Pediatric Surgery

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Introduction:

Pediatric Surgery considers the entire growing organism, from conception through adolescence, and emphasizes the commonality of immaturity on all organ systems rather than emphasizing care according to systems or regions.

A large focus is on the care of the neonate, whose heat-loss propensity, limited ventilatory and cardiac reserves, poorly developed immune system, and inability to communicate are just as important as its disease. An example of this is seen in gastroschisis and diaphragmatic hernia, where the neonate's core temperature and limited pulmonary reserve loom large as predictors of success.

There are lesions, such as esophageal atresia and imperforate anus that are seen only in infancy and if not successfully treated do not allow the patient to be treated later in life by surgeons treating only adults.

In later childhood, there are age-specific diseases such as intussusception's, pyloric stenosis and foreign body ingestion and aspirations that must be appreciated and treated by surgeons in training. hydroceles, testicular mal-descent and torsion.

Goals

Upon completion of the Pediatric Surgery rotation, the students will be able to:

1. Demonstrate an understanding of the diagnosis and treatment of common surgical problems seen in childhood, such as abdominal pain, and masses of the groin, scrotum, and neck.
2. Understand the general care of the small growing infant, considering fluid requirements, ventilation, nutritional and drug needs.
3. Understand basic physiologic differences germane to the pediatric surgical patient, specifically fluid and electrolyte requirements, normal vital signs, normal blood volume, and normal urine output.
4. Understand the embryological and genetic origins of some of the more common birth defects such as Hirschsprung's Disease, the VACTERL Syndrome, Malrotation, and Intestinal Atresia.

A. Neonatal Physiologic Characteristics

1. Water metabolism

- ♠ Water represents 70 to 80% of the body weight of the normal neonate and premature baby respectively. Total body water (TBW) varies inversely with fat content, and premature have less fat deposits. TBW is distributed into extracellular fluid (ECF) and intracellular fluid (ICF) compartment.

- ♠ The ECF compartment is one-third the TBW with sodium as principal cation, and chloride and bicarbonate as anions.

- ♠ The ICF compartment is two-third the TBW with potassium the principal cation.

- ♠ The Newborn's metabolic rate is high and extra energy is needed for maintenance of body temperature and growth.

- ♠ A change in body water occurs upon entrance of the fetus to his new extra-uterine existence. There is a gradual decrease in body water and the extracellular fluid compartment with a concomitant increase in the intracellular fluid compartment. This shift is interrupted with a premature birth. The newborn's body surface area is relatively much greater than the adults and heat loss is a major factor.

- ♠ Insensible water loss is from the lung (1/3) and skin (2/3). Transepithelial (skin) water is the major component and decreases with increase in post-natal age. Insensible water loss is affected by gestational age, body temperature (radiant warmers), and phototherapy.

- ♠ Neonatal renal function is generally adequate to meet the needs of the normal full-term infant but may be limited during periods of stress.

♠ Renal characteristics of newborns are a low glomerular filtration rate and concentration ability (limited urea in medullary interticium) which makes them less tolerant to dehydration. The neonate is metabolically active and production of solute to excrete in the urine is high.

♠ The kidney in the newborn can only concentrate to about 400 mOsm/L initially (500-600 mOsm/L the full-term compared to 1200 mOsm/L for an adult), and therefore requires 2-4 cc/kg/hr urine production to clear the renal solute load. The older child needs about 1-2 cc/kg/hr and the adult 0.5-1 cc/kg/hr.

2. Fluid and Electrolytes Concepts

- Cellular energy mediated active transport of electrolytes along membranes is the most important mechanism of achieving and maintaining normal volume and composition of fluid compartments. Infants can retain sodium but cannot excrete excessive sodium.
- Electrolytes requirements of the full-term neonate are: Sodium 2-3 meq/kg/day, potassium 1-2 meq/kg/day, chloride 3-5 meq/kg/day at a rate of fluid of 100cc/kg/24 hrs for the first 10 kg of weight.
- The daily fluid requirements can be approximated too:
  - ☆ premature 120-150cc/kg/24 hrs
  - ☆ neonates (term) 100cc/kg/24 hrs
  - ☆ Infants >10kg 1000cc+ 50cc/kg/24 hrs.

Special needs of preterm babies' fluid therapy are:

- Conservative approach, consider body weight changes, sodium balance and ECF tonicity. They are susceptible to both sodium loss and sodium and volume overloading. High intravenous therapy can lead to patent PDA, bronchopulmonary dysplasia, enterocolitis and intraventricular hemorrhage.

- Impaired ability to excrete a sodium load that can be amplify with surgical stress (progressive renal retention of sodium).

Estimations of daily fluid requirements should take into consideration:

(1) urinary water losses,

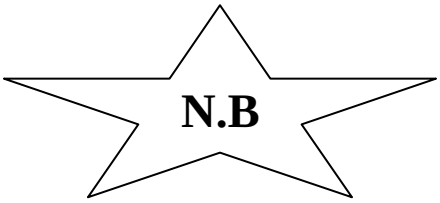
(2) gastrointestinal losses,

(3) insensible water losses, and

(4) Surgical losses (drains).

Blood Volumes estimates of help during surgical blood loss are:

- Premature 85-100 cc/kg
- Full Term 85 cc/kg
- Infant 70-80 cc/kg.



Newborn infants require 100-200 calories/kg/day for normal growth.

This is increased during stress, cold, infection, surgery and trauma. Minimum daily requirement are 2-3 gm/kg of protein, 10-15 gm/kg of carbohydrate and small amount of essential fatty acids.

Pediatric and Neonatal Critical Care Transport

A transport team member will:

- ☆ Undertake transfer.
- ☆ Contact the ambulance service at the earliest opportunity to book the most appropriate ambulance. Correctly use all equipment for each baby transported.
- ☆ Promote effective communications with base hospitals and parents.

Principles of safe transport

• Team composition

- A transport service has organizational and operational teams. Intensive care unit (ICU) staff can be members of either or both. The organizational team is made up of senior ICU staff and is tasked with the setting up and management of the transport service. By convention, the service director is a senior ICU consultant and the service coordinator a senior ICU nurse.
- The operational team is the medic, nurse, paramedic and driver or pilot out on an inter-hospital patient transfer. If a child requires only level 1 care it is possible that a nurse, without a medic, may fetch him or her.
- Critically ill patients require a transport team of at least two personnel, both adequately experienced and accompanied by the ambulance, helicopter or airplane staff. The team leader will usually be a doctor, although advanced neonatal nurse practitioners are increasingly leading neonatal transports.
- The seniority or grade of the medic and nurse is not an absolute—experience and competence are also considered, as is the patient's diagnosis and perceived condition.

- In instances of extremely severe illness, it might be prudent if possible to take two senior medics.
- The ambulance or aircraft personnel are an integral part of the operational team, and not just drivers. Experienced vehicle personnel, familiar with the needs of the transport team, can save valuable time.

• Mode of transport

Patient transfers are made *both within and between hospitals*. The principles for the safe and effective movement of a sick patient, particularly where the patient *is dependent on support devices* such as intravenous pumps and ventilators, apply to any transfer out of the intensive care environment irrespective of the distance needed to travel.

Hence, even when a patient is being moved for an investigation or to the operating theatre, the team should adhere to their practiced safety procedures.

Three options are available for inter-hospital patient transfers. The patient may be:

• Sent via the local emergency services

• Sent via the local (referring) emergency medical services, i.e. via ambulance with the referring physician and/or nurse

• Fetched by a specialized critical care transport team.

The first two modes are referred to as one-way transports and the third as two-way transport. The only advantage of sending a patient, or one-way transport, is time.

Inter-hospital patient transfers are either by road or by air in a helicopter or fixed wing aircraft.

Individual intensive care transport teams will have protocols as to which form is used based on various deciding factors.

These factors or criteria include:

• *Distance between referring and receiving hospital*. If the distance exceeds two hours' travel by road, serious consideration has to be given to transferring the patient by helicopter

• *Traffic density*. This can be partially overcome with the help of the emergency services, including the police who can clear the road. It should not be a reason to summon a helicopter for a short distance

• *Buildings in the town or city where the hospitals are located.* Transferring a patient to or from a big city by air has the added difficulty of high rise buildings affecting transit and landing

• *Weather*. Safety is the prime concern in any inter-hospital patient transfer. There is nothing to be gained from endangering the lives of a helicopter crew and the transport team by sending them out into poor unsafe weather conditions. A timely decision to use an ambulance may save precious time whilst you wait for the weather to clear.

Two-way transport (or retrieval) of critically ill patients by trained and experienced staff is the best and most desirable form of interhospital transport.

• Equipment

The operational team must carry all equipment, supplies and drugs necessary for stabilization and transfer. The team should not rely on the referring hospital for supplies. Pediatric patients are often "held" in adult facilities waiting for the transport team to arrive. These may carry limited pediatric supplies.

The equipment must be regularly checked and serviced. Supplies and drugs (i.e. contents of medical bags) must be replenished after each use; a contents checklist is essential. Checked and restocked boxes and bags should then be sealed, rendering them ready for immediate use. These should be stored in an accessible but protected and restrained space.

Equipment required during transport of a critically ill child

Patient movement

- Trolley
- Incubator for children < 5 kg
- Metal pole or shelf system to secure ventilator, pumps, monitors
- Adjustable belts (safety belts) to secure patient in transfer
- Equipment bags: multiple compartments to allow access to individual items without unpacking
- Drug boxes

Airway management

- Equipment to establish and maintain a secure airway bag-valve device (for example, Ambu bag) with selected mask sizes endotracheal tubes (all sizes), stylet and Magill forceps laryngoscope with assorted size blades
- Portable mechanical ventilator small, lightweight with economical gas usage capable of ventilating infants and children of all ages disconnection and high pressure alarms .
- Portable oxygen supply provide high pressure supply with low pressure meter flow sufficient to last duration of transfer with reserve, usually 1–2 hours
- Suction—portable, battery powered Intravenous infusions
- Equipment to establish and maintain venous and arterial access
- Resuscitation drugs.
- Infusion pumps—small, lightweight, long battery life

Monitoring

• Monitor: portable, battery powered, clear illuminated display ECG, oximetry, blood pressure and temperature .

Document folder

• Recording chart, audit form, consent form

• Infusion charts and crash drug charts—filled in prior to transfer

• Information for parents, i.e. maps and telephone numbers

Mobile phone

Warm protective clothing for staff

• Communication

Communication has many facets.

• Hotline. A direct line (not through the hospital switchboard) or "hotline" into the ICU that is dedicated to incoming calls (and hence not continuously engaged in a busy ICU) but also open to outgoing national calls will make conversations between consultants considerably easier and more effective. Advertise the number widely in the catchments area.

• Essential information. The ICU must inform their regular referring institutes about the information they will need to ensure that the referring doctor has this essential information to hand when making the telephone call.

Continuous contact: The transport physician or consultant on call should maintain continuous contact by regularly (every half hour) telephoning the referring physician for an update and further advice, if needed

Inform ambulance crew. The operational team must inform the ambulance crew about the patient's condition and the projected time required for stabilization and hence expected time of departure for home. This form of communication is particularly important with helicopter crews in air retrievals.

Talk to staff and family:

The family needs relevant information and reassurance, without misplaced optimism. Their child is being taken from them in an ambulance to a strange city, which they might never have visited before. A statement to the effect that the child is very sick but stable and that the team undertakes many transfers successfully every year will provide great comfort.

• Documentation

A good nurse is only as good as the records he or she keeps. A clear and concise written clinical record is important with any patient treatment, but possibly more important during inter-hospital patient transfers as these are patients who are usually critically ill and who in addition, for the duration of the transfer, are not managed in an optimal environment.

