

Congenital defects of the urinary system

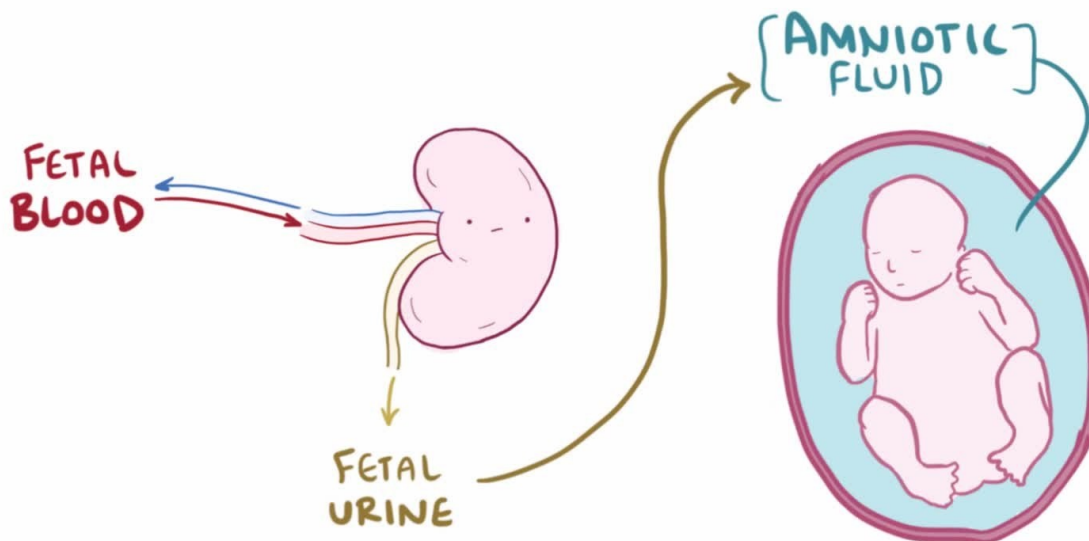
Congenital developmental defects of the kidneys

- relatively frequent congenital developmental defects;
- hypoplasia, dysplasia, cystic kidney diseases;
- often asymptomatic, sometimes prone to urinary tract infection;
- secondarily they can lead to the formation of a wrinkled kidney.

The Potter sequence

- malformation characterized by hypertelorism, broad nasal root, epicantha, low-set auricles, hypognathia and lung hypoplasia due to oligo- to anhydramnios in developmental defects of the kidneys.^[1]

Renal agenesis and hypoplasia



Agenesis of the kidney

- disorder in the development of primitive drainage tracts of the urinary tract and metanephrogenic blastema;
- **bilateral** renal agenesis is not postnatally compatible with life (incidence 1:4000);
- **unilateral** kidney agenesis
 - 1:1000; more often in boys; more often the left kidney is absent; sometimes associated with other VVV;
 - compensatory hypertrophy of a solitary kidney;
 - asymptomatic.

Kidney hypoplasia

- reduction of kidney parenchyma below 50% of normal or below 30% of normal in both kidneys;
- **simple form**: unilateral or bilateral reduction in the number of normally established nephrons; clinically asymptomatic;
- **oligomeganephronia**: significant reduction in the number of nephrons that are hypertrophic; the kidneys are reduced in size and have an irregular surface; leads to progressive chronic renal insufficiency;
- **segmental hypoplasia**: hypoplasia limited to one kidney segment; a typical finding is arterial hypertension.^[1]

Cystic kidney disease

Cystic kidneys

- AR hereditary (ARPKD) and AD hereditary (ADPKD) polycystic kidney disease;
- **ARPKD** (1:20,000)
 - only the collecting ducts of the kidneys are dilated; proliferation and dilatation of intrahepatic and later extrahepatic bile ducts

- severe forms – oligohydramnios and lung hypoplasia, Potter's sequence; resistance can be felt in the abdominal cavity; arterial hypertension; chronic renal failure with anemia and growth failure; liver fibrosis.
- **ADPKD** is the most common monogenic hereditary nephropathy (1:1000)
 - formation of cysts in all parts of the nephron; often also cysts in the liver parenchyma;
 - clinical manifestation often only in adulthood.

Multicystic dysplasia

- early embryonic kidney development disorder, usually not hereditary;
- usually only one kidney is affected;
- the most common VVV of the kidneys; more often in boys;
- compensatory hypertrophy of the second kidney to a functionally solitary kidney;
- back pain, abdominal tumor, vomiting, hematuria, arterial hypertension, recurrent urinary infections.^[1]

Anomalies of location and fusion of renal parenchyma

- disturbance of the physiological rotation and migration of the renal parenchyma during the fetal period;
- diagnosis: urine examination, ultrasound, i.v. excretory urography (IVA), voiding cystourethrography (MCUG), renal scintigraphy.

Double kidney (ren et ureter duplex)

- Weigert-Meyer rule: the ureter draining the upper pelvis opens into the bladder medially and caudally from the ureter draining the lower pelvis – the ureters cross;
- mostly asymptomatic, rarely leads to recurrent urinary infections; sometimes associated with VUR, ureterocele or other VVV.

