CLINICAL PRESENTATION OF CONGENITAL URINARY TRACT ANOMALIES IN CHILDHOOD

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ABSTRACT

Congenital urinary tract anomalies (CUTAs) are common pathology of the urinary tract in childhood. They present with a variety of symptoms in most patients. Some CUTAs can be diagnosed as an accidental finding during the ultrasound examination on another occasion. CUTAs can accompany other genetic syndromes in children, too. The significance of CUTAs is determined by the complications caused by them. They underlie the urinary tract infections and nephrolithiasis. CUTAs can lead to a chronic renal disease in a significant percentage of cases. Therefore, they require a sufficient physician’s knowledge about their clinical presentation, appropriate diagnostic approach and timely prevention of complications. In the present brief survey, some features of the most common CUTAs in children are presented.

Key words: congenital urinary tract anomalies, symptoms, diagnosis, complications, childhood

Congenital urinary tract anomalies (CUTAs) are common pathology of the urinary tract in childhood (1,17). One could find information about them in Aristotle’s works, Leonardo da Vinci’s paintings and Eustachian’s anatomical descriptions. So, one can establish that this issue is topical and significant even today. Knowledge of the anatomy and embryology of the urinary tract provides responses to a series of questions, however, numerous issues remain unsolved yet. The division of CUTAs into syndrome-related and not syndrome-related, the investigations of the genes involved in their pathogenesis and the expansion of the scope of image examinations represent a true clinical interest, indeed.

Antenatal ultrasonic examination underlies the proof of some CUTAs (10,14). Knowledge of the postnatal clinical presentations of CUTAs enables the correct diagnosis and the selection of the appropriate therapeutic approach (1,12,15,16,21).

The following pathology can be visualized after the 17th gestational week:

❖ number and size of the kidneys. According to some authors, the empty fossa in the lumbar region is a sign of renal agenesis.
❖ pyelon, ureters and urinary bladder. The different degrees of drainage disturbances are visualized.
❖ oligohydramnios. Its presence suggests the search for CUTA.
❖ anomalies of other organs and systems.

The most common clinical symptoms of CUTA are the following: abdominal pain as anxiety is its equivalent in sucklings and infants, manifestations of urinary tract infections, arterial hypertension as well as diurnal and nocturnal enuresis in children. The clinical examination of the patient can detect enlarged kidneys as well as positive succusio renalis. The single CUTAs present with a different spectrum of clinical manifestations. Sometimes a given CUTA could remain unrecognized at all. This could delay the proper diagnosis of the disease and...
its complications. The evidence of familial history in the patients with some CUTAs such as renal polycystic disease requires the screening examination of these families in order to enable the early diagnosis and timely prevention of these diseases.

**CLINICO-DIAGNOSTIC ANALYSIS OF CUTA**

**Renal agenesis**

Recently, new information about it has accumulated that defines the approach to this apparently harmless anomaly. In the past, one has assumed that the patients with a single kidney do not necessitate any specific treatment at all. Recent observations of the patients with renal agenesis indicate that with the purpose of preventing the chronic renal disease, arterial blood pressure should be monitored and urinary tests for microalbuminuria should regularly be performed.

**Renal hypoplasia**

In dependence on the histological variations of this CUTA, clinical spectrum can vary between absent complaints and present arterial hypertension, on the one hand, and manifestations of a chronic renal disease, on the other hand.

**Nephroptosis**

There is an outlined mobility of the kidneys because of the poor binding apparatus and the long pedicle. This leads to usual complaints of chronic relapsing abdominal pain. Sometimes, drainage disturbances accompanied by common infections can occur, too. Besides, periodic proteinuria and haematuria can be observed.

**Renal malrotation**

The pathology is due to disturbed intrauterine rotation of the kidney. Clinically, it presents with abdominal pain and urinary tract infections because of the drainage disorders.

**Renal dystopia**

The disturbances of the migration of the kidneys lead to their pathological location, most commonly, in the pelvic region. In many cases, this is related to drainage disorders resulting in common urinary tract infections. The children often complain of abdominal pain that is of chronic relapsing nature.

**Horseshoe-shaped kidney**

This CUTA is associated with disturbed kidney interaction. Most often, both kidneys are coalesced through their upper poles. Usually, abdominal pain and relapsing urinary tract infections can be observed. The children at older age can present with manifestations of nephrolithiasis and arterial hypertension as well.

**Solitary renal cyst**

Usually, the solitary renal cyst does not provoke any complaints. In case of a large cyst, the child complains of abdominal pain.

