World Journal of Surgery and Surgical Research

Case Report
Published: 21 Nov, 2022

Uterine Isthmus Atresia Conservatively Treated by Laparoscopy and Transvaginal Utero-Cervical Anastomosis

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Abstract

Uterine Isthmus Atresia (UIA) is an unusual complete separation of uterine body from cervix, often excessively treated (hysterectomy). This report aims to describe a uterine-sparing technique for UIA treatment. This is the case of a 14-year-old girl referred for primary amenorrhea and cyclic pelvic pain. Imaging study was suggestive for UIA, hematometra and bilateral hematosalpinx. The patient was submitted to a combined laparoscopic-transvaginal end-to-end utero-cervical anastomosis.

A stable anastomosis was obtained. At 3-month follow up the patient referred regular painless menses. Ultrasound identified the new connection between the cervix and the uterine body. Hysteroscopy revealed a short but regular cervical canal, widely permeable and regularly connected with the uterine cavity. For UIA treatment, when the cervical segment is intact, an end-to-end utero-cervical anastomosis is feasible and effective.

Keywords: Uterine isthmus atresia; Mullerian anomalies; genital anomalies; utero-cervical anastomosis; Laparoscopy

Introduction

Uterine Isthmus Atresia (UIA) is a rare abnormality of female reproductive tract, characterized by the complete separation between the uterine body and the cervix due to the failure of the isthmic part of the uterus to develop (Figure 1).

The relative rarity of these conditions favors confusion among physicians with delay of diagnosis [1] and serious consequences in management of these patients [2].

Generally confused with cervical dysgenesis, only few cases of UIA are correctly diagnosed [3] and frequently submitted to demolitive interventions (i.e., hysterectomy), while a conservative surgical management is feasible and effective, as we aim to demonstrate in the case described in this report.

Case Presentation

Patient characteristics

A 14-year-old woman referred to the Adolescent Gynecological Unit of Vita-Salute San Raffaele Hospital of Milan complaining of primary amenorrhea and cyclic pelvic pain lasting 1 year. For 3 months the patient was treated with continuous hormonal therapy (levonorgestrel 1 mg/ethinyl estradiol 0.02 mg) in order to avoid endometrial proliferation.

A detailed medical history was obtained to determine pubertal development and low abdominal pain features (onset, timing, and nature). She had no sexual intercourse and no history of in utero exposure to teratogenic drugs was reported.

Physical examination was conducted. Development of secondary sexual characteristics was normal (Tanner stage IV for breast and pubic hair development). Careful perineal inspection, pelvic and rectal examination were performed. At perineal inspection no evidence of obstructive abnormalities, such as imperforate hymen or transverse vaginal septum, was identified. At rectal examination no tumescence proximal to the introitus was observed. At pelvic evaluation the uterine body was easily palpable, mobile and enlarged due to the probable presence of hematometra.

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Received Date: *31 Oct 2022*
Accepted Date: *17 Nov 2022*
Published Date: *21 Nov 2022*

Citation:
Candiani M, Fedele F, Dolci C, Grecchi G, Petrone M, Ruffolo AF. Uterine Isthmus Atresia Conservatively Treated by Laparoscopy and Transvaginal Utero-Cervical Anastomosis. World J Surg Surgical Res. 2022; 5: 1422.

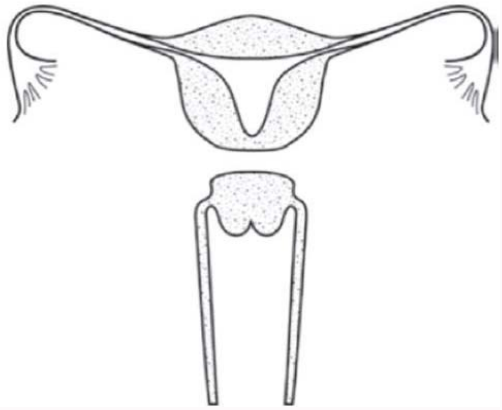

Figure 1: Uterus isthmus agenesis. Class of Complex anomalies. ASRM 2021.

Serum hormone levels were within normal limits.

