STERNOMASTOID `TUMOUR' (FIBRO-MATOSIS COLLII) AND TORTICOLLIS

Clinical evidence
This may present in three different ways depending upon the age of the patient.
- In the neonate 2 - 3 weeks old a firm swelling is noticed in the neck which is localized to the sternomastoid muscle. Usually detected because of torticollis - `wry neck'. Occasionally presents with facial and cranial asymmetry.
- In the older infant there may be no history of a swelling but the sternomastoid muscle is now firm and foreshortened. `Wry neck'
- The older child may present with torticollis. The face is rotated away from the affected side and the head tilted towards the affected side. This is due to the shortening and fibrosis in the muscle with facial and cranial asymmetry are usually apparent.

Pathology
The cause is unknown. However, it may well be present in utero thus causing obstetric problems. There is a 20% incidence of abnormal presentation. Histological changes are the same at all ages; a typical endomycial fibrosis around individual muscle fibres which then undergo atrophy and fibrous replacement. The effect of this is to shorten the sternomastoid muscle, which over a period of time results in facial asymmetry and plagiocephaly.

Other causes of torticollis should be excluded: localized pathology in the neck which may be inflammatory or neoplastic causing tilting of the head, ophthalmodologic conditions such as squints and nystagmus, and skeletal abnormalities of the cervical spine.

Management
A combination of active and postural physiotherapy is successful in the vast majority of cases. Active: the child's head is taken gently through a full range of movement (chin to shoulder, ear to shoulder on each side and extension/flexion.) Postural: positioning such that the child is encouraged to turn the head towards the affected side e.g. cot position, car seat, etc. Facial asymmetry cannot be expected to correct itself after the age of 5 years. In those cases which present late or where no progress is obtained, the muscle is surgically divided. In the older child the investing deep cervical fascia on that side is also divided. Less than 5% of all cases diagnosed in infancy require surgery. It is important to continue with physiotherapy after surgery.

THYROGLOSSAL CYST AND FISTULA

A common problem. Most appear before 5 years of age.

Clinical evidence: The cyst usually lies in the midline below the hyoid bone. It is a discreet, firm cystic structure. It may become inflamed and subsequent discharge or drainage will lead to a persistent fistula. The diagnosis is suggested when, with the mouth opened thus fixing the platysma and mandible, the tongue is protruded and the cyst moves upwards.

Embryology: At the 4th week the tongue develops from several growth buds in the floor of the primitive pharynx. Just caudal to this is the foramen caecum from which the thyroid diverticulum develops. During the 7th week this diverticulum descends to the front of the neck and remains connected to the foramen caecum by the thyroglossal duct. The
Branchial arches are developing at the same time, the ventral 2nd and 3rd arch cartilages forming the hyoid bone. There is thus an intimate relationship between the thyroglossal duct and the hyoid bone. The cyst may be found anywhere from the thyroid to the base of the tongue, the cyst arising from the thyroglossal duct. Although the cyst is usually connected by a single primary duct structure there may be several small accessory ducts running parallel particularly above the hyoid bone. The duct is lined with stratified squamous or ciliated columnar epithelium with associated mucus secretion. There may be an incidence of ectopic thyroid tissue, usually dysplastic, in relation to the duct or cyst.

The treatment is to remove the cyst or sinus and the thyroglossal duct in its entirety up to the base of the tongue. This will include the mid-third of the hyoid bone (Sistrunk's operation). If only the cyst is removed morbidity is significant with persistence of secretion local recurrence (30%) and sinus formation. Recurrence is also attributed to failure of removal of inferior extension of the tract; or in the supra-hyoid area multiple fistulous tracts.

**EPIDERMOID / DERMOID CYST**

These cystic structures occur in lines of fusion. Ectodermal elements become buried beneath the skin surface leading to the formation of cysts. Sebaceous glands, hair follicles and connective tissue may be present in the cyst wall giving rise to the typical sebaceous material. There may be a deeper penetration through underlying bone giving rise to a `dumb-bell' configuration. These inclusion cysts are differentiated from the so-called dermoids that occur in ovaries and testes which are in fact teratomas.

