MINISTRY OF HEALTH OF UKRAINE
Higher National Educational Institution of Ukraine
"Ukrainian Medical Stomatological Academy"

«Approved»
on meeting the
department of Pediatric Surgery
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The Head of the department  O.V. Pelypenko

METHODICAL INSTRUCTIONS

FOR STUDENTS` SELF-WORK

WHILE PREPARING FOR PRACTICAL LESSONS

Educational discipline	Pediatric Surgery
module №5	Congenital Anomalies in Children
Theme of the lesson	Congenital anomalies of the gastrointestinal tract responsible for low intestinal obstruction. Hirschprung"s disease. Malformations of the anus and rectum
Course	VI
Faculty	foreign students preparation

POLTAVA 2020

1.The topic basis: The diseases of colon and malformations of the anus and rectum occupy one of leading places among reasons of disability for children. Actuality of theme is conditioned prevalence among other innate anomalities (48-50%), by plenty of diagnostic errors, complications and unsatisfactory consequences of treatment.

The topic "Congenital anomalies of the gastrointestinal tract responsible for low intestinal obstruction. Hirschprung"s disease. Malformations of the anus and rectum" is very important for future doctors in their professional activity, positively influences the students in their attitude to the future profession, forms professional skills and experience as well as taking as a principle the knowledge of the subject learnt.

2.The aims of the training course:

1.A student must have general presentations of the basic stages of embryogenesis of alimentary canal

2.A student must know:

ØPrincipal reasons of low innate intestinal obstruction;

ØClassifications of low innate intestinal obstruction;

ØClinical displays, features of clinical picture depending on a level and reason of low innate intestinal obstruction;

ØModern methods of diagnostics at innate intestinal obstruction, principles of differential diagnostics;

ØBasic questions of treatment, principles of grant a help on the different stages of treatment.

3. A student must seize:

ØBy the method of examination of children with low innate intestinal obstruction;

Øby the method of digit rectal research;

Øby the technique of sounding of stomach and its washing;

Øby a method pneumo- and contrasting irrigography;

Øby a method high cleansing and hypertensive microclysters;

Øby the method of contrasting research of alimentary canal with a barium dredge;

4. A student must be able:

Øto diagnose low innate intestinal obstruction;

Øto distinguish the features of clinical picture depending on a level and reason of low innate intestinal obstruction;

Øto conduct differential diagnostics;

Øto analyse and estimate the results of laboratory and instrumental researches;

Øto appoint therapy a child with low innate intestinal obstruction on the different stages of treatment;

Øto estimate efficiency of therapy which is conducted.

5. A student must develop the creative capabilities:

Øin the process of clinical inspection and treatment of children with low innate intestinal obstruction, conducting research in obedience to by the conducted department by research work;

Ø to conduct the scientific analysis of sources on this issue.

3.Basic knowledge, skills, habits necessary for studying the subject (interdisciplinary integration).

Names of previous disciplines	**Obtained skills**
1. Anatomy	Describe the structure of the abdominal organs. To assess the features of possible variants of the anatomical structure of the abdominal cavity organs. Determine the possible level of obstruction, determine

	the location of operative access
2. Physiology. Pathological Physiology	Describe the physiology of the gastrointestinal tract. Determine the features of the digestive system in children of different age groupsIndicators of a general analysis of blood, urine, biochemical blood test, coagulogram of acid-base state in norm and pathology. To distinguish between normal and pathological indicators, to interpret changes. .
3. Faculty pediatrics.	Differential diagnosis of acquired obstruction
4. Surgical diseases, operative Surgery and topographic anatomy.	Surgery and topographic anatomy. Identification of priority research methods and indications for surgical intervention. Principles of care for surgical patients. Clinic, diagnosis, treatment of intestinal obstruction. Measure CVP, establish gastric tube, urinary catheter, catheterize veins
5.Propedeutics of childhood diseases Faculty and hospital pediatrics, neonatology	Examination of a child with intestinal obstruction. Symptoms of pathology of the abdominal organs in infants Measure blood pressure, heart rate, the number of respiratory movements per minute, conduct a survey of the abdomen (palpation, percussion, auscultation), rectal examination
6.Pharmacology and Clinical Pharmacology	Pharmacology, pharmacodynamics and pharmacokinetics of drug groups that are used in the surgical pathology of abdominal organs in infants Calculate doses of drugs in the provision of emergency care and different routes of administration
7. Radiology	Interpretation of X-ray examination data. Recognize on the roentgenograms of the Clauber Bowl, free gas, free fluid in the abdominal cavity, evaluate the airway intestine Radiological anatomy of the abdominal cavity organs is normal. X-ray changes in intestinal obstruction, peritonitis, perforation of hollow organs.