Study imaging was assessed. Abdominal two-dimensional ultrasound revealed an enlarged uterine body with an endometrial cavity distended by hyperechoic material compatible with hematometra, a modest bilateral increase of Fallopian tubes dimension evocative of bilateral hematosalpinx, and a modest fluid flap in the pouch of Douglas suggestive of retrograde menstruation. No renal or urinary anomalies were identified. The transrectal ultrasound confirmed the distended uterus with hematometra and showed a morphologically normal cervix. In a sagittal plane no connection was observed between the cervical canal and the endometrial cavity, as they were separated by 20 mm of fibrotic tissue (Figure 2A). The MRI confirmed the agenesis of the uterine isthmus (Figure 2B) and the absence of associated malformations.

In order to surgically treat the uterine isthmus agenesis a combined laparoscopic and transvaginal approach was decided. Before the operation, an appropriate counselling session was conducted with the patient and her parents. The anatomical malformations and the therapeutic possibilities available were illustrated in detail. The advantages and disadvantages of definitive surgical therapy and those of conservative surgical therapy were illustrated, emphasizing that experience with the latter approach is limited. The primary outcomes were the stable recovery of menstrual function and secondly the possibility of allowing future fertility (Figure 3).

The reported risks of the surgical procedure are perioperative bleeding, bladder and rectal injuries, infections, reoperation and hysterectomy. The alternatives remain elective hysterectomy or continuous hormone therapy to avoid menstrual cycles.

Surgical procedure

A first laparoscopic step was performed with a standard technique. An accurate survey of the entire abdominal-pelvic cavity was carried out. The uterus was enlarged, highly mobile, ball-shaped with normal peritoneal profile (Figure 4.1). However, at an accurate inspection the isthmic area of connection between the uterus and Douglas's peritoneum lacked. When the adnexa were bilaterally evaluated, both of them resulted loosely adhered to the posterior surface of the uterus and to the homolateral ovarian fossa. Left ovary presented normal characteristics, while right ovary showed small superficial endometriotic foci. The fallopian tubes had a regular course but resulted moderately enlarged due to a condition of bilateral hematosalpinx.

Firstly, diathermal-coagulation of pelvic endometriosis and lysis of ovarian adhesions were performed. Subsequently, a small myometrial incision (of approximately 1 cm) of the uterine fundus was made with a monopolar needle and the previously described hematometra was drained. A graduated probe was then inserted into the uterine cavity.

A following vaginal step started with the recognition of the extreme elasticity of the hymen that allowed the surgeon to avoid its sharp incision. Despite a normal view of the vagina and of the external cervical os, an hysteroscopic approach failed to enter into the endometrial cavity that resulted inaccessible; therefore, a uterine internal view was impossible to obtain. In fact, the cervical canal abruptly ended, and a transverse cervical diaphragm that occluded the route was identified. Cervical dilation was tried with Hegars dilators (up to number 8), but no cervical canal patency was obtained (Figure 4).

Under simultaneous laparoscopic vision, a small transversal incision was made vaginally in correspondence of the anterior vaginal fornix, leading to the opening of the posterior peritoneum of the Douglas pouch.

The cervical canal was then bluntly opened in a cranial direction, where it was previously blocked by the transverse cervical diaphragm. Then, the lower end of the ball-shaped uterine body, on the guide of a hysterometer laparoscopically inserted into the endometrial cavity from the fundal incision, was pushed down towards the previously opened anterior vaginal fornix. The first operator grasped the uterus base from below and on the guide of the lower end of the intracavitary

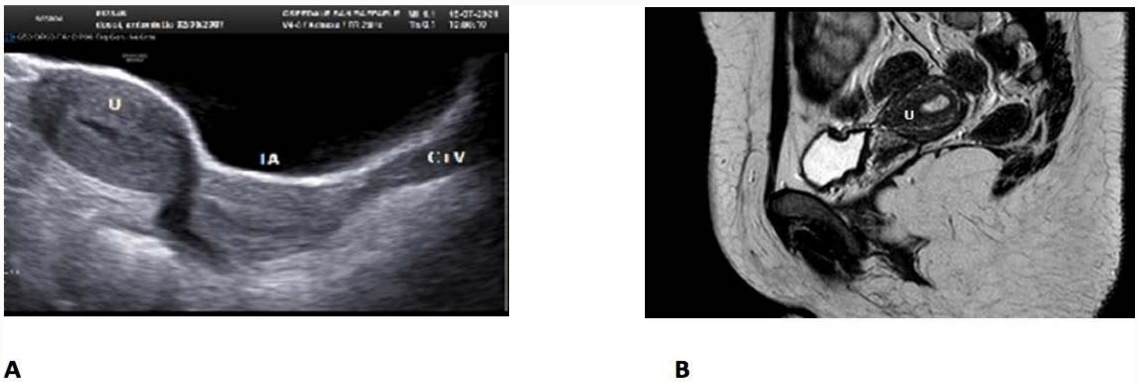

Figure 2: Preoperative transrectal ultrasound (A): U: Uterine body, IA: Isthmus Absent, C+V: Cervix + Vagina and MRI (B): U: Uterine body.

