

Laparoscopic Bilateral Gonadectomy and Inguinal Hernia Repair with Mesh for Complete Androgen Insensitivity Syndrome: A Case Report



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ABSTRACT

Background: Complete androgen insensitivity (CAIS) is a difference of sex development, in which there is complete androgen receptor resistance, leading to a female phenotype, despite the presence of normal testes. There is a recognized risk of testicular germ cell tumor in adulthood as high as 15%, and gonadectomy is usually performed in late adolescence or early adulthood.

Case: A 20-year-old patient, with a known diagnosis of CAIS since the age of 16, underwent magnetic resonance imaging of the pelvis, which revealed the testes lying deep into the inguinal canal. The patient was explored laparoscopically and using the technique of transabdominal preperitoneal hernia repair both testes were successfully reduced into the peritoneal cavity and resected. In addition, the defect of the internal inguinal ring was covered and reinforced using a prosthetic polypropylene mesh.

Summary and Conclusion: Minimally invasive gonadectomy is feasible even for gonads situated deep into the inguinal canal. Established benefits of laparoscopic surgery such as reduced postoperative pain, rapid recovery, aesthetically small incisions, and early return to normal activities are very important for CAIS patients. Multidisciplinary team work, among gynecologists and general surgeons, provides excellent results in these complex cases.

Key Words: Gonadectomy, Complete androgen insensitivity syndrome, Laparoscopic inguinal hernia repair

Introduction

Complete androgen insensitivity (CAIS) is a form of difference of sex development, in which there is complete androgen receptor resistance, leading to a female phenotype, despite the presence of normal testes. Diagnosis of the condition is usually made during investigations of primary amenorrhea, in girls with normal breast development. The condition is X-linked and typically, there are other affected women on the maternal side of the individual.¹

There is a recognized risk of testicular germ cell tumor, presumably due to cryptorchidism, which is estimated to be approximately 10%-15% in adults.^{2,3} However, the risk of malignant change is low in childhood and adolescence, therefore gonadectomy can be considered after completion of secondary sex characteristics. This allows for natural progression of puberty, while giving the opportunity for the affected individual to make an informed decision about the procedure.

Gonadectomy is increasingly performed laparoscopically and, in intra-abdominal gonads the procedure technically resembles, that of an oophorectomy.⁴ Nevertheless, testes can be positioned at any point of the embryological descent

route, and if located within the inguinal canal, their retrieval and removal can be technically challenging.

Case

We present a case of a 20-year-old woman, with a known diagnosis of CAIS since the age of 16 years. She had normal breast development, scant pubic and no axillary hair. The vagina was blind ending, measuring 5 cm and there was no uterus identified on ultrasound scan or magnetic resonance imaging of the pelvis, whereas her karyotype was confirmed 46XY. Imaging noted the presence of testes positioned near the internal inguinal ring on either side and into the inguinal canal, with only the left being palpable on clinical examination.

The woman was counseled about the risks associated with retaining her testes, along with consequences of removing them, including short-term surgical risks and long-term requirements for estrogen replacement therapy. She was listed for a laparoscopic gonadectomy, which was performed jointly with a general surgeon, with experience in transabdominal laparoscopic inguinal hernia repair and a specialist in pediatric and adolescent gynecology.

The patient was positioned supine with a slight (10%) Trendelenburg tilt, and an indwelling bladder catheter was inserted. A 3-port laparoscopy was performed, out of which the two 5-mm lateral ports were inserted in the right and left midclavicular line at the umbilicus level.

The authors indicate no conflicts of interest.

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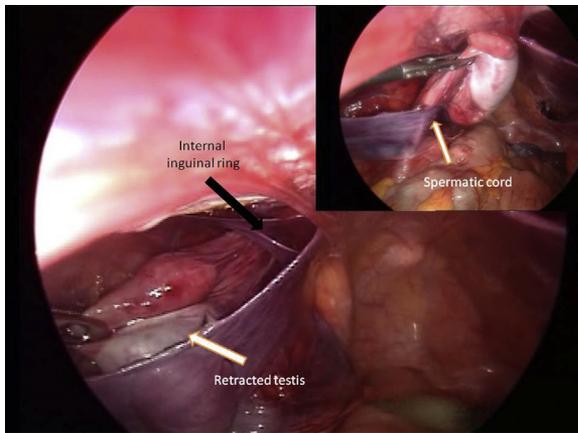


Fig. 1. Retraction of the left testis from the inguinal canal.

The surgical approach followed the classic trans-abdominal preperitoneal bilateral inguinal hernia repair. Dissection of the peritoneum, after a long horizontal incision starting from the anterior iliac spine to the outer edge of the homolateral umbilical ligament, was performed. Dissection started laterally (Borgos' space) and proceed medially behind the symphysis pubis and the iliopubic branch, achieving parietalization of the spermatic fascia. The internal inguinal ring was revealed and then following the spermatic cord, the testis was found in the inguinal canal and pulled into the peritoneal cavity, while gentle push was applied externally at the level of the external inguinal ring (Fig. 1). The spermatic cord vessels and vas deferens were secured en mass using Endoloop Vicryl 0 (Ethicon Endosurgery) and diathermy electrocautery before transection. A similar approach was carried out bilaterally and specimens were removed using an Endobag (Ethicon Endosurgery) through the umbilical port. The 1.5-cm defect at the internal ring was reinforced with a preperitoneal nonabsorbable polypropylene mesh 10 cm high and 15 cm wide (Fig. 2). Mesh fixation and final peritoneum closure were facilitated with the use of laparoscopic tacking staples (Fig. 3).