Data elements of a transport log

Data

• Patient: name date of birth sex weight (*very important for drug calculations*)

• Referring institute: referring physician contact telephone number and pager number referring hospital and ward or unit where patient will be located

Operational data

• Staff member receiving call

• Times: of receiving call of embarkation of arrival at referring hospital of departure from referring hospital of arrival at ICU at receiving hospital

Clinical data

• Pre-departure:

brief incident-related history, provisional diagnosis, reason for transfer request initial vital signs and pertinent physical findings relevant laboratory results, for example, arterial blood gas (ABG) treatment given, for example, ventilator settings, infusions, antibiotics recommendations given

• Assessment on arrival at referring hospital:

Clinical findings by transport team, including treatment, for example, ventilator settings recent laboratory investigations, for example, ABG, CXR with ETT position interventions at referring hospital

• In transit:

Vital signs (monitoring) during transfer medication administered by transport team problems encountered and treatment given

• On return to PICU:

Vital signs ventilator settings

Checklists

• For referring hospital

• Pre-departure (*focuses on equipment needed*)

• Pre-return (*focuses on patient care*)

Pre & Post operative Nursing Care

Surgery Related Differences between Young Children and Adults:

1. The metabolic rate of the infant and young children is much greater proportionately than that of adult. Children are growing and need to be fed more frequently.
2. The body tissues of the child heal quickly because of his rapid rate of metabolism and growth.
3. The child usually needs proportionately less analgesic than adult patient to obtain relative comfort after surgical procedures.
4. The child lacks the reserve physical resources that are available to the adult. His general condition may change very rapidly, almost without warning.
5. Abnormal fluid loss is more serious in the infant and young child than in the adult. Fluid intake and output must be calculated very carefully.

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Prepare children for surgery

Infants

☆ Infants (birth to 1 year) develop relationships based on trust and depend on parents and caregivers to meet their needs. Separation from parents can cause distress, which is primarily expressed through crying. Encourage parents to remain with the infant through as much of the process as possible, as they derive comfort from familiar faces and voices.

☆ Encourage parents to bring a security object such as a blanket or stuffed animal to help soothe the infant when separated from his parents.

- ☆ Reunite the infant with his parents as soon as possible after the procedure to minimize distress.
- ☆ Limit the number of caregivers, if possible, as strangers also cause fear.
- ☆ Other comfort measures such as soft music, a pacifier, cuddling, gentle stroking, or rocking can help calm infants. Infants also enjoy nursery rhymes and simple rhythmic play.
- ☆ Preparation at this stage focuses on the parent due to the limited cognitive abilities of the child. Inform the parents about the procedure and provide support; their infant is likely to be less anxious if the parents are calm.

Toddlers

- ☆ Toddlers (1 to 3 years) differ from infants as they're just beginning to gain autonomy by attempting to do things for themselves. This is an active period with short attention spans. Although verbal skills are limited, they understand more than they're able to say.
- ☆ Provide simple, concrete explanations about what the toddler will hear, feel, smell, taste, and see. It isn't necessary to cover all details of the surgery.

- ☆ In general, preparation should be done a day or two before surgery at a level the child will understand Toddlers possess a limited concept of time, so use explanations the child is familiar with. For example, tell the child, "The surgery will be before lunch." Unfamiliar faces, routines, and scary equipment may cause stress.
- ☆ Encourage family members to bring his favorite activities or toys to distract the child from boredom, fears, and the new routine. Toddlers fear abandonment; as a result, separation from family can be extremely stressful.

- ☆ Minimizing separation from a parent can further a toddler's ability to cope in a stressful situation. A favorite toy, stuffed animal, or photograph that the child can hold while going to surgery can offer comfort. Toddlers may sometimes temporarily lose newly acquired skills such as walking or toilet training.
- ☆ It is important to answer questions about pain and separation truthfully. Also, provide simple choices whenever possible to heighten a toddler's sense of independence and control. For example, allow the toddler to choose his favorite of two character adhesive bandages for use after the I.V. is removed.

Preschoolers

- ☆ Preschoolers (3 to 5 years) develop their own identities, are very inquisitive, and have active imaginations. Fantasies and misconceptions related to surgery are common. Offer simple explanations and descriptions, allowing the child to see and handle some of the medical equipment that will be used.
- ☆ Although their vocabulary is developing rapidly, they don't have the full ability to reason. Therefore, explanations about a procedure should
- ☆ be kept simple and matter-of-fact, focusing on what the child will hear, feel, smell, taste, and see, as well as what the child is expected to do.
- ☆ As a preschooler's concept of time is limited, the nurse must review and reinforce the sequence and duration of all events. Use time concepts the child is familiar with: "The operation will take less time than it takes to watch a cartoon." Preparation for surgery for this developmental stage is recommended a day or two in advance
- ☆ Preschool children fantasize and may feel that their operation is punishment for bad behavior. Reassure the child that he isn't to blame and that the surgery is to fix something specific. Be honest, especially about separation and potential pain.

- ☆ Playing during the preoperative phase can help distract children who are distressed. As with all children, this age group will benefit from having a familiar toy, stuffed animal, or photograph. Offer choices whenever possible to promote a sense of control

School-age children

- ☆ School-age children (5 to 12 years) devote most of their energy to school and peers. Mastery of skills, experiences, and self-esteem are increasingly important issues for this age group. Although still engaged in some imaginary thinking, they're capable of concrete, logical reasoning and are gaining an increased understanding of cause and effect. They are better able to cooperate with treatment because they can think before they act
- ☆ Preparation should begin a week in advance of the surgery. Allow the school-age child to participate in care when possible. For example, to provide a sense of control and encourage acceptance of treatment, ask the child to help hold the anesthesia mask.5 Provide choices when possible such as asking, "Which arm should we use to measure your blood pressure?"
- ☆ School-age children may fear other body parts will be hurt during the operation. Offer a simple explanation of what part of the body the operation will affect. Body outlines, pictures, or dolls may be helpful.
- ☆ For example, a child's understanding of a surgical site can be quickly assessed by asking him to mark the site on the doll. Any misconceptions revealed can then be corrected.

Adolescents

- ☆ During adolescence (12 to 18 years), abstract thinking begins and adolescents can understand how the body functions, the nature of the problem, and the reason for surgery.

- ☆ Provide honest, detailed explanations about the diagnosis and need for surgery, including what the adolescent will hear, feel, smell, taste, and see. Adolescents often want to know the results of surgery and what'll happen next. Involve them in decisions about their health as much as possible.
- ☆ Common fears include waking during the procedure, pain, and the possibility of death. They may need reassurance in their anesthesia provider's ability and an explanation that they won't wake up during the procedure but will wake up afterward
- ☆ Nurses should respect an adolescent's need for privacy. Always knock on the door or announce your presence before opening a curtain to avoid embarrassing them if they're undressing. This kind of courtesy can lead to increased cooperation.

Preparing parents

- ⏳ Preoperative preparation processes have been affected by changes in healthcare delivery systems and economic constraints. Preoperative hospital visits are now often replaced with information shared via telephone or Web sites.
- ⏳ Parents are becoming increasingly responsible for preparing their child for surgery. Therefore, parents should be partners in the preparation process, receiving not only information about the procedure and sequence of events, but also the guidance and tools to provide support and prepare the child for the experience.

Immediate Postoperative Care

The postoperative care of surgical neonates and children begins upon completion of wound closure. The level of postoperative care administered is dependent upon the procedure performed but some general guidelines are

provided below. Specific guidelines for postoperative management of many pediatric surgical conditions are provided throughout this handbook.

Immediate Postoperative Care

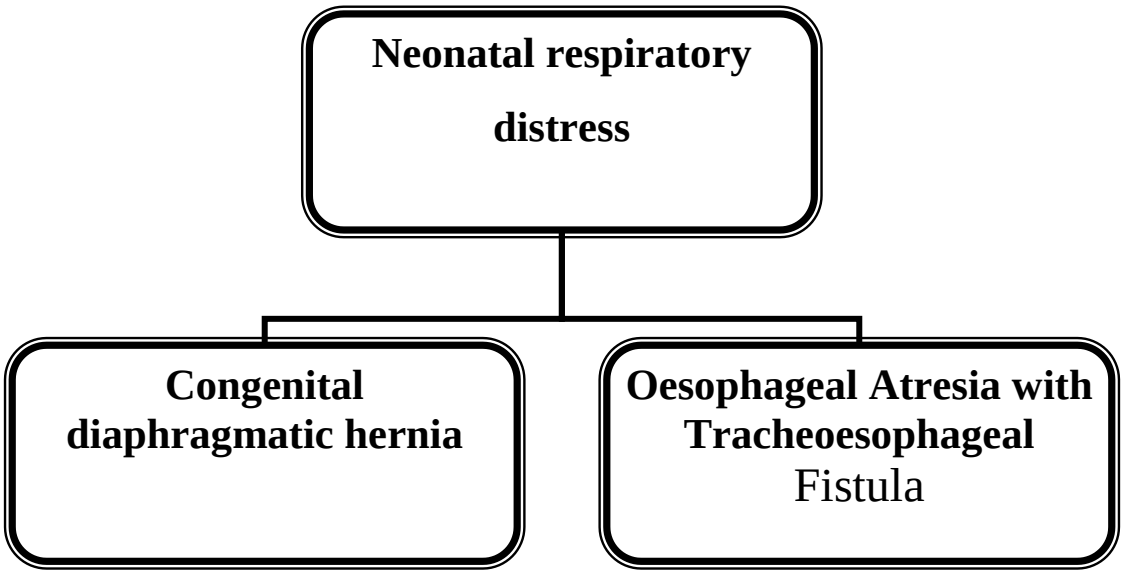
1. After return from the operating room, the child's general condition must be closely observed **(a)** Vital signs, especially temperature. **(b)** Airway must be kept patent newborn babies must be kept in warm cot or incubator.
2. Unit the child is responsive and alert; he should be kept on his side (for secretion and vomitus to get out from mouth).
3. Observe conditions and placement of dressing. Check and mark any apparent drainage from wound.
4. Intravenous fluids should be checked for correct rate of flow and for possible infiltration.
5. The child should be carefully handled and should be protected from harming himself by use of appropriate restraints.
6. Any urinary catheter should be connected to drainage bag and stabilized properly to bed.
7. Observe patient's skin color and temperature as well as any sings of shock **(a)** low blood pressure **(b)** rapid pulse **(c)** cold moist pale or cyanotic skin **(d)** dilated pupils **(e)** restlessness.
8. Oral fluids may be started often after the following criteria are observed **(a)** color of aspirate is clear **(b)** peristaltic movements are heard **(c)** flatus or gases are passed. Oral fluids should be started while infusion still on. If well tolerated then infusion is gradually discontinued. Routine postoperative diet is modified according to child's age, but in general it changes from clear of liquid, full liquid, soft and then regular diet.
9. Sedatives are used according to prescribed orders and child's needs.
10. For children who can walk, early progressive ambulation is the rule (expect in few cases), this will help to restore gastrointestinal function

and prevent complications of pneumonia, the thrombosis, and pressure areas. If too young to go out of bed, the nurse should turn the child frequently and give him good skin care and help him to breath deeply at intervals.

Specific Postoperative Care

- Wound and Dressing Care
- Extubation and Transfer
- Pain Management

Neonatal respiratory distress



Esophageal Atresia and Tracheoesophageal Fistula: (T.E.F.)

It is congenital malformation represent a failure of the esophagus to develop a continuous passage. These defects may occur separately or in combination.

