MINISTRY OF HEALTH OF UKRAINE
Ukrainian Medical Stomatological Academy

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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	Developmental abnormalities of the kidney and urinary tract and congenital malformations of the urinary collecting system. Anomalies of the male genital organs and urethra
Course	VI
Faculty	foreign students preparation

POLTAVA 2020

1.The topic basis: The developmental abnormalities of the kidney and urinary tract make 45% all innate malformations. Among obstructive uropathies in child's age hydronephrosis is on the I place. The topic "Developmental abnormalities of the kidney and urinary tract and congenital malformations of the urinary collecting system" 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.

Actuality of theme of employment is conditioned distribution of violations of genesial function for men as a result of innate vices and consequences of inflammatory processes of privy parts. Absence of adequate treatment at this pathology results in high-frequency of atrophy of testicle, losses by it of the endocrine and genesial function.

The topic "Anomalies of the male genital organs and urethra" 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:

To master the list of diseases which cause violation of urodynamics and act of urination.
To recognize the basic clinical displays of violations of urodynamics and act of urination.
To differentiate violation of urodynamics depending on the level of obstacle.
To interpret the auxiliary methods of research, laboratory and biochemical analyses.
To show the cannulation of urinary bladder, characterize composition of urine.
To identify the features of flow of separate malformations which result in violation of urodynamics.
To analyse the reasons and consequenses of origin of violation of urodynamics, syndrome of leucocyturia for separate patients, to ground and formulate a previous clinical diagnosis.
To offer the algorithm of action of doctor at violation of urodynamics and violations of act of urination.
To offer tactic of conduct of patients with disorders of act of urination and violation of urodynamics.
To interpret general principles of treatment of diseases which are accompanied violation of urodynamics and disorders of act of urination.
To define a testimony to conservative and operative treatment of patients with floggings of development of the urinary and sexual systems.
A student must have general presentations about embryogenesis of privy parts.
A student must know: classification, clinical displays, diagnostics, complication and principles of treatment of anomalies of position of testicles
A student must seize: by the method of review of children with the malformations of privy parts by the method of diaphanoscopy, by the technique of raising of urinary catheter, by the method
A student must be able: to diagnose the innate malformations of privy parts, to distinguish the features of clinical picture depending on nosology unit, to conduct differential diagnostics.
A student must develop the creative capabilities: in the process of clinical inspection and treatment of children with the malformations of privy parts, conducting research in obedience to by the conducted department by research work

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

Names of previous disciplines	**Obtained skills**
1. Anatomy, operative Surgery, Topografic Anatomy	Anatomy and topographic anatomy of genital organs . Set the places of execution of operative accesses .

2. Physiology.	Anatomico-physiological properties of the genitourinary system
3. Pathological Physiology	Indicators 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.
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	Symptoms of the pathology of the urinary system Measure blood pressure, heart rate, the number of respiratory movements per minute, conduct a survey of the abdomen (palpation, percussion, auscultation), rectal examination
6. Urology	Principles of care for urological patients. General semiotics of urological pathology Measure CVP, establish urinary catheter, catheterize veins

4. Thereotical questions for the lesson:

1. Definition of the terms hydronephrosis, ureterohydronephrosis.
2. Define the concept of naming the classification of bladder and urethral anomalies.
3. To explain the pathogenesis of urodynamic disorders due to defects in the development of the kidney, ureter, bladder.
4. To characterize the clinical manifestations of nosology of the topic.
5. To substantiate the need to choose diagnostic measures for different forms of pathology.
6. To determine the indications for the use of ultrasound, cystoscopy, cystography, excretory urography, computed tomography, MRI, laparoscopy.
7. Determine the timing and extent of surgery for various forms of urinary tract pathology.
8. To prescribe antibacterial therapy for infection of the urinary system.
9. Algorithm of the doctor's action in determining birth defects and tactics of patient management in the first months of life.
10. Algorithm of action of pediatric surgeon in determining congenital malformation of bladder and urethra and tactics of patient management.

