Management of Vaginal Agenesis

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A B S T R A C T

Rokitansky syndrome and complete androgen insensitivity syndrome are the most common causes of vaginal agenesis. Treatment should be deferred until adolescence to allow informed consent and compliance. The best treatment for vaginal agenesis remains controversial although vaginal dilation therapy is still widely considered the first line treatment because success rates are high and associated risks are low. A variety of surgical options are also available, each with enthusiastic proponents. Long-term outcome studies on most surgical techniques, however, are still lacking and until recently most studies have reported on success rate in terms of anatomical success only, without including sexual function. Moreover, the medical literature lacks prospective comparative outcome studies, meaning that current choice of surgical procedure relies greatly on the surgeon’s preference and experience.

Key Words: Vaginal agenesis, Vaginal hypoplasia, Vaginal aplasia, Vaginal dilation, Vaginoplasty, Rokitansky syndrome, Complete androgen insensitivity syndrome

Introduction

Congenital vaginal agenesis is most commonly seen in women with Rokitansky syndrome (also referred to as Mayer-Rokitansky-Kuster-Hauser syndrome or Müllerian aplasia), and complete androgen insensitivity syndrome (CAIS). In addition it can be associated with other rarer complex conditions affecting the urinary and gastrointestinal tracts, such as cloacal and anorectal anomalies. Women with Rokitansky syndrome have a XX karyotype and normal functioning ovaries but an absent or rudimentary uterus and a short vagina resulting from failed embryonic development of the Müllerian duct. It occurs in one in 5,000–10,000 female births; however, the exact etiology is still unknown. Women with CAIS, which affects one in 13,000–40,000 live births, have 46,XY karyotype, testicular gonads, absent Müllerian structures, and a short vagina. This is attributed to a mutation in the androgen receptor gene which renders the body insensitive to testosterone and hence results in a female phenotype with female external genitalia. Since patients with Rokitansky syndrome and CAIS have normal breast development either from normal ovarian hormones in the former condition or from peripheral conversion of testosterone to estradiol in the latter one, these patients typically present in adolescence with primary amenorrhea. Detailed investigation is required to diagnose and differentiate these conditions; however, in a clinical setting the vaginal findings are identical with an absent or short vaginal dimple. Surgical and non-surgical treatments are available to lengthen the vagina and facilitate penetrative sexual intercourse.

Timing of Treatment

In the majority of cases these conditions present in adolescence which means the patient can be fully involved in decisions about the type and timing of treatment. Where presentation is earlier in childhood, it is accepted that both non-surgical and surgical methods of vaginal creation are best deferred until adolescence or even adulthood when the patient has reached physical and psychological maturity. This allows for proper decision making and also increases the compliance with vaginal dilation therapy whether used as primary treatment or post-operative adjuvant treatment to prevent vaginal stenosis.

The requirement for vaginal lengthening before intercourse is of course only part of the devastating implications of these conditions. Both conditions result in infertility. In CAIS disclosure of an XY karyotype and the decision about gonadectomy are difficult concepts for an adolescent girl to manage. As the risk of malignancy is found to be low before puberty, the current standard treatment is to delay gonadectomy until puberty. This allows pubertal changes to occur as a result of peripheral conversion of testosterone to estradiol. In addition, delaying gonadectomy will provide the time for the patient to be involved in the decision. All patients with disorders of sex development should be cared for by a multidisciplinary team which includes a psychologist.