**Common sites:** Epidermoid cysts - midline supraclavicular area of the neck and on the scalp over the anterior fontanelle. Dermoids over the eyebrow (external angular dermoid) very uncommonly have a deeper extension. Those on the scalp also only rarely have a deep extension but may be mistaken for other pathology, e.g. haemangiomas, hamartomas and lipomas.

**Treatment:** Excision. Pre-op x-ray and ultrasound to exclude deep extension. Dermal sinuses and posterior cysts nearly always have a deep extension into the CNS, and require pre-op CT scan.

**BRANCHIAL FISTULAE, CYSTS AND CARTILAGINOUS REMNANTS**

These occur in the head and neck and result from embryonic structures that have failed to completely disappear after their temporary foetal presence in early embryonic life. To understand these lesions the embryology of the head and neck should be studied. (See Chapter: The Anatomy of the Abnormal, Part II. A synopsis of Surgical Anatomy - Lee McGregor, DJ du Plessis; or The Developing Human, Clinically Orientated Embryology - KL Moore.).

**Sinuses or fistulae related to second arch.** These are the commonest branchial remnants. Fistulae discharging mucus are present from birth and are evident along the anterior border of the sternomastoid. There is a 15% incidence of bilaterality. They may also occur in families. The lining of the tract on the inner two-thirds is columnar epithelium and the outer one-third is squamous epithelium. The course of this tract can be predicted because it arises from the cervical sinus where the second branchial arch overgrows the third and fourth. The track is related above to the IXth nerve, the stylopharyngeus muscle extending into the tonsillar fossa, and more inferiorly it traverses the fork of the carotid bifurcation, the XIith nerve, internal jugular vein and goes beneath
the posterior belly of digastric and stylohyoid to finally open on the skin along the lower part of the anterior medial border of the sternomastoid.

**Sinuses or fistulae related to the first arch** are uncommon and classically present inferiorly as an opening on the skin below the ramus of the mandible, with a tract extending upwards to the junction of the bony and cartilaginous part of the external auditory canal. The track is intimately related to the facial nerve, the branches of which may have a variable relationship to the sinus tract.

**Pre-auricular sinuses** are probably ectodermal inclusions related to the development of the auricle, which forms from 6 cartilaginous tubercles. **Cartilaginous remnants** usually arise along the anterior border of the sternomastoid and extend into the fascia of the muscle. These also commonly have a family history and are bilateral. Surgery is only advised if symptomatic.

**Branchial cysts** tend to present in later childhood or early adult life and are again related to the anterior border of sternomastoid, usually lying deep to the muscle. They are typically described as feeling like a "half-filled hot-water bottle" with a thick wall and contain a yellow creamy substance with cholesterol crystals. They are usually unilateral. Branchial cysts related to the first arch may occur within the parotid gland - often misdiagnosed as a pleomorphic adenoma.

**Treatment** of all these is careful excision, being aware of the related structures. Surgery of branchial fistula is usually conducted before the third year of life as the neck is shorter and the whole fistulous track can be removed through a single incision. In the older child two separate "step ladder" incisions may become necessary.

**HAEMANGIOMAS / VASCULAR MALFORMATIONS**

Although grouped under a single term, haemangiomas encompass a wide range of pathology. Histological classification is based on the size of the vascular structure in the lesion: capillary, mixed, cavernous, A-V fistulae. Clinically they can be divided into two groups. Haemangiomas and malformations.

**Haemangiomas** usually occur in females, are visible at birth in only ±40%, but subsequently undergo tremendous **endothelial proliferation** with rapid increase in size. Histology is mainly capillary. This group can be expected to subsequently **involute**. The commonest of this variety is termed a **strawberry haemangioma** (capillary histology). Treatment is thus expectant and involution should be complete by about 5 years of age. In those areas where the lesion is constantly traumatized, causing infection and bleeding, e.g. the scalp and perineal area, or where its site affects vital functions e.g. over the eye where complete closure may rapidly result in amblyopia, excision may be attempted.

**The stork bite or neonatal staining** may appear shortly after birth and is classically related either to the nape of the neck, over the forehead or sacrum. It can be expected to spontaneously fade over several months.

