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

«Approved»
on meeting the
department of Pediatric Surgery
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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 high intestinal obstruction, Anomalies of rotation
Course	VI
Faculty	foreign students preparation

POLTAVA 2020

1.The topic basis: 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 high intestinal obstruction, Anomalies of rotation" 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 about the basic stages of embryogenesis of alimentary canal

2. A student must know:

Ø Principal reasons of high innate intestinal obstruction;
Ø Classifications of high innate intestinal obstruction;
Ø Clinical displays, features of clinical picture depending on a level and reason of high innate intestinal obstruction;
Ø Modern methods of diagnostics at innate intestinal obstruction, principles of differential diagnostics;
Ø Basic questions of treatment, principles of grant a help on the different stages of treatment.

3. A student must seize :

Ø By the method of examination of children with high innate intestinal obstruction;
Ø by the method of digit rectal research;
Ø by the technique of sounding of stomach and its washing;
Ø by a method pneumo- and contrasting irrigography;
Ø by a method high cleansing and hypertensive microclysters;
Ø by the method of contrasting research of alimentary canal with a barium dredge.

4. A student must be able:

Ø to diagnose high innate intestinal obstruction;
Ø to distinguish the features of clinical picture depending on a level and reason of high innate intestinal obstruction;
Ø to conduct differential diagnostics;
Ø to analyse and estimate the results of laboratory and instrumental researches;
Ø to appoint therapy a child with high innate intestinal obstruction on the different stages of treatment
Ø to estimate efficiency of therapy which is conducted.

5. A student must develop the creative capabilities:

Ø in the process of clinical inspection and treatment of children with high innate intestinal obstruction, conducting research in obedience to by the conducted department to 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. Congenital pylorostenosis. Clinic. Diagnostics. Treatment.
2. Classification of congenital intestinal obstruction.

3. Atresia of duodenum. Clinic. Diagnostics. Treatment.

4. Atresia of iliac colon. Clinic. Diagnostics. Treatment.

5. The Ladd syndrome. Clinic. Diagnostics. Treatment

4. Maintenance of the subject:

INFANTILE HYPERTROPHIC PYLORIC STENOSIS

Pathophysiology. Diffuse hypertrophy and hyperplasia of the smooth muscle of the antrum of the stomach and pylorus proper narrow the channel, which then can become easily obstructed. The antral region is elongated and thickened to as much as twice its normal size. In response to outflow obstruction and vigorous peristalsis, stomach musculature becomes uniformly hypertrophied and dilated. Gastritis may occur after prolonged stasis. Hematemesis is occasionally noted. The patient may become dehydrated as a result of vomiting and develop marked hypochloremic alkalosis.

Frequency. Pyloric stenosis is a common cause of gastric outlet obstruction in infants. The prevalence of HPS ranges from 1.5-4/1000 live births among whites, although it is less prevalent among African and Asian Americans.

Clinical features. The peak incidence for the onset of symptoms is between the 3rd and 6th weeks of life. However, the condition can commence before that time, and as late as the 7th week. The pillars upon with the diagnosis rests are:

Vomiting is the presenting symptom in all cases and withing 2 or 3 days it becomes forcible and progectove. Bile is not present in the vomiting. Immediately after vomiting the baby is often very hungry.

Visible peristalsis. After the child has been fed, peristaltic waves may be seen passing from the left to right across the upper abdomen. It is the so-cold symptom of “sand clock”. A ggod light is essential for this. The abdomen should be examined throughout a feed until vomiting occurs.

The presence of a lump. The palpation of the hypertrophyed pylorus is the most essential step in reaching a diagnosis. The surgeon should palpate under the liver with a warm hand. It may be helpful to examine the child more than once. The lump is most easily felt when the child is given a feed.

Constipation is usually present, and when a stool is passed it is small and dry. It is important to ask the mother about napkins. If the child is dehydrated, they are not wet and the case is correspondingly more urgent.

One of the most striking signs of infants sufferinf from IHPS is loss of weight. Moreover, it is not long before the infant begins to look emaciated and dehydrated.

Often a change from one type of feeding to another brings about a remission. Consequently a series of changes in diet are sometimes made before the diagnosis is established, by with time the infant`s condition may be pitiable.

