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Direttore Nicola Picardi

# A giant inguinal hernia in a patient with Ehlers-Danlos syndrome



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Gennaro Quarto, Anna D'Amore, Sara Vertaldi, Pietro Anoldo, Giacomo Benassai, Giovanni Domenico De Palma, Michele Manigrasso, Marco Milone

Department of Clinical Medicine and Surgery, University "Federico II" of Naples, Naples, Italy

## A giant inguinal hernia in a patient with Ehlers-Danlos syndrome.

BACKGROUND: Inguinal hernia formation is a common event in patients with Ehlers-Danlos syndrome. Minimally invasive surgical technique for inguinal hernia repair is the same used in patients without EDS but it is related to more intraoperative and postoperative complications.

AIM: Inour study, we present a case of inguinal hernia in a EDS patient successfully treated with a robotic transabdominal preperitoneal procedure (TAPP procedure).

MATERIAL AND METHODS: We decided to perform a robotic TAPP with the DaVinci Xi<sup>®</sup> platform (Intuitive Surgical, Sunnyvale, USA) under general anaesthesia. A robotic docking was performed and three arms were positioned in the abdomen. Total operative timing was 45 mins.

RESULTS: During the robotic procedure no intraoperative complications were recorded and no drains were applied. The postoperative period was uneventful and the patient was discharged in the first postoperative day.

DISCUSSION: Inguinal hernia occurs more frequently in patients with EDS, mainly men. Many surgeons believe that EDS may have a negative effect on the clinical outcome of hernioplasty because of postoperative complication and recurrence rates. Our strategy has been robotic technology to facilitate the surgical approach.

CONCLUSION: Robotic technology is feasible and associated with a shorted recovery and better cosmetic results. The endowrist movement of the robotic arms allows wide instrument articulation in a confined space, bypassing the limitis of laparoscopic instruments. In this way, it is possible to realize an accurate dissection of important elements, to reduce operative timing and intraoperative and postoperative complications.

KEY WORDS: Ehlers-Danlos syndrome, Inguinal hernia, TAPP

#### Background

Inguinal hernia formation is a common event in patients with Ehlers-Danlos syndrome, mainly in men.

Minimally invasive approach to inguinal hernia repair is the same used in patients without EDS but it is related to more intraoperative and postoperative complications. In our study, we present a case of inguinal hernia in a EDS patient treated successfully with a robotic transabdominal preperitoneal procedure (TAPP procedure).

### **Case** Presentation

A 61-year old male with EDS presents a left inguinal hernia since six years. His skin has been elastic since infancy and he has been able to hyperextend it. His joints has shown more than the normal range of movement since babyhood. Furthermore, he suffers from heart failure, coronary artery disease, varicose veins, hypertension, hypercholesterolemia and diabetes mellitus.

The patient previously underwent stripping of varicose veins of the lower limbs, according to current literature<sup>1</sup>.

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Correspondence to: Prof. Gennaro Quarto, Department of Clinical Medicine and Surgery, University of Naples "Federico II", Via Sergio Pansini, 5, 80131, Naples, Italy (e-mail: gquarto@unina.it)



Fig. 1: Giant inguinal hernia (the laxity of the patient's skin is evident in the photo).

Technical developments in lower limbs varicose veins treatment, endovascular and combined procedures, have led to less invasive approaches, also with the aim to reduce the recurrence rate. Recurrences treatment is enriched in more recent years with innovative procedures. According to the patient, in left inguinal region, a bulge, engaging the scrotum, had gradually developed over a 6-year period.

The voluminous bulge caused physically and cosmetically discomfort to the patient with marked pain in inguinal region (Fig. 1).

The swelling increased on straining and coughing, it did not disappeare spontaneously or with Taxis maneuver.

Thus, the patient was hospitalized and we decided to perform a robotic transabdominal preperitoneal procedure (TAPP procedure).

Perioperative antiplatelet drugs were administrated according to current literature <sup>2</sup> and elastic bandage of the lower limbs was made.

The procedure was performed with the DaVinci Xi<sup>®</sup> platform (Intuitive Surgical, Sunnyvale, USA) under general anaesthesia with the patient in supine position and in Trendelenburg position, just slightly. Anaesthesia management of these minimally invasive techniques is very different and challenging from open technique <sup>3</sup>. The obtainment of an adequate anaesthesia and the postoperative pain control are really important to patient and in outpatients hernia repair (Lichtestein technique) they could be achieved with the combination of TAP block and local anaesthesia, according to current literature<sup>4</sup>.