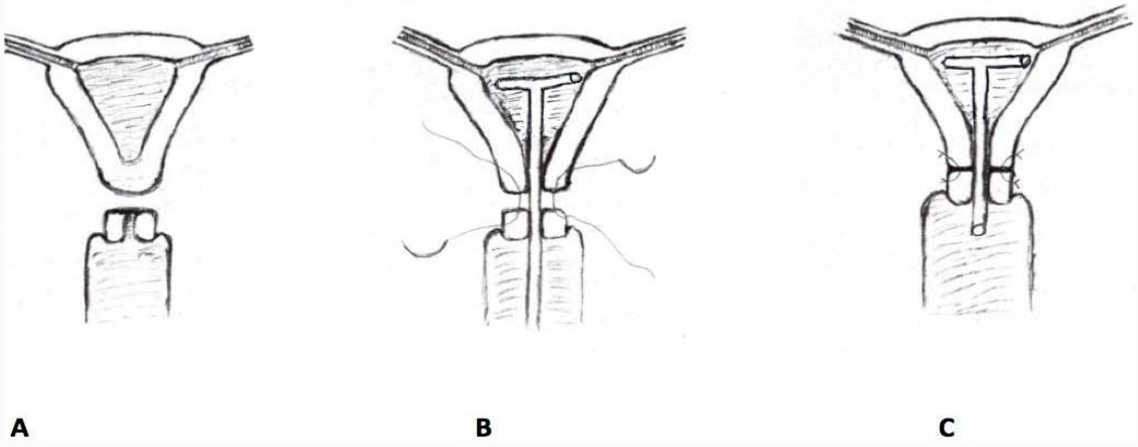

Figure 3: Surgical steps of operation. (A) Initial findings: the uterine body, site of hematometra, is completely separated from the cervix and vagina by agenesis of the isthmus. (B) The edges of the open caudal uterine body and the edges of the cervix are anastomosed on the guide of a suitably shaped Kherr tube. (C) Last time: the stitches are tied and the Kherr tube, cut at the external orifice of the cervix, is left in place until after the first menstruation.

