Rectopexy*: Although many of them are described, e.g., (laparoscopic suture rectopexy, posterior sagittal rectopexy, prosthetic/mesh fixation) these are rarely performed in children and are usually reserved for complicated or recurrent prolapse in elderly women.

³Karl Thiersch (1822–1895)—German surgeon who also pioneered use of split-skin grafts.

Further Reading

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