A robotic docking was performed and three arms were positioned in the abdomen. The adopted robotic instruments were Permanent Cautery Hook<sup>®</sup>, Fenestrated Bipolar Forceps<sup>®</sup>, Large Driver Needle<sup>®</sup> to close the peritoneum and 30° endoscope.

The preliminary laparoscopy confirmed the presence of a wide peritoneal defect with the herniation of the part of small bowel in inguinal canal and in scrotum. There were also adhesions between bowel loops.



Fig. 2: Robotic polypropylene mesh positioning.

First, the peritoneum was incised starting 2 cm above the iliac spine and 5 cm above defect. Then we proceeded to preperitoneal dissection and creation of an adequate peritoneal flap, ensuring, in this way, the identification of several anatomic landmarks such as epigastric vessels, bladder, pubis, Cooper's ligament, Gimbernat's ligament, medial part of ilio-pubic tract, external iliac vessels and internal inguinal ring. Afterwards the hernia sac dissection was performed using traction and countertraction maneuvers and avoiding the injuries of the above mentioned structures and of the vas deferens and spermatic vessels.

Thus a polypropylene mesh was placed in the appropriate position and the peritoneum was closed by a running barbed suture (Fig. 2).

Total operative timing was 45 mins, no intraoperative complications were recorded and no drains were applied. The postoperative period was uneventful and the patient was discharged in the first postoperative day.

### Case discussion

Ehlers-Danlos syndrome (EDS) is a group of hereditary disorders caused by abnormal collagen synthesis.

Patients with EDS have hyperelastic or fragile skin, poor wound healing, hypermobile joints, clotting abnormalities, spontaneus pnuemothoraces, recurrent hernias, bowel perforation and vascular complications. In 1682 Meekeren first described a patient with hyperelastic skin. Ehlers and Danlos described the syndrome as being characterized by hyperelasticity of the skin and overextensibility of joints. In 1936 Ronchese established that Ehlers-Danlos syndrome is characterized by "dermatorrhexis and fragititas cutis" and "dermatochalasis and arthrochalasis" <sup>5</sup>. However Schumpelick et al. have been hypothesized that tenascin, a family of glycoproteins, could be linked to hernia formation <sup>6</sup>.

These clinical features and a familiar history of EDS are

often enough to have an exact diagnosis but genetic tests only can confirm the diagnosis and help to rule out other diseases.

Originally, EDS was divided into numbered subtypes. In 1998, the Villefranche classification scheme divided EDS into six subtypes, based on clinical features, biochemical and genetic findings and mechanism of inherence: classic (type I and II), hypermobility (type III), vascular (type IV), kyphoscoliosis (type VI), arthrochalasia (type VIIA and VIIB) and dermatosparaxis (type VIIC). The incidence is one in 5000 people, of which the hypermobility subtype is most common.

The recently published international classification of the Ehlers-Danlos syndrome describes the genetic basis for each type of EDS. The classic, vascular and arthrochalasia types have been linked to either type I or type III collagen disorders. The hypermobility type is linked to tenascin X alterations.

EDS can interest every part of the body where connective tissue is present, causing a disturbed balance between mature and immature collagen. There are twenty different types of collagen which make up the human extracellular matrix and most represented are type I and III collagen. Collagen type I is mature and stable, while collagen type III is immature and instable.

Hernia formation occurs when fatty or intestinal tissues push through a weakness in the abdominal wall and, in fact, it is more frequently in patients with EDS. Another problem for these patients is recurrent hernia formation, probably due to a combined problem of biology and technique. Moreover many surgeons believe that EDS may have a negative effect on the clinical outcome of hernioplasty because of postoperative complication and recurrence rates.

EDS is frequently undiagnosed in patients who present for inguinal hernia or repair of ventral abdominal wall hernias and it could be related to the fact that the syndrome presents heterogeneity and phenotypic variability. The literature on the relationship between EDS and hernia development is scarce and anedoctal.

