Anorectal anomalies (imperforate anus)

Anorectal anomalies are the most common cogenital malformations of the newborn that are incompatible with life. Minor abnormalities of the anus and rectum occur in 1 in 500 living newborns, while major abnormalities occur in 1 in 5000 living infants.

Anorectal malformations comprise a wide spectrum of diseases that affect boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tract.

Malformations range from minor , easily treated defects that carry an excellent functional prognosis, to complex defects that are difficult to treat, are often associated with other anomalies, and carry a poor functional prognosis.

Incidence :

A major anomalies occur in 1 of 5000 living infants. Minor abnormalities occur in 1 of 500 living newborns. These malformations may range from simple imperforate anus to include other associated complex anomalies of genitourinary and pelvic organs, which may require extensive treatment of fecal, urinary and sexual function.

Anorectal malformations may occur in isolation or as a part of VACTERL association. The enormity of the problem can be judged by the fact that 1 out of every 5000 newborn babies are liable to have this condition. On rough estimates nearly 15,000 babies with anorectal anomalies are born every year in densely populated state of uttar Pradesh,with one of the highest birth rates in the country.

Etiology:

Although the etiology remain unknown, a slight genetic predisposition appears to exit.
Familial incidence
Closely associated with other congenital anomalies

V- vertebral
A- anorectal
C- cardiac
T- tracheoesophageal
R-renal and
L-limb abnormalities.

Embryology:

Early in the embryonic life there is a commom chamber called the cloaca- in to which open the hindgut and allantois. The cloaca soon shows a subdivision in to a broad ventral part and a narrow dorsal part.

These two parts are separated from each other by formaton of the urorectal septum , which is first formed in the angle between the allantois and the cloaca.

The ventral subdivison of the cloca is now called the primitive urogenital sinus and gives origin to the some part of the urogenital system. The dorsal part is called the primitive rectum. It forms the rectum, and a part of anal canal. The urorectal septum grows towards the cloacae membrane and eventually fuses with it . The cloacae membrane is now divided in to a ventral urogenital membrane, related to urogenital sinus and a dorsal anal membrane related to the rectum.

Mesoderm around the anal membrane becomes heaped up with the result that the anal membrane comes to lie at the bottom of a pit called the anal pit or proctodeum. The anal pit contributes to the formation of the anal canal. Failure of the membrane separating the lower rectum, anus and urogenital tract at the 8^{th} week of fetal life ranges from mild and stenosis to complex deformities such as rectal atresia and fistula.

Classification:

Anorectal malformations are classified according to the newborn's gender and abnormal anatomic features, including genito urinary defects.

WING'S SPREAD CLASSIFICATION IN USA IN 1984: Anorectal malformations present with a wide spectrum & therefore, there have been numerous attempts at classification. To overcome the drawback of the various classification systems, some of which were too simple and others too complicated, a group of surgeon from all over the world met at wing spread in USA in 1984 and formulated a classification which is reproduced the essential of a classification for a preliminary student are as follows.

- The terms high, intermediate and low are in relation to the terminal end of the bowel remaining above(high), with in intermediate or below the levator ani muscle(pelvic floor)which is the main muscle of the continence.
- Most female babies have low or intermediate anorectal malformation while the reverse is for males.
- Female babies usually have a fistula from the terminal end of the bowel opening externally, which in male this fistula is usually well hidden and therefore , investigations are required to determine the level of termination of bowel.

High:

- Anorectal agenesis with rectoprostatic fistula or recto vaginal fistula
- Urethral fistula, fistula with or without rectal atresia

Intermediate:

- Recto bulbar recto vestibular urethral fistula

- Recto vaginal fistula
- Anal agenesis with or without fistula

Low:

- Anocutaneous and vestibular fistula
- Anal stenosis, anocutaneous fistula
- Anal stenosis

Rare malformations

- Cloacae formations

BASED ON SEX OF CHILD ARM IS AS FOLLOWS:

Males:

- Perineal fistula- no colostomy is needed
- Recto urethral fistula
- Recto vesicle fistula
- Imperforate anus without fistula
- Rectal atresia

Females:

- Perineal fistula
- Vestibular fistula
- Persistent cloaca
- Imperforate anus without fistula
- Rectal atresia

a. Perineal fistula:

- Perineal fistula is the simplest defect in both sexes. Patients have a small orifice located in the perineum, anterior to the center of the external sphincter, close to the scrotum in the male or to the vulva in the female.
- Male patients frequently have in their perineum “a bucket handle” type malformation or block ribbon type structure that represents a sub epithelial fistula with meconium.
- These patients usually have a well formed sacrum, a prominent anal dimple, the diagnosis is established by simple perineal fistula.

b. Male recto urethral fistula:

- In cases of recto urethral fistula, the rectum communicates with the lower part 0f the urethra(bulbar urethra)or upper part of urethra(prostatic urethra)
- The sphincter mechanism is satisfactory

- The sacrum may have different degrees of hypo development , particularly in case of recto urethral prostatic fistula.
- Most of these patients have a well formed midline perineal groove and an anal dimple.
- Those with a recto prostatic fistula have a poorly developed sacrum and frequently a flat perineum.

c. Recto vesical fistula:

- In this condition , the rectum communicates with the urinary tract at the level of the bladder neck
- The rectal sphincter mechanism is poorly developed
- The sacrum is frequently deformed and is often absent.
- The perineum looks flat

d. Imperforated anus without fistula:

- The defect is same in the both sexes
- The rectum is completely blind and is usually found approximately 2cm above the perineal skin.
- The sacrum and sphincter mechanism is well developed.

e. Rectal atresia:

- These patients will have a normal anal canal and a normal anus.
- The obstruction is present about 2cm above the skin level.
- These patients require protective colostomy.

Female patients:

A. Ano vestibular fistula:

- The rectum opens in the vestibula of the female genitalia immediately outside the human orifice
- The sacrum is normal
- Perineum shows a prominent midline groove a noticeable anal dimple . All these features indicate that the sphincter mechanism is intact.
- A protective colostomy must precede the complete repair.

B**. Persistent cloaca:**

- In this anomaly cloaca, rectum, vagina & urinary tract meet & fuse into a single common channel
- The perineum shows a orifice located immediately behind the clitoris
- The length of the common channel varies from 1-10 cm. if its length less than 3 cm the patient will have developed sacrum & good sphincter . If the length is more than 3cm, it

indicates poor prognosis with poor sphincter mechanism & poor sacrum

- Most patients will have large vagina filled with mucous secretions (hydrocolpos)

Patho physiology :

During the embryonic development the cloaca becomes the common channel for developing urinary, genital and rectal systems. The cloaca is divided at the 6^{th} week of gestation in to an anterior urogenital sinuses and posterior intestinal channel by an urorectal septum. After the lateral folds joins the urorectal septum, seperation of urinary and rectal segments take place. Further differentiation results in anterior genito urinary system & the posterior anorectal channel. An interruption of this development will lead to incomplete migration of the rectum to its normal perineal position.

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Further differentiation results in genito urinary channel & ano- rectal channel

Interruption in these development

Incomplete migration of rectum

Anorectal malfromation

Clinical manifestations:

- Absence of anal opening
- Other symptoms include:
- abdominal distension
- Vomiting

- Absence of me conium or presence of me conium in the urine
- Flat perineum
- Absence of interlineal groove
- The appearance of perineum alone does not accurately predict the extent of defect & associated anomalies; genitourinary & spinal/ vertebral anomalies associated with anorectal malformations should be considered when an anomaly is noted.
- Esophageal atresia with or without tracheo-esophageal fistula , cardiac defects & spinal or vertebral anomalies may occur in association with anorectal malfromations, & the infants should be carefully evaluated for presence of those & other anomalies.
- A perineal fistula may be diagnosed by clinical observation. The presence of prominent anal dimple and a band of skin tissue commonly known as a bucket handle is indicative of a perinea fistula

Conditions is usually discovered immediately after birth or within several hours:

- Absence of anal opening
- Misplaced anal opening
- Anal opening near vaginal opening in female
- Thermometer , small finger or rectal tube cannot be inserted in to the rectum
- me conium stool is absent
- Stool passed by way of vagina or urethra may appear as green- tinged urine
- Progressive abdominal distension
- Fistula likely to be present
- Vomiting if infant is fed.

Diagnostic evaluation:

- The diagnosis of an anorectal malformation is based on the physical findings of an absent anal opening & for other signs & symptoms such as abdominal distension, vomiting etc.