In prematures infants, in whom the condition is not uncommon, the symptomatology is often paradoxical. There is anorexia instead of voracious appetite; the vomiting is regurgitant rather than projectily, and so frequently is peristalsis normally visible. None the less, amids this sea of bewilderment one diagnostic rock remains – a hypertrophied pylorus can be felt through the poorly-developed abdominal wall with comparative ease.

Imaging Studies

Ultrasonography has become the criterion standard imaging technique for diagnosing HPS. It is reliable and easily performed. An experienced ultrasonographer increases the test's predictive value. Necessary measurements include pyloric muscle thickness and pyloric channel length. Muscle wall thickness 3 mm or greater and pyloric channel length 14 mm or greater are considered abnormal in infants younger than 30 days.

Barium upper gastrointestinal (UGI) study is an effective means of diagnosing HPS. It should demonstrate an elongated pylorus with antral indentation from the hypertrophied muscle. The

classic "railroad track" sign of two thin parallel streams of barium traversing the pylorus is pathognomonic. Confirming the diagnosis of HPS is impossible if barium does not leave the stomach. Sufficient patience, however, usually demonstrates the above findings. An advantage of a barium UGI contrast study is the ability to identify gastroesophageal reflux, a frequent differential diagnosis of HPS. After UGI, irrigating and removing any residual barium from the stomach is advisable to avoid aspiration.

Although UGI ***endoscopy*** would demonstrate pyloric obstruction, physicians would find it difficult to differentiate accurately between HPS and pylorospasm. Endoscopic dilatation has rarely been employed as a method of treatment. This treatment is not standard for HPS; endoscopy should be used rarely, if ever.

Treatment

Preoperative resuscitation: If necessary, administer an initial fluid bolus of 10 mL/kg with lactated Ringer solution or 0.45 isotonic sodium chloride solution. Continue intravenous (IV) therapy at an initial rate of 1.25-2 times the normal maintenance rate until adequate fluid status is achieved.

Adequate amounts of both chloride and potassium are necessary to correct metabolic acidosis. Unless renal insufficiency is a concern, initially add 2-4 mEq of KCL per 100 mL of IV fluid. Adequate chloride for resuscitation can usually be provided by 5% dextrose in 0.4% sodium chloride solution. Avoid adding hypertonic chloride or ammonium chloride.

Urine output and serial electrolyte determinations are performed during resuscitation. Correction of serum chloride level to 90 mEq/L or greater usually is adequate to proceed with surgical intervention.

Surgical Care.

A nasogastric tube must be placed before the induction of anaesthesia if the tube was not placed pre-operatively. And if the barium meal study has been carried out prior to surgery, it may be necessary to remove the residual barium meal by gastric aspiration and irrigation. The patient is placed in the supine position. After the induction of anaesthesia and endotracheal intubation, careful abdominal palpation will usually identify the site of the pyloric tumour. A 2.5-to 3-cm long transverse incision is made lateral to the lateral border of the rectus muscle. The incision is deepened through the subcutaneous tissue and the underlying external oblique, internal oblique and transverse muscles are split. The peritoneum is opened transversely in the line of the incision. When supra-umbilical skin fold incision is employed, a circumumbilical incision is made through about two-thirds of the circumference of the umbilicus. The skin is undermined in a cephalad direction above the umbilical ring and the linea alba is exposed. The linea alba is divided longitudinally in the midline from the umbilical ring to as far cephalad as necessary to allow easy delivery of the pyloric tumour.

The stomach is identified and is grasped proximal to the pylorus with non-crushing clamp and brought through the wound. Then, the greater curvature of the stomach can be held in a moist gauze swab, and with traction inferiorly and laterally, the pylorus can be delivered through the wound. Grasping the duodenum or pyloric tumour directly by forceps often results in serosal laceration, bleeding or perforation, therefore should be avoided.

The pylorus is held with surgeon’s thumb and forefinger to stabilize and assess the extent of hypertrophied muscle. A seromuscular incision is made over the avascular area of pylorus with a scalpel, commencing 1~2 mm proximal to the pre-pyloric vein along the gastric antrum. The incision should go far enough onto the gastric antrum at least 0.5~1.0 cm from the antropyloric junction where the muscle is thin.