Definition:

- ☆ **Oesophageal atresia** is defined: as an interruption in the continuity of the esophagus with or without fistula to the trachea.
- ☆ ***Tracheoesophageal Fistula*** is defined as abnormal fistulous communication between the trachea and the esophagus with Atresia of the esophagus singly or in combination.

Etiology:

Unknown.

Embryogenesis:

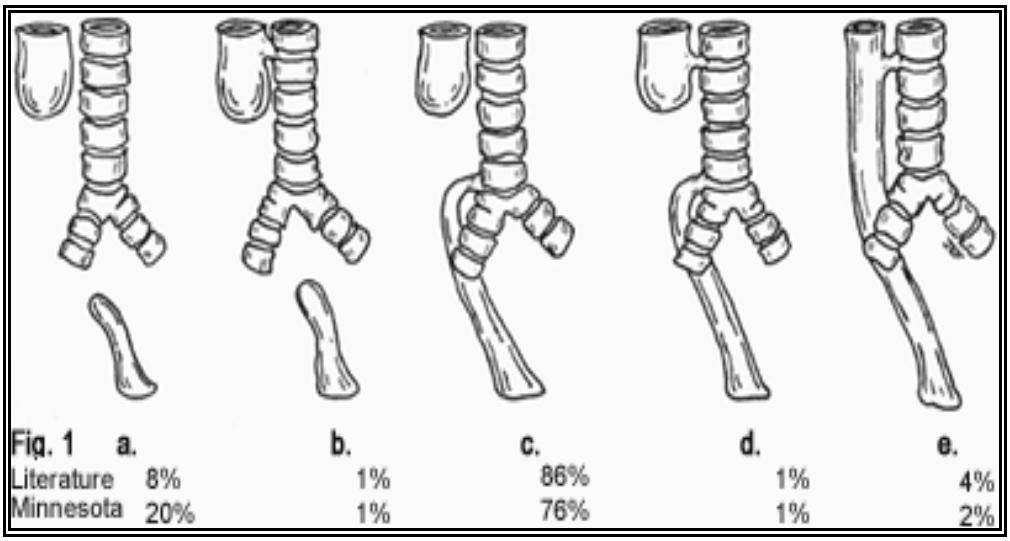
- ♦ Separation of the esophagus from the trachea occurs by the development of ridge grow inward to separate the two tubes within the fourth week of gestation. (Interruption of this separation leads to ***fistula*** formation).

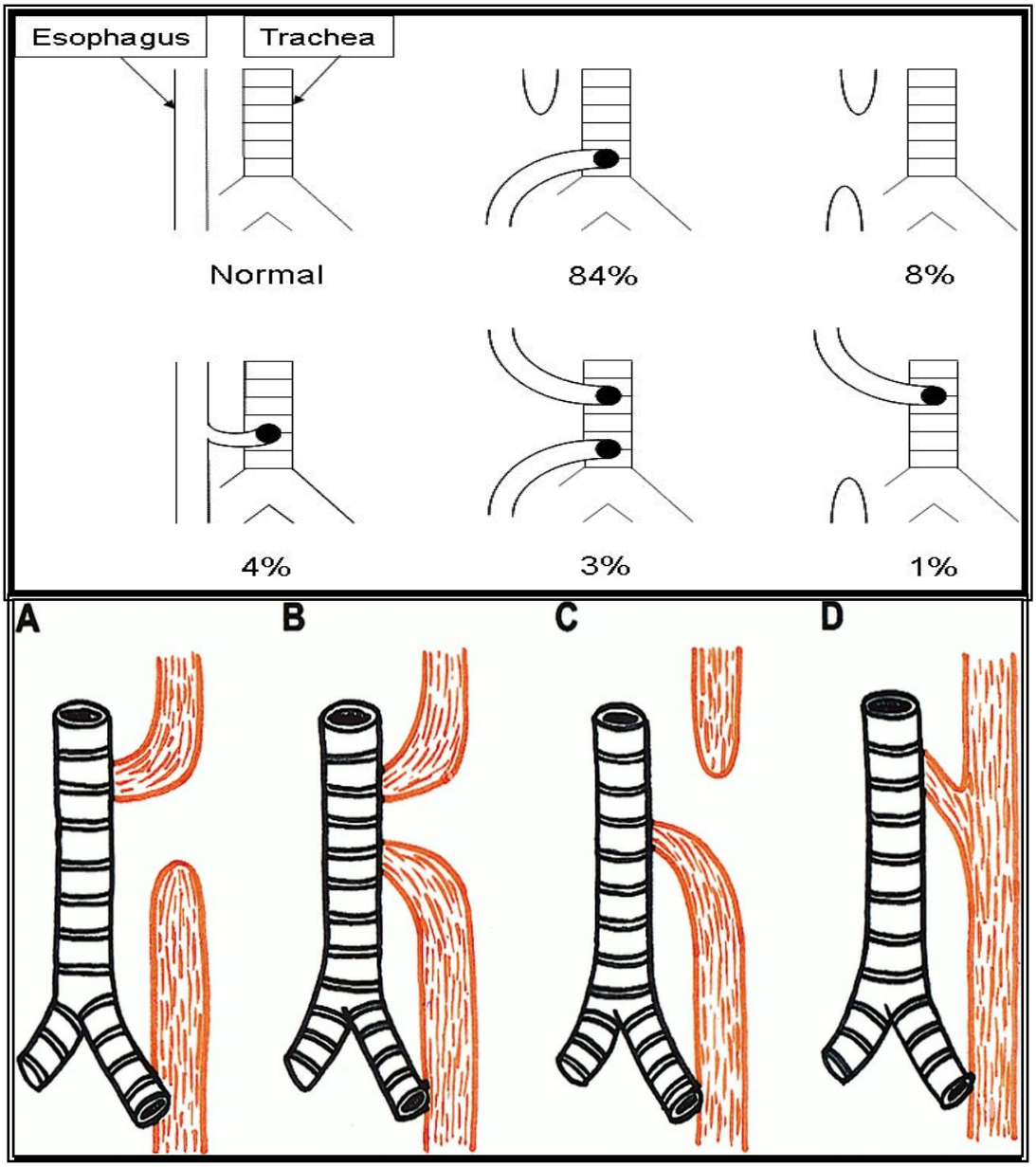
- The mechanism of Artesia may be due to:

2. The mechanism of Artesia is more difficult to explain but recently; atresia was attributed to extremely hyper flexion of the embryo.

Types:

1. Atresia with proximal blind pouch and fistula of the distal oesophagus pouch to the bifurcation of the trachea (85%).
2. Atresia with out fistula (6%).
3. Atresia with fistula of both pouches (5%).
4. Atresia with blind distal pouch and fistula in the proximal pouch (2%).
5. Fistula withoutbatresia (H fistula).





N.B VATER association:

V = Vertebral anomalies

A = Anal anomalies

TE = Tracheoesophageal Fistula

R = Renal deformaties

Clinical Manifestation of TEF:

- prematurity and polyhydramnios are common association.
- Excessive salivation and drooling
- Three Cs of TEF on feeding:

-Coughing.

-Choking.

-Cyanosis.

-and, regurgitation

- Apnea.
- Increased respiratory distress following feeding.
- Abdominal distention.

N.B causes of R.D:

1. Regurgitation of saliva and milk from the obstructed upper pouch into the trachea.
2. Reflux of acidic content through the fistula into the lung leading to pneumoniatis.
3. Gastric distention by air from trachea leading to diaphragmatic elevation.

Diagnostic Evaluation:

- Insertion of catheter gently into the esophagus meets with resistance if the lumen is blocked but passes unobstructed if the lumen is patent.
- Radiographic visualization of opaque catheter inserted into the hypopharynx helps to determine patency and / or alternate channels.
- Endoscopy for isolated fistula.

Objective of Therapeutic Management:

(1) Prevent pulmonary complications.

(2) Provide nutrition.

(3) Repair defect surgically

Preoperative evaluation and treatment:

☒ Assess the infant fitness and his need for pre-operative resuscitation.

☒ General lines for neonates with surgical emergency :

1. Transfer to ICU
2. Intubation (Regulate oxygen ,temp, and humidity)
3. I.V fluid for correction of dehydration and electrolyte disturbance

☒ Prevention of pulmonary Complications:

1. Continuous suction from the upper pouch
2. Minimize gastric reflux by elevation of head of the bed.

☒ Treatment of the established pneumonia:

1. Humidified oxygen inhalation.
2. Broad-spectrum antibiotic.
3. Tracheal aspiration through endotracheal tube.

Surgical treatment:

The choice and timing of operative procedure depends largely on the distance between the two oesophageal pouches and risk classification of the infant.

Postoperative care and treatment:

☒ The preoperative scheme is continued in the postoperative.

☒ Gastrostomy feeding could be started in the third postoperative day.

☒ Esophagogram on the fifth postoperative day to exclude the presence of leakage or stenosis of the suture line.

☒ Gradual oral feeding could be started on the 7^{th} postoperative day.

Complications:

Anastmosis → Leakage ,Stricture and recurrent.

Pleural → Pnumothorax and empyema.

Dysphagia → From disordered esophageal peristalsis.

Congenital Diaphragmatic Hernia

Bochdalek Hernia (60%):

- ☆ Congenital diaphragmatic hernia (CDH) is a malformation characterized by a defect in the posterolateral diaphragm, through which the abdominal viscera migrate into the chest during fetal life.
- ☆ Most common type due to early reduction of physiological hernia (normal at the 10^{th} week) before closure of the foramen of Bochdalek by the pleuroperitoneal membrane at the 8^{th} week.
- ☆ It is more common on the Lt. Side because the left side usually closes later than the Rt. Side does. The sac is absent in 90% of cases.

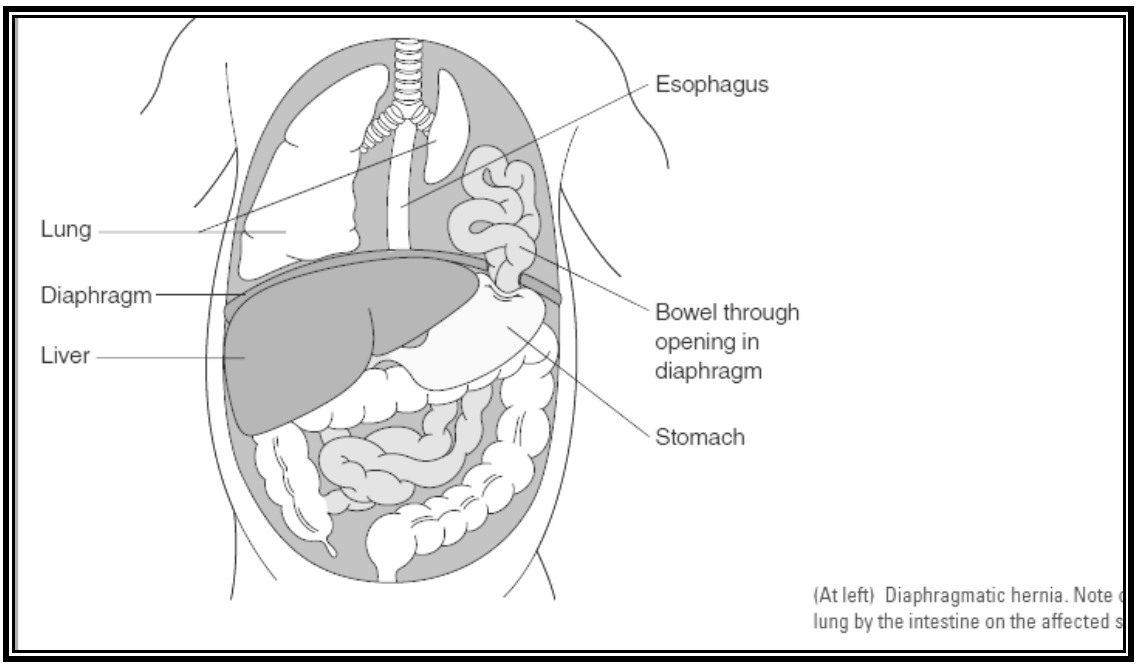
Clinical Picture:

- ☆ Postnatally, the most severely affected babies present with respiratory distress (cyanosis, tachypnoea and sternal recession) at birth.
- ☆ Other infants develop cyanosis, tachypnoea and grunting respirations within minutes or hours after birth.
- ☆ Physical examination reveals a scaphoid abdomen, an increased anteroposterior diameter of the thorax and mediastinal shift. Breath sounds are absent on the affected side.

