Thyroid Diseases

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Thyroglossal cyst & fistula

Thyroglossal cyst

Etiology: Congenital, due to persisting part of thyroglossal duct.

Incidence:

- **Age of onset:** Although congenital it is not present since birth but appears in childhood (It needs time to enlarge).

Pathology: **Structure of Thyroglossal cyst:**

- The cyst is lined with columnar epithelium.
- Its wall is rich in lymphatic tissue communicating with cervical nodes making the cyst liable to infection.
- The cyst contains clear mucoid fluid rich in cholesterol crystals.

Clinical Picture:

1. Size : Small (2-3 cm diameter).

2. Site : Commonly subhyoid, less commonly on thyroid cartilage and rarely suprahyoid. It lies in midline, but if on thyroid cartilage it is pushed by the central prominence of larynx to one side, usually the left.

3. Mobility :

- Being attached to foramen caecum, it moves up with protrusion of tongue (diagnostic).
- In addition it moves up with swallowing as goitre, being inside pretracheal fascia.

N.B.:

- **It is attached to hyoid bone by a fibrous cord.**

D.D.: From midline neck swellings (cystic and solid), but mainly from cystadenoma of thyroid isthmus.

Complications: **Liable to repeated infection.**

N.B.:

- Thyroglossal cyst & subhyoid bursitis moves with deglutition & protrusion of tongue.

Thyroglossal fistula

Never congenital, but results from :

1. Incomplete removal of thyroglossal cysts.
2. Rupture or incision of the inflamed cyst.

Clinically :

1. History of pre-existing cyst.
2. The fistula opens in midline, low in neck.
3. The opening shows a characteristic semilunar fold of skin due to retraction of the fistula as the neck elongates.
4. It discharges mucus and, if infected, pus.
5. The track of the fistula can be felt as cord attached to body of hyoid bone and moves up with protrusion of tongue and also with swallowing.

D.D.: From fistulae and sinuses in neck.

Structure :
The thyroglossal fistula is lined by columnar epithelium and discharges mucus. Its wall is rich in lymphoid tissue and is liable to recurrent inflammation.

Treatment :
The same for cyst of fistula. The idea is to remove all remnants of thyroglossal duct up to foramen caecum **(Sistrunk's operation),** the fistula or cyst is dissected and the track is followed up to hyoid bone. The body of hyoid bone is excised with the track, being adherent to it. Finally, the track is followed up to foramen caecum in tongue.

N.B. Recurrence after operation occurs as a thyroglossal fistula. This is due to incomplete removal of thyroglossal duct.

Lingual thyroid

Pathology :
Failure of descent of the thyroid can lead to an intralingual development of the gland.

Clinical picture :
Patients may present with a tongue swelling with obstructive manifestations i.e. dysphagia, difficult breathing or change of quality of voice.

Investigations :
Thyroid scans: show absence of thyroid tissue in the normal side in the neck.

Treatment :
1. Full replacement with L-thyroxine to induce reduction in size.
2. Surgical excision
3. Radioactive-iodine ablation

Mechanism of formation of thyroid hormones

T Iodine **T**rapping.
O **O**xidation by peroxidase enzyme (inorganic → organic).
B **B**inding to tyrosine → Mono & di-iodotyrosine (MIT, DIT)
C **C**oupling of MIT & DIT → T3, T4
S **S** torage as thyroglobulin.
R **R**elease.

N.B :
- **Perchlorate** → prevents T
- **Carbimazol & Thyouracil** → depress B
- **Lugol's iodine** → depress R
- **T.S.H** → +++T, O, B, C, S, R & produces hyperplasia of the gland

N.B. : The Pituitary–Thyroid Axis :
- T.S.H secretion is controlled by blood level of thyroxin. A low blood level of thyroxin stimulates the release of T.R.H (thyrotropin releasing hormone), which stimulates the anterior pituitary to produce T.S.H. The later increases the size of the thyroid gland, its vascularity and its production of thyroid hormones.
- A high level of thyroxin on the other hand inhibits TRH & T.S.H and reduces the release of thyroxin.

Goitre

What is goiter?

In Latin-gutter=the throat
It is any generalized enlargement of thyroid gland.

Types of Goitre:

1. Simple:
(a) Physiological.
(b) Colloid.
(c) Nodular.

2. Toxic:
(a) Primary.
(b) Secondary.
(c) Toxic nodule.

3. Neoplastic: Thyroid carcinoma (benign tumors are rare).

5. Autoimmune goitre as Hashimoto's thyroiditis and collagen diseases as Riedel's thyroiditis
6. Hypothyroid goitre.

Simple Goitre

Definition :

Non-toxic, non-neoplastic, non-inflammatory thyroid enlargement. Essentially the patient is euthyroid.

Incidence :

Sex : Commoner in females.
Age : Usually starts at a young age especially in endemic areas.
Residence : Certain areas are endemic (oases), but sporadic cases appear anywhere.

Cause :

Simple goitre is due to persistent stimulation of thyroid gland by high TSH level. Increased TSH secretion is due to decreased production of thyroid hormones from:

[A] Iodine deficiency :

- This may be absolute or relative.
- The normal daily requirement of iodine is 100-125 µg.

1. Absolute deficiency : Simple goitre is endemic in regions with low iodide content of the water.
2. Relative deficiency : Occurs during periods of stress e.g. females during the menarche, pregnancy and lactation.

[B] Enzymatic deficiency :

- Responsible for many cases of sporadic goitre.
- Positive family history is common.
- Examples :

1. Pendred's syndrome : It is due to deficiency of peroxidase enzyme. There are goitre, deafness and mutism.
2. Deficiency of dehalogenase enzyme

[C] Goitrogens : e.g.: Thiocyanates in cabbages, drugs as para-amino salicylic acid and antithyroid drugs. Large doses of iodides inhibit iodine binding, thus resulting in iodide goitre.

Pathogenesis (How does simple goitre develop ?)

Stages of goitre formation :

[1] Deficient synthesis of thyroid hormone leads to persistent TSH elevation that produces diffuse homogenous hyperplasia. All follicles are active **(Diffuse hyperplastic goitre).**

- This stage is reversible, i.e. If the iodine requirements are satisfied, the follicles undergo involution.

[2] Sometimes, when the iodine requirements are satisfied the follicles undergo hyperinvolution and become large in size, lined by flattened epithelium and full of colloid **(Simple colloid goiter).**

[3] Repeated fluctuations in the level of TSH due to repeated stresses which produce :

(a) Repetition of this process of hyperplasia and involution.

(b) Heterogenecity of the gland, with areas of active follicles and others with inactive follicles.

(c) Nodular goiter : most nodules being inactive, the active follicles being present in the internodular tissue.

Immunoglobulins is one of the factors (other than TSH), which play a role in etiology of nodular goitre.

1. Physiological Goitre (Diffuse Hyperplastic Goitre)

Incidence : Usually in female at puberty (10-20y) during pregnancy and lactation.

Pathogenesis : See before.

Gross Features : Regular, moderate, smooth, soft enlargement of the whole gland cut section is fleshy brown.

Histology : Diffuse hyperplasia. Acini are increased in number, lined with columnar epithelium and contain little colloid.

Clinical picture : The only symptom is fullness of lower part of the front of neck **(Venus neck).**

Signs : (Thyroid swelling)

1. Regular enlargement of the whole gland, which is butterfly shaped.
2. Moderate size.
3. Smooth surface.
4. Soft consistency.

2. Colloid Goitre

Is an intermediate stage between physiological and nodular goitre.

Gross features : The gland is regularly or irregularly enlarged, soft and smooth. Cut section shows wide spaces filled with colloid.

Histology : Hyperinvolution. Acini are hugely distended with colloid and lined with flat epithelium. Adjacent acini may rupture and fuse together forming large cysts filled with colloid.

Clinical Picture :

The gland is enlarged with irregular surface and soft consistency.

3. Simple Nodular Goitre

Clinically there are 2 types :

Diffuse simple nodular goitre = More than one nodule is felt.
Solitary simple nodular goitre = One nodule is felt.

Incidence :

Frequency : It is the commonest type of goiter. **Locality :** sporadic or endemic.
Age : 30-40 (younger if endemic). **Sex :** Commoner in females.

Pathogenesis : See before.

Gross features :

- Usually multiple nodules are grossly present.
- A dominant nodule may be present but microscopic changes are present throughout the gland.
- Cystic degeneration, hemorrhage and calcification are common.

Histology :

- Dense fibrous stroma around nodules.
- Nodules contain hyperinvoluted acini (colloid nodules which are bulky & gelatinous), distended with colloid and lined with flat epithelium.

N.B.: Most nodules are inactive. Active acini exist only in internodular tissue.

Clinical picture :

Symptoms : There are 2 groups of symptoms :

1. Disfiguring swelling : In lower part of neck of insidious onset, long duration & gradual enlargement.

2. Pressure symptoms (by large goitre) :

[A] Dyspnea from pressure on trachea :

It increases by sleeping on high or low pillow, so that it is called postural dyspnea. **It results from :**

1. Lateral kinking of trachea in unilateral goitre,
2. Compression from both sides in bilateral huge goitre (scabbard trachea)
3. Anteroposterior compression in retrosternal goitre.
4. Prolonged pressure on trachea produces softening and easy collapse (tracheomalacia).

[B] Dysphagia from pressure on esophagus : only retrosternal goitre.

Signs : The swelling shows the following characters :

1. **Butterfly shape** (Diffuse enlargement) or **Localized lump** (adenoma).
2. **In lower part of front of neck** (Muscular triangle).
3. **Moves up with deglutition** (being enclosed in pretracheal fascia).

N.B.: These 3 signs prove that the swelling is goiter

> Through pretracheal fascia, the thyroid gland is suspended to thyroid cartilage & hyoid bone, which moves with deglutition. Ligament of Berry is a thickening of posterior part of pretracheal fascia that fixes the back of each lobe to the cricoid cartilage.

Swellings move with deglutition	Thyroid swellings don't move with deglutition
Thyroglossal cyst, cervical ectopic thyroid, prelaryngeal & pretracheal L.N.s, subhyoid bursitis, parathyroid swelling.	Fixed by malignancy, Reidel's thyroiditis, scarring of previous operation, huge goiter.

4. **Size :** Variable.
5. **Surface :** Nodular.
6. **Consistency :** Firm or tense cyst.
   - If hard = calcification, malignancy or Reidl's thyroiditis.
   - If cystic = cystic degeneration, hemorrhage or suppuration.
7. **Border :** Well-defined (in retrosternal goitre: lower border can not be felt).

8. Tenderness : Not except if infection occurs in a nodule.
9. No thrill or murmur over the gland (only in toxic goitre).
10. Transmitted pulsations may exist (expansile in toxic goitre).
11. Percussion over manubrium sterni = dullness in retrosternal goitre.
12. Relation of the swelling to surrounding structures :
- **Absent carotid pulse denotes malignant infiltration = Berry's sign.**
- **Fixity of goitre to skin, sternomastoid or trachea denotes thyroiditis, malignancy or postoperative adhesions.**

Investigations :

1. Routine lab & ECG.
2. Indirect Laryngoscopy.
3. Plain X-ray : may show calcification and tracheal deviation or compression.
4. Ultrasound : to detect cystic from solid lesions and can it can detect retresternal goiter
5. Thyroid function tests : to exclude, hyper or hypothyroidism(subclinical case of toxic goiter).
6. Thyroid antibodies estimation : to exclude autoimmune (Hashimoto's) thyroiditis : not routine.
7. Biopsy (FNAC) : not routine.

Complications of simple nodular goiter :

1. Malignancy : Recent rapid enlargement + hard consistency (3%). Usually follicular type.
2. Toxicity : producing secondary toxic goitre (30%).
3. Tracheal obstruction by compression : (see c/p).
4. Infection in a nodule : Rapid enlargement, pain, warmth & tenderness.
5. Cyst formation (cystadenoma) from :
(a) Degenerated nodule.
(b) Rupture and fusion of adjacent colloid acini.
6. Hemorrhage in a nodule or cystadenoma :
Produces sudden enlargement of preexisting goitre. Severe dyspnea results if impaction occurs at thoracic outlet **and urgent treatment is essential :**
(a) Cold applications. (b) Aspiration.
(c) Incision in skin and deep fascia allowing the gland to bulge freely in the wound releasing the trachea until urgent thyroidectomy is done.
7. Calcification in a nodule : hard, simulating malignancy.
8. Retrosternal extension : occurs usually in men due to stronger pretracheal muscles and shorter neck.

Treatment of simple goiter :

1. Prevention: The use of iodized table salt (potassium iodide 1:10,000)

2. Treatment of diffuse hyperplastic goitre:

- Usually reversible by the use of L-thyroxin, usually 0.2 mg/d for several months to be tapered to 0.1 mg/day for several years.
- The patient is reassured and is advised for regular follow-up.

3. Treatment of simple nodular goiter:

Surgery is indicated for :
1. Evidence of compression manifestations mainly upon the trachea.
2. Cosmetic reasons.
- Both lobes affected = Partial thyroidectomy (subtotal).
- Only one lobe affected = Hemi-thyroidectomy.

N.B.:
1. Removed gland is sent for histological examination (biopsy).

N.B: Histological surprise:
Accidentally discovered malignancy from histological examination of removed benign thyroid swelling.

2. After operation 0.1mg L-thyroxin is given daily for life to avoid recurrence by inhibiting TSH stimulation.
3. Surgery should not be advised below the age of 25 unless very much indicated as it may be followed by recurrence.

Retrosternal Goitre

Definition :

- Goitre in superior mediastinum.
- It is an anatomical, not pathological diagnosis.

Pathology :

- Retrosternal goitres usually arise in a normally placed thyroid gland. The sternohyoid and sternothyroid muscles prevent forward expansion, and assisted by gravity and the negative intrathoracic pressure, they direct the swelling into the mediastinum.
- In most cases, the intrathoracic swelling occupies the superior mediastinum, but occasionally it lies in the posterior mediastinum behind the trachea, and sometimes behind the esophagus.

Retrosternal goitre is classified into 3 types :

1. Plunging goitres : which rise with deglutition and descend again through the thoracic inlet.
2. Mediastinal goitres : which lie wholly in the chest, but are connected with the thyroid by a relatively narrow band of tissue and derive their blood supply from the thyroid vessels.
3. Intrathoracic goitres : which lie wholly in the chest, completely separate from the gland. They probably arise from congenitally misplaced thyroid tissue, deriving independent blood supply from mediastinal vessels.

Clinical features :

- An intrathoracic goiter is more common in men particularly short-necked individuals.
- It often remains symptomless for years.

1. **Dyspnea** from compression of the trachea, it gets worse at night and aggravated with lying down or flexion of the neck. Some patients are diagnosed as asthamtics.
2. **Cyanosis and edema** of the face and neck.
3. **Dilatation of superficial veins of the upper part of the chest** due to obstruction of the innominate veins **(D.D.** congestive heart failure**).**
4. **Dysphagia** may be present.
5. **Palpation of the neck** may reveal an enlarged thyroid gland (the lower edge cannot be felt).
6. **Percussion of the sternum** may reveal retrosternal dullness.
7. **Symptoms and signs of toxicity or malignancy** may be present.

Special tests : (Pemberton's sign)

Ask the patient to raise up both arms or to tilt her/his head to one side strongly & keep this position for a while → Congestion of face & distress occurs due to obstruction of great veins & trachea at thoracic inlet.

Investigations :

1. Chest X-ray shows a soft tissue shadow in superior mediastinum.
2. Sonar.
3. Thyroid scan.
4. C.T. Scan of thorax will relieve the exact level of the retrosternal extension and its anatomic relations.

Treatment :

Surgery is always essential. Cervical part of goitre is dealt with as usual and its vessels ligated. This step avoids the risk of hemorrhage, as retrosternal goitre takes its blood supply from neck.

Retrosternal goitre is dealt with by :

1. Gentle pulling to deliver it in neck.
2. Piecemeal removal from within the capsule to collapse the goitre and then pull it up in neck.
3. Only if retrosternal goitre is malignant or intrathoracic, sternal split is needed (median sternotomy).

N.B.: Preparation of toxic cases is only by Inderal, as antithyroid drugs are (goiterogenic).

Toxic Goitre (Thyrotoxicosis)

The term thyrotoxicosis is better used, as not all manifestations are a direct effect of elevated thyroid hormones, e.g. True Exophthalmos and pretibial myxedema.

Common types :

- Diffuse toxic goitre (Primary toxic goitre = Grave's disease) 75%.
- Toxic nodular goitre (Secondary toxic goitre = Plummer's disease) 15%.
- Toxic nodule 5%.

Rare causes of hyperthyroidism :

1. Thyrotoxicosis factitia : due to excess intake of L-thyroxin (> 0.2 - 0.3 mg per day).
2. Jod-Basedow thyrotoxicosis : Occurs in cases of physiological goitre upon the administration of large doses of iodine (iodine induced toxic goitre). Temporary
3. Subacute thyroiditis : De Quervain's diseases.
4. Hashitoxicosis : 5% of patients with Hashimoto's disease may have hyperthyroidism in the early stages of the disease.
5. Functioning thyroid carcinoma.
6. Neonatal thyrotoxicosis : may occur in newborns of thyrotoxic mothers due to transmission of thyroid antibodies across the placenta. Subsides in 3-4 weeks
7. TSH secreting adenomas of pituitary gland
N.B:T4 pseudothyrotoxicosis: there is increased T4 and decreased T3 secondary to failure of peripheral conversion of T4 to T3.

Primary toxic Goitre

There is diffuse enlargement of the whole thyroid gland accompanied by the clinical picture of thyrotoxicosis.

Incidence :

- Females : Males = 8:1.
- Usually 20 - 40 years.
- Usually associated with unilateral or bilateral exophthalmos in 85%of cases.

Etiology :

- It is an autoimmune disease, which leads to formation of abnormal thyroid stimulating antibodies leading to stimulation of T.S.H. receptors on the thyroid cell membrane resulting in an action on the thyroid cell similar to T.S.H but prolonged.
- The disease is often precipitated by psychic trauma, emotional stress, acute infections, pregnancy or labor.

Pathology :

[1] Thyroid :

- The gland is highly vascular and friable.

Gross picture : There is diffuse small or large, smooth, soft or firm enlargement of thyroid gland.
Cut section : Homogenous, reddish brown & fleshy appearance.
Microscopic picture :

The acini	The stroma
1. Hyperplasia and hypertrophy. 2. The acini contain no or little colloid.	1. There is increased vascularity with A-V shunts. 2. Dense lymphocytic infiltration.

[2] On other organs : • There is hyperplasia of the R.E.S .

Clinical picture :

[A] Local manifestations :

1. Moderate, regular diffuse thyroid enlargement, with minimal pressure effects.
2. Smooth surface.
3. Soft consistency (less commonly firm).
4. Evidence of high vascularity :
(a) Systolic expansile pulsations.
(b) Thrill (on palpation) and bruit (on auscultation) maximum at upper poles, where superior thyroid arteries lie superficial.

[B] Systemic manifestations :

[1] Metabolic :

1. Loss of weight in spite of increased appetite.
2. Excessive sweating.
3. Intolerance to hot weather.
4. The hands of the patient are warm and moist.

[2] Nervous system :

Symptoms : Nervousness, insomnia or nightmares.
Signs :
- The patient looks tense and irritable.
- Tremors in the outstretched hands.
- Exaggerated reflexes.

[3] Cardiovascular system :

Symptoms :
- Palpitation.
- There may be anginal pains.
- Orthopnea, pain in the right hypochondrium and edema of the lower limbs (in case of HF).

Signs :
- Tachycardia with a sleeping pulse more than 90/minute.
- Water hammer pulse is due to high systolic (due to increased cardiac output) and low diastolic blood pressure (due to decreased peripheral resistance).
- **Arrhythmias :** all types of cardiac arrhythmias except heart block may occur. Arrhythmia is usually started by extrasystoles.

[4] Muscular :

- Muscular weakness with rapid exhaustion.
- In severe cases proximal myopathy and muscle atrophy may occur.

[5] Skin :

(a) Patchy pigmentation may occur.
(b) Pretibial myxedema :
- Presents as multiple orange yellow patches of thickening of the skin over the shin of the tibia.
- They are due to deposition of mucin like substances.

[6] Gastrointestinal tract : Diarrhea is due to increased c-AMP which increases the permeability of cells of mucous membranes.

[7] Endocrine system :

- **In fem ale :** Menstrual abnormalities.
- **In males :** Abnormal libido, gynecomastia or impotence.

[8] Urinary system : Polyuria and glycosuria.

[9] Eye manifestations :

Symptoms :

1. Blurred vision.
2. Diplopia.
3. Pain and photophobia (corneal ulcers).
4. Excessive tearing.
5. Swollen black eyelids.
6. Bulging.

Signs :

I- Tremors : Detected in lightly closed eyelids.

II- Exophthalmos : May be false or true.

[A] False exophthalmos :

- It accompanies both 1ry and 2ry toxic goitres.

Etiology : It is due to contraction of the levator palpebrae superioris (Muller's muscle) pulling the upper eyelid's tarsus and superior conjunctival fornix upwards.

- The high level of thyroid hormones sensitizes this muscle to the effect of circulating catecholamines.

Treatment : It usually disappears when the hyperthyroidism is treated.

[B] True exophthalmos :

- There is actual protrusion of the eyeball.

Etiology : Deposition of fat and round cell infiltrate in the retrobulbar tissues or it may be an autoimmune disease due to thyroid stimulating antibodies affecting the ocular muscles.

Results :

1. Ophthalmic vein compression, leading to edema, conjunctival congestion and chemosis.
2. Weakness of extra -ocular muscles results in diplopia.
3. In severe cases papilledema and corneal ulceration may occur. The severe progressive form is known as **malignant exophthalmos.**

Treatment :

- Usually self-limiting and may even regress.

1. Sleeping in the semi-sitting position and lateral tarsorrhaphy may help to protect the eyes.
2. High doses of systemic prednisone may be helpful.
3. Orbital decompression may be indicated when the eye is endangered.

N.B.: Hypothyroidism may increase the condition.

Clinical detection of exophthalmos

1. Naffziger's method :

- The examiner stands behind the patient, with the head tilted backwards.
- In true exophthalmos the eyeballs protrude beyond the plane of the superciliary ridge.

2. Russell Frazer's method :

- Examining the patient from the side with the eyes lightly closed, will determine the depth of the groove between the orbital margin and the covered globe (shallow in true exophthalmos).

3. Ruler test :

- Examiner uses ruler & tries to rest its edge in front of eyeball on the supraorbital edge & zygoma.
- Normally, it doesn't touch the cornea.

