

## RENAL AGENESIS - PAST AND FUTURE

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### ABSTRACT

**Congenital urinary tract anomalies (CUTAs) are common among children and represent approximately 30% of all the prenatally diagnosed malformations. They are the primary reason for a chronic renal failure. Most anomalies are manifested by urinary tract infections, changes of urine colour, abdominal pain while some malformations are asymptomatic. A particular anomaly such as renal agenesis is of interest for pediatric nephrologists. In the past, it has not troubled the patients. Nowadays it is accepted that this anomaly relates with a hidden risk for the development of kidney failure and thus requires a special attention. In the present survey, some issues of the epidemiology, diagnosis and treatment of CUTA are considered.**

**Key words:** *renal agenesis, childhood, epidemiology, diagnosis, treatment*

Congenital urinary tract anomalies (CUTAs) are common in childhood. They are divided into malformations of the kidney, pyeloureteral segment, ureters, uretero-vesical segment and urinary bladder. Renal anomalies represent 3% of all the malformation uropathies (20,23). Clinical practice proves that their number is greater than that of the diagnosed cases as some of them are not manifested by complaints and are occasionally detected during certain examination of the patient. Here belongs renal agenesis that can be either unilateral, or bilateral. It is characterized by unilateral or bilateral absence of renal structure and blood supply (14,20).

Bilateral renal agenesis occurs in one of 4000 births (3,15,20). There is a severe oligohydramnios, birth weight is usually below 2500 g, and there is no

nitrogen retention. Sometimes there are still-births, or kidney failure develops already in the first days after delivery. The diagnosis can be done by antenatal echography after the 17<sup>th</sup>-20<sup>th</sup> gestational week or by magnetic resonance imaging (MRI). It represents an indication for pregnancy interruption (8,11,16,19).

Unilateral renal agenesis occurs in one of 1000 births (3,15,17,20). It is characterized by complete absence of renal parenchyma in the corresponding side of the organism. In the embryo, there are neither nephroblastic cells, nor blood circulation in them that causes the necrosis of these cells. Sometimes at the site of absent kidney there is accumulation of adipose sclerotic tissue and an obliterated ureter. In case of unilateral renal agenesis, there is a working hypertrophy of the contralateral kidney (20,23). According to some hypotheses, in infants with a single functioning kidney, certain compensatory mechanisms emerge already in the intrauterine period such as a greater number of nephrons. The diagnosis is done accidentally during the examination of the patient on the occasion of another disease of the single kidney. Following the introduction of the antenatal echography for pregnant women in the course of pregnancy monitoring, the foetus is purposefully examined in terms of the congenital anomalies. The diagnosis of

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renal agenesis can be done during the antenatal period, the so-called 'empty fossa symptom' (1,11,19).

Postnatal ultrasonic examination provides information about the unilateral renal agenesis. Echographically proved kidney absence at the usual site, either dystopic, or ectopic, along with the presence of a contralateral kidney of a larger size testifies in favour of renal agenesis (5,6,8). Scintigraphy, renovasography, MRI and miction cystography belong to the modern diagnostic methods used in the clinical practice in order to confirm the renal agenesis and reveal the alterations of the single kidney. The ultrasound examination helps in excluding the polycystic renal dysplasia. Such a kidney does not exhibit any excretory function and its echographic image represents a multilocular abdominal mass containing thin-wall cysts surrounded by a denser matter and not communicating one with another (5,8). This pathology is usually combined with a congenital ureter atresia.

Genital, skeletal and gastrointestinal anomalies are more common among the children with a single kidney (2,9,11). Such a pathology could be due to a series of syndromes of congenital malformations such as VACTER association (combination of malformations of vertebrae, anus, heart and kidney as well as tracheoesophageal fistula), Mayer-Rokitansky syndrome (subtotal or partial symmetrical aplasia of both Müllerian canals along with a congenital absence of vagina and uterus).

Urinary tract anomalies co-occur more commonly in children with renal agenesis (3,9,20). The investigation of 46 patients with renal agenesis demonstrates the presence of vesicoureteral reflux and obstruction of the pyeloureteral segment in 48% of the cases. According to numerous authors, the most common association is that of renal agenesis and vesicoureteral reflux (9,20).

The rising incidence rate of the chronic kidney disease not only in adults but also in children results in a better management of the children with proved CUTA (14,21). This has changed the behaviour towards the children with renal agenesis, too (4,10).

The concept that nothing takes place in cases with a single kidney remains far in the past (4).

Early recognition and treatment of urinary tract anomalies in the patients with a single kid-

ney reduces the long-term risk of kidney damage (4). One should regularly follow-up the urine status of the children with proved renal agenesis in terms of microalbuminuria, glomerular filtration velocity (GFV) and arterial pressure (13,18,22,24).

The children who are not liable to further examinations should meet the following criteria: normal urine analysis (absent proteinuria), normal GFV and normal arterial blood pressure values.

The analysis of the results from the observation of the patients with renal agenesis enables the elaboration of the following practical approach:

- in case the patient does not exhibit any aberrations of the laboratory parameters he should be followed-up every two years up to the puberty and thereafter every three-five years;
- in case of changes of the urine status, GFV and arterial pressure, the patient should be followed-up every year. This approach underlies the early detection of the patients with a chronic kidney disease and the prevention of this disease as well.

The prognosis of the children with renal agenesis is determined by the following factors:

- ❖ presence of unilateral or bilateral renal agenesis;
- ❖ presence or absence of association with other urinary tract anomalies;
- ❖ incorporation of renal agenesis into a series of congenital syndromes;
- ❖ correct follow-up of the children born with a single kidney without any other malformations and syndromes, and
- ❖ normal values of the arterial blood pressure and GFV as well as absence of microalbuminuria.

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