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Antenatal Diagnosis Of Multicystic Renal Dysplasia: A Case Report And Review Of The Literature

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Abstract: *Multi-cystic renal dysplasia (MCRD) is an anomaly of renal development, usually unilateral, characterized by a large cystic kidney and a totally reworked, non-functional parenchyma. It is the most frequent clinical expression of Congenital Abnormalities of Kidney and Urinary Tract (CAKUTDiagnosis of CKUT is almost always prenatal in developed countries, thanks to ultrasound, but often post-natal in Africa, when a large abdominal mass is detected. Long-term prognosis is generally good, but nephron reduction requires long-term follow-up and the introduction of measures to prevent cardiovascular and nephrotoxic risks. We report a case of multi-cystic renal dysplasia diagnosed on second-trimester morphological ultrasonography with a good post-natal evolution, and review the diagnostic, etiological, prognostic and therapeutic aspects of this pathology.*

Keywords: antenatal diagnosis, multicystic renal dysplasia, ultrasound, prognosis.

Observation :

This was a 29-year-old patient, III gesture II pare, received in our department for the follow-up of a pregnancy of 21 weeks of amenorrhea (SA).

The patient's obstetrical history included two deliveries, the last of which resulted in a newborn weighing 4700 grams following a scheduled caesarean section for unbalanced gestational diabetes despite insulin therapy.

Clinical examination was normal. Nuchal translucency was measured at 1.2 mm on first-trimester ultrasound.

Morphological ultrasound examination at 21 weeks' gestation revealed an abdominal circumference of 165mm, at the P50-P90th percentile of 21 weeks' amenorrhea. A multiloculated cystic mass measuring 6.16 mm by 48.1 mm, with the largest cyst measuring 3.3 mm in diameter, was visualized in the right renal pelvis (figures 1, 2 and 3).

The homolateral ureter was not dilated. The left kidney had a normal morphological appearance.

Pregnancy progress was normal, and delivery was by vaginal delivery at 37 weeks' gestation and 2 days in spontaneous labor.

She gave birth to a male infant weighing 4,000 grams and measuring 52 cm, with an Apgar of 10/10/10 at 5 minutes. Postpartum care was straightforward. The neonatal examination confirmed the diagnosis of right multi-cystic renal dysplasia, and a scintigraphy performed at 6 months of age confirmed its non-functional nature. The evolution of the child was normal after 24 months.

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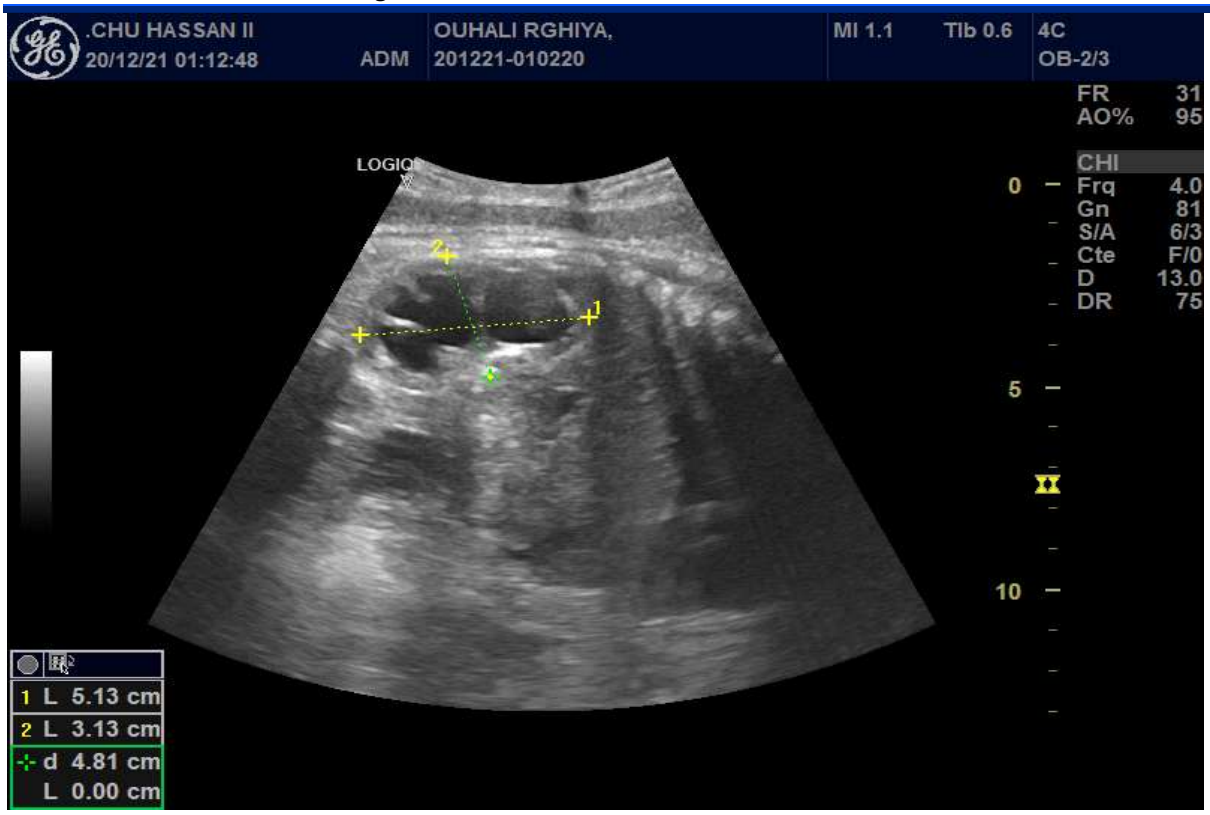

