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ORIGINAL ARTICLE

# Ectopic pelvic kidney: Prenatal diagnosis and management

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## ABSTRACT

**Objective:** Ectopic pelvic kidney (EPK) is one of the most frequent renal anomalies detected in newborns as 1/500–700. Although it is generally asymptomatic, its association with recurrent urinary infections, vesicoureteral reflux, predisposition to stone formation, and genital anomalies has been shown. This study aimed to present the prenatal findings and postnatal outcomes of cases diagnosed with unilateral EPK.

**Material and Methods:** Twelve cases were recruited for this study between January 2018 and June 2020 in Van Training and Research Hospital. EPK diagnosis was achieved if the kidney was located in the pelvis, the separation of the kidney-specific cortex-medulla was present, and the renal pelvis was detected. EPK diagnosis was confirmed by renal USG in all cases postnatally. Long-term results of all cases were analyzed retrospectively from patients records. Statistical analysis was achieved by calculating the mean and standard deviation values with the Statistical Package for Social Sciences (SPSS 20), Chicago, USA.

**Results:** Mean gestational age at diagnosis week was 25.2±4.2 weeks. Left EPK was detected in 7 (58.3%) and right EPK in 5 (41.7%) cases. Pelvis dilatation was detected in EPK in 2 (16.6%) cases and the kidney versus 3 (25%) fetuses. An additional structural anomaly was observed in 1 (8.3%) case. A genital anomaly was not observed in any case during the prenatal period. The mean follow-up interval was 11.2±2.8 months. 7 (58.3%) were female, and 5 (41.7%) were male. Renal functions were normal in all cases. A total of 3 (25%) cases, including Grade-1 vesicoureteral reflux in one case, recurrent urinary infection in 1 case, and hypospadias in one case, anomaly associated with EPK was detected.

**Conclusion:** The presence of EPK should be investigated in all fetuses with an empty renal fossa in the prenatal period. All cases should be followed up after delivery in terms of accompanying anomalies.

**Keywords:** Anomaly, fetus, kidney, ultrasonography.

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## INTRODUCTION

Kidneys develop in three stages in fetal life, including pronephros, mesonephros, and metanephros. The metanephros is located in the pelvis in front of the sacral somites during the first 6 weeks of fetal life, but then it migrates to the pelvis and takes the final position by rotating 90°. This process is completed in the 8th week of pregnancy.[1] Ectopic pelvic kidney (EPK) occurs when migration stops earlier, and the kidney cannot reach its final location in the retroperitoneal renal fossa. A simple pelvic kidney is defined as the kidney located on the right side of the body but out of place. If the kidney migrates to the opposite side of the body, it is defined as a crossed ectopic kidney. Crossed ectopic kidneys may fuse with the normally located kidney or separate from it.[2]

Although the incidence of EPK varies between countries, it is estimated to occur with a frequency of 1/1000. In a study conducted in Taiwan and involving 132000 school-age children, it was detected in 1/5000.[3] Yüksel and Batukan examined 13,701 fetuses and found that its frequency was 1/571.[4]

EPK is seen quite frequently; however, it is rarely diagnosed in the prenatal period in low-risk patients. The main reason for this low detection rate is that the amniotic fluid is expected in these fetuses and usually occurs as an isolated anomaly. The first clue for the prenatal diagnosis of EPK is to demonstrate that one of the kidneys is not in its standard location. The finding of lying down in the adrenal gland and the absence of renal vessels in color Doppler lead us to diagnose the empty renal fossa. EPK is observed in 30–40% of fetuses diagnosed with empty renal fossa. EPK is often located in the midline anterior to the aortic bifurcation and above the bladder.

It is generally diagnosed at the end of the second trimester or in the third trimester. Although the postnatal prognosis is uneventful in most cases, EPK may rarely cause hydroureteronephrosis, recurrent urinary infections, and stone formation in these infants. It is associated with hypospadias and cryptorchidism in boys, vaginal and uterine anomalies in girls. This study aimed to present the prenatal findings and postnatal outcomes of 12 cases diagnosed with unilateral EPK.

