Congenital anomalies

Prepared

By

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Congenital Anomalies

Definition:

An anomaly is a structural defect present at birth. Some defects may be compatible with life but need treatment that could be delayed. Others are in compatible with life and must be repaired immediately.

Causes of Fetal malformations:

Several factors are known to produce malformations of the developing fetus. These are environmental factors including:

1- **Drugs**
2- **Radiation**
3- **Viruses like rubella virus**
4- **Genetic traits**

Common anomalies, of the different systems are as follows:

Gastrointestinal system:

Anomalies of the Intestine:

Hirshsprung's disease: (congenital A ganglionic Mega colon).

-There is congenital absence of parasympathetic ganglion nerve cells of a part of intestine usually in the distal end of the descending colon/the affected portion has no peristalsis.

-It is common in males & females. It then hypertrophies with accumulation of feace.

Symptoms and signs:

- -Failures to pass meconium within 24-48 hours after birth.
- -vomiting.
- -Abdominal distention.
- -Food refusal.

Diagnosis:

- In neonate: usually based on clinical signs of intestinal obstruction and failure to pass meconium.
- On examination, the rectum is empty of faces.
- Rectal biopsy
- Anorectal manometery.

Treatment:

- -Surgical removal of Aganglonic portions of the bowel in order to permit normal bowel motility and establish continue by improved functioning of the internal anal sphincter.
- Surgery consists of first stage and second stage.

Acquired Intestinal Defect: Intussusceptions: It is an acquired type of intestinal obstruction. It is the invagination of a portion of the bowel in to the portion immediately distal to it; thus the blood supply is out off and if the condition continues, the bowel will become gangrenous. Death may result if the condition is not recognized and corrected sometimes a simple reduction occurs.

Incidence:

- -Intussusceptions is one of the most frequent causes of intestinal obstruction in children between the ages of 3-5 years.
- -The male to female incidence is 2:1.
- -The peak incidence is in summer due to increased gastroenteritis.
- -This may explain the frequent occuranous of intussusceptions at the age of 3-12 months (age of weaning).

Symptoms and signs:

Symptoms:

- -The infant awakens from sleep by severe abdominal colic, screams and draws his knees up into the abdomen. These attacks alternate with intervals of apparent well-being during which the infant asks for feeding.
- -With each attack of pain there is pallor, apathy and lethargy.
- -Vomiting follows the attacks of colic in 85% of cases.
- The infant passes mucous and blood per rectum (red currant jelly stool).

***Signs*:**

- -Emptiness in the right iliac fossa (Signe de Dance).
- -A sausage shaped mass may be felt.
- -Distention is usually absent in early cases. If it occurs it denotes possible perforation or gangrene.
- -Digital rectal examination revels bloody mucous in 60% of cases.

Investigations of infantile ilocacal intussusceptions:

- -Blood picture shows anemia.
- -Gastro Raphine enema. (claw sign).

- -Ultrasound examination.

Treatment of infantile ilocacal intussusceptions:

- -Resuscitation of the I.V. infusion of dextrose and saline.
- -Antibiotics are prescribed.
- -A nasogastric tube is inserted.
- -In early cases a trial of hydrostsstic reduction is performed:

*The pressure should not exceed120cm of sterile water, air, gastrographine and normal saline.

Contraindication hydrostsstic reduction:

- -Doubtful diagnosis.
- -Late cases.
- -Presence of abdominal distension or rigidity.

Surgery:

-Is needed when hydrostsstic reduction fails.

-A lapartomy.

Prognosis of infantile ilocacal intussusceptions:

-Mortality high in dangerous cases.

- Intussusceptions: may occur in 2% of cases.

Congenital Heart Diseases

1- Congenital heart disease with no shunt

- Coarcutation of aorta
- Aortic stenosis
- Pulmonary stenosis

2- Congenital heart disease with shunt and cyanosis

- Fallout's tetralogy
- Transposition of great vessels.