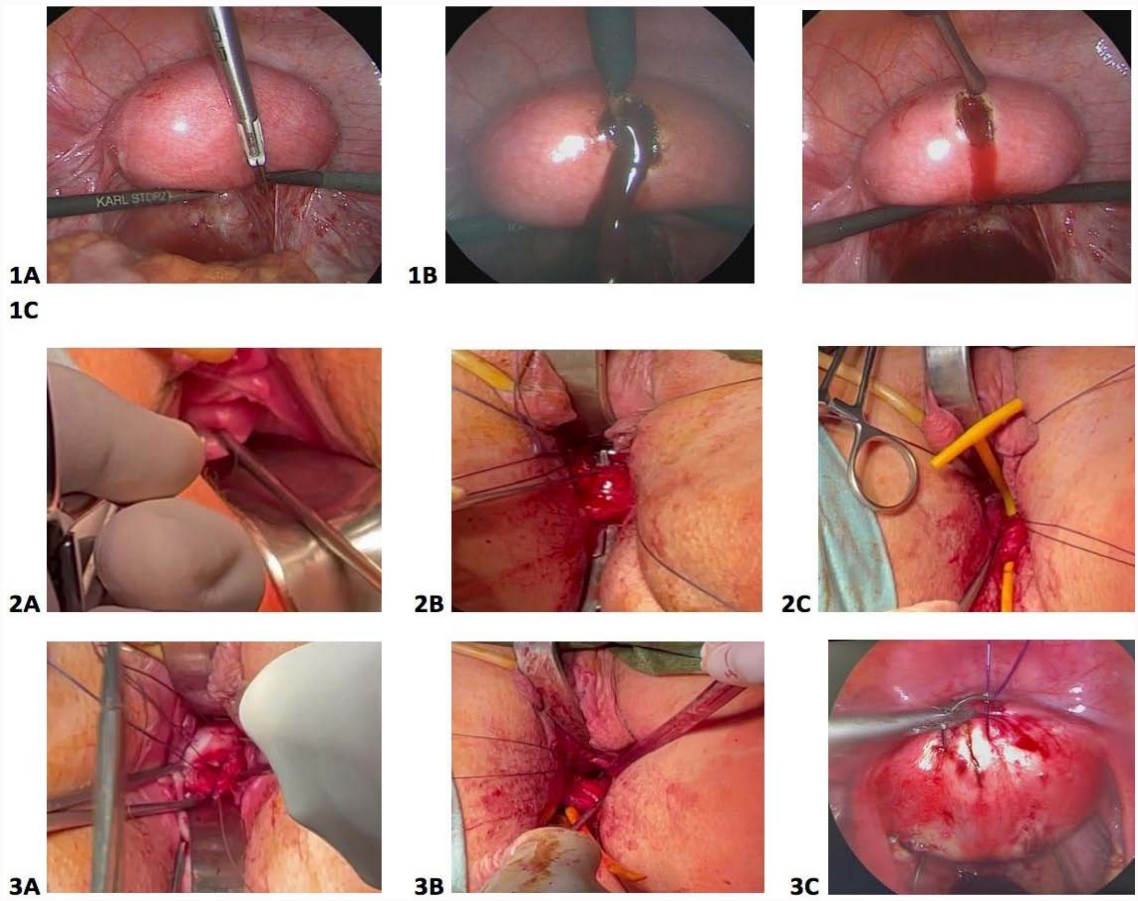

Figure 4: Photo sequence of the intervention.
1. Laparoscopy: (A) The uterus is enlarged, ball shaped, without isthmic zone; (B) A small fundal myometrial incision is made to empty the hematometra; (C) A graduated probe is inserted into the uterine cavity and the uterus body is pushed down.
2. Vaginal step: (A) The cervix is present but completely blind; (B) After opening the anterior vaginal fornix, the cranial end of the cervical canal is opened; (C) The long arm of the appropriately shaped Kherr drainage is inserted into the cervical canal.
3. Uterocervical anastomosis: (A) The uterine stump is prepared after opening the caudal end of the uterine body; (B) The T-arm of the Kherr drainage was inserted into the uterus and the anastomosis was completed. It remains to close the anterior vaginal fornix; (C) The myometrial breach made at the beginning is sutured laparoscopically.

probe performed a circular myometrial incision of the uterine caudal body reaching the endometrial cavity. The excision of a circular portion of the myometrium allows a spontaneous maintaining of the patency of the endometrial cavity. The subsequent discharge of a small amount of blood from the uterine cavity was observed. Subsequently, the edges of the caudal uterine body were anastomosed with the edges

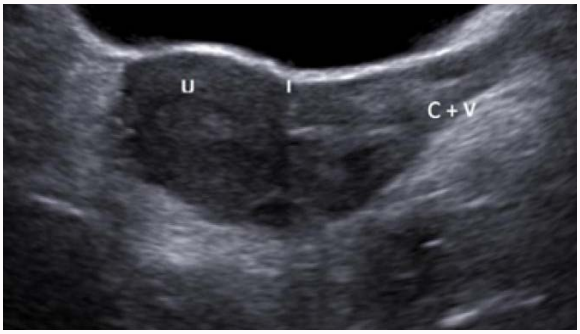

Figure 5: At three months follow-up transabdominal ultrasound confirms the new connection between the cervix and the uterine body.
U: uterine body; I: Isthmus, C+V: Cervix + Vagina

of the cranial extremity of cervix previously opened, on the guide of a Kherr tube inserted into the uterus and into the cervical canal. A circular series of stitches, with single interrupted full thickness bites, was used to firmly fix the anastomosis. Six stiches in total were used, 3 for the rear wall and 3 for the front wall. Anterior fornix was sutured by interrupted stiches.

Patient received broad-spectrum antibiotics (i.e., cephalosporins of the last available generation) the day before and the day of surgery. The Foley bladder catheter was removed 24 h after the surgery and the patient was discharged after 48 h, without pain or postoperative complications.

