Lee, J Clin Case Rep 2017, 7:10
DOI: 10.4172/2165-7920.10001031

Journal of Clinical Case Reports

Case Report Open Access

Two Cases of Successful Laparoscopic Hernia Repair and Repositioning of the Testes into the Abdominal Cavity in Pediatric Patients with Androgen Insensitivity Syndrome

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Abstract

Some infants with androgen insensitivity syndrome (AIS) reportedly develop inguinal hernia (IH). These infants' testes are mostly located in the proximal deep inguinal ring or abdominal cavity. There are few reports on testes present in the labia majora (distal to the superficial inguinal ring) in newborns; however, there is no report on testes fixed to the labia majora that were preserved and moved into the abdominal cavity. Here two cases of successful laparoscopic hernia repair and orchiopexy to move the testes from the labia majora into the abdominal cavity in pediatric patients with AIS having IH are reported.

Keywords: Androgen insensitivity syndrome; Laparoscopy; Inguinal hernia

Introduction

Androgen insensitivity syndrome (AIS) is a recessive genetic disorder inherited through X chromosomes. Morris first described testicular feminization syndrome in 1953 [1]. If the testes are located in the labia majora, it is a problem for AIS individuals who have to live as females. If the testes are not in the abdominal cavity, but in contact with the labia majora, they should be preserved and moved into the abdominal cavity. Then, after the normal external genitalia appear during puberty, individuals can undergo orchiectomy. Although various medical and ethical problems are associated with orchiectomy, laparoscopic orchiectomy is relatively easy to perform in AIS patients [2,3]. However, it is difficult to transfer the testes attached to the labia majora into the abdominal cavity as this report is the first to describe this to the best of my knowledge. Most AIS patients develop into females with external genitalia, a feminine face, and breast development during puberty. AIS patients may be diagnosed on admission because of amenorrhea and infertility since puberty, but the diagnosis may be delayed in children unless specific abnormalities are noticed. One of the most common reasons for pediatric AIS patients to be hospitalized is the occurrence of IH [4,5]. Hernia repair should be performed for inguinal hernia (IH) in infants with AIS. Most AIS patients have testes located in the abdominal cavity or deep inguinal ring. Orchiectomy is commonly recommended after puberty and breast development because the risk of testicular tumor development is low before puberty and the presence of testes may prevent hormone therapy before puberty [6,7]. If the testes are present in the inguinal canal of an infant with AIS before performing a physical examination or ultrasonography, it may be difficult to distinguish it from ovarian inguinal hernia because ovarian hernia in patients aged less than 2 years is not uncommon [8]. In laparoscopic surgery, the uterus and ovary status in girls and vas deferens and spermatic cord in boys can take the advantage of being confirmed by the camera in the abdominal cavity. Here successful laparoscopic hernia repair and one-stage vessel preserving orchiopexy (moving the testes from the labia majora into the abdominal cavity) is reported for pediatric patients with AIS having IH.

Case Report

A retrospective study was conducted of eight phenotypically female AIS pediatric patients who underwent hernia repair at Damsoyu Hospital (Seoul, South Korea) from September 2012 to December 2016. Two of the eight AIS patients had a bowel-incarcerated IH. Two infants were admitted to the hospital (on May 1, 2015 and May 31, 2017, respectively) with left inguinal bulging at the age of 30 days (Figures 1A and 1B). Ultrasonography revealed a left incarcerated IH, thus surgery was planned (Figure 1C). Ultrasonographic findings of one patient showed that the labia majora appeared as bilateral 1.0-cm round mass (Figure 1D), while findings of the second infant revealed a 1.0-cm round mass in the left labia majora and a 0.5-cm round mass in the right inguinal canal. Both infants were diagnosed with an incarcerated IH and underwent emergency surgeries, which were performed by the same surgeon (Figures 1E and 1F).

Laparoscopic surgery was performed in the Trendelenburg position under general anesthesia. A 2.9-mm 0° rigid camera and a 2.7-mm laparoscopic instrument were used as laparoscopic equipment. After the umbilicus was pulled out, a 3-mm longitudinal incision was made with a no. 15 scalpel and a trocar was inserted. After inducing pneumoperitoneum at 6-8 mmHg, two 3-mm stab incisions were made on the lateral abdominal wall and the laparoscopic instrument was inserted directly into the incision without the use of a trocar. Organ incarceration was reduced in the abdominal cavity and the deep inguinal ring was observed (Figures 2A and 2B). AIS was suspected because there were no uterus and ovaries (Figure 2C). After removal of the hernia sac, the spermatic cord and vas deferens were confirmed (Figure 2D). The testes were pulled and the spermatic cord and vas deferens were preserved. Finally, the testes were placed into the abdominal cavity (Figures 2E-2G). Intracorporeal high ligation was performed (Figure 2H).

