

TEXTBOOK OF CORE GENERAL SURGERY

PRACTICE AND PRINCIPLES

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(PRINCIPLES AND PRACTICE)

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PUBLISHER:

AMBIK PRESS

FOR

HANAKS MEDICAL PUBLISHERS

**Headquarters: BLEMA HOUSE, 17, Paul Omorodion Street, BDPA, Ugbowo, Benin City,
Nigeria**

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Printed in Nigeria by:



AMBIK PRESS LTD.

**#4, Otike-Odibi Avenue, Isiohor,
Via Ugbowo Old Lagos Road,
P.O. Box 5027,
Benin City, Edo State.
052-465518 & 08074009192**

CONTENTS

Preface	vii
Acknowledgements.....	ix

CHAPTERS	PAGES
1 Thyroid gland	1
2 Tracheostomy	21
3 Non-thyroid swellings of the neck	26
4 Appendicitis	35
5 Enterocutaneous fistula	44
6 Biliary system	50
7 Obstructive jaundice	58
8 Hepatic neoplasms	65
9 Hepatic abscesses	69
10 The spleen	71
11 Peptic ulcer disease, upper GI bleeding	77
12 Carcinoma of the stomach and GIST	95
13 Small bowel tumours	104
14 Colorectal carcinoma	108
15 Lower GI bleeding	117
16 Haemorrhoids	120
17 Anal surgical conditions	123
18 Colostomy	129
19 Inflammatory bowel disease	132
20 Intestinal obstruction	138
21 Peritonitis	158
22 Hernia	166

23	Hydrocoele	166
24	The breast	181
25	Diseases of the oesophagus	186
26	Pancreatic diseases	227
27	Haematuria, bladder outlet obstruction, posterior urethral valves	242
28	Urinary calculi	257
29	Bladder tumours	261
30	Benign prostatic hypertrophy	263
31	Prostate cancer	268
32	Urethral stricture	277
33	Maldescended testis, prune belly syndrome	280
34	General principles of management of trauma	283
35	Abdominal trauma, abdominal compartment syndrome	288
36	Traumatic head injury	293
37	Hydrocephalus	304
38	Management of burns	313
39	Chronic leg ulceration	316
40	Wound healing	328
41	Metabolic response to trauma	340
42	Fluid and electrolyte balance in surgical practice	344
43	Shock, SIRS	358
44	Blood transfusion in surgical practice	365
45	Disseminated intravascular coagulation	372
46	Surgical haemostasis	376
47	Deep venous thrombosis and pulmonary embolism	378
48	Sickle cell disease in Surgery	393

49	Nutrition in the surgical patient	397
50	Surgical site infection	406
51	Principles of antimicrobial therapy	413
52	Principle of Day Case Surgery	415
53	Surgical biopsy	420
54	Tumour markers in surgical practice	424
55	Drains in Surgical practice	427
56	Abdominal incisions	430
57	Minimal access surgery	435
58	Principles of cancer management	442
59	Principles of radiotherapy	449
60	Lasers in Surgery	453
61	Diabetes in Surgery	456
62	Hypertension in the surgical patient	458
63	Principles of fracture management and compartment syndrome.....	463
64	Tetanus and tetanus prophylaxis	474
65	Principles of amputation	476
66	Bariatric Surgery	480
67	A 'pot-pourri' of common short cases	483
68	Surgery in general practice	499

DEDICATION

This book is dedicated to

- **All my teachers**
- **All my students**

PREFACE

"To study the phenomenon of disease without books is to sail an uncharted sea, while to study books without patients is not to go to sea at all"

Sir William Osler (1849 – 1919)

Professor of Medicine, Oxford, UK

My experience over the past three decades as a clinician, medical teacher, and examiner at all levels of Surgery has shown that our medical students and surgical residents need to have more basic information as regards common clinical conditions in General Surgery. This is evident when you take them on teaching ward rounds. The 'sea' (medical facilities) is getting shallower. This situation therefore calls for a handy compass (book) to ease navigation through this peculiar sea. This book is written to act as a handy compass as the young trainee navigates through the 'surgical sea'.

We have a cliche in Medicine: "Common things occur commonly". This book, to all intents and purposes, is not a comprehensive textbook of Surgery. It is, however, aimed at highlighting the core topics in General Surgery. As we all know, General Surgery remains the mother of all surgical disciplines and has been accorded a pride of place in both the undergraduate and postgraduate training and examinations in Surgery.

If the knowledge derived from reading this book is able to tide the trainee over the hurdle of surgical examinations and more importantly prepare him adequately for clinical practice (fortunately or unfortunately one has to pass the examinations before one is licenced to practice either as a medical doctor or as a surgical specialist!), then I can beat my chest and with a loud voice exclaim: "Hurray!! Glory be to God for this modest contribution to medical training. This game is worth the candle after all!"

Chibundu Emmanuel Ohanaka

(January, 2018)

The following experts painstakingly read through various aspects of the original manuscript and made invaluable contributions

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ACKNOWLEDGEMENTS

First and foremost, I wish to appreciate the Almighty God, the Original Source and Giver of knowledge for granting me the wisdom, courage and strength to be able to put these words together. The invaluable contribution of my colleagues who painstakingly read through various aspects of the original manuscript and made useful suggestions is highly appreciated. My thanks also go to my colleagues in the Department of Surgery, University of Benin Teaching Hospital, Benin City, Nigeria for encouraging me to write this book. Dr Chukwunonso Agogbua and Dr John Ukwuoma, my surgical mentees, were gracious enough to supply me with most of the photographs used in the text. I am very grateful.

I am equally grateful to my good old friend and classmate, Dr Dele Oluwatade, who is a successful general practitioner. He suggested the idea and indeed encouraged me to write the last chapter on "Surgery in General Practice". Writing this chapter was most exhilarating as I was made to recall and share my experience in carrying out surgical procedures in the 'workshop' (a euphemism for the operating theatre of a general practice clinic)!

I will never forget to appreciate all the medical students and resident doctors I have been privileged to teach in various medical schools and teaching hospitals in Nigeria. My interaction with them stimulated me into writing this book. Thank you for giving me the honour and privilege to contribute to your success in your chosen profession.

Finally, I am most grateful to my wife, Dr (Mrs) Blessing Ohanaka and our children: Barrister Onyekachukwu Ohanaka, Dr Chukwuemeka Ohanaka, Dr (Mrs) Ivuoma Ohanaka, Dr Adaeze Ohanaka and Dr Chidinma Ohanaka for their patience, love and encouragement in the course of writing this book.

May the Almighty God bless you all and continue to enlarge your coast.

PRACTICE OF SURGERY

CHAPTER ONE

THE THYROID GLAND

Anatomy:

The thyroid gland is a bilobed structure located on either side of the neck. Each lobe extends inferiorly from the side of the thyroid cartilage to the sixth tracheal ring. Both lobes are linked by an isthmus which runs between the 2nd and 3rd tracheal rings. An inconstant pyramidal lobe may project upwards from the isthmus. The pyramidal lobe represents a remnant of the embryological descent of the thyroid gland and usually projects on the left side. The average weight of an adult thyroid gland is 20 grams to 25 grams.

Relations: Anteriorly – Skin, platysma muscle, anterior jugular veins and the strap muscles. The thyroid gland is enclosed by the pretracheal fascia.

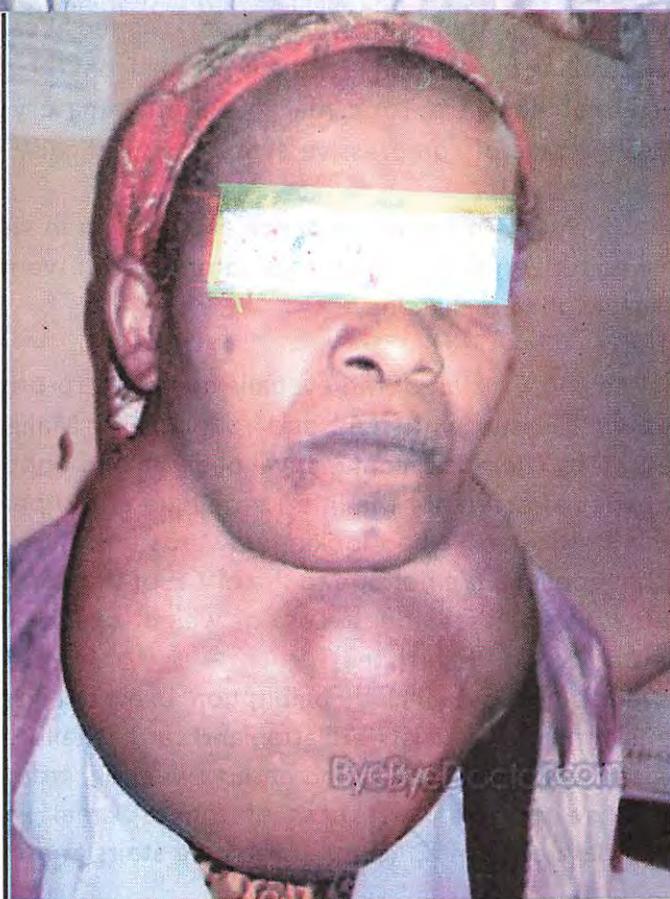
Posteriorly – Larynx and trachea and more posteriorly, the pharynx and oesophagus. The carotid sheath lies on the lateral aspect of the gland.

The recurrent laryngeal nerve lies in the groove between the trachea and oesophagus. It has an intimate relationship with the inferior thyroid artery. The recurrent nerve is usually deep to the vessel. It may, however, be superficial or pass between its branches. The external branch of the superior laryngeal nerve runs in close relationship with the superior pole.

Blood supply: This is from three sources - superior thyroid artery (branch of external carotid); inferior thyroid artery (offshoot of the thyrocervical trunk of the subclavian artery); and the inconstant thyroid ima artery (from the aortic arch of the brachiocephalic artery).

The veins are the superior, inferior and middle thyroid veins. The superior thyroid and middle thyroid veins which drain the upper pole and the lateral aspects of the gland respectively drain to the internal jugular vein. The inferior thyroid vein, on the other hand, drains the lower pole and empties into the brachiocephalic vein. In addition to its specific blood vessels, the thyroid gland has supplementary blood supply from the neighbouring structures (pharynx and trachea).

Embryology: The thyroid develops at the junction between the anterior two-thirds and the posterior one-third of the tongue and descends to its definitive position in the neck. The thyroglossal duct which marks the line of descent eventually atrophies.



GIANT GOITRES

GOITRE; This is an enlarged thyroid gland and is classified as follows

- **Diffuse:** Smooth with no palpable nodule. It is sometimes referred to as a hyperplastic goitre
- **Multinodular:** More than one palpable nodule
- **Solitary nodule:** Single palpable nodule resident in a lobe. The rest of the lobe is presumed to be normal. The clinical importance lies in the fact that about 10% of solitary nodules are malignant
- **Toxic:** Primary or secondary. Primary thyrotoxicosis is said to occur when the thyroid swelling and toxic symptoms manifest simultaneously. The thyroid swelling is usually diffuse in nature and eye signs feature more prominently. Secondary thyrotoxicosis, on the other hand, occurs as a complication in an already existing simple goitre. The onset is insidious and the goitre is usually nodular in nature. Cardiovascular features are usually more prominent than those of the eyes.
- **Malignant (follicular, papillary, medullary and anaplastic)**
- **Inflammatory (Riedel, de Quervain, Hashimoto)**
- **Simple:** Diagnosis of exclusion (non-toxic, non-malignant, non-inflammatory)

MANAGEMENT OF GOITRE

Initial management (history, examination and investigation) is aimed at establishing the following

- That the neck swelling in question is actually arising from the thyroid gland (goitre)
- The clinical state of the goitre: Toxic, malignant, inflammatory or otherwise euthyroid
- Presence or otherwise of compressive symptoms: Difficulty with breathing and/or swallowing

HISTORY – Personal data (including place of residence and religion in view of the possibility of residence in an endemic goiter zone). It is five times commoner in women. Goitre in a middle aged man should raise suspicion of malignancy

- **Toxic symptoms:**
 - **Exophthalmos:** Protrusion of the eyeballs due to retro-orbital fat and infiltration of retro-bulbar tissue with fluid and round cells. Malignant exophthalmos is characterised by chemosis, conjunctival oedema and ophthalmoplegia. The main ocular muscles affected are the inferior oblique and superior rectus. This results in diplopia particularly when looking upwards and outwards. Orbital proptosis and ophthalmoplegia are not due to the effect of the hormone
 - **Gastrointestinal manifestations:** Loss of weight despite an increased appetite, and hyperdefaecation (diarrhoea)
 - **Cardiac symptoms:** These include palpitation, exertional chest pain and dyspnoea and other symptoms of atrial fibrillation and cardiac failure.
 - **Neurological symptoms** include increased sweating, trembling especially of the upper and lower limbs. Weakness of the proximal limb muscles (proximal myopathy) may result in difficulty in climbing stairs and in standing up from the sitting posture
 - **Others** are menstrual irregularities and pretibial myxoedema

- Malignant features: Loss of weight, anorexia, voice change, other associated neck swellings (lymphadenopathy) and bony protrusions especially involving the skull
- Obstructive symptoms: Difficulty with breathing and/or swallowing; change in voice (compression of the recurrent laryngeal nerve)
- Family and social history: Prevalence of goitre in residential community (endemicity), source of drinking water, ingestion of goitrogens and drugs that may predispose to goitre
- Gynaecological history: menstrual irregularities as may occur in toxicity
- Reason for requesting surgery: Cosmetic, social, or presence of obstructive symptoms

Clinical examination: The following are the aims of the clinical examination in a patient with suspected goitre. The clinician should

- Establish that the lesion is arising from the thyroid gland and is therefore a goitre
- Determine the nature of the goitre: toxic, malignant, inflammatory or euthyroid
- Establish the presence or otherwise of complications such as those due to obstruction of the neighbouring structures (trachea, oesophagus, recurrent laryngeal nerves)
- Rule out retrosternal extension of the goitre

It is important to examine the patient without a head-gear in view of the possibility of metastasis to the skull from a follicular thyroid carcinoma.

General examination: Assess the composure of the patient (restlessness in thyrotoxicosis); jaundice and weight loss in patients with thyroid cancer or thyrotoxicosis; pretibial myxoedema in thyrotoxicosis.

Cardiovascular assessment: Cardiac signs of thyrotoxicosis include

- High volume and collapsing pulse
- Wide pulse pressure
- Arrhythmias: Paroxysmal atrial tachycardia, extrasystole, paroxysmal/persistent atrial fibrillation and cardiac failure. For this reason, the ECG is an important preoperative investigation particularly in patients above the age of 40.

Examination of the abdomen: Hepatomegaly in metastatic thyroid malignancy

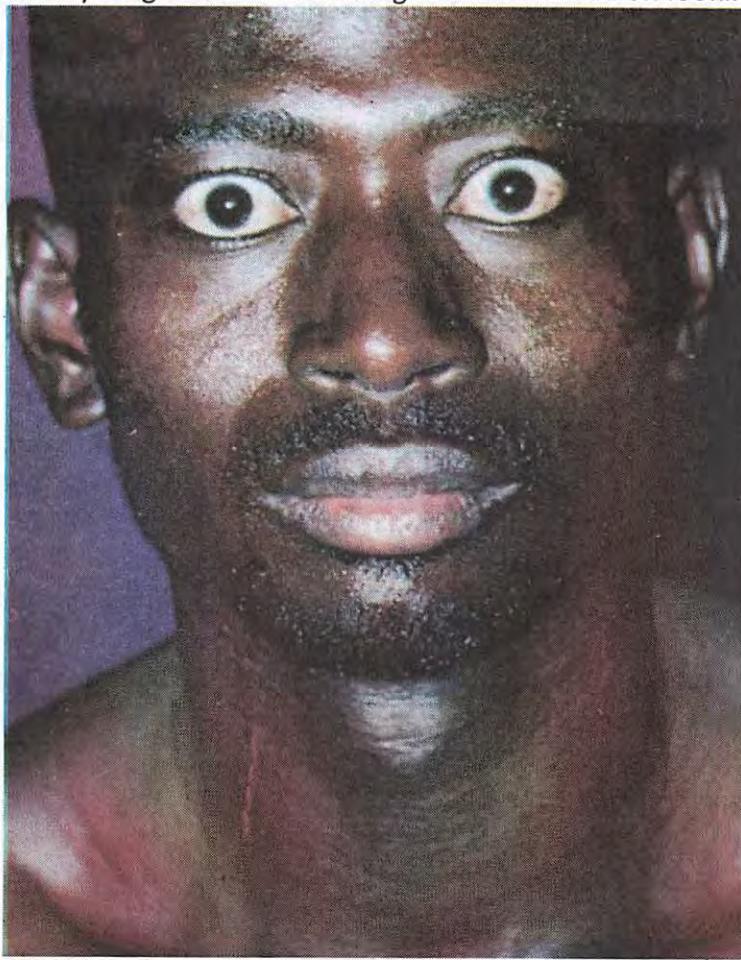
Examination of the neck: Based on the aims of clinical examination outlined above, the following should be carried out

Establish that the neck swelling is arising from the thyroid gland and is therefore a goitre

- Patient is requested to swallow: Both goitre and thyroglossal cyst will move up and down with deglutition. Other neck swellings that move with deglutition include prelaryngeal or pretracheal lymph nodes and indeed any swelling arising from the larynx or trachea. It is, however, important to note that there are some notable thyroid swellings that may not move with deglutition. These include retrosternal goitre with intrathoracic impaction and thyroid cancer with local infiltration. Others include Riedel's thyroiditis and a very huge goitre
- Patient is then requested to open the mouth. This is followed by tongue protrusion. Thyroglossal cyst, unlike goitre, moves upwards with protrusion of the tongue. This movement may be better appreciated by palpation with the hand placed over the swelling.

Signs of toxicity

- Feel palms and check pulse rate: Warm and moist hands/ tachycardia and collapsing pulse are features of toxicity.
- Fine tremors of the hands with the outstretched arms (more obvious with a paper on top of the dorsum of the hand). Fine tremors are equally elicited in the protruded tongue as it lies on the lower lip.
- Eye signs:
 - Exophthalmos: This is demonstrated by the Nafziger's sign (protrusion of the eyeball beyond the superior orbital margin on extension of the neck and viewing the latter directly from the posterior aspect of the head). Under normal circumstances, only the lower sclera of the eye is visible. Ability to visualise both the upper and lower sclera is indicative of exophthalmos
 - Von Graefe's sign (Lid lag): The upper eyelid lags behind the movement of the eyeball during rapid up and down eye movement
 - Joffroy's sign: Loss of wrinkling of the forehead on looking up



THYROTOXIC PATIENT: NOTE THE THYROID SWELLING AND THE ASSOCIATED EXOPHTHALMOS

- Mobius' sign: Absence of medial convergence of the eyeballs
- Ophthalmoplegia: Due to weakness of the orbital muscles
- Presence of thrill and bruit over the thyroid mass: Best elicited over the upper pole of the gland
- Pretibial myxoedema: It is due to deposition of mucin-like material in the subcutaneous tissue of the lower limbs and manifests as swelling of both lower limbs in patients with thyrotoxicosis. The early manifestation may involve just the ankle and the foot. Severe cases may, however, involve the whole leg up to the level of the knee. Clinically it manifests as thickened, shiny skin with coarse hair.
- Finger clubbing has been seen in some cases of thyrotoxicosis

Signs suggestive of compression of the neighbouring structures and retrosternal extension

- Clinical evidence of tracheal deviation
- Inability to get below the swelling on deglutition
- Dullness to percussion over the superior retrosternal area
- Pemberton's sign: Congestion of the face and clinical evidence of respiratory distress on raising both arms close to and above the head for 2 to 3 minutes

Signs suggestive of thyroid malignancy

- Berry's sign: Inability to palpate the carotid pulsation. This is due to local infiltration of the carotid sheath by a malignant goitre
- Cervical lymphadenopathy: Due to spread particularly from a papillary cancer
- Clinical evidence of spread: Protrusion over the skull bone; jaundice and presence of a hard, nodular hepatomegaly.

Palpate and describe the clinical features of the swelling (site, size, shape, consistency etc). This is best carried out while standing at the back of the patient

INVESTIGATION OF GOITRE:

This exercise is still aimed at addressing the aforementioned issues

* Thyroid function test: T3, T4, and TSH levels. In thyrotoxicosis, T3, and T4 levels are high while TSH level is low; the reverse is the case in hypothyroidism. TSH level therefore tends to mirror the serum levels of T3 and T4. Both T3 and T4 are bound to thyroglobulin. Free T3 and T4 estimation may be employed in the evaluation of a patient with suspected thyrotoxicosis. In the latter condition, the free T3 and T4 may be elevated despite normal levels of thyroid hormones. A high TSH level is consistent with a hypofunctioning nodule while a low TSH level indicates hyperactivity of the nodule. Whereas hyperactivity of the nodule is associated with only 1% of carcinoma, hypoactivity is associated with a much higher rate of 10% to 20%. It follows therefore that a patient with a solitary thyroid nodule whose TSH level is either normal or high should be further investigated for thyroid cancer

* Ultrasound scan of the neck: This is a very important modality of assessment of goitre

- May detect multiple nodules in an apparent clinical solitary nodule
- Helps to distinguish between a solid and a cystic mass. Some thyroid lesions consist of a 'mixed bag' of both solid and cystic lesions. The clinical importance of this lies in the fact that such complex cysts are highly suggestive of malignancy

- May be employed as a guide in fine needle aspiration cytology (FNAC) particularly when dealing with an impalpable nodule
- May be employed in the progress- monitoring of a clinically detected nodule
- Aids in the detection of occult cervical lymphadenopathy
- Monitoring of postoperative thyroidectomy patients with a view to detecting recurrence
- May detect early vascular invasion which is highly suggestive of malignancy
- * Fine needle aspiration cytology (FNAC): This aids in determining the pathological nature of a thyroid swelling and is best carried out under ultrasound guidance. It has been found useful in the following ways
 - Evaluation of a thyroid nodule: A definite tissue diagnosis may raise the option of non-operative management
 - Differentiation between a benign and malignant thyroid nodule. There is, however, a caveat: FNAC can differentiate between papillary adenoma and papillary carcinoma but not between follicular adenoma and follicular carcinoma. This is because follicular carcinoma is characterised by capsular and vascular invasion by the neoplastic follicular cells. These obviously cannot be demonstrated by FNAC.
 - Diagnosis of a colloid goitre

Fine needle aspiration cytology is currently regarded as the most important test in the diagnostic evaluation of a dominant thyroid nodule

* Biopsy: Core needle biopsy is indicated only when FNAC result is inconclusive. It is also helpful in the evaluation of suspected anaplastic and locally advanced thyroid carcinoma. There is a risk of haemorrhage/haematoma, trauma to the recurrent laryngeal nerves and tracheal injury. Frozen section biopsy plays a minimal role and is employed only when the nature of the goitre cannot be established intraoperatively. This is most pertinent when the lesion is highly suspicious of malignancy

* X-ray of the neck and thoracic inlet: Tracheal deviation/compression and retrosternal extension respectively

*Radioactive iodine uptake test: This involves an oral intake of radioactive iodine (^{123}I , ^{131}I , or ^{99}Tc). The resultant uptake is thereafter assessed by means of a gamma camera. It is considered to be warm in euthyroidism and hot in thyrotoxicosis. About 10 to 20% of cold nodules have been found to be malignant. It is no longer carried out routinely for benign goitres but is still relevant in the follow-up of patients with thyroid malignancy. It may also be useful in the evaluation of toxic nodular goitre. The toxic nodule takes up radioactive iodine at the expense of the rest of the gland which does not take up any. The increased uptake is evident at both the 4th and 24th hours of assessment

* Chest x-ray and electrocardiography: Usually carried out in patients above 40. They are also mandatory in patients with thyrotoxicosis in view of the cardiovascular complications. Presence of the latter may necessitate further investigation by way of echocardiography

* Thyroid antibodies: May be positive in patients with autoimmune thyroiditis. Estimation of the antibodies against thyroglobulin and microsomes is carried out.

* CT scan and MRI: Not routinely employed but may, however, be useful in the evaluation of a large cervical or retrosternal goitre

- *Serum calcium and phosphate: The baseline values should be known in view of possible postoperative hypocalcaemia due to parathyroid insufficiency. The latter is a known complication of thyroidectomy
- *Indirect laryngoscopy: Preoperative vocal cord assessment with a view to detecting an already existing recurrent laryngeal nerve paralysis. This has a medicolegal implication.
- *Full blood count: Preoperative evaluation is important in view of the anticipated primary haemorrhage.
- *Group and crossmatch blood: Patient may require perioperative blood transfusion
- * Serum electrolytes, urea and creatinine
- * Fasting blood sugar especially when diabetes mellitus is suspected
- * Urinalysis

GENERAL TREATMENT OF GOITRE

It is important to note that not all patients with goitre will require surgical treatment. The initial management of early hyperplastic goitre is by the administration of L-thyroxine particularly in those that demonstrate elevated TSH. Many of such patients will respond to this mode of conservative treatment. In the management of thyrotoxicosis, it is equally imperative to ensure that patient is euthyroid before surgical treatment. Achieving this state of euthyroidism in thyrotoxicosis is a bit more complex as there are different modalities of treatment. These are

- * Antithyroid drugs: May be the only mode of treatment particularly in primary thyrotoxicosis. As discussed below, these are supplemented with the concomitant administration of beta blockers
- * Surgery: Usually indicated in large diffuse primary thyrotoxicosis and also in secondary toxic nodular goitre. Experience has shown that the recurrence rate after treatment with antithyroid drugs is quite high in toxic nodular goitre. The patient, however, needs to be made euthyroid by the preoperative administration of these drugs. Surgery has the unique advantage of rapid control of the toxic state. One of the most feared perioperative complication of thyrotoxicosis in an ill-prepared patient is thyrotoxic crisis (thyroid storm). Recurrent thyrotoxicosis may equally occur later in these patients following surgical management. Both conditions are indications for treatment with antithyroid drugs
- * Radioiodine therapy: May be of use in thyrotoxic patients who are above the age of 25. It is also one of the treatment modalities in recurrent thyrotoxicosis.

Antithyroid drugs

* Carbimazole: It inhibits the production of thyroxine and also exhibits an immunosuppressive effect on the production of thyroid stimulating antibodies. Treatment in thyrotoxicosis is commenced with an initial total daily adult dose of 20 to 40 mg. This is continued until patient is certified euthyroid. Thereafter, one of two regimens would be applied in a bid to prevent hypothyroidism. The dose-titration regimen entails a gradual reduction of the dosage of carbimazole down to a maintenance dose. On the other hand, the block and replace regimen involves the addition of 100 to 150 nanogram of levothyroxine sodium to the original dose of carbimazole. Treatment in primary thyrotoxicosis is continued for about 18 months followed by re-evaluation of the thyrotoxic state. On the other hand, antithyroid treatment is continued in secondary thyrotoxicosis until the definitive surgical treatment is carried out.

*Propylthiouracil: Equally inhibits the production of thyroxine. Treatment is commenced with a starting dose of 200mg to 400mg daily and is reduced to 50mg to 100 mg daily when patient is euthyroid

Side effects of antithyroid drugs include

- Common complications: Skin rashes, pruritus, arthralgia and hepatitis.
- Agranulocytosis: This is the most serious and fortunately rare complication of antithyroid drugs. The patient should be warned to report to the hospital at the earliest sign of infection such as sore throat. Fortunately this complication abates with discontinuation of therapy.
- Hypothyroidism: This can be prevented by employing the above-mentioned dosing regimens.
- Recurrent thyrotoxicosis: There is a 50% chance of recurrence of thyrotoxicosis even after prolonged treatment with antithyroid drugs.
- Antithyroid drugs have been found to aggravate exophthalmos

* Propranolol: This is a beta-blocker which acts by inhibiting the adrenergic effects of thyroxine on the end organs. It also inhibits the peripheral conversion of T₃ to T₄. In situations of adverse reactions to antithyroid drugs, beta-blockers may be used as a sole agent in preparing the patient for surgery.

The euthyroid state should be confirmed by both subjective (clinical) as well as objective (repeat thyroid function test) assessments before surgery to prevent perioperative thyrotoxic crisis. It should be pointed out that thyrotoxicosis, particularly the primary variety, may be managed solely by the administration of antithyroid drugs. Radioactive iodine may also be employed in selected cases. Contraindications to the use of radioactive iodine include pregnancy and the presence of pressure symptoms.

SURGICAL TREATMENT OF GOITRE

Indications for surgery

- *Social: Patient's relatives/friends may be embarrassed by the presence of the goitre. It must be pointed out that in some communities, goitre is associated with witchcraft
- * Cosmetic: The patient, as an individual, does not like what she sees of herself in the mirror
- * Nodular goitre: Longstanding solitary or multinodular goitre may be complicated by tracheal obstruction, secondary thyrotoxicosis (35%) or malignant transformation
- * Hyperplastic goitre that fails to respond to conservative management with L-thyroxine
- * Presence of obstructive symptoms with particular emphasis on respiratory distress which may be secondary to a retrosternal extension of the goitre
- * A rapid progression or sudden increase in size of a long-standing goitre: This may connote either haemorrhage into the lesion or malignant transformation
- *Giant goitre: This is defined as one greater in volume than the head that carries it. Olurin defined it more quantitatively as one weighing at least 10gm/kg body weight.
- *Malignant goitre: Either already established or suspected
- * Goitre in a middle aged male patient should raise a suspicion of malignancy

Surgical treatment involves thyroidectomy (excision of the thyroid gland). The extent of surgery, however, depends on the pathophysiological state of the goitre

- * Subtotal thyroidectomy: Carried out in both euthyroid and toxic goitres. The thyroid tissue left behind is just enough to fill the trachea-oesophageal groove (amounts to a thumb-size or 4 grams of tissue on each side). For obvious reason, a much smaller weight of thyroid tissue is left behind during surgery for thyrotoxicosis.
- * Lobectomy: This entails complete excision of one lobe. Lobectomy may be carried out in a confirmed case of solitary nodular goitre provided the contralateral lobe is adjudged normal
- * Hemithyroidectomy: Unilateral lobectomy and isthmusectomy
- * Hartley Dunhill procedure: Lobectomy on the ipsilateral side and subtotal thyroidectomy on the contralateral side
- * Near-total thyroidectomy: This involves leaving only a rim of posterior thyroid tissue after almost total excision of the thyroid gland. The aim is to avoid iatrogenic injury to both the parathyroid glands and the recurrent laryngeal nerves.
- * Total thyroidectomy: Complete excision of the thyroid gland (lobes and isthmus). Indicated in malignancy and when entire thyroid tissue is considered to be diseased.

Modified Olurin's guidelines in the operative management of giant goitre

- * Informed consent: Discuss the problem, its surgical management and the attendant complications with the patient before surgery. The possible role of tracheostomy in the course of management should be highlighted. The role of replacement therapy after total thyroidectomy should be pointed out.
 - * Multidisciplinary approach: Anaesthetists (intubation and postoperative ventilation), ENT surgeons (tracheostomy may be necessary), endocrinologist (management of toxic goitre), speech therapist (related nerve injuries) and surgeon.
- * Positioning in theatre: Anti-trendelenberg. This reduces the blood flow to the neck (thyroid) and consequently reduces primary haemorrhage
- * Incision: it should be at a higher level than that for the average-sized goitre. The incision is placed even higher on the side where the goitre is bigger. This forestalls a situation where the healed scar will fall very low in the neck (and probably into the upper chest wall)
- * Haemostasis: Surgery reveals a highly vascular lesion with 'angry-looking' vessels. It is important to dissect with care and ensure adequate intra-operative haemostasis
- * Strap muscles: These are usually stretched and attenuated in giant goitres. They should be retracted, bunched together and plicated in order to increase their bulk. For better exposure, the strap muscles may need to be divided. Such division should be carried out at the superior half of the muscles in order to preserve their nerve supply (ansa cervicalis) which runs along their inferior half
- * Surgical procedure: Subtotal thyroidectomy is ideal for benign goitre. If both lobes are COMPLETELY diseased, however, there is no alternative to carrying out a total thyroidectomy. The latter is even more relevant when malignancy cannot be ruled out particularly when operating in elderly patients. The possibility of thyroid replacement therapy should have been discussed with the patient before surgery.
- * Preserve the recurrent laryngeal nerves and the parathyroid glands: Both structures may be visualised at surgery. The rule of thumb, however, is to aim at intracapsular dissection of the thyroid gland with possible preservation of the posterior aspect of the capsule

- * Wound drainage: This is necessary as it prevents haematoma formation with its highly morbid sequelle of tension haematoma and rarely thyroid abscess
- * Trachea: The severe tracheal displacement in giant goitres coupled with attenuation of the strap muscles may culminate in what is referred to as a 'floating trachea'. This adverse effect may be minimised by an external stitch fixation of the trachea. This manouvre may obviate the need for post-operative endotracheal intubation and tracheostomy.
- * Postoperative ventilation: This may be necessary in view of tracheomalacia which may complicate a long-standing giant goitre. Tracheostomy may be indicated if ventilation will be prolonged, particularly if there is diagnosed injury to the recurrent laryngeal nerve. The nerve injury is usually detected by the anaesthetist by direct laryngoscopy after surgery.
- * Hypocalcemia: Watch out for clinical signs of hypocalcemia. Serum calcium estimation should be carried out 48 hours after surgery.
- * Hypothyroidism: Thyroid function tests should be carried out two weeks after surgery

COMPLICATIONS OF THYROIDECTOMY

- 1 Reactionary haemorrhage: Occurs within twenty-four hours of surgery and may result in tension haematoma and respiratory obstruction. It should be treated as an acute emergency
 - * Open up the wound by removing all wound clips and/or sutures BY THE BEDSIDE OF THE PATIENT IN THE WARD. Evacuate the haematoma
 - * Administer intravenous infusion, analgesics and antibiotics
 - * Reassure patient, group and crossmatch blood and book the operating theatre for re-exploration
 - * In theatre: Re-explore wound, control haemostasis and insert a drain
- 2 Damage to the recurrent laryngeal nerve: This may be spotted by the anaesthetist during extubation. Bilateral recurrent laryngeal nerve paralysis with adduction of both cords is a clear-cut indication for endotracheal intubation and ventilation. If prolongation of the duration of endotracheal ventilation is indicated, it is more expedient to continue ventilation by way of tracheostomy.
- 3 Tracheomalacia: Tracheal collapse may occur particularly after dealing with a huge, longstanding goitre. Endotracheal intubation and ventilation are required. Again, tracheostomy may be indicated if ventilation is unduly prolonged.
- 4 Thyrotoxic crisis (thyroid storm): This is an acute exacerbation of the clinical features of thyrotoxicosis. It occurs when a euthyroid state is not achieved before surgery. Clinical features are those of severe thyrotoxicosis and include fever, tachycardia, confusion and dehydration. General treatment includes tepid sponging/fanning, administration of intravenous infusion and intravenous hydrocortisone. Specific treatment involves administration of carbimazole and propranolol. Potassium iodide has also been found useful in the management of thyroid crisis. In the short term, potassium iodide causes a prompt reduction in the release of thyroid hormone while on the long term it inhibits the formation of thyroid hormones and iodine trapping by the thyroid gland. It must be emphasised, however, that thyroid storm is a preventable complication. As mentioned earlier, the euthyroid state should be confirmed by both subjective (clinical) as well as objective (preoperative thyroid function test) assessments before embarking on definitive surgery.

5 Hypocalcaemia: This is due to either direct damage to the parathyroid gland or operative injury to its blood supply. Administer intravenous calcium gluconate

6 Recurrent thyrotoxicosis: Treat with antithyroid drugs. Radioactive iodine is an option in selected patients.

7 Hypothyroidism: May be transient. Administer L-thyroxine for about 4 to 6 weeks after surgery; then re-evaluate by repeating the thyroid function tests.

8 Complications related to the wound: Infection, hypertrophic scar, and keloids

THYROID CYSTS

Thyroid cysts are not uncommon as about 30% of solitary thyroid swellings are found to be cystic in nature. Of these, 10% – 15% are malignant. Most of the latter consist of papillary carcinoma. A complex thyroid cyst is one that has both solid as well as cystic components. The clinical significance of complex cysts is their association with a high rate of malignancy

Management: Diagnosis is confirmed by ultrasonography while initial treatment is aspiration. The cyst may refill after aspiration. There is provision for three consecutive aspirations. Further recurrence (refilling) and established complex cysts are indications for surgery (ipsilateral thyroid lobectomy). Intra-cystic ethanol injection has been found effective in the ablation of recurrent, benign thyroid cysts

THYROID TUMOURS

Thyroid tumours are the most common endocrine tumours. Primary epithelial thyroid cancers may arise from either the follicular cells that secrete thyroxine or the parafollicular cells that secrete calcitonin. Lymphoma is the only non-epithelial thyroid cancer. Secondary thyroid cancers are rare

Classification of thyroid cancer

Epithelial cancers

- Papillary carcinoma (70%): This includes microcarcinoma, follicular variant and sclerosing variant
- Follicular carcinoma (15%): Minimally invasive, widely invasive and poorly differentiated
- Undifferentiated or anaplastic carcinoma (10%)
- Medullary carcinoma (5%): Arises from the C cells. Includes mixed medullary and follicular carcinoma
- Non epithelial tumours: Benign (haemangioma) and malignant (angiosarcoma and others)
- Malignant lymphoma

Secondary cancers: Rare but kidney, colon and melanoma may be the primary foci.

Aetiological factors of thyroid cancers

- External irradiation: This may cause genetic mutation. Incidence of thyroid cancers increased in Ukraine after the Chernobyl nuclear incident in 1986. A solitary thyroid nodule in a patient with a prior history of low-dose head or neck irradiation has a 40% chance of being malignant.

- Chromosomal abnormalities: Both follicular and papillary cancers may result from activation of the RAS oncogene while papillary carcinoma is specifically associated with activation of the RET oncogene. As mentioned above, external radiation may be the factor that finally pulls the trigger
- Long term stimulation by thyroid stimulating hormone (TSH): Thyroid cancers are common in endemic goitrous zones
- Familial predisposition: Medullary carcinoma may be associated with type 2 multiple endocrine neoplasia (MEN) which consists of medullary carcinoma, parathyroid neoplasia and adrenal phaeochromocytoma. Mutation of the RET proto-oncogene has been found in association with medullary carcinoma
- Premalignant lesions: Autoimmune thyroiditis and Hashimoto's thyroiditis may lead to the development of malignant lymphoma

Papillary carcinoma: It is the most common thyroid cancer. An admixture with colloid-filled follicles is found in most of them. Spread is usually slow and is by lymphatics. Haematogenous spread which occurs in extrathyroidal lesions is rare. Histology shows papillary projections and presence of pale, empty nuclei referred to as Orphan Annie-eyed nuclei. Fine needle aspiration is useful in the laboratory diagnosis. It has a good prognosis as it is a slow-growing tumour. Lymphatic spread of an occult papillary carcinoma to the lymph nodes gives rise to what was erroneously referred to as the lateral aberrant thyroid. The presence of positive cervical lymph nodes does not, however, affect the prognosis. Papillary microcarcinoma refers to lesions less than 1cm in diameter

Follicular carcinoma: It is more aggressive than papillary carcinoma and spreads through the blood stream. Macroscopically, it appears encapsulated but microscopically there is evidence of capsular and blood vessel invasion. The most common site of distant metastases is bone. The latter may involve the skull. Tissue diagnosis by histological examination has been found to be more useful than FNA in the management of follicular carcinoma. The prognosis is not as good as in papillary carcinoma. Hurthle cell cancers are characterised by the predominance of oxyphil cells (Hurthle or Askanazy cells). They originate from follicular cells but have a worse prognosis than pure follicular cancer.

Medullary carcinoma: This arises from the parafollicular C cells which are derived from the neural crest. The C cells form part of the APUD system (amine precursor uptake and decarboxylation). The upper pole of the thyroid gland harbours the highest concentration of these cells. Medullary carcinoma may be associated with type II MEN which is autosomal dominant. The RET proto-oncogene in chromosome 10 is the genetic marker. Family members should be screened particularly when occult lesions are diagnosed in children and young adults. Medullary carcinoma secretes calcitonin. Secretion of the latter may be stimulated by administration of calcium or pentagastrin and is associated with a high level of carcinoembryonic antigen (CEA). There is associated diarrhoea in 32% of cases. This may be due to a high output of 5-hydroxytryptamine in type II MEN. Other



PATIENT WITH FOLLICULAR CARCINOMA: NOTE THE SWELLING IN THE SKULL WHICH IS METASTATIC FROM THE OBVIOUS GOITRE

secretory products from C cells in medullary thyroid carcinoma are chromogranin, somatostatin, prostaglandin, vasoactive intestinal peptide (VIP), substance P, ACTH and TSH. The familial variety is usually bilateral, multicentric with C cell hyperplasia. On the other hand, the sporadic form is solitary. Histology shows a characteristic amyloid stroma that contains calcitonin. Spread is by both lymphatic and haematogenous routes. Prognosis is good with impalpable lesions but poor when lesions are clinically palpable.

Anaplastic carcinoma: About 75% of anaplastic carcinomas arise from previously differentiated thyroid cancers especially follicular carcinoma. It is more common in elderly women. Spread is by both lymphatic and haematogenous routes. Histologically, anaplastic thyroid cancer is associated with spindle and giant cells. It has a dismal prognosis as most patients present late.

Microcarcinoma (occult carcinoma): Small-sized occult tumours of up to 36% have been reported as autopsy finding in some series. They are usually of the papillary variety and measure less than 1.0 cm in diameter. Clinically, some may present with cervical lymphadenopathy with no palpable thyroid lesion. Histology of the lymph nodes will show presence of thyroid tissue. In the past, such lesions were erroneously referred to as lateral aberrant thyroid. Like all papillary cancers, however, it has a good prognosis.

Clinical features of thyroid cancer

Patients present with a history of painless neck swelling. Thyroid cancer is commoner in females but the diagnosis should be considered in any male adult who presents with goitre. There may be a family history of goitre. The suspicion may be heightened if there has been exposure to external irradiation and if patient is coming from an endemic goitre zone. A relatively short history of neck swelling and/or an acute increase in size of an already existing goitre should equally raise a suspicion of malignancy. The latter scenario may also be due to haemorrhage into an existing benign goitre. There may be associated anorexia, loss of weight and symptoms of compression/invasion of the neighbouring structures. Symptoms of metastatic spread include respiratory symptoms (spread to lungs and pleurae), yellowness of the eyes (spread to the liver) and associated skull swelling (spread to the skull by a follicular carcinoma). See the photograph above

Clinical examination may reveal a chronically ill-looking patient who is pale and jaundiced. The neck swelling may be irregular in shape and with a firm-to-hard consistency. It may be fixed to the superficial and/or deep structures and so may not move with deglutition.. There may be associated cervical lymphadenopathy especially in papillary carcinoma. Late presentation may show signs of spread to the bones (including the skull), lungs and liver (hepatomegaly with associated jaundice).

Generally speaking, the following clinical features should raise a high index of suspicion of thyroid cancer

- Age: Both extremes – children and elderly
- Rapid growth of a thyroid mass of recent onset or a sudden increase in size of a long-standing goitre
- Symptoms suggestive of vocal cord paralysis: A change in voice may be indicative of local invasion of the recurrent laryngeal nerves
- Symptoms suggestive of compression of adjacent structures: Dyspnoea and dysphagia may be due to invasion of the trachea and oesophagus respectively
- Consistency of the gland: A hard, fixed thyroid nodule is highly suspicious of malignancy
- Presence of palpable cervical lymph nodes

Clinical features peculiar to specific thyroid carcinomas

- Anaplastic thyroid carcinoma:
 - Rapid increase in size
 - Presence of compressive symptoms
 - Gland: hard, diffuse, irregular, fixed to underlying structures
 - Positive Berry's sign: Inability to palpate the carotid artery in the superior pole of the gland due to infiltration by cancer
 - Palpable cervical lymph nodes
- Medullary thyroid carcinoma: In addition to the neck swelling and obstructive clinical features, the following raise suspicion of medullary thyroid cancer
 - Family history of thyroid cancer (30%)

- Presence of other features of multiple endocrine neoplasia such as hypertension
- Cervical lymphadenopathy

Investigations for thyroid cancer

- Thyroid function tests: T3, T4 and TSH. Levels may be normal in thyroid cancer
- Serum calcium, calcitonin and parathormone assay particularly in patients suspected of having medullary carcinoma
- Thyroid autoantibodies: The level is usually raised in Hashimoto's thyroiditis. Of note is that the latter may be associated with malignant thyroid lymphoma.
- Fine needle aspiration cytology (FNAC): Helps to identify malignant cells. May prove inadequate, however, in the diagnosis of follicular and anaplastic carcinoma. Follicular carcinoma is characterised by vascular and capsular invasion which can only be demonstrated by histology
- Tissue diagnosis: Core biopsy and incisional biopsy may be required if FNAC proves inadequate as in the diagnosis of follicular and anaplastic carcinoma
- Isotope scan: Most thyroid cancers do not take up radioactive iodine. They constitute the so called 'cold nodules'. Scans should, however, be interpreted with caution as majority of benign nodules may equally show up as cold nodules. To further compound the issue, a few carcinomas may take up radioactive iodine. Iodine¹²³I and technetium⁹⁹Tc are the isotope scanning agents of choice. Medullary and anaplastic carcinomas have poor uptake of radioactive iodine. Indium-111 labelled octreotide scanning has been found effective in the detection of medullary carcinoma while ^{99m}Tc sestamibi is useful in suspected cases of Hurthle cell tumour
- Imaging studies: USS, MRI and CT are indicated when there is evidence of either lymphatic and/or haematogenous spread.
- Frozen section biopsy: May be indicated in papillary carcinoma with cervical lymphadenopathy in order to determine the extent of surgery.
- Urinary estimation of catecholamines, vanillylmandelic acid (VMA) and metanephrine in patients with medullary carcinoma. This is aimed at ruling out an associated phaeochromocytoma in patients with MEN 2 syndrome

Treatment of thyroid cancer

There are various modalities of treatment: Surgery, thyroxine, radioactive iodine and external irradiation. More often than not, patients are managed by combination of two or more of these. Treatment of thyroid cancer therefore involves a multidisciplinary approach

1 Surgery: The extent of surgery depends on size of tumour, histological variety, degree of differentiation and presence or otherwise of any extrathyroid involvement.

- Lobectomy: Small (<2 cm), well differentiated tumours
- Hartley-Dunhill procedure: Ipsilateral lobectomy and contralateral subtotal thyroidectomy
- Total thyroidectomy: Aims at dealing with any synchronous lesion in the contralateral lobe. This is important when dealing with medullary carcinoma. Total thyroidectomy

is considered the ideal operation even in patients with well differentiated thyroid cancer for the following reasons

- Decreases the incidence of recurrent cancer as it not only removes all the thyroid tissue in the ipsilateral lobe but also takes care of any occult cancer that may be present in the contralateral lobe. This obviates the need for a future completion thyroidectomy which is a much more difficult surgical procedure.
- Improves the efficiency of ablation of residual thyroid cancer by radioiodine and also the authenticity of whole body scanning with radioactive iodine
- Increases the sensitivity of thyroglobulin in the detection of recurrent or metastatic disease
- Central neck dissection: This involves dissection of the functional cervical lymph nodes of the anterior neck. The latter is bounded superiorly by the hyoid bone, laterally by the internal jugular vein and inferiorly by the suprasternal notch. The main indication for central node dissection is suspicion of lymph node involvement by cancer. The practice of routine prophylactic central lymph node dissection, particularly in papillary cancer, is controversial owing to the high rate of postoperative hypoparathyroidism (temporary and permanent) associated with it. It is, however, indicated in the more aggressive medullary thyroid carcinoma
- Prophylactic total thyroidectomy: May be carried out in unaffected family members of patients with mutation in the RET-oncogene who equally test positive for same oncogene. Total thyroidectomy and replacement therapy are offered at a much younger age
- Anaplastic lesions are rarely operable at time of diagnosis

2 Thyroxine: Plays a dual role in the treatment of thyroid cancer

- As a replacement-therapy after total thyroidectomy
- As a suppressor of TSH secretion: TSH is a promoter of thyroid growth and its secretion can only be suppressed by thyroxine. The aim is to administer a good enough dose of thyroxine in order to reduce the TSH level to the barest minimum. Thyroxine therapy is for life.

3 Radioactive iodine: The thyroid gland has a peculiar avidity for iodine. On administration therefore, radioactive iodine is concentrated by the thyroid cancer cells with minimal absorption by other organs of the body. The concentration of radioiodine by the cancer cells is further enhanced by the near-total or total thyroidectomy that is carried out prior to the administration of this radiopharmaceutical. It has the following effects

- Destroys the thyroid cancer cells (tissue)
- Facilitates the future use of radioactive iodine as a tracer element in the diagnosis of residual tumour or recurrence.

Thyroxine therapy should be discontinued for a short period prior to the administration of radioactive iodine. This increases the circulating TSH level and therefore the avidity of thyroid tissue for iodine. It is important to note that medullary and anaplastic cancers are much less responsive to radioactive iodine. On the other hand, follicular carcinoma exhibits a high uptake

of radioactive iodine. Whole body scanning is indicated after total thyroidectomy particularly when there is clinical evidence of recurrent or metastatic disease. T_3 has a shorter half life of 7 days and is the replacement agent of choice when whole body scanning is contemplated. It is administered 4 weeks before radioactive scanning and discontinued for 7 days prior to the procedure. ^{123}I is administered orally and the body is scanned with a gamma camera after 48 to 72 hours in order to determine its uptake of radioactive iodine.

4 External irradiation: This is the sole treatment for malignant lymphoma

Follow up of patients after treatment: This involves regular assessment by way of

- Palpation of the neck
- Monitoring the serum levels of thyroxine and TSH
- Monitoring the level of thyroglobulin. An increasing level may indicate tumour recurrence. Further treatment with radioiodine may be necessary in the latter situation

Indications for completion thyroidectomy

- Recurrent thyroid swelling
- Rising levels of thyroglobulin

Peculiar treatment for specific thyroid cancers

- Anaplastic thyroid cancer: Curative resection is not possible in majority of patients with anaplastic thyroid cancer. Guidelines to treatment are
 - Isthmusectomy or isthmusotomy carried out to relieve tracheal compression
 - Avoid tracheostomy
 - Palliative radiotherapy may be useful
 - Consider reoperation and surgical ablation if whole body radioiodine scanning reveals substantial residual thyroid tissue
- Hurthle cell tumour: It is a variant of follicular neoplasm and arises from the oxyphylic cells of the thyroid gland. Only 20% of Hurthle cell tumours are malignant. Treatment is as for follicular neoplasm

Follow-up after management of differentiated thyroid cancer

- Whole body radioactive scanning after 4-6 weeks; repeat after 6 months
- Estimate thyroglobulin level at 6 weeks and 6 months
- Continue suppressive therapy with L-thyroxine if the above tests are normal
- Continue with routine clinical assessment and thyroglobulin estimation at 3 months interval for 2 years, then at 6 months interval for 3 years, then yearly
- Whole body radioiodine scanning is indicated when there is rising level of thyroglobulin. Metastases are managed with ablative dosages of radioiodine

Prognostic factors in thyroid cancer

- Age at presentation: The older the patient, the worse the prognosis. It is worse in females and males above 40 and 50 respectively.
- Grade of tumour: Well differentiated tumours have a better prognosis than the anaplastic variety

- Extent of tumour: Extrathyroid spread (local or metastases) worsens prognosis
- Size of the primary tumour: Tumours less than 2cm have better prognosis
- Histological type: Survival is best with papillary and worst with anaplastic cancer

Prognostic classification of thyroid tumours

1 Low grade malignancy

- Papillary carcinoma
- Low grade follicular carcinoma
- Low grade malignant lymphoma

2 Intermediate grade malignancy

- Tall cells and columnar variants of papillary carcinoma
- Widely invasive follicular carcinoma
- Medullary carcinoma
- Mixed medullary and papillary carcinoma
- Malignant lymphoma (large cell type)

3 High grade malignancy

- Undifferentiated carcinoma
- Angiosarcoma
- Other sarcomas

CHAPTER TWO

TRACHEOSTOMY

Tracheotomy is an operation to make an opening in the trachea. Tracheostomy, on the other hand, is the creation of an opening directly into the trachea which is maintained by means of a tracheostomy tube or a stoma in order to bypass the upper airway with a view to enhancing respiration. It is one of the life-saving procedures in the field of Medicine. Tracheostomy should be timely in order to achieve the purpose for which it is meant. A peculiar adage says that 'the time to do a tracheostomy is when you first think it may be necessary'. The procedure may be carried out as an emergency or elective procedure. New techniques of performing a tracheostomy such as percutaneous dilatation are now common place.

Relevant anatomy

The trachea lies in the midline of the anterior neck. It extends from C6 (level of cricoid cartilage) to the suprasternal notch inferiorly; then passes retrosternally to the tracheal bifurcation at the level of T5. It comprises of 16 – 20 'C'-shaped rings anteriorly with the trachealis muscle lying posteriorly. The cricoid is the only complete tracheal ring. The tracheal lumen is lined by respiratory ciliated epithelium while the external surface of trachea is covered by the pre-tracheal fascia.

Functions of tracheostomy

- It reduces the anatomical dead space by at least 50%
- It serves as an alternative pathway for breathing
- It improves alveolar ventilation by reducing the work of breathing
- It protects the airway
- It facilitates tracheobronchial toileting
- It facilitates intermittent positive pressure ventilation
- It may aid in the administration of drugs

Side effects of tracheostomy

- There is loss of the humidificatory function of the upper airway leading to dessication of the tracheal epithelium and loss of ciliary activity
- Mucus becomes thick and more viscid. This may result in blockage of the tracheostomy tube
- The vocal cords are not set into vibration, hence patient is unable to speak unless the tube is occluded

Indications for tracheostomy

1 Acute upper airway obstruction

- Foreign body in upper airway
- Trauma: Mandibular fracture (blunt or penetrating), laryngeal and tracheal injuries
- Infections: Acute laryngotracheobronchitis in children, acute epiglottitis, angioneurotic oedema, Ludwig's angina
- Bilateral recurrent laryngeal nerve paralysis which may complicate thyroidectomy
- Tracheomalacia: Post-thyroidectomy for a huge, long-standing goitre
- Compression by a large pharyngolaryngeal tumour

2 Anticipated upper airway obstruction: Tracheostomy may act as an adjunct procedure to surgeries involving the head and neck. It prophylactically secures the airway prior to the following surgical procedures

- Operations in the larynx and pharynx
- Major surgeries involving the neck such as thyroidectomy for a huge, long-standing goitre. Bearing in mind that the resultant weakening of the trachea may result in its collapse after surgery, a preliminary, prophylactic tracheostomy may be carried out
- Operations in the oral cavity: Mandibulectomy and retropharyngeal abscess

3 Protection of the lower airway and tracheobronchial toileting: This is best achieved by the use of a cuffed tracheostomy tube in the following conditions

- Unconscious patients particularly those in coma: head injuries, cerebrovascular accidents, narcotic overdose
- Severe head and facio-maxillary injuries
- Bulbar poliomyelitis
- Tetanus
- Paralysis of respiratory muscles: Spinal injuries, Guillen Barre's syndrome, myasthenia gravis

4 Patients requiring prolonged artificial respiration due to respiratory insufficiency: The typical example is severe chest injuries such as flail chest. Tracheostomy in this condition not only reduces the dead space but equally reduces the air-flow resistance.

Indications for paediatric tracheostomy

Infants:

- Subglottic haemangioma
- Subglottic stenosis
- Laryngeal cyst
- Laryngomalacia
- Tracheomalacia
- Choanal atresia
- Congenital laryngeal web.
- Bilateral vocal cord paralysis
- Children:
 - Acute epiglottitis
 - Diphtheria
 - Acute laryngotracheobronchitis
 - Laryngeal oedema secondary to thermal or chemical injury
 - External laryngeal trauma
 - Juvenile respiratory papillomatosis

Surgical procedure for tracheostomy

- The procedure is best performed under elective conditions in an operating room. In emergent conditions, it may be performed by the bedside. Tracheostomy may be carried out under local or general anaesthesia.
- The patient is positioned supine with neck extended by means of a sand-bag under the shoulders and head supported on a head-ring.
- After cleaning and draping, a horizontal incision is made two finger's breath above the suprasternal notch. The incision lies within the two anterior borders of the sternocleidomastoid muscles. The incision is further deepened into the subplatysmal layer with dissection of strap muscles. The isthmus of the thyroid gland is retracted superiorly and the perichondrium is cleared off the second and third tracheal rings. A smaller knife is used to make a vertical incision on the trachea. The trachea is exposed with the aid of a tracheal dilator. This is followed by the insertion of a tracheostomy tube and the inflation of its cuff. The flanges of the tube are then strapped to the neck by means of a tape or suture.

Postoperative care of tracheostomy

1 Nurse the patient in a warm, well-ventilated room

2 Provide a trolley by the bedside containing a bell, spare tracheostomy tube, tracheal dilator, retractor, and dressing materials



CUFFED PORTEX TRACHEOSTOMY

3 Ensure humidification of oxygen by means of a continuous thermostatically-controlled humidifier

4 Regular suctioning of the tube: This prevents encrustation and hardening of secretions. The latter may result in tracheal obstruction. Suction should be applied only on withdrawal of the catheter. The latter should be soft and sterile. Prolonged or too frequent suction should be avoided. Instillation of sodium bicarbonate will help to reduce the viscosity of the secretions

5 Intermittent deflation of a cuffed tracheostomy tube in order to prevent tracheal mucosal ischemia and stenosis. The cuff is deflated for 5 minutes every hour for the first 24 hours.

6 Prevent the displacement of the tube by securing it with tapes. When in doubt, the position of the tube may be confirmed by a soft tissue lateral X-ray of the neck.

7 The tracheostomy tube may be changed not earlier than the 4th postoperative day. If a cuffed tube is to be retained, then a tube with a long cuff should be selected and inflated to produce an air-tight seal. A short cuff or inflation to an unduly high pressure should be avoided.

8 Voice: After a few days following surgery, an inner tube with an inspiratory valve may be inserted to enable the patient to speak. It is, however, important to ensure that the patient is unlikely to develop emphysema due to resistance following expiration or coughing

9 Appropriate use of antibiotics to prevent infection and its consequences

Complications of tracheostomy

1 Intra-operative

- **Haemorrhage:** Bleeding may involve the anterior jugular veins, the isthmus of the thyroid gland and even the tracheal wall. Prevention is the key to its management. As much as possible, haemostasis should be secured before opening the trachea
- **Injury to other structures:** Carotid artery, recurrent laryngeal nerve, oesophagus and even the trachea itself can be injured.

2 Early postoperative

- **Apnoea and hypotension:** This is due to an abrupt decrease in CO₂ and leads to a loss of stimulus for breathing. This results in apnoea. Treatment is by the administration of 5% CO₂ in oxygen for some hours. Avoid giving patients 100% oxygen.
- **Subcutaneous emphysema, pneumomediastinum and pneumothorax.** It is commoner in children. Prevention is by avoiding unnecessary dissection of tissue
- **planes during surgery and also by maintaining a clear airway during and after surgery.** Any inadvertent rent into the pleura at surgery should be closed
- **Displacement of the tube into the pretracheal space.** This is best prevented by correct siting of the tracheostomy opening and by the use of a well-fitted tube
- **Obstruction of the tracheostomy tube:** This is due to blockage by tenacious secretions. Prevention is by adequate humidification and regular suctioning. Secretions may be made less viscous by the instillation of sodium bicarbonate solution. Frequent changing of the tube also reduces the incidence of tubal obstruction.
- **Pulmonary infection:** This is due to irritation of the tube and endotracheal aspiration. If the tip of the tube is too long and it abuts on the main stem bronchus, atelectasis

of the opposite lung may occur. Pneumonia is prevented by the appropriate use of a sterile suction catheter

- **Secondary haemorrhage:** This is best prevented by a careful selection of the size and shape of the tracheostomy tube. An incorrect-fitting tube may ulcerate through the trachea and erode into a large artery. Infection should also be prevented by the appropriate use of antibiotics. The respiratory tract is best protected in this situation by the use of a cuffed tracheostomy tube.

Late postoperative complications

1 Tracheal stenosis: This may occur either around the site of tracheostomy or at the subglottic region. Prevention is by ensuring that the correct size of opening is made into the trachea below the level of the second tracheal ring. It is important to avoid the use of ill-fitting tubes and ensure a regular deflation of the tracheostomy cuff to prevent mucosal ischemia.

2 Difficulty with decannulation: This is more pronounced in children due to the need to readjust the tube in order to redirect the flow of air through the larynx. It may be prevented in adults by a gradual reduction in the size of the tube. In children, same objective is achieved by partial corking of the tube

3 Tracheo-cutaneous fistulas: This is due to epithelialisation of the tract over a long period. It is managed by allowing the fistula to contract for some weeks after decannulation. This is followed by excision of the epithelialised tract and closure of the wound in layers.

4 Fistulation into surrounding structures: Tracheo-oesophageal fistula and trachea-inanimate arterial fistula. The latter results in severe haemorrhage

5 Wound infection and keloid formation.

CHAPTER THREE

NON-THYROID SWELLINGS OF THE NECK

In order to appreciate the management of non-thyroid swellings of the neck, it is important to review the compartmental anatomy of the neck.

Anatomy: The sternomastoid muscle divides each half of the neck into two triangles - anterior and posterior. The anterior triangle is bounded by the anterior border of the sternomastoid posteriorly, the lower edge of the jaw superiorly and the midline anteriorly. The upper part of the anterior triangle, below the jaw but above the digastric muscles is referred to as the digastric or submandibular triangle. The posterior triangle is bounded by the posterior border of the sternomastoid anteriorly, the anterior border of the trapezius muscle posteriorly and the clavicle inferiorly.

Neck swellings may be congenital or acquired. Congenital neck masses are commoner in children and may appear at birth and indeed at any age. The various pathological neck swellings may be deduced from the possible pathology of its component structures.

- * Skin: sebaceous cyst, lipoma
- * Lymph nodes: infective, lymphomas, lymphocytic leukemia
- * Lymphatics: Cystic hygroma
- * Muscle: Sternomastoid tumour
- * Artery: Carotid aneurysm, carotid body tumour
- * Pharynx: Pharyngeal pouch
- * Branchial arch remnant: Branchial cyst
- * Thyroglossal tract remnant: Thyroglossal cyst
- * Nerves: Neurofibromas, Schwannomas
- * Glands: Salivary glands

Swellings of the neck may also be classified according to the anatomical location in the neck: Lateral, midline and entire neck. This is helpful as a landmark in clinical diagnosis

Lateral:

- Lymph nodes
- Submandibular gland
- Branchial cyst
- Carotid body tumour
- Lipoma
- Neurofibroma
- Sebaceous cyst
- Parathyroid cysts
- Cystic hygroma
- Soft tissue tumours
- Infections: Tuberculosis, syphilis, HIV
- Laryngocoele

- **Midline:**

- Thyroglossal cyst
- Dermoid cyst
- Sebaceous cyst
- Pretracheal node (Delphian)
- Thyroid isthmus swelling
- Plunging ranula

- **Entire neck:**

- Lymphangioma
- Haemangioma

As complex and varied as the causes may seem, they can be classified according to the mnemonic 'KITTEENS' for easy remembrance

K – Congenital; developmental: Sebaceous cyst, branchial cyst, lymphangioma, haemangioma, dermoid cyst, laryngocoele, pharyngeal diverticulum, thymic cysts

I – Infections/inflammatory: Lymphadenitis (bacterial, viral, granulomatous, tuberculosis, cat-scratch, fungal)

T – Traumatic: Haematoma, sternomastoid tumour

T – Toxins (unlikely)

E – Endocrine: Parathyroid, carotid body, MEN

N – Neoplastic: Metastatic – Unknown primary; melanoma, breast, lung, kidney, GIT, lymphoma, lipoma, salivary gland tumours, angioma, carotid body tumour, rhabdomyosarcoma

S – Systemic: HIV, sarcoidosissssss

Generally speaking, neck swellings may be referred to as 'lymphadenopathy and others' as cervical lymphadenopathy constitutes the commonest neck swelling.

Cervical lymphadenopathy:

The causes of cervical lymphadenopathy can be grouped into local and general. In lymphadenopathy due to a local cause, only the cervical nodes are involved. On the other hand, when it follows a general cause, other groups of body lymph nodes are equally enlarged. Either way, the causes can be categorised into infective and neoplastic.

Local causes of cervical lymphadenopathy

- * Infections: Tonsilitis (acute), tuberculosis (chronic)
- * Neoplastic: Metastatic spread from primaries in the head, neck chest and abdomen

General causes of cervical lymphadenopathy

- * Infective: Acute (septicaemia, infectious mononucleosis), chronic (HIV-AIDS, secondary syphilis), tuberculosis, toxoplasmosis
- * Reticuloses: Lymphatic leukemia, Hodgkin's disease
- * Sarcoidosis

The initial step to be taken in the clinical examination of any patient with established cervical lymphadenopathy therefore lies in establishing its origin: local or generalised. This can be achieved by paying attention to the following

*Thoroughly examine the areas drained by the cervical nodes: Head and neck including the oral cavity, ears and larynx (may require laryngoscopy)

*Examine the other groups of lymph nodes in the body: If other groups are involved, then it connotes lymphadenopathy due to a generalised cause

*Examine the abdomen for evidence of hepatosplenomegaly. Its presence is suggestive of reticulosis, sarcoidosis or glandular fever (infectious mononucleosis)

Tuberculous cervical lymphadenopathy and abscess: This commonly results from tonsillar tuberculosis (oropharyngeal) and most often affects the upper deep cervical lymph nodes. Nasal and pharyngeal tuberculosis are also the other known primary sites of tuberculous cervical lymphadenopathy. Laryngeal tuberculosis tends to mimic laryngeal carcinoma. There is usually no associated systemic effect. Initially, the nodes are discrete but they later mat together and undergo caseation necrosis. An abscess forms which eventually discharges more superficially through a hole in the deep cervical fascia. It is characterised by having two components, each on either side of the deep fascia and connected by a small central track. This is the so-called collar-stud abscess that is characteristic of advanced cervical tuberculosis.

Clinically, the patient presents at the early stage with a history of a painless mass in the neck. Subsequent necrosis and abscess formation will, however, render it painful. There may be constitutional symptoms including loss of weight and a possible history of exposure to a carrier. Examination may reveal a tender mass of indistinct, matted cervical lymph nodes. The skin temperature over the mass is usually normal. The slow process of caseation and formation of pus engenders minimal constitutional symptoms; hence its description as a 'cold abscess'. Other body systems, especially the lungs, should be examined for clinical features of tuberculosis. Treatment is with anti-tuberculous drugs.

Metastatic cervical lymphadenopathy: It is the commonest cause of cervical lymphadenopathy in adults. In children, however, papillary carcinoma is a known common cause. The latter was initially thought to be an 'aberrant thyroid tissue'. The lumps are usually painless and slow-growing. It is important to carry out a comprehensive examination of the head and neck including the oral cavity and the larynx in search of an occult primary. The thyroid should equally be examined particularly in children to rule out papillary carcinoma. General examination should include the breasts, lungs and abdominal structures (stomach, pancreas). Metastatic deposits from the pancreas may result in left supraclavicular lymphadenopathy. This is referred to as Virchow's node while the overall phenomenon is referred to as Troisier's sign. Treatment is of the primary condition.

Lymphomas:

There are basically two types of lymphoma: Hodgkin's and non-Hodgkin's lymphoma. Both may present as discrete, smooth, rubbery and painless lumps in the neck. Lymphomas may also involve the tonsils and nasopharynx. Constitutional symptoms include malaise, fever, night sweats, weight loss and weakness due to anaemia. Bone pain may occur due to skeletal infiltration. Alcohol consumption may result in abdominal pain. General examination may reveal

evidence of jaundice and pallor. There may be generalised lymphadenopathy. Abdominal examination may show that the liver and/or spleen are enlarged. Treatment involves referral to the Haematologist for expert management.

Branchial cyst:

It is cystic swelling arising from a persistent cervical sinus. The latter is the product of the fusion of the second and the sixth branchial arches. It is lined by stratified squamous epithelium and contains a large amount of lymphoid tissue and cholesterol crystals. Clinically it presents in the young adult (20-25) as a painless, fluctuant mass bulging from beneath the upper one-third of the sternomastoid muscle in the anterior neck triangle. There is no associated cervical lymphadenopathy except when infected. Treatment is by surgical excision. An acquired branchial sinus may arise from inadvertent incision over an infected branchial cyst. It may also arise from incomplete excision of a branchial cyst

Carotid body tumour:

It is a rare, slow-growing tumour of the chemoreceptor tissue of the carotid body at the carotid bifurcation. It is made up of large chromaffin cells in a vascular fibrous stroma. Clinically, patient presents with a long history of swelling located in the upper one-third of the anterior triangle of the neck. There may be a history of associated pulsation over the mass and symptoms suggestive of transient ischaemic attacks. The pulsation may either be transmitted from the adjacent carotid arteries or palpable from the external carotid artery which may be running over the superficial aspect of the lump. It is important to listen for associated bruit. Diagnosis is confirmed by arteriography. Treatment is surgical and involves dissecting out the tumour away from the carotid sheath. Difficult cases may require graft replacement of the carotid artery. Owing to the inherent dangers involved in the surgical management, these slow-growing tumours may be managed by way of 'masterly inactivity' particularly in the elderly.

Cystic hygromata (Cavernous lymphangioma)

When a cluster of lymphatic channels fail to connect to the mainstream lymphatic pathways, it may result in a mass of lymphatic sacs that contain lymphatic fluid. This is referred to as cystic hygroma. As to be expected, it is most common in childhood and particularly after birth when it presents as a congenital disorder. Occasionally, however, it could manifest in adult life. In this case, the filling up of an apparently empty sac is precipitated by infection or trauma. Clinically, it presents as a swelling most commonly at the base of the posterior triangle of the neck. The massive variety may extend to involve the entire subcutaneous tissue of the ipsilateral side of the neck. Unusual sites include the axilla, groin, cheek and mediastinum. It consists of multiple intercommunicating cysts. These cysts are lined by a single layer of columnar epithelium and covered externally by a rim of lymphoid tissue. The cystic fluid is straw-coloured and contains cholesterol crystals. Cystic hygroma is soft, fluctuant and dull to percussion due to the fluid content. Characteristically, it is brilliantly translucent.



CYSTIC HYGROMA IN A BABY

Complications of cystic hygroma

- Compression of neighbouring structures such as trachea (respiratory obstruction) and oesophagus (dysphagia)
- Superior mediastinal syndrome in mediastinal cystic hygroma
- Obstructed labour at birth due to a large cystic hygroma
- Infection: May be spontaneous or complicate aspiration

Cystic hygroma may end up in any of the following ways

- Undergo spontaneous resolution
- Become infected with associated pain
- Rapid expansion with obstructive features such as respiratory obstruction.

Investigations include a chest radiograph to exclude mediastinal involvement and full blood count (leucocytosis may be of infective origin)

Treatment: Owing to the possibility of spontaneous regression, in the absence of obstructive symptoms, patient should be observed for two years. Active treatment modalities include

- Aspiration of the cyst: Not quite effective as recurrent rate is high and may be complicated by infection. Aspiration may, however, be beneficial as a temporary relief of obstruction
- Aspiration followed by sclerotherapy: Sclerosing agents include bleomycin and hypertonic saline. This may be used as a prelude to definitive surgery. Preoperative treatment with sclerosants have, however, been found to distort the normal tissue planes
- Surgical excision
- Tracheostomy: For the relief of respiratory distress in severe respiratory obstruction.

Sternomastoid 'tumour':

It is not actually a tumour in the true sense of the word but rather an ischaemic contracture of a segment of the sternomastoid muscle. It commonly occurs as a result of birth injury and is situated around the middle third of the anterolateral surface of the muscle. As to be expected, it is associated with painful distress and torticollis in late stages (head turned to one side). Initially, the lump is firm, solid and can be felt easily. Subsequently, it shrinks, becomes harder and may become impalpable. There may be an associated squint. Most resolve with early stretching exercises. Late presentation may require surgical excision of the affected segment of the muscle.

Thyroglossal cyst

Thyroglossal cyst is a cystic swelling of the neck that arises as a result of a developmental anomaly of the thyroid gland. The latter develops at the foramen caecum which lies at the base of the tongue. Thereafter it descends through the thyroglossal duct into its natural position in the neck. The duct subsequently closes before birth and later atrophies. Persistence of the duct after birth results in the formation of a cystic lesion known as thyroglossal cyst. It follows that thyroglossal cyst may be found anywhere between the base of the tongue and the isthmus of the thyroid gland. The most common sites, however, are below and above the hyoid bone respectively. The cyst contains a thick jelly-like fluid that may contain cholesterol crystals while the wall is lined by columnar epithelium surrounded by a rim of lymphoid tissue. Clinically, it may appear at any age but is most common between 15 and 30.



THYROGLOSSAL CYST

Examination will reveal a midline neck swelling that has the following clinical features

- Unlike a goitre which moves only on swallowing but not with protrusion of the tongue, the thyroglossal cyst characteristically displays both features. Its movement with protrusion of the tongue is owing to its close attachment to the hyoid bone. This feature is better felt by palpation.
- Swelling is globular in shape, cystic and has a smooth surface
- Swelling is mobile from side to side but with restricted up and down mobility
- It is important to examine the base of the tongue to rule out a rare lingual thyroid (ectopic thyroid tissue).

Differential diagnosis of thyroglossal cyst include



THYROGLOSSAL FISTULA

- Swellings that move up with protrusion of the tongue: sublingual dermoid and subhyoid bursal cyst
- Dermoid cyst of the neck
- Lymphadenopathy of the pretracheal and prelaryngeal lymph nodes
- Ectopic thyroid gland
- Solitary thyroid nodule involving the thyroid isthmus

Investigations for thyroglossal cyst include

- Ultrasound scan of the neck: Demonstrates the cyst and equally confirms the presence of the thyroid gland and its isthmus
- ^{123}I radionuclide scanning: Demonstrates all thyroid tissue including the ectopic variety. More importantly, it helps to identify situations where the ectopic thyroid is the only functioning thyroid tissue and whose excision will spell disaster.

Treatment is surgical and aims at both excision of the cyst as well as the entire thyroglossal tract remnant in order to forestall recurrence. Owing to the close relationship with the hyoid bone, it is prudent to equally excise the central aspect of this bone in order to ensure complete excision of the thyroglossal tract. This is a prophylactic measure against recurrence and iatrogenic thyroglossal fistula. The procedure is referred to as Sistrunk operation.

Complications of thyroglossal cyst are

- Thyroglossal fistula: This may result from an attempt to carry out an incision and drainage of what was erroneously thought to be an 'abscess'. It may also arise from an incomplete excision of the thyroglossal tract during surgery for thyroglossal cyst. Examination will reveal a cervical fistulous opening that moves with deglutition and

- also on protrusion of the tongue. Treatment is surgical and involves Sistrunk's procedure
- Recurrent infection
- Papillary carcinoma has also been reported in a thyroglossal cyst
- Laryngeal obstruction: Rare

Pharyngeal pouch:

In the true sense, this is a pulsion diverticulum of the pharynx. It is the protrusion of the pharyngeal mucosa through a defect between the thyropharyngeus which has an oblique disposition and the cricopharyngeus with transverse fibers. It is this outpouching of the pharynx through a weakness between these muscles (Killian's dehiscence) that is referred to as pharyngeal pouch. This diverticulum eventually grows into a pouch (sac). Pharyngeal pouch occurs more commonly on the left side and at middle and old age. Clinically it presents as regurgitation of food and halithosis. Aspiration pneumonitis may occur giving rise to cough that may be associated with a choking sensation. Lung abscess is a known complication and a large pouch may compress the oesophagus resulting in dysphagia. Owing to its deep-seated nature behind the sternomastoid muscle, it may not be very obvious. Confirmation is by barium meal. Chest X-ray may show features of pneumonia and lung abscess. Treatment is surgical excision through a transverse neck incision. The pouch is identified by means of a nasogastric tube/endoscopy. Currently, endoscopic excision (Dohlman's method) is the procedure of choice.

CHAPTER FOUR

APPENDICITIS

Appendicitis is the commonest abdominal surgical emergency while appendicectomy is the most commonly performed emergency abdominal surgery.

ANATOMY: The appendix is an integral part of the large intestine. It arises from the posteromedial aspect of the base of the caecum and approximately 2.5 cm below the ileo-caecal valve. Its length ranges between 1.25cm to 22cm. The appendix could, however, be so long as to abut against the duodenum.

Positions: Retrocaecal (75%), pelvic (21%), retroileal (1%), preileal (1%), and subileal (1%)

The appendicular mesentery carries with it the vascular supply which is the appendicular branch of the ileocolic artery

The appendix has a lumen which is relatively wide in the infant but frequently obliterated in the elderly while its wall consists mainly of lymphoid tissue

Pathophysiology: Appendicitis occurs as a result of bacterial infection secondary to intraluminal obstruction. The latter may be caused by lymphoid hyperplasia or faecaliths. Other aetiological obstructive factors are parasitic worms (*oxyuris vermicularis*) and tumours of the appendix and caecum. Intraluminal obstruction results in accumulation of secretions distal to the obstruction. The resultant stasis predisposes to bacterial proliferation and infection. The pressure within the lumen of the appendix is further heightened by an increased peristaltic movement in a bid to overcome the obstruction. Distension results in venous congestion with subsequent ischemic necrosis of the appendiceal wall, perforation and peritonitis. The rising incidence of appendicitis in the developing world may be due to increasing westernisation of the native diet. This includes a change in the dietary pattern from the original high-fibre native diet to the western low fibre diet. Ironically, the incidence is currently reducing in the developed world owing to the increasingly higher fibre content of their diet engendered by aggressive health education.

PATHOLOGICAL PATHWAYS OF APPENDICITIS

A Resolution: This may be followed by recurring episodes of appendicitis. The latter occurs in about 5% of patients and commonly results from administration of antibiotics to patients with early appendicitis

B Acute appendicitis: This is the acute form of presentation and may progress to stage of gangrene, perforation and peritonitis

C Appendix mass: This mass may resolve or progress to form an appendix abscess. The latter occurs when the inflamed appendix ruptures within the 'cocoon' of the appendix mass.

D Perforation and peritonitis



COMPOSITE PHOTOGRAPH OF AN INFLAMED APPENDIX (UPPER)
HISTOLOGICAL APPEARANCE OF INFLAMED APPENDIX (LOWER)

CLINICAL FEATURES OF APPENDICITIS

Appendicitis can occur at any age. The peak incidence, however, is in young adults between 20 and 30. Appendicitis is uncommon in the extremes of life owing to the wide lumen of the appendix in the infant and the almost complete obliteration of the lumen in the elderly. All the above-mentioned clinical forms will be preceded by the following basic clinical features of appendicitis

Abdominal pain: Initially midline especially periumbilical (referred or visceral pain) as the appendix shares the same T10 innervation with the cutaneous innervation of the periumbilical region. The pain subsequently shifts to the right iliac fossa (parietal pain) due to irritation of the overlying parietal peritoneum. This classical pain of appendicitis is usually accompanied by the following

- *Nausea and vomiting
- *Low grade pyrexia
- *Anorexia which may be associated with a change in bowel habit
- *A detailed gynaecological history is important in order to rule out pelvic inflammatory disease (PID) and ectopic gestation in the female while urinary symptoms should be ruled out in view of the relevant differentials.

Examination: This will reveal an ill-looking patient who may be dehydrated and may have low grade pyrexia.

Abdominal examination: There may be positive pointing sign in which the patient points to the right iliac fossa and even at the McBurney's point as the site of maximal pain. Guarding, tenderness, and even rebound tenderness may be elicited at the right iliac fossa. Tenderness is

usually most marked at the McBurney's point (junction between the lateral one-third and the medial two thirds of a line that runs between the anterior superior iliac spine and the umbilicus). The clinical examination is completed by carrying out a digital rectal examination. The importance of a pelvic examination in the female patient cannot be overemphasised as this could go a long way in the detection of gynaecological pathologies. The clinical features of appendicitis as presented above may vary depending on the position of the appendix.

Rare clinical signs of appendicitis include

- Rovsing's sign: Deep palpation of the left iliac fossa gives rise to pain in the right iliac fossa. It is due to retrograde displacement of gas from the left colon
- Psoas sign: Pain is elicited by extending the hip with the knee in full extension. This sign is due to inflammation of the psoas muscle (on which a retrocaecal appendix sits) by an inflamed retrocaecal appendix
- Obturator sign: Pain is elicited by internal rotation of the leg with both the knee and the hip flexed. This may be positive in a patient with an inflamed pelvic appendix.

It is important to point out that the above signs are rarely elicited and are therefore unreliable in the clinical diagnosis of appendicitis.

The Alvarado (MANTRELS) score

This is a 10-point scoring system designed to evaluate the relevant symptoms, signs and laboratory findings with a view to assisting in the diagnosis of acute appendicitis. It may be remembered by the mnemonic MANTRELS

Symptoms	Score
Migratory right iliac fossa pain	1
Anorexia	1
Nausea and vomiting	1
Tenderness (right iliac fossa)	2
Rebound tenderness	1
Elevated temperature	1
Laboratory (leucocytosis) (With a selective neutrophilia)	2 (white blood count: 10,000 – 15,000)
Shift to the left	1
Total	10

The Alvarado scoring system may aid the diagnosis of appendicitis as follows

- Scores 0 to 4: Low probability
- Scores 5 to 8: Probable appendicitis
- Scores 9 to 10: Highly probable appendicitis

The Alvarado scoring system may equally aid in management decisions regarding appendicitis

- Scores 0 to 4: Observation
- Scores 5 to 8: Imaging (CT or ultrasound) for definitive diagnosis
- Scores 9 to 10: Appendicectomy

Differential diagnosis of acute appendicitis

Any abdominal emergency may mimic appendicitis. Appendicitis should therefore be considered in any patient that presents with an acute abdomen. Common differentials include

- * Perforated duodenal ulcer: The effluent from the stomach and duodenum may trickle down to the right iliac fossa via the right paracolic gutter.
- * Other acute abdominal conditions: These include acute pancreatitis, acute cholecystitis, acute intestinal obstruction, typhoid perforation and Meckel's diverticulitis. Others are gastroenteritis, acute Crohn's ileitis, psoas abscess and right ureteric colic
- * Specifically In the female: Pelvic inflammatory disease, Mittelschmerz (ovulation, mid-cycle) pain, and ruptured ectopic pregnancy. Others are complicated right ovarian cyst (rupture or torsion) and degenerating fibroids.
- * Specifically In childhood: Mesenteric adenitis and right-sided basal pneumonia. Mesenteric adenitis is an inflammatory condition of viral origin that results in painful lymphadenopathy of the mesenteric lymph nodes which drain the small intestine
- * Medical conditions: Abdominal crisis of sickle cell disease, acute intermittent porphyria, diabetes mellitus and acute myocardial infarction

Investigation: Diagnosis is basically clinical. Diagnostic investigations may be employed to confirm the clinical diagnosis particularly in difficult cases. This is in a bid to reduce the negative appendectomy rate. Other investigations are made to aid in the preparation of the patient for surgery and include

- * Full blood count: Packed cell volume and white blood count which may reveal polymorphonuclear leucocytosis
- * Electrolytes, urea and creatinine: This is very relevant in a patient who has been vomiting
- * Urinalysis: Microscopic haematuria may occur with appendicitis. Pus cells in the urine may either be due to bladder irritation from a pelvic appendicitis or secondary to urinary tract infection. Microscopy haematuria, on the other hand, raises the possibility of ureteric colic as a differential diagnosis
- * Abdominal ultrasound scan (USS): This has a high sensitivity (80%) as well as specificity (81%) in the diagnosis of acute appendicitis. It is, however, less specific for perforated appendicitis. In addition to aiding the diagnosis of appendicitis, abdominal USS helps to rule out other differentials particularly in young females. USS helps in the screening of the pelvis in the latter group of patients
- * Computer tomography (CT) scan: Though highly sensitive (94%) as well as specific (95%), CT is rarely employed as a routine investigation in the diagnosis of appendicitis. This is because it has a limited sensitivity both for early appendicitis as well as pelvic pathology. It may, however, be useful when there is a diagnostic dilemma in an inflammatory process not related to pelvic pathology
- * Diagnostic laparoscopy: This is especially useful in young females with suspected appendicitis in order to rule out pelvic pathology. Diagnostic laparoscopy will help to reduce the negative appendectomy rate in this group of patients. It is, however, a surgical procedure with its attendant morbidity. Diagnostic laparoscopy is therefore regarded as a last resort in the evaluation of patients with non-specific clinical or radiographic evidence of

inflammation or pathology. Provision should be made to proceed to surgical management on confirmation of diagnosis

* Human chorionic gonadotrophin assay in women of child bearing age to rule out ectopic pregnancy

Treatment of uncomplicated acute appendicitis: In the Accident and Emergency Unit

- *Administer intravenous infusion (in the process take blood for full blood count, electrolytes, urea and creatinine).
- * Commence on nil orally
- * Nasogastric aspiration if patient is vomiting
- * Commence on intravenous antibiotics (include metronidazole for anaerobic infection)
- * Administer adequate intravenous analgesic. Contrary to a long-held view, adequate analgesia rather aids in enhancement of the abdominal clinical signs as reduction of abdominal pain tends to elicit more cooperation from the patient.

Definitive treatment of uncomplicated acute appendicitis: The treatment of acute appendicitis is emergency appendicectomy. This may be carried out either by the open method or laparoscopically. The latter has the unique advantages of affording an extra ability to inspect the abdominal organs (especially the pelvis in the female adult), as well as ensuring a more rapid recovery. Some studies have, however, shown that the incidence of intra-abdominal abscess is higher in laparoscopic than open appendicectomy. The open approach may therefore be a better option when there is a high suspicion of significant contamination. To prevent recurrent symptoms, it is important to ensure that the entire length of the appendix is excised. This has been reported as one of the complications of laparoscopic appendicectomy. Some authors are reluctant to recommend laparoscopic appendicectomy in children.

It is adviseable to remove an apparently normal-looking appendix found at surgery in order to prevent a future diagnostic dilemma. This is important as studies have shown that about 58% of apparently normal-looking appendix have histological features of inflammation.

Perforated appendicitis

Clinical features: The history is initially as for acute appendicitis but the abdominal pain which initially was localised in the right iliac fossa later becomes generalised. Nausea and vomiting are equally more prominent. Abdominal distension and high grade fever result from the ensuing bacterial peritonitis.

On examination, patient is ill-looking and may present with clinical features of dehydration in the early stage or of hypovolemic shock in a later stage.

The abdomen may be full or distended with generalised guarding and tenderness most prominent at the right iliac fossa. Bowel sounds may be reduced or absent

Digital rectal examination is mandatory.

Emergency management of perforated appendicitis is as follows:

- Commence on nil orally
- Naso-gastric aspiration is mandatory as it helps to decompress the gastrointestinal system.
- Intravenous infusion: Commence with normal saline. Take blood samples for electrolytes/urea/creatinine and group and crossmatch blood as the definitive treatment entails exploratory laparotomy
- Urethral catheterisation is carried out in order to monitor the urinary output. This acts as a guide in the rehydration exercise
- Plain abdominal x-rays (erect and supine) should be carried out after resuscitation and may demonstrate the presence of gas under the diaphragm.
- Surgical management: Entails exploratory laparotomy, appendicectomy, peritoneal lavage and insertion of tube drains
- Postoperative management: Continue intravenous antibiotics and infusion, nasogastric aspiration and analgesia. Remove tube drain when effluent is minimal.

APPENDIX MASS

Appendix mass is an inflammatory mass formed by the wrapping-round of the inflamed appendix by a 'cocoon' consisting of the caecum, coils of small intestine, and loop of the greater omentum ('the abdominal policeman'). It is nature's way of trying to contain the infection and limiting it from spreading to the general peritoneal cavity.

History in perforated appendix is essentially as for acute appendicitis but the duration of ailment is longer as the patient could have been experiencing right iliac fossa pain for 72 hours or more prior to presentation.

Findings on clinical examination are as for acute appendicitis. In addition, there is a palpable, tender mass in the right iliac fossa. Diagnosis should ideally be made before the commencement of treatment. Particularly in the obese patient, the mass may initially be vague/obscure and only distinctly palpable after administration of adequate analgesia or when the patient is more relaxed under anaesthesia.

Differential diagnosis of a right iliac fossa mass includes masses that are located in both the intra- and retro-peritoneal compartments of the abdomen

Intra-abdominal masses

- * Appendix mass
- * Appendix abscess
- * Carcinoma of the caecum or ascending colon (especially in the elderly)
- * Crohn's ileitis
- * Ileocaecal tuberculosis
- * Right ovarian cyst
- * Pedunculated fibroid
- * Huge, distended gallbladder

Retroperitoneal masses

- Retroperitoneal sarcoma
- Iliopsoas abscess
- Tumour in an undescended right testis
- Pelvic kidney: May be normal, hydronephrotic or neoplastic.
- Paraganglioma

Investigation: As for acute appendix. In addition, serial abdominal ultrasound scan is essential both for confirmation of diagnosis as well as to rule out other differentials. Serial ultrasound scan is also useful in monitoring the progress of the mass during the period of conservative management.

Treatment: This is essentially conservative as unguarded surgical intervention at this early stage may predispose to enterocutaneous fistula. The principle of conservative management is generally referred to as the Ochner-Sherren's regimen and consists of the following

- Stop oral intake if patient is vomiting but continue with oral fluids if there is no vomiting
- Nasogastric aspiration if patient is vomiting
- Intravenous infusion if patient is vomiting and cannot tolerate oral intake
- Antibiotics including metronidazole for anaerobic organisms
- Monitor vital signs: 3-hourly monitoring of the pulse rate, blood pressure, temperature and respiration
- Symptomatic monitoring of patient's clinical progress: Pain, vomiting, ambulation and tolerance of oral intake
- Delineate the perimeter of the mass with a felt pen and serially monitor same by abdominal examination. Serial ultrasound scan is more ideal in this situation.
- Serial ultrasound scan: Monitors the size of the mass. Demonstration of presence of pus is indicative of progression to appendix abscess.

Positive clinical progress is indicated by a gradual reduction in the size of the mass as well as stabilisation of the vital signs (especially by a reduction in the pulse rate). An improvement in the general wellbeing of the patient (decreased vomiting, improved appetite and ambulation) is also an indication of a positive response to the conservative treatment. Serial abdominal USS will show a reduction in the size of the appendix mass

It is worth emphasising that if the diagnosis is made on abdominal palpation when patient is already under anaesthesia, he should be reversed and managed conservatively in the ward. Same principle applies to a diagnosis made at surgery. In this case, the abdomen should be cleaned and patient closed up for continuation with the conservative regimen in the ward. Appendicectomy is not adviseable even at this stage, as it still bears a high risk of a postoperative enterocutaneous fistula. The only exception is when a 'free-floating' appendix is encountered at surgery and literally 'begging' for removal

Most patients respond to conservative management as evidenced by the resolution of the mass. Non-resolution, as evidenced by a progressive increase in size of the mass, a rising pulse rate coupled with degeneration in the overall wellbeing of the patient connotes deterioration of the pathology to formation of an appendix abscess (see below). This calls for surgical intervention.

Particularly in older patients, it is important to further investigate the patient after resolution of the mass. This is meant, among others, to exclude some of the more sinister differentials listed above. The relevant investigations include abdominal ultrasound scan, computerised tomography and colonoscopy. Barium enema is an alternative investigation when there are no facilities for colonoscopy.

Traditionally, the patient is readmitted 6 to 12 weeks after resolution of an appendix mass for an interval appendicectomy. This trend has, however, been reviewed as the patient is currently made to 'earn' the subsequent surgery ONLY if symptoms persist. Studies have shown that most patients remain symptomless after a successful conservative management of an appendix mass (only 3 to 15% of patients have recurrent symptoms). This has helped to reduce the problems associated with interval appendicectomy. At surgery, the appendix is usually found to be enmeshed in fibrosis with associated difficulty in its accessibility and excision. Even when eventually located, the appendix is found to be shrivelled. It is most probable that the high degree of inflammation associated with the pathogenesis of an appendix mass may have literally 'eaten up' the appendix.

Management of appendix abscess: The age-long surgical rule of drainage being the best way of managing an abscess equally applies in the case of appendix abscess. Patient requires an incision and drainage procedure which may be carried out either percutaneously under ultrasound guidance or by open surgery through the traditional appendicectomy incision. The Rutherford Morrison's muscle splitting incision is preferred in this instance as it makes room for adequate drainage. It involves the cutting of the internal oblique and transverses muscles in the line of the skin incision. Again, no deliberate effort aimed at appendicectomy should be made as this equally carries a high risk of postoperative enterocutaneous fistula. Not uncommonly, however, the appendix may be found floating in the pool of pus. Under this situation, a careful appendicectomy should be carried out with clearance of any observed faecalith. Owing to the fragile nature of the base of the caecum in this situation, no attempt should be made to bury the stump of the appendix. Ability to remove faecaliths and clean up the periappendiceal region constitutes the main advantage of open drainage over its percutaneous, ultrasound-guided counterpart in the surgical management of appendix abscess.

Drainage may be carried out either transrectally or transvaginally when pelvic abscess complicates pelvic appendicitis

Recurrent appendicitis: Ultimately managed by elective appendicectomy

Note the four types of appendicectomy

- Elective appendicectomy for acute appendicitis
- Emergency appendicectomy for recurrent appendicitis
- Interval appendicectomy following a recurrence of symptoms after the management of an appendix mass or abscess (no longer routinely practised)
- Incidental appendicectomy: When an apparently normal appendix is excised in the course of laparotomy for an entirely different surgical condition. It is no longer practised routinely as the healthy appendix may prove useful in some other procedures, especially in Urology.

Complications of appendicectomy

- 1 Haemorrhage: Ranges from bleeding into the wound to severe intraperitoneal haemorrhage due to slipping of the ligature over the stump of the appendicular artery. The latter may result in hypovolemic shock. Treatment involves the following
 - Intravenous infusion
 - Group, crossmatch and transfuse if necessary
 - Serial monitoring of the packed cell volume
 - Re-exploration, identification and transfixtion of the bleeding stump of the appendicular artery
- 2 Infection: Ranges from mild to moderate
 - Wound infection particularly after dealing with a gangrenous appendix. Treatment entails drainage of any abscess, regular wound dressing and use of appropriate antibiotics. Consider delayed primary closure in moderate to severe contamination.
 - Intra-abdominal abscesses (pelvic and subphrenic): Treatment involves drainage either by the open method or percutaneously under ultrasound guidance. The transrectal route may be employed when dealing with a pelvic abscess
 - Portal pyaemia: This results from the spread of infection from a gangrenous appendix to the liver via the portal vein. Clinical features of portal pyaemia may manifest either before or after an emergency appendicectomy. The cardinal features are jaundice, fever and hepatomegaly. Treatment involves the administration of intravenous infusion and antibiotics (including metronidazole).
- 3 Enterocutaneous fistula: See below
- 4 Damage to neighbouring structures

CHAPTER FIVE

ENTEROCUTANEOUS FISTULA

A fistula is an abnormal communication between two epithelial surfaces lined by granulation tissue. An enterocutaneous fistula is therefore an abnormal communication between a segment of the intestine and the skin. It is associated with a high degree of morbidity and mortality (10%). Management of enterocutaneous fistula is challenging, frustrating but ultimately rewarding. A good knowledge of the relevant pathophysiology is essential for proper management of these very ill patients.

Aetiology:

- Postoperative: Commonly occurs following an inadvertent damage to the intestine in the course of an abdominal surgery. Enterocutaneous fistula may occur following failed resection and anastomosis of gut and operations requiring extensive adhesiolysis. It may also complicate surgery for peritonitis such as typhoid perforation and drainage of pelvic and appendix abscesses. The commonest predisposing surgery is, however, appendicectomy. It is usually sequel to an ill-advised appendicectomy in the presence of an appendix mass. In the process of delivering the appendix which is deeply buried in the inflammatory mass, an inadvertent damage could be inflicted on the adjoining intestine. This will manifest a few days after surgery as an enterocutaneous fistula. It is commoner following emergency surgeries. This may result from poor operative management
- Intestinal neoplasms: Colonic cancer
- Radiotherapy
- Chronic inflammatory conditions: Crohn's disease and tuberculosis
- Diverticulitis.
- Spontaneous enterocutaneous fistulae have been reported

Fistulas related to malignancy, irradiation and inflammatory bowel disease are less likely to close spontaneously. Surgery for small bowel volvulus and strangulated hernia may also be complicated by enterocutaneous fistula. The ileum is the most commonly involved part of the gastrointestinal system.

Classification of enterocutaneous fistula: This may be anatomical or physiological

- Anatomical: This depends on the anatomical segment of the involved viscera: Gastric, jejunal, ileal or colonic. Location of the fistula may suggest its aetiology, management strategy and prognosis. The higher the anatomical level of the fistula, the higher the output and the more difficult the management.
- Physiological: This is based on the fluid output of the fistula - high output (400mls and above in 24hours), moderate output (200-400mls in 24hours) and low output (less than 200mls in 24 hours). Moderate to high output fistulae are most likely small intestinal in origin while the low output variety is usually of colonic origin. Higher

output fistulae are more likely to be associated with electrolyte imbalance and malnutrition.

Clinical features: Patient may complain of staining of the wound dressing by faeculent, foul-smelling discharge. This is noticed a few days after surgery. Relevant information should be sought in relation to the other predisposing factors. There may be fever due to associated infection, loss of weight and dehydration due to fluid loss. The typical history is that of fever and persistent discharge occurring 5 to 6 days after an index abdominal surgery. Examination may reveal clinical features of a 'wound abscess'. There is an apparent resolution of the fever after drainage. This is, however, followed 24 hours later by clinical manifestation of the fistula as the dressing over the wound becomes stained with obvious faeculent discharge. It is important to know if there is an associated change in bowel habit since the onset of illness. Constipation may connote a distal obstruction. This is one of the factors that may hinder closure of an enterocutaneous fistula.

Examination will reveal an ill-looking patient who may be pale and dehydrated. In extreme cases, patient may manifest with evidence of hypovolemic shock. The dressing over the wound will be stained with faeculent effluent.