Treatment Choice

The best treatment for vaginal agenesis is still controversial. In the USA and UK vaginal dilation therapy is considered the best first-line treatment2 and surgery is reserved for cases when vaginal dilation therapy fails or when a patient is ineligible for vaginal dilation due to previous perineal surgery. However, in many European
countries, surgical vaginoplasty such as a laparoscopic Vecchietti or Davydov procedure is the first line treatment and vaginal dilation is only used post-operatively. There is a lack of long term outcome data on surgical and non-surgical options. In addition there are no comparative studies of different techniques. It must be remembered that the main objective of surgery is not just to create a passageway for penetration but to facilitate enjoyable sexual intercourse. Only recently has sexual function data been included in outcome studies. At present the choice of the method depends on the genital configuration, previous surgical attempts, and, above all, the surgeon’s preference and expertise.3

Nonsurgical Methods

Vaginal Dilation

Vaginal dilation therapy for vaginal agenesis was first described by Frank in 1938.4 It involves the use of vaginal molds, initially made from pyrex and currently made from plastic, of increasing width and length. The patient is asked to apply gentle pressure on the vaginal dimple for at least 30 minutes daily for several months with the aim to achieve a vaginal length of 7–8 cm. This therapy works by the pressure effect which acts to progressively stretch the vagina with time. Thus the prerequisite for this treatment is first a healthy non-scarred vaginal dimple which is amenable to stretching and second, but more important, a motivated patient who is ready to adhere to such a lengthy treatment.

Because vaginal dilation is a non-invasive and inexpensive method with a high success rate, of up to 90%,5 it has been recommended as first line treatment for vaginal agenesis by several medical bodies including the American College of Obstetricians and Gynecology Gynecologists (ACOG).2 However, the definition of “success” in earlier studies has been criticized6 as it was either vaguely defined or mainly focused on the “anatomical success”.5,7,8 Later studies included sexual function as an integral element of success, which highlighted the difficulties encountered with this type of therapy. These include low compliance and patient dissatisfaction9 as the regime was described by patients as distasteful,10 painful and a constant reminder of their abnormality.6 Despite the fact that vaginal dilator therapy is mainly patient driven, psychological support and supervised programs are necessary to maintain patients’ motivation. The obstacles to dilation therapy can be overcome when treatment is delivered by a multi-disciplinary team which acts to boost patients’ confidence as described by Ismail-Pratt et al11 who reported non-painful sexual intercourse in 81% of participants with a background anatomical success rate of 86%. Another study also concluded that despite the difficulties encountered with lubrication, pain, and orgasm, the degree of satisfaction with the overall sexual life was comparable to the control population.12 However, the condition itself does have a negative impact on sexual and emotional wellness and anxiety levels are higher in women with Rokitansky syndrome who have undergone vaginal dilation treatment compared to those who did not require vaginal dilation.13

The timing to commence dilation therapy is elective and is best planned when the patient is emotionally mature.2 The question remains whether the patient should be given the option to choose between vaginal dilation or first line surgical option, bearing in mind that vaginal dilation might still be needed as adjuvant treatment after surgical reconstruction.

In fact the merits of vaginal dilation therapy extend beyond its noninvasive nature to other important aspects. First of all, even when dilation therapy fails, Routh et al14 have proven that initial progressive perineal dilation followed by vaginoplasty would still be more cost-effective than initial vaginoplasty in 99.99% of simulations. In addition, until now the key focus of clinical management has been to increase vaginal size and to achieve penetrative sexual intercourse. However, the functional success of vaginoplasty is not necessarily proportional to the length of the new vagina. Initiating vaginal dilation therapy can prompt concomitant psychological support which should not only improve the outcomes of any treatment but would also orient patients towards realistic expectations even when surgery is still needed.

Surgical Methods

Surgical vaginoplasties can be subdivided into the following categories:

1. Creation of a perineal pouch. This includes the Williams vaginoplasty and subsequent modifications.
2. Lining a neovaginal space. This includes procedures based on the McIndoe technique where a neovaginal space is dissected between the bladder and rectum, then lined with different types of tissue.
3. Laparoscopic procedures. These methods include the Vecchietti procedure and Davydov procedures.
4. Intestinal vaginoplasty.