4. Exophthalmometer:measures the distance between lateral orbital margin and apex of the cornea:normally it is 15-17 mm.

III- Eye signs, other than exophthalmos, include :

1. Stellwag's sign : staring look with infrequent blinking (normal blinking 5-8 /min).

2. Von Grafes' sign : the upper eyelid lags behind the moving eyeball as the patient looks down without moving the head.

3. Dalrymple's sign : appearance of a rim of sclera between the upper eyelid and the cornea.

4. Joffroy's sign : lack of forehead wrinkling on looking upwards without moving the head.

5. Moebius sign : defective convergence due to muscle paresis (for 30 cm at least).

Thyrotoxicosis should be suspected in :

1. Children with growth spurt, behavioral problems or myopathy.
2. Elderly persons with unexplained tachycardia or arrhythmia (apathetic hyperthyroidism).
3. Unexplained diarrhea.
4. Unexplained loss of weight.
5. Resistant heart failure.

Differential diagnosis :

1. Primary thyrotoxicosis should be differentiated from secondary thyrotoxicosis :

Items	Primary thyrotoxicosis	Secondary thyrotoxicosis
1. On top of	Normal thyroid gland.	Simple nodular goitre.
2. Exophthalmos	True in 85% of cases.	Only apparent due to thyroxin.
3. Histology	Diffuse hyperplasia.	Hyperplasia of internodular tissue.
4. Age of onset	Younger (20-40).	Older (over 40).
5. Onset	Rapid, after stress.	Gradual.
6. Course	Fluctuating.	Progressive.
7. Degree	Usually severe.	Usually mild.
8. Main toxic presentation	Metabolic & nervous.	Cardiovascular, due to older age.
9. Local signs	Smooth regular goitre, small or moderate, with no pressure effects. High vascularity.	Nodular goitre may be huge. High vascularity.
10. Treatment	Drugs, surgery, radioiodine.	Surgery is the best.

2. Other causes of goitre.
3. Other causes of loss of weight in spite of increase of appetite e.g. parasitic infestations & D.M.
4. Anxiety neurosis.

Investigations :

- **Thyroid function tests T_3, T_4, TSH and ^{123}I uptake test** confirm the diagnosis.
- **T_3 thyrotoxicosis** is identified by elevated free T_3.
- **Toxic adenoma** shows as a hot nodule on ^{123}I scanning with suppression of the uptake of the surrounding thyroid tissue.

N.B: measurement of serum TSH: The test is routinely performed with normal range of 0.5-5 Mu/l. It is no possible to do **ultrasensitive TSH** and this test now is the most sensitive for assessment of thyroid functions.

Thyroid functions tests

[1] measurement of serum TSH

[2] Free T_3 & T_4 in the serum : (measured by radioimmunoassay)

Normal values :

- Free serum T_4 = 8 - 26 pmol/L.
- Free serum T_3 = 3-9 pmol/L (essential if T_3 thyrotoxicosis is suspected).

[3] Total T_3 & T_4 in the serum: (measured by radioimmunoassay).

Normal value:

- Total serum T_4 = 55 - 150 nmol/L.
- Total serum T_3 = 1.2 - 3.1 nmol/L.

[4] T.R.H. test :

- I.V. Thyrotropin releasing hormone.

Normal : Rise of T.S.H. level in the serum.

In thyrotoxicosis : No rise in T.S.H level in the serum.

[5] Thyroid antibodies: raised in Hashimoto's thyroiditis and Grave's disease.

[6] Radioactive iodine studies :

^{123}I is ideal as it has short half life (11 hrs.) while it is 8 days in ^{131}I.

Radioactive iodine studies include :

(A) ^{123}I uptake by thyroid gland and excretion in urine : (ordinary iodine is ^{127}I)

- 5 microcuries of ^{123}I is given orally → the uptake by the thyroid gland and the amount excreted in urine are measured at 4, 24 and 48 hours.
- In thyrotoxicosis : Very high dose of ^{123}I is taken rapidly by the thyroid gland with little excretion in urine → ^{123}I is discharged rapidly from the gland.

(B) Scanning of the thyroid gland :

- Using a special apparatus after giving a dose of $1^{123}I$, Technetium 99m or recently florescent scanning.

Values :

1. It gives an idea about the size and the shape of the gland.
2. Differentiate hot nodule i.e. increased activity (toxic) from a cold nodule i.e. decreased activity (cyst or malignant nodule).
3. Detect ectopic thyroid tissue e.g. thoracic or lingual.
4. Detect functioning thyroid metastasis.
5. It detects retrosternal extension.

[7] Blood examination :

- **In thyrotoxicosis there are :**

1. Hypercreatinemia : N: less than 1 mg%.
2. Hypocholesterolemia : N: 150 - 250 mg%.
3. Hyperglycemia : N: 120 mg%.

Treatment of thyrotoxicosis

[I] Non-specific measures : For all cases :

1. Rest. 2. Sedatives. 3. Nutritious diet and excess fluids.

[II] Specific measures to control thyrotoxicosis :

- Include anti-thyroid drugs, surgery and radioiodine.
- **Each patient is considered separately and one measure or a combination of measures is chosen for him.**
- **Choice depends on the following factors :**

1. Intelligence. 2. Socioeconomic factors. 3. The available medical facilities.
4. Age. 5. Pregnancy. 6. High thyroid antibodies titer.
7. General fitness. 8. Recent exophthalmos. 9. Cardiac complication (thyrocardiacs).
10. **Type of thyrotoxicosis :** examples :

(a) Primary toxic goiter : Surgery, anti-thyroid or radioiodine.
(b) Secondary toxic goiter : Surgery is always better because :
- Incomplete response to anti-thyroid drugs and radioiodine.
- Usually large with cosmetic and pressure effects and liable to all complications of nodular goitre.

(c) Toxic nodule : Surgery or radioiodine.
- Excision is easy and safe.
- Radioiodine is theoretically ideal, as suppressed thyroid tissue around the nodule will not take radioiodine, with no risk of delayed thyroid insufficiency.

(d) Recurrent thyrotoxicosis after adequate surgery : Radioiodine or anti-thyroid drugs.
(e) Failed anti-thyroid drugs or radioiodine : Surgery.
(f) Suspected malignancy : Surgery.
(g) Retrosternal goitre : Surgery, anti-thyroid drugs enlarge the goitre causing suffocation.

[A] Medical Treatment

Aim : to restore the patient to an euthyroid status, then to prescribe a maintenance dose for a prolonged period in the hope that a permanent remission occurs.

Indications :

1. Primary thyrotoxicosis. 2. Mild thyrotoxicosis.
3. Children and young patients. 4. Pre-operative preparation of patients.
5. Post-operative recurrence. 6. Refusal of surgery.
7. Small gland.

Contraindication :

1. Toxic nodular goitre. 2. Huge goitre causing pressure symptoms.
3. Retrosternal goitre. 4. Suspicious of malignancy.
5. Leucopenia or agranulocytosis. 6. Pregnancy and lactation.

Methods :

1. Sedative : ensure complete mental rest & good night sleep.
2. Inderal (B-blocker) : to decrease the HR and palpitation and partially inhibit conversion of T4 to T3.
Dose : 10 - 40 mg T.D.S. orally.
3. Anti-thyroid drugs :
Onset : Anti-thyroid drugs start to give their clinical effect after 2 weeks, thereafter it control thyrotoxicosis gradually till euthyroid state is reached.

Preparations :

[1] Lugol's iodine (5% I+ 10% KI) :

Indication : Used only in preoperative preparation.

Effect :

1. Reduces the effect of TSH on the thyroid gland and inhibits iodine binding.
2. Reduces the vascularity of the thyroid gland.
3. Leads to storage of colloid within the acini.
4. Its action declines after 2 weeks.

Dose : 10 drops tds with milk or juice to mask the bitter taste.

[2] Carbimazol : (The commonest anti-thyroid drug used)

- It blocks iodine binding to tyrosine and decreases antibody titers.

Dose :

- 10 mg tds (maximum dose 60 mg/d) till euthyroid state is reached then decrease the dose to 5 mg T.D.S for 12 - 18 months.
- If thyroxin 0.1 mg/day with anti-thyroid drugs → less danger of producing iatrogenic hypothyroidism or an increase in the size of the gland.

[3] Propylthiouracil :

- It blocks iodine binding to tyrosine and prevents peripheral conversion of T4 to the more active T3.

[4] Potassium perchlorate : It inhibits iodide trapping by the thyroid.

Advantages : Avoiding surgical risks and the possible hazards of radioactive materials.

Disadvantages :

1. There is no way to predict which patient is liable to pass into remission.
2. High relapse rate : 60% within 2 years from stopping treatment.
3. Further enlargement of the gland may occur.
4. Adverse effects of the drugs :
   - Thyouracil & Carbimazol may cause gastrointestinal upset, rashes & arthralgia.
   - Reversible bone marrow depression leading to agranulocytosis. Blood picture should be performed every two weeks.

[B] Radioactive iodine

Aim : It destroys the thyroid cells thus reducing the mass of functioning thyroid tissue.

Dose: 160 uCi per 1 gm of thyroid tissue. Improvement occurs by 8 - 12 weeks, if not a second dose may be required.

Indications :

1. Diffuse toxic goitre (above 25 y. old).
2. Thyrocardiac patients.
3. Refusal of surgery.
4. Recurrence after surgery.

Contraindication :

1. Below the age of 25 years.
2. During pregnancy & lactation.
3. Huge goiter, retrosternal goitre and suspicious of malignancy.

Advantage : Avoidance of surgery and prolonged medical therapy.

Disadvantage :

1. Thyroid insufficiency in up to 75-80% of cases after 10 years and so an indefinite follow-up is mandatory.
2. It is also inadequate for the treatment of secondary toxic goitre.

[C] Surgery

Aim : Surgery cures by reducing the mass of overactive tissues or removing all the overactive tissues in case of toxic nodule.

Indications of surgery are :
1. Secondary toxic goitre.
2. Severe primary thyrotoxicosis.
3. Retrosternal toxic goitre, huge goitre or suspicious of malignancy.
4. Failure of medical treatment.
5. Occurrence of side effects due to medical treatment.

Contraindications : Young patient, mild cases or recurrence after surgery.

Advantage : A rapid cure with a high rate of success.

Disadvantage :
1. Morbidity and mortality (negligible in experienced hands).
2. Recurrence rate less than 5%.
3. Thyroid insufficiency at an incidence of 20-45%.

Preoperative preparation :

[1] Routine Method :

Medical treatment :

- Neomercazole is given till the euthyroid state is reached then surgery is performed.
- Some surgeons add Lugol's iodine 10 - 14 days immediately prior to surgery to decrease the vascularity and friability of the gland.

[2] Rapid preparation :

By B-adrenergic blocker :

- The action of thyroid hormones especially on the CVS is proved to be through activation of adrenaline.
- Therefore inderal (propranolol) is now used for rapid control of CV manifestations before operation.
- It acts on target organs and not on the thyroid itself.
- So, it must be continued for one week after surgery.

Operation :

- The following precautions should be considered :

1. Pethidine and hyoscine are given before anesthesia (atropine is not used as it causes tachycardia).

2. The incision should be wide with division of pretracheal muscles, thus manipulation is minimal to avoid thyrotoxic crisis.

3. The operation done is **SubTotal Thyroidectomy** by which the main mass of the gland is removed leaving only a thin posteromedial wedge of each lobe to maintain an euthyroid state, preserve the parathyroids and protect the R.L.N. The amount of the gland left is 4-5 gm on each side.

4. Perfect hemostasis and free drainage to avoid thyrotoxic crisis.

Special problems in management

Thyrotoxicosis with pregnancy :

- **Radioiodine is absolutely contraindicated** because destruction of the fetal thyroid may result.
- **Antithyroid drugs** at usual doses would result in fetal hypothyroidism with the development of goitre that may obstruct the airway. Minimum dose antithyroid drug, preferably ß- blockers, reduces this risk.
- **Surgery after a short course of antithyroid drugs and propranolol** proved to be safe during the second trimester.

Thyrotoxicosis in children :

- **Radioiodine is contraindicated for children.**
- **Surgery is followed by a high recurrence rate** due to high activity of the cells in the young.
- It is preferable to treat them by **antithyroid drugs** until the late teens.

The thyrocardiac patient :

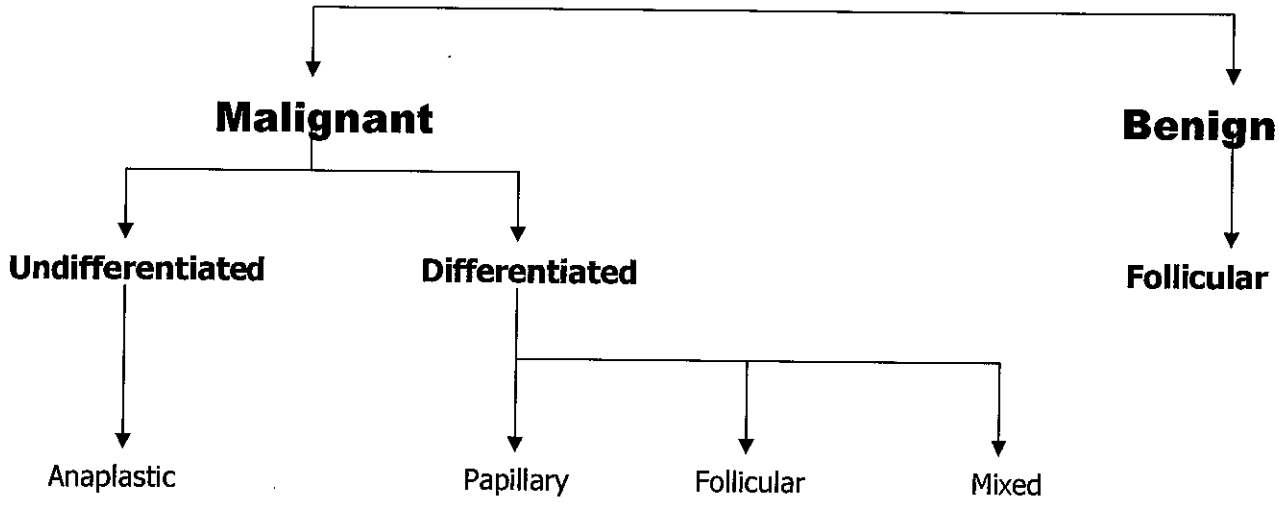
- **Thyroidectomy is ideal after control of the cardiac status.**
- **In unfit patients :** radioiodine is used with antithyroid drugs until the effect of radioiodine appears (6 weeks).

Proptosis of recent onset :

- It is not preferable to terminate the toxic status abruptly by surgery or radioiodine for fear of the risk of malignant exophthalmos.
- Antithyroid drugs are used until the proptosis has been stable for 6 months after which thyroidectomy is permissible.

Tumors Of The Thyroid Gland

1. Tumors arising from follicular epithelium :



2. Tumors arising from parafollicular epithelium : Medullary carcinoma.

3. Tumors arising from lymphoid elements : Malignant lymphoma

4. Rarely the thyroid gland is infiltrated by metastatic deposits or by local infiltration from a nearby lesion.

Follicular Adenoma

- Presents as a solitary nodule.
- Fine needle aspiration is not reliable to differentiate it from carcinoma as the main differentiating point in presence of histological evidence of capsular or vascular invasion.

Treatment :

Lobectomy plus isthmectomy.

(Many authors now consider any papillary adenoma as carcinoma).

Malignant Neoplasms

Incidence : More in females.

Predisposing factors :

1. External irradiation of the head and neck region in children : was previously used for treatment of hemangiomas and thymic gland enlargement.

2. Simple nodular goitre 3%.

3. Follicular carcinoma is the commonest type to develop in endemic goitrous areas : due to continuous stimulation by TSH.

4. Genetic : the lesion is more common in certain families. Hereditary factor is marked in medullary carcinoma.

5. Malignant lymphoma may occur on top of Hashimoto's disease.

6. Anaplastic cancer thyroid may occur DE NOVO.

Pathology :

[1] Tumors arising from follicular epithelium :

Items	Papillary	Follicular	Anaplastic
Incidence	60%	17%	13%
Age	May occur in children and young adults	Middle age	Elderly
Sex (F : M)	3.5 : 1	2 : 1	1 : 1.3
Microscopic picture	• Papillary projections composed of a stalk of connective tissue covered by a single layer of epithelial cells. • Laminated calcified bodies **(Psammoma bodies)** are often present in the stroma. The tumor shows characteristic pale empty nuclei **(orphan Annie-eyed nuclei)**.	• Thyroid follicles with a variable degree of differentiation. • Solid sheets may be present. • Diagnosis depends upon finding capsular or vascular invasion or the presence of metastases.	• It may take the form of spindle cell, small cell or giant cell types.
Multiplicity	Multiple foci may be present in the same or the other lobe in up to 80% of cases. These are due to either multicentricity or intrathyroidal lymphatic spread.	Rarely seen.	
Spread	Mainly by lymphatics	Mainly by blood stream	Early direct lymphatic or blood
10 year survival	90%	Encapsulated:97% Invasive :70%	Most of patients die within 1-2 years

N.B.: If the tumor contains both papillary and follicular elements it is considered papillary

[2] Tumors arising from parafollicular cells : (Medullary carcinoma 6%)

• The tumor may occur **alone**, or it may be associated with hyperparathyroidism and pheochromocytoma constituting **(MEN IIa).**

• **The tumor may be familial** and in this case, it is more common in children and young adults.
• **It secretes calcitonin** (Tumor marker).
• **Microscopically :** Sheets of neoplastic cells in a hyaline stroma, which may contain amyloid material.
• **Spread :** Lymphatic or blood.
• **There is diarrhea in 30% of cases.**

MEN (multiple endocrinal neoplasia) syndrome

Inherited condition, autosomal dominant, all family members should be screened.

[1] MEN1 (Wermer's syndrome) :

1. **P**arathyroid Hyperplasia
2. Islet cell tumors of **p**ancreas (e.g. gastrinoma).
3. **P**ituitary tumor.

[2] Type II Sipple's syndrome :

• **Type IIa :**

1. Parathyroid hyperplasia.
2. Pheochromocytoma.
3. Medullary Carcinoma (thyroid).

• **Type IIb:** Pheochromocytoma, Medullary Carcinoma and mucosal neuromas.

[3] Lymphoma.

TNM staging

T1: limited to thyroid gland, 1cm or less
T2: limited to thyroid gland, 1-4 cm.
T3: limited to thyroid gland,> 4 cm.
T4: extended beyond the capsule

N0 no nodal metastasis **M0** no distant metastasis
N1 nodal metastasis **M1** distant metastasis

N.B: microcarcinoma is a term used for cancers less than 1 cm in diameter. they carry excellent prognosis.

Clinical Picture :

- May be history of irradiation.

[A] Early or Suspicious presentations :

1. If on top of pre-existing goiter :
(a) Faster growth rate.
(b) Harder consistency.

2. If a lump appears de novo : The rule is that any single thyroid nodule (especially in young males) is considered malignant until proved otherwise, especially if rapidly growing, hard, irregular and ill defined, considering that carcinoma can occur at any age from 15 to over 60y.

3. Rarely, the patient presents with metastases while the primary is impalpable. This occurs particularly in papillary type and the patient presents with enlarged deep cervical nodes. In the past, before knowing the nature of such nodes they were called **lateral aberrant thyroid.**

[B] Late (sure) presentations : Goitre with spread

I- Local spread :

1. Hoarseness of voice (infiltrated RLN). However, laryngitis, tracheitis and RLN damage by past fever or previous thyroid surgery should be excluded.
2. Pain radiating to ear (infiltrated vagal thyroid branches). However hemorrhage or infection in a nodule should be excluded.

3. Dysphagia (infiltrated esophagus). Rarely it occurs in retrosternal simple goitre.
4. Severe dyspnea (infiltrated trachea).

5. Fixity to skin, sternomastoid or trachea.
6. Surrounding carotid sheath, biding carotid pulse instead of pushing it backwards **(Berry's sign).**

II- Lymph node metastasis :

- Mainly in prelaryngeal, pretracheal and lower deep cervical nodes.
- First lymph node affected in thyroid carcinoma is RLN then pretracheal and prelaryngeal (Delphic).

III- Blood metastasis :

N.B bone metastases present by: Painful, tender, pulsatile lump or pathological fractures (Common sites are spine, skull & neck of femur).

N.B.: Medullary carcinoma produces diarrhea in 30% of cases due to production of 5-hydroxytryptamine or prostaglandins.

D.D :Other causes of goitre especially hard nodules e.g. Tense cyst, calcified nodule & Reidel's thyroiditis.

Investigations :

[A] Laboratory Test : (For Tumor markers)

1. Serum levels of thyroglobulin : It is elevated in carcinoma as well as other types of goitres. It is only useful for following patients after total thyroidectomy to detect metastasis or recurrence.

2. Detection of serum calcitonin : elevation in medullary carcinoma.

[B] Radiological :

1. Thyroid scanning (of limited diagnostic significance). Malignant lesion usually appears as cold areas in the scan, but it should be remembered that many benign lesions as cysts or hemorrhage in a cyst or degenerative nodules or thyroiditis appear also as cold areas.

2. Ultrasound examination : can prove whether the lesion is cystic, solid or cystic with solid projection.

3. C.T. & M.R.I.

[C] Aspiration of cyst :

A benign cyst is characterized by :	A cystic malignant lesion is suspected when :
The aspirate is clear.	The aspirate is hemorrhagic.
Complete disappearance of the cyst.	A residual lump is present.
No reaccumulation of fluid.	Rapid reaccumulation of fluid in the cyst.
Cytological examination is free.	Cytological examination may be positive for malignant cells.

[D] Biopsy :

1. Fine needle aspiration cytology (FNAC) :

- An experienced pathologist can give accurate results in 90% of cases.
- This is a simple, fast, inexpensive investigation, which depends upon cytological examination of nodules 0.5 cm or more.
- False positive results occur in 2% while false negative results occur in 3% of cases.
- In follicular carcinoma, FNAC may not be conclusive as the diagnosis of this lesion depends upon demonstration of capsular or blood vessel invasion.

2. Pre-operative L.N. biopsy : may be done from an enlarged cervical node.

3. Frozen section examination during open operation.

4. Excision biopsy (hemithyroidectomy).

5. Incision biopsy is absolutely contraindicated as it results in seedling of malignant cells.

[E] Investigations to detect spread :

1. Chest X-ray : to detect pulmonary deposits.

2. Laryngoscopy : to check the mobility of the vocal cords.

Treatment :

I- Differentiated carcinoma

[1] Surgical treatment :

- In operable and if possible in inoperable cases (as palliation) either total thyroidectomy or lobectomy plus isthmectomy.