Pathophysiology and challenges in the management of enterocutaneous fistula

- Loss of fluid and electrolytes: The higher the anatomical location of the segment of the intestine involved, the higher the degree of fluid loss; hence the output of a jejunal fistula will be higher than that of its colonic counterpart.
- Malnutrition due to loss of nutrients which rather than being absorbed are lost through the fistula. Again, the higher the fistula anatomically, the worse the degree of malnutrition
- Anaemia: This is sequel to malnutrition.
- Wound infection: There may also be occult intra-peritoneal abscesses
- Excoriation of the skin: This is the effect of digestive enzymes on the adjacent skin
- Psychological impact of the clinical state may result in depressive illness

MANAGEMENT OF ENTEROCUTANEOUS FISTULA

This can be divided into five phases

- Phase 1: Recognition and stabilisation
- Phase 2: Investigation
- Phase 3: Stage of decision
- Phase 4: Definitive surgical treatment
- Phase 5: Healing

PHASE1 - RECOGNITION AND STABILISATION:

Clinical diagnosis can easily be made from the history and clinical examination. Clinical evidence of infection and loss of fluid and electrolytes should be ascertained. There may be fever, and clinical signs of dehydration/shock. The detection of faeculent staining of the wound dressing completes the clinical diagnosis. It is most important to address the urgent clinical problems

outlined above. Initial treatment is aimed at addressing the above-mentioned challenges and should be conservative as immediate surgical intervention may worsen the prognosis. The natural reaction when challenged with a case of enterocutaneous fistula is to re-explore the patient. This urge should be resisted as surgery at this stage entails major adhesiolysis with the attendant risk of haemorrhage and further damage to the intestine. Furthermore, intestinal re-anastomosis at this stage carries a high risk of anastomotic breakdown. The following are the principles of conservative management of an enterocutaneous fistula

- Fluid and electrolytes: Prior to the administration of fluids, blood samples should be taken for FBC, electrolytes, urea and creatinine. Equally send a blood sample for grouping and crossmatching. Pass a urethral catheter for adequate monitoring of the fluid input.
- Correct existing anaemia by blood transfusion
- Rule out the presence of intra-abdominal collections by carrying out abdominal USS, CT scan and water-soluble contrast examination.
- Drain any intra-abdominal abscess detected. Take a swab of the purulent discharge for microscopy, culture and sensitivity (m/c/s). Commence on broad-spectrum antibiotics. This should include metronidazole in view of possible anaerobic infection. The antibiotic regimen should be reviewed on receiving the report of the m/c/s.
- Care of the surrounding skin starts by the proper collection and estimation of the effluent. This is important both for monitoring of the progress of the condition as well as for prognostication. A tube drain or a colostomy bag may be employed. In the alternative, cotton wool completely wrapped up with gauze (gamgee) may be used and the frequency of change of dressing aids in monitoring the progress of the condition. Meanwhile, a barrier cream such as zinc oxide or petroleum jelly (Vaseline) is applied over the skin adjacent to the fistulous opening. This protects the surrounding skin from the macerating effect of the gastrointestinal digestive enzymes that are present in the fistula effluent. The quantity of the latter may be reduced by the administration of Octreotide which is a somatostatin analogue. The latter acts by reducing the volume of gastrointestinal secretions. This facilitates skin care and enhances closure of fistula especially those of biliary and pancreatic origin.
- Nutritional support: In addition to the ongoing nutritional loss, enterocutaneous fistula sets up a hypercatabolic state. The baseline nutritional status can be readily assessed by estimating the total serum protein, albumin and transferrin. Determination of the caloric requirement is based on patient's activity level and can be estimated by indirect calorimetry. Protein requirement is output-dependent: low output 1-1.5G/day; high output 2-5G/day.

Ideally patient should be commenced on a form of nutritional support. Closure rate is lower with enteral than with parenteral nutrition. Parenteral nutrition completely rests the gut and enhances closure. Most times, however, it is neither available nor affordable. In this case, patient could be placed on a low residue diet particularly if the fistula is anatomically of the low variety. This may be supplemented with intravenous glucose and Astymin which contains protein supplements. Essential vitamins, magnesium, zinc and other trace elements should be given.

Despite its limitations, the enteral route of nutrition is preferred where possible as it provides both immunological and hormonal functions. A minimum small intestinal length of 120cm is required. Enteral nutrition is, however, contraindicated in severe ileus and proximal gut fistula. Nasogastric intubation should be avoided except when absolutely indicated (ileus or mechanical obstruction). Apart from the associated discomfort, it may predispose to aspiration pneumonia, oesophageal stricture and reflux oesophagitis. Gastrostomy is preferred for long-term decompression.

It is important to clear stool that may be present in the distal bowel in order to ensure patency and prevent distal obstruction. This is accomplished by the administration of soap and water enema.

- Psychological care: Enterocutaneous fistula is a highly depressing condition. The patient will benefit from psychological support in order to maintain good morale. In addition to reassurance by the attending clinician, an enterostomal therapist and/or a psychiatrist should be consulted if necessary.

Progress is assessed by monitoring the 24-hour effluent as well as the general condition of the patient. Positive progress is demonstrated by a consistent decrease in the quantity of effluent as demonstrated by an absolute measurement or a decrease in frequency of change of wound dressing.

PHASE 2: INVESTIGATION

In addition to confirming the presence of a fistula, it is equally important to define its anatomical location. As stated earlier, this will help both in the management and in determining the possible prognosis.

- Dye test: Patient ingests a dye which is ultimately detected in the effluent. Congo red, methylene blue or charcoal may be used.
- Fistulogram: Involves both the surgeon and the radiologist. Fistula is cannulated with a small feeding tube or catheter into which a water-soluble contrast (gastrograffin, lipiodol) is injected. This procedure defines the tract.
- Gastrointestinal series: Barium enema and small bowel enema may be employed to define the tract in difficult cases
- CT scan and abdominal ultrasound scan

Emerging questions from this assessment include

- What segment is involved?
- Is bowel wall defect larger than 1cm?
- Is bowel completely disrupted?
- Does fistula communicate with distal bowel?
- Does fistula arise from the lateral bowel wall?
- Is there an associated abscess cavity?
- Is adjacent bowel damaged, strictured or inflamed?
- Is there a distal obstruction?
- What is the length of the fistula?

The answers to these questions will assist in identifying fistulas whose anatomic features may hinder spontaneous closure of the fistula. These include

- Fistulae involving the stomach, ileum and jejunum
- Fistulae with tract length greater than 2cm and with defects larger than 1cm
- Fistulae with complete disruption of bowel wall
- Poor quality of adjacent bowel
- Presence of a large abscess cavity
- Fistulae with distal obstruction

PHASE 3: STAGE OF DECISION

The main aim of management is to create a favourable environment for closure of fistula and reestablishment of gastrointestinal continuity. Spontaneous closure is ideal. It is therefore important to persevere with conservative management which may drag on for a while. This is because the expected time of healing of a fistula varies with its anatomical site. Whereas colonic fistulas are expected under favourable condition to heal in 30 to 40 days, the duration for the small intestinal variety is 40 to 60 days. In addition, surgical intervention in the management of fistula is not a particularly easy surgery due to adhesions and haemorrhage encountered in the course of adhesiolysis. The golden rule therefore remains: 'MAKE HASTE SLOWLY'.

Factors that affect the healing of a fistula can be remembered with the mnemonic "FRIENDS" viz

F - Foreign body,

R - Radiation,

I - Infection,

E- Epithelialisation

N - Neoplasms,

D - Distal obstruction

S - Sepsis/steroids

PHASE 4: Definitive surgical treatment

Indications:

- Anatomic features based on initial clinical and radiological assessment precluding spontaneous closure
- Anatomic favourable fistula that does not close in expected time (4-5 weeks) despite a sepsis-free environment and adequate nutritional support. This manifests as minimal/no reduction in the volume of the effluent despite properly supervised conservative management.
- Suspicion of intraperitoneal collection

Definitive surgery: Surgical options are

- Drainage of abscess
- Resection and anastomosis: preferred if possible
- Exteriorisation of the fistulating segment
- Wedge excision of the fistulating segment

Preoperative management

- Appropriate bowel preparation: mechanical as well as chemical
- Prophylaxis for deep venous thrombosis
- Ensure adequate nutrition; meanwhile discontinue enteral nutrition
- Group and cross -match blood

- Discuss the possibility of a stoma, mark the possible site
- Secure an informed consent.

Operative surgery

- Conduct a thorough search for any pocket of pus and ensure adequate drainage
- Access through a fresh incision (median or paramedian). May use previous scar if well healed.
- Meticulously dissect and free bowel both proximal and distal to the affected site
- Resect the affected segment of bowel and reconstitute the tract with an end-to-end anastomosis
- As much as possible, minimize the addition of other procedures except when such supplementary procedures are absolutely necessary.

PHASE 5: Healing

Postoperative management

- Nasogastric aspiration as prolonged ileus is common
- Continue antibiotics for at least 72 hours
- Nutritional support: Gradually (re)introduce enteral nutrition as the parenteral route may prove inadequate later
- Wean and resume oral intake. The dietitian and family may be of assistance.

Prognosis of enterocutaneous fistulae

For prognostication, Ajao et al have classified enterocutaneous fistula into four anatomical types

Class	Mortality
Type I: Gastroduodenal fistulae	20%
Type II: Small bowel fistulae	33%
Type III: Large bowel fistulae	17%
Type IV: Any fistula coexisting with significant skin involvement ('entero-atmospheric')	60%

From their experience, large bowel fistulae (type III) have the best prognosis followed by the gastroduodenal variety. The 'enteroatmospheric' (Type IV) has the worst prognosis

Prevention of enterocutaneous fistula

- Strict adherence to established surgical principles in performing intestinal anastomosis and management of appendix mass
- In the surgical management of intestinal obstruction, as much as possible, one should endeavour to decompress the gut without an enterotomy. All intestinal tears should be addressed at surgery
- Always remember to make haste slowly in the management of established enterocutaneous fistula as most of them will close spontaneously on strict conservative management. An ill-advised, hasty surgical re-intervention may worsen an already difficult situation.

CHAPTER SIX

THE BILIARY SYSTEM

The biliary system is important both in health and in disease. It is responsible for the manufacture and secretion of bile.

Anatomy of the biliary system

Bile is produced in the liver and secreted into the biliary canaliculi located in the substance of the liver. It exits from the liver through the right and left hepatic ducts. At the hilum of the liver, the hepatic ducts confluence to form the common hepatic duct. A little down the line, the hepatic duct receives the cystic duct which drains the gallbladder and thereafter becomes the common bile duct. This passes posterior to the second part of the duodenum and is joined by the main pancreatic duct to form a common channel of opening into the second part of the duodenum after forming the ampulla of Vater. The sphincter of Oddi marks the point of this opening.

Pathophysiology of bilirubin

Bilirubin is the product of destruction of effete red blood cells. The red blood cells are split into the two component parts of haem and globulin. Bilirubin is a product of biliverdin which is formed from the haem component. The unconjugated bilirubin is transported to the liver where it is conjugated and rendered soluble. Thereafter, it forms a major component of bile which is initially stored in the gallbladder and later secreted into the second part of the duodenum by the stimulatory effect of cholecystokinin. Within the intestine, bilirubin is degraded into stercobilinogen which imparts the natural colour to stool. Stercobilinogen is also absorbed into the blood and is excreted in the urine as urobilinogen.

THE GALLBLADDER

The gallbladder is a pear-shaped hollow viscus that is situated in the gallbladder bed below the right lobe of the liver. Its inferior relation is the second part of the duodenum while its contents drain into the biliary system through the cystic duct.

The gallbladder stores bile into which it secretes mucus. On stimulation by the hormone cholecystokinin, it contracts and discharges its contents into the biliary system through the cystic duct.

CHOLELITHIASIS (GALLSTONES)

Gallstones are formed in the gallbladder but their consequences far transcend the boundaries of this organ. It is commoner in females and was said to occur mainly in "fair, fat, fertile, females at forty". This is, however, not entirely true

Types of gallstones:

- Pure cholesterol stones: Usually single and may be large. They are usually radiolucent and cast an acoustic shadow on ultrasonography

- Pigment stones: Of two types – black and brown. The black pigment stones are formed in the gallbladder and are multiple. They are associated with haemoglobinopathies and are precipitates of bile supersaturated with calcium bilirubinate. Brown pigment stones, on the other hand, are primary bile duct stones and are usually associated with biliary infection due to gram-negative organisms such as E.coli and bacteroides. These organisms elaborate beta-glucuronidase which hydrolyses conjugated bilirubin resulting in the formation of calcium bilirubinate. They contain calcium bilirubinate, calcium palmitate and small quantity of cholesterol. Infestation of the biliary tract with ascariasis sets up a similar pathology and equally results in the formation of brown stones.
- Mixed stones: The main component of mixed stones is cholesterol. In addition to a protein matrix, other components are calcium carbonate, calcium bilirubinate and calcium palmitate. About 10% of mixed stones are radiopaque

Factors that predispose to formation of gallstones

A Metabolic derangement: Cholesterol is kept in solution by bile salts and lecithin. A delicate balance is maintained between bile salts/lecithin on one hand, and cholesterol on the other. Hence, either a deficiency of bile salts/lecithin or a high concentration of cholesterol will result in precipitation of cholesterol stones. The normal ratio of concentration of bile salts/lecithin and cholesterol is 10:1. Some cholesterol will remain in solution as vesicles. Aggregation and fusion of the vesicles will result in crystallisation of cholesterol. Further deposition of cholesterol crystals results in the formation of gallstones. Metabolic factors in the formation of gallstones can be summarised as follows:

- High concentration of constituents (lithogenic bile): Contraceptive pills, clofibrate, obesity, total parenteral nutrition and hyperlipidemia. Chronic haemolysis as occurs in chronic haemolytic conditions such as haemoglobinopathy and haemochromatosis may result in the formation of pigment stones.
- Deficiency of bile salts: Bile salts are reabsorbed in the terminal ileum. Ileal disease or ileal resection will interfere with the enterohepatic circulation of bile salts and therefore result in deficiency of bile salts available for keeping cholesterol in solution. Cholestyramine therapy, by mopping up bile salts, also has the same effect

B Stasis: Truncal vagotomy, pregnancy, total parenteral nutrition and prolonged immobilisation.

C Infection: E.coli, ascariasis, and clonichis sinensis. Remember Moynihan's aphorism "A gallstone is a tombstone erected in memory of the organism within it". Microorganisms trapped by the liver are excreted through bile. They will continue to proliferate even when trapped by cholesterol crystals. The glucuronidase secreted by these organisms results in deconjugation of bilirubin. The end result is precipitation and formation of mixed stones

PATHOLOGICAL EFFECTS OF GALLSTONES

As pointed out earlier, gallstones are formed within the gallbladder but their effects reverberate far beyond the organ of origin. The pathological effects can be categorised according to the structure in which they are lodged.

Gallbladder:

- **Asymptomatic gallstone:** May be an incidental finding when investigating for an unrelated abdominal pathology
- **Biliary colic:** It is due to the presence of gallstone in the infundibulum. It presents as intermittent attack of spasmodic right upper quadrant pain which may be aggravated by fatty meals. Biliary colic is due to cholecystokinin (CCK)-stimulated gallbladder contraction following food ingestion. The average duration of this pain is usually less than 6 hours. It is less commonly due to gallbladder dysfunction. Pain may radiate to the shoulder region. Characteristically, the patient is afebrile and the white cell count and liver function tests are normal. On the other hand, in acute cholecystitis/acute cholangitis, the patient is febrile and the laboratory results are abnormal. Treatment of biliary colic is by elective cholecystectomy
- **Inflammation of the gallbladder (cholecystitis):** May be acute or chronic. As mentioned above, the term 'biliary colic' has been used to describe the clinical condition that results from impaction of a gallstone within the gallbladder infundibulum or the cystic duct. The current thinking is that biliary colic and acute cholecystitis are part and parcel of the same clinical spectrum.

The initial inflammatory process in acute cholecystitis is sterile in nature. Obstruction and stasis later result in bacterial colonisation. The pressure effect on the wall of the gallbladder may result in gangrene and perforation of the gallbladder. The latter will result in biliary peritonitis. If there is impaction of stone at the Hartman's pouch, the gallbladder could distend with fluid whose nature will depend on the microbiological state of the on-going pathological process. It will either form an empyema when there is an underlying infection or a mucocoele when there is no infection.

- **Carcinoma of the gallbladder:** This may arise from the presence of longstanding gallstones. The resultant chronic irritation may induce a metaplastic change
- **Common bile duct:** Impaction of a stone in the lumen (choledocholithiasis) will result in obstructive jaundice. Dilatation and inflammation of the proximal duct will result in ascending cholangitis
- **Pancreas:** Gallstones constitute one of the most common causes of acute pancreatitis
- **Intestine:** Translocation of gallstones into the small intestine may result in gallstone ileus. The latter has an interesting pathogenesis. The resultant necrosis from a severe acute inflammation of the gallbladder wall may spread to involve the adjoining duodenal wall. This results in the creation of a fistulous communication between the gallbladder and the duodenum. The ingress and subsequent movement of these gallstones from the duodenum to the terminal ileum may result in impaction and mechanical obstruction at this narrow part of the gut. In addition to demonstrating the features of intestinal obstruction, a plain abdominal X-ray may demonstrate the presence of a calculus in the terminal ileum as well as the presence of gas in the biliary tree. This condition is, however, rare.

ASYMPTOMATIC (SILENT) GALLSTONES: Not all patients with gallstones will manifest with symptoms. It is not unusual to pick up the diagnosis of gallstones on abdominal ultrasound scan in the course of investigation for an unrelated ailment. The incidence is 10% in men and 20% in

females. Only about 1% of patients will develop symptoms per annum. In view of the possible complications of gallstones, the following are the indications for surgery in patients with silent gallstones

- Large gallstones (> 2.5 cm)
- Multiple small gallstones
- Diabetic patients: High risk of emphysematous cholecystitis
- Patients on immunosuppressive therapy
- Porcelain gallbladder: Calcified gallbladder may undergo metaplastic change resulting in carcinoma of the gallbladder
- Patients listed for renal transplantation
- Patients living in areas where the incidence of carcinoma of the gallbladder is high.
- Surgery is advised in the younger age group who may live long enough to develop complications.
- Patients with haemoglobinopathies

ACUTE CHOLECYSTITIS

There are three varieties of acute cholecystitis

- Calculous acute cholecystitis: This is initiated by the presence of gallstones. The gallbladder is distended with thick and oedematous wall. Dilated vessels and areas of patchy gangrenous change may be visible in the latter. The wall of the gallbladder is infiltrated by acute inflammatory cells. The pathogenic organisms include E.coli (commonest), proteus and salmonella. Others are non-haemolytic streptococci pseudomonas and clostridium. The main route of infection is biliary. Haematogenous spread may also occur via the cystic artery. Complications of acute cholecystitis include gangrene and perforation (at fundus or the site of impaction of the stone). Others are mucocoele and empyema of the gallbladder respectively
 - Mucocoele of the gallbladder: Obstruction of the cystic duct by a gallstone may result in the absorption of the contained bile by the gallbladder epithelium. This is followed by secretion of mucus by the latter resulting in distension of the gallbladder. The contained mucus is usually sterile
 - Empyema of the gallbladder: Same pathophysiology as mucocoele and may indeed result from infection of the latter.
- Acalculous cholecystitis: Accounts for about 5% of acute cholecystitis. Predisposing factors include gallbladder ischemia (patients with hypovolemic shock from any cause including multiple trauma), and biliary stasis (prolonged fasting and hyperalimentation). It may follow major surgery, burns or trauma. No wonder it is common in critically ill patients on admission in the intensive care unit. The acute inflammation of the gallbladder may be as a result of microvascular ischemia.
- Emphysematous cholecystitis: This is both uncommon and insidious. It is secondary to gas-forming organisms and is characterised by the presence of gas in the lumen and wall of the gallbladder. In severe cases, there may be extension of gas into the pericholecystic soft tissue as well as the biliary ducts. About one-third of patients have associated diabetes mellitus. It is associated with high morbidity and mortality

Clinical features of acute cholecystitis: There is acute onset of abdominal pain located around the right hypochondrial region. The pain may radiate to the tip of the right shoulder (Boas sign) or through to the back. There may be associated fever, nausea and vomiting. The pain is spasmodic initially but later becomes constant.

On examination, the patient is usually ill-looking, tachycardic and pyrexic. There may be clinical features of dehydration and even shock due to loss of fluid through vomiting. Jaundice may be present. Abdominal examination will reveal guarding and tenderness over the right hypochondrium. With a hand placed over the gallbladder area in the right hypochondrium, the patient is likely to catch his breath on deep inspiration. This mid-inspiratory arrest is referred to as Murphy's sign. It is due to the movement of the gallbladder on deep inspiration into the waiting hand of the examiner.

There are some peculiar modes of presentation associated with the rare forms of acute cholecystitis. For instance, the diagnosis of acalculous cholecystitis should be entertained in postoperative or acutely ill patients with right upper abdominal pain, and unexplained fever occurring 2 to 4 weeks after injury.

Emergency management of acute cholecystitis

- Set up an intravenous infusion.
- Take blood samples for full blood count (leucocytosis), electrolytes, urea and creatinine.
- Pass a nasogastric tube if patient is vomiting and place on nil orally. On the other hand, oral fluids may be permitted in a patient who is not vomiting.
- Administer intravenous antibiotics: Broad spectrum antibiotic plus metronidazole. The latter takes care of the anaerobes. Clostridium perfringens infection of the necrotic gallbladder is more common in diabetes mellitus.
- Administer Intravenous analgesic but avoid opioids which may cause spasm of the bile duct. Non-steroidal anti-inflammatory agents are quite effective as they are thought to reduce the degree of inflammation in addition to the analgesic effect.

When the clinical condition of the patient is stable, request is made for plain abdominal x-rays (erect and supine). This will, however, demonstrate calcified gallstones in only 10% of cases.

Transabdominal ultrasound scan is the hallmark of diagnosis. In addition to demonstrating gallstones, it also displays the entire hepatobiliary system. Doppler sonography gives a more accurate diagnosis. The key to the diagnosis of emphysematous cholecystitis is the presence of air on abdominal X-ray or ultrasound examination.

Biliary scintigraphy: This is the study of gallbladder function and biliary patency with the use of an intravenous radiotracer. Non-visualisation of the gallbladder in a symptomatic patient is an indication of gallbladder dysfunction caused by either acute or chronic cholecystitis.

Patients that manifest clinical features of obstructive jaundice should be investigated along that line after managing the acute situation.

Blood investigations include full blood count (leucocytosis) and liver function tests which may be deranged due to associated inflammation of the liver.

Most patients improve with conservative treatment. The prognosis is, however, worse with emphysematous cholecystitis. The latter is associated with a five-fold increased risk of gallbladder perforation as well as a ten-fold increase in mortality in patients younger than 60

years. By and large, most patients will recover from acute cholecystitis and will subsequently require cholecystectomy.

Some patients may, however, fail to respond to conservative treatment. Since emergency cholecystectomy may not be advised for fear of damage to vital structures due to the fulminating inflammation, cholecystostomy should be employed as a stop-gap measure. Essentially, this is a form of incision and drainage of the gallbladder. After evacuating the stones and infected fluid from the gallbladder, a tube drain is inserted for continuous drainage. The procedure can be carried out either by the open method or percutaneously under ultrasound guidance. An elective cholecystectomy is planned for 6 – 12 weeks later.

There is some controversy associated with the timing of definitive surgery after the initial management of acute cholecystitis. The proponents of elective surgery argue that emergency surgery is not advised because of the high risk of damage to the associated vital structures as a result of oedema and fibrosis ensuing from the acute inflammation. On the other hand, the proponents of emergency cholecystectomy believe that surgery is even easier if carried out within 24 to 48 hours of onset of acute cholecystitis. The initial oedema and fibrinous adhesions, they argue, even facilitate the procedure. Studies have compared early and late laparoscopic cholecystectomy. Some studies have shown that the delayed-surgery group had a greater need for conversion to open cholecystectomy as well as a longer average total hospital stay and convalescence. For obvious reasons, high risk patients with acalculus or emphysematous cholecystitis and those with associated systemic toxicity should have early cholecystectomy. The current thinking is that early laparoscopic cholecystectomy is a safe procedure and should be encouraged because of its socioeconomic benefits. Laparoscopic cholecystectomy is now carried out prior to discharge of the patient managed for acute cholecystitis

Owing to the critically-ill nature of patients with acalculous acute cholecystitis, initial surgical treatment should be by percutaneous cholecystostomy. Cholecystectomy should only be contemplated in the absence of rapid clinical improvement.

CHRONIC CHOLECYSTITIS

The clinical features of chronic cholecystitis are similar to those of the acute variety except that they are milder and recurrent. Chronic cholecystitis results from repeated bouts of acute cholecystitis and/or biliary colic leading to gallbladder wall inflammation and fibrosis. The gallbladder wall is thickened and shrunken with atrophy of the muscle coat. Projection of the mucous membrane into the lumen results in the formation of clefts. This is referred to as Rokitansky-Ashoff's sinuses. There is remission of symptoms with the use of antibiotics. Abdominal examination may elicit right hypochondrial tenderness

Diagnosis is confirmed by transabdominal ultrasound scan.

The ultimate treatment is elective cholecystectomy. Traditionally, this was carried out by the open method. Currently, the gold standard is laparoscopic cholecystectomy. The advantages of the latter over the open method include minimal blood loss, better cosmetic outcome and rapid recovery and discharge.

Complications of cholecystectomy include complications of anaesthesia, haemorrhage and infection. Others are damage to the common bile duct resulting in stricture/obstructive jaundice and biliary peritonitis. The latter may follow slipping of the ligature over the stump of the cystic duct.

Post-cholecystectomy syndrome:

This is the relapse of preoperative morbid symptoms after cholecystectomy. It may be due to complication of the index surgery or a missed diagnosis of an unrelated pathology. The common causes of post-cholecystectomy syndrome are

- Missed stone either in the cystic duct stump or in the common bile duct
- Iatrogenic damage to any part of the biliary tree
- Gastro-oesophageal reflux disease
- Peptic ulceration
- Hiatus hernia

Management of postcholecystectomy syndrome lies in carrying out relevant investigations in order to determine the fundamental cause

- Clinical evaluation: History and physical examination
- Liver function test
- Upper gastrointestinal endoscopy
- Barium swallow/meal
- Abdominal ultrasonography

The definitive treatment consists of addressing the underlying cause

CARCINOMA OF THE GALLBLADDER

This is a rare neoplasm. It is commoner in women than men (4:1). Cholelithiasis. The risk factors are

- Long-standing gallstones: This is the most common risk factor
- A porcelain (calcified) gallbladder may undergo metaplastic change.
- Gallbladder polyps,
- Cholecystoenteric fistula
- Chronic typhoid carriage. The fundus is the most common site (60%). They are mainly adenocarcinomas (90%)
- Choledochal cysts
- Adenomyomatosis of the gallbladder
- Carcinogens such as nitrosamines
- Anomalous pancreatico-biliary duct junction: May cause reflux of pancreatic juice into the biliary tree

The histological types of gallbladder carcinoma are adenocarcinoma (80 to 95%), squamous cell carcinoma and the mixed adenosquamous carcinoma. Rare types are carcinoid and undifferentiated carcinoma. Spread of gallbladder carcinoma is by direct, lymphatic and haematogenous routes (liver, lungs and bones). Tumour markers are nonspecific and include CEA and CA 19-9

Clinical symptoms include right hypochondrial pain, jaundice, and loss of weight. While some patients may present with features of acute cholecystitis, others may be asymptomatic and only diagnosed on histology of routine cholecystectomy specimen for chronic cholecystitis. A recent change in the symptomatology of a patient with longstanding gallstones should raise suspicion of metaplasia resulting in carcinoma of the gallbladder.

Examination may reveal an ill-looking patient with evidence of jaundice and weight loss. Abdominal examination may reveal a nodular, right hypochondrial mass with associated hepatomegaly and ascites.

Diagnosis is confirmed by abdominal ultrasound scan, CT scan, ultrasound-guided FNAC, and liver function test. Contrast enhanced CT scan is valuable for further evaluation (staging and operability). There is a place for diagnostic laparoscopy.

Treatment is surgical. The extent of surgery depends on the stage of the disease. It ranges from simple cholecystectomy to radical cholecystectomy. The latter involves cholecystectomy with a 2 cm wedge of liver tissue at the gallbladder bed to ensure a tumour free margin. Some authorities advocate resection of segments V and IVb. This is accompanied by a wide clearance of associated lymph nodes (pericholedochal, periportal, hepatoduodenal and up to the peripancreatic lymph nodes). To forestall trochar site tumour implantation, open cholecystectomy is advised if diagnosis is confirmed before surgery. The prognosis is poor. Obstructive jaundice and acute pancreatitis are discussed in the relevant sections.

CHAPTER SEVEN

OBSTRUCTIVE (SURGICAL) JAUNDICE

This is post-hepatic jaundice that is amenable to surgery. It results from obstruction of the post-hepatic biliary system.

AETIOLOGY

Like all luminal structures, the causes of obstructive jaundice can be categorised into intraluminal (within the lumen), intramural (involving the wall) and extramural (outside the wall).

- Intraluminal: Gallstones, worm infestation (ascariasis)
- Intramural: Congenital biliary atresia, biliary strictures, sclerosing cholangitis, choledochal cyst, and cholangiocarcinoma. Klatskin's tumour occurs at the confluence of the hepatic ducts just above the point of entry of the cystic duct.
- Extramural: Common causes are carcinoma of the head or periampullary region of the pancreas and cholangiocarcinoma. Others include external compression of the common bile duct by tumours and lymphadenopathy especially at the porta hepatis

CLINICAL FEATURES

- Jaundice: This is due to hyperbilirubinemia resulting from obstruction of the natural pathway. Acute cholangitis may result from obstruction of the common bile duct and presents with the classical Charcot's triad: upper abdominal pain, jaundice and rigors. The complete triad is, however, said to be present in only 25% of cases. Severe cases may present with clinical features of septic shock and multiple organ dysfunction. In addition to the features of Charcot's triad, Reynold's pentad incorporates mental status changes and evidence of shock (hypotension). It is the hallmark of severe cholangitis with septicaemia
- Itching: Due to excess bile salts in circulation
- Passage of pale stools (steatorrhoea): This is due to the absence of stercobilinogen which is a by-product of bilirubin in the stool
- Passage of dark coloured urine: This results from excretion of bilirubin in the urine
- Easy bruising and bleeding from mild trauma: Bleeding diathesis due to nonabsorption of vitamin K which is essential for the synthesis of clotting factors II, V, VII, IX and X

Other features such as fever and weight loss will depend on the cause of the jaundice

The clinician should endeavour to elucidate the aetiology of jaundice from the history. The commonest causes of obstructive jaundice in our environment are gallstones and pancreatic cancer. Whereas jaundice due to gallstones is painful, intermittent and occurs predominantly in the younger age group, that due to peri-pancreatic neoplasms is initially painless, progressive and occurs in the older age group. Previous history of biliary surgery, such as cholecystectomy, may suggest biliary stricture.

It is important to rule out other causes of jaundice such as previous blood transfusion, history of recent exposure to a jaundiced patient, and multiple intramuscular injections as in drug addiction.

Examination will reveal an ill-looking patient who may be pale and febrile. Loss of weight may signify a neoplastic aetiology. Obstructive nature of the jaundice may be suggested by the greenish-yellow hue of the conjunctiva.

Abdominal examination may reveal a distended abdomen due to ascites. Courvoisier's rule states that "if in a jaundiced patient the gallbladder is palpable, then the jaundice is not likely due to stones". The rationale behind this is that a gallbladder that harbours stone(s) is unlikely to distend due to fibrosis that occurs from the resultant chronic cholecystitis. Exceptions to this rule include a situation where an impacted stone in the cystic duct coexists with one in the common bile duct. Also in Mirrizon's syndrome, a stone in the cystic duct may indirectly impact on the common bile duct resulting in both obstructive jaundice as well as distension of the gallbladder. Another exception to this rule is when obstructive jaundice is due to a primary biliary duct stone.

INVESTIGATION OF OBSTRUCTIVE JAUNDICE

- Liver function test: There is increased serum bilirubin level that tilts in favour of conjugated bilirubin; elevation of all the liver enzymes and with exceptionally high levels of alkaline phosphatase.
- Clotting profile: Prothrombin time will be deranged. This is basically due to nonabsorption of vitamin K which is a fat-soluble vitamin and therefore requires the presence of bile for its absorption. Since this vitamin is required for the production of clotting factors 11, V11, 1X and X, which are essential for blood clotting, this patient will bleed unless steps are taken to rectify the anomaly. The international normalised ratio (INR) should be monitored.
- Transabdominal ultrasound scan: This outlines the hepatobiliary system and may define the point of obstruction with the attendant proximal dilatation of the common bile duct. The pancreas and its lymph nodes are equally visualised



ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATOGRAPHY SHOWING STONE IN THE COMMON BILE DUCT

- Endoscopic Retrograde Cholangio Pancreatography (ERCP): Identifies the site of blockage. Can equally be therapeutic as it may be employed in the extraction of the offending stone in the distal common bile duct
- Magnetic Resonance Cholangio Pancreatography (MRCP): Unlike ERCP, it is non-invasive



T-TUBE CHOLANGIOGRAM SHOWING A FILLING DEFECT

- Computer Tomography (CT) scan: This is found to be more useful in neoplastic disease.
- Percutaneous Transhepatic Cholangiography (TPC): This involves the cannulation of an intrahepatic duct followed by injection of a contrast. It is particularly useful in defining the hepatobiliary system in a heavily jaundiced patient. It can equally be employed therapeutically in decompressing the biliary system preoperatively
- Urinalysis: Will show excess bilirubin and absence of urobilinogen. The latter situation is due to non-production of stercobilinogen in the gut. Glucosuria may indicate the presence of diabetes mellitus which may be associated with a pancreatic neoplasm.
- Fasting blood sugar estimation will be necessary in the presence of glucosuria
- Full blood count
- Serum electrolytes, urea and creatinine
- Group and crossmatch blood

Depending on the age and clinical state of the patient, chest Xrays and electrocardiography may be carried out.

PREOPERATIVE PREPARATION OF THE JAUNDICED PATIENT

Patients who present with clinical features of acute cholangitis require preliminary treatment with intravenous infusion, analgesics and antibiotics that will deal with both aerobic as well as anaerobic organisms. This is followed by decompression by non-surgical biliary drainage. The latter can be achieved by either endoscopic (ERCP) or percutaneous (PTC) drainage.

The aim of preoperative preparation of a jaundiced patient for surgery is to address any identified preoperative problem(s) and forestall any perceived perioperative complications. It consists of the following

- Administration of parenteral Vitamin K to correct any existing clotting defect. Administer 10mg parenterally for 5-7 days. It is imperative that the clotting profile (prothrombin time, partial thromboplastin time and international normalised ratio) is crosschecked to ensure that it is near normal before surgery in order to prevent both intraoperative as well as postoperative haemorrhage.
- Administration of prophylactic antibiotics: Jaundiced patients are prone to infection. A combination of a broad spectrum antibiotic and metronidazole should be administered at induction of anaesthesia. It has been shown that gram-negative septicaemia may be partially responsible for the postoperative acute renal failure seen in jaundiced patients
- Adequate hydration of the patient is important in order to prevent acute renal failure. It is believed that the high bilirubin load may result in the blockage of the renal tubules. Liberal fluid intake should be encouraged on admission and intravenous fluid administration commenced just before surgery. After ensuring that the patient has been reasonably hydrated, intravenous mannitol is given at the induction of anaesthesia to stimulate osmotic diuresis. A urethral catheter should be put in place to monitor the urinary output during the perioperative period.
- Replenishment of hepatic glycogen store by preoperative administration of oral glucose drinks
- Correction of preoperative anaemia by blood transfusion
- Preoperative malnutrition should be corrected by either the enteral or parenteral route
- Evaluation of pulmonary function and correction of any anomaly by chest physiotherapy

TREATMENT OF OBSTRUCTIVE JAUNDICE

This depends on the cause:

- Stone in the biliary duct: Exploration of the common bile duct either by the open method or laparoscopically. Cholecystectomy is an integral part of the procedure. The stone can also be extracted endoscopically via ERCP. T-tube drainage may be necessary after exploration of the common bile duct.

- Biliary stricture: Roux-en Y hepatico-jejunostomy is indicated in a case of established stricture. If the injury is, however, detected at surgery, choledocho-jejunostomy should be carried out. Stenting may also be feasible when the strictured segment is short. Malignant strictures as in Klatskin's tumour may be amenable to radical resection.
- Periampullary and pancreatic head cancer: Whipple's operation in the early stage and palliative triple bypass in the late disease. Whipple's operation is an extensive procedure that involves resection of the entire duodenum, up to 19cm of proximal jejunum, head and neck of pancreas and distal half of stomach. Other structures involved in resection are gallbladder, lower end of common bile duct and all the anatomically-related lymph nodes. Continuity is restored by gastrojejunostomy, pancreaticojejunostomy and hepaticojejunostomy. Traditionally, triple bypass consists of cholecysto-jejunostomy, jejuno-jejunostomy and gastro-jejunostomy. Recently, however, choledocho-jejunostomy is gradually replacing cholecysto-jejunostomy. Currently, some surgeons deliberately omit the jejuno-jejunostomy component of triple bypass with no adverse consequences.
- Others include Kasai's operation or liver transplantation for congenital biliary atresia.

CHOLANGIOPANCREATIC DISEASE

This is a rare tumour of the biliary ducts. It accounts for 1-2% of malignant tumours. About 60% of patients are above the age of 65. There is association with the following conditions: sclerosing cholangitis (20 fold increase risk), hepatolithiasis, choledochal cyst, ulcerative colitis and clonichis infection. It is thought that parasites produce carcinogens and free radicals which induce DNA mutations in the ductal epithelial cells.

Sites: Intrahepatic (10%), perihilar (65%), and distal (25%). Klatskin tumours occur at the hilar region at the confluence of the hepatic ducts and the common hepatic duct.

Clinical features of cholangiocarcinoma include abdominal pain, early satiety, weight loss and progressive clinical features of obstructive jaundice. Examination will reveal a palpable liver which may be either soft and smooth (hydrohepatitis) or hard and nodular (metastatic). The gall bladder will be palpable in lesions involving the distal bile duct below the level of the cystic duct (Courvoisier's rule)

Investigations include

- Tests to confirm the presence of obstructive jaundice: bilirubin, alkaline phosphatase, gamma-glutyl transferase.
- Abdominal USS and CT to determine the level of obstruction and the extent of disease.
- Percutaneous Transhepatic Cholangiography (PTC): Both diagnostic and therapeutic as it could be used to decompress the biliary system. PTC is more useful in proximal lesions
- Endoscopic Retrograde Cholangio-Pancreatography (ERCP): Useful in distal tumours. Endobiliary stent may be inserted to maintain the luminal channel. Specimens for cytology and biopsy may equally be taken.

Treatment: Surgical intervention is minimally effective as only 10-15% of lesions are operable. The procedure of choice is dictated by the site of the tumour. Proximal lesions may require partial

hepatectomy and hepaticojejunostomy while distal lesions may require pancreaticoduodenectomy.

Adjuvant therapy (chemotherapy and radiotherapy) have not been found useful. Doxorubicin, cisplatin and I¹³¹ anti CEA antibodies have, however, been tried.

Prognosis is poor as 90% die within the first year of diagnosis. Prognosis is better with distal tumours.

CHAPTER EIGHT

HEPATIC NEOPLASMS

There are both benign as well as malignant varieties.

BENIGN TUMOURS:

Of three histological types – haemangioma, hepatic adenoma and focal nodular hyperplasia

Hepatic haemangioma:

Hepatic haemangioma is a benign vascular tumour and the most common primary tumour of the liver. It consists of an abnormal plexus of veins and is commonly solitary but may be multicentric. The lesion presents with right upper quadrant pain and a right hypochondrial mass over which may be heard a bruit on auscultation. The Kasabach - Merrith syndrome comprises of haemangioma, thrombocytopenia and fibrinogenemia. Spontaneous rupture is fortunately rare with hepatic haemangioma.

Investigations include USS, CT with intravenous contrast and MRI. Angiography is the gold standard investigation. Though invasive, it has a high sensitivity and specificity. Needle biopsy is contraindicated as it may result in life-threatening haemorrhage.

Treatment: As this tumour has a minimal malignant potential, no further treatment is required in incidental lesions. Indications for surgery are inability to rule out malignancy, severe symptoms and rupture. Initial radiotherapy and hemihepatectomy may, however, be employed in giant and symptomatic lesions.

Hepatic adenoma

It is common in females and is associated with sex hormones (including contraceptive pills). Hepatic adenoma has a propensity to produce symptoms, undergo malignant transformation and cause haemorrhage. Malignant transformation of adenoma to hepatocellular carcinoma is reported to occur in 5% to 11% of cases. Since regression may follow withdrawal of hormonal therapy, patients with small symptomless lesions may initially be treated with cessation of oral contraceptives and close surveillance. Molecular biology has demonstrated three forms of hepatic adenoma with their respective clinical characteristics: inflammatory (increased risk for bleeding but with low malignant potential), those containing hepatocyte nuclear factor 1 (HNF-1) alpha mutations with no risk of bleeding or of malignant transformation, and those with beta-catenin activation (often seen in patients with glycogen storage disease, it has an increased risk of malignant transformation).

Investigations include USS, CT and angiography. Laparoscopic ultrasound and biopsy is of a high diagnostic value. Owing to its malignant potential, it is pertinent to monitor the alpha fetoprotein level.

Treatment is by surgical resection.

Focal nodular hyperplasia (FNH):

It is the second most common benign tumour of the liver and most commonly occurs in females in the reproductive age group. Its aetiology is unknown and it has no malignant potential. As the name suggests, there is a focal overgrowth of functioning liver tissue supported by fibrous tissue. The association with birth control pills is not as strong as with adenoma.

Diagnosis: Most FNHs are asymptomatic and are incidental findings. Diagnosis is by way of USS and CT. Angiography is highly sensitive and specific. The main radiographic feature is that of a liver mass with a central scar. Histology shows normal hepatocytes and bile ducts. Hepatic adenoma, on the other hand, does not demonstrate the presence of bile ducts.

Treatment: Since it has no malignant potential, the asymptomatic cases may be followed up with discontinuation of contraceptive pills if indicated. Symptomatic cases will require surgical resection. Embolisation has been found useful as the tumour is usually fed by one major vessel.

PRIMARY MALIGNANT LIVER TUMOURS

There are two main types of primary malignant liver tumours in the adult: Hepatocellular carcinoma (90%), and cholangiocarcinoma (5%). Mixed carcinoma accounts for 2% to 6%. Other rare types include Kupffer cell carcinoma and haemangioendothelioma. Hepatoblastoma is a childhood tumour that occurs within two years of life. It is characterised by the high sensitivity to chemotherapy (vincristine, doxorubicin and 5FU)

HEPATOCELLULAR CARCINOMA (HCC)

It used to be regarded as a cancer of poverty because of its prevalence in the developing world

Aetiology

- Arising from infections: Hepatitis B, C, and D viruses
- Aflatoxin: Arises from a mould by fungus *Aspergillus*. Mechanism of action is not clear but may act by immunosuppression. Malnutrition may act as a cofactor
- Lifestyle: Alcoholic cirrhosis and smoking
- Premalignant condition: As in hepatic adenoma
- Haematological: Haemochromatosis
- Chemicals: DDT, nitrates and nitrites related to food processing, herbicides and solvents.
- Hormones: Experimental studies have implicated sex hormones like oestrogens and testosterone

Pathology: Has a male preponderance (M: F=4:1) and more commonly affects the right lobe.

Of three macroscopic types: Hanging, pushing and infiltrative.

Histological types: Well, moderately and poorly differentiated respectively.

Spread is by direct route (diaphragm and neighbouring structures); via lymphatics (liver, portohepatic and abdominal nodes, cysterna chyli) and haematogenous (lungs, bones and adrenals).

Clinical features: Painful hepatomegaly is the early presentation while weight loss (cachexia), weakness and jaundice appear later. Features of chronic liver disease (ascites, encephalopathy), and signs of portal hypertension are seen in advanced cases. Child's classification of hepatocellular function may be employed in categorising the severity of the condition.

Child's classification of hepatocellular dysfunction

Group designation	A	B	C
Bilirubin	< 2.0	2-3.0	> 3.0
Albumin (g/dl)	>3.5	3.0-3.5	< 3.0
Ascites	None	Easily controlled	Poorly controlled
Neurological disorder	Nil	Minimal	Advanced
Nutrition	Excellent	Good	Wasting

Investigations:

- Blood investigations include FBC, LFT, Hepatitis B, Hepatitis C, and alpha-foetoprotein. The latter is a tumour marker in which a serum level above 100 IU is considered significant. Much higher levels (1000 IU) are not uncommon.
- Radiological investigations include USS, CT scan; contrast enhanced CT, and CT angiography. Others are coeliac angiography, chest X-ray, chest CT, and bone scan.
- Other investigations are liver biopsy, ascitic tap and laparoscopy.

Treatment: Surgery is aimed at excising the lesion with 1-2 cm of unaffected normal liver tissue. Resectability is related to the morphology of the tumour. Whereas both the hanging and pushing types may be resectable, the infiltrative type is more difficult to resect. Hemihepatectomy (limited to one lobe) may be feasible in non-cirrhotic patients who are in Child's grades A and B. Cirrhotic patients require total hepatectomy and liver transplantation. In order to benefit from liver transplantation, it is preferable that the HCC should be less than 5cm, not more than 3 in number, and without portal vein or extrahepatic involvement.

Modes of palliative treatment of hepatocellular carcinoma are

- Radiofrequency ablation
- Percutaneous ethanol or acetic acid injection
- Intra-arterial injection of cytotoxic chemotherapy (cisplatin, Adriamycin, mitomycin) through the gastroduodenal artery
- Intra-arterial embolization (gelfoam, gelatin sponge),
- Ligation of hepatic artery
- Microwave or cryoablation.

Adjuvant therapy involves the use of cytotoxic drugs (cisplatin, carboplatin, mitomycin, 5FU). The somatostatin analogue, octreotide, has been found to reduce tumour size. Neoadjuvant chemotherapy may be useful. Others include intralesional alcohol injection, cytokines and heat therapy

Follow up is by serial alpha-foetoprotein estimation and imaging

Prognostic features include wasting, jaundice, ascites and presence of venous collaterals

Secondary liver tumours:

The liver is a common site of metastasis of many malignant lesions. Common sites of primary lesions include the colorectum, pancreas and ovaries. Others are testes, prostate, breast and lung. Colorectal carcinoma is the commonest lesion that spreads to the liver.

It is imperative to endeavour to elucidate the primary site of a secondary in the liver. This entails obtaining a comprehensive history coupled with detailed clinical examination bearing in mind

the common sites of primary tumours that have the potential for hepatic spread. This is supported by relevant laboratory and radiological investigations. Application of immunohistochemical techniques have been found very useful in the elucidation of the primary site. Tumour markers have been found useful in the identification of the origin of secondary liver tumours. Some are known to be specific for specified organs. For example while prostate-specific antigen (PSA) is specific for prostate cancer, carbohydrate antigen 125 (CA 125) is specific for ovarian cancer. Identification of the primary site is useful not just for its treatment, but also for decision-making regarding the treatment modality of hepatic secondaries. Furthermore, though it is indicative of late stage disease, some liver secondaries are still amenable to surgical ablation. For example, a few hepatic secondaries in a patient with advanced colorectal cancer do not preclude resection of the primary tumour as well as possible resection of the hepatic secondaries. Resection of the primary tumour is regarded as the best form of palliation and improvement in quality of life. Liver transplantation has no role in the treatment of patients with secondary tumours.

CHAPTER NINE

HEPATIC ABSCESES

The term, hepatic abscess, refers to a collection of pus in the hepatic parenchyma. It consists of two varieties: pyogenic and amoebic. Whereas the pyogenic form usually presents as multiple abscesses and is of bacterial origin, the amoebic variety is usually solitary and is a known complication of infection with *Entamoeba histolytica*.

PYOGENIC ABSCESES

The following are the routes of infection:

- Direct spread from contiguous biliary tract infection such as cholangitis
- Portal spread (portal pyaemia) from gastrointestinal infections such as appendicitis and diverticulitis
- Systemic spread: Bacteremia from any cause.
- Hepatic trauma: Blunt injury, stab wound
- Cryptogenic from an unknown source

Offending organisms include *E. coli*, *proteus*, *staph pyogenes*, and *haemolytic streptococcus*:

Clinical features of pyogenic abscess

In addition to the clinical features of the primary condition, there is associated fever, rigors, and right upper quadrant pain. Other symptoms are anorexia, nausea and vomiting.

Examination will reveal an ill-looking patient who may be jaundiced. Abdominal examination will demonstrate a tender hepatomegaly.

Relevant investigations include FBC, blood culture, abdominal USS, and abdominal CT in difficult cases.

Treatment of pyogenic abscesses: Initial treatment is by the administration of intravenous antibiotics (including metronidazole for anaerobic organisms). The ideal drainage procedure is by USS or CT guided percutaneous drainage. Indications for open drainage include failure of percutaneous drainage, and presence of multiple and/or loculated abscesses.

AMOEBOIC LIVER ABSCESS

The causative agent is the protozoa, *Entamoeba histolytica*. It is spread by the faeco-oral route and the primary organ of infection is the intestine. Infection spreads to the liver via the portal vein from intestinal amoebiasis. It is common in the developing world with a male predilection (M: F= 9:1). The right lobe of the liver is usually more affected than the left.

Pathogenesis: As stated above, spread is from focal ulcers in the colonic mucosa through the portal vein to the liver. Histiolysin is released by the trophozoites and is responsible for the ensuing amoebic hepatitis. The latter consists of liquefactive necrosis, thrombosis of vessels, and breakdown of red cells. The end-product is the so called "anchovy sauce" pus. The hepatic lesion demonstrates 3 zones: a central zone of necrosis, a middle zone showing various degrees of liver

parenchymal damage and an outer zone of apparently normal cells. The outer zone may harbour some amoeba.

Clinical features: Patient will present with right upper quadrant pain, fever, diarrhoea, and yellowness of the eyes. There may be a preceding history of passage of bloody, mucoid stools. Examination will reveal an ill-looking patient with clinical evidence of jaundice. Abdominal examination will confirm a tender hepatomegaly. Digital rectal examination may show evidence of loose, mucoid and bloody stool.

Complications: There may be rupture of the abscess to adjoining structures with grave implications. It may rupture into the lungs, bronchial tree, and pericardial cavity (especially left hepatic lobal lesions) resulting in cardiac tamponade. Rupture into the peritoneal space will result in peritonitis. There may be associated secondary bacterial infection.

Differential diagnoses include hydatid cyst, hepatitis and primary liver carcinoma. Others are subphrenic abscess, lung abscess and empyema thoracis.

Investigations include FBC, ESR, stool examination, proctoscopy/sigmoidoscopy, and indirect haemagglutinin test. Others are chest and abdominal X-rays, abdominal USS and CT, diagnostic aspiration and therapeutic trial in difficult cases.

Treatment of amoebic liver abscess: There is a positive response to metronidazole. This may either be given as a large bolus dose of 2.4 gm or a much lower dose over a period (800mg tds for 5 days or an even lower dose of 400mg tds for 10 days). Refractory cases are treated with chloroquine and those with secondary infection will have tetracycline in addition to the primary drugs. Emetine hydrochloride which used to be the first line drug treatment is now only reserved for proven resistant cases.

Initial surgical treatment consists of aspiration at the 9th or 10th intercostal space or point of maximal tenderness. Aspiration is best guided by USS. There may be recourse to open surgical drainage in cases of failure of aspiration and in difficult cases such as multiple abscesses and presence of very thick pus.

Indications for surgery

- Repeated aspiration
- Thick and/or multiloculated abscess
- Multiple abscess
- Left lobar abscess
- Ruptured abscess

Complications of surgery include anaesthetic complications, bleeding and liver failure. Others are fistulation, intraperitoneal abscesses and biliary peritonitis from biliary leakage.

CHAPTER NINE

THE SPLEEN

Anatomy:

The spleen is located in the left hypochondrium and tucked into the lower thoracic cage at the level of the 9th, 10th and 11th ribs. It is related to the left kidney posteriorly and anteriorly to the left hepatic flexure and the descending colon. The pancreatic tail abuts at the hilum of the spleen. Blood supply: It is rich in blood supply. The main supply is from the splenic artery which is a branch of the celiac trunk. The vasa brevia, which fans out at the splenic hilum, arises from the left gastro-epiploic artery. The venous drainage follows the arterial supply and ultimately drains into the portal vein.

Functions of the spleen:

The spleen is a part of the reticuloendothelial system and is ascribed with the following functions

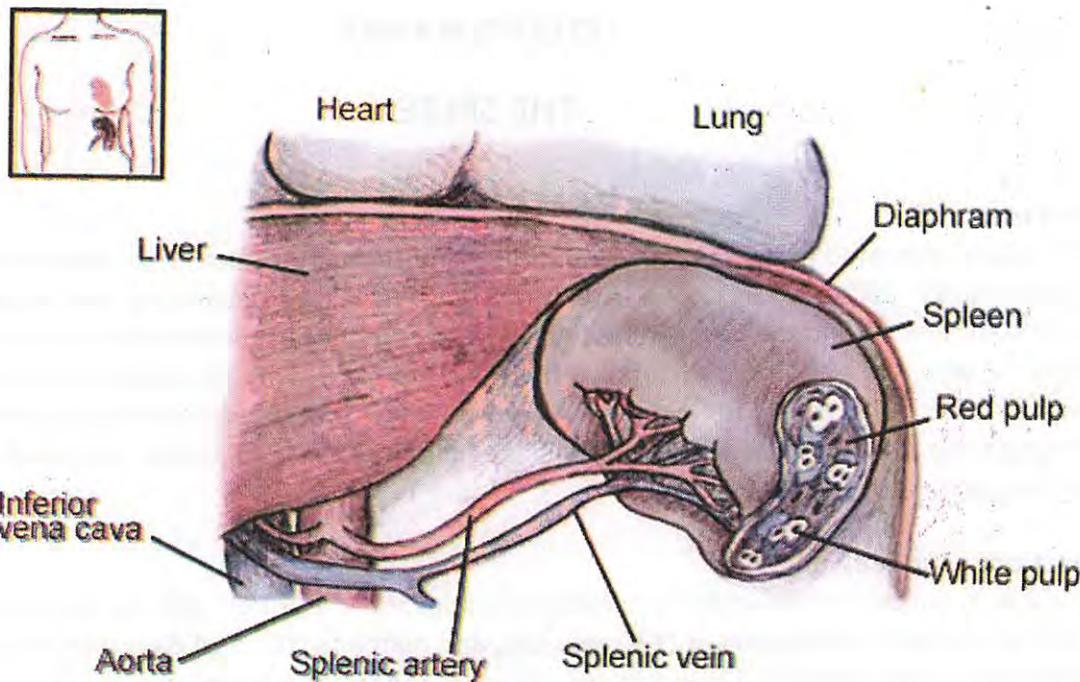
- * Destroys effete red blood cells (120 days old) and platelets (10 to 14 days old). It also removes erythrocytes with abnormal membranes and also abnormal intracellular erythrocyte particles (Howell-Jolly bodies, Heinz bodies and Pappenheimer bodies)
- * Produces opsonins (tuftsin and properdin) that are important in immunity
- * Produces antibodies especially IgM
- * It is a site for phagocytosis
- * It is an extramedullary organ of haemopoiesis
- * It is a site for the storage of blood elements like platelets and lymphocytes

Splenectomy:

The first splenectomy was performed in 1916 by an Australian surgeon called Herman Schloffer.

The following are indications for splenectomy

- Severe splenic rupture (grades 4 and 5). Splenic conservative surgery is employed in the management of milder degrees of splenic trauma (grades 1 to 3)
- Hypersplenism causing pancytopenia: Leukemias and lymphocytic infiltration
- Chronic haemolytic diseases: These include hereditary spherocytosis, autoimmune haemolytic anaemia, hereditary elliptocytosis and medullary fibrosis. Others are myeloid metaplasia, sickle cell anaemia and thalassemia



THE SPLEEN. INSET: SURFACE ANATOMY OF THE SPLEEN

- Thrombocytopenia: Idiopathic thrombocytopenic purpura and thrombotic thrombocytopenic purpura. In these patients, surgery is delayed until the patient is six years old in order to reduce the risk of overwhelming post-splenectomy infection. Patients with idiopathic thrombocytopenic purpura who respond to corticosteroid therapy are most likely to obtain long-term remission from splenectomy.
- Miscellaneous: Splenic abscess, Gaucher's disease, splenic cysts, Felty's syndrome (rheumatoid arthritis, splenomegaly and neutropenia) and sarcoidosis
- Cancers: leukemia and lymphoma. In the management of Hodgkin's disease refractory to medical therapy, splenectomy reduces pain, abdominal fullness and associated hypersplenism
- As part of other surgeries: Resection of the body and tail of the pancreas in insulinoma, splenorenal shunt in portal hypertension and as an integral part of total gastrectomy in the management of gastric cancer.
- Was previously used as a staging procedure in the management of lymphoma. CT scan has replaced splenectomy in this regard.

SPLENIC RUPTURE

Aetiology

- Trauma: RTA, fall from heights, direct blow at the right hypochondrium (marital disharmony has been reported as a not uncommon cause), and penetrating injury
- Spontaneous rupture may occur in an already diseased spleen. Malaria infection is a known predisposing factor.

Presentation: This depends on the nature and severity of the injury

- Patient neither rallies nor able to get to the hospital. Occurs in very severe injuries
- Initial recovery after injury, followed by clinical features of hypovolemic shock
- Delayed rupture may occur in cases of subcapsular haematoma
- Spontaneous rupture may occur in an already diseased spleen.

Clinical features: Usually, there is a preceding history of trauma. Clinical examination reveals an ill-looking patient who is pale and manifests the clinical features of shock. Abdominal examination may reveal

- Fullness with left hypochondrial tenderness and evidence of haemoperitoneum
- Kehr's sign: Left shoulder pain on elevation of the foot end of the bed
- Ballance's sign: Non-shifting dullness in left upper quadrant of the abdomen due to clotted blood around the spleen and shifting dullness in the right flank due to unclotted blood in the peritoneal cavity. Seen in 25% of cases of splenic rupture.
- Saegesser's sign: Phrenic nerve compression causing tenderness around the lower posterior triangle of the neck between the left sternomastoid and scaleneus medius muscle above the clavicle.
- Cushen's sign: Redness around the umbilical region. It is a reflection of underlying haemoperitoneum.
- Cullen's sign: Redness over the left flank: Equally indicates haemoperitoneum

A thorough clinical examination is mandatory to rule out associated injuries to the chest and other abdominal viscera.

Treatment of splenic injuries

- Institute the ABC of resuscitation
- Initial resuscitation: Intravenous infusion with normal saline or Ringer's lactate
- Take blood specimen for investigations: Full blood count, electrolytes/urea/creatinine, grouping and cross-matching of blood
- Urethral catheterisation: Monitor the urinary output
- Naso-gastric aspiration may be necessary particularly if there is abdominal distension.
- Radiological investigations:
 - Plain abdominal X-rays (erect and supine): Findings may include elevation of the left hemidiaphragm, obliteration of the left psoas shadow and downward displacement of the left colonic gas shadow. Others are indentation of the fundic gas shadow, associated left lower rib fractures and left basal pleural effusion
 - Chest X-ray: To identify associated chest injuries
 - Abdominal USS: Findings may include a subcapsular haematoma, laceration of the splenic capsule and haemoperitoneum
 - Abdominal CT: Indicated if abdominal USS findings are inconclusive. It is more efficient in the detection of relatively small splenic tears
- Other investigations: Diagnostic peritoneal lavage, and four-quadrant tap. These are usually positive in splenic trauma with intraperitoneal haemorrhage. Despite the

advent of abdominal USS, these investigations are still relevant particularly in situations where USS facilities are not readily available.

Definitive treatment:

Conservative management (non-operative management): This has been described particularly in children. The advantages include

- Maintenance of the immune function of the spleen: It also eliminates the risk associated with overwhelming postsplenectomy infection (OPSI)
- Associated with a lower morbidity when compared to operative treatment

Non-operative treatment is, however, associated with the following disadvantages:

- Requires multiple blood transfusion with its attendant risk
- Haemodynamic instability resulting from continued bleeding is a sign of failure of conservative management and will ultimately require operative treatment

Contraindications to non-operative treatment are

- Haemodynamic instability requiring multiple blood transfusions to maintain cardiovascular stability. Beyond a transfusion threshold is an indication for operative treatment
- Patients with moderate to severe brain injury: Any spell of hypoperfusion of the brain will worsen the clinical state of the patient
- CT scan -confirmed high grade splenic injury

An analysis by the National Trauma Data Bank (NTDB) found high failure rates and prolonged hospital stays when high-grade splenic injuries were managed conservatively (non-operative management). The importance of very close monitoring of patients who are managed nonoperatively cannot be overemphasised.

Indications for surgical treatment in splenic rupture include

- Haemodynamic instability or shock on arrival
- Presence of associated injuries that require open intervention
- Failure of non-operative treatment

Currently there are two methods of surgical intervention: laparoscopy and open surgery.

Laparoscopic approach: Advantages include

- Less postoperative pain
- Shorter hospital stay
- Early return to a regular solid food diet
- Quicker return to normal activities
- Rarely complicated by incisional hernias

Contraindications to laparoscopic splenectomy:

- Large spleen,
- Scarred spleen
- Obesity.

Open splenectomy approach is preferred when splenectomy is for trauma or for haematological consideration.

Surgery may involve either 'total' or partial splenectomy. The latter entails salvaging and retention of some splenic tissue for the sustenance of the immune functions of the spleen. Studies have shown that at least 25% of splenic mass is critical for immunological function. This is important in the prevention of Overwhelming Postsplenectomy Infection (OPSI). Splenorrhaphy entails repair of a damaged spleen and in a way includes partial splenectomy. The following are indications for splenorrhaphy

- Grades I to III splenic injuries
- Patient should be haemodynamically stable
- No associated moderate to severe head injury nor indeed any potentially life threatening injury

Methods of splenorrhaphy include mesh wrap and omental patch techniques.

The main indication for partial splenectomy is when splenic damage or vascular injury is confined to one pole of the spleen. This is the usual situation with iatrogenic injuries. It may also be employed in the diagnosis of idiopathic splenomegaly, splenic cysts, benign tumours and splenic infarcts. Splenorrhaphy is necessary after an anatomical partial splenectomy.

Indications for splenectomy in trauma

- Uncontrollable bleeding
- Grades IV and V injuries
- Very ill patient: Haemodynamic instability with associated acidosis and coagulopathy
- Associated moderate to severe traumatic brain injury: Hypotension predisposes to secondary brain damage

The following is the grading of splenic injuries and a suggestion of the management regime. Grades I and II are regarded as low grade injuries while grade III is of moderate grade. Grades IV and V are managed as high grade injuries.

Grade I: Subcapsular haematoma < 10% surface area or capsular tear/laceration < 1cm deep. This grade of splenic injury is best managed nonoperatively if patient is not bleeding and is haemodynamically stable

Grade II: Includes subcapsular haematoma involving 10 to 50% of surface area and laceration of 1 to 3cm but without blood vessel involvement. Also included is intra-parenchymal haematoma < 5cm. Suggested management is as for grade 1 injuries

Grade III injuries: This includes subcapsular haematoma > 50% surface area/expanding subcapsular haematoma and intraparenchymal haematoma > 5cm. Also included in grade III injuries are splenic lacerations > 3 cm with involvement of the blood vessels. Initial treatment may be nonoperative but there should be no hesitation in resorting to surgical treatment if this fails.

Grade IV injuries: Devascularisation of the spleen (>25%) resulting from laceration of the segmental or hilar vessels. Treatment is either partial or total splenectomy.

Grade V injuries: Shattered spleen or complete devascularisation of the spleen from injury to the hilar vessels. Splenectomy is inevitable

Complications of splenectomy

- Complications of anaesthesia
- Bleeding: Ensure adequate haemostasis during surgery
- Thrombocytosis: May result in thromboembolism
- Infection: Overwhelming post-splenectomy infection (OPSI) is an uncommon but well recognised and most feared complication of splenectomy. This is because the spleen is known to play a major role in the immune response to encapsulated organisms such as streptococcal pneumoniae, haemophilus influenza and neisseria meningitidis. OPSI is more common when splenectomy is performed for haematologic disease compared to splenectomy for trauma. It is commonest in the paediatric age group. The incidence is 0.3% in adults as opposed to 0.6% in children. Although it may occur later, OPSI most commonly develops within the first 2 years after splenectomy. The initial symptoms are non-specific and include malaise, nausea, headache and confusion. There may be rapid progression to shock and death. It goes with a high mortality rate of over 50% in children and 20% in adults. Prevention of OPSI is therefore the most essential key to its management.
 - Conservation of splenic tissue (splenorrhaphy/partial splenectomy) should be encouraged. Total splenectomy should only be carried out in extreme situations. Transplantation of slices of the splenic specimen into the omentum may help to subserve splenic function after splenectomy
 - Immunisation with pneumococcal conjugate vaccine (Pneumovax), haemophilus vaccine and meningococcal vaccine. For elective splenectomy, patient should be immunised 2 weeks prior to the procedure. The timing of administration after emergency splenectomy is, however, under debate. While some authorities believe in administering the vaccine during the immediate postoperative period, others defer it for 7 to 10 days after surgery.
 - Daily antibiotic administration is necessary particularly in children.
- Pancreatic injuries: Owing to its anatomical relationship, the tail of the pancreas is at risk following splenic surgery. Complications include acute pancreatitis, peripancreatic abscesses and pancreaticocutaneous fistula.
- Left colonic injury
- Acute gastric dilatation
- Sub-diaphragmatic abscess: Predisposing factors include peritoneal contamination due to associated bowel injury and pancreatic fistula
- Pleural effusion (left side)
- Disseminated intravascular coagulation

CHAPTER ELEVEN

PEPTIC ULCER DISEASE (PUD), UPPER GASTROINTESTINAL HAEMORRHAGE AND ZOLLINGER – ELLISON'S SYNDROME

An ulcer is a breach in the continuity of epithelium. Peptic ulcer disease occurs under the influence of acid and is a common gastrointestinal problem. The lifetime risk is approximately 10%. The incidence is related to that of *Helicobacter pylori* (*H. Pylori*). The peak age incidence of peptic ulcer disease is 30-40 in the developing world and 40-50 in America. It is currently regarded as a medical disease with specific indications for surgical intervention. The rate of surgical intervention has reduced drastically since the advent of antisecretory drugs such as omeprazole. Currently, the surgeon is mainly called in either when there is failure of medical treatment or in the event of complications arising from the primary disease.

PATHOPHYSIOLOGY

There are three phases of the regulation of gastric acid secretion. They are

- The cephalic phase that is mediated via the vagus nerves. The sight, smell, taste, and even thought of food will stimulate the secretion of acid by the stomach
- The gastric phase that is stimulated by the presence of food in the stomach. It is mediated via the hormone, gastrin which is produced by the G- cells of the gastric antrum
- The intestinal phase whereby the presence of the products of digestion will lead to the secretion of somatostatin. This has a negative feedback mechanism that inhibits the gastric acid secretion.

The parietal cells of the stomach secrete acid. The mucosa of the stomach is, however, protected from the effect of the acid by the presence of a thick mucus barrier which is lined by bicarbonate. The high gastric blood flow equally contributes to the protection of the stomach. Any condition, therefore, that reduces the efficacy of any of these protective mechanisms, will naturally predispose to peptic ulceration. In line with the role of acid secretion in the pathogenesis of PUD, reduction of gastric acid secretion is the key to surgical management. Surgery either by vagotomy or gastric resection is aimed at reduction of gastric acid secretion.

AETIOLOGY:

Peptic ulcer disease arises from an imbalance between the gastroduodenal defence mechanisms and the damaging forces. By and large, there are currently two principal causes of peptic ulcer disease: Infection with *Helicobacter pyloris* and following the use of non-steroidal anti-inflammatory drugs (NSAIDS).

H. pylori: It is a gram-negative flagellate, and spirochaetal organism first discovered by Bircher in 1874. Warren and Marshall discovered its role in the pathogenesis of peptic ulcer disease. *H. pylori* is transmitted by the faeco-oral route and is more prevalent in low socioeconomic communities. It is the major causative factor in PUD (60% gastric and 90% duodenal). In addition to PUD, it is also implicated in the pathogenesis of type B gastritis, and gastric carcinoma. Other

pathologies associated with *H. Pylori* infection include intestinal metaplasia, gastroesophageal reflux disease (GERD), mucosal associated lymphatic tissue (MALT) and lymphoma. The organism elaborates the enzyme, urease which generates ammonia from urea. Ammonia being a potent alkali stimulates the secretion of gastrin by the antral G cells leading to hypergastrinaemia. This results in excessive secretion of gastric acid. It also releases some cytotoxins (CAG A and VAC A genés) which cause direct damage to the gastric epithelial cells, stimulate an inflammatory reaction and may also predispose to malignancy. *H.pylori* does not directly colonise the duodenum. Exposure to the increased gastric secretion, however, induces a gastric metaplastic change in the duodenal epithelium. The latter creates a conducive environment for the colonisation and proliferation of *H.pylori* with its pathophysiological consequences.

Non-steroidal anti-inflammatory agents (NSAIDS) adversely affect the production of the protective gastric mucus. Mucus secretion is stimulated by prostaglandins. NSAIDS inhibit the production of cyclo-oxygenase1 which is the key enzyme in the production of prostaglandin. The protective role of prostaglandin is diminished, thereby predisposing the gut mucosa to peptic ulceration. Due to the direct mucosal injury by NSAIDS, there may be diffusion of non-ionised weak acid into the gastric mucosal cells leading to subsequent haemorrhage and cell necrosis. Presently there are COX- 2 selective agents which have minimal effects on the gastrointestinal tract.

Other risk factors include

- Smoking: its role is controversial. It is said to prevent the healing of an ulcer and may act by decreasing gastroduodenal prostaglandin and bicarbonate production.
- Alcohol: Stimulates gastric acid secretion and increases mucosal permeability.
- Stress ulceration: May cause mucosal ischemia and increased acid secretion. Curling's ulcer is a known complication of severe burn injury while Cushing's ulcers arise following head injury and neurosurgical procedures
- Zollinger-Ellison's syndrome: This is caused by a gastrinoma. See below

SITES OF PEPTIC ULCER DISEASE:

PUD occurs in areas that are in contact with acid. The sites of peptic ulcer disease from above downwards are

- Gastro-oesophageal junction
- Stomach, especially around the lesser curvatur
- The first part of duodenum.
- Unusual sites include the jejunal part of a gastro-jejunostomy and Meckel's diverticulum (due to the presence of heterotopic gastric mucosa). Multiple ulcers in the jejunum may suggest Zollinger-Ellison's syndrome.

Modified Johnson classification of PUD

- Type 1: Body of stomach along the lesser curve at or proximal to the incisura angularis accounts for 60 to 70% of PUD, (associated with low acid secretion)
- Type II: Body of stomach in combination with DU accounts for about 20% of cases; (associated with high acid secretion)
- Type III: Pyloric channel within 3 cm of pylorus (associated with high acid secretion)

- Type IV: Proximal gastro-esophageal ulcers : cardiac ulcers (low acid secretion)
- Type V: Can occur anywhere in the stomach and is associated with chronic use of NSAIDS

There is also a wide spectrum classification into acute and chronic ulcers

PATHOLOGY:

This is essentially the same in both duodenal and gastric ulceration. The ulcer will stimulate a chronic inflammatory reaction which penetrates the mucosa and muscle coat resulting in the formation of fibrous tissue. An excess production of fibrous tissue by a peptic ulcer may result in the narrowing of the lumen and may ultimately end up with gastric outlet obstruction. A penetrating ulcer may eventually lead to destruction of the subserosal layer, perforate the wall of the viscus and result in peritonitis. Anteriorly-placed ulcers are more prone to perforation. In like manner, a penetrating ulcer may encounter a blood vessel in the process resulting in haemorrhage. This is more common with posteriorly-placed ulcers. Thus, the complications of peptic ulcer disease are perforation, bleeding and gastric outlet obstruction. Malignancy is not regarded as a complication of the disease as malignant ulcers are thought to be malignant ab initio. It is most unusual for an originally benign ulcer to metaplate to malignancy. Malignant ulcers are commoner in the stomach. Microscopy of benign ulcer bed reveals fibrosis, basal granulation tissue, chronic inflammatory cells and endarteritis obliterans.

SPECIAL FORMS OF PEPTIC ULCER

- Cushing's ulcer: associated with neurological conditions such as head injury, and neurosurgical procedures
- Curling's ulcer: associated with major burn injury
- Marginal or stomal ulcer: occurs at the site of a gastro-jejunal anastomosis
- Dieulafoy's ulcer: associated with an underlying vascular malformation
- Zollinger-Ellison's syndrome: Due to an underlying gastrinoma

CLINICAL FEATURES: Attempts have been made in the past to distinguish between the clinical features of gastric ulcer from those of duodenal ulcer. They essentially have a common mode of presentation consisting mainly of abdominal pain. The patient may also present with the clinical features of any of the above mentioned complications which are common to both forms of the disease.

The abdominal pain of PUD has the following peculiar characteristics

- Epigastric:: May radiate to the back due to penetration of the ulcer :
- Relationship to meals: Aggravated by hunger and relieved by eating.
- Wakes the patient up at night
- Demonstrates periodicity: Waxing and waning of pain. The typical ulcer pain is interspaced with pain-free intervals. Attempts at ulcer healing will result in temporary remission, while subsequent breakdown of the healing process will manifest as relapse of symptoms

Passage of dark, tarry stools (melaena) is due to chronic blood loss. Peptic ulcer disease is one of the common causes of unexplained anaemia due to chronic blood loss.

Examination may reveal a patient who is pale from chronic blood loss. The uncomplicated case may only demonstrate epigastric tenderness on abdominal examination. Rectal examination may show evidence of melaena stool. The complicated PUD will demonstrate the relevant clinical features

Differential diagnoses of peptic ulcer disease include gastric cancer, gastritis, worm infestation, gallbladder disease, chronic or recurrent pancreatitis and hiatus hernia. Others are reflux oesophagitis, carcinoma of the transverse colon, intestinal parasites and non-ulcer (functional) dyspepsia.

Investigations: Specific and non-specific

- Oesophagogastroduodenoscopy: This is the gold standard. In addition to visualising the ulcer, biopsies can equally be taken for histology. This is even more important when dealing with a gastric ulcer which may be malignant in nature. It is equally of importance in monitoring the outcome of medical treatment.
- Barium meal and follow-through: May reveal an ulcer crater. It has a 90% accuracy and diagnostic features may be made more obvious with an air-contrast study.
- Helicobacter pylori screening tests: biopsy specimen, serology (ELISA), rapid urease test and urea breath test.
- Stool tests: ova, cysts and protozoa; screen for occult blood
- Full blood count: Patient may be anaemic from chronic blood loss.
- Serum gastrin level: especially when diagnosis of Zollinger-Ellison's syndrome is being entertained
- Tests for gastric acid secretion are no longer popular.

TREATMENT OF PEPTIC ULCER DISEASE

As mentioned earlier, peptic ulcer disease is basically a medical disease. Medical treatment is aimed at reducing the level of gastric acid secretion. Duodenal ulceration (Johnson's types II and III) is associated with an increased secretion of acid while the level of secretion may be normal or even low in gastric ulceration (Johnson's types I and IV). Irrespective of the degree of acid secretion, however, all peptic ulcers are known to heal in a minimal acid environment. The eradication of H.pylori which is an established causative agent, forms an integral part of the medical treatment of peptic ulceration. Treatment is aimed at relieving pain, enhancing healing of ulcer and preventing complications associated with peptic ulcer disease.

General treatment is in the form of counselling: Stop smoking and/or alcohol abuse, and discontinue with the use of NSAIDS.

Eradication therapy: It is targeted at the eradication of H. Pylori and consists of the administration of a combination of clarithromycin (or amoxycillin), metronidazole and omeprazole for a period of 10 to 14 days. Three regimens are currently in use:

- Omeprazole, amoxicillin and clarythromycin (OAC)
- Omeprazole, metronidazole and clarythromycin (OMC)
- Omeprazole, amoxicillin and metronidazole (OAM)

It should be noted that proton pump inhibitor is the common denominator in all the above regimens. At the end of treatment with the combination therapy, omeprazole therapy should be continued for two weeks as monotherapy.

Reduction of gastric acid secretion: This is achieved by the administration of H₂ receptor blockers (cimetidine and ranitidine) and proton pump inhibitors (omeprazole, esomeprazole, lansoprazole)

Other drugs include sucralfate (complexes with bile salts and pepsin and binds to proteins in mucosa) and antacids (neutralise gastric acidity and decrease activity of pepsin). Antacids and H₂-receptor blockers should not be combined with omeprazole. The alkaline medium created by the use of antacids is unconducive for the action of proton pump inhibitors. Co-therapy with misoprostol, a prostaglandin analogue may be considered in NSAID-dependent patients. In the alternative, a safer NSAID that selectively inhibits the inducible isoform of cyclo-oxygenase (COX-2 inhibitor) could be considered. Treatment with antisecretory drugs should be continued for six months to one year in order to prevent relapse.

In addition, patient should be counselled on relevant lifestyle changes such as cessation of smoking and curtailing the intake of alcohol. NSAIDs should either be discontinued completely or substituted with drugs that have less ulcerogenic effects.

INDICATIONS FOR SURGICAL MANAGEMENT:

Essentially failure of drug treatment and complications

- Failure of properly supervised medical treatment. Ensure patient's compliance with prescribed treatment before establishing failure of treatment.
 - Symptoms persist for more than 3 months despite active and appropriate drug therapy
 - Ulcer recurs within one year after initial healing despite maintenance therapy
 - Persistent periodicity: Cycles of prolonged activity with brief remissions
- Presence of complications: Bleeding, perforation, gastric outlet obstruction
- Revision of previous surgery

Principle of surgical treatment: Like its medical counterpart, surgery is aimed at reducing the level of gastric acid secretion. This may be achieved by any of the following

- Reduction of vagal stimulation (vagotomy): The vagus nerves (anterior and posterior) on exiting from the gastro-oesophageal sphincter give out two main branches. The coeliac branch from the posterior vagus innervates the gastrointestinal system up to the level of the proximal one-third of the transverse colon while the anterior vagus gives off a hepatic branch that innervates the hepatobiliary system. Thereafter, they run along the lesser curvature of the stomach giving off twigs that innervate the stomach. The vagi are secreto-motor to the stomach. The vagal trunks finally terminate in a crow-foot manner at the pylorus. The terminal 'crow foot' twigs are motor nerves and are responsible for the rhythmic contraction of the pylorus. This regulates the mechanism by which gastric contents are discharged into the duodenum. Vagotomy is aimed at eliminating the influence of the cephalic phase of gastric acid secretion. In addition to reducing the secretion of gastric acid by the stomach, truncal vagotomy also paralyses the muscles of the pylorus thereby creating

a challenge with gastric emptying. For this reason, truncal vagotomy is usually accompanied by a drainage procedure. The latter includes pyloroplasty (widening of the pyloric channel) and gastrojejunostomy. Highly selective or proximal gastric vagotomy involves careful transection of the vagal secretory fibers that supply the stomach and the lower 5 cm of the oesophagus while sparing the motor nerve (nerve of Latarjet) that supplies the pyloric antrum. The latter constitute the terminal branches of the anterior and posterior vagal nerve trunks. This procedure conserves the motor function of the pylorus thereby obviating the need for a drainage procedure. It is the most physiological acid-reducing procedure. Unfortunately it is a much more technical procedure and is attended with a higher recurrence rate (2 to 10%) when compared to that of truncal vagotomy (2 to 7%).

- Reduction of the parietal cell mass: This can be achieved by way of distal partial gastrectomy. In addition to this, antrectomy equally excises the gastric antrum which is the site of gastrin secretion

As pointed out earlier, the advent of very potent antisecretory drugs has reduced the rate of elective surgery in the management of peptic ulcer disease. Whereas surgery for duodenal ulcer is generally conservative, that in gastric ulceration is more radical. This is due to the higher incidence of malignancy associated with gastric ulceration. The following is a summary of the surgical treatment of uncomplicated peptic ulcer disease

- Duodenal ulcer: Truncal vagotomy and drainage (pyloroplasty, or gastrojejunostomy) or highly selective vagotomy (does not require a drainage procedure)
- Gastric ulcer: May involve antrectomy. The nomenclature attached to the procedure depends on the mode of reconstitution of the gastrointestinal system after resection. Reconstitution by anastomosing the duodenum to the gastric stump (gastroduodenostomy) is referred to as Billroth 1 procedure. On the other hand, anastomosing the jejunum to the gastric stump (gastrojejunostomy) is referred to as Billroth 2 procedure. Antrectomy is usually accompanied by a truncal vagotomy.
 - Type 1 gastric ulcer: Distal gastrectomy with Billroth 1 anastomosis
 - Type 1 1/2 gastric ulcer: Antrectomy that includes the gastric ulcer plus truncal vagotomy. Reconstruction may be by either of the Billroth anastomoses. Alternatively, patients could have truncal vagotomy and either pyloroplasty or gastrojejunostomy
 - Type 1 1/2: Antrectomy and truncal vagotomy
 - Type 1V: Difficult to treat. Surgical management depends on the size of the ulcer and the degree of surrounding inflammation. Equally important is the distance of the ulcer from the gastro-oesophageal junction.
 - Type V: Rarely requires surgery. Malignant disease should be suspected if ulcer fails to heal with medical treatment.

COMPLICATIONS OF PEPTIC ULCER DISEASE

PERFORATION

: As earlier mentioned, there is current decline in the rate of elective surgery for peptic ulcer disease. This is, however, associated with a corresponding increase in the rate of surgery for perforated peptic ulcer. This change in epidemiology is attributed to three factors: an ageing population, a rising incidence of *H. pylori* infection and chronic use of NSAIDs. Perforation due to *H. pylori*-induced peptic ulceration tends to be commoner in the younger age group and has a male preponderance

As pointed out earlier, this complication is more common with anteriorly-disposed ulcers. It ultimately results in peritonitis. With the perforation, gastric contents (including bile) are discharged into the peritoneal space. There are three clinical phases of peritonitis due to perforation from PUD

- Phase of initial chemical peritonitis: This is due to flooding of gastric contents and bile into the peritoneal cavity and is usually accompanied by severe abdominal pain. The patient may be able to tell the exact time of perforation. The osmotic activity of this peritoneal fluid will result in the absorption of fluid into the peritoneal space leading to dilution of acid and its painful effect.
- Phase of illusion: The raised osmotic activity of the pathological peritoneal fluid will result in the absorption of fluid into the peritoneal space leading to dilution of acid and its painful effect. This manifests clinically as a marked reduction in pain. This situation is illusory as there is silent invasion and proliferating of bacteria
- Phase of bacterial peritonitis: This marks the manifestation of bacterial peritonitis and is heralded by the recrudescence of symptoms. Patient may manifest with clinical features of 'mixed shock' (bacterial and hypovolemic) at this stage.

Clinical features: Perforation is heralded by a sudden onset of epigastric pain. As pointed out earlier, the patient may be able to recall the exact time of perforation. There may be a past medical history suggestive of peptic ulcer disease. Vomiting, abdominal distension and features of shock may be present. It should be noted that sometimes the patient will present with right iliac fossa pain. This is due to the trickling down of gastroduodenal effluent down the right iliac fossa through the right paracolic gutter. The clinical features may in this instance, mimic those of acute appendicitis. It is important to note that 'silent' perforation is known to occur in elderly and very ill patients.

Examination will reveal an ill-looking patient who is in obvious distress and may manifest with clinical features of dehydration and shock. Abdominal examination may reveal a distended abdomen that does not move with respiration. There may be generalised guarding and tenderness especially around the epigastrium. Bowel sounds may be diminished or absent. Digital rectal examination should always be carried out. Late presentation may manifest with clinical features of septic shock and acute renal failure which are known complications of perforated peptic ulcer disease.

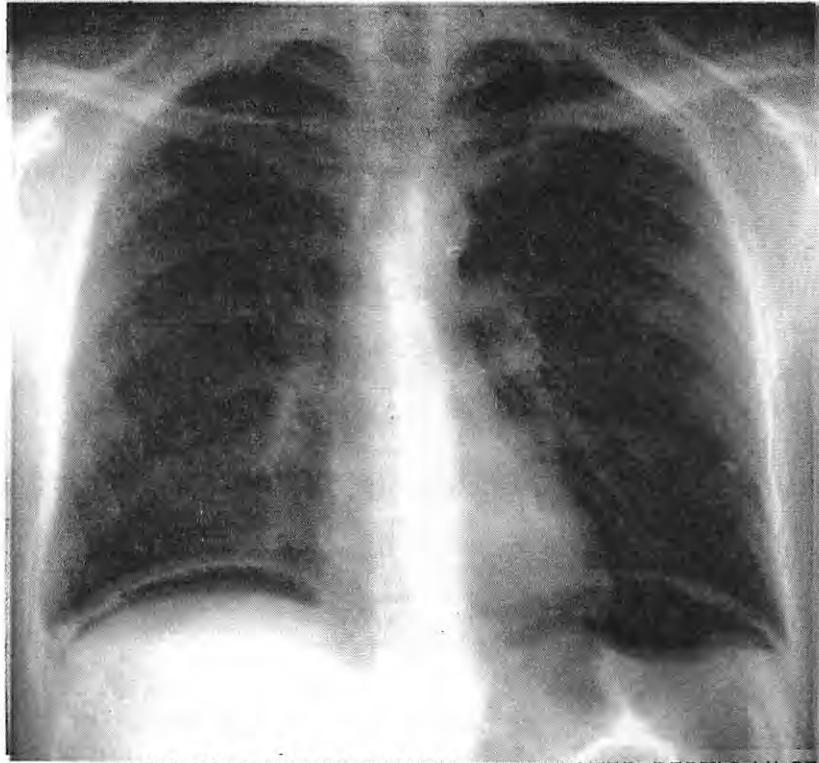
Differential diagnoses include acute cholecystitis, acute pancreatitis and intestinal obstruction
Treatment: The ultimate treatment is surgical. It is, however, necessary to resuscitate the patient and carry out some basic investigations before surgery

Resuscitation:

- Place patient on nil orally
- Set up an intravenous line for replacement of fluid loss. Intravenous fluids of choice are Ringer's lactate and normal saline
- While setting up the infusion line, take blood samples for some basic investigations: Full blood count, electrolytes, urea and creatinine. Group and cross-match two units of blood
- Catheterise the urethra and monitor the urinary output. This monitors the degree of resuscitation with intravenous infusion
- Commence on intravenous antibiotics. This should include metronidazole in view of the possible presence of anaerobic organisms.
- Administer intravenous analgesics.

The following investigations should be carried out when the clinical state of the patient has been stabilised:

- Plain abdominal x-rays (erect and supine): The erect film may demonstrate presence of air under the diaphragm. This may be more obvious in an erect chest radiograph. This sign may be absent in about 50% of cases.
- Contrast series: This is achieved by the use of a water soluble medium which is either administered orally or through a nasogastric tube. It helps in the detection of an intraperitoneal leak. It should be noted that about 50% of the perforations would have healed at presentation.
- Other tests include serum amylase (to rule out acute pancreatitis) and abdominal ultrasound scan to rule out other acute abdominal conditions.



CHEST RADIOGRAPH OF A PATIENT WITH PERFORATED PEPTIC ULCER DISEASE SHOWING THE PRESENCE OF GAS UNDER THE DIAPHRAGM

Surgical treatment: This entails an exploratory laparotomy after adequately resuscitating the patient. At surgery, purulent fluid is sucked out of the peritoneum and the perforation is identified. Perforation is closed transversely with interrupted non-absorbable sutures. Closure of perforation may be reinforced by tacking a viable segment of omentum over the repair. This is referred to as Graham-Douglas procedure. Other modes of closure include 'sutureless' repair with fibrin glue and pedicled ligamentous teres repair. Fibrin glue repair is less time-consuming. This line of treatment applies to the treatment of both perforated duodenal as well as gastric ulcers. Biopsy of the ulcer is advised in gastric ulcers due to the high incidence of malignant ulcers in the stomach. Adequate peritoneal lavage is carried out with warm saline followed by the insertion of a tube drain in the pelvis. Postoperatively, the patient is continued on intravenous fluids and antibiotics.

The conventional surgical treatment of truncal vagotomy is not carried out in the surgical treatment of perforated peptic ulcer disease for two reasons. The first has to do with the high risk of mediastinitis sequel to the cephalad spread of infection in the course of manipulating the lower oesophagus to gain access to the vagus nerves. The second reason the availability of highly effective antisecretory agents and the knowledge of the role of *H. pylori* in the pathogenesis of peptic ulcer disease. Postoperatively, the patient is commenced on antisecretory therapy and is screened for *H. pylori* infection. The urea breath test is quite simple and convenient. Eradication therapy is commenced if the test proves positive for *H. pylori*.

There is a place for conservative management in selected cases that present early and in whom there is radiological evidence of sealing of the perforation. This non-operative approach has, as would be expected, been found to have a higher incidence of sepsis and intra-abdominal abscess. Surgical treatment may be carried out via a laparoscopic approach.

Prognostic factors: The following are known to adversely affect the outcome of management of perforated peptic ulcer

- Age over 75 years
- Delay in diagnosis and treatment (worse after 24 hours)
- Presence of shock
- Coexistent medical disease

GASTRIC OUTLET OBSTRUCTION:

The most common causes of gastric outlet obstruction are PUD and ~~antral~~ carcinoma. Other causes are

- Congenital hypertrophic pyloric stenosis
- Adult pyloric stenosis
- Advanced carcinoma of the head of the pancreas
- Pyloric mucosal diaphragm

Pathophysiology: Healing of a peptic ulcer by fibrosis may result in obstruction of the outlet of the stomach at the pyloro-duodenal region. This leads to the accumulation of ingested food material in the stomach as only very little is able to trickle beyond the point of obstruction. In response to this, the stomach distends and may be so distended as to get into the pelvis. Subsequently, there is vomiting with loss of electrolytes. The chronic loss of gastric fluids results in a biochemical state of hyponatremic, hypokalemic, hypochloremic, metabolic alkalosis. If this state of affairs persists, the reduction in the renal blood flow may stimulate the renin-angiotensin mechanism resulting in the release of aldosterone. The latter aids the reabsorption of sodium in the distal renal tubules. This is an exchange mechanism as the sodium is exchanged for potassium which is secreted into the urine. In the presence of severe hypokalemia, hydrogen ions are made to substitute for potassium. This results in the secretion of acidic urine albeit in the presence of metabolic alkalosis. This paradoxical situation is referred to as paradoxical aciduria. Similarly, the patient may present with clinical features of hypocalcaemia. This is due to a reduction in the level of ionised calcium resulting from metabolic alkalosis. The total serum calcium level in this condition may be deceptively normal.

CLINICAL FEATURES: These are fallouts from the above pathophysiology

- There may be a past medical history of peptic ulcer disease
- Vomiting: Projectile, non-bilious vomitus consisting of stale, undigested food. Patient may be able to recognise food eaten a few days earlier
- Weakness and loss of weight: Loss of fluid and electrolytes
- May present with features of dehydration and shock
- Abdominal examination may reveal visible peristalsis and a peculiarly dilated stomach that can be seen (outline), felt (by palpation) and heard (by way of succussion splash).
- Clinical features of tetany in severe cases

Initial management after history and physical examination entails active resuscitation.

- Set up an intravenous infusion line and commence resuscitation with normal saline. Ringer's lactate is contraindicated as it may worsen the already existing metabolic alkalosis.
- While setting up the intravenous line, take blood samples for the following investigations: full blood count, electrolytes/urea/creatinine and grouping and cross-matching of blood. The initial PCV may be deceptively high due to haemoconcentration. The actual reading will manifest after resuscitation with intravenous infusion
- Pass a wide-bore nasogastric tube and lavage the stomach with normal saline until the effluent is clear. In addition to clearing the stale contents of the stomach, nasogastric aspiration and gastric lavage equally shrink the stomach. By this manoeuvre, the stomach is cleaned up with improved handling qualities at surgery
- If patient is severely dehydrated or in shock, pass a urethral catheter and monitor the urinary output. An adequate urinary output signifies adequate rehydration
- Correct any electrolyte anomaly detected. Hypokalaemia is a common electrolyte anomaly that is fraught with anaesthetic complications. It should be corrected via the intravenous route when the urinary output improves. As pointed out earlier, the initial haemo-concentration will be corrected with adequate intravenous infusion thereby reflecting the true PCV level. Estimation of serum haemoglobin level is believed to give a better reflection of the true haematological state of the patient.
- Diagnosis can thereafter be confirmed by endoscopy or barium meal. The two-hour film in the latter will show minimal presence of contrast medium beyond the point of obstruction.

The ultimate treatment is surgical and involves truncal vagotomy and drainage. As regards the accompanying drainage procedure, some surgeons insist on carrying out pyloroplasty as it is thought to be a more natural pathway than gastrojejunostomy. The problem with this lies in the difficulty associated with creating an effective, patent channel in the presence of a severely scarred pyloro-duodenum. This has tilted the scale in favour of gastrojejunostomy as the drainage procedure of choice in the surgical management of gastric outlet obstruction due to peptic ulcer disease. Some work has been done in relation to balloon dilatation of the obstructed lumen of the pylorus particularly in early cases. Balloon dilatation may have to be repeated for effective outcome. Duodenal perforation is a known complication of balloon dilatation..

UPPER GASTROINTESTINAL BLEEDING

This is defined as bleeding into the lumen of the upper gastrointestinal system proximal to the ligament of Treitz. It accounts for 80% of all significant gastrointestinal haemorrhage and requires prompt diagnosis and treatment.

AETIOLOGY: Common causes include

- Peptic ulcer disease (duodenal and gastric): This is the most common cause. Bleeding duodenal ulcer accounts for 25% of cases while bleeding gastric ulcer has an incidence of 20%

- **Acute erosive gastritis:** Accounts for 20% of significant upper GI haemorrhage. It is due to stressful stimuli. Common causes include head injury (Cushen's ulcer), burns (Curling's ulcer), hepatic failure and shock. The common denominator is impaired mucosal blood flow. Drugs such as NSAIDs and steroids equally predispose to acute erosive gastritis. About 10% of patients on daily NSAIDs develop an acute ulcer. The main difference between acute erosive gastritis and chronic gastritis is the absence of bleeding in the latter.
- **Oesophageal varices:** This is one of the clinical manifestations of portal hypertension sequel to cirrhosis of the liver
- **Oesophagitis:** Due to mucosal erosions from gastro-oesophageal reflux, infections or medications (NSAIDs)
- **Mallory-Weiss syndrome:** Accounts for 15% of significant upper GI haemorrhage. It results from a longitudinal mucosal tear around the gastro-oesophageal sphincter. It is sequel to vigorous retching, coughing or vomiting in the presence of a closed sphincter. A full-thickness tear (oesophageal rupture) may follow repeated vomiting (Boerhaave's syndrome). Mallory-Weiss syndrome usually follows a bout of alcoholic intake.
- **Advanced gastric cancer:** Carcinoma and lymphoma
- **Gastrointestinal stromal tumours (GIST):** These may present primarily with upper gastrointestinal bleeding. Leiomyoma and leiomyosarcoma may also present with upper gastrointestinal haemorrhage
- Other less common causes include Dieulafoy's ulcer (due to an underlying aberrant submucosal artery), aortoenteric fistula, angiodyplasia, and oesophagitis.
- It is important to consider systemic causes of bleeding diathesis such as thrombocytopenia, disseminated intravascular coagulation (DIC) and anticoagulant therapy

Clinical features: Patient will present with haematemesis and melaena. Haematochezia (passage of fresh blood in stool) may be present if the rate of bleeding is very high. There may be associated epigastric discomfort/pain. In severe upper upper GI bleeding, patient may present with symptoms of shock such as weakness and syncopic attacks. There may be a past medical history suggestive of peptic ulcer disease. A positive history of ingestion of NSAIDs, and a social history of chronic alcoholism would be useful in elucidating the aetiology.

Examination will reveal a highly distressed, ill-looking patient with clinical features of hypovolemic shock. Digital rectal examination may show evidence of melaena or haematochezia. Immediate resuscitation is the hallmark of the clinical management of a patient presenting with upper gastrointestinal haemorrhage.

- Prompt attention should be given to the airway, breathing and circulation of the patient
- Quick assessment to know if bleeding is acute (haematemesis, coffee-ground emesis, bleeding per rectum or melaena) or chronic (occult bleeding with clinical features of anaemia)
- Assess the severity of blood loss
- Commence on nil orally

- Set up an intravenous line using a wide bore cannula. Commence resuscitation with either normal saline or Ringer's lactate
- Before commencing the infusion, take blood samples for some baseline investigations: Full blood count (including the platelet count), liver function test, electrolytes, urea and creatinine; group and crossmatch at least 3 units of fresh whole blood. The clotting profile (prothrombin time and partial thromboplastin time) should be assessed to rule out bleeding diathesis.
- Pass a urethral catheter to monitor the effectiveness of the resuscitation exercise
- Depending on the amount of blood loss, commence blood transfusion as soon as cross-matched blood is available
- Pass a wide-bore nasogastric tube and irrigate the stomach with room-temperature water or normal saline until the effluent is clear. This measure not only clears already existing blood clots from the stomach but equally alerts the attending clinician of any recurrent bleeding.
- Commence on intravenous antisecretory drugs: Administer either H₂-receptor blockers (cimetidine and ranitidine) or proton pump inhibitors (omeprazole, pantoprazole). Octreotide, a somatostatin analogue which reduces both splanchnic blood flow and acid secretion has been found effective in the management of non-variceal bleeding. Misoprostol, a prostaglandin analogue, has also been found useful.

It is only when the patient has been adequately resuscitated and the bleeding brought under control that an exhaustive clinical history should be taken with a view to identifying the aetiological factor. Fortunately, bleeding tends to be self-limiting in approximately 80% of patients with upper gastrointestinal haemorrhage. Comprehensive clinical assessment is followed by relevant specific investigations aimed at confirming the source of the bleeding.