- Search for associated congenital anomalies like genito urinary, spinal/ vertebral anomalies and esophageal atresia with or without TEF.
- Perineal fistula may be diagnosed by **clinical observation** - the presence of prominent anal dimple and a band of skin tissue commonly known as bucket handle is indicative of perinea fistula.
- **Abdominal & pelvic ultrasosnography** - is performed to further evaluate associated anomalies involving the urinary tract.
- Other diagnostic examinations that may be performed include
    - **MRI**
    - **Radiography**
    - **Fluoroscopic examination of pelvic anatomic contents**

    - The most important decision regarding a newborn with a anorectal malfromation

is whether the patient needs a diverting decompression colostomy and emergency urinary diversion for an associated uropathy.

Diagnosing male patient:

- Good clinical evaluation and a urinalysis will provide sufficient information in 80-90% of patients.
- At birth the bowel is not distended, so, the clinical and radiological evaluations are not reliable before 16-24 hours
- A piece of gauze is placed around the tip of penis and check for the me conium filtered through the gauze.
- The presence of meconium in the urine and a flat bottom is considered indications to create a protective colostomy.
- Clinical findings consistent with the diagnosis of a perineal fistula represent an indication for an amplest without a protective colostomy.
- Sometimes none of the clinical signs already described becomes evident after 24 hours of observation; a radiologic evaluation is indicated to determine the position of rectal pouch.
- Invertogram - with the infant inverted (prone) and an opaque marker at the anal dimple, air ascending in to the rectum and lower bowel outlines the location of the pouch in relation to the anal depression.
- When this is separated from the skin by more than 1cm, the patient needs a colostomy.
- During the first 24 hours of life, all these patients need an obstructive uropathy.
- Wangensteen - Rice X-ray (upside down position)- a) limited accuracy in locating rectal pouch. B) useful only after infant is 24 hours of age.

Diagnosing for female child:

- More than 90% of the time the diagnosis can be established by a meticulous perineal inspection. These patients must be observed during the first 16-24 hours of life.
- The presence of single perineal orifice is pathognomonic of a cloaca.
- A palpable pelvic mass (hydrocolpos) reinforces the suspicious of a cloaca.
- Vestibular fistula can be diagnosed by separating the labia to see the vestibule. The rectal orifice is located somewhere between, the female genetalia and the centre of the sphincter.
- Anterior anus: the rectal orifice is located between the female genetalia and center of the sphincter and is surrounded by skin.
- invertogram
- Ultra sound study of the abdomen during the first 24 hours of life, because patients with persistent cloaca have highest urologic defects.

Therapeutic management:

The primary management of anorectal malformation is surgical. Once the defect is identified steps are taken to rule out associated life -threatening defects, which needs immediate surgical intervention, provided there are no immediate life-threatening problems, the newborn is stabilized and made nil per oral for further evaluation.

- I.V fluids are provided to maintain glucose and fluid balance.
- A current recommendation is that surgery be delayed atleast 24 hours to properly evaluate for the presence of fistula and possibly other anomalies.
- For the newborn with a perinea fistula an amplest is performed, which involves moving the fistula opening to the centre of the sphincter and enlarging the rectal opening.
- A program of anal dilations is usually initiated when the child returns for the 2weeks checkup.
- Feeding is started as soon after surgical repair and breast feeding is encouraged, because it causes less constipation.
- In neonates with anomalies such as cloaca (females),recto urethral prostatic fistula (males), & vestibular fistula (females), a descending colostomy is performed to avoid fecal contamination of the distal imperforate section and subsequent urinary tract infection in infants with urorectal fistulas.
- The posterior sagittal anorectoplasty (PSARP):-
    - It is a common surgical procedure for the repair of anorectal malformations in infants approximately 1 month after initial colostomy. In this procedure the repair is made via a posterior midline sacral approach unordered to direct the different muscle groups involved without damaging strategic innervation of pelvic structures , so that optimal postoperative bowel continence is achieved.
- A laprotomy may be required if rectum is unidentifiable by the posterior approach.
- Additional management following successful repair involves a program of anal dilations, colostomy closure and a bowel management program.

Prognosis:

- The long term prognosis dependes on such factors as the type of defect, anatomy of sacrum 7 rectum, quality of muscles, & the success of the surgery.
- Parents are instructed in perineal & wound care or care of colostomy as needed
- Anal dilations may be necessary for some infants.
- Parents are advised to observe stooling pattern & to observe signs of anal stricture or complications.
- Information on dietary management & administration of medications is included in counseling
- Nurses have a vital role in assisting families of children with anorectal malforamtions to provide optimal care for the child so that bowel management is successful & quality of life enhanced for the child & family.