## MATERIAL AND METHODS

The prenatal and postnatal features of the cases diagnosed between January 2018 and June 2020 in the Van Training and Research Hospital were analyzed retrospectively. The study was conducted under the Declaration of Helsinki. Ethics committee approval was obtained. All prenatal ultrasonography examinations were performed by a single perinatology specialist (G.T). In all cases, the examination was performed with convex 3.5–7.2 MHz transabdominal and transvaginal probes. (Voluson E8, GE. TX USA). Abdominal organs were examined in axial, sagittal and coronal planes. When a kidney could not be seen in the renal fossa, a lying down sign in the adrenal gland on that side and the absence of the renal artery on that side in colour Doppler were described (Fig. 1a). In cases with unilateral empty renal fossa, the fetal pelvis was examined in detail on the axial, coronal and sagittal planes to detect the presence of EPK. The diagnosis of EPK was confirmed by the location of the kidney in the pelvis, the kidney-specific cortex-medulla separation, and the presence of the renal pelvis (Fig. 1b). Then, EPK was investigated to reveal renal anomaly. Furthermore, the presence of hypertrophy and anomaly in the normally located contralateral kidney was examined. According to the gestational week, the diagnosis of hypertrophy was defined as the long axis of the kidney above the 95th percentile. Anteroposterior diameters of both EPK and contralateral renal pelvis were measured in the axial plane, and renal pelvis dilatation was diagnosed in <4 mm below 28 weeks and >7 mm above 28 weeks.[5]

Detailed anatomical examination and fetal echo were performed for the presence of accompanying anomaly in all fetuses. If there are no additional risk factors such as advanced maternal age and increased risk in screening tests in isolated EPK, karyotype analysis is not recommended. Karyotype analysis was performed in cases with additional structural anomalies. All fetuses were followed up with fortnightly.

Renal ultrasound (USG) confirmed the diagnosis of EPK in all cases after birth (Fig. 1c). All cases were examined in terms of associated genital anomalies. Renal USG was repeated in all cases 1–2 weeks after delivery, and voiding cystourethrography (VCUG), magnetic resonance imaging (MRI), or static kidney scintigraphy (DMSA) was performed in cases that detected a renal anomaly in EPK or normally located contralateral kidney. All children were investigated in terms of recurrent urinary infections for 12 months. Patients with creatinine values >95 percentile were considered high risk in abnormal kidney functions, and further investigation was planned.[6]

Statistical analysis was performed with SPSS 20, Chicago, USA. The mean and standard deviation values were calculated.

## RESULTS

Twelve cases diagnosed with EPK were included in this study. EPK was unilateral in all cases. The mean gestational age at diagnosis was 25.2±4.2 weeks. Left EPK was detected in 7 (58.3%) cases, and right EPK was detected in 5 (41.7%) cases. Pelvic dilatation was revealed in EPK in 2 (16.6%) fetuses. Renal hypertrophy was shown in the contralateral kidney in 4 (33.3%) cases. Pelvic dilatation was detected in the contralateral kidney in 3 (25%) fetuses, and the mean pelvic anterior-posterior diameter of these three fetuses was 8.8±2.2 mm. Amniotic fluid volume was normal in 91.6% of the cases. An additional structural anomaly was observed in only 1 (8.3%) case. In this fetus, mild bilateral ventriculomegaly (11 mm) was detected, and karyotype analysis was normal. During follow-up, mild ventriculomegaly regressed. No genital anomaly was observed in the prenatal period in any of the cases. The prenatal characteristics of the fetuses are summarized in Table 1.

Twelve children were followed up for an average of 11.2±2.8 months postnatally. Mean gestational age at delivery was 38.2±3.1 weeks, and mean birth weight was 3160±370 grams. 7 (58.3%) cases were female, and 5 (41.7%) cases were male. A renal anomaly was detected in 3 (25%) cases in EPK. The mean EPK pelvis diameter of these cases was 13.3±2.6 mm. VCUG was performed in these three children, and grade-1 vesicoureteral reflux was detected in 1 case. In 1 baby, the contralateral renal pelvis was measured 12 mm, but the renal pelvis diameter decreased to normal ranges. Antibiotic prophylaxis was required in 1 (8.3%) case due to recurrent urinary infections. Kidney stones were not observed in