The gold standard investigation remains endoscopy (oesophagogastroduodenoscopy). It has been found to establish diagnosis in more than 90% of cases. Upper GI endoscopy also assesses the current activity of bleeding. This core investigation should ideally be carried out within the first 24 hours of admission as it serves a dual purpose - diagnostic as well as therapeutic. It has been shown to be effective in achieving initial haemostasis, and also in the reduction of both clinical rebleeding and the need for urgent surgical intervention. In addition to locating the site of the bleeding and its possible aetiological factor, haemostatic therapeutic procedures can be carried out. These include the use of

- Bipolar diathermy: Heat generated by an electrical current causes coaptive coagulation of bleeding vessels
- Adrenaline injection (1 in 10,000 solution): Causes vasospasm and activation of platelets. Platelet and fibrin thrombus are formed within the lumen of the vessel. It is particularly useful where the bleeding point is obscured by an adherent clot.
- Injection sclerotherapy: Sclerosants include 3% sodium tetradecyl sulphate (STD), 5% ethanolamine, absolute alcohol and 1% polidocanol. Whereas alcohol acts by dehydration and fixation of tissue, the others are detergents and act by causing endothelial damage.
- Heat probe: Does not require electric current

- **Laser photocoagulation:** Delivers energy which is converted to heat on contact with tissue. Complications include perforation and exacerbation of bleeding if improperly applied.

Newer endoscopic procedures include the use of fibrin glue injection and microclip application. Studies have shown the better efficacy of combination of endoscopic modalities of treatment over single modality

- Selective mesenteric angiography may be employed to elucidate the site of massive haemorrhage when endoscopy proves ineffective.

Surgical intervention: Most of the patients with bleeding peptic ulcer disease will respond to conservative management. They are thereafter placed on eradication therapy and continued on proton pump inhibitors. Some, however, will continue to bleed despite the above measures.

Indications for surgical intervention include

- Failure of endoscopic therapy
- Massive, persistent or recurrent upper GI bleeding such that the patient requires more than three units of blood to stabilise the cardiovascular system: The older the patient, the earlier the requisite intervention. This is due to the low cardiovascular reserve in the elderly. A subsequent major bleed in the elderly may spell disaster. There is a place for repeat endoscopy in the management of rebleeding. This has been shown to reduce the rate of surgical intervention
- Haemodynamic instability inspite of aggressive fluid replacement
- When there is insufficient blood in the bank and patient continues to bleed. In this situation, it may be more prudent to identify and plug the 'leaking tap' early by way of surgical intervention. This may be regarded as a relative indication for surgery
- Endoscopic finding of non-healing or giant ulcer (>3cm)

As mentioned earlier, bleeding is quite common with posterior ulcers. Therefore at laparotomy after due resuscitation, a longitudinal incision is made over the pylorus and the first part of the duodenum. This exposes the posterior aspect of the pyloroduodenal region. Bleeding here is usually due to erosion of the gastroduodenal artery. The bleeding point is underrun with two appropriately placed sutures (one above and one below the bleeding point) in order to establish haemostasis. This is accomplished by the use of an absorbable suture that is long-lasting and with reasonable strength such as vicryl. The incision is then closed transversely in the form of a pyloroplasty. The standard operative management of the peptic ulcer disease is thereafter completed with the addition of a truncal vagotomy.

In the event that no ulcer is found on opening the duodenum, the incision should be extended superiorly into the stomach. If a gastric ulcer is located, it is treated by local excision, and closure of the incision is with a pyloroplasty as described above. As usual, surgery is completed with a truncal vagotomy. Partial gastrectomy is an alternative procedure in the case of a bleeding gastric ulcer.

Occasionally, one is confronted with a situation of generalised oozing of blood from the mucosal surface of the stomach and duodenum. This is usually sequel to erosive gastritis. Rather than contemplate on a possible total gastrectomy in a very ill patient, it may be more prudent to carry out the basic truncal vagotomy and pyloroplasty.

Angiography has a place in the management of upper GI bleeding. In addition to identifying the bleeding vessel, haemostasis may be achieved by arterial embolisation with gel foam, metal coil springs or a clot. Arterial vasopressin has also been used to control bleeding due to peptic ulcer disease

STRESS ULCERS: As the name implies, they occur under stressful conditions and present as superficial erosions. The aetiology includes burns (Curling's ulcer), and following head injury or neurosurgical procedures (Cushing's ulcer). Other causes include shock, and trauma (reduced gastric blood flow). Stress ulcers usually manifest as drainage of frank blood from the nasogastric tube. The situation is best prevented by early management of shock, and by the prophylactic administration of intravenous antisecretory drugs such as omeprazole and H₂-receptor blockers such as cimetidine. Confirmation of diagnosis is by oesophagogastroduodenal endoscopy. Initial conservative treatment involves cold saline lavage and administration of intravenous proton-pump inhibitor. Refractory cases will require surgical intervention (vagotomy and pyloroplasty) or total gastrectomy in difficult cases with uncontrollable haemorrhage.

Prognostic factors associated with increased bleeding and mortality in upper GI haemorrhage:

These are categorised into clinical and endoscopic

Clinical factors

- Shock on admission
- Age greater than 60: Increased mortality but no increase in rebleeding
- Prior history of bleeding requiring transfusion
- Admission haemoglobin less than 8 grams/dl
- Continued bleeding as revealed by the bloody nasogastric aspirate
- Transfusion requirement of 5 units and above

Endoscopic factors

- Visible vessel in base of ulcer
- Oozing of bright blood form ulcer base
- Adherent clot at ulcer base
- Location of ulcer (worse prognosis when located near large arteries such as posterior duodenal bulb or lesser curve of stomach)

Rebleeding rates in order of frequency are: Oesophageal varice (60%), gastric cancer (50%), gastric ulcer (28%) and duodenal ulcer (24%). Others are gastric erosions (15%), Mallory-Weiss tear (7%) and no identified source (1.5%).

COMPLICATIONS OF SURGERY FOR PEPTIC ULCER DISEASE

- Haemorrhage: most likely from the site of anastomosis
- Duodenal stump blow-out: usually affects the afferent limb of the Billroth anastomosis. Patient presents with abdominal pain and features of peritonitis. Treatment involves resuscitation, re-exploration and repair over a duodenostomy tube for drainage
- Stomal obstruction
- Acute pancreatitis

- **Early and late dumping syndrome (Postvaginal syndrome):** Both arise from the loss of the storage capacity of the stomach following a drainage procedure. Under this situation, ingested food is 'dumped' as it were directly into the small intestine due to loss of the regulatory activity of the pyloric sphincter mechanism. The high carbohydrate load increases the intestinal osmolarity. Consequently, fluid is drawn from the circulation into the intestine by the principle of osmosis. This reduction in the circulatory blood volume is responsible for the early dumping syndrome. The symptoms are similar to those of hypovolemic shock. It is heralded by epigastric fullness, colicky abdominal pain, sweating, tachycardia, and weakness shortly after a meal consisting mainly of carbohydrates and fluid. Typical of a state of shock, it is relieved by lying down.

Late dumping syndrome, on the other hand, occurs about an hour after a meal. It is a form of reactive hypoglycaemia. The 'dumping' of an unusually large amount of carbohydrate leads to a rapid absorption of glucose into the circulation. The resultant hyperglycaemia stimulates the pancreas to secrete a correspondingly high amount of insulin. This induces a hypoglycaemic state (blood sugar may be as low as 50 mg %). Clinically, the patient will manifest with features of hypoglycaemia (tremors, fainting and sweating) about an hour after a meal. Typical of hypoglycaemia, it is relieved by the ingestion of high-calorie drink or food.

Management of dumping syndrome involves advising the patient to take rather small but frequent meals. Fluids with high carbohydrate content should also be avoided. Administration of octreotide (a somatostatin analogue) before meals may be of help. Revisional surgery has been found useful in intractable cases. Conversion of an earlier gastrojejunostomy to a Roux-en-Y anastomosis has been found to be effective.

- **Recurrent ulceration:** This may result from incomplete vagotomy. Recurrence rate is higher with highly selective vagotomy. Management is with antisecretory drugs. Only in refractory cases is re-exploration advised with the aim of identifying and transecting the hitherto unidentified vagal nerve fibers. Recurrent ulceration may also result from the presence of an occult gastrinoma and persistence with smoking habit after surgery.
- **Stomal ulceration:** Usually occurs at the jejunal side of the anastomosis. Treatment is with antisecretory drugs
- **Small stomach syndrome:** Gastric resection results in a decreased gastric capacity. There is easy satiety which may result in inadequate intake and nutritional deficiency. The latter may manifest as weight loss, iron deficiency anaemia and steatorrhoea.
- **Postvagotomy diarrhoea:** This is the fallout from both the rapid gastric emptying as well as the denervation of the upper gastrointestinal tract sequel to vagotomy. The severity of the diarrhoea ranges from mild discomfort to an almost unbearable situation. Treatment is difficult and may consist solely of the patient being advised to take relatively dry, small but frequent meals. Antidiarrhoeal agents like codeine may be helpful.

- Malignant transformation: This may result from the reflux of bile into the gastric remnant following antrectomy. The chronic irritation may subsequently result in a metaplastic change in the gastric remnant. Anacidity following vagotomy and atrophic gastritis may also be contributory factors.
- Gall stones: Truncal vagotomy denerves the gall bladder resulting in stasis. Stasis is one of the known factors that predispose to cholelithiasis.
- Anaemia: Megaloblastic anaemia may result from loss of intrinsic factor normally secreted by the stomach. This aids the absorption of Vitamin B₁₂ in the terminal ileum. Megaloblastic anaemia may therefore follow gastric resection. It is usually a late development due to the appreciable body store of Vitamin B₁₂. Iron deficiency anaemia is due to both reduced absorption of iron as well as chronic blood loss associated with peptic ulceration.
- Bile vomiting: Can be quite distressing. Severe cases may require revisional surgery by way of a Roux-en-Y anastomosis.

ZOLLINGER-ELLISON SYNDROME (Z-E SYNDROME)

This clinical condition results from an excessive secretion of gastrin. This gives rise to multiple sites of ulceration in the gastrointestinal system. Z-E syndrome is due to an underlying gastrinoma. The latter most commonly occurs in the pancreas (non beta islet cells 80%). Other locations include the third part of the duodenum and around the so-called gastrinoma or Passaro's triangle. This is the area bounded by the cystic duct/common bile duct junction, the junction of the second and third parts of the duodenum and the neck of the pancreas. About 80% gastrin-secreting tumours are located in this region. In about 25% of cases, this syndrome constitutes part of the Multiple Endocrine Neoplasia 1 (MEN1) syndrome. The other associated neoplasias include those of the pituitary and the parathyroid glands.

The excessive secretion of gastrin results in multiple gastrointestinal ulcers particularly in unusual sites such as the jejunum. In like manner, there is an exaggeration of all the complications of peptic ulceration such as upper GI bleeding, perforation and gastric outlet obstruction. The malignant variety may metastasise to the liver.

The following are pointers to a possible gastrinoma in a patient with peptic ulcer disease

- Unusual sites of ulceration such as the distal duodenum and the jejunum
- Associated diarrhoea: Due to excessive gastric secretion
- Associated hypercalcaemia or pituitary pathology in the patient or family

The patient presents with epigastric pain, and diarrhoea. The latter is due to the effects of acid on the digestive enzymes. The relevant investigations include serum gastrin level (normal level: 200pg). In this condition it could be up to 1000pg. In the secretin test, the basal acid secretion is increased following the injection of secretin. In Zollinger-Ellison syndrome, however, there is no appreciable increase with the administration of secretin as the parietal cells are already secreting at an optimal level.

Serum calcium estimation may help to rule out an associated parathyroid adenoma. Special investigations to identify the site of the tumour include endoscopic ultrasound scan, CT scan, MRI

and pancreatic CT. The usual investigations for peptic ulceration (endoscopy and barium meal) are equally carried out.

Treatment: The ideal treatment should entail localisation and resection of the primary tumour. This is, however, not an easy exercise as most of these tumours are small and multifocal. Until recently, treatment was targeted at surgical treatment of the secondary peptic ulcer disease. This involved total gastrectomy with Roux-en-Y oesophagojejunostomy. The potency of current anti-ulcer drugs, however, has given them a pride of place in the treatment of Z-E syndrome. The antisecretory drugs include H₂ receptor blockers and omeprazole. Surgical excision may be necessary in the management of the malignant variety.

CHAPTER ELEVEN

CARCINOMA OF THE STOMACH AND GASTROINTESTINAL STROMAL TUMOURS (GIST)

The incidence of carcinoma of the stomach is rising. It is the second leading cause of cancer-related deaths and the fourth most commonly diagnosed cancer. Gastric cancer is associated with a high degree of morbidity and mortality and accounts for about 800,000 deaths worldwide annually. It has therefore been infamously referred to as 'one of the captains of the men of death'. Japan has the highest global incidence rate (78/100,000) as it accounts for 75% of her national annual cancer-related deaths. The poor prognosis (5-year survival of 5-15%) is due to late presentation.

Risk factors: These are multifactorial and include precancerous lesions, environmental and genetic factors

- Precancerous lesions include atrophic gastritis, chronic gastritis, and pernicious anaemia.
- Previous gastric surgery particularly for peptic ulceration (partial gastrectomy, gastrojejunostomy and pyloroplasty). This is attributed to duodeno-gastric reflux of bile.
- H.pylori infection: Results in gastritis, gastric atrophy and intestinal metaplasia which could later give rise to gastric malignancy. The latter affects the distal part of the stomach.
- Environmental factors: Lifestyle (smoking, alcohol, and obesity), low vitamin A and C consumption and excessive consumption of red meat. Smoked fish may predispose to it. This may be due to exposure to benzpyrene. It is believed that these food substances generate nitrates which are metabolised to nitrites. The end-product, nitrosamine, is a confirmed carcinogen.
- Genetic: Mainly due to genetic mutations.
 - The Adenomatous Polyposis Coli (APC) is an autosomal dominant gene that is equally implicated in the pathogenesis of colonic cancer
 - E. cadherin gene, on the other hand, is implicated in relation to diffuse gastric cancer.
 - Mutation of p53 gene which acts as a cancer-protective gene has also been implicated. --Hereditary diffuse gastric cancer (HDGC) is autosomal dominant.
 - Mutation of H-ras oncogene and overexpression of c-erb2 gene have also been associated with gastric cancer.
 - There is an increased incidence of gastric cancer in families of patients with hereditary nonpolyposis colorectal cancer (HNPCC). There may be a common genetic link

As regards the socioeconomic stratification, distal gastric cancer is commoner in the lower socioeconomic class, occurs in the older age group and is associated with H. pylori infection. On the other hand, the proximal gastric cancer is commoner in the higher socioeconomic class,

occurs in the younger age group and there is no association with H pylori infection. There is a rising incidence of proximal gastric cancer in developed countries.

Pathology of gastric cancer

Location

- Pyloric (antral) region (about 50%): Associated with the lower socioeconomic class and the older age group
- Body (about 30%)
- Fundus and cardia (about 20%): Occurs in the upper socioeconomic class and in the younger age group

Macroscopically, there are various forms:

- Ulcerative (50%): Most common variety. The ulcer has rolled and everted edges and is indurated
- Protruding or proliferative (20%): Projects into the gastric lumen
- Colloid or mucoid type: Rare; the cells are filled with mucoid material
- Superficial spreading, Infiltrative or limitis plastica (30%): The cancer cells spare the mucosa but infiltrate into the mucosa and submucosa with associated proliferation of fibrous tissue. It is of two varieties: localised and generalised. Whereas the generalised variety gives the entire stomach a leather-water bottle appearance, the localised variety on the other hand, is commonly restricted to the pyloric antrum and predisposes to gastric outlet obstruction.

Lauren's classification of gastric carcinoma: Of two types

- Type 1: Intestinal gastric cancer. Sequel to intestinal metaplasia; ulcerative or polypoid
- Type 2: Diffuse gastric cancer. This is the limitis plastica variety described above

Microscopically, most are adenocarcinomas. Other cell types are squamous cell carcinoma, mixed type (adenosquamous carcinoma), and undifferentiated carcinoma.

Modes of spread: Whereas there is an early intra-abdominal spread, distant metastases occur late. Unlike breast cancer, nodal involvement does not imply systemic spread.

- Direct spread to adjacent structures such as the liver, pancreas, mesocolon
- Lymphatic spread: The stomach is well endowed with lymph nodes which constitute a major channel of tumour spread. Spread is both by permeation as well as embolisation. The disposition of the lymph nodes are as follows
 - Perigastric lymph nodes (lymph node stations 1 to 6)
 - Nodes around the main and arterial trunk (lymph node stations 7 to 11)
 - The terminal group (stations 12 to 18)
 - The left supraclavicular nodes may be affected (Virchow's node) and constitutes the Troisier's sign. Abdominal nodes are involved in 60-80% of cases.

- Spread through the lymphatics that run along the falciform ligament may result in the formation of subcutaneous nodules around the umbilicus (Sister Mary Joseph's nodule)
- Haematogenous spread through the bloodstream to distant structures such as the liver, lungs, and later to the bones and brain. The liver is involved in about 25-40% of cases.
- Transcoelomic spread to the peritoneal space (ascites), ovaries (Krukenberg's tumour) and pelvis (Blummer's shelf). The peritoneum is involved in 60-80% of cases.

Staging of carcinoma of the stomach (International Union against Cancer UICC)

T1 Tumour invades the lamina propria

T2 Tumour invades the muscularis or subserosa

T3 Tumour invades the serosa

T4 Tumour invades adjacent organs

N0 No lymph nodes

N1 Metastasis in 1-6 regional nodes (nodes within 3cm)

N2 Metastasis in 7-15 regional nodes (nodes more than 3cm)

N3 Metastasis in more than 15 regional nodes (distant nodes)

M0 No distant metastasis

M1 Distant metastases: This includes the peritoneum and distal lymph nodes

From the above, it is obvious that staging can only be complete after surgery.

Clinical features: This depends on the stage of the lesion at presentation. Diagnosis is usually late owing to the fact that early gastric cancer presents mainly with non-specific dyspeptic symptoms. Patient initially may present with

- Mild epigastric discomfort and indigestion.
- Weakness due to anaemia resulting from chronic occult loss of blood
- Loss of weight

Later, patient may present with symptoms of late disease

- Epigastric mass
- Features of gastric outlet obstruction in antral lesions: vomiting, dehydration
- Dysphagia in lesions located around the oesophago-gastric junction
- Jaundice if there is spread to the liver
- Abdominal distension due to ascites
- Palpable umbilical swelling: Sister Mary Joseph's nodule
- Cough and difficulty with respiration if there is spread to the chest
- Symptoms attributable to migrating thrombophlebitis (Trousseau's sign)

On the other hand, there may be a paucity of signs in the early stage. Examination may reveal an ill-looking patient who may be pale and jaundiced in the metastatic stage. There may be an

umbilical nodule and patient may show clinical evidence of dehydration and even shock when there is associated gastric outlet obstruction. Other signs include

- Left supraclavicular lymphadenopathy (Virchow's nodes)
- Epigastric mass and tenderness
- Hepatomegaly and clinical signs of ascites (fluid thrill and shifting dullness)
- Pelvic mass in case of metastatic spread to the ovaries (Krukenberg's tumour)
- Rectal examination may show evidence of melaena stool and Blummer's shelf (solid peritoneal deposits anterior to the rectum)
- Clinical evidence of gastric outlet obstruction: Visible peristalsis and succussion splash
- Chest examination may reveal evidence of pulmonary spread or pleural effusion.
- **DIAGNOSIS ENTAILS A HIGH INDEX OF SUSPICION.**

Investigations include both specific and non-specific tests:

- Endoscopy (oesophagogastroduodenoscopy): In addition to visualising the lesion, multiple biopsies (up to 10) can also be taken
- Endoscopic luminal ultrasound scan: Determines the extent of the lesion in relation to the depth. Can also detect secondaries to the liver and lymph nodes.
- Contrast enhanced CT scan: Detects intra-abdominal metastatic disease in peritoneum, liver and pelvis
- MRI: as accurate as endoluminal USS
- Positron Emission Tomography (PET) scan: Detects occult secondaries. May be useful in the evaluation of response to neoadjuvant chemotherapy
- Barium meal and follow through: Quite useful in *linitis plastica* where it demonstrates the very narrow gastric lumen. It may also demonstrate the distended stomach associated with gastric outlet obstruction.
- Abdominal ultrasound scan: Detects ascites, lymph node and hepatic involvement
- Chest X-rays to rule out pulmonary secondaries and pleural effusion
- Diagnostic laparoscopy to detect hepatic and peritoneal secondaries
- Tumour markers: Carcino-Embryonic Antigen (CEA), and Carbohydrate Antigen (CA especially CA 72-4). Levels correlate with extent of metastases especially to the liver and also correlate with cure rate.
- Others: Full blood count, electrolytes/urea/creatinine, liver function tests and stool (occult blood)

TREATMENT

The major difficulty in management of gastric cancer lies with late diagnosis. Treatment entails various modalities and depends on the stage at presentation. It is therefore best addressed by a multidisciplinary team. Decision as regards the modality of treatment of an index patient will depend on the following factors

- Stage of disease
- Surgical fitness of the patient
- Patient's preference
- Patient's comorbidities

Preoperative preparation

- Rehydrate with intravenous infusion and correct any detected electrolyte anomaly, particularly when dealing with gastric outlet obstruction
- Nasogastric aspiration and lavage in case of gastric outlet obstruction
- Transfuse with blood in order to build up the packed cell volume if found to be low
- Adequate counselling in order to obtain an informed consent

Surgical treatment: The aim of surgical treatment is to excise the entire tumour with a margin of normal tissue. It equally entails associated lymphadenectomy. The type and extent of surgery is dependent on the location and extent of the disease.

- Proximal oesophagogastric tumours: Total gastrectomy with a Roux-en-Y oesophagojejunal anastomosis. Some surgeons will offer the patient a proximal gastrectomy with an oesophagogastric anastomosis
- Tumours around the body of the stomach: total gastrectomy with a Roux-en Y oesophagojejunal anastomosis
- Antral tumours will require a distal subtotal gastrectomy (Billroth 2 procedure)
- Linitis plastica: Total gastrectomy with oesophago-gastric anastomosis

The following are therefore the indications for total gastrectomy in gastric cancer

- Proximal gastric cancer involving the cardia
- Tumours involving the body of the stomach
- Diffuse gastric cancer

Three forms of resection have been described with respect to the resection margins

- R0 resection: Adequate resection with both macroscopic and microscopic free margins
- R1 resection: Macroscopic but not microscopic free margins
- R2 resection: No free margins detected (both macroscopic and microscopic)

Endoscopic Mucosal Resection (EMR): This was pioneered in Japan. The tumour and the underlying mucosa are excised from the wall of the stomach using an electric wire loop through an endoscope.

Endoscopic Submucosal Dissection (ESD): Resects a large area of submucosa in one piece. If microscopy reveals incomplete resection, or deep invasion, then formal gastrectomy is carried out.

Currently, surgery is curative in less than 40% of cases.

It is a common experience to open up these patients and discover an advanced, inoperable tumour. The most common palliative surgery carried out is an anterior gastrojejunostomy. Others are Devine's exclusive procedure in which the transected distal tumour is left behind and the system reconstituted by way of gastrojejunostomy. Endoscopic stenting has also been applied in a bid to reopen the channel.

Adjuvant therapy

Chemotherapy: It must be stated that gastric cancer does not particularly respond to chemotherapy. The latter may, however, help in achieving the following: decreasing tumour size, relieving symptoms and increasing the survival time. It may be employed either as an adjuvant or neoadjuvant system and involves combination chemotherapy. The drugs that have been used include intravenous 5-fluorouracil, mitomycin C, and adriamycin. The FAM regimen is a combination of the three drugs. Other drugs are epirubicin, leucovorine, oxaliplatin, cisplatin and docetaxel. Oral capecitabine (oral 5-FU) has also been found

Biological therapy: using a combination of Trastuzumab (Herceptin), capecitabine and cisplatin may be useful in HER₂ overexpressed metastatic disease. It has been approved by the US and European regulatory authorities. The epidermal growth factor receptor inhibitor (EGFR), Cetuzinab may also be used either alone or in combination with 5-fluorouracil. These drugs, if given preoperatively in a bid to render a relatively inoperable tumour operable, constitute the neoadjuvant therapy. Intraperitoneal mitomycin-C impregnated with charcoal has been tried in Japan in a bid to tackle sites of local recurrence.

Radiotherapy: Plays a limited role in the management of gastric cancer due to the presence of radiosensitive structures around the gastric bed. Chemotherapy may be combined with radiotherapy as adjuvant therapy. In combination with chemotherapy (chemoradiation) it has been employed in the management of inoperable gastric cancer. It relieves pain by shrinking the tumour and so may be useful for palliation in inoperable cases. Radiotherapy may also be useful in the management of painful bone secondaries.

Multimodal therapy has been found to improve survival

Summary of the general treatment recommendations for gastric cancer:

Stages 0 to 1A: This is early gastric cancer. Patient will benefit from either endoscopic mucosal resection or complete surgical resection. The latter offers potential for long-term survival

Stages 1B to 111C: These tumours are potentially resectable. Choice of treatment, however, depends on the medical fitness of the patient

- Medically fit patients: Perioperative neoadjuvant chemotherapy or chemoradiotherapy followed by surgery. Postoperative chemoradiotherapy has been shown to have a clear survival benefit.
- Medically unfit patients: Chemotherapy or chemoradiotherapy

Stage IV: This connotes metastatic disease. Patients will benefit from chemotherapy

Chemotherapy regimens: Generally speaking, two-drug regimens are preferred to its triple counterpart. This is due to the lower toxicity of the former. Triple regimens are reserved for patients who are medically fit and have access to frequent follow-ups. The regimen will depend on the preferred period of administration as well as the extent of the disease: Neoadjuvant, perioperative or metastatic

Neoadjuvant chemotherapy regimens

- Paclitaxel + carboplatin
- Cisplatin based: Combine with either 5-FU or capecitabine
- Oxaliplatin + Leucovorine

Perioperative regimen

- Epirubicin based: Combine with either cisplatin or oxaliplatin

Metastatic gastric cancer

- Docetaxel-based: Combine with any of the following – cisplatin, leucovorin, oxaliplatin or carboplatin
- Epirubicin-based: Combine with either cisplatin or oxaliplatin
- Oxaliplatin-based: Combine with either capecitabine or leucovorin
- For metastatic adenocarcinomas that overexpress HER2-NEU, trastuzumab may be added to a combination of cisplatin and a fluoropyrimidine (5-FU or capecitabine). Another option is to administer a combination of ramicumbab and paclitaxel

Prognosis: The prognosis of gastric cancer is poor because most patients present late due to delayed symptomatology. There is also a high recurrence rate (40 to 80%) following surgical treatment

Prognostic factors include

- Degree of invasion of gastric wall
- Nodal status
- Extent of spread of tumour
- Duration of symptoms
- Degree of differentiation

Early diagnosis should be the ultimate aim. Routine screening as practised in Japan where the disease is quite common is the cornerstone of early diagnosis. Any patient above the age of forty who presents with dyspepsia of recent origin should be screened endoscopically. Ideally, no patient should be commenced on any specific anti-ulcer regimen without prior confirmation of the diagnosis by way of endoscopy.

GASTROINTESTINAL STROMAL TUMOURS (GIST)

These are rare connective tissue tumours originating in the muscularis mucosa. They account for 1% to 3% of all gastrointestinal tumours. About 40 to 60% originate in the stomach where they account for 2% of all gastric tumours. They constitute the most common mesenchymal (non-epithelial) tumour of the gastrointestinal system. They arise from the interstitial cells of Cajal (the pacemaker cells of the GIT) that regulate intestinal contraction and reside between the inner circular and the outer longitudinal layers of the muscularis propria. The primary problem is the mutation of the tyrosine kinase c-kit oncogene. They occur in the 50-70 age groups and the incidence is equal in both sexes.

GIST may occur either sporadically or familialy. The latter is associated with a germline KIT mutation in exon 11. It may occur in association with type 1 neurofibromatosis.

Pathology: GIST may occur in any part of the gastrointestinal tract from oesophagus to anus and may be benign or malignant in nature. Malignancy is highest with small intestinal GIST (40%) and lowest with the gastric variety (20%). It is characterised by an apparently normal-looking mucosa but may, however, have an eroded or ulcerated centre. It has the following distribution: stomach (50%), small intestine (25%), rectum (15%), and colon

(10%). The fundus is the most common site in the gastric variety. It is normally surrounded by a pseudocapsule. Spread of the malignant variety is haematogenous (liver, lungs and bone) and transperitoneal (resulting in ascites). Lymphatic spread is rare. The risk of tumour spread is directly related to tumour size and mitotic activity.

GIST may be associated with other pathologies. For example, Carney's triad consists of

- Functioning extradural paragangliomas
- Pulmonary chondromas
- Gastric GIST

Clinical features: These are similar to those of carcinoma of the stomach. Gastric GIST may be asymptomatic and only discovered incidentally. This is very common with relatively small lesions. As GISTs grow, they may cause erosion/ulceration in the overlying mucosa resulting in bleeding. They may also compress adjacent structures resulting in abdominal pain. The commonest mode of presentation is upper gastrointestinal haemorrhage due to mucosal ulceration. The patient may present with clinical features of chronic blood loss (unexplained iron deficiency anaemia) or those of acute upper gastrointestinal haemorrhage (haematemesis and melaena). Compressive symptoms include early satiety, nausea, vomiting, abdominal pain and abdominal distension.

Investigations:

- Upper GI endoscopy,
- Contrast-enhanced CT scan
- Endoluminal ultrasonography
- FNAC and biopsy
- Endoluminal ultrasonography-assisted tissue biopsy

Treatment: Lesions greater than 2 cm are best managed by surgical resection with the adjacent bowel. Smaller lesions are amenable to endoscopic enucleation.

GIST is not amenable to chemotherapy. Gemcitabine and doxorubicin have been tried but with minimal therapeutic effect. Imatinib mesylate is a drug that inactivates tyrosine kinase c-kit resulting in a decrease in the phosphorylation of receptors with the consequent reduction in the proliferation of the tumour. Imatinib may be employed both in the adjuvant and neoadjuvant setting. It is also useful in the management of metastatic or unresectable tumours expressing c-kit mutation. A newer drug, Sunitinib, is employed in cases refractory to Imatinib. Sunitinib inhibits vascular endothelial growth factor amongst other tumour growth factors. Other tyrosine kinase inhibitors include Vatalanib and Dasatinib.

Prognostic factors: Related to the site and size of the tumour. The mitotic index also plays an important role in prognosis.

Leiomyomas: They were previously categorised as GISTs. It has, however, been found that unlike GISTs they are typically c-kit-negative tumours. Their patterns of growth are, however, similar. Both can grow inwardly and outwardly to form a dumbbell shape. Notwithstanding, whereas leiomyomas are more likely to grow intramurally, GISTs expand predominantly in an extramural fashion.

Other gastric polyps include

- Fundic gland polyp: More common in those on proton pump inhibitors (PPI)
- Hyperplastic polyps: Associated with gastritis.
- Adenomas (Raised intraepithelial neoplasia): Related to H.pylori infection
- Inflammatory fibroid polyps
- Gastric neuroendocrine polyps (carcinoids)

CHAPTER THIRTEEN

SMALL BOWEL TUMOURS

The small intestine stretches between the first part of the duodenum and the distal ileum, terminating at the ileal sphincter and is about 6 meters (20 feet) long. This is approximately 75% of the entire length of the gastrointestinal system. The primary function of the small intestine is to digest food and absorb nutrients. For this, it is endowed with a wide surface area that accounts for about 90% of the surface area of the entire gastrointestinal system. Despite these impressive statistics, neoplastic lesions of the small intestine are rare and account for only 2% of all gastrointestinal cancers. Secondary tumours are equally rare. Melanoma is the only malignancy that classically spreads to involve the small intestine. The following may contribute to the rarity of small bowel neoplasms

- * Liquid nature of contents which may be less irritating to the mucosa
- * Rapid transit time of contents: Reduces the exposure to carcinoma-inducing agents
- * Relatively low bacterial count
- * Others: Large lymphoid tissue, alkaline pH, presence of enzyme benzpyrene

The following is a table of small intestinal tumours in order of frequency

Benign	Malignant
Leiomyoma	Leiomyosarcoma
Adenoma (villous)	Adenocarcinoma
Lipoma	Lymphoma
Hamartoma (Peutz-Jeghers)	Carcinoid tumour
Haemangioma	
Neurogenic tumours	
Lymphangioma	

About 64% of small bowel tumours are malignant and approximately 40% of these are adenocarcinomas. The overall and locational frequencies are shown below (in percentage)

Tumour type	Overall	Duodenum	Jejunum	Ileum
Adenocarcinoma	40	40	40	20
Carcinoid	30	10	40	50
Lymphoma	20	10	10	80
Leiomyosarcoma	10	Rare	40	60

Epidemiology of small bowel cancers

Small bowel cancers are generally most common in western countries. In these countries, blacks are more affected than whites. There is, however, a predominance of lymphomas in less developed countries. It has an incidence rate of 1.5 per 100,000 and a mean age incidence of 60 years. The incidence tends to increase with age. Small bowel cancers are slightly commoner in males than females (1.4:1).

General clinical presentation: Malignant neoplasms, though statistically less common, are more likely to produce symptoms. The following is a table of comparison between the clinical presentation of benign and malignant small intestinal lesions

Parameter	Benign	Malignant
Weight loss	No	Yes
Abdominal pain	Yes	Yes
Obstruction	Yes	Yes
Gastrointestinal bleeding	Yes	Yes
Abdominal mass	Rare	Yes
Acute perforation	No	Yes
Obstructive jaundice	No	Periampullary
Malabsorption	No	Extensive lymphoma
Carcinoid syndrome	No	Yes
Asymptomatic	Yes	Rare

Abdominal pain and bleeding are the commonest symptoms. Abdominal pain is related to the associated subacute intestinal obstruction. The latter could be due to narrowing of the intestinal lumen, volvulus or intussusception.

Specific investigations for diagnosis of small bowel neoplasm

- Enteroclysis: An intestinal tube is passed through the stomach into the duodenum or jejunum. Dilute barium is slowly and steadily infused under fluoroscopy
- CT with oral contrast
- Upper GI endoscopy: Will detect duodenal neoplasms
- Colonoscopy: To rule out a colonic lesion as a cause of haemorrhage. Terminal ileum may be accessible for inspection
- Capsule enteroscopy: This involves the swallowing of a small camera which relays digitalised images to a computer recorder over an 8-hour period
- Mesenteric angiography
- Radioactive-labelled RBC scan
- Exploratory laparotomy: Diagnosis made at surgery in most cases

The following is the summary of the management peculiarities of the respective neoplastic lesions.

Benign tumours:

May present with complication(s) notably intussusception and haemorrhage. Peutz-Jegher's syndrome consists of intestinal hamartomatosis and perioral melanosis. It is an autosomal dominant disease. Benign lesions should be treated by primary resection and anastomosis.

Adenocarcinoma:

Commonest site of occurrence is the duodenum. This raises the question as to whether bile is carcinogenic. Adenocarcinoma may be associated with Crohn's disease, familial adenomatous polyposis (FAP) and adult celiac disease. It has also been associated genetically with K-ras mutation, and p53 overexpression. Diagnosis is by endoscopic examination and biopsy. Treatment is surgical

- Wide segmental small bowel resection including the mesenteric lymph nodes

- Right hemicolectomy and ileotransverse anastomosis: Involvement of the terminal ileum
- Side to side entero-enteric bypass: Advanced tumour
- Whipple's pancreaticoduodenectomy: Duodenal tumours

Carcinoid tumours:

They arise from the enterochromaffin cells of the GIT. The cells are pleuropotential, neuroendocrine in nature and form part of the amine precursor uptake and decarboxylation (APUD) system. They therefore synthesise vasoactive amines and regulatory peptides. The most common site of occurrence is the appendix.

Carcinoid syndrome: The clinical components are diarrhoea, flushing of the face and upper trunk and bronchospasm. This may be precipitated by foods, alcohol and emotional stress. In addition, there may be features of pellagra (dementia, dermatitis and diarrhoea). Right sided endocardial valvular fibrosis may result in tricuspid valve insufficiency which may culminate in right sided heart failure. The causative circulatory vasoactive amines include serotonin, bradykinin, prostaglandin and histamine. Others are dopamine, tachykinin and dopamine. Carcinoid syndrome is also associated with neoplastic lesions of the lungs, testes and ovaries. Symptoms are aggravated by the presence of hepatic metastases. Treatment of carcinoid syndrome is by the administration of octreotide (sandostatin)

Surgical management of carcinoid tumours: Depends on the site and size of the tumour

- Appendiceal carcinoid < 2 cm: Appendectomy
- Appendiceal carcinoid > 2 cm: Right hemicolectomy
- Small bowel carcinoid: Segmental resection with en-block mesenteric resection and primary anastomosis

Chemotherapy: Adriamycin, 5-fluorouracil and streptozotocin

Prognosis is good as it is a slow-growing tumour. Liver metastases, however, carries a gloomy outcome.

Gastrointestinal stromal tumours (GIST)

GIST is a mesenchymal neoplasm derived from the interstitial cells of Cajal in the gastrointestinal tract. Tyrosine kinase activity is encoded by the protogene, c-kit. The latter facilitates tyrosine kinase activity which results in uncontrolled cell proliferation (present in about 60% of GISTs). This appears to play a central role in tumorogenesis.

GISTs most commonly occur in the stomach; only 30% occur in the small bowel. It has been shown, however, that small intestinal GISTs are rather more aggressive than their gastric counterparts and have a worse prognosis. Metastases are to the liver, bone and lungs. GISTs are encapsulated, grow extraluminally and are usually subserosal.

Clinical presentation: Presents as an abdominal mass in 50% of cases. Commonly presents with complications – haemorrhage, perforation and intussusceptions.

Treatment: Adequate surgical resection and anastomosis. Advanced cases have been found to respond to the tyrosine kinase inhibitor, Imatinib (Glivec).

Prognosis depends on the stage of the disease and the histological grade (cellularity). Presence of distant metastases adversely affects the prognosis.

Lymphomas

They may either occur as primary gastrointestinal tumours or as part of a generalised lymphoma. In the primary form, it is limited to the area of the involved bowel without evidence of systemic disease (peripheral lymphadenopathy, splenomegaly, mediastinal involvement, and typical features in the peripheral blood film). The ileum is the most affected part of the small intestine (80%). The peak incidence is the 5th decade of life. It is more common in patients with Crohn's disease, celiac disease, and immunosuppressive conditions such as AIDS.

Clinical presentation: The early symptoms include nausea, bloating, and anorexia. Late symptoms include abdominal mass, loss of weight, malabsorption and clinical features of complications: haemorrhage, perforation, and intestinal obstruction. The diagnostic workup is as outlined above.

Treatment: Mainly surgical

- Resection in early tumours and debulking in late stages
- Surgical management of complications such as intestinal obstruction
- Chemotherapy in metastatic and/or recurrent tumours: Adriamycin-based
- Radiotherapy in advanced or recurrent tumours

Prevention of small bowel neoplasm: Identification and regular screening of the population at risk. This includes patients with Crohn's and celiac diseases. Also for screening are patients who manifest with clinical features of Peutz-Jegher's and Gardner's syndromes and the immunosuppressed patients such as those with AIDS.

CHAPTER FOURTEEN

COLORECTAL CARCINOMA

Colorectal cancer is the commonest cancer of the gastrointestinal tract and the second most common cause of cancer death in the United States. It is predominantly a disease of middle age and elderly population. Only about 5% of colorectal cancers occur in patients below the age of 40. The rectum is involved in 30-38% of cases.

Risk factors in the pathogenesis include:

* **Genetic:**

- Familial adenomatous polyposis (FAP)
 - Hereditary nonpolyposis colonic cancer (HNPCC) or Lynch syndrome. Transmitted as an autosomal dominant trait
 - Peutz Jegher's syndrome: Intestinal hamartomatosis affecting the whole small bowel and colon with associated melanosis of the oral mucous membrane and the lips
 - Positive family history (first degree relatives).
- About 70% of colorectal cancers develop from adenomatous polyps through a series of mutations in proto-oncogenes and tumour suppressor genes. Patients with FAP are equally at high risk for development of adenomas and adenocarcinomas of the duodenum. This calls for surveillance with upper GIT endoscopy in such patients.

* **Lifestyle:**

- Low fibre diet hampers evacuation of colonic contents thereby increasing the contact duration of the colonic mucosa with potential carcinogens.
- Ingestion of red meat and animal fat equally results in the formation of the carcinogenic nitrosamines.
- Fruits, vegetables and vitamins (A, C, and E) tend to reduce the risk.
- High calcium diet is preventive: Calcium combines with bile salt thereby reducing the bile salt concentration in the colon. Acting directly on the mucosal cells, it reduces their proliferative potential
- Alcohol and smoking are both regarded as risk factors.

* **Premalignant conditions:**

- Inflammatory bowel disease (ulcerative colitis, Crohn's disease). The risk in ulcerative colitis is about 10% at 20 years.
- Previous colonic surgery
- Cholecystectomy and ileal resection may also predispose to colonic cancer from the unregulated flooding of the colon with bile salts.
- Uretero-sigmoidostomy is considered a high risk factor.

* **Radiation:** Exposure to radiation is a risk factor in mucinous adenocarcinoma

Synchronous lesions are multiple and diagnosed at the same clinical sitting while metachronous lesions on the other hand, are diagnosed at separate occasions.

PATHOLOGY: May follow an adenoma-carcinoma sequence.

Macroscopically there are four types: annular, tubular, ulcerative and polypoid (proliferative or cauliflower).

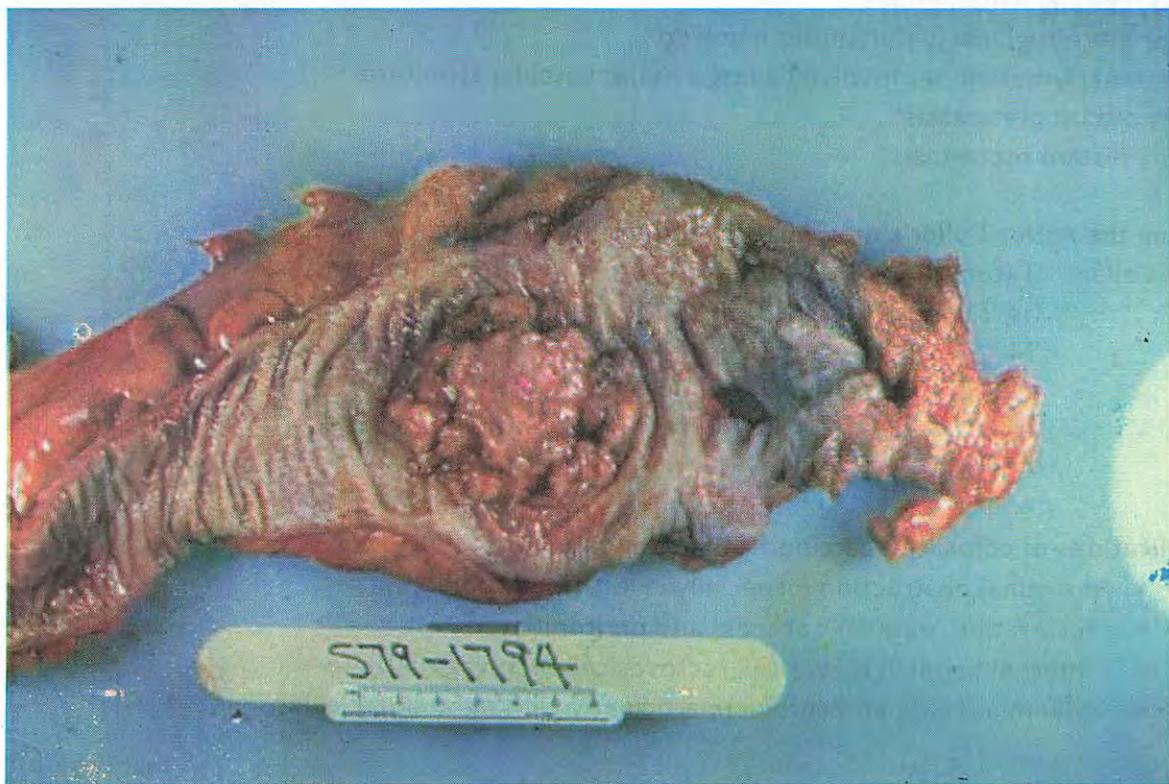
- The annular and tubular varieties spread around the inner wall, present with obstructive symptoms and are more common on the left side of the colon.
- The ulcerative and polypoid types are more common on the right side of the colon and may present with bleeding and anaemia

Microscopically, they are mainly adenocarcinoma (90%). Other less common histological types include mucinous adenocarcinoma (5%), squamous cell carcinoma, signet ring carcinoma, oat cell carcinoma and undifferentiated carcinoma. Duke graded them histologically into four grades

- Low (grade I): Well differentiated),
- Average (grade II): Moderately differentiated),
- High (grade III): Poorly differentiated),
- Anaplastic (grade IV): Undifferentiated

Spread of colonic cancer

- Direct spread: Circumferential and through the bowel wall to adjoining structures (bladder, ureter, ovaries, uterus and the psoas muscle).
- Haematogenous spread: Via the mesenteric and portal veins to the liver (33%), lungs (22%), bones (10%) adrenals (11%), kidneys and brain
- Lymphatic spread: Involves the peri-colic, epicolic and the para-aortic lymph nodes



POLYPOID CARCINOMA OF THE COLON.

- Lymphatic spread: Involves the peri-colic, epicolic and para-aortic lymph nodes.
- Trans-peritoneal spread: Results in ascites, Blummer's shelf and ovarian involvement.

Current staging is by the Astler-Coller's modification of the original Duke's staging (Duke, a pathologist in St Mark's Hospital, London originally described this in relation to rectal carcinoma in 1932. His original staging comprised of 3 stages: A, B and C).

A Invasion up to submucosa, no lymph node involvement

B1 Invasion up to muscularis propria; no lymph node involvement

B2 Invasion through wall; no lymph node involvement

C1: B1+ involvement of lymph nodes

C2: B2 + involvement of lymph nodes

D Distant metastasis

TNM (Tumour, Node, Metastasis) staging system

Tx: Primary tumour cannot be assessed

TO: No evidence of primary tumour

Tis: Carcinoma in situ

T1: Tumour invasion into submucosa

T2: Tumour invasion into muscularis propria

T3: Tumour invasion through muscularis propria

T4a: Tumour perforation of visceral peritoneum

T4b: Tumour invasion of adjacent structure

NX: Regional lymph nodes cannot be assessed

N1: 1-3 regional lymph nodes

N2: 4 or more regional lymph nodes involved

N3: Regional lymph nodes involved along a major vascular structure

M0: No distant metastasis

M1: Any distant metastasis

Relating the Astler-Coller's modification and the TNM staging

Astley-Coller	T (tumour)	N (nodes)	D (distant metastasis)
A	Tis, T1, T2, T3	N0	M0
B	T4	N0	M0
C1	T1, T2, T3	N1, N2, N3	M0
C2	T4	N1, N2, N3	M0
D	Any T	Any N	M1

Complications of colorectal carcinoma include

- Intestinal obstruction (especially in left-sided tumours)
- Perforation, paracolic abscess and peritonitis
- Internal fistula (colovesical, rectovesical, rectovaginal)
- Anaemia (acute and chronic) particularly with right-sided lesions.

CLINICAL PRESENTATION: Commonly presents in the adult age group (40 and above). Patients with genetic predisposition may present much earlier. Owing to the disparity in the calibre and contents of the various parts of the large intestine, clinical presentation tends to differ from one part to the other. Whereas the right side of the colon has a large calibre and the stool at that point is semisolid, the left side of the colon is of a smaller calibre and the stool is well formed.

- General presentation: Passage of blood and mucus in stool, change in bowel habit, abdominal mass (especially in caecal carcinoma), chronic anaemia (especially in right-sided lesions) and weight loss
- Right-sided lesions: Late diagnosis is quite common. They are mainly of the ulcerative variety and result in chronic blood loss. Presentation is most often with melaena in association with anaemia of unexplained origin. Patient may have presented earlier to the physician and/or haematologist and may have received multiple blood transfusions. The blood film may reveal evidence of hypochromic, microcytic anaemia. It follows that any male or postmenopausal female adult that presents with unexplained anaemia should be screened for right-sided colorectal carcinoma. Late presentation may be with a right iliac fossa mass, faecal fistula with associated weakness and loss of weight. Carcinoma of the caecum may present as acute appendicitis and may serve as the lead point of an intussusception (a variety of intestinal obstruction). Presence of ascites, hepatomegaly and lesions in the lungs and brain signify advanced disease with metastatic spread.
- Left sided lesions: Due to the high occurrence of the annular variety, presentation commonly involves a change in bowel habit (constipation alternating with spurious diarrhoea) with passage of mucoid, bloody stool. Patient may require increasing doses of purgatives to evacuate the bowel. Late presentation may be with an abdominal mass. Left sided colonic cancer may also be complicated by intestinal obstruction.
- Rectal and distal pelvic lesions: Present with haematochezia, tenesmus (feeling of incomplete bowel emptying), and a rectal mass. Some patients may present with haemorrhoids. The latter could be a red herring, particularly in the elderly. Such patients should be thoroughly screened for colorectal carcinoma.
- Emergency presentation: Results from complications such as acute-on-chronic intestinal obstruction, perforation, paracolic abscess, peritonitis and acute haemorrhage.
- Advanced cases may present with clinical evidence of spread to the liver, lungs and peritoneum (ascites).

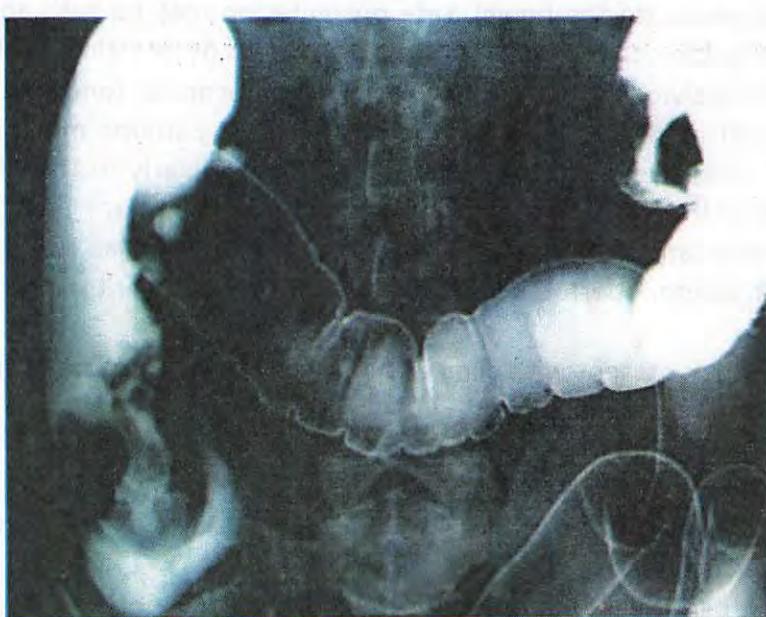
INVESTIGATIONS: Diagnostic investigations are

- Sigmoidoscopy, colonoscopy and biopsy. Colonoscopy is also useful in the detection of synchronous cancer and associated polyps.
- Double-contrast barium enema: This is useful in the absence of colonoscopy. It may demonstrate a stenosing lesion and an irregular filling defect due to an underlying malignancy. This is referred to as the apple-core sign (also known as napkin ring sign). This image appearance may also be seen in other colonic lesions such as chronic

ulcerative colitis, colonic amoebiasis and lymphoma with colonic involvement. The barium enema may also reveal associated polyps and synchronous lesions.

- Contrast-enhanced CT scan: has been found useful. It assesses the degree of direct invasion by the primary tumour. It is also useful in the detection of any intra-abdominal metastasis.
- Endorectal USS: Helps to determine the depth of a rectal malignancy.
- Abdomino-pelvic USS
- Positron Emission Tomography (PET) scan: Detects distant metastases
- Chest X-ray: For the detection of pulmonary metastasis
- Virtual colonoscopy (also called CT colonography or CT pneumocolon): This is a relatively new imaging procedure which uses CT scan to produce two- or three-dimensional images of the entire colon and rectum (from the rectum to the caecum). It is employed in the diagnosis of colorectal diseases including polyps, diverticulosis and cancer. It may also be performed via MRI. It is possible to provide 3D endothelial views of the large intestine by the use of virtual endoscopy. Conventional endoscopy is, however, required for further management of any detected lesion. The following are the advantages of virtual colonoscopy
 - More comfortable than conventional endoscopy since no tube is passed into the colorectum
 - Early return to work as the procedure is shorter and does not require sedation
 - Gives clearer images than the conventional barium enema
 - Examines the whole colon: The caecum is not examined in about 10% of patients undergoing conventional colonoscopy

Disadvantages of virtual colonoscopy

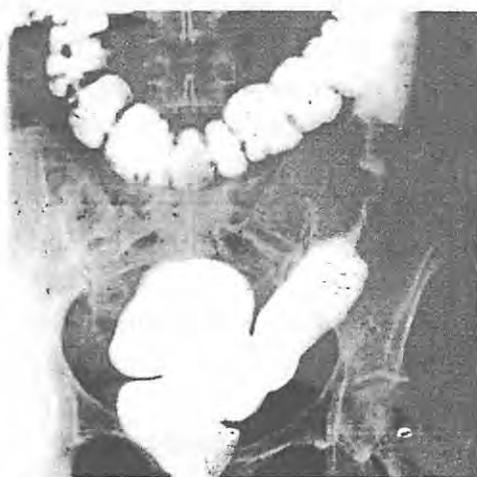


BARIUM ENEMA SHOWING COLORECTAL CARCINOMA

- Inability to collect tissue samples. These will ultimately require conventional colonoscopy
- Patient is exposed to the adverse effects of ionising radiation. Magnetic Resonance Colonography (MRC) is as effective as virtual colonography but does not expose the patient to the hazards associated with ionising radiation. MRC is highly specific but is less sensitive than conventional colonoscopy

Blood investigations include

- Full blood count: In preparation for surgery and/or chemotherapy. There may be an elevation of the ESR.
- Liver function test: For the detection of derangement in liver function due to metastatic deposits.
- Carcino-Embryonic Antigen (CEA). This is a tumour marker and its estimation at this stage is to establish a baseline for the detection of recurrence after surgery. The CEA level is elevated in patients with colorectal cancer that has penetrated the wall of the gut. The level normalises after surgical resection provided there is no metasatasis. A rising level after surgery is in keeping with recurrent tumour even at the occult stage. CEA is not necessarily specific for colorectal cancer as it may be elevated in some other clinical conditions. These include cancers involving the lung, breast, stomach and pancreas. It may also be elevated in benign conditions such as ulcerative colitis, cirrhosis of the liver, renal failure and pancreatitis. It may also be



APPLE-CORE DEFORMITY OF COLORECTAL CARCINOMA
(also referred to as the napkin-ring sign)

elevated in smokers. From the above, it can be surmised that serum CEA estimation is a non-specific test

- Electrocardiography (ECG): This is mandatory before surgery in patients above 40
- Others: Electrolytes/ urea/ creatinine, urinalysis and fasting blood sugar

TREATMENT: Surgery is the mainstay of treatment. This is preceded by adequate bowel preparation (mechanical and chemical). The ultimate goal of bowel preparation is to maximally

reduce the bowel content and achieve the least possible bacterial count in the colorectum. This reduces the incidence of surgical site infection as well as the rate of anastomotic breakdown in the event of bacterial spillage during surgery. The mechanical component is ideally carried out by the ingestion of a large volume of polyethylene glycol. Metoclopramide may be administered before the administration of polyethylene glycol in order to reduce the nausea associated with the latter. In the alternative, patient is commenced on low residue diet coupled with daily administration of soap and water enema about three days before surgery. A high rectal washout is carried out on the morning of surgery. Prophylactic administration of broad spectrum antibiotics (including metronidazole) has replaced the oral administration of nonabsorbable antibiotics as the chemical form of bowel preparation. It is administered at the induction of anaesthesia and continued for 24 hours. In the event of Intraoperative contamination, however, the intravenous antibiotic regimen is continued for 3 to 5 days. Other methods of bowel preparation include the administration of oral solution of picosulphate (Picolax) on the day prior to surgery and whole gut irrigation with 8 to 12 litres of normal saline through a Ryle's tube. The latter method is contraindicated in the elderly and patients with cardiac disease.

Surgical principle of the management of colorectal carcinoma: This involves excision of the tumour with appreciable tumour-free margin (at least 5cm proximal and 2 cm distal) together with the associated regional lymphatic drainage. Same principle applies in the case of rectal carcinoma. Radical excision of the rectum together with the mesorectum and associated lymph nodes should be the aim. Even in advanced cases, excision is regarded as the best form of palliation. The extent of resection depends on the site of the lesion.

- Right-sided colonic lesions (caecum, ascending colon and hepatic flexure): Right hemicolectomy (from the terminal ileum up to the proximal one-third of the transverse colon), Hepatic flexure tumours require an extended right hemicolectomy (resection up to the mid-transverse colon). The gut continuity is reconstituted with an ileo-transverse anastomosis
- Transverse colonic lesions: Transverse colectomy (entire transverse colon and the sleeves of the adjoining colon on both flexures). Anastomosis is carried out between the ascending and descending colon
- Splenic flexure and descending colonic lesions: Left hemicolectomy (descending colon and distal two thirds of transverse colon). Reconstitution is by anastomosing the transverse and sigmoid colon.
- Pelvic colon: Pelvic colectomy with or without left hemicolectomy
- Rectal lesions: The aim as much as possible is to restore gastrointestinal continuity and continence by the preservation of the anal sphincter. Lesions in the upper part of the rectum are treated with anterior resection. Previously, all lower rectal lesions were managed by way of abdomino-perineal resection with a permanent colostomy. With the advent of the stapling machine, however, "upper-lower" lesions can now be managed by low anterior resection while "lower-lower" lesions must, of necessity, be managed by abdomino-perineal resection (resection of the rectum and anal canal including the anal sphincter complex) with a permanent colostomy

- Palliative treatment for patients unfit for surgery includes transanal excision, laser destruction, and interstitial radiation.

Turnbull's no-touch technique of colonic resection: In a bid to prevent haematogenous spread of tumour during the handling of the lesion at surgery, the first step in this technique involves ligation of the blood vessels draining the site of the tumour. This early ligation is said to reduce the chance of dissemination of colorectal cancer.

Liver metastasis: About 20% of patients will have liver metastasis on presentation. This, however, does not preclude surgery. Management is by resection of liver metastasis with at least 1cm margin followed by chemotherapy.

Laparoscopic surgery in colonic carcinoma: Laparoscopic resection of colonic carcinoma requires skill in advanced laparoscopy. The principles are same as for open surgery. The unique advantages include safety, reduced pain and early return of bowel functions. These culminate in a shorter hospital stay when compared with the open technique. In the hand-assisted laparoscopic technique, after an initial laparoscopic dissection, the relevant anastomosis is hand-assisted and carried out manually outside the peritoneal cavity.

Adjuvant therapy: 5FU, Leucovorine (folinic acid) and Levamisole have been in use. Others are Oxaliplatin and Irinotecan. The current standard regimen is the FOLFOX4: combination of 5-FU (fluorouracil), leucovorin and oxaliplatin. Chemotherapy may be indicated as the sole treatment in patients with relatively asymptomatic stage IV disease. Combination therapy of capecitabine and oxaliplatin may also be used

Biological therapy: This involves the use of monoclonal antibodies. Bevacizumab (Avastin) is a monoclonal antibody which binds and neutralises the vascular endothelial growth factor (VEGF). There is an improvement in the survival rate when it is combined with the FOLFOX4 regimen. About 60% to 80% of patients with colorectal carcinoma over-express for the endothelial growth factor receptor (EGFR). Cetuximab is a monoclonal antibody that binds and inhibits EGFR. It is known to be effective in tumours that do not have K-RAS gene mutation. This necessitates testing for this gene prior to treatment with cetuximab.

Complicated cases such as those presenting with intestinal obstruction and perforation will, of necessity, require the appropriate surgical management after resuscitation. The aim of the emergency management is to relieve the obstruction. This is achieved by resection and the use of a temporary colostomy. In very ill patients, however, colostomy alone may suffice to pull through the emergency situation. Definitive surgery is later carried out when the general condition is much improved and the bowel adequately prepared.

Palliative surgical procedures include Hartman's procedure, fulguration and colostomy. Preoperative radiation therapy with 5FU as a radiosensitiser may also be useful in advanced cases.

Prognostic factors include

- Stage at presentation,
- Biological characteristics of the tumour
- Duration of symptoms (worse with short history)
- Age at presentation (worse in the younger age group)
- Presence of complications

Follow-up is by serial monitoring of the CEA level, faecal occult blood testing and colonoscopy.

Screening for colorectal cancer: This is important for early detection. Screening for those with a negative family history is by way of annual digital rectal examination and faecal occult blood test starting at the age of 40 and colonoscopy at the age of 50. Screening for those with a positive family history commences earlier than 40 and entails a more aggressive approach. They should, in addition to the other basic screening methods, have both colonoscopy and double-contrast barium enema at 40. Thereafter such patients with a positive family history should be made to undergo a three-yearly colonoscopy

Patients with familial adenomatous polyposis (FAP): Recommendations are:

- Screening colonoscopy at the age of 10 to 12 and performed yearly until 40; thereafter 3-yearly
- Initial upper gastrointestinal endoscopy at the age of 20 for early detection of duodenal disease
- Surveillance of all first degree relatives of FAP patients with upper GI endoscopy and 3-yearly colonoscopy

CHAPTER FIFTEEN

LOWER GASTROINTESTINAL HAEMORRHAGE

By definition, lower gastrointestinal haemorrhage is bleeding from the gastrointestinal tract that occurs distal to the ligament of Treitz

Aetiology:

Common causes of lower GI bleeding are

- Diverticulosis
- Colonic polyps and neoplasm
- Inflammatory bowel disease: Ulcerative colitis and Crohn's disease
- Angiodysplasia: An acquired degenerative condition in which the submucosal blood vessels become small, thin-walled and dilated. It occurs most commonly in the caecum and ascending colon. About 50% of patients have associated heart disease.
- Meckel's diverticulum
- Ischemic bowel disease: Mesenteric ischemia
- Small bowel tumours
- Intussusception
- Ruptured aortic aneurysm
- Amoebic dysentery and colitis
- Haemorrhoids
- Anal fissure

Anatomical classification of causes of lower GI bleeding

- Small intestine: Meckel's diverticulum, polyps, mesenteric ischemia and intussusceptions. Others are small bowel tumours, inflammatory bowel disease (Crohn's) and amoebiasis
- Colon: Carcinoma, polyps, angiodysplasia, and diverticulosis. Others are mesenteric ischemia and inflammatory bowel disease (mainly ulcerative colitis)
- Anorectal: haemorrhoids, fissure in ano, and carcinoma

In addition, it is important to point out that lower intestinal haemorrhage may be part of generalised bleeding disorder due to haematological conditions such as thrombocytopenia or disseminated intravascular coagulation.

Clinical features:

Bleeding may be acute or chronic. Whereas the former may present as an acute emergency with features of hypovolemic shock, the latter will present with features of chronic anaemia. Presentation is with passage of either maroon-coloured stool or frank haematochezia. Passage of melaena stool is usually associated with upper gastrointestinal or small intestine haemorrhage. It is, however, important to point out that severe upper gastrointestinal bleeding may equally present with passage of frank blood in stool. On the other hand, melaena may also occur with

bleeding from the ascending colon. Passage of formed stool streaked with fresh blood or the passage of fresh blood at the end of normal defaecation exercise is in keeping with an anorectal condition. A comprehensive history and clinical examination is therefore necessary in order to determine the aetiology of lower gastrointestinal haemorrhage. A digital rectal examination is invaluable as it may not only reveal the causative lesion, but may also give an idea of the likely source of bleeding based on the nature of the stool.

Studies have shown that the causative factor of acute lower GI bleeding may be age-related

- Children and adolescents: Meckel's diverticulum, intussusception, polyps and inflammatory bowel disease
- Adults (20 to 60): Diverticulosis, inflammatory bowel disease and neoplasm
- Older adults (above 60): Diverticulosis, neoplasia and angiodyplasia

Most causes of lower GI bleeding do not have associated abdominal pain. Presence of abdominal pain will raise the possibility of the following conditions: Inflammatory bowel disease, ischemic bowel, intussusception and ruptured aneurysm.

Treatment of acute lower gastrointestinal haemorrhage

As usual, the first priority is resuscitation. This is followed by efforts aimed at localising the site of the bleeding.

- Commence on nil orally
- Set up an intravenous line and commence volume resuscitation with normal saline or Ringer's lactate infusion
- Take blood samples for baseline investigations: Full blood count, electrolytes/urea / creatinine, grouping and crossmatching of blood. Coagulation profile is necessary in order to rule out a haematological aetiological factor
- Pass a urethral catheter and monitor the urinary output
- Pass a nasogastric tube: The sole aim is to rule out upper gastrointestinal bleeding. Aspiration of non-bloody gastric contents rules out an upper GI pathology as the cause of the bleeding. Gastric lavage should equally be carried out in order to confirm that the index bleeding is not of upper GI origin.

Following cessation of bleeding, the following investigations will aid in the localisation of the source of bleeding. It is important to note that this is most important prior to any surgical intervention.

- Rigid proctosigmoidoscopy: This is a bedside procedure employed in the visualisation of the most distal 25 cm segment of the lower gastrointestinal tract
- Diagnostic colonoscopy: it is quite informative. The patient should, however, be haemodynamically stable. Apart from its diagnostic function, colonoscopy also has a therapeutic advantage since identified bleeding vessels may be tackled by either coagulation or injection with adrenaline. This is important particularly when dealing with angiodyplasia
- Contrast enhanced CT scan: Demonstrates intravenous pooling of contrast at the site of gastrointestinal bleeding

- Mesenteric angiography: Selective angiography of the superior and inferior mesenteric vessels will localise bleeding from the midgut and hindgut. Active bleeding may be controlled by intravenous vasopressin or the application of gelfoam. Angiography is not applicable when the bleeding has stopped as a bleeding rate of at least 0.5 to 1.0 ml/min is required for accurate visualisation of the bleeding vessel. The main advantage of angiography over colonoscopy is that it is feasible in haemodynamically unstable patients.
- Oesophagogastroduodenoscopy: May be required to rule out a duodenal source of haemorrhage. As stated above, severe upper GI bleeding may present with frank haematochezia.
- Tagged red blood cell scan: This utilises red blood cells tagged with technetium 99m. It is highly sensitive and can locate even very minimal bleeding at a rate of 0.1 ml/minute. May also be used as an initial screening test.
- Small bowel enema: May detect lesions involving the small intestine
- Abdominal ultrasound scan
- Barium enema: May be useful after cessation of acute bleeding particularly when there are no facilities for colonoscopy
- Video capsule endoscopy: Visualises the entire GIT but has no therapeutic advantage
- Faecal occult blood test: Relevant in patients that present with chronic anaemia

Definitive treatment: This addresses the underlying cause of bleeding. It must be reiterated that blind surgical intervention is not advised in the management of lower gastrointestinal bleeding and should rather be preceded by location of the site of bleeding.

The following treatment may be applicable

- Endoscopic coagulation and injection of vasopressin to secure haemostasis
- Endoscopic polypectomy for colonic polyps
- Resection of colonic neoplasm: Extent of surgery depends on the site of the lesion. For instance a right colonic tumour will require right hemicolectomy.
- Small bowel mesenteric ischaemia: May require massive resection of the small bowel.
- Haematological consultation for bleeding diathesis.

CHAPTER SIXTEEN

HAEMORRHOIDS

Haemorrhoid (pile) is a common clinical condition. It is basically due to the abnormal sliding down of the anal cushions with associated weakness of the supporting structures. The resultant loss of elasticity of the cushions could hinder their natural spontaneous retraction after defaecation. Straining is the most common cause. The end-result is engorgement of the associated vessels.

Classification of haemorrhoids:

Classically divided by its site of origin into two main types – internal and external

- Internal: Arises above the dentate line and is characteristically located at the 3, 7, and 11 O'clock positions. This is in keeping with the disposition of the superior haemorrhoidal vessels (right anterior, right posterior and left lateral).
- External: Arises below the dentate line and involves the inferior haemorrhoidal vessels
- Interno-external: Regarded as the external extension of the internal haemorrhoids. It involves both plexuses.

Haemorrhoids have also been classified into primary and secondary depending on the location. Primary haemorrhoids are those located at the classical sites of 3, 7, and 11 o'clock while secondary haemorrhoids occur in-between the primary sites.

Classification by degrees:

This takes the clinical stage into consideration

- First degree haemorrhoid: Bleeding only; no prolapse
- Second degree : Prolapse on defaecation but reduces spontaneously
- Third degree: Prolapse on defaecation or any form of straining and requires active, manual reduction
- Fourth degree haemorrhoid: Permanent prolapse that cannot be reduced.

Aetiology of primary haemorrhoids:

- Man's erect posture may be a predisposing factor coupled with lack of valves in the portal system. Ageing may result in weakening of the relevant structures.
- Diet and stool consistency: Hard faecal mass resulting from constipation will result in obstruction of the venous return and engorgement at straining. Straining due to diarrhoea will have a similar effect.
- Other causes include the gravid uterus, pelvic masses (fibroids, uterine and cervical cancers, ovarian neoplasm, and bladder cancer), ascites, colorectal carcinoma and portal hypertension.

Clinical features: Initially it presents with bright red, painless bleeding. Later, there is anal prolapse that may be associated with mucus discharge; itching and mild pain/discomfort. Clinical examination may reveal evidence of anaemia. Haemorrhoids constitute one of the causes of unexplained anaemia in an adult. The abdomen is examined for presence of an abdominal mass. Rectal examination will reveal prolapse in the case of a third degree haemorrhoid. The earlier stages may be visualised by proctoscopy.

Complications of haemorrhoids: These include haemorrhage (acute and chronic), strangulation, ulceration, and fibrosis. Others are gangrene, and pylephlebitis (portal pyaemia).

Management of haemorrhoids

It is important to rule out any underlying lesion by way of thorough clinical examination and appropriate investigations. This is most important when dealing with the older age group.

Investigations: Full blood count, colonoscopy and barium enema.

General treatment: Avoid constipation by taking high fibre diet and increasing fluid intake. In addition

- **1st degree:** Injection therapy with phenol in almond or vegetable oil. This is carried out via proctoscopy with the aid of special equipment such as Gabriel's syringe. The active ingredient is the phenol while the vegetable oil only acts as a vehicle that prolongs the action of the phenol. Injection is not made directly into the haemorrhoid but rather into the submucosal plane above the anorectal ring.
- **2nd degree:** Rubber band ligation (not more than 2 piles at a time), and cryosurgery (using nitrous oxide or liquid nitrogen). Infrared coagulation (heat) and laser have also been used.

Indications for haemorrhoidectomy

- 3rd and 4th degree haemorrhoids
- Failure of non-operative methods in 2nd degree haemorrhoids
- Others: Fibrosis, well-defined interno-external haemorrhoids
- When bleeding results in anaemia

Complications of haemorrhoidectomy

Early:

- **Reactionary haemorrhage.** Occurs within the first 24 hours of surgery. May be occult when the patient bleeds into the rectum without any external evidence. It may be due to loosening of ligatures. May also occur when the blood pressure starts rising during recovery from an initial postoperative hypotension. Patient may lapse into hypovolemic shock. Initial treatment is by resuscitation with intravenous infusion and blood transfusion when necessary. Do not hesitate to re-explore surgically if bleeding persists.
- **Pain:** Though a natural accompaniment of any surgical procedure, it deserves a special mention post-haemorrhoidectomy owing to its magnitude and the ensuing complications such as acute urinary retention. It is therefore important to give adequate analgesia post haemorrhoidectomy.

- **Acute urinary retention:** This is due to severe post-operative pain. Prevention by administering adequate analgesia after surgery is the key to management. It usually responds to conservative management. Refractory cases will require urethral catheterisation and relief of urinary retention. It is important to de-catheterise the patient after relieving the acute retention of urine. The only indication for leaving an indwelling catheter is recurrence of urinary retention. Even in the face of the latter, the urinary catheter should be removed after 24 hours in a bid to prevent urinary tract infection

Late:

- **Secondary haemorrhage:** Occurs between the 7th and 10th days of surgery. It is usually due to infection. Initial treatment consists of intravenous infusion/blood transfusion and intravenous antibiotics. Bleeding in this case is best controlled by instituting a tamponade-effect over the anorectal mucosa in view of the generalised oozing nature of the bleeding. This can be achieved by the insertion of a Foley urethral catheter with a large-capacity balloon into the anal canal. The tamponade effect of the inflated balloon over the anal mucosa will aid in controlling the bleeding. This is analogous to the situation after prostatectomy.
- **Anal stricture:** Results from fibrosis that follows excessive dissection. The latter leaves no mucocutaneous bridges for proper healing in-between the excised haemorrhoids. Treatment is by anal dilatation.
- **Anal incontinence:** Results from injury to the sphincter complex. Most commonly result from 'over enthusiastic' manual anal dilatation which is the preliminary step during haemorrhoidectomy. Pelvic exercises may be helpful in the management.
- **Submucous abscesses.**

External haemorrhoids

Aetiology: May be the external component of an internal pile, the sentinel pile of an anal fissure or just an anal skin tag

Treatment: Address the underlying cause, sitz bath, excision

Complications: Pruritus ani, perianal haematoma.

Perianal haematoma (Thrombosed external haemorrhoid):

This is a painful, perianal, subcutaneous swelling of sudden onset. It consists of clotted blood. Milligan aptly referred to it as "a 5-day painful, self-curing lesion". Treatment at the early stage consists of incision and evacuation of the clot under local anaesthesia. A conservative approach is adopted in early stages as resolution is spontaneous. The latter may be aided by regular sitz bath..

Prolapsed (strangulated) haemorrhoid: This happens when a prolapsed haemorrhoid is gripped at the anal margin by a tight anal sphincter. It becomes irreducible and oedematous due to venous congestion. There is associated pain. Treatment is by administration of analgesics, sitz bath, antibiotics to prevent portal pyaemia and anal dilatation to effect reduction of the prolapsed haemorrhoid. Surgery is best deferred until after recovery from the acute phase as emergency surgery may be complicated by portal pyaemia and bleeding owing to the friable nature of the tissues.

CHAPTER SEVENTY

ANAL SURGICAL CONDITIONS

MALIGNANT ANAL TUMOURS

Anal malignancy accounts for less than 2% of all large bowel tumours.

Classification of anal tumours: This is based on the location in relation to the dentate line.

- Below the dentate line: Usually squamous carcinoma
- Above the dentate line: Also referred to as basaloid, cloacogenic and transitional. They consist of adenocarcinoma, melanoma, lymphoma and sarcoma. The term, epidermoid carcinomas is occasionally used to distinguish them from those of non-squamous cell origin.

SQUAMOUS CELL CARCINOMA

This occurs around the anal margin and the anal canal.

Aetiology: Risk factors include

- Viral infections: Human papillo virus (HPV)
- Premalignant lesions: Anal intraepithelial neoplasia (AIN)
- Immunosuppression: HIV, organ transplantation (especially renal)

Clinical features of anal carcinoma: These are

- Anal pain
- Bleeding,
- Mass around the anus and pruritus
- Faecal incontinence when the sphincter is involved
- Ano-vaginal fistulation in females.
- Late presentation may present with constipation, features of intestinal obstruction and inguinal lymphadenopathy due to lymphatic spread.

Management:

Confirmation of diagnosis is by FNAC and biopsy.

Combined modality therapy (CMT) which involves chemoradiotherapy is the current treatment modality. 5-FU is used in combination with either cisplatin or mitomycin C.

Surgery in the early stage involves excision of small marginal tumours. Defunctioning colostomy may be employed as a palliative measure in late stages.

Radiotherapy may be the only option in the treatment of advanced growths

Other anal malignancies:

Adenocarcinoma: This is usually an extension of a rectal carcinoma. It may also arise from the anal glands in a pre-existing fistula-in-ano. Treatment is by abdominoperineal excision of the rectum (APER) and permanent colostomy.

Melanoma: Occurs in the transitional zone as a hyperpigmented anal mass. Prognosis is poor.

Perianal Paget's disease: Has a poor prognosis.

Anal intraepithelial neoplasia (AIN): Usually sequel to HPV, HIV and anorectal intercourse. It presents as a raised, white/pigmented, scaly lesion. Confirmation of diagnosis is by biopsy and treatment is by excision, and topical imiquimod

ANAL FISSURE

It is a common, painful clinical condition of the anal canal due to a tear of its mucosal lining. All age groups may be affected. It is, however, more common in the younger age group. Anal fissure is usually acute in presentation but is regarded as chronic if it lasts beyond 8 -12 weeks.

Aetiology: Constipation is the commonest predisposing factor. The anal tear is sustained while trying to evacuate hard stool. Thereafter, a vicious cycle is set up as the patient becomes afraid to stool owing to aggravation of the pain during the act of defaecation. The situation is, however, worsened as the longer the stool remains unevacuated, the harder it becomes and consequently the more difficult and painful it is to pass out. This is a vicious cycle. Other causes include repeated diarrhoea and childbirth. The induced spasm of the anal sphincters may reduce the blood flow to the region. This adversely affects healing of the fissure.

Clinical features: Anal fissure is usually preceded by severe constipation. In an attempt to evacuate, patient experiences a sharp anal pain that may be associated with haematochezia. There may be recurrent pain on defaecation, bleeding and itching. Any attempt at digital rectal examination is usually resisted as it is associated with excruciating pain. A 'sentinel pile' usually located around the 6 o'clock position may be obvious. It is referred to as sentinel pile as it appears to 'guard' the anal lesion.

Treatment: Initial conservative treatment includes management of constipation by high residue diet, increased fluid intake and regular sitz bath. The latter, in addition to soothing and promoting healing of the torn tissue also relaxes the sphincters. These physical measures may be enhanced by the application of Nitroglycerin cream and the gel form of Nitrofipine. Botulinum toxin (Botox) has also been found useful. It should be noted that a nonhealing anal fissure and one located anywhere other than the posterior area of the anus should raise the possibility of malignancy or Crohn's disease as differential diagnosis

Surgical treatment is employed when conservative measures fail. It involves lateral sphincterotomy. Advancement anal flaps have been employed in the management of chronic as well as pregnancy-induced fissures and those resulting from injury to the anal canal.

ANAL ABSCESES

The underlying pathology is obstruction of the crypts that drain the anal glands. This results in infection of the anal glands and formation of abscess. Delay in drainage or non-drainage of the abscess could result in fistulation into the anus or rectum (ano-rectal fistula). Anal abscesses are classified according to their location

- Perianal abscess
- Submucous abscess
- Ischiorectal abscess: Occurs in the pyramidal-shaped ischiorectal fossa and constitutes about 30% of anal abscesses. There is a posterior communication between the right and left fossae.

- Pelvirectal abscess: Situated above the levator ani muscle and is just a little short of being a pure pelvic abscess

Clinical features: Anal abscess presents with painful anal swelling. Patient is restless and unable to sit down due to pain. There is associated fever. Digital rectal examination reveals a tender, fluctuant perianal swelling or ulcer associated with fever and/or purulent discharge.

Treatment: This involves incision and drainage of the abscess under local anaesthesia if small or under sedation/ general anaesthesia if large. In particular, ischiorectal abscess requires adequate drainage. This is achieved by making a cruciate incision over the site of proposed drainage. It is subsequently 'deroofed' by excision of the triangular flaps. The abscess cavity is laid open for adequate drainage. A specimen of pus is sent for microscopy/culture/sensitivity. After drainage, the patient is commenced on a broad spectrum antibiotic regimen that includes metronidazole for anaerobic infection. The antibiotic regimen may need to be revised later in line with the culture result. The patient is commenced on regular sitz bath (twice daily and after defaecation). Follow-up aims at ensuring healing of the incision site and early detection of fistula-in-ano which is a well known complication of anal suppuration.

FISTULA – IN –ANO

By definition, a fistula is an abnormal communication between two epithelial surfaces lined by granulation tissue. Fistula-in-ano is therefore a fistulous communication between the perianal skin and the anus or rectum.

Aetiology of fistula-in-ano: This is multifactorial

- Commonest cause is a poorly-treated perianal abscess. If drainage of the abscess is delayed or not carried out at all, it will rupture into the anus thereby creating a fistulous track between the superficial abscess and the anus
- Chronic inflammatory conditions such as Crohn's disease and ulcerative colitis
- Tuberculosis
- Colorectal malignancy
- Lymphogranuloma venereum
- Hydradenitis suppurativa
- Medical conditions that predispose to suppuration such as diabetes mellitus and HIV

Classification of fistula-in-ano: There are two types of fistula-in-ano based on the relationship of the internal opening to the levator ani muscle

- Low fistula-in ano: The internal opening is below the levator ani muscle
- High fistula-in ano: The internal opening is above the levator ani muscle

It can also be classified on the basis of the relationship of the fistulous tract to the anal sphincteric muscles. The latter comprise of an inner internal sphincter and an outer external sphincter. The following is referred to as Park's classification of fistula-in-ano

- Intersphincteric: Fistula runs between both muscles
- Trans-sphincteric: Fistula runs across both muscles
- Suprasphincteric: Runs above the sphincter complex, originating at the dentate line
- Extrasphincteric: Above the sphincter complex but originating in the rectum.

Suprasphincteric and extrasphincteric varieties obviously constitute the high fistula-in-ano.

Diagnosis: This is aimed at establishing the presence of the fistula as well as identifying any predisposing factor(s). The history should enquire about the following

- History of intermittent seropurulent discharge from a perianal opening with soiling of the undergarments. Patient complains of 'hardening' of the surrounding skin. The discharge may temporarily remit with the administration of antibiotics only to relapse later
- Past history of clinical features suggestive of perianal abscess: perianal pain and swelling followed by discharge of purulent material around the anus. The purulent discharge is usually followed by relief of pain and attendant constitutional symptoms such as fever. This is later followed by intermittent perianal purulent discharge
- Rule out presence of any predisposing medical condition such as polydypsia and polyuria (diabetes mellitus); chronic cough (tuberculosis) and change in bowel habit (neoplasm)
- History of any treatment received prior to index consultation

Clinical examination may reveal a chronically ill-looking patient who may show evidence of weight loss. There may be associated pallor. A digital rectal examination is the ultimate in the clinical examination of a patient with fistula-in-ano

- Inspection will reveal the external opening of the fistula. This may be in the form of a reddish, fleshy dimple which currently may or may not be discharging. Occasionally, the external opening may undergo temporary healing and so may not be very obvious.
- Digital examination: There is induration of the skin surrounding the external opening. Effort is made to feel the internal opening of the fistula in the anal canal. It is usually indurated and may be visible in the low type on patting the anus. The Salmon-Goodsall's rule acts as a guide in the location of the internal opening of a fistula. It states that a fistula whose external opening is anterior to an imaginary transverse line drawn across the midline of the anus in the lithotomy position is most likely to run a straight course into the anus. On the other hand, fistulae that are located posterior to this line run a horse-shoe course and open in the posterior midline. An anterior fistula whose external opening is more than 3 cm from the anal margin may, however, run a long tortuous course and may end up opening in the posterior midline. This is therefore an exception to the Salmon-Goodsall's rule

Investigation: This is aimed at confirming the presence and nature of the fistula as well as elucidating any aetiological factor

- Fistulography: This involves injection of contrast through the external opening in order to define the track. It helps in categorising the fistula into high or low. It is not a routine investigation
- Proctosigmoidoscopy
- Colonoscopy: Screens the colon for any predisposing pathology particularly neoplasm
- Barium enema: It is an alternative when there are no facilities for colonoscopy

- Swab from the discharge for microscopy, culture and sensitivity
- Chest X-ray: Aims at ruling out tuberculosis and also as a preoperative investigation
- Mantoux test: For the diagnosis of an underlying tuberculosiss
- Fasting blood sugar: To rule out diabetes mellitus
- Retroviral screening for HIV
- Full blood count: As part of the preparation for surgery

Treatment of fistula-in-ano

This involves treatment of any known underlying cause such as control of blood sugar in diabetes mellitus and institution of relevant therapy for tuberculosis. Crohn's disease and malignancy should be managed appropriately.

Definitiye surgical treatment is preceeded by bowel preparation. The principle of surgical management is to identify and open up the tract with a view to removing all element of chronic inflammation while sparing the anal sphincteric mechanism. The nature and extent of surgical treatment is therefore guided by the type of fistula: high or low. Whereas one can safely dissect out a low anal fistula, same cannot be said of a high fistula. This is because of possible damage to the anal continence mechanism when dealing with a high fistula

Management of low fistula-in ano: A gentle anal stretch is carried out as a preliminary procedure. Digital rectal examination is carried out with the aim of feeling for the internal opening of the fistula. The latter is usually indurated on digital palpation. This is followed by the introduction of a well-lubricated malleable probe into the external opening. The probe is gently negotiated through the entire length of the fistulous tract and exits through the internal opening. The subsequent procedure consists of either of two forms

- Fistulotomy: Cutting down on the probe with a view to laying open the entire tract. This is followed by curetting of all elements of chronic inflammation located in the floor of the tract.
- Fistulectomy: Essentially same objective as fistulotomy but more radical in nature. This involves complete excision of the entire tract over the malleable probe.

It is ideal to send the specimen of the granulation tissue/tract for histology and microscopy, culture and sensitivity. The raw area is packed with gauze lubricated by a wrap of sofratulle. The latter is removed after 24 hours or whenever the patient wants to stool (whichever comes first). Thereafter, the patient is commenced on sitz bath (twice daily and whenever he stools). This is aimed at keeping the wound clean thereby enhancing the healing process. Patient should be reassured as healing is usually slow but sure. It is pertinent to point out that primary closure of the wound after fistulectomy is another option which is practised in some centres with success. The main advantage of this method is that it reduces the time of recovery. There is, however, a high incidence of recurrence if infection is not completely eradicated at the primary surgery. It is therefore only suitable in relatively mildly infected cases. It is prudent to cover the procedure of primary closure with appropriate antibiotics.

Management of high fistula-in-ano

Unlike the low variety, any attempt at formal fistulectomy/fistulotomy of a high fistula-in-ano may result in anal incontinence due to disruption of the anal sphincter control mechanism. Surgical treatment is by any of the following methods

- Seton stitch: After dissecting the external part of the fistula, a stout nonabsorbable suture made of silk or silicone (Seton stitch) is guided through the internal part of the fistula into the rectum. Both ends of the suture are brought together and knotted loosely around the remaining part of the sphincter. The suture is left in place for weeks to months until the drainage resolves and the fistula closes. Closure of the fistula may be facilitated by a preliminary temporary colostomy that rests the distal bowel and minimises contamination. Spontaneous healing of the fistula may follow removal of the Seton stitch. This procedure is meant to safeguard the sphincter mechanism. Despite these measures, anal incontinence still occurs in about 20% of cases.
- Temporary diverting colostomy: This completely rests the distal bowel, markedly reduces its contamination and thereby creates a favourable environment for healing of the fistula. It may be complemented by the insertion of a Seton stitch. Most fistulae will heal by this combined measure. Healing of the fistula is followed by closure of colostomy.

Prognosis: There is a recurrence rate of 30% to 40%. This may be attributed to the following factors

- Inadequate excision of infected granulation tissue thereby creating room for recurrent infection. Since surgery is being carried out on a contaminated area, the procedure is usually fixed at the tail-end of the operation. If carried out by a less-experienced resident doctor (surgeon may be tired!), the recurrence rate due to inadequate excision may be high
- Premature bridging of the anal skin may result in improper healing of the wound from below. This mitigates adequate wound drainage and encourages recurrent postoperative infection

CHAPTER EIGHTEEN

COLOSTOMY

Colostomy is a surgically-created communication between the colon and the skin of the abdominal wall for the sole purpose of drainage of stool and flatus. In plain terms, it is an artificial anus.

Indications for colostomy: May be temporary and permanent

Temporary colostomy: It is employed to tide over a critical situation in the course of management of a complex primary condition. Temporary colostomy is subsequently closed after the primary condition is fully addressed. Indications include

- Paediatric Surgery: Hirschsprung's disease, high imperforate anus with an anorectal fistula
- Colonic injuries: Immediate definitive management may not be possible in the presence of an unprepared bowel. Proximal colostomy diverts faeces in distal colonic trauma
- Proximal colostomy may be instituted in order to protect a distal anastomosis
- To divert faeces and enhance healing in benign rectovesical or rectovaginal fistula. It may also be employed in the management of a high fistula-in-ano
- To rest a severely inflamed distal gut in chronic conditions such as proctocolitis and lymphogranuloma venerum

Permanent colostomy: Low anorectal carcinoma and severe anorectal stricture are the indications for permanent colostomy. In this regard the institution of a permanent colostomy forms an integral part of an abdominoperineal resection.

Types of colostomy:

Colostomy is only feasible in a segment of large intestine that is endowed with mesentery. For this reason, the transverse colon and the sigmoid colon are the segments available for use as colostomy.

Transverse colostomy: This is the temporary colostomy of choice in the emergency relief of a distal colonic obstruction and also in the management of a distal colorectal injury. Apart from diverting the faecal stream, it also permits efficient cleansing and preparation of the obstructed colon proximal to the lesion. It is usually fashioned as a loop colostomy. In the latter situation, a loop of transverse colon is brought out through an upper transverse incision and stripped of omentum. The colon is opened and its edges are sutured to the adjacent skin margin. This relieves obstruction and rests the distal gut thereby facilitating the healing of a more distal injury. Closure of a temporary transverse colostomy for trauma should ideally be preceded by a contrast enema (distal loopogram) in order to ensure adequate healing of the lesion and to

rule out distal obstruction. When carried out for malignant colonic obstruction, it may be retained after the definitive surgery in order to protect the temporarily 'fragile' anastomosis. Closure may be carried out later. Currently, most surgeons close the colostomy simultaneously with the definitive surgery provided there is adequate preoperative bowel preparation.



PAUL-MICULICZ DOUBLE-BARREL COLOSTOMY

Double-barrel colostomy: In this case, there is complete division of the colon and both ends are sutured separately to the skin. The more distal opening is regarded as a mucus fistula. Double-barrel colostomy ensures complete defunctioning of the colon.

Hartman's procedure: This is usually carried out as an emergency procedure and involves complete transection of the colon (usually the pelvic colon). The distal part is closed and 'dropped' in the pelvis while the proximal stump is brought out as a temporary colostomy. When the condition of the patient improves, the gut is reconstituted after dealing with the primary condition. It is sometimes difficult to identify the distal stump during surgery for closure of colostomy. This may be facilitated by tagging the distal stump to the anterior parietal peritoneum with non-absorbable sutures in the course of the primary procedure. The alternative is to pass a urethral catheter with a large capacity balloon into the rectum during the restorative surgery. The inflated balloon will ease the detection of the distal stump at surgery.

Permanent colostomy: This is carried out as an integral part of abdomino-perineal excision of the rectum for low rectal malignancy. Ideally patient should receive counselling from an expert stoma nurse. The site of the proposed colostomy should be decided and discussed with the patient preoperatively. It should not be sited near a bony prominence, specifically the anterior superior iliac spine. An informed consent is obtained prior to surgery. During surgery, it is important to close the lateral space between intraperitoneal sigmoid colon and the peritoneum of the pelvic floor. This prevents internal herniation through the defect.

Complications of colostomy

- Prolapse
- Retraction and stenosis
- Strangulation around the paracolic gutter
- Excoriation of the surrounding skin
- Paracolostomy herniation
- Diarrhoea



DOUBLE-BARREL COLOSTOMY

- Faecal impaction
- Psychological: Depression

CHAPTER NINETEEN

INFLAMMATORY BOWEL DISEASE (IBD)

Inflammatory bowel disease is an umbrella term that covers two chronic inflammatory diseases of the gastrointestinal system. These are ulcerative colitis and Crohn's disease.

ULCERATIVE COLITIS

Ulcerative colitis is one of the inflammatory bowel diseases. There is a general incidence of 10-15 per 100,000 of the population. The incidence is, however, much higher in the Jewish population. The aetiology is unknown but it is, however, thought to be an autoimmune disorder in individuals with a yet unknown hypersensitivity to an external antigen. It could equally be of genetic origin as it is known to run in families. About 20% of patients have a positive family history. No specific gene has, however, been associated with it. Males are more susceptible. It is most common in the 20-30 age group.

Pathology: Lesion starts in the rectum and spreads proximally. Only the mucosa and submucosa are involved. There are multiple minute ulcers with the formation of pseudopolyps. On microscopy, the lamina propria is infiltrated by chronic inflammatory cells. It is an established precancerous condition. The cancer risk is 3.5% but is much less in early cases. The cancer risk increases with the duration of disease and involves the colon rather than the rectum.

Clinical features: Ulcerative colitis presents with recurrent watery/bloody diarrhoea with the associated loss of fluid and electrolytes. In severe cases, patient may have up to 20 bouts of motion in one day. Features of anaemia and hypoproteinemia are not uncommon.

Extraintestinal manifestations include

- Joints: Arthritis of large joints, ankylosing spondylitis
- Skin: Erythema nodosum, pyoderma gangrenosum, aphthous ulcers
- Eye: Iritis
- Liver: Sclerosing cholangitis, cholangiocarcinoma

Grading of disease severity: Mild, moderate and severe

Mild: Maximum of four motions per day with no systemic sign of disease.

Moderate: More than four motions per day but no systemic disease.

Severe: More than four motions per day plus one or more signs of systemic disease. The latter include fever (above 37.5), tachycardia (above 90), hypoalbuminaemia (less than 3G %), and weight loss (above 3 kg).

Complications of ulcerative colitis

Acute:

- **Toxic megacolon** (more than 6cm in diameter with associated systemic symptoms). All the muscle layers of the intestine are affected in this situation. Symptoms include severe abdominal pain, weight loss and dehydration. Diagnosis is by serial monitoring of the intestinal size by plain abdominal X-rays. An increasing colonic diameter is an indication for surgical intervention
- **Perforation:** May complicate toxic megacolon and has a high mortality rate
- **Haemorrhage:** Could be torrential with high mortality

Chronic: Cancer and extraintestinal manifestations (see above)

Investigations

- Plain abdominal X-ray
- Barium enema: Will show the classical lead pipe colon with loss of haustration. In an emergency situation, a water-soluble medium that does not require prior bowel preparation may be employed.
- Sigmoidoscopy/ colonoscopy and biopsy: It is not applicable to acute cases for fear of aggravating disease and/or perforation.

Treatment: Entails a multidisciplinary approach and tailored to the severity of the condition.

- Mild: Oral Prednisolone plus 5-aminosalicylic acid (5-ASA)
- Moderate: As in mild
- Severe: Intravenous hydrocortisone, azathioprine, and cyclosporin A

Indications for surgery

- Failure of medical treatment particularly in severe disease
- Intractable symptoms such as debilitating diarrhoea, and chronic anaemia
- Dependence on high dose of steroids for maintenance
- Metaplastic change
- Extraintestinal manifestations
- Severe haemorrhage
- Stenosis

Surgical procedures

- Proctocolectomy and ileostomy
- Restorative proctocolectomy with an ileo-anal pouch (Parks)
- Colectomy and ileorectal anastomosis. In addition, patient may require intravenous infusion and blood transfusion

CROHN'S DISEASE

This chronic inflammatory lesion was first described in 1932. The pathology was originally thought to be restricted to the terminal ileum and hence the term, 'terminal ileitis'. The latter

was later replaced with 'regional enteritis' when it was discovered that apart from the ileum, other parts of the gastrointestinal tract, from the mouth to the anus, may be involved. All layers of the intestinal wall are involved in the form of sharply demarcated segments with skip lesions. Currently, the term 'granulomatous colitis' is also attached to it as it was discovered that the colon is equally involved in the pathology. The most affected part of the GIT still remains the segment of the terminal ileum just proximal to the ileocaecal valve. Ulcerative colitis is the other form of inflammatory bowel disease. In contrast to Crohn's disease, however, ulcerative colitis affects only the mucosa and submucosa, primarily involves the rectum and extends proximally in a continuous fashion. On the other hand, they both have systemic manifestations.

Epidemiology: All age groups may be affected but there are two peaks of presentation: second and third decades of life and the elderly. It is commoner in whites. The Jews are more commonly affected in the USA.

Aetiology: This is unknown but genetic as well as environmental factors may be involved. It may be due to mutation of the NOD2 gene. Environmental factors include

- Low residue diet
- Autoimmune reaction
- Infection with *Mycobacterium paratuberculosis* and/or *Yersinia enterocolitica*
- Smoking: May initiate the primary process and may also give rise to recurrence after treatment

Pathology: Crohn's disease presents with the following macroscopical features

- Sharp demarcation of the diseased bowel segments from the adjacent uninvolved bowel. Hence, it is said to demonstrate the presence of skip lesions
 - There is a transmural involvement of the bowel wall. The latter is thick and rubbery as a result of oedema, hypertrophy and fibrosis
 - There is fissuring with the formation of fistulae, perforation, adhesions, and abscesses
- Histology reveals transmural inflammation affecting all layers of the affected gut in association with non-caseating granulomas

Clinical presentation: This can be categorised into intestinal and extra-intestinal. The intestinal manifestations include

- Diarrhoea, fever, abdominal pain and loss of weight
- Clinical features reminiscent of acute appendicitis: Right iliac fossa pain and tenderness
- Right iliac fossa mass: It falls into the differential diagnosis of an appendix mass. The mass is usually 'sausage-shaped', tender and mobile
 - Features of intestinal obstruction due to fibrosing strictures (especially the terminal ileum)
- Features of both internal and external fistulation. The former includes fistulation into the bladder or other loops of bowel while the latter may manifest as fistula-in-ano. Enterocutaneous fistula usually follows operative intervention
- Clinical evidence of malabsorption: Protein losing enteropathy, megaloblastic anaemia (due to vitamin B12 deficiency) and steatorrhoea (malabsorption of bile salts)
 - Ano-rectal sepsis and abscesses
 - Higher incidence of gastrointestinal cancers

The extraintestinal manifestations include

- Joint involvement: Migratory polyarthritis, ankylosing spondylitis, sacro-ilitis
- Liver: Sclerosing cholangitis (association not as strong as in ulcerative colitis)
- Eye: Uveitis and episcleritis
- Others: Erythema nodosum, finger clubbing, and renal disorders resulting from the trapping of the ureters by adhesion



BARIUM ENEMA SHOWING THE LEAD-PIPE APPEARANCE OF ULCERATIVE COLITIS

Investigations: These include

- Plain abdominal X-rays: May show evidence of small intestinal obstruction
- Small bowel enema: Multiple skip lesions; narrowing of the terminal ileum (Cantor's sign)
- CT scan: Detects complications

- MRI: Demonstrates abnormality of the gut lumen as well as abscesses
- Stool: Occult blood and culture
- Liver function tests: Low protein/albumin level; enzymes may be elevated
- Full blood count and blood film: Low PCV, leucocytosis and evidence of iron deficiency anaemia
- Elevated erythrocyte sedimentation rate and C-reactive protein
- Electrolytes, urea and creatinine: To rule out dehydration and renal impairment
- Biopsies: Small intestine (Crosby capsule), and rectum (sigmoidoscope)

Treatment: Basically conservative but surgical intervention may be indicated

Medical management is categorised into treatment of the acute episode and maintenance of remission.

