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Case Series

Antenatal Diagnosis of Bilateral Multicystic Kidney Dysplasia: Two Case Reports

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Abstract

Multicystic kidney dysplasia (MCKD) is the most common clinical expression of Congenital Abnormalities of Kidney and Urinary Tract (CAKUT). Its etiopathogeny is still debated and the obstructive hypothesis is generally accepted. It can be found in isolation or as part of a genetic syndrome.

The aim of this work is to demonstrate the role of obstetrical ultrasound and MRI in the antenatal diagnosis of multicystic kidney dysplasia and the search for other malformations, through two cases diagnosed in the pediatric radiology department.

Keywords: *Multicystic; Kidney; Dysplasia; Ultrasound; MRI*

Abbreviations

MCKD: Multicystic Kidney Dysplasia; CAKUT: Congenital Abnormalities of Kidney and Urinary Tract

Introduction

Multi-cystic dysplastic kidney (MCDK) is one of the most common congenital anomalies of the urinary tract, with an estimated incidence of 1 in 3,640 births [1]. It is characterised by a general structural disorganisation of the renal parenchyma with multiple non-communicating cysts throughout the dysplastic renal tissue. It is usually sporadic, but some familial cases may exist. It can be isolated, associated with other genitourinary malformations or part of a genetic syndrome [2].

It is described as most often unilateral (75%) with a location in the left kidney in 53% of cases. While the bilateral form is usually lethal. It affects males in 60% of cases [2]. Its pathogenesis is incompletely elucidated. In the classical vision, it results from a defect in the induction of the metanephric blastema by the ureteral bud. Another hypothesis suggests that MCDK is secondary to a major abnormality of fetal urine flow occurring early in renal development.

Antenatal ultrasound and MRI play a crucial role in the diagnosis of CDKD and the search for associated malformations and also provide important diagnostic information for the follow-up of these patients.

In this work we report two cases of antenatally diagnosed multicystic kidney dysplasia.

Case Reports

Case report 1

A 34 year old woman, gravida 3, para 2, presented to the mother and child radiology department for routine obstetric ultrasound at 24 weeks gestation. Her first two pregnancies were carried to term, with vaginal delivery, without any other particularities. The patient and her partner have no history of renal disease. The ultrasound scan showed an anamnios, intrauterine growth retardation of the fetus, enlarged kidneys contain pure anechoic cystoid formations, with thin walls, without communication between them, with no visualization of the bladder (Figure 1).

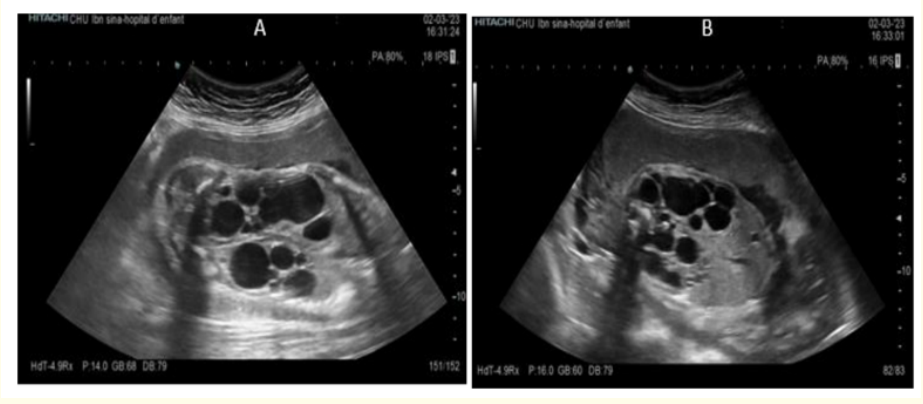
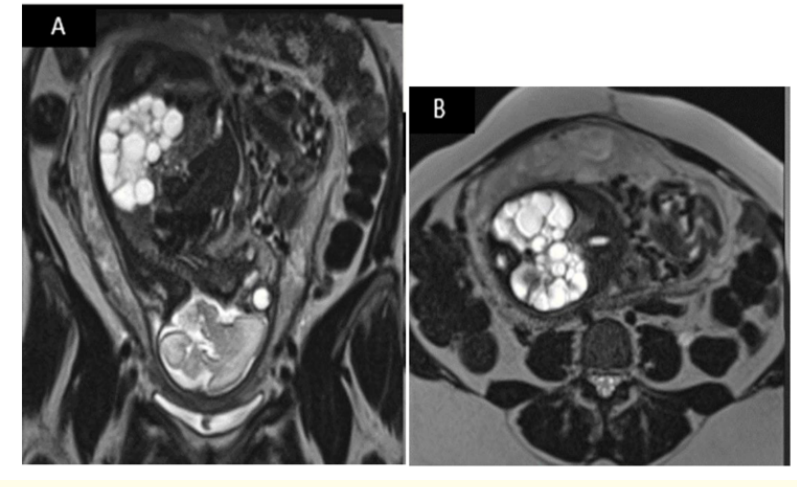


Figure 1: *Transverse (B) and longitudinal (A) image of both kidneys at 24 weeks gestation showing an anterior insertion placenta, multiple simple cysts of both kidneys, absence of amniotic fluid.*

A fetal MRI was performed to look for other associated malformations, showed a fetus in cephalic presentation with an anteriorly inserted placenta, an enlarged right and left kidney measuring (RK: 62 mm, LK: 63 mm), containing multiple simple cysts of varying size, an empty bladder and no amniotic fluid with no other malformations (Figure 2).



