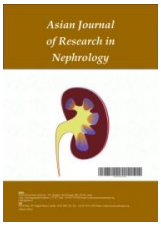

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Diagnostic Accuracy of Antenatal Ultrasound in Diagnosis of Multicystic Dysplastic Kidneys

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Objective: To establish the diagnostic accuracy of prenatal detection of unilateral and bilateral MCDK at our tertiary foetal medicine center and identify the association of MCDK with other renal or extra-renal abnormalities.

Methods: This was a retrospective observational study of all the MCDK cases diagnosed either antenatally or postnatally between January 2009 and December 2018 in our tertiary center.

Results: We identified 70 cases that were suspected or diagnosed with MCDK in the 10 year period. 11 cases were suspected to have bilateral disease antenatally opted for termination of pregnancy. However these could not be confirmed as post-mortem data was not available. 2 cases had miscarriages due to associated co-morbidities.

Correct antenatal diagnosis was made in 43 cases, 7 cases were false positive as alternative postnatal diagnosis was made and remaining 7 cases were found to have normal scans. Amongst false positive cases hydronephrosis was found in 3, Cortical Cysts in 2 cases and no abnormality in 2 cases postnatally. Associated renal or extra renal abnormalities were found in 13 patients.

Conclusions: The diagnostic accuracy of antenatal ultrasound in our center was 86% with a positive predictive value of 0.8. Associated anomalies were found in 19% of patients. This is comparable to published figures and can be used to guide antenatal counselling accurately.

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1. INTRODUCTION

Multicystic dysplastic kidney (MCDK) is defined as a variant of renal dysplasia with multiple non-communicating cysts separated by dysplastic parenchyma [1].

MCDK is one of the most common renal anomalies detected antenatally during ultrasound examination, with an incidence around 1 in 4300 live births [2] Most MCDK cases are unilateral, with the left kidney more commonly affected [3] Features of MCDK are characteristic, however they can be mistaken for other renal anomalies, in particular pelvicalyceal dilatation due to pelviureteric junction obstruction. [4] Bilateral MCDK is a lethal anomaly and is incompatible with life. The condition is usually detected at the mid-trimester scan; however certain reports suggested that it can be seen as early as 15 weeks of gestation [5].

Our aim is to identify the diagnostic accuracy of mid-trimester USS in our institution and evaluate the association of MCDK with associated anomalies and chromosomal abnormalities.

2. MATERIALS AND METHODS

This is a retrospective study of all patients diagnosed with MCDK in a 10 year period from 2009-2018 whether unilateral or bilateral. The midtrimester antenatal ultrasound scans performed by foetal medicine consultant was reviewed and compared to their post-natal scans.

3. RESULTS

We identified 70 cases with suspicion or a diagnosis of MCDK during the study period. Antenatally, 11 patients were suspected to have bilateral disease and decide to abort the foetus. Termination of pregnancy occurred in 2 cases due to other associated anomalies. Since post-mortem data was not available, these patients were excluded from the study.

Confirmed cases of MCDK amounted to 50 with 43 cases diagnosed correctly on antenatal scans while 7 cases were diagnosed postnatally. Right sided disease was found in 26 patients while 24 cases had left sided affection. Male to female ratio was 1.2:1. In the 7 remaining cases suspected with MCDK antenatally, hydronephrosis was found in 3, Cortical Cysts in 2 cases and no abnormality in 2 cases on their post-natal scans.

The diagnostic accuracy therefore in our study was 85% with a Positive predictive value of 0.8. In 13 patients, associated anomalies and chromosomal abnormalities were found. Extra-renal abnormalities found included VSD, ASD, Double-Outlet right ventricle, Anomalous Single Coronary, Cleft lip and Palate, duodenal atresia, Biliary atresia, Imperforate anus, oesophageal atresia and Skeletal abnormalities. Chromosomal abnormalities found included Chime Syndrome and Cri-Du-Chat.

4. DISCUSSION

The overall incidence of unilateral MCDK is estimated to be around 1 in 4300 births, with male predominance and left side is more frequently affected [3].