Drinking water or milk was introduced 2 h postoperatively if there were no complications, in accordance with the procedures of Damsoyu Hospital. Since there was no problem with the ability to drink fluids, both patients were discharged on the day of surgery. A surgical adhesive

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Received August 19, 2017; **Accepted** October 22, 2017; **Published** October 27, 2017

Citation: Lee SR (2017) Two Cases of Successful Laparoscopic Hernia Repair and Repositioning of the Testes into the Abdominal Cavity in Pediatric Patients with Androgen Insensitivity Syndrome. J Clin Case Rep 7: 1031. doi: 10.4172/2165-7920.10001031

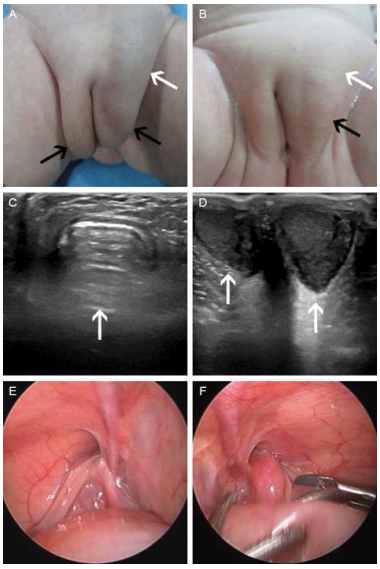

Figure 1: Appearance of the inguinal area of a patient with androgen insensitivity syndrome having inguinal hernia and ultrasonographic findings and laparoscopic views
(A) Left incarcerated inguinal hernia (white arrow) with a palpable round mass (black arrow) in both labia majora of an androgen insensitivity syndrome patient. **(B)** Left incarcerated inguinal hernia (white arrow) with a palpable mass (black arrow) in the left labia majora of an androgen insensitivity syndrome patient. **(C)** Pre-operative ultrasonography of the inguinal canal showing bowel incarceration (arrow). **(D)** Pre-operative ultrasonography of the labia majora showing 1.0-cm round mass (arrow) on both sides. **(E)** The testes are not visible due to the location in the labia majora. **(F)** The testes are located in the deep inguinal ring.

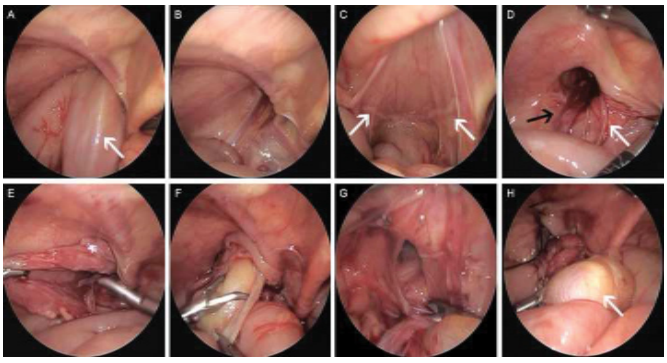

Figure 2: Laparoscopic procedures of hernia repair and orchiopexy.
(A) Laparoscopic view of the left internal inguinal ring. Bowel (arrow) incarceration is visible. **(B)** Laparoscopic view after the reduction of incarceration. **(C)** At the bottom of the bladder, both vas deferens (arrow) are visible, but the uterus and adnexa are not visible. **(D)** After removal of the hernia sac, the spermatic cord (black arrow) and vas deferens (white arrow) are visible. **(E)** Pulling dissection of the spermatic cord and vas deferens. **(F)** Pulling of the testis into the abdominal cavity. **(G)** The vas deferens and spermatic vessels were preserved. Shape of the deep inguinal ring after full dissection. **(H)** High ligation of the peritoneum. The preserved testis (arrow) is visible.

was used to cover the wound and no outpatient treatment for wound management was needed postoperatively. Follow-up was conducted on postoperative day 7. Ultrasonography showed well-preserved intraabdominal testes. Karyotype analysis revealed the 46, XY karyotype as AIS.

Results

Patient characteristics are shown in Table 1. By preoperative ultrasonography, AIS was suspected in seven patients. Seven patients had complete AIS and one had partial AIS. Six patients had a palpable inguinal mass without IH and two patients had testis in labia majora. In one infant, both testes were located in the bilateral labia majora (Figure 1A). In the other infant, the left testis was located in the labia majora (Figure 1B) and the right testis was located in the deep inguinal canal (Figure 1F). The surgical duration for the two patients was 17 and 23 min, respectively, and they were discharged 7 and 22 h after surgery, respectively. There was no specific complication in either patient and both were transferred to the Department of Pediatrics after surgery.