5. Maintenance of the subject

Simple Renal Ectopia

When the mature kidney fails to reach its normal location in the "renal" fossa, the condition is known as renal ectopia. The term is derived from the Greek words ek ("out") and topos ("place") and literally means "out of place." It is to be differentiated from renal ptosis, in which the kidney initially is located in its proper place (and has normal vascularity) but moves

downward in relation to body position. The ectopic kidney has never resided in the appropriate location.

An ectopic kidney can be found in one of the following positions: pelvic, iliac, abdominal, thoracic, and contralateral or crossed. Only the ipsilateral retroperitoneal location of the ectopic kidney is discussed here. Thoracic kidney and crossed renal ectopia (with and without fusion) are described later.

With the increasing use of radiography, ultrasonography, and radionuclide scanning to visualize the urinary tract, the incidence of fortuitous discovery of an asymptomatic ectopic kidney is also increasing. The steady rise in reported cases in recent years attests to this fact.

Most ectopic kidneys are clinically asymptomatic. Vague abdominal complaints of frank ureteral colic secondary to an obstructing stone is still the most frequent symptom leading to discovery of the misplaced kidney. The abnormal position of the kidney results in a pattern of direct and referred pain that is atypical for colic and may be misdiagnosed as acute appendicitis or as pelvic organ inflammatory disease in female patients. It is rare to find symptoms of compression from organs adjacent to the ectopic kidney. Patients with renal ectopia may also present initially with a urinary infection or a palpable abdominal mass. Several cases of a rare association of renal and ureteral ectopia causing urinary incontinence have been reported. The difficulty in diagnosing this condition is related to the poor function of the ectopic kidney. Dimercaptosuccinic acid (DMSA) scintigraphy or computed tomography with contrast will delineate these unusual cases.

Horseshoe Kidney

The horseshoe kidney is probably the most common of all renal fusion anomalies. It should not be confused with asymmetric or off-center fused kidneys, which may give the impression of being horseshoe-shaped. The anomaly consists of two distinct renal masses lying vertically on either side of the midline and connected at their respective lower poles by a parenchymatous or fibrous isthmus that crosses the midplane of the body. It was first recognized during an autopsy by DeCarpi in 1521, but Botallo in 1564 presented the first extensive description and illustration of a horseshoe kidney. In 1820 Morgagni described the first diseased horseshoe kidney and since then more has been written about this condition than about any other renal anomaly. Almost every renal disease has been described in the horseshoe kidney.

Generally, the isthmus is bulky and consists of parenchymatous tissue with its own blood supply. Occasionally it is just a flimsy midline structure composed of fibrous tissue that tends to draw the renal masses close together.It is located adjacent to the L3 or L4 vertebra just below the origin of the inferior mesenteric artery from the aorta. As a result, the paired kidneys tend to be somewhat lower than normal in the retroperitoneum. In some instances, the anomalous kidneys are very low, anterior to the sacral promontory or even in the true pelvis behind the bladder.The isthmus most often lies anterior to the aorta and vena cava, but it is not unheard of for it to pass between the inferior vena cava and the aorta or even behind both great vessels.

Symptoms: Almost one third of all patients with horseshoe kidney remain asymptomatic. In most instances, the anomaly is an incidental finding at autopsy. When symptoms are present, however, they are related to hydronephrosis, infection, or calculus formation. The most common symptom that reflects these conditions is vague abdominal pain that may radiate to the lower lumbar region. Gastrointestinal complaints may be present as well. The so-called Rovsing sign—abdominal pain, nausea, and vomiting on hyperextension of the spine—has been infrequently observed. Signs and symptoms of urinary tract infection occur in 30% of patients, and calculi have been noted in 20% to 80%. Five percent to 10% of horseshoe kidneys are detected after palpation of an abdominal mass. Horseshoe kidneys have been detected after angiography for evaluation of an abdominal aortic aneurysm.

UPJ obstruction causing significant hydronephrosis occurs in as many as one third of individuals. The high insertion of the ureter into the renal pelvis, its abnormal course anterior to the isthmus, and the anomalous blood supply to the kidney may individually or collectively contribute to this obstruction.