- **Surgeons who perform total thyroidectomy favor this procedure because :**

1. There is a high incidence of muiticentric lesions in papillary carcinoma.
2. Total thyroidectomy ablates all thyroid tissue and makes it possible to detect metastasis by scanning and to treat them by radioactive iodine.
3. It is easy to assess recurrence by serum thyroglobin.

- **Surgeons who perform lobectomy and isthmectomy favor this procedure because :**

1. No statistical difference was shown between the results of the two procedures.
2. Total thyroidectomy has a high incidence of post-operative hypoparathyroidism.

- **Management of lymph node metastases : (only in operable cases)**
  - If there are no palpable nodes: only the central lymph nodes are excised.
  - If there are metastasis in the posterior triangle: modified block dissection is done(spinal accessory nerve, internal jugular vein and sternomastoid muscle are preserved).

[2] Postoperatively L-thyroxin :

- 0.1-0.2 mg/day is prescribed to suppress TSH production, as most differentiated carcinomas are TSH dependent.

[3] Management of blood borne metastasis :

- Thyroxin is stopped for few weeks or substituted by tri-iodothyronine (to raise TSH) and then iodine scan is performed.
- If there is evidence of local recurrence or metastases, a therapeutic dose of radioactive iodine is prescribed.

II- Treatment of medullary carcinoma

- This tumor does not concentrate radioactive iodine and is not under the control of TSH.
- Treatment is by total thyroidectomy and resection of metastatic lymph nodes.

III- Treatment of anaplastic carcinoma

- The majority of these patients are inoperable when first seen.
- Rarely the lesion is operable and an attempt at cure by total thyroidectomy,
- The usual treatment is external irradiation and chemotherapy. If there is tracheal compression, resection of the isthmus is performed

VI- Treatment of lymphoma

1. Radiotherapy & chemotherapy.
2. Surgery only :
   (a) To establish the diagnosis.
   (b) Relieve a compressed airway.

Features of inoperability

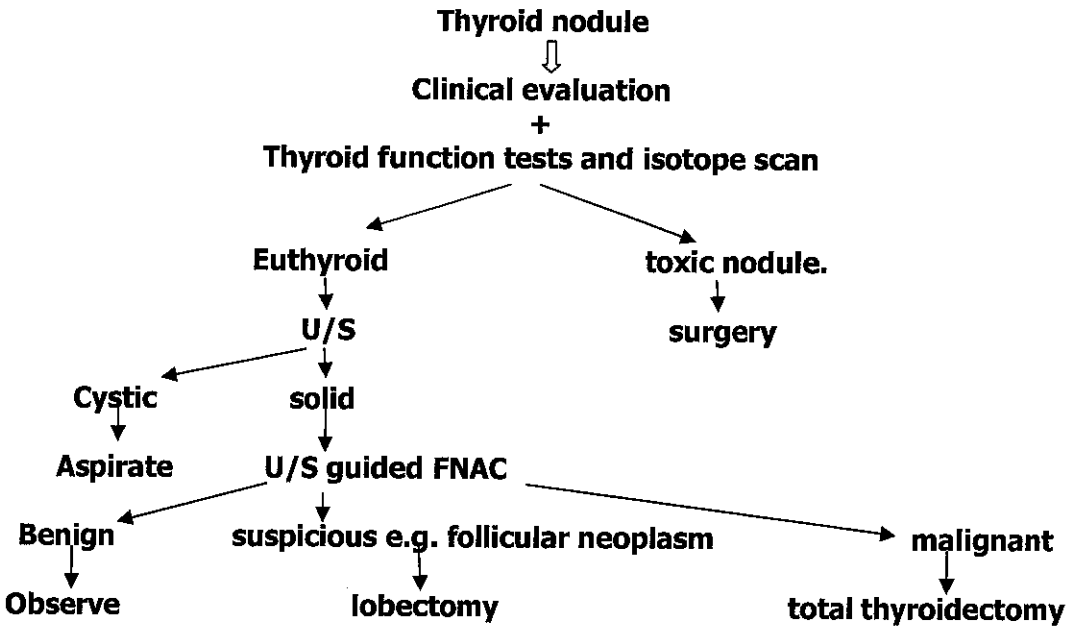
1. Infiltration of vital structures as carotids, trachea, bone etc.
2. Blood spread.
3. Adherent L.N. to vital structures.
4. Unfit patient
5. Anaplastic carcinoma is usually inoperable.

Prognosis of differentiated thyroid carcinoma

Certain factors increase the risk including :

1. **Age of the patient :** Males above 40 and females above 50 years of age.
2. **Sex :** Females has better prognosis.
3. **Size of the lesion :** If more than 5 cm in diameter.
4. **Presence of capsular or blood vessel invasion.**
5. **Presence of distant metastases.**
6. **Histological type.**

Algorithm for the management of a solitary thyroid nodule



Solitary Thyroid Nodule

Definition :

A goitre, which on clinical examination appears to be a single nodule in an otherwise normal gland.

Incidence :

More common in females and the commonest site is at the junction of the isthmus with the lateral lobe.

Etiology :

1. Simple nodular goitre with one palpable nodule (the commonest).
2. Toxic nodule.
3. Colloid nodule.
4. Adenoma.
5. Malignant nodule.
6. Inflammatory nodule (thyroiditis).

Clinical picture :

[1] Malignant manifestations : (In case of malignant nodules)

How to suspect malignancy in a solitary nodule ?

1. History of previous irradiation.
2. Young & elderly patients.
3. Recent onset & rapid growth.
4. Pain.
5. If the nodule is hard, irregular, with limited mobility.
6. Presence of local invasion, lymphatic or blood born metastases.

[2] Toxic manifestations : (In case of toxic nodules)

[3] Cases which are not Frankly malignant or toxic : The case may be :

1. Early malignant nodule.
2. Early toxic.
3. Simple nodule.
4. Adenoma.
5. Inflammatory nodules.

- **In these cases, investigations are essential to diagnose the case.**

Investigations :

1. Thyroid scan :

- May reveal the presence of multinodular goitre.
- If the nodule is hot, it is toxic and the possibility of malignancy is nearly excluded.
- If the nodule is warm, it is a functioning adenoma and the possibility of malignancy in only 3.5%.
- If the nodule is cold, the possibility of malignancy is 10-16%.

2. Ultrasound.

3. Biopsy (see carcinoma).

4. Aspiration.

5. Thyroid function test : Not routine.

6. Investigation of carcinoma : Not routine.

Treatment : Depend on the cause :

1. Malignant nodule: see before.

2. Toxic nodule : see before.

3. Cases which are not frankly malignant or toxic :

- Hemithyroidectomy and histological examination i.e. excision biopsy.
  (a) If not malignant → nothing more is needed.
  (b) If malignant → see treatment of thyroid carcinoma.

Thyroiditis

I- Acute Thyroiditis :

- The thyroid gland is rarely involved by bacterial infection.

Clinical picture : Sudden onset of severe neck pain with dysphagia, fever.
Treatment : antimicrobial drugs, however, surgical drainage of suppuration may be required.

II- Subacute Thyroiditis : (Granulomatous thyroiditis, De Quervain Thyroiditis)

Etiology : Most probably a viral infection (usually self-limited).
Clinical picture :

- Usually follows an upper respiratory infection.
- The thyroid becomes congested, swollen, firm, irregular & slightly tender.
- Mild manifestations of thyrotoxicosis may occur due to libration of hormones from destroyed tissue
- Pain with fever.

Rapid response to oral prednisolone is diagnostic

Investigations :

- Elevation of ESR with a normal or depressed leucocytic count.
- Iodine-uptake by the gland is depressed in the presence of a slight elevation of serum T^4.

Treatment : Oral prednisone & anti-inflammatory may be used.

III- Chronic Thyroiditis : due to tuberculosis or syphilis is rare.

Autoimmune thyroiditis (Hashimoto's Thyroiditis)

Incidence :

- The most common form of thyroiditis.
- Usually affects females at menopause

Pathologically :

- There is infiltration of the thyroid gland by lymphocytes and plasma cells.

Clinical picture :

- **Onset :** Insidious and asymptomatic or sudden and painful in others.
- The goitre usually lobulated, diffuse or localized to one lobe, soft or firm, flat topped nodularity.
- Mild hyperthyroidism may be present initially but hypothyroidism is inevitable.

Investigation :

- Serum titers of antimicrosomal and anti-thyroglobulin antibodies are elevated
- Biopsy. *FNAC or true cut biopsy may be required to differentiate it from nodular goiter or carcinoma*

Treatment :

1. Full replacement dosage of thyroxin.
2. Surgical treatment : Indications :

- Large-sized goitres.
- Suspected development of papillary carcinoma, lymphoma or both.

Riedel's Thyroiditis

- 0.5% of goitres.
- The thyroid is hard due to extensive fibrosis, which extends even beyond the gland.
- It is probably a collagen disease.
- Hypothyroidism is usually present.
- The differentiation from anaplastic carcinoma may need an open biopsy where a wedge of the isthmus is removed to free the trachea.

N.B:thyroid incidentinoma:
It is clinically unsuspected and impalpable thyroid swelling.

Thyroidectomy

Indications :

1. Unilateral lobectomy :
(a) All solitary nodules.
(b) Multinodular disease in one lobe only.

2. Subtotal thyroidectomy :
(a) Diffuse toxic goitre.
(b) Bilateral toxic multinodular goitre.
(c) Bilateral simple multinodular goitre.

3. Total thyroidectomy : Papillary, follicular & medullary carcinoma.

4. Isthmectomy :
(a) Anaplastic carcinoma & lymphoma to free airway & obtain tissue diagnosis.
(b) Riedel's thyroiditis.
(c) Nodule in the isthmus.

5. Near total thyroidectomy=lobectomy+isthmectomy+near total lobectomy:

Complications after thyroidectomy :

• Apart from general surgical complications (shock, hemorrhage, infection, pulmonary complications and deep vein thrombosis).

[1] Post-operative thyroid crisis (Storm) :

> This is a condition of **"acute hyperthyroidism"**

Incidence : Rare nowadays due to the good preparation & control of toxicity before operation.

• It can occur during the course of the disease excited by severe nervous disturbance or a minor operation e.g. extraction of a tooth or incision of an abscess.

• More commonly it occurs after subtotal thyroidectomy. It usually starts within the 1st few hours to 48 hours after the operation.

Manifestations :

1. Hyperpyrexia : Temperature may rise to 41 °C or more.
2. Rapidly rising pulse rate : 160 or more, may be irregular.
3. BP rises greatly, systolic & diastolic (e.g. 250/120). The pulse pressure is high, so HF may occur.
4. Muscular excitability up to convulsion.
5. Dyspnea due to lack of O_2 as the metabolic rate is greatly increased.

Fate : The condition, if severe, and if left untreated, death occurs in short time.

Treatment : Should be urgent :

1. Ice packs to the limbs, head & abdomen. In severe cases the whole patient may be immersed in cold water or ice.
2. ß-adrenergic blockers (Inderal) : I.V. drip until the pulse drops to about normal should be given carefully under ECG control.
3. Morphia for sedation.
4. O_2 inhalation is essential & I.V. hydrocortisone (25mg).

[2] Reactionary hemorrhage :

• Results from continuous venous ooze following bad hemostasis. This hemorrhage is serious because it collects under deep fascia, compressing trachea. Suffocation may occur.

Management :

1. Immediate reopening of the wound.
2. Then transferring patient to theatre to explore the operation field under anesthesia and ligate bleeders.

[3] Dyspnea : May be due to :
1. Reactionary hemorrhage.
2. Bilateral incomplete RLN injury = Spasm of both cords = Severe dyspnea= Urgent tracheostomy (immediately after extubation).
3. Laryngeal edema from rough intubation (= tracheostomy).
4. Tracheal collapse from tracheomalacia (= tracheostomy).
5. Thyrotoxic heart failure (propranolol - digitalis).

[4] Change of voice :

1	Unilateral RLN injury	Hoarseness
2	Bilateral complete RLN injury	Aphonia
3	External laryngeal nerve injury	Loss of high-pitched voice
4	Tracheitis and laryngitis	May cause temporary change in voice

[5] Injury of nerves related to thyroid :

Effects of RLN injury (on voice and respiration)			
1	Unilateral complete injury	Paralysis of one cord	Hoarseness + Dyspnea on exertion
2	Unilateral incomplete injury	Spasm of one cord	Hoarseness + Dyspnea on exertion
3	Bilateral complete injury	Paralysis of both cords	Aphonia + Dyspnea on exertion
4	Bilateral incomplete injury	Spasm of both cords	Aphonia + Stridor

- 3% of people have RLN paralysis (without operation) may be due to viral infection → neuritis. So do preoperative laryngoscopy (medico-legal).

[6] Tracheitis and laryngitis = Temporary change of voice + dyspnea.

[7] Surgical emphysema = Tracheal injury = Immediate suturing.

[8] Hypoparathyroidism :
- Rarely occurs if all parathyroid are removed, or their vessels injured.
- Cramps (tetany) start few days after operation.

Emergency treatment : 20 ml of 10% calcium gluconate IV slowly.
Maintenance treatment : Ca + Vit D.

[9] Progressive exophthalmos :
Management :
1. Thyroxin to inhibit pituitary.
2. Preventing progression by sleeping semi-sitting and cortisone.
3. The eye is protected by :
   (a) Tarsorrhaphy.
   (b) Orbital decompression in malignant exophthalmos.

[10] Hypothyroidism : If enough thyroid tissue is not left.

[11] Recurrence : Of toxic or nodular goitre.

[12] Keloid in the scar :
Occurs in low wounds on sternum. It is avoided by placing the incision in the lower neck crease.

Hyperparathyroidism

Types :

1. Primary hyperparathyroidism : Autonomous production of parathormone with loss of feedback mechanism.

2. Secondary hyperparathyroidism : Due to compensatory hypersecretion of parathormone secondary to low serum calcium.

3. Tertiary hyperparathyroidism : Starts as secondary type but develops autonomous dysfunction.

Primary hyperparathyroidism

Incidence :

- 0.1-0.3 % of the general population.
- More in females, particularly in the 5th & 6th decades of life.
- Familial predisposition & sometimes a part of multiple endocrine neoplasia (MEN).

Etiology :

1. Single adenoma (92%).
2. Multiple adenomata (4%).
3. Hyperplasia (3%).
4. Parathyroid carcinoma (1%).
5. Rarely ectopic PTH producing cancers, especially that of lung, kidney & bladder.

Clinical presentation :

- Usually detected by laboratory investigations.
- Patients below 60 years present by urological manifestations.
- Patients above 60 years present by neuromuscular & psychiatric manifestations.
- In general 80% present with renal involvement, 35% skeletal involvement.

1. The earliest complaints : muscle weakness, anorexia, nausea, constipation, polyuria, & polydipsia.

2. Renal presentation :

- Nephrolithasis (30_80%).
- Nephrocalcinosis (5_10%), irreversible & may lead to renal failure & hypertension.
- Renal stones with its presentation.

3. Bone diseases : Subperiosteal resorption especially of the phalanges & in severe cases bone cysts.

4. GIT manifestations : Peptic ulcer.

5. Emotional disturbances.

6. Articular & soft tissue manifestations : Chondrocalcinosis (3_7%).

7. Hyperparathyroid crisis : In severe hypercalcemia (16_20mg/dl). It presents by rapidly developing muscular weakness, nausea, vomiting, weight loss, fatigue & drowsiness.

D.D of hypercalcemia :

1. Primary hyperparathyroidism.

2. Malignancies :

- Group I (30%) : Hematologic malignancies as multiple myeloma & lymphomas.
- Group II (50%) : Solid tumors with lytic bone metastasis as cancers of lung, kidney & pancreas.
- Group III (20%) : Ectopic PTH producing cancers, especially that of lung, kidney & bladder.

3. Others :

- Milk-alkali syndrome.
- Hyperthyroidism.
- Paget's disease of bone.
- Vitamin D intoxication.
- Sarcoidosis.

Laboratory diagnosis :

1. Serum calcium : Total level is elevated except in cases of severe hypoalbuminemia.
2. PTH essay : Elevated level but in presence of hypercalcemia only.
3. Serum chloride to phosphate ratio above 33 suggests hyperthyroidism.
4. Increased urinary calcium 24 hours.
5. Increased serum alkaline phosphatase with skeletal lesions.

Localization :

- The most accurate method is neck surgical exploration.
- Preoperative methods include :

1. High resolution U/S with 76% accuracy.
2. CT scan has 50% accuracy & is helpful in medistinal lesion.
3. Thalium-Technetium scan (thyroid-parathyroid subtraction scan) : 72% accuracy.
4. In patients with previous neck exploration, **selective venous sampling** at certain points along big veins for serum PTH.

Treatment :

- **Treatment is surgical.**

1. Parathyroid adenoma : The adenoma is excised. Operative localization can be done by giving radioactive technetium & tracing the highest radiation density in the neck by a hand–held probe. The others are exposed to ensure that they are of normal size. One of them is biopsied.

2. Parathyroid hyperplasia :
(a) Subtotal thyroidectomy : i.e. excision of three & half glands.
(b) Other alternative include : total parathyroidectomy with heterotropic auto-transplantation of very thin slices in the forearm muscles. Postoperative hypercalcemia requires removal of few parathyroid slices.

- 80% of parathyroid tumors are attached to the posterior capsule of thyroid gland, symmetrically placed & overlie the recurrent laryngeal nerve.
- The superior glands are placed far dorsally at level of upper 2/3 of thyroid lobes.
- The tissues inferior to thyroid lobe are cleaned down to trachea to expose recurrent laryngeal nerve.
- It is helpful to follow the branches of the inferior thyroid artery especially if one is enlarged.
- If both lower glands cannot be found, the thymus pedicle should be carefully examined.
- The glands should be gently handed as their blood supply is easily damaged & the field should be bloodless to allow for search.
- Postoperative care : serum calcium should fall to normal within 48 hours. If hypocalcemia occurs, oral calcium supplementation or even IV calcium can be administrated.

Secondary & tertiary hyperparathyroidism

Secondary hyperparathyroidism : Occurs in :
1. Renal osteodystrophy.
2. Malabsorption syndrome.
3. Rickets & osteomalacia with low serum phosphorus.

Tertiary hyperparathyroidism :

- Prolonged chief cell hyperplasia cause them to be autonomus with elevated serum PTH & calcium.

Treatment of both : Essentially medical :
1. Vitamin D.
2. Calcium & phosphate binders.
3. Surgery in failure response to medical treatment by removal of 50 gm of parathyroid or 15 slices 1mm each.

Breast Diseases

The Breast

Congenital Anomalies

Congenital anomalies of the breast may involve the nipple or the breast itself.

Anomalies of the nipple :

1. Athelia : Absence of the nipple is very rare and is usually associated with absence of the breast (Amazia).

2. Polythelia : Supernumerary nipples occur anywhere along either or both the mammary ridges (mammary lines) which extend from the axillae to the groins. An accessory nipple may be mistaken for a mole or a wart.

Anomalies of the breast :

1. Amazia : Absence of the breast is usually unilateral and is often associated with absence of the sternal portion of the pectoralis major.

2. Polymazia : Supernumerary breasts are due to persistence of extramammary portions of the mammary ridges. They may occur in the axilla (accessory breast), groin, or even the thigh. They may function during lactation.

3. Small breast (Micromazia) : Treatment : Augmentation mammoplasty.

4. Diffuse Hypertrophy : Treatment : reduction mammoplasty.

5. Infantile gynecomastia : Diffuse enlargement of the breast, which may be unilateral or bilateral. It is caused by the effect of circulating maternal sex hormones. The condition is usually reversible within 6 months and therefore requires no treatment.

Inflammations of the breast (Mastitis)

Acute lactational mastitis & breast abscess

Incidence : acute mastitis is most common during lactation.

Etiology :

- **Organism :**

Staphylococcus aureus : It induces clotting of milk in the ducts producing obstruction and stasis.

- **Route of entry :**

1. Organisms from the mouth of the suckling infant gain access through nipple cracks and openings of lactiferous ducts.
2. Much less common is the blood born infection.

- **Predisposing factors are :**

1. Milk engorgement caused by inspissated milk and epithelial debris that block the ducts.
2. Abrasions to the nipples.
3. Poor hygiene.

Pathology :

- Infection is at first diffuse.
- The disease usually starts by milk engorgement, which if not properly treated will lead to acute mastitis.
- Staphylococci tend to produce necrosis and pus formation causing a **multilocular abscess.**

Clinical Features :

[A] Stage of milk engorgement :

1. **Pain :** Dull aching.
2. **Fever :** Persistent pyrexia.
3. **Examination reveals :** Enlargement and induration of the breast but there are no physical signs of inflammation.

[B] Stage of acute mastitis :

1. **Pain :** Gets worse.
2. **Fever :** Continuous higher pyrexia.
3. **Examination reveals :** Diffuse swelling, redness, induration and tenderness.

[C] Stage of acute abscess : Suppuration is diagnosed if :

1. **Pain :** Throbbing.
2. **Fever :** Hectic.
3. **Examination reveals :**
   - Physical signs get localized in the breast.
   - Pitting edema is elicited in the area of the skin overlying the abscess.
   - Persistence of the local signs of inflammation for more than 5 days or severe systemic upset for more than 2 days after full antibiotic treatment.

Breast abscess is one of the sites where the surgeon should not wait for fluctuation to diagnose it.

[D] Stage of chronic breast abscess.

Differential Diagnosis : The most serious is acute mastitis carcinomatosa.

Investigations : Total leucocytic count will show leucocytosis.

Treatment :

[A] Before the development of an abscess :

- The condition can be medically treated.

1. **An antibiotic** against staphylococci e.g. flucloxacillin, cephalosporin.
2. **Support of the breast** helps to lessen pain.
3. **Local heat.**
4. **The advisability of weaning is controversial, but a reasonable approach is to :**
   - Advise cessation of breast-feeding if the baby has been nursed for more than 9 months. The agents in common use are bromocriptine (Parlodel) 2.5 mg twice daily or stillbesterol 10 mg three times / day.
   - If the baby is younger, the patient is asked to use the healthy breast for feeding and to regularly empty the inflamed breast by squeezing and by a breast pump. After resolution of infection the baby can be refed by both sides.

[B] Abscess formation is treated by incision and drainage :

- **Anesthesia :** general.
- **Incision :** A radial or more cosmetic circumferential over the most tender area to release the pus, which is sampled for culture and sensitivity.
- The surgeon's finger **breaks down the septa** between the loculi to form a single cavity.
- **A drain** is brought out through the wound or at the most dependant part of the breast through a **counter incision.**
- **Antibiotic** therapy may be continued for a few days.

N.B: some surgeons advise in selected cases of breast abscess repeated aspiration guided by ultrasonography.

