

## Ultra-mini percutaneous nephrolithotomy surgery in a pediatric patient with osteogenesis imperfecta

Osteogenesis imperfecta tanılı pediatrik hastada ultra-mini perkütan nefrolitotomi

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### Özet

Osteogenesis imperfecta (OI), genetik ve klinik çeşitlilik gösteren nadir, kalıtsal bir hastalık grubudur. Tip 1 kollajende defekt vardır, kemik fragilitesinde artış, osteopeni ve iskelet deformiteleri görülebilmektedir. OI hastalarında cerrahi uygulamalar, kemik yapılarında yaralanma ve kırık gibi komplikasyonlara neden olabilir. Bu nedenle pediatrik ürolitiazis yönetiminde girişim kararı dikkatle alınmalı, güvenli ve işlem süresini kısaltacak teknikler tercih edilmelidir.

Perkütan nefrolitotomi (PCNL) operasyonu erişkin hastalarda büyük böbrek taşlarında ilk tercih olarak kullanılmaktadır. Teknolojik gelişmelerle birlikte mini-PCNL, ultra mini-PCNL ve mikro-PCNL gibi daha minimal invaziv yaklaşımlar ortaya çıkmıştır ve bu yöntemler düşük morbiditeleri sayesinde çocuk hastalarda başarıyla kullanılmaktadır.

Olası riskleri nedeniyle literatürde OI hastalarına yönelik üriner taş cerrahisi nadir olarak bildirilmiştir. Bu raporda böbrek taşına ultra mini-PCNL uyguladığımız OI tanılı 4 yaş 4 aylık çocuk hasta sunulmuştur.

**Anahtar Kelimeler:** endouroloji, ürolitiazis, pediatrik üroloji, ultra mini pcnl

### Abstract

Osteogenesis imperfecta (OI) is a rare, inherited disease group with genetic and clinical diversity. There is a defect in the type 1 collagen structure. Increasing bone fragility, osteopenia, and skeletal deformities can be seen. Surgical applications in patients with OI may cause complications such as spinous process injury in vertebrates or bone fracture. Therefore, in the management of pediatric urolithiasis, the decision of surgery should be carefully considered, and safe techniques that shorten the procedure time should be preferred.

Percutaneous nephrolithotomy (PCNL) operation is the first choice for large kidney stones in adult patients. Along with the technological advances, more minimally invasive approaches such as mini-PCNL, ultra-mini-PCNL, and micro-PCNL have been developed, and these methods are safely used in pediatric patients due to their low morbidity.

Because of its potential risks, urinary stone surgery in OI patients has rarely been reported in the literature. This study presents a 4-year-4-month-old girl with OI and kidney stone, whom we performed ultra-mini-PCNL.

**Keywords:** endourology, urolithiasis, ultra mini pcnl, pediatric urology

## INTRODUCTION

Osteogenesis Imperfecta (OI) is a rare, inherited group of diseases with genetic and clinical diversity. There is a defect in type 1 collagen, and it is seen with a frequency of 1/10000 to 1/20000. Increased bone fragility, osteopenia, skeletal deformities, short stature, blue sclera, and early age deafness may develop (1). When surgery is required in these patients, complications such as hyperthermia, injury to the cervical vertebra, tooth and bone fractures may occur due to position and anesthesia applications. The choice of surgical technique is of great importance, especially in the management of pediatric urolithiasis (2).

Percutaneous nephrolithotomy (PCNL) is a procedure that has been used successfully in adults. However, with technological advances, more minimally invasive methods such as mini-PCNL, ultra-mini-PCNL, and micro-PCNL have emerged and are used safely in pediatric patients (3).

There are very few reports in the literature about urinary stone surgery on OI patients. In this study, a pediatric patient has been diagnosed with OI and has a kidney stone, whom we performed ultra-mini-PCNL is presented.

## CASE REPORT

A 4-year-4-month-old girl presented to our clinic with right flank pain. She had no known disease and urological history other than OI. Physical examination findings were unremarkable, and laboratory tests were within the normal range for age. Urinary ultrasonography detected a stone in the right renal pelvis. Intravenous pyelography (IVP) was applied to evaluate the urinary system anatomy and stone location more clearly. In imaging, the left kidney and collecting system were normal. The right kidney was larger than normal, and its collecting system was dilated, with approximately 15 mm of stone in the renal pelvis (Figure 1).

Ultra mini-PCNL was decided due to the large stone load. General anesthesia was given to the patient. In the lithotomy position, the right ureter was catheterized with a 4 fr open-end ureteral catheter, and then the patient was turned to the prone position (Figure 2). Access was obtained to the right kidney collecting system from the middle calyx with C-arm fluoroscopy (Figure 3), and a 13 fr sheath was placed. After holmium laser lithotripsy in the renal pelvis with a 7.5 fr nephroscope,

a 3 fr 14 cm double-J catheter was placed into the right ureter from the inside access sheath. Finally, an 8 fr nephrostomy tube was placed into the kidney.

It was seen that the patient was stone-free on USG and x-ray imaging at 3 months post-op (Figure-4a and 4b).