Thereotical questions for the lesson:

1. Clinical manifestations of low intestinal obstruction.
2. Diagnostic of congenital intestinal obstruction.
3. Surgical treatment of congenital intestinal obstruction.
4. Hirschsprung's Disease. Classification. Clinic. Treatment.
5. Anorectal malformation. Classification. Clinic. Treatment.

4. Maintenance of the subject:

Hirschsprung's disease.

Hirschsprung disease is characterized by a congenital absence of ganglion cells in the distal colon resulting in a functional obstruction.

Causes. Although the precise mechanism responsible for Hirschsprung disease is unknown, the following basic theories have been proposed: (1) the neuroblasts fail to migrate into the affected segments of bowel; (2) the neuroblasts that were once present fail to mature; or (3) normal development of the cells occurs, but some insult leads to degradation or destruction of the cells.

Pathophysiology. Congenital aganglionosis of the distal bowel defines Hirschsprung disease. Aganglionosis begins with the anus, which is always involved, and continues proximally for a variable distance. Both the myenteric (Auerbach) and submucosal (Meissner) plexus are absent, resulting in reduced bowel peristalsis and function. The precise mechanism underlying the development of Hirschsprung disease is unknown.

Three neuronal plexus innervate the intestine: the submucosal (ie, Meissner) plexus, the intermuscular (ie, Auerbach) plexus, and the smaller mucosal plexus. All of these plexus are finely integrated and involved in all aspects of bowel function, including absorption, secretion, and motility.

Normal motility is primarily under the control of intrinsic neurons. Bowel function is adequate, despite a loss of extrinsic innervation. These ganglia control both contraction and relaxation of smooth muscle, with relaxation predominating. Extrinsic control is mainly through the cholinergic and adrenergic fibers. The cholinergic fibers cause contraction, and the adrenergic fibers mainly cause inhibition.

In patients with Hirschsprung disease, ganglion cells are absent, leading to a marked increase in extrinsic intestinal innervation. The innervation of both the cholinergic and adrenergic systems is 2-3 times that of normal innervation. The adrenergic (excitatory) system is thought to predominate over the cholinergic (inhibitory) system, leading to an increase in smooth muscle tone. With the loss of the intrinsic enteric inhibitory nerves, the increased tone is unopposed and leads to an imbalance of smooth muscle contractility and a functional obstruction.

Frequency. Hirschsprung disease occurs at an approximate rate of 1 case per 5400-7200 newborns.

Mortality/Morbidity. Untreated aganglionic megacolon in infancy may result in a mortality rate of as much as 80%. Operative mortality rates for any of the interventional procedures are very low. Even in cases of treated Hirschsprung disease, the mortality rate may be as much as 30% as a result of enterocolitis.

Age. The age at which Hirschsprung disease is diagnosed has progressively decreased over the past century. In the early 1900s, the median age at diagnosis was 2-3 years; from the 1950s to 1970s, the median age was 2-6 months. Currently, approximately 90% of patients with Hirschsprung disease are diagnosed in the newborn period.