Follow-up

Kherr's tube was removed after the first menstruation and an estrogen-based vaginal cream was prescribed for three months.

At 3-month follow up the patient referred regular painless menses. An abdominal two-dimensional ultrasound identified the new connection between the cervix and the uterine body (Figure 5). An explorative hysteroscopy revealed a short but regular cervical canal, widely permeable and regularly connected with the uterine cavity.

Discussion

We described one of the few cases of Uterine Isthmus Agenesis (UIA) management reported in literature. While the incidence of cervical agenesis ranges from 1/80.000 to 1/100.000 [4], the report of a selective agenesis of the isthmus of the uterus without other associated anomalies is extremely rarer. A hypothesis can be that UIA is poorly reported in literature as it is frequently confused with cervical dysgenesis. Indeed, both conditions are characterized by a delayed diagnosis at the age of menarche, when the obstruction to the outflow of menstrual blood leads to a blood collecting firstly into the uterus and then retrograde into the fallopian tubes and the peritoneal cavity.

The classification of the female reproductive tract anomalies remains a constant issue, and the UIA is not an exception. In fact, The American Fertility Society Classification of the Mullerian Anomalies [5] does not precisely contemplate this malformation. The Vagina Cervix Uterus Adnexa-associated Malformation (VCUAM), includes UIA among congenital hypoplasia of uterine cervix (i.e., VCUAM type U4a ESGE U6/unclassified) while the ESHRE/ESGE classification [6] places the anatomical defect of the upper part of the cervix in subclass C4. Only the new classification of the American Society for Reproductive Medicine (ASRM) of 2021 includes specifically the UIA of the uterus among the complex anomalies [7].

A careful preoperative study of the patient provides great benefit to the scheduled surgery, with MRI preferred in pediatric patients when transvaginal ultrasound is not practicable [8].

In cervical atresia different attempts at “conservative” therapy (i.e., recanalization around stents of the rudimentary cervix) resulted in peritonitis and fatal septic shock [9,10], reinforcing advocates of the so-called "definitive" therapy (hysterectomy). These infectious complications were attributable to the lack of a cervix which normally protects the endometrium from the vaginal environment. In the UIA, on the other hand, part of the cervix is usually preserved and can play its protective role. This concept made the attempt of a conservative intervention mandatory in our case [11].

However, the technical difficulties of a utero-cervical anastomosis and the choice of the route of surgery forward remain considerable factors in UIA management and different approaches have been described (Table 1).

In our opinion, the chosen approach achieved the best compromise between invasiveness of the surgical access and strength of the performed anastomosis. In fact, as laparoscopy allows to obtain an overall view of the abdomen-pelvic anatomy, the vaginal route leads to a meticulous preparation and a precise approach of the uterine abutments to be connected. Finally, the manual closing of the anastomosis sutures is certainly safer and more stable than what an excellent laparoscopist can do endoscopically.

Conclusion

When a UIA is diagnosed, and a part or the whole cervix with its mucus is present, a surgical recanalization of the genital tract through an end-to-end anastomosis between the cervical canal and the uterine body by an integrated laparoscopic-vaginal approach is a feasible option.

Table 1: Characteristics of previous studies describing the surgical management of uterine isthmus agenesis or cervical agenesis.

	Age (years)	Malformation	Surgical procedure	Surgical approach
Grimbizis, 2004 [12]	15	Cervical agenesis	Utero-vaginal anastomosis	Laparoscopic first attempt converted in laparotomy
Wright, 2011 [13]	14	- Uterine isthmus agenesis - Bicorn uterine with only one cavitated horn	Bilateral supra-cervical hemi-hysterectomy	Laparoscopy
Yang, 2015 [14]	15	Uterine isthmus agenesis	Cervical-uterine anastomosis	Laparoscopy
Richards, 2018 [3]	15	Uterine isthmus agenesis	First surgery: Evacuation of the hematometra Second surgery: Cervical-uterine anastomosis	First surgery: Laparoscopy Second surgery: Laparotomy
Carreras, 2020 [15]	14	Uterine isthmus agenesis	Cervical-uterine anastomosis	Laparoscopy

Ethical Approval

The Institutional Review Board (IRB) Approving and the Patient Consent were obtained (GARA protocol 73/INT/2021).

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