The scalpel handle is used to further split the hypertrophied muscle down to the submucosal layer. Then pyloric muscle is spread widely. Spreader is placed at the midpoint of incision line and muscle is spread perpendicularly and spreading must be continued proximally and distally. Gentle spreading is required to obtain a complete myotomy. Mucosal tears are most common at the pyloroduodenal junction because of the attempt to split all remaining muscle fibres. In order to reduce the risk of mucosal tear, care should be taken when spreading pyloric muscle fibres at the

duodenal end. Loose prolapsing of intact mucosa is evidence of a satisfactory myotomy. To test the mucosal injury, the stomach is inflated through the nasogastric tube, and passage of air through the pylorus to duodenum is confirmed. Then the pylorus is dropped back into the abdomen. Bleeding from the myotomy edge or submucosal surface is frequently seen; however, it is generally venous and always stops after returning the pylorus to the abdominal cavity. Posterior rectus fascia and peritoneum is approximated with a running 4/0 absorbable suture material and anterior fascia is closed with 5/0 absorbable suture material.

For the laparoscopic procedure the patient is placed in the supine position at the end of the operating table (or 90C to the anaesthesiologist). The video monitor is placed at the head of the table, and the surgeon stands at the end of the table with the assistant to the patient's right. The abdomen is scrubbed and draped in a sterile fashion.Attention must be paid to ensure the appropriate preparation of the umbilicus. The access sites are injected with local anaesthetic (0.25% bupivacaine) with epinephrine,which is used to reduce the post-operative pain and reduce the risk of bleeding from the stab wound. The author prefers an open procedure for insertion of the primary port. A 4.0- to 5.0-mm curvilinear supra-umbilical incision is made and carried down to the peritoneal cavity. At the level of umbilical fascia, 4/0 absorbable suture material is placed circumferentially to anchor the port and to use for closure of the peritoneal cavity after laparoscopic pyloromyotomy is completed. Intra-abdominal pressure is maintained at 8 mmHg, and insufflation rate is set at 0.5 l/min. In the right mid-clavicular line just below the costal margin (just above the liver edge), a no. 11 scalpel blade is used to make a 2- to 3-mm stab incision under direct vision. Also using the no. 11 scalpel blade, a second stab incision is made under direct vision, just below the costal margin in the left mid-clavicular line. An atraumatic grasper is placed directly through the right upper quadrant stab wound and is used to retract the inferior border of the liver superiorly and expose the hypertrophic pylorus. A retractable myotomy knife (retractable arthrotomy knife or Endotome) is inserted directly through the left stab wound. Working ports are usually not necessary and instruments are directly introduced through these stab wounds.

The working instruments, retractable myotomy knife, atraumatic laparoscopic grasper are used to assess the extent of the hypertrophied pylorus by palpating the margins of the pylorus as one would use with thumb and forefinger in the open procedure. The duodenum is then grasped just distal to the pyloric vein (pyloroduodenal junction) and retracted using the atraumatic grasper to expose the avascular surface of hypertrophic pylorus. The tips of positioning the pylorus for myotomy is that lateral and slightly anterocephalad retraction of the distal pylorus achieve excellent exposure of the avascular surface of hypertrophic pylorus. This manoeuvre also exposes the proximal margin of hypertrophied muscle that is seen as a deep fold in the wall of stomach. A seromuscular incision is made over the hypertrophic pylorus with retractable myotomy knife commencing at 1–2 mm proximal to the pyloroduodenal junction extending to the gastric antrum. The incision should go far enough onto antrum at least 0.5~1.0 cm proximal to antropyloric junction. Care must be taken at this stage that this incision is deep enough to allow the insertion of the pyloric spreader blades and must penetrate the pyloric muscle somewhat deeper than is usual with the conventional open procedure.

After the muscle is incised, the blade is then retracted and the sheath of the knife is used to further split the hypertrophied muscle fibre, as the scalpel handle is used in open procedure, until mucosa is visualized. The retractable myotomy knife is removed and a laparoscopic pyloromyotomy spreader is introduced into abdominal cavity directly through the left stab wound to complete the pyloromyotomy. The spreader is placed in the midpoint of the seromuscular incision line and the muscle is spread perpendicularly. Once the initial spread reaches the mucosa, spreading must be continued proximally and distally. Pushing the spreader towards the mucosa or rapid spreading can result in mucosal tear. In order to avoid the mucosal tear, the spreader should not be placed at the proximal and distal edges of the incisional (myotomy) line. To test for the mucosal injury, the stomach is inflated through the nasogastric tube (160–180 ml) as is usually done in open techniques. Bulging of the mucosal layer with no evidence of defect should be confirmed. Greenish or yellowish fluid at the myotomy area is a sign of mucosal tear. After the successful

myotomy, the instruments are withdrawn under direct vision and the pneumoperitoneum is evacuated. The nasogastric tube is also removed after completing the surgery. The umbilical fascia is reapproximated with 4/0 absorbable suture material,which is already in place, and the skin of all the wound is reapproximated with skin adhesive tapes.