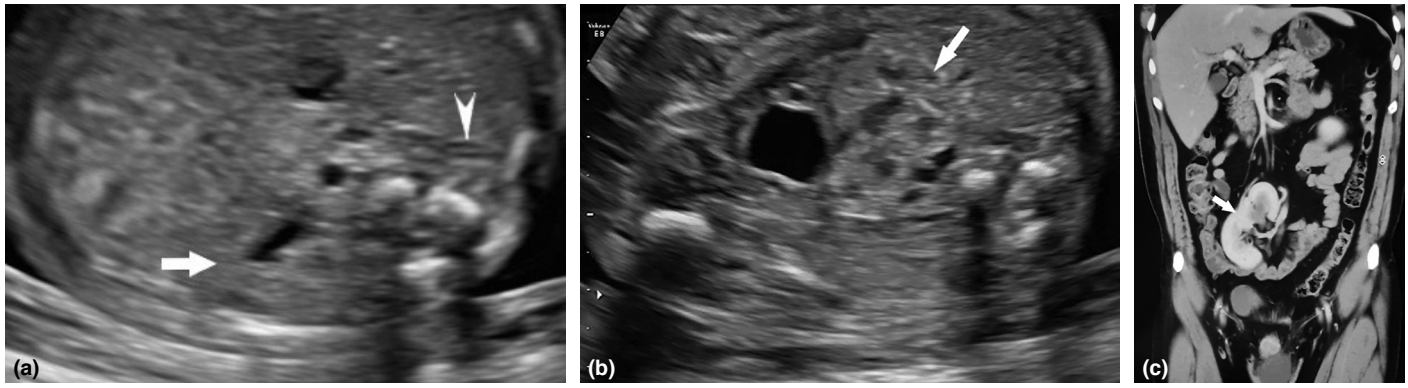


**Figure 1: (a)** Normal located kidney (arrow) and lying down sign with empty renal fossa (arrowhead) in ultrasonography. **(b)** Appearance of EPK in ultrasonography at the axial plane of the 22-weeks of gestation fetus (arrow). **(c)** Right EPK view in the coronal plane in computed tomography at 6th postnatal month (arrow).

EPK: Ectopic pelvic kidney.

**Table 1: Prenatal characteristics of 12 cases diagnosed with EPK**

| Prenatal findings | n=12 Mean±SD (%) |
|---|---|
| Gestational age at diagnosis | 25.2±4.2 |
| Location of EPK | |
| Left | 7 (58.3) |
| Right | 5 (41.7) |
| Anomaly in EPK | 2 (16.6) |
| Contralateral kidney hypertrophy | 4 (33.3) |
| Anomaly in the contralateral kidney | 3 (18.7) |
| Amniotic fluid volume | |
| Normal | 11 (91.6) |
| Oligohydramnios | 0 |
| Polihidramnios | 1 (8.3) |
| Extrarenal anomaly | 1 (8.3) |
| Genital anomaly | 0 |

SD: Standard deviation; EPK: Ectopic pelvic kidney.

**Table 2: Postnatal findings of 12 cases diagnosed with EPK**

| Postnatal findings | n=12 Mean±SD (%) |
|---|---|
| Follow-up interval (month) | 11.2±2.8 |
| Gestational age at delivery | 38.2±3.1 |
| Birthweight | 3160±370 |
| Route of delivery | |
| Vaginal | 9 (75) |
| Cesarean | 3 (25) |
| Sex | |
| Female | 7 (58.3) |
| Male | 5 (41.7) |
| Need for further examination (VCUG, DMSA, MR) | 4 (33) |
| Anomaly in EPK | 3 (25) |
| Anomaly in contralateral kidney | 1 (8.3) |
| Recurrent urinary infection | 1 (8.3) |
| Kidney stone | 0 |
| Genital anomaly | 1 (8.3) |
| Abnormal renal function | 0 |

SD: Standard deviation; EPK: Ectopic pelvic kidney; VCUG: Voiding cystourethrography; MRI: Magnetic resonance imaging; DMSA: Static kidney scintigraphy.

any of the cases. Hypospadias was detected in 1 (8.3%) cases and required an operation. Renal functions of all patients were normal during follow-up. A total of 3 (25%) cases, including Grade-1 vesicoureteral reflux in 1 case, recurrent urinary infection in one case, and hypospadias in one case, were found to have an accompanying problem with EPK during follow-up. The results of 12 children are shown in Table 2.

