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## RARE RENAL ECTOPIA IN CHILDREN – INTRATHORACIC ECTOPIC KIDNEY

### RZADKI PRZYPADEK EKTOPII NEREK U DZIECI – NERKA W KLATCE PIERSIOWEJ

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#### Abstract

*Renal ectopia occurs in 1:3000-1:7000 children. Simple ectopia refers to the abnormal kidney location on the same side; in crossed ectopia the ureter crosses the midline of the body. In most cases ectopic kidney is found in the pelvis minor, in 5% of cases in the thoracic cavity. Kidney displacement does not give any clinical symptoms in the majority of patients, sometimes it may cause abdominal pain, urinary tract infection, chest pain or promote nephrolithiasis. Renal ectopia is usually discovered during routine abdominal ultrasound. We describe a boy with prenatal ultrasound diagnosis of left kidney agenesis. Right normal kidney and displaced to the thoracic cavity, smaller, with normal echostructure left kidney were found in postnatal ultrasound. Left kidney ectopia was confirmed in renal scintigraphy, radiography and computed tomography of the chest. Voiding cystourethrography (VCUG) excluded vesicoureteral reflux. Conclusions: In case of kidney's absence in typical localization in the abdominal ultrasonography, the ectopic kidney should be suspected. Diagnosis of ectopic kidney requires further additional imaging examination to check renal function and other urinary tract anomalies.*

**Keywords:** renal ectopia, crossed renal ectopia, thoracic kidney, scintigraphy, CAKUT

#### Streszczenie

*Ektopia nerek występuje z częstością 1:3000-1:7000 dzieci. Ektopia prosta dotyczy nieprawidłowego położenia nerki po tej samej stronie; w ektopii skrzyżowanej moczowód przecina linię pośrodkową ciała. Nerka ektopowa położona jest najczęściej w miednicy małej, w 5% przypadków leży w klatce piersiowej. Przemieszczenie nerki u większości dzieci nie daje objawów klinicznych, czasami nerka ektopowa może powodować bóle brzucha, zakażenia układu moczowego lub sprzyjać wystąpieniu kamicy. Ektopia nerki wykrywana jest najczęściej w rutynowych badaniach ultrasonograficznych jamy brzusznej. Przedstawiamy przypadek chłopca, u którego na podstawie prenatalnych badań ultrasonograficznych rozpoznano agenezję nerki lewej. W badaniach ultrasonograficznych jamy brzusznej wykonanych po urodzeniu stwierdzono prawidłową nerkę prawą i przemieszczoną do klatki piersiowej nerkę lewą, mniejszą od prawej, o prawidłowej echostrukturze. Ektopię potwierdzono scyntygrafią nerek, badaniem radiologicznym klatki piersiowej i tomografią komputerową klatki piersiowej. W cystografii mikcyjnej wykluczono odpływy pęcherzowo-moczowodowe. Wnioski: W przypadku braku nerki w miejscu typowym w badaniu ultrasonograficznym należy poszukiwać nerki ektopowej. Obecność nerki ektopowej wymaga wykonania dodatkowych badań obrazowych w celu oceny jej funkcji i towarzyszących wad układu moczowego.*

**Słowa kluczowe:** ektopia nerki, skrzyżowana ektopia nerki, nerka w klatce piersiowej, scyntygrafia, CAKUT

## BACKGROUND

Anatomically, human kidneys are located in abdomen, retroperitoneal, on the level of two last thoracic vertebrae (Th11-Th12) and first three lumbar vertebrae (L1-L3). Left kidney is generally placed higher than the right one. It measures about 10-12 cm in adults, average weight is 120-200 g [1]. Kidneys exhibit respiratory mobility. Renal ectopia means the displacement of kidney from its normal lumbar location. It is connected with the abnormal prenatal kidneys migration from sacral spine to the lumbar region, which appears about 5-9 week of gestation. Ectopia may touch one or both kidneys. In the simple ectopia, kidney is placed on the same side of body in the pelvis minor or in the chest. In crossed ectopia, kidney is placed on the opposite side of the body and the ureter crosses the midline of the body and reaches bladder in the normal place. In 90% of crossed renal ectopia kidneys are fused. About 60% of ectopic kidneys are placed in pelvis minor, 35% paravertebrally, 5% in thoracic cavity [2]. The incidence of ectopic kidney in pelvis minor is 1:3000, crossed renal ectopia 1:7000, the solitary kidney in pelvis minor 1:22 000 births [3]. In most cases of ectopic kidney does not give any clinical symptoms. However, the abnormal position of kidney and ureters can cause urinary tract infection, nephrolithiasis, abdominal pain, chest pain, tachypnoe or hypertension [4, 5]. Renal ectopia may appear in children with trisomy of 21 or 18 chromosome, Turner syndrome, Beckwith-Wiedemann syndrome, be part of VACTERL or CHARGE sequence [6].