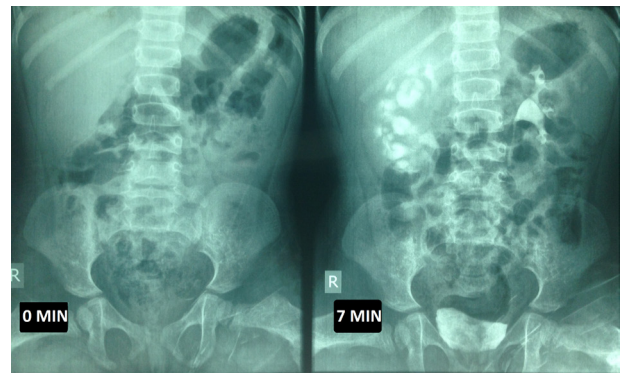


Figure 1. Intravenous Pyelography (0 Min and 7 Min)



Figure 2. Patient In Prone Position

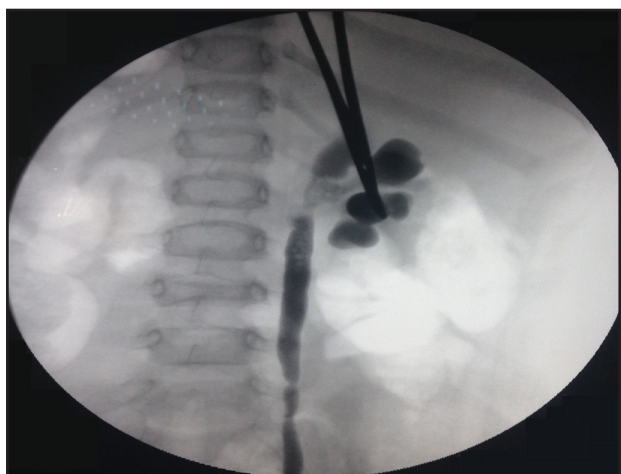
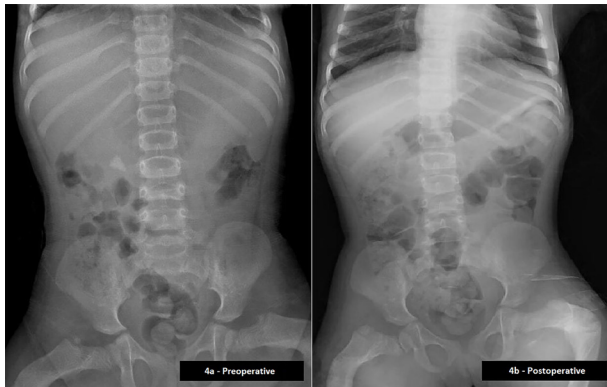


Figure 3. Right Kidney Collecting System Under Fluoroscopy



**Figure 4a and 4b.** Preoperative (4a) and Postoperative Third Month (4b) X-ray Image

## DISCUSSION

OI may affect the urinary passage by causing anatomical deformities at the outlet of the bony pelvis, increasing the risk of stone development (4). However, the information about this in the literature is limited. In two different series, the incidence of stones in pediatric patients with OI was 4.7% and 6.9%, which is similar to the general population (5, 6). In a study, 47 OI patients were evaluated, the rate of hypercalciuria was found at 36%, but there was no increased stone risk (7).

In OI, SWL use has not been reported due to fracture risk. Flexible renoscopy, PCNL, or open surgery are alternative approaches, and PCNL has been successfully used in these patients (8, 9).

PCNL surgery was first described in 1976, and traditionally, 26-32 fr width access is obtained. In the course of time, new techniques have been developed to reduce morbidity, and the mini-PCNL (11-20 fr) was performed for the first time in the pediatric group in 1998. Afterwards, micro-PCNL (4.85 fr) was defined in 2011 and ultra mini-PCNL (11-13 fr) in 2013 (10). In this case, we obtained 13 fr access and used the ultra-mini-PCNL technique with 7.5 fr nephroscope.

In percutaneous stone surgeries performed in the prone position, caution should be exercised when turning from the lithotomy position to the prone position. Complications such as injury or fracture of bone structures associated with general anesthesia and surgical position have been reported in OI patients (2). That is why extra care should be taken, especially in bone fracture predisposition such as in OI patients.

## CONCLUSION

We think that preoperative planning is important in cases with comorbid diseases that may increase operational risk. Stone surgery should be aimed to be completed in a little while and in one session; therefore, the surgical technique and anesthesia application should be preferred accordingly. Low morbidity methods such as ultra-mini-PCNL can be successfully performed in experienced centers.

## Conflict of Interest

All authors declared that there is no conflict of interest.

## Financial Disclosure

The authors declared that this study has received no financial support.

## Author Contributions

Conception and design; BK, Data acquisition; AO, SÇ, ECB, Data analysis and interpretation; SÇ, ECB, Drafting the manuscript; AO, Critical revision of the manuscript for scientific and factual content; SÇ, ECB, Supervision; BK

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