



## Local control for vaginal botryoid rhabdomyosarcoma with pre-rectal transperineal surgical resection and autologous buccal graft vaginal replacement: A novel, minimally invasive, radiation-sparing approach

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### ABSTRACT

**Purpose:** Localized vaginal rhabdomyosarcoma (RMS) is associated with a favorable prognosis, but strategies for local control remain controversial. The use of radiotherapy (RT) can have important long-term sequelae, while traditional resection involves major reconstructive surgery. We describe a new surgical approach employing a minimally-invasive resection and immediate reconstruction.

**Materials and methods:** Records from 4 consecutive patients with localized vaginal RMS managed in 4 major pediatric referral centers were reviewed. All cases were performed with a standardized technique.

**Results:** Patients were diagnosed at a median age of 24 months. Each underwent a total/subtotal vaginectomy with autologous buccal graft vaginal replacement. Final margins were focally positive in one patient and negative in three. None received radiotherapy. To date, all patients have patent buccal neovaginas, enjoy a favorable aesthetic result, and remain disease-free at a median follow-up of 35 months.

**Conclusions:** We report 4 cases of localized vaginal RMS successfully treated with a minimally invasive surgical approach. All patients have avoided radiation and remain disease-free. Our initial data suggest that surgical local control and immediate reconstruction are feasible and can spare these patients the long-term complications of RT. Longer follow-up is critical to ensure disease-free survival with a functional, successfully reconstructed neovagina.

**Type of study:** Case series.

**Level of evidence:** Level IV.

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Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma of childhood. Outcomes have improved significantly over the past few decades with the implementation of a multidisciplinary approach, which relies on the selective combination of surgical resection, chemotherapy, and radiotherapy (RT) [1]. RMS arising in the female genital tract is rare, accounting for approximately 3.5% of cases, with approximately half of these cases arising in the vagina [2]. This location is

associated with a favorable prognosis, and is considered to be one of the most curable forms of RMS [3].

Despite great advances in management, cure often comes at a price. Addressing the primary tumor with either surgery or radiation can be associated with important complications. To date, an optimal and preferred approach for local control is lacking. Owing to the vagina's anatomical location and limited space for dissection within the prepubertal pelvis, these tumors are usually considered to be difficult to resect while preserving adjacent organ integrity. Therefore, RT – either as external beam or brachytherapy – is often favored. Despite good oncological outcomes, this strategy can be associated with important late effects [4]. Additionally, even many years after pelvic RT for RMS, surgical repair of these problems can be exceptionally challenging, and carries a considerable risk for further complications [4].

**Abbreviations:** ASTRA, Anterior sagittal trans-rectal approach; CAH, Congenital adrenal hyperplasia; COG, Children's Oncology Group; RT, Radiotherapy; RMS, Rhabdomyosarcoma; VAC, Vincristine, actinomycin-D (dactinomycin) and cyclophosphamide.

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To minimize side effects from local therapy, a response-based approach to local control was offered to females with vaginal RMS in a recently completed Children's Oncology Group (COG) study for newly-diagnosed, low-risk RMS. RT was eliminated for patients who achieved a complete response following chemotherapy and/or surgical resection. However, higher than expected local failure rates were observed for patients with Group IIA or III vaginal RMS [5]. This unfortunate study outcome triggered modifications to the proposed treatment approach, with the recommendation that local control is critical for these patients. In accordance with protocols for other primary RMS sites, in girls with nonresected RMS of the vagina, RT remains mandatory.

Given the frequency and severity of late effects from vaginal RT, along with possible unforeseen issues that may arise as this patient population ages, it appears prudent to reconsider the value of surgery. Complete surgical resection may allow us to preserve excellent oncological outcomes while sparing these young patients from radiation while preserving function. With that goal in mind, our group has pioneered and developed a minimally invasive approach to widely resect affected vaginal tissue and perform immediate reconstruction employing autologous grafts. Herein we present extended follow-up of the first 4 cases of localized vaginal RMS treated with a novel method of radical transperineal surgical resection (vaginectomy) followed by vaginal reconstruction using buccal mucosa grafts, without adjuvant radiation.

## 1. Materials and methods

The complete medical records from 4 patients with localized vaginal RMS surgically managed between November 2013 and June 2016 were retrospectively reviewed. All cases were consecutively planned and conducted with the involvement of the senior author of this report (AJL). Data including chemotherapy protocol, surgical approach, pathology, and current disease status were captured.

