

Anomalies of development, incorrect position of the female genital organs

Department of obstetric and
gynaecology No 1 KNMU

Anomalies of development of female sexual organs



Anomalies of development of female sexual organs

- it is congenital violations of anatomic structure as uncompleted organogenesis, violation of sizes, form, proportions, symmetry, topography, and also presence of formations not inherent to the woman organism in a postnatal period;
- frequency of anomalies of development of genital organs makes 1 case (1%), however the last years there is its rise.



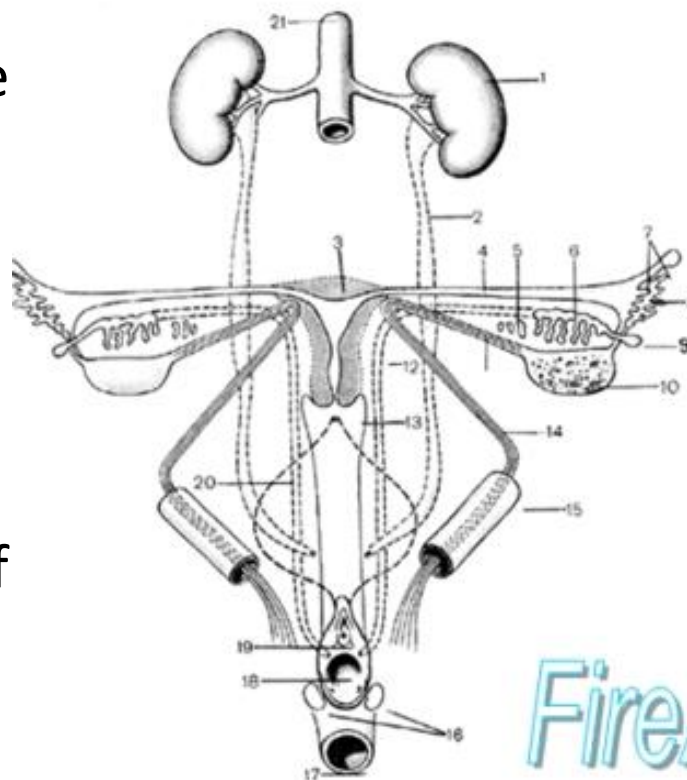
Aetiology

- among reasons of development of anomalies of genital organs are endogenous, exogenous, inherited, multifactor;
- malformations are formed in the process of embryogenesis under influence of harmful factors, breaking the process of embryogenesis;
- to such factors belong:
 - diseases of mother(infectious, endocrine);
 - intoxications(alcohol, narcotic and chemical substances);
 - it is a radiation;
 - complications of pregnancy and labor;
 - the inherited factors(chromosomal and genic mutations);
 - external factors(stress situations, psychologic traumatic experience, long starvation).

Pathogenesis

- related to violation of confluence of paramesonephral (müller's) canals, their recanalisation, defects of forming of urogenital system, and also by pathology of organogenesis of gonads, which depends on the characteristics of development of the middle kidney and timeless gonocytes migration in the embryonic gonad rudiment.

Рис. 19. Развитие внутренних женских половых органов (схема)



- 1 – ren;
- 2 – ureter;
- 3 – uterus;
- 4 – tuba uterina;
- 5 – paroophoron;
- 6 – epoophoron;
- 7 – fimbriae tubae;
- 8 – ostium abdominale tubae uterinae;
- 9 – appendix vesiculosa;
- 10 – ovarium;
- 11 – lig. ovarii proprium;
- 12 – ductus mesonephricus;
- 13 – vagina;
- 14 – lig. teres uteri;
- 15 – canalis inguinalis;
- 16 – gl. vestibulares majores;
- 17 – rectum;
- 18 – ostium vaginae;
- 19 – ostium urethrae externum;
- 20 – vesica urinaria;
- 21 – aorta.

FireAiD - все по медицине.

- in the 50 cases forming of genital organs combines with other anomalies (the malformations of the urinary system, locomotor system, GIT and other.

Classification

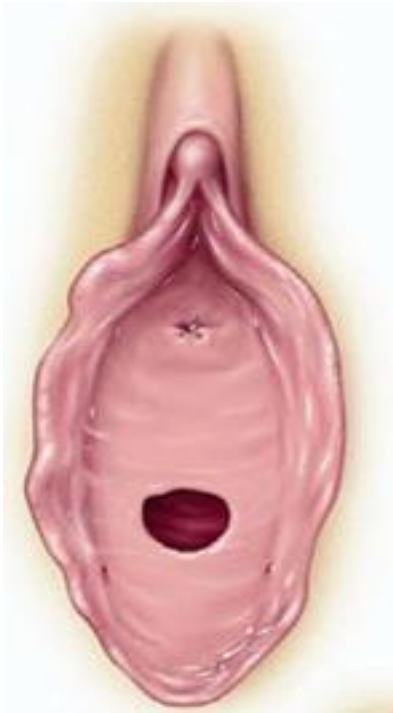
- 1 group is violation of sexual development (infantilism);
- 2 group is violation of pubescence;
- 3 group is anomalies of sexual embryonization (dysgenesis of gonads);
- 4 group is anatomic defects of genital organs.

Terminology

- Agenesis is absence of organ;
 - an aplasia is absence of part of organ;
 - an atresia (imperforation) is absence of opening or canal;
 - a gynatresia is fusion of certain department of the woman sexual system;
 - a hypoplasia is an underdevelopment of organ;
 - a dysraphia is lack of fusion or closing of parts of organ;
 - multiplication is an increase of amount of parts of organ;
 - heterotopia or ectopia is development of organs or tissues in those places, where they in a norm are absent

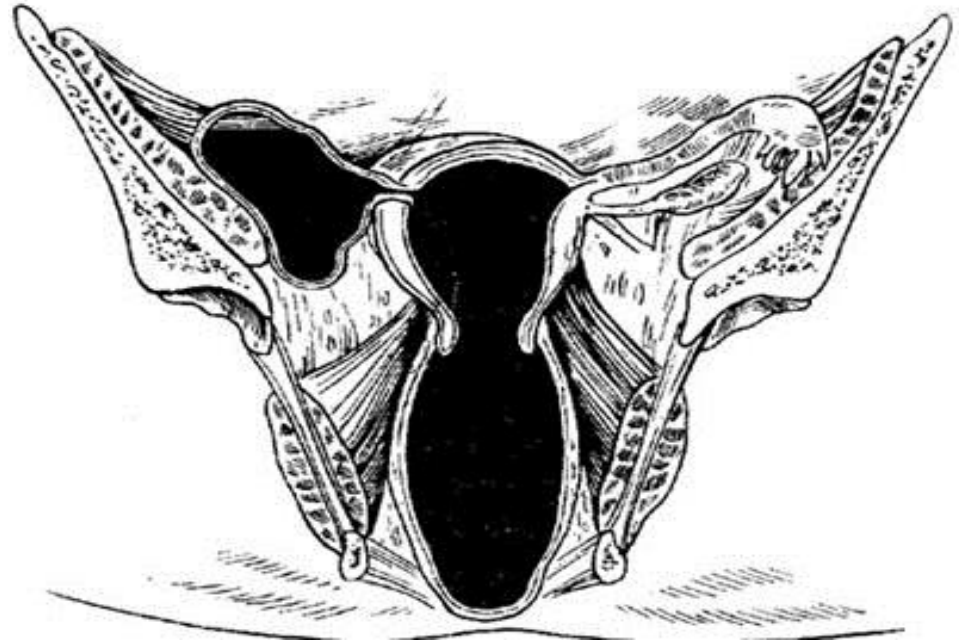
Anomalies of development of hymen and vulva

- at normal development a hymen has opening for the outflow of menstrual blood.
- Atresia of hymen - one of the most widespread innate anomalies of vulva



Anomalies of development of hymen and vulva

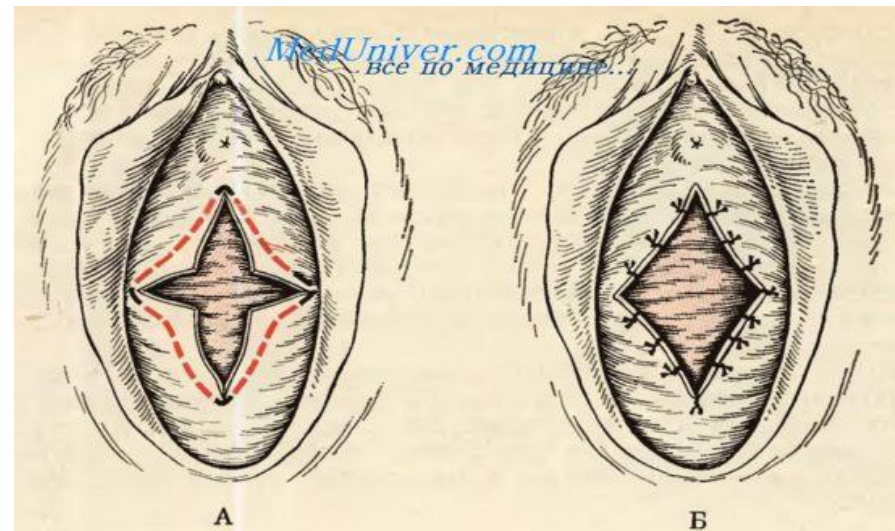
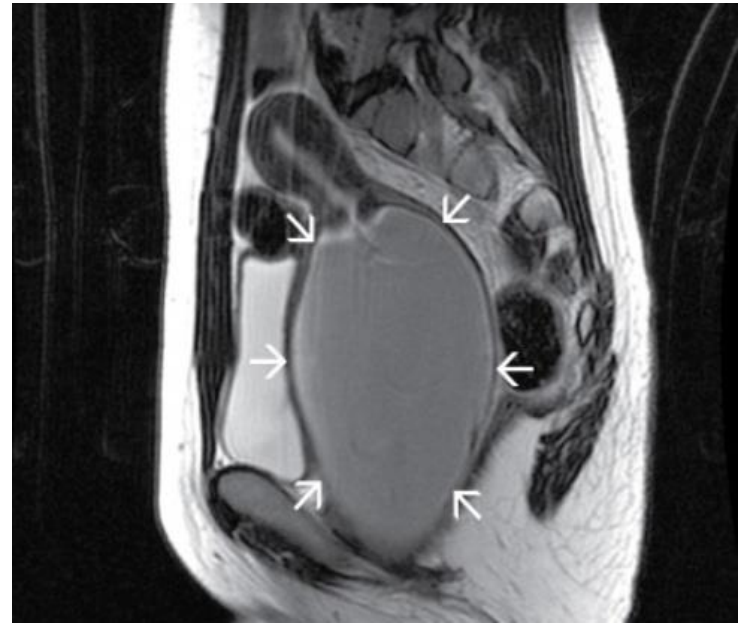
- pathology shows up in the period of pubescence; during menstruation blood fills a vagina, uterus and salpinx gradually; blood also can get to the abdominal region, simulating the clinic of acute abdomen; clinical presentation is characterized by periodic pain in lower abdomen in default of menstruations(pseudoamenorrhea).