## CASE REPORT

The boy was born at term, with birth weight 3340g, in good general condition. Agenesis of left kidney was diagnosed in prenatal ultrasonography. Postnatal abdominal ultrasound showed correctly placed right kidney (55 mm) and displaced to the chest left kidney (35 mm), both with normal echostructure. The control abdominal and chest ultrasound performed at the age of three years showed the normal right kidney of 76mm and hypoplastic left kidney in the thoracic cavity, which measured 57 mm (Figure 1).

Renal scintigraphy DMSA described physiologically placed right kidney and intrathoracic left kidney. Static scintigraphy (DMSA) revealed normal function of right kidney (58%), and decreased function of the left kidney (42%). Voiding cystourethrography (VCUG) excluded vesicoureteral reflux. Four years later the chest radiography was done, because of the suspicion of pneumonia. It showed a shadow of 70x30 mm placed over the diaphragm, which could correspond to the left ectopic kidney (Figure 2). Because of this suspicious mass in the chest, the computed tomography was performed and it confirmed the ectopia of left kidney located in the thoracic cavity (Figure 3). During the seven-year follow-up arterial blood pressure, kidney function and urinalysis are normal.

## DISCUSSION

Congenital abnormalities of kidney and urinary tract (CAKUT) are one of the most common malformations in

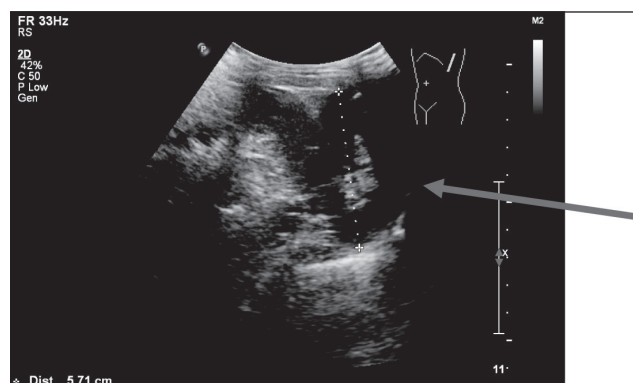


Fig. 1. Ultrasonography of the chest; arrows shows thoracic ectopic kidney.

Ryc. 1. Badanie ultrasonograficzne klatki piersiowej; strzałka wskazuje nerkę ektopową w klatce piersiowej.

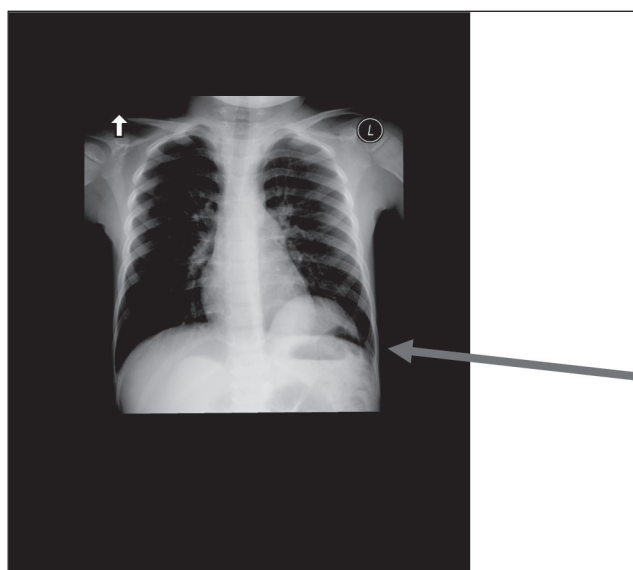


Fig. 2. X-ray of the chest in patient with intrathoracic ectopic kidney.

Ryc. 2. Zdjęcie radiologiczne klatki piersiowej u pacjenta z ektapią nerki w klatce piersiowej.

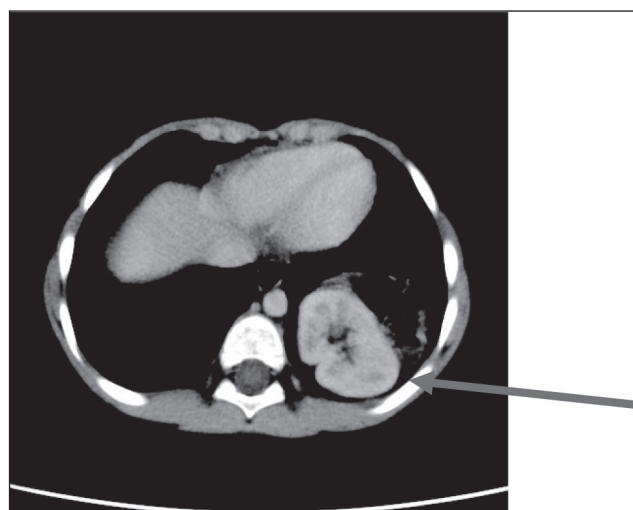


Fig. 3. CT of the chest, arrows shows ectopic kidney.