### 1.1. Surgical technique

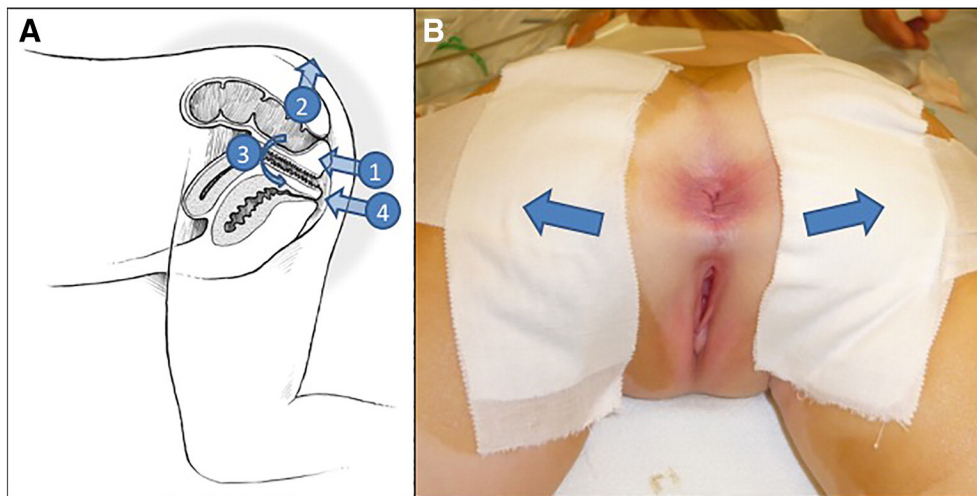
The buccal mucosa can be procured at the beginning of the case, as the remainder of the intervention will be done prone, or after resection is completed and the proper amount of tissue needed is measured. Nasotracheal intubation is favored to facilitate unhindered access to the whole oral cavity. The mucosa of each cheek is exposed with the aid of a mouth retractor and the area infiltrated with bupivacaine and epinephrine solution. Care is taken to avoid injury to the parotid gland

duct. Symmetric, bilateral, rectangular-shaped grafts are obtained in cases where total vaginal replacement is required. A more limited excision of the labial buccal mucosa of the lower lip was employed in one case where less tissue was needed. The donor sites are left open, and hemostasis is revised prior to extubation.

The patient is then positioned prone, in a modified "jackknife" position (Fig. 1A and B). Vaginoscopy is performed to confirm the location of the tumor while prone. Indwelling Foley catheters are inserted in the bladder and vaginal cavity to provide traction and aid with identification during dissection. A midline (sagittal) incision is made between the vagina and the rectum, extended posteriorly in an "inverted lambda" fashion around the anterior aspect of the rectum and anal sphincter complex (Fig. 2A). The incision can also be extended anteriorly, around the vagina, distal to the hymeneal ring (Fig. 2B).

Dissection starts by developing the prerectal space. Retraction of the anterior rectal wall delivers a good plane for exposure of the posterior vaginal wall. Alternatively, the anterior wall of the rectum can be incised following the ASTRA (anterior sagittal transrectal approach), as described for vaginoplasty in patients with congenital adrenal hyperplasia (CAH) [6] and high urogenital sinus [7]. None of the patients in this series required incision of the rectal wall, however this technique may allow for greater exposure in selected cases. The lateral walls of the vagina are subsequently developed by blunt dissection, a step that is straightforward as this is a relatively avascular plane (Fig. 3A). The most difficult part of the resection involves the anterior wall. The surgeon needs to exercise caution developing the space between the vagina and the urethra (Fig. 3B). The vagina can then be mobilized circumferentially up to the level of the cervix (Fig. 3C).

The extent of the dissection can be tailored based on the location of the tumor and assessment of margins. Unless felt to be involved (based on presentation features, vaginoscopy and assessment of margins by frozen section), the cervix is spared along with unaffected fornical mucosa near the vaginal dome. The vagina is resected *en bloc*, and margins are confirmed intraoperatively by frozen section. The buccal grafts are then sewn in place, first at the level of the vaginal dome, anteriorly and posteriorly, and then laterally to each other. Vaginal reconstruction is concluded by anastomosing the buccal grafts to the incision distal to the hymeneal ring (Figs. 4 and 5). If a more limited resection is performed, the buccal graft is sewn into place to cover the newly created defect in the vagina. The perineum is then approximated in the midline, and a pediatric chest tube tip is left in place for 10–14 days to act as a mold. The Foley catheter is removed 1–2 after surgery.



**Fig. 1.** Patient positioning for prerectal prone approach. Child is placed on the modified "jackknife" position, with gentle elevation of the pelvis (A). This allows dissection to be sequentially performed from posterior to anterior, employing the following steps: 1. Define the prerectal space, 2. Posterior retraction of the rectum, 3. Dissection of the posterior and lateral vaginal walls, and 4. Dissection of the vaginal wall from the periurethral and bladder neck area. Legs are abducted and the perineum is exposed by placing traction tape strips directed laterally and slightly superior (arrows, B).

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