**Polycystic renal disease**

Polycystic renal disease of adult type manifests itself later on in childhood. The children complain of abdominal pain as a manifestation of nephrolithiasis and arterial hypertension. In some cases, common urinary tract infections are observed.

**Congenital hydronephrosis**

In dependence on the degree of hydronephrosis, common urinary tract infections are observed. Urine colour is often changed, too. Abdominal pain occurs in cases of high-grade hydronephrosis.

**Fraley’s syndrome**

The clinical manifestations of Fraley’s syndrome consist of common infections, abdominal pain as well as of evidenced micro- and macrohematuria.

**Congenital megaureter**

This CUTA consists in upper urinary tract obstruction. Its course is characterized by frequent urinary tract infections, sometimes accompanied by abdominal pain.

**Ureterocele**

Ureterocele represents a cystic dilatation of the terminal part of the ureter. It is located in the lumen of the urinary bladder and is accompanied by stenosis of the mucous part of the ureteral ostium. Clinically, it presents with common urinary tract infections, urine retention, and urinary tract obstruction.

**Vesicoureteral reflux**

Vesicoureteral reflux is one of the most common CUTAs in childhood. Its clinical manifestations consist of common urinary tract infections and postmiction pain in the pelvic region. Although in recent years the term of ‘reflux nephropathy’
has been revised, it is nowadays accepted that the vesicoureteral reflux represents a CUTA causing the development of a chronic kidney disease.

Some other anomalies of the lower ureteral segment such as stenosis of the neck of the urinary bladder, posterior urethral valvae and urethral stenosis cause a subvesical obstruction and thus lead to the development of a megaureter and vesicoureteral reflux. They are clinically manifested by miction disorders and frequent urinary tract infections.

Table 1 shows the constellation of clinical manifestations of CUTAs in infants and children.

<table>
<thead>
<tr>
<th>Abdominal pain</th>
<th>Urinary tract infections</th>
<th>Hematuriae</th>
<th>Arterial hypertension</th>
<th>Enuresis</th>
<th>Acute renal failure</th>
<th>Chronic renal failure</th>
</tr>
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<tbody>
<tr>
<td>Hydronephrosis</td>
<td>Hydronephrosis</td>
<td>Hydronephrosis</td>
<td>Hypoplasia with dysplasia</td>
<td>Vescicoureteral reflux</td>
<td>Obstructive uropathies in newborns and infants</td>
<td>Hydronephrosis</td>
</tr>
<tr>
<td>Horseshoe-shaped kidney</td>
<td>Vesicoureteral reflux</td>
<td>Renal polycystic disease</td>
<td>Renal polycystic disease</td>
<td>Posterior urethral valvae</td>
<td></td>
<td>Renal polycystic disease</td>
</tr>
<tr>
<td>Dystopia Syndrome</td>
<td>Congenital megaureter</td>
<td>Horseshoe-shaped kidney</td>
<td>Unilateral cystic aplasia</td>
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<td>Obstructive uropathies</td>
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<tr>
<td>Renal polycystic disease</td>
<td>Double drainage system</td>
<td></td>
<td>Renal agenesis</td>
<td></td>
<td>Vescicoureteral reflux</td>
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The issue of CUTAs is a challenge to the general practitioner, pediatric nephrologist and urologist.

Comprehensive knowledge of the clinical manifestation and variety of symptoms of a given CUTA shortens the way from the symptom to the diagnosis and ensures fewer complications for the patient (2-4,6-9,11,13,22). This enables the elaboration of a diagnostic algorithm consisting of a set of paraclinical and image examinations. The ultrasound examination plays a crucial diagnostic role in the patients with abdominal pain, urinary tract infections, enuresis, arterial hypertension, and hematuria (5,18-20).

In conclusion, the analysis of the problem enables the pediatric nephrologist to answer the following important questions concerning CUTAs in childhood:

What do we know? We can reveal the disturbances of the renal anatomy and embryology as well as CUTAs type and clinical manifestations.

What can we do? We can elaborate a correct individualized diagnostic and therapeutic approach. We can apply the echographic examination in the clinical practice as a primary means for the precise diagnosis of CUTAs. We can apply the renal scintigraphy with the purpose of detecting the discriminative function of the kidneys and the degree of obstruction as well as of verifying the scars.

What do we hope for? We should cover all the pregnant women and implement the antenatal ultrasound examination with the purpose of warranting the early diagnosis of CUTAs. In this way, a successful postnatal prevention of the chronic kidney disease could be achieved.

REFERENCES
Clinical presentation of congenital urinary tract anomalies in childhood


