Bowel vaginoplasty in children

Y. K. Sarin, D. Pathak, M. Sengar
Department of Pediatric Surgery, Maulana Azad Medical College, New Delhi - 110 002, India

Correspondence: Yogesh Sarin, Department of Pediatric Surgery, Maulana Azad Medical College, New Delhi - 110 002, India. E-mail: yksarin@hotmail.com

ABSTRACT

Objectives: To describe our experience with bowel vaginoplasty done in children.

Materials and Methods: This is a retrospective study of eight children aged 10 months to 8 years, who underwent bowel vaginoplasty over a period of 5 years (2000-2005). The indications of bowel vaginoplasty included anorectovestibular fistula (ARVF) associated with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome (n=6) and cloaca (n=2). The bowel segment used for vaginoplasty included colon (n=3), ileum (n=2) and duplicated rectum (n=1). In two patients of ARVF associated with uterovaginal agenesis, the distal-most part of ARVF was transected at the level of peritoneal reflection and left as neovagina, whereas the proximal bowel was pulled through at the proposed neo-anal site. All the patients were advised daily home dilatation of the neo vaginal orifice with Hegar’s dilators, for a period of six weeks.

Results: Bowel vaginoplasty was done in eight patients. None had any significant per-operative complication. Two patients had abdominal wound dehiscence, requiring secondary suturing. Two patients had mucosal prolapse of the neovagina, which required trimming. One patient died two months after discharge, because of meningitis. Out of the eight patients, seven are in regular follow-up. Six patients have neovagina, cosmetically acceptable to the parents; all have been radiologically proven to have adequate length. One patient had unacceptable perineal appearance with nipple-like vaginal orifice and scarred perineal wound, that merits a revision. None of the patients had vaginal stenosis and excessive mucus discharge, during follow-up visits. Although post surgical results are acceptable to the parents cosmetically, the sexual and psychological outcome is yet to be assessed.

Conclusions: Bowel vaginoplasty is a safe and acceptable procedure to treat the pediatric patients of uterovaginal agenesis and cloaca.

KEY WORDS: Uterovaginal agenesis, bowel vaginoplasty, vaginal reconstruction, mayer-rokitansky- kuster-hauser syndrome, cloaca

The surgical management of absence of the vagina in pediatric patients constitutes a significant technical challenge, the outcome of which affects both the physical and psychological health of the patients and the parents.[1] Typically, congenital syndromes such as vaginal agenesis (MRKH syndrome) and intersex conditions such as androgen insensitivity syndrome, congenital adrenal hyperplasia and gonadal dysgenesis, are the most common indications for vaginal reconstruction in the pediatric patients. However, defects acquired from extensive resection of the pelvic tumors also may require vaginal reconstruction.[1] Different therapeutic options available in agenesis of vagina, include non-operative method (Frank technique),[2] free skin graft or McIndoe method, [3] sigmoid vaginostomy,[4] amnion graft,[5] pedunculated skin graft[6] and pelvic peritoneal graft.[7] The use of enteric segment was reported as early as 1904.[8] The use of ileal, ileocecal and sigmoid colon has been reported.[9,10] We report our experience of bowel vaginoplasty in eight patients.

MATERIALS AND METHODS

Eight children aged 10 months to 8 years underwent bowel vaginoplasty, over a period of 4½ years (2000-2005). The indications of bowel vaginoplasty, included anorectovestibular fistula (ARVF) associated with uterovaginal agenesis (n=6) and cloaca (n=2).

The bowel segment used for vaginoplasty included colon (n=3) and ileum (n=2). In one patient of ARVF associated with uterovaginal agenesis, there was a duplicated blind rectum opening in the vestibule; the same was used mobilized anteriorly and left as future vagina. In two patients of ARVF with uterovaginal agenesis, the diagnosis was suspected preoperatively. In these two patients, distal 8-10 cms of the ARVF was left undissected, as a future vagina and bowel proximal to that, was pulled through as neoanus.

The procedural details of bowel vaginoplasty have been...
discussed before in literature\cite{11} [Figure 1] and are not being repeated here. The important steps that need to be highlighted include: isolation of bowel segment on vascular pedicle, closing one end and rotating the isolated segment in to the pelvis and sutureing the pulled-through isolated vascular segment to the perineal skin, to act as the neovagina.

One of the patients with ARVF and uterovaginal agenesis had three orifices in the vulva to start with and was clinically diagnosed as the usual ARVF. Intraoperatively, the middle orifice was seen to emit feculent returns, whereas the posterior orifice in the vestibule led to a blind rectal duplication measuring 8 cm in length. The duplicated blind rectum was mobilized anteriorly and the ARVF was pull-throughed as neo-anus in the perineum [Figure 2].

Of the two girls with cloaca in this series, it is important to mention that both the patients had concomitant congenital pouch colon. One of the patients having type III congenital pouch colon (CPC) and cloaca, had undergone excision of pouch and end colostomy and the distal end was left as a 2 cm deep blind anorectum. She underwent urethrovaginoplasty through the posterior sagittal route at two years of age; the end colostomy was pulled down as neoanus. Postoperatively, she developed a large urethrovaginal fistula. Six months, later she underwent transpubic closure of urethrovaginal fistula. The large defect in the anterior wall of vagina necessitated reconstruction of most of the vagina, using a segment of the proximal colon on a vascular pedicle.