Non-lactational mastitis

- The commonest type of non-lactational mastitis is that which complicates mammary duct ectasia.
- Inflammation produces a painful tender periareolar swelling.
- Anaerobes are commonly responsible for mastitis. Therefore, it is customary to use metronidazole (400mg 4 times per day) with flucloxacillin (250 mg 4 times per day) for 5 days.
- If an abscess forms, it needs surgical drainage.
- Mammilary fistula is a possible complication that requires surgical excision.

Pubertal Mastitis

- This condition affects adolescent boys leading to pain and swelling of the breast, which becomes indurated and slightly tender, but suppuration never occurs.
- The tenderness usually subsides in two years, but may persist for a prolonged period.

Chronic Inflammatory Conditions of the Breast

1. Mammary duct ectasia (plasma cell mastitis).

2. Chronic breast abscess :

- **Etiology :** Acute abscess, which was treated by, prolonged use of antibiotics or was drained by a small incision.
- **Clinical picture :** See carcinoma.
- **Treatment :** Treatment is excision not only simple incision.

3. Tuberculosis of the breast :

- **Clinical picture :** see carcinoma.
- **Treatment :** Is by antituberculous drugs. Mastectomy is indicated in patients with resistant infection.

Mammary Duct Ectasia

- The disease is characterized by dilatation of the major ducts, which fill with creamy secretion.
- The etiology is unknown.

- Duct ectasia may be asymptomatic or may present by one of the following :
  1. **Nipple discharge :** blood stained, serous, creamy white or yellow.
  2. **Retraction of the nipple :** due to shortening of the ducts.
  3. **Acute inflammation**
  4. **Recurrent and chronic inflammation :** see breast carcinoma.

- **Treatment :**
  Troublesome cases of duct ectasia are treated surgically by **excision of the major ducts** through a circumareolar incision.

Fibrocystic disease of the breast

Incidence :

- **This is the most frequent disorder of the breast.**
- The upper outer quadrant of the breast is the commonest site of affection.

Other names :

Mammary dysplasia, **ANDI** (aberrations of normal development and involution), fibroadenosis.

Etiology :

- The exact cause is not known, but the prevalence of the disorder in women aged 30-50 years and its rarity after the menopause suggests that it is related to ovarian activity.

Pathology :

Mixture of the following with a variable proportion :

1. Adenosis : Glandular hyperplasia- causing multiplication of acini.

2. Epitheliosis : Solid epithelial hyperplasia within small ducts. In rare cases there is **"Atypical epithelial hyperplasia"** which is accompanied by higher possibility of developing breast cancer.

3. Fibrosis : When fibrosis is extensive it clinically resembles schirrhous carcinoma, and is called **"sclerosing adenosis".**

4. Cyst formation :

- **Structure :**

(a) Lined with epithelium & are filled with clear yellow or sometimes brownish fluid.
(b) Cyst may be large, distended with clear fluid and the cyst is now called (Blue doomed cyst of Bloodgood)

- **Etiology :**

(a) Abnormal involution of the supporting connective tissue in the lobules allows the acini to dilate.
(b) Obstruction of the ducts by papillomatosis and by the surrounding fibrosis.

Clinical Features :

1. May be asymptomatic.

2. Accidentally felt lump. This may be caused by a prominent cyst, aggregation of small cysts, or by the sclerosing adenosis.

3. The most frequent complaint is of multiple, sometimes painful, small lumps that may be unilateral or bilateral **(painful nodularity).** The lady usually observes this in relation to her cycles; on examination they are felt by **fingertips** not by flat of the hand.

4. Mastalgia (breast pain) : Usually cyclical. The pain typically occurs few days premenstrually and is accompanied by enlargement and increased nodularity of the breasts.

5. Nipple discharge : Is usually clear or yellow, but sometimes brown or green.

Differential Diagnosis :

1. Other causes of breast mass.
2. Other causes breast cysts.
3. Other causes of mastalgia.

Investigations :

- Investigations are usually not required, but the following are indicated to rule out breast cancer in suspected cases :

1. Ultrasound and mammography : show the cysts.
2. A cyst is fully aspirated. It is considered benign if :
(a) The fluid is not blood stained.
(b) The mass disappears completely after aspiration.
(c) Does not recur within two weeks.
(d) Cytology of the fluid may be performed to exclude malignancy, but the results are not conclusive.
3. A solid mass is aspirated for cytology.
4. Open biopsy : is indicated if cytology is not available, or if its result is not conclusive.

Treatment :

Exclusion of malignancy and reassurance of the patient are the most important.

- **No symptoms :** No treatment.
- **Cysts :** Are treated by aspiration. A recurring cyst is excised for biopsy.
- **Cyclic mastalgia :**

1. In mild cases : reassurance and wearing (night and day) a brassiere is usually enough.
2. Giving up caffeine consumption (coffee, tea and chocolate) may be useful.
3. Analgesics : NSAID's
4. Prolactin inhibitor : as bromocriptine 2.5 mg b.d. gives good results in many patients.
5. Danazol : which is a synthetic androgen, is effective in controlling cyclic pain.

- **N.B. Cases with atypical epithelial hyperplasia need regular medical follow up.**

Cysts of the breast

Classification :

[A] Acinar Cysts : (From the duct system)

1. Retention cysts : See fibroadenosis.
- Large cysts distended with clear fluid may have a blue green color (blue domed cyst of Bloodgood).
- It represents more than 75% of breast cysts.

2. Galactocele (milk cyst) :
- Develops during or shortly after lactation from obstruction to one of the principal ducts.
- The cyst forms a solitary painless swelling situated close to the nipple; a little milky fluid may be expressed from the nipple by pressure on the cyst.

3. Intracystic papilliferous carcinoma.

[B] Inter-acinar cysts : (in the stroma)

- **Cysts arising in the stroma are rare and include :**

1. Dermoid cyst.
2. Lymph cyst.
3. Blood cyst : arise either as traumatic hematoma or due to bleeding into a cyst.
4. Hydatid cyst.
5. Sebaceous cysts.

Treatment:

1. Aspiration : Mention criteria of benign and suspicious cyst.
2. Excision : Retention cysts, which recur after aspiration, and all other cysts should be excised.

Nipple discharge

Etiology :

[A] Physiological :

1. Milk production with lactation.
2. Serous discharge during pregnancy.

[B] Pathological :

Cause	Nature of discharge
Duct ectasia (the commonest)	Serous, creamy white, yellow, blood stained, brown-green or black
Fibrocystic disease	Clear, yellow, brown or green
Duct papilloma	Blood or blood stained discharge from the duct
Duct carcinoma	Rare cause of blood or blood stained discharge from one duct
Contraceptive pills	Serous or milky discharge from multiple ducts
Hyperprolactinemia	Milky discharge (galactorrhea) from multiple ducts

Diagnosis : History and examination should provide the following information :

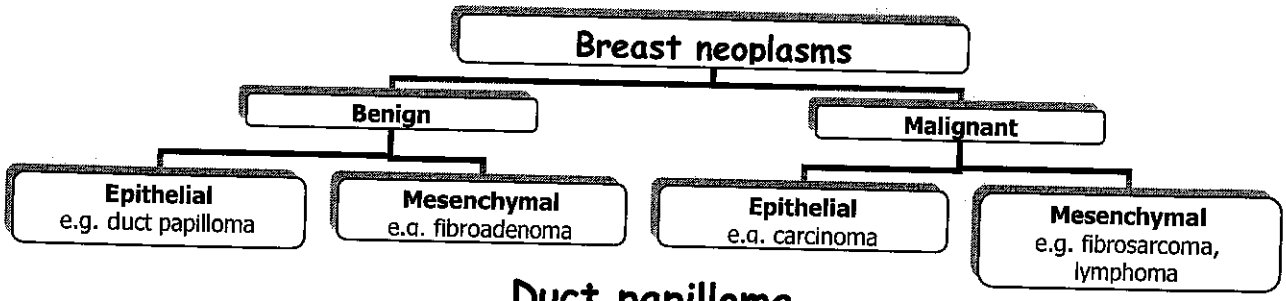
1. Nature of discharge.
2. Association with a mass.
3. Unilateral or bilateral.
4. Single duct or multiple duct discharge (Differential pressure test).
5. The use of contraceptive pills.

Investigations :

1. Test for occult blood : in the discharge if it is not apparent.
2. Cytology : of the discharge for exfoliated cancer cells.
3. Soft tissue mammography.
4. Duct galactography : in case of single duct discharge.
5. Serum prolactin estimation : in suspected cases of galactorrhea.
6. Ultrasound : to detect solid or cystic swelling.
7. Biopsy : if mass was detected with mammography or ultrasound.

Treatment :

1. A palpable mass should be **excised** for histology and treated accordingly.
2. A single duct bloody discharge calls for excision of this duct **(microdochectomy).** A needle with a blunt tip is introduced in to the affected duct & acts as a guide for the surgeon. This duct, with a rim of the surrounding tissues is excised and is sent for histological examination.
3. Discharge from multiple ducts with no palpable or mammographic mass is treated **conservatively by observation.** Rarely the discharge is persistent and troublesome where it is surgically treated by **excision of the major ducts.**



Duct papilloma

- A duct papilloma is usually situated in one of the main ducts near the nipple in a young woman.

Clinically : 1. Bloody or blood stained discharge (the commonest).
2. Retention cyst : Behind the mass.
3. Palpable mass : Pressure on the mass may produce discharge.

Ductography : Shows filling defect.
Treatment : Is by excision of the affected duct (microdochectomy).

Fibroadenoma

- Fibroadenoma is the commonest breast mass of young women. The usual age is between 15-30 years.

Pathology :

A fibroadenoma is a benign mixed neoplasm of the breast, which affects both the fibrous and the glandular tissues, but the fibrous element predominates.

Grossly :

- The tumor may be solitary or multiple, firm, with smooth surface that may be lobulated in big lesions. It is well circumscribed and is never attached to surrounding tissue.
- The cut surface reveals lobules of whirled white fibrous tissue, which bulges out of its capsule.

Histologically : There are two types :

- **Pericanalicular (hard) fibroadenomata :** Are the usual form. These tumors are formed mainly from fibrous tissue that surrounds a few small tubular glands. They tend to be small.
- **Intracanalicular (soft) fibroadenomata :** Contain more glands. They are usually larger and softer than the pericanalicular type.

Clinical Features :

- **Age :** Hard fibroadenoma occurs commonly in young women 20-30 years of age while soft fibroadenoma occurs between the ages of 30-50.
- **Painless lump** accidentally discovered usually small (few centimeters), not tender, well circumscribed with smooth surface that may be lobulated with high mobility within the breast **" breast Mouse ".**

Items	Hard fibroadenoma	Soft fibroadenoma
Frequency	Commoner	Less common
Age	20-30 years	30-50 years
Size	Small	Large
Shape	Rounded	Lobulated
Consistency	Firm	Softer
Mobility	Free (breast mouse)	Restricted
Complications	Never turn malignant	Liable to turn to sarcoma

Investigations : Soft tissue mammography reveals well-circumscribed lesion.

Treatment : Treatment is by **excision** and histological confirmation of the diagnosis.

Cystosarcoma Phylloides

Pathology : Highly cellular type of fibroadenoma that tends to grow rapidly.

The name : It was so named by Brodie who used the term phylloides because the cut surface resembles a leaf.

- The term Cystosarcoma however is a **misnomer** as many are not cystic and it is rarely malignant.
- It is therefore better termed **Phylloides tumor.**

A big tumor may cause skin ulceration. The tumor is not attached to the skin.

Treatment : Is by wide local excision to prevent recurrence. If the tumor is occupying the whole breast, simple mastectomy is indicated.

[A] Carcinoma of the ducts

1. Non-infiltrating (ductal carcinoma in situ) (6%)

- **Non-infiltrating comedocarcinoma :** Central necrosis of cancer cells leads to the extrusion of a sebaceous like material from the cut surface of the tumor.
- **Non-infiltrating papillary duct carcinoma :** May cause bleeding from the nipple.

2. Infiltrating ductal carcinoma

- **Schirrhous carcinoma : (70-75%) (NOS = Not Otherwise Specified)**

The tumor : Is hard. The cut surface is concave, rough, gritty and pale gray.
Microscopically : All degrees of differentiation from anaplasia to well differentiated tumors with attempts at adenoid formation may occur.
Spread : Early lymphatic spread.

- **Encephaloid carcinoma : (10%)**

The tumor : Is Large, soft and brain like.
Microscopically : It contains less fibrous tissue than **NOS**. The tumor shows lymphocytic infiltration.
Spread : To lymph nodes occur later than **NOS**. The prognosis is better than **NOS**.

- **Mucinous carcinoma : (3%)**

The tumor : Has a gelatinous consistency and is usually bulky.
Microscopically : Mucin is produced by the tumor cells and is present either intra or extra cellular.

- **Inflammatory carcinoma (mastitis carcinomatosa) :**

The tumor : Is a rare type, but is the most malignant. It is a fulminant form that resembles mastitis.

[B] Carcinoma of the lobules

- **Non-Infiltrating lobular carcinoma : (0.7 %)**

Frequently multicentric.

- **Infiltrating lobular carcinoma : (7.5 %)**

1. Grossly and microscopically : The tumor is indistinguishable from scirrhous carcinoma and the diagnosis can be certain only when the preinvasive lobular nodule of a tumor can be identified histologically.
2. Bilateral in 25 % of cases.

[C] Paget's disease of the nipple (1%)

1. Nature : An intraduct carcinoma which begins in the epithelium of a main duct and spreads to the skin of the nipple and down into the breast substance.
2. When the nipple epithelium is infiltrated, it produces nipple erosion. In early stages a mass may not be palpable, a mass may only appear after 2 years from the start of the disease.
3. Histologically : there are :
(a) Paget's cells : Clear vacuolated cells, with small dark staining nuclei, which occur alone or in clusters in the deep layers of the epidermis.
(b) Hyperplasia of all layers of the epidermis.
(c) Round cell infiltration of the dermis.
N.B:James Paget(1809-1892) is a pathologist-university of cambridge.

Hormone receptors :

- About 60% of breast cancers have receptors for estrogen & are termed ER-positive.
- These tumors are hormone dependant & respond more to hormonal treatment.
- Tumors may also have progesterone receptors.
- Recently: tumors are stained for ***c-erb 52*** (growth factor receptor). if proved to be positive,monoclonal antibodies against these receptors could be used.

N.B: *li-fraumini syndrome:* Mutation in tumor suppressor gene p53 leading to: Malignancies in breast, ovary, colon and lymphomas.

Spread :

- Previously it was thought that breast cancer spreads locally at first, then by lymphatics and lastly by the blood stream.
- This view is no longer accepted and is now well realized that carcinoma of the breast may spread by the blood stream very early producing **micro-metastases** in distant organs.

[A] Local spread :

- Can occur through the breast substance, overlying skin- underlying pectoralis major and serratus anterior muscles and the chest wall.

[B] Lymphatic spread :

- **By embolism and permeation** is mostly to the axillary nodes, next common is the internal mammary chain.
- **Involvement of the supraclavicular node** is considered an advanced disease.

1. Axillary lymph nodes are classified into 3 levels :

- **Level I** : Below the pectoralis minor muscle.
- **Level II** : Behind the pectoralis minor muscle.
- **Level III** : Above the pectoralis minor muscle.

2. Mammary lymph nodes lying along the internal mammary vessels in the first three intercostal spaces. They receive part of the lymph from the medial half of the breast.

3. A few lymphatics pierce the pectoralis major muscle to drain into the inter-pectoral L.N. **(lymph nodes of Rotter)** and pass to the posterior intercostal lymph nodes.

4. Lymphatics from the lower inner quadrant : may pierce the rectus sheath to reach the peritoneal lymphatic plexus.

5. Supraclavicular lymph nodes : may be affected by retrograde lymphatic spread.

6. Obstruction of the dermal lymphatics by malignant cells produces breast skin edema that is marked in the dependant part, i.e. the lower half of the breast. The edematous skin is pulled at the sites of hair follicles, sebaceous glands and sweat glands giving the appearance of an orange peel, hence the French name **Peau d' Orange.**

[C] Blood stream spread :

- Produces metastasis in the lungs, bones, brain and liver.

N.B: part of the venous drainage of the breast goes to intercostal veins which drain into the azygos system and communicates with valveless vertebral venous plexus. This explains the tendency of breast cancer deposits to affect the axial skeleton

Staging :

[A] International TNM staging :

T = Tumor

Tis : Carcinoma in situ. Paget's disease with no palpable tumor.
T0 : No evidence of primary tumor.
T1 : 2 cm diameter or less.
T2 : 2-5 cm diameter.
T3 : Tumor larger than 5 cm.
T4 : Any size with direct extension to chest wall or to skin.

N = Nodes

N0 : No palpable axillary nodes.
N1 : Mobile palpable homolateral axillary nodes.
N2 : Fixed homolateral axillary nodes.
N3 : Palpable homolateral supraclavicular nodes. Edema of the arm.

M = Metastases

M0 : No evidence of distant metastases.
M1 : Distant metastases.

[B] Staging of the UICC (union international center cancer) is now internationally approved. It is based on the TNM staging:

Stage UICC		description	category	5 year survival (%)
I		T1,N0,M0	Early breast cancer	93
II	IIA	T2,N1,M0	Early breast cancer	72
	IIB	T3,N0,M0		
III	IIIA	T1-3,N0-2,M0	Locally advanced Breast cancer	41
	IIIB	T4,any N,M0		
IV		Any T,any N,M1	metastatic	18

[C] Manchester classification :

Stage I :
- A mobile mass in the breast.

Stage II :
- A mobile mass in the breast with or without skin tethering.
- Palpable mobile homolateral axillary nodes.

Stage III :
- Peau d' orange larger than the tumor but still limited to the breast.
- Tumor fixed to pectoral muscle but not to the chest wall.
- Homolateral axillary lymph nodes matted together or fixed to chest wall.
- Homolateral supraclavicular lymph nodes.
- Edema of the arm.

Stage IV :
- Skin involvement wide of the breast (cancer en cuirasse).
- Fixation to the chest wall.
- Distant metastases.
- Involvement of the opposite breast or axilla.

Clinical Features :

I- Symptoms :

1. The patient accidentally notices **a painless lump in the breast.** An accidental trauma to the breast may attract the attention of the patient to presence of the lump.
2. Much less frequently the disease is discovered because of **mild breast pricking pain, nipple retraction** or **blood stained nipple discharge.**
3. Picture of metastasis (lymph node or blood).
4. The disease may be discovered by **routine screening mammography.**

II- Signs : Some of the following signs may be detected :

[A] Breast :

1. Asymmetry. 2. Enlargement.
3. Skin dimpling. 4. Skin puckering.

- **Dimpling and puckering of the skin are evident when the patient is sitting and elevates her arms.**

5. Peau d'orange (orange peel). 6. Skin nodules. 7. Skin ulceration.

- **Peau d'orange, skin nodules, and ulceration indicate a locally advanced disease.**

[B] Mass :

1. Hard. 2. Irregular. 3. Ill-defined.
4. Restricted mobility within the breast substance.
5. Fixation to the skin, underlying muscles, or chest wall, if present, is diagnostic of carcinoma.

[C] Nipple :

1. Recent retraction. 2. Change of direction.

[D] Axillary and supraclavicular nodes :

- **Number and mobility of palpable nodes are assessed.**

[E] Distant metastases :

1. Chest examination. 2. Hepatomegaly. 3. Ascites.
4. Pelvic examination for hard deposits or Krukenberg tumor.

Skin manifestations of breast cancer

1. Dimpling and puckering of the skin : Overlying the tumor. This is the earliest manifestation of superficial tumors. It is due to infiltration of Cooper's ligaments by the tumor, pulling the skin toward the tumor.(Cooper's ligament : Are conical fibrous tissue normally connecting the skin to the pectoral fascia).
2. Fixation.
3. Fungation.
4. Ulceration : Ulcer is always indurated.
5. Skin nodules : Near or far from the tumor, even around the umbilicus. They are due to lymphatic spread.
6. Peau d' orange: non-pitting edema due to obliteration of the skin lymphatics by malignant permeation & surrounding fibrosis. The pits giving skin the orange appearance are due to hair follicles, sweat glands & sebaceous glands anchored to the skin.
7. Cancer en cuirasse : In very late cases, due to malignant infiltration and fibrosis of the skin as well as lymphatic obstruction, the skin is very hard, very thick, and non-mobile like a shield.
8. Nipple : Is retracted.
9. Paget's disease of the nipple.

III- Special clinical forms :

1. Paget's disease of the nipple : (1 % of breast cancers)

- The first symptom is often an abnormal pricking sensation of the nipple, with superficial erosion.
- A tumor mass may not be palpable.
- The lesion is commonly mistaken for eczema.
- The diagnosis is established by biopsy of the erosion

	Paget's disease	Eczema
1	Unilateral	Commonly bilateral
2	Usually occurs at menopause	Commonly occurs at lactation
3	No itching	Itching
4	No vesicles-not oozing (dry)	Vesicles-oozing
5	Nipple is eroded	Intact nipple
6	Well-defined margin	Ill defined margin
7	A breast lump may be felt	No lump
8	No response to eczema treatment	Responds to treatment
9	Starts in the nipple	Starts in the areola

2. Inflammatory carcinoma :

- This is a rare aggressive form of breast cancer.
- Usually occurs during pregnancy or lactation.
- There is a rapidly growing, sometimes painful, breast swelling.
- The overlying skin becomes red, edematous and warm. Often there is no distinct mass, since the tumor infiltrates the breast diffusely.
- The picture clinically resembles acute mastitis.
- **Prognosis :** is poor as it is usually advanced at the time of diagnosis.

Inflammatory carcinoma	Acute bacterial mastitis
Gradual onset with no or low grade fever	Acute onset with high fever
Progress is slower	Rapid progress
Involves more than one third of the breast	One breast sector is affected
Skin is dusky red	Skin is rosy or bright red
Mildly tender or non-tender lesion	Markedly tender lesion
Non-tender axillary nodes	Tender axillary nodes
No response to antibiotics in one week is an indication for biopsy	The lesion is either cured by antibiotics or forms an abscess

3. Carcinoma in situ:

	Duct carcinoma in situ (DCIS)	Lobular carcinoma in situ (LCIS)
Frequency	More common	Less common
Bilaterality and multicentricity	Rare	common
Microcalcifications	Present	absent
Early detection	Possible	Less likely
Potential for invasive cancer	30-50%	It is a marker of increased risk of malignancy in the same or other breast
Treatment	As invasive cancer	Strict follow up.

