# Holding Water: Congenital Anomalies of the Kidney and Urinary Tract, CKD, and the Ongoing Role of Excellence in Plumbing

Lars J. Cisek

**Congenital anomalies of the kidneys and urinary tracts can result in diminished natal kidney function, possibly through common embryologic pathway disruption or as a result of development taking place in the face of disordered 'post-renal' drainage. Impaired conduit and reservoir function present potential for an ongoing assault leading to further deterioration and progression of chronic kidney disease, a risk that extends to adults with these conditions, even after "correction". The drainage and storage aspects of the urinary system that can impact kidney function are reviewed with attention to correctable or manageable problems including: Bladder dysfunction wherein the low pressure storage of urine is compromised requiring the kidney to work against a pressure gradient, the classic post renal failure problem. The kidney in the aftermath of obstruction which may have lost concentrating capacity leading to a tendency to dehydration ('pre-renal' failure) and through polyuria which exacerbates bladder pressure problems. Further there is an added challenge in evaluation for ongoing or reemergent obstruction in a significantly dilated system where the capacious system leads to slow turnover of urine often requiring a ureteral stent or nephrostomy to clearly establish clinical significance of delayed drainage. Stasis where slow urine flow leads to buildup of debris (stone) or potentiates infection. Vessicoureteral reflux which allows for introduction of lower urinary tract bacteria to the kidney and can lead to pyelonephritis. Conditions which combine problems such as posterior urethral valves where the bladder outlet obstruction compromises kidney function potentially impairing concentrating ability, creates bladder compromise often reducing emptying efficiency or elevating bladder storage pressures, as well as dilating the system potentially promoting stasis. Cognizance of the potential for plumbing problems to further kidney deterioration as patients with congenital urinary tract anomalies, even after they have been repaired is incumbent on those caring for these patients as they age. Thoughtful evaluation of those patients in whom kidney compromise maybe aggravated by drainage and storage disorder will optimize native renal function.**



**Key Words:** CAKUT, Urinary anomaly, Congenital, CKD, Transitional care

> The society which scorns excellence in plumbing because plumbing is a humble activity and tolerates shoddiness in philosophy because it is an exalted activity will have neither good plumbing nor good philosophy. Neither its pipes nor its theories will hold water.
>
> John W. Gardner.[1]

Where to draw a line between nephrology and urology is not completely clear. For kidney excretory function the system considered as a whole works as a unit, "downstream" plumbing problems can "back up" and compromise upstream function. This may occur during development where presumably potentially normal kidneys at their inception fail in normal development and manifest dysplasia with compromised function as a result of impaired drainage.[2,3] The absence of low pressure drainage and storage can compromise immediate function ("postrenal") and lead to permanent deterioration of the kidney if persistent over time. Integrity problems, such as backflow, a bane in conventional plumbing where it is prohibited[4] because of contamination/infection risk, similarly creates these same risks for patients when urine backwash occurs.[5,6] Although early embryological theories such as that advanced by Mackie and Stephens[7] (anomalous embryology of the mesonephric duct/ureteral bud leads to faulty induction of the metanephric blastema generating dysplasia) may bring commonality to some problems, namely that reflux is integrally related to dysplasia, the added risk posed by failure to physiologically effectively isolate the kidney from the bladder is well established and provides a correctable target for management to prevent further kidney function deterioration.[5,8,9] More recently the understanding that any congenital anomalies in the urinary tract may reflect a more globally disordered system has led to recognition that as a whole congenital anomalies of the kidney and urinary tract (CAKUT) confer an increased risk of eventual CKD.[10,11] As in the example of reflux it is likely that as a whole CAKUT can engender renal compromise through a variable contributions of genetics, disordered development as a result of shoddy plumbing, and similarly promoted injury through shabby plumbing continuing after birth.

In this discussion, I will principally view urology from the standpoint of plumbing and focus most attention on "plumbing problems" that compromise function and when dismissed as nugatory contribute to progression

*From the Division of Pediatric Urology, Department of Pediatric Surgery, The University of Texas Health Science Center at Houston, Houston, TX.*

*Financial Disclosure: The author declares that he has no relevant financial interests.*

*Address correspondence to Lars J. Cisek, Division of Pediatric Urology, Department of Pediatric Surgery, The University of Texas Health Science Center at Houston, 6431 Fannin Street, MSB 5.220, Houston, TX 77030. E-mail: Lawrence.J.Cisek@uth.tmc.edu*



*1548-5595/$36.00*
*https://doi.org/10.1053/j.ackd.2017.09.012*

or even more tragically to recidivism when kidney replacement is required.[12,13] The discussion will focus on long-term management issues in the setting of early renal compromise, with limited review of currently recommended early management where the latter impacts long-term care. Congenital anomalies are far more common contributors to renal compromise in children.[13,14] However, as these children transition to adulthood ongoing cognizance and management of a compromised urinary system cannot be forgotten for optimal outcomes.