- The presence of a flat or rocker bottom or no midline groove will usually have a poor prognosis for bowel continence because of associated neurologic, muscular & anatomic problems. When the internal anal sphincter is absent, incontinence is a common longterm problem.
- These children may achieve socially acceptable continence overtime with the aid of a bowel management program.
- Other potential complications following surgical treatment of anorectal malformations include strictures, recurrent recto urinary fistula, mucosa prolapse and constipation.

Nursing considerations:

After the diagnosis of occlusion has been made gastric suction may be used . If surgery is to be done , there may be no specific preoperative preparation except for withholding oral feedings & giving parental hydration.

Complete obstruction in a male infant must be relieved at once because the stool cannot be passed. An emergency is not likely in a female since the fistula probably exits in to vagina, perineum, or fourchette. The outlet of the fistula can be found on inspection.

The surgical procedure depends on the type of anomaly. If anal membrane atresia is present, the physician is able to incise the membrane or perforate it with a blunt instrument. No further treatment except dilation to prevent scar formation is required. If the distance between the anal dimple & the blind end of the colon is not more than 1.5cm, correction is made through the perineum. Fecal continence can be expected after such repair, unless surgery was not successful. If a true low rectovaginal fistula is present, the anus will have to be transplanted dorsally.

In the high types of imperforate anus (a distance greater than 1.5cm between the anal dimple & blind end of the colon, a colostomy is done , with further intestinal repair & closure planned in 6-12 months. Alternatively, the colon can be brought down through the anal dimple by an abdominal perineal procedure.

Nursing care plan:

Assessment:

1. Perform physical assessment of newborn for abnormalities.
    A. presence of perineal fistula.
    B. meconium coming from vagina or presence of me conium- stained urine.
    C. no anal opening or inability to pass thermometer in to the rectum.
2. Perform thorough examination for other congenital anomalies.
3. Assess parents level of understanding of condition & ability to cope with infants surgery

Nursing diagnosis:

Preoperative:

Risk for injury of infant related to inability to evacuate rectum with anomalies before surgery.

Postoperative:

- Risk for infection related to surgical incision of anoplasty.
- Risk for impaired skin integrity related to ostomy.
- Risk for fluid volume deficit related to restricted intake
- Altered nutritional status less than body requirement related to inability to feed.
- Family coping: potential for growth related to increased needs for infants
- Risk for injury after definitive repair surgery.

Nursing intervention:

Preoperative:

Nsg diagnosis: Risk for injury of infant related to inability to evacuate rectum with anomalies before surgery.
Goal: maintaining stability before surgery.
Interventions:

- Withholding feedings. Note any vomiting; colour and amount
- Minimize energy expenditure due to altered nutritional status.
- Maintain nasogastric tube passed to decompress the stomach. Measure the abdominal girth.
- Observe the patient carefully for any signs of distress & report. Check vital signs frequently.
- Use an isolate or radiant warmer to maintain temperature stability.

Postoperative:

Nsg diagnosis: Risk for infection related to surgical incision of anoplasty.
Goal: preventing infection of suture line
Interventions:

- After anoplasty, do not put anything in rectum.
- Expose perineum to air
- Position the infant for easy access to perineum for cleansing and minimal irritation to site (ie place the infant on abdomen possibly with hips elevated, to prevent pressure on perinea surfaces; turn side to side.
- Observe for redness, drainage, poor healing.

Nsg diagnosis : Risk for fluid volume deficit related to restricted intake

Goal: maintaining fluid and electrolyte balance
Interventions:

- Start oral feedings as ordered (usually within hours after an anoplasty)
- Monitor for rectum of peristalsis. When primary repair is done: nasogastric suction may be maintained until feedings are started.
- Monitor potential fluids and discolouration when oral intake is sustained.
- Report any vomiting or stooling.

Nsg diagnosis : Family coping: potential for growth related to increased needs for infants
Goal: strengthening coping.
Interventions:

- Ensure the parents that colostomy is temporary.
- Encourage the parents to participate in case of the child & to provide emotional security for the child.
- Provide thorough teaching programme for special case needed at home.

- colostomy care
- anal dilation to prevent a stricture at site of anastomosis from scar tissue.
- Initiate referral to community nurse, especially if the parents are particularly anxious about carrying for the child at home.
- Encourage the parents to talk about their concerns
- If mother prefer to breast- feed, encourage frequent pumping to establish milk supply.

Family education;

- Help the parents to understand situation that may be encountered as a result of imperforate anus as the baby gets older.

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