Differential Diagnosis :

I- Case presenting by a lump (mass):

Items	1. Carcinoma	2. Solitary cyst	3. Fibrocystic disease	4. Fibroadenoma
Age	Usually > 35 yrs	35-55 years	20-55 years	15-30 years
Pain	Painless	Occasionally painful	Occasionally painful	Painless
Surface	Irregular	Smooth surface	Indistinct surface	Smooth may be lobulated
Consistency	Stony hard	Fluctuation is difficult to elicit, so it feels soft to hard	Firm, ill defined areas of thickening	Firm, highly mobile
Lymph nodes	Probably axillary node enlargement	Free axilla	Free axilla	Free axilla

The above lesions constitute 95 % of breast lumps.

Other less common causes are :

5. Traumatic lesions : hematoma and fat necrosis.

Breast hematoma	Traumatic fat necrosis
• If there is no external bruising, a deeply seated old hematoma may form a hard mass that greatly resembles a carcinoma.	• Blunt breast trauma may cause death of some of the fat cells. • The liberated fatty acids combine with calcium to form calcium soaps. • The result is one of two forms : 1. A cyst that contains thick oily fluid is formed. 2. Less frequently a hard mass, which resembles a carcinoma, is formed.

6. Inflammatory lesions : Chronic abscess, mammary duct ectasia & T.B.

Chronic breast abscess	T.B.	Mammary duct ectasia
• This is the result of improper treatment of acute abscess. • There may be (with the mass) nipple retraction and skin puckering. • However it is more painful than carcinoma and is accompanied by low-grade pyrexia.	• Presents as : 1. Multiple cold abscesses & sinuses. **OR** 2. Multiple nodules in the breast substance.	• The affected area is hard & may be associated with skin dimpling & nipple retraction simulating carcinoma. • It is called **plasma cell mastitis** due to predominance of plasma cells.

7. Other cysts : e.g. Galactocele.
8. Other tumors : Duct papilloma accumulating blood behind it and sarcoma (very rare).
9. Swellings arising from the chest wall : Tuberculosis or tumor of a rib or lipoma.

II- Paget's disease : Should be differentiated from eczema of the nipple.

III- Inflammatory carcinoma : Should be differentiated from an acute breast abscess.

IV- Nipple retraction : Two types of nipple retraction should be identified :

- **Longstanding type** dates back to the puberty is an innocent finding. It poses difficulty only during lactation. It predisposes to mastitis and resolves spontaneously after lactation.
- **Recent nipple retraction** in womanhood is considered seriously. One cause is carcinoma. Other causes are *mammary duct ectasia, TB* and *chronic breast abscess.*

Investigations :

[A] Laboratory: CEA and Cancer antigen CA 15-3.it is prognostic rather than diagnostic.

[B] Mammography :

- Is soft tissue radiology of the breast. In expert hands it is 95% accurate in diagnosing breast cancer.
- It is useful in detecting multifocal lesions in the same or other side.
- A cancer appears as a dense opacity containing microcalcifications and has an indefinite outline.
- **N.B1:** only 20% of microcalcifications are malignant.

N.B2: xero-radiography: as mammography but more accurate.

[C] Ultrasonography:

- A speculated hypo-echoic mass more deep than wide suggests a malignant lesion.
- It differentiates between chronic abscess & carcinoma.
- It differentiates between solid & cystic lesions.
- Malignant lesion receives blood flow from all around with turbulent speed.
- It is particularly useful in young women inwhom mammography is not helpful.

[D] Magnetic resonance imaging (MRI): it is indicated with contrast in certain situations ***e.g.***

- Postoperative scarring to differentiate between fibrosis and local recurrence of malignancy.
- After neoadjuvant therapy to monitor response.
- In the presence of breast implants

E] Biopsy:

1. **Excision biopsy:** Is the most reliable and provides a big specimen to allow for hormone receptor estimation as well.
2. **Frozen section biopsy :** The biopsy is frozen and slides are prepared from the frozen block. A diagnosis is obtained within 20 minutes. Patient is kept under general anesthesia. If the result is positive, the surgeon proceeds with radical surgery. Consent for mastectomy should be obtained preoperatively.
3. **True-cut biopsy:** done under local anesthesia with a special needle that cuts a core of tumor tissue.
4. **Fine needle aspiration cytology (FNAC) :** It depends on examination of cells to detect criteria of malignancy in them-The aspiration can be done in the outpatient clinic using fine needle. A skilled cytologist is needed.
   - Advantages of FNAC :
     1. Very simple, inexpensive and accurate procedure.
     2. Give a definite diagnosis in 90 % of cases.
5. **Biopsy from impalpable breast masses:** the radiologist can place a wire inside the lesion under mammographic guidance. At operation, the mass is removed with the wire for histological assessment.

[F] Aspiration of a cyst :

- **A breast cyst is considered benign if :**
  1. The fluid is not blood stained.
  2. The cyst disappears completely with aspiration.
  3. Does not recur within 2 weeks.
  4. In case of doubt, the fluid is subjected to cytological examination.

[G] Investigations to detect distant metastasis: routinely done by:

1. Chest x-ray.
2. Ultrasound examination of the pelvis and abdomen
3. Alkaline phosphatase (a high level indicates bone or liver deposits)
4. Further tests (needed in special situations):
   a) Bony pains require x-rays and isotope bone scan.
   b) Suspicion of cerebral secondaries is an indication for CT examination.
   c) PET (positron emission tomography) scan.

Triple assessment

It means comparing the results of

1) **Clinical examination**
2) **Mammography or ultrasonography**
3) **FNAC**

- ***If the three parameters are concordant, the surgeon can rely on the diagnosis.***

Early detection

1. Breast self examination :

All women over age of 20 years should be advised to examine their breasts monthly, one week after the menstrual period. The physician instructs the woman as how to conduct a systematic inspection and palpation.

2. Screening programs :

In some Western countries, high risk women are subjected to regular clinical examination and mammography, every one or three years.

Treatment

I. Early (potentially curable) breast cancer

This is defined as stages I and II in UICC staging.

[A] Surgery:

Different surgical options are available (all of them give almost equal results):

[1] Conservative therapy:

It includes:

1. Local wide excision with a 2cm safety margin. If the lesion is close to skin, part of it may be excised.
2. The sentinel lymph node: is the first node in the axilla to be affected. injection of patent blue violet or a radioactive sulphur colloid near the tumor will allow identification, excision and immediate pathological examination of the sentinel node
   - If the node is positive for metastasis: axillary clearance is done.
   - If the node is negative for metastasis: no further excision of lymph nodes.
3. Postoperative radiotherapy for 4 weeks directed to the breast.

Advantages :

1. If done for the properly indicated patients, the operation provides good results that are equal to radical mastectomy.
2. The breast is preserved minimizing psychological trauma.

Indications :

1. Small tumor ≤ 4cm.
2. Sometimes, large lesions (up to 5 cm) in large breasts.
3. Peripheral lesions.

Contraindications :

1. Pregnancy.
2. Large or central tumors in small breasts (no cosmetic advantages).
3. Multicentric disease as detected by soft tissue mammography.
4. Collagen vascular disease (poor tolerance to radiotherapy).
5. In situ breast cancer more than 20% due to the common incidence of multicentricity.

[2] Modified radical mastectomy (of Patey):

- It involves removal of the following structures in one block:
  - Skin ellipse over the tumor with at least 5 cm safety margin including the areola and nipple.
  - The whole breast including the tumor.
  - The pectoralis minor muscle is either removed or its tendon is cut to open the axilla.
  - All axillary lymph nodes and axillary fat medial to the axillary vein.

NB. Axillary vessels, axillary nerve, nerve to serratus ant. and nerve to latissimus dorsi are spared

After Patey's mastectomy radiotherapy is advised for patients with :
1. Positive axillary lymph nodes. 2. Tumors in the medial half of the breast.

[B] Adjuvant chemotherapy and hormonal treatment:

1. **Hormonal therapy: For all hormone receptor positive cases.**
   It reduces ipsilateral and contralateral breast recurrence by 40%.**tamoxifen** blocks estrogen receptors and **anastrazole** is an aromatase inhibitor which inhibits peripheral conversion of androgen to estrogen.
2. **Chemotherapy: is indicated for:**
   1. Positive axillary nodes.
   2. All patients below 70 years.
   3. Tumors more than 1 cm.
   4. Hormone receptor negative and Her2/neu positive tumors (denote aggressive tumors).
3. **Targeted therapy:** for Her2/neu positive tumors. Monoclonal antibodies are given against Her2/neu receptors **(herceptin)**.

[C] Follow up :

- After treatment, patients are reviewed at regular intervals, usually 3 months for the first 2 years, 4 monthly for the next 3 years, and annually thereafter. **This is required to :**

1. Detect and treat complications of mastectomy :

(A) Psychiatric morbidity : caused by loss of the breast.

(B) Arm edema results from :

- Excision of lymphatics.
- Lymphatic obstruction by radiotherapy.
- Lymphangitis caused by infection.
- Malignant axillary recurrence blocking them.
- Thrombosis of the axillary vein.

How to avoid ?

- Avoidance of radiotherapy to the axilla, which has been surgically evacuated of its nodes, reduces the possibility of lymphatic edema.
- The patient is warned to avoid minor trauma to the ipsilateral hand & should wear gloves when carrying out rough work in order to avoid infection & lymphangitis.

Treatment : (difficult) Arm elevation, massage & elastic or pneumatic arm compression are partially effective.

N.B1: brawny edema: hard but still pitting edema due to infiltration of lymph vessels.
N.B2: elephantiasis surgica: lymphedema secondary to block dissection of lymph nodes.
N.B3: stewart-treves disease: it is lymphangiosarcoma on top of long-standing lymphedema.

2. Detect local recurrence or distant disease.

3. To do annual mammography of the contralateral breast.

4. Ladies who are free of the disease after two years can be considered for reconstruction of breast using either a synthetic implant or a myocutaneous flap (e.g. Transverse Rectus Abdominus Myo-cutaneous flap TRAM). Some surgeons prefer immediate reconstruction for low risk patients.

5. Patients are instructed : Not to get pregnant for at least three years & to use non-hormonal contraception.

II. Treatment of Intermediate disease (locally advanced breast cancer)

- **It includes** tumors larger than 5 cm or fixed axillary or internal mammary nodes.
- Distant metastasis should be excluded.
- **Treatment:** Neoadjuvant (pre-operative) chemotherapy with the aim of down staging of the tumor. Then either conservative surgery or modified radical mastectomy is done according to the response to treatment.

III. Advanced (incurable) breast cancer

- It includes **stages III** and **IV** in UICC staging
- fungation, ulceration & inflammatory carcinoma are included in this category.
- **Endocrine therapy and chemotherapy are the two main lines of treatment.**
- Surgery and radiotherapy are of secondary value.

[A] Endocrine therapy:

Patients who are more likely to respond are :

1) Postmenopausal women.
2) Estrogen receptor positive tumors.
3) Progesterone receptor positive tumors.

The methods in common use are :

1. **Tamoxifen** (Nolvadex tablets): the first line hormone therapy. It is an antiestrogen that blocks estrogen receptors preventing the hormone from activating cancer cells. The dose is 10 mg twice daily. It is ***not*** given for more than 5 years to avoid risk of endometrial cancer or thrombogenecity.
2. **Aromatase inhibitors.**
3. **Raloxifen**:as tamoxifen with less side effects.

[B] Chemotherapy : *It is indicated for the following cases :*

1) Rapidly progressive disease.
2) Premenopausal women.
3) Receptor negative cases.
4) Hormone failure.
5) Liver metastases.

Combinations commonly used include : **C**yclophosphamide, **M**ethotrexate and 5-**F**lurouracil **(CMF).**

N.B: it can be used as a targeted therapy for Her2/neu positive cases.

[C] Radiotherapy: *It is indicated for the following cases:*

1. **Pain** whether due to bone or soft tissue involvement.
2. To control tumor fungation.

[D] Surgery : *It is indicated for the following cases :*

- *Radical mastectomy* may be done for advanced local disease (stage III). It provides good local control.
- *Simple mastectomy* to remove fungating tumor.

[E] Management of specific problems :

1. Pleural effusion : Commonly responds to systemic therapy and chest tube drainage. If not, local instillation of the cytotoxic bleomycin through the tube may be required.

2. Pathological fractures :
Internal fixation is used. The patient is given radiotherapy to the fracture site.

3. Spinal cord compression : requires urgent surgical cord decompression, with stabilization followed by radiotherapy.

4. Cerebral metastases : are treated by a combination of corticosteroids and radiotherapy. Occasionally a solitary brain metastasis is suitable for surgical excision.
The requirements are:
(a) Well-controlled breast primary. (b) A long life expectancy. (c) An accessible area in the brain.

5. Liver metastases : Chemotherapy.

6. Hypercalcemia :

Treatment: Is correction of dehydration by IV fluids + frusemide + prednisolone.

7. Superior vena caval obstruction : Requires urgent radiotherapy.

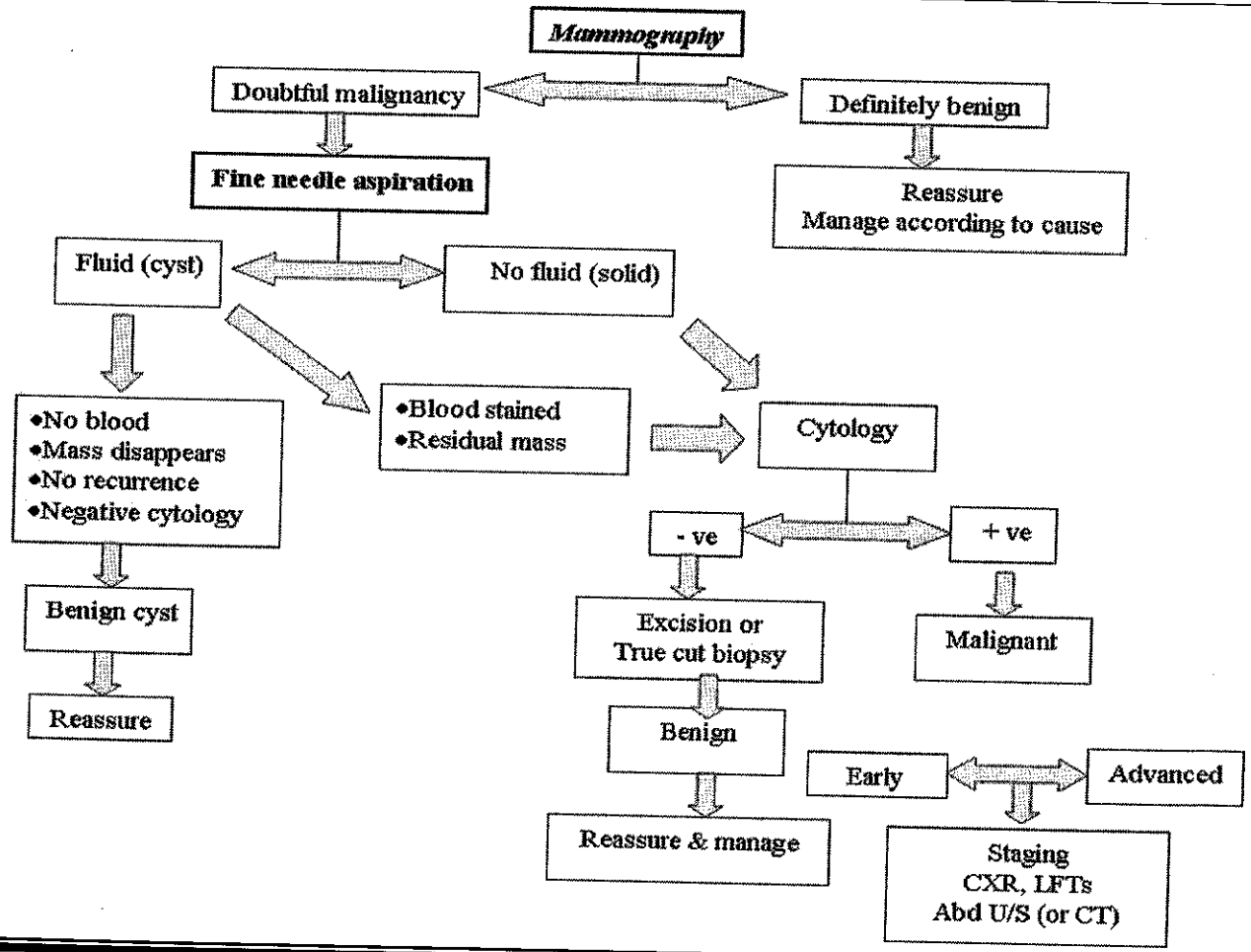
Prognosis:

➧ **Prognosis of patients with breast carcinoma depends on the following factors :**

1. **The type of the tumor:** The best prognosis is provided by the in situ carcinoma and Paget's disease, while the worst is the inflammatory carcinoma.
2. **The T stage of the primary tumor:** The higher the T stage, the worse is the prognosis.
3. **The site of the tumor:** Medial half tumors have a worse prognosis than those of lateral half due to early involvement of the internal mammary lymph nodes.
4. **The involved lymph nodes :** Size, mobility, number and location of the involved lymph nodes.
   Assessment of lymph node involvement depends on the histological examination :
   (a) Large fixed nodes are of bad prognosis.
   (b) The number of involved nodes largely affects the prognosis :
   - Patients with negative axillary nodes have a 10-year survival rate of 65 %.
   - Patients with 1-3 positive axillary nodes have a 10-year survival rate of 38%.
   - Patients with more than 4 positive axillary nodes have a 10-year survival rate of 13%.

   (c) The prognosis worsens the higher the level of the affected nodes in the axilla. High-level nodes carry a bad prognosis.
5. **The presence of distant metastasis**
6. **Hormone receptor status: Receptor positive tumors respond more often to hormonal therapy and have a better prognosis than those that are receptor negative.**

Investigations of clinically doubtful breast mass



Diseases of the male breast

Gynecomastia

Definition : Generalized enlargement of the male breast.

Etiology :

[A] Physiological :

1. **Infantile gynecomastia :** Due to circulating maternal sex hormones.
   - It usually resolves within six months.
2. **Pubertal gynecomastia :**
   - It occurs in up to 70% of normal pubertal boys.
   - It is often asymptomatic.
   - It usually resolves within 2 years.
3. **Senile gynecomastia :** Due to the reduction of testicular function.

[B] Secondary gynecomastia :

1. Reduced production of testosterone e.g. after orchidectomy, testicular atrophy.
2. High levels of estrogen caused by feminizing testicular tumors and suprarenal tumors.
3. Failure to metabolize estrogen in cases of chronic liver disease.
4. Drugs as estrogen, cimetidine, digoxin, spironolactone, phenothiazines. and cannabis.
5. Ectopic hormonal production in bronchogenic carcinoma.

Clinical features :

- **The usual complaint is breast enlargement** which may be unilateral or bilateral, and with or without tenderness.
- **Examination reveals a subareolar mass (a disc of tissue),** which is soft and mobile. Any induration, or fixation should raise the suspicion of malignancy (this is not a possibility in children and adolescents).
- **Examination should include the abdomen and testes.**

Investigations : These are not required for neonatal and adolescent cases.

1. Liver function tests.
2. Biopsy in suspected cases of malignancy.

Treatment :

- **The neonatal and adolescent types :** usually resolve spontaneously.
- **Secondary gynecomastia :** treatment of the underlying condition.
- **Persistent gynecomastia :** can be treated surgically by subcutaneous mastectomy.

Carcinoma of the male breast

- **Incidence :** male : female carcinoma = 1:100.
- **Staging & treatment :** similar to that for carcinoma of the female breast, but castration is the principal mean of hormonal control of advanced cases.
- **Prognosis :** is worse than that of female breast cancer as it rapidly becomes attached to the skin & chest wall.

Differential Diagnosis of Breast Pain

1. Itching pain :	a. Ordinary eczema. b. Scabies.
2. Burning pain :	Cracked nipple.
3. Acute dull pain :	a. Milk engorgement. b. Acute mastitis.
4. Throbbing pain :	Acute abscess (suppuration).
5. Dull ache related to menstruation :	Fibroadenosis.
6. Carcinoma is painless, rarely painful from :	a. Associated abscess. b. Nerve infiltration. c. Mastitis carcinomatosa.

Differential diagnosis of retracted nipple

[A] Congenital :

- Present since childhood. Nipple fails to develop.
- It is due to failure of the nipple to follow the growth of the breast.
- The retracted nipple possess difficulty only when the lady is lactating, and is a predisposing factor for the development of mastitis. The problem may be spontaneously corrected during pregnancy or lactation.

[B] Recent :

- Nipple was normal. Retraction resulted from excess fibrosis in :

1. **Carcinoma :** it is the most important cause.
2. **Chronic inflammation :** Chronic abscess, mammary duet ectasia and tuberculosis.
3. **Fibroadenosis :** rare.

Causes of huge breast

1. Sarcoma.
2. Mastitis carcinomatosa.
3. Encephaloid carcinoma.
4. Colloid carcinoma.
5. Soft fibroadenoma.
6. Diffuse massive hypertrophy.

Salivary Disorders

Salivary glands

Non-Neoplastic Salivary Gland Diseases

This term includes different disease groups, which are :

1. Congenital diseases : as ectopic parotid tissue, cystic hygroma & sialectasis.

2. Infections:

(a) Acute : viral, bacterial.

(b) Recurrent subacute or chronic.

(c) Rare : T.B.

3. Salivary stones.

4. Salivary fistula.

5. Degenerative diseases (sialectasis).

6. Autoimmune salivary diseases.

7. Drug-induced enlargement of the salivary glands has been reported e.g. iodide containing compounds, Thiouracil, hypotensive drugs & contraceptive pills.

8. Metabolic & endocrinal causes : include liver cirrhosis, diabetes mellitus, alcoholism &malnutrition.

Sialectasis

(Degenerative disease)

Definition : Sialectasis means abnormal dilatation of the small branches of the salivary ducts.

Etiology : Not known, but the childhood type is known to be a familial disease.

Clinical picture : The usual presentation is unilateral recurrent attacks of sialadenitis.

Investigations : Sialography "snow storm appearance".

Treatment :

1. Conservative: Stimulate salivary flow and massage the affected gland with antibiotics during attacks of inflammation.

2. Surgical excision : of the gland.

Autoimmune salivary diseases

1- Mikulicz's disease: Consists of enlargement of the salivary and the lacrimal glands and dryness of the mouth.

2- Sjogren's disease

(a) Primary Sjogren's disease : consists of dryness of the mouth (xerostomia) and dryness of the eye (keratoconjunctivitis sicca).

(b) Secondary Sjogren's disease : as well as rheumatoid arthritis, salivary gland discomfort, but only 30 % have sialomegaly.