Postoperative management

Continue IV maintenance fluid until the infant is able to tolerate enteral feedings. In most instances, feedings can begin within 8 hours following surgery. Graded feedings can usually be initiated every 3 hours, starting with Pedialyte and progressing to full-strength formula.

Complications

Undetected mucosal perforation: Perform a diligent search for mucosal transgressions at the time of operation and examine the infant again before initiating feedings. In those rare cases where a perforation was not detected, the infant develops fever, tenderness in the abdomen, and abdominal distention. Return to the operating theater if perforation is suspected.

Bleeding: In most instances, venous oozing from the myotomy site is self-limited and is not a concern in the postoperative period.

Persistent vomiting: Incomplete pyloromyotomy is rare in the hands of an experienced pediatric surgeon. Less experienced surgeons occasionally commit this error, but they are more likely to cause injury to the mucosa from injudicious spreading during the myotomy. This problem is confounded when repeat studies performed after surgery provide a confusing picture. Patient observation resolves the problem in most cases

CONGENITAL DUODENAL ILEUS

Embryology The hepatobiliary system and pancreas form during the third week of gestation, as the second portion of the duodenum gives rise to biliary and pancreatic buds at the junction of the foregut to the midgut. The duodenum also undergoes a solid phase during this time; between the eighth and tenth weeks of gestation, the duodenal lumen is reestablished by the gathering of vacuoles, and recanalization occurs. Insults during this crucial period of development are believed to result in failure of recanalization and consequent atresias, stenosis, and webs. In addition, duodenal atresias have been associated with a closely surrounding piece of pancreatic tissue. Whether this tissue is an annular pancreas or merely a failure of duodenal development is debatable.

Frequency. Duodenal atresia is present in approximately 1 out of 6,000 newborns.

Mortality/Morbidity. If duodenal atresia or significant stenosis is left untreated, the condition is rapidly fatal owing to electrolyte loss and fluid imbalance.

One half of the neonates with duodenal atresia or stenosis are born prematurely. Polyhydramnios is present in approximately 40% of neonates with duodenal obstruction.

Duodenal atresia or stenosis is most commonly associated with trisomy 21. About 22-30% of patients with duodenal obstruction have trisomy 21. Other problems associated with trisomy 21 include cardiac defects (most commonly ventricular septal defect and endocardial defects) as well as Hirschsprung disease.

Sex: The incidence of duodenal atresia and stenosis is approximately equal in males and females.

Age: Infants with duodenal atresia present with vomiting in their first few hours of life, but patients with duodenal stenosis present at various ages. The clinical findings depend on the degree of stenosis.

Classification. Duodenal obstruction is the result of atresia, stenosis, and duodenal web, annular pancreas, or peritoneal bands secondary to incomplete intestinal rotation. Intrinsic anomalies of the duodenum occur in several forms. There can be atresia with continuity of the bowel wall, atresia with a fibrous cord joining the segments, atresia with complete loss of continuity of the wall and of the blood supply, and all but complete diaphragm with a small fenestration, or a membranous ring within the duodenum which peristalsis from above forces into the development of a "wind sock". The dilating effect of the wind sock may produce the appearance of obstruction distal to the actual annulus of the wind sock.

In general, duodenal obstructions may be either preampullary or postampullary; however, most are considered periampullary. The degree of obstruction dictates the amount of resulting pathology. The obstruction causes dilation of the proximal duodenum and stomach as well as hypertrophy and distension of the pylorus. A common variation is the windsock anomaly, in which the duodenum is dilated distal to the point of obstruction because of a prolapsing membrane or web. This may be confused with a more distal duodenal obstruction.

Diagnosis

Prenatal ultrasonography may indicate structural and associated abnormalities, such as a dilated stomach and proximal duodenum. Polyhydramnios indicates that the fetus may be having difficulty swallowing the amniotic fluid, and it is suggestive of GI tract obstruction. Common associated anomalies and chromosomal defects may be assessed by screening maternal serum and amniotic fluid.