The second group can be termed **malformations** in that the lesions are present to their total extent at birth and will not fade but will grow at the same rate as the child. These show normal endothelial mitotic activity. The **port wine stain** consists of a malformation located within the dermis - histologically it is a capillary vascular malformation. These lesions correspond to the skin distribution of various sensory nerve branches, e.g. on the face the branches of the trigeminal nerve. Those more deeply
situated in the dermis tend to have a darker colour and in time become hyperkeratotic with the development often of troublesome eczema. The salmon patch is fainter in colour and blanches on pressure but is not elevated above the skin.

The cavernous haemangioma can also be regarded as a malformation and appears as an isolated, puffy, spongy, soft swelling with a blue colour. These may also occur deep to the subcutaneous tissue and may extend into muscle. They consist of mainly large venous spaces. Involution tends not to occur.

A combination of both groups occurs where there is some degree of capillary proliferation but usually the major element is cavernous. These malformations may be more extensive with large fields of greatly increased purpura and arterio-venous shunting. Some involution may be expected to occur. The local increased blood supply may lead to regional gigantism, varicose veins, and systemic complications such as cardiac failure and consumption coagulopathy, with most commonly a thrombocytopaenia.

Congenital arteriovenous fistulae are multiple between small arteries and veins. Rarely evident at birth, but become apparent in childhood, enlarging at irregular rates with elevation of skin temperature, red colour if superficial and progressive enlargement of the involved area. Arteriography reveals multiple AV shunts. These tend to be refractory to treatment. Embolization of feeding arteries may help temporarily and amputation is a last resort.

Haemangiomas in Internal Organs
Haemangiomas may occur anywhere in the body including the gastro-intestinal tract and other abdominal viscera, e.g. the liver, leading to massive hepatomegaly, portal hypertension and heart failure; in the lung resulting in haemoptysis or recurrent pulmonary infections; and in the gastro-intestinal tract, with bleeding.

Complications of Haemangiomias
A. Local: Disfigurement, infection, pain, ulceration, bleeding, recurrent trauma, regional increase in growth, e.g. limb varicosities, pressure effects, airway compromise.
B. Systemic: Cardiac failure, consumption coagulopathy.

Summary of treatment
1. Observation.
2. Await involution in those cases of the proliferative group or where there is a proliferative component.
3. High dose short course steroids in those larger proliferative lesions with complications (4-8 mg/kg/day Prednisone).
4. Interferon may assist involution.
5. Intermittent compression if feasible.
6. Surgical excision or local steroid (triamcinolone) injection for those small malformations causing local complications.
7. Embolisation, in some instances. Difficult because often multiple feeding vessels.
8. For extensive localized lesions causing life-threatening complications - surgical excision with hypothermia and cardiac standstill or bypass.
9. Radiotherapy is contra-indicated.
10. Pulsed flash lamp laser therapy of port wine stains has had good results. Treatment of capillary haemangioma with laser has been less rewarding but worth attempting in young infants with prominent lesions.

Where consumption coagulopathy and thrombocytopaenia is a primary problem, low dose aspirin and persantin has success when used in some cases.
**LYMPHANGIOMA / CYSTIC HYGROMA**

**Cystic Hygroma**
This is a multicellular cystic malformation of the lymphatic system. Incidence 1:12000, 60% present at birth, 90% detected before the 2nd year. During development of the lymphatic system, isolated cell buds fail to achieve venous connections. These continue to produce lymph and this results in fluid-filled endothelial cysts. These may enlarge and infiltrate surrounding tissue. Many have a solid component with bleeding and infection.

**Lymphangioma / Mixed mesodermal mal-formation**
There is an ill-defined gradation of malformation from pure cystic hygroma to those with an increase in angiomatous element - so called lymphangioma. 75% occur in the lateral neck and they are twice as common on the left side. Extension into the axilla or mediastinum is common. Secondary infection, which produces massive swelling, often prolonged due to poor lymphatic drainage, is common and if this is in the neck, it may lead to suffocation.

**Treatment** is early excision remembering that the lesion is benign and therefore attempting to avoid surrounding major structures, particularly the facial nerve. The use of sclerosant injection, Bleomycin in an oil emulsion or OK432 an anti-endothelial cell antibody is occasionally successful but aspiration, drainage or radiotherapy are contra-indicated. In those cases where the anastomosing lymph channels are cystic spaces (small, almost honeycombed) and where infiltration surrounds vital structures - there being a predilection to tongue, cheek, pharynx and retroperitoneum - the operations should be staged. The commonest form is involvement of the tongue with extension into the floor of the mouth. This often results in multiple cystic vesicles over the tongue with gross enlargement. Again staged excision is the treatment, remembering that there will be a period of prolonged post-operative oedema in the remaining tissue which may compromise the airway for some time.