- at examination there is the stretched hymen of blue-purple color, absence in it opening is well visible..

Anomalies of development of hymen and vulva

- at rectoabdominal research of patient find out tumular formation of elastic consistency in area of vagina, on the apex of that it is located small dense uterus;
 - in case of difficulty at raising of diagnosis apply ULTRASONIC;
 - surgical treatment consists of providing of outflow of gathering menstrual blood the method of cruciform section of hymen; the edges of cut take in the knotted catgut guy-sutures.



Hermaphroditism

- presence of male and female sexual glands for the same individual;
 - veritable hermaphrodites are persons with two functioning glands of both sexes (phenomenon extraordinarily rare) or with combination in one sexual gland of elements of testicle and ovary (ovotestis);
 - pseudohermaphrodites are persons with the sexual glands of one sex and secondary sexual characters of opposite;
 - in connection with that sex is determined exceptionally by the structure of sexual glands distinguish a male and female pseudohermaphroditism.



Pseudohermaphroditism

- there is a sexual gland masculine at a masculine pseudohermaphroditism, and the structure of external genitals approaches to a female;
 - at a woman - sexual internalss(ovaries, uterus, vagina) are formed correctly, a clitoris is megascopic and reminds a masculine penis, large sexual lips acquire the type of scrotum

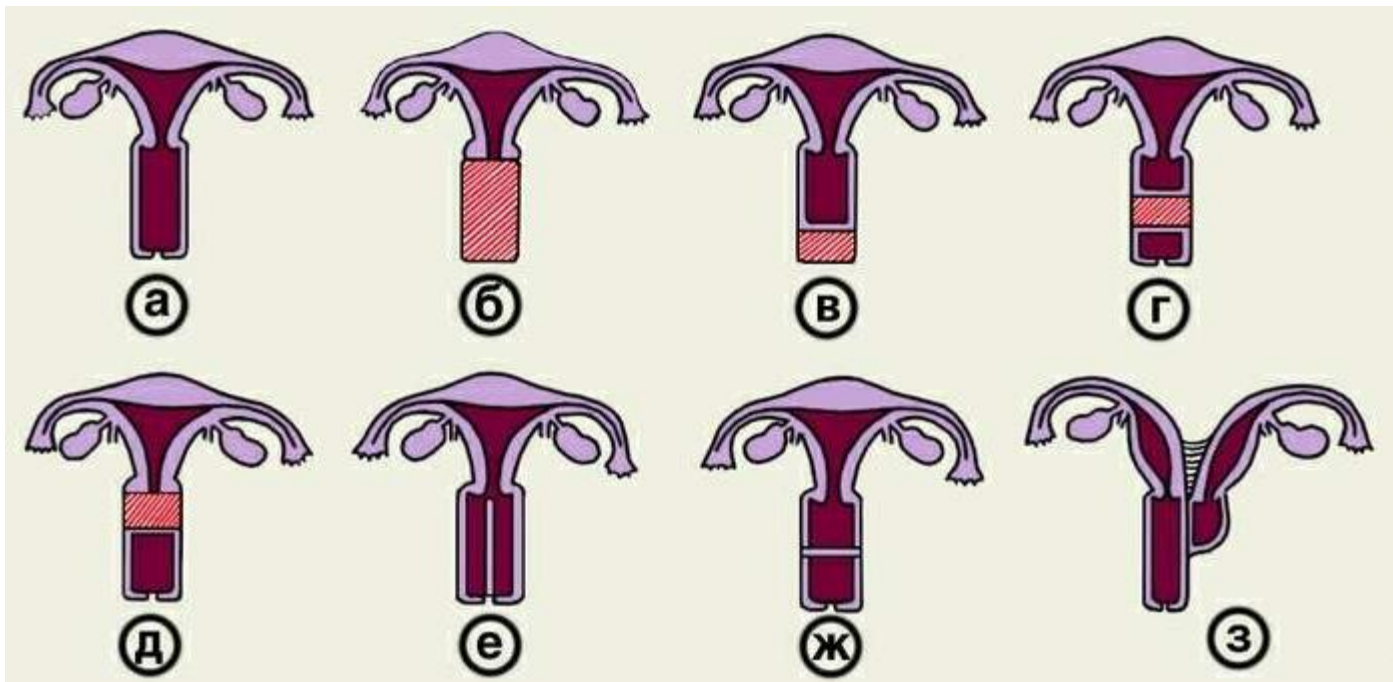


Female pseudohermaphroditism

- develops:
 - on a background of innate adrenogenital syndrome;
 - because of take the androgens or progesterone in high doses during pregnancy;
 - on a background of virilizing tumours of adrenal glands and ovaries;
- treatment:
 - it is amputation of clitoris and section of urogenital bosom in combination with therapy of glucocorticoids;
 - it is ectomy of virilizing tumours

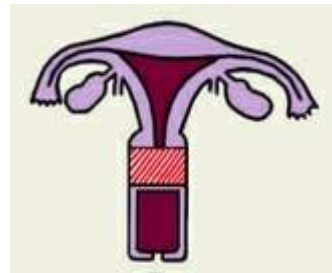
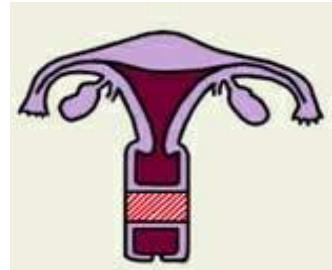
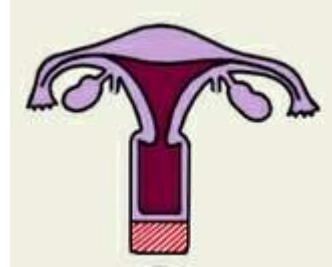
Defects of developments of vagina

- it can be isolated or combine with defects of development of uterus; the variableness of anomalies of development of vagina is related to it forming from different embryonic rudiments: it overhead one third - their meeting paramesonephral channels, lower two one third - from an urogenital bosom;
- the anomalies of development of vagina show up as an imperforation, agenesis and aplasia



Atresia of vagina

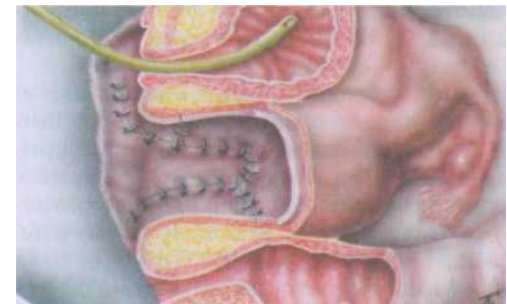
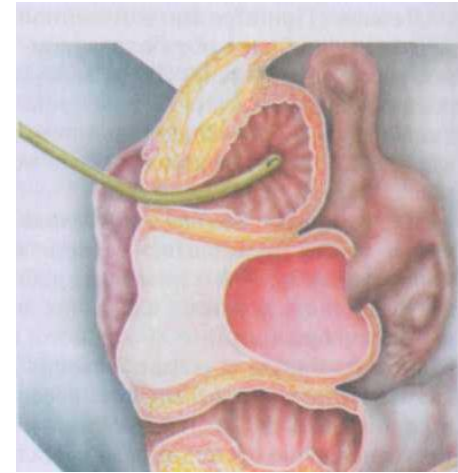
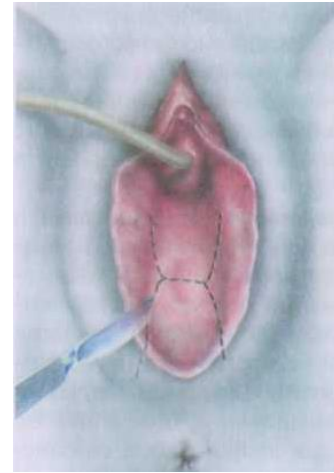
- the complete or partial imperforation in the overhead one, middle or lower departments; arises up on a background of the formed paramesonephral canals that did not meet with an ectoblast urogenital bosom;
 - among clinical displays is periodic pain in a lower abdomen and absence of menstruations; blood accumulates higher than area of imperforation, gradually filling the cavity of uterus, a hematometra is formed;
 - a diagnosis and level of imperforation set the methods of rectal research, ULTRASONIC and sounding of vagina



• .