Ryc. 3. Tomografia komputerowa; strzałka wskazuje nerkę ektopową w klatce piersiowej.

children. Kidneys reach the final correct position in the early stages of embryogenesis, by the 9<sup>th</sup> week of gestation. Prenatally, there are a lot of risk factors such as genetic, teratogenic which may disturb the physiological development of kidneys and lead to congenital defects. Absence of kidney in its typical location in the ultrasound may be associated with agenesis or ectopia in a different location, most often in pelvis minor. Intrathoracic ectopic kidney is a rare kidney abnormality. The incidence of intrathoracic ectopic kidney in autopsies was rated 1:13 000 (Campbell, 1930). It occurs more often in males and on the left side of the body. Renal function is usually correct, proportional to the kidney's size. Ectopic kidney is generally discovered accidentally during routine abdominal ultrasounds [7]. In the literature, one can find descriptions of patients with ectopic intrathoracic kidney diagnosed in the adulthood [8]. In every case, when the kidney cannot be found in its typical location in ultrasound, it is necessary to look for it outside the lumbar area. Most commonly, ectopic kidney is placed in pelvis minor, but it is highly recommended to always consider the thoracic location during the diagnosis. On the other hand, seeing the mass in the chest, the ectopic kidney should be taken in account in the differential diagnosis. Not only ultrasound may be useful in the diagnosis of renal ectopia, but also renal scintigraphy and voiding cystourethrography may apply. In case of absence of kidney in its typical location, renal scintigraphy can detect the ectopic kidney. Scintigraphy does not measure accurately the share in filtration of ectopic and normal kidney. The reason of this difficulty is the difference in distance kidneys from gamma camera. Ectopic kidney is usually rotated and placed deeper than the normal one, so it may understate its share in filtration [9]. In some cases computed tomography is performed to confirm the diagnosis. In this case, the ultrasound was repeated after birth because of the prenatal suspicion of renal agenesis and it showed the left kidney's displacement to the thoracic cavity. Renal scintigraphy and computed tomography confirmed this ectopia. In the ultrasound examination, the ectopic kidney was smaller than the normal one, but its functioning was correct and proportional to size. Voiding cystourethrography excluded the presence of vesicoureteral reflux. Routine chest radiography proceed because of the pneumonia exposed suspicious mass in chest, which proved to be the ectopic kidney. During the long observation, the boy did not report any complaints and his psychomotor development is correct.

## CONCLUSIONS

1. In case of kidney's absence in its typical location in the ultrasonography, an ectopic kidney should be sought.
2. Ectopic kidney needs further additional imaging examination to check the renal function and other urinary tract abnormalities.

## REFERENCES

1. Dudek RW. High-Yield Kidney. LWW 2007, Chapter 2.
2. Jhun BW, Lee K-J. A right thoracic kidney with a pulmonary vascular malformation in an adult. *Respirol Case Rep.* 2013;1(2):55-57.
3. Cinman NM, Okeke Z, Smith AD. Pelvic kidney: associated disease and treatment. *J Endourol.* 2007;21 (8):836-842.
4. Fadaii A, Rezaian S, Tojari F. Intrathoracic kidney presented with chest pain. *Iran J Kidney Dis.* 2008;2(3):160-162.
5. Sesia SB, Haecker FM. Late-presenting diaphragmatic hernia associated with intrathoracic kidney: tachypnoea as unique clinical sign. *BMJ Case Rep.* 2012 Apr 23;2012.
6. Stein JP, Kurzrock EA, Freeman JA, Esrig D, Ginsberg DA, Grossfeld GD, Hardy BE. Right intrathoracic renal ectopia: a case report and review of the literature. *Tech Urol.* 1999 Sep;5(3):166-168.
7. Sumner TE, Volberg FM, Smolen PM. Intrathoracic kidney – diagnosis by ultrasound. *Pediatr Radiol.* 1982;12(2):78-80.
8. Liddell R, Rosenbaum D, Blumhagen J. Delayed radiologic appearance of bilateral thoracic ectopic kidneys, *AJR* 1989;152:120-122.
9. Allen D, Bultitude MF, Nunan T, Glass JM. Misinterpretation of radioisotope imaging in pelvic kidneys. *Int J Clin Pract Suppl.* 2005 Apr;(147):111-112.

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### Authors' contributions/Wkład Autorów

According to the order of the Authorship/Według kolejności

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