**NEUROFIBROMA**
This is commonly an expression of Von Recklinghausen's disease, a dominant inherited genetic disorder of variable penetrance. Isolated tumours may occur in the skin, subcutaneous tissue or in relation to spinal nerves. They are soft, grey, fleshy and slightly mucoid. The subcutaneous lesions lie in the dermis, producing an elevated nodule with compression of the epidermis and stretching of the overlying skin. The deep aspect has an ill-defined edge. Where multiple nerve twigs are involved, this is called a plexiform neurofibroma. There is a definite incidence of malignancy in the lesion itself and an association with neural crest tumours. In addition in those related to spinal nerves there may be intraspinal extension with spinal cord compression. Neurofibromatosis is suggested when the following are present:
1. Five or more café au lait macules >0.5 cm in diameter.
2. Cutaneous neurofibromas.
3. Other neural crest tumours, e.g. phaeochromocytoma.
4. Leish nodules in the iris.
5. A relative with neurofibromatosis plus 2 of the above.

Diagnosis can be made by skin biopsy, looking for macromelanosomes.

**FIBROLIPOMA**
This is a common hamartomatous lesion, which usually presents as a subcutaneous swelling with a poorly defined edge. Treatment is excision biopsy. There is a 15% incidence of local recurrence. Beware of those occurring over the sacrum - usually
associated with a meningocoele that may extend into the spinal cord.

THE BREAST

Neonatal mastitis
The majority of infants, both male and female, develop enlargement of one or both breasts in the first weeks of life. Swelling may be tender and is often accompanied by secretion of a few drops of milky material, `witch's milk'. This is thought to be due to maternal gonadotrophic stimulation of foetal prolactin and/or oestrogen. Withdrawal of this stimulation at birth allows the swelling and secretion to subside. No treatment is required. Any attempt to express the breasts may prolong secretion with increased milk production and greatly predisposes to infection. Infection itself may be devastating with involvement and destruction of the breast bud and chest wall. Overwhelming septicaemia may lead to death. Abscess formation requires formal drainage and systemic antibiotic therapy against the organisms most involved - Staphylococcus and Streptococcus.

Premature thelarche
A condition of bilateral breast development usually occurring before puberty with no other signs of sexual maturation nor evidence of oestrogen effect on vaginal cells. Although both breasts are involved one may develop before the other. There is breast enlargement but the nipple shows little or no change. The condition is believed to be the result of increased sensitivity to circulating gonadotrophin and oestrogen. Treatment is expectant with reassurance. Do not biopsy. This must be distinguished from precocious puberty where there is early generalized maturation with evidence of secondary sex characteristics and is usually the result of ovarian, or more rarely, intracranial or adrenal tumours.

Gynaecomastia or pubertal mastitis
implies excessive development of the male breast and usually occurs in adolescent boys. There is often a tender, uncomfortable granular disc under the nipple, with variable soft tissue enlargement. This is believed to be a result of altered testosterone: oestrogen ratio, which originates from the testis. The disorder is benign and self-limiting. Carcinoma extremely uncommon in children.

THE UMBILICUS

Umbilical hernia
Twenty percent in the newborn. The defect is round and most close spontaneously. The skin may look thin though it never ruptures. Complications are rare. If the diameter is less than 1 cm at 1 year, the defect can be expected to spontaneously close; if the defect is greater than 2 cm at 4 years, it is unlikely to close and an operation is usually attempted after 5 years of age. Irreducibility may be caused by inspissated sand in the small bowel due to pica. Most reduce spontaneously with bed rest and sedation, and are then electively repaired.

Para-umbilical hernia/Supra-umbilical hernia
This is differentiated from umbilical hernia by the defect being elliptical and in the linea alba adjacent to the umbilicus. These will not close and require surgical repair. Irreducibility is also more common.