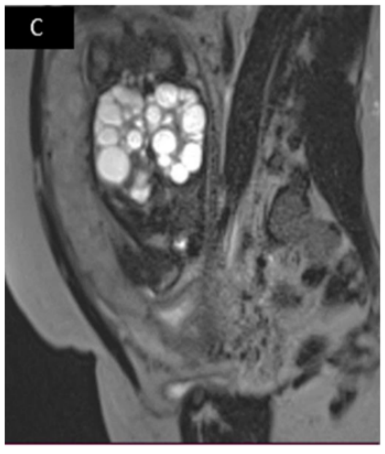


Figure 2: *Fetal MRI in T2 sagittal (A) axial (B) and Coronal (C) sequences showing: renal parenchyma replaced by multiple thin-walled T2 hypersignal cystic formations, without septa or vegetations and absence of amniotic fluid.*

Case report 2

A 28 year old woman, gravida 1, para 0, with no medical history, presented to the mother and child radiology department for routine obstetric ultrasound at 27 weeks gestation. The ultrasound scan showed a large left kidney occupying the entire abdominal cavity, with multiple small cysts within its parenchyma with no visualization of the right kidney (Figure 3).

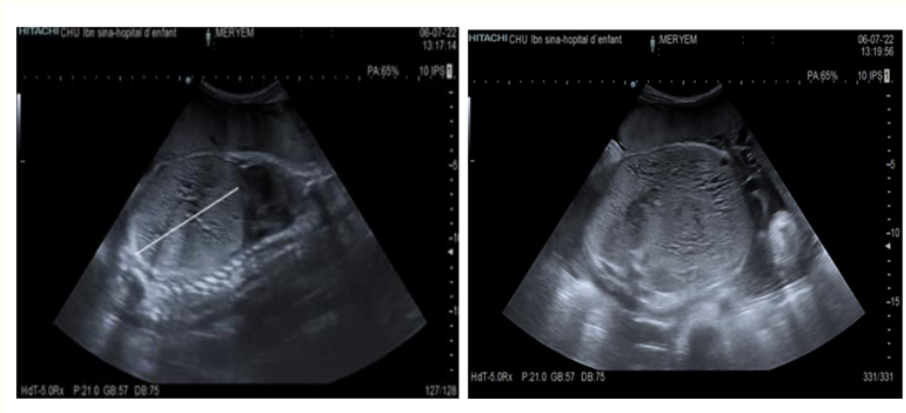


Figure 3: *Longitudinal (A) and Transverse (B) image of the left kidney at 28 weeks gestation showing an anterior insertion placenta and a large left kidney occupying the entire abdominal cavity, with multiple small cysts within its parenchyma.*

A fetal MRI was released to characterise the appearance found on the ultrasound and to look for other associated malformations, showed a fetus in cephalic presentation with an anteriorly inserted placenta, a large left kidney occupying the entire abdominal cavity,

with the parenchyma replaced by microcysts. The right kidney has a normal size and the same characteristics, an empty bladder and a an occipital myelomeningocele (Figure 4).