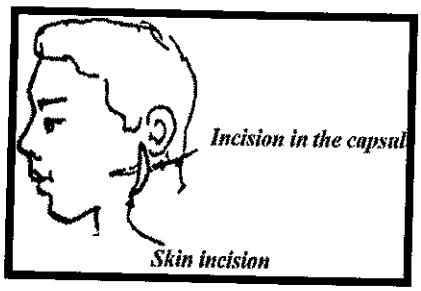
- **Complications :**

Patients with Sjogren's disease are 44 times prone to the development of lymphoma than the general population.

Acute parotitis

Definition :

Acute inflammation of the parotid gland.



Etiology :

Causative agent : pyogenic organisms usually staph. Aureus.

Route of infection :

1. Retrograde infection from the mouth through the duct.
2. Blood born infection.

Predisposing factors :

1. Dehydration.
2. Debilitating medical diseases as typhoid and cholera (dry infected mouth).
3. Obstruction of the parotid duct.
4. As a complication of septicemia.

Pathology :

- As the inflammatory process proceeds, the resultant edema and exudate will increase the tension inside the closed space formed by the parotid capsule and fascia.

Clinical picture :

1. **Severe toxemia.**
2. **A painful swelling** on the side of the face. **Edema** appears early and **fluctuation** appears later.
3. **Pus** can be expressed from the duct of the gland in the oral cavity.

Treatment :

1. Prophylactic : correct dehydration, and care for oral hygiene.

2. Early cases : respond to antibiotic therapy.

3. In fulminating cases : Early decompression of the gland is needed **(even before appearance of fluctuation)**. A curved skin incision is done following the angle of the mandible. The skin flap is reflected. The capsule of the parotid is opened transversely by **Hilton's method** (this avoids injury of facial nerve).

Salivary calculi

Incidence : Stone may occur in the parotid gland, which is very rare, but commonly occurs in the submandibular gland **because:**

1. The parotid secretion is watery while the submandibular secretion is viscid.
2. Drainage of the parotid gland is free, while the submandibular duct ascends upwards towards the floor of the mouth making drainage inadequate.
3. Foreign particles are liable to lodge in the submandibular duct.

Pathology :

- The stone may be single or multiple occurring in the duct or in substance of the gland.
- The stone is formed of calcium and magnesium phosphate and carbonate (radio-opaque).

Clinical picture :

1. Attacks of pain and swelling : in the region of the submandibular gland during meals. Pain is colicky in nature and may be referred to the ear.

2. On examination : The gland is enlarged and tender. Stone in the duct can be felt or even seen in the floor of the mouth.

3. The duct opening : Is congested and pus may be expressed from it.

Investigations: X-ray : a stone in the submandibular gland is radio-opaque in 80% of cases and best seen on a plain occlusal view. More parotid stones are radio-lucent.

Treatment :

1. Stone at orifice is removed through meatotomy.
2. Stone in the duct is removed by cutting directly over it.
3. Stones in the gland are treated by total excision of the gland (sialadenectomy).

N.B:*parotidectomy for calcular disease is difficult as recurrent inflammation makes identification of facial nerve difficult.*

Salivary Neoplasms

Incidence :

Salivary neoplasms constitute 5 % of head and neck tumors. The majority of these neoplasms are benign and most commonly arises in the parotid gland.

Classification :

Salivary Neoplasms

Benign	Malignant
1. Pleomorphic adenoma (mixed salivary tumor)	1. Mucoepidermoid carcinoma
2. Warthin's tumor (adenolymphoma)	2. Adenoid cystic carcinoma (cylindroma)
3. Oncocytoma (oxyphil adenoma)	3. Acinic cell carcinoma
4. Monomorphic adenoma	4. Adenocarcinoma
	5. Carcinoma ex pleomorphic adenoma.
	6. Lymphoma

- **Hemangiomas & lymphangiomas may affect the parotid, but these are pathologically hamaratomas rather than true neoplasms.**

Pathology of benign salivery neoplasm :

1. Pleomorphic adenoma (Mixed parotid tumor) :

- The commonest tumor of salivary glands.
- It occurs in the fourth decade of life, but any age and any sex may be affected.

Pathology :

- It is a pleomorphic adenoma; epithelial cells arranged in sheets, duct like structures, mucoid material (thought to be cartilaginous).
- The tumor is apparently well encapsulated, but the capsule is incomplete (this explain the frequency of recurrence after enucleation).

2. Adenolymphoma = Warthin's tumor :

- **It is an epithelial tumor arising within a parotid lymph node.**
- **Macroscopically :** Multiple cysts of variable sizes are seen.
- **Microscopically :** Epithelial lined spaces in lymphoid stroma.

Pathology of carcinoma of salivary glands :

- It occurs in elderly age, commoner in females.
- It may start de-novo or on top of a mixed salivary tumor.

1. Mucoepidermoid carcinoma :

- Arises from the duct epithelium and is the commonest member of the group.
- It usually affects the parotid.
- Three grades are described; low grade, intermediate grade, or high-grade tumors. The low-grade type is the most frequent and is known to affect children.

2. Adenoid cystic carcinoma (cylindroma) :

- **Histologically** alternating layers of cylinders of cells and hyaline material are arranged around a central cavity forming cylindroma.
- It is the commonest tumor of minor salivary glands.

3. Acinic cell carcinoma :

- The cells of this uncommon tumor resemble the acinic cells of the parotid gland.
- They may metastasize to local lymph nodes or distant sites.

4. Miscellaneous adenocarcinomas :

- They vary according to their histologic pattern.
- They may be trabecular, anaplastic, mucous carcinomas. They are highly malignant.

Clinical picture of benign salivary neoplasm :

Symptoms :

- **Painless, slowly growing swelling in the parotid region.**

Signs :

[A] Inspection shows :

1. Swelling over the angle of the mandible.
2. Elevate the lobule of the ear.
3. The swelling is superficial to masseter muscle.
4. Facial nerve is NOT affected by the tumor.

[B] On palpation :

1. The swelling is not hot, not tender, not attached to the skin, masseter or bone.
2. The swelling is well-defined.
3. Consistency : varies from firm to cystic (but never hard).
4. Pulsation of the superficial temporal artery : is felt normal.
5. Tumors arising in the deeper part of the parotid may bulge in the oropharynx behind the tonsil, hence the importance of examining the mouth cavity.

N.B: ***take care that swelling in the oropharynx may be a carotid body tumor(highly vascular tumor).***

Clinical picture of carcinoma of the parotid :

1. **The tumor forms a rapidly growing swelling, hard in consistency, ill defined and nodular.**
2. **It becomes fixed to skin and deeper structures.**
3. **Local pain, which may be referred to the ear.**
4. **Facial palsy and lymph node enlargement.**
5. **Loss of superficial temporal pulse.**

Differential diagnosis :

1. Extra parotid swellings :

- Lymph nodes (parotid or upper deep cervical), sebaceous cyst & lipomas may resemble pleomorphic adenomas.
- Mandibular, maxillary & infratemporal fossa tumors may produce the false appearance of a parotid enlargement.
- Hypertrophy of the masseter : The condition is sometimes so difficult to differentiate from a true parotid enlargement.

2. True parotid enlargement : that is caused by non-neoplastic salivary gland disease. The gland is diffusely enlarged with no definite lump

Investigations :

Not routine as in most cases clinical diagnosis is reliable enough to proceed to treatment and the pathological diagnosis is not necessary for tumors that behave in a benign way.

1. Biopsy : FNAC is allowed (Reliable investigation which needs expert cytologist).

N.B: ***open surgical biopsy of the major salivary glands is contraindicated.***

2. CT-scan : To show extent of the tumor.

3. Isotope scanning with technetium : Salivary neoplasm shows as a cold spot. Adenolymphoma and oncocytoma show as hot spots.

Treatment :

[1] Tumors that are clinically benign :

- **Enucleation** of a pleomorphic adenoma is easy, but is followed by recurrence that is caused by the left tumor extensions through the defective capsule. The tumor should therefore, be excised with a safety margin.
- The standard operation is **"superficial parotidectomy".** Early in the operation the facial nerve trunk is exposed. All the parotid tissue that is superficial to the nerve and its branches is excised, taking great care not to injure them.
- Tumors arising in the deep part of the gland are treated by **"total conservative parotidectomy"** i.e. excising the whole gland while preserving the facial nerve.
- Benign tumors of the submandibular gland are treated by **"submandibular sialoadenectomy".**

[2] Tumors that are clinically malignant :

- If a pathological diagnosis has not been obtained prior to the operation, **frozen section examination** during surgery is helpful.
- **The standard treatment is radical excision,** which includes wide surgical clearance with cervical lymph node dissection if they were enlarged. For the parotid, this usually necessitates excision of the facial nerve, and probably part of the masseter or the mandible.
- **Radiotherapy** is of limited value, but is administered as a postoperative adjuvant therapy for tumors of high-grade malignancy.

N.B: Salivary neoplasms

1. Carcinoma ex pleomorphic adenoma presents as a painless swelling that has been stationary for years and is now getting bigger.
2. Pleomorphic adenoma of parotid raises the lobule of ear and does not affect facial nerve.
3. Open surgical biopsy of the major salivary glands is contraindicated. FNAC is safer.
4. For tumors of the minor salivary glands of the mouth cavity, excision biopsy is feasible.
5. CT and MRI are the most useful method for assessment.
6. Unlike other salivary neoplasms, adenolymphoma & oncocytoma show a hot spot on isotopic scanning.
7. Even if the facial nerve is preserved in parotid surgery, nerve weakness commonly develops because of neurapraxia that recovers spontaneously few months after the operation.
8. The majority of the tumors arise in the parotid gland are superficial to facial nerve.
9. During submandibular sialadenectomy, 3 nerves can be injured (mandibular division of facial N, inguinal & hypoglossal nerves).
10. The commonest cause of submandibular sialadenectomy is stone in it.
11. For low grade mucoepidermoid carcinoma an attempt to preserve the facial nerve is warrented.
12. For tumors that are clinically malignant, treatment is modified according to the aggressiveness of it.

Swellings in the parotid region

[A] Skin & S.C :

1. Hemangioma. 2. Sebaceous cyst. 3. Lymphangioma.
4. Preauricular dermoid cyst. 5. Melanoma 6. Warts, Boils, Keloids.
7. Lipoma. 8. Hematoma, abscess. 9. Papilloma.

[B] Preauricular L.N.s.

[C] Parotid salivary gland :

1. Inflammation : Mumps, suppurative parotitis, endemic parotitis.
2. Sialectasis. 3. Stones. 4. Tumors.

[D] Masseter : (become fixed on muscle contraction) e.g. Fibrosarcoma or hypertrophy.

[E] Mandible.

[F] Artery : Aneurysm of superficial temporal artery.

[G] Nerve : Neurofibroma.

Swellings in the submandibular region

[A] Skin & S.C. : As before.
[B] Submandibular L.N.s. : Multiple, can be rolled over the lower border of mandible & only felt toward the skin.
[C] Submandibular salivary gland : Single, cannot be rolled over lower border of mandible & felt in the floor of mouth.
[D] Mandible.
[E] Muscle.
[F] Artery : Aneurysm of facial artery.
[G] Nerve : Neurofibroma.

IMPORTANT NOTES:

1-operative treatment of facial nerve injuries:

a) Nerve suture: if nerve graft is needed, the easiest source is the great auricular nerve.
b) If repair is impossible, hypoglossal anastmosis may be done.
c) In hopeless cases, a plastic operation may be done.

2-frey's syndrome:

- It may follow surgery in the region of the parotid gland.
- After injury of auriculo-temporal nerve, parasympathetic fibres from otic ganglion become united to the sympathetic nerves from superior cervical ganglion (cross regeneration).
- Flushing and sweating of the skin innervated by the auriculo-temporal nerve when salivation is stimulated.

Salivary fistula

- Def: fistula that discharges saliva to the skin of the cheek.
- Etiology: injury of the parotid gland or duct resulting from trauma or after surgery.
- Fistula in the gland heals spontaneously.
- Fistula in the duct
  - unlikely to heal on its own (high rate of salivary flow)
  - Sialogram is indicated to role-out distal obstruction.
- Treatment: Surgical treatment is usually necessary.(repair of the fistula or excision of the gland)

Benign lymphoepithelial lesions

- It is uncommon autoimmune disease
- characterized by lymphocytic infiltration & diffuse enlargement of salivary glands particularly parotid & submandibular gland.
- The term "benign" is misleading as 20% of cases develop lymphoma (pre-lymphomatous potential)
- Investigated by Lip biopsy & parotid sialogarm.

Skin & Subcutaneous tissues

Skin & subcutaneous tissues

Sebaceous (epidermoid) cyst

Definition :
A sebaceous cyst is a retention cyst due to blockage of the duct of a sebaceous gland.

Structure :
It is lined by stratified squamous epithelium and contains a foul smelling, white material composed of keratin, epithelial cells and granular debris.

Clinical features :
1. Age : Slowly growing and are, therefore, rarely seen before adolescence.

2. Site : Most commonly in the scalp, face, neck or scrotum but they can occur anywhere except the palm of the sole of the foot, which are devoid of sebaceous glands.

3. On examination:
Small (less commonly large), well defined, cystic swelling, which is usually attached to the skin at one point, which is the site of the duct.
A **punctum** may be seen.

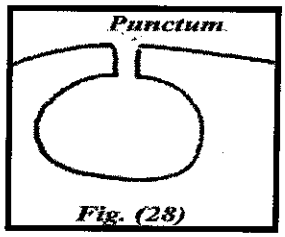


Fig. (28)

Complications :
1. Infection : It may subside by antibiotics, otherwise if an abscess forms, it should be drained and the wall of the cyst curetted. Infection makes the cyst more difficult to excise

2. Sebaceous horn : The contents of the cyst may come out slowly and become inspissated in successive layers over the base.

3. Ulceration : An infected cyst may undergo ulceration leading to the appearance of an ulcer with raised edges. This ulcer is called **(Cock's peculiar tumor)** and it may be mistaken for a carcinoma, but it lacks the induration of malignant lesions. If in doubt a biopsy should be taken.

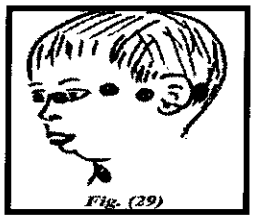
4. Localized alopecia.

Treatment :
Complete excision of the cyst with an ellipse of overlying skin containing the punctum.
The operation can be done under local anesthesia if the cyst is small.

Dermoid cyst

This is a cyst lined by stratified squamous epithelium and contains sebaceous material. Sometimes hairs or sebaceous glands may grow from the wall of the cyst.

Types of Dermoid cyst :

Types	Due to	Sites	Structure
1. Sequestration dermoid  Fig. (29)	Subcutaneous inclusion of portions of the surface epithelium along the lines of fusion of cutaneous segments during fetal life. They may not appear clinically except after few years when the cyst begins to distend.	e.g. external angular dermoids at the outer angle of the eye, at the root of the nose, around the ear or along the midline of the body.	A well-defined, globular, cystic swelling, which is not attached to the skin. The underlying bone may be hollowed out and there may be a pedicle connecting the deep aspect of the cyst to the dura matter.
2. Tubulodermoids	Distension of remnants of embryonic ducts such as branchial cyst.		
3. Inclusion dermoids	Due to inclusion of the epidermis during closure of a cavity.	Sublingual.	
4. Teratomatous dermoid	These are benign forms of teratomas.	Mostly in the ovary but are occasionally found in the testis or posterior mediastinum.	The cyst is lined by squamous epithelium but contains teeth, hairs, bone, cartilage or glands.
5. Implantation dermoids	Puncture wounds which displace some epithelial cells into the subcutaneous tissues. The displaced cells retain their viability and form a dermoid cyst.	Mainly in the fingers, palm or sole.	

Treatment :

The only way of treatment is by **surgical excision.**

NB :

1. In children with a dermoid cyst in the scalp it is better to wait until closure of the skull sutures because some cysts may communicate with the dura.

2. Surgery for a dermoid cyst is more difficult than that for a sebaceous cyst, because a dermoid cyst is deeper.

Vascular anomalies

Classifications:

1-hemangioma
2-vascular malformations
- ✓ Low flow lesions
  - Capillary malformation
  - Venous malformation
- ✓ High flow lesions
  - Arterial
  - Arteriovenous.
  - lymphatic malformations

N.B: differentiation between high and low flow lesions is done by duplex and MRA.

[1] Hemangioma : (previously known as strawberry hemangioma)

1. It affects 10% of white infants and it is three times more common in females.
2. It usually presents by three stages (proliferation, involution & involuted phase).

[2] capillary malformation(port-wine stain)

Items	1. Hemangioma Strawberry angioma	2. capillary malformation Port wine stain (nevus flammeus)
On examination	The lesion is **red** in color and slightly **raised** above the surface.	The lesion is dark purple in color and is **not** raised above the surface. Pressure causes blanching but the color returns immediately after release of pressure.
Site	The commonest site is the face and the head.	May take the distribution of one of the branches of the trigeminal nerve, but the lesion does **not** cross the midline. Sometimes, a port wine stain of the face is associated with similar lesions in the meninges **(Sturge-Weber syndrome).**
Natural history	After the age of one year the lesion starts to undergo involution. Eventually the color fades and flattening occurs so that there may be complete involution at the age of 7 to 8 years. This course is not predictable and sometimes, the lesion continues to grow.	This lesion is present since birth and it does **not** undergo involution.
Complications	The only danger of this lesion is the liability to bleeding if the angioma is in an area liable to friction and ulceration	
Treatment	**(a) Better left for spontaneous involution.** **(b) If problems occur :** Surgery or laser therapies are performed. A course of corticosteroid therapy may help the involution of the lesion.	**(a) Laser ablation.** **(b) Surgery and grafting** may be difficult as the lesion may involve a large area of the face.

[3] venous malformations : previously known as cavernous hemangioma

On examination	Presents as a compressible swelling with some discoloration of the overlying skin.
Site	Occur in the **s**ubcutaneous tissue or the submucous lining of the cheek, lips or tongue. It may involve the internal organs as the liver.
Natural history	The lesion is present **s**ince birth and it has no tendency to involution.
Complications	(a) It may lead to thrombocytopenic purpura due to **s**equestration of a large number of platelets. (b) Atrophy of the overlying skin may lead to **s**evere hemorrhage. (c) **S**epticemia from invasion by microorganisms.
Treatment	(a) Laser therapy. (b) Injection of **s**clerosing solutions as 20 % sodium chloride. (c) **S**urgical excision. *N.B: Compression therapy for venous malformations may relieve pain & edema.*

[4] Arterial malformations (cirsoid aneurysm) :

On examination	The lesion appears as soft, compressible and pulsating swelling with a marked bruit.
Site	It occurs most commonly in the scalp especially in the temporal or occipital regions.
Nature	This lesion is a sort of venous plexus with multiple feeding arteries and so; it is a sort of an arterio-venous fistula.
Complications	Ulceration of the overlying skin may lead to serious hemorrhage, as the bleeding is arterial.
Treatment	Difficult as the swelling is supplied by multiple feeding arteries which have to be ligated. Preliminary embolization of the feeding vessels may be tried.

[5] Arterio-venous malformations:

- It is high flow lesions with an abnormal connections between arteries & veins without intervening capillary bed.
- It may present at birth but may not be evident until late childhood.
- It may be localized or diffuse.
- It may results in growth disturbance or skeletal distortion. There may be local gigantism.

[6] Lymphatic malformations: (see cystic hygroma)

- The commonest cause of congenital macroglossia, macrocheilia & macrotia.

N.B1: Glomus tumor (angioneuromyoma) :

True tumor arising from A-V shunts (glomera) in the distal extremities. It is composed of vascular, neural and smooth muscle fibers. It causes severe paroxysmal pain induced by pressure or temperature changes. It appears as tender purple spot under the nail. Treatment is by excision.

N.B2: Spider nevi and **hereditary hemorrhagic telangiectasia** are considered forms of capillary malformations.

Hereditary hemorrhagic telangiectasia: These are multiple small angiomas in the skin and mucosa with a strong tendency to hemorrhage.

Lipoma

Definition : Benign tumor of the fatty tissue.

Incidence : Common in adult, single or multiple.

Classification :

[A] According to composition :

1. Ordinary lipoma : Mainly fat with little fibrous tissue. Soft.
2. Fibrolipoma : Contains excess fibrous tissue rendering it firm.
3. Angiolipoma : Contains angiomatous tissue rendering it bluish.
4. Myxolipoma : Contains myxomatous tissue.

[B] According to presentation : Lipomas may present as :
1. A solitary swelling.
2. Multiple lipomatosis : in which the limbs or the trunk are the seat of multiple lipomas **(differential diagnosis of multiple swellings).**
3. Diffuse lipomatous deposits : These can occur in certain areas, e.g. patients with myxedema have supraclavicular fatty deposits, elderly persons may develop lipomatous deposits below the chin and sometimes females may develop painful fatty deposits in the thigh **(Dercum's disease).**

[C] According to the site of origin : Solitary lipomas are classified into :
1. Subcutaneous lipomata : These are the commonest and they have the following characters :
(a) Slowly growing tumor in the subcutaneous tissue.
(b) The tumor is painless and is not tender.
(c) **Lobulated surface**, which may be attached to the skin at multiple points.
(d) The consistency is **soft**, although some lipomata may give **pseudofluctuation**; this is due to mobility of the tumor in its bed and because fat at warm temperature may undergo liquefication.
(e) It has a well-defined **slippery edge** due to movement of the tumor inside its capsule.
(f) The swelling is mobile over deep structures.

2. Subfascial lipomata : Occur under the deep fascia.
3. Intermuscular lipomata : Difficult to diagnose and has to be differentiated from a fibrosarcoma. The latter is hard in consistency and grows rapidly.

4. Submucous lipomata : A submucous lipoma in the larynx may cause respiratory obstruction. A submucous lipoma in the intestine may initiate intussusception causing intestinal obstruction.

5. Parosteal lipomata : Arise in relation to cranial bones and cause erosion of the underlying bones.
6. Extradural lipomata : Are sometimes found within the spinal canal and may cause paraplegia (never in the skull, no fat).

7. Intra-articular lipoma.
8. Retroperitoneal lipoma.

Complications : Are rare and include :
1. Degenerative changes leading to liquefication and calcification.
2. Malignant transformation is very rare, but it can occur only in a retroperitoneal lipoma.
3. Submucous lipoma and extradural lipoma (see before).

Treatment : Surgical excision.

Callosity

Area of superficial skin thickening due to continuous friction and pressure, usually not painful.
Treatment : should avoid the predisposing factor and it should be shaved with a razor blade and repeatedly painted with salicylic acid.

Corn

Similar to callosity but caused by pressure as well, compressing the sensory nerve endings causing **pain** with similar treatment.