Umbilical sepsis
Infection in the neonatal period is not uncommon and usually presents with erythema, oedema and swelling of the umbilicus with extending oedema into the abdominal wall. Infection here should never be taken lightly as rapid progression to septicaemia or necrotising fasciitis may also occur. Infection may spread via superficial lymphatics to the inguinal lymph nodes. Portal vein thrombosis can
occur with subsequent development of portal hypertension. Common organisms are staphylococci, haemolytic streptococci and clostridium.

An umbilical granuloma presents as heaped up grey red granulations and is a pyogenic granuloma, which can be treated with either ligation if there is a stalk, or daily applications of silver nitrate with care taken to protect the surrounding skin. Infection in an obliterated umbilical artery may also present with intermittent purulent discharge.

**Umbilical discharge**
Discharge from the umbilicus may be purulent, watery, mucoid or faecal. **Purulent discharge** is most commonly related to localised infection of the umbilicus. An inflammatory response at the umbilicus is part of the normal mechanism of shedding of the cord. Prolonged adherence of the cord should make one suspicious of either a generalised condition relating to the inflammatory response e.g. immune deficiency, or a localised underlying defect at the umbilicus.

**Watery discharge:** This is usually due to a persistent communication with the urachus and bladder and may suggest lower urinary tract obstruction. Thus the urinary tract should be investigated with ultrasound and cystogram prior to surgical excision of the urachus remnant. A partly patent urachus may present as a midline sub-umbilical mass or if infection has occurred, as an abscess. This requires initial drainage if infected and subsequent excision.

**Mucus or faecal discharge:** These are due to abnormalities of the vitello-intestinal duct. Embryonic communication between the midgut and the yolk sac leaves the yolk stalk. This usually completely obliterates. Anomalies of the above may be:-

(i) Enterotoma. This is a sequestrated nodule of alimentary mucosa, leaving a pseudopolyp, which presents as a shiny, red (Cherry tumour) nodule in the umbilicus with a mucoid discharge present from birth.

(ii) Patency of the whole tract leaves a fistula, which discharges bile-stained small bowel material from the umbilicus.

(iii) Enterocystoma. Where there is partial obliteration, a cyst may be present which usually lies beneath the umbilicus within the abdomen. This may become infected, resulting in an abscess with intermittent discharge of pus.

**Treatment:** Exploration of the umbilicus. A check for an intraperitoneal remnant, and excision of the ectopic mucosal cyst.

**MECKEL'S BAND**
A Meckel's band is where there has been obliteration of the omphalomesenteric duct but a residual fibrous band remains, attaching the umbilicus to the mid-ileum. This may be associated with an antimesenteric diverticulum, Meckel's diverticulum, which is the patent inner segment of the duct. This attachment leads to twisting and knotting of the bowel around the band, resulting in small bowel obstruction and strangulation.

**ANORECTAL PATHOLOGY**

**Fissure-in-ano**
This, the commonest cause of a minor fresh rectal bleed occurring in infants and toddlers, is due to passage of a large stool, splitting the mucocutaneous lining of the anal canal. Usually associated with pain and a streak of blood on the outside of the stool. Most often occurs in the midline posteriorly. Many heal quickly. It is a common cause of rectal bleeding and realisation that if not treated, the complication of fear of defaecation, stool retention and subsequent persistent constipation and faecal soiling may occur. Treatment is to treat the constipation with a stool
softener and a mild laxative, having attended to any predisposing dietary factors. A local anaesthetic cream may be used for a short period. The rectum must be emptied of hard stool (disposable enema). In resistant cases or where a large faecaloma is present, under GA the faecaloma is removed and the anus forcibly dilated to temporarily paralyze the sphincters, thus preventing sphincter spasm (pain +). This allows normal bowel function to resume. Partial internal sphincterotomy is rarely necessary. Nitric oxide paste applied locally has some benefit.