Anomalies of Rotation

The adult kidney, as it assumes its final position in the "renal" fossa, orients itself so that the calyces point laterally and the pelvis faces medially. When this alignment is not exact, the

condition is known as malrotation. Most often, this inappropriate orientation is found in conjunction with another renal anomaly, such as ectopia with or without fusion or horseshoe kidney. This discussion centers on malrotation as an isolated renal entity. It must be differentiated from other conditions that mimic it and are caused by extraneous forces such as an abnormal retroperitoneal mass.

It is thought that medial rotation of the collecting system occurs simultaneously with renal migration. The kidney starts to turn during the 6th week, just when it is leaving the true pelvis, and it completes this process, having rotated 90 degrees toward the midline, by the time ascent is complete, at the end of the 9th week of gestation.

The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent. Weyrauch (1939), in an exhaustive and detailed study, outlined the various abnormal phases of medial and reverse rotation and labeled each according to the position of the renal pelvis.

Rotation anomalies per se do not produce any specific symptoms. The excessive amount of fibrous tissue encasing the pelvis, UPJ, and upper ureter, however, may lead to a relative or actual obstruction of the upper collecting system. Vascular compression from an accessory or main renal artery or distortion of the upper ureter or UPJ may contribute to impaired drainage. Symptoms of hydronephrosis (namely, dull, aching flank pain) may be experienced during periods of increased urine production. This is the most frequent cause of symptoms. Hematuria, which occurs occasionally within a hydronephrotic collecting system from jostling of sidewalls, may be noted as well. Infection and calculus formation, each with its attendant symptoms, may also occur secondary to poor urinary drainage.

Abnormalities of the Testis

Undescended Testicles

The most common anomaly in the male genital tract is cryptorchidism. Maternal HCG stimulates the fetal testis to secrete testosterone which influences testicular development and descent. The normal male with descended testes demonstrates a transient increase in testosterone at sixty days after birth. This testosterone response seems to be blunted in children with undescended testes. Many other factors both hormonal and mechanical can result in undescended testes. Subsequent fertility and testicular tumor formation are the most common problems related to cryptorchid testes.

Undescended testes may be truly cryptorchid or ectopic. Cryptorchid testes may be an abdominal, inguinal or suprascrotal position. Ectopic testes can assume any position outside of the inguinal canal (Figures1 and 2). Three percent of term infants and thirty three percent of premature babies will have a cryptorchid testis. Virtually all testes that eventually descend will do so by six months of age, therefore, children with undescended testes after six to eight months of age should be considered for surgical correction. HCG stimulation is usually reserved for bilateral undescended testes. Even though it may not result in complete descent, subsequent orchidopexy is often technically easier after hormonal manipulation. Laparoscopy as both a diagnostic and therapeutic tool is being utilized more often especially in cases of non-palpable testes.

Retractile Testes

Retractile testes result from hyperactivity of the cremaster muscle that causes retraction of the testicle into the inguinal canal. On examination with gentle manipulation the affected testis can be manipulated into its respective hemiscrotum where it will remain when the cremaster muscle is relaxed. Testicular development and maturation is normal and surgical intervention is not indicated. Children with retractile testes should have a yearly examination until the age of puberty at which time the cremaster reflex lessens and the testes assume their normal scrotal position.

Ascending Testis

An ascending testicle has been described recently in which a previously documented descended scrotal testis later comes into an extrascrotal position. This is a result of a delayed resorption of the processus vaginalis and a "pulling up" of the testicle. The affected testis feels tethered and cannot be manipulated into a dependent scrotal position. Ninety-eight percent of ascended testes are associated with a patent processus vaginalis and, therefore, a potential inguinal hernia and surgical correction is necessary.

Vanished Testis

A vanished testis describes the child with a nonpalpable undescended testicle who upon examination may have a small hard pea sized structure in the affected hemiscrotum which represents an atrophic calcified testicular remnant. This condition is most often related to a neonatal or prenatal testicular torsion or other early vascular accident with subsequent testicular atrophy (Figure 3). Treatment includes surgical exploration and excision of the testicular remnant. Often laparoscopy is used to search for an intraabdominal testis and at laparoscopy a blind ending spermatic vessel and vas deferens can be seen entering the internal inguinal ring.