Management of the acute episodes involves

- Administration of steroids and immunosuppressive agents such as azathioprine. Oral prednisolone has been found effective in majority of cases
- Enteral or parenteral nutrition in very ill patients
- Administration of intravenous fluids and antibiotics
- Reduction of gastric secretion: Proton pump inhibitor/H₂ receptor blocker
- Anti-tumour necrotic factor-alpha drugs: When there is failure of response to the above treatment
- If diagnosis of the acute phase is made at exploratory laparotomy, no further manipulation is advised as a good proportion will completely subside without further acute episodes

Maintenance of remission: Crohn's disease being a chronic condition requires maintenance therapy

- Sulphasalazine: This is a combination of 5-aminosalicylic acid (5-ASA) and sulphapyridine. The active ingredient is the 5-ASA which is released by bacterial action in the terminal ileum and the colon. These are the most common sites of the lesion.
- Monoclonal antibodies to tumour necrotic factor-alpha: Infliximab and adalimumab have been found useful in achieving remissions
- Immunosuppressive agents: Azathioprine, methotrexate and 6-mercaptopurine
- Antibiotics: Ciprofloxacin and metronidazole
- Control of diarrhoea: Imodium and codeine phosphate
- Supplements: Folic acid, B12 and iron tablets

Surgical treatment: This is indicated in the management of complications which include fistulae and abscesses. Emergency surgical intervention is rarely indicated except in the management of intestinal perforation with peritonitis. At all times, conservative surgery is indicated as patients, more often than not, require further surgical interventions in the future. **Surgical procedures for**

Crohn's disease include

- Resection of strictured segment of bowel with end-to-end anastomosis
- Stricturoplasty: Reminiscent of pyloroplasty to widen the lumen thereby forestalling intestinal resection
- Bypass of narrow or strictured segments of bowel

Complication of surgery for Crohn's disease include

- Short bowel syndrome
- Infection: Wound infection and abdominal abscesses
- Fistula formation
- Anastomotic dehiscence
- Recurrent disease

Comparison between Crohn's disease and ulcerative colitis

Macroscopic features

Criterion	Crohn's disease	Ulcerative colitis
Affected gut	Small intestine and colon	Colon only
Distribution	Skip lesions	Diffuse
Stricture	Early	Late/rare
Lymphoid reaction	Marked	Mild
Fibrosis	Mild to moderate	Mild
Granulomas	Present	Absent
Fistula/sinuses	Present	Absent
Clinical		
Fat/vitamin malabsorption	Present	Absent
Malignant potential	Present	Present
Response to surgery	Poor/fair	Good

CHAPTER TWENTY

INTESTINAL OBSTRUCTION

The gastrointestinal tract can be likened to a natural ever-flowing stream. The peristaltic movements of the intestines ensure this free flow of intestinal contents (food, gastrointestinal secretions and gas) from proximal to distal gut. Intestinal obstruction is said to occur when there is failure of this distal propagation of intestinal contents. Most often, this results from physical obstruction of the lumen of the gut. On the other hand, it may be due to an intrinsic inability of the intestinal musculature to perform its natural function. It should be noted that in the latter situation, there is no evidence of physical luminal obstruction.

Clinical types of intestinal obstruction

- 1 Mechanical (dynamic) obstruction: This is characterised by an obvious obstruction of the lumen of the gut. The gut proximal to the point of obstruction undergoes more vigorous contraction (peristaltic movement) in a bid to overcome and possibly expel the obstructing agent. This accounts for the dynamic tag to this type of intestinal obstruction.
- 2 Paralytic ileus (adynamic obstruction): There is failure of the contracting mechanism of the intestinal musculature. There is no evidence of physical obstruction as the lumen of the gut is quite patent. It is associated with gross dilatation of the gut.
- 3 Strangulation: This is one of the most feared complications of intestinal obstruction. The blood supply to the involved segment of gut is compromised. This results in ischaemic necrosis and ultimately, gangrene of the affected segment of gut.
- 4a Acute intestinal obstruction: This is of sudden onset and the symptoms are more severe. A typical example is an obstructed hernia
- 4b Chronic intestinal obstruction: This is of insidious onset and the symptoms are rather slow in progression. The typical example is large bowel obstruction
- 4c Acute-on-chronic obstruction: An apparently chronic form of obstruction suddenly becomes acute. A typical example is large bowel obstruction which, though partial initially, suddenly develops acute symptoms at the onset of complete intestinal obstruction.
- 5 A high intestinal obstruction involves the upper part of the gut (small intestine) while a low intestinal obstruction involves the colon.
- 6 Closed loop obstruction: This is said to occur when there is occlusion of both the proximal and distal ends of the affected segment of the intestine. The pressure buildup within this isolated segment tends to enhance the onset of gangrene and perforation. A typical example of a closed loop obstruction is sigmoid volvulus.
- Mesenteric vascular disease: This results from a thromboembolic phenomenon involving the blood supply to a segment of intestine.

MECHANICAL INTESTINAL OBSTRUCTION

The aetiology of obstruction of any hollow viscus, including the intestine, can be categorised into intraluminal, intramural and extramural

Intraluminal (within the lumen): Causes are

- Impacted faeces
- Foreign body
- Parasites (ascariasis)
- Bezoars: Phytobezoars and trichobezoars
- Gallstone
- Pedunculated tumour

Intramural (within the wall): Causes include

- Congenital atresia
- Hirschsprung's disease
- Tumours: Primary (small bowel, colonic), and secondary
- Strictures (inflammatory): Postoperative, Crohn's disease, radiation enteritis, scleroderma
- Intussusception

Extraluminal (outside the wall): Causes include

- Obstructed hernias: Primary (congenital as in diaphragmatic and acquired as in groin hernias) and secondary (incisional and internal postoperative as in lateral space and mesenteric hole)
- Bands and adhesions (congenital and acquired as in postoperative, inflammatory, chemical and pharmacological). Chemical causes include starch and talc while the beta blocker, practolol is a classical example of a pharmacological cause.
- Abdominal tumours (masses)

By and large, bands/adhesions and obstructed hernias constitute the commonest causes of intestinal obstruction. They are closely followed by malignancy

Pathophysiology of intestinal obstruction

The cardinal symptoms of intestinal obstruction are: abdominal pain, vomiting, constipation and abdominal distension. These features may be explained by a good knowledge of the pathophysiology.

- Pain: With the advent of obstruction, the gut proximal to the point of obstruction contracts vigorously in a bid to overcome the obstruction. The contraction is rhythmical in nature. This accounts for the colicky nature of the abdominal pain associated with uncomplicated intestinal obstruction
- Vomiting: An enormous amount of fluid passes through the gastrointestinal tract daily. This consists of saliva, gastric juice, bile, pancreatic and intestinal secretions. In the presence of obstruction, these secretions are held back in the segment of gut that

is proximal to the point of obstruction. Vomiting ensues when this fluid 'overflows' into the stomach. The higher the level of obstruction, the closer it is to the stomach and hence the earlier the onset of vomiting. This accounts for the fluid and electrolyte loss which is an important contributory factor in the morbidity and mortality associated with intestinal obstruction.

- Constipation: In complete obstruction, the gut distal to the point of obstruction is blocked from receiving intestinal contents from the proximal gut. This results in constipation. The patient may, however, still be able to pass some stool for a while after the onset of intestinal obstruction. This is due to the evacuation of the pre-existing contents of the distal gut. This is subsequently followed by absolute constipation.
- Distension: Intestinal contents and gas (mainly swallowed air) are held up in the proximal gut. There is subsequent dilatation of the latter in a bid to accommodate the intestinal contents whose movement is restrained by the 'hold up' resulting from the intestinal obstruction. Just as the jam worsens with time in a traffic situation, so also the distension associated with intestinal obstruction increases with time. The more distal the obstruction, the more room there is for distension to occur. It follows therefore that the more distal the obstruction, the more room there is for distension to take place. This explains the predominant status of abdominal distension in the symptomatology of colonic obstruction
- Strangulation: The pressure exerted on the intestinal wall is proportional to the degree of distension. Venous occlusion occurs when this surpasses the venous pressure. The resultant congestion further increases the pressure on the intestinal wall. Unrelieved, the pressure may surpass the arterial pressure thereby cutting off the blood supply to the involved segment of intestine. The resultant ischemia ultimately results in intestinal necrosis and gangrene.
- Shock in intestinal obstruction: There is a combination of hypovolemic and endotoxic shock. The hypovolemic component is multifactorial. This is a combination of fluid and electrolyte loss by way of vomiting and third space loss due to the 'locking up' of fluid within the intestinal lumen. The third space loss is further worsened by the seepage of fluid through the weakened wall of the grossly dilated gut into the peritoneal cavity. Bleeding from rupture of the capillaries equally exacerbates hypovolemic shock. Endotoxic shock results from the proliferation of bacteria in the proximal gut. These organisms eventually translocate through the intestinal wall into the peritoneal cavity. This explains the genesis of secondary bacterial peritonitis in intestinal obstruction. Further delay in management will result in full-blown septic shock.

Clinical presentation of intestinal obstruction

As mentioned above, the patient presents with the symptom quartet of abdominal pain, vomiting, constipation and abdominal distension. The relative severity and period of onset of each of these will depend on the level (high or low) of obstruction.

- Upper intestinal obstruction: Pain and vomiting occur early and predominate. The pain is located in the central abdomen due to the midgut embryological origin of the small intestine. Constipation and abdominal distension occur later

- Lower intestinal obstruction: Constipation and abdominal distension occur early and are the predominating symptoms. Abdominal pain and vomiting are late events
-

Other important aspects of the history include

- Previous abdominal surgery (laparotomy): This raises the suspicion of intestinal obstruction due to bands and adhesions. It should be pointed out, however, that inflammatory conditions of the abdomen, even without a concomitant laparotomy, may predispose to the formation of bands and adhesions
- Presence of groin swelling: This is suggestive of an obstructed hernia as a likely cause of the intestinal obstruction
- Chronicity and associated loss of weight: This is suggestive of a longstanding underlying neoplasm
- Strangulation: Simple obstruction is associated with colicky abdominal pain. The pain, however, tends to become continuous with the advent of strangulation. There may also be associated systemic symptoms such as fever and progressive deterioration of the clinical state. Other features of strangulation are tachycardia and leucocytosis.

Examination will reveal an ill-looking patient who may manifest with clinical signs of dehydration (sunken eyeballs, dry tongue and decreased skin turgor) or shock (cold and clammy extremities, tachycardia and hypotension). There may be associated pyrexia. Abdominal examination will reveal the following

- Abdominal distension: Distension due to small intestinal obstruction is centrally-located while distension due to large intestinal obstruction is located at the flanks.
- Previous laparotomy scar is suggestive of obstruction due to bands and adhesions
- Visible peristaltic movements: Indicative of intestinal obstruction. Note, however, that this is best seen in a thin, healthy patient
- Examine the groin for the presence of an obstructed hernia. In particular, look out for an apparently small bulge in the area of the femoral canal. This may be indicative of an obstructed femoral hernia. This latter condition is commoner in females
- Abdominal palpation will reveal generalised guarding and tenderness. Rebound tenderness may indicate the onset of strangulation. A palpable mass may occur in intussusception and obstruction due to an intestinal neoplasm.
- Bowel sounds are increased in intestinal obstruction. This may later diminish or even become absent when intestinal obstruction is complicated by peritonitis
- Rectal examination may reveal faecal impaction, the apex of an intussusception or a rectal or pararectal growth.

General management of intestinal obstruction

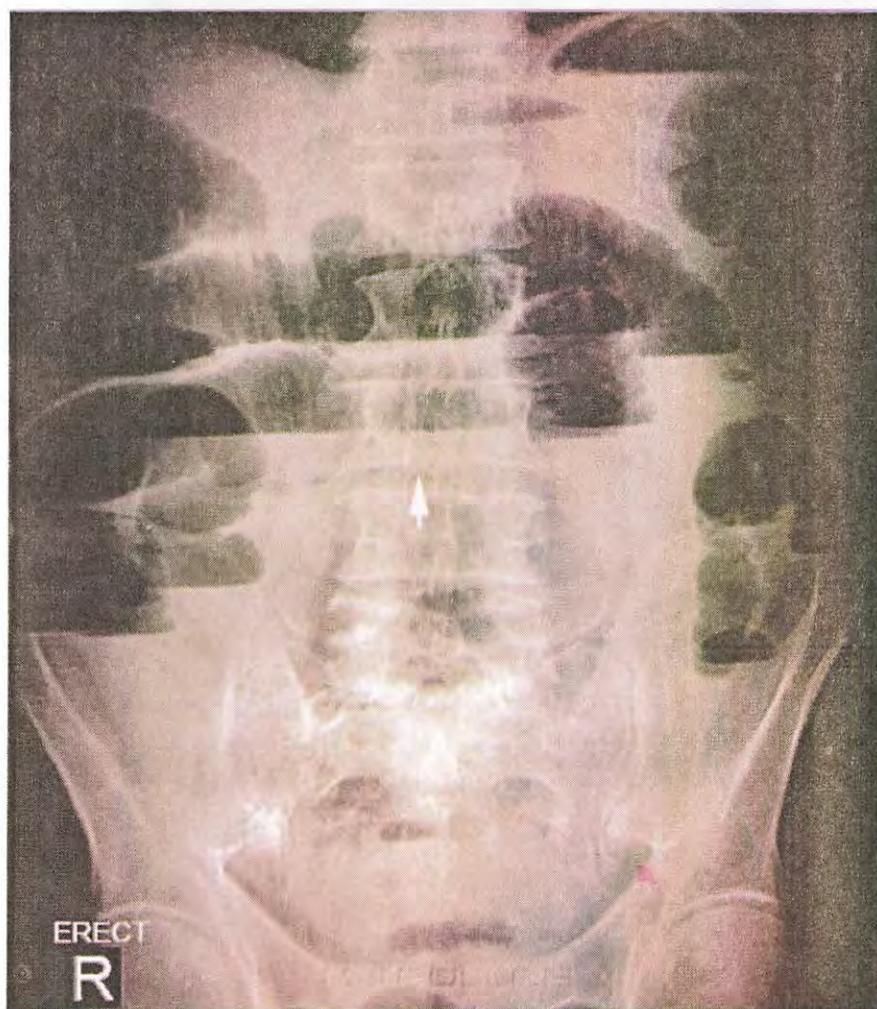
Resuscitation takes priority of management in all patients with intestinal obstruction

- Set up an intravenous line and resuscitate with either normal saline or Ringer's lactate infusion.
- In the course of setting up the infusion line, take samples of blood for the following investigations: Full blood count, electrolytes/urea/creatinine. A sample of blood is also taken for grouping and cross-matching
- Place the patient on a regime of nothing by mouth
- Pass a nasogastric tube to decompress the GIT. In addition to reducing the intra-abdominal pressure, nasogastric aspiration also helps in getting rid of the toxic contents of the GIT. Note that the tube is meant to suck out both fluid and gas. It should therefore be left open for drainage even when there is apparently little or no effluent
- Catheterise the bladder and monitor the urinary output. This helps in monitoring the rehydration process
- Commence on intravenous antibiotics to cover for both aerobic as well as anaerobic organisms
- Commence on intravenous analgesia. Rather than masking the abdominal signs, adequate analgesia will relax the patient and make it easier to detect some apparently occult signs like the presence of an abdominal mass

Having resuscitated the patient, one can now confidently send for the following specific investigations

- Plain abdominal X-ray (erect and supine views): The erect view will demonstrate multiple air-fluid levels while the supine film will show dilatation of the gut proximal to the site of obstruction. The lower the point of obstruction, the more the number of demonstrable air-fluid levels. Small intestinal distension appears more centrally-located on plain X-ray while that involving the colon is more peripheral. Similarly, the identity of the dilated gut can be inferred from the radiological features. Valvular conniventes of the jejunum span the entire width of the gut while haustrations which do not traverse the entire width of gut are in keeping with colonic dilatation. The ileum has been described as being 'characterless' by Wangesteen.
- Contrast studies: These have been found to be more useful in large intestinal than small intestinal obstruction. It is difficult to distinguish mechanical and pseudo-colonic obstruction by plain abdominal X-rays. Contrast studies should, however, be carried out with caution as inspissation of barium may worsen the intestinal obstruction. A water soluble medium such as gastrograffin is more acceptable and may demonstrate evidence of intestinal obstruction particularly with the aid of fluoroscopy. Dilute barium enema may be both diagnostic as well as therapeutic in intussusception
- Abdominal USS: More informative than plain X-rays. It is, however, operator-dependent. Ultrasound scan will demonstrate dilated bowels as well as evidence of peritoneal fluid

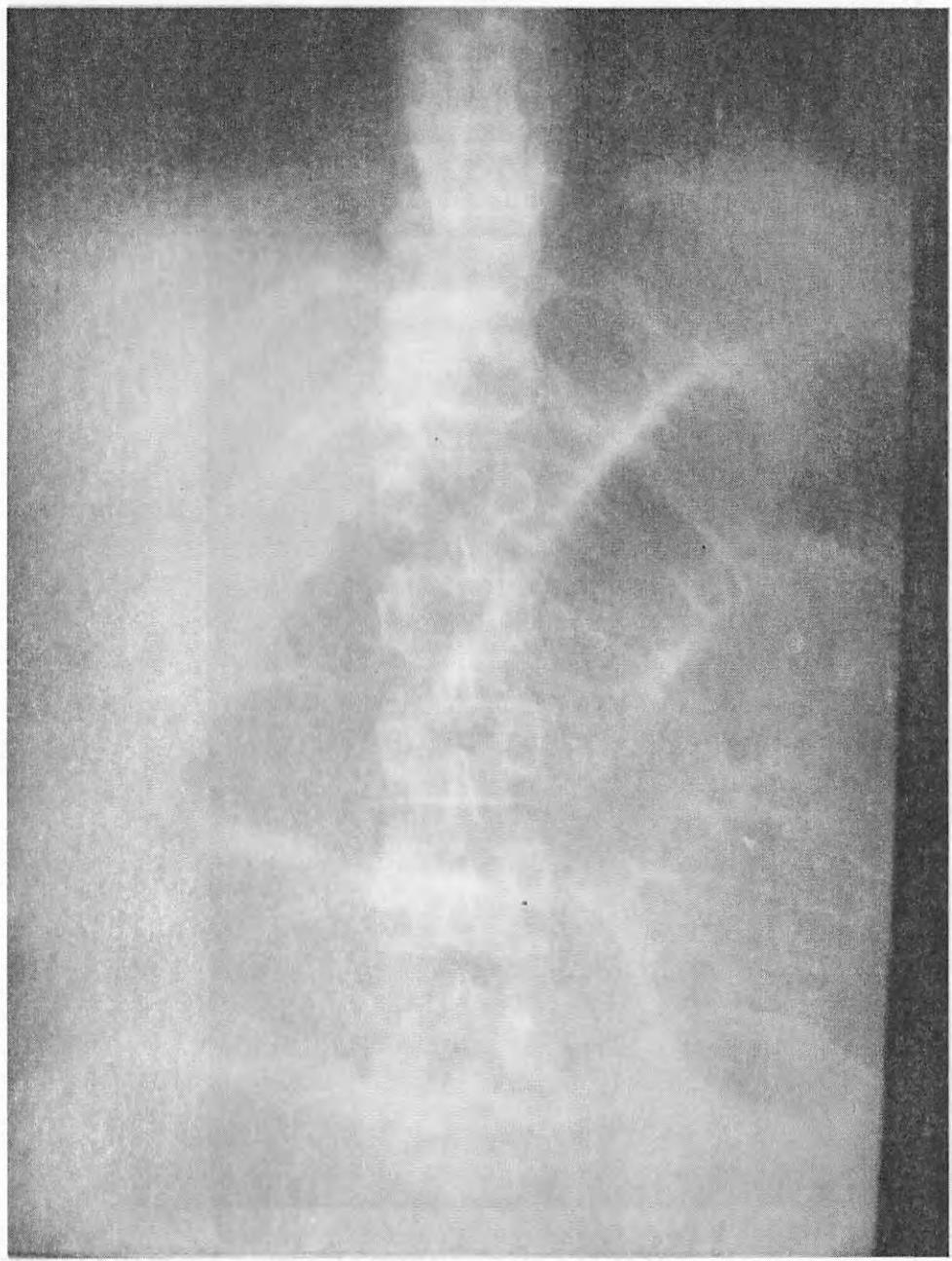
- CT scan: This is the most informative radiological investigation. Unfortunately, however, it is the least accessible and affordable
- Flexible sigmoidoscopy/colonoscopy: In addition to identifying the index cause of the colonic obstruction, it helps to exclude synchronous adenoma or carcinoma below the level of obstruction.



PLAIN ABDOMINAL X-RAY (ERECT FILM) SHOWING MULTIPLE AIR-FLUID LEVELS

The subsequent management is more specific and depends on the aetiology and nature of the intestinal obstruction. As earlier mentioned, obstructed hernias and bands/adhesions constitute the commonest causes of intestinal obstruction.

A Obstructed hernia: Femoral hernia is more likely to obstruct than its inguinal counterpart in view of the anatomy of the femoral canal. The femoral ring is almost completely surrounded by ligaments (inguinal, lacunar and pectineal). The lone 'breathing space' is in the lateral wall which is formed by the femoral vein. Femoral hernia both in its normal and obstructed forms is commoner in females. Nevertheless, it is important to note that inguinal hernia remains the commonest hernia in females. The ultimate treatment is surgery (herniotomy/herniorrhaphy).



PLAIN ABDOMINAL X-RAY (SUPINE) SHOWING DILATED LOOPS OF BOWEL

This is discussed in greater detail in the section on hernias. Richter's hernia in which only a part of the circumference of the gut is involved has a peculiar form of clinical presentation when obstructed. It is associated more with diarrhoea rather than constipation. In Maydl's hernia involving two adjacent loops in the sac, it is the intervening intra-abdominal portion that first gets obstructed. This raises a diagnostic dilemma.

B Bands and adhesions:

Bearing in mind that these might have arisen as a result of a previous abdominal surgery, it is not adviseable to reoperate on these patients in a hurry. This is because any subsequent surgical intervention may result in the formation of more bands and adhesions. There is therefore no end to repeat surgeries with the attendant risks and complications. It is therefore advised to 'make haste slowly'. Management is essentially conservative but with some caveat which will be mentioned subsequently

- Admit patient into the ward for close monitoring
- Resuscitate as outlined above
- Monitor vital signs closely: Pulse, blood pressure, temperature and respiratory rate
- Examine the abdomen regularly: Look out for abdominal distension (monitor the abdominal girth), tenderness and rebound tenderness
- Monitor the general condition of the patient with a view to eliciting any progress or decline in the general state

The main aim of close monitoring is the early detection of the onset of strangulation. As pointed out above, no definite symptoms or signs can be ascribed to the latter. A high index of suspicion is the key to its detection. A suspicion of possible strangulation should be raised if the patient develops any of the following: fever, tachycardia, hypotension and rebound tenderness. This scenario calls for emergency exploration of the abdomen. The mantra should be: 'if in doubt, explore'. Thus, though it is regarded as a conservative form of management, it is better dubbed an 'active-conservative management'. It is analogous to the conservative management of an appendix mass by the Ochsner-Sherrin's regimen. Failure of conservative management is a strong indication for exploratory laparotomy. The latter involves the release of bands and adhesions. The affected gut is examined for evidence of gangrene. Resection and anastomosis of any segment of strangulated gut is carried out.

C Sigmoid volvulus:

it accounts for 76% of all colonic volvulus and is a classical example of closed-loop obstruction. The pathology is that of the sigmoid colon twisting around its mesentery. Predisposing factors to this form of intestinal obstruction are

- Redundant loop of sigmoid colon
- Long mesentery of the sigmoid colon with the limbs closely attached and fixed at the base
- Associated chronic constipation resulting in gross colonic dilatation

The closed-loop nature of the obstruction results in gross dilatation that may culminate in gangrene if urgent relief is not given. The torsion is either clockwise or anticlockwise in nature and is about 180 to 720 degrees. The pedicle for rotation is provided by the narrowed sigmoid mesocolon. Redundancy of the colon is seen in Hirschsprung's disease and Chaga's disease. Sigmoid volvulus may therefore occur in both conditions. It is commoner in males and is usually of sudden onset. Sigmoid volvulus presents with abdominal pain, constipation and abdominal distension. The latter may be so gross as to cause respiratory distress. Examination reveals abdominal tenderness, tachycardia and hypotension.

Initial management is as outlined earlier. The plain abdominal X-ray is diagnostic and demonstrates the grossly dilated sigmoid colon. It extends in an inverted U-pattern from the pelvis to the right upper quadrant of the abdomen. There is loss of haustration and evidence of fluid levels in both limbs of the loop in the erect film.

Relief of obstruction may be effected by the passage and negotiation of a rectal tube through the point of obstruction. Its success is heralded by an explosive sound due to the rapid gushing out of faeces and gas. A success rate of 80% has been reported. It has been suggested that when successful, the rectal tube should be secured by a suture to the perineal skin and left in-situ for 24 to 48 hours. The risks inherent in this non-surgical method of decompression include the inadvertent reduction of a gangrenous sigmoid colon and the rather small risk of colonic perforation.

Surgical management: This involves an exploratory laparotomy and should be the treatment of choice when the clinical features are suggestive of colonic ischemia. Other indications for surgical management are

- Failure of non-operative management
- When drainage from the rectal catheter is blood-stained
- Sigmoid colonic mucosa shows evidence of gangrenous patch

Non-operative treatment should ideally be followed up with elective sigmoid colectomy to forestall recurrent volvulus whose incidence has been reported as between 40% and 60%. There is a place for open detorsion of a viable colon followed by elective sigmoid colectomy. This is quite helpful in the management of very ill patients. Sigmoidectomy in general is currently been carried out as a one-stage procedure.

Caecal volvulus accounts for 22% of colonic volvulus. It may be due to congenital incomplete rotation of the midgut that leaves the caecum 'hanging' on a long mesentry. Pregnancy is a risk factor. It involves a clockwise twist around the ileocolic vessels and the movement of the caecum to the superior-left of its natural position. It presents with abdominal pain, vomiting and abdominal distension. The supine film of the abdomen will show a grossly dilated caecum while the erect film will reveal a single, long fluid level. The ultimate definitive treatment is right hemicolectomy. There is, however, a place for caecopexy and caecostomy.

Ileo-sigmoid knotting (also referred to as double or compound volvulus) is a special variety of volvulus in which the ileum twists around the base of the sigmoid mesocolon. The remote predisposing factors include redundant loops of both the ileum and the sigmoid colon with associated long mesentry. Constipation is the triggering factor. The patient will present with clinical features of intestinal obstruction. Plain abdominal X-rays will demonstrate air-fluid levels in both the small intestine and the colon. Treatment involves resuscitation and emergency laparotomy. The recommended surgical procedure is resection and anastomosis of the ileum coupled with hartman's procedure. Ileosigmoid knotting is associated with a high mortality rate (1.5 to 73%) owing to the high incidence of gangrene.

Volvulus involving the transverse colon is rare (2%). Treatment is by either transverse colectomy or an extended right hemicolectomy.

D Intussusception:

This refers to a situation where a proximal segment of gut invaginates (telescopes) into a distal segment. The invaginating proximal segment is referred to as the intussusceptum while the receiving distal part is the intussuscipiens. This results in an incomplete subacute intestinal obstruction and venous congestion of the intussuscipiens. Unrelieved, the end result is strangulation. This condition is common in children between the ages of 3 months and 2 years (peak incidence: 6 to 9 months). This coincides with the period of weaning and may be due to a change in the bacterial flora of the gut. Lymphoid hyperplasia at the Peyer's patch of the distal ileum may be the triggering factor. On the other hand, intestinal polyp, tumour or Meckel's diverticulum may act as the lead point of intussusceptions in the adult.

Types: Intussusception usually builds up from the ileum and progresses in a retrograde direction. Varieties include ileo-ileal, ileo-caecal and ileo-colic (commonest). Others are caeco-colic, and colocolic (rare). Intussusception may progress down to the anus where it may prove difficult to differentiate from rectal prolapse.

Clinical presentation is with features of intestinal obstruction and is age-related. There is a peculiar form of presentation in childhood. The initial symptom in the latter consists of mucoid, bloody diarrhoea (stool is referred to as anchovy sauce). The latter is explained by the incomplete nature of the obstruction and the associated venous congestion. Abdominal



INTUSSUSCEPTION

examination in this irritable child will reveal a sausage-shaped mass around the right iliac fossa. The clinical triad in childhood intussusception consists of paroxysmal abdominal pain, rectal bleeding and a palpable abdominal lump.

Investigations: These include

- Plain abdominal X-rays (erect and supine): Will show multiple air-fluid levels. The head of the intussusception may be outlined with an associated absence of gas at the right iliac fossa
- Ultrasound scan: Particularly when diagnosis is doubtful

- Barium enema: Both diagnostic as well as therapeutic (may reduce the telescoping gut). Intussusception demonstrates a characteristic coiled spring appearance.

As in all cases of intestinal obstruction, resuscitation with intravenous infusion and nasogastric aspiration takes pride of place. The first line of definitive treatment in the uncomplicated case is reduction by either air or contrast enema under fluoroscopic or ultrasound control. Indications for surgical intervention are

- Failure of reduction with air or contrast enema
- Presence of complication such as perforation
- Clinical evidence of strangulation

Definitive treatment is surgery and consists of open reduction in the cephalad direction. 'Milking' of the intussusciens commences at the most distal part of the complex. The last portion of the latter usually proves more difficult to reduce and may necessitate resection and anastomosis, especially if gangrenous.

E Large intestinal obstruction: Colonic tumours constitute the commonest cause.

Constipation and gross distension are the dominant symptoms. The priority of treatment is the relief of obstruction by exploratory laparotomy after the initial management. The definitive treatment depends on the part of the colon that is affected

- Caecum and ascending colon: Right hemicolectomy with an ileo-transverse anastomosis. A temporary diversion for the relief of obstruction may be effected with a diversionary ileo-transverse anastomosis in a very ill patient. This is followed later by definitive resection when the patient is considered fit.
- Transverse colon: Extended right hemicolectomy. Lesions close to the splenic flexure and those with extensive involvement of the transverse mesocolon will benefit from a more extensive resection with primary anastomosis between the ileum and the upper descending colon
- Splenic flexure: Subtotal colectomy
- Left colon (including sigmoid colon): Current management is by way of intra-operative irrigation and segmental colectomy. This is referred to as the one-stage procedure. The two-stage procedure consists of initial resection coupled with a protective colostomy. Hartman's procedure falls into this category. The original management regimen consists of a three-stage procedure: initial colostomy to relieve the obstruction followed by an elective resection and anastomosis. Closure of the colostomy constitutes the last leg of the 'race'. The one-stage procedure is ideal for relatively fit patients while the other options are reserved for ill patients whose priority of the initial treatment is relief of obstruction.

F Ascariasis:

Ascariasis can cause intestinal obstruction in endemic areas. This may result from occlusion of the terminal ileum by a bolus of worms. Treatment is by exploratory laparotomy and the milking of the latter into the caecum. Enterotomy and manual evacuation of ascaris worms is employed

when milking into the caecum proves difficult. Patient is further treated with a course of antihelminthic therapy when healing is presumed to have occurred.

G Bezoars:

Bezoars are of two types – phytobezoars and trichobezoars. Phytobezoars are composed of vegetable materials and are common in edentulous patients and those that have undergone partial gastrectomy. Trichobezoars, on the other hand, consist of swallowed hair and are common in psychiatric patients. Both can result in a bolus of concretion that can occlude the intestinal lumen and result in intestinal obstruction. Treatment is by enterotomy

H Gallstone ileus:

This is a very rare form of intestinal obstruction. It is sequel to calculous cholecystitis that is complicated by a cholecysto-duodenal fistula. The gallstone is discharged into the duodenum through the fistulous communication and subsequently impacts in the terminal ileum. Diagnosis requires a high index of suspicion. The possibility is raised by the diagnosis of intestinal obstruction in a patient with already confirmed cholelithiasis. This is corroborated by the outlining of the biliary duct by air (pneumobilia) coupled with an opacity in the right iliac fossa (but recall that only 10% of gallstones are radio-opaque). Definitive treatment consists of emergency exploratory laparotomy and enterotomy. This is followed later by elective cholecystectomy.

I Faecal impaction:

Faecal impaction may occur in elderly bed-ridden patients and those with fissure-in-ano. The latter is associated with painful defaecation which makes natural defaecation a dreadful event. Faecal impaction has also been associated with mental ailment. Diagnosis is made from the history and the finding of an indentable mass on abdominal examination. Digital rectal examination will reveal a rectum that is loaded with hard, inspissated faeces. In addition to the features of intestinal obstruction, plain X-rays will show stippling of faeces in a dilated gut. Treatment is by a combination of manual evacuation and enema saponis

J Mesenteric vascular disease

It is a rare form of intestinal obstruction. Of two types: occlusive and non-occlusive. The occlusive variety is secondary to thromboembolism and atherosclerosis. The embolus may arise from the heart as a result of myocardial infarction or atrial fibrillation. Atherosclerosis of the portal vein may result in thrombosis. Portal hypertension, portal pyaemia and sickle cell disease all predispose to thrombosis. Non-occlusive mesenteric vascular occlusion results from a sluggish circulation as occurs in cardiac failure, hypotension and hypoperfusion due to shock. The superior mesenteric artery and vein are the most affected vessels in mesenteric vascular disease.

Presentation: Mesenteric vascular disease results in ischaemic necrosis of the involved bowel. This culminates in strangulation, gangrene and perforation. The patient will present with clinical features of intestinal obstruction and shock. The latter arises from a combination of hypovolemic (bleeding into the wall of the gut and third space loss) and septic shock (peritonitis and septicaemia). There is associated bloody diarrhoea. The infarcted bowel may be felt as a vague tender mass on abdominal examination. The cardiovascular system should be clinically evaluated for evidence of atrial fibrillation and myocardial infarction. It is important to look out for clinical evidence of liver disease. The triad of acute colicky abdominal pain, rectal bleeding and shock in an elderly patient with clinical features of atrial fibrillation is classical for mesenteric vascular

disease. A milder form may present with what has been referred to as 'intestinal angina'. The symptoms are associated with meals. This makes the patient afraid to eat with the attendant loss of weight.

Investigations: These include

- Plain abdominal X-rays (erect and supine): Will show features of intestinal obstruction
- Abdominal ultrasound scan
- Doppler studies
- Selective angiography of the superior mesenteric vessels: Presence of gas bubble in the mesenteric vein is pathognomonic
- Chest X-ray, ECG and echocardiography: To diagnose heart disease
- Full blood count, grouping and cross-matching of blood
- Liver function tests
- Serum electrolytes/urea/creatinine and phosphate (rising level as small bowel is rich in phosphate)

Treatment: Initial management of intestinal obstruction as outlined above. Blood transfusion is required in the early management. Patients with the non-occlusive form will require conservative treatment for the management of the underlying cause (heart failure, hypoperfusion or hypotension). The occlusive variety will require exploratory laparotomy for assessment of the intestinal viability. Gangrenous bowel, which is usually massive, will require resection and anastomosis. A second-look surgery is essential to determine the viability or otherwise of marginally-perfused gut left behind during the primary surgery. There may be need for re-resection. Arterial thrombosis requires on-table embolectomy and anticoagulation therapy while its venous counterpart will benefit from anticoagulation therapy alone. Thrombolytic therapy may be carried out with the catheter placed in the affected vessel.

Complications of mesenteric vascular disease

- Short bowel syndrome: Patient requires at least 1.2 meters of small intestine for survival. Patient may benefit from total parenteral nutrition and intestinal transplantation
- Acute renal failure from hypovolemic shock
- Adult respiratory distress syndrome
- Disseminated intravascular coagulation (DIC)

PARALYTIC ILEUS

Paralytic ileus is also referred to as adynamic obstruction. Unlike the mechanical form of intestinal obstruction, there is no physical blockage of the lumen of the gut. It is due to paralysis of the intestinal musculature due to failure of its nervous supply (Auerbeck's and Meissner's plexuses). It is on this basis that it is sometimes referred to as neurogenic obstruction. The motility of the intestine is governed by two opposing forces: The stimulatory influence of the parasympathetic nervous supply and the inhibitory influence of the sympathetic nervous system. Either neuropraxia of the parasympathetic system and/or an increased stimulation by the sympathetic system will result in a decrease in the propulsion of the intestine.

Aetiological factors

- Postoperative: This is the most common cause as it is almost a natural sequel to gastrointestinal surgery or procedures. May be due to neuropraxia sequel to both manipulation of the gut as well as peritoneal irritation
- Peritonitis: May be due to toxic paralysis of the intrinsic nerve supply
- Metabolic conditions: Electrolyte imbalance (hypokalemia and hypercalcaemia), diabetic ketoacidosis and other causes of metabolic acidosis, uraemia and hypothyroidism
- Reflex causes: Retroperitoneal haemorrhage or surgery, fracture of the spine or pelvis, spinal cord injury (above T5 level) and application of a plaster cast. Other rare causes in this group are parturition and ureteric colic. May be secondary to sympathetic overactivity
- Drugs: Anticholinergic drugs (usually administered preoperatively), narcotics (administered postoperatively), vincristine, loperamide and calcium channel blockers.
- Others: Acute pancreatitis, systemic sepsis and shock

Pathophysiology: The absence of peristaltic movements will prevent the downward propulsion and reabsorption of gastrointestinal fluids. This results in the loculation of fluids and electrolytes within the lumen of the gut. This is referred to as a third space loss. Unrelieved, it may result in dehydration and hypovolemic shock. The resultant abdominal distension engendered by the accumulation of intestinal fluid is further worsened by the presence of swallowed gas in the intestinal lumen. The resultant distension may cause splinting of the diaphragm and respiratory insufficiency. Most cases, particularly those that follow surgical intervention, usually improve within three to four days. Beyond this period, fibrinous adhesions may progress to form fibrous bands. The presence of the latter will insidiously convert the paralytic ileus to a state of mechanical obstruction. It is therefore imperative to review the diagnosis of paralytic ileus if symptoms persist beyond the fourth day of onset.

Clinical presentation: A good history is important in order to determine the causative factor. The patient presents with abdominal distension, constipation, and vomiting. Characteristically, paralytic ileus is associated with minimal abdominal pain. Discomfort from the abdominal distension as well as the surgical wound may, however, give the impression of pain from the underlying paralytic ileus. Examination will reveal an ill-looking patient who may manifest with clinical signs of dehydration and shock. There may be laboured respiration and tachypnoea as a result of the abdominal distension. Bowel sounds are characteristically absent. This is in contrast to the increase in bowel sounds found in mechanical intestinal obstruction. Digital rectal examination may show absence of stool in the rectum. Clinical signs suggestive of an underlying cause may also be present.

Investigations:

- Plain abdominal X-rays (erect and supine): Will show dilated gut with uniform distribution of gas throughout the bowel including the colon and rectum. This differs from the radiological findings in mechanical obstruction that demonstrate distended proximal and collapsed distal segments
- Electrolytes/urea/creatinine
- Full blood count

Treatment involves

- Nil per oral and nasogastric aspiration
- Intravenous infusion: Ringer's lactate or normal saline may be used.
- Steps are taken to correct any identified electrolyte anomaly
- Closely monitor the progress of the patient by serial monitoring of the vital signs, abdominal girth and general wellbeing
- Address the underlying cause
- It has been realised that gentle enteral feeding may help to restore motility by triggering the normal gut feedback mechanism. Persistence of the paralytic ileus on attempt at enteral feeding calls for its discontinuation, resumption of nasogastric aspiration and commencement of parenteral nutrition.
- Acute colonic pseudo-obstruction: There is no evidence of mechanical obstruction. It is due to dysfunction of the sacral parasympathetic nerves. Most patients will respond to conservative treatment. There is a place for neostigmine in carefully selected cases. Colonoscopic decompression has been found to be effective in majority of the patients who fail to respond to conservative management. Re-laparotomy is reserved only for patients who fail to respond to the more conservative approach and when perforation is suspected. The latter is usually heralded by a caecal diameter above 12 cm. Surgical treatment is by tube caecostomy, resection and anastomosis. Ogilvie's syndrome is massive non-obstructive colonic dilatation. Disease conditions that may be associated with this include diabetes mellitus, hypothyroidism, sepsis, myopathies and psychiatric disorders. Others are scleroderma, chronic renal failure, orthopaedic procedures and radiotherapy. The management of Ogilvie's syndrome is essentially as outlined above.
- Bowel movement may be stimulated by the administration of lactulose or erythromycin
- Chewing gum (also referred to as sham feeding) has been found to be of help in the restoration of intestinal mobility. The mechanism of action is unknown.

PAEDIATRIC INTESTINAL OBSTRUCTION

Intestinal obstruction is a common paediatric surgical emergency. As usual, the aetiology may be classified into intraluminal, intramural and extramural.

Intraluminal cause of childhood intestinal obstruction is meconium ileus in the newborn and ascariasis in older patients

Intramural causes include

- Atresia/stenosis
- Hirschsprung's disease
- Anorectal anomalies

Causes of paralytic ileus in infancy

Septicaemia and necrotising enterocolitis

Extrinsic causes are

- Malrotation with or without volvulus
- Duplication of gut
- Inguinal hernia

General principles of childhood intestinal obstruction

Clinical features: The cardinal clinical feature of neonatal intestinal obstruction is projectile vomiting. This is bile-stained in lesions below the ampulla of Vater. The lower the level of obstruction, the more gross the abdominal distension. There is associated inability to pass meconium. Unrelieved, it may progress to gangrene, perforation and peritonitis.

Plain abdominal X-ray is the investigation of choice as it shows features of intestinal obstruction. An upper gastrointestinal and contrast enema may aid in the diagnosis of upper and lower intestinal obstruction respectively.

Treatment: Such patients should ideally be managed in a paediatric surgical unit

- Naso-gastric aspiration with a size 8 to 10F tube
- Check PCV, electrolytes/urea/creatinine
- Intravenous infusion: Correct deficit and give maintenance
- Address the underlying cause

Meconium ileus

It is a clinical manifestation of cystic fibrosis. About 10% to 15% of these patients will present with meconium ileus due to obstruction with the viscid, tenacious meconium obstructing the terminal ileum and colon. This may be complicated by atresia, volvulus, or perforation with meconium peritonitis.

Investigation: Plain abdominal X-ray will show dilated loops of bowel, absence of the typical air-fluid levels and a soap-bubble appearance in the right lower quadrant of the abdomen. Contrast enema may reveal a small calibre colon (microcolon) containing meconium.

Treatment: Initial resuscitation as outlined above. The uncomplicated case may be managed conservatively by the administration of gastrografin in the form of an enema. Gastrografin (diatrizoate meglumine) is a hyperosmolar contrast radiological agent. Following adequate fluid rehydration, the diluted agent is carefully instilled in the colon under fluoroscopic control until it is seen to make an inroad into the dilated ileum. This procedure which has a success rate of 55% may be repeated in the uncomplicated case. Indications for surgical intervention include failure of conservative treatment and those complicated by volvulus or atresia. Surgery involves resection of the affected segment and clearance of both the proximal and distal segment of meconium followed by end to end anastomosis. Some difficult cases may require ileostomy

Duodenal atresia

Duodenal atresia may be associated with other congenital anomalies such as Down's syndrome, oesophageal atresia, anorectal anomalies and congenital cardiac malformations. Diagnosis is clinical and supported by plain abdominal radiograph which will demonstrate the classical 'double-bubble' appearance. Treatment is by side-to-side duodenoduodenostomy

Malrotation

Intestinal rotation and fixation is meant to be completed by the 12th week of intrauterine life. Failure of this normal development will result in malrotation. In this condition, caecum and appendix are located at the right hypochondrium or at the upper midline of the abdomen while the duodeno-jejunal flexure lies to the right of the midline. Volvulus is a potential complication in view of the resultant narrow-based mesentery.

Clinical features: In the infant, presentation may be with just bile-stained vomiting whereas in the older child there is associated anorexia, abdominal pain and failure to thrive. When complicated by volvulus, there is associated passage of bloody stool which rapidly degenerates to a state of shock

Investigations: The following are indicated

- Plain abdominal radiograph: Typical gasless abdomen in volvulus
- Upper gastrointestinal series: Abnormally-placed duodenojejunal flexure with small intestinal loops located on the right side of the abdomen. Will show a cork-screw appearance when complicated by volvulus

Treatment: Initial treatment is as outlined earlier. This is followed by emergency laparotomy and Ladd's procedure. During this procedure, the volvulus is untwisted and the bowel repositioned after widening the mesentery of the midgut

Necrotising enterocolitis

This is an acquired (not congenital) condition that affects premature neonates. Immaturity, coupled with formula feeds play complimentary roles as breast milk is known to be protective. No part of the gut is immune but it most commonly affects the terminal ileum and the colon. The pathogenesis is three-fold: intestinal ischemia, bacterial colonisation and the presence of a substrate milk formula in the intestinal lumen

Clinical features: Bilious vomiting, abdominal distension, refusal of feeds and lethargy. These are later followed by bleeding per rectum and features of septicaemia.

Diagnosis: is clinched by the pathognomonic presence of intramural gas on plain X-ray – pneumatosis intestinale. There will be radiological sign of pneumoperitoneum if there is perforation.

Treatment: Initial treatment is as outlined earlier. Patient is commenced on antibiotics. About 70% to 80% of cases will resolve on conservative management coupled with parenteral nutrition. Failure to respond to conservative treatment and complications such as perforation are indications for surgical treatment. Surgery involves resection of gangrenous bowel and primary anastomosis.

Small intestinal atresia

This is believed to be due to intra-uterine interference with the vascular supply of the affected segment of gut. Essentially, there are four varieties of small intestinal atresia

- Type 1: Distal and proximal segments are in continuity but only separated by a web. The associated mesentery is intact
- Type 1I: Both the proximal and distal ends end blindly but are joined by a band
- Type 1Ia: Blind ends are disconnected with a V-shaped gap in the mesentery
- Type 1Ib (classical 'apple peel' or Christmas tree): Deformity as in type 1Ia but with an extensive mesenteric defect, the distal ileum receiving its blood supply from a collateral vessel from the ileocolic artery
- Type 1V: Multiple atresias

Types 1I and 1V may be associated with defective peristaltic activity. Plain abdominal X-ray is the main diagnostic tool but contrast enema is employed in ascertaining the level of obstruction in difficult cases

Initial general treatment consists of resuscitation as outlined earlier followed by definitive surgery. The latter consists of resection of the grossly-distended proximal bowel followed by an end-to-end anastomosis



ABDOMINAL RADIOGRAPH SHOWING THE TYPICAL 'DOUBLE-BUBBLE' SIGN OF DUODENAL ATRESIA

Hirschsprung's disease

The alternative name, congenital aganglionic megacolon, is a reflection of its pathology. It is due to the congenital absence of ganglion cells within the wall of the intestine. The fundamental problem is the failure of migration of vagal neural crest cells into the developing gut. The pathology commences at the internal sphincter of the anus and extends for a variable distance

proximally. In most cases (75%), the aganglionosis is restricted to the rectosigmoid (short segment) but in 15% of cases it is extensive and involves the proximal colon (long segment) and may occasionally extend to involve the terminal ileum (10%). A transition zone exists between the dilated, proximal, normally innervated large gut and the narrow, distal aganglionic segment. Hirschsprung's disease may be genetic and the affected segment may contain the RET-proto-oncogenes

Clinical features: Varied as it presents in various ways

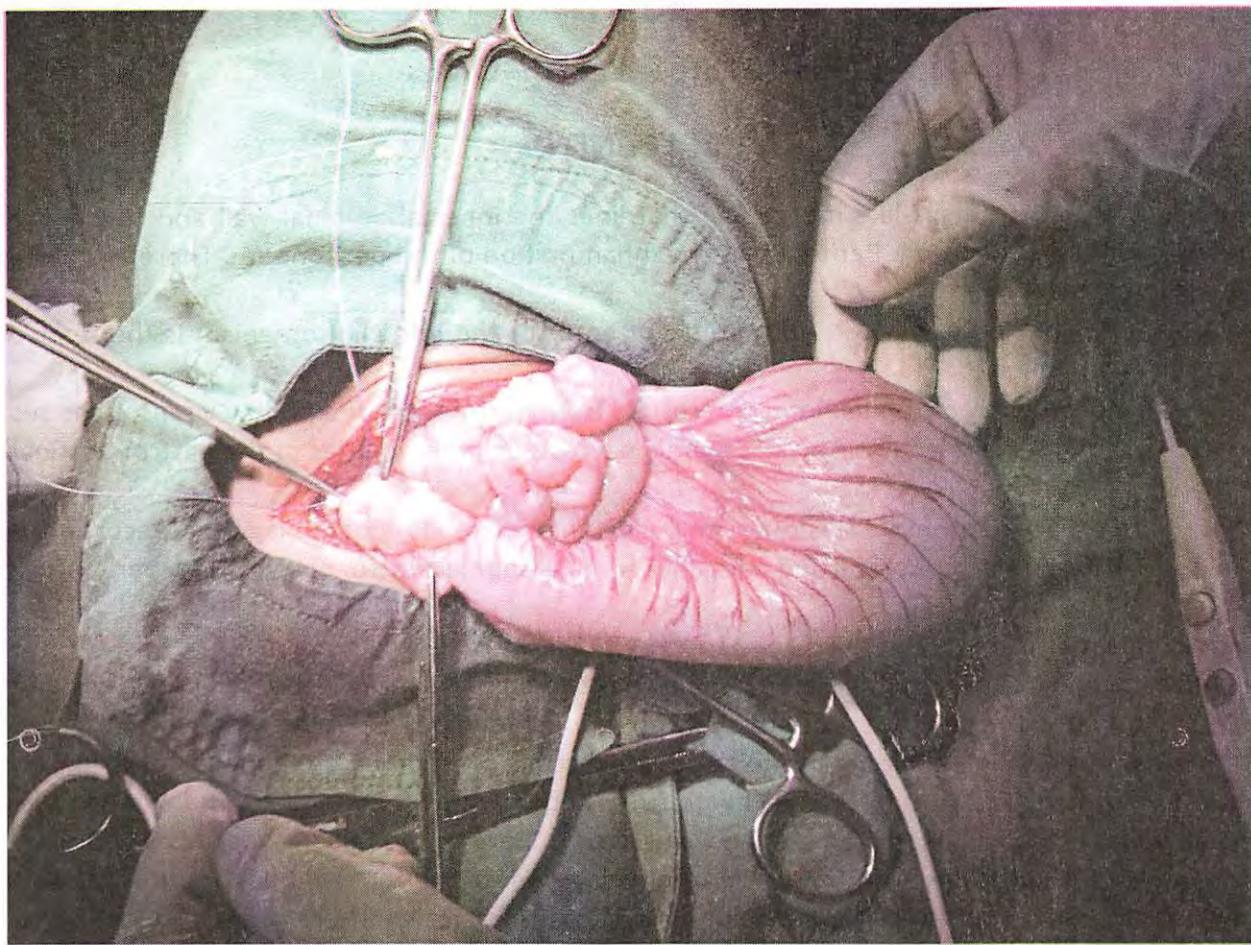
- Delayed passage of meconium within the first 24 hours of birth (80%). This may be relieved by digital rectal examination or a saline rectal washout. Symptoms may recur after a few days of relief
- Clinical features of enterocolitis: Bloody diarrhoea, bilious vomiting, dehydration and abdominal distension.

* Severe chronic constipation: This may be the mode of presentation in late childhood and even in adult life

Investigations

- Plain abdominal X-ray: Radiological features of intestinal obstruction
- Triple rectal suction biopsy: Will demonstrate absence of ganglion cells in the submucosa, presence of large nerve trunks and an increase in acetylcholinesterase-stained nerve fibres in the lamina propria and muscularis mucosa of the spastic segment. The transitional cone-like zone will show few ganglia while the dilated upper segment will demonstrate normal features
- Barium enema: Demonstrates the extent of the lesion. Will show the narrow aganglionic segment, a cone-shaped transitional zone followed by the proximal dilated colon
- Anorectal manometry: Absence of recto-anal reflex is diagnostic

Definitive treatment: This is surgical and is aimed at anastomosing a proximal healthy segment of proximal bowel to the anus. The various operations are generally referred to as pull-through procedures. The aganglionic segment is either bypassed (Duhamel, Soave) or excised (Swenson, Rehbein). Traditionally, treatment is commenced with a proximal defunctioning colostomy using the



Hirschsprung's disease: Note the narrow distal and the dilated proximal segment normal proximal gut as the initial part of a staged-repair. This is followed, when the child is bigger (at least 10 kg) by resection of the involved segment and restoration of continuity. The final phase is the closure of colostomy. Currently, a one-stage repair is feasible and is the preferred method of treatment in some centres.

Other forms of paediatric intestinal obstruction

- Intussusception: Described in the general discussion of intestinal obstruction
- Congenital groin hernias: Discussed in the section on hernias

CHAPTER TWENTY-ONE

PERITONITIS

The peritoneum is the silk-like membrane that lines the inner abdominal wall and covers the organs within the abdomen. Peritonitis is the inflammation of the peritoneum. There are three types of peritonitis

- Primary peritonitis: Primary route of entry of the infecting organism is the female genital tract
- Secondary peritonitis: Infection results from perforation of a hollow viscous, ischemic necrosis, anastomotic leak, ischemic necrosis or other injuries of the gastrointestinal tract
- Tertiary peritonitis: This refers to persistent intra-abdominal soilage that recurs after 48 hours of successful and adequate surgical source control of secondary peritonitis.

Primary peritonitis

The route of entry of organism(s) is the female genital tract and spread is through the vagina and fallopian tubes to the peritoneal cavity. It is common in young girls between the ages of 4 and 10. The commonest infecting organism is pneumococcus. Infection by the gonococcal organism is not uncommon. The patient presents with fever and lower abdominal pain. Examination will reveal lower abdominal tenderness and guarding. There is equally tenderness on rectal examination. Management of primary peritonitis is as described for secondary peritonitis. It is difficult to differentiate this from secondary peritonitis clinically. There are, however, some peculiar features that help to clinch the diagnosis at laparotomy: profuse collection of greenish pus in the pelvis with no evidence of primary disease.

Secondary peritonitis

As mentioned above, it results from a definite intra-abdominal pathology. Aetiology includes

- Direct injury: Penetrating injury, infection following laparotomy
- Leakage of infection through an abdominal source of inflammation: Acute appendicitis, gangrenous bowel, acute cholecystitis
- Perforation of an intra-abdominal viscous: ileum in typhoid and appendix in appendicitis
- Rupture of an erstwhile circumscribed abscess: Appendix, subphrenic and pelvic abscesses
- Intestinal obstruction: There is translocation of organisms from the proximal dilated gut to the peritoneal cavity
- Complication of abdominal surgery: Anastomotic breakdown, biliary peritonitis and laparotomy
- Systemic: As part of septicaemia

By and large, the leading causes of peritonitis are acute appendicitis, perforated peptic ulcer and postoperative complications.

Pathophysiology: Peritonitis will result in the exudation of fluid into the peritoneal cavity. The resultant loss of fluid and electrolytes is worsened by both the outpouring of fluid from a ruptured viscus and the loculation of fluid in the intestine as a result of paralytic ileus. The latter is a known complication of peritonitis. All these will culminate in what is referred to as the third space loss. This fluid and electrolyte loss coupled with the associated vomiting are responsible for dehydration and hypovolemic shock which are common clinical features of acute peritonitis. The initial phase of peritonitis may be purely chemical in nature. Subsequent proliferation of organisms will result in the infective phase. This is the case when the pathology involves the upper GIT as exemplified by perforated peptic ulcer disease. The lower the site of perforation in the gastrointestinal tract, the worse the degree of peritoneal contamination. The subsequent bacterial proliferation and widespread absorption of toxins from the peritoneum will result in septic shock. Hence, the shock associated with peritonitis results from a combination of hypovolemia and sepsis. The resultant septic shock may result in acute renal failure, systemic inflammatory response syndrome (SIRS) and multiple organ dysfunction syndrome (MODS).

Clinical features of secondary peritonitis: The initial clinical features are in keeping with those of the causative factor. For example, peritonitis secondary to ruptured appendix will commence with clinical features of acute appendicitis (right iliac fossa pain and tenderness) while that secondary to perforated duodenal ulcer will commence with epigastric pain and tenderness. With time, pain, guarding and tenderness become generalised. Careful examination, however, will reveal that these signs are most elicited around the primary site of origin. Rebound tenderness is characteristic of inflammation of the peritoneum and strongly suggests onset of strangulation in a case of intestinal obstruction. Bowel sounds are either diminished or absent in full-blown peritonitis. Digital rectal examination will elicit fullness and tenderness particularly when there is associated pelvic abscess. Cervical excitation tenderness will be positive in pelvic inflammatory disease. The term, 'Hippocratic facies', is used to describe the very ill and gaunt-looking patient with advanced peritonitis. Characteristically, the patient is in severe shock and has a moist, cold and cyanosed skin.

Investigations for peritonitis are

- Full blood count: May show anaemia and leucocytosis. There may be thrombocytopenia when peritonitis is associated with coagulation anomalies
- Plain abdominal X-rays (erect and supine): These may demonstrate intestinal dilatation as seen in paralytic ileus; gas under the diaphragm as seen in perforation of an abdominal hollow viscus
- Abdominal ultrasound scan: May show evidence of intra-abdominal collection
- Electrolytes, urea and creatinine
- Group and cross-match blood

Treatment of peritonitis:

This is multidisciplinary and involves the following teams: Surgery, Gastroenterology, Critical care and Nutrition. The general principles of treatment of all types of peritonitis are as follows:

- Correct the underlying process

- Eliminate bacteria and toxins by the administration of systemic antibiotics
- Provide adequate supportive therapy to prevent or limit secondary complications due to organ systemic failure

The initial treatment consists of resuscitative measures

- Set up an intravenous line and commence rehydration with either Ringer's lactate or normal saline. Blood samples for the above investigations are taken before the commencement of intravenous infusion.
- Correct any identified electrolyte anomaly
- Commence on nil orally
- Pass a nasogastric tube: This decompresses the GIT, reduces the load of toxin and prevents aspiration
- Catheterise the urethra: Monitoring the urinary output aids in the assessment of the degree of rehydration.
- Adequate antimicrobial therapy: Combination systemic antibiotic therapy is administered to cover the gram positive, gram negative and anaerobic organisms. The duration of antibiotic therapy will depend on the nature of the underlying pathology, the speed and effectiveness of source control, the severity of the infection and the patient's response to therapy
- Relieve pain by administering adequate analgesia
- Address and rectify any identified coagulation anomaly
- Control the source of infection: This can be achieved either by nonoperative or operative means

Nonoperative control: Methods of nonoperative treatment include percutaneous abscess drainage and endoscopic stent placements. It is useful in the drainage of localised, non-loculated abscesses and is best carried out under ultrasound or CT guidance. Nonoperative treatment may not be effective in the presence of anastomotic dehiscence, infected clot or multiloculated abscesses

Operative methods: This involves surgical intervention and aims at addressing the primary problem. Surgery may involve appendicectomy, closure of a perforated peptic ulcer or resection/closure of typhoid perforation. A specimen of pus/peritoneal fluid is taken for microscopy, culture and sensitivity. It is important to ensure that the patient is reasonably resuscitated prior to operative intervention. Before closing the abdomen, adequate peritoneal lavage should be carried out with warm normal saline in a bid to drastically reduce the bacterial load. The insertion of an abdominal drain is controversial. While some authors believe it helps to clear the peritoneal cavity of infected fluid, others believe it is unnecessary as it is impossible to drain the entire peritoneal cavity. It is even argued that peritoneal lavage may enhance spread of infection.

Postoperative management

The patient may require an intensive care management with pulmonary, renal and haemodynamic support. Intravenous fluids and antibiotic administration are continued postoperatively. A change of antibiotics based on the result of the specimen obtained during the operative treatment may be indicated. The vital signs, urine output, and blood gases are monitored closely. Also to be serially monitored are haemoglobin, electrolytes/ urea/creatinine. Paralytic ileus is a known complication of peritonitis and results in some degree of dysfunction of the gut after an operative management. In addition, sepsis increases the nutritional demands. The patient who is in a hypercatabolic state requires at least 25 – 35 kcal/kg/day. Nutritional support is therefore helpful in ensuring early recovery of the patient. The enteral route of alimentation is as effective as the parenteral route. In a very ill patient, however, the parenteral route may be preferred. A high protein and isocaloric diet is recommended.

Postoperative complications

- Surgical site infection: This could be a tell-tale sign of a residual intra-abdominal condition that may require re-operation
- Tertiary peritonitis
- Decompensation and even worsening of pre-existing organ dysfunction
- Iatrogenic infections: Urinary tract infection (2-8%), pneumonia (30%)
- Abdominal compartment syndrome: Intra-abdominal hypertension is due to an increased intra-abdominal volume coupled with a diminution in abdominal wall compliance. Limiting fluid administration, adequate analgesia and removal of constrictive dressings may help to increase the abdominal wall compliance. Secondary abdominal closure may be the only solution in severe cases

Tertiary peritonitis: As mentioned above, this is the intra-abdominal infection that persists or recurs after 48 hours of a supposedly successful and adequate surgical control of secondary peritonitis. The overwhelming risk factors for tertiary peritonitis as documented by the Mannheim Prognostic Index are

- Faecal peritonitis
- Age (>50)
- Female sex
- Malignancy
- Diffuse generalised peritonitis
- Organ failure.

C-reactive protein (Procalcitonin) is a recently-discovered parameter of infection and sepsis. A rising level after surgery is now regarded as a tell-tale evidence of recurrent infection. Its main advantage is the easy detectability and reproducibility.

SPECIAL FORMS OF PERITONITIS

Biliary peritonitis:

This connotes the spillage of bile into the peritoneum. The following are the predisposing factors

- Acute cholecystitis: Includes seepage of infected fluid from the wall of an inflamed gallbladder as well as rupture of a gangrenous gallbladder
- Postoperative leakage of bile: Cholecystectomy may be complicated by biliary peritonitis due to slipping of the ligature over the stump of the cystic duct. It may also be due to direct bile secretion through the accessory hepato-cholecystic ducts (ducts of Luschka).
- Traumatic rupture of the gallbladder or its ducts: May be due to either direct penetrating injury or iatrogenic (liver biopsy, percutaneous transhepatic cholangiography)
- Idiopathic

Clinical presentation: Patient presents with right hypochondrial pain. This may be preceded by clinical features of acute cholecystitis. Its occurrence during the postoperative period connotes an iatrogenic aetiology. Examination will reveal an ill-looking patient in obvious distress. There may be clinical features of hypovolemic shock. If massive, the accumulated bile may impinge on the inferior vena cava and reduce the venous return to the heart. The resultant diminution in cardiac output gives rise to hypotension and tachycardia. This phenomenon is referred to as the Walter-Walmann's syndrome. Abdominal examination will demonstrate clinical features of peritonitis.

Treatment: Initial resuscitation is in line with the general management of peritonitis. Definitive management involves exploratory laparotomy. The cause is identified and addressed at surgery.

Tuberculous peritonitis

There is a rising incidence of tuberculous peritonitis. This is attributed to the increasing incidence of the known predisposing conditions such as immunocompromised ailments (especially HIV/AIDS), liver disease and chronic kidney disease. The origin of tuberculous peritonitis is mostly from the mesenteric lymph nodes located around the ileocaecal region. Direct spread may also originate from tuberculous pyosalpinx while haematogenous spread may originate from pulmonary foci. It presents either in the acute or chronic forms. The various types of chronic presentation are

- Ascitic form: Patient presents with massive ascites
- Encysted form: Differential diagnoses include mesenteric cyst and ovarian cyst in the female
- Fibrous form: There is widespread adhesion leading to matting together of loops of intestine. This predisposes to subacute/acute intestinal obstruction and pockets of blind loop.
- Purulent form: This is usually secondary to tuberculous salpingitis. There may be pockets of pus amidst the mass of adherent intestines and omentum.

Clinical presentation: Diagnosis of tuberculous peritonitis requires a high index of suspicion particularly when one is confronted with a case of unexplained ascites. This is most important when dealing with high risk patients. In general, the acute variety will present with clinical

features of acute peritonitis, while the chronic form presents with abdominal swelling especially ascites (78.1%), fever and night sweats (53.8%), and anorexia (35.9%). Others are weight loss (44.1%) and abdominal pain (35.9%). It should be pointed out that the ascitic form may be painless. There may be an alteration in bowel habit with diarrhoea alternating with constipation. Some patients may present with faecal fistula. Characteristically, such fistulae prove difficult to heal. Tuberculous peritonitis is more common in men than women and is most frequently encountered in the 3rd and 4th decades of life.

Investigation of tuberculous peritonitis:

- Relatively simple and non-invasive tests such as culture and staining of the ascitic fluid for acid and alcohol fast bacilli are slow and associated with a low yield. The ascitic fluid is an exudate, pale yellow in colour, and with a specific gravity of at least 1020. It is unusually rich in lymphocytes and has a serum/ ascites albumin gradient (SAAG) of < 11 grams/litre. *Mycobacterium tuberculosis* is rarely cultured from the ascitic fluid except by guinea-pig inoculation.
- Chest X-ray: Only 38% give positive information
- Abdominal ultrasound scan and computerised tomography: It is more informative when both tests are combined. While the ultrasound scan outlines the thickened peritoneum and mesenteric lymphadenopathy, the CT scan will demonstrate the omental thickening. The fine, mobile septations in the peritoneum are best seen on the ultrasound scan
- Peritoneal biopsy: This is the most sensitive test. Specimens may be obtained by various methods. These are minilaparotomy, laparoscopy and image-guided percutaneous peritoneal biopsy. The latter method is most preferred.
- Other tests include retroviral screening, mantoux test (not quite informative in adults), erythrocyte sedimentation rate and full blood count

Treatment of tuberculous peritonitis: Diagnosis of the acute form may be made during laparotomy for acute peritonitis. The peritoneum will be found to be riddled with military tubercles. Treatment involves drainage of any ascitic fluid with a sample sent for microscopy, culture and sensitivity. Omental and peritoneal biopsies are taken and the abdomen is closed without drainage. The chronic variety may present with challenges in management due to gross abdominal distension from ascites and splinting of the diaphragm. This may be palliated by graduated ascitic tap coupled with adequate fluid replacement to prevent hypovolemic shock. Definitive treatment of all forms of tuberculous peritonitis is with combination anti-tuberculous chemotherapy. This comprises of rifampicin, isoniazid, ethambutol and pyrazinamide. Treatment is for at least six months

Subphrenic abscess

Subphrenic abscess connotes the presence of pus under the diaphragm. The subphrenic region lies between the diaphragm above and the transverse colon and mesocolon below. It has been divided into two anatomical entities: the subphrenic space proper (between the liver and the diaphragm) and the subhepatic space (under the liver). The falciform ligament of the liver separates the right and left subphrenic spaces. The right subhepatic space is referred to as

Morrison's pouch while the left is the lesser sac. The space between the bare area of the liver and the diaphragm is referred to as the right extraperitoneal space. The right side harbours most abscesses (about 70%). Bilateral abscesses are rare.

Aetiology of subphrenic abscess: Usually secondary to an intra-abdominal infective condition and may therefore result from any of the above-mentioned causes of secondary peritonitis. It can also occur as a complication following the operative management of any of these conditions. Most notable causes are peritonitis sequel to an upper abdominal condition such as perforated duodenal ulcer and postgastrectomy. Rarely, infection may spread through the haematogenous route or via a contiguous empyema thoracis.

Clinical features: Subphrenic abscess occurs days or weeks after managing a patient for secondary peritonitis. A high index of suspicion is necessary for its diagnosis as the initial symptoms may be vague and nonspecific

- 'Feeling unwell': Malaise, anorexia and loss of weight
- Fever: May present as pyrexia of unknown origin and hence the aphorism 'pus somewhere, pus nowhere else, pus under the diaphragm'. There is swinging temperature which usually commences from the tenth day after the aetiological infective condition.
- Right hypochondrial pain that may radiate to the right shoulder
- Chest symptoms: Right-sided chest pain, cough and dyspnoea due to direct spread of the infection to the right lower chest

Examination will reveal

- An ill-looking patient who may be pale and pyrexic
- Abdominal examination: Right hypochondrial tenderness. There may equally be a palpable right hypochondrial mass in late presentation
- Chest: Right lower chest tenderness with evidence of crepitations, pleural effusion and collapse of the base of the right lung
- CVS: There may be associated tachycardia and hypotension

Investigations

- Full blood count: The PCV may be low and there may be marked polymorphonuclear leucocytosis
- Chest X-ray: Best carried out under fluoroscopy. The following features are characteristic
 - Elevation of the right hemidiaphragm
 - Diminished/absent mobility of the diaphragm on fluoroscopic examination
 - Presence of an air-fluid level below the right diaphragm
 - 'Sympathetic' pleural effusion of the right base
 - Collapse of the base of the right lung

Other investigations are abdominal ultrasound scan and CT scan

It is important to point out that the apparently localised subphrenic abscess may rupture to involve other organs. This may result in empyema, lung abscess, and generalised peritonitis. Septicaemia may also occur.

Treatment: This is commenced with a course of antibiotics. This may aid resolution at the early stage of the infective process

There is no alternative, however, to drainage of an established subphrenic abscess. This is best carried out by either ultrasound or CT guided percutaneous drainage. Surgical drainage is indicated either on failure of guided percutaneous drainage or absence of relevant facilities for percutaneous drainage. The ideal route of surgical access is extraperitoneal depending on the location of the abscess. An anterior subphrenic abscess is drained via a subcostal incision while its posterior counterpart is drained through the bed of the 12th rib.

Pelvic abscess

Just like its subphrenic counterpart, it is usually sequel to generalised peritonitis. The most common predisposing conditions are gangrenous/perforated appendicitis and perforated peptic ulceration (due to percolation of gastric/biliary fluid through the right paracolic gutter into the right iliac fossa). Pelvic infection in females, notably primary peritonitis and septic abortion, are also common predisposing factors. In the male, the abscess is located between the urinary bladder and the rectum while the pouch of Douglas (space between the uterus and the posterior fornix of the vagina anteriorly and the rectum posteriorly) is the seat of the abscess in the female. It may rupture and spread to other structures if not promptly managed. It may rupture posteriorly into the rectum or into the general peritoneum and may equally discharge through an unhealed laparotomy wound.

Clinical features: There is usually a preceding history of an intra-abdominal infective process. It is usually associated with swinging temperature, anorexia, loss of weight and mucoid diarrhoea. Examination will reveal an ill-looking patient with suprapubic tenderness. There may be a palpable lower abdominal mass. Rectal examination will reveal the presence of a boggy, tender mass in the anterior rectal wall.

Investigations: These include FBC (low PCV and polymorphonuclear leucocytosis) and more specifically, an abdominal ultrasound scan. CT scan will help to further delineate the mass.

Treatment: May respond to a course of antibiotics in the early stage. Drainage is of essence once pus has formed. The transrectal approach is the route of choice. Spontaneous rupture through the rectum is a common occurrence. This may equally be achieved in the established case by a firm pressure over the anterior wall of the rectum.

CHAPTER TWENTY TWO

HERNIAS

A hernia is defined as the protrusion of a viscus or a part of it through an abnormal opening in the wall of its containing cavity. Generally speaking, there are two broad groups of hernia: internal and external. Internal hernias occur within the body and include paraoesophageal hernias. External hernias, on the other hand, are found externally particularly around the groin. They constitute a considerable part of the workload of a general surgeon. Approximately 75% of all abdominal wall hernias occur around the groin.

Common hernias include

- Inguinal hernias (direct and indirect): Arise from the inguinal canal
- Femoral hernias: Arise from the femoral canal
- Epigastric hernias: Arise from defects in the linea alba
- Umbilical hernias: Arise from the umbilical defect
- Incisional hernias: Arise from previous surgical scars

By and large, inguinal, femoral and umbilical hernias are the commonest external hernias and account for 75%, 8.5% and 15% of all hernias respectively.

Uncommon hernias include Spigelian, lumbar, gluteal and obturator varieties

Aetiology of hernias

- Congenital: The origin of the indirect inguinal hernia is normally traced to the presence of a patent processus vaginalis. This accounts not just for congenital hernias but also for congenital hydrocoelous in children. It may also be associated with congenital anomalies such as cryptorchid testis, hypospadias, epispadias and extrophy of the bladder
- Circumstances surrounding birth: Meconium peritonitis, prematurity and low birth weight
- Weakness of the abdominal wall as a result of sustained raised intraabdominal pressure. Involuntary causes include chronic cough (bronchitis, tuberculosis), straining at micturition (prostatic enlargement, urethral stricture), chronic constipation (inappropriate diet, colonic tumours) and pregnancy. Voluntary causes include lifting of heavy loads as found in farmers, mechanics and weight lifters. Some clinical conditions which result in an increase in the intraabdominal pressure also predispose to hernia formation. They include ascites from any cause (especially liver disease), continuous ambulatory peritoneal dialysis, and ventriculoperitoneal shunts
- Issues arising from previous surgeries: As in incisional hernias (see below)
- Experimental evidence suggests that collagen derangement may be an aetiological factor in the pathogenesis of inguinal hernia

General characteristics of a hernia

- The uncomplicated hernia is reducible and exhibits a positive cough impulse (visible and/or palpable)
- A hernia consists of the hernia sac, coverings of the sac and contents of the sac
- The hernial sac consists of a mouth, proximal neck, body, and fundus
- Contents of a hernia sac: These may include intestine (enterocoele), omentum (omentocoele), a portion of the circumference of gut (Richter's hernia) and the appendix. Others are sigmoid colon, urinary bladder, Meckel's diverticulum (Littre's hernia), fallopian tube and fluid (secondary to ascites)
- Complications of a hernia include irreducibility or incarceration due to adhesion formation between the hernia sac and its contents. There is, however, no clinical evidence of intestinal obstruction when a hernia is irreducible. Others are obstruction (a common cause of intestinal obstruction), which may proceed to strangulation (compromise of the blood supply to the gut) if unrelieved.
- Hernioplasty: This involves hernia repair by reinforcement with either a natural material such as fascia lata or a synthetic substance such as prosthetic mesh

Diagnosis of hernias: This is mainly clinical. Herniography is rarely indicated in obscure cases especially in the obese. A radio-opaque dye instilled in the peritoneal cavity will define the hernia sac either on fluoroscopy or by a plain abdominal X-ray. It may also aid in the diagnosis of recurrent hernia whose clinical manifestation may be marred by fibrosis. An occult contralateral hernia may be revealed by herniography.

Nyhus classification of groin hernias

- Type 1: Indirect hernia with normal internal abdominal ring (infants, children, and young adults)
- Type 2: Indirect hernia in which the internal ring is enlarged (patulous) but without impingement on the floor of the inguinal canal. Hernia does not extend into the scrotum
- Type 3A: Direct hernia; size is not taken into consideration
- Type 3B: Indirect hernia that has enlarged enough to encroach upon the posterior inguinal wall. Includes sliding and scrotal hernias
- Type 3C: Femoral hernia
- Type 4: Recurrent hernia: Modifiers A,B,C, and D are sometimes added to Type 4 corresponding to indirect, direct, femoral and mixed respectively.

Inguinal hernias

They arise within the inguinal canal. For a good appreciation of the pathophysiology, it is pertinent to review the anatomy of the region

The inguinal canal is a channel in the groin that runs between two rings: the internal and external rings. It has the following boundaries

- Anteriorly: The skin and the external oblique aponeurosis; the lateral one-third is strengthened by the internal oblique as it arises from the inguinal ligament

- Posteriorly: The transversalis fascia and the conjoint tendon. The conjoint tendon consists of the common insertion of the internal oblique and the transversus muscles into the pubic tubercle.
- Roof: The arching fibers of the internal oblique and transversus muscles
- Floor: The inguinal ligament which runs between the anterior superior iliac spine and the pubic tubercle

The canal transmits the spermatic cord and the ilioinguinal nerve in the male. In the female, the cord is replaced by the round ligament. The spermatic cord is composed of the following

- Three coverings: external spermatic fascia (from the external oblique), cremasteric fascia (from the internal oblique), and the internal spermatic fascia (from the transversalis fascia). The coverings of an inguinal hernia are direct derivations of the coverings of the spermatic cord
- The vas deferens
- Three arteries: testicular artery, cremasteric artery, and the artery to the vas deferens
- Others are: pampiniform plexus of veins, sympathetic nerves, and lymphatics

The Hasselbeck's triangle is bounded inferiorly by the inguinal ligament, laterally by the inferior epigastric artery (which is medial to the deep inguinal ring), and medially by the lateral border of the rectus abdominis muscle. Its importance is two-fold. Firstly, peritoneum and transversalis fascia are the only components of the anterior abdominal wall in this triangle. This renders it a weak spot and accounts for its vulnerability to the development of acquired hernia (direct inguinal hernia). Secondly, it is the anatomical landmark of differentiation between direct and indirect inguinal herniae at surgery. Whereas the indirect hernia exits from the deep ring which is lateral to the inferior epigastric artery, the direct hernia is medial to it as it originates from a defect in the Hasselbeck's triangle. The anatomical relationship between the spermatic cord and the hernia sac equally differs in both forms of inguinal hernia. Whereas the sac of a direct inguinal hernia is posteromedial to the spermatic cord, that of an indirect hernia is anterolateral to it. The clinical differentiation between direct and indirect inguinal hernia is equally based on this anatomical relationship. The hernia is reduced with the patient in the recumbent position. This is followed by identification and occlusion of the deep inguinal ring which is located 1.5 cm above the midpoint of an imaginary line that runs between the pubic tubercle and the anterior superior iliac spine (mid-inguinal point). On reduction of the hernia and occlusion of the deep ring, the patient is asked to cough. The result is that a cough impulse will still be positive in direct inguinal hernia (does not exit from the deep ring) but negative in indirect inguinal hernia since the deep ring is its exit point. The exception to this rule is the so called pantaloan or saddle-bag hernia which has both lateral and medial components like the two legs of a pair of trousers. It must be pointed out, however, that the ultimate mode of differentiating between these varieties of inguinal hernia is at surgery and is hinged on the relationship of the neck of the hernia to the inferior epigastric artery. The neck of indirect hernia is lateral to the inferior epigastric artery while that of direct hernia is medial to it.

Clinical features: On the basis of age of occurrence, inguinal hernia can be divided into two clinical types

- Inguinal hernia in infants and children
- Adult inguinal hernias

Adult inguinal hernias

They are commoner in males than females (10:1) with a peak age incidence in the sixth decade. The indirect variety is commoner and accounts for 65% of all inguinal hernias. It is slightly commoner on the right (55%) than the left. Bilateral inguinal hernia is four times more common in direct than indirect hernias. Inguinal hernias may be classified based on the location of the hernial fundus into complete and incomplete

- Complete: Fundus reaches the bottom of the scrotum with difficulty in palpating the testis separate from the hernia
- Incomplete: Fundus falls short of the bottom of the scrotum. The incomplete inguinal hernia is further subdivided into bubonocoele and funicular
 - Bubonocoele: hernia is confined to the inguinal canal; does not get beyond the superficial ring
 - Funicular: Fundus of hernia lies between the superficial inguinal ring and the upper pole of the testis

The relevant clinical history will comprise of history of the hernia, its aetiology and complications (if any)

- Duration, site, progression, reducibility,
- Aetiology: congenital; presence of any condition that raises the intra-abdominal pressure
- Complications: irreducibility, and obstruction (pain and vomiting)

Examination involves a full clinical examination to identify any of the above-mentioned aetiological factors. Thorough examination of the chest and abdomen (including a rectal examination) should be carried out. Examination of the groin should begin with the patient in the standing position.

- Identify the swelling and elicit a cough impulse (visible and palpable)
- For a scrotal swelling, try to get above it by palpating for fullness or otherwise at the neck of the scrotum. You will be able to get above a purely intrascrotal swelling. The reverse is the case for an inguinoscrotal swelling (hernia).
- Patient now lies down and a gentle attempt is made at reducing the hernia. The patient may be requested to help accomplish this if any difficulty is encountered.
- Palpate for the pubic tubercle. This is a landmark that helps to differentiate the inguinal from the femoral hernia. When the hernia is reduced and the patient is made to cough, the hernia is found to exit superior and medial to the pubic tubercle in the case of inguinal hernia but inferior and lateral in a femoral hernia.
- The deep inguinal ring is identified as described earlier and occluded
- A cough impulse is then attempted with the deep ring still occluded. As mentioned above, it will remain positive in direct inguinal hernia but negative in indirect inguinal hernia. The cough impulse is restored in the indirect inguinal hernia with the release of the finger from the deep ring. It is worth reiterating, however, that the ultimate differentiation between the direct and indirect inguinal hernia is at surgery and is based on the relationship of the neck of the hernia to the inferior epigastric vessels.

Differential diagnosis of a groin swelling: As usual, relate this to the local structures in the groin

- Skin: Lipoma
- Canals: Inguinal and femoral hernias; hydrocoele of the cord or canal of Nuck
- Lymph nodes: Lymphadenopathy
- Blood vessels: Femoral aneurysm, saphena varix
- Testis: Ectopic testis, undescended inguinal testis
- Muscle: Psoas bursa or abscess

Investigation: The diagnosis of inguinal hernia, like all hernias, is essentially clinical. Investigation is aimed at further confirming any predisposing factor that may have been identified during the clinical evaluation. Relevant investigations are also carried out as an integral part of the preoperative workup. It includes the following

- Chest X-ray: Quite relevant in the investigation of a patient with chronic cough. It is also an integral part of the preoperative evaluation of any patient above the age of 40
- Investigation of the lower gastrointestinal and urinary systems if suggested by the clinical assessment
- Packed cell volume
- Serum electrolytes, urea and creatinine particularly if there is associated vomiting from obstruction
- Urinalysis

Treatment: This should take into consideration the size of the hernia, the age of the patient as well as comorbid factors such as diabetes mellitus. For example, a small hernia in an old, frail diabetic may be managed by just reassurance and observation. Any sign of complication such as irreducibility and impending obstruction will, however, require surgical treatment.

There are two forms of hernia repair: Open repair and laparoscopic repair. Open repair, as the name implies, entails the use of open surgical incision and dissection. Laparoscopic repair, on the other hand, is a 'closed' method that involves the use of a laparoscope. In addition to better cosmetic outcome, the recovery rate is faster with laparoscopic repair compared to open repair.

Open surgical repair: The type of surgery carried out depends on the age of the patient. Any underlying precipitating factor such as chronic cough, constipation or bladder outlet obstruction should be addressed prior to surgery in order to forestall recurrence. For example, prostatic enlargement should be addressed prior to herniorrhaphy. In practice, however, both problems are usually tackled at the same surgical sitting

Treatment: In adults, the inguinal canal is well formed. There is also a degree of weakness of the posterior wall of the inguinal canal. In addition to herniotomy (excision of the hernia sac), the posterior wall is further strengthened by way of a reconstructive procedure. In herniorrhaphy, this is achieved by the use of a monofilament, non-absorbable suture such as nylon. The main drawback to this procedure is the resultant increased tension in the site of repair. This is an established cause of recurrence. The problem may, however, be overcome, by bridging the gap between the structures to be apposed with the use of a synthetic mesh. Reconstruction with the use of a synthetic mesh is referred to as hernioplasty.

Methods of open surgical repair of inguinal hernia

- Bassini repair: This is the original method of repair. It involves approximation of the conjoined tendon to the lower edge of the inguinal ligament by means of a non-absorbable suture such as monofilament nylon. The tension over the repair may be relieved to an extent by an incision over the anterior rectus sheath (Tanner's slide). Bassini repair is no longer as popular as it used to be due to the relatively high recurrence rate engendered by the tension at the site of repair
- Macvay (Lothiessan's or Cooper's) repair: After a formal herniotomy, the Cooper's ligament is dissected by dividing the iliopubic tract. A series of sutures are used to appose the transversalis fascia and the transversus aponeurosis on one hand and the Cooper's ligament on the other. This procedure commences at the pubic tubercle and terminates at the medial edge of the femoral vein. It is not a popular procedure.
- Shouldice repair: This involves a careful dissection and double-breasting of the layers of the canal beginning with the transversalis fascia. It is believed to have a lower recurrence rate when compared to the Bassini repair
- Lichtenstein's tension-free repair: This involves the use of prosthetic mesh in order to accomplish a tension-free hernia repair. The gap between the conjoint tendon and the inguinal ligament is bridged by the use of a prosthetic mesh. It is a tension-free method of repair. The relatively low recurrence rate associated with mesh repair has made it the current gold standard in the repair of adult inguinal hernia.

Special types of hernia

- Sliding hernia: A part of the sac of the hernia is formed by a viscus. It may involve the caecum in a right-sided hernia, or the sigmoid colon in a left-sided inguinal hernia. The urinary bladder may form part of the hernia sac on either side of the groin. The importance lies in the early identification of this state of affairs in order to prevent injury to the associated structure. Repair of sliding hernia entails excision of the part of the hernia sac distal to the viscus, reperitonising and reducing the viscus in question back into the peritoneal cavity prior to repair of the posterior wall of the inguinal canal..
- Richter's hernia: One sidewall of the intestine is trapped in the hernia. There is no appreciable clinical manifestation of intestinal obstruction even when the affected part is gangrenous. Rather than being constipated in the presence of intestinal obstruction, patient may even have diarrhoea owing to the partial patency of the lumen of the gut. Richter's hernia can pose a diagnostic dilemma
- Littré's hernia: Involves Meckel's diverticulum
- Maydl's hernia: This involves two adjacent loops of bowel with an intervening, intra-abdominal segment of gut. The latter will be the first casualty in the event of obstruction of the hernia
- Pantaloons hernia: The single hernia sac is split like the legs of a pair of trousers which straddle the inferior epigastric vessels. It conveys the impression of a combination of both direct and indirect hernia.

- **Spigelian hernia:** Occurs at the lower limit of the posterior rectus sheath (semilunar line) just lateral to the rectus sheath
- **Obturator hernia:** More common in multiparous women with a history of recent weight loss. Herniation is through the obturator canal alongside the obturator vessels and nerves. Demonstration of this hernia is by the Howship-Romberg sign. When the hip is flexed, externally rotated and abducted, the hernia comes up as a palpable mass in the medial aspect of the thigh.

Laparoscopic repair:

Much as laparoscopic repair may be employed in the management of any form of inguinal hernia, it is, however, best suited for the following

- **Recurrent hernias:** Repair by the open method is difficult owing to fibrosis and haemorrhage. The preperitoneal space is, however, relatively intact. This makes it easy to effect a mesh repair of the hernia
- **Bilateral hernias:** Both sides are repaired with no additional stress on both the surgeon and the patient
- A clinically occult hernia detected at surgery may be repaired at the same operative sitting without additional stress.

Procedures for laparoscopic inguinal hernia repair: Like open repair, laparoscopic hernia repair entails reduction or excision of the sac followed by insertion of a mesh in the preperitoneal space. There are three laparoscopic methods of repair of inguinal hernia:

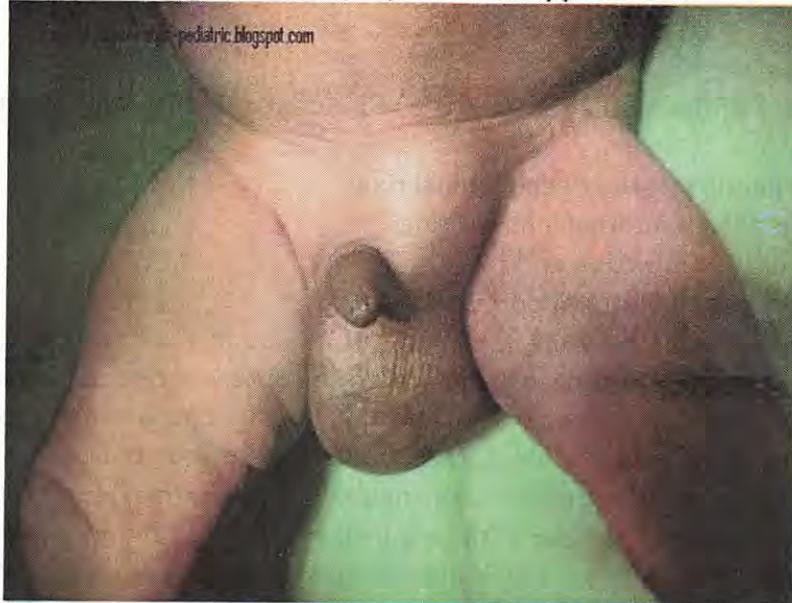
- **Intraperitoneal repair:** The prosthetic mesh is placed as an inlay graft within the peritoneal cavity. Compared to other approaches, it is less time-consuming and requires less dissection of the preperitoneal space. The main disadvantage is the exposure of the prosthetic material to the peritoneal cavity. This may result in formation of adhesions and intestinal obstruction. It is also associated with a high recurrence rate.
- **Transabdominal preperitoneal repair (TAPP):** The peritoneum is dissected and a mesh is inserted in the preperitoneal space. Thereafter, the peritoneum is closed over the mesh with staples or sutures. It permits inspection of the opposite groin and so bilateral hernia repairs can be carried out if indicated. It, however, involves a wider dissection and a greater chance of injury to the intraperitoneal structures. Furthermore, the peritoneal incision increases the potential for adhesion formation and intestinal obstruction
- **Totally extraperitoneal prosthetic repair (TEP):** This involves the insertion of a laparoscope and inflation of a balloon in the extraperitoneal space between the abdominal wall and the peritoneum through a paraumbilical incision. A mesh is inserted in the space so created. The main advantage is the avoidance of the peritoneal cavity and subsequent intraperitoneal adhesion formation. It is, however, more tedious and time consuming as the operating space is more limited when compared to trans-abdominal procedures.

Inguinal hernia in infants and children

Management of congenital inguinal hernia constitutes a substantial part of the workload of the paediatric surgeon. Clinical inguinal hernia is diagnosed in about 3 to 5% of full-term infants with about one third presenting in the first 6 months of life. The incidence is much higher in pre-term infants (10%). It is slightly more common on the right (60%) while 15% have bilateral presentation.

Clinical presentation: Congenital inguinal hernia presents as an intermittent bulge in the groin or scrotum which is more prominent on severe straining such as crying and defaecation and reduces spontaneously when the child is relaxed. An older child is examined in both supine and upright positions. It is important to examine the scrotum in order to confirm the presence of the testis. Absence of testis on the ipsilateral side will raise suspicion of either ectopic or undescended testis. The latter is usually associated with a congenital inguinal hernia. Both conditions are addressed in one composite surgical procedure. There is a high risk of incarceration, obstruction and strangulation of inguinal hernia in the paediatric age group. This warrants early treatment of inguinal hernia in children.

Treatment of paediatric inguinal hernia: Surgical repair is usually carried out as a day-case procedure under an ambient environment. The hernia in this case is usually of the indirect variety and is due to persistence of the processus vaginalis. In this age group, the inguinal canal is not quite formed, with both rings of the inguinal canal almost apposed to each other. Surgery in children involves simple excision of the hernia sac. This is referred to as herniotomy. Laparoscopic hernia repair has no role in children. Adolescents with recurrent hernia may, however, benefit from a laparoscopic repair through a preperitoneal approach



CONGENITAL INGUINAL HERNIA

Complications of hernia repair: Classified into general and local complications respectively

General complications

- Pulmonary: Atelectasis, pneumonia and pulmonary embolism
- Urinary retention: May occur in a patient with prostatic enlargement. The latter may have been asymptomatic prior to surgery. Overzealous fluid administration may result in diuresis and atony of the overfilled bladder
- Complications of regional anaesthesia

Local complications

- Haemorrhage and haematoma formation particularly following repair of large hernias
- Infection particularly when mesh repair is carried out. Prophylaxis may be carried out with a single perioperative dose of a broad spectrum antibiotic. Occasionally stitch abscess may occur from a subcutaneous stitch. Incidence of wound infection is 1 to 5%
- Testicular infarction: Ischemic orchitis is due to a combination of damage to the testicular artery and venous congestion from compression of the pampiniform plexus of veins
- Persistent groin pain: May be due to damage to the ilioinguinal, iliohypogastric or genital branch of genitofemoral nerves
- Injury to the intestine and urinary bladder especially when dealing with an unexpected case of sliding hernia
- Injury to the vas deferens
- Recurrence: This is highest with Bassini repair and lowest with laparoscopic approach (intraperitoneal 3.2%, TAPP 0.8% and TEP 0.0%). The following are the risk factors for recurrence:
 - A missed hernial sac
 - Low 'amputation' of the hernia sac leaving a redundant cuff below the deep inguinal ring
 - Broken suture ligature at the deep inguinal ring.
 - Failure to repair a large internal inguinal ring,
 - Severe postoperative infection and
 - Sustained increase in the intra-abdominal pressure.Prosthetic mesh repair is now the technique of choice in the repair of recurrent hernias. The preperitoneal approach is the preferred route when repair is carried out laparoscopically. It can be surmised that the most important cause of early recurrence is technical failure while tissue failure may account for late recurrence
- Scrotal haematoma: More common with inguinoscrotal hernia repair. It can be prevented by transection of the sac at the inguinal canal rather than tracing it all the way to the scrotum. The surgeon should not hesitate to put in a drain if necessary.
- Postoperative hydrocele
- Penile oedema: may be due to injury to the superficial external pudendal vein

Femoral hernia

It constitutes about 5% of all hernias and exits through the femoral canal. It is commoner in females (85%) than in males. Inguinal hernia, however, still remains the commonest hernia in females. The importance lies in the fact that of all hernias, it is the most prone to obstruction.

Anatomy of the femoral canal: The femoral canal is approximately 1.5 cm in length and stretches between the femoral ring above and the saphenous opening below. It barely admits the tip of the little finger. The femoral canal is larger in females because of the wider width of the pelvis. This accounts for the higher occurrence of femoral hernia in females. Apart from its lateral boundary, the femoral ring is bounded by rigid structures

- Laterally is the femoral vein
- Medially is the lacunar ligament (Gimbernat's ligament): This is the sharp free edge of the pectineal part of the inguinal ligament
- Anteriorly is the inguinal ligament
- Posteriorly is the pectineal ligament (of Astley Cooper). It is the thickened portion of the periosteum along the pectineal border of the superior pubic ramus and is continuous medially with the pectineal part of the inguinal ligament.

The femoral canal usually contains a plug of fat, lymphatic vessels and a lymph node referred to as Cloquet's node or the node of the femoral canal and plays a dual role: Firstly, it is a potential dead space for expansion of a distended femoral vein and secondly it acts as a lymphatic pathway for lymphatic fluid from the lower limb to the external iliac lymph nodes

Clinical features: As mentioned above, femoral hernia is commoner in females. The swelling may be barely visible and palpable and may project above the inguinal ligament thereby raising a diagnostic dilemma as it is difficult to differentiate this from an inguinal hernia. The pubic tubercle is the landmark of diagnosis of a femoral hernia. On cough impulse, the neck of a femoral hernia lies below and lateral to the pubic tubercle. This differs from the sac of an inguinal hernia that has a superior-medial relationship to the pubic tubercle.

Surgical treatment of femoral hernia: Traditionally, there are three approaches to the open procedure. There is currently a laparoscopic approach. The open approaches are

- Lockwood's approach: This is a direct approach through a transverse crease incision that is inferior and parallel to the inguinal ligament. After dealing with the hernia, repair is carried out by approximating the inguinal ligament to the iliopectineal ligament. It is a direct approach to the sac and is the approach of choice in uncomplicated femoral hernia. The main disadvantage is the tension that results from approximation of two tendinous structures. This may predispose to recurrence. Recurrence may be prevented by inserting a plug or cylinder of polypropylene mesh into the canal and tagging it to the inguinal ligament anteriorly and the iliopectineal ligament posteriorly. Access may be improved by dividing the inguinal ligament which can be repaired later. This manoeuvre comes to bear when surgery is carried out for obstructed femoral hernia.
- Lotheissen's approach: Through an inguinal incision, the sac is dissected from above. It was initially considered a good approach particularly when dealing with an

obstructed femoral hernia. The drawbacks, however, are that it is a more elaborate procedure than the Lockwood's approach and involves going through the inguinal canal which gets weakened in the process. Owing to this inherent disadvantage, it has fallen out of favour.

- **MacEvedy's approach:** This involves tackling the sac through an abdominal incision that is vertical to the femoral canal and continued upwards above the inguinal ligament. It is said to be quite useful particularly when dealing with a strangulated hernia. It is not commonly used as it entails a lot of dissection.
- **Laparoscopic approach:** Same approach (TAPP and TEP) as inguinal repair. The myopectineal opening is fully visualised

Ventral hernias: This is a blanket term that embraces all the anterior abdominal wall hernias and includes umbilical, paraumbilical, epigastric and incisional hernias. Apart from the latter which is iatrogenic, the others involve a congenital or acquired defect through the abdominal wall. Obesity and conditions that increase the intra-abdominal pressure may be predisposing factors. Management, as usual, involves addressing any identified predisposing factor followed by hernia repair. This is exemplified by the management of incisional hernias as discussed below.

Incisional hernia

It is a ventral abdominal hernia that occurs over the scar of a previous surgical operation. All the deep layers of the anterior abdominal wall would have given way. The skin therefore constitutes the only covering over the scar. It may be regarded as a complication of wound failure and diagnosis is quite straight forward. There is a history of previous surgical operation which is subsequently followed by development of a reducible protrusion over the surgical scar. Cough impulse is positive and introduction of the finger(s) into the defect will help to determine its size. The latter is an important deciding factor as regards the method of repair

Aetiology of incisional hernia: It is basically due to poor wound healing which results in weakness of the surgical scar. The aetiology may be divided into preoperative, operative and postoperative factors

Pre-operative factors: These include morbid obesity, conditions that give rise to raised intraabdominal pressure (chronic cough, constipation, and urinary obstruction), anaemia and cancer. Other factors are those that adversely affect wound healing such as administration of steroids, chemotherapy and jaundice. Advanced age is known to negatively affect wound healing.

