



Unilateral Renal Agenesis with Contralateral Multicystic Dysplastic Kidney in a Neonate

David Myers*

Department of Urology, Cyril and Methodius University, Skopje, BulgariaZealand

✉ David Myers

Department of Urology,

Cyril and Methodius University,

Skopje, Bulgaria

E-mail: dmyers@northhealth.org

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Description

Unilateral Renal Agenesis (URA) and Multicystic Dysplastic Kidney (MCDK) are Congenital Anomalies of the Kidney and Urinary Tract (CAKUT), representing distinct developmental disorders with significant implications when occurring together. While each anomaly alone can be compatible with a relatively normal life if the contralateral kidney is healthy, the simultaneous presence of URA and contralateral MCDK is a rare and severe condition often incompatible with long-term survival. This combination leads to a functional absence of both kidneys and, consequently, end-stage renal failure in the neonatal period.

MCDK, on the other hand, is a nonhereditary, usually unilateral renal anomaly characterized by a non-functioning kidney replaced by multiple non-communicating cysts and absence of normal renal parenchyma. It is one of the most common causes of an abdominal mass in neonates. MCDK arises from early obstruction of the ureteric bud, which prevents normal nephron formation, leading to abnormal metanephric

differentiation. Typically, the affected kidney is non-functional and involutes over time. Bilateral MCDK is fatal due to the absence of functioning renal tissue and resultant oligohydramnios and pulmonary hypoplasia.

In the rare circumstance when URA coexists with contralateral MCDK, the neonate is left with no functioning renal tissue—a condition known as renal aplasia or bilateral renal agenesis in functional terms. This condition results in oligohydramnios during fetal life, leading to pulmonary hypoplasia, Potter sequence, and severe postnatal complications including respiratory failure, anuria, and metabolic imbalances. The estimated incidence of this specific combination is exceedingly low, with few cases reported in medical literature.

Diagnosis of this condition can often be suspected antenatally through routine obstetric ultrasonography. In the second trimester, the absence of one kidney combined with a multicystic appearance of the contralateral renal fossa raises concern for this anomaly. Oligohydramnios low amniotic fluid volume due to impaired fetal urine production is often a significant finding and a prognostic marker of poor outcome. Fetal MRI may be used to confirm the absence of renal tissue and to assess associated findings such as pulmonary hypoplasia.

Postnatal evaluation begins with a thorough clinical examination. Infants may present with features of Potter sequence, including flattened facial features, low-set ears, limb deformities, and respiratory distress due to underdeveloped lungs. Abdominal palpation

may reveal a cystic mass if the MCDK is significantly enlarged. More importantly, the neonate will typically exhibit signs of renal failure shortly after birth absent urine output (anuria), rising blood urea nitrogen and creatinine, electrolyte disturbances (hyperkalemia, hyponatremia, metabolic acidosis), and fluid imbalance.

The prognosis in neonates with unilateral renal agenesis and contralateral multicystic dysplastic kidney is extremely poor. The absence of functional renal tissue results in End-Stage Renal Disease (ESRD) from birth. These neonates require immediate intensive supportive care, often in a Neonatal Intensive Care Unit (NICU). Management is focused on addressing life-threatening complications such as hyperkalemia and acidosis, respiratory failure, and fluid overload. Haemodialysis or peritoneal dialysis is required in the first days of life if the infant survives long enough to initiate renal replacement therapy.

However, initiating dialysis in neonates with this condition is challenging. Peritoneal dialysis, the preferred modality in neonates due to size and vascular access limitations, may be complicated by technical issues, including catheter-related infections, poor drainage, and respiratory compromise due to abdominal distension. Moreover, the presence of pulmonary hypoplasia often leads to early respiratory failure that is not easily reversible with dialysis alone. Even if dialysis is initiated successfully, the infant remains dependent on long-term renal replacement therapy until kidney transplantation can be considered, typically after the

first year of life.

Advances in fetal imaging and the use of fetal interventions, such as serial amnioinfusions or fetal shunting, have been explored to mitigate the effects of oligohydramnios and pulmonary hypoplasia in similar conditions. However, these interventions are still experimental and have not demonstrated consistent success in changing outcomes in cases with complete bilateral renal non-function. Gene testing and evaluation for syndromic associations (e.g., branchio-oto-renal syndrome, renal-coloboma syndrome, or Fraser syndrome) may be appropriate, especially if there are other anomalies present.

Conclusion

Unilateral renal agenesis with contralateral multicystic dysplastic kidney in a neonate represents a critical and life-threatening condition due to the complete absence of functional renal tissue. Antenatal detection through imaging and associated findings such as oligohydramnios and pulmonary hypoplasia can guide prognosis and management decisions. Postnatally, these neonates present with anuria and signs of renal failure, often compounded by respiratory distress. Despite advances in neonatal care and renal replacement therapy, the prognosis remains poor, and care should be centered on multidisciplinary support and ethical decision-making tailored to each family's values and wishes. Continued research in fetal interventions, neonatal dialysis technologies, and transplantation may one day offer better outcomes for this devastating condition..