figure 1

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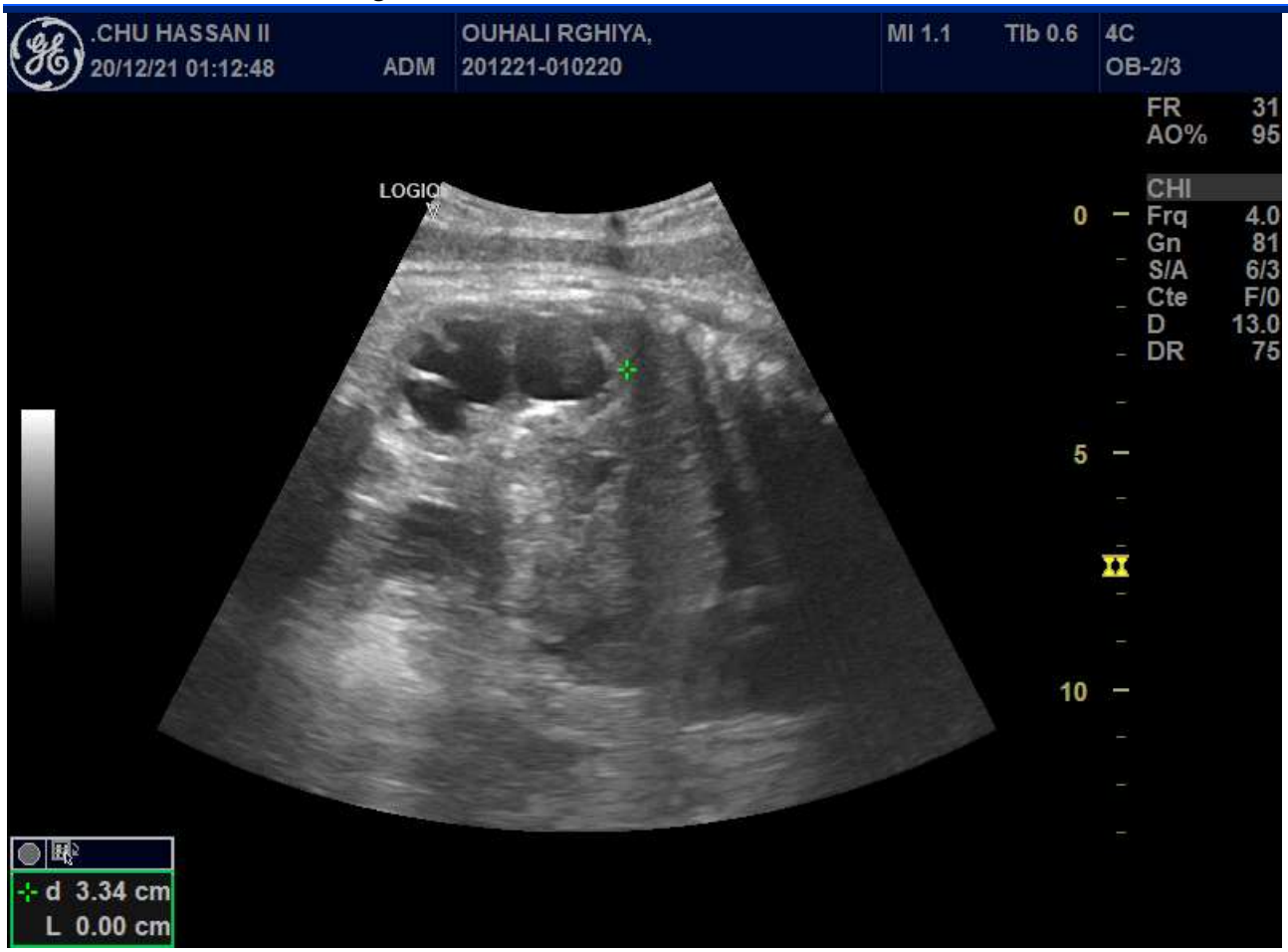