Operative factors: Poor surgical technique, inadequate haemostasis resulting in haematoma formation and inappropriate use of absorbable sutures. No appreciable difference has, however, been found between continuous and interrupted suture closure in the pathogenesis of incisional hernia. The suture length (SL) versus wound length (WL) ratio has been examined. It has been shown that a SL:WL ratio of 4 or more is associated with a low incidence of incisional hernia while a lower ratio may be associated with a threefold increase. Ironically, any ratio

higher than 5 is associated with a higher rate of wound infection and incisional hernia. This may be due to incorporation of a large chunk of tissue into the suture.

Postoperative factors: Raised intra-abdominal pressure (postoperative ileus, vomiting, and prolonged coughing), wound infection, and any unresolved preoperative factor.

Treatment: Small asymptomatic hernias may require reassurance as the outcome of surgery is unpredictable. Recurrence rate of 30-50% has been reported in some series. It is therefore best prevented by addressing the above predisposing factors prior to any elective abdominal procedure. This is equally relevant prior to an elective repair of an incisional hernia. Weight reduction in an obese patient is mandatory. The services of a dietician may be necessary. Chronic bronchitis and other chest conditions should be addressed. A known smoker should be advised to stop smoking for at least four weeks prior to an elective repair. It is advisable to delay surgical repair for at least three months from the onset of symptoms. This is to enable the inflammation and fibrosis resulting from the initial surgical operation to subside before effecting repair. Surgical repair is strengthened as the dissection and approximation of tissue planes are made easier.

Surgery: Aims at bridging the gap between the tissues. An elliptical skin incision is made over the previous scar with the aim of excising it. Care should be taken to prevent damage to the underlying viscera which lie directly under the skin layer. This is followed by teasing of the viscera off the hernial wall by means of both blunt and sharp dissection. A careful layered dissection of the abdominal wall is then carried out

For a long time, the Mayo's method of repair was mostly employed in the surgical treatment of incisional hernia. This entails a careful layered dissection of the anterior abdominal wall over the hernia followed by double-breasting of the rectus abdominis aponeurosis in the 'pant over vest' manner. The high recurrence rate (over 30%), particularly with large defects, has rendered this method of repair less popular. It is, however, still relevant in the repair of small hernial defects. Other methods of repair such as direct suturing and Keel's repair (use of successive lines of non-absorbable sutures) are effective when applied to hernias with small defects (< 2 cm) but have proved less effective in the management of those with wider defects. The latter are best managed by the use of prosthetic mesh.

Mesh repair of incisional hernia: This is now considered the most effective method of repair of moderate to large incisional hernias. Polypropylene mesh is used to bridge the gap. The defect is repaired with less tension thereby reducing the chances of recurrence. In the inlay method, the mesh is applied posterior to the peritoneum and directly on the abdominal viscera. In the onlay method, on the other hand, the mesh is applied anterior to the peritoneal layer. The overlay method involves the application of the mesh over the musculoaponeurotic layer. Of these methods, the onlay method is the most preferred. Unlike the inlay method that predisposes to the formation of adhesions, bands and fistulation, the onlay method is free of these since it has no direct contact with the abdominal viscera. The stability of the mesh is also more secure in the onlay method as it is naturally fixed by the intra-abdominal pressure. The mesh seals the defect mechanically and also induces the formation of scar tissue of appreciable strength. Efforts should be made to prevent infection during mesh repair by ensuring the sterility of the mesh and also by the administration of intravenous prophylactic antibiotics.

Laparoscopic repair may also be employed in the repair of ventral hernias. A mesh is inserted over the defect from within the abdomen. Its advantages include better cosmesis, low complication and faster recovery rates.

Umbilical and paraumbilical hernias:

Congenital umbilical hernia: This paediatric surgical emergency is also referred to as omphalocoele. It results from herniation of the abdominal viscera through the tissue of the umbilical cord. The umbilical viscera are covered by amniotic membrane.

Infantile umbilical hernia: It is due to failure of the umbilical vessels to fuse with the urachal remnant and the umbilical ring. The hernia presents with a protrusion of the umbilicus. Unlike the congenital variety, however, it is always covered with skin..The incidence is about one in five births and has been found to be commoner in blacks. There may be a familial predisposition. The diagnosis is clinically obvious. It rarely enlarges over time; rather spontaneous resolution occurs in about 90% of cases by the age of two. Those that fail to resolve at pre-school/nursery are candidates for surgical management. The approach is via a curvilinear incision made at the inferior aspect of the hernia. The hernia defect is repaired after excision of the sac by either simple apposition of the fascia or by Mayo's repair ('vest-over-pants'). Since it is a congenital defect, repair can be carried out with absorbable suture material. The skin is closed after securing adequate haemostasis.Umbilical hernioplasty rarely has attendant complications. The latter include infection and haematoma/seroma formation. Recurrence may occur if there is tension associated with repair of large defects or if there is an undiagnosed paraumbilical hernia.

Adult umbilical hernia: Unlike infantile umbilical hernia, the adult variety is a spectrum from the partially unfolded cicatrix to indirect herniations through the umbilical cord. The umbilical cicatrix may give way in conditions associated with persistent elevation of the intra-abdominal pressure such as ascites from any cause (congestive cardiac failure, cirrhosis of the liver and renal pathology). True herniation occurs through the umbilical canal. The latter is bounded anteriorly by the linea alba, posteriorly by the umbilical fascia and the medial edges of the two rectus sheaths on either side. Adult umbilical hernias are prone to obstruction and strangulation. They are more common in obese and multiparous women and present with an umbilical protrusion that may be difficult to reduce. Association with intermittent colicky abdominal pain may be as a result of incomplete intestinal obstruction. Umbilical hernias due to the medical conditions mentioned above do not require surgery except when complicated by incarceration. Surgery for true umbilical hernia is carried out under local or regional anaesthesia. Small hernias (less than 4 cm diameter) are managed by simple repair with nonabsorbable suture. Larger hernias (more than 4 cm diameter) require reinforcement with extraperitoneal prosthetic mesh. The patient should be forewarned about the possibility of excising the umbilicus in the course of hernia repair. Complications of surgery include infection and haematoma/seroma. Cardiovascular and respiratory complications may also occur with the return to the peritoneal cavity of the contents of large umbilical hernias.

Paraumbilical hernia: They occur secondary to dehiscence of the linea alba. Most cases occur in the supraumbilical position. It is commoner in females than males (female to male ratio is 5:1).

This is attributable to obesity and pregnancy. The patient presents with a swelling that is either above or below the umbilicus. There may be associated intermittent abdominal pain due to dragging of the contents culminating in intermittent intestinal obstruction. It should be differentiated clinically from umbilical hernia. The skin over a paraumbilical hernia is wholly or partially covered by skin of the anterior abdominal wall. It should be noted that both types of hernia may coexist. Whereas repair of a solitary paraumbilical hernia may be approached via a transverse incision, a midline approach is preferred in the compound variety. Being an acquired condition, repair is carried out with nonabsorbable suture material.

Epigastric hernia: Occurs due to a fascial defect in the linea alba between the xiphoid process and the umbilicus. There is a male preponderance (male to female ratio of 4:1). Majority of patients are asymptomatic. Symptoms include vague epigastric pain and nausea. These, coupled with epigastric tenderness on examination raises a differential diagnosis of peptic ulcer disease. Diagnosis is made by palpation of a midline mass. The latter may not be palpable in obese patients. Diagnosis in such cases is confirmed with the aid of ultrasound or CT scan. Surgical repair of the symptomatic case is carried out through either a transverse or vertical incision over the mass. The latter is preferred when dealing with multiple defects or when strangulation is suspected. Lesions less than 4 cm in diameter are repaired with simple suturing with nonabsorbable suture material while those measuring 4 cm and above require reinforcement with prosthetic mesh. Complications are rare and include infection and haematoma/seroma.

Management of complicated hernia

As mentioned earlier, the complications of a hernia include irreducibility, obstruction and strangulation. The latter is sequel to an already existing obstruction and is said to occur when the blood supply to the segment of the entrapped viscus is compromised. It is associated with high morbidity and mortality.

Obstructed hernia: The hernia suddenly becomes irreducible, painful, with associated clinical features of intestinal obstruction such as abdominal pain and vomiting. If unrelieved, the abdominal pain could become generalised. Examination reveals an irreducible, tender swelling with negative cough impulse. It is important to institute emergency care urgently in order to prevent strangulation. This is necessary because there are no clear-cut clinical features that herald the onset of strangulation. Management of obstructed hernia involves the following:

- Good history and clinical examination
- Set up an intravenous infusion to replace the fluid and electrolyte loss.
- Take blood samples for electrolytes, urea and creatinine while setting up the infusion
- Commence on nil orally
- Nasogastric aspiration to decompress the gastro-intestinal system
- If the hernia is adjudged to have developed a sudden state of irreducibility but without clinical evidence of obstruction, attempt should be made to achieve reduction. This is accomplished by administering a strong analgesic and sedating the patient with diazepam. These, coupled with elevation of the foot of the bed, may result in spontaneous reduction of the hernia. Surgery in this case could then be elective, preferably in the next elective list. Late presentation with clinical evidence of

obstruction, however, requires emergency exploration after adequate resuscitation as there is associated risk of strangulation.

The aim of surgery is three-fold: relieve the obstruction, ascertain the viability of the gut and repair the hernia. The incision employed is as for the routine surgical management of the hernia in question. For the inguinoscrotal hernia, however, a more oblique incision that curves into the scrotum (the so called 'hockey-stick' incision) is employed. For the obstructed femoral hernia, the low or McEvedy's approach is employed. As earlier mentioned, access may be improved in the low approach by division of the inguinal ligament which is reconstituted after dealing with the obstructed hernia. The fluid in the hernia sac should be carefully sucked out to prevent tissue contamination as it is naturally riddled with a high bacterial load. Care is taken to ensure that the involved segment of gut is available for inspection to determine its viability.

Determination of viability of a segment of gut

Criterion	Viable gut	Non-viable gut
Appearance	Pink, glistening and shiny	Dark/grey and dull in appearance
Pulsation	Mesenteric pulsation present	Mesenteric pulsation absent
Peristalsis	Peristaltic wave present	Peristaltic wave absent

The final 'acid test' to determine the viability of the segment of gut involves the following

- Wrap up the segment of gut with an abdominal mop soaked with warm saline
- Request the anaesthetist to administer 100% oxygen to the patient
- Wait for between 5-10 minutes
- Then unwrap the gut and look out for any changes in favour of viability.

While a viable segment of gut is returned into the abdomen, its non-viable counterpart should of necessity be resected coupled with primary anastomosis.

CHAPTER TWENTY THREE

HYDROCOELE

A hydrocoele is an abnormal accumulation of fluid between the parietal and visceral layers of the tunica vaginalis. The aetiology of hydrocoele is multifactorial.

- Congenital hydrocoele is due to the trickling of peritoneal fluid into the tunica vaginalis through its direct communication with the peritoneal cavity.
 - It may also result from an imbalance between fluid production and absorption
- Primary hydrocoele: This is mostly due to defective absorption of fluid probably resulting from degeneration of the wall of the tunica vaginalis
- Secondary hydrocoele: Due to excessive production of fluid secondary to an underlying pathology such as epididymo-orchitis, testicular torsion, trauma or tumour
- Defective lymphatic drainage of the scrotal organs may also play a role. This may explain the development of hydrocoele following inguinal herniorrhaphy
- Hydrocoele of the hernia sac: May be due to obstruction at the level of the neck of the sac by omentum. This results in the collection of fluid in the hernia sac.

Embryological basis of hydrocoele:

The testis is formed in the abdomen and descends through the inguinal canal into the scrotum. The processus vaginalis is pulled down along with the testis during this process. The processus vaginalis, however, closes before birth. The tunica vaginalis is the part of the processus vaginalis that covers the anterolateral aspect of the testes. A potential space that is continuous with the intra-abdominal cavity is thereafter created. Hydrocoele is said to occur when fluid accumulates anywhere along the processus vaginalis. Hydrocoele fluid is amber-coloured and its contents include water, fibrinogen, salts and albumin. Others are tyrosine and cholesterol crystals. It has a specific gravity of 1012 to 1024.

Classification of hydrocoele:

There are two forms of classification – anatomical and pathological

Anatomical classification: This is based on the location of fluid along the processus vaginalis and whether it is continuous into the peritoneal cavity.

- Congenital hydrocoele: The entire length of the processus vaginalis is patent right into the peritoneal cavity. The opening into the latter at the internal inguinal ring is through a narrow orifice which makes it possible for the trickling of peritoneal fluid into the scrotum. A wider opening at this level will admit a viscous resulting in congenital inguinal hernia. It is therefore obvious that both congenital hydrocoele and congenital inguinal hernia have the same embryological basis. This equally explains the basis for the common surgical treatment for both conditions
- Infantile hydrocoele: The accumulation of fluid extends from the testis to the deep internal ring but does NOT communicate with the peritoneal cavity

- Encysted hydrocoele of the cord: This is the isolated accumulation of fluid anywhere between both ends of the tunica vaginalis (between the deep ring and the testes). The most common site is just below the inguinal canal
- Vaginal hydrocoele: The fluid in this case is confined to the tunica vaginalis that partially surrounds the testis (2/3rds around the anterolateral aspects). There is no connection whatsoever with the peritoneal cavity. This is the commonest form of hydrocoele in adults

The above are the common forms of hydrocoele. The rare forms are as follows

- Bilocular hydrocoele (Hydrocoele-en-bisac): This is a variant of encysted hydrocoele of the cord imbued with two intercommunicating sacs. The sacs are respectively located superiorly and inferiorly to the neck of the scrotum.
- Hydrocoele of the Canal of Nuck: This occurs in females in relation to the round ligament. As to be expected, it is located in the inguinal canal
- Hydrocoele of the hernial sac: This is the accumulation of fluid in a hernia sac due to adhesions of its contents

Pathological classification of hydrocoele: Of two forms – primary and secondary

- Primary hydrocoele: It is also referred to as idiopathic hydrocoele. There is no underlying lesion of the testis
- Secondary hydrocoele: It is secondary to an underlying testicular lesion. Predisposing conditions are
 - Trauma: Direct injury, post-herniorrhaphy (due to damage to the lymphatic vessels of the tunica vaginalis)
 - Infection: Filariasis, syphilis and tuberculosis
 - Neoplasm: Testicular tumours

Clinical features of hydrocoele: The mode of presentation depends on the type of hydrocoele

- Congenital hydrocoele: It occurs in the paediatric age group. There is gradual emptying of the swelling on elevation of the scrotum or on lying down due to its communication with the peritoneal cavity. The swelling reappears on standing due to the gravitational aid of the refilling process.
- Infantile hydrocoele: It also occurs in the paediatric age group. The swelling does not empty on elevation of the scrotum as it does not communicate with the peritoneal cavity.
- Hydrocoele of the cord: The swelling is pulled down by downward traction on the testis

Vaginal hydrocoele: It presents as an entirely scrotal mass. The examining fingers will be able to get above it. This is in contrast to an inguinoscrotal hernia where it is not possible to get above the scrotal swelling owing to it being an extension of an inguinal pathology.

Bilocular hydrocoele: Characterised by cross-fluctuancy

- Hydrocoele of the canal of Nuck: Occurs only in females and is located in the inguinal canal
- Hydrocoele of the hernia sac: Associated with a hernia that may be irreducible due to adhesions

Despite the peculiarity in the clinical features of the various types of hydrocoele, some features are common to all varieties of uncomplicated primary hydrocoele:

- Vaginal hydrocoele: Ability to get above the swelling and inability to palpate the testis due to the disposition of the tunica vaginalis
- Do not demonstrate a positive cough impulse
- They are brilliantly translucent. This attribute is lost when there is associated infection or presence of blood. A long-standing hydrocoele may not be translucent just from mere thickening of the dartos muscle.
- They are cystic in nature and demonstrate fluctuancy

Differential diagnosis of vaginal hydrocoele: Essentially from other scrotal swellings

- Inguinal hernia: Usually the inguinoscrotal variety. As noted above, one cannot get above the latter at the neck of the scrotum. Unlike a hydrocoele, a hernia demonstrates a positive cough impulse
- Testicular tumour: There may be evidence of malignancy both on clinical and radiological grounds
- Epididymal cyst: Can be palpated separate from the testis
- Varicocoele: 'Worm-like' in consistency
- Haematocoele: Due to bleeding into the tunica vaginalis. Common causes include trauma and complication following tapping of a hydrocoele
- Chylococoele: Accumulation of lymphatic fluid in the tunica vaginalis. It is a complication of lymphatic obstruction
- Scrotal filariasis

Investigations:

The diagnosis is usually clinical. The following investigations may be necessary both to confirm the diagnosis but more importantly to rule out any underlying disease. This is even more pertinent when dealing with hydrocoele in the adult age group.

- Ultrasound scan of the scrotum
- Analysis of the hydrocoele fluid: This applies to secondary hydrocoele and is aimed at identifying the underlying problem. The fluid is analysed for microscopy, culture and sensitivity (bacterial infection), acid and alcohol fast bacilli (for tuberculosis), and fine needle aspiration cytology (to rule out malignancy). The hydrocoele fluid is obtained by aspiration. The testis becomes palpable after aspiration making it easier to detect any inherent abnormality. The complications of hydrocoele aspiration include introduction of infection and haemorrhage (haematocoele). Aspiration should be avoided in a suspected case of underlying testicular tumour as the path of the needle may create a portal for direct spread of the lesion to the scrotal skin. This may predispose to inguinal lymphadenopathy

Complications of hydrocoele

- Infection: Will result in pyocoele
- Bleeding: Will result in haematocoele
- Testicular atrophy: May result in infertility
- Herniation of a hydrocoele: Rare
- Calcification of the hydrocoele sac
- Rupture: May be due to trauma

Treatment of hydrocoele: This depends on the age of the patient and the nature of any underlying pathology

Paediatric age group: Hydrocoele in childhood is diagnosed soon after birth. Since there is still a possibility of spontaneous closure, it is advised that the child should be under observation until the age of two. Surgery is recommended if it does not close within this period. The recommended procedure is herniotomy. This involves a transinguinal incision, inspection of the testis and high ligation/excision of the processus vaginalis at the deep inguinal ring. It is essentially the same treatment that is offered in the management of congenital inguinal hernia

Adults: Relatively small hydrocooles may be placed under observation. Indications for surgical treatment are

- Large/expanding hydrocoele
- Patient is uncomfortable
- Infected hydrocoele
- Hydrocoele associated with a hernia

The common surgical procedures for vaginal hydrocoele include

- Subtotal excision of the tunica vaginalis
- Lord's procedure: Plication of the tunica vaginalis by inserting a series of interrupted absorbable sutures. The resultant mass will later undergo fibrosis
- Jabouley's procedure: Partial excision and eversion of the tunica vaginalis. The everted edges of the tunica vaginalis are sutured together with an absorbable suture.

It is important to insert a drain in order to forestall haematoma formation. A scrotal support should equally be applied in order to reduce bleeding and discomfort.

Encysted hydrocoele of the cord is treated by simple excision under local anaesthesia

Aspiration and sclerotherapy: Simple aspiration of the hydrocoele fluid is usually complicated by recurrence and infection. It may, however, be the only option in

- Patients who have symptomatic hydrocoele but are unfit for surgery.
- Patients who reject surgery.
- Men in whom fertility is no longer an issue (relative indication)

It has the advantage of offering the opportunity for a more clinical evaluation of the testis which becomes more easily palpable after aspiration of the hydrocoele fluid. Post-aspiration

sclerotherapy may be instituted in order to forestall the reaccumulation of fluid in the tunica vaginalis. Recommended sclerosing agents include tetracycline, sodium tetradecyl sulphate and doxycycline solutions.

Currently there is a place for laparoscopic percutaneous extraperitoneal closure

Complications of surgery include

- Bleeding with formation of scrotal haematoma
- Infection.
- Recurrence of hydrocoele is a rare complication.

Surgery for hydrocoele is now carried out as a day case procedure

CHAPTER TWENTY – FOUR

THE BREAST

The study of the breast and its diseases forms a very important part of Surgery. This is due to the high morbidity and mortality associated with breast cancer.

Anatomy: The breast, while rudimentary in the male, is well developed in the female. It lies between the second and sixth ribs. The breast is bigger than it appears as it extends into the upper part of the rectus sheath inferiorly and up to the lower edge of the clavicle superiorly. The horizontal span is between the lateral edges of the sternum medially to the mid axillary line laterally. The extension into the axilla is referred to as the axillary tail of Spence. It lies mainly on the pectoralis major muscle.

Structure: Consists of about 15 lobules, each of which is drained by a lactiferous duct. The ducts ultimately drain into the nipple which is surrounded by the pigmented areolar.

Lymphatic drainage of the breast: Knowledge of this is of utmost importance in the understanding of the pathophysiology of breast cancer. The following groups of lymph nodes drain the lymphatic fluid from the breast and hence may be involved in the spread of breast cancer

- Axillary group of lymph nodes: Drains about 75% of lymphatic fluid
- Internal mammary group of nodes: 20% drainage
- Others are: Supraclavicular, cephalic, posterior intercostals, subdiaphragmatic and subperitoneal lymph nodes: 5% drainage

Most of the lymphatics from the lateral half drain to the axillary group while most from the medial half drain to the internal mammary group of lymph nodes. In practice, there is no clear distinction as lymph from any part may drain to either group of nodes. In other words, lymph from the lateral half may still flow into the internal group of nodes and vice versa.

Berry's classification of axillary nodes

- Level 1: Lateral and inferior to the pectoralis minor muscle. Includes the pectoral (anterior), humoral (lateral), and subscapular (posterior) groups.
- Level 11: Nodes posterior to the pectoralis minor muscle (central group of lymph nodes)
- Level 111: Nodes superior and medial to the pectoralis minor muscle
- Interpectoralis nodes (Rotter's nodes): Lie between P.major and P.minor. Involved only when axillary nodes are obstructed

Others are

- Infraclavicular and supraclavicular groups: Receive drainage from both the axillary and internal mammary nodes. Involvement of the supraclavicular nodes signifies N3 nodal status.

- The contralateral breast or the opposite axilla may be involved when the primary lymphatic channels of the ipsilateral breast are blocked
- Hepatic involvement: Through rectus abdominis and falciform ligament
- Peritoneal lymphatics: Lymph from the lower inner quadrants may communicate with the subdiaphragmatic and subperitoneal lymphatics through the lower internal mammary nodes
- Subareolar plexus of Sappey: Deep to the areolar. Lymphatics pass through the P.major muscle and clavipectoral fascia to reach the apical and also the internal mammary nodes

BREAST CANCER

Worldwide, breast cancer is the commonest cancer in females. The exception is the United States of America where lung cancer is most common. Owing to the high morbidity and mortality associated with breast cancer, any breast lesion in a female is regarded as cancer until proved otherwise. Studies have, however, shown that only about 25% of breast lesions turn out to be cancerous at the end. The above principle of management is aimed at reducing the incidence of mis-diagnosis of breast cancer to the barest minimum. Any missed case of breast cancer is regarded as one too many. Breast diseases can therefore be divided into two broad groups: breast cancer and the rest

In the discussion of breast diseases therefore, we shall discuss the problem of breast cancer in details. This will then be followed by analysis of other common breast diseases as they differ from breast cancer.

Risk factors for breast cancer

Like most cancers, the exact aetiology of breast cancer is unknown. There are, however, some associated risk factors. A risk factor is defined as anything that increases one's chances of getting a disease such as cancer. There are quite a number of factors associated with the pathogenesis of breast cancer.

- Sex: It is a predominantly female disease as it is uncommon in males. Whereas only 1% of breast cancers occur in males in western countries, in Africa the percentage is 2-5%.
- Age: The chances of developing breast cancer tend to increase with age. The peak age incidence in our environment is 40 to 50. This is a decade lower than what obtains in developed countries (50-60).
- Early menarche and late menopause: Early menarche (before the age of 12) and late menopause (after the age of 55) connote a prolonged menstrual lifespan. We know that the breast, under hormonal influence, undergoes a progressive increase in size after ovulation only for it to gradually reduce in size at the end of menstruation. This waxing and waning of the size of the breast as a result of hormonal influence makes the breast more vulnerable to changes that may 'spark off' neoplastic or cancerous changes in the breast. The group of benign lesions that may arise from this phenomenon is generally classified as 'Abnormalities of Normal Development and Involution' (ANDI) and will be discussed later.

- Age at first full-term pregnancy: It is thought that the earlier a woman achieves her first full-term pregnancy, the lower the risk of breast cancer. When this happens at the age of 30 and above, the risk is thought to be higher. Indeed, it is thought that probably the relatively late age at which women achieve their first full-term pregnancy these days owing to educational and career pursuits may account for the increasing incidence of breast cancer worldwide.
- Nulliparity: It is thought that a woman who has not had a biological child may be more susceptible to breast cancer. This is not, however, the experience in Nigeria as most of our patients have had children.
- Absence of lactation: It is thought that a woman who has not breastfed a baby is more prone to breast cancer. This may be due to the protective influence of lactational amenorrhoea which results in reduction of the lifetime summative menstrual cycle. Again, this is not the experience in Nigeria as most of our patients here breastfed their children.
- Hereditary breast cancer: Familial predisposition is believed to account for about 10% of breast cancer cases. This has been linked to mutation in certain genes such as BRCA1, BRCA2 and p53. BRCA1 gene was isolated in 1990 and is associated with the pathogenesis of breast and ovarian cancers. It has a cumulative risk of 56% to 80%. On the other hand, BRCA2 has a cumulative risk of 20%. These are genes which under normal circumstances help to protect against the development of breast cancer. Hereditary breast cancer is characterised by early age at onset, bilaterality (affectation of both breasts), and multiple affection of first degree relatives such as mother, sisters and daughters. Between the two genes, they carry an over 60% lifetime risk of a second primary breast cancer. Mutation of p53 oncogene has been implicated in 7% to 37% of intraductal breast cancer. It has also been found to equally contribute to the genesis of both lung and colon cancer.
- Use of exogenous hormones such as oestrogen replacement therapy in the management of postmenopausal symptoms. The use of oral contraceptive pills is known to increase the chances of developing breast cancer.
- Certain non-breast malignancies such as ovarian and endometrial cancers may be associated with breast cancer.
- Environmental factors such as fatty diet and lifestyle characteristic of developed western nations: Being overweight (obesity), alcoholism, smoking, and lack of exercise have all been associated with high risk of breast cancer. For instance, the risk of breast cancer is 1.5 to 2 times higher in obese postmenopausal women than in their non-obese counterparts. It is believed that the rising incidence of cancers which was hitherto rare in the developing world may not be unconnected to the westernisation of their lifestyle.
- Exposure to irradiation such as X-rays: The incidence of breast cancer, and indeed cancers in general, was found to be higher in survivors of Hiroshima and Nagasaki. Diagnostic chest X-rays may potentiate the development of breast cancer. Adjuvant radiotherapy may increase the risk of breast cancer in the contralateral breast
- High breast density,

- Previous breast cancer
- Breast conditions such as atypical hyperplasia.

Pathology of breast cancer

The two main histological forms are ductal and lobular carcinoma. Each of these may present either in the form of insitu carcinoma (no breach of the basement membrane) or the invasive form (with breach of the basement membrane). Other histological forms include mucinous, squamous cell, tubular and medullary forms of carcinoma as well as papillary and cribriform varieties

Special types of breast cancer include the inflammatory carcinoma and Paget's disease of the nipple. The inflammatory variety presents clinically with features suggestive of acute mastitis and histology reveals the presence of inflammatory cells. It is notorious for exhibiting a rapid degree of progression. Paget's disease of the breast, on the other hand, presents as an ulcer around the nipple-areolar complex. Underlying this apparently benign eczema-like lesion, however, is an intraductal carcinoma. Histology reveals the characteristic Paget cells.

Spread of breast cancer: As propounded by Halsted, breast cancer was thought to be a loco-regional disease which is held up by the axillary nodes before spreading to other parts of the body. This was the principle behind the operation of radical mastectomy as it was believed that the more radical the local excision, the better the prognosis. This was proved wrong, however, by the occurrence of metastatic lesions even after such radical and highly mutilating procedures. This led to the conclusion that breast cancer is probably a systemic disease and that even before surgery, some cancer cells would have detached from the primary organ, spread through the blood stream and got lodged in distant organs. These cells were only bidding their time to proliferate before manifesting in the respective organs of spread. The routes of spread are

- Local spread: Skin (peau d'orange), underlying muscles (pectoralis major and minor)
- Lymphatics: Internal mammary nodes, and especially the axillary group of nodes
- Haematogenous: Lungs (15% - 20%), pleura (10% - 15%) and bones (50% - 65%). Others are liver (5% to 15%) and brain
- Transcoelomic: Peritoneal space (ascites)

STAGING OF BREAST CANCER

The purpose of staging of breast cancer, as in all cancers is three-fold

- To plan a therapeutic strategy that is most appropriate for the patient
- To allow for more intelligent prognostication of the disease status of the patient
- To permit comparison of therapeutic results obtained from different sources by different means

There are three staging methods: The TNM (WHO or International Union against Cancer) and the Manchester stagings are the ones most commonly used in clinical practice. The less commonly used, which ironically is quite simple and straight forward is the Columbia Clinical Classification (CCC)

TNM staging: This was formulated by the international Union against Cancer (UICC) and accepted by the American Joint Commission on cancer staging is a world standard. T stands for primary tumour; N stands for regional lymph node while M stands for distant metastasis

T (Primary tumour)

T0: No demonstrable tumour.

C is carcinoma in situ

T1 Tumour < 2cm in its widest diameter

T1a not fixed to underlying fascia or muscle

T1b fixed to underlying fascia or muscle

T2: Tumour of 2cm – 5cm diameter

T2a Not fixed to underlying fascia or muscle

T2b fixed to underlying fascia or muscle

T3 Tumour > 5cm in its widest diameter

T3a not fixed to the underlying fascia or muscle

T3b fixed to underlying fascia or muscle

T4 Tumour of any size but with extension to skin, underlying fascia or muscle

T4a Fixed to chest wall

T4b with peau d'orange, ulceration or skin nodules

T4c when T4a and b are present

T4d inflammatory carcinoma

N0: No palpable node in ipsilateral axilla

N1: Palpable mobile nodes in ipsilateral axilla

N2: Matted nodes in the ipsilateral axilla

N3: Involvement of ipsilateral supraclavicular lymph nodes

M0: No distant metastasis

MX: Distant metastasis is undiagnosed or cannot be assessed

M1: Distant metastasis present

MANCHESTER CLASSIFICATION (Four stages)

Stage 1: Tumour is confined to the breast, not attached to underlying muscle or overlying skin but if attached to the overlying skin, must be less than the diameter of the tumour. There are no palpable axillary lymph nodes

Stage 1A: Same as stage 1 but with palpable and mobile axillary lymph nodes

Stage 1B: There is tumour in the breast in addition to the following

A The area of attachment to the skin is larger than the widest diameter of the tumour

B The tumour is attached to the underlying fascia/muscle

C The axillary nodes are present and are fixed

Stage 1C: Tumour in the breast with

A Lymphatic spread beyond the ipsilateral axilla eg supraclavicular nodes, internal mammary nodes

B Haematogenous metastases to liver, lungs, bone etc

Columbia Clinical Classification (CCC) of breast cancer

Haagensen and Stout in a review of the clinical features of patients with breast cancer in their hospital in the 1940s described some grave signs associated with a 0% possibility of a 5-year cure and 50% risk of local recurrence. The grave signs are: skin ulceration, fixation of tumour to the chest wall, axillary nodes > 2.5cm in diameter, oedema of 1/3rd of the skin of the breast and presence of fixed axillary lymph nodes. These features were later encapsulated in what is now referred to as the Columbia Clinical Classification (CCC) of breast cancer. This classification consists of four stages, A to D, as follows

Stage A: No skin oedema, ulceration or solid fixation of the tumour to the chest wall. Axillary nodes are not involved clinically

Stage B: No skin oedema, ulceration or solid fixation of the tumour to the chest wall. Clinically involved nodes, but less than 2.5cm in transverse diameter and not fixed to overlying skin or deeper structures of the axilla

Stage C: Any one of the five grave signs of advanced breast carcinoma

- 1 Oedema of the skin of limited extent (involving less than one-third of the skin over breast)
- 2 Skin ulceration
- 3 Solid fixation of the tumour to the chest wall
- 4 Massive involvement of axillary lymph nodes (measuring 2.5cm or more in transverse diameter)
- 5 Fixation of the axillary lymph nodes to the overlying skin or deeper structures of the axilla

Stage D: All other patients with more advanced breast cancer including

- A combination of any two or more of the five grave signs listed under stage C
- Extensive oedema of the skin (involving more than one-third of the skin over the breast)
- Satellite skin nodules
- The inflammatory type of breast cancer
- Clinically-involved supraclavicular lymph nodes
- Internal mammary metastases as evidenced by a parasternal tumour
- Oedema of the arm
- Distant metastases

It is important to note that the CCC staging, though not much emphasised, is still popular in the United States of America

Symptoms of breast cancer

*Breast lump: This is the commonest mode of presentation. The history of its duration and progression should be obtained. A dominant breast mass is a three-dimensional breast lump that persists all through the menstrual cycle.

- * Associated pain in the breast: Most malignant lumps are painless.
- * Associated fever may suggest an inflammatory condition such as acute mastitis
- * Associated nipple discharge: May be bloody, milky, pus-like, serous or watery
- * Ulceration around the breast tissue. In Paget's disease of the nipple, the ulcer is located around the nipple-areolar complex.

*Identification of any risk factors as outlined above: An example is by probing into the obstetric/gynaecological, family and social history.

* Note any feature of metastatic disease such as cough, chest pain, back pain, jaundice, and paresis or inability to walk. The general effects of cancer such as loss of weight, weakness and anorexia should be ruled out

Clinical examination: Aims at examining the breast for clinical features of cancer and determining the extent of the lesion. At the end of the clinical examination, the lesion is categorised into either early or late. This is important both in the prognostication, as well as the determination of the management strategy.

General examination may reveal a chronically ill-looking patient who may be pale and jaundiced (hepatic spread). There may be axillary and supraclavicular lymphadenopathy (usually reserved as an integral part of the breast examination). The various systems should be examined both for the purposes of determination of the extent of the lesion, as well as the overall management of the patient.

- Cardiovascular system: To determine the fitness of the patient for any subsequent surgical procedure
- Chest: To determine any involvement of the lungs and pleura (pleural effusion)
- Abdomen: For the detection of hepatomegaly, and ascites.

Breast examination: Essentially comprises of the following

- Sitting position with arms raised: Check for asymmetry of both breasts and for any obvious swelling. Inspect the nipple and areolar for skin changes and ulceration
- In the lying position and with the hands tucked under the head (patient gives you a 'salute'), palpate the breasts starting with the apparently normal breast. The latter gives the baseline texture of the breast for the index individual. Very occasionally, a hitherto occult lesion may be detected in an apparently normal contralateral breast.
- Palpate the breast systematically from quadrant to quadrant and feel for the presence of a breast lump.
- Palpate the nipple and areolar: Peau d'orange, nipple discharge and any cutaneous ulcer over the breast should be noted.
- Any palpable lump should be thoroughly evaluated in the usual manner. In particular, its attachment to the skin and to the deep structures should be ascertained.
- Fixation of the lump to the skin is carried out by attempting to fold the skin over the lump. Inability to achieve this connotes attachment to skin. Attachment to the underlying pectoralis muscle can be ascertained by a disparity in the mobility of the mass when the muscle is relaxed and when it is tensed. The muscle is tensed by gripping the waist with the ipsilateral hand.
- Examine the axilla and the supraclavicular fossa for associated lymphadenopathy
- Examine the lumbo-sacral spine and lower limbs for paresis. Ability to walk should be ascertained.

The following is the pathophysiology of the common signs of breast cancer

- Peau d'orange: This is due to lymphoedema resulting from obstruction of the lymphatic drainage by cancer. It may result in deviation of the nipple and thickening of the skin. The orange peel appearance is as a result of puffy skin between dimpled pores of sweat glands and hair follicles
- Nipple retraction: results from infiltration and subsequent fibrosis of the lactiferous ducts
- Skin tethering: Due to shortening and subsequent traction of the suspensory ligaments resulting from cancerous infiltration and fibrosis of the glandular tissue

Investigation of a patient with breast lump:



RIGHT BREAST CANCER: NOTE THICKENING OF THE SKIN AND ASSOCIATED PEAU D'ORANGE

As stated earlier, the principle of management is to regard all breast lumps as malignant until proved otherwise. Therefore, the aim of the initial assessment of a patient with a breast lump is to determine its pathology (benign or malignant). This is carried out by way of triple assessment:

- Clinical: History and clinical examination
- Radiological: Mammography and ultrasonography



ADVANCED BREAST CANCER WITH ULCERATION AND NECROSIS

- Pathological: Fine needle aspiration and/or histology of biopsy specimen.

The following investigations are carried out

* Mammography: This is a soft tissue X-ray of the breast. The primary aim of mammography is the detection of non-palpable breast masses. The X-rays are taken in two planes: cranio-caudal and medio-lateral with the breast compressed between two plates. Mammography is more informative in the older women (above 40) but less reliable in relatively young women due to the dense fibrocystic nature of their breast tissue. Cancer is indicated by the presence of a dense mass with associated speculation, parenchymal distortion and microcalcifications. The latter result from the deposition of calcium flakes in the clump of dead cancer cells and appear like grains of salt. Mammography is employed in the routine screening for breast cancer. It may not, however, demonstrate the presence of all cancers as lobular breast carcinoma does not readily absorb X-rays. Overall, mammography has about 90% sensitivity; 6 to 8% false-negative and 10% false-positive rates.

* Ultrasonography of the breast: Quite useful in younger women with dense breasts and does not expose patients to ionising radiation. The main advantage of USS is that it is readily available and affordable. It aids mainly in the determination of the nature of the lump (solid or cystic). Benign cysts usually have clearly demarcated edges whereas cancers have indistinct outlines. It is useful in the screening of solid breast masses. In addition, ultrasound-guided breast biopsy may be employed in the determination of the nature of impalpable lumps. The drawback is that it cannot detect lesions less than 1 cm in diameter

* Magnetic Resonance Imaging (MRI): It is an evolving trend particularly in the diagnosis of lobular carcinoma and ductal carcinoma insitu. MRI is quite sensitive in the identification of small cancers of the breast and in defining the local extent of the disease. It, however, lacks specificity as lesions identified as malignant may eventually turn out to be benign. It is useful in the detection of nonpalpable lumps in young females with dense breasts and as an adjunct to

mammography. MRI has also been found useful in the detection of metastasis to the vertebral body.

The American College of Radiology has analysed breast imaging reporting with a view to standardising reports as well as institution of quality assurance. This applies to all forms of breast imaging: mammography, breast ultrasound scan and breast magnetic resonance imaging (MRI). Radiological images are categorised from 0 to 6 with specific recommendations attached to each categories of the BI-RADS system

- Category 0: Assessment is incomplete; additional imaging assessment is needed
- Category 1: Negative. Routine annual screening mammography is recommended for women older than 40
- Category 2: Benign findings; routine annual mammography is recommended for women older than 40-
- Category 3: Probably benign finding; initial short-term imaging follow-up (usually 6-month) recommended. Malignancy rate < 2%
- Category 4: Suspicious abnormality; consider biopsy. Malignancy rates in this category ranges between 3% to 94%. Depending on the malignancy rating, category 4 can be divided into three sub-categories: 4A, 4B and 4C
 - 4A: Low suspicion of malignancy
 - 4B: Intermediate suspicion of malignancy
 - 4C: Moderate concern but not classic for malignancy

Category 5: Highly suspicious of malignancy (> 95% malignancy). Requires biopsy or surgical excision

Category 6: Known biopsy-proven malignancy

*Fine needle aspiration cytology (FNAC): Diagnosis is made by the examination of cells aspirated from the lesion. It has the unique advantage of producing speedy results. FNAC has a sensitivity of 80% to 98% and a false negative rate of 2% to 10%. Its specificity is very high (almost 100%). It is, however, operator-dependent

* Tissue biopsy: Provides the specimen for a complete histological diagnosis. This is accomplished either by the close or open methods. The close method is by way of core biopsy. The latter is aimed at obtaining small pieces of tissue for histological examination by the use of special needles (Tru-cut is the most commonly used variety). In the open method, the tissue is obtained through a cosmetic (preferably circumareolar) breast incision. Surgical excision of the whole lump for histological examination is referred to as excisional biopsy. It is indicated when the lump is small (< 4 cm). On the other hand, a partial excision is referred to as an incisional biopsy. The latter is indicated when dealing with bigger breast lumps particularly when the result of core-biopsy is inconclusive. Incisions for open biopsy should be positioned in such a manner that will not encroach on the incision for a subsequent mastectomy. Frozen-section biopsy is not encouraged in the management of breast cancer as the diagnosis may be confusing particularly when dealing with in-situ disease or atypical hyperplasia. A tissue biopsy will aid in the resolution of conflicting situations under the following circumstances:

- Clinical suspicion of malignancy even when FNAC and mammogram are normal
- Suspicion of malignancy on FNAC or mammography, even though the breast appears clinically normal
- Finding of atypical cells on FNAC

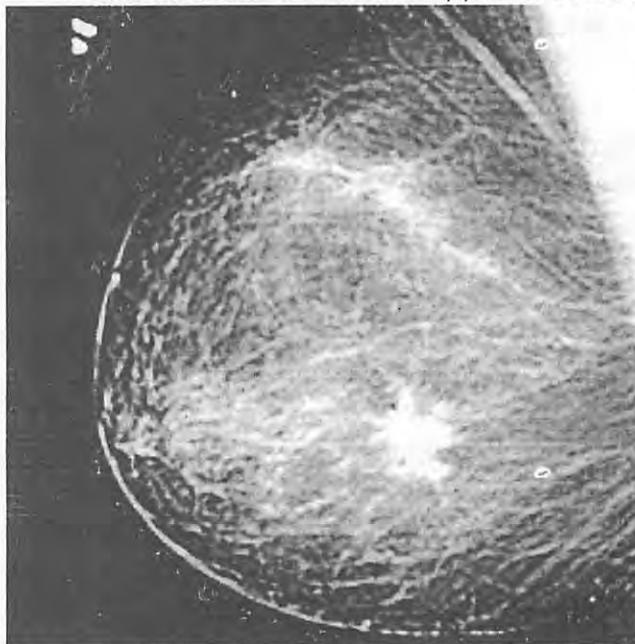
- Request by the patient for open biopsy in order to prove the diagnosis of breast cancer beyond all reasonable doubt

* Oestrogen and progesterone receptor assay should be carried out on the tissue. A positive result is an indication that the tumour will respond to anti-oestrogen hormonal therapy. On the other hand, a negative result indicates a low response to hormonal manipulation. Whereas 60% of ER-positive tumours respond to endocrine ablation, only 10% of ER-negative tumours respond.

* HER-2 neu assay: It is human epidermal growth factor receptor associated with the HER-2 gene. It occurs in one out of four breast cancers. A positive result indicates a likely response to the use of monoclonal antibodies such as Herceptin and Tykerb (lapatinib). Positive HER2-neu in breast tissue is associated with poor prognosis.

If the diagnosis of breast cancer is confirmed by way of the triple assessment (clinical, radiology and pathology), the next step in the management is to further determine the extent of the lesion. The following, aptly referred to as staging investigations, are carried out.

- Chest X-ray: Look out for pulmonary metastasis and pleural effusion
- X-ray of the lumbosacral spine: To detect metastases to the bone
- Isotope bone scan: Preferred to X-rays for detection of bony metastasis
- Abdominal ultrasound scan: To detect liver metastasis and ascites.
- CT scan of the thorax and upper abdomen for lung and liver metastases



MAMMOGRAM OF THE BREAST SHOWING MICROCALCIFICATION

- CT scan of the brain: Carried out only if there are clinical features suggestive of metastatic spread to the brain.
- Magnetic resonance imaging
- Positron emission tomography (PET): It is more sensitive than conventional imaging in identifying distant metastases. When combined with brain MRI, it may be the sole method of staging in breast cancer

- Liver function tests: Raised alkaline phosphatase may indicate involvement of the liver and/or bone
- Serum calcium: Raised levels may indicate bone secondaries
- Genetic screening: Recommended in young patients (30s and 40s) with newly diagnosed breast cancer who have a positive family history of breast cancer particularly when a first degree relative is involved.

Treatment of breast cancer:

This is multidisciplinary and involves various specialists such as the medical oncologist, radiotherapist, pathologist, psychologist and specially trained nurses. While the surgeon should be at the centre as a coordinator, the input from these other professionals is indispensable for the overall management of the patient. The first step in the treatment of a patient with breast cancer entails proper counselling with a view to obtaining an informed consent. This involves a heart-to-heart discussion with the patient preferably in the presence of a close relative. Details of the diagnosis and treatment options are extensively discussed. The importance of this cannot be overemphasised as it not only allays fear and anxiety thereby eliciting the cooperation of the patient during the long and difficult period of treatment. It is also important for medicolegal reasons.

Generally speaking, there are two aims of treatment of breast cancer depending on the stage at presentation. If a patient presents early in the course of the disease, all things being equal, the treatment is aimed at a possible cure. It is, however, a different kettle of fish when the presentation is late. In this case, treatment is only palliative and is primarily aimed at improving the quality of life of the patient. The importance of early diagnosis and treatment cannot therefore be overemphasised.

The current treatment of breast cancer employs a combination of methods, all aimed at attacking the cancer cells.

- Surgery cuts them out
- Chemotherapy poisons these rapidly dividing cells wherever they may be residing
- Radiotherapy burns them
- Hormonal therapy creates an unconducive environment for cancer growth.
- Targeted therapy disorganises the process of cell proliferation

Surgery and chemotherapy are regarded as locoregional treatment modalities while chemotherapy, hormonal therapy and targeted therapy are systemic modalities.

In early breast cancer, surgery is the primary line of treatment. The other forms of treatment play a secondary role and collectively constitute the adjuvant therapy. On the other hand, surgery plays a secondary role in the management of advanced breast cancer. In this case, chemotherapy and hormonal therapy are administered initially in a bid to reduce the size of the tumour. This results in the downstaging of the tumour, thereby rendering it amenable to surgery. In extreme cases, the tumour could be so downstaged as to render the primary cancer amenable to breast conserving surgery.

Early breast cancer

Surgery: Surgical treatment for breast cancer is no longer as radical as was the situation some decades ago. This is because breast cancer is now regarded as a systemic disease. Depending on the stage at presentation of early breast cancer, the options include

Breast conservative surgery: This includes wide local excision for very small tumours and quadrantectomy for slightly bigger lesions. QUART is an acronym for quadrantectomy, axillary dissection (levels 1 to 111), and radiotherapy. In both situations, the pathological confirmation of the tumour edges being free of tumour is imperative. The following are criteria for breast conserving surgery

- Small breast lumps that are subject to wide excision,
- Clinically negative axillary lymph node and
- Breast of adequate size to allow adequate dosage of radiation
- Positive consent from the patient
- Availability of facilities for postoperative radiotherapy. The latter constitutes an integral part of any breast conservative regimen.

Mastectomy: Widely practised in the developing world due to late presentation of patients for treatment. The indications for mastectomy in early breast cancer include

- When the tumour is large in relation to the size of the breast
- Tumour is situated close to the nipple
- When a multifocal disease is suspected as in lobular carcinoma and
- When the patient with an apparently small lesion requests for mastectomy in order to allay the fear of a possible recurrence.

Currently, both mastectomy and breast conservation surgery are regarded as viable options in the management of early breast cancer since the extent of local surgery generally does not impact on overall survival. Clinical trials have shown that the outcome of breast conservation surgery is better following initial administration of neoadjuvant chemotherapy.

There are two types of mastectomy: Simple (total) and modified radical

- Total (simple) mastectomy removes the breast tissue, nipple, areolar and skin but not the lymph nodes
- Modified radical mastectomy removes the entire breast including the breast tissue, skin, areolar and nipple. In addition, it aims at removing most of the axillary lymph nodes. May also remove pectoralis minor while preserving pectoralis major.

There are three forms of modified radical mastectomy based on the extent of clearance of the axillary lymph nodes as well as the extent of manipulation exerted of the pectoralis minor muscle. The latter as noted above divides the axillary lymph nodes into levels 1, 11 and 111

- Patey's modified radical mastectomy: Levels 1, 11 and 111 cleared after removing pectoralis minor
- Scanlon's modified radical mastectomy: Levels 1, 11, and 111 cleared. The coracoid process attachment of the pectoralis minor is incised for Level 11 clearance but is reattached thereafter
- Aunchicloss modified radical mastectomy: Only levels 1 and 11 are cleared.

The axilla: It is important to determine the status of the axillary lymph nodes in relation to invasion by cancer. Involvement of the nodes is synonymous with a possible metastatic spread.

This may be carried out either pre- or intra-operatively. The former is carried out either by axillary node sampling via an axillary incision or through a sentinel lymph node biopsy (SLNB). The main objective of SLNB is to help curtail the rate and extent of axillary lymph node dissection. SLNB ensures that this procedure is carried out only when absolutely necessary in view of the associated complication of lymphoedema which poses a lot of management challenges. It has equally been found useful in the prognostication of breast cancer. SLNB involves an intratumoral injection of a radioisotope and/or a dye (isosulphan blue dye) in order to locate the sentinel node which is the first axillary node that receives the lymphatics from the cancer. Its histological status as regards involvement by malignancy is a predictor of the involvement or otherwise of the more distant lymph nodes. The axilla is incised and the sentinel node is detected either by a hand-held Geiger counter in the case of radioisotope scanning method or by staining when a dye is used. A positive result is an indication for incorporating axillary dissection as an integral part of the definitive surgery. Axillary node dissection is, however, fraught with lymphoedema which is a very distressing complication. Numbness and paraesthesia have also been reported. This is due to damage to the intercostobronchial nerves. It must be stressed that the axillary node status is the most important prognostic factor in breast cancer. It is envisaged that in future, more conservative methods of assessing the state of the axilla such as positron emission tomography scanning (PET scanning) will replace SLNB. An even much simpler method involving DNA technology is being worked on. At surgery, the sentinel node is identified and sent for histological examination to determine the involvement or otherwise by cancer. The outcome will thereafter dictate the extent of the surgery.

Complications of mastectomy (modified radical/total)

As in all surgeries, these can be categorised into general and specific complications. The general complications include those due to anaesthesia and atelectasis. The specific complications include

- Seroma formation
- Lymphoedema
- Flap necrosis
- Shoulder dysfunction
- Axillary hyperesthesia
- Winged scapula

Prominent among these specific complications of mastectomy are postmastectomy seroma and lymphoedema. These will be discussed in more detail.

Postmastectomy seroma: This is a collection of serous fluid in the dead space of postmastectomy skin flap, axilla or breast following modified radical mastectomy (MRM) or breast conserving surgery (BCS). The incidence is 50% to 70% and is proportional to the degree of surgery and the number and extent of axillary lymph node involvement. It is believed that mastectomy and axillary node dissection open up a large raw surface area with the attendant opening up of several blood vessels and lymphatics. The oozing of blood and lymphatics from the latter is responsible for the subsequent seroma. The adverse effects of the latter include delayed wound healing, wound infection and wound dehiscence. Others are prolonged hospital stay and delayed

recovery. The latter causes delay in the commencement of adjuvant therapy. Prevention remains the key to management of postmastectomy seroma. The following measures have been applied

- Surgical technique: Avoid the use of electrocautery in making skin flaps. It has been found that this technique while reducing the degree of blood loss tends to increase the rate of seroma formation.
- Use of sealants and sclerotherapy: Fibrin glue has been used to secure haemostasis
- Compressive dressing: Associated with discomfort and therefore engenders low level of tolerance by the patient
- Use of drains: Has been found useful. Associated controversies, however, include use of suction versus passive drainage and application of single or multiple drains. Whatever draining technique is applied, it is important to leave the drain long enough for seroma to subside before removal. This may be when drainage is less than 20 to 50 mls in 24 hours
- Reduction or obliteration of the dead space within the flaps has been found effective. Closely spaced interrupted sutures are applied for this purpose. The main drawback is the associated increase in the operation time.
- Delay in the commencement of shoulder exercises has been found to reduce the incidence of postmastectomy seroma
- Role of octreotide: This is a somatostatin analogue that reduces gastrointestinal secretions by a reduction in splanchnic blood flow. It equally reduces both the production of lymphatic fluid as well as the degree of local inflammatory reaction. Some studies have shown that administration of octreotide postmastectomy helps to reduce the incidence of postoperative seroma

Lymphoedema: Four factors are responsible for lymphoedema which develops in the course of breast cancer management

- The primary lesion with involvement of the axillary lymph nodes
- Axillary dissection during surgery with excision of the lymph nodes
- Radiotherapy which 'burns off' the lymphatic nodes and drainage channels
- Recurrent disease causing lymphatic infiltration

As in seroma, prevention is the key to management of lymphoedema

- Axillary dissection should be carried out only when absolutely indicated. As stated above, sentinel lymph node biopsy may serve this purpose
- Radiotherapy should not be applied after an axillary dissection as it increases the chances of developing lymphoedema
- Early detection

Treatment of an established case of lymphoedema is challenging. The following may be helpful

- Elevation of the affected limb to enhance lymphatic drainage
- Application of elastic arm stockinette or crepe bandage from the metacarpals to the shoulder
- In practice, the above two measures are combined
- Application of pneumatic compression device
- Avoid the use of the ipsilateral arm for intravenous infusion lines and blood sampling

Chemotherapy of breast cancer:

Chemotherapeutic agents act on rapidly dividing cancer cells. They have been found to be quite useful in oestrogen receptor-negative and node-positive patients. In general, chemotherapy helps to improve the disease-free and survival rates. Owing to the high risk of recurrence after a long follow-up, the majority of patients with stage II disease or more advanced lesions are offered chemotherapy. Combination chemotherapy has been found to be more effective than monotherapy. One of the original combinations comprises of cyclophosphamide, methotrexate and 5-fluorouracil (CMF). In the currently used CAF regime, methotrexate is replaced with Adriamycin. The AC regimen (adriamycin and cyclophosphamide) is also effective. It has been shown that the combination of Paclitaxel (taxane) and AC (PAC) is even more effective. Dose dense chemotherapy involves the shortening of the interval between doses from the usual 3 to 4 weeks to 1 or 2 weeks. Though found to be more effective, the main drawback is the increased severity in the chemotherapeutic associated complications. Chemotherapy as an adjuvant treatment is more effective in the younger age group. In addition to the action on cancer cells, it also affects other rapidly dividing cells of the body. These include the blood cells (bone marrow depression), hair follicles (alopecia), and liver cells (jaundice). Cardiotoxicity is also associated with the use of adriamycin. Cytotoxic drugs induce nausea and vomiting. This accounts for the use of metoclopramide and ondansetron in the course of chemotherapy. It is therefore imperative to carry out some baseline investigations prior to the administration of each cycle of these cytotoxic drugs.

- Packed cell volume (minimum accepted value is 30%)
- White cell count (minimum accepted value is 3000/cubic mm)
- Platelet count (minimum accepted value is 100,000/cubic mm)
- Liver function test
- Electrolytes/urea/creatinine

Combination chemotherapy is administered on a four-weekly cycle after ensuring that the blood counts are within the acceptable range. Signs of toxicity include sore throat (leucopenia), bleeding on minimal trauma such as during tooth-brushing (thrombocytopenia) and jaundice. The patient may require transfusion with fresh whole blood either before or after chemotherapy.

Neoadjuvant chemotherapy: This implies the use of chemotherapy before expected surgical treatment. The main advantage is that it makes the tumour smaller (down-staging) and can therefore render a hitherto inoperable tumour, operable. When applied in the management of moderately sized early breast cancer, it may shrink the tumour thereby making a subsequent breast conserving surgery feasible. It also assesses the effectiveness or otherwise of the first-line chemotherapeutic agents with a view to switching over to the second-line drugs if there is no appreciable clinical response. The main disadvantage is the possible potentiation of drug resistance.

Newer drugs include taxanes (docetaxel and paclitaxel) and gemcitabine which are reserved for cases that fail to respond to first line drugs and in metastatic disease. Recently, taxanes have been used as a first-line drug in combination with other drugs.

Hormonal treatment: This is quite effective in the management of patients with oestrogen receptor-positive cancers. Since oestrogen enhances the growth of such breast growths, a decrease in the rate of growth can be achieved by effecting either a reduction in the amount of oestrogen in circulation, or by blocking the receptor sites. The former is achieved with the use of aromatase inhibitors (anastrazole, letrozole, and exemestane) and luteinising hormone-releasing hormone agonists (goserelin). Oestrogen receptor blockers include tamoxifen and raloxifene. Hormonal manipulation, which includes surgical oophorectomy in premenopausal women with breast cancer, is useful in the following situations

- As a postoperative adjuvant treatment in patients with early breast cancer.
- As an integral part of the neoadjuvant treatment in the management of late breast cancer
- As the sole treatment in the management of metastatic postmenopausal disease.
- As a prophylactic treatment to reduce the risk of breast cancer in high risk patients

Hormonal therapy is made to last for 5 years. The most common drug employed is tamoxifen.

Tamoxifen (Nolvadex): This is a selective oestrogen receptor modulator (SERM) that competes for some oestrogen receptor sites with oestrogen. It is regarded as a 'pretender' as it lacks the intrinsic oestrogenic properties that enhance tumour-growth. This ultimately results in the slowing-down of tumour growth. It has been found useful in the above-mentioned situations. In the uterus, tamoxifen acts like oestrogen and encourages the growth of the uterine endometrium. Tamoxifen is known to have a weak oestrogenic effect. The normal dose is 20mg daily. Higher doses may be given when used as a sole therapeutic agent. The duration of treatment is five years. Tamoxifen has been found to

- Improve disease free survival
- Reduce the incidence of contralateral breast cancer in women whose primary breast cancer was ER positive
- Reduce the incidence of breast cancer by 30% to 40% following 5 years of therapy as compared to placebo. This prophylactic effect has been put into use in the management of young people who, by genetic constitution, are prone to the development of breast cancer

Side effects of tamoxifen include are

* Nausea and hot flushes
* Increased risk of thromboembolism.
* Endometrial hyperplasia and carcinoma have been reported in patients after long term usage. This is ascribed to the oestrogenic effect on the endometrium. The risk of uterine cancer in premenopausal women taking tamoxifen is very low compared to postmenopausal women. On a positive note, however, it lowers the cholesterol level and decreases the incidence of postmenopausal osteoporosis.

Raloxifene is a second generation antioestrogen. It has all the therapeutic effects of tamoxifen. A recent study revealed that raloxifene has a similar rate of reduction rate of breast cancer compared to tamoxifen. Equally important is the fact that the beneficial effect appears to be maintained even after discontinuation of treatment. It reduces the resorption and overall turnover of bone and has been used in the treatment of osteoporosis. It is associated with a lower risk of thromboembolism compared to tamoxifen. Raloxifene continues to offer a reasonable alternative to tamoxifen for prevention of breast cancer in high risk women.

Aromatase inhibitors: Oestrogen is synthesised from cholesterol via a series of biochemical reactions ending with the conversion of androgens to oestrogen. Aromatase is an enzyme that facilitates this final pathway. Aromatase inhibitors bind to aromatase and thereby prevent conversion of androgens to oestrogen. They therefore decrease oestrogen production and ultimately decrease the rate of oestrogen-related breast carcinogenesis. They are more useful in postmenopausal women who no longer produce oestrogen of ovarian origin. Whereas anastrozole and letrozole (non-steroidal AIs) are effective only during the duration of treatment, exemestane (steroidal AI) induces a more permanent effect. The major side effect of this group of drugs is osteoporosis. There is a higher risk of fracture particularly of the hip, spine and wrist. They are therefore contraindicated in patients with osteoporosis or those susceptible to it.

Lutenising hormone-releasing hormone agonist (goserelin) has replaced the hypophysectomy of old. It has the same effects as the aromatase inhibitors.

Bisphosphonates: This is a non-hormonal drug approved for the treatment of osteoporosis. It has also exhibited a unique benefit of reducing metastases to bone. Evidence is emerging that the addition of a bisphosphonate, specifically zoledronate to adjuvant therapy in early breast cancer is associated with improved survival as evidenced by the reduced incidence of contralateral breast cancer

Ovarian ablation: This can be achieved surgically by means of oophorectomy. It is only relevant in premenopausal patients whose main source of oestrogen is ovarian. It may also be achieved by medical therapy or radiotherapy

Clinical application: The mechanism of action of chemotherapy and hormonal treatment on the cancer cells seems to be in opposite extremes. Whereas chemotherapy is more effective on rapidly dividing cells, hormonal treatment acts by slowing down the rate of growth. It is therefore logical that both modes of treatment should not be applied simultaneously. In practice, it is adviseable to complete the cycles of chemotherapy prior to the commencement of hormonal treatment.

As regards the hormonal treatment of postmenopausal women, some authorities give tamoxifen for three years, and then administer an aromatase inhibitor for the remaining part of the duration of treatment. This combination tends to maximise the clinical effects of both groups of antioestrogens while at the same time minimising their side effects.

Targeted therapy:

Breast cancer targeted therapies involve substances or drugs which block the growth of cancer by interfering with the formation of specific molecules responsible for tumour cell proliferation and survival. These drugs work differently from chemotherapy drugs which attack all rapidly dividing cells (including cancer cells). About 20% of breast cancers are HER2 positive. They tend to grow and spread more aggressively. HER2 receptors cause abnormal cell growth and division through the use of protein signals called kinases. The following drugs target HER2 positive breast cancer:

- Trastuzumab (Herceptin): Acts on the cell surface. May be used alone or in combination with chemotherapy drugs in the treatment of both early and late breast cancer. Herceptin is administered intravenously for 52 weeks
- Pertuzumab (Perjeta): It is a newer drug that only differs from Herceptin in relation to the site of action on the HER2 receptor. It is administered intravenously and can be applied as a complimentary therapy to herceptin. In combination with herceptin and docetaxel, pertuzumab has been used in the management of HER2-positive metastatic breast cancer.
- Lapatinib (Tykerb): This is a kinase inhibitor. It functions by interfering with HER2-related kinases inside the cell thereby limiting the amount of energy available for the growth and multiplication of breast cancer cells. The oral preparation is employed in the management of advanced breast cancer. Lapatinib can be administered with Xeoda in the treatment of advanced HER2 positive cancer that is no longer responding to anthracyclines, taxanes and herceptin. It can also be used in combination with letrozole (femara) in the treatment of hormone positive, HER2 positive metastatic postmenopausal breast cancer
- Neratinib : This is also a kinase inhibitor that comes as oral pills. Neratinib is given for early breast cancer after one year of initial treatment with trastuzumab. The treatment with Neratinib is for another year

Side effects of HER2- targeted therapy

- Cardiotoxicity: This applies to all the drugs in this category and may result in congestive cardiac failure. It is therefore prudent to screen the cardiovascular status of patients by means of ECG and echocardiography prior to commencement of treatment with these drugs
- Severe diarrhoea: This is peculiar to treatment with Pertuzumab, Lapatinib and Neratinib
- Hand-foot syndrome: This is peculiar to treatment with Lapatinib. The hands and feet become sore, red and may blister and peel

Bevacizumab (Avastin). This drug works against the action of vascular endothelial growth factor (VEGF) which normally stimulates the formation and growth of new blood vessels. It may be combined with paclitaxel in the treatment of advanced breast cancer.

Targeted therapy for hormone-receptor-positive breast cancer: These drugs block proteins in the cell called cyclin-dependent kinases (CDKs) especially CDK4 and CDK6. This inhibits the process of cell division in hormone receptor positive breast cancer thereby slowing cancer growth.

The drugs include Palbociclib (brance), ribociclib and abemaciclib (Verzenio). They are indicated in postmenopausal and advanced hormone receptor positive/HER2 negative breast cancer and come in form of oral pills. Treatment is for 3 weeks. The side effects are pancytopenia (patient is prone to infection), nausea/vomiting and fatigue.

Everolimus (Afinitor): This blocks mTOR, a protein in cells that normally helps them grow and divide. Everolimus blocks the energy supply to the cells and may also prevent the development of new blood vessels. It is indicated in postmenopausal and advanced HR+and HER2-ve breast cancers. It may be used in combination with aromatase inhibitor, exemestane (aromasin). It comes in once daily oral preparation. Side effects include mouth sores, nausea, diarrhoea and pancytopenia. Others are increased cholesterol/triglycerides and increased blood sugar level.

Radiotherapy in early breast cancer has been found useful in the following situations

- As an adjunct treatment after breast conserving surgery
- As an adjunct treatment in node-positive patients: There is an ever present risk of lymphoedema of the arm coupled with damage to the brachial plexus
- As the primary treatment in inflammatory breast cancer
- As an adjunct treatment of the chest wall after mastectomy particularly in the presence of risk factors for recurrence: large tumour, grade 3 tumour, axillary lymph node involvement and evidence of vascular invasion.
- As a palliative treatment in the management of metastatic bone lesions
- As neoadjuvant treatment prior to surgery

Radiotherapy reduces the risk of recurrent invasive breast cancer to 10%. There are three modes of administration of radiotherapy in breast cancer

- External breast irradiation: This involves the use of linear accelerator. The total dose of adjuvant radiotherapy is 4,500 to 5000 cGy divided into 200 fractions per day and 5 days a week over a period of 5 to 7 weeks. A local boost therapy (at a dose of 1000cGy) may be preferred following breast conservation therapy.
- Internal partial breast radiation (brachytherapy): This involves the temporary placement in the breast of seeds or pellets of radioactive materials. Compared to external irradiation, the duration of treatment is shorter
- External partial-breast irradiation: This zeroes in on the site of cancerous growth. Duration of treatment is quite short: 5 to 10 days. This modality is still undergoing research.

Management of advanced breast cancer

The term, advanced breast cancer, refers to both metastatic disease as well as recurrent breast cancer. The latter is defined as the return of cancer in the same or contralateral breast or chest wall after a period of time when there was no evidence of cancer. Recurrence of breast cancer may be local, regional or locoregional. As opposed to early breast cancer whose aim of treatment is curative, management of advanced breast cancer is solely palliative. In this case, adjuvant treatment which in principle is administered as a secondary measure after surgery in the treatment of early breast cancer now plays a more frontline role. It is administered before the

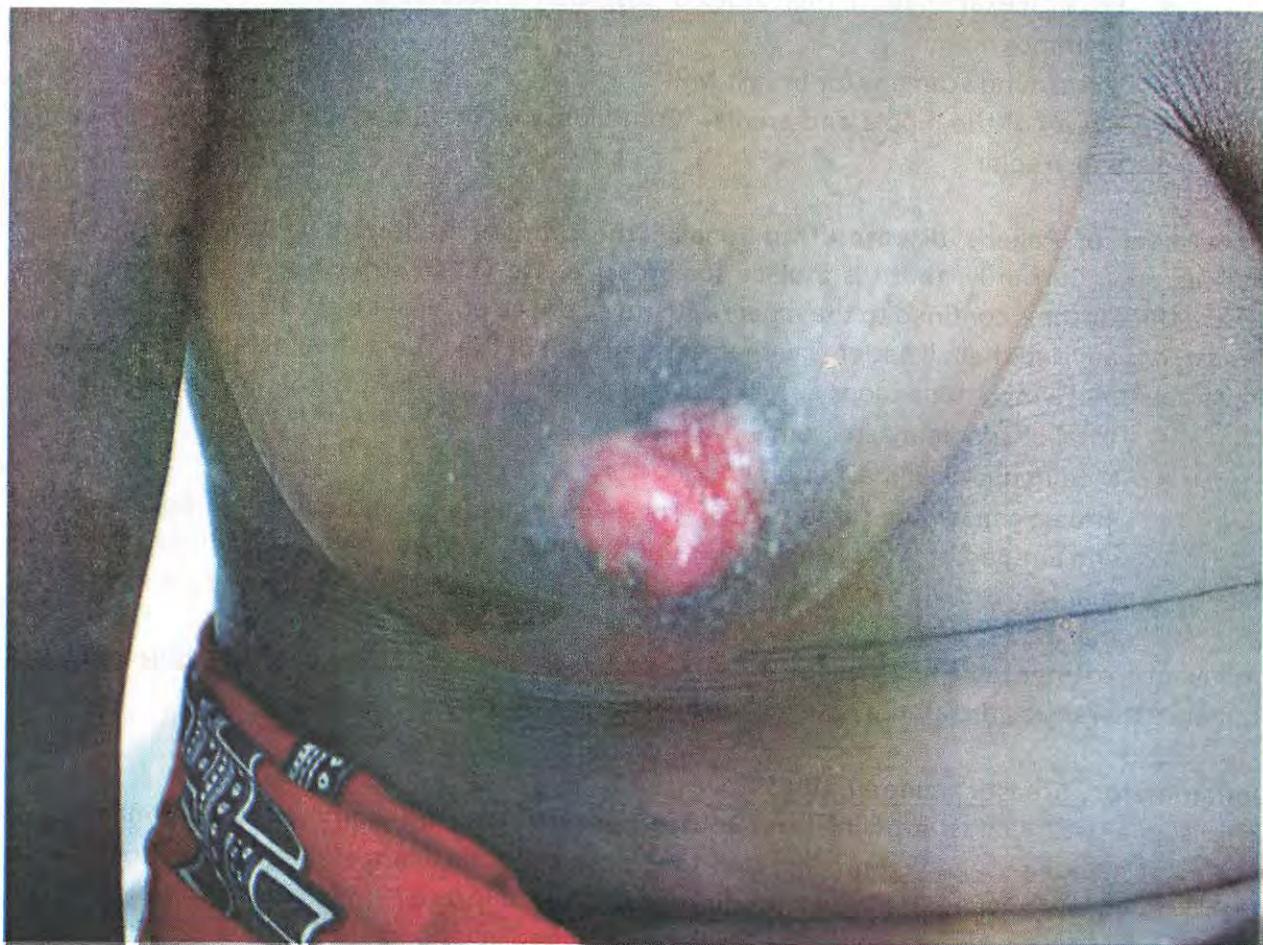
surgical treatment of advanced breast cancer. This is the principle of neoadjuvant therapy as chemotherapy is administered before surgery. Neoadjuvant therapy has been found useful in situations where the tumour is very gross and covers the entire breast (high tumour/breast ratio). Pre-operative chemotherapy tends to shrink the size of the tumour thereby downstaging and rendering an initially inoperable tumour, operable. In the case of smaller tumours, neoadjuvant therapy downstages and reduces the size of the tumour thereby rendering it more amenable to breast-conserving surgery. The subsequent surgical treatment in the case of large tumours is essentially palliative and consists of a total (simple) mastectomy. Chemotherapy and hormonal manipulation are continued after surgery.

Management of the metastatic complications constitutes an integral part of the treatment of late breast cancer. It usually involves a joint management with other surgical specialists.

- Malignant pleural effusion is drained by either chest aspiration if minimal, or by thoracostomy if massive. Pleurodesis may be considered if recurrent. This aims at obliterating the pleural space by the instillation of a substance such as tetracycline or talc that induces an inflammatory reaction. Pleurodesis is associated with transient pain and occasional pyrexia
- Metastasis to the bones (usually of the lower spine or cervical spine) may result in compression of the spinal cord. This presents as either paraparesis or paraplegia. Management of this condition may involve both the orthopaedic and the neurosurgical teams as the treatment may range from spinal decompression (laminectomy) to internal fixation. Irradiation of the spine is treatment of choice for pain associated with spinal involvement.
- Metastasis to the CNS: CT and MRI are useful in the evaluation of focal neurological symptoms. The initial treatment is to reduce the oedema by the use of steroids. Fractionated radiotherapy may help to stabilise the condition. Solitary metastasis may be excised by neurosurgery. Palliative chemotherapy may be useful for the control of both cerebral and systemic metastases. Sadly, CNS metastasis is an end-game phenomenon as most patients do not live long.
- Pathological fracture: This is heralded by a sharp increase in pain over the affected area. Treatment is by internal fixation followed by radiotherapy. It is best treated prophylactically before the fracture occurs by same method.
- Hypercalcaemia: Occurs with metastases to the bone: Cytokines released by the breast cancer cells activate the osteoclastic activity in the bone and release calcium into the circulation. The resultant hypercalcaemia can result in renal failure, pancreatitis, cardiac arrhythmias and psychosis. Treatment consists of adequate hydration and administration of bisphosphonates and chemotherapy. The affected bone may require external beam irradiation.

SPECIAL FORMS OF BREAST CANCER

Paget's disease of the nipple-areolar complex:



PAGET'S DISEASE OF THE NIPPLE-AREOLAR COMPLEX

Paget's disease of the nipple and areolar accounts for less than 5% of all breast cancers. The latter may be in the form of either a local ductal carcinoma in situ (intraductal carcinoma) or invasive breast cancer elsewhere in the ipsilateral breast. There are two theories in relation to the pathogenesis of Paget's disease of the breast

- Spread of invasive ductal carcinoma located elsewhere in the ipsilateral breast to the nipple-areolar complex resulting in the clinical condition seen in the latter
- Primary lesion starts locally around the nipple-areolar complex. Any other synchronous cancer in the ipsilateral breast develops independent of the cancer around the nipple-areolar complex

Clinical features: It is a disease of middle aged and elderly as it develops after the age of 50. The mean age of occurrence differs in both sexes: 62 for men and 69 for women. The initial presentation is with a red, sore, scaly or flaky recurrent lesion around the nipple. This is later followed by an itching, burning and/or tingling sensation. There is associated pain, scaling and flattening of the nipple. These may be accompanied by a yellowish or bloody nipple discharge. The clinical picture raises a differential diagnosis of eczema. It should, however, be noted that

whereas Paget's disease involves both the nipple and the areolar and is usually unilateral, eczema only affects the areolar and may be bilateral. Diagnosis is by way of the following

- Physical examination: High index of suspicion is necessary
- Mammography
- Ultrasound scan and/or breast MRI
- Biopsy of the nipple and areolar: Core biopsy or preferably punch biopsy of the skin and areolar

Treatment of Paget's disease: Traditionally, the primary treatment is surgical by way of mastectomy. Currently, there is a place for breast conservative surgery in about 5% of cases where the cancer is confined to the nipple with no evidence of cancer elsewhere in the ipsilateral breast. It is advised that all patients with Paget's disease of the breast should as much as possible have a sentinel lymph node biopsy irrespective of nodal status. The surgical options are

- Total or simple mastectomy
- Modified radical mastectomy
- Breast conserving surgery (BCS): This involves excision of the nipple-areolar complex and lumpectomy. As usual, this is followed up with radiotherapy. Studies have shown that in carefully selected cases, the outcome of BCS and radiotherapy is as good as mastectomy

Adjuvant treatment like in other forms of breast cancer involves chemotherapy, hormonal and targeted therapy as dictated by the intrinsic properties of the primary lesion.

Inflammatory breast carcinoma (IBC)

This is an aggressive fast-growing tumour that presents like an inflammatory lesion (mastitis). It occurs at a younger age when compared to the typical breast cancer and is more common in blacks. Being overweight is a known predisposing factor. It is rare in men. There is redness of the whole breast with associated oedema. The latter is due to invasion of the dermal lymphatics. Just like mastitis, no definite breast lump is discernable. Unlike mastitis, however, there are skin changes (peau d'orange) with no associated fever in inflammatory breast cancer.

Diagnosis is made by a high index of suspicion. Investigations are

- Skin punch biopsy: There may be no discrete lump
- Ultrasound-guided core-needle biopsy
- Breast MRI: Considered the most reliable test for IBC
- Staging investigations: Chest X-ray, CT scan (chest, abdomen and pelvis), bone scan and liver function tests. The use of PET/CT scan is still under investigation.
- Hormone receptor status: Most are receptor negative
- HER2 receptor status: Many are HER2 positive

Treatment of inflammatory breast cancer: It must be pointed out that surgery is not the first-line treatment. Neoadjuvant therapy is imperative as it shrinks the cancer, reduces the swelling and restores normal skin texture. These render the breast tissue easier to handle during the subsequent surgery. Wound healing is also enhanced.

Chemotherapeutic agents: Combination of anthracycline and taxane preferred. Anthracyclines include adriamycin (doxorubicin) and epirubicin (ellence) while taxanes include paclitaxel (taxol), docetaxel (taxotere) and nab paclitaxel (abraxane-albumin bound)

Targeted therapy: Herceptin (blocks growth effect of HER2 on the cell surface) and Lapatinib (blocks the effect of HER2 protein from inside the cell), slow or stop the growth of malignant cells

Surgery: Modified radical mastectomy. Breast conserving surgery is contraindicated

Radiotherapy: Administered after surgery to the breast and chest wall

Prognosis: The prognosis of inflammatory breast cancer is poor

Pregnancy associated breast cancer (PABC):

This is defined as breast cancer that occurs during pregnancy or within one year of delivery. It constitutes 1-2% of all breast cancers. Diagnosis is made by history and physical examination. The extensive parenchymal changes associated with pregnancy renders mammography less reliable in the diagnosis of PABC. Contrary to the popularly held view, pregnancy does not worsen the prognosis of breast cancer. Rather, the engorgement of the breasts during pregnancy and lactation makes it more difficult to pick up a breast lump. Stage for stage, the prognosis is not worse in pregnancy.

Breast surgery can be done safely in all trimesters of pregnancy. Nevertheless, many surgeons will rather wait until the end of the first trimester when the rate of spontaneous abortion is lower. Chemotherapy, radiotherapy and even hormonal manipulation are contraindicated in the first trimester of pregnancy as these modalities of treatment are associated with risk of foetal damage. Termination of pregnancy has not been shown to improve the prognosis.

In the second trimester, PABC can be treated with surgery. Chemotherapy may be administered but radiotherapy should be avoided in this trimester and indeed at all times during pregnancy.

In addition to treating the mother for breast cancer, treatment during the third trimester of pregnancy aims at the delivery of a relatively mature live baby. Treatment may therefore be delayed till 32 weeks of gestation when patient is delivered of the baby. Full treatment is thereafter commenced by surgery, chemotherapy, hormonal manipulation, and radiotherapy. PABC is usually associated with ER/PR negative tumours. Therefore hormonal treatment is not effective in most patients with PABC. Since recurrence is usually evident within the first two years of treatment for breast cancer, patients treated for breast cancer in pregnancy are advised not to get pregnant within this period. This is to facilitate early diagnosis of recurrence.

Bilateral breast cancer:

This form of breast cancer may suggest a genetic aetiology. A previous breast cancer is also a risk factor for cancer involving the other breast. Two forms of this condition have been described based on the timing of the manifestation: synchronous and metachronous. Synchronous bilateral breast cancer is said to occur when both cancers are diagnosed during the same hospital visit while the metachronous variety is said to occur when they are diagnosed separately. The importance of keeping a close watch on the other breast after management of cancer in one affected breast cannot be overemphasised.

Male breast cancer:

Male breast cancer constitutes about 1-2% in Western countries and 2-5% in Africa of all breast cancers. The risk factors for male breast cancer are Klinefelter's syndrome, gynaecomastia and

mutation in BRCA 2 gene. The clinical presentation is not different from that of their female counterparts. Just like females in our environment, male breast cancers equally present late. Spread is, however, faster owing to the rudimentary nature of the male breast. Clinicians should therefore look beyond the diagnosis of gynaecomastia when attending to male patients with breast lesions. As benign as a male breast lesion may appear, there may be an underlying, more ominous pathology.



Male right breast cancer

Treatment: As in the female variety, definitive treatment is total mastectomy. Adjuvant treatment involves chemotherapy and radiotherapy. Most male cancers are hormone receptor positive (70% ER positive and 65% PR positive) and therefore respond well to adjuvant tamoxifen therapy. Other forms of hormonal therapy in male breast cancer are luteinising hormone releasing hormone analogue (goserelin) and aromatase inhibitors (letrozole or anastrozole)

Ductal carcinoma in-situ (DCIS):

Also known as intraduct carcinoma, DCIS is the most common type of non-invasive breast cancer. This is a very early form of breast cancer in which the basement membrane is intact. About 60,000 new cases are diagnosed in the USA annually. This accounts for 1 in every 5 new breast cancer cases. The increased incidence may be attributed to the following

- Increased longevity: Risk of breast cancer increases with age
- Mammography: There is an increased rate of mammography of even better quality. 80% of cases are diagnosed on mammography

There is a risk of subsequent development of invasive breast cancer in the ipsilateral breast. Usually non-symptomatic but occasionally, patient may present with breast symptoms. Clinical diagnostic indicators are bloody nipple discharge, Paget's disease of the nipple and a palpable lump. Most often, however, DCIS is only detected as microcalcification confined to the ducts on routine mammography. It shows as a fine linear or branching pattern on mammography.

Confirmation of diagnosis is usually made by either fine needle aspiration cytology or core biopsy (may be ultrasound-guided). The following are indications for open biopsy

- Failure of needle biopsy to extract sufficient/representative tissue for proper histological examination
- Inconclusive histology report

Granted that all DCIS are regarded as stage 0 disease, the pathologist is still expected to report on the type/grade of the DCIS as well as its hormone receptor status. The various grades are

- Normal cells
- Ductal hyperplasia: has too many cells
- DCIS proper: Too many cells having features of cancer but these are confined to the ducts
- DCIS-M: This is DCIS with microinvasion. A few of the cancer cells have started to break through the wall of the duct. Slightly more serious than DCIS proper
- Invasive ductal carcinoma: Cancer cells have eventually broken through the wall of the duct

Cell grading: Like all breast cancers, the cells are categorised into

- Low or grade 1
- Moderate or grade 11
- High or grade 111

Low grade and high grade DCIS have the lowest and highest rates respectively of developing into invasive ductal carcinoma

Treatment: DCIS being a very early stage of breast cancer has raised a management dilemma. Much as the clinician would want to pre-empt the development of a full-blown invasive ductal carcinoma, he faces the possible dilemma of 'over-treating' the patient. The following are the treatment options of DCIS

- Lumpectomy (with wide excision): Traditionally this involves a wide excision with an adequate margin of at least 10mm. Histological confirmation of the edges being free of cancer is mandatory. This is naturally followed by irradiation of the breast. The latter may be by external beam radiation or brachytherapy. External partial-breast irradiation that zeroes in on the area of DCIS has been successfully carried out

* Simple or total mastectomy: Indications are

- Large area of DCIS
- Multifocal disease
- Acceptable margin not achievable without deforming breast
- Strong family history of breast cancer
- Positive gene mutation
- Contraindication to radiotherapy: Previous radiation, 1st trimester of pregnancy and hypersensitivity to radiotherapy

There is no indication for axillary node dissection. Sentinel node biopsy may, however, be indicated in the younger age group (< 40), microinvasion, high grade and widespread DCIS

- Lumpectomy alone without radiotherapy: May be indicated when there is a small area of DCIS and when the lesion occurs in patients above 70 with medical conditions. Patient should be closely followed up with mammography, USS and MRI

- Hormonal therapy: As indicated by the receptor assay

Clinicians at the University of Southern California, Van Nuys have developed a formula that attempts to objectively determine the aggressiveness of DCIS in terms of the likelihood of local recurrence following 'breast conserving' surgeries. Factors that influence the aggressiveness of DCIS are

- Overall tumour size
- Pathological 'nuclear grade' including presence or absence of necrosis
- Width of clear 'surgical margins': This is the thickness of the unaffected tissue
- Age of patient: A recent addition to the above three criteria

Van Nuy Prognostic scoring system

Criteria	1 score	2 score	3 score
Tumour size (diameter in mm)	Less or equal to 15	16-40	> or = 41
Margin width	< or = 10	1 - 9	< 1

Pathological classification

	Non-high grade (1,2) No necrosis	Non-high grade (1,2) With necrosis	High grade (3) + or - necrosis
Age	61 or older	40 - 60	< or = 39

Final score is between 4 and 12. This is related to the ultimate prognosis

Score	Chance of recurrence	5 year survival	Recommended treatment
4 - 6	1%	99%	Solely wide local excision
7 - 9	20%	84%	Excision + radiotherapy
10 - 12	50%	51%	Mastectomy

Other indications for mastectomy in DCIS are

- Tumour larger than 4cm
- Multicentric tumours occurring in more than one breast quadrant
- Location of tumour: May be such that wide excision with a negative margin is difficult
- Associated Paget's disease

Follow-up: Regular physical examination (6 to 12 monthly for 5 years, then annually). This is complemented by a 12 monthly mammography

Inherited breast cancer: Features include

- A strong family history
- BRCA1 and BRCA2 genes located on chromosomes 17 and 13 respectively are thought to account for the majority of inherited breast cancers. The estimated lifetime risks of developing breast cancer among BRCA1 and 2 are 47%-66% and 40-57% respectively compared to a 12.5% risk among the population. Patients harbouring these genes are also at a higher risk for the development of ovarian cancers, bilateral breast cancers and male breast cancer (BRCA2).

Intensive surveillance is therefore recommended for high risk. This involves

- * Self breast examination from the age of 18
- * Clinical breast examination 6 to 12 monthly from the age of 25
- * Annual mammography from the age of 25
- * Annual bilateral breast magnetic resonance imaging (MRI) as an adjunct to mammography.

Management options include chemoprevention and prophylactic surgery (bilateral subcutaneous mastectomy, and bilateral salpingo-oophorectomy). Surgery is performed at 5 years younger than the youngest family relative to have developed breast cancer. Tamoxifen remains the only approved chemoprophylactic agent

Phyllodes tumour of the breast (Cystosarcoma phyllodes):

It is a rare fibroepithelial breast tumour and accounts for less than 1% of all breast tumours. Its name is derived from the Greek word 'phyllodes' due to its leaf-like pattern. The overall incidence is 2.1/million while its peak age incidence is 45–49. Preoperative diagnosis of phyllodes tumour is usually difficult as it is clinically indistinguishable from fibroadenoma. Phyllodes tumours are indeed considered to be on a disease spectrum that consists of fibroadenoma, fibroadenoma variant and benign phyllodes. Some have even tried to extend it to include malignant phyllodes and frank sarcoma.

Aetiology: Many have come to the conclusion that phyllodes tumour is a hyperplastic rather than a neoplastic lesion. It is thought that in a proportion of fibroadenoma, a somatic mutation may result in a monoclonal proliferation histologically indistinguishable from the polyclonal element, but with a propensity to local recurrence. This eventually progresses to a phyllodes tumour. Endothelin-1, a stimulator of fibroblast growth in the breast may be responsible. Tumour growth may be stimulated by trauma, pregnancy, lactation and indeed any factor that increases the secretion of oestrogen.

Pathology: Phyllodes tumour involves the connective tissue (stroma) of the breast. It is histologically graded according to the degree of cellular atypism and mitotic figures into benign (low grade), borderline and malignant (high grade). The greater the degree of cellular atypism, mitotic figures, and stromal cellularity, the more malignant the clinical course and consequently the more aggressive the treatment required. The nature of the tumour margin is also taken into consideration. Azzopardi et al and Salvadori et al have come up with a modification of the WHO classification based on the above criteria

Criteria	Benign	Borderline	Malignant
Tumour margin	Pushing	Pushing and infiltrative	Infiltrative
Stromal cellularity	Low	Moderate	High
Mitotic rate	< 5	5 – 9	10 or more per 10 HPF
Pleomorphism	Mild	Moderate	Severe

About 85 to 90% of phyllodes tumours are benign while 10 to 15% are malignant. The latter are characterised by a high incidence of recurrence and metastatic propensity to bones, liver, lungs and heart. It rarely spreads to the axillary lymph nodes.

Phyllodes tumour differs from fibroadenoma by virtue of its high stromal cellularity and increased mitotic activity. On the other hand, there is an exaggeration of the intracanalicular growth pattern in fibroadenoma.

Clinical presentation: As mentioned above, the clinical dilemma is to differentiate phyllodes tumour from fibroadenoma. The history is short relatively to the size of the lesion as phyllodes tumours are fast-growing. They are usually painless and occasionally ulcerate. Fibroadenoma is commoner in the younger age group and has a lower peak age incidence of 30 to 40 while that of phyllodes is 40 to 50. Any lesion of 10 cm diameter and above is referred to as giant phyllodes tumour.

Differential diagnosis: This includes giant fibroadenoma, lipoma, adenoma, and hamartoma. Others are carcinoma, juvenile papillomatosis, virginal hypertrophy, sarcoma and metastatic tumour

Investigations: Radiological and histological

- Mammography: Will demonstrate a large, round or oval mass with well-defined edges. There may be associated calcification
- Ultrasound scan of the breast: Well-defined cystic masses
- MRI: Will yield additional information on the nature of the lesion
- Biopsy: Excisional biopsy is preferred as core biopsy may not provide enough specimen. Histology will reveal a bunch of rapidly dividing cells incorporating an overgrowth of stromal cells. There may be no evidence whatsoever of epithelial cells. The lesion demonstrates an intermediate appearance between a benign condition and sarcoma. Three histological forms exist depending on the degree of cellularity: benign, intermediate (borderline) and malignant

Treatment of phyllodes tumour: Surgery is the mainstay of treatment and entails a wide local resection with at least a 1 cm margin. The latter should ideally be confirmed by the pathologist. Indications for mastectomy include a

- Large tumour size in a relatively small breast
- Recurrent tumour. These make it impossible to have a wide enough resection whose margins are clear of tumour. Mastectomy is also indicated in
- Proven malignant phyllodes tumour

Mastectomy may either be partial (segmental), or total (simple). Axillary lymph node dissection is not indicated. Hence, the patients with the low grade variety who present with clinical features suggestive of fibroadenoma are managed by wide excision with subsequent confirmation of tumour free margins. On the other hand, patients with either the borderline or the malignant variety usually present with clinical features suggestive of breast cancer and are managed with mastectomy. It is advised that even when there is an apparent low degree of cellular atypism and mitotic figures, a generous wide local excision is advocated owing to the very high recurrence rate. The gloomy aspect of recurrence is the likelihood that histological transformation may take place. Progression to a histologically more aggressive-looking tumour may augur a correspondingly more aggressive clinical behaviour. It has, however, been argued that the 'recurrence' may arise from an area of malignancy in the original lesion that was missed during the initial assessment. Thus, local recurrence may be regarded as 'failure of the original surgery'

Adjuvant therapy does not play a significant role in the management of phyllodes tumour. Postoperative radiotherapy should be considered after mastectomy if there is involvement of the resection margin. It may also be of use in the prevention of local recurrence. Chemotherapeutic combinations such as cisplatin and etoposide or doxorubicin and ifosfamide have been tried in metastatic disease but generally found to have no survival advantage. Even though oestrogen and progesterone receptors have been identified in 40 to 50% of phyllodes tumour, the use of hormonal therapy has not been fully investigated.

Role of tumour markers in phyllodes tumour: An increased expression of p53 protein and Ki-67 has been detected. This may eventually be of help in differentiating fibroadenoma from phyllodes tumour. Their levels have been found to correlate with negative prognostic factors. There are presently no reliable indices for the latter.

Prognosis of breast cancer:

As discussed above, it is only the early breast cancer that is amenable to curative treatment. It follows therefore that the earlier the diagnosis is made, the better the prognosis. The following affect the outcome of treatment of breast cancer.

- Initial size of tumour at presentation: the smaller the size, the better the prognosis
- Axillary nodal involvement: prognosis is worse with axillary nodal involvement
- Histological grade of the tumour
- Vascular invasion: connotes a poor prognosis
- Hormone receptor status: oestrogen/progesterone receptor positive tumours tend to have a better prognosis as there are more treatment options. On the other hand, HER-2 neu positivity is a poor prognostic factor
- Histological type: Specific histological types such as mucinous, medullary, papillary and medullary types of cancer seem to have a better prognosis than the purely invasive ductal carcinoma.

Nottingham Prognostic Index (NPI): This aims at prognosticating the outcome of a particular case of breast cancer by combining the above factors. It is a sum total of

- One-fifth the size of the tumour in centimetres
- Lymph node stage: 1=no node involved; 2=one to three nodes involved; 3=four or more nodes involved
- Histological grade of tumour: grade1-low grade; grade2-intermediate grade; grade3-high grade

Interpretation of NPI analysis

3 or less: good prognostic outcome

6 and above: poor prognostic outcome

The prognostic index acts as a guide to the clinician as to the direction of treatment of the individual patient. Whereas he would aim at a curative treatment for patients with good prognostic outcome, the aim is only palliative when the indices are only compatible with a poor prognostic outcome.

Molecular subtypes of breast cancer

The classification of breast cancer into intrinsic and molecular subtypes is based on the genes a cancer expresses. The latter will influence the receptor status of a breast neoplastic lesion. On this basis, there are five subtypes

1 Luminal A: Has the following characteristics

- Hormone receptor (oestrogen and progesterone) positive
- HER2 negative
- Low levels of Ki-67 which helps to control the rate of growth of cancer cells.
Luminal A breast cancers are low grade, tend to grow slowly and have the best prognosis

Luminal B breast cancer

- Hormone receptor (oestrogen and progesterone) positive
- Either HER2 positive or HER2 negative
- High levels of Ki-67: This is a nuclear protein associated with and necessary for cellular proliferation.

Luminal B breast cancers generally grow slightly faster than luminal A cancers and their prognosis is slightly worse

Triple-negative/baseline breast cancer

- Hormone-receptor (oestrogen and progesterone) negative
- HER2 negative
- This type of breast cancer is more common in young African women with BRCA1 gene mutation and is discussed in more detail below

HER2- enriched breast cancer

- Hormone receptor (oestrogen and progesterone) negative
- HER2 positive
- Grows faster than luminal carcinomas
- Worse prognosis than luminal carcinomas
- Often successfully treated by targeted therapy aimed at HER2 protein

Normal-like breast cancer

- Similar to luminal A
- Hormone receptor (oestrogen and progesterone) positive
- HER2 negative
- Low levels of Ki-67
- Good prognosis but not as good as that of luminal A

TRIPLE-NEGATIVE BREAST CANCERS (TNBC): These are breast cancers found to be negative for all the usually assayed receptors: oestrogen, progesterone and HER-2 neu. They account for 15% to 20% of breast cancers and are found to have the following characteristics:

- Occur before the age of 45 (premenopausal)
- Common in Africans and Hispanics
- Represent about 10% to 15% of all breast cancers
- Lower socioeconomic status

- Exhibit high histological grade
- Have a high risk of relapse irrespective of histological and clinical grade
- Account for a large proportion of metastatic cancer: Brain and viscera

It has been found that while increased parity and early age at first childbirth have historically been associated with reduction in breast cancer risk, these factors on the other hand, are associated with an increased risk of TNBC.

Cytotoxic chemotherapy is the mainstay of treatment for TNBC as they are chemosensitive. A number of studies have demonstrated the efficacy of chemotherapy in the adjuvant, neoadjuvant and metastatic disease settings. Options include the combination of anthracyclines and taxanes. Platinum based chemotherapy and capecitabine have been found to be effective in the management of TNBC. Some targeted molecular therapies have been found to be effective in the management of TNBC. They include polyadenosine diphosphate ribose polymerase (PARP)-1 inhibitors and tyrosine kinase inhibitors. The former has been found to be more effective when combined with the above chemotherapeutic regimen.

Prognosis of TNBC: Despite the fact that it accounts for only 10% to 15% of breast cancers, it is associated with a disproportionate number of breast cancer-related deaths. Even after adjusting for size, grade and nodal-status, TNBC is an independent predictor of poor outcome

EARLY-ONSET BREAST CANCER:

In the United States, breast cancer occurring over the age of 45 is commoner in whites than in blacks. On the other hand, black women under 35 have more than double the incidence of invasive breast cancer and thrice the breast cancer mortality of young white women. The following features are associated with early breast cancer

- Positive family history (BRCA1, BRCA2 and p53 mutations)
- High body mass index: Obesity, high energy intake and sedentary lifestyle in premenopausal women
- Recent oral contraceptive use is a risk factor for early-onset breast cancer unlike in older women. This is a relevant factor especially in oestrogen-negative tumours
- Early childbearing and multiparity particularly for women below 35. There appears to be a short-term elevation of breast cancer risk for several months immediately following childbirth.
- Prior mantle irradiation for Hodgkin's disease
- Early age at menarche
- Heavy consumption of alcohol
- High intake of red meat
- High mammographic density
- Low calcium or vitamin D intake

Fruits and vegetables have been associated with a decreased incidence of breast cancer risk in premenopausal women.

Clinicopathological features

- Triple-negative phenotype: more aggressive, larger, of higher cellular grade, and associated with a high lymph node positivity rate
- Gene Set Enrichment Analysis (GSEA) shows 367 significant sets enriched among young women's tumours, specifically distinguishing them from tumours arising in older women: defective immune function and apoptosis, hypoxia, and BRCA1 stem cells. Others include multiple targeted oncogenic signalling pathways, and mammalian target of rapamycin (mTOR).

Treatment: Breast-conserving surgery, whenever feasible, is obviously the preferred option in early breast cancer involving young women. The principal consideration, however, is the higher risk of local recurrence after conservative surgical management in patients below 35 as compared to older patients above 60. Options of adjuvant treatment include radiotherapy, cytotoxic chemotherapy, ovarian ablation (surgery, irradiation, chemical) and anti-oestrogen therapy.

Anthracycline-based chemotherapy (CAF) has been found to reduce recurrence by 35% and death by 27%. Clinical studies have shown that patients with node-positive early breast cancer given four cycles of AC followed by four cycles of paclitaxel demonstrated a 17% reduction in the risk of recurrence and an 18% reduction in the risk of death compared with four cycles of AC alone.

Challenges of adjuvant treatment in early-onset breast cancer

- Induction of an early menopause: Induced amenorrhoea may have a better prognosis than those retaining their menstrual cycle. It follows that amenorrhoea may be an important factor in the action of chemotherapeutic agents
- Fertility impairment
- Adverse effects on bone marrow density with chemotherapy and endocrine therapy
- Development of second malignancy with radiotherapy.

Psychological issues in early onset breast cancer: Depression following the diagnosis of breast cancer cuts across all age groups. It is, however, more pronounced in younger patients. It presents with a variety of unique psychosocial and emotional changes including but not limited to interactions with spouse/children, body image, sexuality and loss of fertility/premature menopause. The younger the patient at the time of diagnosis, the higher the negative psychological impact of breast cancer. It is thought that psychological issues may enhance cancer recurrence. There is therefore a need for the services of a psychologist as part of the oncology team.