Clinical signs

***History*.** Hirschsprung disease should be considered in any newborn with delayed passage of meconium or in any child with a history of chronic constipation since birth. Other symptoms

include bowel obstruction with bilious vomiting, abdominal distention, poor feeding, and failure to thrive. Older children with Hirschsprung disease have usually had chronic constipation since birth. They may also show evidence of poor weight gain.

Despite significant constipation and abdominal distension, children with Hirschsprung disease rarely develop encopresis. In contrast, children with functional constipation or stool-withholding behaviors more commonly develop encopresis.

Physical. Physical examination in the newborn period is usually not diagnostic, but it may reveal a distended abdomen and/or spasm of the anus. A low imperforate anus with a perineal opening may have a similar presentation to that of a patient with Hirschsprung disease. Careful physical examination differentiates the two. In older children, however, a distended abdomen resulting from an inability to release flatus is not uncommon.

Diagnosis

Plain abdominal radiographs may show distended bowel loops with a paucity of air in the rectum.

Barium enema. Avoid washing out the distal colon with enemas before obtaining the contrast enema because this may distort a low transition zone. Radiographs are taken immediately after hand injection of contrast and again 24 hours later. A narrowed distal colon with proximal dilation is the classic finding of Hirschsprung disease after a barium enema.

However, findings in neonates (ie, babies aged <1 mo) are difficult to interpret and often fail to demonstrate this transition zone, which takes time to develop.

Another radiographic finding suggestive of Hirschsprung disease is the retention of contrast for longer than 24 hours after the barium enema has been performed.

Anorectal manometry. Anorectal manometry detects the absence of the relaxation reflex of the internal sphincter after distension of the rectal lumen.

Because cardiac malformation (2-5%) and trisomy 21 (5-15%) are associated with congenital aganglionosis, ***cardiac evaluation*** and ***genetic testing*** may be warranted.

Rectal biopsy. The definitive diagnosis of Hirschsprung disease is confirmed by rectal biopsy, ie, findings that indicate an absence of ganglion cells. The definitive method for obtaining tissue for pathologic examination is by a full-thickness rectal biopsy. The specimen must be obtained at least 1.5 cm above the dentate line because aganglionosis is not present below this level.

The abdomen is opened via the Pfannenstiel incision. The biopsy site is selected by observing the apparent transitional zone. In the usual case of rectosigmoid aganglionosis, three seromuscular biopsies are taken along the antimesenteric surface without entering the lumen. One biopsy is taken from the narrowed segment of bowel, a second biopsy from the transition zone and a third biopsy from the dilated portion above the transition zone.Biopsies are assessed intra-operatively by frozen section, to determine the level of ganglionic bowel.

Treatment

Medical Care. The general goals of medical care are to treat the complications of unrecognized or untreated Hirschsprung disease, to institute temporary measures until definitive reconstructive surgery can take place, and to manage bowel function after reconstructive surgery.

Management of complications of recognized aganglionosis is directed toward reestablishing normal fluid and electrolyte balance, preventing bowel overdistension (with possible perforation), and managing complications such as sepsis.

Intravenous hydration, nasogastric decompression, and, as indicated, administration of intravenous antibiotics remain the cornerstones of initial medical management.

Colonic lavage, consisting of mechanical irrigation with a large-bore rectal tube and large volumes of irrigant, may be required.

Balanced salt solutions may help prevent electrolyte imbalances.

Colonic lavage may also be used in postoperative patients who develop enterocolitis as a complication. Injecting the nonrelaxing internal sphincter mechanism with botulinum toxin

(Botox) has recently been shown to induce more normal patterns of bowel movements in postoperative patients with enterocolitis.

Surgical Care. Surgical management of Hirschsprung disease begins with the initial diagnosis, which often requires a full-thickness rectal biopsy. In most cases, treatment also includes creating a diverting colostomy at the time of diagnosis. Once the child grows and weighs more than 10 kg, the definitive repair is performed.