In the newborn, clear or bilious emesis is evident within hours of birth. Abdominal distension may or may not be present. An output of more than 20 mL of gastric contents is indicative of possible obstruction (normal gastric capacity is 7 – 10 ml). Patients with a stenosis or web may present later with dehydration or failure to thrive.

Plain radiography is helpful and may reveal the classic double bubble, ie, air in the stomach and duodenum, which is associated with complete or near complete duodenal obstruction. Upright and contrast radiography using air or contrast may confirm the diagnosis. Malrotation with volvulus may also result in duodenal obstruction and a consequent double bubble. Duodenal atresia and malrotation may coexist.

Treatment

Gastric decompression is essential to prevent aspiration, and thermoregulation should be monitored at all times. When fluid resuscitation has been accomplished, the neonate may proceed to surgery.

The baby is placed supine on the table with a small roll under his upper abdomen and on a warming blanket. Endotracheal anaesthesia is used. A nasogastric tube is passed to decompress the stomach. An intravenous infusion is set up. The abdominal skin is prepared by cleaning with prewarmed povidoneiodine. A transverse supra-umbilical abdominal incision is made 2 cm above the umbilicus starting in the midline and extending laterally into the right upper quadrant. A small incision is made in the posterior fascia and peritoneum after these are drawn up with forceps. To enlarge this initial incision, two fingers are inserted and the fascia and peritoneum are cut along the length of the wound. The underlying structures are retracted.

After exposing the peritoneal cavity, the surgeon inspects the entire bowel for the presence of other anomalies. There may be an associated annular pancreas or malrotation in one-third of the patients. If the colon is in a normal position, malrotation is probably not a coexisting factor.The stomach and first portion of the duodenum are usually thickened and dilated. The liver is carefully retracted superiorly. The ascending colon and the hepatic flexure of the colon are mobilized medially and downwards to expose the dilated duodenum. The duodenum is then adequately mobilized and freed from its retroperitoneal attachments – Kocher manoeuvre. Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The tube in the stomach is then passed distally into the dilated duodenum and helps to locate the point of obstruction and determine if a “windsock” abnormality is present. The type of atresia as well as any pancreatic abnormality (annular pancreas) or the presence of a rare preduodenal portal vein are noted. In patients with an annular pancreas, the pancreatic tissue should never be divided and should be bypassed.The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments. If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum is performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension.

Duodenoduodenostomy is the procedure of choice for patients with duodenal atresia, stenosis and annular pancreas. The two surgical techniques, either side-to-side duodenoduodenostomy or proximal transverse to distal longitudinal – "diamond-shape" anastomosis – may be performed. Diamond-shaped duodenoduodenostomy has been reported to allow earlier feeding, earlier discharge and good long-term results. With two traction sutures, the redundant wall of the proximal duodenum is pulled downward to overlie the proximal portion of the distal duodenal segment. A transverse incision is made in the distal end of the proximal duodenum and a longitudinal incision is made in the smaller limb of the duodenum distal to the occlusion. These are made in such a position as to allow good approximation of the openings without tension. The papilla of Vater is located by observing bile flow. This is performed by gentle compression of the gall bladder. The orientation of the sutures in the diamondshape anastomosis and the overlapping between the proximal transverse incision and the distal longitudinal incision are shown. At this stage a small Nelaton catheter is passed distally through the opening made in the distal segment. 20–30 ml of warm saline is injected to rule atresias distally. The catheter is then removed.

A single layer anastomosis using interrupted 5/0 or 6/0 Vicryl sutures with posterior knots tied inside the posterior wall of the anastomosis and interrupted sutures with anterior knots tied outside the anterior wall. Before completion of the anterior part of the anastomosis, a transanastomotic feeding tube (5F silicone) may be passed down into the upper jejunum for an early post-operative enteral feeding.

After abdominal exploration and the diagnosis of duodenal web (identified by the advancement of the gastric tube into the proximal dilated duodenum) two stay sutures are placed at the anterior dilated duodenal wall. A longitudinal incision of 2.5–3 cm is performed above the "transitional zone" between the wide and the narrow segments of the duodenum, and the duodenum is opened.