Creation of a Perineal Pouch

Initially described by Williams in 1964, this technique fell into general disuse until modified by Creatsas et al in 2001.15 It involves a U-shaped incision extending from the medial side of the labia at the level of the external urethral meatus down across the perineal body to form a skin flap. The tissues are then mobilized and sutured in layers to form a pocket in the perineum to allow coitus.3 In a study on 178 Rokitsansky patients, anatomical success was achieved in 96% while 94% reported sexual satisfaction in a non-validated follow-up assessment.16 Possible complications of the Creatsas vaginoplasty are hematoma, wound opening, and infection in addition to excessive hair growth from the skin flap. It is difficult to explain the success of the Creatsas modification as the Williams procedure fell into disuse because the perineal pouch was often too short and superficial for comfortable penetration. It is likely that the
Creatasas procedure supports the posterior fourchette to allow coital dilation to extend the vaginal length.

Lining of a Neovaginal Space

This approach is based on the McIndoe procedure which was first pioneered by Abbe and further popularized by McIndoe and Banister in 1938. The use of split-thickness skin graft to line a neovagina has become one of the most commonly used techniques especially in the United States although used less frequently in Europe. The procedure involves a perineal approach to create a space between the rectum and urethra followed by the use of a split-thickness skin graft, usually harvested from the buttock area, to form the epithelium of the neovagina. The graft is usually introduced into the neovagina wrapped over a mold which is left in the vagina for a variable period of time to allow the graft to take. The patient is then required to dilate the vagina post-operatively to prevent stenosis. Good anatomical results have been reported in large series with several modifications introduced to the original procedure described,[17,18] although studies were retrospective with no objective sexual function outcome data. The main advantages of this procedure are the simplicity of the technique and the avoidance of an abdominal incision. However, drawbacks to this procedure have been reported, including the visible donor scar, inadequate lubrication causing dyspareunia, and the high rate of neovaginal stenosis necessitating the need for long-term dilation.[19] Long-term sexual function results have not been validated; however, sexual satisfaction rates have been reported to be between 80–90%.[20]

Other tissues have also been used in order to avoid some of the disadvantages of a split skin graft. In 1934, Brindeau described a novel approach using chemically processed and sterilized freeze-dried human amnion to line a neovaginal cavity created according to the McIndoe procedure. The main aim was to substitute the use of split-thickness skin graft which leaves significant scars at the donor site with human amniotic membranes that are considered immunologically inert and thus have low risk of graft rejection. Various authors have reported on this technique,[21–23] however, all these studies include only small numbers of patients and this technique has fallen into disuse especially with the theoretical risk of viral transmission through the allograft.

Various other authors have reported on the use of several artificial or biologic materials to cover the neovaginal cavity and induce epithelialization including buccal mucosa,[24,25] artificial dermis and recombinant basic fibroblast growth factor,[26] oxidized cellulose,[27] acellular human dermal allograft,[28] and autologous in vitro–cultured vaginal tissue.[29]

Vecchietti Procedure

Initially described in 1965 as an open operation, the Vecchietti procedure is now usually performed laparoscopically.[31] It has gained popularity in Europe although the instruments appear not to be FDA approved in the United States[8] despite the fact that the ACOG lists it as one of the surgical options for neovaginal creation. Unlike vaginal dilation therapy which requires active pressure applied by the patient, the Vecchietti procedure dilates the vagina by passive traction on an ovoid bead or mold placed in contact with the vestibule and attached to the abdominal wall by traction wires that are threaded retroperitoneally. The threads are pulled by 1 cm each day to create a neovagina of 10–12 cm in 7–8 days. The neovagina re-epithelializes to form a normal mucosa with mild structural and ultra-structural modifications.[32]

Several studies have reported the success of this procedure.[31,33–35] In a large study including 110 patients, anatomic and functional success was achieved in 97% and 98% respectively.[36] The anatomic success was defined as a vagina of at least 6 cm in length and which admits two fingers within 6 months after surgery. However, the inclusion criteria for this study are not clearly described to show whether these patients were counselled for vaginal dilation therapy as first line treatment. In other words, if a subgroup of these patients were eligible for vaginal dilation then the success rate might be falsely inflated. This is not to say that the Vecchietti procedure is unsuccessful but to stress the importance of counselling patients on the best option and least invasive procedure that would suit their condition.