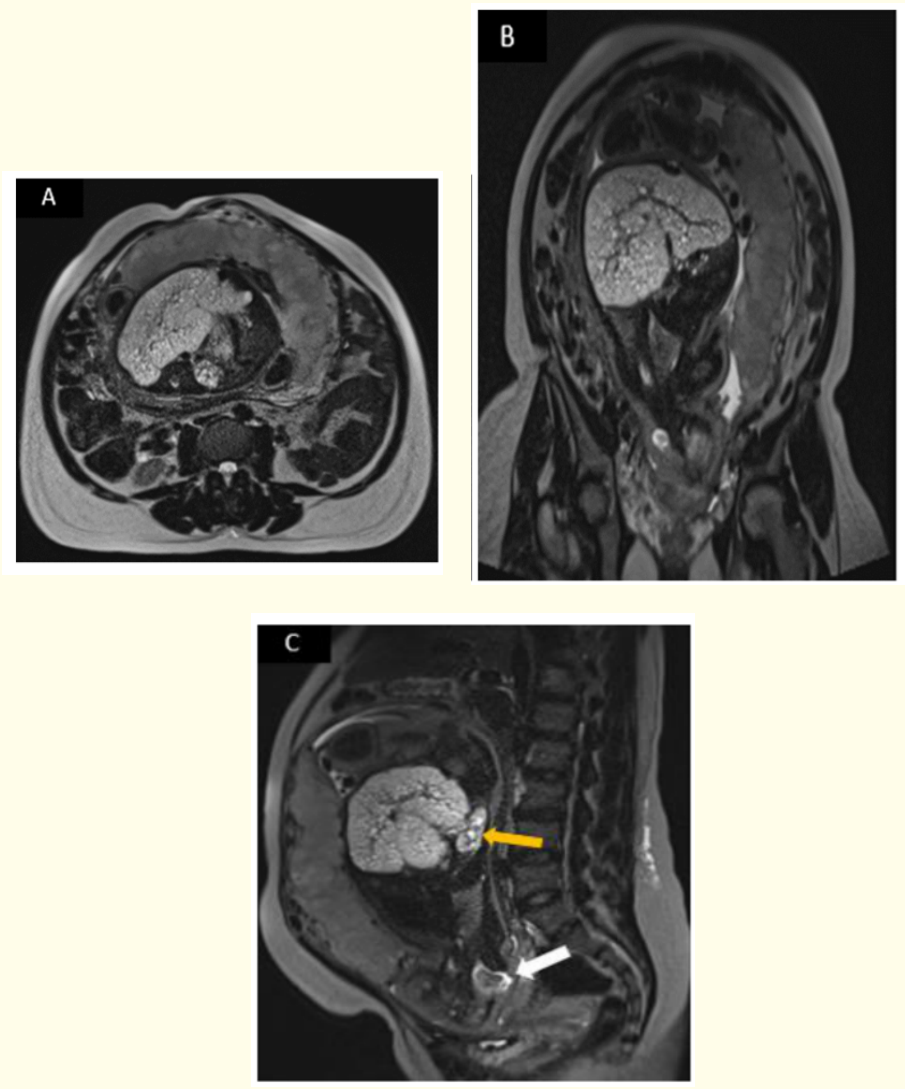


Figure 4: *Fetal MRI in T2 axial (A) Coronal (B) and sagittal (C) sequences showing a cephalic position fetus with a large left kidney occupying the entire abdominal cavity, with the parenchyma replaced by microcysts. The right kidney (Yellow arrow) is of normal size with the same characteristics. Microcephaly with occipital myelomeningocele (white arrow).*

Discussion

Multi-cystic kidney dysplasia (MCKD) is one of the most common malformations of the urinary tract, classified as "Congenital Abnormalities of Kidney and Urinary Tract (CAKUT)". Around 40% of end-stage renal disease in children is secondary to CAKUT [3], with an estimated incidence of 1 in 3,640 births [1].

There are multiple theories explaining the development of MCDK. The ureteral bud theory suggests that multi-cystic kidney dysplasia is due to an abnormal communication of the ureteral bourgeon with the mesencephalic mesenchyme [4]. This theory has also been suggested for the pathogenesis of other renal abnormalities, especially vesico-ureteral reflux and renal agenesis. In support of this theory, In support of this hypothesis, there are mutations that play a role in MCDK, as well as other forms of renal dysplasia, including EYA1, SIX1 and PAX2 [5].

MCKD is distinct from polycystic kidney disease (ADPKD) which is an autosomal dominant disease, the presence of a single cyst is sufficient to establish the diagnosis in a child under 5 years with a known family history of ADPKD [6].

MCKD is usually diagnosed on routine prenatal ultrasound examination, which will show clusters of non-communicating grape-shaped cysts in enlarged kidneys (Figure 1 and 3) with no visualisation of the bladder due to lack of urine accumulation causing a severe oligohydramnios [7].

There may be associated non-renal, cerebral, cardiac or other urogenital anomalies. Antenatal ultrasound monitoring of MCDKD may reveal changes in the appearance of the affected kidneys, The kidneys may enlarge or decrease in size (due to involution of the bowel), rarely disappearing [8]. Both our cases had severe bilateral kidney involvement with extreme oligohydramnios. The absence of a bladder and the presence of another extra-renal malformation in the second case, an occipital myelomeningocele, were also noted, both cases had a growth delay of 3 - 5 weeks.

The prognosis of unilateral MCDK is generally favourable, the normal kidney will grow a little more than average, to “compensate”. The dysplastic kidney will evolve in three ways:

1. Decrease in size, or even disappear completely 55 to 95% of cases.
2. The dysplastic kidney stays stable: 15 to 30% of cases.
3. The dysplastic kidney increases in size: 0 - 2% of cases.

While the literature reports a pejorative prognosis for the bilateral form describing it as lethal [9].

The parents were informed of the gravity of the prognosis. Both cases ended in spontaneous in utero fetal death within 5 - 6 weeks of diagnosis.

Conclusion

Multicystic kidney dysplasia (MCKD) is a large category of congenital malformations that may be secondary to obstructive uropathy. Its antenatal diagnosis depends essentially on obstetric ultrasound. Thanks to the progress made in this field and the qualification of ultrasound technicians, screening can be carried out at an early stage. The search for associated renal or extra-renal anomalies is necessary in view of the possibility of chromosomal or gene anomalies. The prognosis of bilateral MCKD remains unpredictable because of the inability to quantify residual functional parenchyma, while that of unilateral MCKD remains good when it is isolated.

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Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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