Wart

Due to viral infection in an abrasion causing localized overgrowth of the epidermis and papillae of the skin. It is small horny projection and may be multiple. Its treatment includes :

1. Curettage & diathermy.
2. Repeated application of glacial acetic acid.
3. Cryosurgery.

Papilloma

Benign tumor, usually pigmented commonly develops in large numbers in the face, arms and upper trunk. Treatment is excision, cryosurgery or cautary.

Hypertrophic scar

- It is an increase of both intensity and duration of the active phase of scar formation.
- It is common in young ages after burns.
- The scar is very thick, red, itchy for 3 to 6 months then the condition regresses gradually
- **Treatment :** Excision & Plastic repair.

Keloid

- A rare condition, commonly over the sternum.
- There is inhibition in maturation and stabilization of collagen fibers.
- The same picture as hypertrophic scar but keloid may spread among nearby healthy tissues and continues to get worsen even after years.
- High rate of recurrence after excision or X-ray therapy.
- **Management :**
  1. Shaving of excess tissue then resurfacing by thin skin graft.
  2. Steroid injection.

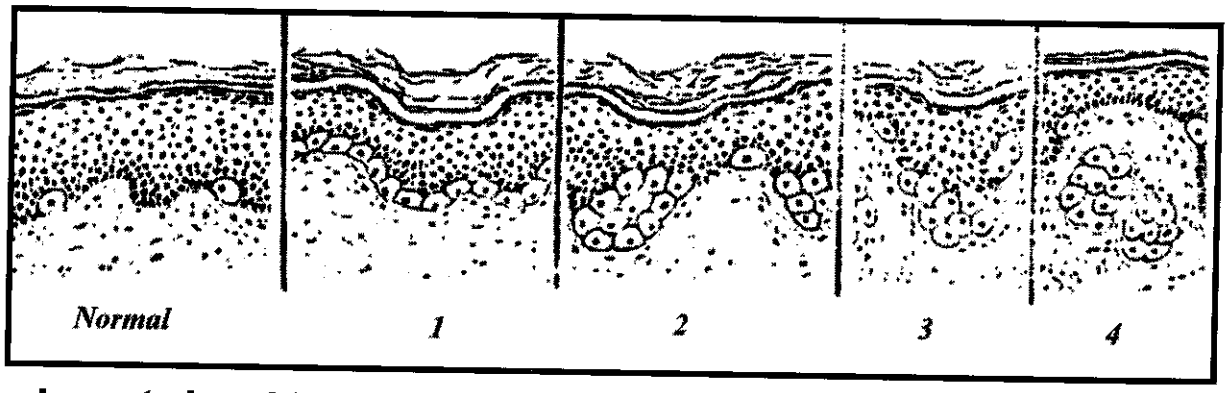
Items	Hypertrophic scar	Keloid
Definition	Delayed maturation of collagen fibers in a wound.	Failure of maturation of collagen fibers in a wound.
Induration	Limited to the scar.	Extends beyond the scar.
Course	Stationary after 6 months.	Progressive even after 6 months.

Melanocytic Tumors

• Melanocytes lodge in between the basal cells of the epidermis. Various hamaratomas or true tumors can arise from the melanocytes.

Benign lesions of melanocytes :

1. Lentigo : Melanocytes replace the basal layer of the epidermis over a certain area.
Clinically : It appears as a spot varying in color from pale brown to black.
2. Junctional-pigmented nevus : More proliferation of melanocytes at the junction of epidermis and dermis.
Clinically : Looks like a lentigo.
3. Compound pigmented nevus : Some melanocytes pass to the dermis. These melanocytes lose their capacity to form melanin and they are called nevus cells.
Clinically :
• Lesions are raised above the surface and vary in color from pale brown to black.
• Sometimes dark hairs are seen growing from the surface of the lesion. Rarely, an extensive area of the skin is replaced by such hairy-pigmented nevus **(Giant hair nevus).**
4. Intradermal pigmented nevus : The commonest.
Clinically : They look like compound pigmented nevus.



Can pigmented nevi turn malignant ?

1. The giant hairy-pigmented nevi can give rise to metastases.
2. After puberty any pigmented nevus with junctional activity has the potential to undergo malignant changes, but this is rare.

Indications for surgical excision :

1. For cosmetic reasons.
2. If they are subjected to repeated trauma e.g. during shaving.
3. If there is suspicion of malignant transformation.

Adequate surgical excision should be performed and the specimen should be examined histologically.

Precancerous skin lesions

Squamous keratosis (actinic or senile) :

Present by dry, rough, inelastic and irregular pigmented areas of skin with hyperkeratosis. It is the most important precursor of squamous cell carcinoma.

Bowen's disease :

Arises in non exposed skin with sharply demarcated, rounded reddish patches which enlarge slowly with marked cellular hyperplasia.

Melanoma

Incidence :

• In Western countries the incidence of malignant melanoma is increasing, which may be due to defective ozone layer. Malignant melanoma is almost unknown before puberty.

Etiology :

1. Prolonged exposure to ultraviolet rays of the sun.
2. Albinism and xerodermia pigmentosa.
3. On top of a benign nevus (discuss).

Criteria of malignant transformation of a benign nevus :

1. Increase size or volume of the tumor.
2. Increased pigmentation or depigmentation.
3. Ulceration and bleeding.
4. Pain.
5. Induration.
6. Surrounded by halo of pigmentation or multiple satellites.

Pathology and clinical types :

Items	Superficial spreading melanoma	Nodular melanoma	Lentigo melanoma	Acral lentiginous melanoma	Amelanotic melanoma
Incidence	64 %	12-25 %	1-15 %	Rare	Rare
Site	In any part of the body.	In any part of the body.	Usually in the face.	In the palms and soles. It may occur beneath the nail.	
Age	Usually in middle age.	In younger age groups.	Elderly persons.		
Clinically	1. Raised above the surface. 2. Edge : Irregular.	1. Raised above the surface. 2. Surface : Smooth. 3. Color : Gray or Black. 4. Liable to ulceration.	Some areas may regress.		It is not pigmented.
Prognosis		Bad.	The least malignant type.	Poor.	Very poor.

• Malignant melanoma may rarely arise in the eye, in the meninges, or at the mucocutaneous junction as the anal canal.

Factors, which have a major influence on the prognosis, are :

[A] The thickness of the tumor **(Breslow classification).**

[B] The depth of invasion of the skin **(Clark's level of invasion)** as follows :

1. Epidermis.
2. Dermo-epidermal junction.
3. Superficial papillary dermis.
4. Deep papillary dermis.
5. Subcutaneous tissue.

Spread :

1. Direct : To subcutaneous tissues and the deep fascia.

2. Lymphatic : By permeation or embolism. Lymphatic permeation leads to satellite nodules around the tumor or between the tumor and the regional lymph nodes.

3. Blood : Malignant melanoma can spread to any organ. Secondary deposits are usually black.

Differential diagnosis:

1. Granuloma.
2. Pigmented basal cell carcinoma.
3. Hemangioma.
4. Compound or junctional nevus.

Investigations :

- The only sure method of diagnosis of malignant melanoma is **Biopsy & Histological Examination.**
- The biopsy should include the whole skin & S.C. tissue with safety margin 3mm at least.

Treatment :

1. The primary lesion : Should be excised with a safety margin 2-3 cm together with the subcutaneous fat but not including the deep fascia.

- If the tumor thickness < 1mm → safety margin is 1 cm.
- If the tumor thickness 1-4mm → safety margin is 2 cm.
- If the tumor thickness > 4mm → safety margin is 3 cm.

2. The lymph nodes :

- If the regional lymph nodes are enlarged and firm, a radical block dissection is performed.
- If the lymph nodes are not frankly malignant on clinical examination, FNAC is performed.
- Prophylactic block dissection is no longer performed.

3. Metastases : Are treated by chemotherapy, interferon and by interleukin-2.

Items	Basal cell carcinoma (Rodent ulcer)	Squamous cell carcinoma (Epithelioma)
Definition	• This is a locally malignant lesion, which arises from the basal cells of the epidermis or from the equivalent cells of hair follicles and sweat or sebaceous glands.	• Malignant tumor of stratified squamous epithelium.
Incidence	• The disease is more common in males above the age of 40 years. It is the commonest malignant lesion of the skin.	• The tumor is more common in males especially elderly persons.
Etiology	1. Prolonged exposure to ultraviolet rays of sun (most important). 2. Persons with a light colored complexion are more liable (presence of melanin pigment protects against the effects of ultraviolet rays). 3. Albinism and xerodermia pigmentosa predispose to multiple basal cell carcinomas all over the body. 4.patient receiving immunosuppression	1. Prolonged exposure to ultraviolet rays of the sun. 2. Previous irradiation. 3. Albinism and xerodermia pigmentosa. 4. Long standing irritation of the skin as : Chronic ulcers, sinuses, and old burn scars. A carcinoma arising on top of a scar is called Marjolin's ulcer. 5. Carcinogenic agents as : polycyclic hydrocarbons, coal tar derivatives, or mineral oils.
Pathology : **1. Site**	• 90 % of the lesions occur in the face especially above a line from the lobule of the ear to the angle of the mouth. • The commonest sites are the outer or inner canthus of the eye and the nasolabial fold.	• The tumor occurs more in the upper part of the face, the lower lip, and the dorsum of the hands. • Sometimes, the tumor arises at the mucocutaneous junctions.
2. Macroscopic picture	**1. The lesion usually starts as a small nodule** covered by thin epidermis. The nodule ulcerates after sometimes with serous discharge and bleeding. Healing sometimes occur in one area of the lesion while further ulceration occurs. **2. Later the patient presents by an ulcerated mass :** • **The edge :** rolled-in and beaded. • **Size :** Small. • **The floor :** red & granular, and often covered with a dry crust or scab. • **The base :** Soft with late fixation to underlying structures. **3. Excavating type :** Where the ulcer erodes deeply into the underlying structures leading to destruction of the nose and infiltration of the nasal sinuses. **4. Other less common types include :** the cystic, the pigmented (containing melanocytes) or the flat superficial spreading type resembles psoriasis or eczema and presents as a red scaly patch.	• **An ulcerated mass (malignant ulcer) :** **1. Size :** Grows rapidly, may reach large size. **2. The edges :** Are raised and everted. **3. The floor :** Malignant fungating tissue. **4. The base :** Is indurated and it becomes rapidly fixed to the underlying tissues. **5. Secondary infection :** May occur and there may be blood stained discharge.

3. Microscopic picture	• The tumor cells are arranged so as the outer layer of these groups of cells is composed of low columnar cells arranged side by side like **a palisade.** • Inside this layer the cells are polyhedral with large basophilic nuclei. • There is no tendency to keratinization. • The stroma shows infiltration by lymphocytes.	• In well-differentiated tumors each mass shows the same layers as the normal stratified squamous epithelium with keratin inside and basal cells outside. • These cell masses with central keratin are called cell nests (epithelial pearls). *Presence of keratin & prickle cells indicates the differentiated nature of the tumour. Undifferentiated tumors, cells are anaplastic with no cytological evidence of their origin* • Broder's classification is used in squamous cell carcinoma and depends upon the number of cell groups with cell nest appearance : **Grade I :** 75-100 % Keratinized groups **Grade II :** 50-75 % Keratinized groups **Grade III :** 25-50 % Keratinized groups **Grade IV :** 0-25 % Keratinized groups
4. Spread	**1. Direct spread :** To the surrounding and underlying structures. **2. Lymphatic spread :** Does not occur. If the draining lymph nodes are enlarged, this is usually due to secondary infection or the presence of a focus of basosquamous carcinoma. **3. Blood stream spread :** Does not occur.	**1. Direct spread :** The tumor rapidly infiltrates the adjacent structures. **2. Lymphatic spread :** To the regional lymph nodes. Sometimes the nodes are enlarged as a result of secondary infection. In patients with Marjolin's ulcer, the fibrosis associated with the scarring makes lymphatic dissemination late. **3. Blood stream spread :** Is very uncommon.
Differential diagnosis	1. Basal cell carcinoma. 2. Squamous cell carcinoma. 3. Malignant melanoma. **4. Keratoacanthoma :** It commonly occurs due to sun exposure, in the face or hands of middle aged people as a firm rounded red papule, which may grow to 2 cm or more in a few weeks. The swelling often ulcerates & may undergo spontaneous healing in 6 monthes. • **Treatment :** Surgical excision is recommended to permit adequate histological examination.	
Investigations	• Diagnosis should be confirmed by a biopsy. • If the lesion is small, excision biopsy is performed; otherwise a small piece of the edge with an area of adjacent skin is excised.	

Treatment

[A] Radiotherapy :

- ✓ Basal cell carcinoma is very radiosensitive.
- ✓ The dose of radiotherapy is fractionated over several weeks to diminish the scaring and necrosis.

Contraindications :

1. If there is involvement of an underlying cartilage or bone
2. If the lesion is near the eyes

[B] Surgery Is indicated for :

1. **Small lesions.**
2. **Infiltration of underlying cartilage or bone :** Because :
   (a) The bone cells will protect the malignant cells from the effect of radiotherapy.
   (b) Radiation is likely to cause necrosis of involved bone & cartilage
3. **Radio resistant lesions.**
4. **Recurrences after previous radiotherapy.**

- Surgery should include an **adequate safety margin** ½ cm at least regarding the surface and the depth.
- The pathologist should check that an adequate safety free margin has been excised, and if not, **revisional surgery** or **radiotherapy** should be applied.
- **The defect created after excision** can be closed by primary suture or by various plastic rotational flaps.

[C] Other possible lines for treatment include :

1. **Cryosurgery.**
2. **Local application of cytotoxic drugs :** as 5-flurouracil ointment.

- **Prognosis :** Excellent and provided that the lesion has been excised completely, the cure rate is 100 %.
- **Recurrence** of the lesion is due to leaving behind foci of malignant tissue and this is more liable to occur if there is infiltration of bone or cartilage.

Two modalities of treatment are available :

[A] Surgery Is indicated for :

1. **Small lesions :** Treated by excision biopsy with an adequate safety margin.
2. **Infiltration of underlying cartilage or bone.**
3. **Radioresistant lesions.**
4. **Recurrences after previous radiotherapy.**
5. **Marjolin's ulcer.**
6. **Block dissection of metastatic lymph nodes.**

- The safety margin for epithelioma is at least 2 cm except in the face where it is 0.5 inch.

[B] Radiotherapy :

The main indication of radiotherapy is for tumors of the head and neck particularly for poorly differentiated lesions.

- **Prognosis :** 90 % five years cure rate.

Hernia

Diseases of the Umbilicus

1. Umbilical fistula :

(a) Fecal fistula : Is either congenital from a patent vitello-intestinal duct, traumatic, inflammatory from tuberculosis of small intestine or malignant from carcinoma of the transverse colon ulcerating through the umbilicus.

(b) Urinary fistula : Is either congenital from patent urachus or is rarely acquired.

(c) Biliary fistula : Is due to perforation of an inflamed gall bladder.

2. Umbilical sinus : Discharging pus from abdominal wall abscess or umbilical infection. Pilonidal sinus of the umbilicus is rare.

3. Umbilical stone.

4. Enteroteratoma (Umbilical polyp).

5. Umbilical granuloma.

6. Umbilical hernia.

7. Tumors of umbilicus :

(a) Squamous cell carcinoma.

(b) Secondary carcinoma nodules.

Desmoid tumor

It is a locally malignant fibrosarcoma. Arises from the anterior abdominal wall muscles. It may occur on top of scars or incisions.

Treatment : Is by excision with a safety margin of at least one inch and reconstruction of the defect.

Complications of abdominal incisions

1. Hematoma : May be due to bleeding tendency, more commonly due to careless surgical hemostasis. It causes painful indurated wound. If of large size, it should be evacuated to avoid secondary infection.

2. Infection.

3. Wound disruption (burst abdomen) : May cause mortality.

4. Incisional hernia : Deep layers disruption of wound during the immediate or early post-operative period.

5. Desmoid tumor.

N.B: rectus sheath hematoma:

May be secondary to trauma or iatrogenic(following abdominal operations) with injury of superior or inferior epigastric vessels.**treatment:** conservative,aspiration or surgical evacuation and ligation of bleeding vessels.

Burst abdomen

Predisposing factors: see incisional hernia.

Types:

- **Partial**. The deep layers burst but the skin is intact.
- **Complete**
- If the intestine prolapses out of the wound, it is called **evisceration**.
- If the intestine is retained inside the abdomen, it is called **dehiscence**.

Clinical features:

- A warning sign to the occurrence of burst is "**the red sign**" where a serosanguinous discharge soaks the dressing.
- The burst usually occurs on **6th** to the **8th** day post-operatively.
- Symptoms of intestinal obstruction may be present.

Treatment: is by urgent surgical closure.

- Cover the prolapsed bowel by a sterile dressing.
- Insert a nasogastric tube and start IV infusion.
- The abdominal wall is closed as one layer by through-and-through sutures using strong non-absorbable suture material, e.g., polypropylene. These are called "**retention sutures**".

Hernia

Definition :

Protrusion of a viscus or part of a viscus (usually within a sac) through a defect in its containing cavity

Types :

[A] External abdominal hernia : It is protrusion of a viscus or part of a peritoneal sac through a defect in the abdominal wall.

1. Inguinal hernia. 2. Incisional hernia. 3. Femoral hernia. 4. Epigastric hernia.
5. Paraumbical hernia. 6. Umbilical hernia. 7. Divarication of recti. 8. Rare types.

[B] Internal hernia :

1. Diaphragmatic hernia. 2. Paraduodenal hernia. 3. Paracecal hernia. 4. Paracolic hernia.
5. Hernia through a defect in the broad ligament of the uterus.
6. Hernia through the foramen of Winslow.

Etiology :

1. Congenital factor : Due to the presence of a preformed sac (as congenital inguinal hernia).

2. Acquired :

(a) Any condition that raises the intra-abdominal pressure : As whooping cough, chronic cough, pregnancy, straining at micturition (stricture or prostatic obstruction), straining at defecation (constipation) and powerful muscular effort.

(b) Weakness of abdominal wall : As a result of obesity, pregnancy, old age and previous operation.

Anatomy :

The hernia consists of a sac, contents and coverings.

[A] Sac : Consists of a diverticulum of peritoneum which is divided into : neck, body and fundus. Neck is usually narrow and well defined.

[B] Contents : May be any of abdominal viscera except the pancreas e.g.:

1. Omentum (Omentocele)	2. Bowel (Enterocele)
Doughy.	Soft.
On reduction → no gurgle sensation.	On reduction → give gurgle sensation.
Percussion → dull.	Percussion → resonant.
Easy reducible at first but difficult at the end.	Difficult reduction at first but easy at the end.

3. A part of the circumference of the intestine = Richter's hernia.
4. Meckel's diverticulum = Littre's hernia.
5. Two loops of the bowel = Maydl's hernia (W hernia)

[C] Coverings : Are derived from the layers of abdominal wall Through which the sac passes.

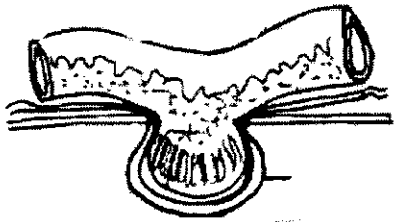


Fig. (52)
Richter's hernia

N.B.:

Richter's hernia :

1. Common in femoral hernia due to small sac.
2. Strangulation occurs without intestinal obstruction (gives picture of gastroenteritis).

Clinical picture :

- Patient complains of an intermittent swelling at the site of a hernial opening.
- The swelling increases in size on standing, straining or cough and is reducible on lying down.

On Examination : Swelling in an anatomical site of hernia

1. The swelling increases in size in all directions when the patient cough ; **Expansible impulse.**
2. The swelling is reduced in the abdomen ; **Reducibility.**
3. The hernial orifice can be palpated. Its obliteration with the thumb after reduction of the hernia prevents its descent.
4. Consistency : See table.
5. Percussion : See table.
6. Transillumination : Opaque.
7. A non complicated hernia is not hot, not red, not tender, not tense and expansile impulse on cough is always present.

Investigations :

1. Routine pre-operative :
2. Investigations to detect precipitating factors e.g. Plain X-ray chest, abdominal US and I.V.U.

Treatment :

1. Correct any predisposing factor.

2. Truss : Is a palliative treatment and is indicated when the patient is unfit or hernia is very liable to recurrence. Truss can only be applied to a reducible hernia after reduction.

3. Operative treatment : Is the only method of cure :

[A] Herniotomy (better called herniectomy) : Removal of the sac as far as the neck.
- By itself it is sufficient for the treatment of inguinal hernia in children.

[B] Herniorrhaphy : It consists of excision of the sac and reconstruction of the defect by local tissues.

[C] Herniplasty : It is indicated in :
- Very wide hernial defects (can not be closed by sutures).
- Very poor muscles (can not protect the defect).
- Repeated recurrence.

Complications of Hernia :

1. Irreducibility. 2. Obstruction. 3. Inflammation. 4. Strangulation.
5. Rupture due to trauma. 6. Hydrocele of hernial sac.
7. Torsion of the omentum. 8. Interference with normal activity in huge hernia.
9. Complications of treatment : (a) Truss. (b) Taxis. (c) Surgery.

Irreducible Hernia

The contents cannot be returned to the abdomen.

Causes :

1. Adhesion between sac and contents. 2. Over crowding of contents within the sac.
3. Sliding hernia. 4. Narrow neck.

Clinical Picture : The hernia can not be reduced by patient or surgeon, but the hernia is not hot, not tender, nor tense and impulse on cough is still present.
Complications : Obstruction and strangulation.
Treatment : Operation is always indicated (not an emergency). No truss can be used.

Obstructed Hernia

A hernia containing intestine the lumen of which is obstructed either from without by band of adhesion or from within the lumen by a mass of stool (this type is termed **incarcerated hernia**).

Clinical picture :

1. Patient complains of repeated attacks of **abdominal colic, vomiting, abdominal distension** and **absolute constipation.**
2. The hernia is irreducible, soft, not hot, not red and not tense.

Treatment : Distinction between obstruction and strangulation in hernias may be difficult, thus, it is safe to treat it as strangulation and early surgery should be performed.

Inflammation

Causes :

1. Inflammation of contents as appendix.
2. Trauma by attempts at taxis or pressure of truss.

Clinical picture :

1. General manifestations of inflammation.
2. The hernia is painful, hot, red and tender and impulse on cough can be felt.

Treatment : Operation is essential as strangulation can be missed.

Strangulated Hernia

Definition :

A hernia in which the blood supply of the contents is impaired leading to gangrene follows (may occur within 5-6 hours).