## DISCUSSION

Renal anomalies constitute the most common group after cardiac and central nervous system anomalies, but the rate of EPK among prenatally diagnosed renal anomalies is relatively low. The main reasons for this are that it is difficult to separate the kidney tissue from the intestines, especially in the second trimester. The EPK is partially hypoplasic and abnormal compared to the normal kidney. Yüksel and Batukan[4] found its frequency as 1/713 in a reference prenatal diagnosis center in our country. Many studies have reported lower incidence rates in prenatal series in the literature. Hill et al.[7] have screened 25000 fetuses, and they found the frequency of EPK is 1/2000. Meizner et al.[8] calculated the incidence of EPK as 1/100 in their series in which they screened 29000 fetuses. In postnatal studies, the frequency of EPK is considerably higher than in the prenatal series. Arena et al.[9] screened more than 32000 newborns, and they

found the frequency of EPK to be 1 in 500 and showed that only one of the three cases was diagnosed prenatally. The literature shows us that many EPK cases are missed in the prenatal period.

EPK is often an isolated anomaly; however, the frequency of other system anomalies increases. Yuksel and Batukan examined 36 fetuses diagnosed with EPK observed additional extrarenal anomalies in 10 fetuses. EPK was associated with a diaphragmatic hernia in two cases, microcephaly in one case, and hemivertebrae and truncus arteriosus in one case. The most frequent associated anomaly was the single umbilical artery.[4] We detected mild bilateral ventriculomegaly as an additional structural anomaly in one fetus. Detailed fetal anatomical examination and fetal echocardiography should be performed to detect other accompanying system anomalies in all fetuses with EPK.

Renal anomalies can be detected in both normal located kidneys and EPK. Yüksel and Batukan showed urogenital system anomalies in half of 36 cases. EPK hypoplasia and multicystic dysplastic formation in EPK were the most common urogenital anomalies. They also found concomitant anomalies, most commonly renal pelvis dilatation, in 30% of normally located contralateral kidneys.[4] We also observed structural renal anomalies in EPK in two fetuses and in the contralateral kidney in three cases in our series. Both EPK and contralateral kidney must be followed up until delivery in all fetuses.

Isolated EPK cases are not associated with aneuploidies, and karyotype analysis is not recommended. In recent years, prenatal chromosomal microarray analysis has been widely used instead of conventional karyotyping. This technique can detect minor chromosomal changes that cannot be detected in standard karyotyping. Sagi-Dain et al.[10] performed chromosomal microarray in 220 fetuses diagnosed with isolated EPK and observed pathological submicroscopic chromosomal changes in two cases. We did not perform karyotype analysis in any of the fetuses in our series.

The first step examination of newborns diagnosed with EPK in the prenatal period is to confirm the diagnosis by pelvic ultrasonography and evaluate the contralateral kidney's structure. Serial follow-ups are recommended in cases without hydronephrosis and with normal creatinine values. If there is an abnormal finding in these first-line examinations, further tests such as VCUG, MRI, or DMSA are required.[11] In our series, prenatal diagnosis was confirmed in all cases, and advanced examinations were required in 33% of the children.

EPK is rarely associated with renal dysfunction in childhood, but complications may occur, primarily due to changes in the ureteral anatomy of EPK. The most common problem is vesicoureteral reflux due to the ureter of EPK entering the pelvis higher instead of the expected location and dilatation of the ureter. Calisti et al.[12] examined 50 children with isolated EPK and found the frequency of vesicoureteral reflux significantly higher than the normal population. Gleason et al.[13] showed the frequency of vesicoureteral reflux was 17% in their series of 77 patients. The frequency of kidney stones in EPK cases is unknown, but it is thought to be more common than a normally located kidney.[14] Moreover, hypospadias and cryptorchidism are seen more frequently in boys, and vaginal agenesis and uterine anomalies in girls.[15] In our series, there was vesicoureteral reflux in 1 case and hypospadias that could not be diagnosed in the prenatal period in one child.

## CONCLUSION

EPK is one of the most common renal anomalies, and its presence should be investigated when one of the kidneys is not seen in the routine fetal examination. Although the overall prognosis is excellent, the frequency of hydronephrosis, recurrent urinary infections, and stones in these kidneys increases, and genital anomalies may accompany.

### Statement

**Ethics Committee Approval:** The Van Training and Research Hospital Clinical Research Ethics Committee granted approval for this study (date: 20.05.2021, number: 2021/10).

**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept – GT; Design – GT; Supervision – BÇ; Resource – BÇ; Materials – GT; Data Collection and/or Processing – BÇ; Analysis and/or Interpretation – GT; Literature Search – GT; Writing – GT; Critical Reviews – GT.

**Conflict of Interest:** The authors have no conflict of interest to declare.

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