Prevention of breast cancer

It is difficult to completely prevent the development of breast cancer as the condition is already genetically programmed in some individuals. This is with reference to persons with a family history of the disease. They are likely to carry the predisposing genes such as BRCA1 and BRCA2. The aim in such persons, as in all adult women, consists of early detection of breast cancer and

improved survival in view of early treatment. On a positive note, the patient is reassured with confidence if screening shows no evidence of cancer. Routine screening for breast cancer is carried out by a combination of the following procedures:

- **Breast self examination (BSE)**
- **Clinician breast examination (CBE):** Can detect cancer of about 1.5 cm in diameter
- **Routine screening mammography:** Can detect cancer as small as 0.5 cm in diameter
- **Yearly breast MRI in high risk women**

It has been found that healthy lifestyle which entails eating healthy foods, avoiding smoking and alcohol, and regular exercise help to reduce the development of cancers in general. Modalities that have been employed in preventing the genesis of breast cancer in patients whose genetic constitution carries a high risk of cancer include bilateral mastectomy with immediate reconstruction, oophorectomy, and the prophylactic use of hormone manipulating drugs such as tamoxifen and aromatase inhibitors.

Breast cancer risk assessment tool (Gail)

This is an interactive tool to help estimate a woman's risk of developing breast cancer. It was designed by scientists at the National Cancer Institute (NCI) and the National Surgical Adjuvant Breast and Bowel Project (NSABP) to estimate a woman's risk of developing invasive breast cancer during the next 5-year period and up to 90 (life time risk). It is a statistical model named after Dr Mitchell Gail, Senior Investigator in the Biostatistics Branch of NCI's Division of Cancer Epidemiology and Genetics. It is upgraded periodically in light of new data and research. The following criteria are used to derive what is popularly referred to as the Gail Index

- Age (risk for women 35 or older)
- Age at the time of first menstrual period (menarche)
- Age at first live birth
- Family history of breast cancer or of ductal carcinoma in situ (DCIS) or lobular carcinoma in situ (LICS)
- Number of first degree relatives that have had breast cancer (mother, sisters, daughters)
- Previous radiation to the chest for treatment of Hodgkin's lymphoma
- Mutation in either BRCA1 or BRCA2 gene or diagnosis of a genetic syndrome that may be associated with elevated risk of breast cancer
- Previous breast biopsy (number, diagnosis of atypical hyperplasia)
- Race, subrace and ethnicity

The outcome of analysis should be regarded solely as predictions as it is difficult to accurately predict precisely which woman will develop breast cancer based on the application of this instrument. In fact, some women who do not develop breast cancer have been found to have higher risk estimates than some women who do.

Chemoprevention of breast cancer:

These are medications used to reduce the risk of breast cancer in women at high risk of developing the disease. Such women may remain cancer-free while on these medications. As mentioned earlier, they are hormone manipulating drugs such as tamoxifen and aromatase inhibitors.

Tamoxifen in breast cancer chemoprevention

- it reduces the risk in both pre- and post-menopausal women at high risk of breast cancer by 50%.
- It is more effective in women with atypical hyperplasia as it reduces the risk by 86%.
- Due to the small sample size, women with LCIS have been shown to have a nonstatistically significant reduction of 56%
- Raloxifene is 76% as effective as tamoxifen in preventing breast cancer but has lower risks than tamoxifen

Aromatase inhibitors in breast cancer chemoprevention

- Exemestane reduces risk by 65%
- Anastrozole reduces risk by 56%
- Neither is associated with increased risk of thromboembolism, cardiovascular complications or other cancers

Barriers against chemoprevention

- Patient: Fear of side effects of chemopreventive drugs
- Clinician: Not enough time for proper counselling; challenges with selection of potential beneficiaries

Prophylactic surgical procedures (Breast cancer reducing surgery):

It comprises essentially of two procedures: Bilateral mastectomy and bilateral salpingo-oophorectomy. Bilateral mastectomy reduces the breast cancer risk by at least 95% in BRCA I and II mutations and 90% in those with strong family history. Bilateral salpingo-oophorectomy on the other hand reduces breast cancer risk by 50% and ovarian cancer risk by 90%

Bilateral mastectomy: Involves either of two procedures

- Total mastectomy
- Subcutaneous mastectomy (with or without nipple-sparing)

Total mastectomy offers greater breast cancer risk reduction than subcutaneous mastectomy as less breast tissue is left behind. All said and done, none of the procedures is sure-proof as it is difficult to completely excise breast tissue

Bilateral salpingo-oophorectomy (prophylactic oophorectomy): This may be combined with bilateral mastectomy in premenopausal patients with high risk for breast cancer. The main complication remains premature menopause (surgical menopause) with all its associated problems.

Women at high risk with ipsilateral breast cancer may consider contralateral prophylactic mastectomy. This practice should, however, be discouraged in patients who are not at high risk

BENIGN BREAST CONDITIONS

As stated earlier, any patient presenting with a breast complaint should be investigated in a bid to rule out breast cancer. The bulk of breast lesions are, however, benign in nature.

Fibroadenoma:

It is the most common breast lump in young ladies below the age of 30. It is a slow-growing tumour. Histologically, it consists of an overgrowth of the stromal cells and made up of both fibrous and glandular tissue. There are two histological forms of fibroadenoma: Pericanalicular (hard on palpation, consists more of fibrous tissue and freely mobile) and intracanalicular (consists more of glandular tissue and relatively soft on palpation). The collagen is arranged into whorls.

Clinically, it presents as a well circumscribed lump which is freely mobile in the breast. In this regard, it is commonly referred to as the 'breast mouse'. Occasionally it could be quite big in size and consequently referred to as a giant fibroadenoma. Diagnosis is clinical and confirmation is by ultrasonography, FNAC and core biopsy. Treatment depends on the size of the tumour. Small lesions are closely observed and only excised if they demonstrate evidence of continuous growth.

Indications for surgery include

- Patients with large tumours (> 4 cm) ab initio. Giant fibroadenomas are more than 5 cm in diameter
- Small lesions at initial presentation which are found to increase in size during follow-up.
- Suspicious lumps that show atypical hyperplasia
- High suspicion of malignancy on clinical assessment.
- In situations where the patient is anxious and there are limited facilities for diagnosis and follow-up.

It is important to note that whereas conservative management of fibroadenoma is the norm in Western countries where there are facilities (FNAC) for follow up, this is not the case in our environment (Nigeria and Africa). Management of fibroadenoma in the latter is by excisional biopsy owing to paucity of facilities for FNAC

Mastitis:

This is an inflammation of the breast and consists of two types – puerperal/lactating and non-lactating. The puerperal/lactating form is the acute variety and is common in lactating mothers. Infection, usually staphylococcal, spreads to the breast tissue through a crack in the nipple. This sets up an inflammatory reaction. The affected breast thereafter manifests all the clinical features of inflammation - red, swollen, hot, and painful. If inadequately treated, it progresses to formation of a deep-seated abscess. Treatment involves administration of antibiotics and analgesics with the provision of adequate support for the affected breast. Breastfeeding should be suspended in the ipsilateral breast and the breast milk expressed either manually or by the use of a breast pump. Failure to respond to conservative management may indicate the formation of a breast abscess. This is confirmed by needle aspiration. Treatment entails

aspiration for small abscesses and open drainage for more florid ones. Care should be taken to break down all the loculi by the use of the finger during open drainage.

The non-lactating form is the chronic variety and arises sequel to mismanagement of acute mastitis particularly when an abscess is not adequately drained. Merely administering antibiotics in a case of breast abscess will lead to 'mummification' of the breast. The ensuing clinical picture may be confused with breast cancer. The abscess now forms an inflammatory swelling commonly referred to as an 'antibioma'. This may be clinically indistinguishable from breast cancer. Investigations should therefore be carried out to rule out breast cancer. Treatment involves a wide excision of the lump.

As pointed out earlier, mastitis commonly occurs in the lactating breast. Occurrence in the non-lactating breast should alert the clinician about the possibility of an inflammatory breast cancer. A classical example of chronic mastitis is tuberculous mastitis which presents with features of breast cancer (generalised swelling of the breast, peau d'orange and sinus formation). Treatment is with antituberculous drugs

Traumatic fat necrosis:

This results from trauma to the breast. The ensuing inflammatory reaction results in a clinical picture that may be confused with breast cancer. Diagnosis is made from the history. A core biopsy may be necessary in difficult cases. Treatment consists of reassurance, analgesics, adequate breast support and antibiotics. This is one of the breast lesions that are clinically indistinguishable from breast cancer. Others in this category include chronic mastitis and Mondor's disease of the breast

Aberration of Normal Development and Involution (ANDI):

This refers to a group of breast lesions that was previously referred to by many names such as fibrocystic disease of the breast, fibroadenosis and later benign mammary dysplasia. These names constitute an embodiment of the histological features – fibrosis, adenosis and formation of breast cysts. For quite some time the aetiology was not clear. It has now been found to be related to the cyclical hormonal changes that occur in the breasts. It is most prominent during the luteal phase of the menstrual cycle. The patient may present with mastalgia, and bilateral lumpiness of the breasts. Management involves ruling out breast cancer by way of proper history taking, and adequate clinical examination supported by ancillary investigations if necessary. The first line of treatment entails reassurance about the benign nature of the lesion. Various treatments have been employed. These include the administration of non-steroidal anti-inflammatory agents, danazol, oil of evening primrose (contains essential fatty acids), and adequate breast support. Others include the use of contraceptive pills with low dose oestrogen (20mg desogestrel) and goserelin (gonadotrophin-releasing hormone agonist). It has been discovered that avoidance of caffeine-containing products proves to be useful in some patients. Regardless of the treatment modality employed, adequate follow-up is necessary. Any subsequent palpable lump of doubtful origin should be biopsied for histological examination to rule out breast cancer.

Mastalgia of non-breast origin:

The breast lies on the chest wall and pain may originate due to pathology in any of the structures that have a relationship with the chest wall. These include

- Pain (Localised or diffuse) arising from the lateral chest wall
- Mondor's disease
- Ischemic heart disease
- Radicular pain
- Pain arising from pathology of the liver and gallbladder
- Tietze's syndrome: This is commoner in females. It is costochondritis of the second costal cartilage.

Cystic breast swellings

The commonest cause is benign mammary dysplasia. Management involves a diagnostic aspiration of the cyst. Whereas the aspirate is usually straw coloured or greenish in benign mammary dysplasia, it usually contains milk in galactocoele. A biopsy to rule out an underlying breast cancer is advised under the following circumstances

- Initial bloody aspirate
- Palpable residual mass after aspiration
- Refilling of the cyst after initial aspiration.

Mondor's disease:

This results from thrombosis of the superficial veins of the chest wall. Presentation is that of a painful, cord-like swelling, that runs longitudinally over the chest wall. Treatment involves reassurance, anti-inflammatory drugs and adequate breast support. It normally runs a self-limiting course.

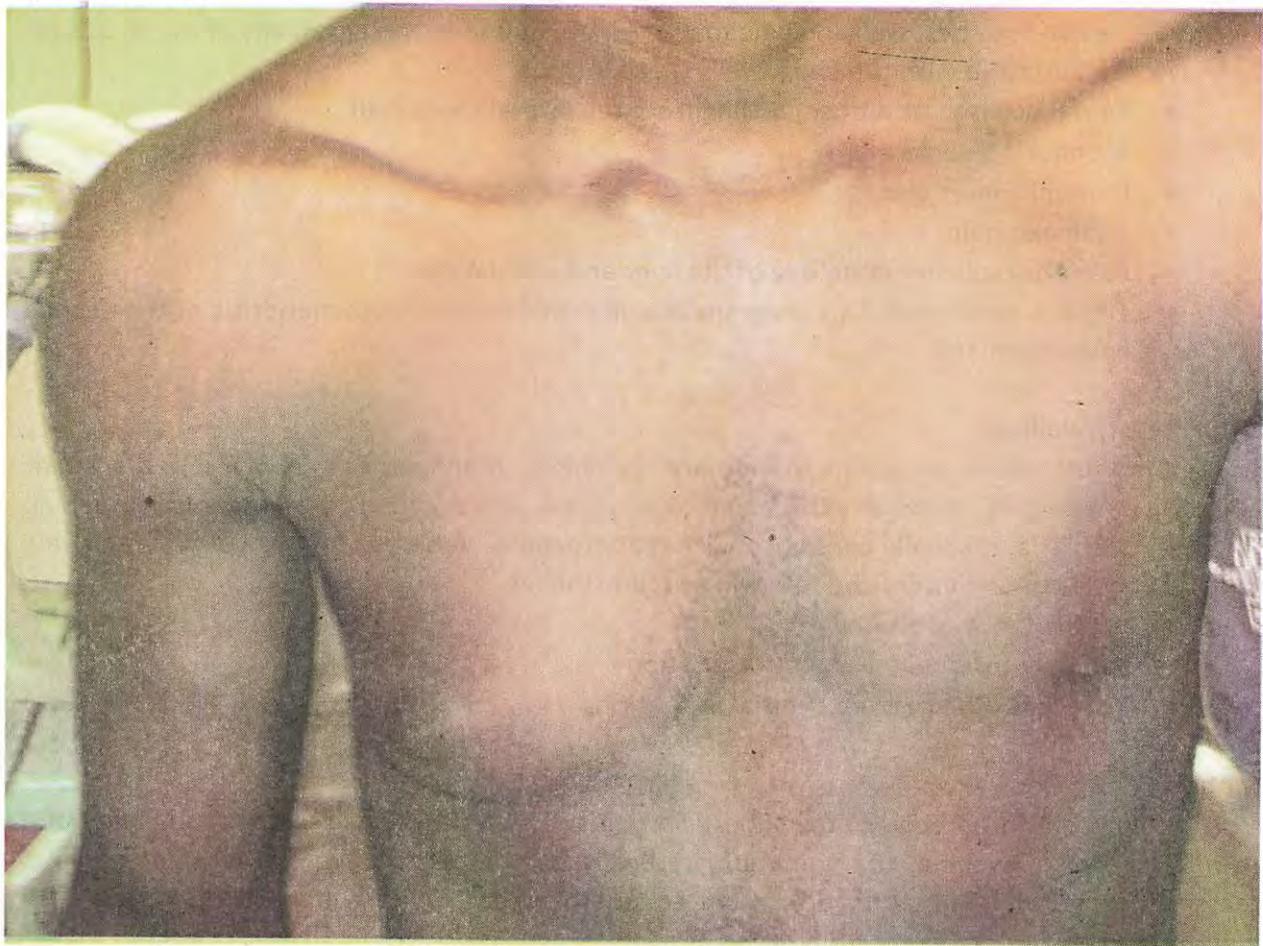
Galactocoele:

This is a clinical condition in which the breast cyst contains milk. It is associated with lactation and may be due to increased production of prolactin and/or thyrotropin. Treatment is as outlined for cystic breast swellings.

Mammary duct ectasia:

This is dilatation of the mammary duct arising from a distal obstruction. There is a subsequent inflammation of the ductal wall (periductal mastitis). A breast abscess may ensue. It may also present with nipple discharge. Treatment involves the administration of antibiotics and an excision biopsy of the duct and surrounding tissue.

Gynaecomastia:



GYNAECOMASTIA OF THE RIGHT BREAST

Gynaecomastia (also referred to as man boobs) is the female-like, non-cancerous enlargement of the male breast due to proliferation of the glandular component of the breast. It is a completely different entity from pseudogynaecomastia (lipomastia) which arises from deposition of fat in the breast. The lipomastia may be an integral part of generalised obesity. Gynaecomastia may result from either a hormonal imbalance (relative preponderance of oestrogen) or as a side effect of some drugs. Causes include testicular lesions/atrophy (leprosy, mumps and Klinefelter's syndrome) and liver diseases which result in a derangement of oestrogen metabolism. Systemic conditions that have been associated with gynaecomastia are chronic renal failure, bronchial carcinoma, adrenocortical carcinoma and thyrotoxicosis. Drugs implicated in the pathogenesis of gynaecomastia include oestrogen preparations (as was originally employed in the management of prostatic carcinoma), antipsychotic drugs, marijuana, and cimetidine. Idiopathic gynaecomastia refers to one with no obvious aetiological factor. Senescent gynaecomastia occurs in the elderly age group and may be due to a relative preponderance of oestrogen resulting from reduced production of testosterone. The major differential diagnosis, particularly in the older patient, is breast cancer. It is important to note that both lesions may coexist. Diagnosis is made from the history, and confirmed histologically by either a core or excisional biopsy.

Treatment involves addressing the underlying cause. Surgical excision (submammary mastectomy with preservation of the nipple and areolar) may be indicated if there is no response to conservative management. Indications for surgery are cosmesis and when cancer is suspected. Either a submammary or a periareolar incision will give the desired cosmetic effect.

Nipple discharge: Causes include

- Duct ectasia
- Galactocoele
- Intraduct papilloma
- Ductal carcinoma in situ
- In association with a cyst

Management entails making a definite diagnosis. The breast is examined to rule out associated lump or cyst. A bloody discharge may be worrisome but it is important to note that 90 – 95% of bloody nipple discharge is due to intraduct papilloma which is a benign condition. It is equally important to know if the discharge is coming from multiple ducts or from a single duct. If in doubt, the discharge should be tested for the presence of blood. Where neither a lump nor cyst is palpable, triple assessment (clinical, radiological and FNAC) is advised. While duct ectasia will demonstrate the presence of macrophages and chronic inflammatory cells, full blown malignant cancer will show the presence of malignant cells. Nipple discharge involving a single duct requires investigation by way of ductography. Excision of the affected duct may be carried out by way of microdochectomy.

POSTMASTECTOMY RECONSTRUCTION OF THE BREAST

The loss of a breast can be psychologically devastating to a woman. While some are able to accept the situation as the price to pay for remaining alive, others, though appreciative of the restoration of health, will wish to address the resultant cosmetic deformity. The simplest form of restoration is the insertion of a moulded plastic breast or foam under the brassier of the ipsilateral chest. A permanent solution to this problem lies in surgical breast reconstruction. The latter neither enhances nor masks recurrence and could be immediate (alongside the mastectomy) or delayed (later after mastectomy). Immediate breast reconstruction is indicated in patients undergoing mastectomy either for in-situ carcinoma or prophylaxis as in hereditary cancer. If given the opportunity, most patients will prefer the combined procedure. The following are the optional methods for postmastectomy breast reconstruction:

- 1 Insertion of silicone gel prosthesis: After tissue expansion, the silicone gel prosthesis is inserted under the pectoralis major muscle. This is more suitable for patients with small breasts
- 2 Transverse rectus abdominus myocutaneous flap (TRAM flap): The rectus abdominis flap is based on the superior epigastric artery. TRAM flaps are bulky and do not require the insertion of an implant
- 3 Latissimus dorsi myocutaneous flap (LD flap): This involves the use of the lateral half of the latissimus muscle and the skin over it. The flap is based on the thoracodorsal neurovascular bundle. The flap is not as bulky as the TRAM flap and so may require the insertion of prosthesis in order to augment the size of the 'breast'.

On restoration of the shape of the breast, a nipple is fashioned to sit on top of the mound.

Complications of breast reconstruction

- **Infection:** About 3% of prosthesis is removed because of infection.
- **Formation of a fibrous, hard capsule around the prosthesis**
- **Flap necrosis:** More common with TRAM flaps
- **Implant rupture:** About 1% of all implants will rupture. This may be due to either trauma or silicon fatigue. Most inserted silicon prosthesis are complicated by small but insignificant leakage.

CHAPTER TWENTY-FIVE

DISEASES OF THE OESOPHAGUS

The oesophagus is a tubular structure that forms the uppermost part of the gastrointestinal system. Anatomically, it is divided into three parts: upper, middle and lower thirds. Two sphincters, upper and lower oesophageal sphincters, at the two opposite ends help to regulate its function. Whereas the upper sphincter aids in swallowing, the main function of the lower oesophageal sphincter is the prevention of gastro-oesophageal reflux. The oesophagus is approximately 25cm long in the adult and 40 cm from the teeth to the lower oesophageal sphincter.

DYSPHAGIA

This refers to difficulty in swallowing. Odynophagia, on the other hand, connotes pain on swallowing. Presbyphagia is a form of functional dysphagia that occurs in old age. Dysphagia may result from either local or general conditions.

Local causes of dysphagia: Obstruction to any luminal structure may be categorised as intraluminal (within the lumen), intramural (within the wall) and extramural (outside the wall)

- Intraluminal: Foreign body
- Intramural
 - Congenital atresia
 - Pharyngeal pouch
 - Corrosive stricture
 - Motility disorders: Achalasia, diffuse oesophageal spasm
 - Tumours of the oesophagus and gastric cardia
 - Reflux oesophagitis
 - Plummer-Vinson syndrome
- Extramural
 - Retrosternal goitre
 - Thoracic aortic aneurysm
 - Bronchial carcinoma
 - Pressure of enlarged lymph nodes, lymphoma
 - Mediastinal tumours

General causes

- Stroke
- Myasthenia gravis
- Presbyphagia
- Parkinson's disease
- Cerebral palsy
- Bulbar poliomyelitis

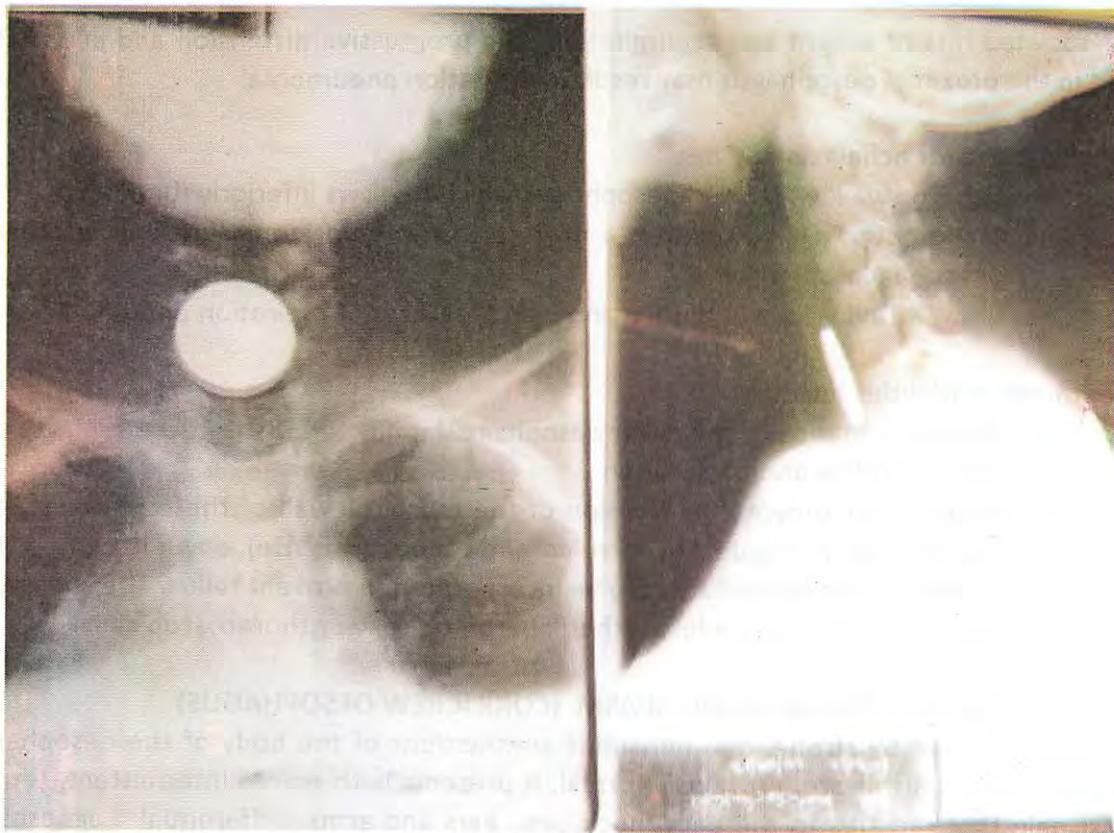
- Multiple sclerosis
- Hysteriae

History from a patient with dysphagia aims at identifying the causative factor and will includes

- Duration of symptoms: A short history particularly with associated weight loss is suggestive of a neoplastic lesion
- Associated weight loss
- Level at which patient thinks that the food gets stuck on swallowing (does not quite correlate with actual level of obstruction)
- To solids and/or liquids and history of progression
- Any history of deliberate or accidental ingestion of corrosives
- Heartburn or regurgitation soon after eating, on bending forward or in the recumbent position: suggestive of reflux oesophagitis
- Presence of cough: May indicate aspiration
- Jaundice: May indicate spread of an oesophageal neoplasm to the liver

Investigation of oesophageal diseases

- Endoscopy: Both diagnostic (visualisation/biopsy) and therapeutic (removal of foreign body/insertion of stents)
- Barium swallow: Single contrast studies are useful in the diagnosis of strictures, achalasia, diverticular and hiatus hernia. Double contrast studies are useful in the detection of mucosal lesions such as nodules, neoplasm and ulcers
- Chest X-ray and CT of the chest and abdomen: For staging of malignant lesions
- Endoscopic ultrasound: Assesses the depth of penetration of tumours and any concomitant lymphadenopathy
- Laparoscopy: For the detection and biopsy of peritoneal and hepatic secondaries
- Oesophageal manometry: Assesses the function of both the body of the oesophagus as well as that of the lower oesophageal sphincter.



FOREIGN BODY IN THE OESOPHAGUS

- Oesophageal pH monitoring (24 hour): it is employed in the investigation of gastro-oesophageal reflux. A pH of less than 4 for greater than 4% of the time is accepted as normal
- Full blood count: Iron deficiency anaemia may predispose to Plummer-Vinson's syndrome
- Others: Serum protein and transferrin aid in the assessment of the nutritional state of the patient

ACHALASIA OF THE CARDIA

This is a clinical condition that results from failure of the lower oesophageal sphincter to relax during swallowing. There is an associated loss of peristalsis, progressive dilatation and hypertrophy of the proximal oesophagus due to accumulation of ingested food material.

Pathology: Achalasia is thought to be of neurologic origin due to ganglionic degeneration of the Auerbach's plexus and/or the vagus nerve. This may be of infective origin as seen in South America where it is associated with Chaga's disease. The stasis of food may predispose to Barrett's oesophagus which is an established forerunner of oesophageal adenocarcinoma.

Clinical: Achalasia is most common in the 30 to 40 age group and commoner in females (female to male ratio is 3:2). The cardinal symptom is dysphagia to both liquids and solids. Dysphagia to liquids is, however, worse. Some patients find it easier to swallow in the standing position. There

is associated loss of weight and regurgitation. The progressive distension and accumulation of food in the proximal oesophagus may result in aspiration pneumonia.

Investigations for achalasia

- Barium swallow: Dilated oesophagus which narrows inferiorly (bird peak)
- Manometry: Demonstrates a high pressure and non-relaxation of the gastro-oesophageal sphincter
- Chest X-ray: Widened mediastinum and features of aspiration pneumonia

Treatment modalities include

- Balloon dilatation of the lower oesophageal sphincter: Disrupts the sphincter but may result in reflux and perforation
- Heller's cardiomyotomy: Division of the muscular wall of the cardia and the lower oesophagus (longitudinal, circular and smooth muscle) down to the mucosa. A fundoplication procedure may be incorporated to prevent reflux. The current trend is to carry out this procedure either laparoscopically or thoracoscopically.

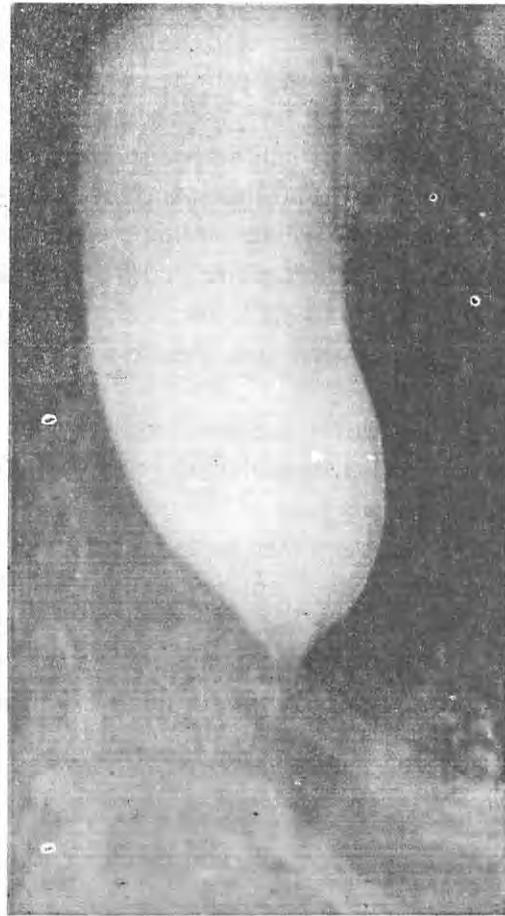
DIFFUSE OESOPHAGEAL SPASM (CORKSCREW OESOPHAGUS)

It is characterised by strong, non-peristaltic contractions of the body of the oesophagus. The sphincter mechanisms are, however, normal. It presents with severe intermittent, retrosternal chest pain that radiates to the back, neck, jaw, ears and arms. Differential diagnoses include angina pectoris, and nutcracker oesophagus.

Diagnosis: Requires a high index of suspicion. Investigations include

- Barium swallow: 'Corkscrew' oesophagus characterised by distorted peristaltic waves
- Oesophageal manometry: Repetitive high amplitude response and a raised pressure in the lower oesophagus
- Endoscopy: Findings as in barium swallow

Treatment: Initially medical. Surgery is, however, indicated on failure of medical treatment



BARIUM SWALLOW FILM OF ACHALASIA SHOWING THE 'BIRD PEAK' NARROWING AT THE DISTAL OESOPHAGUS

- Medical treatment: Calcium channel blockers, proton pump inhibitors, H₂ receptor antagonists and tricyclic antidepressants
- Surgical treatment: Long oesophagomyotomy

Nutcracker oesophagus is an uncommon condition that shares the same pathology as well as clinical features with diffuse oesophageal spasm. The management is essentially the same.

GASTRO-OESOPHAGEAL REFLUX DISEASE (GERD)

This condition is characterised by the reflux of acid gastric contents into the lower oesophagus. It is due to a functional abnormality of the lower gastro-oesophageal sphincter.

Pathophysiology: Under normal circumstances, gastroesophageal reflux and its resultant effect on the lower oesophagus are prevented by the following physiological mechanisms

- An intra-abdominal length of the oesophagus: Abnormal location of the oesophagus wholly in the chest will result in oesophageal reflux. This is the situation with achalasia
- The phreno-oesophageal ligament: This is an extension of the crural diaphragm and is attached to the oesophagus. It is an anatomical reflux mechanism

- The oesophageal sphincter undergoes physiological intermittent relaxations. An abnormal increase in the number and duration of relaxation may predispose to oesophageal reflux
- The lower oesophageal sphincter serves as a zone of increased pressure between the positive pressure in the stomach and the negative pressure in the chest. A hypotensive or incompetent valve will result in increased reflux
- There are inherent mechanisms in the oesophagus that enable it not only to clear acid but also to withstand its effects on the mucosa. Failure of these physiological protective mechanisms will result in the deleterious complications of gastro-oesophageal reflux

Gastroesophageal reflux exposes the lower end of the oesophagus to acid mucosal injury. This results in inflammation (reflux oesophagitis). On long term, this may result in a metaplastic change whereby the squamous cells of the oesophagus are replaced with columnar epithelium (intestinal metaplasia). This is referred to as Barrett oesophagus. The latter is associated with a high risk factor for oesophageal adenocarcinoma. Longstanding GERD may also result in oesophageal stricture and also increases the risk for pharyngeal reflux and silent aspiration. The latter is responsible for the extraoesophageal complications of GERD. These are laryngitis, reactive airway disease and recurrent pneumonia. Persistent inflammation of the lung tissue may result in pulmonary fibrosis.

The following are the risk factors for GERD

- Decreased lower oesophageal sphincteric tone
- Decreased oesophageal motility
- Hiatal hernia: May not be the sole cause
- Gastric outlet obstruction: Diminished gastric emptying
- Excess gastric acidity
- Nasogastric intubation
- Scleroderma
- Exacerbating lifestyle factors: Obesity, alcoholism, smoking, caffeine

Complications of GERD are

- Oesophageal stricture,
- Barrett's oesophagus and carcinoma.
- Aspiration pneumonia: Known complication of reflux oesophagitis

Clinical presentation: Patients present with heartburn, retrosternal pain, and regurgitation. Symptoms are worsened by meals and assumption of the supine position. Aspiration may result in clinical features of pneumonia such as chronic cough and difficulty with respiration. Longstanding cases may present with features of oesophageal stricture and oesophageal carcinoma (complication of Barrett's oesophagus).

Investigations: Not all patients with clinical features of GERD will require diagnostic workup. Patients with relatively mild condition may be managed medically. Indications for a detailed diagnostic workup include

- Failure of medical therapy to relieve symptoms (patients on maximal dose of PPI)
- Recurrence after initial response to medical treatment
- Longstanding atypical symptoms such as chronic cough, hoarseness and wheezing

The diagnostic workup for GERD involves

- Endoscopy: Will demonstrate features of oesophagitis as well as those of its complications such as stricture, Barrett's oesophagus and oesophageal carcinoma. Relevant biopsies are taken. From the therapeutic angle, strictures may be managed by dilatation and stenting.
- Oesophageal manometry and pH monitoring: This is a combined procedure. In preparation for this test, the patient should discontinue all medication. The patient is made to swallow a small electronic pressure transducer. This is positioned in the vicinity of the lower oesophageal sphincter. With the use of a 24 hour pH monitoring device, the pH at the level of 5 cm above the lower oesophageal sphincter is measured. This test will reveal both the presence and timing of acid reflux in the lower oesophagus. Diagnosis of silent aspiration may also be made by monitoring the pharyngeal pH. The latter is a reflection of the pH at the proximal oesophagus. Oesophageal manometry evaluates the competence of the lower oesophageal sphincter and can diagnose motility disorders such as achalasia or diffuse oesophageal spasm. Oesophageal manometry quantifies the degree of the GERD and aids in decision-making as regards surgical intervention
- * Barium swallow/meal: may reveal the following:
 - The location of the gastrooesophageal junction in relation to the diaphragm
 - Associated hiatal hernia and stricture.
 - Associated gastric outlet obstruction
 - Trendelenburg position or a Valsalva manoeuvre may demonstrate reflux at the lower oesophageal sphincter

Treatment: Conservative treatment consists of changes in lifestyle and use of drugs.

Lifestyle changes

- Elevation of the head of the bed and/or sleeping propped up with multiple pillows
- Feeding: Avoid spicy foods and eating large, heavy meals at night. Early dinner is advised (at least 2 to 3 hours before bedtime) as well as small, multiple meals.
- Reduction of weight in the obese: May require bariatric surgery
- Reduction in the rate of smoking, alcohol and caffeine intake

Drug treatment includes

- Acid-reduction prescriptions: antacids, H₂-receptor blockers, and proton pump inhibitors (PPI). The latter are the most effective in the relief of GERD symptoms and constitute the sheet-anchor of acid-suppression therapy. Very high doses of PPI may be necessary for relief of GERD symptoms.
- Prokinetic drugs that enhance gastric emptying such as metoclopramide are also helpful

Surgical treatment: Indications for surgery are

- Failure of conservative treatment: Persistence of symptoms inspite of treatment with maximal doses of PPI
- Barrett's oesophagus: Replacement of the squamous epithelium of the lower oesophagus with columnar epithelium secondary to gastro-oesophageal reflux. It is a documented premalignant lesion
- Patients who for personal reasons voluntarily opt for surgery rather than the long period associated with medical treatment

Laparoscopic fundoplication is now the procedure of choice. The classical Nissen fundoplication is a 360 degrees wrap of the fundus of the stomach around the lower oesophagus. The Belsey Mark IV is a 270 degrees wrap.

Complications of surgery include

*Gastric perforation and bleeding.

Dysphagia and abdominal gas bloat: May result from a tight repair These may require revision surgery.

- Recurrence is not uncommon and will require fresh reassessment and treatment (medical and surgical).

Two endoscopic endoluminal techniques have been introduced in the surgical management of GERD. They are

- Application of radiofrequency energy to the lower gastro-oesophageal junction: This aims at increasing the pressure in a bid to prevent reflux
- Endoluminal suturing of the gastro-oesophageal junction.

These have not been put to routine use

CORROSIVE OESOPHAGEAL STRICTURES

Ingestion of corrosive may be accidental or suicidal. The ingested material may be acidic or alkaline. Alkaline agents predominantly affect the oesophagus while acidic agents paradoxically spare the oesophagus and affect the distal part of the stomach resulting in gastric outlet obstruction. That acid solution tends to spare the oesophagus and causes damage more distally is related to the pathophysiology of corrosive ingestion. Whereas the ingestion of alkali results in liquifactive necrosis with saponification of all the layers of the wall of the oesophagus, acid burn results in coagulative necrosis and eschar formation. This tends to limit the depth of penetration and thus allows the acid to pass down quickly into the stomach, coursing along the lesser curvature on to the pylorus where it pools and causes pylorospasm. This situation may give rise to ulceration, fibrosis and perforation. Similar complications may also occur in the oesophagus particularly when the quantity of acid ingested is much. This is the situation in attempted suicide as opposed to accidental ingestion when small quantities of acid are involved.

Emergency management of corrosive stricture involves

- Drinking a lot of water as part of first aid measure immediately after ingestion
- Strictly nil orally after the early stage with avoidance of vomiting
- Administration of intravenous infusion
- Administration of parenteral analgesic, antibiotics and hydrocortisone (anti-inflammatory and so reduces the rate of fibrotic reaction)
- Gentle and cautious oesophago-gastro-duodenoscopy within 24 hours of injury to assess severity and extent of damage. Scoping should terminate just above the level of injury

Late management includes

- Nutrition: Gastrostomy, feeding jejunostomy, or total parenteral nutrition

Definitive treatment depends on the extent of the injury and includes

- Bouginage
- Balloon dilatation,
- Oesophageal resection (oesophagectomy) with colon interposition or gastric pull up.
- Gastric outlet obstruction may require a bypass procedure (gastrojejunostomy)

Patient follow up is important as they run the risk of oesophageal carcinoma

HIATUS HERNIA

Of two main varieties: Sliding and paraoesophageal

- Sliding: Superior pulling up of the cardiac part of the oesophagus through the diaphragmatic oesophageal hiatus. Referred to as Type 1 variety
- Paraoesophageal: Fundus migrates beside the apparently intact gastro-oesophageal junction into the mediastinum. Referred to as Type 11 variety
- Combination of the above two: Referred to as Type 111 variety
- Rare type containing other abdominal viscera: Referred to as Type IV variety

Clinical: Sliding oesophageal hernia presents with features of reflux oesophagitis. Pressure effect following a large paraoesophageal hernia may result in hiccapping, vomiting, dyspnoea, palpitations and arrhythmias

Investigations

- Chest X-ray: Wide mediastinum and an air-fluid level
- Barium swallow: Stomach in the chest; reflux on tipping the patient
- Endoscopy: Confirms barium enema findings
- pH monitoring : Findings in keeping with reflux

Treatment is essentially same as for reflux oesophagitis. Surgical treatment will include reconstruction of the diaphragmatic defect. The latter may require the use of a mesh

CARCINOMA OF THE OESOPHAGUS

The incidence of oesophageal cancer is rising rapidly and has been found to be the sixth most common malignancy in the United States of America.

Most common in the sixth decade of life and has a male preponderance. The commonest site is the mid-oesophagus.

Aetiology: May result from lifestyle or secondary to established premalignant conditions

- Lifestyle: Tobacco and alcohol. Consumption of nitrite- and nitrate-containing food also constitutes a potential risk factor due to the production of carcinogens (Nitrosamine)
- Gastro-oesophageal reflux disease: May predispose to Barrett's oesophagus which is an established fore-runner of gastroesophageal junction adenocarcinoma
- Premalignant conditions: These include caustic stricture, Plummer-Vinson's syndrome (resulting from iron deficiency anaemia) and achalasia

Pathology: Tumours may be ulcerative, papilliferous (exophytic), or annular. Most are squamous cell carcinoma. Whereas squamous cell carcinoma is the most common histological type encountered in developing countries, adenocarcinoma is the most common in developed countries. In fact, the incidence of squamous cell carcinoma is decreasing in the western world. Adenocarcinoma usually occurs at the lower oesophagus either as a complication of Barrett's oesophagus or resulting from local spread of a gastric cardiac neoplasm.

Spread: Like most neoplasms, spread is by local, lymphatic and blood stream

- Local: Involves the mediastinal structures (trachea, pericardium, major vessels)
- Lymphatic: Supraclavicular and cervical lymph nodes for upper third tumours, Tracheobronchial and paraoesophageal nodes for middle third tumours and subdiaphragmatic and coeliac lymph nodes for lower third tumours
- Blood: Lungs and liver

Staging of oesophageal cancer

Stage I Tumour invades lamina propria or submucosa; negative lymph nodes

Stage IIa Tumour invades muscularis propria or adventitia; negative nodes

Stage IIb Tumour: Any tumour that invades up to the muscular propria or adventitia

Node: Positive regional lymph nodes

Stage III Tumour invades adventitia

Nodes: Positive regional nodes OR

Tumour: Invades adjacent structures

Nodes: Positive or negative

Stage IV: Distant metastasis

Clinical evaluation: This entails obtaining a comprehensive history and proper clinical examination. The basic symptom and whose analysis is very important is dysphagia.

- A history of previous corrosive ingestion is suggestive of caustic stricture

- A history of heartburn and long-term medication with proton pump inhibitors suggests gastro-oesophageal reflux disease
- Progressive dysphagia of rapid onset with associated loss of weight is highly suggestive of neoplastic disease.
- Dysphagia of a fairly long duration (months to years) may be suggestive of benign strictures and achalasia.
- It is worthy of note that although they have different approaches to treatment, carcinoma of the distal oesophagus may present with same clinical features as that of the proximal stomach.

The clinical features of oesophageal cancer, like all other cancers, can be categorised into those of local disease, those related to neoplastic spread, and general features of cancer.

- Local: Progressive dysphagia initially to solid dry food but eventually even to liquids. There may be associated features of aspiration pneumonia (cough, dyspnoea, crepitations). In Plummer-Vinson's syndrome, there is associated pallor and koilonychias
- Spread: Retrosternal pain following mediastinal spread, cervical and supraclavicular lymphadenopathy, jaundice and hepatomegaly
- Weight loss, anorexia and features of anaemia

Investigations:

The first and foremost priority is to determine the location and the nature of the oesophageal obstruction. This is followed by evaluating the extent of the lesion in terms of spread. The relevant investigations are

- Oesophagogastroduodenoscopy and tissue biopsy: This is the gold standard
- Barium swallow and follow-through: This is still relevant particularly where there are no facilities for endoscopy. It will demonstrate the strictured malignant segment with 'shouldering'
- Endoluminal ultrasound scan (EUS): Determines the depth of the lesion. May also detect involvement of the lymph nodes. It is therefore important in the determination of the TNM staging of the lesion (TN). The lymph node staging may be confirmed by carrying out an EUA-directed fine needle aspiration of the identified lymph node. Like most advanced methods of investigation, EUA results are operator-dependent
- Chest X-ray and bronchoscopy: To rule out a primary lesion of the lung and to detect mediastinal involvement. May also demonstrate pulmonary complications of silent aspiration
- CT scan of chest, liver and upper abdomen: Detection of metastases
- Transabdominal USS: For the detection of metastatic spread to the liver
- Positron emission tomography (PET) CT scan: This is the imaging modality of choice and is replacing standard CT scan. The main drawback, however, is that it is neither readily available nor affordable
- Full blood count: Patient may be anaemic and will require preoperative blood transfusion

- Electrolytes/urea /creatinine
- Liver function test: May show evidence of hepatic involvement. It is also important in the evaluation of the patient's nutritional state by way of the serum albumin level.
- Fasting blood sugar
- Electrocardiography: Patient is likely to be above the age of 40 and may require surgical treatment.

Treatment of oesophageal carcinoma

If diagnosed at a potentially curable stage, oesophagectomy (resection-restorative) surgery is the primary treatment. Unfortunately, however, many patients present with advanced growth as less than 50% of patients are found eligible for curative surgery. Multimodality treatment has been found to improve the prognosis.

- Preoperative preparation of the patient involves
- Intravenous infusion: Patient may be dehydrated due to inability to drink enough water as a result of his clinical condition. It is equally important to correct any detected electrolyte anomaly



BARIUM MEAL OF OESOPHAGEAL CARCINOMA

NOTE THE 'SHOULDERING'

- Optimisation of the nutritional state: If the patient is tolerating liquids, nutritional supplementation should be carried out with oral administration of high calorie, high protein food supplements. These will not only replenish the losses but will equally

supply his ongoing nutritional requirements. For patients who are unable to tolerate orally, other means of enteral feeding distal to the point of obstruction should be considered. Total parenteral nutrition may not be necessary since there is no contraindication to enteral nutrition

- Blood transfusion may be necessary if patient is anaemic as evidenced by a low PCV and haemoglobin
- Counselling of patient as regards the nature of the illness. This is followed by obtaining an informed consent

Radiotherapy is the treatment of choice for tumours involving the upper third of the oesophagus. Based on the location of the neoplasm, various types of surgery have been described for oesophageal carcinoma.

- **Transthoracic oesophagectomy:** This involves a thoracoabdominal approach and involves a laparotomy incision (or laparoscopy) and a right thoracic incision. Oesophagogastric anastomosis is carried out in the chest after dissection and resection of the oesophagus. The main disadvantage is the high rate of pulmonary complications. This is due to severe pain arising from incisions in both the chest and upper abdomen
- **Transhiatal oesophagectomy:** Approach is by laparotomy via an abdominal incision (or by laparoscopy) coupled with a cervical incision. Through the abdomen, the stomach is mobilised and the distal oesophagus is equally mobilised after enlarging the hiatal opening. Mobilisation of the cervical oesophagus and dissection of the proximal thoracic oesophagus is carried out through the cervical incision. This is followed by resection of the entire thoracic oesophagus and the proximal stomach. The remaining stomach is brought up to the neck through the posterior mediastinum. The ensuing oesophago-gastric anastomosis is carried out in the neck. The main advantage of this approach is the reduction in morbidity and mortality associated with anastomotic leakage when compared to transthoracic oesophagectomy. In addition, there is a comparative reduction in pulmonary complications when compared with transthoracic oesophagectomy.
- **McEwan three-stage thoracotomy:** Utilises three incisions – left neck, thoracotomy and laparotomy. Anastomosis is carried out in the neck.
- **Total gastrectomy and Roux-en-Y oesophagojejun al reconstruction:** Carried out by a thoracoabdominal approach. Suitable for cardiac tumours

Stewart classification of gastro-oesophageal junction adenocarcinoma: This aims at relating the treatment of these lesions to their location

- **Type 1 tumours:** Located more than 1 cm proximal to the junction. Surgery involves oesophagectomy
- **Type 11 tumours:** Located within 1cm proximal and 2 cm distal to the junction. In addition to oesophagectomy, surgery involves partial resection of the proximal stomach

- Type 111 tumours: Located more than 2 cm distal to the junction. Surgical treatment consists of total gastrectomy

Complications of surgery include

- Anastomotic leakage (5 to 10%)
- Chyle leakage due to thoracic duct injury
- Others are atelectasis and cardiac arrhythmias.

Contraindications to surgery include

- Cervical or celiac node involvement
- Evidence of haematogenous metastases
- Broncho-oesophageal fistula
- Evidence of vocal cord paralysis
- Invasion of pericardium
- Invasion of the major vessels in the mediastinum: Superior venacava and the aorta

Other treatment modalities

- Chemotherapy: Combined therapy with epirubicin, cisplatin and 5-fluorouracil (ECF) has been found to be beneficial. This may be administered either as adjuvant or neoadjuvant regimen. There have been controversial reports of an improved curative as well as survival rates when administered both pre- and post-surgery. Chemotherapy can also be applied as a palliative measure
- Radiotherapy: Oesophageal squamous carcinoma is highly sensitive to radiotherapy. This has been applied in the treatment of cancers involving the upper one-third of the oesophagus. Unfortunately, the recurrence rate after radiotherapy is high.
- Chemoradiation has been found to be effective
- Targeted molecular therapy: This applies to gastro-oesophageal junction adenocarcinoma. Those that demonstrate overexpression for human epidermal growth factor receptor 2 (HER2 neu) have been found to respond to the monoclonal antibody Trastuzumab (Herceptin). Studies have shown that outcome is better when Herceptin is combined with the standard chemotherapy regimen.

Palliative therapy

This is aimed at improving the quality of life in patients with advanced disease and is achieved by the relief of dysphagia which constitutes the most debilitating symptom. It equally tries to forestall bleeding, perforation and tracheoesophageal fistula. The following are modalities of palliative treatment

- Endoscopic stenting: Provides rapid relief of dysphagia. Recurrence may occur due to stent migration or tumour overgrowth
- Nd:YAG laser and photodynamic therapy: Quite effective with exophytic neoplasia as they achieve endoluminal destruction of obstructing lesions. The main drawback is that the facilities are neither readily available nor affordable

- Single-dose brachytherapy: This delivers intraluminal radiotherapy. Although it acts slowly particularly when compared with stenting, it tends to achieve a much longer relief of dysphagia
- Chemotherapy: Response to obstruction is variable. This raises the need for a supplementary palliative treatment

Prognosis: An overall 21% survival rate for early cases that qualify for surgical treatment. This may be further divided into 40% to 50% survival for node-negative resections and 10% to 20% for node-positive resections. Cancers arising from Barrett's oesophagus tend to have a better prognosis.

CHAPTER TWENTY – SIX

PANCREATIC DISEASES

Anatomy of the pancreas: The pancreas is a horizontal organ which is tucked into the C-loop of the duodenum. It is divided into three parts – the head which is in direct relationship to the duodenum, body and tail. The neck separates the head from the body while the tail abuts at the hilum of the spleen.

The blood supply of the pancreas is from the three branches of the coeliac trunk mainly through the pancreaticoduodenal vessels. The common blood supply with the duodenum makes it imperative to resect the duodenum along with the pancreas whenever the head of the pancreas is to be excised.

The pancreas has both endocrine as well as exocrine functions. This is carried out by three groups of cells. The islet cells of Langerhans are responsible for the exocrine functions of the gland as they produce the digestive enzymes. These include amylase, lipase, trypsinogen, chymotrypsin and elastase. On the other hand, the endocrine functions are carried out by the alpha and beta cells which play major roles in glucose metabolism. The alpha cells produce glucagon which increases the blood sugar level while the beta cells produce insulin which does the opposite (decreases the blood sugar level).

PANCREATITIS:

This connotes an inflammation of the pancreas. There are two forms – acute and chronic. The acute form as to be inferred, presents as an abdominal emergency. The pancreatic function tends to recover fully after such attacks. In the chronic form, however, the pancreatic function rarely recovers; rather there is progressive deterioration.

Classification of pancreatitis (Marseille's)

- Acute pancreatitis
- Acute relapsing pancreatitis
- Chronic pancreatitis
- Chronic relapsing pancreatitis

As stated above, acute pancreatitis is characterised by acute reversible pathological changes as opposed to the chronic variety which exhibits morphologically irreversible changes.

ACUTE PANCREATITIS:

Acute pancreatitis is defined as an acute inflammatory process of the pancreas, with variable involvement of other regional or remote organ systems. Moynihan described acute pancreatitis as "the most terrible of all calamities that occur in connection with the abdominal viscera". There are two clinical varieties of acute pancreatitis: mild and severe. Mild acute pancreatitis is associated with minimal organ dysfunction and an uneventful recovery. Severe acute pancreatitis, on the other hand, is associated with organ failure and/or local complications such as necrosis, abscess or pseudocyst.

Atlanta classification of acute pancreatitis:

- Acute oedematous pancreatitis (80%): This is the milder form and has a mortality of 1%
- Acute necrotising pancreatitis (20%): Associated with pancreatic and peripancreatic necrosis and has a mortality of 20%

Inflammation in acute pancreatitis is sparked off by the endogenous pancreatic enzymes which in an incongruous and paradoxical manner start digesting the pancreatic tissue. It is a form of autodigestion.

Aetiology of acute pancreatitis

The underlying factor is the inability of the pancreatic duct to discharge the pancreatic juice containing digestive enzymes into the duodenum. This may be either due to pancreatic duct spasm or obstruction: The aetiological factors are myriad and include

- Biliary tract disease especially gallstones: This accounts for about 50% of cases of acute pancreatitis. By obstructing the lower segment of the common bile duct, gallstones may equally hinder the free flow of pancreatic juice into the duodenum.
- Alcoholism: This accounts for 25% of cases and may be due to ductal spasm
- Trauma (surgery, sphincterotomy, biliary manometry and ERCP)
- Hypercalcaemia
- Viral infections (mumps, coxsakie, HIV).
- Drugs (steroids, INH, loop diuretics, and azathioprine),
- Parasites: Biliary ascariasis
- Autoimmune diseases
- Diabetes mellitus
- Infectious mononucleosis
- Scorpion bites.
- Idiopathic: No known cause can be attributed in 20 to 25% of cases. Application of more exhaustive investigations aimed at identifying known causative factors will reduce the incidence of the 'pack' categorised as idiopathic acute pancreatitis

Pathology: All the aetiological factors act by one or a combination of the following

- Spasm of the sphincter of Oddi resulting in reflux of bile into the pancreatic duct and into the pancreatic parenchyma
- Increased secretion of pancreatic enzymes
- Act directly as toxins to the pancreas.

The process involves the activation of trypsinogen to trypsin within the substance of the organ. This then goes on to activate other pancreatic enzymes and ultimately results in the digestion of the pancreatic tissue. This may end up as acute haemorrhagic pancreatitis. While the amylase digests the carbohydrate component, lipase and trypsin act on the fat and protein components respectively. Elastase, on the other hand, digests the elastin of the blood vessels resulting in severe haemorrhage. Lipase splits the fat into fatty acid and glycerol. The fatty acid chelates the serum calcium by the process of saponification to form 'calcium soap'. This results in hypocalcaemia. This acute inflammation is associated with exudation and loss of fluid into the

peritoneal space and constitutes a third space loss. This reduces the circulatory blood volume and results in hypovolemic shock. A systemic inflammatory response is triggered by the systemic activation of leucocytes and the profuse secretion of proinflammatory cytokines. On the other hand, there is reduction in the activity of the physiological anti-inflammatory mechanisms. This may explain the development of multiple organ dysfunction that is a known feature of acute pancreatitis.

Clinical features of acute pancreatitis

The patient presents with a sudden onset of epigastric pain that radiates to the back. There is associated nausea, retching and vomiting. This patient assumes a leaning-forward posture as this helps to ameliorate the abdominal pain by temporarily lifting the pancreas off the posterior abdominal wall. On examination, the patient is usually ill-looking and may manifest the clinical features of jaundice, dehydration and hypovolemic shock. The abdomen may be distended with generalised guarding and tenderness especially around the epigastrium. There may be clinical evidence of ascites. Abdominal examination in severe acute pancreatitis may manifest with Cullen's sign (periumbilical bruising) and Grey-Turner's sign (flank bruising). The other systems of the body such as respiratory, cardiovascular and genitourinary systems should be carefully evaluated in view of the complications that may arise.

The differential diagnoses of acute pancreatitis include

- Perforated duodenal ulcer,
- Cholecystitis,
- Intestinal obstruction, and
- Ruptured ectopic pregnancy.
- Others are mesenteric ischemia, diabetic ketoacidosis and salpingitis.

Management: Having made an impression of acute pancreatitis, the next step involves emergency resuscitation of the patient and confirmation of the diagnosis. The following should be instituted immediately

- * Set up an intravenous line and commence resuscitation with either normal saline or Ringer's lactate solution. Resuscitation with intravenous infusion and intensive monitoring constitute the most important elements in prevention of multiple organ failure
- * Commence patient on nil orally
- * In the course of setting up the intravenous line, take blood samples for the following investigations:
 - Serum amylase: Marked elevation of this enzyme is the hallmark of diagnosis
 - Full blood count: PCV, white cell count (they have prognostic value)
 - Electrolytes, urea and creatinine
 - Serum calcium
 - Grouping and cross-matching of blood
 - Liver function tests
 - Random blood sugar
 - PO₂ and PCO₂

- Coagulation profile
- * Pass a urethral catheter to monitor the urinary output. This acts as a guide in the fluid resuscitation of the patient
- * Administration of adequate analgesia: Avoid the use of opiates which may cause further spasm of the pancreatic duct
- * Administer oxygen if necessary, particularly if the PO₂ is low
- * The role of antibiotics is controversial. Whereas some authorities will not administer antibiotics since they believe that acute pancreatitis is not basically an infective process, most others will rather give it in order to forestall any infective complication. More recent trials have, however, shown improved therapeutic outcome following antibiotic prophylaxis in severe acute pancreatitis
- * Intensive care management may be required.
- * Send for radiological investigations after stabilising the patient:
 - Plain abdominal X-rays: Will demonstrate sentinel loop and colon cut-off sign
 - Transabdominal ultrasound scan: Gallstones and presence of pseudocyst
 - Abdominal CT scan: Useful when the diagnosis is doubtful as well as diagnosis of pseudocyst and pancreatic necrosis
 - Contrast-enhanced CT scan: Quite relevant when the diagnosis is in doubt
 - ERCP may be useful in patients with confirmed pancreatitis due to gallstones
 - Other radiological investigations that may be helpful include MRI and magnetic resonance cholangio pancreatography (MRCP).

As mentioned above, a marked elevation of the serum amylase level is consistent with the diagnosis of acute pancreatitis. There are, however, two caveats to this. The first is that the serum amylase level may be elevated in other acute abdominal conditions. The rise in such cases, however, is only marginal. It is only in acute pancreatitis that one could have a very high elevation (more than three times the upper limit of normal) in the level of serum amylase. The second is that serum amylase has a high clearance rate via the kidneys. There is a marked drop in the level after 24 to 48 hours. This means that to get an accurate result, the blood sample must be taken soon after the onset of the pathological process. The serum lipase level may be useful after 48 hours as it has a lower clearance rate and therefore stays longer in the blood. Recent studies have shown the reliability of serum lipase estimation in the diagnosis of acute pancreatitis. It is believed to be as good as, or even more reliable, than serum amylase in the diagnosis of acute pancreatitis. Trypsinogen activation peptide (TAP) and serum carboxypeptidase activation peptide B (CAPAP-B) are newer markers that are both diagnostic as well as prognostic. It is important to note that the serum levels of amylase and lipase do not correlate with the severity of acute pancreatitis. Very high levels of serum amylase on admission are suggestive of a gallstone aetiology.

The subsequent management of acute pancreatitis will depend on the investigation results

- Intravenous administration of calcium gluconate if there is hypocalcaemia
- Insulin therapy is instituted if there is hyperglycaemia
- Administration of parenteral nutrition or post-pyloric tube feeding in view of the severe metabolic upset. An attempt is made at oral feeding in mild cases.

- Administration of intravenous H₂-receptor blockers and omeprazole to reduce gastric acid secretion and hence pancreatic stimulation. It equally acts as a prophylaxis against upper gastrointestinal haemorrhage in severe acute pancreatitis.
- Correct any detected electrolyte anomaly
- Commence thromboprophylaxis with either unfractionated or low-molecular weight heparin
- Octreotide, a somatostatin analogue may help in reducing pancreatic secretion
- Prophylactic antibiotics: They are quite useful in the management of necrotising pancreatitis and have been shown to significantly reduce the incidence of peripancreatic infections. On the flip side, antibiotics have been known to increase the risk of infections by resistant bacteria and fungal species.

Indications for surgical treatment in acute pancreatitis: Acute pancreatitis usually presents as an acute abdomen. Diagnosis is difficult and requires a high index of suspicion. Any confirmed case should be managed conservatively as surgical intervention may worsen the prognosis. All said and done, surgical intervention may be indicated under the following circumstances

- When it is difficult to arrive at a definite clinical diagnosis. In this case, it is better to carry out an exploratory laparotomy with a view to achieving a correct working diagnosis.. When the diagnosis is finally made intraoperatively, it is not advisable to carry out an extensive procedure. Rather, one should do the basic minimum: suck out haemorrhagic fluid, replace fluid and blood loss, insert abdominal drains, close up the patient and continue with conservative management.
- When clinical response proves inadequate despite the administration of appropriate treatment in a clinically diagnosed case of acute pancreatitis
- In gallstone acute pancreatitis: Diagnosis is usually made by abdominal ultrasound and CT scan. Conservative management should be instituted and an interval cholecystectomy carried out soon after recovery. This may be carried out before discharge from the index admission if patient is considered fit.
- When there are complications such as pancreatic pseudocyst, pancreatic necrosis, pancreatic abscess and haemorrhage (see below).

Surgical procedures at laparotomy: May include excision of necrotic tissue, drainage of abscess or pseudocyst, and peritoneal lavage. There is a place for early ERCP/endoscopic sphincterotomy in patients with severe gallstone-induced acute pancreatitis. Early cholecystectomy, preferably during the index admission, is advocated for gallstone-induced pancreatitis. Newer concepts in the treatment of acute pancreatitis include

- Early enteral nutrition by jejunostomy: This has proved useful in the management of acute pancreatitis. It is thought to improve gut barrier function, reduce bacterial translocation and also reduce the overall inflammatory response.
- Anti-cytokine therapy: There is growing evidence that increased production of pro-inflammatory cytokines plays a significant role in the pathogenesis of acute pancreatitis. Anti-cytokine therapy is therefore aimed at reducing the development of systemic inflammatory response and preventing organ dysfunction. The platelet

activating factor antagonist, lexipafant, has demonstrated a beneficial effect. In addition, lexipafant has been found to act as a down-regulator of the proinflammatory cytokine response. Its use is, however, still under investigation by multicentre trials.

Complications of acute pancreatitis

General complications include

- Acute renal failure from hypovolemic shock,
- Respiratory complications: These include adult respiratory distress syndrome, respiratory complications of splinting of the diaphragm resulting from raised intra-abdominal pressure and pleural effusion
- Multiple organ failure
- Encephalopathy
- Coagulopathy and disseminated intravascular coagulopathy
- Severe hypocalcaemia
- Diabetes mellitus
- Recurrent attacks of acute pancreatitis

The local complications that may require surgical management include

A Pancreatic pseudocyst: This is a collection of fluid and necrotic tissue enclosed in an inflammatory capsule composed of fibrous or granulation tissue. It is referred to as a pseudocyst because unlike true cysts, the capsule is not lined by epithelium. It presents as an epigastric swelling soon after an attack of acute pancreatitis. Diagnosis is usually confirmed by abdominal ultrasound and CT scans. About 50% will resolve spontaneously.

Complications of pseudocyst include rupture, infection and bleeding. The others are duodenal obstruction, obstructive jaundice (obstruction of the bile ducts) and segmental portal hypertension (thrombotic).

Indications for surgical intervention are

- Failure of spontaneous resolution
- Cysts larger than 5 cm in diameter. Such cysts may not resolve spontaneously
- Suspicion of bleeding into the cyst
- Calcified cyst wall
- Thickened cyst wall

A waiting period of at least six weeks is advised before attempting drainage of pancreatic pseudocyst. This is to give room for maturation of the cyst wall prior to any form of intervention. Drainage can be achieved either by an ultrasound-guided percutaneous method or by open drainage. The route of drainage is dictated by the structure that is most adjacent to the cyst. The options include cystogastrostomy, cystoduodenostomy, cystojejunostomy, and Roux-en-Y drainage. A biopsy of the wall of the pseudocyst should be taken in order to rule out the presence of cystadenocarcinoma.

B Pancreatic necrosis: This consists of non-viable pancreatic tissue following severe acute pancreatitis. There is associated peripancreatic fat necrosis. Diagnosis is made by contrast-enhanced abdominal CT scan. The non-viable pancreatic tissue will not take up contrast.

Treatment is conservative initially with intravenous infusion and antibiotics. Surgical treatment is indicated if there is no clinical response to medical treatment and consists essentially of debridement and drainage. Patients with pancreatic necrosis are usually ill and need to be supported with enteral nutrition and glycaemic control

C Pancreatic abscess: This connotes the collection of pus in the peripancreatic region. Diagnosis is made by the presence of fever and an epigastric mass. Confirmation is by CT scan. Treatment consists of antibiotics and drainage. The latter may be by way of either percutaneous or open drainage complemented by the insertion of drains.

D Splenic/mesenteric/portal vessel rupture: This is due to pseudoaneurysm arising from the action of elastase on these blood vessels. The end result is aneurysmal dilatation, rupture and severe haemorrhage. Treatment consists of blood transfusion and laparotomy to control bleeding and achieve haemostasis.

E Pancreatic fistula: This is due to enzymatic disruption of the ductal wall. The fistula may be internal (involving adjacent abdominal viscera such as the rectum or bladder) or external. Investigations for the confirmation of diagnosis include fistulography, CT scan and ERCP. Like all fistulae, initial management is conservative. Surgery is indicated when there is failure of conservative management and may involve pancreatic resection and pancreaticojejunostomy.

Prognosis in acute pancreatitis

Acute pancreatitis is a serious clinical condition. The prognosis depends on the degree of inflammation. Some scoring systems have been used to prognosticate the outcome of the disease. The scoring systems include Ranson, Modified Glasgow scale and APACHE II. These are referred to as the multiple factor scoring systems.

Ranson's Early Objective Signs of Severity of Acute Pancreatitis

On Admission	After initial 48 Hours
Age > 55 years	Serum calcium < 8 mg/dl
Glucose > 200mg/dl	Arterial PO ₂ < 60 mm Hg
WBC > 16,000/mm ³	Base deficit > 4 mEq/dl
LDH > 350 iu/dl	Serum urea nitrogen increase > 5 mg/dl
AST > 250 u/dl	Haematocrit fall > 10%
	Fluid requirement of > 6,000 mls

AST: aspartate aminotransferase; PO₂: oxygen tension; WBC: white blood cell count

Note: Less than 3 signs = mild pancreatitis; 3 signs and above = severe pancreatitis

Modified Glasgow Criteria

P _a O ₂	< 60mmHg
Albumin	< 32g/L
Calcium	< 2.0mmol/L
White cell count	> 15 per 10 ⁶ cells/L
Aspartate transaminase	> 100units/L
Lactate dehydrogenase	> 600units/L
Glucose	> 10 mmol/L (in non-diabetic patients)
Urea	> 16mmol/L

Pancreatitis is regarded as severe if three or more of the severity score criteria are present within the first 48 hours of the onset of the attack. Other scoring systems based on different criteria include the following

- APACHE 11 (Acute Physiology and Chronic Health Evaluation 11)
- Biochemical scoring: C-reactive peptide levels
- Hong Kong scoring: Based on glucose and urea
- Immunological scoring: Interleukin
- Radiological scoring

Both the sensitivity and specificity of these newer prognostic systems are comparable to the Ranson criteria.

CHRONIC PANCREATITIS

This is a persistent, progressive, irreversible damage of the pancreas resulting from a chronic inflammation.

Aetiology:

- Alcoholism is the major cause and accounts for 70% of cases.
- Hypercalcaemia (hyperparathyroidism),
- Trauma,
- Hyperlipaemia,
- Gallstones
- Malnutrition
- Familial predisposition: May be found to run in families without any associated risk factor.
- Iatrogenic: No known cause in about 15% of cases.

Pathology: There is a progressive loss of function in chronic pancreatitis. This affects both the endocrine and exocrine pancreatic functions. Two forms of the disease have been described – chronic calcific pancreatitis and chronic obstructive pancreatitis. The pathological features include focal necrosis, segmental or diffuse fibrosis, parenchymal calcification and stricture or dilatation of the pancreatic duct.

Complications of chronic pancreatitis include

- Diabetes mellitus: Due to destruction of beta cells
- Malnutrition: Due to lack of pancreatic enzymes resulting from deficiency of exocrine function
- Obstructive jaundice: Obstruction of common bile duct
- Pancreatic pseudocyst/abscess.
- Splenic vein thrombosis, splenic artery aneurysm,
- Pancreatic ascites/pleural effusion
- Pancreatico-enteric fistula
- Carcinoma of the pancreas
- Narcotic addiction: Due to frequent dosing of analgesics for pain relief

Clinical features: The cardinal symptom is recurrent episodes of unrelenting chronic epigastric pain that radiates through to the back. As mentioned above, there are clinical features of deficiency of both the endocrine as well as exocrine pancreatic function. There is therefore clinical evidence of type 1 diabetes mellitus (endocrine) as well as steatorrhoea and malnutrition (exocrine). The obstructive type may present with clinical features of obstructive jaundice.

Differential diagnoses include peptic ulcer disease, gallstones, angina pectoris and pancreatic cancer.

Investigation: The following will help to confirm the clinical diagnosis of chronic pancreatitis

- Serum amylase and lipase: These may be normal due to extensive loss of pancreatic tissue. They are therefore not diagnostic
- Liver function tests: Serum bilirubin and alkaline phosphatase may be raised in obstructive jaundice
- Fasting blood sugar: The blood sugar is raised due to insulin deficiency. This test may be supplemented with the glucose tolerance test.
- 72 hour faecal fat estimation: Will confirm steatorrhoea. Should not be higher than 5 grams daily on a normal diet
- Secretin/cholecystokinin test: This assesses the volume and bicarbonate content of gastrointestinal juice. A duodenal tube is passed and pancreatic secretion is stimulated by the injection of secretin and cholecystokinin. Chronic pancreatitis is diagnosed by a low volume output ($> 2\text{ml/kg}$) and normal bicarbonate level
- Plain abdominal X-ray: May show pancreatic calcification.
- Abdominal ultrasound and CT scan: May demonstrate pancreatic calcification, multiple cystic collections, stones, pseudocyst and anomalies of the common bile duct.
- Endoscopic retrograde cholangiopancreatography (ERCP): May confirm the above findings - ductal irregularities with multiple stenosis and dilatation (beaded appearance).
- Other radiological investigations are: MRCP, endosonography and CT-guided FNAC
- Coagulation tests: Prothrombin time, INR and APTT

Treatment: This comprises of both medical and surgical modes of treatment

Medical treatment involves

- Discontinuation of alcohol abuse: This may reduce frequency of attacks but not has no effect on existing pancreatic damage
- Pain management: Narcotic analgesia initially but may eventually require celiac or splanchnic plexus block
- Insulin administration for diabetes mellitus
- Supplements: Pancreatic enzymes should be administered in order to improve absorption of nutrients and ultimately enhance the nutritional status. Supplements of vitamins, minerals, and medium chain fatty acids are also helpful.
- Diet: Should consist of low fat, high carbohydrate, high protein diet. Patient is advised to take small but frequent meals

- Ascitic tap to reduce intra-abdominal pressure and splinting of the diaphragm associated with pancreatic ascites.

Surgical treatment: The main indications for surgical treatment are

- Severe and intractable pain: This may be the main indication for surgical treatment
- Severe malabsorption
- Suspicion of malignant transformation
- Multiple relapses
- Biliary obstruction.
- Complications such as pseudocyst, ascites, fistula and ductal stenosis may also benefit from surgery.

Surgical treatment is aimed at draining the dilated pancreatic ductal system with or without excision of necrotic pancreatic tissue. This is useful in the relief of pain. The possible options include

- Coeliac or splanchnic block for pain relief
- Puestow's procedure: longitudinal pancreaticojenostomy
- Frey's procedure: as in Puestow's procedure but with additional core resection of the pancreatic head
- Duval's procedure: distal pancreaticojejunostomy
- Pancreatectomy: Usually a last resort and may be either total or near total.

Prognosis: Generally poor; only about 60 – 70% of cases remain free of pain despite radical surgical treatment. The mainstay of management lies in the treatment of alcohol addiction. The Mental Health team may prove helpful in the achievement of this goal.

PANCREATIC TUMOURS

Neoplasm may involve either the endocrine or the exocrine pancreatic tissue. Benign tumours are rare and include cystadenomas which may be mistaken for pseudocyst of the pancreas. The latter is a known complication of acute pancreatitis. Malignant neoplasm usually involves the ductal system by way of ductal adenocarcinoma. Other malignant tumours include acinar cell carcinoma and cystadenocarcinoma.

DUCTAL ADENOCARCINOMA:

Accounts for about 80% of pancreatic carcinomas and involves the pancreatic ductal system. Any part of the pancreas may be involved – head, body or tail. The pancreatic head is, however, most commonly involved (about 75 %). Cancer of the head of the pancreas belongs to a group of malignant tumours collectively referred to as periampullary carcinoma. The latter consists of

- Cancer involving the pancreatic head within 2 cm of the ampulla (40 to 60%)
- Carcinoma involving the distal bile duct (10%): Cholangiocarcinoma
- Tumour arising from the ampulla of Vater (20 to 40%)
- Periampullary duodenal carcinoma (10%)

Late presentation of pancreatic cancer is literally the rule rather than the exception. This is evidenced by the extent of the growth at presentation

- Growth confined to the pancreas: 10%
- Locally advanced disease: 40%
- Evidence of distant spread: 50%

Risk factors for pancreatic carcinoma:

- Smoking: Responsible for about 25% of pancreatic cancer. The risk is related to both the intensity and the duration of smoking
- Chronic alcoholism: may result in chronic pancreatitis which on its own increases the risk of pancreatic cancer by 10 to 20 fold
- Diet rich in fat and protein has been incriminated. On the other hand, diet rich in fruits and vegetables has been found to be protective against the development of pancreatic cancer
- Diabetes mellitus, notably the type II variety increases the risk of pancreatic cancer by 60%. Interestingly, diabetes mellitus may be the presenting symptom of pancreatic cancer
- Chronic pancreatitis: Increases the risk of pancreatic cancer by 10 to 20 fold
- Genetic predisposition: Familial in 10 to 20% of cases. Sporadic pancreatic cancer has been linked to the BRCA2 gene in 7 to 10% of cases
- H. pylori: This infection has been associated with pancreatic cancer. Further research on this is ongoing
- Others include haemachromatosis with calcification

It is commoner in males (M: F=3:2) and in the older age group (above 60).

Pathology: Tumour sites are head and neck (70%), body and tail (30%). There are solid (infiltrating ductal carcinoma, acinar cell carcinoma and pancreaticoblastoma), and cystic (mucinous, serous, and intraductal papillary) neoplasms.

As usual, tumour spread is by various routes – direct, lymphatic, blood stream and transcoelomic.

- Spread via the blood stream is to the liver and the lungs
- Transcoelomic spread: Peritoneum and results in ascites.
- Lymphatic spread: Accounts for associated left supraclavicular lymphadenopathy (Virchow's node; Troisier's sign). Similar spread to the umbilicus will present as a nodule – Sister Joseph's nodule

CLINICAL FEATURES:

These depend largely on the anatomical location of the tumour.

- Head of pancreas: Presents as obstructive jaundice – painless and progressive at onset. It is later associated with epigastric pain that radiates to the back. The subsequent abdominal pain may be due to obstruction of the pancreatic duct with its attendant stasis. It is important to note that the main difference between the clinical presentation of carcinoma of the head of the pancreas and that of its closest differential, obstructive jaundice due to stone in the common bile duct lies in the mode of presentation of the common key symptoms of pain and jaundice. Whereas carcinoma of the head of the

pancreas presents initially with painless progressive jaundice, choledocholithiasis presents with right hypochondrial pain and intermittent jaundice. Involvement of the retropancreatic nerves contributes to the subsequent abdominal pain of pancreatic cancer. Carcinoma of the head of the pancreas manifests with features of obstructive jaundice described earlier: pruritus, passage of pale stools and darkish urine. As with all cancers, there is associated weight loss, anorexia and weakness. Occasionally there will be migratory thrombophlebitis (Trousseau's sign). Examination may reveal a palpable gallbladder in the right hypochondrium – Courvoisier's rule. The latter states that if in a jaundiced patient the gallbladder is palpable, the jaundice is not likely due to gallstones. This is because the resultant inflammation would have resulted in a fibrosed, and shrunken gallbladder that would not be able to distend. Troisier's sign (left supraclavicular lymphadenopathy) may be positive. A rare form of presentation is with upper gastrointestinal haemorrhage due to either invasion of the wall of the duodenum or portal hypertension arising from obstruction of the portal vein.

- Body and tail of the pancreas: This may present with loss of weight, and abdominal pain. The growth may be silent in nature and this may account for the late presentation.

Diabetes mellitus may be associated with tumour at any site. Patients may equally present with evidence of metastatic spread to distant organs such as the lungs and the liver. Peritoneal involvement will result in ascites.

Differential diagnosis: This includes gastric carcinoma, chronic pancreatitis, and other pancreatic tumours (insulinoma, glucagonoma, and vipoma). Others are cystadenocarcinoma, cholangiocarcinoma, sclerosing cholangitis and the rare duodenal carcinoma.

Investigations: Like all cases of obstructive jaundice, this is aimed at both confirmation of diagnosis as well as preparation of the patient for surgery.

- Liver function test: Conjugated hyperbilirubinaemia; very high level of alkaline phosphatase
- Urinalysis: absence of urobilinogen
- Fasting blood sugar: Raised level in associated diabetes mellitus
- Clotting profile: Deranged due to the inability to absorb vitamin K
- Tumour markers: Carbohydrate Antigen 19-9 (CA 19-9) may be useful
- Abdominal ultrasound scan: Will detect biliary duct dilatation, demonstrate pancreatic head cancer and the distended gallbladder. Dilatation of both the pancreatic as well as the common bile duct is referred to as the 'double duct sign'
- Abdominal CT scan: Will confirm the ultrasound findings. Contrast-enhanced CT will not only delineate the pancreatic lesion but will also determine the relationship between the mass and the portal vein and superior mesenteric vessels.
- Endoscopic ultrasonography (EUSG): It demonstrates the relationship of the tumour to the vasculature, as well as the regional lymph nodes. An EUSG-guided FNAC may be carried out.
- Endoscopic Retrograde Cholangio Pancreatography (ERCP): Currently employed in stenting to relieve obstructive jaundice. May cause acute pancreatitis

- Magnetic Resonance Cholangio - Pancreatography (MRCP): Differentiates between obstruction due to gallstones and cholangiocarcinoma.
- Cytology of the pancreatic juice: Specimen may be obtained in the course of stenting the common bile duct
- Laparoscopy: Aids in staging of disease. Also detects spread to the peritoneum and liver

Treatment: The preoperative treatment is as discussed in the management of obstructive jaundice. The result of specific treatment of pancreatic cancer is not encouraging because of its late presentation. Only 10-15% of cases are operable at presentation. Surgery depends on the site of the tumour and the stage at presentation. In the case of cancer affecting the head of the pancreas, the treatment is as follows

- Early: Whipple's procedure (Pancreaticoduodenectomy): This is aimed at curative treatment. It is an extensive procedure that involves excision of the following structures
 - Head and neck of pancreas including the uncinate process
 - The entire duodenum down to 10 cm of proximal jejunum
 - Distal 40 to 50% of stomach
 - Gallbladder and lower end of bile duct
 - Peripancreatic, periduodenal and pericholedochal lymph nodes

Reconstitution of the upper GIT is as follows

- Pancreaticojejunostomy: End to side
- Hepaticodochojjunostomy: End to side
- Gastrojejunostomy: Beyond hepaticodochojjunostomy

Extended Whipple's procedure: This is indicated when the superior mesenteric vessels and adjacent lymph nodes are involved by pancreatic cancer and entails the resection of more tissue than those detailed above. This is made possible by resecting a segment of the superior mesenteric vessels coupled with a bypass graft and dissection of adjacent lymph nodes.

Postoperatively, patients should be managed in an intensive care unit with nasogastric decompression and adequate intravenous infusion

Complications of Whipple's procedure (25%) include

- Delayed gastric emptying
- Anastomotic leakage
- Pancreatic/biliary fistula
- Postgastrectomy syndromes
- Wound infection
- Pancreatitis.

It should be pointed out that the duodenum is removed with the pancreatic head because they share a common blood supply.

- Late presentation: Will benefit from palliative treatment:
 - ERCP and stenting is the current trend. It is gradually replacing
 - Triple bypass which consists of Roux-en-Y cholecystojejunostomy (or Roux-en-Y choledochojjunostomy), jejunojejunostomy, and gastrojejunostomy.

Jejunojejunostomy is no longer routinely carried out. As pointed out earlier, triple bypass is gradually been replaced by ERCP and stenting

- Pain relief by analgesics; coeliac plexus block may be helpful
- Adjuvant chemotherapy has not been found to be effective. Cytotoxic drugs such as 5-fluorouracil and cyclophosphamide have been tried. Newer drugs include gemcitabine, cisplatin, vincristine and mitomycin are been tried
- Radiotherapy has not proved to be useful

Lesions involving the body and the tail of the pancreas require distal pancreatic resection

Prognosis: As mentioned earlier, the prognosis of pancreatic cancer is generally poor owing to late presentation and diagnosis. About 90% of patients die within one year of diagnosis. It is not much better with the apparently early cases as only 3% of patients are alive 5 years after resection.

OTHER PANCREATIC TUMOURS

Insulinoma

This tumour arises from the pancreatic beta cells and secretes excess insulin. The patient presents with clinical features of hypoglycaemia. These include sweating, palpitations and tremors during a period of fasting. There may also be neuropsychiatric symptoms. These symptoms are relieved by ingestion of glucose. This is the basis of the Whipple's diagnostic triad which consists of the following

- Hypoglycaemia (glucose level < 50 mg %)
- Symptoms of hypoglycaemia: as mentioned above
- Relief of symptoms after administration of glucose

Serum insulin levels remain high in the presence of fasting induced hypoglycaemia

Abdominal and multislice CT scans may pick up small tumours. Intraoperative and endoscopic ultrasound scan may equally aid diagnosis.

Surgical treatment involves tumour resection. Diazoxide is useful in the postoperative control blood sugar. Streptozocin has been tried for inoperable tumours.

Vasoactive Intestinal Polypeptide tumour (VIPoma)

This is also referred to as the Verner-Morrison syndrome. It is a pancreatic tumour that secretes vasoactive intestinal peptide. The latter inhibits the secretion of gastric acid and results in the WDHA (Watery Diarrhoea Hypokalemia Achlorhydria) syndrome.

Diagnosis is made by imaging of the pancreas: CT, MRI and angiography.

Medical treatment of VIPoma involves administration of octreotide and streptozocin. Surgery aims at resection of the tumour and may involve subtotal pancreatectomy.

Glucagonoma is a tumour that involves the alpha cells of the pancreas. They elaborate glucagon which results in diabetes mellitus, glossitis, stomatitis, necrotizing dermatitis and associated loss of weight. Investigation and treatment is as for VIPomas.

Gastrinoma

As discussed elsewhere, these tumours secrete gastrin and may result in the Zollinger-Ellison's syndrome. The increase in the serum gastrin level results in diarrhoea, and recurrent peptic ulceration at unusual sites such as the jejunum. The investigations are as for VIPomas but also include the estimation of the serum gastrin level. Surgical treatment involves tumour resection. Proton pump inhibitors have proved very useful in the management of multifocal tumours.

CHAPTER TWENTY-SEVEN

HAEMATURIA, BLADDER OUTLET OBSTRUCTION, POSTERIOR URETHRAL VALVES

HAEMATURIA

Haematuria denotes the passage of blood in urine. By definition, it is the presence of more than three red blood cells per high-power microscopic field in the urine. It may be obvious to the naked eye that the urine is blood-stained (macroscopic haematuria). On the other hand, it may only be detected on analysis of the urine specimen under a microscope (microscopic haematuria). It should be managed with the utmost degree of seriousness.

Aetiology of haematuria: Can be classified into two broad groups – systemic and local

Systemic causes include any condition that predisposes to bleeding diathesis.

- Purpura
- Sickle cell disease
- Thrombocytopenia
- Anticoagulant therapy

Local causes: This depends on the part of the urinary system

- Kidneys: Trauma, tuberculosis, stones, acute glomerulonephritis and infarcts. Neoplastic causes are transitional cell carcinoma (TCC), hypernephroma, and Wilm's tumour.
- Ureters: Stone, neoplasm (TCC), trauma
- Bladder: Cystitis, tuberculosis, and schistosomiasis. Others are stones and tumours
- Prostate: Both benign and malignant enlargements, prostatitis
- Urethra: Trauma and neoplasm

Management of haematuria: A good history is of utmost clinical importance

- Rule out the presence of bleeding diathesis: Is there bleeding from any other orifice? Is patient on anticoagulant?
- Timing of haematuria in respect to voiding: Initial, total or terminal
 - Initial haematuria is usually associated with a urethral cause while
 - Terminal haematuria with severe pain and frequency is usually due to bladder calculus.
 - Total haematuria is usually associated with lesions involving the upper part of the urinary system (kidneys and ureters)
- Associated pain: Haematuria of neoplastic origin is usually painless while that due to presence of stone in the kidney or ureter is said to be colicky in nature. Renal colic radiates from the loin to the groin. This is strictly speaking not the true picture as there is an intermittent exacerbation over a background of continuous pain.
- In acute glomerulonephritis, there may be a preceding history of sore throat

Clinical examination: General examination may reveal pallor when there is massive blood loss. The blood pressure of the patient should be recorded both for the assessment of blood loss as well as diagnosis of glomerulonephritis. The kidneys should be balloteted as presence of a renal

mass in association with haematuria may suggest diagnosis of adenocarcinoma (Hypernephroma). The lower abdomen should also be examined for a palpable bladder. The latter is suggestive of urinary retention due to bladder outlet obstruction. A digital rectal examination may reveal an enlarged prostate.

Investigations for haematuria

- Urine (mid-stream): Haematuria may either be macroscopic or microscopic. Presence of casts or ova of schistosoma haematobium on microscopy is suggestive of nephritis and bilharziasis respectively. Urine culture may reveal presence of infection
- Intravenous urography (IVU): Preliminary plain film may reveal urinary stones and local lesions such as bladder tumours (filling defects in the bladder) and evidence of bladder outlet obstruction due to prostatic enlargement
- Abdominal computerised tomography (CT) scan
- Transabdominal ultrasound scan
- Cystoscopy: Will reveal any bladder cause of haematuria
- Selective angiography: Will outline any bleeding tumour
- Clotting profile: Platelet count, prothrombin time, PTT and INR
- Full blood count: May demonstrate the degree of blood loss (low PCV) and evidence of on-going infective process (leucocytosis)

Treatment: Address the underlying cause. Patient may require admission and blood transfusion.

GENERAL FEATURES OF BLADDER OUTLET OBSTRUCTION

Bladder outlet obstruction is a clinical condition in which the urinary bladder is unable to efficiently discharge its urinary content due to an obstructive condition around or distal to the bladder neck.

Aetiology includes

- Benign prostatic hypertrophy
- Prostatic cancer
- Urethral stricture
- Posterior urethral valves
- Urinary stones

Pathophysiology

Bladder neck obstruction results in obstruction to the free flow of urine resulting in distension of the bladder and subsequent hypertrophy of the detrusor muscle. This is followed by trabeculation, sacculation and diverticular formation within the urinary bladder. The resultant stasis results in infection of the urine and formation of bladder stones. There is congestion of veins in the wall of the bladder which on rupture will result in haematuria. The increased intravesical pressure may result in kinking and/or transmission of pressure to the ureters resulting in bilateral hydroureters and hydronephrosis. The latter may also result from vesicoureteric reflux. Chronic renal failure may be the ultimate result.

Symptomatology: May be classified into obstructive and irritating symptoms.

- Obstructive symptoms: These include hesitancy, weak stream and dribbling (interrupted stream). Others are straining and incomplete bladder emptying.
- Irritative symptoms : Urgency, frequency, dysuria and nocturia

Unrelieved, the following may occur

- Acute urinary retention: Usually of sudden onset and is associated with suprapubic pain
- Chronic urinary retention: Usually painless but may present with overflow incontinence
- Prolonged bladder outlet obstruction will result in hydroureters and hydronephrosis. Long term hydronephrosis will result in chronic renal failure
- Straining at micturition may predispose to the development of hernias, haemorrhoids and rectal prolapse

POSTERIOR URETHRAL VALVES

This is an abnormality that results from the presence of congenital symmetrical valves at the posterior urethra just below the verumontanum. They are one-way valves in that even though the patient is not able to pass urine freely due to the effect of these valves, a urethral catheter can be passed freely into the urinary bladder. In other words the valves result in antegrade but not retrograde urinary obstruction. This results in gross dilatation of the posterior urethra. The child will manifest with all the clinical features of bladder outlet obstruction

Clinical features: Posterior urethral valves may present during the neonatal period or within the first year of life with difficulty with micturition. This may progress to urinary retention which is usually complicated by urinary tract infection, hydroureters and hydronephrosis. Patient may therefore present with renal failure and failure to thrive

Investigations for posterior urethral valves are

- Micturating cystouregraphy: This is the diagnostic tool of choice. It will demonstrate the dilated posterior urethra with its characteristic 'spinning top' appearance
 - Abdominal ultrasound scan: Will demonstrate any attendant complications such as hydroureters and hydronephrosis
- Intravenous urography: Will demonstrate the dilated posterior urethra
- Urine: Urinalysis, microscopy/culture/sensitivity
- Others: Full blood count, electrolytes/urea/creatinine

Treatment

- As an interim measure, decompress the bladder by either urethral catheterisation or vesicostomy for long-term management.
- Institute treatment for urinary tract infection
- Temporary ureterostomy when there is gross hydroureters
- Definitive treatment: Transurethral resection of the posterior urethral valves

Prognosis: The main determining factor is the degree of renal dysplasia. About 50% of patients who present at infancy do not survive. Late presenters tend to do better.



Micturating cystourethrogram of a patient with posterior urethral valves. Note the 'spinning top' appearance of the dilated posterior urethra

CHAPTER TWENTY-EIGHT

URINARY CALCULI

Stones can occur in any part of the urinary tract. There are various types of urinary calculi:

- Calcium oxalate calculi (oxalate calculi): They account for about 60% of urinary calculi and are hard with a sharp spiky surface. They traumatisse the underlying epithelium and evoke haematuria which is responsible for the darkish hue of oxalate calculi.
- Phosphate calculi: These account for about 33% of urinary calculi and are often incorporated with ammonium, magnesium phosphate and calcium (triple phosphate stone). Phosphate calculi are usually of infective origin and classically present as 'staghorn calculi' in the renal pelvis
- Uric acid calculi: They account for about 5% of urinary calculi and are usually multiple and radiolucent
- Cystine calculi: About 1% of urinary calculi are of cysteine origin. They are commonly found in patients that have cystinuria due to a congenital error of metabolism and are usually multiple.
- Xanthine calculi: These are rare.

Aetiology of urinary calculi: Predisposing factors include

- Urinary stasis: Hydronephrosis, bladder diverticulum, neurogenic bladder and bladder outlet obstruction
- Solute/solvent disproportion: May result from
 - Dehydration due to inadequate fluid intake particularly in the tropics.
 - Hypercalcaemia may result from hyperparathyroidism and prolonged immobilisation.
 - Elevation of the serum uric acid level following gout
 - Chemotherapy for leukemia and polycythaemia may result in the formation of uric acid stones
- Foreign bodies:
 - Epithelial sloughs (bacterial infections)
 - Nonabsorbable sutures
 - Broken-off catheter tip
 - Sloughing of urinary epithelium (hyperkeratosis following vitamin A deficiency)

Clinical features of urinary calculi:

These depend on the part of the system involved. It must be pointed out that stones may be silent and symptomless, particularly when embedded in the renal parenchyma. Attention may only be drawn to their presence when bilateral involvement results in uraemia.

- Pain: Loin pain when stone is lodged in the renal calyx. Stone in the pelvis and ureter will result in 'renal colic' with the pain radiating to the groin and pelvis. As pointed out earlier, it is not strictly colicky as there is an intermittent exacerbation over a

background of continuous pain. Bladder calculi will present with strangury (pain radiating to the tip of the penis occurring at the end of micturition). Pain is usually referred to the tip of the penis or labia majora. Bladder calculi in childhood may result in a child screaming and pulling at his penis towards the end of micturition.

- **Haematuria:** Usually microscopic in urinary calculus disease.
- **Clinical features of urinary tract infection**
- **Interruption of the urinary stream in bladder calculus**

Investigation of urinary calculi

- **Urine:** The urine is examined for presence of blood, crystals and casts
- **Plain abdominal X-ray:** About 90% of urinary calculi are radio-opaque. Note that the opposite is the case with gallstones in which only 10% are radio-opaque
- **Ultrasound scanning**
- **Contrast enhanced CT**
- **Intravenous urography:** Will also screen the functional activity of the contralateral kidney
- **Serum calcium assay to rule out hypercalcaemia**
- **Serum uric acid assay in uric acid stones**
- **Stone analysis:** Carried out on any excreted stone
- **Full blood count:** PCV, WBC
- **Electrolytes, urea and creatinine**

Treatment of urinary calculi

A small silent stone in the kidney should be placed under surveillance. This also applies to the treatment of small calculi in other parts of the urinary system as they are likely to be passed out spontaneously. Surgical removal, however, becomes imperative if there is evidence of complication (infection and hydronephrosis). The following may be employed in the treatment of urinary calculus disease

- **Liberal intake of fluids**
- **Extracorporeal shock wave lithotripsy:** The shock waves disintegrate the calculi
- **Endoscopic stone removal:** Dormia basket and ureteric meatotomy for ureteric stones
- **Litholapaxy:** Employed for bladder stones. Includes ultrasound lithotripsy, laser lithotripsy and percutaneous suprapubic litholapaxy
- **Open surgery:** Percutaneous nephrolithotomy, pyelolithotomy, ureterolithotomy and cystolithotomy
- **In association with surgery for the predisposing condition such as benign prostatic hypertrophy**

Recurrent stones should be investigated in order to identify any underlying primary condition such as parathyroid adenoma.

CHAPTER TWENTY- NINE

BLADDER TUMOURS

Bladder tumours consist of benign and malignant varieties. Benign tumours consist of transitional cell papillomas while there are various types of malignant tumours. As a rule of thumb; however, any bladder tumour should be considered malignant until proved otherwise. This is analogous to the clinical management of breast lesions.

Primary malignant tumours

- Transitional cell carcinoma: 90%
- Squamous cell carcinoma: 5%. Common in bladders with underlying bilharziasis and stones due to metaplastic change from chronic irritation.
- Adenocarcinoma: 2%. Common at the fundus of the bladder. Usually located at the site of the urachal remnant. It can also result from glandular metaplasia in other sites.

Secondary bladder tumours: Primary sites are neoplastic lesions involving the neighbouring structures such as sigmoid colon, rectum, prostate, uterus, and ovaries.

Aetiology of bladder cancer: Multiple risk factors include

- Smoking: Found in about 60% of cases. Smoking reduces the age of onset and also increases the degree of invasiveness of bladder cancer
- Chemicals and dyes: Aromatic amines and dyes such as 2-naphthylene, benzidine, and toluidene. Occupations at risk include textile, rubber, petroleum and paint industries. Hairdressing as a profession is also a risk factor
- Drugs: Cyclophosphamide and phenacetin-containing analgesics
- Schistosoma haematobium: Associated with squamous cell carcinoma
- Stones: Associated with squamous cell carcinoma
- Activation of dormant oncogenes: E2F3, ras, c-erbB
- Inactivation of tumour suppressive genes: p53, p21 and p16
- Activation of cancer potentiating factors: lysosomal enzymes, urinary plasminogen activator, angiogenic factors and epidermal growth factors

Clinical features of bladder cancer

- Painless haematuria. May be microscopic initially and macroscopic (gross) later
- Loin pain and features of ureteric colic when there is ureteric obstruction and hydronephrosis
- Pain around the lower part of the body (suprapubic region, groin, perineum, anus and thighs). This is sequel to the involvement of the pelvic nerves
- Lower urinary tract symptoms especially irritative symptoms: Frequency, urgency and dysuria.

- Retention of urine: Bladder outlet obstruction from clots or growth involving the bladder neck
- Symptoms of metastatic disease: Weight loss, anorexia, and anaemia from chronic blood loss. There may be symptoms of local and metastatic spread (liver and lungs)

Complications of bladder cancer include

- Anaemia due to blood loss
- Retention of urine: Due to urinary obstruction from blood clots and/or tumour
- Cystitis: This is sequel to urinary stasis
- Intractable pain: Due to tumour infiltration of plexuses of pelvic nerves
- Priapism: Occurs with involvement of the base of the penis
- Oedema of the lower limbs: May be due to venous or lymphatic obstruction
- Fistulation to neighbouring structures: Rectum in the male and vagina in the female

Staging of bladder carcinoma: Carried out by bimanual palpation under general anaesthesia and biopsy (Examination under anaesthesia and transurethral resection)

The TNM staging for Transitional Cell Carcinoma (TCC)

Stage	Definition
Superficial tumour	
Ta	Confined to mucosa
T1	Invasion into lamina propria
Tis	Carcinoma in situ
Invasive tumour	
T2a	Invasion of inner half of muscularis
T2b	Invades deep muscle (outer half)
T3a	Microscopic invasion of perivesical fat
T3b	Macroscopic invasion of prevesical fat
T4a	Invasion into prostate, uterus or vagina
T4b	Invasion of pelvic or abdominal wall
Lymph node (N)	
N0	No regional lymph node
N1	Single regional lymph node < 2cm
N2	Single regional node >2cm, <5cm
N3	Node > 5cm
Distant metastases (M)	
M0	No distant metastasis
M1	Distant metastasis

Bladder tumours may also be graded histologically from 1 to 3 (low to high grade).

The clinical importance of both staging and grading lies in the prognostic implication

- Low grade tumours: Do not invade muscle; rarely metastasise nor progress to higher grade or stage lesions. They, however, tend to recur after treatment.
- Muscle invasive tumours: Tend to metastasise and therefore have a poor prognosis

Investigations

- Urine cytology: More relevant in high grade lesions
- Blood: FBC, electrolytes, urea and creatinine
- Ultrasound scan of the full bladder and abdomen will demonstrate degree of local and visceral spread respectively
- Cystourethroscopy: Biopsy is carried out in two parts – superficial portion and base with some underlying muscle respectively
- Intravenous urography: Will show filling defects and urinary obstruction
- CT scan of the abdomen and pelvis: For staging in muscle-invasive cancer
- Chest X-ray: To detect chest metastases in invasive bladder cancer
- Bimanual examination: Carried out both pre- and post-endoscopic resection under general anaesthesia. Assesses the clinical features of the bladder mass.