In terms of sexual function outcomes, no significant difference was found between patients and controls with respect to desire, arousal, and satisfaction; while patients had lower scores for the domains on lubrication, orgasm, and comfort compared to controls.[37] The complication rate including bladder and rectal wall injury is reported to be low[38] even in the presence of a pelvic kidney.[39] However, a more common drawback for this procedure is the pain resulting from the sustained traction which necessitates hospital stay throughout the whole traction process.[3] Overall, laparoscopic Vecchietti procedure is considered a safe and successful surgical option for the creation of a neovagina in patients who failed dilation therapy and have not had any previous reconstructive surgery.

Balloon Vaginoplasty

This approach mimics the Vecchietti procedure. A foley catheter introduced laparoscopically into the rectovesicle space is used instead of the traction wires, while the catheter balloon is inflated progressively to stretch the vagina. It is claimed that compared to the small and solid acrylic “olive”, balloon distension would stretch the vagina not only in length but also in width with less pain and discomfort to the patient. Successful anatomical results in addition to non-validated sexual satisfaction have been reported in patients with Rokitansky syndrome and CAIS.[38,39] However, these studies included only a small
number of patients and therefore larger studies with long term follow-up are still needed.

**Davydov Procedure**

As with the Vecchietti procedure, the Davydov procedure was initially described as an open procedure. It was then modified to a laparoscopic approach which is more commonly used today although the former is still used in some countries. This method consists of a three-stage operation involving dissection of the rectovesical space with abdominal mobilization of the peritoneum, attachment of the peritoneum to the created introitus, and finally closure of the neovaginal vault with purse-string suture. After the operation patients are instructed either to keep a mold in the vagina or to perform regular vaginal dilation to avoid vaginal collapse before the complete epithelialization has occurred. Unlike the Vecchietti procedure, the Davydov technique can be used for patients who have genital scarring due to previous perineal surgery because it does not require the vaginal skin to be elastic.

Several series have been published on the successful use of the Davydov procedure. Giannesi et al reported anatomic success in 26 out of 28 patients studied while Fedele et al reported 97% and 96% rates for anatomic and functional success, respectively. Reported complications of this procedure include bladder and ureteric injury, vesicovaginal fistula, and septic peritonitis.

Conflicting outcomes have been described in terms of sexual function. On one hand the overall sexual function scores in the Giannesi study were comparable to controls with a slight trend towards less satisfaction in the domains of comfort and pain only. Fedele et al reported similar findings where no significant differences were found in the domains of arousal and satisfaction, although slightly lower scores were found in the domain of lubrication, desire, orgasm, and pain. On the other hand, a small study on the psychosexual outcome of the Davydov procedure reports Female Sexual Function Index scores that were statistically significantly lower, both as a total value and in the domains of arousalability, lubrication, orgasm, and comfort in a group of XY women who had failed previous vaginal dilation and surgical procedures. This last paper highlights the need for careful reading of the literature especially in terms of patient selection. Women with Rokitansky syndrome who have not had prior reconstruction have a normal vaginal introitus and supple vaginal skin and will do well with dilation and most surgical procedures. Women with complex psychological requirements and previous genital reconstruction will have poorer outcomes from any treatment. It can be argued that the risks of the Davydov procedure make it an inappropriate first line treatment to offer women who would have been successful with vaginal dilation where the associated risks are extremely low.

