http:// jpp.mums.ac.ir
Letter to Editor (Pages: 19747-19750)

CAKUT Conundrums: Tailoring Kidney Transplants for Congenital Anomalies in Kids

Iman Nosratabadi [1] , * Reza Abdollahi [2]

[1] Department of Nursing, Faculty of Nursing and Midwifery, Sirjan University of Medical Sciences, Sirjan, Iran.

[2] Patient Safety Research Center, Clinical Research Institute, Nursing & Midwifery School, Urmia University of Medical Sciences, Urmia, Iran.

Abstract

Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) represent a spectrum of structural malformations present at birth, affecting approximately 4.2 to 1000 per 10,000 births worldwide. These include conditions such as posterior urethral valves (PUV), vesicoureteral reflux (VUR), renal hypoplasia, and neurogenic bladder (NGB), which collectively account for nearly half of end-stage renal disease (ESRD) cases in children. CAKUT disrupts normal renal development and urinary flow, leading to progressive chronic kidney disease (CKD) and often necessitating kidney transplantation (KTx) as the optimal renal replacement therapy. While pediatric KTx offers superior outcomes compared to dialysis, including improved growth and quality of life, children with CAKUT face unique conundrums due to underlying anatomical and functional abnormalities. These challenges complicate surgical procedures, elevate post-transplant risks, and demand tailored multidisciplinary strategies. Tailoring kidney transplants for CAKUT in children navigates a labyrinth of anatomical, functional, and infectious pitfalls through rigorous evaluation, adaptive surgery, and vigilant post-op care. This multidisciplinary approach not only salvages grafts but also elevates quality of life, though prospective studies are imperative to refine protocols and resolve lingering conundrums. As pediatric nephrology evolves, addressing these congenital anomalies promises brighter futures for affected kids.

Key Words: CAKUT, Congenital Anomalies, Kidney Transplants, Kids.

* Please cite this article as: Nosratabadi I, Abdollahi R. CAKUT Conundrums: Tailoring Kidney Transplants for Congenital Anomalies in Kids. J Ped Perspect 2025; 13 (11):19747-19750. **DOI: 10.22038/jpp.2025.92049.5607**

**** Corresponding Author:***

Reza Abdollahi, Patient Safety Research Center, Clinical Research Institute, Nursing & Midwifery School, Urmia University of Medical Sciences, Urmia, Iran. Tel: 09019753320; Email : rezaabdollahi97@yahoo.com

DEAR EDITOR

Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) represent a spectrum of structural malformations present at birth, affecting approximately 4.2 to 1000 per 10,000 births worldwide. These include conditions such as posterior urethral valves (PUV), vesicoureteral reflux (VUR), renal hypoplasia, and neurogenic bladder (NGB), which collectively account for nearly half of end-stage renal disease (ESRD) cases in children. CAKUT disrupts normal renal development and urinary flow, leading to progressive chronic kidney disease (CKD) and often necessitating kidney transplantation (KTx) as the optimal renal replacement therapy. While pediatric KTx offers superior outcomes compared to dialysis, including improved growth and quality of life, children with CAKUT face unique conundrums due to underlying anatomical and functional abnormalities. These challenges complicate surgical procedures, elevate post-transplant risks, and demand tailored multidisciplinary strategies (1).

Pre-transplant evaluation emerges as a critical battleground in addressing CAKUT-related challenges, where incomplete assessment can doom the graft to failure. Unlike non-CAKUT recipients, children with these anomalies often harbor dysfunctional bladders or obstructions that impair urine drainage and foster infections. A thorough multidisciplinary approach, involving nephrologists, urologists, and surgeons, is essential. Key tools include ultrasound for anatomical mapping, voiding cystourethrography (VCUG) to detect reflux or obstructions, and urodynamic studies (UDS) to evaluate bladder compliance, capacity, and emptying mandatory for lower urinary tract obstructions (LUTO) or NGB. Genetic testing for syndromic CAKUT provides prognostic insights and reproductive counseling, while functional scans like dimercaptosuccinic acid (DMSA) assess differential kidney function. The analytical crux lies in recognizing that CAKUT's heterogeneity, upper versus lower tract issues dictates risk stratification; for instance, PUV or NGB heightens urinary tract infection (UTI) susceptibility due to residual urine and high-pressure voiding (2). Tailoring here involves proactive interventions: pharmacotherapy with anticholinergics like oxybutynin to reduce detrusor overactivity, or clean intermittent catheterization (CIC) to ensure complete emptying. However, evidence remains retrospective, highlighting a conundrum: while these steps mitigate risks, their timing and intensity vary across centers, potentially introducing selection bias in outcomes (3).