Treatment of bladder tumours

Initial management entails transurethral resection of the tumour. The resected specimen is made to include the underlying muscle. This is analysed histologically to determine its involvement in the malignant process. Bimanual palpation is carried out both pre and post resection in order to detect any residual growth. The findings help to categorise bladder tumours into two groups: superficial and invasive cancers. This has management implications

Superficial tumour

Initial transurethral resection is usually curative. Regular cystoscopic examinations are, however, indicated for the early detection of recurrence. The latter is minimised by the intravesical instillation of mitomycin C.

Immunotherapy: This is achieved by a weekly intravesical instillation of a live-attenuated Bacille Calmette-Guerrin (BCG) over a 6 – week period. It has been found to stimulate an anti-tumour immunological response in over 60% of cases. Subsequently, it is administered six-monthly and its effect monitored by serial cystoscopy. BCG administration is known to halt/delay the muscle-invasive propensity of high-grade tumours.

Invasive tumours

Treatment involves surgery, radiotherapy and systemic chemotherapy

Surgery: The rare tumour occurring in the vault of the bladder can be treated by partial cystectomy. Following partial cystectomy, the patient is still able to pass urine as only a part of the bladder is resected. Radical cystectomy, however, is indicated in inoperable lesions. This entails the surgical removal of the bladder, prostate and seminal vesicles in the male and the uterus, ovaries and vagina in the female. The surgery is completed by the addition of a pelvic

lymphadenectomy. Urinary diversion forms an integral part of the procedure. Initially, this was achieved by the transplantation of the ureters into the colon. It was, however, found to be associated with a high risk of ascending infection, uraemia and hyperchloraemic acidosis due to the excessive reabsorption of chlorides from the urine passed into the colon. Currently, urinary diversion is achieved by the implantation of the ureters into an ileal conduit. An orthotopic neobladder can also be constructed from a bowel segment.

Radical radiotherapy: This is fractionated over a 5- to 6-week period. Complications include haemorrhagic cystitis and bladder fibrosis. Others are radiation proctitis and damage to the small intestine. There is poor response to radiotherapy.

Systemic chemotherapy: There are four ways of applying chemotherapy in the management of bladder cancer

- Before surgery (neoadjuvant): Helps to shrink the tumour in order to render it more amenable to surgical treatment
- Postoperative (or sometimes post irradiation): As an adjuvant treatment to reduce recurrence
- During radiation therapy (chemoradiation): To enhance the effect of radiotherapy
- As the main treatment for advanced tumours that have spread to distant sites

The most commonly used drug combinations for bladder cancer are

- Methotrexate, Vinblastine, Adriamycin and Cisplatin (MVAC)
- Cisplatin, Methotrexate, and Vinblastine (CMV)
- Gemcitabine and Cisplatin
- Carboplatin and either paclitaxel or docetaxel for patients with poor renal function

Drugs that are used for single therapy include gemcitabine, carboplatin, docetaxel and cisplatin. Others are paclitaxel, adriamycin, 5FU, methotrexate, vinblastine and ifosfamide.

Immunotherapy: This involves the use of drugs to aid an individual's immune system in the recognition and destruction of the ravaging cancer cells. It has been applied in the management of bladder cancer by the use of the following

- **Intravesical BCG (see above):** Of use in the management of early bladder cancer
- **PD-1 inhibitors:** Of use in patients with advanced bladder cancer particularly those that recur after chemotherapy. PD-1 is a protein that is found on the surface of T cells. They help keep the body's immune system in check. When PD-1 attaches to another protein, PDL-1 that is present on the surface of the cancer cell, it equally halts the destruction of the cancer cell by the T cell. PD-1 inhibitors increase the vulnerability of the cancer cells by attaching to PDL-1. By this mechanism, they facilitate the destruction of the cancer cells by the T cells. They are used to shrink tumours by slowing their growth. PD-1 inhibitors include Atezolizumab and Pembrolizumab. They are given as intravenous infusions two- or three- weekly. Their side effects include fatigue, nausea, fever, anorexia, and rash. Others are urinary tract infection, diarrhoea and constipation. Occasionally, the immune system goes 'haywire' with the use of these drugs and starts

attacking other parts of the body such as the lungs, intestine, liver and hormonal-producing glands.

- **Synthetic interferon:** Interferon is naturally produced by the immune system and helps to fight infection. A synthetic version, alfa-2b (intron A) may be combined with BCG in the management of bladder cancer.

CHAPTER THIRTY

BENIGN PROSTATIC HYPERPLASIA (BPH)

The prostate is located at the bladder neck and contributes to semen production. It is about the size of a walnut and completely encircles the urethra. The main physiological function of the prostate is the production of semen which acts as a vehicle for the transportation of spermatozoa. It almost invariably hypertrophies as a part of the ageing process. Statistics have shown that about 50% of men demonstrate histological evidence of BPH by the age of 50 and 75% by the age of 80. The human prostate is the only mammalian prostate with a capsule. The latter tends to restrict the expansion of the prostate as it hypertrophies thereby giving rise to bladder outlet obstruction. Whereas the benign enlargement affects the periurethral (inner) aspect of the gland, prostatic cancer occurs around the outer part (periphery).

Pathogenesis of benign prostatic hypertrophy

There are two phases of prostatic growth in males. At puberty, the prostate doubles in size. The second phase of growth begins around the young adult stage of 25 and continues into late adulthood. It is this second growth phase that results in BPH later in life. BPH is due to hyperplasia of both glandular and stromal cells and affects the periurethral region of the gland. This results in the formation of discrete nodules which when sufficiently large, compress the urethra and give rise to clinical features of bladder outlet obstruction. Certain factors are thought to be responsible for the development of BPH

- Dihydrotestosterone (DHT): This is formed from testosterone and the conversion process is potentiated by the enzyme 5-alpha reductase. DHT is a known facilitator of prostatic growth. Despite the relatively low level of testosterone at old age, DHT production does not abate. This accumulation of DHT in the prostate is responsible for the glandular and stromal hyperplasia found in BPH
- Oestrogen stimulation: With age, there is a reduction in the production of testosterone. This creates a relative rise in the oestrogen : testosterone ratio. It is thought that oestrogen sensitises the prostatic tissue to the influence of dihydrotestosterone thereby enhancing prostatic hyperplasia.
- The neoplastic theory regards BPH as a benign neoplastic lesion

Risk factors for BPH

- Age: BPH is a disease of old age and the peak incidence is 60 – 65. It is rare in men younger than 40
- Family history: The risk of BPH is higher if a close relative (father, brother) has suffered from it.
- Marital status: Studies have shown that married men are more prone to BPH than their single counterparts. No reason is adduced for this as there appears to be no link between sexual intercourse and prostate growth.

- Lifestyle : Unhealthy factors include cigarette smoking, lack of exercise and unhealthy diet. It is believed that diets rich in lycopene and zinc such as tomatoes are protective.
- Medical conditions: These include diabetes mellitus, obesity and high blood pressure. Others are peripheral artery disease and low levels of HDL cholesterol.

Clinical presentation of benign prostatic hypertrophy

Patient manifests the clinical features of bladder outlet obstruction. It is worthy of note that whereas straining improves the stream in urethral stricture, it has no such effect on prostatic enlargement. Urethral stricture may be preceded by urethral trauma and clinical features of urethritis. Diagnosis of BPH is made by way of digital rectal examination. Usual findings are

- Uniformly enlarged prostate that is firm in consistency and has a smooth capsule
- Presence of a median sulcus
- Rectal mucosa moves smoothly over the enlarged prostate

The International Prostate Symptom Score (I-PSS)

This is an internationally standardised screening tool aimed at evaluating the symptoms of prostatic obstruction. It takes into consideration the patient's assessment of the severity of seven key symptoms of bladder outlet obstruction. These comprise of both obstructive and irritating symptoms. In addition, the last question borders on the patient's assessment of his quality of life in respect of the prevailing symptoms of the ailment. Each question is provided with five answers which are graded in order of severity of the symptoms. This tool is employed in the determination of the severity of the condition with a view to taking management decisions.

The questions refer to the following symptoms

Question	Symptom
1	Incomplete emptying
2	Frequency
3	Intermittency
4	Urgency
5	Weak stream
6	Straining
7	Nocturia

The answers are assigned points from 0 to 5. The total score ranges from 0 to 35 (asymptomatic to very symptomatic).

The 8th question is more or less a measure of the patients overall assessment of his clinical condition in relation to the ailment. This quality of life question may go like this: "If you were to spend the rest of your life with your urinary condition the way it is now, how would you feel about that – delighted, pleased, mostly satisfied, mixed (equally satisfied/dissatisfied), mostly dissatisfied, unhappy, terrible?". In addition to evaluating the patient's perception of the severity of the illness, this last question may go a long way in marking a starting point in a patient-doctor relationship.

This symptom score was developed by the International Scientific Committee under the patronage of the World Health Organisation (WHO) and the International Union against Cancer

(UICC). The first seven questions are identical to those developed by the American Urological Association Measurement Committee. The Symptom Index categorises the symptoms based on the overall score:

- Mild (symptom score less than or equal to 7)
- Moderate (symptom score range between 8 and 19)
- Severe (symptom score range between 20 and 35)

In the extreme situation, urinary retention (acute or chronic) may be the sole presenting clinical feature

Differential diagnosis of benign prostatic hyperplasia

- Prostatic carcinoma
- Urethral stricture
- Neurogenic bladder
- Acute prostatitis
- Urinary tract infection

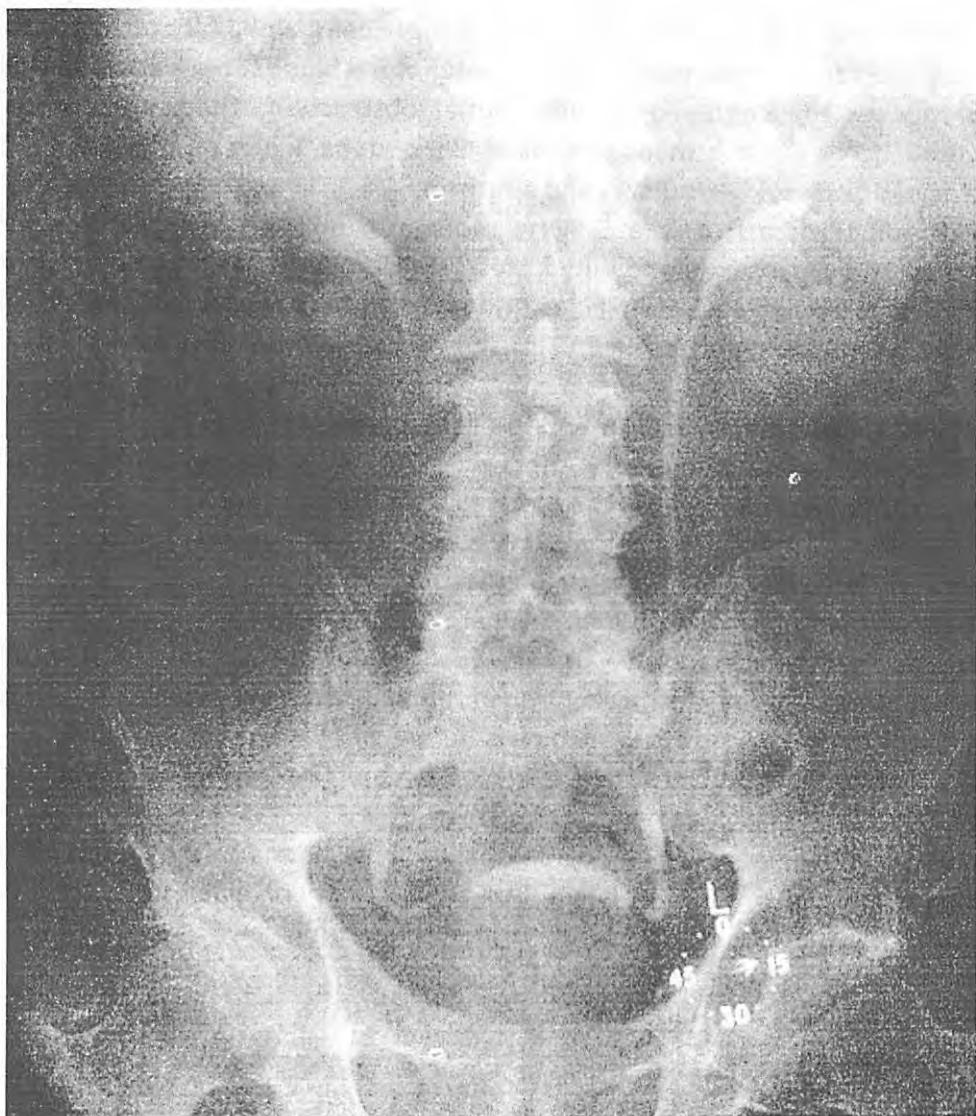
Investigations for BPH

- Full blood count
- Electrolytes, urea, creatinine:
- Prostate-Specific Antigen (PSA). Aids in raising suspicion of prostatic cancer. Levels above 10 nanogram are suggestive.
- Abdominopelvic ultrasound scan of the kidney, ureter and bladder (KUB). It also assesses the size of prostatic enlargement
- Chest X-ray: Required in the preoperative assessment of any patient above the age of 40
- Urine: Urinalysis and microscopy, culture and sensitivity. In addition to guiding the preoperative treatment of urinary tract infection, knowledge of the pathogenic organism and its antibiotic sensitivity will equally guide in the treatment of postoperative urinary tract infection.
- Electrocardiogram: As a preoperative investigation in patients above the age of 40
- Postvoidal residual volume (PVRV): Rises to a significant value as symptoms progress. Urinary flow values below 10mls/sec and residual volumes above 100mls are indicative of significant urinary obstruction
- Urodynamics: Tests the function of the bladder. The detrusor pressure at micturition helps to determine the cause of a poor urinary stream. A high detrusor pressure suggests a bladder outflow obstruction while a low pressure is more in keeping with detrusor failure
- Intravenous urography: It is no longer as popular as it used to be. It demonstrates the pathognomonic fish hook ureters (also known as J-shaped ureters; hockey stick ureters). It describes the distal ureters as seen in bladder outlet obstruction due to prostatic hypertrophy. It may be due to prostatic median lobe enlargement.

Treatment of BPH

This depends on the stage of the disease after careful evaluation of symptomatology. The International prostate scoring system has proved a useful guide in the categorisation of treatment

- Stage 1: Mild symptoms that do not necessarily disturb patient's lifestyle. Besides, there is no clinical evidence of significant urinary obstruction. They are managed by close observation – 'watchful waiting'
- Stage 2: Symptoms are such that the patient is worried but there is no evidence of significant obstruction. These patients will benefit from medical treatment



Intravenous urography in BPH showing the fish hook distal ureters

- Stage 3: There is evidence of significant urinary obstruction (urine flow of less than 10mls per second and persistent residual urine of more than 100mls). Such patients will benefit from surgical intervention
- Stage 4: Patients presenting with complications such as chronic urinary retention and bladder stones. There is no alternative to surgery in such patients

Treatment may be either medical or surgical

Medical treatment: This is employed in the management of the elderly, patients who are unfit for surgery and those who refuse surgery. It is also indicated in patients with mild symptoms without evidence of significant urinary obstruction

- Alpha-1 receptor blockers: Alpha-adrenergic receptors are responsible for the contraction of the myofibroblasts (smooth muscles) present in the prostatic stroma. By decreasing the sympathetic resting tone of these cells, alpha blockers relax the smooth muscle and cause an increase in the diameter of the bladder neck and prostatic urethra. This reduces the degree of bladder outlet obstruction. They are used as first-line medical therapy in the management of BPH. Alpha-1 blockers in use are terazocin, prazocin, doxazosin, tamsulosin and alfuzosin.
- Finasteride (Proscar): It is a 5-alpha reductase inhibitor and therefore blocks the conversion of testosterone to dihydrotestosterone. The latter is a potent stimulator of benign prostatic hyperplasia. Finasteride therefore slows down the progression of benign prostatic hypertrophy by causing an involution of the prostate glandular tissue. The following are the possible mechanism of action of finasteride
 - Inhibits the activity of vascular endothelial growth factor (VEGF)
 - Androgens have a stimulatory effect on VEGF. By reducing the level of circulatory androgen, finasteride reduces the blood flow to the prostate
 - Reduces the level of VEGF in the kidneys and the microvessel density
 - Reduces the level of TNF – alpha and PSA
 - Reduces systemic inflammation and overexpression of PSA

Due to its action on prostatic blood flow, it has equally been found to be useful in reducing the blood loss during prostatectomy and prevention of rebleeding following surgery. This is achieved by daily administration of the drug for seven days before surgery.

A combination of drugs from both groups (alpha blockers and finasteride) has been found to have a synergistic effect.

- Sawpalmetto (Prostacare): This traditional drug has been used since the 19th century in the management of BPH but has only recently been universally accepted. It has been found to relieve the symptoms of BPH safely and effectively.
 - Relieves lower urinary tract symptoms
 - Improves urinary peak flow
 - Improves patient's quality of life.
 - Has been found to have no significant side effects

Indications for surgery

- Failure of medical therapy
- Intractable symptoms of prostatism
- Recurrent acute urinary retention
- Chronic urinary retention
- Haemorrhage
- Presence of urinary stones
- Established/impending renal failure: Hydronephrosis with a high or rising level of creatinine
- Recurrent urinary tract infection
- Uroflowmetry: Rising post-void residual urine volume; maximum flow rate < 10 ml/sec

Surgical approach: May be closed or open.

- The closed approach is by way of transurethral resection of the prostate (TURP) and is currently regarded as the 'gold standard' of surgical treatment. Prostatic tissue is 'chipped' out by means of a specially designed cutting element attached to a scope. The cut prostatic tissue is flushed out by water irrigation. The sole aim of TURP is to resect the adenoma endoscopically just enough to reopen the urinary passage. The recovery rate is fast. The earlier technique was best suited for mild to moderate prostatic enlargements. With technological advancement, however, even larger glands can now be tackled by TURP.
- Open prostatectomy: Still practised widely in developing countries due to the unaffordability and inadequate human capacity in the handling of TURP equipments. When the latter is even available, it is beyond the financial reach of the average citizen due to the high poverty level. Surgical approach is either transvesical (Freyer's technique) or retropubic (Millin's technique). The latter is ideally preceded by cystoscopy to rule out the presence of complications such as stones and diverticulum. Complicated cases are best approached transvesically.
- Newer alternative methods of prostatectomy include transurethral laser ablation of prostate and transurethral plasmakinetic resection of the prostate. A combination of high intensity diode laser with bipolar transurethral resection of the prostate may be a new strategy for the treatment of large prostates.

Transurethral plasmakinetic resection of the prostate: Also known as

- Transurethral vaporisation of the prostate
- Plasmakinetic resection of the prostate (PRP)

Transurethral resection of the prostate (TURP) is regarded as the gold standard in the endoscopic management of benign prostatic hyperplasia. TURP resection is carried out by a monopolar electrical system. Plasmakinetic resection of the prostate (PRP), on the other hand, is carried out with a bipolar system. The latter enables the use of normal saline as the irrigation fluid instead of sterile water, hyponatremic glycine or mannitol solutions which are the fluids of choice in TURP. This singular measure thus avoids the risk of transurethral resection (TUR) syndrome. PRP

offers rapid tissue removal and haemostasis during resection with better vision under saline irrigation while eliminating the risk of TUR syndrome. The advantages of bipolar PRP include

- Low Intraoperative and postoperative complications
- Excellent Intraoperative haemostasis
- Short convalescence
- Absence of fluid absorption and consequently TUR syndrome

On the flip side, urethral injuries and meatal strictures appear to be more with PRP

Transurethral resection syndrome:

This is a clinical condition of fluid overload and iso-osmolar hyponatremia during TURP arising from large volumes of irrigation fluid. The latter is required to maintain visibility despite bleeding from tissue beds. It occurs as a result of the absorption of the irrigation fluids into the prostatic venous plexuses. It can also occur during other endoscopic procedures such as uretero-renoscopy (URS), percutaneous nephrolithotomy (PCNL) and transcervical resection of endometrium (TCRE). The incidence of TURP syndrome ranges between 2.5 and 20%

Pathophysiology: Normal saline is not used in TURP because it disperses high frequency current from the resectoscope. Glycine (1.5%) and sterile water are the most widely used irrigation fluids in urological endoscopic surgeries. Absorption of fluid will overload the circulation and results in the following

- Water intoxication
- Hyponatremia
- Visual disturbances
- Coagulopathies (DIC)
- Bacteremia, septicaemia
- Hypothermia
- Glycine toxicity
- Ammonia toxicity

Clinical features: TURP syndrome can occur any time from few minutes after commencement of surgery to several hours after surgery. The symptoms of TURP syndrome include

- Headache, dizziness, confusion, restlessness and lethargy
- Tight feeling in chest and throat, shortness of breath
- Nausea, retching and abdominal pain

Examination may reveal

- An increase in both the systolic and diastolic blood pressure
- Reduced heart rate
- Dilated pupils which react sluggishly to light
- Short tonic and clonic seizures which may result in coma
- Delayed intervention may lead to cyanosis, hypotension and ultimately cardiac arrest

A patient under anaesthesia may manifest the following

- Unexplained rise in blood pressure followed by hypotension
- Refractory bradycardia
- ECG changes: Nodal rhythm, ST changes, U waves and widening of the QRS complex
- Delayed recovery from general anaesthesia and muscle relaxants

Prophylaxis for TURP syndrome

- Correct preoperative hyponatremia
- Close monitoring of the central venous pressure (CVP)
- Restrict the duration of the TURP to one hour
- Ideal height of fluid should be 60 cm and maintain continuous flow
- Endeavour to preserve the prostatic capsule
- Estimate the serum sodium level half-hourly
- Administer frusemide prophylactically in order to prevent overload
- Packed red cells should be preferred to whole blood
- Increase the theatre ambient temperature
- Prewarm irrigation fluids
- Regional block is preferable to general anaesthesia

Treatment of TURP syndrome

- Terminate surgical procedure as soon as diagnosis of TURP syndrome is made
- Administer intravenous frusemide 1 mg/kg
- Give 15% mannitol
- Administer oxygen by nasal cannula
- Pulmonary oedema: Tracheal intubation and IPPV with 100% oxygen
- Monitor the arterial blood gases, Hb concentration and sodium
- Administer hypertonic saline: 0.5 mEq/hour
- Administer intravenous calcium for acute cardiac disturbances
- Seizures: Manage with diazepam/ imidazolam/barbiturates
- Replace blood loss with packed cells
- Disseminated intravascular coagulation (DIC): Administer fibrinogen followed by heparin. Fresh frozen plasma and platelets may be administered depending on the coagulation profile
- Perforation: Surgical drainage

Complications of prostatectomy

- Haemorrhage which may result in clot retention: Aetiological factors in post-prostatectomy haemorrhage are incomplete haemostasis and bladder spasm. These cannot be completely avoided even with improved surgical skills. They are best prevented by ensuring adequate haemostasis at surgery and by judicious bladder lavage after surgery with normal saline. Studies have shown that perioperative administration of finasteride helps to reduce the degree of both Intraoperative and postoperative blood losses.

- **Infection:** Wound infection and urinary tract infection. The latter is likely to respond to antibiotics as dictated by the preoperative urinary assessment.
- **Retrograde ejaculation**
- **Impotence**
- **Epididymo-orchitis**
- **Urinary incontinence**
- **Damage to the ureter(s)**

CHAPTER THIRTY-ONE

PROSTATE CANCER

Prostate cancer is the commonest cancer in males. It is a disease of old age and peaks at the 70–90 age groups. Quite a good percentage of this lesion is discovered at post-mortem.

Pathology:

Prostate cancers involve the peripheral glands and so can still occur after prostatectomy for benign prostatic hyperplasia. They are mainly adenocarcinoma.

Spread: This involves direct

- Spread to the adjoining structures
- Lymphatic spread
- Haematogenous spread to distant organs such as the liver, lungs, brain, adrenal and bone. The latter is unique in that it results in osteoblastic secondaries in the lower spine.

An important point worthy of note is that benign prostatic hypertrophy and prostate malignancy can coexist in the same patient.

Staging (AJCC)

Stage I: Clinically inapparent tumour; no nodes, no metastasis

Stage II: Tumour within prostate; no nodes, no metastasis

Stage III: Tumour through prostate capsule; no nodes, no metastasis

Stage IV: Tumour extends into adjacent structures or presence of nodes/metastasis

Clinical presentation

Owing to the peripheral disposition of the lesion anatomically, presentation is rather late. It is usually asymptomatic. Attention may only be drawn to its presence following the palpation of a prostatic nodule in the course of a digital rectal examination for an unrelated condition. It eventually manifests with clinical features of bladder outlet obstruction as outlined previously. In addition, there is associated loss of weight and low back pain due to involvement of the spinal vertebra. Sequel to the latter, the patient may present with paraparesis/ paraplegia.

Examination will reveal a chronically ill-looking patient with evidence of loss of weight. Patient may be pale and jaundiced (hepatic spread). There may be evidence of metastatic spread to distal organs when presentation is late. digital rectal examination (DRE) may act as the 'whistle blower'. The following is a comparison of the DRE findings in benign and malignant prostatic enlargements

Criteria	Benign	Malignant
Nature	Smooth	Craggy
Consistency	Firm	Hard
Nodularity	Non-nodular	Nodular
Rectal mucosa	Moves freely over it	Does not move freely over

Midline sulcus	Preserved	May be obliterated
In addition, prostate cancer may present with a palpable Blumer's shelf on rectal examination. It is pertinent to point out that benign prostatic hyperplasia is, strictly speaking, a histological rather than a clinical diagnosis		

Investigations

- Prostate-Specific Antigen (PSA): See below
- Transrectal prostatic biopsy: This provides the specimen for histological confirmation and staging. Indications are
 - Palpable prostatic nodule: Most important indication
 - PSA elevation in the face of an apparently normal prostate on digital rectal examination.

Guided by ultrasound and under local anaesthesia, multiple biopsies (at least six) are taken from the prostate for histology. The Prostate Cancer Gleason Score is based on the degree of disruption of the glandular architecture. It is the sum of two independent scores (majority and minority) with each ranging between 1 and 5. The total score ranges between 2 and 10. If only one pattern is seen, it is counted twice. A high Gleason score such as 10 is indicative of a high grade tumour. A rapid growth and spread is more likely in high grade tumours than in the low grade variety.

Grading of Prostate Cancer Gleason Score

Prostate cancer: Description of biopsy result

Gleason Score 1: Cancerous cells look very much like normal prostate cells

Gleason Score 2-4: Pattern of cells in these grades varies. Some cells do look like normal prostatic cells, others do not

Gleason score 5: Abnormal cells which do not look like normal prostate cells and appear to scatter haphazardly throughout the prostate

The Pathologist grades the two most common patterns. The overall Gleason score is the sum of both assessments. If only one pattern is seen, it is counted twice.

- Magnetic resonance imaging: Detects local infiltration and lymphatic spread
- Bone scintigraphy with isotope labelled compound: Demonstrates bony metastasis
- Skeletal survey: Detects sclerotic metastasis to the spine
- Transrectal USS: Assesses the depth of the lesion
- Chest X-ray: Detects metastases to the lungs and pleura
- Abdominal USS: Determines intraperitoneal spread
- Others: FBC, electrolytes/urea/creatinine, and urinalysis/mcs.

Prostatic Specific Antigen (PSA)

- This blood enzyme is produced by the prostatic cells no matter their location. The clinical implication is that both normal and malignant prostate cells produce PSA even if they are outside the prostate. Normal level of PSA in the blood ranges between 0 – 2.5ng/ml.

- PSA is a protein-bound enzyme. It is a glycoprotein produced by the prostate cells and detectable in the serum. Levels of 10 and above are highly suggestive of prostate cancer. It should be noted, however, that PSA levels should be interpreted with caution as they are neither specific nor sensitive enough for the definitive diagnosis of prostatic cancer. High levels of PSA may be found in prostatitis, following urinary retention and even following a digital rectal examination. PSA level may also be directly related to the size of the prostate gland; hence the PSA level may be elevated in large benign prostates. PSA assay has replaced acid phosphatase estimation as the diagnostic tool of choice in the management of prostatic cancer.
- Most of the circulating enzyme is bound to proteins in the blood. The remaining unattached PSA is referred to as 'Free PSA'. It has been found that the percentage of free PSA has an inverse relationship to the degree of risk of prostatic cancer. In other words, men with a lower percentage of free PSA have a higher risk for prostate cancer. A high percentage level of free PSA is therefore encouraging. Free PSA estimation has been found useful in taking management decisions when assessment results are on borderline. A typical example is when the PSA level is only slightly elevated in the presence of a negative prostatic biopsy result. In this case, a repeat biopsy is indicated much sooner in the presence of a lower free PSA level as opposed to when the level is high.
- PSA is meant to complement digital rectal examination (DRE) as a screening tool for prostatic cancer. Occasionally, there may be an elevated PSA level despite a normal DRE finding and vice-versa. Consequently, none of the findings should be taken in isolation. PSA estimation has been found useful in the diagnosis, treatment and follow-up in the management of prostate cancer. It equally plays an important role in the comparison of results from various centers

Treatment of prostate cancer

Like most cancers, this is dependent on whether the lesion is early (confined to the prostate) or late (metastatic disease). Curative treatment is the aim in the former, whereas palliation is the goal in the treatment of the latter.

Early cancer: Treatment is by surgery or radiotherapy

- Radical prostatectomy: may be by either open or laparoscopic surgery. Complications include impotence and incontinence
- Radiotherapy: May be by either external beam or implantation of radioactive seeds (brachytherapy)

Late cancer: Treatment is aimed at suppressing the production of endogenous testosterone. This may be achieved by the use of luteinising hormone releasing hormone agonists such as goserelin. Surgical castration by way of bilateral orchidectomy is usually the last resort. Ultimately, most relapse owing to 'hormonal escape'. This is an end-game phenomenon from which the patient rarely survives. Chemotherapy with docetaxel is rarely beneficial.

CHAPTER THIRTY-TWO

URETHRAL STRICTURE

Urethral stricture is the narrowing of the urethra following a process of healing by fibrosis. It is a common cause of infravesical obstruction.

Aetiology of urethral stricture

- Congenital
- Post-urethritis: The commonest is post-gonococcal urethritis
 - Post-gonococcal urethritis: This results in periurethritis and affects mainly the bulbar urethra. The resultant inflammation heals by fibrosis. Urethral narrowing occurs within one year of the healing process but it may take about 10 to 15 years for voiding challenges to manifest
 - Tuberculous urethritis
- Trauma: Urethral injuries due to road traffic accident and falling- astride injuries
- Post-instrumentation: Urethral catheterisation and cystoscopy. Contributory factors include trauma, infection and pressure necrosis
- Postoperative: Prostatectomy (open and endoscopic), amputation of the penis

Clinical features: Urethral stricture presents with clinical features of bladder outlet obstruction as detailed earlier. The difference in symptomatology between urethral stricture and prostatic hypertrophy is that straining improves the urinary stream in urethral stricture but has no effect on its prostatic counterpart. Late presentation may be with clinical features of complications

Complications of urethral stricture

- Retention of urine: Acute, chronic
- Urinary tract infection
- Bladder: Stones, diverticular formation
- Periurethral abscess, periurethral fistula, 'watery-can' perineum
- Hydroureters, hydronephrosis, renal failure
- Complications of straining at micturition: Hernias, haemorrhoids, rectal prolapse

Investigations

- Retrograde cystourethrography
- Micturating cystourethrography
- Serum electrolytes, urea and creatinine
- Urine: microscopy, culture and sensitivity
- Abdominal ultrasound scan
- Peak urine flow rate



RETROGRADE URETHROCYSTOGRAPHY SHOWING URETHRAL STRicture

- Urethroscopy
- Post-void residual urine
- Full blood count

Treatment:

Particularly in a complicated stricture with retention of urine, initial treatment will involve relief of obstruction. Urethral catheterisation may be attempted by the use of an introducer. This must be done carefully with a moderately-sized urinary catheter and introducer with prior liberal lubrication of the urethra. Suprapubic cystostomy is a more predictable procedure. The following are the various modalities of treatment

- Bouginage (Intermittent dilatation): Instruments employed include gum elastic bougies, filiform bougies and metal sounds. Intermittent self-bouginage is now feasible with the use of Nelaton catheter urethral dilator. Bouginage is usually a regular affair in order to prevent restructuring. Initially dilatation is carried out once a week for one month; then once a month for one year. It is then carried out six-monthly for three years and

thereafter once a year. This is in keeping with the age-long notion that 'bouginage can only ameliorate but rarely cures a stricture'. Complications of bouginage include bleeding, infection, creation of false passage, fistula formation and re-stricture.

- Internal visual urethrotomy: The fibrosed stricture is 'slit' preferably at 12 O'clock under vision. The urethral diameter is maintained with the aid of an indwelling catheter. The latter is removed after two days of urinary drainage. Additional slits, if necessary, may be employed in addition to the primary one at the 12 O'clock position
- Surgical excision and end-to-end anastomosis: This applies to the treatment of short segment strictures
- Urethroplasty: This is a staged procedure and entails excision of the stricture and reconstruction of the urethra. Indications for urethroplasty are
 - Failure of the aforementioned conservative treatments (urethral dilatation and internal visual urethrotomy)
 - Strictures with extensive spongiositis
 - Very long/complete strictures
 - Complicated strictures (periurethral fistula/abscess and calculi)

CHAPTER THIRTY-THREE

MALDESCENDED TESTIS AND PRUNE-BELLY SYNDROME

The testis develops from the genital ridge of the mesonephros in the retroperitoneal region of the abdomen between the 4th and 6th weeks of gestation while the epididymis and vas deferens develop from the mesonephric duct. The testis eventually descends into the scrotum traversing the abdomen and the inguinal canal. By the seventh and ninth months of intrauterine life, it has descended into the deep inguinal and superficial inguinal rings respectively. At the end of the ninth month, it descends into the scrotum. It is preceded in its descent by a fold of peritoneum called the processus vaginalis. Any arrest or deviation of the testis along its journey to its ultimate natural destination (the scrotum) will result in the testis being absent in the scrotum. It is important to appreciate the following terminologies in relation to evaluation of the absence of the testis in the scrotum during a clinical examination:

1 **Undescended testis:** There is an arrest of the movement of the testis along its natural pathway of descent. It occurs in about 3% of full-term male newborns. The annual incidence is 1% of all newborn males. The arrest of testicular descent may occur within the abdominal cavity (abdominal testis) or within the inguinal canal (inguinal testis). About 90% of the latter have associated congenital indirect inguinal hernia. Factors responsible for maledescent of the testis include

- Short vas deferens
- Adhesions: Retroperitoneal and at the level of the deep inguinal ring
- Short testicular vessels
- Inefficient pull by the gubernaculum testis: This fibrous tissue is attached to the lower pole of the testis
- Intrinsic maldevelopment of the testis
- Inadequate hormonal stimulation: Androgens have been found to play a positive role in the descent of the testis. This is corroborated clinically by the response of some patients to hormonal stimulation
- Bilateral cryptorchidism is an integral component of prune belly syndrome

2 **Ectopic testis:** In the course of its descent, the testis may deviate from its natural pathway into an abnormal location outside the scrotum. The most common locations of an ectopic testis are

- Superficial inguinal pouch (lies between the fascia of Scarpa and the external oblique aponeurosis)
- The perineum
- Femoral triangle
- Opposite hemiscrotum
- Root of the penis at the suprapubic region

Lockwood's theory of ectopic testis states that the gubernaculum testis may be endowed with various tails that point respectively to the areas of possible descent (superficial

inguinal, scrotal, perineal, femoral and pubic). The natural pull should be towards the scrotum. An unusual pull of the strand related to an abnormal site, however, will result in an ectopic location of the testis. Deviation may also be due to differential growth of the tail related to a particular ectopic location.

It is pertinent to point out that in both the undescended and ectopic testis the scrotum is under-developed since it has never housed a testis

- **Retractile testes:** This occurs in a naturally well-descended scrotal testis which occasionally retracts from the scrotum due to an exaggerated cremasteric reflex. The latter can occur when the child is cold or frightened. The distinguishing feature of retractile testis is that the scrotum is well-developed since the abnormal relocation of the testis is only temporary and short-lived. It can also be manipulated back to the scrotum and maintains this position after the exercise

Complications of undescended testis

- Subfertility or infertility: The scrotal temperature is lower than the normal body temperature. This creates a favourable environment for spermatogenesis. The higher temperature subsisting at any location outside this natural position will result in subfertility
- Malignancy: An undescended testis is more prone to malignancy than its scrotal counterpart. It is estimated that about 10% of testicular tumours occur in undescended testis. The risk is about 1 in 80 in inguinal testis and 1 in 20 in abdominal testis. Seminoma is the most common tumour that occurs in an undescended testis. Embryonal cell carcinoma (choriocarcinoma and teratocarcinoma) may also develop
- Testicular atrophy
- Trauma: The abnormal location renders it more prone to repeated trauma
- Torsion of the testis: This is due to its long mesorchium
- Associated with a congenital inguinal hernia
- Psychology: Undescended testis adversely affects the psychology of a growing child particularly amongst his peers

Clinical features

The patient is usually a baby boy brought by the parents with the chief complaint of absence of the testis in one and occasionally in both hemiscrotum. Presentation may, however, be in late childhood and occasionally in adulthood. An essential part of the history is to enquire from the mother if the testis was ever present in the scrotum at any moment after birth. A positive reply is suggestive of a retractile testis. On the other hand, congenital absence of the testes from birth connotes either an undescended or an ectopic testis. Other key points in the history include

- Enquiry as to the feeling of any swelling along the natural line of descent: Right iliac fossa and inguinal canal
- Location of any swelling at the common sites of ectopic testis: suprapubic, superficial inguinal pouch, femoral triangle, perineum and the contralateral hemiscrotum
- Pain over the swelling which may be a tell-tale evidence of repeated trauma or torsion

- Ask for symptoms of an associated inguinal hernia: Groin swelling that is made more obvious by straining and less so by lying down
- In older patients and bilateral pathology, enquire about infertility and features of secondary male characteristics

On clinical examination

- Compare the size of the ipsilateral hemiscrotum with the contralateral one. It is normal and well-developed in retractile testis but smaller and undeveloped in both undescended and ectopic testes. If the pathology is bilateral, both hemiscrotum will be underdeveloped
- Palpate the scrotum to confirm absence of the testis
- Palpate the groin along the path of the inguinal canal with both hands working in synergy. While one hand is trying to 'milk' down any felt or occult inguinal swelling towards the direction of the superficial inguinal ring, the other is positioned in such a way as to 'grab' such detected lump. It is possible to drag the latter down to the bottom of the scrotum and maintain this position in retractile testis. This is not possible in undescended testis.
- Examine the common locations of ectopic testis for a palpable swelling
- Examine the groin for an associated inguinal protrusion: Apply pressure over the child's abdomen so as to elicit the presence of an inguinal protrusion. If one is lucky and the child either coughs or cries in the course of the clinical examination, the inguinal swelling will become more obvious. This is in keeping with an associated congenital inguinal hernia
- Examine for other associated congenital anomalies

Management of undescended testis

Owing to the above-mentioned inherent complications of undescended testis, the earlier it is located and brought down to its natural location in the scrotum, the better. Location of the testis can be achieved by means of the following

- Clinical examination: Relevant only in the case of an inguinally-located testis
- Abdominal ultrasound scan of the groin, pelvis and retroperitoneum: The main limitation of USS is that it is difficult to locate an abdominal testis with the aid of this test.
- Diagnostic laparoscopy: This is now the investigation of choice particularly in the diagnosis of an abdominally-located testis. The location of the vas deferens with reference to the deep ring is the key element in the diagnostic laparoscopy of undescended testis. If the cord structures are seen to emerge from the deep ring, the testis is inguinal. On the other hand, if it is not visualised in this situation, then the testis is most probably located in the abdomen.
- Other investigations to prepare patient for surgery include baseline investigations such as full blood count and urinalysis

Timing of surgery: It is believed that the earlier the surgical intervention, the less the chances of developing complications associated with undescended testis. An undescended testis in the newborn may be expected to descend into the scrotum spontaneously by the age of one year. The patient can therefore be followed up during this period. Undescended testis presenting after infancy therefore needs to be surgically treated. It is equally desirable to carry out surgery before the child commences schooling in order to prevent the psychological embarrassment he may experience at school. Surgical treatment will

- Reduce the risk of torsion and trauma
- Enhance spermatogenesis
- Fix the associated inguinal hernia
- Reduce the chances of neoplastic change. It equally enhances the diagnosis should this occur
- Enhance the quality of life owing to the psychological satisfaction of having both testes in the scrotum. This will grant the patient a 'manly feeling'

Surgery for undescended testis:

This is referred to as orchidopexy and is carried out through an inguinal approach. The inguinal testis is located and the cord structures are identified and freed in order to create enough length for delivery of the testis into the scrotum. The volume of the scrotum is enhanced by inserting the index finger into it with simultaneous stretching of the scrotal wall.

The cord structures may be short and require to be lengthened for unhindered delivery into the scrotum. This can be achieved at the level of the deep ring by the following techniques

- Medial expansion of the deep inguinal ring: The inferior epigastric vessels may need to be sacrificed and the transversalis fascia incised medially. An adequate cord length may be achieved by high dissection of the cord into the retroperitoneum
- Fowler Stephen's procedure: Division of the testicular artery in order to lengthen the cord. There is the firm belief that the testis will have alternative blood supply from the cremasteric artery and the artery to the vas deferens.
- Silbar procedure: Involves microvascular anastomosis – the testicular artery to the inferior epigastric artery and the testicular vein to the inferior epigastric vein
- Occasionally, orchidopexy may be carried out in two stages in order to achieve the set objective.

The testis is eventually relocated into a sub-dartos pouch. This is the standard procedure. Other points of relocation include the contralateral scrotum (Ombredann's technique) and temporarily at the ipsilateral thigh (Keetley Torek technique).

Finally, it is important to identify the sac of an associated congenital inguinal hernia with a view to carrying out a formal inguinal herniotomy

PRUNE BELLY SYNDROME

Prune belly syndrome (also called Eagle-Barrett syndrome) is a rare congenital disorder that is characterised by

- Partial or complete absence of muscles of the anterior abdominal wall
- Failure of both testes to descend into the scrotum (bilateral cryptorchidism)

- Urinary tract malformations such as megaureter, hydronephrosis and vesicoureteric reflux.

In addition, it is associated with a high incidence of congenital cardiac anomalies such as tetralogy of Fallot and ventricular septal defects

Prune belly syndrome is associated with trisomy 18 and 21 and has an incidence of 1 in 40,000 births. It is thought to be due to a congenital developmental arrest

Surgical treatment is multidisciplinary and involves

- Reconstruction of the anterior abdominal wall
- Bilateral orchidopexy
- Addressing the relevant urological issues



PRUNE BELLY SYNDROME: Note the defect in the anterior abdominal wall and the undeveloped scrotum

CHAPTER THIRTY-FOUR

GENERAL PRINCIPLES OF MANAGEMENT OF TRAUMA

Management of trauma is based on the principle of the Advanced Trauma Life Support (ATLS) as enunciated by the American College of Surgeons. Essentially, there are three stages of care of the injured patient

- Primary survey
- Secondary survey
- Definitive care

Primary survey

This is carried out at the scene of the injury. It is aimed at ensuring that the patient arrives at the hospital or trauma centre alive. It identifies and addresses any life-threatening problems discovered during the initial assessment. The action taken during this phase is generally referred to as the ABCDE of resuscitation; each alphabet denoting the emergency procedure to be carried out at this stage.

- A: Airway (and C-spine stabilisation)
- B: Breathing
- C: Circulation
- D: Disability
- E: Exposure and environment

Airway: Secure the airway and stabilise the spine in general and the cervical spine in particular. The latter can be achieved by gentle handling of the spine with the aid of a rigid support and the application of a rigid cervical collar to curtail movement of the neck. The airway is assumed to be satisfactory if the patient can vocalise. The following are useful in the maintenance of a clear airway.

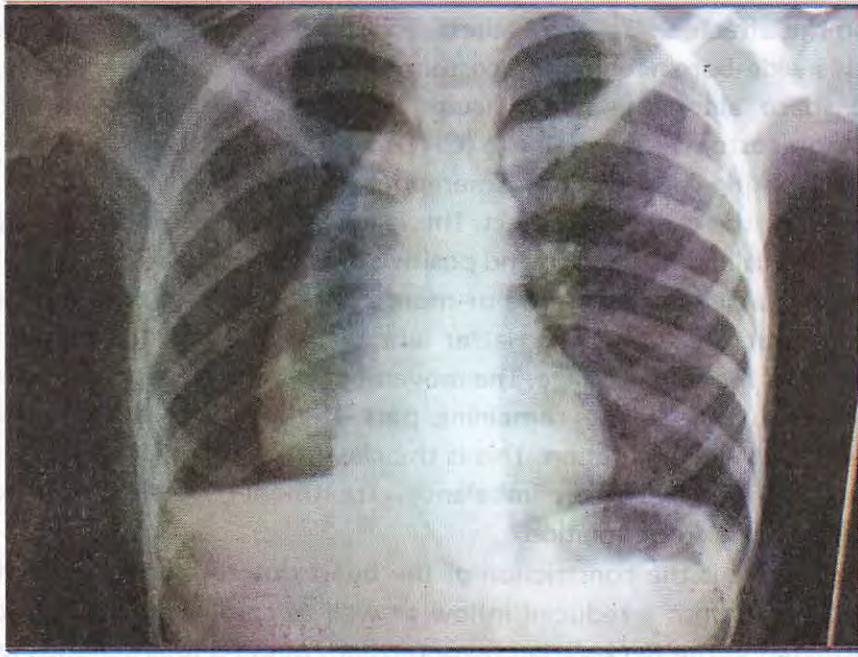
- Insert a finger or sucker to clear the airway of any obstructing agent
- Chin lift, jaw thrust manoeuvre to prevent the tongue from falling back and obstructing the airway
- Insertion of an oropharyngeal airway such as the Guedel variety to secure the tongue. A nasopharyngeal airway may be preferable in patients with massive oral trauma and associated maxillary fracture with its attendant severe bleeding into the mouth
- Endotracheal intubation : In deeply unconscious patients with absence of the gag reflex
- Surgical cricothyroidotomy: This could be a last resort when other methods of restoring the airway fail. Needle cricothyroidotomy is more appropriate in children
- Emergency tracheostomy may prove useful

Breathing: This is achieved by ensuring adequate ventilation and oxygenation. It is aimed at identifying and addressing such life-threatening injuries like pneumothorax of various types, haemothorax, flail chest and cardiac tamponade.

- Tension pneumothorax: Characterised by increasing dyspnoea and tachypnoea, distension of the jugular veins, ipsilateral decrease or absence of breath sounds and

hyper-resonance on the affected side of the chest. This condition requires immediate decompression with a wide-bore needle thoracostomy. This is inserted in the ipsilateral second intercostal space along the midclavicular line. Definitive treatment is by insertion of closed thoracostomy tube drainage (CTTD)

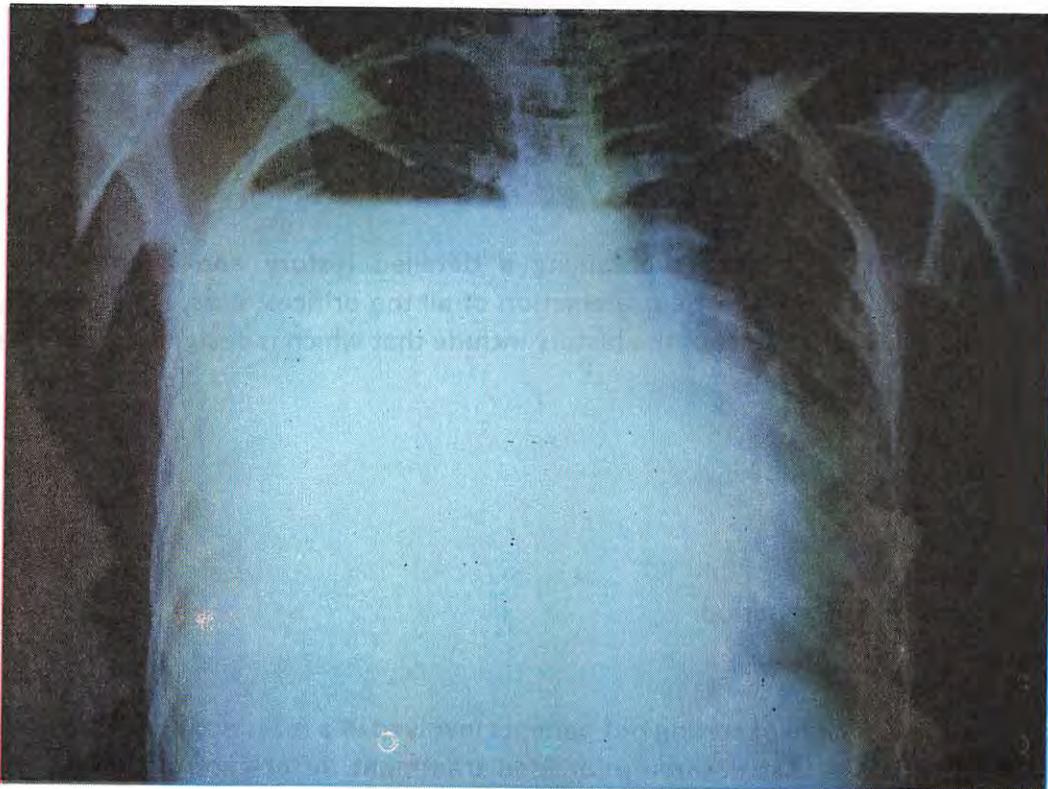
- Open pneumothorax (sucking chest wound): Emergency treatment is by application of an occlusive dressing over the chest wall defect. This is followed by insertion of a chest tube. This condition may call for intubation and positive pressure ventilation
- Flail chest: This involves a segment of three or more consecutive ribs with each rib having two separate fracture sites. The latter are so aligned as to create an 'independent' segment of the thoracic cage. The movement of this flail segment during respiration is opposite to that of the remaining part of the chest: sucks in with inspiration and pushes out with expiration. This is the classical paradoxical respiration which results in a ventilation/perfusion imbalance. Treatment is by endotracheal intubation with positive pressure ventilation
- Pericardial tamponade: This is the constriction of the heart due to bleeding into the pericardial sac. It results in both a reduced inflow as well as cardiac output. Clinical features are encapsulated in the Beck's triad (hypotension, muffled heart sounds and jugular venous distension) and the Kussmaul's sign (jugular venous distension with inspiration). Diagnosis may be confirmed by echocardiography. Emergency treatment is by ultrasound-guided needle pericardiocentesis
- Haemothorax: This is bleeding into the pleural space. Clinical signs are diminished breath sounds, dullness to percussion over the ipsilateral chest wall and hypotension due to diminished blood volume. Diagnosis is confirmed by chest X-ray. Treatment is by intravenous infusion (and blood transfusion if necessary) and tube thoracostomy. Emergency thoracotomy is, however, indicated if the initial drainage via the chest tube is 1500 mls or more and the hourly drainage is recorded to be up to 200 mls in four consecutive hours



CHEST X-RAY OF A PATIENT WITH HAEMOTHORAX OF THE RIGHT HEMITHORAX
NOTE THE AIR-FLUID LEVEL

Circulation: Aims at controlling external haemorrhage and ensuring adequate perfusion.

- Control of external haemorrhage: This is initiated by the application of pressure. In severe haemorrhage affecting the limbs, however, a tourniquet may be applied with adequate precautionary measures taken to prevent complications. These include ensuring that it only overcomes the diastolic pressure and hence ensuring some degree of arterial inflow and tissue perfusion. Other precautionary measures are proper recording of the time of application and rapid movement of the patient to ensure that the period of application is not unduly prolonged.
- Treatment of hypovolemic shock: This is initiated by the administration of intravenous fluids while awaiting grouping and cross-matching of blood for patients with severe blood loss. Crystalloids (normal saline) and Ringer's lactate are the infusions of choice. The latter is preferred for its lactate component which helps to buffer the metabolic acidosis associated with hypovolemic shock. One school of thought discourages the overenthusiastic administration of intravenous infusion in cases of ongoing haemorrhage in a bid to sustain a normal blood pressure. Rather, it is thought that a permissible controlled systolic blood pressure of between 70 and 85 mm Hg is preferable. In addition to preventing the recurrence of the bleeding episode, it



HAEMOPNEUMOTHORAX OF THE RIGHT CHEST

encourages the sustenance of the natural homeostatic mechanisms. The required volume of intravenous infusion depends on the estimated blood loss as well as the type of proposed infusion. The "3-for-1" rule states that the trauma patient in hypovolemic shock requires 3 litres of crystalloid for every litre of blood loss. On the other hand, replacement with colloid is calculated on a 1-for-1 basis. Resuscitation is monitored by way of the urinary output and central venous pressure.

Disability: This involves the determination of

- Mental status: By way of the Glasgow Coma Score (see below). If patient is intubated, however, the verbal evaluation is omitted, overall score becomes 11 and V is replaced by T.
- State of the pupils: Pupils are monitored in order to detect deteriorating head injury

Exposure and environment

- Exposure: Remove all clothing in order to ensure examination of the whole body for early detection and treatment of associated injuries
- Environment: Ensure the provision of a relatively warm environment as hypothermia may predispose to coagulopathy

In addition to the initial resuscitation, some relevant information depicted by the acronym MIST should be obtained as part of the primary survey

MIST

- M: Mechanism of injury
- I: Injury sustained
- S : Signs and symptoms (Prehospital)
- T: Treatment so far

SECONDARY SURVEY: This entails obtaining a detailed history and a complete physical examination. The latter includes the examination of all the orifices: ears, nose, mouth, vagina, and rectum. The relevant aspects of the history include that which is designated by the acronym AMPLE

AMPLE

A: Allergy

M: Medication(s)

P: Past medical history

L: Last meal/Last menstrual period

E: Events (of injury etc)

TRIAGE: This is the principle of sorting out patients involved in a mass accident/disaster with the aim of identifying those that deserve prioritised treatment. After careful clinical assessment based on the information given by the paramedics and the ABCDE assessment, the patients are sorted out into the following categories

- P1 : Life threatening
- P2 : Urgent
- P3 : Minor
- P4 : Dead

It is only rational to give priority to the management of the patients in the P1 and P2 categories. This aspect of triage management should be carried out by the most experienced member of the trauma team.

CHAPTER THIRTY-FIVE

ABDOMINAL TRAUMA AND ABDOMINAL COMPARTMENT SYNDROME

The abdomen is a common site of trauma and could be the primary target of injury. This is the scenario in penetrating injuries caused by a stab or a gunshot. On the other hand, it may be involved as part of trauma in multiple injured patients.

Aetiology of abdominal injuries

- Blunt injuries: Commonest cause is road traffic accidents. Others are falls from heights, blows and compressive injuries
- Penetrating injuries: Stab and gunshot injuries

Management of abdominal trauma

Initial management at the scene of trauma is along the Advanced Trauma Life Support (ATLS) line of primary survey. The ABCDE of management is instituted prior to the movement of the patient to the hospital. Secondary survey is aimed at obtaining a full history as to the mechanism of injury. A full clinical examination is carried out in order to identify other associated injuries. In addition, one should note the following:

- A chest injury at or below the nipple line may have an abdominal component
- Seatbelt sign of abdominal injury: Ecchymosis on lower abdomen from wearing a seatbelt is associated with small bowel and pancreatic injuries in about 10% of cases
- Rectal examination: A reduced sphincteric tone may connote associated spinal injury while presence of blood in the rectum is in keeping with a colonic or rectal injury. A high-riding, ballotable prostate is a pointer to urethral trauma.

Life-threatening injuries are addressed. In the hospital, resuscitation is continued with fluid resuscitation. An indwelling urinary catheter aids in the monitoring of the urinary output which acts as a guide to the degree of cardiovascular resuscitation.

The following investigations are carried out

- Full blood count: The PCV may be normal but a baseline is established
- Serum electrolytes, urea and creatinine
- Group and crossmatch blood
- Trauma X-ray series: Neck (A-P and lateral views), chest (A-P) and pelvis (AP). These may reveal associated injuries. In particular, fracture of the lower ribs may suggest a thoraco-abdominal injury
- Plain abdominal X-rays: Supine film is considered more useful than the erect
- Focused Assessment using Sonography in Trauma (FAST): This is useful in the detection of intraperitoneal as well as pericardial haemorrhage. Ultrasonography is fast and non-invasive. It is, however, operator dependent and is unhelpful in the evaluation of injury to solid viscera.
- CT scan: Now regarded as the gold standard. In addition to the detection of intraperitoneal haemorrhage, it is able to detect retroperitoneal bleeding. It is also

organ specific. The disadvantages include the high cost and the fact that it is best carried out on stable patients. Serial scanning is useful in the progressive evaluation of an abdominal injury.

- Four quadrant tap: As 'crude' as it seems, this simple test is still useful especially when more sophisticated investigative aids are not readily available. It may act as a rapid screening aid in the diagnosis of intraperitoneal haemorrhage
- Diagnostic peritoneal lavage (DPL): Has been virtually replaced by FAST in the developed world. Again, like the four quadrant tap, it should not be completely discarded. It is still very relevant in resource-poor situations. It is aimed at diagnosing injury to the abdominal viscera. Positive criteria include aspiration of more than 10mls of frank, non-clotting blood, red blood cell count $> 100,000/\text{dl}$ and WBC $> 500/\text{dl}$. Other pointers to visceral injury include the aspiration of faeces, bile and vegetable matter. It is important to pass a nasogastric tube and a urethral catheter prior to diagnostic peritoneal lavage in order to prevent injuries to the stomach and the bladder respectively. DPL does not detect retroperitoneal haematoma. It has 97% positivity. The advantages of DPL include its quick and inexpensive nature. The down side is that it is invasive, too sensitive and has limited specificity
- Laparoscopy: Of limited use due to cost and problems associated with pneumoperitoneum and anaesthesia in an unstable patient. It is, however, useful in determining the degree of peritoneal involvement in penetrating injuries. It is also useful in the detection of peridiaphragmatic injuries.
- Exploratory laparotomy: it is both diagnostic and therapeutic. It should, however, be handled with caution when employed for diagnosis as a morbidity of 5 to 22% has been associated with negative laparotomies. It has been posited that the more the uncertainty in the clinical assessment and the more inexperienced the assessor, the more aggressive the tendency towards laparotomy.

Other tests that may be found useful in special situations include

- Transurethral cystography: Useful when rupture of the bladder is suspected. About 400 to 500 mls of contrast is introduced into the bladder in order to ensure adequate distension
- Intravenous urography: Useful in suspected renal injury
- Contrast studies: Useful in isolated organ injury

Treatment of abdominal trauma

While resuscitation is ongoing, plans should be made for the definitive management of specific organ injuries. It had been the norm to surgically explore the abdomen on confirmation of injury to an abdominal viscus. This has, however, resulted in undue surgery (unnecessary laparotomy) in cases that could have been successfully managed conservatively. The advent of sophisticated monitoring gadgets such as USS and CT scan has contributed to the actualisation of conservative management of blunt abdominal trauma. The importance of close clinical monitoring during this mode of treatment cannot be overemphasised. Patients on selective non-operative management who, however, show no evidence of improvement within a specified period both clinically and

radiologically should be subjected to exploratory laparotomy. A delay in aggressive fluid management until rapid operative intervention has occurred has been found to improve the prognosis in patients with penetrating thoracoabdominal injuries.

Essential elements of a trauma laparotomy

Trauma laparotomy should be carried out in a well-equipped operating theatre. The initial priority of treatment should be to*

- Stop bleeding
- Limit contamination

The essential elements of trauma laparotomy include the following

- Large, warm and ambient operating environment
- Wide area of skin preparation: From nipple line to mid-thigh and to table laterally
- Generous incision for excellent exposure: preferably from xiphisternum to pubic symphysis
- Generous packing particularly around the edges of the four quadrants of the abdomen. This will be useful both for the tamponade effect as well as in the identification and control of ongoing bleeding
- Control of bleeding should be regarded as a priority. The most common sources of bleeding in blunt abdominal trauma are the liver, spleen and mesentery. Penetrating injuries may, in addition to the latter structures, also involve vascular and retroperitoneal structures. Initial packing of a bleeding site helps both in the identification as well as the management of bleeding vessels.
- Identification of injuries: Carried out methodically by gradual removal of packs quadrant by quadrant
- Control of contamination especially by injured gut: The entire bowel is examined beginning from the ligament of Treitz to the rectum. Both the mesenteric and antimesenteric surfaces of intestine are examined. Any leakage from the affected bowel is clamped to prevent further soiling of the peritoneum
- Repair/reconstruction of affected viscera as deemed appropriate

Management of specific organ injuries

- Liver: The liver is the most commonly injured organ in blunt abdominal trauma and the second most commonly injured in penetrating abdominal trauma. Its proneness to injury is due to its large size as well as its relatively fixed position. The posterior portion of the right lobe is the most common site of hepatic injury in blunt trauma.

Grading of liver injuries

Grade I:

- Nonexpanding subcapsular haematoma, < 10% of the surface area;
- Capsular tear, nonbleeding, < 1cm in depth

Grade II: ss

- Nonexpanding haematoma, subcapsular or intraparenchymal, 10 to 15% of surface area or < 10cm in diameter
- Bleeding capsular tear
- Laceration 1 to 3 cm in depth, < 10 cm in length

Grade III

- Subcapsular haematoma, > 50% of surface area expanding or ruptured with bleeding
- Intraparenchymal haematoma > 10 cm or expanding
- Laceration > 3 cm deep

Grade IV

- Ruptured intraparenchymal haematoma with bleeding
- Parenchymal disruption involving 25 to 75% of lobe or 1 to 3 segments

Grade V

- Parenchymal disruption of >75% of lobe or more than 3 segments
- Juxtahepatic venous injury

Grade VI

- Hepatic avulsion (rare)

Owing to its generous vascularity, bleeding is of primary concern. Equally important is the anatomical disposition as there may be associated chest injury. It has been shown, however, that about 85% of liver injuries would have stopped to bleed by the time of trauma laparotomy. For this reason, the selective non-operative management (NOM) approach has found a prominent place in the management of liver injuries. A good majority of the patients have grades I, II and III injuries and are successfully treated by NOM. Similarly, about 1/3rd of patients with grades IV and V injuries benefit from NOM. The following are critical in NOM

- Patient must be haemodynamically stable
- Absence of other internal injuries including moderate to severe head injury
- Centre should have facilities for precise diagnosis of the severity of liver injuries and capable of intensive care management
- Centre should have facilities for immediate access to diagnostic CT scan (with intravenous contrast) and interventional radiology (angiography with embolisation)
- Unlimited access to blood for transfusion

Complications of NOM include bleeding, abdominal compartment syndrome, and infections (abscesses). Others are biliary complications (bile leak, haemobilia, biliary peritonitis) and hepatic necrosis. Liver compartment syndrome may follow a large subcapsular haematoma. Decompression is achieved by either laparotomy or laparoscopy

The main candidate for operative treatment is the haemodynamically unstable patient. By the ATLS definition, this unstable patient has a systolic blood pressure < 90 mm Hg, pulse rate > 120 bpm, and evidence of skin vasoconstriction (cold, clammy extremities, and diminished capillary refill). Others are shortness of breath and/or decreased level of consciousness. Patients with associated injuries (including head injury) and those who have failed to respond positively to NOM should have operative management.

Control of haemorrhage in liver injury can be achieved by the following methods

- Manual compression or packing
- Ligation of vessels in the wound
- Application of topical haemostatic agents: Gelfoam, microfibrillar collagen, Surgicel and fibrin glue
- Electrosurgery including the use of bipolar devices
- Omental packing
- Pringle's manoeuvre (application of a vascular clamp across the porta hepatis with the aim of controlling hepatic haemorrhage)
- Hepatic debridement
- Balloon tamponade
- Hepatic vascular occlusion
- Hepatic resection/lobectomy

Packing of the traumatised liver followed by a 'second-look' operation which was once discarded has been reintroduced ('the pack in back'). The surgical management of liver injuries has been summarised as 4Ps: 'push (apply pressure), Pringle (Pringle's manoeuvre to control haemorrhage), plug (plug any hole in the liver) and pack'

Gallbladder trauma: Contrast enhanced CT scan is the investigation of choice. Other investigations include MRI, MRCP and angiography. Treatment is by cholecystectomy.

Spleen: Modalities of treatment of splenic injuries include application of pressure, splenorrhaphy (repair of injury), partial splenectomy and splenectomy depending on the severity (grade) of the splenic injury. Efforts should be made at splenic conservation in order to reduce the incidence of post-splenectomy complications particularly overwhelming post-splenectomy infection (OPSI). See section on Spleen.

Pancreas: Clinical suspicion is best confirmed with operative cholecystocholangiography. Mobilisation of the pancreas is as outlined below (duodenum). The priorities of treatment are: adequate exposure and location of injury, haemostasis, selective debridement and resection when necessary. Conservative surgery may be carried out in mild to moderate injuries with closed drainage. Distal pancreatectomy may be carried out in lesions involving the tail of the pancreas. Pancreaticoduodenectomy (Whipple's procedure) is rarely called for in severely traumatised patients

Stomach: Usually due to penetrating injury. Treatment is by surgical repair and nasogastric aspiration.

Duodenum: Due to the anatomical relationship, duodenal injuries are usually associated with pancreatic injury. Diagnosis of these injuries requires a high index of suspicion. Clinical diagnosis is complemented by plain abdominal X-rays, CT scan, contrast duodenography and ultimately diagnostic laparotomy. The latter requires adequate mobilisation and exposure. This can be accomplished by a combination of the following manoeuvres:

- Kocher's manoeuvre: Division of the peritoneal lining over the lateral aspect of the duodenum

- Cattel Braasch manoeuvre: Incision along the white line of Todd to mobilise the caecum up to the hepatic flexure
- Aird manoeuvre: Division of the lienorenal and lienophrenic ligaments to deliver the spleen and the tail of the pancreas
- Division of the ligament of Treitz to expose the fourth part of the duodenum.

Mild duodenal injuries may respond to NOM with nasogastric intubation for two weeks. Primary repair is carried out using either single/ double layer suturing or stapling. Repair of more severe injuries will require protection. This can be achieved by combination of a sutured closed pylorotomy and gastrojejunostomy. It is important to leave a drain for external drainage.

Complications of pancreaticoduodenal trauma are

- Haemorrhage,
- Pancreatic fistula
- Duodenal fistula
- Pancreatic pseudocyst
- Diabetes mellitus,
- Intra-abdominal abscess, obstruction and malabsorption.
- Pancreatitis
- Uncontrolled sepsis
- Multisystem organ

Small intestine: Surgical treatment ranges from simple repair to resection and anastomosis. Multiple wounds located in a segment of the small bowel require resection of the affected portion followed by end-to-end anastomosis

Large intestine: Lesions involving the right colon are amenable to simple repair in case of minor injuries. More serious injuries will require a right hemicolectomy. Left colonic injuries may require simple repair or left hemicolectomy as the case may be with or without a protective proximal colostomy

Rectum: Primary repair with a protective proximal colostomy

Kidneys: Most will respond to conservative management. The renal function of the opposite kidney should be assessed by IVU/contrast CT scan. Renal conservation may be achieved by application of pressure and partial nephrectomy. Severe renal injuries will, however, require nephrectomy.

Bladder injuries: May be intraperitoneal or extraperitoneal rupture. Intraperitoneal rupture of the bladder will require laparotomy and repair. Extraperitoneal rupture is usually associated with pelvic injuries and is managed conservatively with urethral catheterisation and bladder drainage.

Penetrating injuries

Consist of stab wounds and gunshot injuries. Management of visceral injuries is as outlined above. There are, however, some peculiarities in the management of penetrating injuries.

- Stab injuries: Initial management is hinged on establishing the penetration or otherwise of the peritoneum. This can be achieved by local wound exploration under local anaesthesia. Those that do not penetrate the peritoneum are managed conservatively with proper wound debridement and dressing. Depending on clinical judgement and radiological assessment, non-operative treatment may be employed in the initial management of those that breach the peritoneum. Surgery is only carried out in obvious severe injuries as well as those that fail to respond to non-operative treatment.
- Gunshot injuries: For a long time, operative treatment was the norm. The negative laparotomy rate was, however, on the high side. Currently, there is a place for non-operative treatment. There should be no hesitation, however, in carrying out surgery in cases with obviously serious visceral injuries and those who fail to respond to non-operative treatment.

Principle of damage control surgery

The other names for damage control surgery are 'abbreviated laparotomy', 'staged laparotomy', and 'planned reoperation'. Warren, one of the fathers of American surgery, made an important remark: "Show me an operation that takes more than two hours or requires more than two units of blood, and I will demonstrate a surgical complication". The principle of damage control surgery is adapted from its military application in warfare. Warren's remark is even more apt when dealing with a severely traumatised patient. In our local parlance, it is said that he who fights and runs away today, will live to fight tomorrow. The very ill, traumatised patient may not be able to withstand long surgical procedures due to adverse pathophysiological changes resulting from severe trauma. These include metabolic acidosis, hypovolemia, hypothermia and coagulopathy. Damage control surgery aims at carrying out the basic minimum procedures at the initial surgery when the patient's condition is unstable. The ultimate aim is to sustain life. Thereafter, the patient is sent to the intensive care unit for further resuscitation. During this period, the physiological derangements are addressed. The duration of stay at the ICU varies according to the clinical condition of the patient and may last for between 6 and 48 hours. The earlier the patient is returned to the theatre for definitive surgery, however, the better. All the same, it is prudent to make haste slowly. Initial (damage control) surgery is mainly geared towards addressing critical issues that may jeopardise the patient's chances of recovery. Essentially damage control surgery is carried out in the following stages

Stage 1 (preliminary surgery) consists of

- Control of haemorrhage: Bleeders are identified and haemostasis secured. Control of bleeding in solid organ injuries may involve packing and the application of haemostatic agents such as Surgicel, Avitene and fibrin glue
- Minimisation of the degree of contamination: Identify bowel injuries and apply temporary measures to limit the degree of contamination. The latter includes primary single layer closure and resection without anastomosis

- Application of temporary abdominal closure techniques: These include the use of towel clips, urology irrigation bag and mesh. This measure equally acts as a prophylaxis against abdominal compartment syndrome.
- Commence the correction of any physiological derangement such as hypoxia, hypothermia and acidosis

Stage 2: Correction of physiological derangements in the intensive care unit

- Hypoxia: Ensure optimal oxygen delivery. This may be achieved by volume-loading (to achieve optimum preload), blood transfusion (to optimise the oxygen-carrying capacity), inotropic support (to enhance cardiac output) and correction of acidosis.
- Correction of recognised blood clotting anomalies: May involve transfusion with fresh whole blood and/or relevant blood component
- Hypothermia: Warm and ambient environment, blankets and warmed intravenous infusion fluids
- Early detection of compartment syndrome: Monitor the intra-abdominal pressure by either Foley's catheter or by intragastric technique

Stage 3: Definitive surgery in the operating theatre:

This should be carried out within 24 to 48 hours of the initial surgery. Obviously, patients with seemingly uncontrollable bleeding or abdominal compartment syndrome will require much earlier intervention. Delayed reintervention may tilt the patient towards the development of grave complications such as sepsis, systemic inflammatory response syndrome (SIRS) and adult respiratory distress syndrome (ARDS). The development of any of the latter may preclude further surgery thereby increasing the associated morbidity and mortality of abdominal trauma.

Stage 4: Abdominal wall reconstruction (if necessary): Methods of closing the abdominal wall include

- Primary closure
- Closure with the skin left temporarily open
- Grafts: Gore-Tex and other synthetic sheets and Vicryl mesh
- Silo bags such as Bogota bag with subsequent gradual closure

ABDOMINAL COMPARTMENT SYNDROME (ACS)

This is defined as the adverse pathophysiological changes resulting from an acute elevation of the intra-abdominal pressure (IAP). It is associated with severe intra-abdominal injuries. The following are the consequences of a raised intra-abdominal pressure

- Splinting of the diaphragm by the abdominal viscera: The diaphragmatic stenting results in a decrease in both ventilation as well as lung compliance. It also causes an increase in the peak inspiratory pressure as well as reduction of the tidal volume. The resultant ventilation-perfusion imbalance results in hypercarbia and respiratory acidosis.
- Decreased venous return due to pressure on the inferior vena cava: This results in a decrease in the cardiac output due to reduced preload and increased afterload. The end-result is hypotension which may be aggravated by the associated hypovolemia.

- Renal dysfunction: Results from a combination of hypotension of cardiac origin (reduced cardiac output) and compression of the renal artery (increased renal vascular resistance) due to an elevated intra-abdominal pressure. A minimum IAP of 20mmHg is required to induce renal dysfunction. The latter may degenerate to acute renal failure
- Visceral perfusion: This may be deranged even with a much lower IAP of 15 mm Hg. Intestinal blood flow ranks among the first to be affected.
- Intracranial pressure: Raised IAP has been shown to increase the intracranial pressure with its negative consequences.

These pathophysiological changes are reversed by early decompression of the abdominal cavity.

Intra-abdominal pressure (IAP) in relation to ACS

- Normal: <10 cm water
- Mild ACS: 10 – 25 cm water
- Moderate ACS: 25 -40 cm water
- Severe ACS: > 40 cm water

A diagnosis of ACS is made when at least three of the following are present

- Increased IAP (25 cm water)
- PaCO₂: > 45 mm Hg
- Rise in airway pressure and decrease in tidal volume
- Sudden decrease in urinary output
- Relevant clinical scenario: Large pelvic haematoma or liver packing

Aetiological factors:

The following factors either acting independently or in concert are responsible for a rise in the intra-abdominal pressure

- Blood loss: Capillary oozing may result from coagulation anomalies. The situation is further compounded by third space loss
- Progressive retroperitoneal and bowel wall oedema: Trauma, ischemia and sepsis
- Presence of intra-abdominal packs: These may have been applied as part of damage control procedure
- Primary closure of the abdomen after a damage control procedure
- Non-operated patients with perforated viscus or diaphragmatic injuries with continuous release of gas and fluid into the intraperitoneal compartment
- Ascites
- Ileus
- Pneumoperitoneum

Clinical presentation: The patient is acutely-ill-looking with obvious respiratory distress. Abdominal examination will reveal a distended, tense abdomen. There is associated hypotension and oliguria. On the other hand, it may be diagnosed at surgery by an inability to approximate the wound edges after a trauma laparotomy.

Measurement of the intra-abdominal pressure:

This is carried out by determining the pressure in the urinary bladder. The supine patient is catheterised using a Foley catheter. This is connected to a drainage bag. The bladder pressure is measured by means of a T-piece bladder pressure device. The latter is connected to a pressure transducer which itself is connected to a monitoring system. The urinary tubing is clamped. The pressure transducer is clamped simultaneously at the midaxillary line. About 50 to 100 mls of isotonic saline is introduced into the bladder in order to overcome the resistance of the contracted bladder. This is followed by measurement of the bladder pressure on the monitor after zeroing. The bladder pressure (which is a reflection of the IAP) is usually above 25 mm Hg in abdominal compartment syndrome. The IAP can be measured from the pubic symphysis if the patient is not lying flat.

It is important to identify an impending abdominal compartment syndrome and institute preventive measures. This essentially entails temporary non-fascial closure of the abdomen.

Treatment of raised IAP

- Good intensive care to prevent complications of ACS
- Correct reversible factors: Hypoxia, hypothermia, acidosis and coagulation anomalies
- Treat ileus and acute colonic pseudo-obstruction
- Treat sepsis
- Surgical intervention: Control of persistent haemorrhage; decompression of the abdomen; wound closure with a silo.

Non-operative approach in the management of abdominal trauma

This principle has been applied to solid organ injuries involving mainly the liver and the spleen. Liver: Non-operative approach has been found successful in the management of blunt injuries and selected penetrating trauma. The basic criterion is that the patient must be haemodynamically stable and should be monitored closely. There should be no hesitation in resorting to surgery if there is clinical evidence of ongoing haemorrhage. Fortunately, delayed haemorrhage is rare with the liver when compared to the spleen.

Spleen: Non-operative management of splenic injuries has been quite successful in haemodynamically stable children. It has been extended to adults with grades 1 to 3 blunt splenic and selected penetrating injuries. Non-operative management will help to minimise excessive blood transfusion as well as reduce the morbidity and mortality associated with overwhelming postsplenectomy infection (OPSI).

Management of retroperitoneal haematoma

The retroperitoneal space is divided into superior and inferior compartments. The point of demarcation is the bifurcation of the aorta and inferior venacava. The superior compartment is further divided into the centromedian area (zone 1) and two lateral compartments (zone II). The inferior compartment, the pelvic peritoneum comprises zone III. Management protocol for the respective zones is as follows

- **Zone I: Mandatory exploration in both penetrating and blunt injuries**
- **Zone II: Explore in both penetrating and blunt injuries only if the haematoma is pulsating and/or expanding**
- **Zone III: Explore only in penetrating injury**

CHAPTER THIRTY-SIX

TRAUMATIC HEAD INJURY

Traumatic head injury (THI) has been defined as an alteration in the integrity of the head resulting from an impact. It is a common clinical condition and requires prompt diagnosis and treatment in view of the complications that may arise from it if not well managed. It affects all age groups but is commoner in young adults because of their higher degree of exposure. About half of the patients are aged below 30. Males are more affected than females.

Predisposing factors include

- Road traffic accident
- Fall from heights
- Assaults
- Sports-related injuries
- Penetrating trauma

Of these, road traffic accident tops the list. It is a common occurrence with motor cyclists especially the commercial ones who operate in developing countries. Despite the law making the wearing of crash helmets by motor-cyclists mandatory in developing countries, quite a good number of them, either out of ignorance or sheer carelessness, flout this law. The result is that traumatic head injury in such motor-cyclists is usually severe and is attended by high morbidity and mortality.

Classification of traumatic head injury: Various criteria have been used

- Nature of injury: Open or closed
- Extent of injury: Diffuse or focal
- Mechanism of injury: Coup or contrecoup
- Severity of injury (based on Glasgow Coma Scale): Minor, mild, moderate or severe
- Complication with bleeding: Non-haemorrhagic or haemorrhagic (extradural, subdural, subarachnoid, intraparenchymal or intraventricular)
- Timing: Primary or secondary. Primary injuries include contusions, cortical lacerations and diffuse axonal injury. They occur at the time of the initial impact. Secondary injury, on the other hand, results from the pathophysiological effects of the injury and occurs later

Pathophysiology of traumatic head injury

The skull has been likened to a rigid, inexpandable container that houses the brain. The perfusion of this system is maintained by a balance between the intracranial pressure on one hand, and the systolic blood pressure on the other. The relationship is depicted by the following equation
Cerebral perfusion pressure (CPP) = Mean arterial pressure (MAP) – Intracranial pressure (ICP)
An increase of the volume of the brain within the skull is bound to increase the intracranial pressure. Factors involved in increased intracranial volume are

- Increased blood flow
- Swelling of the substance of the brain
- An increase in the CSF volume
- Presence of an intracranial haematoma.

These factors acting singly or in concert result in an increase in the intracranial volume. The latter is bound to have a negative effect on the cerebral perfusion. Secondary brain injury may therefore result from a raised intracranial pressure, systemic hypotension or hypoxia. Other causes are pyrexia, seizures and electrolyte anomalies. A key element in the management of THI is the prevention or at least the minimisation of secondary brain injury.

Intracranial fractures: These may complicate head injury. Either the vault or base of the skull may be involved.

- Vault fractures: May be depressed or non-depressed; open or closed
- Base: May or may not be associated with CSF rhinorrhoea or otorrhoea

Intracranial haematomas associated with head injury are

- Extradural
- Subdural
- Subarachnoid
- Intracerebral
- Intraventricular

Severity of THI: Grouped into minor, mild, moderate and severe according to the Glasgow Coma Scale(GCS). See below

- Minor: GCS of 15 with no loss of consciousness
- Mild: GCS of 14 or 15 with associated loss of consciousness. It is also referred to as concussion. The latter is defined as physiological injury to the brain without any evidence of structural alteration. Its grading of severity takes into cognisance the length of both antegrade and retrograde amnesia
- Moderate : GCS of 9 to 13
- Severe: GCS of 3 to 8

Management of THI

This begins at the site of the injury. The primary survey is in line with the Advanced Trauma Life Support (ATLS) protocol. Life-saving measures are instituted according to the ABCDE protocol of resuscitation. Hence, the airway, breathing, circulation, assessment of disability (preliminary GCS) and exposure are carried out. Breathing may be ensured by the use of endotracheal intubation. While moving the patient to the hospital for definitive care, precautionary measures should be taken to restrict neck movement particularly when there is suspicion of an associated cervical spine injury. Secondary survey is aimed at identifying and instituting emergency care of other life-threatening injuries. This is then followed by a comprehensive evaluation of the head injury.

Focused head injury evaluation: The following information should be obtained

- A good history aimed at eliciting the type and mechanism of injury
- Alteration in the level of consciousness. Presence of a 'lucid interval' is important
- Associated bleeding or discharge particularly from the orifices (nostrils and ears): This depicts a fracture of the base of the skull
- History of associated seizures and/or vomiting
- Use of alcohol/drug: remote or active
- Current anticoagulant therapy
- Past history of psychiatric disease
- Premorbid history of headaches

Neurological assessment entails an ascertainment of the Glasgow coma score and relating the current value with what was obtained during the primary survey.

Glasgow coma scoring system (GCS) is aimed at eliciting an objective assessment of the level of consciousness of the patient by certain laid-down criteria. They add up to an overall score of 15.

- Eye opening: Score range of 1 - 4
- Motor response: Score range of 1 - 6
- Verbal response: Score range of 1 - 5

Eye opening	Spontaneous	4
	To verbal command	3
	To painful stimulus	2
	Do not open	1
Motor response	Obeys commands	6
	Localises pain	5
	Withdrawal flexion	4
	Abnormal flexion (decorticate)	3
	Extension (decerebrate)	2
	No motor response	1
Verbal response	Normal oriented conversation	5
	Confused	4
	Inappropriate words only	3
	Incomprehensible words	2
	No sounds	1
	Intubated patient	T

Other assessments include

- Cranial nerves: Pupillary reflex, ocular movements and assessment for VII nerve (facial nerve) palsy. Rule out hearing loss and dysphagia
- General/motor examination
- Corneal reflex

The following blood investigations are carried out

- Full blood count
- Electrolytes/urea/creatinine
- Arterial blood gases
- Blood alcohol level

Radiological studies

- Plain X-ray: In addition to the trauma series (cervical, chest and pelvis), an X-ray of the skull may be requested. It must be pointed out, however, that it has been largely replaced by CT in the management of THI. The information that can be obtained from it includes demonstration of a skull fracture and a shift from the midline which is more obvious when the pineal gland is calcified
- CT scan: This is the current gold standard investigation in the management of TBI. The range spans from the base of the occiput to the top of the vertex in 5mm increments
- MRI: Has a limited role in the investigation of these very ill patients. It is, however, superior to CT in the management of diffuse axonal injury
- Monitoring of the intracranial pressure: Normal range is 8 to 12 mm Hg

Treatment of THI

As mentioned earlier, this entails treatment of the primary brain injury as well as prevention of secondary brain injury. Close monitoring is equally essential for early detection of potential complications such as intracranial haemorrhage. As usual in the management of any form of trauma, initial resuscitation and stabilisation of the patient should take precedence over radiological investigations. For a patient in hypovolemic shock, it is important to identify and address the source of blood loss as a priority because the resultant hypotension will expedite the onset of secondary brain injury. Treatment will depend on the degree of head injury

Mild head injury: Most are managed and discharged home from the Emergency Department. This is on the premise that the patient should be brought back to the hospital for reassessment if there is any inkling of deterioration of the clinical condition. The patient must therefore be handed to the care of a responsible adult. The place of routine CT scan in the management of mild head injury must be weighed against the cost of the procedure and more importantly against the risk of undue exposure to irradiation. It is, however, imperative to carry out this procedure on patients who fall into the following criteria

- Age > 65
- Symptoms of raised intracranial pressure: Seizure, vomiting
- Antegrade amnesia > 30 minutes



CT SCAN OF THE BRAIN: NOTE THE BICONVEX, HYPERDENSE EXTRADURAL HAEMATOMA

- Antegrade amnesia > 30 minutes
- History suggestive of a dangerous mechanism of injury
- Patient on anticoagulant therapy
- Clinical evidence of neurological deficit
- Suspicion of a skull fracture

Moderate to severe head injury: After initial stabilisation and investigation, treatment is aimed at preventing secondary brain injury and early detection of potential complications such as intracranial haemorrhage. As outlined above, the main thrust of treatment of moderate to severe head injury lies in the prevention and treatment of raised intracranial pressure. This enables the brain to recover from the baseline injury.

Acute treatment of raised intracranial pressure

- Elevate the head of the bed by 30 degrees (reversed trendelenburg). Exercise caution in patients with cervical spine injury. This encourages venous return. Care should, however, be taken when dealing with patients wearing a cervical collar. Ensure that the collar is not tight as this may be counterproductive by impeding venous return.
- Ensure adequate ventilation in order to prevent hypoxia and hypercapnia
- Administration of diuretics to help 'shrink' the brain: Agents that have been used include furosemide, hypertonic saline (3-23%), urea and mannitol. Mannitol is an osmotic diuretic and is most commonly employed. It is given as a bolus of 0.25-1 gm/kg every 4 to 6 hours with a limit of 4 gm/kg/day. Adequate hydration should be ensured prior to the administration of mannitol because the massive diuresis sequel to its administration may worsen an existing hypovolemic shock in an already underhydrated patient.
- Prevent and control seizures by the use of anticonvulsants
- Administer barbiturates such as thiopentone to suppress cerebral metabolism
- Antipyretic administration for the prevention and control of pyrexia

Ensure adequate fluid and electrolyte balance. Maintain euvoolemia with normal saline. If anything, it is better to err on the side of mild hypervolemia.

Controversial issues in the treatment of raised intracranial pressure

- Decompressive craniectomy and durotomy: This entails an operative removal of part of the skull coupled with incision of the restrictive dura mater covering the brain. It is aimed at increasing the capacity of the skull to accommodate the increased brain volume particularly in situations of refractory raised intracranial pressure in the absence of any form of intracranial haemorrhage. Craniectomy allows swollen brain tissue to herniate upwards through the surgical defect rather than downwards to compress the brain stem. The larger the bone flap removed, the more the reduction in the intracranial pressure. Craniectomy equally improves cerebral perfusion pressure and cerebral blood flow. It is recommended in young patients whose intracranial pressure is uncontrollable by other methods. There is a poorer outcome in those above 50. The bone flap is preserved and replaced at cranioplasty after recovery from the brain injury. Complications of craniectomy include meningitis and brain abscess. This is obviously a desperate measure in the management of TBI. The role of decompressive craniectomy in TBI and in the control of intracranial hypertension remains a matter of debate.
- Ventriculostomy: 'Tapping' of a small quantity of cerebrospinal fluid (CSF) may aid in the reduction of the ICP. Again, its application is controversial. Ventriculostomy, however, may be used for the monitoring of the intracranial pressure (ICP). The ICP may also be monitored through the parenchymal route.

In addition, it is important to ensure adequate nutrition. This could be achieved by either enteral or parenteral route.

Intracranial haemorrhage:

This is divided into two broad groups: intra-axial and extra-axial.

- Intra-axial: Bleeding occurring within the brain itself. Could be intraparenchymal or intra-ventricular
- Extra-axial: Bleeding occurring within the skull but outside the brain tissue: Extradural, subdural and subarachnoid.

Extradural haematoma

This connotes bleeding occurring between the dura mater and the skull and is commonly due to laceration of a meningeal artery, especially the middle meningeal artery. It may also arise from a torn sagittal sinus or oozing from the diploe, bone and stripped dura mater on each side of an associated fracture. One-third of the patients will demonstrate a lucid interval. The latter is a period of temporary loss of consciousness and apparent recovery ('lucid interval') followed some hours later by deterioration in the level of consciousness. It is due to cerebral compression by the extradural haematoma. Clinically, there may be boggy swelling of the scalp over the clot, ipsilateral papillary dilatation and contralateral hemiparesis or hemiplegia. CT scan will show a hyperdense, biconvex swelling between the skull and the brain.

Treatment: Extradural haematoma is a surgical emergency. Treatment entails evacuation of the haematoma and securing of adequate haemostasis via a craniotomy.

Subdural haematoma

This is bleeding into the space between the dura and the arachnoid. Of two types: acute and chronic

Acute subdural haematoma: Results from an overwhelming brain injury usually with pulping and laceration of the brain coupled with a disruption of a cortical vessel. Unlike the lucid interval that may occur in extradural haematoma, there is no episode of 'recovery' from the initial coma; rather there is continuing deterioration as the haematoma expands. On CT, the acute haematoma is hyperdense, concave-shaped with a diffuse spread. Treatment entails evacuation of the haematoma. The poor prognostic outcome is due to the severe nature of the primary brain injury.

Chronic subdural haematoma: This follows an apparently trivial injury and is common in the elderly and patients on anticoagulant therapy. The supposed injury may have been forgotten. There is a minor tear in a cerebral vein as it traverses the subdural space. This results in chronic leakage of blood which is exacerbated by coughing and straining. Subsequently, the haematoma gets encapsulated and by the process of osmosis, absorbs fluid from the tissues, and expands. The resultant 'hygroma' results in compression of the underlying brain tissue. The clinical features of headache, drowsiness, vomiting and papilloedema may raise the possibility of an intracerebral tumour. Diagnosis is by CT which will show a hypodense mass characteristic of a bleed that has spanned a period of at least two weeks. It should be noted that whereas an acute bleed is hyperdense, a subacute bleed (10 days to 2 weeks) is isodense while a chronic bleed is hypodense. Treatment is evacuation of the haematoma. A burr-hole exploration may be carried out under local anaesthesia.

Subarachnoid haemorrhage: There is bleeding between the arachnoid and the pia meningeal layers. Trauma constitutes the commonest cause. Aneurysm is a common cause of spontaneous subarachnoid haemorrhage. Clinically, it presents with features of 'meningism': headache, neck stiffness and positive Kernig's sign. Treatment is conservative with bed rest and analgesics followed by rehabilitation when symptoms subside.

Intracerebral haemorrhage: This usually follows severe brain injury. There are scattered small haemorrhages throughout the brain substance. CT scanning will demonstrate extensive cerebral injury. Management is conservative and requires close monitoring. A delayed haematoma may evolve in the course of conservative management and this will require surgical evacuation.

Intraventricular haemorrhage: May result from either rupture of an intracerebral clot into the ventricle or a choroid plexus tear at the time of injury. It follows a severe head injury and occurs most commonly in children.

Skull fractures: Of two types – vault and base

Skull vault fractures: Compound fractures will require debridement particularly if there is associated underlying injury. Only severely depressed closed vault fractures require surgical elevation.

Basal skull fractures: The 7th and 8th cranial nerves may be affected. CSF otorrhoea and rhinorrhoea are common features of basal skull fractures. The role of antibiotic in the management of basal skull fractures is controversial. To prevent meningitis, a persistent CSF leakage may require delayed craniotomy and anterior fossa dural repair.

Management of penetrating injuries: In addition to the general management of head injury, treatment entails debridement and surgical removal of the penetrating object(s). It must be stressed that the latter procedure should only be carried out in the operating theatre.

Complications of head injury

- Focal neurological deficits: May affect cranial nerves I, IV, VII and VIII
- Post-traumatic seizures particularly in patients with penetrating injuries. Patient should be commenced on prophylactic anticonvulsant therapy
- Post-traumatic hydrocephalus
- Meningitis especially in basal skull fractures with otorrhoea and rhinorrhoea
- Deep venous thrombosis
- Post-concussion syndrome: Characterised by headache, nausea, vomiting, emotional lability, sleep disturbances and memory loss. May last for 2-4 months. Management is multidisciplinary.
- Alzheimer's syndrome: May be a late occurrence.
- Vascular injuries: Arterial transection may result in post-traumatic aneurysms and carotid-cavernous fistulae.

- Brain death: This is an acute, catastrophic, and irreversible brain injury. Diagnosis is hinged on the demonstrable absence of all brain stem reflexes. These include the corneal reflex, gag reflex and the vestibulo-ocular reflex. There is also absence of respiratory movement on disconnection from a respirator.

Prognostic factors in head injury

- Initial GCS of 8 and below adversely affects outcome
- Age: Prognosis worsens with age
- Factors that contribute to raised ICP such as hypoxia and pyrexia adversely affect prognosis

Glasgow Outcome Score (GOS): Grades the functional outcome and the possibility of returning to the pre-injury state after management of head injury.

Outcome	Score
Good recovery	5
Moderate disability	4
Severe disability	3
Persistent vegetative state	2
Dead	1

CHAPTER THIRTY-SEVEN

HYDROCEPHALUS

Hydrocephalus is a clinical condition due to excess accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain. It is due to a disquilibrium between the production and absorption of CSF. The end-result is a raised intracranial pressure and dilatation of the ventricles.

Functions of the CSF

- Regulates the intracranial pressure in the midst of changing cerebral blood flow
- Acts as a protective water-jacket for the intracranial contents

Pathophysiology of CSF: CSF is secreted by the choroid plexuses of the ventricles of the brain. These comprise of the two lateral ventricles on either side of the brain, the third and the fourth ventricles. The fourth ventricle communicates with the subarachnoid space. There is intercommunication between the ventricles: the third and fourth ventricles communicate through the aqueduct of Sylvius while the fourth ventricle connects with the subarachnoid space through the medial and lateral ducts of Magendie and Lushka respectively. Cerebrospinal fluid is reabsorbed into the dural venous system. Total volume of circulating CSF is 150 mls and the rate of production is 20 mls per hour.

Classification of hydrocephalus: There are two types of classification

- Communicating and non-communicating
 - Communicating: There is no physical obstruction; pathways are patent but there is defective absorption (trauma, inflammation such as meningitis, subarachnoid haemorrhage)
 - Non-communicating: Due to an obstruction along the CSF pathway (inflammation, tumours)
- * Congenital and acquired
 - Congenital: Commonly associated with spina-bifida and myelomeningocele. May be due to congenital stenosis at either the foramen of Munro or the aqueduct of Sylvius
 - Acquired: Brain tumours, trauma, meningitis and subarachnoid haemorrhage

Clinical features of congenital hydrocephalus

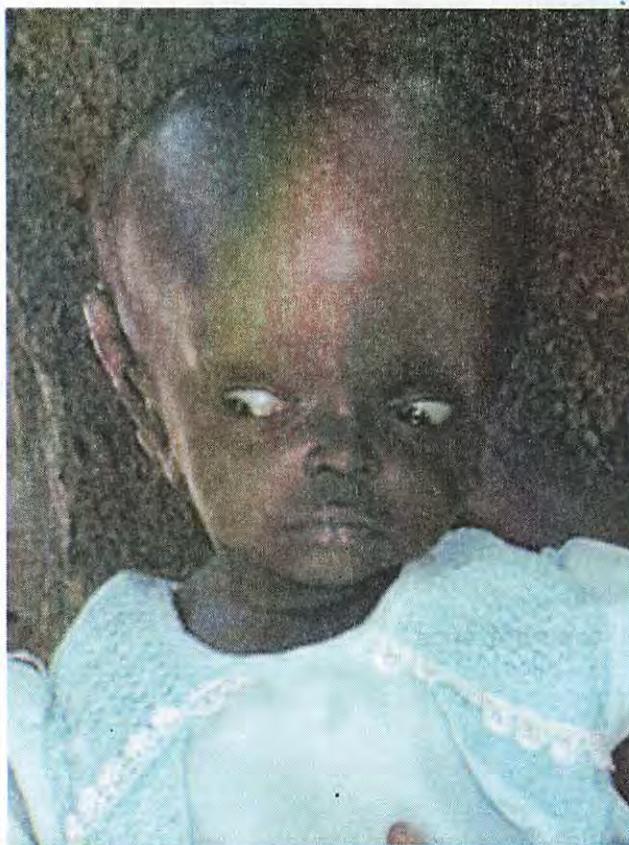
- Increased occipito-frontal (head) circumference
- Sutural diastasis
- Engorged scalp veins
- Bulging anterior fontanelle
- Sun-setting eyes
- May be associated with spina bifida (myelomeningocele), and talipes equinovarus deformity. These should be examined for in every case of hydrocephalus

Investigations for hydrocephalus

- CT scan of the brain: Will demonstrate marked dilatation of the ventricles. In children, the skull X-ray may show a copper-beaten appearance
- MRI of the brain: Quite informative particularly when there is aqueductal stenosis
- Ventriculography
- Lumbar puncture: Diagnostic and therapeutic in communicating hydrocephalus but highly contraindicated in non-communicating variety as it may result in coning

Treatment of hydrocephalus:

- Address any identified underlying cause. This includes surgical excision of a causative mass lesion. Congenital hydrocephalus is treated with CSF shunt procedures. These include
- Ventriculo-peritoneal shunt: This involves insertion of a catheter into the lateral ventricle and tunnelling same subcutaneously for drainage into the peritoneal cavity. The excess CSF is channelled and absorbed by the peritoneal lining of the abdomen
- Other shunts: Ventriculo-atrial and ventriculo-pleural shunts
- Complications of shunts



HYDROCEPHALUS: NOTE THE INCREASED HEAD CIRCUMFERENCE, BULGING ANTERIOR FONTANELLE AND THE 'SUN-SET' EYES

About 15% to 20% of shunts are removed within the first year of insertion due to shunt blockage and infection

- Shunt blockage: May be due to adhesion by the choroid plexus or cellular debris
- Infection: Occurs in 1% to 7%. Common organisms are E.coli and staph epidermidis
- Overdrainage of shunts: May result in subdural haemorrhage
- Others: Seizures, stroke and intracerebral haemorrhage

CHAPTER THIRTY-EIGHT

MANAGEMENT OF BURNS

Burns is a common clinical condition that is associated with a high level of morbidity and mortality. A variety of agents may predispose to burns injury. The commonest is thermal injury. Other agents include chemicals, electricity, and sunburn. Prompt intervention and treatment is necessary both for the survival of the patient as well as the prevention of complications that may arise from delayed treatment.

Classification of burns: This is based on both the thickness of skin involved (partial or full) as well as the extent of skin involvement as depicted by the surface area involved (major or minor). A partial thickness burn leaves a part or the whole of the germinal epithelium intact. On the other hand, a full thickness burn destroys the entire germinal epithelium.

