**CAKUT Information for Parents**

WHAT IS CAKUT?

The term Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) refers to a wide range of structural abnormalities resulting from a perturbation in the embryonic development of the kidney and urinary tract. CAKUT constitute~20–30% of all congenital malformations, and their prevalence has been estimated to range between 3 and 6 per 1000 births.

Congenital kidney malformations are defined macroscopically by changes in kidney size, shape, position, or number, or microscopically by a reduced number of nephrons and/or abnormal histology.

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| **Kidney number** | Renal agenesis | Unilateral or bilateral, kidney and outflow system fail to form |
| **Kidney size and morphology** | Renal hypoplasia | Unilateral or bilateral, kidney shape is normal, but smaller in size and reduced number of nephrons |
|  | Renal dysplasia | Unilateral or bilateral, kidney shape and tissue differentiation is abnormal, reduced number of nephrons |
|  | Multicystic dysplastic kidney | Multiple cysts within a dysplastic kidney giving it an abnormal shape |
| **Kidney position** | Horseshoe kidney | Kidneys are fused posteriorly forming a horseshoe shape |
|  | Ectopic/pelvic kidney | Kidney in an abnormal location, typically pelvic |
| **Outflow abnormalities** | Ureteropelvic junction obstruction | Unilateral or bilateral, junction between kidney and ureter is obstructed, preventing drainage of urine from pelvis of the kidney |
|  | Vesicoureteric reflux | Unilateral or bilateral, junction between ureter and bladder is defective, resulting in urine backflow from bladder |
|  | Duplex collecting system | Unilateral or bilateral, duplication of ureter and kidney pelvis, can be accompanied with duplicated kidneys; outflow system may reflux or exhibit obstruction |
|  | Megaureter | Unilateral or bilateral, distension of ureter resulting in defects in impaired urine Flow |
|  | Posterior urethral valves | Membrane that forms in urethra preventing emptying of bladder, limited to males |

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CAUSES

Causes of CAKUT is complex and include some genetics and enviromental factors. Risks factors associated with CAKUT include folic acid use, low folate, vitamin A deficiency, maternal obesity, maternal diabetes, maternal malnutrition, maternal use of cocaine and/or alcohol, Angiotensin-converting enzyme inhibitors and angiotensin receptor blockers, in vitro fertilization, parents consanguinity.

DIAGNOSIS

Most cases of CAKUT are diagnosed from antenatal ultrasound imaging that correctly diagnose CAKUT in 60%–85% of infants, especially if imaging is performed in the third trimester. The remaining cases of CAKUT are mostly diagnosed after an infant or child presents with a urinary tract infection prompting ultrasound and/or other imaging studies to examine the kidneys and outflow tracts.

TREATMENT

Care of the child will be determined by the nature and severity of the disorder: the spectrum of CAKUT is very broad. Because of the increased risk for progressive CKD in the patient with low nephron endowment, all infants with low birth weight, preterm birth, and/or intrauterine growth restriction should have regular blood pressure measurement and periodic urinalysis.

Periodic ultrasonography should be considered in all patients with CAKUT to document normal and/or compensatory kidney growth, as well as urinalysis and plasma creatinine concentration. Patients with lower urinary tract anomalies may require monitoring for urinary tract infection.

 Avoidance of nephrotoxic drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs), is important—especially in states of volume depletion.

For patients with reduced kidney function, there is increased risk for acceleration of CKD with the onset of the adolescent growth spurt (Figure 1), and episodes of acute kidney injury can accelerate even mild CKD in the patient with CAKUT.

OUTCOMES

CAKUT are the predominant disease group in children, and an important cause of chronic kidney disease (CKD) in children and adolescents.

Distinguishing the upper urinary tract (kidneys and ureters) from the lower tract (bladder and urethra) and unilateral vs bilateral kidney anomalies are importants determinants of the clinical outcomes.

Infants and children with unilateral CAKUT and a normal-appearing contralateral kidney and no evidence of hydronephrosis on renal ultrasound do not require further initial evaluation. Intermittent follow-up is directed to determine normal growth of the contralateral kidney, absence of proteinuria, normal blood pressure, and a normal serum creatinine.

Children with bilateral hypoplasia or those with evidence dysplasia should be offered renoprotective advise and be monitored for evolving renal injury—hypertension, proteinuria, and declining GFR.

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