3- Congenital heart disease with shunt but no cyanosis

- Patent ducts arteriosus P.D.A
- Interatrial septal defect A.S.D
- Interventricular septal defect V.S.D

Congenital Heart Disease With No Shunt

Coarcutation of aorta

A narrowed segment along the aorta it associated with duct arteriosus, characterized by cardiac murmur, hypertension due to left heart failure, weak or absent femoral pulse and left ventricles hypertrophy with or without failure *Investigation:-* E.C.G, x-ray, aortogram

Treatment:- surgical correction in late childhood

Congenital heart diseases with shunt but no cyanosis

Atrial septal defect (A.S.D) is small defect cause minimal change in cardiac function, diagnosed accidentally, large defect are associated with failure to thrive, repeated chest infection and exertional dyspnea, murmur.

Investigation: x-ray, E.C.G, Echo (electrocardiograph) and catheterization.

Treatment: surgery

Ventricular septal defect (V.S.D) is small defect is asymptomatic, and is diagnosed accidentally during examination, charactrized by harsh murmur, loud and always associated with a thrill large defect characterized by recurrent chest infection, failure to thrive and exertionaldyspnea, may be cyanosis on crying or heart failure , pulmonary hypertension, develops reversal of the shunt many occurs leading to persistent cyanosis.

Investigation*:* Echocaidiogram, E.C.G and x-ray

Treatment*:* in small defects antibiotics in septic procedures and in large defect surgical repair

Patent dictus arteriosus

Is persistence of a fetal duct between the pulmonary artery and the aorta.

Clinical picture depends on the size of the duct, small ducts may be asymptomatic and discovered accidentally on routine examination of the heart, wide ducts cause failure to thrive, dyspnea, sometimes heart failure, pulmonary hypertension, murmur heard on the base of the heart.

Investigation E.C.G - x-ray – catheterization.

Treatment ligation as early as possible specially with heart failure.

General Aspects of pre and post-operative pediatric care:

A. Transportation of the newborn:-

1- Safe means of transportation with a heated portable incubator and available oxygen supply is needed so as to maintain the infant's body temperature and 02 level in the blood

2- Equipment for suctioning to remove secretions is needed (as in esophageal atresia).

3- A Nurse should participate in the transfer of the baby to observe him during that time and give appropriate care proper positioning of infant)

4- All pertinent infant information should accompany the infant as he goes from one health agency to another.

B. Pre-operative care:

1- Psychological preparation of the child (according to is age) this is aimed at prevention fears common to children like fear of separations, fear of death. The nurse should explain to the child and his families what is going to occur.

2- Except in emergency situations, children should preferably be free of respiratory complication and sings of malnutrition.

3- Most children must have nothing by mouth before surgery but the length of the time the child remains NPO will depend on age (for newborns & infants 3-4 hours is usually enough).

4- The incision over or the part involved in surgery must be washed and inspected . Shaving may be needed .

5- The mouth should be checked for loose or missing teeth should be charted in the child's record .

6- Remove barretts and pins from the child's hair.

7- Clothing should be warm and loose. The child should be dressed in a hospital gown and under pants only .

8- Check the child's identification band to see that is legible and secure. If not it needs to be replaced prior to surgery.

9- Pre-medication : sedatives and analgesics are usually given two hours before surgery except in emergency situations) .

10- The nurse should chart whether the child has passed urine and had a bowel movement . Enemas are never done routinely but may be ordered in some conditions .

11- Prior to taking the child to surgery, specially in newborns and infants, nostrils should be carefully cleansed (hard crusts may be softened with a

solution of sodium bicarbonate, normal saline, or even warm water) This is necessary because crusts may be obstructing the air way)

12- The child may need to take his favourite toy with him to surgery. Ideally, he should be allowed to keep the toy with him until he is under the anesthetic.

13- Parents should be allowed to accompany their children to the operating site if they so desire.

14- Parents should be told where to wait during surgery, whether the child will go to recovery room after surgery or directly to his unit.

***C*- Post- Operative care:**

1- After return from the operating room, the child's general condition must be closely observed (a) vital signs, especially temperature

(a) Airway must be kept patent

(b) newborn babies must be kept in warm cot or incubator .

2- Until the child is responsive and alert, he should be kept on his side (for secretion and vomits 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 signs of shock:

8- Oral fluids may be started after the following criteria are observed:

(a) color of aspirate is clear

(b) peristaltic movements are heard

(c) flatus or gases are passed.

9- 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.

10- Sedatives are used according to prescribed orders and child's need.

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