Horseshoe kidney (ren arcuatus)

- connection of the parenchyma of both kidneys in front of the abdominal aorta - most often the connection of the lower poles of the kidneys;
- mostly asymptomatic, rarely leads to recurrent urinary infections, VUR, renal pelvis obstruction or lithiasis.

Ectopic kidney

- arrest of the displacement of the kidney in the cranial direction during the fetal period;
- the kidney is located in a small pelvis, most often next to the common iliac artery;
- mostly asymptomatic, rarely leads to recurrent urinary tract infections or VUR.^[1]

Hyperplastic and huge kidney

Congenital defects of the urinary tract

Congenital obstructive defects of the renal pelvis and congenital defects of the ureter

Congenital hydronephrosis

- outflow disorder from the hollow system of cups and pans due to the narrowing of other sections of the outlet system → expansion of the hollow system of the urinary tract and the risk of impaired renal function;
- if there is an obstruction in the area of the ureterovesical transition, dilation of the ureter and the proximal hollow system occurs = megaureter;
- this is usually a congenital defect established already in the prenatal period;
- more than half of prenatal findings normalize during later pregnancy or the perinatal period;
- primary: obstruction in the pyeloureteral transition (congenital: fibrous tissue, aberrant vessel, etc., acquired: concretion, blood coagulum);
- secondary: obstruction below the pyeloureteral transition – e.g. dolichomegaureter or posterior urethral valve in boys;
- obstructive: mechanical obstacle;
- non-obstructive: peristalsis disorder;
- urinary outflow disorder → risk of urinary tract infection – acute pyelonephritis, urosepsis;
- unilateral hydronephrosis – normal blood pressure and normal renal function;
- sometimes asymptomatic; often failure to thrive, recurrent urinary infections, hematuria, abdominal pain, urolithiasis;
- dilatation without obstruction – conservative procedure;
- obstruction – surgical treatment.^[2]

Atresia and stenosis of the ureter

Congenital mega(lo)ureter

- significant enlargement of the ureter;
- primary: based on congenital obstruction at the transition point of the ureter into the bladder;
- enlargement of the ureter and the hollow system of the kidney = ureterohydronephrosis;
- secondary: due to vesicoureteral reflux (VUR) or obstruction of the outflow part of the bladder (eg posterior

urethral valve in boys);^[2]

Congenital ureterocele

Agensis of the ureter

Duplication of the ureter

Position of the ureter

Congenital vesico-uretero-renal reflux (VUR)

- backflow of urine from the bladder into the ureter or the hollow space of the kidney in case of failure of the valve (antireflux) mechanism at the level of the ureteral ostium in the bladder (shortened intramural section, ectopic ureteral ostium);
- occurs quite often in children; genetic background;
- consequences: dilatation of the hollow system, risk of urinary tract infection and post-infectious scars in the kidney parenchyma (deterioration of renal functions, development of hypertension), functional disorders of bladder emptying;^[2]
- VUR grades according to MCUG:
 - I. = VUR to ureter;
 - II. = VUR to the ureter and to the calicopelvic system;
 - III. = II + dilatation of the calicopelvic system;
 - IV. = III + pressure atrophy of the parenchyma;
 - V. = massive VUR with extensive destruction of renal parenchyma.^[3]
- conservative solution (VUR I-III): sterile VUR is not too risky for the kidney → long-term chemoprophylaxis with the aim of preventing the formation of post-infectious scars of the kidney parenchyma; postnatally, the antireflux mechanism matures and VUR tends to resolve;
- operative solution (VUR IV-V): extension of the intramural section of the ureter and ureterocystoneostomy (reimplantation of the ureter);
- endoscopy: cystoscopically, an inert material is applied to the ostium to create an antireflux mechanism.^[2]

Other congenital defects of the urinary system

Epispadias

Hypospadia

Bladder exstrophy

Posterior urethral valve

- the most severe congenital disorder of urine outflow – already prenatally leads to damage to the kidney parenchyma and glomerular filtration disorder;
- cystostomy is necessary immediately after delivery; later, the valve is surgically removed;
- only in boys; the long-term prognosis is not favorable - chronic renal failure develops with the need for dialysis or a kidney transplant.^[2]

Other atresia and stenosis of the urethra and bladder neck

Defects of the brain

- urachu cyst, persistence, hernia

Congenital absence of bladder and urethra

Congenital bladder diverticulum

Links

Related Articles

- Development of the genitourinary system • Examination of the child's uropoietic system

References

1. MUNTAU, Ania Carolina. *Pediatrics*. 4. edition. Grada, 2009. pp. 435-438. ISBN 978-80-247-2525-3.
2. LEBL, J – JANDA, J – POHUNEK, P. *Clinical Pediatrics*. 1. edition. Galen, 2012. 698 pp. pp. 599-603. ISBN 978-80-7262-772-1.
3. MUNTAU, Ania Carolina. *Pediatrics*. 4. edition. Grada, 2009. pp. 442. ISBN 978-80-247-2525-3.

Template:Stump