figure 2

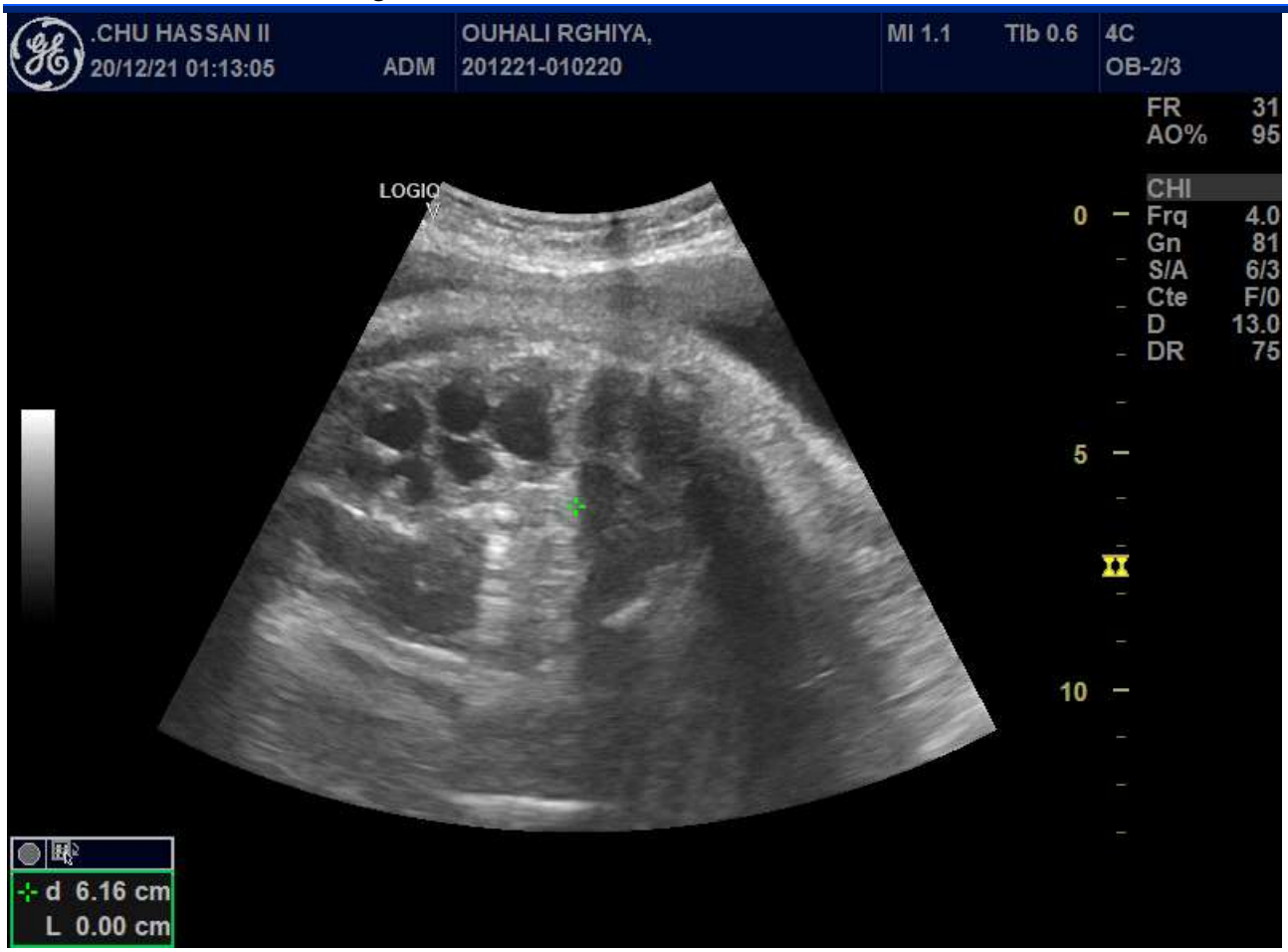

figure 3

DISCUSSION :

1. Epidemiological aspects

Multi-cystic kidney dysplasia (MCKD) is one of the most frequent malformations of the urinary tract, grouped under the term Congenital Abnormalities of Kidney and Urinary Tract (CAKUT) [1], or congenital anomalies of the kidneys and urinary tract. Around 40% of end-stage renal disease in children is secondary to CAKUT [2]. The clinical spectrum is broad, including renal (aplasia, hypoplasia, dysplasia), ureteral (megaureter, pyeloureteral junction syndrome, duplications), bladder and urethral anomalies [2].

DMK is a mostly unilateral anomaly of renal development, characterized by a large, cystic kidney and a totally altered, non-functional parenchyma. Its incidence is estimated at one per 4300 live births. Boys are more often affected, with a sex ratio of 1.48. The left kidney is most frequently involved (55% of cases). Bilateral involvement is rare, so ge'ne'ralement le'tale [3]. Some forms of DMK may eˆtre familial [4-6], but most are sporadic [2].

2. Pathogenesis

The pathogenesis of DMK is still uncertain. For some authors, it could eˆtre la conse'quence d'une obstruction ure'te'rale pre'coce in utero avec anomalie majeure de l'e coulement des urines fœtales [2,7]. For others, it may be secondary to a de'faut of induction of the me'tane'phric blastema by the ure'te'ral bud [8]. Both genetic and environmental factors are likely to be involved, as illustrated by the increased risk of DMK with fetal exposure to certain antie'pileptic drugs (carbamaze'- pine, phe'nobarbital) [1], gestational diabetes [9] or when there is a polymorphism of genes of the re'nine-angiotensin system [10] or of the Glial cell linederived neurotrophic factor (GDNF) cascade, such as PAX2, BCL2 or WT1 [2,8]. In our patient, we noted a history of gestational diabetes in the 2nd pregnancy. DMK can also occur in the context of chromosomal or syndromic abnormalities [2], for