Two other stay sutures are placed at the margins of the duodenal incision. The windsock duodenal web must be clearly identified because the visible transition from the distended proximal duodenum to the small downstream duodenum may be several centimetres distal to the base of the web. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base. The duodenal membrane is usually localized in the second part of the duodenum and occasionally in the third portion. It can be complete or with a central hole. Anatomically, the ampulla of Vater may open directly into the medial portion of the web itself – anteriorly, posteriorly, or with dual openings into the membrane – or it may open close to it. Thus, the close relationship of the membrane to the papilla ofVater makes its identification mandatory, before excision of the web. A single 4/0 Vicryl stay suture is placed at the centre of the membrane. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place, leaving a rim of tissue of 2–3 mm. The medial portion of the membrane should remain intact, thus avoiding damage to the ampulla of Vater. An intermittent bile flow is usually seen via the papilla of Vater indicating to the surgeon the exact line of excision.

Then the resection line is over sewn using interrupted 5-0 absorbable sutures. The duodenum is then closed transversely with interrupted sutures.Because of the pitfalls in cases of lax membrane that may bulge downwards distally into the distended duodenum (the so-called windsock phenomenon), and in order to avoid missing the anomaly, the patency of the distal duodenum must be identified by inserting a catheter through the duodenotomy before its closure. Following completion of the web resection and closure of the duodenum, the abdominal cavity is irrigated with 50 ml sterile warm saline. The wound is closed in layers: the peritoneum and the posterior fascia and the anterior fascia by two layers using continuous 4/0 Dexon or Vicryl sutures. The skin is closed with a running intracuticular suture using 5/0 Vicryl or Dexon suture. A nasogastric tube is left in place for post-operative gastric drainage. A gastrostomy may be performed if the need is anticipated. Intravenous therapy and antibiotics are continued post-operatively. The patient is kept without oral intake until stool is passed and limited clear or pale-green gastric drainage is noted (<1 ml/kg per h). The commencement of oral feeding may be delayed for several days and occasionally for 2 weeks or more. Post-operatively, patients may have a prolonged period of bile-stained aspirate, which is mainly due to the inability of the markedly dilated duodenum to produce

effective peristalsis. Many surgeons therefore use transanastomotic tubes for feeding in the early post-operative period.

Complications. An anastomotic leak, injury to the bile duct, and sepsis are early complications. Late complications include peptic ulceration secondary to alkaline reflux, blind-loop syndrome with duodenal stasis, abdominal pain, diarrhea, and recurrent obstruction.

The **prognosis** is good for patients with a repaired duodenal stenosis or atresia; however, coexisting diagnoses, such as Down syndrome and cardiac anomalies, affect the outcome.

LADD'S SYNDROME

Malrotation is congenital abnormal positioning of the midgut. Intestinal development is traditionally described as a process of elongation, rotation and fixation. The process begins in the fifth week of gestation. Elongation of the bowel exceeds abdominal cavity expansion and the bowel herniates from the abdomen. As the bowel returns to the abdomen, it rotates 270° anticlockwise around the superior mesenteric artery (SMA). Rotation is completed by week 10 of gestation, with the SMA contained within a broad mesenteric base attachment. The distal duodenum comes to lie across the midline towards the left upper quadrant, attached by the ligament of Treitz at the duodeno-jejunal (D-J) flexure to the posterior abdominal wall. The caecum passes to the right and downwards and becomes fixed to the posterior abdominal wall. This latter process may be incomplete at birth giving rise to a "high" caecum, a variant of normal in the neonate. The commonest features of malrotation are: the D-J flexure lies right of midline, the dorsal mesenteric attachment is narrow, and peritoneal folds cross from colon and caecum to duodenum, liver and gallbladder (Ladd's bands), thus possibly obstructing the duodenum.Whether Ladd's bands are substantial enough to cause mechanical obstruction is debatable. The narrowed mesenteric base can lead to midgut volvulus, bowel obstruction and mesenteric vessel occlusion. Antenatal volvulus can result in bowel atresia.

Malrotation is estimated from autopsy studies to occur in 0.5–1% of the population, although only 1 in 6000 live births will present with clinical symptoms. Incidence is slightly higher in males than females. Fifty to 75% of patients become symptomatic in the first month of life and 90% will present before 1 year of age but presentation can occur at any age.Malrotation is present in patients with gastroschisis, exomphalos and congenital diaphragmatic hernia.Coexistent congenital anomalies (cardiac anomalies, bowel atresia, duodenal web, anorectal anomalies, orthopaedic anomalies) are common and affect 50% of children with malrotation. Malrotation is also associated with situs inversus, asplenia and polysplenic syndromes.

