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Editorial on Challenge of Congenital Abnormalities of the Kidney and Urinary Tract

Madison Lyon
Department of Pediatric Surgery, Children Medicak Center, Tehran Univerasity of Medical Sciences Iran

An increasing recognition of congenital abnormalities of the kidney and urinary tract (CAKUT) has led to challenges in their diagnostic evaluation and management. Antenatal ultrasonography is now widely available, and experts in fetal medicine can suspect and detect the presence of various anomalies. They, along with obstetricians and pediatric nephrologists, need to follow the events until delivery. Thereafter, the nephrologists along with urologists and imaging specialists jointly take over the management. CAKUT may be responsible for 34%–59% of chronic kidney disease (CKD). CAKUT comprise a variety of disorders. Antenatal suspicion of abnormalities and their subsequent confirmation has, to some extent, defined their incidence. CAKUT are present in 1%–3% of live births and cause neonatal death in 1 of 2000 births. Some isolated conditions are relatively common; those that are a component of a syndrome are rare. The former include anomalies such as pelviureteric junction obstruction, posterior urethral valve, ureterovesical junction obstruction, polycystic kidney disease, and anomalies of structure and location. CAKUT occurring as part of a syndrome are occasionally serious, but more often other syndromic abnormalities(as in Down syndrome and Turner syndrome) predominate.

As the majority of renal diseases are being satisfactorily managed, the clinical workload of CAKUT has increased at most tertiary centers. The incidence of postinfectious acute glomerulonephritis has decreased in most countries, and a large proportion of children with idiopathic nephrotic syndrome can be adequately treated until permanent remission. Acute kidney injury can be prevented or properly managed until recovery. Children with CAKUT present increasing workload at referral centers. The diagnostic evaluation of CAKUT involves imaging expertise and various modalities that entail considerable time and expenses. Urgent surgical procedures are required for obstructive lesions. Bilateral, severe abnormalities may lead to early CKD, which needs appropriate management with attention to growth and development. Eventually, renal replacement therapy is necessary, which presents serious difficulties in countries with low resources. Infants and children with CKD requiring renal replacement therapy constitute ~5% of all end stage renal failure, but require greater time and resources for their management.

The development of kidney and urinary tract is complex involving precise nephrourogenic programming and timely expression of multiple genes. Crucial developmental events such as obliteration of ureteric duct and its subsequent recanalization occur between 14 and 22 weeks of intrauterine life. Failure of development of ureteric bud results in renal agenesis and that of its recanalization causes obstruction. Factors adversely affecting renal embryogenesis are not well understood. A large number of genetic abnormalities have been reported in various isolated CAKUT and in those associated with syndromes. Only few of these have distinct relationships, e.g., in polycystic kidney disease.[2,3] They help in genetic counseling, but seldom influence the treatment and outcome. Abnormalities of kidney include those of number, size, ascent, rotation, duplication, ectopia, fusion, combinations of these, and cystic disorders. These can be unilateral or bilateral. Common anomalies include pelviureteric junction and ureterovesical junction obstruction, vesicoureteric reflux, megaureter, ureterocele, and posterior urethral valves. Their severity varies considerably. While patients with posterior urethral valves require prompt surgical intervention, others require careful evaluation and continued observation. Several anomalies are asymptomatic and incidentally detected while being investigated for a urinary tract infection or an unrelated complaint.