example in Wiedemann-Beckwitt, Perlman, Simpson-Golabi-Behmel and Kallmann-de Morsier syndromes [11]. In our patient, no other malformations or chromosomal abnormalities were detected during postnatal examinations.

3. Diagnostic aspects

At present, almost all DMKs are diagnosed by prenatal ultrasonography, usually at the morphological examination performed between 20 and 22 weeks' amenorrhea (SA) [12,13]. In our case, the diagnosis was made at 21 weeks' amenorrhea. The ultrasound findings were comparable to those described in the literature [16].

One third of all anomalies detected during ante-natal ultrasound are CAKUT [2]. Of these, DMK is almost always diagnosed prenatally [12,13]. Ultrasound examination reveals anegenous intra-renal cysts, often large, inegal in size and non-communicating. The renal parenchyma is totally altered and fibrous, thinned and sometimes unreliable, which explains the non-functional character of an affected kidney [14]. In unilateral forms, careful consideration must be given to the control kidney, which will determine the final renal prognosis: it may present anomalies such as pye'lo-ureteral junction syndrome or venous-ureteral reflux, or even hypoplasia [14]. Depending on the author, contralateral anomalies are found in 7 to 43% of cases [12]; a recent study of 97 children with DMK found 20% contralateral anomalies (mainly pye'local dilatation) [12]. The quantity of amniotic fluid and the appearance of the bladder must also be studied.

