Management Quandary

Case 2005: Management of Vaginal Agenesis

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Case Presentation

A 17 ½-yr-old female is referred to the multidisciplinary pediatric pelvic reconstructive surgery program for consultation. The local gynecologist was uncomfortable with management of vaginal agenesis. She had a normal +6,XX karyotype. An MRI showed normal kidneys and a normal urinary tract. The MRI suggested a midline uterine structure. An exam under anesthesia and laparoscopy revealed a 1-cm exterior vaginal dimple and bilateral non-distended uterine anlagen (Fig. 1). She and her mother present to discuss management options.

Dr. Dharamsi and Sheldon’s comments:

Vaginal agenesis is an infrequent condition that results from a number of diverse conditions, although it is most commonly associated with Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome. MRKH affects 1 in 5000 live-born females and is characteristically seen with vaginal agenesis and variable müllerian duct/renal, skeletal and auditory anomalies. Management of vaginal agenesis is challenging for both gynecologic and genitourinary surgeons and requires a multidisciplinary approach.

Management Options

Strategies for creation of the neovagina are diverse but all strive to achieve the same final outcome of a functional neovagina. Nonsurgical options, mostly variations of Frank’s (1938) original method, utilize dilators to create an artificial vagina. This technique has been successful in motivated patients with minimal indentation similar to a vaginal dimple. A recent review of 51 patients with vaginal agenesis who underwent a modification of Frank’s technique using a bicycle seat mounted on a stool (Ingram method) showed that 37 of the patients (72%) attempted vaginal dilation, with 92% of those patients attempting dilation achieving functional success (defined as satisfactorily achieving intercourse or accepting the largest dilator without discomfort). Similar studies have also shown no significant differences between patients treated with dilators versus controls in terms of sexual desire, sexual arousal and satisfaction with a sexual relationship. These results suggest that in the motivated, compliant patient, nonsurgical approaches can be highly successful. The reported disadvantages of this technique are: insufficient vaginal lubrication, decreased frequency of orgasms, and dyspareunia (in approximately 20%).

Surgical options for creation of a neovagina are varied, with no consensus regarding the best approach. The goal of the neovagina is to allow satisfying sexual interactions while also providing an egress of menses and the potential for vaginal delivery in patients with a uterus. Ideally, this neovagina has a normal cosmetic appearance, is lubricated, requires minimal maintenance, and does not require a surgically morbid procedure for its creation. At the present time, no single procedure is able to offer all of these outcomes, so controversy exists regarding the best technique.

The patient attempted dilatation of the vaginal dimple, but returned complaining that she is uncomfortable with the procedure and would like to discuss surgical management.

Drs. Dharamsi and Sheldon’s comments:

Vaginal dilation has multiple purposes. Though used more frequently by gynecologists for management of
vaginal agenesis, it also allows the patient to understand what dilation involves and it allows the surgeon to determine the degree of compliance that may be achieved with each patient. Once it is determined that a surgical approach is desired, various options need to be considered. Gynecologic surgeons are most familiar with the Abbe-McIndoe vaginoplasty which utilizes a split

**Fig. 1.** A preoperative perineal photograph demonstrating the absence of the vaginal opening.

**Fig. 2.** Perineal photograph taken intra-operatively after creation of the sigmoid neovagina.
complication rate and 5% reoperative rate found with laparoscopically. The procedure has also been performed via laparotomy however, more recent work has demonstrated that intestinal segments generally have been approached via laparotomy.

Factors to consider include: the availability of a suitable segment of appropriate length, the simultaneous performance of any necessary reconstruction (such as bladder augmentation), past family history of colorectal disease, disruption of the ileocecal valve with subsequent gastrointestinal tract morbidity, and the ability of a segment to reach the perineum without undue tension. Given these factors, sigmoid vaginoplasty has been favored with its close proximity to the perineum and its natural lubrication. Post operatively, patients with either nonsurgical or surgical neovagina construction do not require routine vaginal dilation. The patients are reevaluated under anesthesia at 6–8 weeks after the surgery and if stenosis is present, then a home dilation program may be implemented until the patient becomes sexually active.

Intestinal segments generally have been approached via laparotomy however, more recent work has shown that the vascular pedicle can be mobilized laparoscopically. The procedure has also been utilized at a variety of ages and a variety of diagnoses. A recent meta-analysis of bowel neovaginoplasty shows a complication rate of 35% and a reoperation rate of 4%. This is similar to the 34% complication rate and 5% reoperative rate found with skin graft vaginoplasty with the advantage being that bowel neovaginoplasty complications are most commonly associated with introital stenosis and can be easily managed with dilation or minor surgery.

The young woman underwent a surgical neovaginoplasty. A sigmoid neovagina was created using a combined laparotomy and vaginal approach. Genitourinary surgeons differ in their approach to managing vaginal agenesis. This group advocates first line surgical management, because it immediately corrects the defect, versus a conservative (dilation) approach which takes on average of 11 months to create a neovagina. Genitourinary surgeons are most comfortable with vaginal reconstruction utilizing intestinal segments including ileum, cecum, sigmoid, and rectosigmoid colon (Fig. 2). Each segment has its own inherent advantages and disadvantages and thus utilization of a certain segment in a given patient needs to be individualized. Factors to consider include: the availability of a suitable segment of appropriate length, the surgical morbidity associated with introital stenosis and can be easily managed with dilation or minor surgery.

Dr. Dharamsi and Sheldon’s comments:

A number of issues need to be addressed preoperatively. The patient should be given informed consent about the risks of the procedure. In addition to the surgical risks of bleeding, infection and anesthesia, more specific risks include: the risks of laparotomy with a bowel anastomosis (leakage at the anastomosis site), neovaginal stenosis requiring dilation, excessive vaginal mucus production, dyspareunia, and the small risk of subsequent adenocarcinoma in the neovagina. Preoperative preparation of the bowel is also mandatory. Mechanical and antibiotic preparation before surgery is essential for optimal success of the anastomosis. Other issues of importance include the management of the uterine anlagen with some advocates supporting maintenance of uterine anlagen, while others feel that all uterine tissue be removed.

Post Procedure Care

Patients in whom intestinal segments are utilized for vaginal construction do not require routine vaginal dilation. The patients are reevaluated under anesthesia at 6–8 weeks after the surgery and if stenosis is present, then a home dilation program may be implemented until the patient becomes sexually active.

Post operatively, patients with either nonsurgical or surgical neovaginoplasty have complaints of dyspareunia related to poor vaginal lubrication. This is in contrast to patients with an intestinal neovagina; these patients complain of excess mucus production requiring up to two pads per day. A number of authors have reported that the mucous production decreases within the first few months. However, despite adequate lubrication, authors have reported that sexually active patients with prior intestinal vaginoplasty complain of dyspareunia. The etiology of this dyspareunia is unclear. In addition to other complications associated with either type of repair, both procedures have been associated with carcinoma of the neovagina. These are extremely rare but are associated with neovaginal squamous cell carcinoma in patients with skin grafts and adenocarcinoma in patients with intestinal segment neovagina. Given these concerns, surveillance and counseling are of importance for any patient undergoing neovaginoplasty.

To date, no single procedure meets all the desired goals and new innovations continue to be developed. It is of importance to understand the various options...
available and to be aware of the shortcomings of each. This will allow the patient to be matched with the most ideal procedure. Further investigations are needed to determine the psychosexual and psychosocial impact of this surgery on the patient. Patients must continue to be approached from a multidisciplinary view to ensure optimal management.

References

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