Incidence :

- Strangulated hernia is the commonest cause (with adhesive obstruction) of acute intestinal obstruction.
- It varies according to the type of hernia. It is estimated to be about 2-4 % in inguinal, 25-30% in femoral, 15-20% in paraumbilical and 3-5% in incisional hernias.
- Although the incidence of strangulation is higher in femoral hernias, yet, strangulated inguinal hernias account for more than 50% of all strangulated external hernias.
- Strangulation can occur at any age and it is commoner after prolonged use of a truss.

Predisposing Factors :

1. Irreducibility, obstruction and inflammation.
2. Sudden expulsion of new contents.
3. Repeated attempts at reduction producing edema.
4. The hernia may be strangulated at the time of its first descent, especially in infants.

Pathology :

[A] The constricting agent may be :

1. A resistant structure outside the sac as the superficial or deep inguinal ring or the lacunar ligament.
2. The neck of the sac.
3. Bands of adhesions within the sac.

[B] Contents :

Interference with the venous pressure occurs first (venous pressure being low)
→ congestion & edema → further rise of pressure → impairment of the arterial supply

- Mucosa Sloughs within three hours and bleeds in the lumen.
- Within six hours the whole thickness of the intestinal wall is affected, and exudes serosanguinous fluid in the peritoneum.
- The intestine becomes gangrenous and soon perforates producing peritonitis.

• **Gangrene** (and then perforation) appears at the :
1. Ring of constriction
2. Apex of the loop → peritonitis.

Clinical picture :

[A] General :

• **Picture of acute intestinal obstruction** with colicky abdominal pains, vomiting, distention and constipation.
• The patient is **dehydrated** and has **hypovolemic shock.**
• With the onset of gangrene, peritonitis occurs and produces paralytic ileus and the colic stops **(a bad sign).**
• In late stage, there is **picture of both hypovolemic and septic shock.**

N.B.: No obstruction occurs with omentocele, Richter's hernia or Littre's hernia.

[B] Local :

• Severe pain.
• Sudden enlargement.
• Irreducibility.
• **No impulse on cough.**
• The hernia becomes **tense** and **tender** due to extravasation of fluids and peritoneal irritation.

N.B.:
1. In Maydl's hernia, gangrene affects first the loop which lies in the abdomen.
For this reason during operation for strangulation all loops are explored.
The local tenderness over the hernia of this type is not marked during clinical examination.

2. Strangulated Richter's hernia may be spontaneously reduced and the gangrenous part of the intestine may be overlooked at the operation.

D.D. of strangulated hernia :

1. From other complications.
2. From other causes of acute swellings in the anatomical region of the hernia.
3. From other causes of acute intestinal obstruction.

Treatment of strangulated hernia :

I- Resuscitation : Anti shock measure, fluid replacement, blood transfusion and gastric suction.

II- Taxis : i.e. reduction of strangulated hernia by manipulation.
This is done only within the first 6 hours after the onset and only in a previously reducible hernia.

Complications of taxis of strangulated hernia (better avoided) :

1. Shock.
2. Rupture of intestine.
3. Reduction en masse i.e. the sac with its strangulated contents is reduced as a whole into the abdomen.
4. Peritonitis due to squeezing of harmful fluid of the sac into the peritoneal cavity.
5. Volvulus of reduced bowel.
6. Reduction en bisac : contents are reduced from main sac into secondary sac.

III- Operation : Should be done as soon as possible :

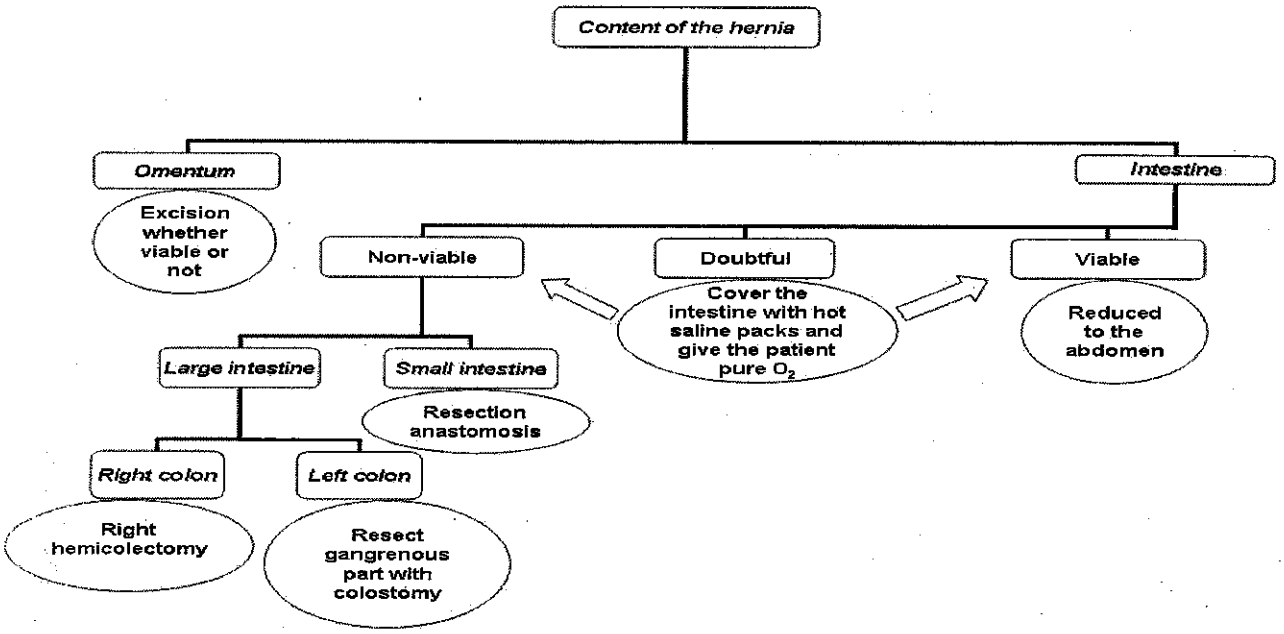
1. Long incision is done reaching down to the fundus of the sac.

2. Fundus of sac is exposed first and is opened before the neck. The fluid in the sac is drained. This avoids escape of the harmful fluid into the peritoneal cavity and avoids peritonitis.

3. Constriction at the neck is relieved.

4. Pull on the loop of intestine present in the sac to reach loops inside the abdomen and examine for strangulated W-hernia **(Maydl's hernia)** in which the strangulated loop of intestine is inside the peritoneal cavity while the loops of intestine in the sac appear normal.

5. Contents of the hernia are dealt with as follow :



Items	Viable intestine	Non-viable intestine
1. Peristalsis.	Present.	Absent.
2. Peritoneal lusture.	Present.	Absent.
3. Mesenteric Pulse.	Present.	Absent.
4. Color	Pink or dark red.	Green, brown or black.
5. Tone	Present (recoil after release ofpressure).	Absent.
6. Fluid in the sac	Yellowish, odorless	Dark, offensive.

Intraoperative tests to detect viability :

1. Doppler ultrasound.

2. Fluroscein test : Inject 1 gm fluroscein I.V., and then inspect the bowel under U.V. rays. If the bowel has good blood supply it will flurescence.

3. Oximeter.

6. Repair of the hernia defect.

7. Subcutaneous drains are usually needed.

Hydrocele of the hernial sac

- **This occurs in narrow necked sacs** if contents return to abdomen and fail to descend into sac again.
- **The neck of the sac** becomes occluded by omentum and serous fluid collects in the sac.
- **Clinically** there is a cystic swelling in the upper part of the cord.

Inguinal hernia

Hernia which traverses the inguinal canal to emerge at the external inguinal ring.

N.B:protective mechanisms of the canal

- The shutter mechanism: contraction (straightening) of conjoint tendon when straining.
- Obliquity of the canal.
- Support of weak points of the canal e.g. conjoint tendon lies behind external ring.
- Bulging mechanism of cremasteric muscle by contraction during straining.
- Increased obliquity of the canal during straining.

(1) Oblique Inguinal Hernia

It is the most common of all hernias. It may be due to the presence of a preformed sac which is part of the patent procesus vaginalis. This is congenital in origin. In old age most of the cases are acquired and are due to muscular weakness and raised intra abdominal pressure.

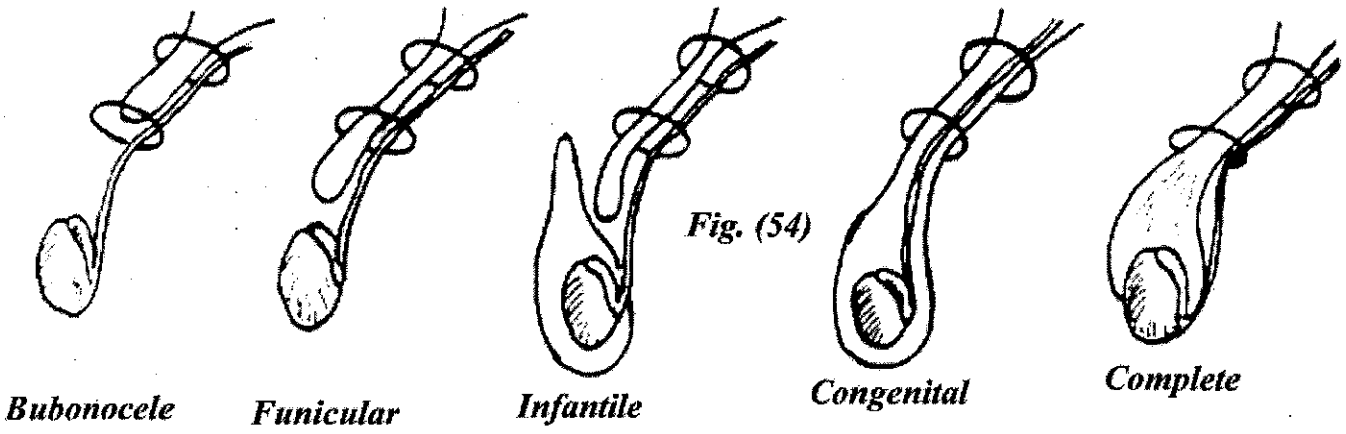


Fig. (54)

Anatomical types :

[A] Congenital type :

The whole procesus vaginalis is patent and thus the general peritoneal cavity is connected to the tunica vaginalis. Though called congenital it may not appear until adult life.

[B] Infantile type :

In which the tunica vaginalis extends up wards towards the inguinal canal. At operation the tunica is liable to be opened in mistake for true sac which will be found behind it i.e. two sacs are found within the cord:
A) Hernial sac.
B) The extension of tunica vaginalis upwards in the cord.

[C] Adult type :

1. Bubonocele : Hernia limited to inguinal canal and stops just distal to superficial inguinal ring.
2. Funicular hernia : Hernia passes with the cord and stops just above the epididymis.
3. Complete inguinal hernia (scrotal hernia) :
Long standing hernia which has reached the bottom of the scrotum. The sac is not continuous with the tunica vaginalis and the testis can be felt separate from the sac (D.D. from congenital inguinal hernia).

Anatomy :

1. The defect : Stretched deep ring.
2. Sac : Escapes from deep ring and lies always inside the cord within the coverings, being anterolateral to the vas and vessels.
3. Contents : See before.
4. Coverings : Skin, superficial fascia, external spermatic fascia from external oblique), cremasteric muscle (from internal oblique), internal spermatic fascia (from transversalis fascia).

Clinical picture : More in males, any age.
Symptoms : Pain, intermittent swelling either inguinal or inguino-scrotal which is reducible.
Signs : As general :

- Direction of reduction is upward laterally and backwards.
- Internal ring test : see clinical notes.
- External ring test : see clinical notes.
- **Transillumination in inguinal hernia :** Opaque (an inguinal hernia may be translucent in infancy or early childhood, but never in adult).

Treatment : See before.

N.B.: Inguinal hernia in children :

1. Herniotomy once diagnosed (for fear of complications).
2. If strangulated : Try taxis if presented early, followed by herniotomy after 48 hours.
3. Bilateral hernias can be operated upon in same setting.

The congenital type of OIH is characterized by :

1. Sudden appearance.
2. Descends into the bottom of the scrotum at once (other types reach the bottom of the scrotum in several years).
3. Completely surrounds the testis and can not be separated from it.
4. Become strangulated at the moment of descent.
5. Found mostly in young children.

(2) Direct Inguinal Hernia

Incidence : 20% of inguinal hernias.

Etiology : Always acquired due to weakness of muscles with associated chronic straining.

Pathology :

The sac enters the inguinal canal behind the spermatic cord.
The sac is small; hemispherical with a wide neck.
The coverings are the skin, external spermatic fascia, conjoined tendon, transversalis fascia and extra peritoneal fat.

Clinical picture and differentiation from the oblique type :

Items	Indirect inguinal hernia	Direct Inguinal hernia
1. Age	Any age.	Usually old.
2. Sex	More in male.	Always in males.
3. Side	Unilateral or bilateral.	Usually bilateral.
4. Shape	Pyriform.	Hemispherical.
5. Descent	Forward medially & downward.	Directly forward.
6. Reduction	Upward, laterally & backward.	Directly backward.
7. Site	Inguinal or inguinoscrotal.	Only inguinal.
8. Internal ring test	(Hernia does not descend).	(Hernia will descend).
9. Defect (seen at operation)	Deep ring, lateral to inferior epigastric vessels.	Hasselbach's traingle, medial to inferior epigastric vessels.
10. Complications	Common.	Rare.

Treatment : See before.

Femoral hernia

Definition :

A hernia which leaves the abdomen through the femoral ring into the femoral canal. It represents 20% of hernias in females and 5% of hernias in men.

Etiology : More in females due to :

1. Larger female pelvis.
2. Less developed ileo psoas muscle.
3. Repeated pregnancies.
4. Pelvic tilt.

Pathology :

- The hernial sac passes down the femoral canal vertically till it reaches the saphenous opening. It is prevented from further downwards descent. The sharp superior edge of the saphenous opening turns the hernial sac superiorly.
- So the hernia passes forwards pushing before it the cribriform fascia, then curves upwards and laterally, towards anterior superior iliac spine.

Clinical picture :

- Patient presents with a small reducible **(frequently irreducible)** swelling at the upper medial part of the thigh below the inguinal ligament.
- The swelling is not tense or tender and an impulse on cough is present.
- The swelling is below & lateral to the pubic tubercle (D.D. inguinal hernia which is above & medial).

Treatment : See operative notes.

Differential diagnosis :

[A] Reducible femoral hernia	[B] Irreducible fem
1. Inguinal hernia.	1. Irreducible inguinal he
2. Saphena varix.	2. Lipoma.
3. Aneurysm of the femoral artery.	3. Inguinal lymphadenopathy.
4. Psoas abscess.	4. Ilioposas bursa.

Comparison between Inguinal hernia & Femoral hernia

Items	Inguinal hernia	Femoral hernia
Site	Above & medial to pubic tubercle.	Below & lateral to pubic tubercle.
External & internal ring tests	+ ve	- ve
Descent	Forwards, medially & downwards.	Downwards, forwards & upwards.
Reduction	Upwards, laterally & backwards.	Downwards, backwards & upwards.

Complications :

- Frequently femoral hernia becomes irreducible and commonly it gets strangulated. This is accounted for by the narrow unyielding femoral ring and sharp border of lacunar ligament.
- Richter's hernia is not uncommon.

Rare types of femoral hernia

1. Cloquet's hernia: The sac lies under the fascia covering the pectineus muscle.Strangulation is likely.The sac may coexist with the usual type of femoral hernia.

2. Laugier's femoral hernia Is a hernia through a gap in gimbernat's ligament. The diagnosis is based on the unusual medial position of a small femoral hernia. Nearly always the hernia is strangulated.

3. Narath's femoral hernia: the hernia lies behind the femoral vessels,occurs only in patients with congenital dislocation of hip.

Umbilical hernia

I- Congenital Umbilical Hernia (Exomphalos)

1. Exomphalos minor :

A small defect (less than 5cm) is present at the umbilicus through which a small peritoneal sac protrudes. The contents are usually intestine. The coverings are a thin layer of Wharton's jelly and a layer of amniotic membrane.

Treatment : The contents are reduced the sac is excised and the defect is repaired in layers.

2. Exomphalos major :

There is a large defect (more than 5cm) in the center of the abdominal wall usually above the umbilical cord. The contents may include many viscera or part of the liver. The covering is only a layer of amniotic membrane. There is a danger of rupture of the sac followed by peritonitis.

Treatment : Urgent operation

- The skin on either sides of the defect is undermined and approximated together over the sac and sutured.
- Release incisions in the flanks are needed.
- After several months the peritoneum and muscles can be approximated and closed in layers.

II- Infantile umbilical hernia

- The neck of the sac is wide and the coverings are extraperitoneal fat and umbilical scar.

Treatment :

1. Correction of the cause of straining if any.
2. Strapping is better avoided as in most cases the defect closes spontaneously within 2 years.
3. Operation is indicated when the defect is more than 2 fingers wide or when the hernia persists after the age of 2 years. The defect in linea alba is closed with few stitches of non-absorbable suture material as prolene.

III- Adult umbilical hernia (Paraumbilical hernia)

Etiology :

It is more common in middle aged females, especially in obese multiparous women usually, paraumbilical and not umbilical hernia.

Surgical pathology :

- **Paraumbilical hernia is more common above the umbilicus** because the linea alba is thin and broader above than below the umbilicus.
- **The sac has a narrow neck.**
- **Adhesions** inside the sac are very common especially at the fundus.
- **Complications** as strangulation and irreducibility are very common due to the narrow neck, sharp edge and adhesions inside the sac.

Treatment : See operative notes.

- **Why in adults it is a para umbilical and not true umbilical hernia ?**

As the umbilical scar is the strongest part of the linea alba which is wider and thinner above the umbilicus than below.

Epigastric hernia

It is herniation of extra peritoneal fat through a small defect in the linea alba. It may be single or multiple.

Anatomy :

- **The commonest site** is midway between umbilicus and xiphoid process.
- The hernia commences as a protrusion of extraperitoneal fat only **(fatty hernia of linea alba).**
- If the protrusion enlarges, it drags a pouch of peritoneum after it **(true epigastric hernia).**
- **The neck of the hernia is narrow,** consequently, either the sac is empty or it contains a small portion of greater omentum.
- **Coverings :** Skin and subcutaneous tissue.

Clinical picture :

- Small swelling which may be irreducible simulating a small lipoma.
- It may be symptomless. Sometimes severe epigastric pain with nausea and vomiting from traction of herniated omentum on the stomach (D.D. peptic ulcer).

Treatment :

- If there is pain, the surgeon should be sure that it is not due to an underlying disease e.g. peptic ulcer or gall stones.
- Operation is performed by excision of hernial sac and closure of the defect in the linea alba.
- If the defect is large, it is repaired by Mayo's operation.

Divarication of recti

This is separation of the recti due to stretching of linea alba by a chronically raised intra-abdominal pressure.

Clinical Picture :

- It is common in middle aged females due to repeated pregnancies and in young males due to Egyptian splenomegaly.
- When the abdomen is relaxed, nothing is visible, but on raising the shoulders from the bed, the linea alba bulges as a longitudinal ridge between the two recti, the fingers can be dipped into the abdomen between the two recti.

Treatment :

- An abdominal belt is satisfactory in symptomless cases.
- If surgery is necessary **Keel operation** is the most suitable. The operation is discussed under the treatment of incisional hernia.

Incisional hernia

Hernia developing after abdominal incision.

Etiology :

[A] Pre-operative causes :

Obesity, malignant disease, jaundice, anemia and hypoproteinemia.

[B] Operative causes :

1. Operations for peritonitis, pancreas with leakage of enzymes and intestinal obstruction.
2. Incision : Muscle cutting, damage to nerve or blood supply to muscles.

[C] Post-operative causes :

1. Post-operative cough, chest complications and distension.
2. Post-operative wound infection.

Clinical picture :

- A swelling through the scar of an operation which is reducible, with an impulse on cough.
- It may contain omentum or intestine.

Treatment : See operative notes.

N.B.:• Direct hernia protrudes through the Hasselbach's Δ which is bounded by :

1. Medially → Rectus muscles.
2. Laterally → Inferior epigastric vessels.
3. Below → Inguinal ligament.
4. Floor → fascia transversalis.

- **The hernia (contra distinct to oblique type)** : Lies posterior to the spermatic cord.
- **Hasselbach's triangle is divided into medial and lateral halves by lateral umbilical ligament so the direct hernia leaves the triangle through its :**

1. Lateral part → lateral direct hernia.
2. Medial part → medial direct hernia (more liable to strangulation).

Pantaloon hernia (Dual or saddle - bag) :

- Here there are 2 sacs, one sac being med. & the other lateral to inferior epigastric vessels.
- One of the sacs may be overlooked at the time of operations.

Sliding Hernia :

Definition : Hernia where a viscus forms a part of the wall instead of contents.

- The commonest viscus implied is the urinary bladder.
- The colon may be implicated on the right side (cecum) or left side (sigmoid).

Common in left side.

When to suspect sliding bladder ?

Hernia is dull on percussion, fluctuant & by pressure on it urine is evacuated or the patient experiences a strong desire to pass urine, partially irreducible, double micturition, and huge long standing hernia.

Treatment of sliding hernia :

A U-Shaped incision is made in the peritoneum at a distance of about 2 cm from the bowel. The gap in the posterior wall of the sac is closed by suture. The bowel is replaced now within the abdominal cavity. The neck of the sac is closed by a purse string suture.

Rare External Hernia

1. Spigelian Hernia :

A hernial sac pass through the apponeurosis of transversus abdominis (Spigelian fascia) which forms the lateral Border of Rectus sheath commonly at the level of arcuate line.

2. Lumbar hernia : May be :

(a) Inferior lumbar hernia : passes through the inferior lumbar triangle of Petit which is bounded below by iliac crest, lateral by the external oblique and medially by latissimus dorsi.

(b) Superior lumbar hernia : passes through the superior lumbar triangle bounded by 12th rib above, medially by sacrospinalis and laterally by posterior border of internal oblique.

3. Perineal hernia : Passes through the pelvic floor.

4. Obturator hernia : Passes through the obturator canal.

5. Gluteal hernia : Passes through the greater sciatic foramen.

6. Phantom hernia : due to muscular paralysis or weakness e.g. Malgaigne's bulges.

7.interstitial hernia: the sac passes between anterior abdominal wall muscles.diagnosis may be difficult and may need CT.

8. Amyand's hernia: inguinal hernia with strangulated appendix.

D.D. of inguinoscrotal swelling :	D.D. of Inguinal Hernia :
1. Inguinal hernia. 2. Infantile or encysted hydrocele. 3. Undescended testis. 4. Cord swelling : (a) Vas : T.B. (b) Veins : Bilharzial, varicocele. (c) Lymphatics : Filariasis.	1. Direct from indirect. 2. Femoral hernia. 3. Hydrocele. 4. Undescended testis. 5. Lipoma of the cord.