Disorders of the Penis

Hypospadias

Hypospadias is a common anomaly occurring in one in two hundred and fifty male children. The exact etiology of hypospadias is unknown; however, worldwide the incidence of hypospadias seems to be increasing. Environmental pollutants with estrogen like affects have been implicated, but not proven in the cause of hypospadias.

Hypospadias occurs as a result of incomplete fusion of the urethral groove producing a urethral meatus located in an aberrant location on the ventral surface of the penis. The meatus can be located anywhere along the ventral penile shaft from the meatus distally to the perineum in the most severe cases of proximal hypospadias. Typically, the corpus spongiosusm is absent or poorly developed surround the hypospadiac urethra. Chordee, or ventral curvature of the penis is a commonly associated finding and should be described as mild moderate or severe.

Hypospadias is best classified according to the location of the urethral meatus: 1) glanular; 2) coronal; 3) distal (Figure 4), mid or proximal shaft (Figure 5); 4) scrotal (Figure 6); or 5) perineal. Coronal and glanular defects account of over 85% of cases hypospadias, whereas perineal hypospadias is present in only two to three percent of children with this anomaly. There does not seem to be a significant increase in the incidence of urinary tract anomalies in children with hypospadias, therefore, routine screening with an IVP or ultrasonography and voiding cysto-urethrography are generally not indicated unless urinary infections are present.

Most cases of hypospadias should be surgically corrected with the goal of producing a straight penis with the meatus at the tip of the glans and a cosmetically pleasing appearance. Even severe degrees of hypospadias can usually be repaired in a one staged procedure on an outpatient basis. The optimal age for correction is between six and twelve months of age.

Retractile Penis

The retractile or concealed penis is most often noted in young boys after circumcision. The penis rather than protruding normally tend to retract beneath the prepubic fat pad resulting in an uncircumcised appearance. Parents often comment on the "lousy job" done by the circumcising physician. In fact, however, this condition has little to do with the technique of the circumcision, but rather relates to the presence of dysgenetic bands of tissue in the dartose layer of fascia that act as "rubber bands" to retract the penis proximally. Surgical correction requires division of these dysgenetic bands in severe cases (Figures 7 and 8). Less severe cases of penile retraction may actually improve at the time of puberty.

Cysts and Webs

Penile cysts can occur anywhere along the penile or scrotal raphe. They are usually asymptomatic, but can get secondarily infected. Cysts are due to entrapment of epithelial cells during fusion of the labial scrotal folds. Epidermal inclusion cysts, sebaceous cysts and mucoid urethral cyst also can occur (Figure 9). In uncircumcised boys entrapped squamous debris between the inner prepuce and glans can accumulate and is often misconstrued as a cyst. The "infantile smegma" will eventually work its way to the surface and gently separate the prepucial-glanular adhesions. No treatment is necessary for this infantile smegma and forcible separation of the prepucial adhesions from the glans should be discouraged.

Penoscrotal webbing describes a fusion of the ventral penile and scrotal skin resulting in a loss of the penoscrotal angle (Figure 10). The penis has a "volcano" appearance and is usually associated with a significantly tight phimosis. Neonatal circumcision is contraindicated and correction should be undertaken at six to eight months of age.

Penoscrotal Transposition And Ectopic Scrotum

The presence of ectopic scrotal tissue is very uncommon and usually occurs in the presence of severe colloquial extrophy. Occasionally ectopic scrotal tissue can be found in a child with otherwise normal genitalia. The ectopic scrotal tissue is usually located in the inner thigh or near the external inguinal ring. Typically the ipsilateral testicle lies within the ectopic scrotum (Figure 11). Surgery involves either transposition or excision of ectopic scrotal tissue with placement of the testicle in a normal scrotal position.

