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Autosomal dominant non-syndromic unilateral renal hypoplasia: A condition not previously reported in the literature

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Abstract
Background: Non-syndromic unilateral renal hypoplasia is a congenital condition associated with one small kidney (hypoplastic) that generally has normal residual parenchyma but smaller calyces. A transmission of unilateral renal hypoplasia from a parent to offspring has never been described in the literature.
Patients and Methods: A boy with unilateral renal hypoplasia whose mother also had the condition suggesting an autosomal dominant inheritance is described.
Results: A boy whose mother known to have unilateral renal hypoplasia was hospitalized at about the age of 18 months because of a gastrointestinal infection presented with vomiting, diarrhea, dehydration, and an elevated urea level of 86 mg/dl. Serum creatinine was 2 mg/ dl. Correction of dehydration resulted in rather rapid lowering of blood urea to normal. However, abdominal ultrasound showed right renal hypoplasia.
Conclusion: The first case of non-syndromic unilateral renal hypoplasia transmitted in an autosomal dominant fashion is reported.

Keywords: autosomal dominant, Non-syndromic, unilateral renal hypoplasia, Iraq

Introduction
Non-syndromic unilateral renal hypoplasia is a congenital condition associated with one small kidney (hypoplastic) that generally has normal residual parenchyma but smaller calyces. The size of the hypoplastic kidney is less than two standard deviations below the expected mean. Clinically, the condition may be asymptomatic or presented with infection tract infections and/or kidney stone formation. Diagnosis of unilateral renal hypoplasia is typically made with ultrasonography. A transmission of unilateral renal hypoplasia from a parent to offspring has never been described in the literature.

Patients and Methods
A boy with unilateral renal hypoplasia whose mother also had the condition suggesting an autosomal dominant inheritance is described.

Results
A boy whose mother known to have unilateral renal hypoplasia was hospitalized at about the age of 18 months because of a gastrointestinal infection presented with vomiting, diarrhea, dehydration, and an elevated urea level of 86 mg/dl. Serum creatinine was 2 mg/ dl. Correction of dehydration resulted in rather rapid lowering of blood urea to below 20mg/dL.After re-hydration all laboratory tests were normal. However, abdominal ultrasound showed unilateral renal hypoplasia. The left kidney (Figure-1A) was normal in size (71 x 37 mm) and position. The outlines, echogenicity, and the pelvi-calyceal system of the left kidney were also normal.

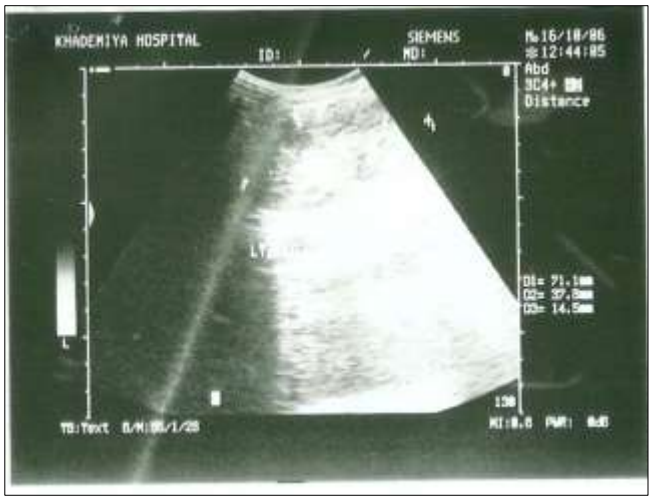

Fig 1A: The left kidney was normal in size and position

The right kidney (Figure-1B) was in normal position, but was hypoplastic, and its size was 38 x 20 mm. The cortical thickness of the right kidney was reduced (8 mm), but, its echogenicity was normal. The pelvi-calyceal system of the right kidney was also normal.
The patents of the mother and the father of didn't have any problem

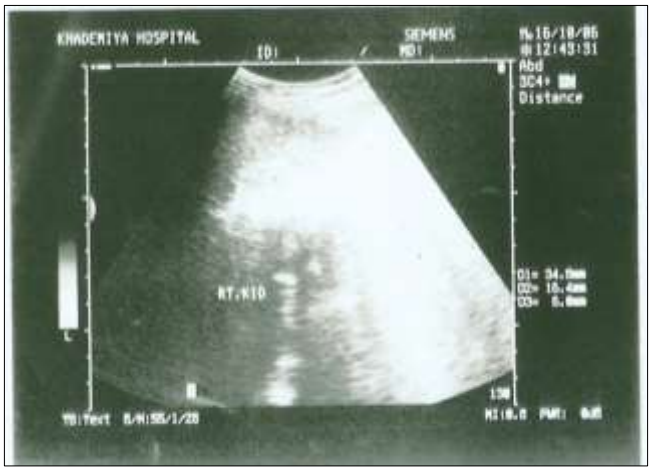

Fig 1B: The right kidney was in normal position, but was hypoplastic

Discussion

The occurrence of non-syndromic unilateral renal hypoplasia has been known early during the 1900s [1]. Taño Pino *et al*. (1996) reported two brothers, not twins with unilateral congenital renal hypoplasia with vesico-urethral reflux. In both, the diagnosis was made at about the age of 24 years and without any urological history, they presented with a dysuria-pollakiuria syndrome [2]. Cain *et al*. (2010) emphasized a genetic basis for the condition [3], but the transmission of the condition from a parent to offspring has never been reported. Autosomal dominant non-syndromic unilateral renal hypoplasia has not been reported in Iraq nor in any other country [2, 8].

Conclusion

The first case of non-syndromic unilateral renal hypoplasia transmitted in an autosomal dominant fashion is reported.

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