Treatment of vaginal atresia

- treatment operative : at the imperforation of lower one third of vagina in area of entrance in it, dissect the lower vault of vagina and sew its mucous membrane to entrance in a vagina;
- if an imperforation is localized in middle one third of vagina, then dissect tissues between lower and overhead of its third and then sew together the mucous membrane of these areas



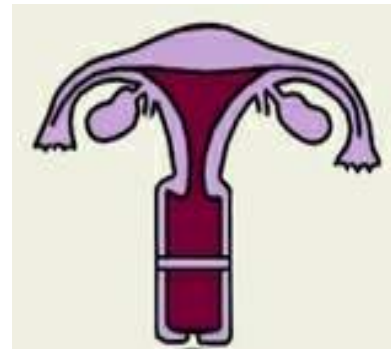
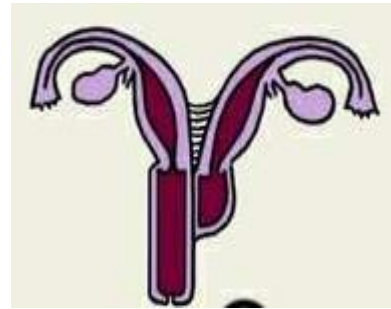
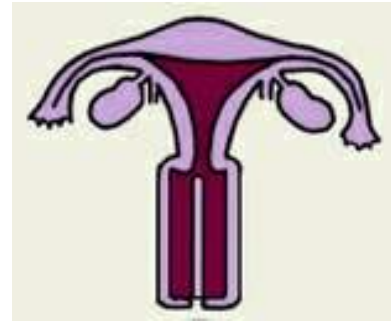
Treatment of vaginal atresia

- at the imperforation of overhead one third of vagina in default of vaginal vaults and location of cervix of the uterus in the tissue of small pelvis operative intervention gives the least effect; an operation is considered effective in case of possibility of emptying hematometra through the chanal of cervix, with the further inserting it in the overhead cupula of the vagina.



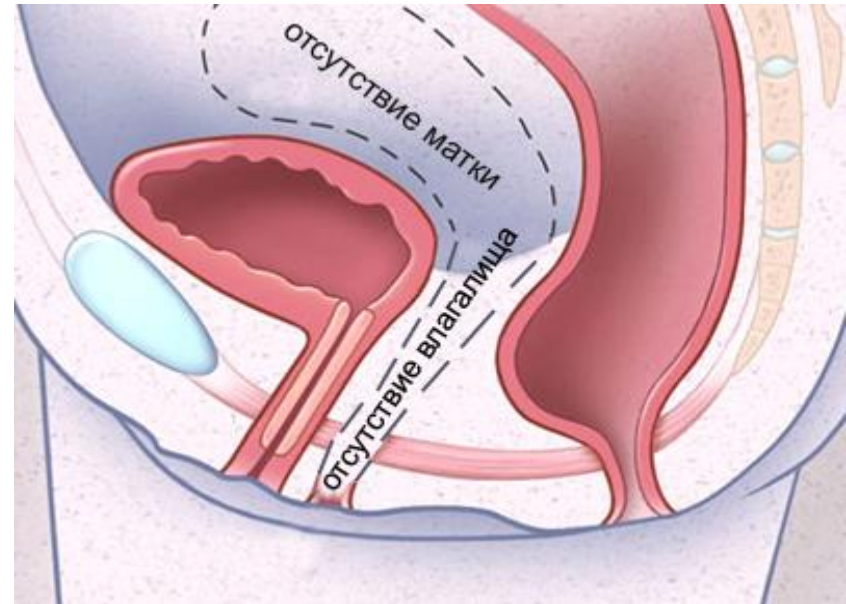
Vaginal septums

- longitudinal vaginal partitions are formed because of incomplete confluence of paramesonephral canals; partitions in a vagina combine with the anomalies of development of uterus(doubling of uterus); sometimes there is an imperforation of one side of vagina and menarche accompanied by development of hematocolpos and hematometra, in this case execute a surgical section or septectomy;
- the transversal partitions of vagina arise up on a background incomplete confluence of paramesonephral canals with an urogenital sinus; such partitions can be complete or incomplete, circular or falx, localized in overhead and middle departments

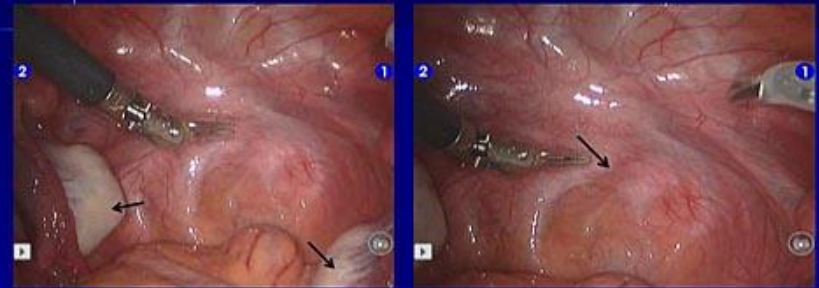


Vaginal agenesis

- complete absence of vagina that arises up because of absence or uncompleted development of paramesonephral channels; agenesis of vagina observed mainly for patients with the syndrome of Rokitansky-Custner, that on a chromosomal set are women with the rudiment of uterus, hypoplastic salpinxs and normal ovaries.



Синдром Майера-Рокитанского-Кюстера

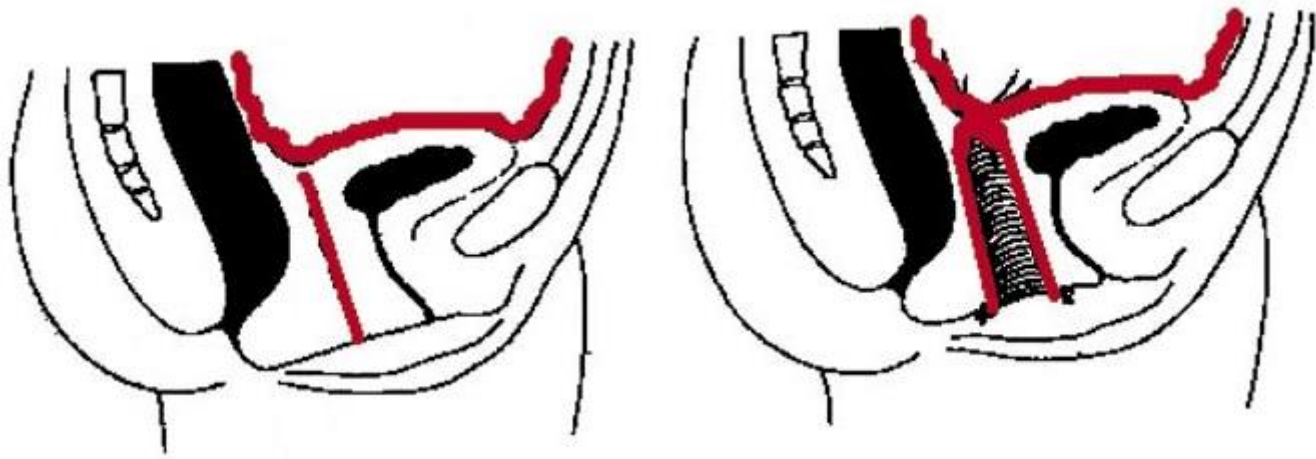


- врожденное отсутствие матки и влагалища
- нормальная функция яичников
- женский фенотип и кариотип
- часто встречающееся сочетание с другими врожденными пороками развития (скелета, органов мочевого выделения)

Aplasia of vagina

- primary absence of part of vagina, conditioned by stopping of the canalisation of vaginal tube on the stage of it forming;
- clinical presentation is characterized by amenorrhea and impossibility to conduct normal sexual life;
- a diagnosis is set on the basis of data of anamnesis and results of gynaecological inspection; from additional methods it is possible to apply laparoscopy ,computer or MRT;
- treatment surgical(a colpopoiesis is creation of artificial vagina); time of realization of operation is determined individually: for patients with the presence of functioning uterus for maintenance of fecundity an operation is executed to the offensive of menarche, in default of uterus an operation is directed for realization sexual function

Colpopoiesis from the peritoneum of small pelvic



Anomalies of uterine development

- arise up because of violation of processes of confluence, wrong forming or incomplete canalisation of paramesonephral channels;
- are reason of sterility, algodismenorrhea, spontaneous abortions, wrong positions of the fetus, discoordination of childbirth, postpartum bleeding, ectopic pregnancy and other