Diagnosis:

Diagnosis of CDH is made postnatally by ***plain radiography*** of the chest and abdomen by demonstration of:

- ☒ Air-filled loops of the bowel in the chest and a paucity of gas in the abdomen.
- ☒ The diaphragmatic margin is absent,
- ☒ There is a mediastinal shift to the opposite side and only a small portion of the lung may be seen on the ipsilateral side.



Treatment:

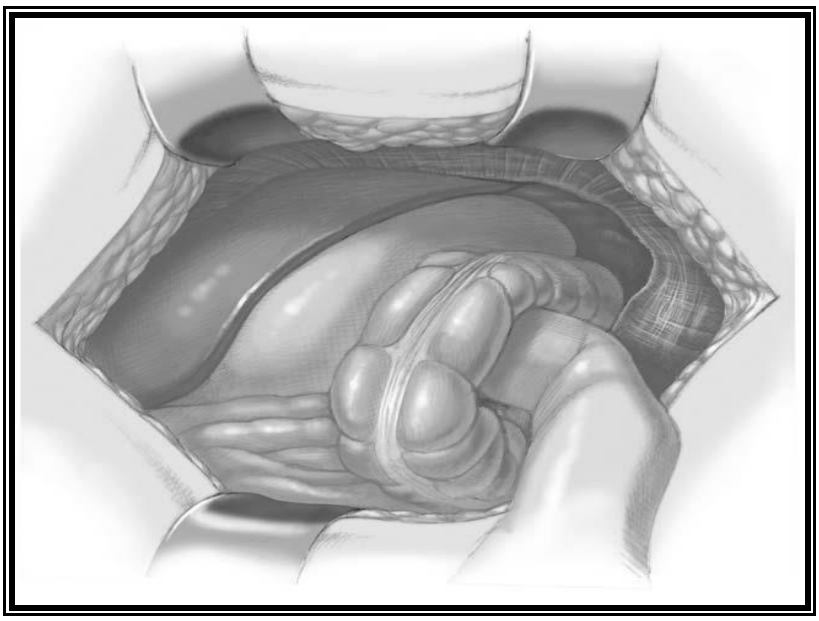
Immediate surgical repair is indicated because of the severe life threatening respiratory distress associated with the condition.

Preoperative measures:

- ☒ O_2 administration to decrease R.D. and cyanosis.
- ☒ Stomach decompression.
- ☒ Adequate hydration.

Operation:

- ☒ The content are reduced, the sac is removed if present.
- ☒ The defect is closed with non absorbable sutures and if large can use a mesh.



Post operative care:

- ☒ O_2 tent
- ☒ Frequent change s in position with pharyngeal aspiration.
- ☒ I.V. fluids till effective peristalsis returns.

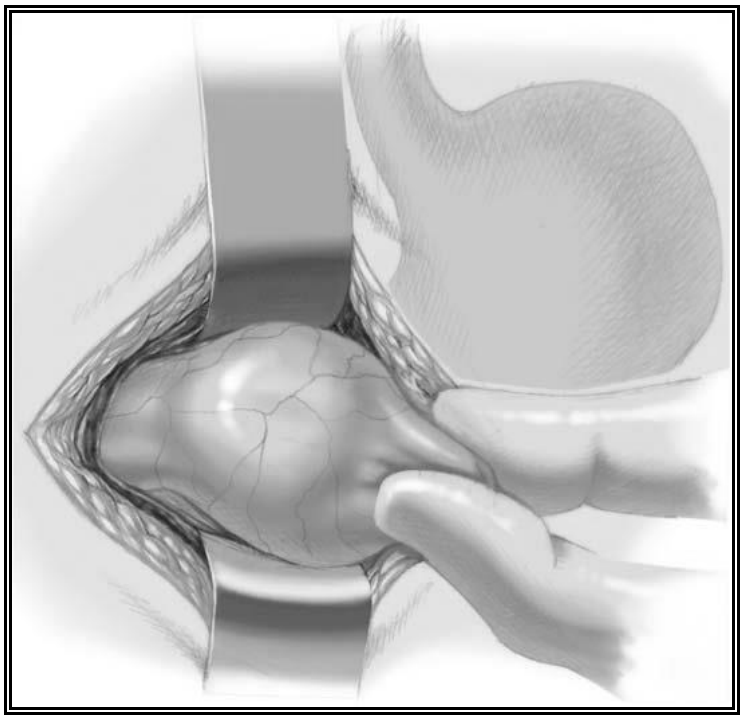
Neonatal Intestinal Obstruction

Types:

1. Infantile hypertrophic pyloric stenosis.
2. Intestinal atresia and stenosis.
3. Mal-rotation and volvulus neonatorum.
4. Meconium ileus,plug syndrome.
5. Meconium peritonitis.
6. Neonatal Hirschsprung Disease.
7. Neonatal entrocolitis.
8. Neonatal Intussusception.
9. Anorectal malformation.
10. Hernias → External and internal.

Hypertrophic pyloric stenosis

- ☆ Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical condition encountered in early infancy, occurring in 2~3 per 1,000 live births.
- ☆ It is characterized by hypertrophy of the circular muscle of the pylorus, leads to constriction of the pyloric canal and obstruction of the gastric outlet.
- ☆ The incidence of disease varies widely with geographic location, season and ethnic origin.
- ☆ Boys are affected four times more than girls.



Pathophysiology:

The circular muscle of the pylorus is grossly enlarged as a result of both hypertrophy (increased size) and hyperplasia (increased mass).

This produces severe narrowing of the pyloric canal between the stomach and the duodenum. Consequently, the lumen at this point is partially obstructed over a period of time, inflammation and edema further reduce

the size of the opening until the partial obstruction may progress to complete obstruction.

Clinical Manifestation:

1. Typical clinical presentation of infants with IHPS is non-bilious vomiting usually occurring at 2–8 weeks of age. Initially there is only regurgitation of feeds, but over several days vomiting progresses to be characteristically projectile.
2. Hungry and irritable later become lethargic.
3. Dehydration.
4. Malnutrition.
5. Constipation.
6. Loss of skin turger.

Diagnostic Evaluation:

1. The diagnosis is usually based on the clinical history and physical examination of a "palpable pyloric tumour".(Palpable olive like mass in right upper quadrant).
2. Visible, peristaltic waves usually form the left to right.
3. Ultrasonographic scanning of abdomen re reveals typical increased muscle thickness.
4. Narrow pyloric sphincter revealed in barium swallow. (Positive string sign).
5. Persistent non-bilious vomiting in these patients results in chloride depletion, metabolic alkalosis and dehydration.

Therapeutic Management:

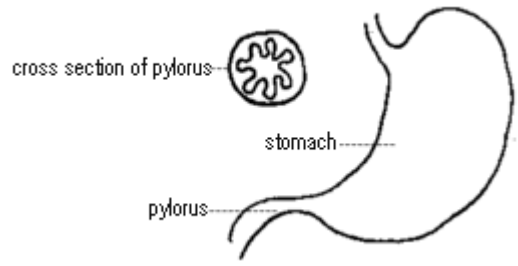
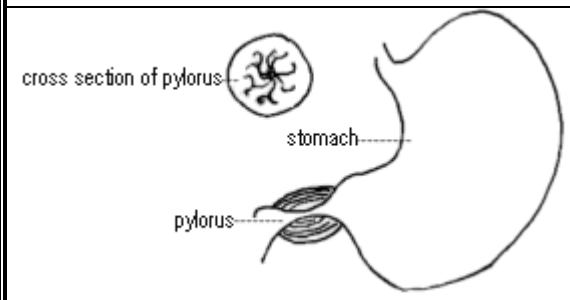
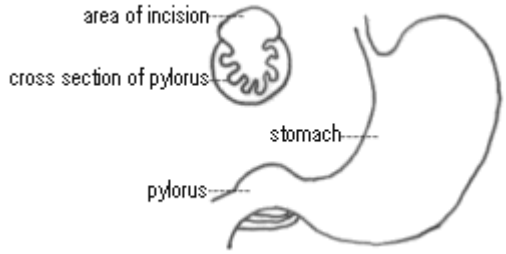
- ☆ Surgical relief of the pyloric obstruction by ***Pyloromyotomy***.
- ☆ Recently, ***laparoscopic pyloromyotomy*** has been advocated. The main advantage of the laparoscopic pyloromyotomy is the superior cosmetic result.

Preoperative measures:

- N.G tube suction and gastric wash with saline to minimize stagnation gastritis, which can lead to post operative vomiting.
- Correction of dehydration, electrolyte and acid –base disturbance.

Postoperative measures:

- N.G tube suction for 8-12 hours, following this oral feeding is started in a very gradual fashion using 10% dextrose every 2 hours in increasing amount.
- If infant tolerances, half formula followed by full milk formula is given.

	Diagram of normal stomach and pylorus. Note the cross-section showing normal pyloric opening.
	Diagram of stomach with pyloric stenosis. Note the cross-section showing how the pyloric opening is very narrowed.
	Diagram of stomach after repair of pyloric stenosis. Note (in the cross-section) how an incision has been made in the muscle, enlarging the pylorus and relieving the obstruction.

Meconium Ileus

Definition:

It is a type of I.O. due to the presence of a thick viscid meconium obstructing the intestinal lumen.

Pathology;

Meconium ileus is the earliest clinical manifestation of cystic fibrosis (CF) and occurs in 8–10% of patients with CF at birth.

The clinical features are mainly due to the presence of abnormal, viscid mucus of intestinal origin.

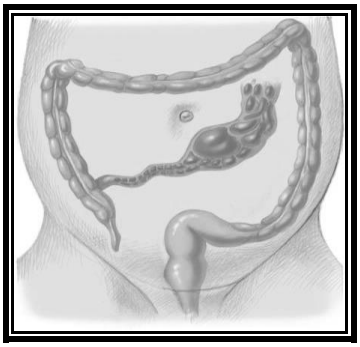
In neonates affected by this condition, the impacted meconium produces an intraluminal obstruction occurring in the midileum, leading to a progressive distension.

Clinical picture:

- ☆ Main symptoms include abdominal distension and visible intestinal loop .
- ☆ Bilious vomiting.
- ☆ Failure of or delayed passage meconium.

Investigations:

- ☆ Plain x-ray abdomen: gas distending small intestinal loops.
- ☆ Barium enema : microcolon with area of dry meconium within the contrasted material.



Management:

Conservative:

- ☆ The first step of the treatment includes a nasogastric tube decompression.
- ☆ Antibiotic prophylaxis.
- ☆ Correction of dehydration, electrolytes and hypothermia.
- ☆ A contrast enema with water-soluble and hyper or iso-osmolar contrast is the medical treatment of choice and mucosal safe, for uncomplicated cases.

Surgical:

surgical procedure such as resection of the dilated meconium-filled ileum and ileal anastomosis

Complications:

About 40% of patients with meconium ileus are complicated by:

- ☆ Intestinal volvolus
- ☆ Atresia,
- ☆ Gangrene and necrosis,
- ☆ Perforation, and,
- ☆ Finally, peritonitis with abdominal calcifications.