Recovery of the patient was uneventful, and she was discharged on the first postoperative day. Estrogen

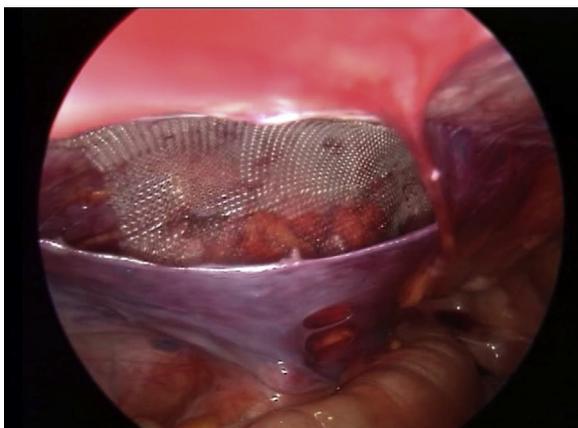


Fig. 2. Mesh is applied over the left internal inguinal ring.

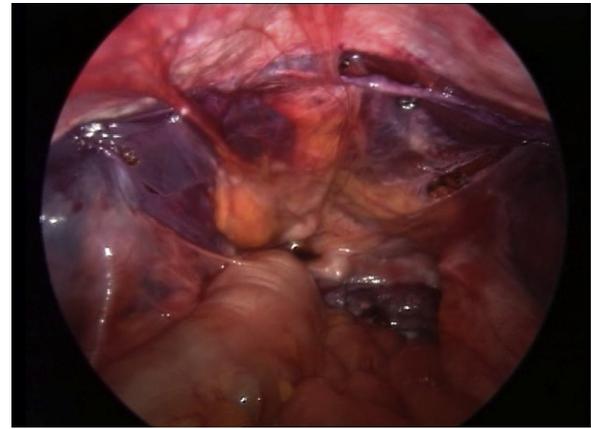


Fig. 3. View of the pelvis after bilateral mesh fixation.

replacement therapy was commenced in the form of 2 mg estradiol valerate orally. She complained of mild soreness over the groin bilaterally, which did not interfere with her mobility. She made a full recovery 2 weeks after the procedure, however, was asked to avoid heavy weight lifting and strenuous exercise for another 4 weeks. Two months postsurgery she is pain-free and has fully returned to her routine activities.

Summary and Conclusion

Gonads in CAIS can be found at any level across their embryological descent route. Inguinal gonads are most commonly identified during childhood, trigger an early diagnosis of CAIS, and are usually managed by pediatric surgeons or pediatric urologists. Traditionally, palpable gonads are removed through an oblique inguinal incision,⁵ during which the internal inguinal ring is usually reinforced using a hernioplasty, to prevent future hernia formation. Current guidelines, however, propose that gonads should be left in situ until completion of puberty, even if identified by chance during hernia repair in childhood.⁶ In a review of CAIS women opting to retain their testes beyond adulthood, fear of surgical complications and postoperative recovery after gonadectomy, concerns regarding side effects of estrogen replacement, and a hope for future fertility prospects were common reasons to defer surgery.⁷ From a medical perspective, despite difficulties in surveillance of intra-abdominal gonads, there might be an advantage in long-term bone mineral density among women who retain their testes longer into adulthood.⁸

Although some women will choose surveillance of gonads, many will opt for gonadectomy to decrease the risk of malignancy. Most of these women will be managed surgically in adulthood and not in a pediatric setting. A proportion of women with CAIS, particularly those with intra-abdominal gonads, will first be diagnosed during investigations of primary amenorrhea in late adolescence and will be primarily managed by gynecologists.⁴

Intra-abdominal gonads are technically easy to remove, and the procedure is preferably performed laparoscopically.⁴ However, gynecological surgical training does not

necessarily involve knowledge of the inguinal region anatomy and exposure. Adequate imaging to locate the gonads, ideally using magnetic resonance, should be performed preoperatively. This is particularly important for nonpalpable gonads, lying deep within the inguinal canal.⁹

In this instance, expertise from a urologist or general surgeon with knowledge in congenital urogenital conditions should be sought. A minimal access approach should again be preferred and although experience in laparoscopic removal of inguinal gonads appears to be scant, it seems reasonable to strengthen the internal inguinal ring, through the placement of a prosthetic mesh preperitoneally after removal of the gonad.¹⁰

The patient described herein made a quick and uncomplicated recovery. We can, however, extrapolate that short- and long-term risks of the procedure are similar to those of a laparoscopic inguinal hernial repair. Postoperative seroma, inguinodynia, and infection, especially of the prosthetic material, are all possible expected complications of laparoscopic transabdominal preperitoneal repair, with complications occurring at similar rates with an open repair approach.¹¹ However, it comes as no surprise that laparoscopic inguinal repair is now preferred, over the open surgery equivalent, particularly because it is associated with shorter postoperative recovery and pain, earlier mobilization, and aesthetically smaller skin scars.

Because CAIS is a relatively rare condition, it is difficult to amass significant surgical experience in laparoscopic inguinal gonadectomy. As with other aspects of difference

of sex development management therefore, it remains crucial that all patients are managed in designated tertiary referral centers.⁶ This will further allow for audit of care and information on long-term effects of gonadectomies to be gathered.

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