Acute bowel obstruction due to Ladd's bands or intermittent midgut volvulus can present with vomiting, typically bilious, as the commonest presenting feature accompanied by colicky abdominal pain and abdominal distention.An infant with abdominal tenderness and blood per rectum is suggestive of bowel ischaemia due to midgut volvulus. Older children without acute volvulus more often present with chronic episodic obstructive symptoms, failure to thrive, malabsorption, diarrhoea and non-specific colicky abdominal pain. Up to 10% of diagnoses of malrotation are made as an incidental finding. Plain abdominal radiograph is often normal but features suggestive of malrotation with or without midgut volvulus are a distended stomach and proximal duodenum with a paucity of gas distally, either throughout or unilaterally.An upper gastrointestinal contrast study is the investigation of choice for any child presenting with bilious vomiting and should be performed urgently. Findings in malrotation are: D-J flexure right of left vertebral pedicle and/or inferior to pylorus, the duodenum passes caudally and anteriorly, and contrast tapering or a "corkscrew" appearance suggests obstruction and/or volvulus. In a recent series, sensitivity and specificity of this test were 92% and 20%, respectively. Caecal position is highly variable and may be normal in up to 15% of cases of malrotation.Contrast enema is therefore not always helpful. Abdominal ultrasound may show reversal in the relationship of SMA to superior mesenteric vein (SMV). In a normal situation the SMV is located to the right of the SMA, while SMV to the left of the artery is suggestive of malrotation.

All symptomatic patients with positive investigative findings should undergo urgent laparotomy. Management of the asymptomatic patient is more controversial. The risk of bowel ischaemia due

to midgut volvulus is invariably present and the majority of surgeons would proceed to prompt operation. The principles of the procedure have remained almost unchanged since originally described by Ladd in 1936. The patient is positioned supine, legs extended. A right upper quadrant transverse incision is made. The umbilical vein is divided and ligated. The peritoneal fluid is examined. Frequently it is clear; bloodstained fluid implies bowel ischaemia and volvulus; faecal staining indicates bowel perforation and should be cultured.

The midgut is delivered from the wound and the base examined. Any volvulus should be derotated anticlockwise, noting the number of turns. The bowel is examined for viability and any ischaemic bowel should be wrapped in a damp swab and re-examined after 5–10 min. Non-viable bowel is resected and a primary anastamosis formed. If extensive ischaemic bowel of doubtful viability is present, a second-look laparotomy is performed after 24 h with the aim of minimizing the extent of bowel resection required. Ladd's bands are divided.

The SMA is identified and mesenteric base broadened as much as possible by division of the peritoneal folds. Care must be taken not to injure the superior mesenteric vessels. The abnormal position of the appendix may cause diagnostic problems in future and, therefore, removal is advocated. The bowel is replaced with the duodenum to the right and the caecum in the left upper quadrant. The abdomen is closed. The nasogastric (NG) tube is aspirated hourly for the first 24 h. Intravenous fluids are continued postoperatively and NG tube fluid loss is replaced, millilitre for millilitre, with normal saline and potassium chloride (20 mmol/l saline). Enteral feeds are restarted when aspirates are clear and reducing in volume, usually after 24 h.

Laparoscopy may be used in non-acute cases of malrotation without volvulus, e.g., in incidentally diagnosed malrotation. The patient is positioned supine with the legs abducted. The surgeon stands between the patient's feet with the assistant to the left of the patient. The umbilical port is placed first. A periumbilical incision is made. The midline fascia is held in two arterial clips, one on either side of the midline. The linea alba is divided and a 5- or 10-mm port placed into the abdominal cavity under direct vision. The port is secured with a purse-string and the ends of the sutures attached to an anchor on the port. Carbon dioxide is insufflated via the port until a final intra-abdominal pressure of 8–10 mmHg is reached in an infant, or 10–12 mmHg in an older child.During insufflation the abdomen is palpated and percussed to ensure adequate pneumoperitoneum is achieved. The flow rate of carbon dioxide is set between 0.5 and 1.5 l/min. The laparoscope is then inserted into this port. Two further 5-mm ports are placed under direct camera vision – left lower quadrant and right lower quadrant.Non-traumatic grasping forceps are inserted into these ports to manipulate the bowel.