Classification based on the depth of burns

- First degree burns: Quite superficial; red, dry and associated with pain
- Second degree burns: Involves the superficial part of the epidermis. The area may have a reddish, mottled appearance with blisters and associated with severe pain. Second degree burn may be divided into two varieties based on the extent of the burn injury: superficial and deep. The latter is a more serious injury, takes a longer time to heal, and may heal with scarring
- Third degree burns: The germinal layer is completely destroyed and healing is by dense scar tissue. Characteristically, it is not painful owing to destruction of the nerves. It has a charred, leathery appearance.
- Fourth degree burns: In addition to the situation in third degree burns, there is involvement of the underlying structures (muscle and bone).

Depth	Clinical characteristics	Common cause
First degree	Erythema, pain, absence of blisters	Sunburn
Second degree	Red/mottled appearance, blisters	Contact with hot liquids
Third degree	Dark and leathery	Fire, electricity, lightning, prolonged exposure to hot liquids

Generally speaking, it is believed that burn depth is usually underestimated at the time of initial assessment. It is not uncommon to find all three degrees of burns in the same burn wound. Moreover, complications such as infection, have been known to increase the depth of the burn injury.

Classification based on the severity of the burns

This takes the following into consideration: depth, surface area, part of the body involved and the age of the patient. It is known that both morbidity and mortality are worse in the very young and the much older age groups. Based on severity, there are two varieties of burns – major and minor. Major burns is more serious and more often than not requires inpatient management.

Major burns includes

- Second degree burns greater than 10% surface area in patients younger than 10 or older than 50
- Second degree burns greater than 20% in persons of other age groups
- Third degree burns greater than 5% in all age groups
- Burns of whatever degree or extent involving the following parts of the body: face, hands, feet, genitals, perineum and major joints
- Electrical burns including those caused by lightning
- Chemical burns
- Burns with associated inhalational injury
- Burns with concomitant mechanical trauma such as fracture
- Patients with pre-existing medical disorders that would complicate management leading to prolonged recovery and increased mortality

Calculation of the total body surface area involved

1 Wallace's Rule of 9: The body is divided into zones with each zone having a surface area calculated as a multiple of 9

- Each lower limb (anterior and posterior surfaces): 18%. Total for both lower limbs is 36%
- Each upper limb (anterior and posterior surfaces): 9%. Total for both upper limbs is 18%
- Trunk: Each surface (anterior and posterior): 18%. Total for both surfaces is 36%
- Head and neck: 9%
- Perineum: 1%

2 Lund and Browder chart: It is more authentic as it is more age-specific and assesses each part individually.

3 Assessment by the patient's hand: The palmar surface of the patient's hand is estimated as 1% of the body surface area. The area of the palm that excludes the fingers is approximately 0.5%.

Pathophysiology of burns

Local: Jackson has described three zones in the local area of burn injury

- An innermost zone of coagulation necrosis
- A middle zone of stasis
- An outermost zone of hyperemia

General effects of burns: Local and systemic

Local effects:

- The coagulation necrosis caused by heat leads to the release of vasoactive peptides. These act by increasing capillary permeability. The resulting fluid loss leads to hypovolemia. Depending on the total body surface area involved, hypovolemic shock may set in with its attendant complications. Fluid loss is at its highest peak within the first 24 hours of injury while fluid exudation is at its peak within the first 8 hours of injury. After 24 hours, there is restoration of the capillary integrity. This, coupled with the formation of a protective coagulum over the wound, lead to a reduction in the fluid loss. As a rule, burns less than 15% of body surface area are not usually associated with an extensive capillary leakage.

- Infection results from loss of the protective skin cover. Common organisms include beta-haemolytic streptococcus, staph aureus, pseudomonas and kiebsiella
- Eschar formation, if circumferential, may result in compartment syndrome
- Contractures appear late and are associated with deformities such as ectropion, and joint deformities
- Hypertrophic scar and keloids may form over the contractures

Systemic effects

- Hypovolemic shock results from fluid loss through the damaged capillaries. It is worse within the first 24 hours and drastically reduces with the formation of a coagulum over the surface of the burn wound
- Renal function: Reduced renal blood flow induces the release of antidiuretic hormone and aldosterone which respectively enhance the reabsorption of water and sodium from the renal tubules. Myoglobin released from damaged muscle tissue coupled with toxins released from the wound may damage the kidneys resulting in acute tubular necrosis.
- Lungs: Inhalational injury may give rise to pulmonary oedema, bronchospasm, and decreased chest and lung compliance. It may be associated with clinical features of adult respiratory distress syndrome (ARDS).
- Metabolism: Initially there is a reduction in the metabolic rate. This is followed by hypermetabolism and negative nitrogen balance. There is accelerated gluconeogenesis, insulin resistance and increased protein catabolism.
- Gastrointestinal system: Mucosal ischemia resulting from hypovolemia may give rise to a specific type of acute gastric ulceration referred to as Curling's ulcer.
- Anaemia: Results both from destruction of red blood cells in the charred capillaries as well as toxic inhibition of bone marrow activity arising from burn wound sepsis

Initial management of burns

This is in line with the primary survey layout of all trauma cases

- A: Maintenance of airway. May involve endotracheal intubation and even tracheostomy in prolonged intubation
- B: Breathing- assess for inhalational injury
- C: Circulation – ensure adequate intravascular access. Send blood specimens for FBC, electrolytes/urea/creatinine, PO₂ and PCO₂.
- D: Disability and level of consciousness
- E: Environment – get patient out of the area of burns. Remove clothing from the burnt area

Secondary survey

- Adequate assessment to determine the extent of the burns: Percentage, degree and type
- Evaluation for inhalational injury
- Admit into a Burns Unit with barrier nursing and aseptic procedures
- Administration of intravenous analgesic
- Commencement of tetanus prophylaxis
- Administration of intravenous antibiotics

- Pass a urethral catheter to monitor urinary output. The latter reflects the degree of fluid replacement
- Commence on anti-ulcer regimen such as omeprazole in severe burn injury

Fluid replacement in burns

The amount of fluid to be given takes into consideration the weight of the patient, the percentage burns and time of injury. Various formulae have been designed in order to ensure timely and adequate fluid resuscitation. The most commonly used formulae are those of Parkland and Muir/Burclay. The preferred replacement fluid is crystalloids: Ringer's lactate and normal saline. The former is regarded as the fluid of choice. In addition to replacing the fluid loss as calculated by any formula, the normal daily requirement of fluid should also be given. In the tropics, this adds up to 3 litres/24 hours. Owing to the increased catabolic rate coupled with reduction in the rate of gluconeogenesis, it is advised that 1 litre of 5% dextrose in water should form a part of the fluid for daily requirement. The remainder should be in the form of either Ringer's lactate or normal saline

- **Parkland formula:** This is an estimate of the fluid loss within the first 24 hours of burn injury. It takes into consideration the weight of the patient and the percentage body surface area involved. The formula is:

$$4\text{ml} \times \% \text{ burns} \times \text{body weight in kg}$$

The maximum weight accepted for this calculation is 50 kg. Half of this fluid is given in the first 8 hours (period of maximal fluid loss) and the balance administered over the remaining 16 hours. The time of injury marks the starting point of calculation

By this formula, a patient who weighs 70 kg and sustains 20% burns should receive a total of: $4 \times 20 \times 70$ mls of fluid in the first 24 hours. This amounts to 5600 mls of crystalloids. He should be given a total of 2800 mls of fluid within the first 8 hours starting from the time he sustained the injury (this should be obtained from the history). The administration of the remaining 2800 mls of intravenous infusion should be spread over the subsequent 16 hours. In addition, this patient should have additional 1 litre of 5% dextrose and 2 litres of Ringer's lactate or normal saline to take care of the normal daily fluid requirement.

- **Muir and Barclay formula:** By this formula, fluid is administered in rations. Each ration is calculated as

$$\% \text{ burns} \times \text{body weight in kg} \times \frac{1}{2}$$

3 rations are given in the first 12 hours

2 rations are given in the second 12 hours

1 ration is given in the third 12 hours

Using the case scenario presented earlier as an example, it follows that each ration in a 70 kg patient whose burn injury involves 20% of the total body surface area will amount to $20 \times 70 \times \frac{1}{2}$. The product of this is 700 mls

He should therefore receive

- 700 x 3 mls within the first 12 hours. This amounts to 2100 mls of crystalloids
- 700 x 2 mls within the next 12 hours. This amounts to 1400 mls of crystalloids
- 700 x 1 mls within the next 12 hours. This amounts to 700 mls of crystalloids. The normal daily requirement as mentioned earlier should be added to the replacement fluid.

By the second day following burns, the fluid requirement is less. The patient should receive half of the calculated loss administered within the previous 24 hours. To this is added the daily fluid requirement. In extensive burns with loss of red blood cells, blood transfusion is imperative and the blood so administered is regarded as part of the replacement fluid. Blood transfusion is usually commenced after initial resuscitation with intravenous infusion.

As important as these formulae are, they should be regarded as guides in relation to fluid replacement in burns. The clinical assessment of fluid replacement takes precedence over the formulae. The vital signs and urinary output of the patient are the best indices of fluid replacement and so should be closely monitored. The minimum urinary output is 1 ml/kg/ hour or 30 to 50mls/hr for adults and 1.2mls/kg/hour for children. Patients passing pigmented urine resulting from high voltage electrical or deep thermal burns may require a higher dose of crystalloids or mannitol in order to prevent acute tubular necrosis.

Nutrition in burns: As mentioned above, there is an increase in the metabolic rate with catabolism taking pre-eminence. Early commencement of oral feeding, if tolerated, is advised. Nasogastric tube-feeding should be initiated if oral feeding is not feasible. Patient may require as much as 6000 kcal/day. Eggs, peanut oil and locally available supplements may be necessary in the management of anaemia and malnutrition. Severe burns, however, is a good indication for total parenteral nutrition.

Management of the burn wound

Role of escharotomy: Circumferential and even near circumferential burns may result in compromise of the blood supply to the affected area by virtue of the formation of eschar. This may end up as compartment syndrome. Escharotomy in such situations is therefore mandatory. This is important both in burns involving the extremities when perfusion may be compromised as well as the torso where ventilation may be threatened.

Local treatment: Initial management entails removing the burnt clothing and flooding the affected area with cold running water preferably for 30 minutes. This will go a long way in mechanically cleaning up the area. Thereafter, wound dressing is commenced by either the open or closed method. **Benefits of wound dressing include**

- Prevention of wound dessication
- Control of pain
- Reduction of wound colonisation and infection
- Prevention of additional trauma

Most topical dressing materials are endowed with the following qualities

- Viscous carrier which helps to reduce dessication
- Broad antibiotic spectrum which reduces the rate of infection
- Guaze wrap which reduces soiling of both clothing and unburned skin. It also protects the wound from the external environment

Agents employed in the management of burn wound include

- Silver nitrate: Antiseptic; depletes electrolytes and stains the local environment
- Silver sulphadiazine (Dermazine, flamazine): Antiseptic and has good penetration ability. Causes neutropenia and wound maceration. Should be avoided in pregnancy, nursing mothers and infants less than 2 months of age since it may result in kernicterus
- Mafenide acetate (Sulfamylon): Very good tissue penetrating ability particularly the eschar; anti-pseudomonal. Painful and causes acidosis
- Petroleum jelly: Bland and non-toxic
- Honey: Promotes wound healing and prevents bacterial contamination

Wound membranes: They are used to

- Provide transient and physiologic wound closure.
- Protect the wound from mechanical trauma and also help to reduce fluid loss by evaporation from the exposed raw surface of the wound.
- Create a barrier to penetration by bacteria thereby reducing the incidence of burn wound infection.

They are usually applied over clean, superficial wounds that are awaiting epithelialisation. It is important to avoid applying these over devitalised tissue as the subsequent submembrane purulence may result in local and systemic sepsis.

Various types of dressing membrane include

- Porcine xenograft
- Split thickness allograft
- Hydrocolloid dressings
- Impregnated gauzes
- Semipermeable membranes

Open method of burn wound management is applicable to burns involving the face, one side of the body and the perineum. It is aimed at enhancing the formation of eschar over the burn wound. Eschar constitutes the most natural covering for the protection of the wound from growth of bacteria as it inhibits their growth. Patient should be nursed in an isolated clean and ambient environment in order to limit cross-infection. The wound is cleaned with chlorhexidine and covered with sulphadiazine cream (dermazine, flamazine). Daily wound inspection is carried out for early detection of cracks within the eschar. Such cracks should be covered with Vaseline gauze or sofratulle.

Closed method of burn wound management: This is carried out by the use of occlusive dressing such as gamgee. The latter consists of a thick pad of cotton wool that is wrapped up with gauze. Gamgee is noted for having a good absorbent ability. Applied like a splint over the wound, it creates a good barrier against bacteria, absorbs wound exudate and minimises fluid loss. In addition, it enhances separation of eschar. The initial dressing is changed after one week and twice weekly thereafter. Application of Vaseline gauze directly on the wound before the application of gamgee will reduce pain and bleeding associated with wound dressing. Biological dressings may be employed as temporary cover for extensive burn wound injuries. The above methods (open and closed) are most suitable for partial thickness burns.

Surgical treatment of the burn wound:

This is useful in the management of full thickness as well as deep partial thickness burns. It helps to reduce pain and infection rate and also encourages early mobilisation of the patient. Surgical intervention should be carried out under general anaesthesia with provision made for blood transfusion as this procedure is associated with significant blood loss. Primary haemorrhage may be minimised by the subcutaneous injection of a dilute solution of adrenaline (1: 500,000 or 1: 1,000,000). Application of a tourniquet will equally prove useful in the prevention of primary haemorrhage. For maximal effect, a combination of both methods of prevention is invaluable. Deep partial thickness burn injury is managed by tangential excision. This entails the 'shaving off' of the dead layer of the wound down to the deeper layer of dermis. In the course of excision, punctate haemorrhage signals the presence of healthy tissue and the termination of excision. On the other hand, full thickness burns demands a more radical excision. Excision in this case is taken down to the subcutaneous layer in order to ensure complete eradication of all necrotic tissue. Skin cover is required immediately after burn wound excision. The ideal natural cover, particularly in full thickness burns is skin. If this cannot be achieved immediately, then a temporary cover in form of a biological membrane will be useful. It is important to point out that, as much as possible burns involving the hand, antecubital fossa and popliteal fossa require immediate skin grafting. Postoperative management involves adequate analgesia, maintenance of fluid balance, regular changing of wound dressing and elevation of the limb to reduce bleeding and oedema.

Electric burns

The degree of injury depends on the electrical voltage. A voltage below 500 volts is likely to result in local injury. Exposure to higher voltage may result in loss of consciousness, fractures, cardiac arrhythmias, myoglobinuria and compartment syndrome. Severe acidosis is a known complication of electrical burns.

Treatment involves

- Administration of large volume of intravenous infusion to avert acute renal failure as a result of myoglobinuria. Patient may be given as much as 2ml/kg body weight/hour. The urinary output should be closely monitored.
- Administration of sodium bicarbonate is necessary in the presence of acidosis
- Fasciotomy should be carried out when indicated for the prevention and treatment of compartment syndrome.
- Severe limb injuries may benefit from amputation

Chemical burns

In addition to causing local skin damage, chemicals may also be absorbed with systemic consequences. The resultant pathological effect depends on the offending chemical. It is therefore important to identify the chemical in question after initial resuscitation of the patient. Management involves

- Immediate removal of clothing and chemicals
- Copious irrigation with tap water for at least 30 minutes
- Apply topical ocular anaesthetics in eye injuries
- Ensure adequate fluid resuscitation in large injuries

Outline of management of burns in special areas

Facial burns: This results in gross oedema within the first 48 hours of injury. Gradually, the oedema fades away. Involvement of the eyes is usually a cause for concern. It is important to address the following issues

- Involvement of the eyelids may require tarsorrhaphy and early skin grafting. The ophthalmologist should be involved in the management of burns involving the eyelids
- The wound is managed by the open method with the application of topical silver sulfadiazine (Dermazine) over the wound.
- Full thickness skin graft is employed in full thickness burns.
- There is a high risk of auricular chondritis in burns affecting the ears. Such wounds require twice daily dressing and application of mefenamic acid (Sulfamylon).
- Burns involving the face may be complicated by inhalational injury. Early diagnosis and treatment is important in the management of inhalational injury (see below)

Neck: Managed by open dressing. Main danger is the development of flexion contractures

- Nurse the patient in a prone position in order to ensure extension of the neck. The latter may be facilitated by applying a pillow under the neck
- When ambulant, a stiff neck collar should be applied

Axilla: Main complication of burns involving the axilla is adduction contractures

- Patient should be nursed with the arms abducted
- Crucifix splint may be used in children

Hand: There is danger of developing compartment syndrome. It is imperative therefore to monitor the circulation for early detection. One should not hesitate to carry out escharotomy or fasciotomy if in doubt

- Splint the hand in a position of function (extension at the wrist, flexion at the metacarpophalangeal joints and extension of the fingers)
- Dress each finger and web separately
- Elevate the limb to reduce oedema
- Institute early physiotherapy
- Full thickness burns requires early excision and grafting

Perineum: Managed by the open method

- Ensure adequate cleaning of the wound
- Consider the use of scrotal support in males

Legs and feet: Treated by the open method. There is a risk of compartment syndrome

- Do not hesitate to carry out fasciotomy if necessary
- Elevate the limb to prevent oedema
- Dress each web and toe separately in burns affecting the feet

Complications of burns

1 Hypovolemic shock: This is due to loss of fluid following the loss of the protective cover of the skin. As pointed out earlier, fluid loss is at its peak during the first eight hours of burn injury. It is best prevented by adequate fluid replacement as detailed earlier.

2 Inhalational injury: This is a serious complication of burns. There is history of exposure to burns in a closed space. Patient may sustain facial burns. The singeing of the nasal hairs and the presence of carbonaceous debris in the mouth, pharynx and sputum help to clinch the diagnosis

Pathophysiology: Inhalational injury results in

- Upper airway oedema
- Bronchospasm
- Occlusion of the bronchioles
- Increased dead space and intrapulmonary shunting
- Infection: Pneumonia may arise due to loss of ciliary activity, and occlusion of the bronchioles
- Other lesions: Laryngo-tracheo-bronchitis, laryngeal oedema and pulmonary oedema

Investigations include

- Chest X-ray: May be normal initially until pneumonia sets in
- Bronchoscopy: May show evidence of erythema, ulceration, and presence of carbonaceous debris
- PO₂ and PCO₂ estimation

Treatment: Initial resuscitation may be followed up with

- Bronchoscopy: Effect vigorous pulmonary toilet
 - Endotracheal intubation and ventilation
 - Tracheostomy if intubation is prolonged. This will help in sucking out unusually thick secretions
 - Intravenous antibiotics
- 3 Genitourinary complications:** Burns may result in the following
- Acute renal failure: Due to diminished renal blood flow and myoglobinuria. It is best prevented by ensuring adequate fluid resuscitation
 - Urinary tract infection: Commonly results from prolonged urethral catheterisation. Prevention lies in discontinuing urethral catheterisation as soon as the patient is conscious enough to void urine naturally
- 4 Infection:** This ranges from burn wound sepsis to septic shock and is a common cause of death in major burns. Again, it is basically due to the loss of the protective cover of the skin.

Burn wound sepsis: Confirmative diagnosis is by carrying out a burn wound biopsy. Minimum bacterial count of 10^5 /gram of tissue is diagnostic. Infective organisms include pseudomonas, E. coli, and proteus. Others are coliforms, staphylococci and streptococci. The burn wound may form a portal for infection by clostridium tetani resulting in tetanus. Prophylactic measures include wound debridement and regular wound dressing. Systemic antibiotics are not very helpful as destruction of blood vessels to the skin coupled with the barrier created by the eschar prevent the antibiotics from getting to the site of burn wound. Topical antibiotics are more effective in the early stages of the injury. Wound cleaning, debridement and excision improve the accessibility of the wound to systemic antibiotics. Tetanus is prevented by adequate wound care and administration of tetanus prophylaxis. Septicaemia and septic shock may complicate burn wound sepsis and will require specific management.

5 Gastrointestinal tract complications are as follows

- Stress ulceration: Gastric mucosal ischemia due to hypovolemic shock may result in a special form of stress ulcer referred to as Curling's ulcer. This is a known cause of upper GI bleeding. Preventive measures include proper fluid resuscitation and prophylactic administration of anti-ulcer drugs such as H₂-receptor blockers (cimetidine or ranitidine) and proton pump inhibitors (omeprazole or esomeprazole). Gastrointestinal haemorrhage should be initially managed conservatively. Refractory cases require surgical intervention by way of vagotomy and drainage or partial gastrectomy
- Acute gastric dilatation and paralytic ileus: These will require nasogastric aspiration and intravenous infusion
- Damage to the liver: Occult hepatic degeneration may occur particularly in major burns. Hepatotoxic drugs are best avoided during management of burn

6 Vascular complications: They include

- Superficial thrombophlebitis following prolonged intravenous therapy
- Deep venous thrombosis: Preventive measures (thromboprophylaxis) should be instituted during prolonged immobilisation
- Pulmonary embolism: Usually a complication of deep venous thrombosis

7 Psychiatric complications: These range from depression to psychosis. Much depends on the psychological, pretraumatic make-up of the patient. Regular reassurance with the aid of a psychologist may be helpful. Do not hesitate to invite a psychiatrist for psychiatric evaluation and treatment in very serious cases

Late complications of the burn wound: These range from contractures and keloids to Marjolin's ulcer. The latter is due to metaplastic epithelial transformation from benign to malignant lesion (mainly squamous cell carcinoma). There may be loss of body parts.

Most of these complications can be prevented with proper clinical management

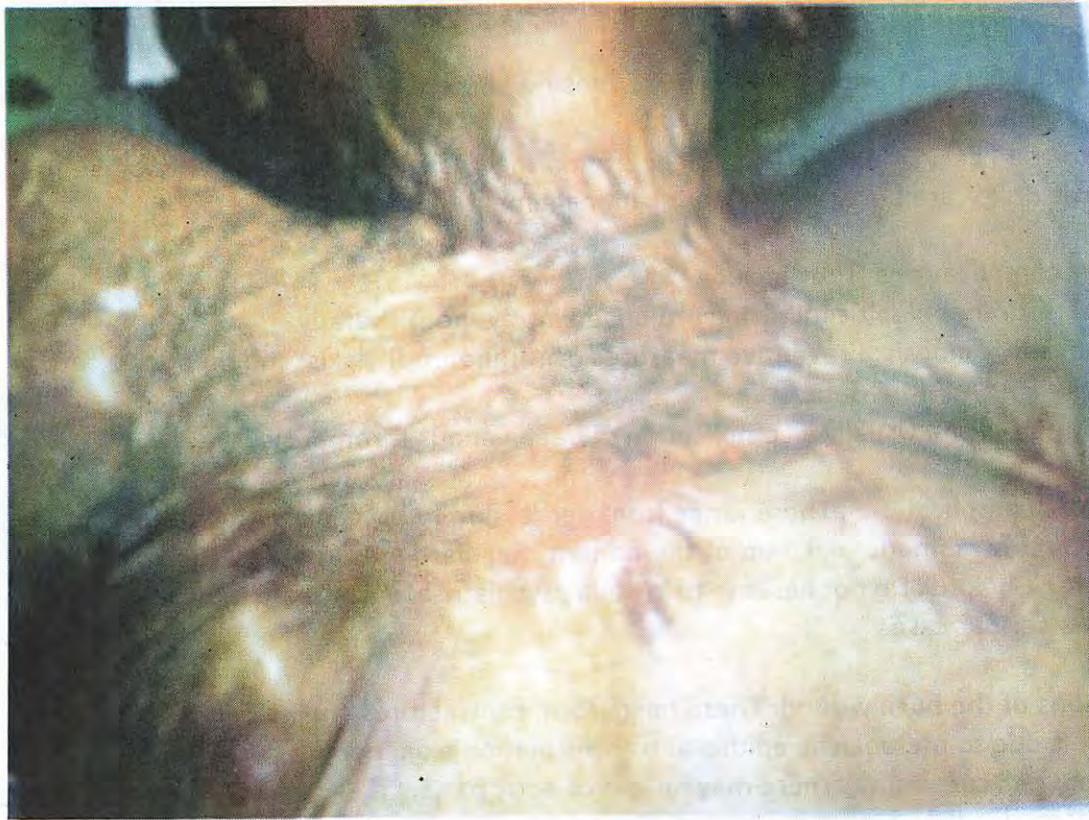
Rehabilitation: This should commence as soon as possible in order to prevent hypertrophic scarring and ensure early reintegration into society. It will also help in ensuring a good quality of

life after treatment. It involves scar massage, use of compression garments and management of pruritus. Others are application of topical silicone and injection with steroids to reduce scarring.

Prevention of burns:

The best form of management is prevention. Burns is associated with high morbidity and mortality. It poses a lot of challenges in management. All efforts must therefore be geared towards prevention

- 1 As much as possible, children should be restrained from entering the kitchen while cooking is taking place. Pot handles should be turned towards the back of the stove or gas-cooker to forestall any accidental tripping.
- 2 Fire extinguishers and sand buckets should be placed near the kitchen. Expired fire extinguishers should be replaced.
- 3 Install smoke detectors and test them once a month to ensure their efficacy. These should mandatorily be replaced after 10 years.
- 4 Keep water heater temperature below 120° F and ensure that the temperature of bath water is tolerable before use.



Post-burns contracture

- 5 Proper handling of matches and lighters
- 6 Do not leave electric naked wires exposed; rather insulate and install electrical outlet covers.
- 7 Keep chemicals out of reach and wear protective gloves while handling such.

- 8 All smoking products should be stubbed out completely.
 - 9 Ensure that fire exits form an integral part of any building plan particularly in places that pull a lot of crowd such as cinema houses, laboratories, and event places.
 - 10 Frequent dress rehearsals of escape manouevres in the event of a fire outbreak.
- 1.1 Provision of well equipped and efficient fire services which should be within reach.
 - 1.2 Adequate communication logistics: Good access roads and telecommunication facilities.

CHAPTER THIRTY-NINE

CHRONIC LEG ULCERATION

The definition of chronic leg ulceration incorporates the following features

- A breach in the epithelium of the skin below the level of the knee
- Ulcer persisting for more than 6 weeks
- Ulcer shows no tendency to heal after three months of appropriate treatment
- Ulcer still not fully healed at 12 months.

In practice, ulcers of the feet are categorised as forms of chronic leg ulceration. It is a common clinical condition that affects about 1% of the adult population in developed countries. The exact incidence in the developing world is not known but is likely to be much higher compared to the incidence in the developed countries. In the latter, the incidence increases with age and is about 4% - 5% in the older generation of 80 years and above. For optimal management, an interdisciplinary approach should be encouraged.

Pathogenesis: This depends on the aetiology. The current view, however, is that non-healing ulcers develop in an environment containing high levels of activated metalloproteinases which may result in chronic tissue turnover and failure in wound closure

Classification of chronic leg ulceration

Infective causes: Also referred to as specific ulcers

- Tropical ulcers: Infective organisms are *Bacteroides fusiformis* (anaerobe) and *Borrelia vincentii* (aerobe). Initially acute in nature but later becomes chronic. Predisposing factors include malnutrition, trauma and walking barefooted.
- Tuberculous ulcers: *Mycobacterium tuberculosis*
- Buruli ulcers: *Mycobacterial ulcerans*
- Syphilitic ulcers: *Treponema pallidum*
- Yaws: *Treponema Pertenuis*

Trauma: This is the commonest cause and a major predisposing factor in the pathogenesis of tropical ulcers. This is more so in people who walk barefooted.

Venous ulcers: Constitute about 70% of all ulcers in the developed world. It affects the part of the leg just above the medial and lateral malleoli. Risk factors include varicose veins, older age, obesity, congenital vein anomalies, and past history of deep venous thrombosis. Others are previous leg injuries, phlebitis and multiple pregnancies. It arises from incompetence of the venous valves and results in venous dilatation, retrograde flow of blood and venous hypertension. There is subsequent leakage and deposition of brownish/red pigments in the skin. The end result is an ulcer around the malleoli. Another theory, the leucocyte entrapment theory, proposes that the reduction in the arteriovenous gradient due to venous hypertension results in sluggish movement of blood. The resultant increased adherence of blood cells to the

endothelium causes the release of inflammatory mediators and microthrombi formation. Ulceration results from capillary occlusion and microangiopathy. Complications of venous ulcers include cellulitis, osteomyelitis and malignant change.

Arterial ulcers: These constitute about 10% of ulcers in the developed world. There is a reduction in the arterial blood flow. It occurs over the toes, heels, shin and bony prominences of the foot. Causes include peripheral vascular disease due to atherosclerosis and diabetes mellitus. The patient may present initially with clinical features of intermittent claudication (pain on walking). It may eventually progress to pain even at rest. The resultant ischemia results in rapid development of gangrene and ulceration.

Pressure (decubitus) ulcers: About 70% of decubitus ulcers occur in the geriatric age group. It is equally a known complication of prolonged immobilisation. The bony prominences are affected. These include the areas of the ischial tuberosities, and the heels. They constitute one category of ulcers that may be prevented by proper clinical management (2-3 hourly turning in bed, use of special mattresses etc). The attendant complications of decubitus ulcers are grave and include septicaemia, osteomyelitis and even death.

Vasculitis ulcers: Rheumatoid arthritis, systemic lupus erythematoses (SLE), polyarteritis nodosa

Metabolic ulcers: Diabetes mellitus may be complicated by ulceration around the foot. It occurs in about 15% of all diabetics and about 10% to 15% of such patients end up with amputation. It is a serious clinical condition as it is estimated that worldwide, a lower limb is lost every 30 seconds as a result of this complication of diabetes mellitus.

Blood dyscrasias: Sickle cell disease, thalassemia, thrombocytopenia and polycythaemia rubra vera. Others are hereditary spherocytosis, thrombotic thrombocytopenic purpura, granulocytopenia and leukemia

Neurotropic ulcers: Leprosy, diabetic neuropathy, cord lesions, peripheral neuropathies, and syringomyelia. Others are alcoholic neuropathy and tabes dorsalis

Malignant ulcers: These include those that are malignant ab initio as well as longstanding benign ulcers that have undergone metaplastic transformation (Marjolin's ulcer). Others include malignant melanoma, squamous cell carcinoma and Kaposi's sarcoma



CHRONIC LEG ULCER

Recently, it has been found that chronic kidney disease (CKD), myocardial infarction and even hypertension may be associated with an increased risk of chronic leg ulceration. Malnutrition and deficiencies of vitamins, zinc, selenium, iron and folic acid have also been implicated, particularly in relation to tropical ulcers.

Clinical presentation

Patient presents with a history of a non-healing leg ulcer. In the history, one should endeavour to identify any associated risk factor as outlined above. Lifestyle predisposing factors such as smoking and obesity should be noted. A past medical history of DVT is important. Examination of an ulcer should be meticulous and includes the following

- Site, size, and shape
- Floor (what you see): May consist of purulent material (connoting an on-going infection), granulating tissue (healing ulcer), or slough with devitalised tissue
- Edge: Punched out (syphilitic), undermined (tuberculous), sloping (healing ulcer), and everted (malignant ulcer)
- Base: Where the ulcer sits. Can only be determined by palpation. May be indurated or hard particularly in longstanding ulcers and those of malignant disposition.
- Surrounding skin: Oedema, discolouration, erythema, maceration, sensation
- Arterial pulsations : dorsalis pedis (between the 1st and the 2nd metatarsal of the foot), popliteal artery (behind the knee), and femoral artery (groin)
- Varicose veins: Common predisposing factor in venous ulceration
- Inguinal nodes at the groin to rule out lymphadenopathy (infection, neoplasms)
- Range of movements in the associated joints of the lower limb: Hip, knee and ankle

Investigations

- Wound swab for microscopy, culture and sensitivity. Commonly cultured organisms include pseudomonas aeruginosa, E.coli, and staph aureus. Others are klebsiella, citrobacter and proteus
- Quantitative bacterial culture: Wound biopsy yield of at least 10,000 count per gram is suggestive of an ongoing infection
- Wound biopsy to rule out malignancy
- Fasting blood sugar: Diabetes is a common cause
- Blood genotype: Sickle cell disease is a common cause particularly in the black race
- Other blood investigations: Full blood count, ESR and lipid profile
- Plain X-ray of the leg including the relevant area to rule out bony involvement (osteomyelitis and malignancy).
- CT and MRI: May be necessary depending on the findings on plain X-ray
- Venography
- Arteriography
- MR angiography and CT angiography: Only when necessary
- Doppler ultrasound scan of the limb to rule out obstruction of a blood vessel.
- Ankle Brachial Pressure Index (ABPI)
- Clotting profile: Prothrombin time, activated partial thromboplastin time and international normalised ratio (INR)

Principles of treatment

It is important to treat the patient holistically as summarised in the saying: "Treat the whole patient and not the hole in the patient". Conservative management involves the creation of an environment that enhances the healing of the ulcer. This includes weight reduction in the obese individual as well as discouragement of smoking in a chronic smoker. There are, however, some indications for surgical intervention. The essential aspects of the conservative management include

- Address the underlying cause
- Promote circulation and enhance venous return
- Promote healing
- Health education

After the clinical assessment and investigations, the following steps are taken

- Wound care: A clean wound is ensured to facilitate wound healing. Wound debridement may be carried out either by wound dressing using appropriate dressing solution and techniques or by way of surgery (surgical debridement). This will be discussed later.
- Infection control: At the beginning of treatment, commence the patient on a combination of a broad spectrum antibiotic and metronidazole. The regimen should be reviewed later in line with the culture and sensitivity result.
- Promote blood circulation and enhance venous return: This is achieved by the use of compressive stockings and elevation of the limb. The latter enhances venous return and so helps to reduce oedema. When in the sitting position, the limb should be elevated by placing it on a stool. In the lying position, venous return is enhanced by placing the leg on one or two pillows. This is very relevant in the treatment of venous ulcers. The minimum recommended period of unit limb elevation is at least 30 minutes three or four times daily.
- Medical treatment: Pentoxyfylline (Trental) and Aspirin (300mg daily) have been recommended as adjuncts to compressive therapy
- Vacuum-assisted closure therapy which works by the creation of a subatmospheric pressure has been shown to enhance wound healing.

Wound dressing in chronic leg ulceration

Types of dressing and solution to be used depend on the nature of the wound. Generally speaking, a dirty wound and one with slough needs to be dressed with a solution that effects desloughing. This is a much more gradual process when compared to surgical desloughing. When the wound gets cleaner, dressing should be carried out with a relatively innocuous solution in order to maintain the integrity of the ingrowing epithelium and granulation tissue. The dressing materials range from highly absorbent materials such as foams, hydrogels and hydrocolloids employed in discharging wounds to plain gauze and paraffin gauze which are employed in relatively clean wounds.

Solutions used in achieving wound debridement include Edinburgh University Solution of Lime (EUSOL), hydrogen peroxide, honey and sugar. Biological debridement with the use of larva has been found effective.

EUSOL: This is calcium hypochlorite solution. It is made up of boric acid and chlorinated lime. Presentation is in separate packs of its components: EUSOL A (boric acid) and EUSOL B (chlorinated lime). The dressing solution is constituted just before use by mixing solutions A and B. It has a strong oxidising property and acts on various types of organisms (bactericidal, fungicidal and viricidal). As mentioned earlier, it is a good debriding and desloughing agent and is therefore one of the solutions of choice at the initial stage of the management of an infected

wound. It is discontinued when the wound is cleaner owing to its toxic effects on fresh epithelium and granulation tissue.

Hydrogen peroxide: Like EUSOL, it is used as a debriding agent. It is, however, more potent in this regard than EUSOL. In practice, it is most suitable in the initial management of very bad necrotic wounds prior to the introduction of EUSOL. It induces a high protease activity which results in the breakdown of devitalised tissues. In addition to its antimicrobial activity, hydrogen peroxide also promotes angiogenesis. Like EUSOL, it is toxic to upcoming granulation tissue and epithelium and should be discontinued after the initial desloughing.

Honey: This is a natural derivative of the honey bee. It is hyperosmolar, acidic and contains hydrogen peroxide and nitric oxide. Its antimicrobial activity is derived from a combination of these properties and contents

- Hyperosmolarity: Absorbs water from the microbes resulting in their death. Similarly, absorption from the tissues helps to dry up the exudates as well as the associated oedema.
- Acidity (pH 3.2 – 4.5): This destroys the organisms, while stimulating leucocyte function and increased fibroblastic activity
- Hydrogen peroxide: Effects as outlined above
- Nitric oxide: Stimulates immunological response

Normal saline: It is the least toxic of all the dressing agents. In practice, it is brought into use after the wound has been rendered much cleaner following the preliminary use of the above 'stronger' dressing agents. The dressing is by 'wet to dry' dressing technique as it promotes mechanical debridement. As the dressing gets drier following the evaporation of fluid, it becomes hyperosmolar and causes the ingress of fluid and exudates into the dressing. In addition to cleaning the wound, this action also reduces tissue oedema. It is the solution of choice during the proliferative phase of wound healing. One major advantage of normal saline over the other dressing agents is that it is most readily available and affordable

Other dressing agents not commonly used include iodine, sugar and larva.

- Iodine in the form of povidone iodine is a broad spectrum antimicrobial agent. Of special note is its action against Methicillin-Resistant Staph Aureus (MRSA). Its use, however, is limited by its toxic effect on tissue.
- Sugar: Like honey, it acts by its osmotic properties
- Sterile larva of the greenbottle Lucilia sericata: It produces proteolytic enzymes that degrade and liquefy necrotic tissue. It feeds on the end-catabolic products of the latter. The drawbacks to its usage include bleeding and migration to other parts of the body. Besides, the mere presence of worms in the body, adversely affects the psychological state of the patient.

When the wound is assessed to be clinically clean as evidenced by the presence of pinkish granulation tissue, coverage of defect is achieved by means of an autologous mesh graft.

Indications for surgery: Categorised as early and late

- Early: Surgical debridement is indicated in necrotic wounds with a good chunk of devitalised tissue. While ensuring that most devitalised tissue is excised, it is important to conserve as much of the skin as possible. The procedure could be carried out under sedation either by the bedside or in the operating theatre depending on the extent of the devitalisation in the wound. Thereafter, wound dressing is commenced using the appropriate agent as outlined above
- Late: Usually to provide skin cover when the wound is adjudged clean and defect has been filled up with granulation tissue. Healing of such wounds is naturally by secondary intention which takes a long time and the end-product may be an unacceptable scar. Indications for skin graft include ulcers that have been refractory to conservative management and those of rather large size. Surgical intervention is also recommended when conservative treatment is envisaged to be unduly prolonged.

Shave therapy: This refers to the radical surgical treatment of an ulcer that covers a wide and contiguous area of the leg. Tangential excision of the entire affected tissue is carried out. Coverage of the defect is carried out subsequently.

THE SURGEON'S DUTY

The surgeon's duty is to bear the results of his surgery. A surgeon who has had a difficult case, who is asked to attend a sick colleague and yet therefore has to leave the theatre to go home to sleep off a hangover will be vulnerable and perhaps a little less than his normal self, but he will still be there, providing excellent efficient medical care.

The good surgeon, however, can be guided by common sense and right judgment from his own experience and knowledge as well as by the words of his colleagues and the advice of his teacher, and find pleasure in his work as well as pride in his patients' recovery.

It is important to remember that the surgeon's responsibility is to his patient, not to himself, and it is his duty to do his best to help him to recover and to give him all the information he needs to do so. He must also be aware of the fact that his patient is not the only one who may benefit from his skill and knowledge, and that his patient's family and friends may also benefit from his care and concern.

PRINCIPLES OF SURGERY

The good surgeon is always looking forward to the day when he can help his patient to recover from his illness or injury, and he does not mind if this takes a long time. He is always willing to help his patient, even if it means staying with him for hours at a time, and he is always ready to do whatever it takes to help his patient to recover.

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CHAPTER FOURTY

WOUND HEALING

Basically, Surgery entails the management of wounds. Much of these wounds are 'iatrogenic' as they are 'inflicted' by the surgeon in the course of carrying out surgical procedures. In a similar manner, it is equally the responsibility of the surgeon to manage wounds inflicted by external trauma (road traffic accidents, gunshot, stabs, burns etc).

Types of wound healing

- Primary intention: This is the method of healing of a surgically incised wound. In this case, the wound edges are apposed and held in place by mechanical devices such as sutures, adhesive strips and clips. The device so inserted is retained until the wound is sufficiently healed and strong enough to withstand stress without external support
- Secondary intention: In a situation of tissue loss, healing is by granulation tissue, contraction and epithelialisation. This is the method of healing in infected wounds particularly when the edges of the wound are far apart with or without skin loss. Wound contraction is by the activity of myofibroblasts while fibroblasts stimulate the formation of granulation tissue
- Tertiary intention (also referred to as delayed primary closure): Wound closure is delayed and later carried out at the proliferative phase of wound healing (see below). Note that wounds closed secondarily heal by tertiary intention

There are four phases of wound healing

- Haemostatic phase: Initial bleeding sequel to an injury is brought under control by the formation of a blood clot. In a similar manner, the initial vasoconstriction is succeeded by the adhesion and aggregation of platelets
- Inflammatory phase (within 2 to 3 days): The aggregated platelets are degranulated. This results in the release of factors that encourage wound healing. The latter include cytokines such as interleukin and tumour necrotic factor (TNF). Others are growth factors such as fibroblast growth factor (FGF), platelet derived growth factor (PDGF), and transforming growth factor-beta (TGF beta). In addition to causing vasodilatation and increased permeability of the blood vessels, these factors also stimulate the activity of fibroblasts and keratinocytes in the wound. They equally attract inflammatory cells (neutrophils, monocytes and macrophages) into the wound site. These help to sterilise the wound by clearing the 'debris' from within while cytokines activate the complement system.
- Proliferative phase (3 days to 3 weeks): There is an increased fibroblastic activity. Fibroblasts secrete a matrix comprising of glycosaminoglycan, proteoglycans and collagen. The existing fibrin is used as the scaffold. Under the influence of vascular endothelial growth factor (VEGF), there is formation of new capillary (angiogenesis).

Epidermal growth factor (EGF) causes re-epithelialisation by the migration of cells from the periphery of the wound

- Maturation and remodelling: This goes on for upwards of 3 months to 1 year. Maturation of collagen takes place. This results from the replacement of type III collagen by type I variety and continues until a ratio of type I to type III of 4:1 is achieved. During this period, there is realignment of collagen fibres along the lines of tension and decreased wound vascularity. There is a gradual but steady increase in the wound tensile strength for approximately 8 to 12 weeks following surgery. At this point in time, it is about 75% to 80% of the tensile strength of normal tissue. The tensile strength continues to increase thereafter for about one year post-injury. On the long run, however, it never reaches the tensile strength of uninjured tissue. The end-product of the ensuing capillary regression is a less vascularised wound.

Factors affecting wound healing: Local and systemic

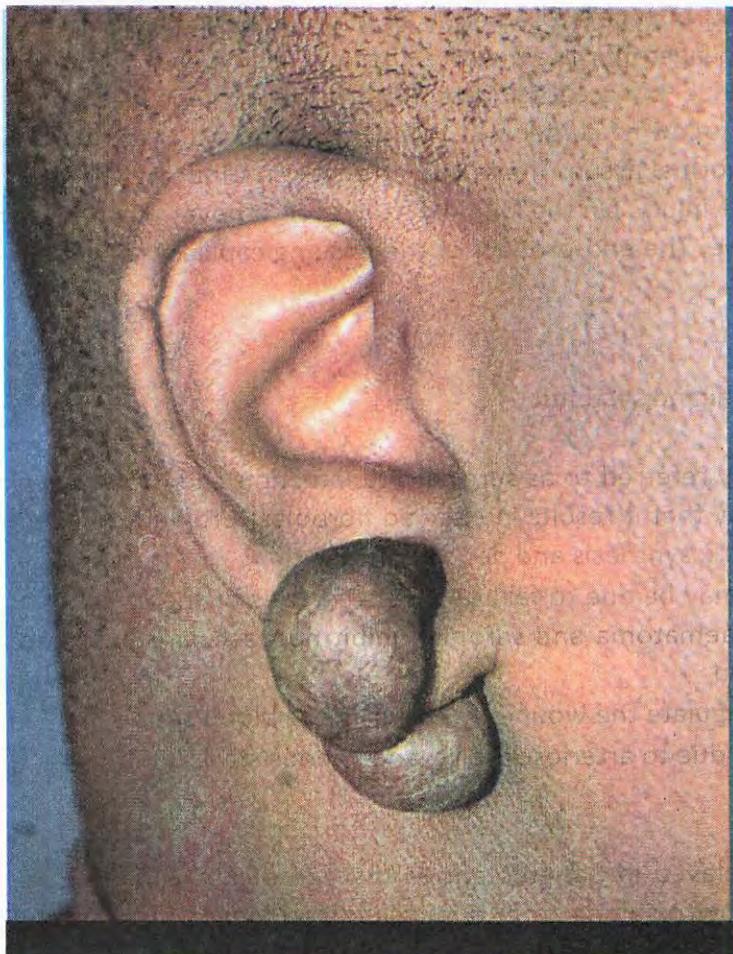
Local causes are

- Wound infection: This is currently referred to as surgical site infection and is discussed under that heading. Suffices to say that it results in delayed fibroblast proliferation and consequently delayed wound matrix synthesis and deposition.
- Haematoma and seroma: These may be due to failure of haemostasis or the effect of a generalised bleeding diathesis. Haematoma and seroma inhibit neovascularisation and predispose to surgical site infection
- Tight sutures: These literally strangulate the wound by reducing the blood supply
- Others are diminished blood flow due to arteriosclerosis, and venous stasis

Systemic factors include

- Age: Wound healing is usually delayed in patients above the age of 60. This may be attributed to diminished blood flow resulting from diminished cardiovascular capacity and atherosclerosis. Relative nutritional deficiency may be an additional factor
- Nutritional deficiency
- Vitamin C deficiency: Results in deficiency of collagen production
- Vitamin A deficiency: Gives rise to defective fibroplasia, collagen synthesis, cross-linking and epithelialisation
- Vitamin B₆ deficiency: Results in impairment of collagen cross-linking
- Obesity: Predisposes to wound separation and dehiscence
- Anaemia: Decreased oxygenation results in reduction of collagen synthesis
- Corticosteroids: Causes a reduction in wound inflammation, collagen synthesis and contraction
- Diabetes mellitus: Resultant microangiopathy leads to poor wound perfusion, impaired keratinocyte growth and platelet-derived growth factor functions in the wound.
- Others are cancer, radiation, hypoxia and sepsis.

Bone healing: Essentially as above. There is periosteal and endosteal proliferation and increased osteoblastic activity. The resultant effect is callus formation. Remodelling of the bone is by osteoclastic activity. This is explained in more details later (see principles of fracture management)



KELOID INVOLVING THE RIGHT EAR LOBE

Complications of wound healing: These include surgical site infection (SSI), wound dehiscence, burst abdomen and incisional hernia. Others are keloids and hypertrophic scars. While the others are discussed in the relevant sections, the differences between Keloids and hypertrophic scars are outlined below.

Criterion	Keloids	Hypertrophic scar
Location	Areas of high vascular supply such as ear lobe, face, neck, chest	Can be found in any part of the body
Age	Not found in extremes of age	Can be found in any age group
Race	Seen mainly in blacks	Found equally in all races
Inheritance	Autosomal recessive	Not familial
Sensitisation	Required as they are not born with	No sensitisation is required

	antibody	
Growth	Grows beyond the edge of the scar	Conffined within the scar edges
Duration of	Continues proliferation	Stops proliferation after
Proliferation	even after 1 year	6months
Shape	Claw-like processes are seen	Usually linear
Duration between	Long duration	Short duration
Injury and formation		
Treatment	Triple: excision, steroids and radiotherapy	None usually required as it regresses spontaneously

339

CHAPTER FOURTY-ONE

METABOLIC RESPONSE TO TRAUMA AND SEPSIS

Following trauma, the body reacts by putting various mechanisms in motion whose sole aim is to restore it back to its pre-injury state. The magnitude of this reaction is directly proportional to the severity of the injury. It may be modified, however, by comorbid conditions and complications such as infection. Though primarily meant to be protective, a major response may have adverse effects on organs distant from the injured site.

The study of metabolic response to trauma was pioneered by Sir Cuthbertson et al in 1932. They reported that trauma is accompanied by a hypercatabolic state which could be as much as 20% to 25% higher than the pre-morbid state. In relation to surgery (which indeed is a form of controlled trauma), they propounded that this natural response could be modified positively by altering the perioperative management of the surgical patient. They referred to the endocrine, metabolic and immunological changes triggered off by injury as 'stress response'. This is in recognition of the roles played by the so called stress hormones in the pathophysiology of major stress such as trauma and sepsis. If left to run its natural course, this body response may deteriorate to the clinical condition of systemic inflammatory response syndrome (SIRS) which may further degenerate to multiple organ dysfunction syndrome (MODS). Clinical management of severe injury is aimed at reducing the catabolic response in order to fast-track recovery.

The metabolic response to trauma is divided into three phases

- Ebb phase: Lasts for 24 to 48 hours
- Flow phase: Duration depends on the severity of the injury. May last for weeks
- Anabolic phase

Ebb phase: This term refers to a reduction in the flow of body metabolism. In this phase, mechanisms are put in motion to maintain tissue perfusion to vital organs while reducing energy consumption and urinary nitrogen excretion. The reduction in the effective blood volume that follows severe injury causes some physiological changes which are meant to increase the cardiac output as well as the delivery of oxygen to the tissues. This is achieved by the following

- Release of antidiuretic hormone by the hypothalamus: This enhances reabsorption of water in the renal tubules
- Release of aldosterone by the adrenal gland through the renin-angiotensin mechanism. This results in the reabsorption of sodium in exchange for potassium in the renal tubules
- Release of catecholamines which results in peripheral vasoconstriction
- Reduced insulin secretion by the pancreas results in hyperglycaemia

Flow phase: The duration of the flow phase is proportional to the severity of trauma. This ranges from 3 to 8 days following uncomplicated elective surgery to weeks after severe trauma and sepsis. There is a catabolic state which affects the peripheral tissues such as muscle, fat and skin while preserving vital organs such as liver and kidneys. This exercise is aimed at maintaining a 'flow' of energy and protein substrates that aid recovery from trauma. There is an increase in the

metabolic rate and a corresponding increase in oxygen consumption. The mediators include glucagon, catecholamines and corticosteroids. Others are cytokines, oxygen radicals and other local mediators. The following are the specific metabolic changes that occur during the flow phase following trauma

1 Carbohydrate metabolism: Insulin is the physiological regulator of carbohydrate metabolism. It is equally the most important anabolic hormone in the body. In addition to the regulation of blood glucose, it also plays an important role in the regulation of protein metabolism. This is achieved by reducing muscle protein degradation on one hand and aiding protein synthesis from amino acids on the other. As regards fat metabolism, it stimulates the formation of triglycerides while reducing its breakdown. Insulin resistance is the main effect of the metabolic response to trauma. The hyperglycaemic state associated with the metabolic response to trauma is multifactorial

- Action of stress hormones: They are cortisol, epinephrine, norepinephrine, glucagon and growth hormone. The activity of these hormones is anti-insulin in nature. For instance, following trauma, there is an increase in the sympathetic activity due to the action of epinephrine and norepinephrine. This increases the rate of hepatic glycogenolysis through the release of glucagon. Glucagon is a naturally recognised anti-insulin as it known to increase the blood sugar level.
- Normal or reduced level of insulin: Reduced level of insulin secretion is mediated by pancreatic alpha receptors, interleukin 1 (IL1), and tumour necrotic factor (TNF)
- Increased peripheral insulin resistance: This results in a reduction in the peripheral utilisation of insulin.

Insulin resistance in sepsis, for instance, is directly proportional to the degree of the stress response. Exogenous insulin therapy may be employed to prevent tissue breakdown.

2 Lipid metabolism: There is an increase in the turnover of fatty acids and glycerol following trauma. Free fatty acids (FFA) constitute the primary source of energy after trauma. They provide energy for the increased rate of gluconeogenesis which is fuelled by lactate and amino acids provided by the liver. Lactate has been found to be the major substrate for gluconeogenesis. It has been found that gluconeogenesis cannot be completely inhibited by exogenous glucose and insulin administration. Breakdown of fat (lipolysis) is stimulated by the secretion of the following hormones: ACTH, cortisol, catecholamines, glucagon and growth hormone. This generates the FFA required to facilitate the metabolic process. Under normal circumstances, the increased FFA has a negative feedback effect on lipolysis. In severe trauma, however, glycolysis continues even in the presence of a high level of FFA.

3 Protein metabolism: The hypercatabolic state following trauma results in systemic proteolysis. Protein catabolism exceeds its anabolism following trauma resulting in a negative nitrogen balance. Glucocorticoids constitute the main driving factor. This results in an increase in the urinary nitrogen excretion which is proportional to the degree of trauma. Skeletal muscle constitutes the major site of protein catabolism. It harbours about 80% of the body's pool of free amino acids. Glutamine constitutes 60% of the latter. The amino acids so generated are utilised in the synthesis of acute phase proteins, albumin, fibrinogen, glycoprotein, and complement factors. They are also used for gluconeogenesis and energy substrates for rapidly proliferating tissues. There is a daily loss of about 1.5% body weight in a traumatised patient who is not

receiving oral intake. While skeletal muscle tissue is utilised in the metabolic response to trauma, vital organs such as the liver and kidneys are preserved. The rate of protein degradation is directly proportional to the degree of trauma. It is particularly high in septic conditions. Muscle catabolism has been found to be reduced by the administration of adequate nutritional support during the flow phase

The following is a comparison of the metabolic response between ebb and flow phases

	Ebb phase	Flow phase
Blood glucose level	Increased	Normal or increased (secondary to insulin)
Gluconeogenesis	Normal	Increased (due to increased gluconeogenesis)
Free fatty acid	Increased	Normal or increased (due to mobilisation)
Insulin concentration	Reduced	Normal or increased (due to insulin resistance)
Catecholamines	Increased	Increased (due to increased sympathetic activity)
Glucagon	Increased	Increased (due to increased sympathetic activity)
Blood lactate	Increased (decreased blood pH)	Normal
Oxygen consumption	Reduced	Increased (due to increased metabolic rate)
Cardiac output	Normal or increased	Normal or increased (due to increased heart rate)
Core temperature	Reduced	Increased (shivering, vasoconstriction, heat generation)

Anabolic phase: This marks the stage of recovery (reparative phase) and takes place at the end of the metabolic response to trauma. There is regeneration and re-synthesis to compensate for the tissue loss.

Factors that influence the extent and duration of the metabolic response

Pain and fear: Stress and pain increase metabolic rate, constant fear, slogan of anaesthesia, etc.

Surgery: type, region, duration and preoperative support: Type of surgery, region, extent of surgery, duration of surgery, preoperative support.

Extent of trauma and degree of resuscitation: Extent of trauma, degree of resuscitation.

Post-traumatic complications: haemorrhage, hypoxia, sepsis, fever, re-operation.

Pre-existing nutritional status: Ability to regenerate. No other basic nutrient apart from oxygen.

Anaesthetic considerations: Least with epidural anaesthesia.

CLINICAL APPLICATION: This is aimed at minimising the negative effects of the metabolic response to trauma.

Consideration is given to both phases of the response (ebb and flow) respectively.

Flow phase: early cooling, rewarming, surgical removal of dead tissue, reduced sweating, etc.

Surgery and anaesthesia: AT the level right is to consider.

Surgical method: Less common with minimal access surgery (MAS).

Anaesthesia: Less common with epidural anaesthesia.

Avoid hypothermia during surgery: Hypothermia leads to coagulopathy, respiratory depression, etc.

Ebb phase: Early resuscitation should include fluid bolus, antibiotics, etc.

Prompt fluid and blood replacement in order to maintain the blood pressure: Early resuscitation.

Adequate oxygen supply and ventilation: Early resuscitation.

Cardiovascular support by the use of inotropes when indicated: Early resuscitation.

- Prompt and judicious use of antibiotics to prevent/treat sepsis

Flow phase

- Adequate nutritional support: Minimise period of starvation
- Avoid hypothermia by nursing patient in a warm environment (room temperature)
- Provide adequate analgesia
- Early mobilisation: Prevents hypostatic pneumonia and deep vein thrombosis
- Avoid tissue oedema: Avoid fluid overload both during and after surgery
- Maintenance of a normoglycaemic state with insulin infusion may help to protect the endothelium in critical illness. This may prevent organ failure and death

Peculiarities of the metabolic response to sepsis: The metabolic response to sepsis is quite similar to that of trauma. The basic difference, however, is that the response is relatively exaggerated in sepsis when compared to uncomplicated trauma. The situation is made worse when sepsis complicates an already existing traumatic situation. In addition, unlike in trauma, there is defective ketogenesis in the presence of sepsis. The latter also affects the physiology of the hepatocyte mitochondria resulting in a reduction of aerobic metabolism of both glucose and fatty acids. This engenders anaerobic metabolism of glucose resulting in the production of lactic acid. Hypoglycaemia may result if alternative sources of production of glucose such as gluconeogenesis are not enhanced. It should be noted that hypoglycaemia is an extremely poor prognostic factor in sepsis.

CHAPTER FOURTY-TWO

FLUID AND ELECTROLYTE BALANCE IN SURGICAL PRACTICE

In a way, surgical practice may be regarded as a 'game of fluids and electrolytes'. This is for the following reasons

- The body is composed mainly of fluids (70%)
- Surgical patients commonly present with one form of fluid and/or electrolyte anomaly. Hypovolemic shock and dehydration, for example, may be due to loss of fluid or blood while patients with gastric outlet obstruction and intestinal fistulae may present with gross fluid and electrolyte anomalies.
- Surgical procedures are associated with loss of fluid and electrolytes: Blood, third space loss
- Additional fluid and electrolyte losses may result from surgical drains (chest and abdomen) and nasogastric aspiration
- Fluid and electrolyte anomalies need to be corrected before surgery for hatch-free anaesthesia. Hypokalemia, for instance, is not compatible with the use of muscle relaxants
- The neuroendocrine and cytokine response to surgery alter the fluid and electrolyte status of the body. This is important in the management of a patient after surgery
- Knowledge of composition and indications for various intravenous infusions is important for optimal management of patients.

Anatomy of body water

Total body water varies with age, gender, body habitus and disposition of adipose tissue (adipose tissue contains less water than muscle).

- Average adult male (70 kg): Total body fluid is about 42 litres . This comprises an average of 60% of total body weight (range 60% to 65%)
- Average adult female (50 kg): Total body fluid is about 27 litres and averages 55% of total body weight
- Neonates: Total body fluid averages 75% of total body weight
- Infants Total body fluid averages 70% of total body weight
- Elderly: Total body fluid averages 45% of total body weight
- Obesity: Less total body fluid per kg than lean body adult

The total body fluid is divided into two broad compartments: Intracellular and extracellular

- Intracellular: Located within the cells. Comprises of 70% of total body fluids (40% and 30% of total body weight in males and females respectively)
- Extracellular: Located outside the cells. Comprises of 30% of total body fluids. The extracellular fluid compartment is comprised of the following component.
- Interstitial: Between cells
- Intravascular: Within the vascular system (composed mainly of plasma)

- Transcellular: Fluid contained in body cavities – cerebrospinal, ocular, synovial, peritoneal and pleural fluids

The osmolarity of a fluid is defined as its capacity to generate osmotic pressure. This is dependent on the concentration of osmotically active substances. The main electrolytes in extracellular fluid are sodium and chloride while potassium is the main electrolyte in the intracellular fluid. Other less osmotically active electrolytes in the latter are magnesium, phosphate and sulphate.

The implication is that the volume of the extracellular fluid is determined mainly by the amount of sodium and chloride while that of the intracellular fluid is determined mainly by potassium. The sodium-potassium pump tends to maintain the gradient of the main electrolytes in both compartments.

Dynamics of body fluid

Methods of fluid input by the body: Sensible and insensible

- Sensible means of fluid input: Oral fluids, solid food
- Insensible means of fluid input: Water from oxidation of food (about 300 mls daily)

Methods of fluid loss by the body

- Sensible means of fluid loss: Urine, intestine and sweat
- Insensible means of fluid loss: Lungs (about 400 mls daily) and skin (about 500 mls daily). Average body insensible loss is about 700 mls daily

Abnormal loss of fluid may occur through the following

- Excessive sweating: 1.0 to 1.5 litres of fluid may be lost
- High fever: 100mls/degree of fever/day
- Tracheostomy with unhumidified air: More than 1.5 litres daily
- Exposed wound surface: Fluid loss from severe burns may be as much as 0.5 to 3 litres of fluid daily
- Third space loss as in acute pancreatitis and intestinal obstruction
- Wound drains
- Nasogastric aspirate: May be up to 1.5 litres daily in severe intestinal obstruction
- Intestinal fistulae such as enterocutaneous fistula
- The diuretic phase of acute renal failure will result in excess loss through the urine

The principle of fluid management is that the total fluid loss should be replaced. This is the sum of normal daily requirement and loss through abnormal channels.

MANAGEMENT OF VOLUME CHANGES

Volume deficit (Hypovolemia): Causes include

- Surgery/anaesthesia
- Gastrointestinal losses: Diarrhoea, vomiting, naso-gastric aspiration
- Drainage from fistulae
- Soft tissue injuries
- Burns
- Third space loss: Peritonitis and intestinal obstruction

Clinical features include thirst, decreased skin turgor, and dryness of the lips and tongue. Others are sunken eyeballs and features of shock.

Treatment: This is discussed in the management of shock. As pointed out earlier, estimation of volume deficit should take the following into consideration

- Maintenance of daily requirement
- Estimate of existing deficit
- Replacement of continuing excessive losses
- Effect of associated conditions: Stress, surgery, age, size, cardiac and renal functions
-

Volume excess (Hypervolemia):

Common cause is iatrogenic from fluid overload. Clinical features include pitting oedema (including anasarca), elevated jugular venous pressure, tachypnoea and other features of pulmonary oedema.

Treatment includes

- Diuretic therapy
- Guarded restriction of fluid therapy and adequate monitoring by way of CVP and PWAP
- Haemodialysis or peritoneal dialysis particularly in severe renal impairment

MANAGEMENT OF CHANGES IN CONCENTRATION

Sodium

Normal serum level of sodium is between 135 and 145 mEq/litre. Excess sodium is excreted through the urine. Normal daily salt intake is 3 – 5 grams of NaCl. Hyponatraemia is said to occur when the level is below 135 mEq/litre and hypernatremia occurs when it is above 145 mEq/litre

Hyponatraemia: Clinical features are

- Musculoskeletal: muscle twitches
- CNS: Confusion, lethargy, stupor, headache, seizure and coma
- GIT: Nausea and vomiting

Estimated sodium deficit 'y' is calculated with the formula:

$(145 - X) \times 60/100 \times \text{body weight}$; where X is the serum sodium level. Correction is aimed at replacing half of this deficit

Hypernatremia

Clinical features include

- CNS: lethargy, restlessness, irritability and ataxia
- Musculoskeletal: Weakness
- Renal: Oliguria
- Metabolic: Fever

Sodium excess 'y' is calculated by the formula:

$(X - 145) \times 60/100 \times \text{body weight}$; where X is the serum sodium level.

Correction is by infusion of 5% dextrose solution. Volume to be infused is calculated as deficit 'y'/154

Potassium

Normal serum level of potassium is 3.5 – 5.1 mEq/litre. It is mainly excreted in the urine. Intracellular potassium is released into the extracellular space in response to severe injury or surgical stress, acidosis and catabolic states. Normal daily intake of potassium is 50 – 100 mEq/day. Hypokalemia is said to occur when the potassium level is less than 3.5 mEq/litre while hyperkalemia connotes a level higher than 5.1 mEq/litre.

Hypokalemia: Aetiological factors are

- Inadequate intake
- Administration of potassium-free intravenous infusions
- Total parenteral nutrition
- Excessive loss of potassium: Gastrointestinal, medications (diuretics), hyperaldosteronism
- Intracellular shift: Insulin therapy, metabolic alkalosis (potassium level decreases by 0.3 mEq/litre for every 0.1 increase in pH above normal)

Clinical features are

- GI: Paralytic ileus, constipation
- Neuromuscular: fatigue, weakness, paralysis, diminished reflexes
- CVS: Arrhythmias, U-waves, T-wave flattening, ST segment changes
- Renal: Polyuria and polydipsia
- Tissues: Dry, sticky mucous membranes; red swollen tongue; decreased saliva and tears

Treatment:

- Replacement of estimated deficits
- Oral supplements potassium rich foods for mild deficits
- Intravenous replacement for moderate to severe deficits. Ensure that patient is making adequate urine before commencing parenteral replacement. Potassium chloride: 10 mEq/litre/hour iv (peripherally) or 20 mEq/litre/hour iv (centrally) Should not exceed 40 mEq/litre
- Correct existing alkalosis
- Correct any identified predisposing cause: Diuretics, fluid losses

Hypokalemia is best prevented by the administration of adequate potassium supplements to patients at risk.

Hyperkalemia:

Serum potassium > 5.1 mEq/litre

Aetiological factors include

- High input: Blood transfusion, potassium supplementation
- Rapid rise of extracellular fluid osmolality: Hyperglycaemia, mannitol
- High level of endogenous release: Crush injury, rhabdomyolysis, gastrointestinal haemorrhage
- Impaired excretion of potassium: Renal insufficiency/failure
- Increased release: Acidosis

Clinical features of hyperkalemia

- GI: Nausea, vomiting, diarrhea
- Neuromuscular: weakness, paralysis
- CVS: Arrhythmias, cardiac arrest. ECG changes are - peaked T, flattened P, widened QRS complex and prolonged PR interval (primary block). It may also result in ventricular fibrillation

Treatment of hyperkalemia

- Discontinue exogenous potassium administration
- Treat underlying cause
- Reverse cardiotoxic effects: With ECG monitoring, administer 1 gram of 10% calcium gluconate
- Administration of glucose/insulin drip
- Rapid alkalinisation of the extracellular fluid; lactate/bicarbonate, cation exchange resins
- Peritoneal dialysis/haemodialysis

Calcium

Serum calcium level is 9 – 11 mg%. Most of the body calcium (1000 – 2000 gm) is contained in bone. Daily intake of calcium is 1 – 3 gm. About 40 – 60% of calcium is bound to albumin. The ionized fraction is responsible for neuromuscular stability. Calcium excretion is via the gastrointestinal system.

Hypocalcaemia: Serum calcium level < 8 mg%.

Aetiological factors include

- Hypoparathyroidism
- Acute pancreatitis
- Acute and chronic renal failure
- Massive soft tissue infection such as necrotizing fascitis
- Pancreatic and small bowel fistula
- Massive blood transfusion

Clinical features of hypocalcaemia are

- Paraesthesia
- Tetany
- Numbness and tingling sensation of the circumoral region and lips
- Carpopedal spasm and convulsions
- Chvostek and Troussseau's signs

Treatment of hypocalcaemia

- Address any underlying cause
- Oral supplement in mild cases
- Intravenous replacement in moderate to severe cases: Give calcium gluconate/lactate slowly under ECG monitoring. For blood transfusion-induced hypocalcaemia, give 10 mls of calcium chloride for every three pints of blood.

Hypercalcaemia: Serum calcium level above 11 mg%

Aetiological factors are

- Hyperparathyroidism
- Milk-alkaline syndrome
- Hypervitaminosis D
- Cancers: some cancers are known to secrete parathyroid hormone-like peptides
- Sarcoidosis

Clinical features of hypercalcaemia are

- General: Fatigue, lassitude, weakness, somnolence, coma
- Thirst and polydipsia
- CVS: Hypertension, polydypsia
- GIT: Constipation, anorexia
- Genitourinary: nephrolithiasis

Treatment of hypocalcaemia

- Fluid replacement
- Administer diuretics (Lasix)
- Administer oral or intravenous inorganic phosphate to inhibit resorption of calcium. This is contraindicated in chronic renal failure. The latter may be given sodium phosphate
- Other medications of use: Steroids, mithramycin

Magnesium

Total body magnesium is 2000 mEq. Half of this is in bone

Daily intake of magnesium: 240 mg

Serum magnesium level: 1.5 – 2.4 mEq

Causes of hypomagnesemia: Starvation, malabsorption and excess loss from GIT. It is a known complication of total parenteral nutrition.

Features of hypomagnesaemia: Similar presentation with hypocalcaemia

COMPOSITION CHANGES

This involves changes in acid-base balance

- Respiratory acidosis
- Respiratory alkalosis
- Metabolic acidosis
- Metabolic alkalosis
-

Respiratory acidosis:

This is primarily due to decreased alveolar respiration resulting in retention of carbon dioxide.

Causes include

- Depression of the respiratory centre (overdosage of narcotics)
- Injuries to the central nervous system
- Pulmonary diseases

The body tries to compensate by renal retention of bicarbonate and increased excretion of acid salts. There is also increased formation of ammonia and chloride shift into the cells.

Treatment involves restoration of the alveolar ventilation to normal

Respiratory alkalosis

This is due to increased alveolar respiration resulting in loss of carbon dioxide. Causes include

- Hyperventilation
- Pain
- Assisted ventilation

The body compensates by renal excretion of bicarbonate, retention of acid salts and decreased production of ammonia

Treatment of respiratory alkalosis involves the following

- Address the underlying cause including alleviation of pain, emotional distress and hypoxia
- Rebreathing and increasing the airway dead space are employed when appropriate

Metabolic acidosis

The fundamental problem is the retention of fixed acids and loss of base bicarbonate. The aetiology includes

- Diabetes mellitus
- Loss from small bowel: Diarrhoea, small bowel fistulas

The body compensates by increasing the rate and depth of respiration

Treatment regimen is as follows

- Remove underlying cause
- Volume replacement for adequate tissue perfusion. This will rapidly reverse lactic acidosis
- Sodium bicarbonate may be necessary in order to forestall cardiac arrest. The replacement formula is $(24 - \text{plasma HCO}_3) \times \text{estimated extracellular volume}$

Metabolic alkalosis

This is basically due to loss of fixed acids and gain of base bicarbonate. The causes of metabolic alkalosis include

- Gastric outlet obstruction
- Diuretic therapy
- Excess intake of bicarbonate
- Hypokalemia

The body tries to compensate by reducing the rate and depth of breathing

Treatment of metabolic alkalosis

- Adequate fluid replacement with normal saline is usually adequate to reverse the fluid and electrolyte derangement
- Administer potassium chloride to correct hypokalemia

Mixed acid – base disturbances

These are difficult to diagnose. The respective effects of acid and base tend to cancel each other and result in minimal changes in pH. Respiratory acidosis may, however, occur in combination with metabolic acidosis in septicaemia and cardiopulmonary arrest

INTRAVENOUS INFUSIONS

Intravenous infusions form a good part of parenteral medication. The word 'parenteral' is a compound word of Greek origin: para (other than) and enteron (intestine). The commonly used intravenous infusions can be categorized according to their clinical applications

- Maintenance fluids: 5% dextrose, 4.3% dextrose with 0.18 normal saline, 5% dextrose with half strength normal saline
- Replacement fluids: Subdivided into crystalloids and colloids
 - Crystalloids (electrolytes): Normal saline and Ringer's lactate
 - Colloids (Large molecules) : Dextran, haemacel, hetastarch
- Special fluids: Mannitol, sodium bicarbonate, 50% dextrose

General indications for intravenous infusion

- When oral intake is not possible owing to underlying ailment
- Severe fluid loss: Vomiting, diarrhea, dehydration, shock
- As a vehicle for the administration of some medications
- To aid nutrition by supplying calories, and electrolytes
- As an aid in the management of critical situations such as poisoning

Advantages of intravenous infusion

- Immediate response as it gains direct entry into the vascular system
- Prompt correction of haemodynamic status
- Administration can be controlled by close monitoring

Disadvantages of intravenous infusion

- Requires strict aseptic conditions
- Requires skilled supervision
- Requires proper selection of fluids; improper selection is dangerous
- Requires proper calibration of volume; administration of improper volume is dangerous
- Requires proper technique; improper technique will result in complications

General complications of Intravenous infusion

- Local: Haematoma, thrombophlebitis
- Systemic: Circulatory overload (pulmonary oedema), rigors and air embolism,
- Fluid contamination: May result in septicaemia
- Others: Human error, infusion set and catheter problems.

Applications of specific intravenous fluids

1.5% dextrose in water:

Supplies 50 grams of glucose and 170 kcal of energy per litre

Pharmacological application: Corrects dehydration and supplies energy

Indications for 5% dextrose in water

- Prevention and treatment of dehydration
- Fluid replacement: Both preoperative and postoperative

- Prevention of ketosis in starvation, vomiting and diarrhea
- Intravenous administration of various drugs
- Correction of hypernatremia
- Adequate glucose infusion protects the liver against toxic substances

Contraindications to the administration of 5% dextrose infusion include

- Cerebral oedema
- Acute ischemic stroke
- Water intoxication
- Hypovolemic shock
- Hyponatremia
- Severe hyperglycaemia
- Uncontrolled diabetes mellitus

2 Normal saline (Isotonic saline – 0.9% NS)

Made up of Na – 154 mEq, and Cl – 154 mEq

Pharmacological basis

- Provides major extracellular fluid electrolytes
- Corrects both water and electrolyte deficit
- Increases the intravascular volume substantially

Indications for administration of normal saline

- Clinical situations of both water and salt depletion: Diarrhoea, vomiting, excessive diuresis
- Hypovolemic shock
- Dehydration with alkalosis: Gastric outlet obstruction
- Hyponatremia with severe salt depletion
- Initial fluid therapy in the management of diabetic ketoacidosis
- Hypercalcaemia
- Fluid challenge in pre-renal failure
- Irrigation of body cavities: Peritoneum, bladder irrigation post-prostatectomy
- As a vehicle for some drugs

Contraindications to the administration of normal saline

- Pre-eclampsia
- Medical conditions: Congestive cardiac failure, renal diseases, cirrhosis of the liver
- Dehydration with severe hypokalemia
- Large volume may result in hyperchloraemic acidosis

Pathophysiology of hyperchloraemic acidosis following saline infusion: This results from excessive administration of chloride in the course of resuscitation with normal saline. The major determinant of hydrogen ion concentration is the strong ion difference (SID) in the body. A normal SID (42 – 46 mmol/litre) is obtained by adding together the concentration of the main cation in solution (Na, K, Ca, Mg) and subtracting the concentration of the main anions (Cl, lactate). A fall in SID level is associated with metabolic acidosis. This can be precipitated by a large volume of saline because renal excretion of Na occurs in preference

to Cl and H. The cause of metabolic acidosis may therefore be erroneously attributed to tissue hypoperfusion and cellular hypoxaemia. Acidemia may then be treated with liberal volume of normal saline infusion. This may worsen rather than correct the acidosis

2 Dextrose-normal saline infusion

This supplies major electrolytes of the extracellular fluid, energy and fluid to correct dehydration.

Indications for the administration of dextrose/normal saline

- Hypovolemia with salt depletion
- Correction of alkalosis and hypochloremia due to vomiting, or nasogastric tube aspiration

Contraindication: Anasarca due to cardiac, renal or hepatic pathology

3 4.3% dextrose/0.18 normal saline and dextrose with half strength normal saline.

This solution contains more water than salt

Indications for their use are

- Fluid management in children and the elderly: Their kidneys are not capable of handling much salt load
- Maintenance fluid of choice immediately after surgery
- Treatment of hypernatremia

Contraindications: Hyponatremia, severe dehydration

4 Ringer's lactate

Ringer's lactate is regarded as the most physiological intravenous infusion. In addition to expanding the intravascular volume, it plays a role as a buffering agent. This is accomplished by the liver which metabolises the lactate component to bicarbonate. The following are the indications for the use of Ringer's lactate.

- Severe hypovolemia
- Fluid replacement in burns and after surgery
- Diarrhoea-induced dehydration in children
- Diabetic ketoacidosis: Provides water, corrects metabolic acidosis and supplies potassium

Contraindications

- Vomiting or nasogastric tube-induced vomiting: Not used in gastric outlet obstruction
- Should not be administered simultaneously with blood
- Liver disease
- Severe congestive cardiac failure

General effects of administering large volume of crystalloid infusion

- Extravascular accumulation in skin, connective tissue, lungs and kidneys
- Delayed healing of anastomosis
- Inhibition of gastrointestinal motility
- Large volume, rapid infusion may result in hypercoagulability

COLLOIDS

These are large molecular weight substances that largely remain in the intravascular compartment where they play a vital role in the generation of oncotic pressure. In this regard, they are more potent than crystalloids. As regards their application in maintaining haemodynamic stability following blood loss, the following subsists

- 1 ml blood loss : 1ml colloid replacement : 3 mls crystalloid replacement

There are various types of crystalloids: Dextran, haemacel, albumin

1 Dextrans

They are glucose polymers produced by the bacterial action of leuconostoc mesenteroides. There are two forms

- Dextran 40 (molecular weight 40,000)
- Dextran 70 (molecular weight: 70,000)

Pharmacology

- Rapidly expand the intravascular volume
- Have a high renal excretion
- Antithrombotic: Inhibit platelet aggregation
- Improve the microcirculation and are therefore useful in the prevention of thromboembolism

Indications

- Correction of hypovolemia
- Prophylaxis of deep venous thrombosis (including postoperative DVT)
- To improve blood flow including microcirculation in impending vascular gangrene

Side effects of dextrans

- Interfere with grouping and cross-matching: it is therefore advised that blood samples should be taken prior to administration of dextrans
- Hypersensitivity reaction
- Acute renal failure

Contraindications to dextrans

- Acute renal failure
- Congestive cardiac failure: May cause circulatory overload
- Severe dehydration
- Bleeding disorders
- Enhance the anticoagulant effect of heparin
- Hypersensitivity to dextran

Gelatin polymers (Haemacel)

This is a combination of degraded polymers and electrolytes

Pharmacological advantages

- Unlike dextrans, they do not interfere with coagulation and blood grouping
- They remain in the blood for a long period (4 to 5 hours)
- Rapidly expand the blood volume: Infusion of 1 litre expands the blood volume by 50%

Indications for haemacel

- Rapid volume expansion in hypovolemic shock
- Employed as a volume preloading agent in general anaesthesia
- Employed in the priming of heart-lung machine

Side effects of haemacel

- Hypersensitivity reaction
- Bronchospasm
- Should not be used with citrated blood

3 Hydroxyethyl starch (Hetzastarch)

Pharmacological advantages

- Generates a high colloidal osmotic pressure
- Greater volume expansion compared to others
- Non-antigenic
- Does not interfere with blood grouping
- Preserves intestinal microvascular perfusion in the presence of endotoxaemia

Side effects

- Causes an elevation of the serum amylase level. This may last for 5 days
- Affects coagulation by prolonging PT, PTT, and bleeding time by lowering fibrinogen level.
It equally decreases the aggregation of platelets

Contraindications to hetastarch

- Bleeding disorders
- Congestive cardiac failure
- Impaired renal function

Special fluids

- Injection potassium chloride (KCl): 10 ml amp contains 20 mEq of KCl
- 25% and 50% glucose in 50 ml pack: Indicated in hypoglycaemia
- Injection sodium bicarbonate (NaHCO₃): 25 ml amp contains 22.5 mEq Na and 22.5 mEq HCO₃ per litre. Indicated in metabolic acidosis
- Mannitol: Comes in 10% and 20% forms. It is an osmotic diuretic
- Isolyte: Rich in potassium. Indicated in fluid and electrolyte loss (hypokalemic metabolic alkalosis)

Resuscitation with intravenous infusion

Crystalloids are recommended as the fluid of choice in the initial phase of resuscitation from haemorrhagic shock. There is no proven advantage of colloids over crystalloids in the management of severe trauma or burns.

Methods of calculation of fluid transfusion rate

- 1 Routine infusion set:
- 15 drops per minute = 1 ml
- 'Rule of ten' for fluid calculation for 24 hours
Volume of iv fluid in litres per 24 hours X 10 = drop rate per minute
- Drop rate calculation for any parameter

Volume to be infused in mls/Duration of infusion x 4 = drops/minute

2 Microdrip infusion set

- 60 drops = 1 ml
- No of microdrips/minute = Volume in ml/hour

PERIOPERATIVE FLUID THERAPY

Proper perioperative fluid management is important in patient management as it helps to reduce the morbidity and mortality associated with surgical procedures. This is based on the physiological reaction to acute stress

- Sympathetic stimuli: Tachycardia and vasoconstriction
 - Release of antidiuretic hormone (lasts for 2 to 3 days): Water retention
 - Adrenal release of aldosterone and corticosteroids (lasts for about 48 hours): Sodium retention and potassium depletion. The latter results in the movement of potassium from the intracellular to the extracellular compartment
 - Surgical stress may cause direct damage to the kidneys, brain and lungs
- The following factors compound the perioperative fluid status of the patient
- Preoperative nil per oral regimen : The fluid deficit requires replacement
 - Pre-, intra-and postoperative fluid loss need to be replaced

Based on the above factors, the following are the guidelines for fluid management during the perioperative period

A Preoperative fluid therapy

- Correct existing hypovolemia as this may adversely affect the physiological compensatory mechanisms when patient is under general anaesthesia
- Correct anaemia: Blood transfusion should be carried out not later than 48 hours before surgery
- Correct any observed electrolyte anomaly such as hypokalemia

B Intraoperative fluid therapy

- Correct fluid deficit due to starvation: The infusion for correction is 5% dextrose in water and the amount of fluid is: Duration of starvation in hours X 2 X weight in kg
- Maintenance volume during the intraoperative period: Again this is given as 5% dextrose in water and the amount of fluid is calculated as: Duration of surgery in hours X 2 X weight in kg
- Correction of intraoperative fluid loss: Takes into cognizance the various means of fluid loss at surgery
 - Surgical sponge
 - Suction container
 - Third space loss

The degree of intraoperative fluid loss has been related to the degree of trauma

- Least trauma: Negligible loss
- Minimal trauma: 4 ml/kg/hour

- Moderate trauma: 6 ml/kg/hour
- Severe trauma: 10 ml/kg/hour

Blood loss is managed by the administration of normal saline infusion at the ratio of 3 volumes of normal saline to 1 volume of blood loss. Blood transfusion should be carried out when necessary.

C Postoperative fluid management

Owing to the salt and water retention during the immediate postoperative period, care should be exercised to avoid overloading of the circulation

- First 24 hours after surgery: Patient should receive the basic maintenance fluid. Should receive an average of two litres of intravenous infusion with restriction of salt input. Fluid input may be in the form of either 2 litres of 5% dextrose in water or a combination of 1.5 litres of 5% dextrose in water and 500 mls of normal saline
- Second postoperative day: Should receive a total of 3 litres of fluid comprising of 2 litres of 5% dextrose in water and 1 litre of normal saline. Blood samples should be taken for estimation of packed cell volume and electrolytes/urea/creatinine if necessary
- Third postoperative day: Fluid regimen as in second postoperative day. If indicated, potassium chloride, 40 to 60 mEq/day may be added
- In addition to the regimen advocated, any additional fluid loss should be replaced. This includes effluent from drains (abdominal, pleural and wound) as well as nasogastric aspirate. It is important to maintain an input-output chart and ensure that the input balances the perceived output. As pointed out earlier, cognizance should be taken of the body temperature as fever is associated with an increase the amount of insensible fluid loss.

CHAPTER FOURTY-THREE

SHOCK; SYSTEMIC INFLAMMATORY RESPONSE SYNDROME

Shock is a clinical state arising from inadequate tissue perfusion resulting in cellular hypoxia and metabolic derangement. A good knowledge of the proper management of shock is important because if not well handled, shock may progress to cellular injury/cell death. Uncorrected, the resultant cellular injury/death may lead to systemic inflammatory response syndrome (SIRS) and multiple organ dysfunction syndrome (MODS). The haemodynamic system has been likened to a water reticulating system consisting of a pump (heart), the pipes (vessels) and the fluid (intravascular fluid). For maintenance of normal perfusion, all components of the system need to be intact.

Pathophysiology of shock

The ineffective blood supply to the tissues results in hypoxia. Anaerobic metabolism sets in, resulting in the formation of lactic acid and metabolic acidosis. Failure of the Na-K pump mechanism arises from depletion of the cellular glucose store. This results in the disintegration of the cell and the release of autodigestive enzymes.

At the microvascular level, hypoxia and acidosis activate the immune and coagulation systems and cause the release of oxygen free radicals and cytokine. These in turn damage the cell membrane resulting in an increased permeability of the capillary endothelium. The end-result is extravasation of fluid. Microthrombi are formed and the resultant consumption of fibrinogen may result in disseminated intravascular coagulation (DIC).

Systemic effects of shock

- **Cardiovascular:** Diminished venous return results in decreased cardiac output. The baroreceptors are activated and in response release catecholamines. The latter are responsible for the tachycardia and generalised vasoconstriction usually associated with shock. This explains the cold, clammy extremities and the increased sweating that are associated with shock. The increased sweating results from stimulation of the sweat glands by catecholamines
- **Respiratory:** Metabolic acidosis and increased sympathetic activity result in tachypnoea and hyperventilation.
- **Renal:** There is reduction in the urinary output due to a diminution in the renal blood flow. The renin-angiotensin-aldosterone system is activated resulting in an enhanced reabsorption of water and sodium.
- **Endocrine:** The reduced blood flow results in the release of antidiuretic hormone (ADH) and cortisol from the hypothalamus and the adrenal cortex respectively. In the renal tubules, while both hormones enhance the reabsorption of water, cortisol in addition promotes the reabsorption of sodium.

Types of shock

Hypovolemic shock: It is due to a diminution in the circulating blood volume. May result either from loss of whole blood (haemorrhagic) or purely due to loss of fluids. Either of them could be revealed when loss is external event or concealed when loss is internal (occult).

- External haemorrhage: Source of bleeding is obvious such as crush injury, laceration or gastrointestinal haemorrhage (haematemesis and melaena).
- Internal haemorrhage: Source of bleeding is concealed within the body tissue or cavity. Examples include haemoperitoneum resulting from ruptured ectopic pregnancy and injury to an intra-abdominal viscous such as the spleen and liver. A fracture of the femur may conceal as much as 1.5 litres of blood without any appreciable change in the circumference of the thigh.

Severe haemorrhage will result in hypovolemic shock. The resultant lactic acidosis coupled with hypothermia due to a reduction in the muscular flow will result in coagulopathy. Hypothermia is made worse by the infusion of cold blood and intravenous fluids in a bid to compensate for blood loss. It is important to note that most commonly used crystalloids are acidic in nature. Normal saline, for instance, has a pH of 6.7. The triad of acidosis, hypothermia and coagulopathy is therefore lethal. The importance of warming up stored blood and indeed intravenous infusion before administration to a patient in shock cannot therefore be overemphasised.

- External fluid loss: Diarrhoea, vomiting, burns etc
- Internal fluid loss: May be sequestered within a body cavity such as the peritoneum as in acute pancreatitis or in a hollow organ such as the gut in intestinal obstruction

Cardiogenic shock: This is categorised into intrinsic and extrinsic depending on the causative factor.

- Intrinsic: Due to an intrinsic inability of the heart to pump blood to the tissues. Causes include myocardial infarction, arrhythmias, valvular heart disease and cardiomyopathy.
- Extrinsic: Also referred to as obstructive shock. It is due to conditions that limit the function of the heart by extrinsic pressure. For instance in tension pneumothorax there is unilateral increase of the intrathoracic pressure and shifting of the mediastinum to the contralateral side. This results in compression of the vena cava and the right side of the heart. Similarly, cardiac tamponade tends to limit the filling of the right side of the heart by compressing the thin-walled right ventricle and shifting the septum toward the more robust left ventricle. This equally makes worse the already decreased the left ventricular end-diastolic volume as a result of external pressure from cardiac tamponade.

Distributive shock: In this form of shock, there is no apparent loss of fluid or blood from the body. Distributive shock is characterised by generalised vasodilatation especially at the microvascular level. Hypotension is the resultant effect of an increase in vascular capacity without a corresponding increase in the intravascular fluid volume. The following forms of shock fall into this category

- **Anaphylactic shock:** This is a hypersensitivity reaction to various agents such as animal protein (such as ATS) and drugs (such as penicillin). It may also arise from the administration of blood products. In addition to the generalised vasodilatation, there is also an increase in capillary permeability. The mast cells present in the antigen-antibody reaction release histamine and prostaglandin. This allergic reaction manifests clinically as urticaria, wheezing, choking sensation and loss of consciousness in association with shock..
- **Neurogenic shock (vaso-vagal syndrome):** Follows fear, anxiety, fright or prolonged standing. May be iatrogenic as in complicated spinal anaesthesia. Acute injury to the cervical or upper thoracic spinal cord may result in the loss of autonomic tone and produce acute hypotension. A major characteristic of neurogenic shock is presence of a normal or a low heart rate in the face of obvious hypotension in a patient who is not on beta-blockers. The relative bradycardia is due to cervical sympathetic denervation.
- **Septic shock:** This is also referred to as endotoxic shock. It arises from moderate to severe infection or tissue damage. Any type of infection affecting any organ of the body may lead to septic shock. The worst culprit, however, is gram-negative septicaemia. Gram-positive bacilli, viruses and fungi may also be responsible. The endotoxin released from the cell wall activates the release of lethal mediating factors. These include tumour necrotic factor- alpha, interleukin and cytokine interleukin 10 (IL10). Others are histamine, serotonin, platelet activating factor and prostaglandin. The latter group will result in vasodilation, increased permeability of the blood vessels and disseminated intravascular coagulation (DIC). This enhanced inflammatory response affects the vital organs. Untreated, it results in Multiple Organ Dysfunction Syndrome (MODS), Multiple Organ Failure (MOF) and finally death.

Mixed shock: This is an infrequently used term. It refers to a patient whose clinical assessment reflects the features of more than one group of shock. It can pose a diagnostic dilemma. A typical example is the patient with advanced and complicated intestinal obstruction. In addition to hypovolemic shock resulting from vomiting and third space loss, there is also a component of septic shock that results from the translocation of bacteria from the intestinal lumen.

- Clinical features of shock:** These can be deduced from the pathophysiology
- Deterioration in the level of consciousness: reduction in cerebral blood flow
 - Cold clammy extremities, sweating: Increased sympathetic activity
 - Tachycardia: increased sympathetic activity
 - Hypotension: Reduced cardiac output due to diminution in venous return
 - Reduced urinary output: Reduction in renal blood flow

Management of shock

Initial resuscitation: After a quick history, the aim is to resuscitate the patient as follows

- Set up an intravenous line with a wide-bore cannula. Before hanging up the infusion, take blood samples for full blood count, electrolytes/urea/creatinine, PO₂ and PCO₂. Blood sample is also sent for grouping and cross-matching. Crystalloids such as normal saline and Ringer's lactate are the most commonly used resuscitation fluids. The ratio of fluid to blood loss is aimed at 3:1. Plasma expanders (colloids) such as human albumin, haemacel, fresh frozen plasma (FFP) and dextran may be used in a ratio of 1:1.
- Pass a urethral catheter to monitor the urinary output. The latter should mirror the degree of hydration of the patient and therefore reflects the effectiveness or otherwise of the resuscitation. There are, however, some potential drawbacks. Hyperglycaemic patients may present with false high urine output triggered by glucose spillage in the urine. On the flip side, acute renal insufficiency following severe or prolonged shock may result in low urinary output.
- Elevate of the lower limb of the patient on bed to enhance venous return
- Administer oxygen to maintain the PO₂ between 80 and 100mm Hg.
- Sodium bicarbonate: Currently it is not so much in use as it is believed that the acidosis which it aims to correct is naturally taken care of by improving tissue perfusion.

When the patient's condition improves, a full history of the ailment is taken followed by a thorough clinical examination. This is aimed at determining the cause and category of the shock. The underlying cause should be addressed promptly. For example, a 'leaking tap' by way of ruptured ectopic pregnancy or a ruptured spleen should be addressed as soon as the patient is fit enough for surgery. This is because the patient's condition cannot be optimal until the 'leaking tap is locked'. Similarly, a patient with a perforated duodenal ulcer who may present with clinical features of both hypovolemic and septic shock will require intravenous infusion, preoperative antibiotics and surgical closure of the perforation. On the other hand, a patient with acute pancreatitis may not require surgery, but will benefit from adequate resuscitation with intravenous fluids.

Management peculiarities of the various types of shock

Hypovolemic shock: As stated above, initial resuscitation is with crystalloids. Shock due to fluid loss does not require blood transfusion. Blood transfusion in haemorrhagic shock is, however, dictated by the severity of haemorrhage and the comorbidity of the patient. In this respect, there are four classes

- Class I haemorrhage: Up to 15% blood loss - does not require blood transfusion
- Class II haemorrhage: 15-30% blood volume loss - may require blood transfusion
- Class III haemorrhage: 30-40% loss with marked features of shock – requires blood transfusion
- Class IV haemorrhage: More than 40% blood loss – requires rapid blood transfusion

Septic shock: In addition to the above general resuscitative measures, patient may require the following

- Intravenous antibiotics to cover both aerobic and anaerobic organisms
- Corticosteroids: They inhibit further synthesis of TNF-alpha and release of prostaglandins, prostacyclines and other mediating factors. Steroids should be used with caution as they may mask some of the symptoms and cause hyperglycaemia.
- Antioxidants such as allopurinol and vitamin C may reduce tissue damage by free radicals.
- Address the underlying focus of infection.

Neurogenic shock: Lie the patient flat as an emergency measure. Intravenous infusion and pressor agents may be necessary.

Cardiogenic shock (Intrinsic and extrinsic)

- Gentle fluid resuscitation with addition of an inotrope for the intrinsic variety
- Address the underlying problem such as myocardial infarction and pulmonary embolism in the case of intrinsic cardiogenic shock.
- Management of the extrinsic causes may include pericardiocentesis and chest tube insertion for pericardial effusion and pneumothorax respectively.

Anaphylactic shock: May require the administration of adrenaline, antihistamine, hydrocortisone or aminophylline if there is associated bronchospasm. Rapid fluid replacement may be necessary. The mainstay of management of anaphylactic shock is prompt diagnosis and administration of adrenaline.

Monitoring of a patient in shock

- ECG
- Pulse oximetry
- Blood pressure
- Hourly urinary output
- Others: Capillary refill, central venous pressure, cardiac output, base deficit and serum lactate

Pitfalls in monitoring of a patient in shock

- Capillary refill: May be normal in septic shock
- Tachycardia: May not be apparent in patients on beta-blockers and those with a pacemaker. There may be a paradoxical bradycardia in young people with minimal tissue damage.
- Hypotension: May be one of the last signs to manifest especially in children and young adults. Hypertensive patients and those on beta-blockers may present with apparently 'normal' blood pressure even in the face of severe shock.
- Urinary output: In the presence of shock, the urinary output may show a normal output in a patient with hyperglycaemia and a low output in a patient who already has acute renal insufficiency.

Severity of shock: In the face of shock, physiological mechanisms are put in place to ensure that the vital organs have adequate supply of oxygen and nutrients. This, coupled with resuscitative measures, gradually result in recovery of the systems. This is referred to as compensated shock. A situation may arise, however, when recovery is no longer possible either because of late intervention or due to inadequate resuscitation. With the onset of multiple organ failure (MOF), recovery becomes unlikely. This is referred to as decompensated shock.

SYSTEMIC INFLAMMATORY RESPONSE SYNDROME (SIRS) AND MULTIPLE ORGAN DYSFUNCTION SYNDROME (MODS)

Severe shock will result in severe inflammatory reaction in the tissues. This is most pronounced in septic shock. The mediating factors include tumour necrotic factor- alpha, and Interleukin-1B. They stimulate the release of prostaglandin and thromboxane A2 from polymorphonuclear and phagocytic cells. If not well managed, SIRS may culminate in Multiple Organ Dysfunction Syndrome (MODS). The following definitions are germane for a good understanding of SIRS in particular and sepsis in general.

SIRS: The diagnosis requires the presence of two or more of the following:

- Temperature > 38 degrees or < 36 degrees (rectal)
- Tachycardia: Heart rate > 90 beats/minute in a patient not on beta-blockers
- Respiratory rate > 20 breaths/minute or PO₂ < 32mm Hg
- White cell count > 12,000 mm³ or < 4000/mm³ or greater than 10% immature band forms/HPF

Infection: An inflammatory response to microorganisms or their invasion of normal sterile host tissue

Sepsis: SIRS + confirmed infective process (focus of infection)

Severe sepsis: SIRS + organ dysfunction, hypotension or hypoperfusion

Septic shock: Sepsis with hypotension or hypoperfusion despite adequate fluid resuscitation

Multiple organ dysfunction syndrome (MODS): Alteration of organ function in acute illness such that homeostasis cannot be maintained without intervention. This is the result of prolonged systemic ischemic injury and results in damage to multiple organs. There are two clinical varieties of MODS: primary and secondary. Primary MODS is a direct consequence of a specific organ insult. It causes an early dysfunction of the organ involved. Secondary MODS, on the other hand, is a consequence of the host response to a remote insult. SIRS is more pronounced in secondary MODS. The organ dysfunction in the latter occurs much later than in primary MODS.

Notable organs affected by MODS and their clinical manifestations are:

- Lung: Adult respiratory distress syndrome. This will manifest as hypoxia, hypercapnia and acid-base disturbances
- Kidneys: Acute renal failure will manifest as loss of renal concentrating power, oliguria, fluid overload and electrolyte/acid-base disturbances
- Liver: Acute liver failure. Manifestations include jaundice and encephalopathy. Others are metabolic acidosis and hypoglycaemia
- Haematology: Coagulopathy, anaemia and leucopenia

- Heart: Cardiovascular failure and fall in peripheral resistance. This will manifest as fluid overload, dysrhythmia and metabolic acidosis
- Gastrointestinal system: Manifestations of dysfunction include gastrointestinal haemorrhage, ileus and pancreatitis. Others include cholecystitis (acalculous) and malabsorption.
- Neurological dysfunction will manifest as altered level of consciousness, neuropathy, convulsions and myopathy

In its most severe form, MODS may progress into Multiple System Organ Failure (MSOF)

The following conditions/co-morbid conditions may lead to the development of SIRS and its sequelae

- Extremes of age
- Immunosuppressive conditions: Primary immunosuppressive diseases (such as HIV/AIDS), postsplenectomy and recipient of organ transplantation
- Underlying malignancy
- Immunosuppressive therapy: Cytotoxic chemotherapy, radiotherapy, corticosteroids and azathioprine
- Intercurrent disease: Renal impairment, diabetes mellitus, hepatic impairment/jaundice and respiratory impairment