**Rectal Polyps**
These may be single, multiple or diffuse. The commonest is the **juvenile polyp**, which is a pedunculated, round, friable, partly cystic lesion composed of regular rectal colonic glands set in a fibrous stroma with mucus retention. The surface is ulcerated, and is covered by a layer of granulation tissue. This is probably an acquired inflammatory lesion although commonly termed a hamartoma. Presentation is almost universally with fresh rectal bleeding and in some cases with prolapse of the polyp through the anus. Auto-amputation is recorded in about 10% of cases. Juvenile polyps have no potential for cancer and treatment is with local excision, >90% being within reach of a sigmoidoscope. Rarely the polyps may be diffuse and these children usually present young, with symptoms of bleeding, anaemia, prolapse, episodes of intussusception and malnutrition. Treatment is to identify the extent of the disease with barium enema, sigmoidoscopy and colonoscopy, and if extensive, resection of the diseased colon should be done early before significant complications develop. Rarely in diffuse juvenile polyposis, a gradation to adenoma may be seen on histology.

**Lymphoid polyps** are not uncommon and are produced by submucosal lymphoid hyperplasia. The cause is unknown. It is important to differentiate these from other intestinal polyps. **Inflammatory polyps** are unusual but may be associated with schistosomiasis and ulcerative colitis - previously described as pseudopolyps.

**Hamartomatous polyps**
**Peutz-Jegher's syndrome**: This is a syndrome of inherited polyposis. The polyps are usually multiple hamartomatous polyps varying in size from 0.5 to 2 cm diameter anywhere in the gastro-intestinal tract, with the majority limited to the small bowel. There is associated external evidence with hyper-pigmentation of the mucosa. Freckles may also occur on the face, hands, feet, digits, and hard palate. Inheritance is autosomal dominant with high penetrance and variable expressivity. The sex ratio is equal and there is no racial predilection. Symptoms are usually due to chronic blood loss and anaemia, or bouts of abdominal pain with incipient or established intussusception. Gastro-intestinal malignancy is rare.(incidence is approximately 2-3%). There is a higher malignancy rate in those polyps occurring in the stomach, duodenum and rectum. In females there is a predisposition to ovarian tumours. Treatment is tailored to the number, size, location and presentation of the polyps and can be defined as conservative surgery.

**Adenomatous polyps**
**Familial polyposis coli**: Autosomal dominant inheritance with varying degrees of penetrance. These polyps are adenomas. The condition is inevitably pre-cancerous but rarely presents in the paediatric age group. The diagnosis is made by histological examination of a resected polyp. The treatment is total colectomy, ileo-rectal anastomosis and subsequent fulguration of rectal polyps with regular follow up.

**Rectal Prolapse**
This may be partial-mucosal or full
thickness. Most are partial and due to faulty bowel habit training - easily treatable. Predisposing factors of full thickness prolapse are poor stool habit, malnutrition with chronic diarrhoea, parasitic infestation, whooping cough, cystic fibrosis, neurological defect, e.g. spina bifida, or anatomical defects, e.g. ectopia vesicae.

Management
Exclude an underlying cause. Buccal smear and sweat test; spine x-ray; stool for parasites. If presenting acutely, conservative treatment with reduction of the prolapse and strapping of the buttocks, thus allowing the anal sphincter to regain tone, may be sufficient. Where rectal prolapse recurs after strapping, treatment is with sclerosant injections (phenol in almond oil). If chronic and persistent a circum-anal suture (Thiersch) or more formal reconstructive operation may be indicated. It is important to differentiate this from an intussusception prolapsing through the anus. The prolapsed rectal mucosa is a pale red colour with an inverted beehive shape whereas the intussusception is usually grossly congested, dark plum coloured with a curved sausage shape. There should always be a sulcus on the edge of the intussusception and should this be easily reduced, diagnosis must be confirmed sigmoidoscopically or with contrast enema. Haemorrhoids rarely occur in children. The other differential diagnosis is that of prolapse of a rectal polyp.

FUSED LABIA
Labial adhesion is often referred by the family physician as an ‘absent vagina’. It is common and is the result of adhesion of the labia minora. This may cover the introitus sufficiently to allow urine into the vagina and may be cribiform in nature. Application of an oestrogen cream usually results in spontaneous separation after ± 10 days. If this fails, digital separation under GA is indicated. There is a tendency to recurrence.

SWALLOWED FOREIGN BODIES
These are very common and rarely present with complications. The length of the object is more important than it's sharpness. As a general rule if the object has reached the stomach it should be passed uneventfully. Occasionally long objects lodge in the duodenum, terminal ileum, or rectum.