Discussion

AIS is a genetic disease of males with 46, XY chromosomes that occurs as a point mutation of androgen intracellular receptors coded on the X chromosome [9]. AIS is classified as complete or partial, depending on the resistance to androgen [10]. The prevalence of AIS is reportedly one case per 10,000-13,000 male births [6,11]. The diagnosis of AIS in pediatric patients is confirmed during hernia surgery, which is reported to be 0.8%–2.4% in phenotypically female patients with IH [6]. However, in most reports, the diagnosis of IH was not an actual hernia causing protrusion of the abdominal organs, but rather contact of the testes with the inguinal canal [12]. In this study, among 1,200 phenotypically female pediatric patients aged under 10 years who visited the hospital because of inguinal bulging or the presence of a palpable mass, 8 (0.67%) patients were diagnosed with AIS. Two patients had actual IH accompanied with bowel protrusion. Adult patients present with symptoms of amenorrhea and infertility, while pediatric AIS patients have few symptoms before puberty. In addition to a physical examination, ultrasonography is used for further preoperative evaluation. On physical examination, AIS patients typically have female external genitalia with a palatable mass or reducible inguinal bulging of the inguinal canal. The possibility of ovarian IH is highly likely in patients aged under 2 years [8]. A previous study reported the use of ultrasonography to distinguish the testes from ovaries [13]. In my experience, a herniated ovary appears as a hypoechoic lesion, while the testes appear more echogenic than the ovaries by ultrasonography. Therefore, the echo pattern of the testes differs from that of the ovaries. Nonetheless, it is sometimes difficult to differentiate between the testicles of a patient from a round palpable mass of AIS.

Laparoscopic surgery of the abdominal cavity can be directly observed with the use of a camera, so that the position of the uterus, ovary,

Variables	Testes located in the labia majora (n=2)	Testes located in the abdominal cavity (n=6)
Age (months)	1.00 ± 0.00 (1)	6.67 ± 5.09 (1–14)
Symptoms		
Palpable mass only	0	6
Inguinal bulging with palpable mass	2	0
Type of AIS		
Complete	2	5
Partial	0	1
Accompanying inguinal hernia	2	0
Location of testes		
Deep inguinal ring	0	6
Labia majora	2	0
Surgical duration (min)	20.0 ± 3.00 (17–23)	22.5 ± 13.8 (10–50)
Hospital stay (h)	14.5 ± 7.50 (7–22)	7.33 ± 0.47 (7–8)
Complication	0	0
AIS: Androgen Insensitivity Syndrome		

Table 1: Characteristics of patients with AIS.

vas deferens, spermatic cord, and the testes can be accurately mapped, while repairing IH simultaneously. If orchiectomy is performed for an infant AIS patient, estrogen replacement therapy should be performed [14]. If the timing of orchiectomy is explained to the parents before surgery, most choose orchiectomy after puberty. In order to check the condition of the testes periodically by ultrasonography, fixation should be performed in a visible position that is easily identified in imaging test. The postponement of eventual orchiectomy to adult age is a major advantage that not only allows puberty to occur spontaneously but also permits informed and authoritative decision-making by patients [15].

Growth of phenotypically females should prevent contact of the testes to the labia majora or inguinal canal. If the testes are located within the abdominal cavity, herniorrhaphy can be performed simply, but if the testes are in contact with the inguinal canal or labia majora, herniorrhaphy should be performed only after the testes are dissected and moved into the abdominal cavity. The author preserved the spermatic vessel during laparoscopic surgery and moved the testes safely into the peritoneal cavity without injury. If the testes are located in the labia majora in an AIS patient, it is as if the testes are normally located in the scrotum in a phenotypic male patient. Therefore, it may be difficult to move the testes into the abdominal cavity by laparoscopic surgery. The dissection of the spermatic cord and vas deferens proceeded from the deep inguinal canal to the external canal allows easy transfer of the testes into the abdominal cavity. For cryptorchidism, orchiopexy is performed by increasing the length of the spermatic cord and vas deferens by pulling the testes from the inguinal canal into the abdominal cavity and then fixing the testes to the scrotum. Therefore, the surgeries of the two patients included in this study were not difficult.

Conclusion

In infants with AIS having IH, it may be difficult to confirm preoperative AIS despite ultrasonographic findings. If an incarcerated hernia is found, emergency surgery should be performed. Laparoscopic surgery for pediatric hernia is now widely performed to accurately assess the condition of the reproductive organs during herniorrhaphy. Laparoscopic surgery for pediatric AIS patients with IH offers the advantage of facilitating herniorrhaphy and transferring the testes into the abdominal cavity. This report suggests that testes preservation may be more possible in patients with infantile AIS who undergo hernia repair.

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