A karyotype is recommended when there are associated extra-renal signs.

A series of 38 fetuses with ante-natally diagnosed DMK found associated re'nal and non-re'nal anomalies in 21% and 5% of fetuses respectively [15]. The fetuses in which DMK was not isolated all had a normal karyotype; however, 4 children were de'ce'de'd due to associated anomalies. The overall prematurity rate was 16%. The quantity of amniotic fluid did not appear to eˆtre a prognostic factor [15]. DRMK should also be distinguished from obstructive renal dysplasia, characterized by the presence of small subcortical cysts with markedly increased echogenicity of the renal cortex, and from hydronephrosis, in which there is communication between the highly dilated calyces and the renal pelvis [16].

4. Prognostic aspects

The evolution of DRMK in utero is variable. An increase in the size and number of cysts is possible, although early regression may be observed, sometimes leading to non-visualization of a kidney [16]. Ultrasound follow-up of children with DRMK shows partial or total involution of the affected kidney in 60 to 89% of cases within 9 months to 10 years [17]. Stabilization is noted in 2 to 37% of cases, and an increase in size in 2%. In the majority of cases, the contralateral kidney shows compensatory hypertrophy with a mean delay of 30 months (95% confidence interval: 15-45 months) [18]. In our case, the size of the functional kidney was still normal at 24 and 9 months respectively.

Approximately 40% of end-stage renal disease in children is secondary to CAKUT [19].

Nevertheless, MRKD is exceptionally responsible for end-stage renal failure, the prognosis depending on the contralateral kidney [19]. During childhood, only nephron reduction and compensatory hypertrophy are observed. It is only in adulthood that lesions can progress gradually and often insidiously towards chronic renal failure. It is also at this age that other factors contributing to the progression of renal failure, such as arterial hypertension, atheroma, dyslipidemia, diabetes, smoking or hyperuricemia, may be associated [19].

These unilateral forms, as described in our observation, account for 76% of cases and have a good prognosis [12,20]. Bilateral forms are rare and usually incompatible with life. They account for a quarter of fetal multicystic dysplasias, are more frequent in girls and are more often associated with oligohydramnios, extrarenal malformations and/or chromosomal abnormalities [2].

5. Therapeutic aspects

There is currently no consensus on monitoring, but some authors believe that it should be essentially ultrasound and clinical [14]. They recommend ultrasonography at birth, essentially to confirm the antenatal diagnosis, then at 1, 5, 10 and 15 years. Cystography and scintigraphy have no systematic indication, and should only be proposed in order to contribute to potential diagnostic and/or therapeutic orientations.

With regard to clinical and biological monitoring, some teams suggest measuring blood pressure, creatinine and microalbuminuria every 5 years [19,13].A systematic review of 29 prospective and retrospective cohorts involving 1115 children with unilateral MRHD under conservative management showed that the probability of developing hypertension in childhood was 5.4 per 1000

(95% confidence interval: 1.9-11.7) [21]. It is therefore necessary to explain to parents, and later to adolescents and adults, the basic nephroprotective measures that should be applied. These essentially involve avoiding nephrotoxic drugs such as non-steroidal anti-inflammatory drugs, smoking and other cardiovascular risk factors, by applying good hygienic and dietary rules. Any lower urinary tract infection must be treated systematically to avoid pyelonephritis, which could damage the single kidney [19]. This abstentionist attitude was adopted in our patient, who is currently progressing well.

Surgery and antibiotic prophylaxis should no longer be proposed as first-line treatment. The indication for surgery, once advocated, is now limited to symptomatic situations, such as the existence of compression, or when the diagnosis of DRMK is not formal and cannot be documented by imaging [14].

CONCLUSION:

Multi-cystic renal dysplasia (MCRD) is one of the most frequent congenital anomalies of the kidney.

In the African context, it is often diagnosed late in the post-natal period, and sometimes only in adulthood during the investigation of renal failure.

In order to improve prognosis in our region, we need to diagnose the condition earlier, through prenatal ultrasound screening and the introduction of nephroprotective measures.

Surgery is considered only in limited clinical situations, such as recurrent urinary tract infection, abdominal pain and sudden or progressive kidney enlargement.

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