Hirschsprung Disease

(Congenital Aganglionic Megacolon)

Definition:

Is an abnormality in which certain nerve fibers are absent in segments of the bowel, resulting in severe bowel obstruction caused by inadequate motility in part of the intestine.

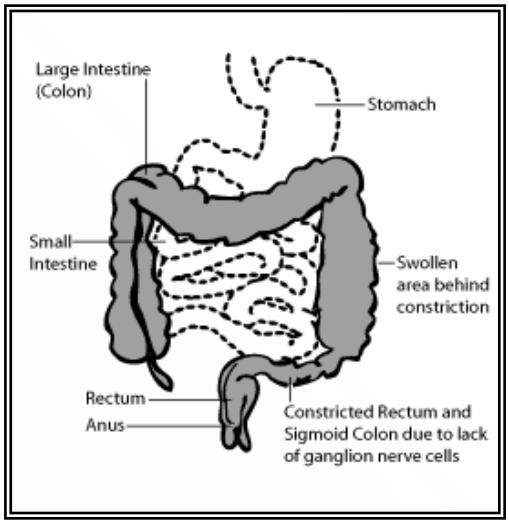
Pathophysiology:

Congenital Aganglionic Megacolon describes the primary defect, which is The absence of autonomic parasympathetic ganglionic cells in the submucosal and myenteric plexuses in one or more segments of the colon.

Lack of innervation produces the functional defect:

- Absence of peristalsis, which cause.
- Accumulation of intestinal contents and bowel distention proximal to the defect.
- Failure of the internal anal sphincter to relax contributes to clinical manifestations of obstruction because it prevents evacuation of solids and gas.

Intestinal distention and ischemia may occur as a result of distension of the bowel wall.



Types:

The A ganglionic segment may be :

1. Short segment affecting the terminal rectum (10%),
2. Extends to the sigmoid colon (65%).
3. Involves more proximal colon (10%)0
4. Affecting the entire colon (15%) and rarely the ileum.

The proximal segment may be :

1. Showing enormous dilatation and hypertrophy.
2. The mucosa shows chronic inflammation and ulceration.

Clinical Picture:

- ☆ Of all cases of HD, 80–90% produce clinical symptoms and are diagnosed during the neonatal period.
- ☆ Delayed passage of meconium is the cardinal symptom in neonates with HD.
- ☆ Over 90% of affected patients fail to pass meconium in the first 24 h of life.
- ☆ Chronic constipation.
- ☆ Poor growth and development.
- ☆ Vomiting and unwilling to feed and irritability.

Rectal examination:

- ☆ Empty rectum (distended with gases mainly) painless P-R.
- ☆ Gush of foul smelling gases and stool after withdrawing the finger, followed by reduction of distention.

N.B.

In older children:

- ☆ Chronic constipation, passage of stools needs effort.
- ☆ Progressive abdominal distention.
- ☆ Stool is of small caliber, tooth-past like.

Investigation:

1. *In neonate*: diagnosis is usually based on clinical signs of intestinal obstruction and failure to pass meconium.
   In infant and children: typical details of chronic constipation.
2. On examination, the rectum is empty of feces, the internal sphincter is tight and leakage of liquid stool.
3. Barium enema
4. Rectal biopsy : shows lack of ganglionic cells.
5. Rectal manometry (Recto-anal reflex test):
   In shows failure of relaxation of the internal sphincter in response to rectal distention.

Complications:

☆ Putrefactive diarrhea (main cause of death).

☆ Perforation.

Treatment:

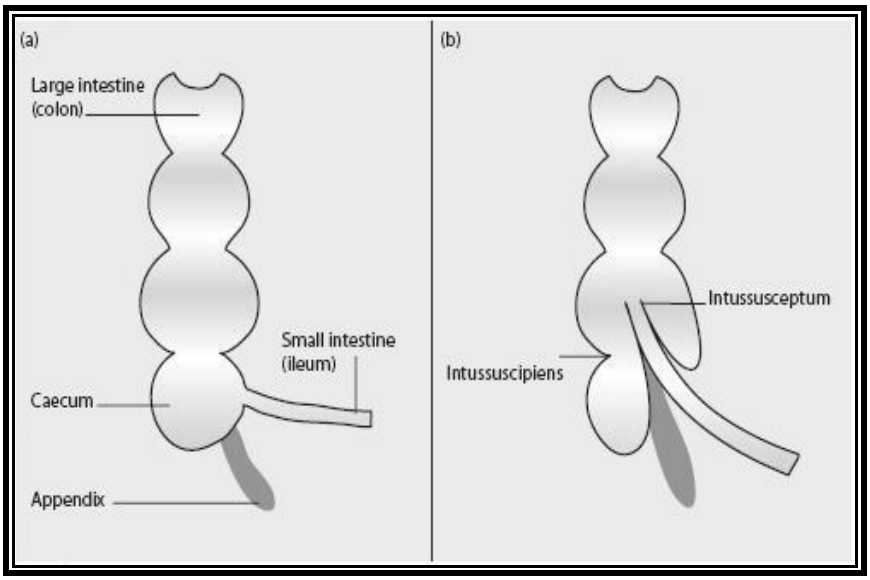
Treatment is primarily surgical removal of Aganglionic portions of the bowel in order to permit normal bowel motility and establish continence by improved functioning of the internal anal sphincter.

Surgery consists of:

1. **First Stage:** Temporary ostomy is created proximal to the Aganglionic segment to relieve obstruction and allow the normal ionervation of dilated bowel to return to its normal size.
2. **Second Stage:** Is complete, correction. Consists of pulling the end of the bowel through the muscular sleeve of the rectum from which the Aganglionic mucosa has been removed.

Intussusception

It is an acquired type of intestinal obstruction. It is the invagination or telescoping of a segment of intestine into the segment immediately distal to it.



Etiology:

Primary:

Exact cause of Intussusception is uncertain but there are several theories as:

1. Greater disparity between the size of the ileum and ileocecal value in infants is believed to encourage telescoping at this point.
2. Distorted peristalsis due to change of diet (weaning).
3. Enlarged lymph nodes along the G.I.T. that occur with respiratory infections, cystic fibrosis, foreign bodies, GI polyps.
4. Hyperperistalsis.
5. Diarrhea, constipation.
6. Mobile ileocecal junction.

Secondary: (5%), rare before age 4years ***due to***

1. Intestinal lymphoma.
2. Submucosal lipoma ,polyp
3. Mackel's diverticulum.

The most common sites:

- Ileocecal value. (Ileocolic)
- Ileoileal.
- Colocolic.

Incidence:

Intussusception is one of the most frequent causes of intestinal obstructions in children between the ages of 3 months and 5 years. Half of the cases occur in children younger than 1 year, and most of the others occur in children during the second year.

Clinical Manifestation:

1. Sudden acute abdominal pain, which becomes progressively more severe.
   - Child screams and draws the knees into the chest.
   - Child is healthy and normal between episodes of pain.
2. Vomiting.
3. Lethargy.
4. Passage of red current jelly stool. (Stool mixed with blood and mucus).
5. Palpable sausage- shaped mass in upper right quadrant.
6. Empty right lower quadrant (dance sign).
7. Anorexia, discomfort.
8. Tender, distended abdomen.
9. Eventually fever, perforation, peritonitis and shock are serious complications of Intussusception.

Late neglected cases:

Prolonged ischemia → infarction of the bowel and subsequent peritonitis.

Intestinal obstruction → bilious vomiting ,distention.

Investigation:

1. Plain abdominal radiograph is only indicated in unclear diagnosis, showing only late specific signs of abdominal gas distribution or faecal contents and, finally, signs of bowel obstruction or perforation.
2. In retrograde barium enema formerly used, the contrast medium is outlining the apex of the intussusceptum.
3. Ultrasonography as well as fluoroscopy with contrast enema is well established.

Therapeutic Management:

1. Conservative (Hydrostatic Reduction):

In this procedure, correction of the invagination is carried out at the same time as the diagnostic testing. The force exerted by the flowing normal saline is usually sufficient to push the invaginated portion of the bowel into its original position. Similar to pushing an inverted finger (out of glove) this observed by fluoroscopy.

N. B *Complications of the reduction maneuver are perforation,*

2. Surgical:

Indication for Surgery

☆ Failure or contraindication of Hydrostatic Reduction.

☆ Older age group over 2 years

Methods:

☆ Resection and anastomosis.

Anorectal Malformation

It is incomplete development or absence of the anus in its normal position in the perineum

(Imperforate anus may include a single abnormality or a combination of abnormalities of the rectum (the end of the intestine near the anus) and anus (the opening of the rectum to the skin).

The incidence of anorectal malformations (ARM) is reported as 2.0–2.5 per 10,000 live births.

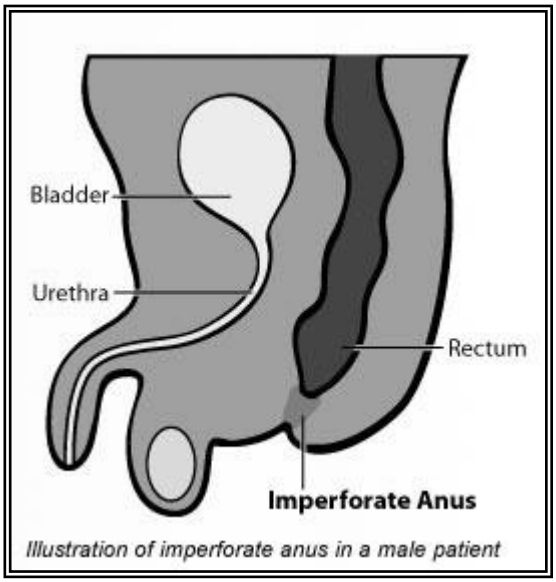


Illustration of imperforate anus in a male patient

There are many forms of this birth defect that may include the following:

1. The absence of an anal opening, which prevents any bowel movements completely.
2. The anal opening in the wrong place that is often too small.
3. A connection or opening called a fistula, between the rectum and the urethra, bladder or vagina. This may cause bowel movements to pass out of these abnormal openings.

4. In girls, the rectum, urethra, vagina can join together to form a single opening. This is called a cloaca and it is very rare.

There is no known cause for imperforate anus and most cases are isolated and do not run in families.

Types of Imperforated Anus:

1. **Anal Stenosis:** Narrowing of the anorectal canal that may occur at any point or extended its entire length.
2. **Imperforated Anal Membrane:** A thin membrane persists across the anal opening.
3. **Anal Agenesis:** The rectum ends in a blind pouch below (Low) or above (High) the levator ani muscles.
4. **Rectal Atresia:** The anus is normal but the rectal canal is not continuous. This type is very rare.

Classification:

ARM represent a wide spectrum of defects and conditions.

A clear understanding of normal anorectal anatomy and the different types of ARM is necessary for both the planning of surgery and the procedure itself.

(1) A narrowed anus,

(2) A closed anal membrane,

(3) Rectum interrupted by a septum at some distance from the opening

(4) Imperforate anus and

(5) The presence of a rectal fistula.

According to the relation of the distal bowel end to the puborectalis of the levator ani.

1. *LOW ANOMLIES :*
   - ☆ Anal memberane obstruction
   - ☆ Anal stenosis.

- ☆ Covered anus → a triangular skin fold covering the anal canal.
- ☆ Ectopic anus: the anal canal is displaced anteriorly.

2. *High anomalies :*

The bowel end above the puborectalis.

- ☆ With fistula
- ☆ Without fistula (in male ~ Recto-urethral or recto-vesical) (in female High recto- vaginal or cloaca = one opening in the perineum for anus,vagina, and urthera).

