



## Fertility sparing technique during pelvic exenteration for recurrent vaginal rhabdomyosarcoma

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### ABSTRACT

We present the first case and describe the surgical technique of anterior pelvic exenteration with uterine preservation in a 17-year-old patient with a recurrent vaginal rhabdomyosarcoma.

Surgical technique included a skeletonization of uterine pedicles and ligation of superior vesical and vaginal arteries, adapting abdominal radical trachelectomy technique. Cervix was transected to avoid vaginal opening and tumor spillage. Uterus was reimplanted to a vaginal reconstruction, created with a DIEP, and a continent urinary diversion was performed. Pelvic filling was completed with an omental J-flap.

Postoperative course was uneventful and the patient was discharged at day 17th. The last days of her stay were dedicated to self-catheterization education and learning the management of Miami pouch. Her menstrual cycle resumed two months after the surgery. Cervix exhibited a normal appearance during clinical examination eight weeks after surgery and postoperative MRI did not show signs of local recurrence. Unfortunately, distant metastatic relapse occurred three months after surgery and the patient died two months later.

Fertility preservation at the time of anterior pelvic exenteration is technically feasible in selected young patients. Further cases are needed to assess the reproducibility of this surgical procedure, the reproductive function of the uterus and the rate of uterine recurrences.

## 1. Introduction

Pelvic exenteration (PE) is an uncommon procedure among young female patients, solely indicated in advanced or recurrent genital tract rhabdomyosarcoma. Decades ago, fertility sparing surgery was not possible after pelvic radiotherapy, as result of damage of reproductive organs. Currently, intensity-modulated radiation therapy (IMRT) allows to spare surrounding tissues (such as ovaries and uterus) from irradiation, which offers the possibility of a subsequently conservative surgery in selected cases [1]. Up to our knowledge, we present the first surgical description of fertility sparing PE with uterine preservation and vaginal reconstruction with a Deep Inferior Epigastric Perforator (DIEP) flap.

## 2. Case report

### 2.1. Patient

In August 2011, a 15-year-old woman was referred to our Comprehensive Cancer Center for a 7-cm vaginal mass. The anatomopathological analysis reported a botrioid rhabdomyosarcoma without identifying PAX/FKHR fusion gene. Bilateral inguinal and right internal iliac lymph node metastasis were detected on preoperative Positron emission tomography/computed tomography (PET/CT) and confirmed histologically. Bone marrow biopsies were found to be negative. After vaginal debulking to relieve symptoms, laparoscopic left ovarian transposition and right oophorectomy –for cryopreservation– were

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performed. The patient underwent 6 cycles of combined intravenous adjuvant chemotherapy, achieving complete magnetic resonance imaging (MRI) response. Finally, according to current guidelines, conservative adjuvant treatment was proposed, using interstitial vaginal brachytherapy and inguinal and right internal iliac IMRT (mean dose of radiation delivered to the uterus was 4.4 Gy).

Ten months after the end of the treatment, a local recurrence was diagnosed. The case was discussed at the French National Board –Multidisciplinary French Sarcoma Group– in order to decide the therapeutic strategy. In January 2013, a partial colectomy with clear macroscopic margins was performed through a vaginal approach. However, microscopical positive margins with lymphovascular space invasion (LVSI) were found in final anatomic-pathological examination, classifying the case as R1. Maintenance chemotherapy with cyclophosphamide and vinorelbine was started concomitantly with vaginal IMRT (66 Gy on R1 area, 56 Gy on the vagina, 44 Gy on the uterus). Her menstrual cycles resumed two months later.

Five months after beginning the maintenance treatment, a second vaginal recurrence occurred. Two courses of combined chemotherapy (intensified doxorubicin plus ifosfamide) were administered, but disease was found to be chemoresistant. Pelvic MRI showed a growing inferior 9-cm vaginal tumor involving the bladder. PET/CT confirmed local recurrence and ruled out distant disease. In September 2013, a salvage anterior PE with total vaginectomy was proposed by the French National Board. Due to her young age, her wish of fertility preservation, the inferior location of the recurrence and normal-functioning uterus and ovaries, a fertility sparing procedure was proposed. Preoperative MRI and ultrasonography confirmed normal uterine morphology and ruled out upper vaginal involvement.