Surgical tailoring during transplantation confronts the anatomical distortions inherent to CAKUT, transforming a standard procedure into a bespoke operation. Native nephrectomy, for example, is not routine but indicated for recurrent UTIs, hypertension, or space-occupying cysts, as in multicystic dysplastic kidneys. Timing pre-, peri-, or post-transplant balances risks like anuria complicating dialysis against benefits like infection prevention. For hostile bladders, augmentation cystoplasty using intestinal segments creates a low-pressure reservoir, though it risks metabolic acidosis and malignancy long-term. Ureteral implantation techniques, such as extravesical anti-reflux reimplantation or ureteroureterostomy, adapt to native anomalies to prevent VUR into the graft. In small children (<10 kg), intraperitoneal placement of adult-sized kidneys addresses vascular mismatches, using the aorta and inferior vena cava for anastomosis to ensure high flow and minimize thrombosis a frequent early graft loss culprit in pediatrics. Analytically, these adaptations underscore a trade-off: while they enable

transplantation in complex cases, they prolong operative time and heighten perioperative complications, such as urine leaks or obstructions. Yet, with meticulous technique and overhydration, outcomes rival non-CAKUT transplants, illustrating how surgical innovation counters congenital barriers (3).

Post-transplant complications amplify CAKUT conundrums, where the interplay of immunosuppression and residual anomalies fosters a vicious cycle of infections and graft dysfunction. UTIs, including pyelonephritis, afflict about one-third of recipients, with febrile UTIs more prevalent in CAKUT due to factors like female gender, pre-transplant UTI history, and persistent VUR. These infections transiently impair graft function but, if untreated, erode long-term survival. Graft VUR, common despite anti-reflux measures, exacerbates risks, necessitating vigilant monitoring via repeat UDS in high-risk groups. Hypertension, often nocturnal non-dipping, further strains the allograft. Tailoring management involves prophylaxis with low-dose antibiotics, hydration protocols, and early stent removal, alongside addressing bowel-bladder dysfunction per International Children's Continence Society guidelines. The analytical lens reveals that while CAKUT does not recur, its legacy of dysfunctional drainage demands lifelong surveillance, contrasting with non-CAKUT cases where complications are more immunologically driven (4).

Despite these hurdles, outcomes for CAKUT children post-KTx are encouraging, with 5- to 10-year graft survival rates of 70-77%, comparable or superior to non-CAKUT peers, particularly with pre-emptive living donor transplants. Subtype variations exist: VUR nephropathy fares better than PUV, yet overall prognosis hinges on pre-transplant optimization. Patient survival nears 98% at 9 years, underscoring transplantation's viability. However, challenges like higher UTI rates in lower tract malformations persist, with no standardized guidelines due to retrospective data limitations (5).

Table-1. Summarizes common CAKUT subtypes, their associated risks, and tailored intervention.

CAKUT Subtype	Associated Risks	Tailored Interventions
Posterior Urethral Valves (PUV)	High UTI susceptibility, residual urine, high-pressure voiding, hypertension	Pre-transplant UDS and CIC; pharmacotherapy (e.g., anticholinergics); native nephrectomy if recurrent UTIs; anti-reflux ureteral implantation
Vesicoureteral Reflux (VUR)	Recurrent UTIs, pyelonephritis, graft dysfunction from persistent reflux	VCUG for detection; anti-reflux surgical techniques; antibiotic prophylaxis post-transplant; vigilant UTI monitoring
Renal Hypoplasia/Dysplasia	Progressive CKD, ESRD, space-occupying cysts	DMSA scans for function assessment; native nephrectomy for cysts; pre-emptive living donor KTx for better outcomes
Neurogenic Bladder (NGB)	Impaired emptying, detrusor overactivity, infections	UDS evaluation; CIC or augmentation cystoplasty; anticholinergics; hydration protocols and bowel-bladder management
Multicystic Dysplastic Kidney	Hypertension, recurrent UTIs, vascular mismatches in small children	Native nephrectomy pre- or peri-transplant; intraperitoneal graft placement for small recipients; genetic testing for syndromic cases

In conclusion, tailoring kidney transplants for CAKUT in children navigates a labyrinth of anatomical, functional, and infectious pitfalls through rigorous evaluation, adaptive surgery, and vigilant post-op care. This multidisciplinary paradigm not only salvages grafts but elevates quality of life, though prospective studies are imperative to refine protocols and resolve lingering conundrums. As pediatric nephrology evolves, addressing these congenital anomalies promises brighter futures for affected kids.

CONFLICT OF INTEREST

The author declares that he has no competing interests to disclose in relation to this paper.

REFERENCES

1. Maringhini S, Pape L. Kidney Transplantation in Congenital Abnormalities of Kidney and Urinary Tract (CAKUT). Biomedicines. 2025 Apr 10;13(4):932.

2. Cornwell LB, Ingulli EG, Mason MD, Ewing E, Riddell JV. Renal transplants due to congenital anomalies of the kidney and urinary tract (CAKUT) have better graft survival than non-CAKUT controls: analysis of over 10,000 patients. Urology. 2021 Aug 1;154:255-62.

3. Aron AW, Dahl NK. Clinical genetic testing in nephrology: core curriculum 2024. American Journal of Kidney Diseases. 2024 Nov 1;84(5):632-45.

4. Morais IS. The role of genetic testing in the diagnosis and management of chronic kidney disease: pathway to tailored nephrology. PQDT-Global. 2023.

5. Schumacher F, Thumfart J, Eckardt K-U, Kleuser B, Bartolomaeus H, Wilck N. Gut dysbiosis contributes to TMAO accumulation in CKD. 2024.