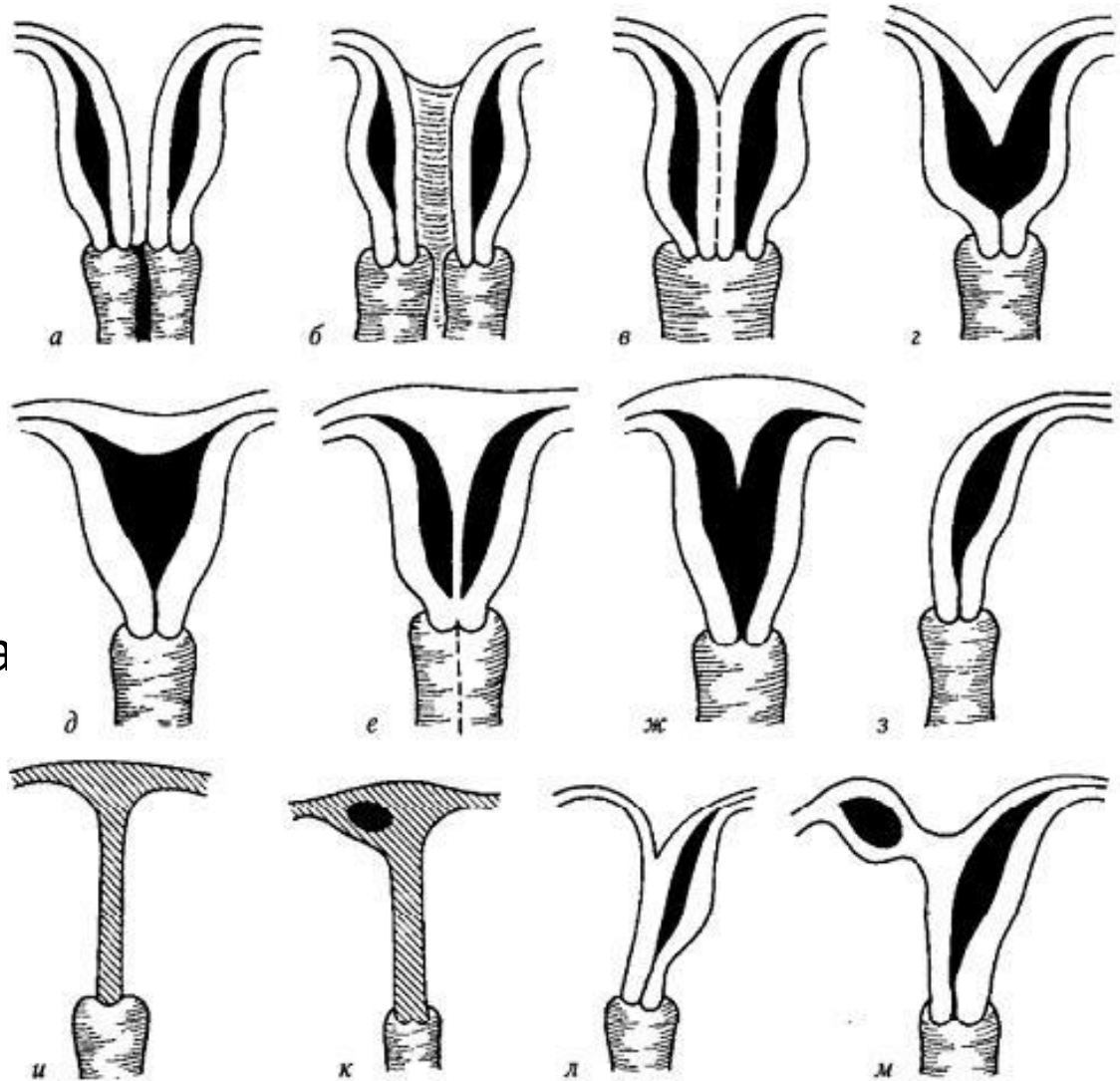
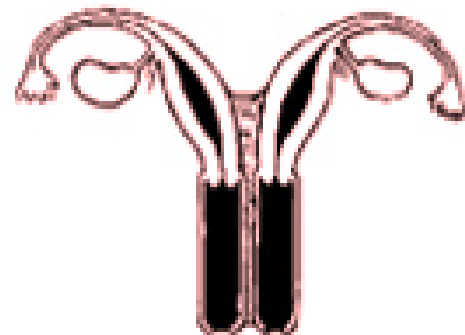


Рис. 10. Аномалии развития матки:

a — uterus didelphus; *б* — uterus duplex et vagina duplex; *в* — uterus bicornis bicollis; *г* — uterus bicornis unicollis; *д* — uterus arcuatus; *е* — uterus septus duplex; *ж* — uterus subseptus; *з* — uterus unicornis; *и, к, л, м* — uterus bicornis rudimentarius

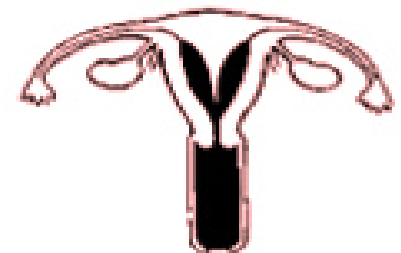
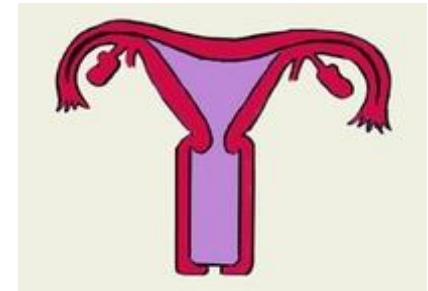
Anomalies of uterine development

- **uterus didelphus** - appears at unconfluence of Muller's channels on all length; two cervix and two vaginas take place, divided by the transversal fold of peritoneum, from every side there are one ovary and one salpinx; both uteruses function well, and pregnancy can come in any of them
- **uterus duplex et vagina duplex** is a presence of two uteruses and two vaginas, connected with each other on certain areas;
- one of uteruses quite often yields other in sizes and functions, thus on the side of exclamation the imperforation of hymen or internal fallopian ostium is *possible*.



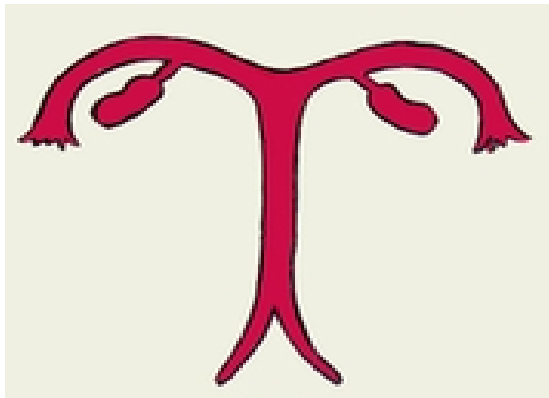
Two horned uterus

- it is result of unconfluence of those parts of paramesonephral channels due to that a single uterus appears usually, thus common vagina develops;
- most expressed variant - **uterus bicornis bicollis**, that shows up the bifurcation of the cervix and body of the uterus;
- uterus **bicornis unicollis** is a bifurcated body of uterus at a common cervix, formed because of confluence of Muller's channels in the area of cervix of the uterus;
- the less expressed variants of the two-horned uterus are a saddle and arched uterus(**uterus arcuatus**), uterus with a complete or incomplete partition(**uterus septus, subseptus**).



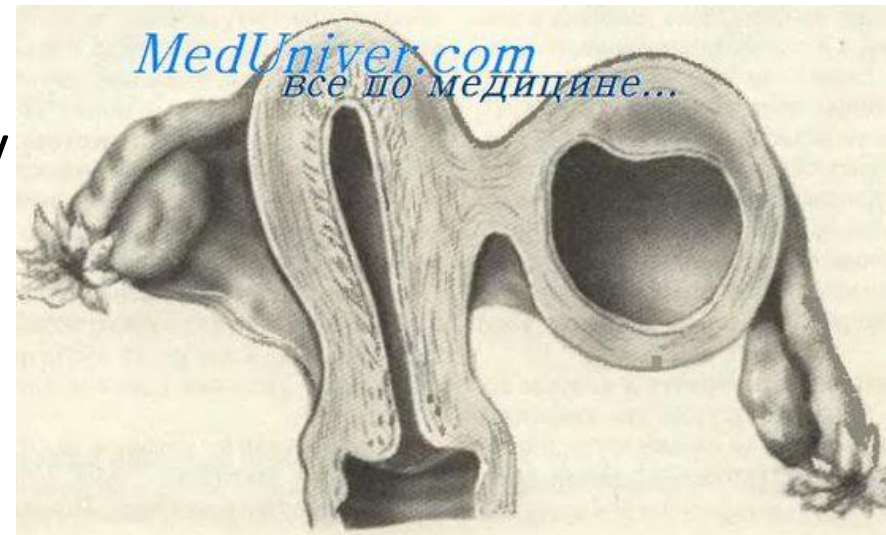
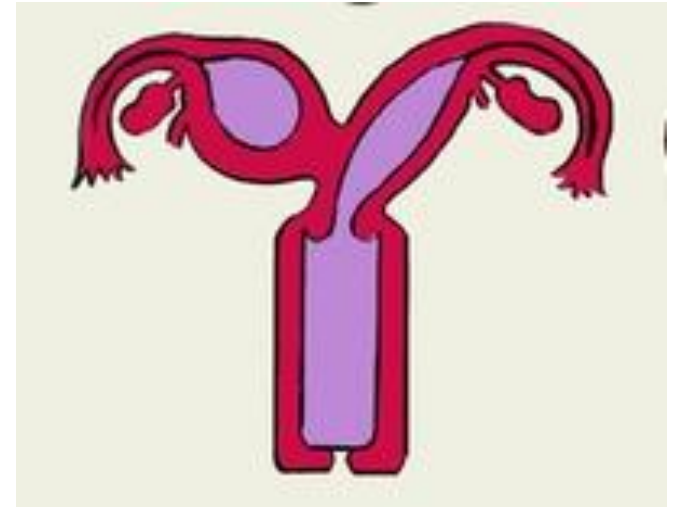
Anomalies of uterine development

- if paramesonephral channels did not meet and does not have a lumen, both uteruses acquire the type of two rudimentary horns of solid structure - **uterus bicornis rudimentalis solidus** or syndrome of Rokytansky-Curstner.
- uterus **unicornis** is the rarest pathology that arises up on a background the ateliosis of one of paramesonephral channels;
- pathology as a rule, shows up a presence only one tube and one ovary



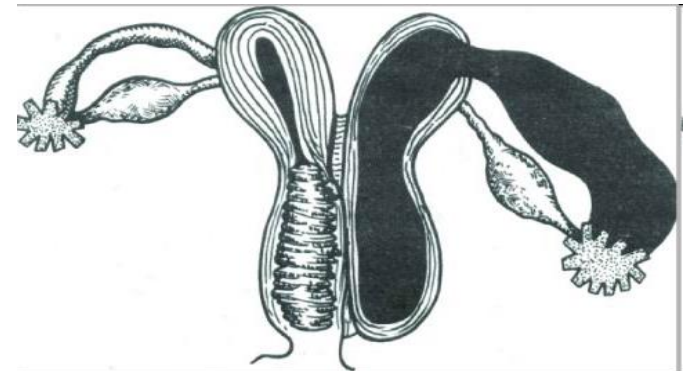
Anomalies of the uterine development

- **uterus bicornis** with a rudimentary horn develops because of considerable exclamation of one of Muller's channels; depending on the state of rudimentary horn distinguish the two-horned uterus with an unfunctioning rudimentary horn and two-horned uterus with functioning rudimentary horn;
- if a rudimentary horn has a cavity, then it is very important to find out where it is connected with the cavity of uterus. The presence of functioning rudimentary horn is accompanied by such complications as polymenorrhea, algodismenorrhea, ectopic pregnancy.