**Intestinal Vaginoplasty**

The use of a section of intestine to replace the vagina was first described by Baldwin in 1904 when he used a segment of the small bowel to create a neovagina. However, due to the high mortality and morbidity rates, this procedure was abandoned and was only revisited in the early 1970s. The procedure involves the isolation of a segment of the bowel, usually 10–12 cm in length, along with its vascular pedicle which is then transposed to the perineum at the level where the vagina should be. The distal sigmoid colon is most commonly used for this purpose due to its location, making the operation technically easier compared with small bowel where the short mesentery at many times prevents tension-free anastomosis. In addition, the less fragile colonic mucosa compared to the ileal mucosa makes it less susceptible to trauma and bleeding. On the other hand, however, advocates for ileal vaginoplasty report that the ileum has a lower risk of diversion colitis and excessive mucous production due to the optimal fluid equilibrium present in the distal ileum. In women who have had multiple reconstructive procedures before, the choice of bowel segment is more pragmatic as it will be dictated by which segment of the bowel will reach into the pelvis without undue traction. Unlike the Vecchietti and Davydov procedures which are now invariably done laparoscopically, laparoscopic intestinal vaginoplasty has not gained popularity as this procedure is still more frequently done through a laparotomy. Laparoscopically assisted sigmoid vaginoplasty is claimed to have better cosmetic results, more rapid recovery of intestinal function and less surgical trauma.

Overall, the main advantages to this operation are the adequate vaginal length, the lack of shrinkage and the natural lubrication provided by the mucous production which obviates the need for artificial lubricants and decreases the risk of dyspareunia. Moreover, long-term vaginal dilation is not required. Several studies have reported excellent cosmetic results and psychosexual outcomes that approached normal controls. However, many of these studies included patients with Rokitansky syndrome and the indication for intestinal vaginoplasty is not clearly stated. This would suggest these patients might have undergone an unnecessary complicated procedure when vaginal dilation could have achieved the same outcome. This is a complex procedure with a complication rate of up to 19–36% and therefore should be reserved only for selected cases such as complex cloacal anomalies with absent vaginal dimple making the use of vaginal dilation therapy or other less complicated surgical approaches inapplicable. The most common complications of intestinal vaginoplasty include excessive mucous production, stenosis of the neovagina, prolapse, and diversion colitis of the bowel vagina. In addition, adenocarcinoma in the grafted bowel has been reported but fortunately with a low incidence.

**Conclusion**

At present there is still no agreement on the gold standard treatment for vaginal agenesis. Most gynecologists in the USA and the UK agree that vaginal dilation therapy should be the first line treatment as it is successful without...
the risks of surgery. However, vaginal dilation is time consuming and some patients find it distressing. Short term outcome data on the laparoscopic Vecchietti procedure has also confirmed good success rates and low morbidity and this may also be an appropriate first line treatment for women with vaginal agenesis without previous failed reconstructive surgery. It is, however, difficult to justify proceeding directly to some of the more complex techniques such as intestinal vaginoplasty with its associated high morbidity and long term side effects.

Choosing the right operation in patients who require surgical reconstruction is integral to success. However, given the current available evidence base, it is very difficult for clinicians to compare the results of the various surgical techniques used and to determine the superiority of one over the other. This is due to various problems including the heterogeneous group of patients included in these studies, the unavoidable reporting bias of surgeons towards the techniques they are mostly experienced in, and the difficulty of long term follow-up. Head-to-head prospective long-term studies are still needed to compare the outcome of the various techniques. Meanwhile, patients should be counselled on the benefits and risks of various techniques including vaginal stenosis or shrinkage, lack of lubrication, prolapse,57–59 diversion colitis,60 and rarely malignancy.61–63 In addition, the essential role of psychologic assessment before and after any treatment should be emphasized.

References

1. Michala L, Cutner A, Creighton S: Surgical approaches to treating vaginal agenesis. BJOG 2007; 114:1455
11. Ismail-Pratt IS, Bikoo M, Liao LM, et al: Normalization of the vagina by dilator and rarely malignancy.61 Meanwhile, patients should be counselled on the benefits of surgery. However, vaginal dilation is time consuming and some patients find it distressing. Short term outcome data on the laparoscopic Vecchietti procedure has also confirmed good success rates and low morbidity and this may also be an appropriate first line treatment for women with vaginal agenesis without previous failed reconstructive surgery. It is, however, difficult to justify proceeding directly to some of the more complex techniques such as intestinal vaginoplasty with its associated high morbidity and long term side effects.

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References
60. Syed HA, Malone PS, Hitchcock R J: Diversion colitis in children with colovaginoplasty. BJU Int 2001; 87:837