Mattel Children's Hospital UCLA

UCLA offers specialized medical care and surgery for congenital anomalies of the kidney and urinary tract

Congenital anomalies of the kidney and urinary tract, or CAKUT, are birth defects that are often discovered during pregnancy in the fetal ultrasound that takes place around the 20th gestational week. Conditions that fall under this umbrella affect about 1 in 600 births, and include hydronephrosis (swelling of the kidney with urine) and unilateral renal agenesis (when one kidney is missing). While some anomalies can resolve on their own after birth, others require close monitoring and corrective surgery, often within the first years of life.

At Mattel Children's Hospital UCLA, specialists from the Division of Pediatric Nephrology and the Pediatric Urology Program work together to provide comprehensive care to expecting mothers, infants and children dealing with these conditions. Because mothers can receive prenatal care and deliver their babies at the same place newborns are seen by specialists, UCLA offers an advantage over other area hospitals.

The importance of early intervention

"While most congenital anomalies are detected before birth, this wasn't always the case," says Steve Lerman, MD, a clinical professor of urology specializing in pediatric cases. "Twenty years ago, we often wouldn't find out about these kids until much later," he says. Sometimes they'd see a 3-year-old with terrible pain because of a completely blocked kidney or a 7-year-old who burst a kidney after falling on the soccer field. Both were likely to have been an undetected congenital abnormality.

Today, high-definition prenatal ultrasounds offer diagnostic imaging of the kidneys and urinary tract as they develop in utero, and as a result, the above types of presentation are rare. The goal is to start treating patients with congenital anomalies of the kidney and urinary tract as early as possible, and correct any deficits before they become catastrophic.

"Our highly specialized and collaborative team has had great success diagnosing, managing and surgically correcting these conditions," Dr. Lerman says. "We can lessen families' anxiety and ensure a smooth transition from pregnancy to infancy and beyond."

A typical timeline

After a congenital anomaly of the kidney and/or urinary tract is found on a prenatal ultrasound, it is typical that a follow-up renal ultrasound is performed within a few days of the child's birth. If the condition has not self-corrected, the family meets with a pediatric urologist and/or nephrologist shortly after birth and the newborn's kidneys, bladder and urinary tract are assessed with an advanced imaging test.

When surgery is needed, physicians try to wait until a child is at least 3 to 6 months old to operate. Two of the more common congenital anomalies, which account for 85 to 90 percent of pediatric kidney surgeries at UCLA, are a kink or blockage in the ureter at the ureteropelvic junction (UPJ) and the backward flow of urine from the bladder up to the kidneys, called vesicoureteral reflux.

In UPJ obstruction, a nuclear medicine scan can be obtained to assess the affected kidney or kidneys' drainage ability in infancy. If only one kidney is blocked, and in the absence of recurring or serious infections, ureter-repair surgery to allow urine to flow freely from the kidney to the bladder can be delayed for a few months. In vesicoureteral reflux, a voiding cystourethrogram (VCUG)study can be obtained to assess the severity of the reflux. For children suffering from recurrent infections in the bladder and/or kidneys, physicians may prescribe antibiotics to try to suppress infections before surgically correcting the problem. If either of these conditions is serious enough that both kidneys become blocked, serious infections cannot be suppressed or there is a risk of dialysis, physicians may recommend prompt surgical intervention.

Posterior urethral valves and prune belly syndrome are rare congenital conditions that affect boys and require surgery as early as possible to prevent serious harm. The former is an anomaly of the urethra that impedes urine from flowing out of the penis and causes a backup in the bladder, while the latter is urinary-tract swelling that causes the abdominal wall to become floppy.

CAKUT patients need ongoing medical management to stay healthy

Whether a congenital anomaly is serious enough to require surgery or not, continuous monitoring is often needed to make sure the kidneys function normally in regulating the body's fluids and electrolytes and to avoid complications from urinary tract infections. With proper medical management and ongoing care, most children born with a congenital anomaly of the kidney or urinary tract can grow normally and enjoy lives much like those of their healthy peers.

Yet even with close monitoring and early intervention, CAKUT are the most common cause of kidney failure in childhood and may require dialysis or a kidney transplant. With one of the most robust pediatric kidney-transplant programs in the country, UCLA is adept in dealing with these patients and managing their care.

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