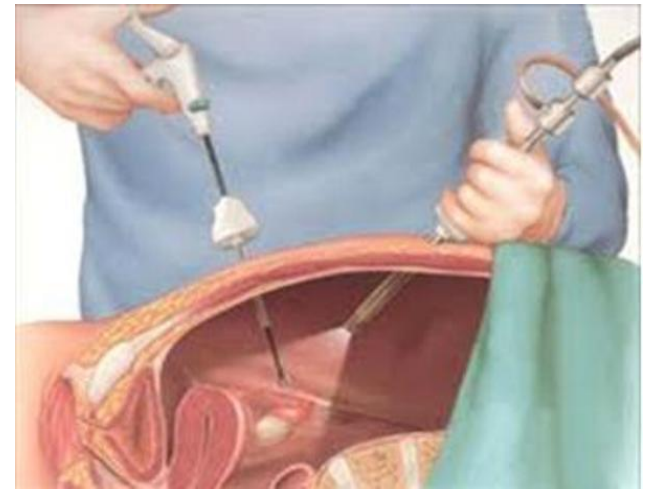
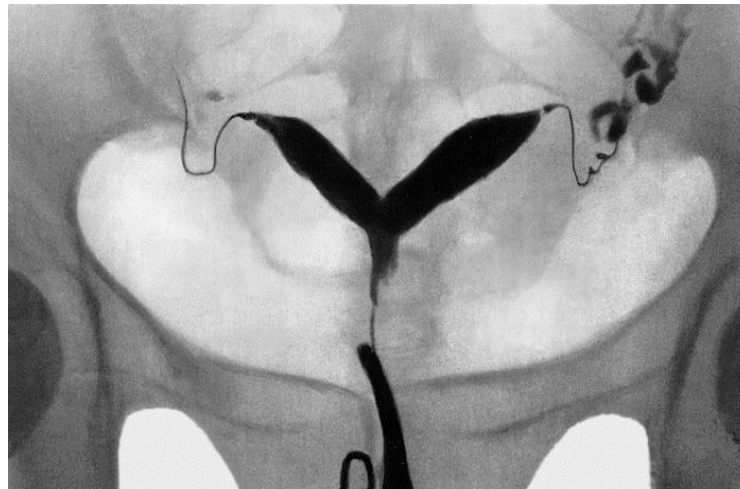
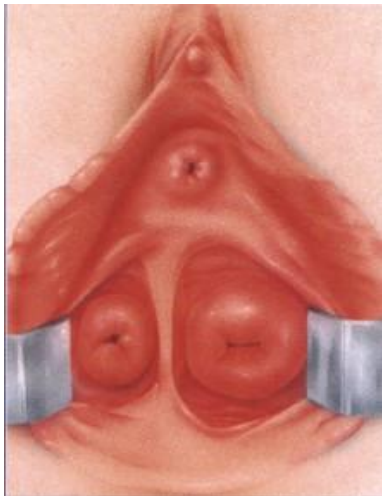
Clinical presentation

- violations of menstrual function as amenorrhea or polymenorrhea;
- there is a abdomen pain (appears in a pubertal period and increases monthly, sometimes accompanied by the unconscious state);
- peritoneal phenomena (in case of infecting of hematometra or hit in the abdominal region of menstrual blood



Diagnostics

- data of anamnesis;
 - clinical displays;
- results of the detailed gynaecological research,
- additional methods of inspection(ULTRASONIC,hysterosalpingography,gysteroscopy, laparoscopy, CT, excretory urography

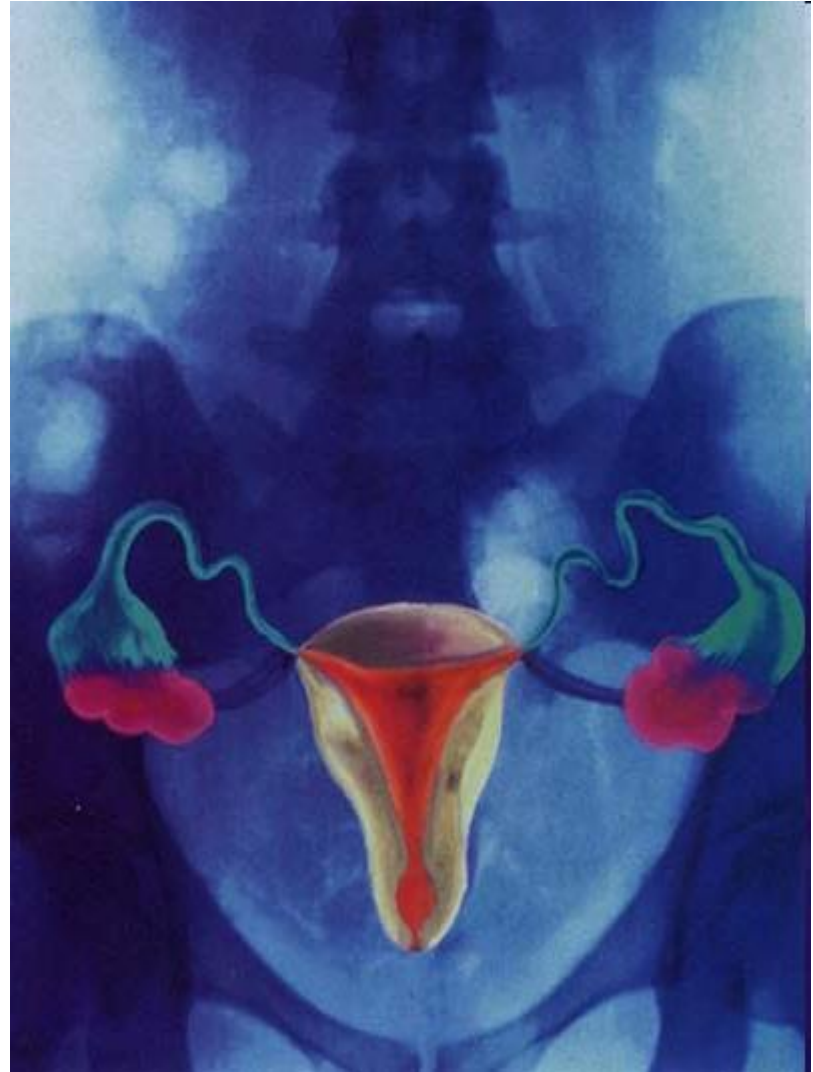


Treatment

- different forms of doubling of uterus and vagina, if they are not accompanied by violation of outflow of menstrual blood, therapy does not need;
- at the delay of outflow of menstrual blood surgical treatment is shown with maximal excision of partition of serried vagina and moving away of functioning horn of uterus;
- surgical treatment(metroplasty) at the two-horned uterus is conducted on testimonies(spontaneous abortions).

Anomalies of the development of uterine tubes

- extraordinarily long salpinx;
- Spiral form of salpinx;
- innate obliteration of tubes;
- imperforation of tubes;
- doubling of tubes (usually attended with doubling of ovaries);
- presence of additional blind ducts, additional openings in the area of the abdominal opening of the tube;
 - innate diverticula of tubes.



Abnormalities of ovaries

- veritable agonadism (aplasia of ovaries);
- pseudogonadism (regressive form);
- veritable hypergonadism (hyperplasia);
- pseudohypergonadism (fibrocystic and polycystic ovaries);
- primary hypogonadism (hypoplasia of ovaries because of anomalies of sexual chromosomes);
- secondary hypogonadism (atrophy of ovaries because of gonadotropic insufficiency of hypophysis);
- ambryogonadism (one and bilateral):
 - one-sided absence of adnexa;
- additional ovaries (is forming as a result of separation of parts of rudiment of ovary on early stages of embryogenesis)

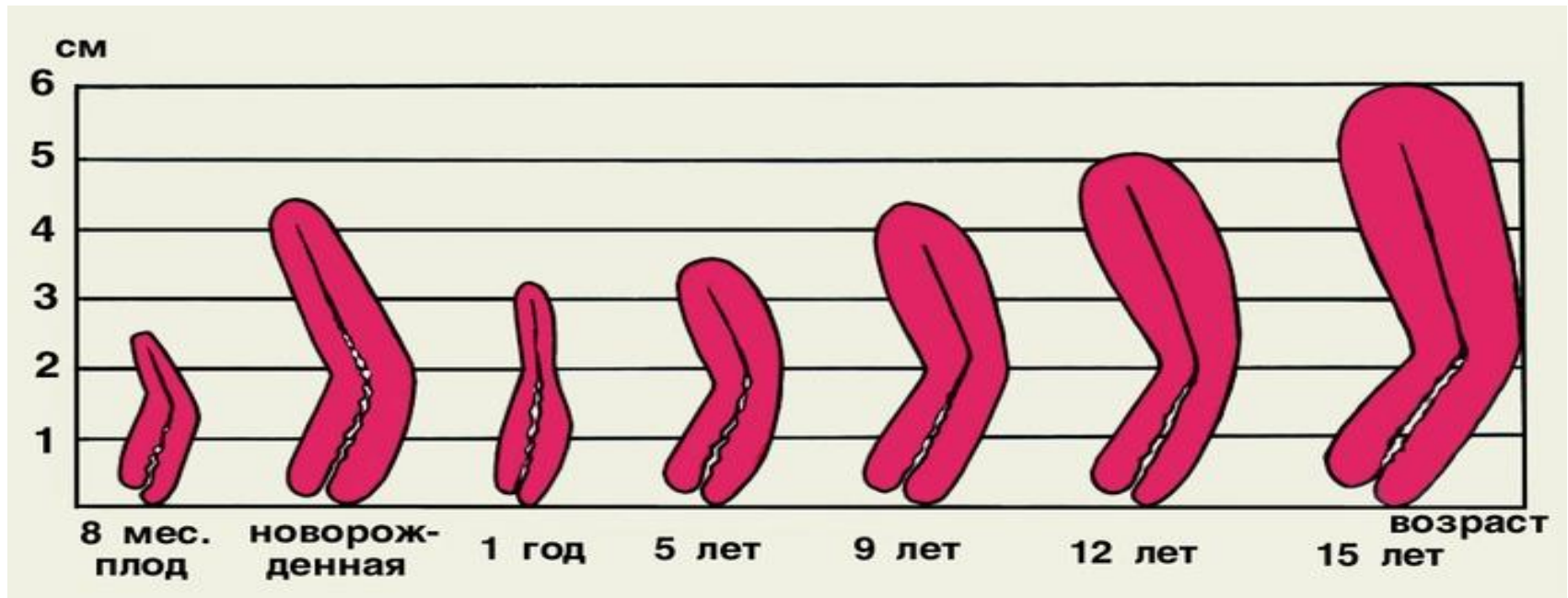


Violations of sexual development

- premature sexual development is appearance of secondary sexual characters for a girl under age 8(because of excessive products of sexual steroids, presence of hormone-productive tumours and other);
- heterosexual development(development on a masculine type);
- delay of sexual development(braking of appearance of secondary sexual characters on 2 and more years: unformed of mammary glands till 13 years, absence of menstrual function and underdevelopment of secondary sexual characters are till 15 years and other).