Typically, neonates diagnosed with Hirschsprung disease are first treated with a diverting colostomy. Identify the transition zone, and place the colostomy proximal to this area. The presence of ganglion cells at the colostomy site must be unequivocally confirmed by a frozen-section biopsy. A diverting colostomy may also be required in older patients to decompress the dilated proximal bowel and allow time for it to return to a normal caliber. Either a loop or end stoma is appropriate, usually based on the surgeon's preference.

Many surgeons prefer right transverse colostomy; others advocate performing colostomy just above the transition to ganglionic bowel. Ileostomy is indicated in patients who have total colonic aganglionosis. A right transverse colostomy is convenient in usual cases. We perform a loop colostomy over a skin bridge.A V-shaped incision is made in the right upper quadrant. The V-skin-flap is reflected upwards. The external oblique is split and the internal oblique and transverse abdominis muscles are divided with diathermy. The peritoneum is opened.

An opening is made in the mesocolon of the selected segment of transverse colon. The skin flap is pulled through the opening in the mesocolon and sutured to the opposite skin margin. A few interrupted absorbable sutures of 4/0 or 5/0 are placed between the peritoneum, the muscle layers of abdominal wall and the seromuscular layer of colon. The colon is opened longitudinally along the antimesenteric border using diathermy. The bowel is sutured to the skin using interrupted 4/0 absorbable sutures.

The 3 most commonly performed repairs are the Swenson, Duhamel, and Soave procedures. Regardless of the pull-through procedure chosen, cleaning the colon prior to definitive repair is necessary.

The Swenson procedure was the original pull-through procedure used to treat Hirschsprung disease. The aganglionic segment is resected down to the sigmoid colon and the remaining rectum, and an oblique anastomosis is performed between the normal colon to the low rectum.

The Duhamel procedure was first described in 1956 as a modification to the Swenson procedure. Key points are that a retrorectal approach is used and a significant portion of aganglionic rectum is retained. The aganglionic bowel is resected down to the rectum, and the rectum is oversewn. The proximal bowel is then brought through the retrorectal space (between the rectum and sacrum), and an end-to-side anastomosis is performed on the remaining rectum.

The Soave procedure was introduced in the 1960s and consists of removing the mucosa and submucosa of the rectum and pulling the ganglionic bowel through the aganglionic muscular cuff of the rectum. The original operation did not include a formal anastomosis, but the procedure has been modified by Boley to include a primary anastomosis at the anus.

Diet. The patient should have nothing by mouth before the operation. Institute tube feeding or formula/breast milk once bowel function resumes. High-fiber diets and diets containing fresh fruits and vegetables may optimize postoperative bowel function in certain patients.

Activity. Limit physical activity for about 6 weeks to allow the wound to heal properly (applies more to older children).

Further Inpatient Care. If a diverting colostomy is created in a newborn, he or she must remain in the hospital until the ostomy is functioning and feeding goals are obtained. Feedings are usually initiated 24-48 hours after the creation of the colostomy.

After the definitive pull-through procedure is performed, the patient is hospitalized until full feedings are possible and evidence of the return of bowel function is obtained. Patients are to take nothing by mouth, with intravenous fluid hydration until they pass flatus or have a bowel

movement. Once this occurs, clear liquids may be started, and the diet may be advanced until feeding goals are obtained. Intravenous antibiotics are also continued until evidence of proper bowel function is observed.

Complications. In general, the complications are anastomotic leak (5%), anastomotic stricture (5-10%), intestinal obstruction (5%), pelvic abscess (5%), and wound infection (10%).

Anorectal malformation.

Anorectal malformations comprise a wide spectrum of disease affecting boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tracts.

Frequency. Anorectal malformations occur in approximately 1 in 5000 live births.

In 80-90% of newborn boys, clinical evaluation and urinalysis provide enough information for the surgeon to decide whether the baby requires a colostomy.

After the baby is born, an intravenous line is placed for fluids and antibiotics. A nasogastric tube is inserted to keep the stomach decompressed to avoid the risk of vomiting and aspiration.