The anatomy is defined and Ladd's bands identified. Care must be taken to correctly identify landmarks such as the duodenum and ascending colon. To gain access to the duodenum, it is useful to raise the head of the operating table and elevate the right flank. The ascending colon falls towards the left side of the abdomen. The duodenum is exposed and Ladd's bands are divided using either an ultrasonic blade or a combination of sharp dissection and electrocautery. After division, the bowel is examined along its length for any further causes of obstruction. The root of the mesentery is broadened by dividing the peritoneal folds. Care must be taken in not injuring the superior mesenteric vein. Appendicectomy is carried out either using an endoloop for intracorporeal ligation or by delivering the appendix through a troacar site and excising it extra-abdominally in smaller patients. Troacar sites are closed.

Conclusion.The outcome of patients undergoing Ladd's procedure for isolated malrotation is very good and the majority make a full recovery. The commonest postoperative complication is adhesional obstruction (3–5%). Midgut volvulus occurs in 45–65% of children with malrotation and still carries a mortality rate of 7–15%; necrosis of more than 75% of the midgut is associated with short bowel syndrome. Up to 18% of children with short bowel syndrome on long term total parenteral nutrition have an original diagnosis of midgut volvulus.

5. Additional materials for the self-control

A. Clinical cases

Case 1.
In a newborn infant vomiting gastric fluid occurred. Meconium stools is in small amounts, in grey colour. The abdomen is distended in epigastrium, scaphoid in lower part. The X-ray film shows two horizontal levels in epigastrium region. What diagnosis have you made?

Case 2. In a 21-day infant 3 days ago gusher-type vomiting occurred after feeding. The vomitus consists curdled milk; it is not bile-stained. The examination results are: loose skin, the patient is poorly nourished. The symptom of "sand-glass" can be seen on the anterior abdominal wall, the abdomen is soft. What diagnosis have you made? What method of diagnostics would you apply?

Case 3. In a new-born 4 hours after birth, there appeared vomiting every 1-2 hours with the content of the stomach and milk with the admixture of bile. The state is not broken. The abdomen is distended in epigastrium. Meconium stool is in small amounts. What diagnosis have you made? What auxiliary methods of examination are to be used?

Case 4.
In a new-born on the second day after birth there appeared vomiting with the intestinal content. The abdomen is distended, soft. On the X-ray film of the abdominal region horizontal levels in the right inguinal area are visible. There was no meconium stool. What diagnosis have you made? What tactics have you developed?

B. Tests

6. LITERATURE FOR STUDENTS

1. Coran AG: Vascular Access and Infusion Therapy. Seminars in Pediatric Surgery 1(3): 173-241, 1992
2. Welch KJ, Randolph JG, Ravitch MM, O'Neill JA, Rowe MI. Pediatric Surgery. 4th edition. Chicago. Year Book Medical Publishers. 1986
3. Ashcraft KW, Holder TM. Pediatric Surgery. 2nd edition. Philadelphia. W.B. Saunders Co. 1993
4. Grosfeld JL. Common Problems in Pediatric Surgery. 1st edition. St Louis. Mosby Year Book. 1991
5. Ashcraft KW. Holder TM. Pediatric Esophageal Surgery. 1st edition. Orlando. Grune & Stratton, Inc. 1986
6. Seeds JW, Azizkhan RG. Congenital Malformations: Antenatal Diagnosis, Perinatal Management and Counseling. 1st edition. Maryland. Aspen Publishers, Inc. 1990
7. Puri P (ed): Congenital Diaphragmatic Hernia. Mod Probl Paediatr. Basel, Karger, Vol 24, 1989
8. Kenneth S. Azarow, Robert A. Cusick. Pediatric Surgery. 2012
9. Lewis Spitz, Arnold G. Coran - Operative Pediatric Surgery, 7th Edition, 2013
10. Peter Mattei-Fundamentals of Pediatric Surgery, 2011

http://6years.net/
http://vk.com/student_unite
http://nmu-s.net/
http://www.amnu.gov.ua/
http://medsoft.ucoz.ua/
http://www.booksmed.com/
http://www.medprizvanic.org/
http://www.medkniga.ukoz.net/
http://www.mednik.com.ua/
http://www.libriz.net/