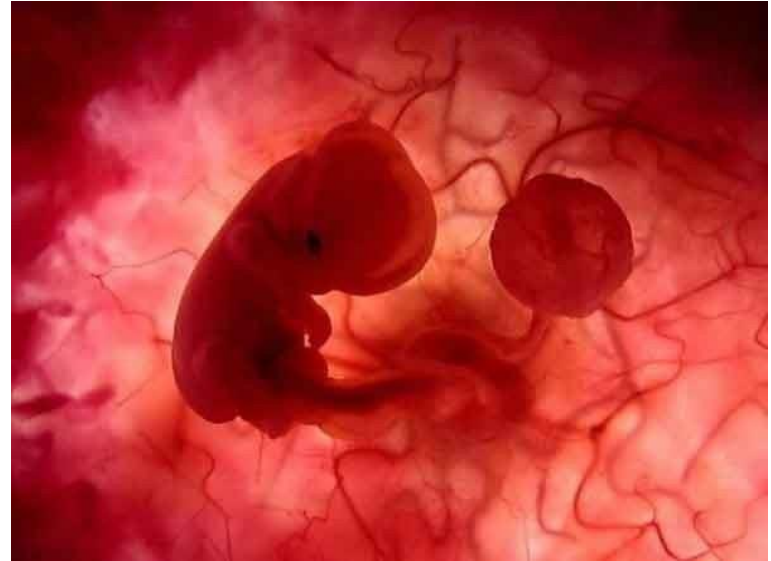
Genital infantilism

- characterized by the underdevelopment (anatomic and histological) of genital organs and hypofunction of ovaries;
- if sexual infantilism combines with general it can be diagnosed in age 13-14; the insufficient expressed of secondary sexual characters (lag from the age-related norm on 1-3) and violation of anthropometric indexes is inherent general infantilism.



Aetiology

- inherited reasons;
- complicated flow of antenatal development;
- hypovitaminosis in a postnatal period;
- child's infections; tonsillitis; rheumatism;
- operations on ovaries.



- infantilism shows up by hypofunction of ovaries, by cyclic changes from the side of endometrium and accompanied by the changes of function of thyroid gland ;
 - in addition, the mionectic sensitiveness of ovaries is marked to the gonadotropic hormones, and organs-targets(uterus, vagina, mammary glands) - to the steroid hormones; thus observed surplus products of FSH and insufficient making of LH;
 - for infantilism the decline of retractive ability of uterus is characteristic, that it is related to the change from the side of estrogen receptors;
 - an important role is played the violation of innervation, and also intraorganic and pelvic hemodynamics.

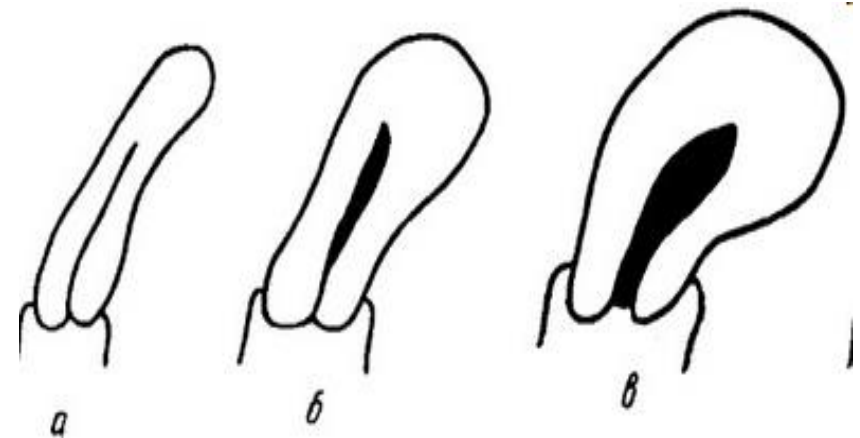
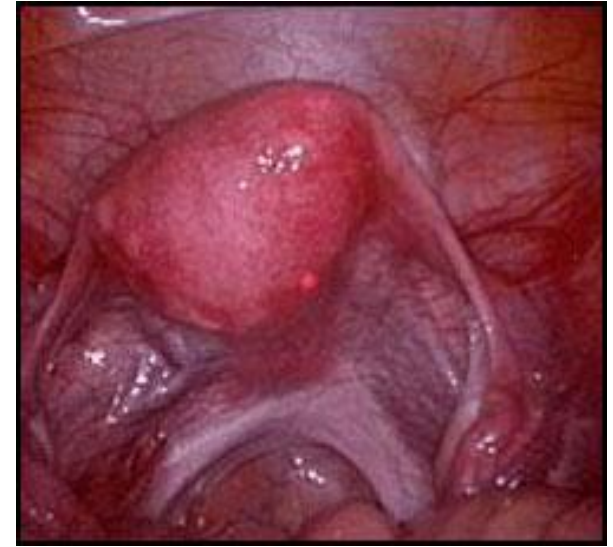
Clinical presentation

- an infantile girl is not high a height, with a thin bone, justomino pelvis, late menarche, hypomenstrual syndrome that quite often combines with algodismenorrhea;
- at genital infantilism there is disparity between a proportional build and exaltation(sometimes expressed) of genital organs.



Clinical presentation

- for determination of expressed of sexual infantilism a considerable role is played by the sizes of uterus that is in a state of hypoplasia;
distinguish three degrees of hypoplasia of uterus :
a) rudimentary;
b) infantile;
c) hypoplastic



- a) a rudimentary uterus has length a from 1 to 3 cm, thus considerable part is occupied by the cervix of the uterus; such variant is observed extremely rarely, anymore close to the anomalies of development of genital organs, what to the exclamation; hypoestrogeny and proof amenorrhea is accompanied; a prognosis in relation to renewal of specific functions of woman organism is unfavorable;
- b) infantile uterus long a more than 3 cm; betweenness by a cervix and body of uterus - 3: 1, as well as for the girl of prepubertate age; at bimanual research find out hyperanteflexio uteruses, weak expressed of vaults of vagina, high placing of ovaries, long and winding salpinxs; menstruations rare, sickly; for renewal of function the protracted therapy is needed;
- c) the length of hypoplastic uterus is 6-7 cm, betweenness by length of cervix and body - 1: 3; formed not only as a result of negative influences in a antenatal period and period of childhood but also as a result of the local inflammatory processes carried in a pubertal period; this pathology can independently disappear after the beginning of sexual life and offensive of pregnancy

Diagnosics

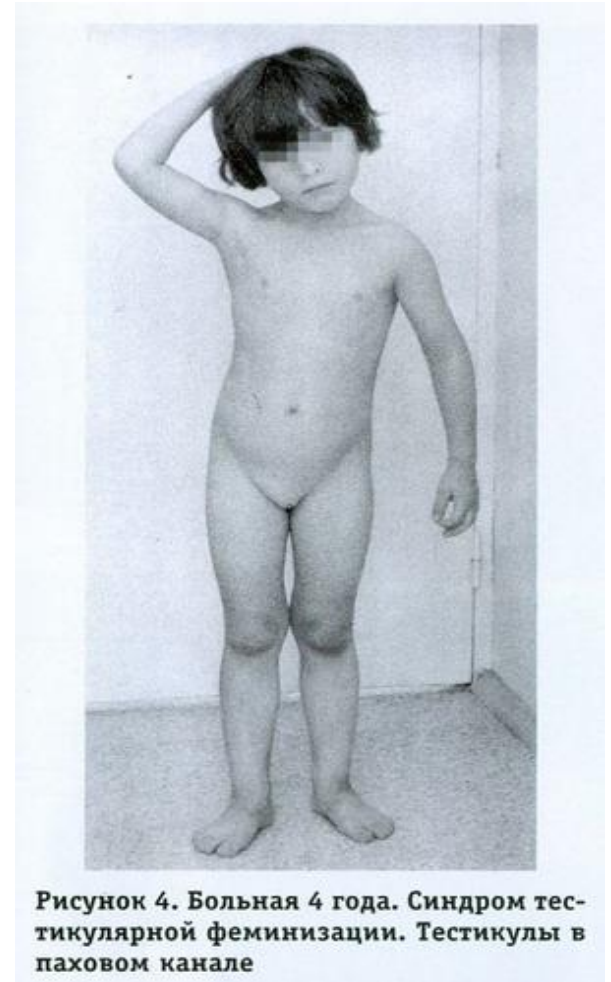
- complaints of patient(late appearance of menstruations, menstrual disorder as a hypomenstrual syndrome,algodimenorrhea);
- data of anamnesis(unfavorable flow of antenatal period, generous amount infectious and other diseases in childhood);
results of objective research (subzero height, formed not enough thorax, excalation of mammary glands, narrowed pelvis, deficit of hair in area of pubic and cavities of arm-pits, hypoplasia of external genital organs;
- Bimanual research (is determined the underdeveloped uterus that is hyperanteflexio) is able;
- additional methods of research (anthropometry, drafting of morphogram, sounding of uterus, sciagraphy,hysterosalpingography, determination of bone age, laboratory researches and others.