## THE URINARY SYSTEM

The upper aspect of the urinary tract is of Wolffian (mesonephric) duct origin and is affixed to the kidney itself at the fornix. This holds the trumpet-shaped calyx, which embraces the papilla and receives urine from the collecting ducts. Just outside the collecting system at the fornix is a vascular lymphatic bed, and in the case of acute obstruction, the fornix will leak/rupture and give rise to pyelovenous and pyelolymphatic backflow and peripelvic extravasation. The infundibulum is a funneled tube, which bridges the calyx to the renal pelvis. The pelvis receives drainage from the calyxes and funnels this to the ureter.[15] There are pacemaker centers that induce contraction of the muscle investing the pelvis and ureter, which propagate contraction distally by myogenic transmission. The frequency of contraction is enhanced by pelvic distention, which will increase peristalsis down the ureter, although will become disordered with prolonged obstruction. The renal pelvis joins to the ureter at the ureteropelvic junction (UPJ)—the region most prone to obstruction in the system. The ureter is a small diameter (hence small volume) tube that traverses the distance from the kidney to the bladder where it perforates the bladder. As it is of Wolffian origin it is tethered distally to the veru, fixed distally, and paired from each side of the body, this forms the triangular trigone. The course through the bladder wall and beneath the mucosa is tangential with bladder muscle behind it and the bladder lumen providing support beneath the ureter. This arrangement compresses the ureter as pressure in the bladder rises serving the role of a backflow preventer valve (for anatomy see https://training.seer.cancer.gov/anatomy/urinary/components/urethra.html).

**CLINICAL SUMMARY**

- Congenital anomalies of the urinary tract can give rise to disordered renal development, promote deterioration of function, and give rise to chronic renal insufficiency.
- Urinary tract anomalies are a common cause of childhood onset renal impairmentt.
- Abnormal urine drainage and storage can lead to ongoing renal functional loss, compromise renal replacement strategies, and often require ongoing management.

The volume of an upper track is normally less than 5 to 10 mL making it a relatively high flow system limiting stasis problems, which further isolates the kidney from both bladder pressure and contaminates. The bladder is a balloon shaped hollow viscus consisting of a transitional epithelial mucosal lining surrounded by a muscular wall, which serves a storage function. This provides adequate capacity and compliance to store urine at low pressure for several hours. Both a competent and a patent egress through the urethra provide for control and complete bladder emptying when coupled with a coordinated bladder contraction. Emptying is a coordinated activity normally comprising relaxation of the pelvic floor, urinary sphincter, and bladder neck followed shortly thereafter by contraction of the bladder itself. The entire process is largely a spinal reflex arc. Volitional control is exercised by supratentorial input, which simply inhibits contraction of the bladder per se and can contract the external sphincter/pelvic floor. Micturition occurs when inhibition is released and adequate tension in the bladder wall activates the spinal reflex arc.[16] The system is normally sterile beyond the distal aspect of the urethra. The system as a whole serves to allow the kidney to remain sterile and subject to only a minimal urine pressure head engendering optimal function.

## UROLOGICAL PROBLEMS OF CONCERN IN RENAL IMPAIRMENT

Two intertwined general problems arise from urinary dysfunction: those that impair function per se and/or that lead to deterioration of function over time (Table 1).

### Bladder Dysfunction

This may be of either myogenic or neurogenic etiology and compromises the ability of the bladder to store urine at low pressure and empty efficiently. High bladder pressure compromises kidney function acutely and will also result in deterioration over time. Furthermore, any problems arising from infection are magnified if coupled with increased bladder pressures, and persistent excessive pressure may lead to dilation and loss of ureteral backflow check valve function over time. Ensuring the bladder continues to provide low pressure storage is among the commonly overlooked problems leading to ongoing functional deterioration and sadly recidivism in the case of kidney transplant.[12,13,17] Principal action items are to maintain active management/monitoring and vigilance in addressing upper tract dilation and bona fide infection. Pressure is best conceptualized as a time average, and that treatment measures such as overnight catheterization can substantively reduce the overall time outside the favorable pressure range.

For adolescents and young adult with a history of spinal cord surgery, realization that their growth may pull previously tethered nerves affixed from their prior corrective surgery ("retethering") and alter seemingly stable bladder dynamics must be borne in mind with periodic monitoring. The mainstay of monitoring is a urodynamics study where a catheter is placed in the bladder and the bladder is filled to generate a pressure volume curve.[16-18]