3. *Intermediate anomalies :*

Defect between the anal canal and the rectum.

The end of bowel is at but not through the puborectalis muscle.

Diagnostic Procedures:

The diagnosis of anorectal malformations (ARM), with the exception of anal stenosis, should be made shortly after birth during the routine neonatal examination.

1. **Diagnostic Guidelines**

The initial management of a newborn baby with ARM should be made only after accurate determination of the exact type and level of the anomaly. A decision can then be made as to whether to do a primary perineal operation, or to perform a colostomy, deferring definitive repair. In addition to determining the exact level and anatomical type, further information is required, such as the integrity of the neuromuscular components of the pelvis and the presence of any associated anomalies, particularly in the urinary tract.

The aims of the initial assessment are threefold:

1. To determine the level of the malformation in relation to the muscular sphincters and the site of any fistulous communications.

2. To determine the integrity of sphincters and their nerve supply.

3. To document any associated anomalies that may affect survival.

2. **Invertogram ;**
   - ☆ Plain X - ray abdomen and pelvic, 2 views the body up side down after 8-12 hours of birth (to give time to the air to reach the rectum).
   - ☆ To assess the level of malformation : comparing the highest level of the intestinal gas to bony land marks coinciding with the level of the levator ani .
3. **Contrasted Studies ;**
   - ☆ For better delineation of the level and the ***fistulo*us track ,** it is done by injection of dye by syringe into the rectal pouch.
4. **I.V.u ;**
   - ☆ For patient with high anomalies and / or suspected ***urinary anomalies.***

Assessment Criteria:

- Failure to pass meconium stool in the first 24 hours after birth.
- Absence or stenosis of the anorectal canal.
- Anal membrane.
- External fistula to the perineum.

Management:

Once the diagnosis is confirmed, immediate surgery is recommended. Preoperative resuscitation is essential with a wide bore nasogastric tube to decompress the abdominal distention, correction of dehydration and electrolyte imbalance, maintenance of body temperature, antibiotic coverage, vitamin K injection, and a urethral catheter to measure urine output as well as to decompress the bladder.

Low anomalies**:** immediate treatment after birth

a. Anal membrane obstruction → simple excision of the membrane
b. Anal stenosis→ Dilatation
c. Covered anus → incision of the skin fold
d. Ectopic anus: cut back operation

High anomalies :

- ☆ First stage colostomy
- ☆ Definitive operation is done at the age of one year [PSARP}
  POSTERIOR SAGITTAL ANORECTOPLASTY
- ☆ Closure of colostomy

Complications:

- ☆ Stool incontinence
- ☆ Anal stricture, due to neglection of post –operative dilatation.
- ☆ Recurrent fistula
- ☆ Sloughing of the rectum → due to ischemia which result of inadequate mobilization and tension.

Abdominal Wall Defects

Exomphalus / Omphalocele

It is a large congenital hernia into the base of the umbilical cord covered by a translucent membrane formed by fused layer of the amniotic membrane and peritoneum.

Definition:

- ☆ Omphalocele is an abdominal wall defect (opening) of the umbilical ring (belly button) that allows intestines and other abdominal contents to grow outside the body into a translucent (see through) sac before birth.

- ☆ Exomphalus (also known as omphalocele) is a condition that is seen in newborn infants, and is thought to result from failure of the intestines to return to the abdomen after the migration into the umbilical cord that occurs between the sixth and tenth week after conception.

Embryogenesis:

At the 4^{th} week of embryonic life, both the mid gut and the liver grow more rapidly than the abdominal cavity.

Thus, some loops of the intestine herniated into the umbilical cord space (physiological hernia.

At the 10^{th} week the abdominal cavity enlages, those loops return back.

N.B

Types : there are major and minor varieties .this depends on the size of the defect rather than the sac:

☆ IF more than 5cm →major.

☆ IF less than 5cm →minor.

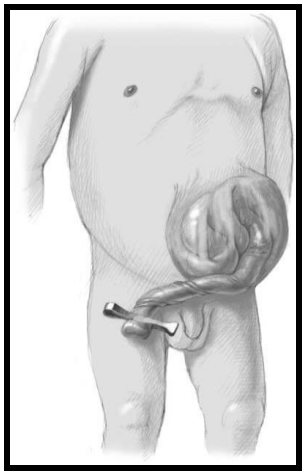
Clinical Picture:

The diagnosis is evident at birth with shiny sac at the base of the umbilical cord containing a variable amount of the abdominal viscera.

In minor type: the sac is relatively small and the umbilical cord is attached to its summit

In major type : the umbilical cord is attached to the inferior aspect of the swelling .it contains small and large intestine and nearly always large portion of the liver.

The covering membrane is shiny and intact at birth, but because of the B.Vs . it begins to die immediately and become opaque and necrotic within 12 hours and the intestine become exposed to outside.



Treatment;

Exmphalos minor :

Must be Rx rapidly as it liable to strangulation due to narrow neck. Doing operative reduction of the content ,then closure of the small defect.

Exmphalos major:

If the hernia is large, can not reduced it so must choose one of the following Rx:

☆ Conservative Rx : use only for large omphaoceles .use a drying agent e.g 2% aqueous mercurochrome or iodine solution were used for this

purpose but there were problems with toxicity; this resulted in the abandonment of this practice. The use of plastic sheeting (“Op-site”) has been described. We currently recommend silver sulfadiazine ,which prevents infection and results in a good bed of granulation tissue

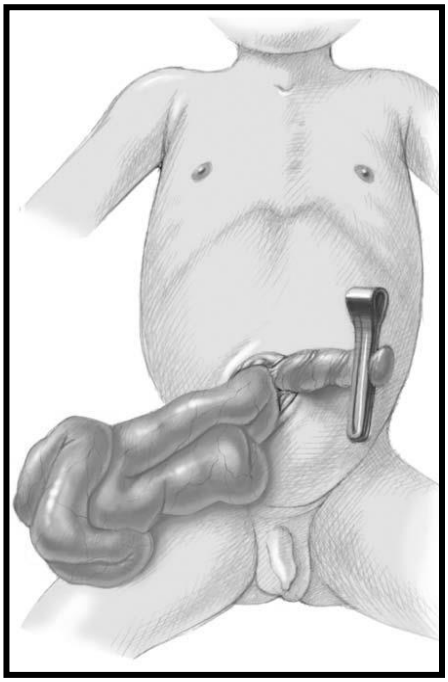
- ☆ Operative Rx : used if the sac is rupture .

Gastroschisis

- ☆ Gastroschisis is an abdominal wall defect (opening) at the base of the umbilical cord that allows intestine (bowel) and other abdominal contents to come outside the body before birth.
- ☆ The anomaly is thought to be the result of a defect at the site where the second umbilical vein involutes.

Definition:

It is herniation of the abdominal viscera through a defect in the abdominal wall that is usually to the right of a normal insertion of the umbilical cord.



Hypospadias

Hypospadias is one of the most common urogenital anomalies, occurring in 3 in 1000 births.

Definition:

It is defined as a hypoplasia of the tissues forming the ventral aspect (ventral radius) of the penis beyond the division of the corpus spongiosum.

There are two main types of hypospadias:

1. The hypospadias with a distal division of the corpus spongiosum with little or no chordee when the penis is erected
2. The hypospadias with a proximal division of thecorpus sponiosum with a marked degree of hypoplasia of the tissues forming the ventral radius, marked by a significant degree of chordee

The causes of hypospadias

The causes of hypospadias remain essentially unknown although several avenues have been explored o explain this congenital defect of the genital tubercle:

- Some endocrine disorders have been described in relation to hypospadias, mainly due to an insufficient secretion of androgens, or insufficient response by the target tissues. However, in very few cases can these disorders be demonstrated.
- Some genetic disorders could explain why hypospadias may be found in several members of the same family.
- Young and old mothers are more prone to carry a baby with hypospadias. Small birth-weight babies and twins also have a higher risk of presenting with a hypospadias. This could be explained by a placental insufficiency.

- The significant increase of hypospadias in the population over the last 20 years raises the role of possible environmental factors such as oestrogen like molecules, pesticides, fertilizers etc.
- Abnormal or insufficient growth factors could also be responsible for these penile anomalies and could also explain the significant complication rate met in this surgery.

Three surgical steps characterize hypospadias surgery:

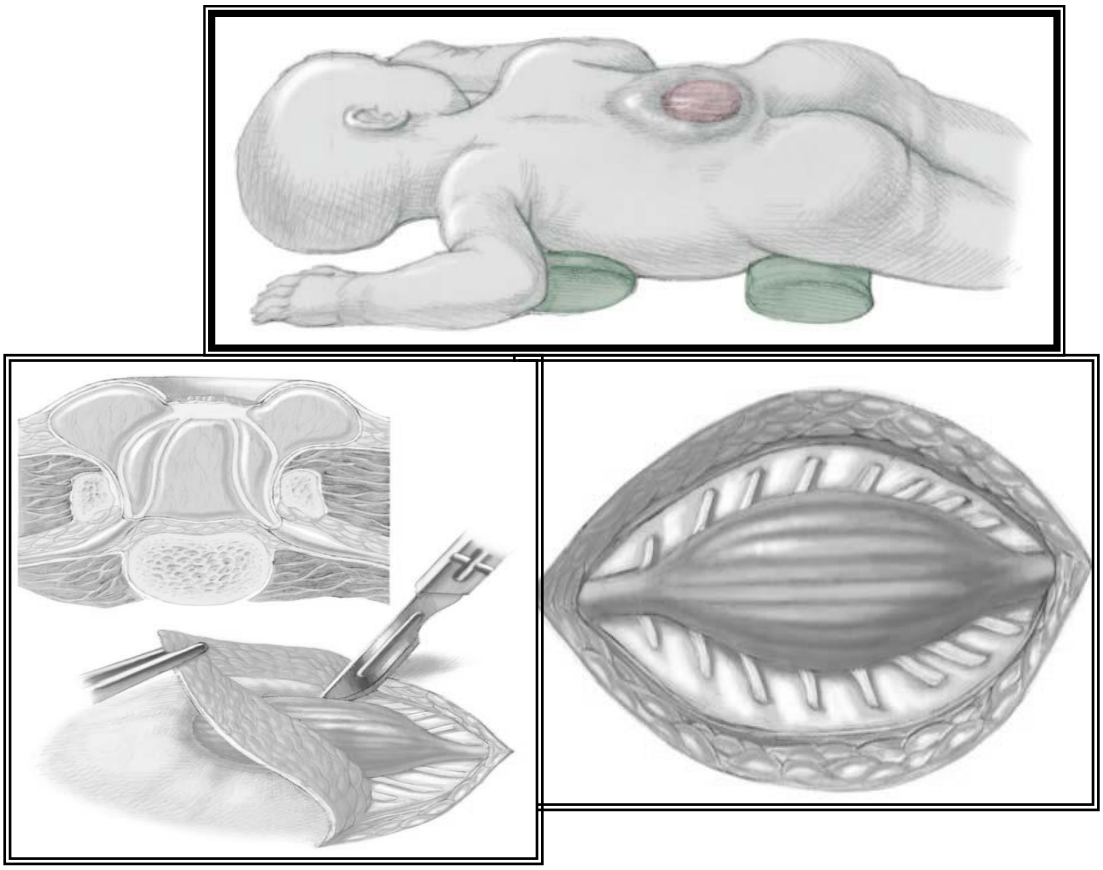
- The correction of chordee, which is essentially the result of the atresia of the ventral radius. Degloving the penis represents the first step of this surgery and straightens the penis in 80% cases.
- Once the penis is straight, the missing urethra should be replaced.
- Once the urethra is repaired, the ventral radius of the penis needs to be reconstructed.