Treatment

- removal of reasons of lag of development of genital organs;
- hormonotherapy(cyclic introduction of estrogens and progesterone is to the minimum doses);
- Vitaminotherapy (E,C, B1, B6, ATP);
- physiotherapy(electrophoresis of copper from 5 to 13 day of cycle, zinc from 14 to 24 days, electro-reflexotherapy(acupuncture, electrostimulation of cervix of the uterus), curative gymnastics,balneotherapy).

Gonadal dysgenesis

- Typical form (Turner's syndrome— 45 XO);
- Purest form (Morris syndrome 46 XY : the sex glands - the testes , producing estrogens) ;
 - Mixed form (chromosomal mosaicism)



Shereshevsky-Terner syndrome

- Wing-shaped skin folds on a neck;
- wide thorax;
- valgus deformation of elbow joints;
- sexual glands as bands;
- infantile genital organs;
- absence of secondary sexual characters

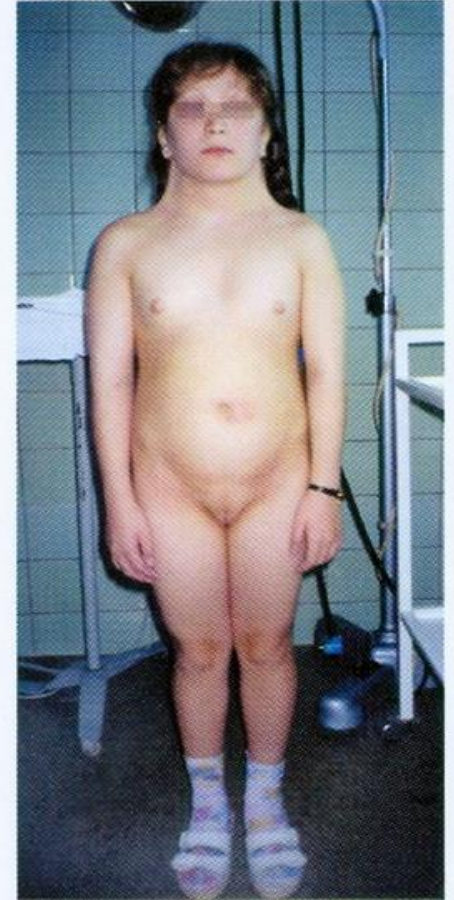


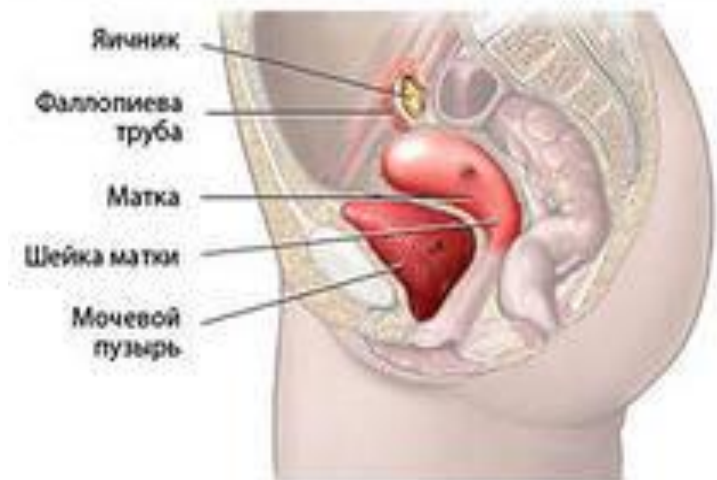
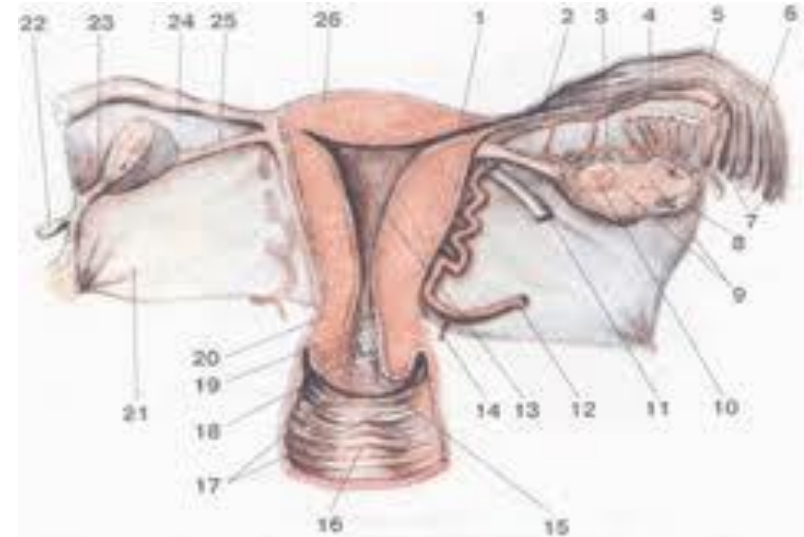
Рисунок 13. Больная 14 лет.
Синдром Шерешевского-Тернера.
Крыловидные складки на
шее "голова сфинкса"

Abnormal position of female
sexual organs



Factors providing normal position of sexual organs

- own tone of genital organs, depending on the level of sexual hormones;
 - concerted activity of diaphragm, front abdominal wall and muscles of pelvic bottom, creating intra-abdominal pressure;
- suspending apparatus of the uterus(round and wide ligaments of uterus, propria ligament of ovaries);
- fixative apparatus of the uterus(sacrum-uterine, cardinal,utero-vesical, vesico -pubic ligaments);
- supporting apparatus(muscles of pelvic bottom).



Determination and etiology

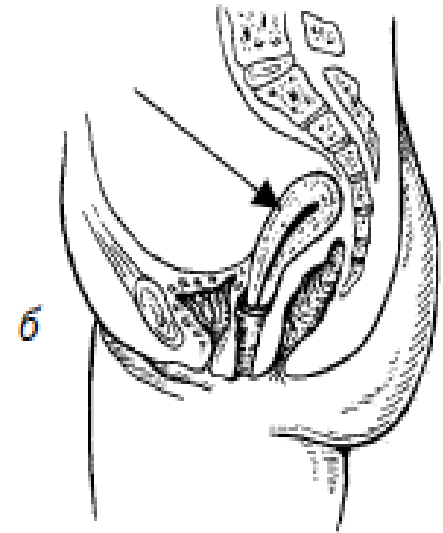
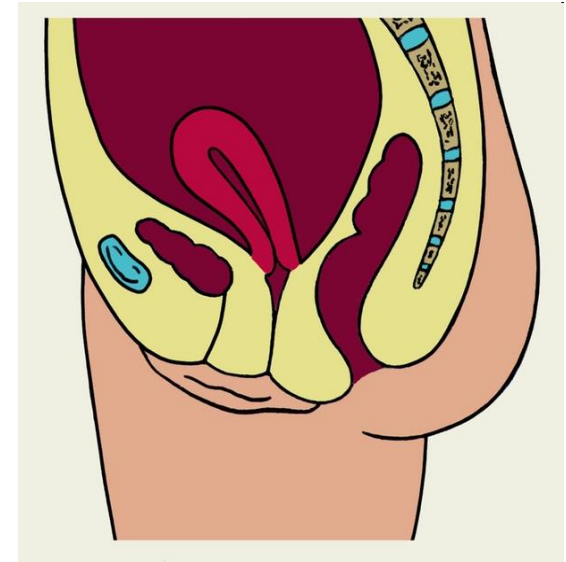
- wrong positions of genital organs is deviations from a norm, carrying persistent character, and also relationships violation by the different anatomic departments of the sexual system;
- reasons of violations of normal location of genitals can be inflammatory processes, tumours, traumas, heavy physical work, pathological labor, infantilism, asthenia, dystrophy.

Classification

- 1. Uterine displacement in the cavity of pelvis :
 - a) in a horizontal plane: displacement anteriorly, posteriorly, to the left, to the right;
 - b) in a vertical plane: raising, descent , prolapsus of uterus(complete and partial);
- 2. Violation of relationships between the different departments of uterus :
 - a) pathological inclinations of uterus anteriorly, posteriorly, to the right, to the left;
 - b) flexio of uterus anteriorly, posteriorly, to the right, to the left;
 - c) turn of uterus;
 - d) torsion of uterus;
 - e) version of uterus.

Uterine displacement in gorizontal plane (positio uteri)

- Antepositio is a metrectopia anteriorly; if it is overcrowded of the rectum is the physiological phenomenon, mainly arises up on a background of development the tumours in the rectal-uterine pouch;
- Retropositio is a metrectopia posteriorly; observed at the overcrowded of urinary bladder, heavy inflammatory processes that cause pulling up of the uterus to the back wall of pelvis, tumours located ahead from an uterus