Meconium is not usually observed at the perineum in a baby with a rectoperineal fistula until at least 16-24 hours of life. Abdominal distension does not develop during the first few hours of life but is required to force meconium through a rectoperineal fistula, as well as through a urinary fistula. This is because the most distal part of the rectum in these children is surrounded by a funnellike voluntary muscle structure that keeps part of the rectum collapsed and empty. The intra-abdominal pressure must be high enough to overcome the tone of the muscles that surround the rectum if meconium is to be expected at the perineum or in the urine. Therefore, the decision of whether to perform a colostomy or an anoplasty must be delayed for 16-24 hours while the surgeon evaluates for clinical evidence of the baby's anorectal anomaly.

Clinical inspection of the buttocks is important. A flat bottom or flat perineum, as evidenced by the lack of a midline gluteal fold and the absence of an anal dimple, indicates that the patient has very poor muscles in the perineum. These findings are associated with a high malformation; therefore, a colostomy should be performed.

Perineal signs found in patients with low malformations include the presence of meconium at the perineum, a bucket-handle malformation (ie, a prominent skin tag located at the anal dimple, below which an instrument can be passed), and an anal membrane (through which one can see meconium).

Newborn boys with rectoperineal fistula do not need a colostomy. They can undergo a posterior sagittal anoplasty.

Baby boys with evidence of a rectourinary tract communication should undergo fecal diversion with a colostomy.

If none of the clinical signs to determine the location of the anorectal anomaly are evident by 24 hours, performing a radiologic test can help. This situation is only necessary in approximately 10% of patients. Crosstable lateral radiography with the baby prone, with the pelvis elevated, and with a radiopaque marker placed on the perineum is performed. Rarely, radiography may show the column of air in the distal rectum to be within 1 cm of the perineum; if this is the case, management can be similar to that for rectoperineal fistula, and a newborn perineal operation can be performed. If the air column is more than 1 cm from the perineum, a colostomy is indicated.

Some authors have performed definitive repair in the newborn period. The advantage to this approach is avoidance of the colostomy and earlier repair of the malformation; however, with this practice, considerable risk to the urinary tract exists because the surgeon does not know the precise anorectal defect. The only way to determine the patient's anorectal defect definitively is to perform distal colostography, which requires the presence of a colostomy. Without this information, an operation in the newborn period is essentially a blind perineal exploration. The surgeon may not be able to locate the rectum and may find and damage other unexpected structures, such as the posterior urethra, seminal vesicles, vas deferens, and ectopic ureters

during the search for the rectum. Finally, without fecal diversion, the risk of dehiscence and infection exists. These complications may compromise the chance of achieving bowel function.

The exception, which is rare, occurs when the crosstable lateral ***radiograph*** taken at 16-24 hours shows that the rectum is located just below the coccyx. In this case, the rectum can be reached from the posterior sagittal approach.

Urinalysis and a gauze placement over the penis can be done to determine the presence of fecal matter in the urine, which is considered evidence of a rectourinary fistula.

Abdominal ***ultrasonography*** must be performed to evaluate for the presence of an obstructive uropathy. At the same time, spinal ultrasonography can be performed to evaluate for spinal anomalies, including the presence of a tethered cord.

Any method of trying to determine the location of the distal rectum before 16 hours of life is flawed because of the contracted state of the funnellike sphincter mechanism. Normally, the funnel-shaped muscle structure is contracted unless overcome with a distending force. Tests such as MRI, ultrasonography, computed tomography scanning, or injection of contrast through the perineum falsely locate the distal rectum as high.

Distal ***colostography*** (performed usually 1 mo after colostomy opening) must have adequate pressure to demonstrate a fistula from the rectum to the urinary tract or this method also falsely locates the distal rectum as high in the pelvis.

Once the patient recovers from colostomy and demonstrates good growth and development, the definitive operation can be planned for 4-8 weeks.