Treatment: Following a history of ingestion the position of the FB must be established by AP and lateral x-ray of neck, chest and abdomen. Those in the oesophagus should be removed immediately and others can be watched expectantly. Surgical treatment is only indicated where symptoms and signs appear. An exception is ingestion of alkaline button batteries (calculator, watch, hearing aid) as these contain concentrated sodium/potassium hydroxide and also poisonous metals. Charged batteries may cause coagulative necrosis of adjacent tissue. Thus if lodged in the oesophagus, a very careful endoscopic assessment must be carried out after removal. Fatalities due to oesophageal perforation have been recorded. Most button batteries swallowed pass through the gastrointestinal tract uneventfully. However an acid environment (stomach) encourages erosion of the plastic grommet seal, leading to disintegration of the battery and leakage of its contents. Thus antacids and aperients should be given. If the battery remains in the stomach for more than 12 hours it should be removed. This can easily be done by endoscopy.

PAROTID SWELLINGS
Diffuse
Inflammation
1. Mumps Pus cannot be expressed from the duct, serum amylase increased, neutropoenic,
bilateral

2. Acute suppurative parotitis; Painful, red, indurated gland associated with duct orifice stenosis, caused by injury from developing teeth, dehydration from diarrhoea and vomiting, cystic fibrosis and AIDS.

3. Acute suppurative lymphadenitis in an intraparotid lymph node

4. Recurrent parotitis of childhood; Common. Male:female ratio 3:2 with maximum incidence after 4 years. Rare in Blacks. Due to immaturity of basement membrane of acini; not congenital sialectasis as previously thought. Chemical parotitis due to liberation of parotid juice into parenchyma which may become secondarily infected.

**Symptoms**

Recurrent acute episodes of pain and swelling in the parotid, made worse by eating. Fairly sudden onset. Lasts 3-7 days with frequency varying 3-6 monthly. There may be constitutional signs of fever and malaise.

**Examination:**

The whole gland is enlarged and tender.

No erythema of overlying skin.

Flocculent pus expressible from the duct orifice.

The orifice is often red and oedematous; 60% one side involved. Most begin with unilateral attacks, the other side becoming involved months or years later. Simultaneous attacks on both sides are rare.

**Investigations**

Pus culture - strep. viridans, staphylococcus, pneumococcus.

Sialogram: punctate sialectasis, probably caused by the technique of sialography on the basis of underlying duct epithelial and basement membrane abnormality with increased fragility and easy rupture.

**Treatment**

Antibiotics for the acute attack where indicated. Prevention of further attacks with improved oral hygiene and gland massage.

Spontaneous remission expected at age of approx. 13 years.

Complications are usually due to ill-advised surgery.

**Lumps within parotid**

1. Lymphadenopathy
   - pyogenic 80%
   - TB
   - lymphoma

2. Mesodermal malformations
   - lymphangioma
   - haemangioma
   - neurofibroma
   - fibrolipoma

3. Cysts - first branchial arch cyst

4. Neoplasms

5. Rarer lesions:
   - Duct stenosis
   - Stone

**Lymphangioma** These may be intracapsular or may be primarily in adjacent tissues in the cervico-facial region, usually unilateral. If small, treatment is conservative, if large and unsightly careful excision is advised after the age of 5 years with preservation of the facial nerve.

**Neoplasms of the parotid:** These are rare - 5% of all parotid tumours occur in childhood. There is commonly a lymph node overlying the parotid gland and there is also significant lymphoid tissue within the gland.

**Presentation**

Age - approximately 5 years.

Long history.

Painless, smooth, localized and usually superficial swelling.

**Investigation**

Sialography - to define its position within the gland.
Distortion of ducts by tumour may suggest malignancy.

**Management**

Excision biopsy with preservation of the facial nerve.

Pathology:

(a) **Benign**: Pleomorphic adenoma. Warthin's tumour (adenolymphoma). Vascular or lymphatic hamartomas.

(b) **Malignant**:

- 1º: Muco-epidermoid carcinoma arising from duct cells. Lymphoma/Rhabdomyosarcoma.
- 2º: Metastases, e.g. retinoblastoma, neuroblastoma.

**THE PIGMENTED SKIN LESION**

Numerous different pathologies, including moles and naevi which are the commonest neoplasms in humans. Histologic assessment essential if excision indicated.