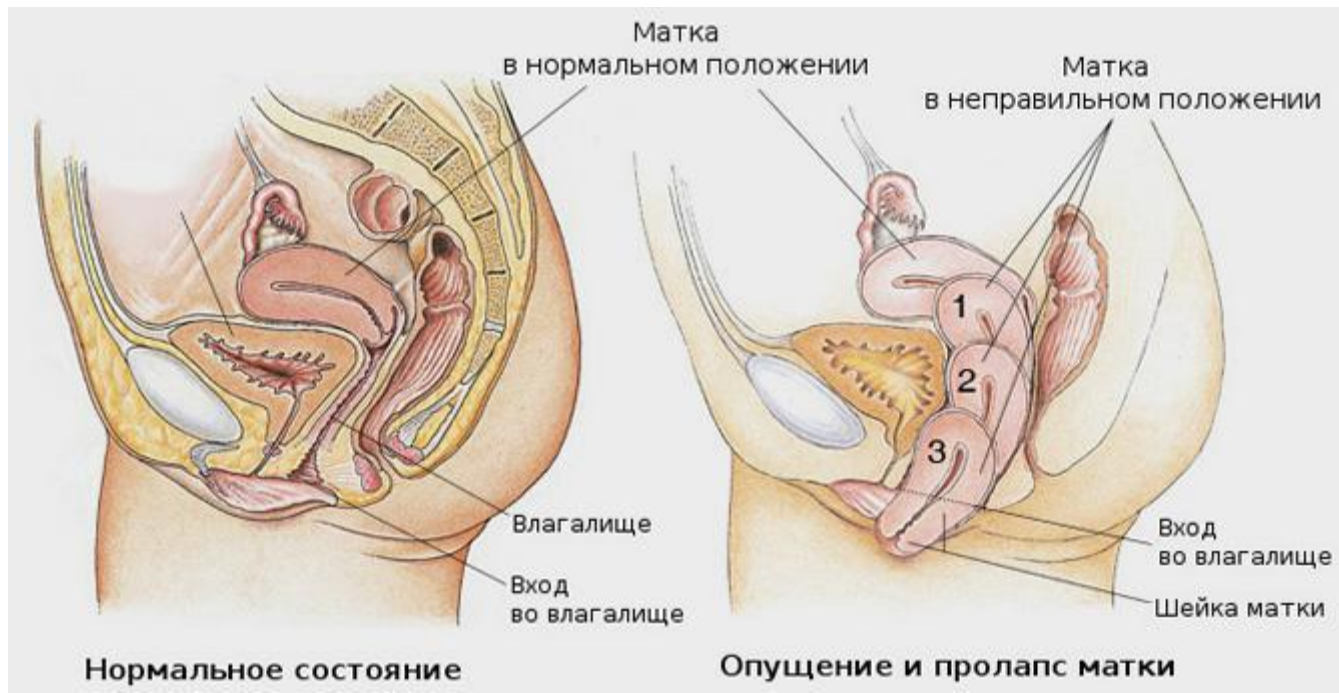


Metrectopia in a vertical plane

- in case of raising of uterus its bottom is situated higher than plane of inlet in a small pelvis, external os of cervix of the uterus - higher than interspine line;
- distinguish the physiological raising of uterus(in child's age, and also on a background overcrowded of urinary bladder and rectum) and pathological(it contingently the tumours of uterus, vagina, rectum, exudate in the rectal-uterine pouch).

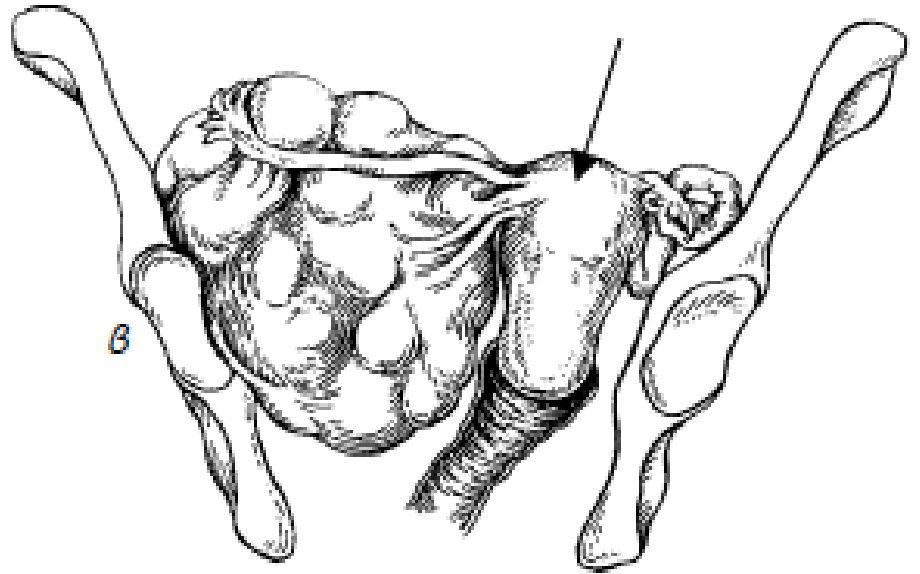
Metrectopia in a vertical plane

- distinguish three degrees of prolapse of uterus :
I is a prolapsus of uterus, at that the cervix of the uterus is situated below than interspine line, but keeps indoors outside a pudendal cleft;
II is an incomplete prolapse of uterus, at that the cervix of the uterus goes out outside a pudendal cleft, and the body of uterus is located in small pelvis;
III is a complete prolapse at that all uterus goes out outside a pudendal cleft together with the walls of vagina.



Metrectopia in gorizontal plane

- Lateroposition is a sidelay of uterus to the right or to the left; mainly conditioned by the presence of inflammatory infiltrates in a parauterine cellular tissuers, tumours of appendages(an uterus is displaced in an opposite side), adhesions process(an uterus is displaced toward localization of joints); it is diagnosed on the basis of bimanual and USE; treatment consists in the removal of reasons that resulted in disposition of uterus.

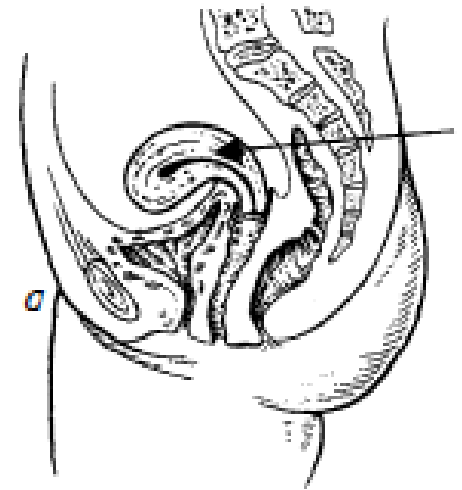


Pathological inclination (versio uteri)

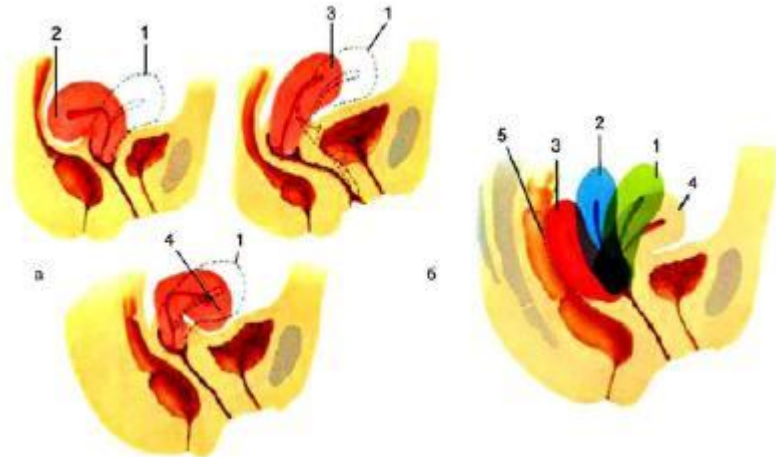
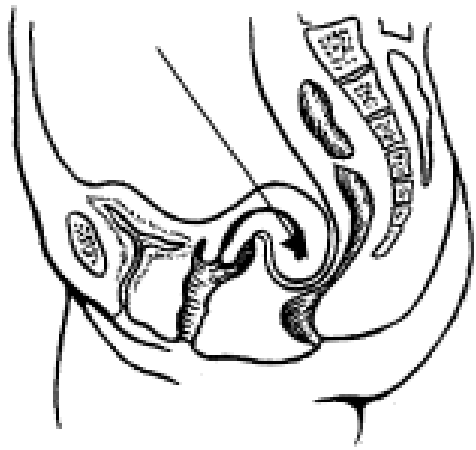
- at pathological inclination the body of uterus is displaced in one side, and cervix- in other, because of the prevailing fixing of cervix by sacrum-uterine and cardinal ligaments at the level of internal os;
- Anteversio is inclination of body of uterus anteriorly, and cervix-posteriorly; at normal position of uterus it insignificant inclination anteriorly is always present; expressed inclination of body of uterus anteriorly, at that a cervix is turned by external os posteriorly and up, testifies to pathological anteversio;-
- Retroversio is inclination of body of uterus posteriorly and down ward, and cervix of the uterus – anteriorly and up;
- Dextroversio is inclination of body of uterus to the right and up, and cervix - to the left and down; at the sinistroversio body of uterus is inclined to the left and up, and cervix of the uterus - to the right and down

Uterine flexio (flexio uteri)

- Hyperanteflexia (pathological anteflexia of uteruses) is a bend of uterus anteriorly, at that an acute angle appears between a body and cervix; it can be investigation of sexual infantilism, inflammatory process in a small pelvis; among clinical displays: pain in a lower abdomen and small of back, violation of menstrual function on the type of hypomenstrual syndrome, algodismenorrhea, primary sterility.



- Retroflexia is characterized by the presence of corner between a body and cervix, open posteriolly;
distinguish movable(at anatomo-physiological violations) and fixed(at presence of unions with a peritoneum because of endometriosis and inflammatory processes in small to the pelvis) retroflexio of uterus;
complaints about pain ina lower abdomen and small of back,hyperpolymenorrhea,algodismenorrhea,constipations, disorders of urination, appearance of whites;
distinguish three degrees of retroflexia :
I is inclination of uterus posteriolly with maintenance of anteflexio;
II is inclination of uterus posteriolly without maintenance of anteflexio;
III is inclination of uterus posteriolly at presence of angle between a body and cervix of the uterus of the expressed corner open



Turn, torsion inversion of uterus

- the turn of body of uterus together with a cervix against longitudinal axis accompanies inflammation of sacrum-uterine ligaments, their shortening, tumour processes located behind and from one side from an uterus, adhesive process;
- Torsion of uterus is a turn of uterus at an immobile cervix, arises up on a background the one-sided tumour of appendages or subserouse located leiomyomatous node;
- Inversion of uterus - a mucous membrane is extrorse, and serosal - inward; distinguish two forms of inversion: puerperal (bind to the wrong conduct of labour) and oncogenetic (arises up at a giving birth fibroid with short and wide crus)