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Editorial

A holistic approach to CAKUT (Congenital Anomalies of the Kidney and Urinary Tract)

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"Structure does not determine Function or vice versa, but both are simply different ways of regarding and describing the same thing."

Jean R. Oliver, *Nephrons and Kidneys*, 1968

Keywords

CAKUT (Congenital Anomalies of the Kidney and Urinary Tract), urinary tract infection, chronic kidney disease, follow-up.

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The urinary tract malformations, today called CAKUT (Congenital Anomalies of the Kidney and Urinary Tract), include a range of morphological and/or functional abnormalities of the kidney and urinary tract diseases, apart from being the main cause of renal failure in children. The frequency of these birth defects is around 2% of pregnancies and many genetic syndromes may include CAKUT: about 500 of them have been described [1]. The CAKUT concern localized alterations at different levels of the urinary system: hydronephrosis, vesicoureteral reflux, duplex collecting system, megaureter, kidney dysplasia. Almost all infants and children with CAKUT have a higher chance to develop a urinary tract infection (UTI), compared to healthy peers, but most children with CAKUT are in good health.

From several case studies it is shown that today the treatment of this group of diseases is mostly conservative and in a constant attempt to preserve the renal function, proposing surgery for cases of severe functional impairment and in patients with recurring UTIs. There is a correlation between CAKUT and chronic kidney disease (CKD). The occurrence of recurrent UTIs in presence of such morphofunctional anomalies can determine CDK. In fact, according to the data of dialysis registers, almost 40% of patients with CKD are suffering from CAKUT. On the contrary, it is demonstrated that the probability that recurrent UTIs can cause isolated CKD is very low.

Prenatal diagnosis with ultrasound of CAKUT has had an important role to improve the natural history of these diseases. It is difficult though to establish prognosis in the prenatal period. The degree of dilatation of the urinary tract does not always correlate with prognosis. The dilatation of the urinary tract is a dynamic process, which not only can vary over time, but is also influenced by other factors.

Every child who suffers from CAKUT was born with a different set of nephrons: our duty as paediatricians is to preserve as long as possible the function, avoiding a worse dysplasia caused by obstruction and recurrent UTIs [2]. On the other hand, prenatal diagnosis of pyelectasis should be interpreted as a marker, not as diagnosis of disease. In our opinion, it is always important to evaluate the possible presence of a visible ureter during the foetal ultrasound and to confirm its presence after birth [3]. Foetal surgery has not achieved the results conjectured in the past, so that the uterine decompression is no longer considered a valid alternative to surgical neonatal correction. Considering the risks related to premature birth, there are no indications to anticipate the date of delivery of foetuses with CAKUT.

After birth, actually, almost all children with CAKUT are in good health. The most sensitive, but fortunately rare, cases are males affected by posterior urethral valves (PUV), which may present acute renal failure at birth. Several authors have proposed recommendations on diagnosis and treatment of CAKUT after birth, suggesting the introduction of antibiotic prophylaxis associated with instrumental investigations such as ultrasound, micturating cystourethrogram (MCUG) and renal scintigraphy [4].

Since several years, following the intuition of Philip Ransley of Great Ormond Street Hospital in London in the ‘90s, the therapeutic approach to CAKUT, when possible, has been conservative. Nevertheless, worldwide these children are still going into surgery without following unique or rational criteria and without proving later if the strategy has been helpful for the patient.

Practical criteria that can make us decide for surgery are: recurrent UTIs, especially in children under 1 year of age, children with VUR of high bilateral grade, worsening hydronephrosis > 3 cm or with noticeable changing during the follow-up, bilateral dilatation and pain symptoms. Surgical correction involves preferably endoscopic, laparoscopic, robotic approach [5].

In children with CAKUT it is necessary to prolong the follow-up after adolescence. Unfortunately, however, a proper transitional care between paediatricians and adult nephrologists/urologists is often missing. Anatomical and functional consolidated alterations could be misinterpreted and bring to a consequent proposal of late surgery with no benefit for the patient.

In conclusion, in the approach to the child with CAKUT it is important to identify a population at risk, distinguishing between CAKUT with and without clinical significance, applying a tailored approach to the individual patient, avoiding unnecessary investigations and treatments.

Declaration of interest

The Author declares that there is no conflict of interest.

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