1. **Naevocellular naevi**

   **Site**: Anywhere on cutaneous and mucocutaneous surfaces.

   **Appearances**: Range from flat through nodular to papillomatous.

   **Pathology**: According to location of naevus cells, viz. Junctional-confined to dermo-epidermal junction. Intradermal - dermis only. Compound - both sites.

   **Treatment**: Nil unless: Cosmetic excision. Potential malignant change - see melanoma. For suspicious features - exclude by careful frequent clinical examinations.

2. **Congenital (Giant) pigmented naevus**

   **Site**: Usually in distribution of dermatomes.

   **Appearance**: Uneven pigmentation, with 95% containing hair.

   **Pathology**: Intradermal or compound naevus, penetrating lower 2/3 of dermis. Significant pre-disposition to malignancy (2-16%).

   **Treatment**: Total excision as early as technically possible. Staged excision required in large lesions.

3. **Café au lait spots**

   **Site**: Anywhere, especially in axilla.

   **Appearance**: Round, oval, flat, light brown pigmentation, found in 10-20% people

   **Pathology**: Pigment in basal layer of epidermis. 90% people with neuro-fibromatosis (von Recklinghausen’s disease) have these but larger and more numerous.

   Highly suggestive if:
   - >5 years old: 6+ spots more than 0.5 cm.
   - <5 years old: 5+ spots more than 0.5 cm.

   **Treatment**: Nil for skin lesion per se.

4. **Malignant melanoma**

   Very rare in children, only ± 100 cases reported.

   **Site**: Anywhere. Danger areas: Back, Arms, Neck, Scapula.
Appearance: Pigmented nodule of variegated colours (red, white, blue) within a single nodule.


Pathology: Proliferation of melanoma cells from dermo-epidermal junction into dermis. Stage and prognosis, related to depth of penetration into deeper layer.

Treatment: Wide surgical excision (>5 cm in adults) including the deep fascia. Expert histological assessment and further therapeutic options.

5. Spindle/epitheloid cell naevus (Juvenile melanoma of Spitz)

Site: Head and neck most commonly.

Appearance: Solitary pink to tan coloured nodule up to 1 cm diameter.


Deeper cells less active than superficial (opposite to melanoma)

Treatment: Excision biopsy if clinically worrying.

THE ENLARGED LIMB

There are multiple aetiologies of congenitally enlarged limbs, many of which are poorly understood. If there is sudden onset of an enlarged limb during childhood, an underlying tumour must be excluded.

Causes of enlarged limb include:

1. Congenital hemihypertrophy: Ipsilateral enlargement of body causing mild asymmetry which may result in orthopaedic problems. (No treatment required.) Increased risk of developing Wilms' tumour (1:32 cf 1:14,000) - requires screening; may be associated with adrenocortical / hepatic tumours; and Beckwith syndrome.

2. Vascular and mesenchymal malformations: Haemangioma or a combination with A-V fistulae e.g. Klippel Trenaunay and Lymphangioma e.g. Osler Rendu Weber Syndrome.

Often associated with overlying vascular markings; may cause disfigurement or haemodynamic complications (see Haemangioma).

Therapeutic problem - if indicated, requires expert investigations and treatment.

3. Neurofibromatosis:

Neurofibromatous overgrowth may cause local enlargement due to nerve involvement, as well as overgrowth of fibrous
tissue, fatty tissue and bone.

Indications for surgery:

- Mass/size.
- Grotesqueness, cosmetic.
- To exclude malignancy if rapidly growing or painful lesions.

4. Lymphoedema:

Primary: Congenital
(Millroy's disease) especially dorsum of foot or hand. Praecox (puberty onset). Female more than male, usually unilateral.


Due to deficient, absent or damaged superficial lymphatic network in the presence of normal arterial and venous systems. Results in pooling of lymph, and secondary venous decompression, in the subcutaneous fat.

Treatment: Non-operative primarily viz. weight loss, elastic stockings, elevation at night - to rid the limbs of as much oedema as possible.

Operation only if:
- Functional improvement needed, e.g. fitting of shoe.
- Recurrent lymphangitis (uncommon).
- Rarely for aesthetic reasons.

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