## 2.2. Surgical technique

Pelvic exenteration was performed through a midline laparotomic approach. Paravesical and pararectal spaces were developed. Both ureters were transected cranially to internal iliac vessels. A complete dissection of internal iliac arteries was performed, along with skeletonization of uterine pedicles (Fig. 1).

Superior vesical and vaginal arteries were ligated. Cervix was

transected in order to avoid vaginal opening and tumor spillage (Fig. 2).

Uterus was mobilized cranially to complete anterior PE. The peritoneal incision was extended anteriorly to reach Retzius space and posteriorly into the Douglas pouch in order to open the rectovaginal septum. Anterior aspect of the rectum was separated from the posterior vaginal wall using blunt dissection. Both paracervical and paravaginal tissues were transected using an ultrasonic and advanced bipolar sealing and cutting device (Thunderbeat®, Olympus, Japan), reaching the pelvic floor. Endopelvic fascia was dissected and opened. After applying cranial traction, paravesical plexus was coagulated, after which the urethra was exposed and transected. A circumferential perineal incision was made including the entire vagina and the urethra, assuring 2-cm macroscopical margins. An en-bloc type II PE according to Magrina classification [2], including total and circumferential vaginectomy, was performed (Fig. 3).

A vertical DIEP flap was harvested for vaginal reconstruction after localizing two medial perforator arteries using a hand-held Doppler flow detector. Lateral part of the skin paddle was sutured to vaginal introitus. Then, the flap was folded on itself and twisted with a spiral shape creating a 13-cm neovagina. Remaining cervix was reattached to the neovagina (Fig. 4).

Pelvic filling was completed with an omental J-flap. Continent urinary reconstruction with a Miami pouch was performed, placing the urostomy in the umbilicus.

To publish this case and accompanying images, written informed consent was obtained from the patient and her parents.

## 2.3. Postoperative outcomes

Postoperative course was uneventful and the patient was discharged after 17 days. Last days of her stay were dedicated to education in self-catheterization and management of Miami pouch.

Anatomic-pathological examination reported a 10-cm vaginal recurrence involving bladder trigone. Cranial vaginal margins were negative whereas vulvar margins were positive with LVSI, hence complementary vulvectomy was performed 10 days after discharge, eventually achieving microscopical in sano margins. Maintenance chemotherapy based on tyrosine kinase inhibitor was proposed. Her

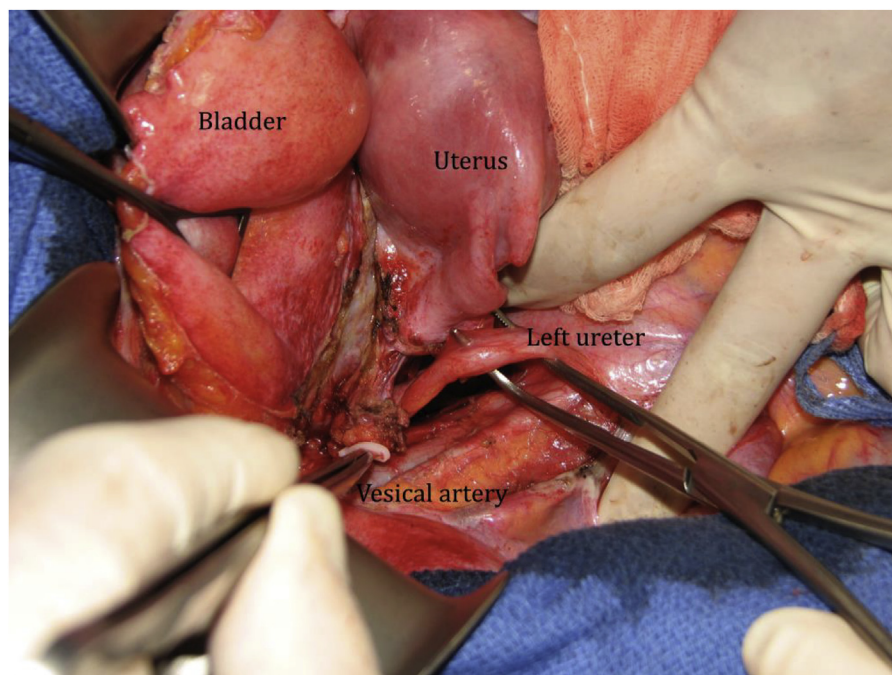


Fig. 1. Complete dissection of left internal iliac artery with skeletonization of the left uterine pedicle and ligation of the superior vesical artery.

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