MCDK can be familial or as part of chromosomal anomalies [6,7]. Environmental factors such as use of antiepileptic drugs have been also associated with MCDK [8]. Mutations of the hepatocyte nuclear factor-1beta (HNF-1B) have also been incriminated in the development of the disease [9].

The most accepted theory for development of MCDK includes primary failure of renal mesenchyme induction during nephrogenesis [10] normally, between 6-8 weeks of gestation; the ureteric bud grows from the mesonephric duct into the meta-nephric blastema .The ureteric bud then branches and differentiates into the collecting ducts and ureter [11]. The failure of induction of nephrogenesis due to non-communication between ureteric bud and metanephric blastema has been proposed to cause MCDK [3].

In a recent systematic review, MCDK was found to be the only anomaly found in 71% of cases. Other renal or extra-renal malformations with MCDK is thought to be present in almost one in five cases (20%) [12,13] This is in line with our results where 19% of our patients had associated anomalies. The most common renal association reported is vesicoureteral reflux which is thought to occur in 30% of patients [3] Pelvi-ureteric obstruction, ureteroceles (in contralateral or ipsilateral side) and MCDK in horseshoe kidneys have also been described. [14,15,16] In our

cohort, Chromosomal abnormalities were found in two patients (3%). This is similar to the reported figure in current literature of 7%. [12] The high association of MCDK with both renal and extra-renal anomalies necessitates the importance of diagnosing this anomaly antenatally.

The classic features for MCDK on two-dimensional (2D) US are multiloculated intra-abdominal masses with multiple non-communicating cysts. Normal renal parenchyma can hardly be identified between cystic walls. The affected kidney is usually enlarged with irregular contour under ultrasonography. The criteria for the diagnosis of MCDK were proposed to include echogenic renal parenchyma, multiple non-communicating cysts with variable sizes at the periphery of the kidney, and no ultrasonic evidence of obstructive nephropathy [17] these signs provide a diagnostic accuracy of antenatal ultrasound between 53.3% and 100% [12]. The variation in the range can be attributed to the variation of individual operator skills together with the technology of Ultrasound machines used in different centers.

The development of 3D US have facilitated to accurately diagnose MCDK. Advantages include reconstruction of areas of interest that may be missed during 2D examinations, better visualisation of the severity of disease and decreasing the scanning time [18].

Some studies advocate the use of fetal MRI in such cases as it was found to add more diagnostic information and diagnose associated or alternate renal disease [19].

Outcomes of unilateral MCDK with a normal contralateral kidney are generally favourable with the natural history of the disease progressing to either complete or incomplete involution. Data from previous studies have shown that the rates of involution at follow-up ranging from 2.6-3.8 years ranges from 35% to 62%. [20,21,22] Complete involution increases with increasing duration of follow-up from 9.8% at 1 year to 53.5% at 10 years of age [23].

Hypertrophy of the contralateral kidney is thought to occur in 24-46% of patients. These patients are thought to have better glomerular filtration rate and creatinine clearance [3,24] On the other hand; glomerular hypertrophy can lead to hyper filtration injury and hypertension. Although the increased risk of hypertension in patients with MCDK is not clearly established, Studies show that hypertension resulting from hyper filtration injury, is significantly higher in children with a normal congenital solitary kidney [25]. This has led to the recommendation of routine blood pressure monitoring assessing for hypertension and proteinuria in patients with MCDK [26].

Besides cases of hypertension, patients may have contralateral renal anomalies (complex MCDK) and have been found to be at an increased risk of urinary tract infections [27] in our cohort, only one patient developed hypertension who is medically managed and four patients with complex MCDK have been suffering from UTIs. Risk of development of Wilm's tumour and renal cell carcinoma have also been described but considered to be low [26].

5. CONCLUSION

In our tertiary center, the diagnostic accuracy of antenatal US was 86%. This figure can be used to guide antenatal counselling. Special importance to assessment of co-existing abnormalities in all cases of MCDK should be given.

CONSENT

In suspected bilateral MCDK cases in women who opted for a termination of pregnancy, diagnosis could not be confirmed as parents did not consent for post-mortem examination.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s). This research study was conducted retrospectively from data obtained for clinical purposes. It is registered with the audit department in the Leicester Royal Infirmary Hospital.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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