Spina Bifida and Hydrocephalus

- ☆ Disorders of neural tube closure are generally either yelomeningocoele or the less common and less severe meningocoele.
- ☆ Although antenatal screening and the widespread use of peri-conceptual folic acid have reduced the incidence of this debilitating condition, it remains a significant part of the pediatric surgeon's workload.
- ☆ While both types are associated with vertebral body defects, the severity of vertebral anomaly is much greater in patients with myelomeningocoele.
- ☆ Similarly, the extent of associated neurological, bladder, bowel and lower limb abnormalities are significantly greater in this group
- ☆ **In myelomeningocoele** the vertebral arches and overlying vertebral fascia is absent over a variable number of vertebral segments. The spinal cord lies superficially at skin level and cerebrospinal fluid usually leaks from the exposed neural plaque.
- ☆ Neurological development to the lower limbs, the bladder and the bowel is incomplete and virtually all children with myelomeningocoele have some degree of paralysis of the lower limbs, with sensory loss in addition.
- ☆ Muscle power is imbalanced and results in flexion deformity of the hips and hyperextension of the knees. Bilateral hip dislocation and clubfoot deformity are common associated problems.
- ☆ **In meningocoele** patients, the spinal cord is well covered by an epithelial lined sac. This sac communicates with the arachnoid space

and contains cerebrospinal fluid (CSF). Leakage from this sac is rare, as are serious associated abnormalities.

☆ Limb innervation is normal and these patients look forward to normal activities. Screening for bladder function is required, however, as a small percentage are at risk of developing a neuropathic bladder.



General Care principle

◎ The baby is nursed prone or in a lateral position.

◎ Careful attention is paid to keeping the area clean.

◎ Feeds are not restricted.

◎ Weekly estimates of head

◎ Circumference is performed and this is supplemented with cranial ultrasound to monitor progress of the associated hydrocephalus. A ventriculo-peritoneal shunt is inserted when the head circumference

rises precipitously or when the ventricular diameter increases beyond 50–60% of the diameter of the skull.

Hydrocephalus

Definition:

Hydrocephalus is defined as an excessive amount of cerebrospinal fluid (CSF) under increased pressure with abnormal enlargement of the ventricular system.

The incidence is approximately 0.4 to 2.5 per 1,000 live births.

The aetiology

The aetiology of hydrocephalus can be obstruction to the flow of CSF, resulting in non-communicating hydrocephalus, and overproduction of CSF or failure of its re-absorption, described as communicating hydrocephalus.

The pathological process that causes hydrocephalus may be congenital or acquired.

Several conditions can result in hydrocephalus, for instance, congenital malformations, neoplasm, bacterial meningitis, and natal infection with toxoplasmosis.

The three most important malformations that cause congenital hydrocephalus due to obstruction of the CSF flow are aqueductal stenosis, Arnold-Chiari II malformation, and Dandy-Walker malformation.

The clinical features

The clinical features of hydrocephalus in neonates and infants comprise enlargement of the head, which can easily be overlooked in mild cases unless the head circumference is measured repeatedly.

In addition, the shape of the head becomes abnormal.

The anterior fontanel is large and may be bulging, cranial suture

separation may be palpated, and the scalp veins may be distended.
The commonest symptoms in infants and young children with hydrocephalus are vomiting, behavioral changes, drowsiness and headache. Other symptoms comprise failure to thrive, irritability, delayed motor and social development, and mental retardation.

Diagnosis:

Ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) distinguish hydrocephalus from other causes of macrocephaly.
Hydrocephalus is frequently diagnosed with antenatal ultrasound.
The investigation of the patient with hydrocephalus also comprises a TORCH screen and chromosomal analysis if a prenatal infection or a chromosomal abnormality is suspected.
The patient should also undergo an ophthalmological examination, and associated anomalies such as a cardiovascular anomaly should be excluded

Treatment:

Mild hydrocephalus caused by intracranial hemorrhage can be monitored by repeated measurement of head circumference, ultrasound scans and clinical signs of progressive hydrocephalus, and it usually resolves.
In cases with progressive ventricular dilatation, abnormally increased head circumference and clinical features of increased intracranial pressure, the condition requires treatment by surgical diversion of CSF. Today ventriculo-peritoneal shunts are the primary option for infants and children.

Pre-operative care:

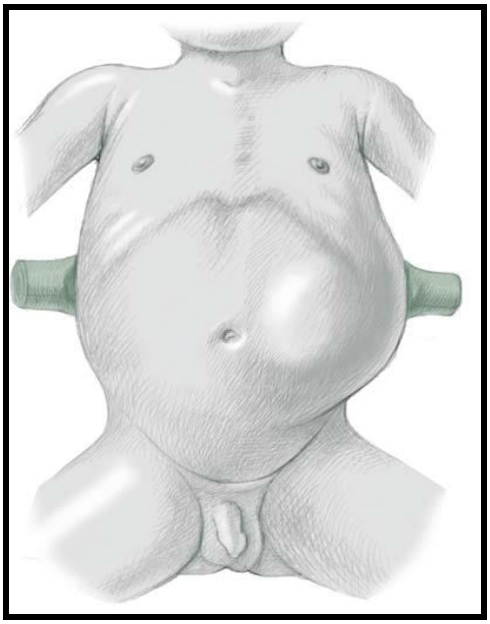
- The pre-operative routines are important to reduce the risk of *Staphylococcus epidermidis* shunt infections to a minimum.
- The patient is carefully washed at least three times during the last 24 h before the operation. The first time ordinary shampoo is used for the hair and the next two times Hibiscrub is used.

- Scurf is treated if necessary. The hair is carefully shaved in the theatre.
- Cloxacillin is administered as prophylaxis at induction of general anaesthesia and post-operatively another two doses are given.

Wilms Tumour

INTRODUCTION

Renal neoplasms in childhood are usually malignant, the most common being nephroblastoma Wilms tumour (WT).



WT presents as a palpable asymptomatic abdominal mass in a toddler, which is discovered when the parents are usually bathing or dressing the child.

Weight loss, malaise, abdominal pain, hypertension and haematuria may be present. Rarely they may present with a varicocoele where the left renal vein is occluded by tumour thrombus.

On examination it is smooth rounded tumour occupying most of the abdomen, and in about 10% of patients the tumour thrombus from the nephroblastoma may invade the inferior vena cava. This may at times

extend to the right atrium causing cardiac dysfunction or even a pulmonary embolus.

The main stay of investigation is imaging; a plain X-ray of the abdomen usually shows a soft tissue mass and calcification may be seen in about 10% of patients.

Abdominal ultrasound confirms that the tumour is renal in origin and can demonstrate a normal contralateral kidney. It evaluates the inferior vena cava for blood flow and for the presence of tumour thrombus within it.

A computed tomography scan will outline the tumour and may show a lesion in the contralateral kidney.Magnetic resonance

image scanning can add a further dimension to renal evaluation with visualisation of blood vessels. Echocardiography may be necessary to exclude the presence of an intra-atrial extension of the tumour thrombus.

The surgical treatment of Wilms tumour involves

Three stages:

(1) Making a diagnosis by biopsy,

(2) Operative excision of the tumour and

(3) Staging of the patient.

BILIARY ATRESIA

- ☒ Biliary Atresia is a rare disease of the liver that destroys the bile ducts, which carry bile from the liver to the intestine. This disease involves the progressive destruction of these bile ducts, especially outside the liver, sometimes inside the liver as well. Because the bile is unable to drain, it builds up in the liver and damages the liver.
- ☒ Biliary atresia is a condition in which the ducts that connect the liver to the intestine and the gall bladder become scarred and blocked. One of the things that the **liver** does is make bile. The bile flows to the intestine through these ducts where it helps **digest** food. The bile blocked in the liver causes scarring in the liver called cirrhosis.
- ☒ The bile trapped in the liver also backs up into the blood and causes the skin and eyes to look yellow. This is called jaundice.

Causes

The cause of biliary atresia is not known. Suggested causes include viral infections or an over-response of the body's own immune system. It is not a hereditary disease and is unlikely to occur more than once in a family. Biliary atresia only occurs about once in every 15,000-20,000 births worldwide.

Symptoms of Biliary Atresia

Symptoms or signs of biliary atresia typically appear within the first two weeks to two months of life. These may include:

- **Jaundice** - A yellow appearance of the skin and whites of the eyes (sclera) can be present in many newborn babies. In Biliary Atresia, this

jaundice does not improve within 1 to 2 weeks. The jaundice is due to the build-up of excess bilirubin throughout the body as the liver isn't able to clear it properly.

- **Urine** - Appears very dark yellow or brown: this is due to the increased bilirubin in the bloodstream, which then passes to the kidneys.
- **Stools** - Appear pale or clay-coloured. This happens because there is little or no bile reaching the intestine to colour the bowel movements.
- **Enlarged Liver** - Feels larger and harder than normal.
- **Poor Weight Gain** - Infants with biliary atresia often do not gain weight. Bile is required to digest and absorb most types of fat.

Diagnosed

Several other liver diseases can give the same symptoms seen in biliary atresia. Therefore a series of tests are required to work out the cause of these symptoms. These tests include various blood tests, urine tests, ultrasound scan of the liver, tests to look at flow of bile through the liver and tests to look directly at the liver tissue (a biopsy).

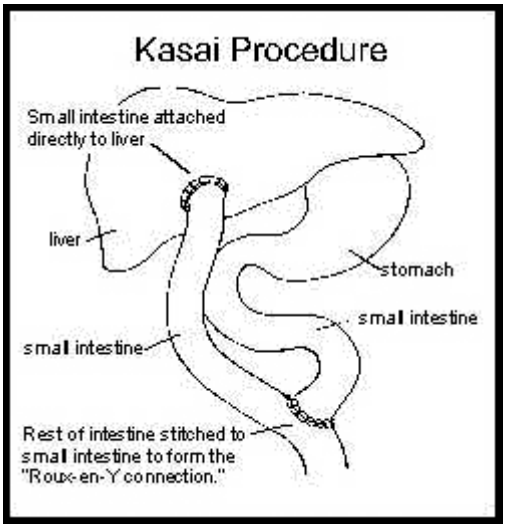
Biliary atresia is usually diagnosed after checking the results of many of these tests.

Treatment

One particular type of operation, the Kasai Procedure, can be done to help re-establish bile flow from the liver to the intestine. This operation involves connecting the liver directly to the small intestine with a loop of the small intestine. This bypasses the blocked ducts, letting bile flow from the small bile ducts inside the liver straight into the intestine.

An early operation can improve the outcome of biliary atresia. 25% of infants will have good bile flow after surgery while 50% will have some bile flow. The remaining 25% will have little or no bile flow and will require liver transplantation. Liver transplantation is the only cure for

biliary atresia and most children needing a transplant do very well long-term



Complications of Biliary Atresia

- **Cholangitis** - An inflammation of the bile ducts caused by bacteria moving up from the bowel.
- **Failure to Thrive** - Lack of bile salts in the intestine, which are needed for fat digestion, result in poor growth and fat-soluble vitamin deficiency.
- **Cirrhosis** - is when scarring to the liver occurs causing irreversible damage. Once cirrhosis develops the functions of the liver start to be interrupted and liver transplantation is considered.
- **Portal Hypertension** - As the liver becomes scarred, veins travelling through the liver become constricted. This impairs the flow of blood and increases the pressure in these veins, specifically in the portal vein (a major vein of the liver system) leading to a big spleen and fluid problems (such as ascites).
- **Ascites** - Ascites is a complication of portal hypertension. This occurs when there is a build up of fluid in the space between the lining of the abdominal wall and the lining of the organs.
- **Varices** - occur when the increased pressure causes blood to flow through smaller veins, which leads them to weaken and swell. Varices can lead to bleeding. This bleeding can cause dark or black-coloured stools and blood-stained vomit. Immediate medical attention is necessary.

