Case report

**Robot-assisted pyeloplasty and pyelolithotomy in patient with osteogenesis imperfecta**

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**Abstract**

Robot-assisted pyeloplasty is currently the technique of choice for cases of pyelo-ureteral junction syndrome. It may be accompanied by pyelolithotomy in cases of associated lithiasis. The presence of body abnormalities, as in the rare case of osteogenesis imperfecta, can make the procedure more difficult, starting from a different way of positioning the robotic ports, therefore we present a case report that can be a guide for similar cases.

**Keywords:** robotic-assisted pyeloplasty, pyelolithotomy, osteogenesis imperfecta,

**Introduction**

The pyelo-ureteral junction syndrome is a condition that includes various anomalies that cause hydronephrosis due to a defect in the passage of urine in the junction between the renal pelvis and the ureter [1]. A frequent complication is the urinary stone formation [2]. Indications for surgery include decreased split renal function, poor drainage function after furosemide administration, increased anteroposterior diameter at ultrasound and grade III and IV dilation. The advantages of conventional laparoscopy compared to open surgery are reduced length of hospital stay, better aesthetics, less postoperative pain and earlier recovery [3]. Robotic-assisted laparoscopic pyeloplasty has the same advantages as laparoscopic pyeloplasty but also greater ease in performing the procedure, so shorter operating times [4]. Osteogenesis imperfecta is a condition which consists in bone fragility and malformations accompanied by short height [5]. We present the case of a patient suffering from Osteogenesis imperfecta and affected by pyeloureteral junction syndrome, complicated with urinary stone formation and successfully treated with a Robotic-assisted pyeloplasty and pyelolithotomy.

**Case Presentation**

The patient was 42 years old, 110 cm tall and weighs 34 kg. He was suffering a form of osteogenesis imperfecta with marked kyphoscoliosis and barrel chest. After pain on the right side of the abdomen and ultrasound detection of consensual hydronephrosis he was subjected to abdominal CT showing hydronephrosis and multiple lithiasis of the renal pelvis. In the suspicion of pielo-ureteral junction syndrome he performed diuretic renal scintigraphy which showed defect of emptying the right renal pelvis even after diuretic stimulus but right kidney function preserved. For the presence of stones and flank pain a DJ stent was positioned. Following an unsuccessful attempt to replace the DJ stent, a percutaneous nephrostomy was placed. After about 3 months the patient underwent robotic pyeloplasty an pyelolithotomy described as below:

Patient in lateral decubitus on the left side. Suprumbilical incision in the right pararectal, open access to the peritoneal cavity, introduction of trocar for airseal and induction of pneumoperitoneum at 12 mmhg ; positioning of only three 8 mm robotic ports in the right pararectal line at a distance of 5-6 cm between them, for anatomical deformities and small abdomen; placement of two 5 mm service accessory ports in the pararectal line on the suprumbilical and subxiphoid level; docking of the DaVinci system.; incision of the right parietocolic shower, medialization of the colon and duodenum; placement of a grasper anchored to the abdominal wall for liver lift; identification of the ureter and isolation of the same in the subjunctional portion; isolation of the right renal pelvis that appears very dilated; section of the stenotic junction tract of the ureter that is placed in endobag along with about 10 stones (about 4-5 mm in diameter each) removed from the renal pelvis; placement of ureteral stent 16 cm x 6 ch on guide wire; pieloplasty suture with detached monofilament 3-0 points; anastomosis sealing control with injection of saline solution from nephrostomy; a laminar drainage was placed in front seat through the most caudal robotic port.

The operating time was 130 minutes. With the exception of hyperpyrexia, successfully treated with antibiotic therapy, the clinical postoperative course was normal. The patient was discharged after nephrostomy removal. The Ureteral stent was removed about 1 month after surgery. After 6 months after surgery the patient was in good condition, without flank pain, with normal renal function and urine tests.

**Discussion**

The robot-assisted pyeloplasty is the surgery management of uretero-pelvic junction (UPJ) syndrome and it can be simultaneous accompanied by pyelolithotomy in stone formation cases [6]. The robotic surgical approach typically involves the use of multiple robotic ports. Generally, using DaVinci system, robotic ports should be positioned at a distance of about 8 cm among them, but in this case the anatomic abnormalities led us to reduce this distance and the number of ports for lack of enough abdominal space.

In addition, when performing the right UPJ pyeloplasty, the presence of the liver can make the procedure more difficult, so you can make the surgery easier using a clamp laparoscopic grasper positioned below the liver that can pull the liver upwards, away from the kidney [7]. This also happened in our procedure where it was necessary to use this clamp to avoid the involvement of one of the only two operative robotic arms. Robot-assisted laparoscopic pyeloplasty is the procedure of choice for its greater safety profile than open surgery [8]. We were also able to perform the simultaneous treatment of renal stones, reducing the morbidity associated with a double surgical treatment. The feasibility of this procedure even in a patient with these anatomical abnormalities allows to make it a first choice approach even in these conditions.

**Conclusion**

There are not similar cases reported in patients with osteogenesis imperfecta, treated as before illustrated. As before shown the specific robotic technique was adapted to the patient’s case for his anatomical deformities. In the way it was described this case represents a good robotic surgery management model.

**Supplement Video:** https://ojs.oss-cn-hongkong.aliyuncs.com/journal/UTJ/Video/UTJ%20video-731.mp4

**Declaration**

**Availability of data and materials** The data used during the current study available from the corresponding author on reasonable request.

**Conflict of interest** None of the contributing authors have any conflict of interest, including specific financial interests and relationships and affiliations relevant to the subject matter or materials discussed in the manuscript.

**Ethical Approval and Informed consent**: Ethical approval and informed consent: The patient has provided an informed consent for publication of images and information in this study

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