M.S.L. Liem et al., quantifying the increased risk for inguinal hernia in both male and female patients with EDS, confirmed the popular belief that inguinal hernia occurs more in patients with EDS, mainly in men. It can due to the fact that the female inguinal anatomy may have a lower risk of inguinal hernia <sup>7</sup>.

Kroese et al. evaluated outcomes of ventral hernioplasty in 14 patients with a median follow-up of 50 months and a recurrence rate of 7.1% (one patient), considering the primary outcome hernia recurrence and the secondary outcome postoperative complications. They used large meshes that reinforce the entire midline. This case series has been the first study which considers patients EDS as a specific group for developing hernia recurrence and it has demonstrated that patients with EDS are prevalent in the ventral abdominal wall hernia population. However, the study by Kroese et al presents some

limitations such as the retrospective method, the fact that only two of the fourteen patients (14%) had the classic EDS type and the relatively short follow-up period for some patients  $^{6}$ .

A mesh overlay has been already recommended by J. A. Girotto et al., who used the components separation to solve the abdominal wall herniations of two patients with EDS. This is the first article that address recurrent ventral abdominal herniation in patients with EDS <sup>8</sup>.

According to DiBello and Moore, the ideal abdominal wall reconstruction should satisfy the following criteria: prevent visceral eventration, provide a tension-free repair, incorporate with the remaining abdominal wall, provide dynamic muscle support and endure over time.

L. d. Weerd et al. described a novel approach to treat a large abdominal intercostal hernia in a patient with vascular type Ehlers-Danlos syndrome, using both a polypropylene mesh and a Vacuum Assisted Closure (VAC) system. They positioned a polypropylene mesh over the peritoneum, the latissimus dorsi and external oblique muscles were approximated and the VAC system was positioned over the muscle, the sponge was removed after 3 weeks when the patient was reoperated, a second polypropylene mesh was positioned over the muscle layer and, after skin closure, the skin was covered with a large semiadhesive hydrocolloid dressing <sup>9</sup>.

Hernia repair is a challenge to the surgeon not only to the characteristic tissue friability but also to the associated life-threatening complications. Surgical complications in patients with ESD can be cutaneous, vascular and gastrointestinal and they could be related to the friability of the soft tissues. The cutaneous complications are: poor suture purchase, wound dehiscence with high risk of infection in postoperative period and wide scar formation. Vascular complication include the friability of large vessels and oozing from small vessels with highbleed risk during surgical treatment. Gastrointestinal complications involve bowel anastomoses which tend to deiscence during the first or the second postoperative week <sup>1</sup>.

Our strategy was robotic technology to facilitate the surgical approach <sup>10-12</sup> and to improve the surgical outcomes <sup>3</sup>. This treatment is feasible and associated with a shorted recovery and better cosmetic results using smaller surgical incision in a patient with very difficult wound healing. Robotic surgery offers a greater view of anatomic structures because of high definition 3D display inserted in the surgeon's console <sup>13</sup>. The surgeon can interacts directly with optic and instruments, driving them as he likes without assistance <sup>14</sup>. Furthermore, the endowrist movement of the robotic arms allows wide instrument articulation in a confined space, bypassing the limits of laparoscopic instruments. In this way, it is possible to realize an accurate dissection of important elements, to reduce operative timing and intraoperative and postoperative complications. In

oncological disease, robotic approach also allows the safety and oncological efficacy with acceptable results and short-term outcomes <sup>15</sup>. Moreover robotic surgery has also shown its safety in critical patients, according to current literature <sup>2,16</sup>.

In our case, postoperative period was uneventful and the patient presents a normal wound healing without vascular or gastrointestinal complications.

Our case is an important demonstration of the benefit of the robotic approach to this type of disease, even if this remains a high cost-related procedure.

#### Key Messages

Hernia formation is more frequently in patients with EDS. EDS is frequently undiagnosed.

Hernia repair, in patients with EDS, is a challenge for the surgeon not only to the characteristic tissue friability but also to the associated cutaneous, gastrointestinal and vascular complications.

Robotic surgery offers a greater view of anatomic structures because of high definition 3D display inserted in the surgeon's console. Furthermore, the endowrist movement of the robotic arms allows wide instrument articulation in a confined space, bypassing the limits of laparoscopic instruments. In this way, it is possible to realize an accurate dissection of important elements, to reduce operative timing and intraoperative complications.

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