Home care after surgery

Care of the incision: The dressing will be off and no special care will be needed.

Bathing: Sponge bathing until 1 week after surgery, then you may give your baby a tub bath.

Medications: You will get a prescription for pain medication and instructions on when to give it. Your baby will have to take several other medications for a long time--a year or longer. These are to encourage bile flow and prevent infection. You will be given prescriptions & instructions. *It is very important to follow the instructions carefully.*

Diet: If you are breastfeeding, the nurses will help you pump and save your milk during the time that your baby can not eat. If you are not breast feeding, your baby will need to have a special formula that is easier to digest than regular formula.

Warning S&S

fever, drainage from the incision, vomiting, pain not relieved by medication, or stools turning white again for more than a day.

UMBILICAL HERNIA

Hernia

A hernia occurs when a section of intestine protrudes through a weakness in the abdominal muscles. A soft bulge is seen underneath the skin where the hernia has occurred.

In children, a hernia usually occurs in one of two places:

- around the belly-button
- in the groin area

A hernia that occurs in the belly-button area is called an **umbilical hernia**. A hernia that occurs in the groin area is called an **inguinal hernia**.

Umbilical hernia

An umbilical hernia occurs when bowel protrudes through an opening in the abdominal wall into the umbilicus (belly button).

Most umbilical hernias do not require surgery because they go away by themselves. It is normal for the bulge to get larger before the hole closes. An umbilical hernia is usually harmless, painless and rarely needs treatment.

Incidence

They are more common in African American children.]

- In about 10 percent of all children.
- More often in African-American children.
- More often in girls than in boys.
- More often in premature infants.

Causes

A hernia can develop in the first few months after the baby is born because of a weakness in the muscles of the abdomen.

Diagnoses

An umbilical hernia is diagnosed by physical exam. No special tests or procedures are required.

Clinical Manifestations

Umbilical hernias appear as a bulge or swelling in the belly-button area. The swelling may be more noticeable when the baby cries and may get smaller or go away when the baby relaxes. If your physician pushes gently on this bulge when the child is calm and lying down, it will usually get smaller or go back into the abdomen.

If the hernia is not reducible, then the loop of intestine may be caught in the weakened area of abdominal muscle. Symptoms that may be seen when this happens include the following:

- A full, round abdomen.
- Vomiting.
- Pain or fussiness.
- Redness or discoloration.
- Fever.

Treatment

By 1 year of age, many umbilical hernias will have closed on their own without needing surgery. Nearly all umbilical hernias will have closed without surgery by age 5.

Home care after surgery

Care of the incision: There is usually a large bulky dressing on the umbilicus after surgery. Keep it dry. Remove it according to your surgeon's instructions.

Activity limitations:

0-8 months: normal activity for age.

8 months-21/2 years: Be careful of toddler accidents.

21/2 years -12 years: Quiet supervised play for 24-48 hours. No running, contact sports or vigorous activity for 4 weeks.

Diet: Your child may have his/her usual diet. It is not unusual for your child to be nauseous after surgery. If this is true, give small frequent amounts of juices or ginger ale. Some children become constipated after surgery. Give plenty of fluids to prevent this. Your child should not go more than 48 hours without a bowel movement.

Bathing: Sponge bathe your child until the dressing is removed.

Medication: Give medicine around the clock for the first 24-48 hours and as needed after that for three to four days. Please call if your child is uncomfortable. Some children require more medicine than others.

Warning S&S

Please call the surgery team of there is bleeding or drainage from the dressing, fever., vomiting or less peeing than usual.

What are the long term consequences?

The most common complications related to this surgery are bleeding and infection. Complications occur in less that 1% of the population. There is no effect on growth and development.

Inguinal Hernia and Hydrocele

Incidence

- Hernias and hydroceles are among the most common pediatric surgical problems.
- The incidence of indirect inguinal hernia in the term neonate is 3.5-5%. Premature infants have a higher incidence of approximately 9-11%. Inguinal hernias more common in boys (male: female ratio is 5:1 to10:1). Sixty percent of inguinal hernias occur on the right side, while about 30% occur on the left. Ten percent occur as bilateral hernias.
- Bilateral hernias are more common in premature infants (45-55%) and females.
- Indirect inguinal hernias and hydroceles are known to have familial tendencies, but true heredity factors have not been clarified.

Etiology

The processes vaginalis is an elongated diverticulum of the peritoneum which accompanies the testicle upon its descent into the scrotum. It pierces the anterior abdominal wall at the deep (internal) inguinal ring which is located just lateral to the deep inferior epigastric blood vessels.

In most individuals, the processes obliterates during the ninth month of intrauterine life or soon after birth. If that channel
remains open, intra-peritoneal fluid will slowly accumulate in the structure forming a communicating hydrocele (also known as hernia/hydrocele) If the processes is wide enough, intestines, ovaries, or omentum can herniate into the inguinal canal forming an indirect hernia. Should the processes vaginalis obliterate near its origin remain patent distally fluid may accumulate forming a non-communicating hydrocele. If the processes obliterates proximally and distally but remains patent in its mid portion then it is known as hydrocele of the cord.

Direct inguinal hernias are occasionally identified in children. The abdominal wall defect is in the floor of the inguinal canal within the confinement of Hasselbach's triangle.

Anatomically, Hasselbach's triangle is that area bordered superolaterally by the inferior epigastric vessels, inferiorly by the inguinal ligament, medially by the rectus abdominus muscle. Direct inguinal hernias are believed to occur secondary to structural weakness. Femoral hernias occur inferior to the inguinal ligament within the femoral canal, just medial to the femoral vessels and are extremely rare in children.

Clinical Presentation

- A bulge (swelling) in the groin which at times may extend into the scrotum is by far the most frequent sign.
- The bulge may appear and then disappear with some regularity especially during straining, crying, or coughing.
- Although sharp pain is usually not associated with herniation, discomfort that occurs in some babies is easily overlooked.
- Occasionally constipation, "colicky-baby" syndrome, and even regurgitation are present.
- In the very young, the initial presentation may be an episode of incarceration.
- In this scenario, the baby is more symptomatic, the bulge is firm and tender to touch, the groin and scrotum may be erythematous, and vomiting or poor feeding are frequent.
- A history of recurring groin swelling which the parents or the pediatrician can reduce is a strong indication that a hernia is present.

Diagnosis

- The physical examination in many is so characteristic that only observation is necessary to make the diagnosis. The examiner palpates the cord to ascertain if bowel or other structures are present.
- Diagnostic confirmation is made when the contents of the hernia are reduced into the peritoneal cavity.
- Hydroceles, even the communicating variety, are difficult to reduce though many reduce spontaneously when the child is recumbent over several hours.
- Palpation of the cord may elicit the "silk glove" sign (rubbing together of the opposing peritoneal membranes of the empty sac).
- The cord may feel thickened in comparison to the contralateral side. Increases in intra-abdominal pressure (i.e., coughing, crying, exhaling against an occlusion such as thumb in mouth, blowing bubbles, etc) may help demonstrate the hernia.
- In the face of a suggestive history but no concrete findings, repeated examination in 2-3 weeks is recommended.
- Plain films are also useful to distinguish between an acute hydrocele, for which an operation can be delayed, and incarceration which requires immediate attention.
- When in doubt—operate! Untreated incarceration leads to bowel necrosis and/or testicular ischemia.

Treatment

The reason for repairing an inguinal hernia is to prevent incarceration. Since the incidence of incarceration is inversely related to age, the younger the patient—the sooner the repair.

Premature babies should have their hernias repaired just prior to discharge from the hospital.

The timing of repair is less clear with hydroceles.

In most centers,

hydroceles are not repaired until the baby is 12-18 months or older.

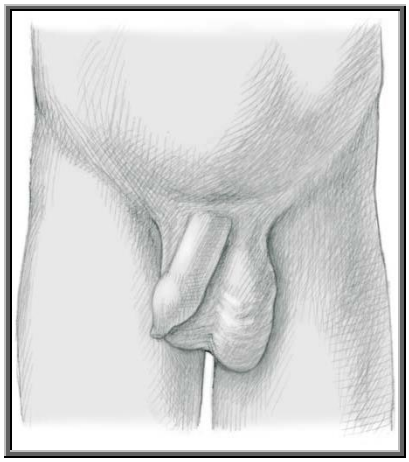
Approximately 90-95% of all hydroceles resolve spontaneously in the first few months of life.

If a hydrocele becomes very large and tense, earlier repair can be considered.

If a hydrocele cannot be differentiated from a hernia, operation is indicated.

Varicocele

- Testicular varicocele is characterized by variceal dilatation of the veins in the pampiniform venous plexus secondary to incompetent valves in the testicular vein.
- Varicoceles are almost always localized on the left side. This is supposedly related to the fact that the left testicular vein drains via the left renal vein,which offers higher resistance to the bloodstream than the right testicular vein, which enters vena cava directly.
- The age group most frequently affected are older boys and adolescents. Symptoms are rare. Sometimes, an ill-defined discomfort in the way of a dragging sensation in the scrotum is reported.



Etiology

Varicocele occurs because of increased hydrostatic pressure within the gonadal veins. This may be due to either incompetent venous valves or venous obstruction.

Its relative greater occurrence on the left side is related to the fact that the left gonadal venous drainage is to the left renal veins rather than directly to the IVC as occurs on the right side.

Pathophysiology

A varicocele increases blood surrounding the testis, thereby raising the testicular temperature as heat dissipates from the venous blood. Spermatogenesis is decreased with an increase in testicular temperature. In addition, hormone-like substances secreted by the affected testicle may adversely influence contralateral testicular function.

Clinical Presentation

- Varicocele occurs predominately in postpubertal teenagers and young adults and occurs almost exclusively on the left side (80-90%).
- Most boys describe enlargement of the cord and testicle after physical activities, coughing or straining.
- The lesion is a complex elongated "cord," at times, described as a "tangle of worms."
- Standing and straining may demonstrate the varicocele. Tenderness is not a common finding. With the patient supine and resting, the swelling will recede.
- The differential diagnosis includes hernia, hydrocele, and tumor. Diagnostic testing is rarely required for varicocele.

Treatment

Operative interruption of the gonadal vein is curative. Traditionally, this is carried out through an inguinal exposure.

Recent reports suggest that these patients can be successfully treated laparoscopically. Using this technique, the main gonadal vein trunk is interrupted in the retroperitoneum.

Some surgeons ligate the gonadalartery as well (with the hope of reducing the recurrence rate) and rely on the collateral blood supply (perivas, gubernacular and pudendal vessels) to maintain testicular viability.

This technique is known as the Fowler-Stevens maneuver, which is commonly used to treat intra-abdominal cryptorchidism.

RECTAL IRRIGATIONS

Rectal Irrigations

Supplies:

Irrigation Catheter
Syringe
Lubricating gel
Normal saline
Cool to room temperature
Make a new solution each time

Fill the syringe with ____ml of normal saline solution.

Lubricate the tip of the catheter and insert it into the rectum.

Gently push the saline through the catheter.

Draw back on the syringe and withdraw the saline.

Do the irrigations _____ times daily.

Make sure all or at least most of the solution is withdrawn.

Signs to watch for and call us or come in to see us:

Vomiting green
Fever
The abdomen is bloated or distended and does not go down after the irrigation
Nothing is expelled with the irrigation
Stools are bloody, very foul smelling, or explosive
Anything about your child's behavior that you are concerned about