Decision-making in the newborn girl with anorectal anomalies

The decision of whether to perform colostomy in newborns girls is less complicated than in newborn boys. In 90% of patients, a meticulous perineal inspection demonstrates the anorectal defect. Waiting 16-24 hours for enough abdominal distension to demonstrate the presence of a rectoperineal fistula or rectovestibular fistula also applies to females.

The most common anomaly in females is a rectovestibular fistula. Perineal inspection shows a normal urethra, normal vagina, and another orifice that is the rectal fistula in the vestibule.

Performing a diverting colostomy is the safest option for a surgeon without extensive experience in anorectal anomalies when faced with a baby who has clinical evidence of a rectovestibular fistula. Colostomy before the main repair avoids the complications of infection and dehiscence. Definitive repair of this anomaly in the newborn period should be reserved for surgeons who have significant experience repairing these defects. This anomaly has an excellent prognosis; therefore, complications that could affect future continence must be avoided.

Unfortunately, the most common referral for reoperations to tertiary centers that care for anorectal anomalies is for patients with rectovestibular fistulas who experienced failed primary repair in the newborn period. Occasionally, the fistulas are large enough to decompress the gastrointestinal tract, and they may be dilated to facilitate fecal drainage until the baby is older and a definitive repair is performed. Definitive repair involves a posterior sagittal approach. The most delicate part of this operation is the separation of the rectum and vagina, which share a common wall.

Like males, females can also have a rectoperineal fistula, which requires anoplasty to be performed in the newborn period.

Like in males, less than 5% of female babies have no clinical evidence of the location of the rectum after 24 hours. They may have imperforate anus with no fistula. Crosstable lateral radiography should be performed, which helps determine the need for a colostomy.

The presence of a single perineal orifice in a patient is clinical evidence of persistent cloaca. Patients with these anomalies also have small genitalia. In patients with persistent cloaca, examination of the abdomen may reveal an abdominal mass that likely represents a distended vagina (hydrocolpos), which is present in 50% of patients with persistent cloaca. Abdominal ultrasonography is helpful to determine the presence of an obstructive uropathy and hydrocolpos.

Unfortunately, a common error in diagnosis occurs during the perineal inspection when a female is thought to have imperforate anus with rectovaginal fistula; however, in actuality, all 3 structures (ie, urinary tract, vagina, rectum) meet in a common channel, and the baby has persistent cloaca. This misconception has important therapeutic implications that are discussed below.

Making the correct determination of persistent cloaca is vital because 90% of babies with persistent cloaca have an associated urologic problem and 50% have hydrocolpos. The urinary tract and the distended vagina both may need to be managed in the newborn period to avoid serious complications.

Missing the diagnosis of persistent cloaca frequently means that an obstructive uropathy is overlooked. The patient may then receive only a colostomy, and subsequently, sepsis, acidosis, and sometimes death may occur.

The other implication of missing the diagnosis of persistent cloaca involves repairing only the rectal component of the anomaly, leaving the patient with a persistent urogenital sinus.

5. Additional materials for the self-control

A. Clinical cases

Case 1.

In a newborn boy during the examination in the maternity hall the absence of the anus has been detected, rectal fistulas are not available. How is the level of atresia of distal part of the intestine detected?

Case 2.

In a one-year old child permanent constipations appeared after introduction of the additional feeding. Emptying takes place once in 4-5 days, more frequently after an enema. In the summer and autumn constipations are rarer. On the irrigography the narrowing of rectosigmoid part of the colon is visible, and above it there is the expansion of the colon. What anomaly could you think about? What tactics have you developed?

Case 3.

A 14 day child since birth has miserable meconium stool, in subsequent days an excrement and gases depart badly - only after an enema through a gas-freeing tube. Three times there was vomiting with the admixture of bile. The abdomen is distended, peristalsis of intestinal loops is visible. After an enema there appeared insignificant stool. What diagnosis have you made? What auxiliary methods of examination are to be used? What tactics have you developed?

B. Tests

6.LITERATURE FOR STUDENTS

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