Penoscrotal transposition is an anomaly frequently associated with severe hypospadias. The penis lies entirely behind the scrotum in complete transposition (Figure 12). In less severe forms, the penis may appear to arise from center of the scrotum or be enveloped by the scrotum. Correction of penoscrotal transposition is usually carried out at the time of hypospadias repair either as a single stage or in multi-staged procedures.

Hernias and Hydroceles

Congenital hydroceles are a very common finding in newborn male children. There are two classifications of hydroceles based upon whether or not the processus vaginalis is patent. Noncommunicating hydroceles represent fluid contained within the tunica vaginalis surrounding the testis after the processus vaginalis has obliterated. Simple noncommunicating hydroceles are very common at birth and usually resolve within the first year of life. Hydroceles that persist after one year of age usually do not resolve and surgical correction is indicated (Figure 13)

Communicating hydroceles represent fluid within the tunica vaginalis associated with a patent processus vaginalis. Communicating hydroceles often fluctuate in size depending upon the gravitational flow of fluid between the abdominal cavity and tunica vaginalis surrounding the testis. These hydroceles typically will enlarge in the evening when the child is supine at night the hydrocele may appear to resolve. Communicating hydroceles that are present after six months of age should be surgically corrected.

Inguinal hernias represent the presence of intraabdominal content that has protruded through the patent processus vaginalis into the inguinal canal and potentially into the scrotum (Figure 14). Inguinal hernias are more common in premature infants and are present in up to three percent of the general population. Because of the potential of incarceration of bowel or injury to the vascular supply to the testis, inguinal hernias should be surgically corrected on an elective basis sometime after diagnosis is made.

Anomalies of the urethra

Among the **anomalies of the urethra** is more common in hypospadias, less epispadias and extremely rare – aplasia and doubling the urethra.

Hypospadias – the most common congenital abnormality that is characterized by underdevelopment of the urethra to the replacement of the missing of dense connective chord, which is the cause of penile curvature. The basis of the numerous classifications of hypospadias put the localization of the external opening of the urethra. Female hypospadias is characterized by the splitting of the rear wall of the urethra and anterior vaginal wall. Epispadias – congenital splitting of the whole or part of the front wall of the urethra. On the dorsal surface of the penile urethra opening is detected.

1. Epispadias glans penis is extremely rare and requires surgical correction.
2. Epispadias penis: external opening of the urethra is located in the crown on the back surface of the penis.
3. Full (total) epispadias – the most severe form in which the external opening of the urethra is at the root of the penis. Hole resembles a wide funnel.
4. Clitoris form of epispadias in girls – a slight splitting of terminal part of the urethra. Most often, this form remains unnoticed.
5. Epispadias which is characterized by the splitting of the urethra to the bladder neck and the splitting of the clitoris.
6. Full epispadias: the front wall of the urethra and the wall of the anterior segment of the bladder neck are missing.

6. Additional materials for the self-control

A. Clinical cases

Case 1.

A 3 year-old girl often complains about pain in the right half of the abdomen. The leukocyturia to 10-30 cells is periodically detected. On the USD considerable expansion of pyelocaliceal complex is educed on the right, the kidney parenchima is thinned, an ureter is not traced. What diagnosis have you made? What tactics have you developed?

Case 2.

A 3-year old girl has been treated conservatively about the recurrent pyelonephritis for the last two years, but the child has not been inspected in the hospital. For clarification of the diagnosis the roentgenologic examination has been carried out. At cystography the reflux of the contrasting matter into the extended winding left ureter and renal pelvis have been detected. What diagnosis have you made? What auxiliary methods of examination are to be used? What tactics have you developed?

Case 3.

The parents complain about the absence of a testicle in scrotum in a 24 month-old boy. At the examination the excalation of the left half scrotum, and a 1,5×1, 0 cm, mobile, densely-elastic, not painful tumular formation in the left inguinal area have been detected. Your diagnosis and tactics?

Case 4.

A boy has the splitting of the prepuce since birth . The urination takes place in a thin flow with a straining effort. The external opening of the urethra is pointed and detected in the area of coronal furrow. Your diagnosis and tactics

6.LITERATURE FOR STUDENTS

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