The other patient of cloaca who underwent bowel vaginoplasty, also had concomitant CPC. She presented to us the first time at 6 years with multiple perineal explorations, done elsewhere. The ‘resident’ surgeon failed to recognize the concomitant type III CPC and performed divided colostomy, just proximal to the pouch. At definitive surgery, the pouch was diagnosed, it was tubularised and pulled through as neoanus; the proximal stoma was left intact and the lower tubularised pouch colon was left in as is described for Hartmann’s procedure. During the surgery, the child was also diagnosed to have long urogenital sinus and right unicorneate uterus. The urogenital sinus was left as urethra, while a segment of ileum on the vascular pedicle was mobilized to reconstruct the vagina [Figure 3].

All the patients were advised daily home dilatation of the neo vaginal orifice with Hegar’s dilators for a period of six weeks; the parent’s compliance to this advice was excellent.

---

Figure 1: Surgical steps of bowel vaginoplasty: segment of bowel being isolated. Inset depicts rotation of the isolated segment

Figure 2: Line diagram of patient of ARVF with uterovaginal agenesis with duplicated rectum having three perineal openings. Inset depicts the duplicated rectum used as neo-vagina, ARVF pulled through as neoanus

Figure 3: Line diagram depicting ileal vaginoplasty done in a patient of cloaca associated with congenital pouch colon
RESULTS

None of the eight patients undergoing bowel vaginoplasty, had any significant per-operative complication. Immediate post-operative period was also uneventful for four of them. Two patients had abdominal wound dehiscence that required secondary suturing. One patient who had rectal duplication died two months after discharge, because of meningitis.

All the seven survivors are in regular follow-up, ranging from 1 month to 5 years. Two patients had mucosal prolapse of the neovagina, that required trimming. Six patients have neovagina, which is cosmetically acceptable to the parents. One patient had unacceptable perineal appearance with nipple like vaginal orifice and scarred perineal wound [Figure 4]. Vaginograms have been performed postoperatively for all of them; and they have been proven of adequate length radiologically. None of the patient had vaginal stenosis during follow-up visits. Also, none of them had excessive mucus discharge.

Although post surgical results are acceptable to the parents cosmetically, the sexual and psychological outcome is yet to be assessed.

DISCUSSION

The major indications for vaginal reconstruction include, varying degrees of mullerian failure such as Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome and intersex states and previous pelvic exenteration for tumor (e.g., rhabdomyosarcoma)

Congenital vaginal agenesis (MRKH syndrome) with reported occurrence of 1/4000 to 1/5000 live births, is the most common indication of replacement vaginoplasty.[11] Characteristically, the uterus is rudimentary in these patients and lacks a lumen. Vagina is also absent, although normal fallopian tube and ovaries may be present. These patients are seldom diagnosed at an early age; diagnosis is frequently not made until adolescence, when the patients present with amenorrhea.[12] No wonder that these patients are seen by the gynecologists and not by pediatric surgeons!

However, occasionally MRKH syndrome is associated with an anorectal malformation (ARM); the latter is too obvious an anomaly to be overlooked and a pediatric surgeon is consulted. It is pertinent to highlight here, that traditionally, a female child with imperforate anus and two orifices in the vulva, was believed to have anorectal agenesis and rectovaginal fistula. But going by the experience of the principal author, as well as the recently available literature, the incidence of anorectal agenesis and rectovaginal fistula and that of ARVF associated with uterovaginal agenesis, is comparable.[12] In fact, the principal author didn’t witness a single case of anorectal agenesis and rectovaginal fistula, but came across half a dozen of patients with ARVF associated with uterovaginal agenesis, in the study period. Although less then 25 cases of MRKH syndrome associated with ARM have been reported in the world literature,[13] this association is probably not that rare and is grossly under-reported. In the experience of the principal author, as many as 10% of all patients of ARVF have associated MRKH syndrome (unpublished data). Unfortunately, in only two out of six of such patients included in this series, the diagnosis of ARVF associated with uterovaginal agenesis was suspected preoperatively. In the other four, standard dissection of ano-rectum was started before uterovaginal agenesis was recognized and the authors were left with the only option of a concomitant bowel vaginoplasty, either using colon or ileum.

The options for treatment of vaginal agenesis include non-operative and operative methods. The ideal reconstructive procedure should provide a vagina that has an appropriate length and that requires minimal, if any, dilatation. The ideal neovagina should not scar, stenose, or contract and should be cosmetically acceptable.

The non-operative Frank’s technique involves sequential dilatation of the vaginal remnant. The split skin graft (Mc Indoe) technique is popular with the gynecologists and plastic surgeons. Rotational flaps have also been used to create a neovagina. These have included pudendal thigh flaps,[14] gracilis myocutaneous flaps,[15] and labia minora flaps.[16] All these techniques are associated with

Conclusion: The ideal solution for congenital vaginal agenesis is a neovagina that is cosmetically and functionally acceptable. The present experience provides an interesting insight into the surgical and medical field of vaginal reconstruction and a safer approach to the patients of MRKH syndrome and ARM.

Figure 4: Perineum after bowel vaginoplasty-good and bad results
discomfort and requires a long period for adequate results. Moreover, it is associated with significant dyspareunia, because of dryness during intercourse. There is also concern regarding cosmetic appearance, after harvesting large amounts of tissue from the groin and thigh with rotational flaps. The experience of the principal author has been, that rotational flaps result in a rather shallow vaginal pit!

Bowel vaginoplasty is another widely accepted method for neovagina formation. Either ileum or sigmoid colon can be used for bowel vaginoplasty. Sigmoid colon is considered better than the ileum, because of presence of longer mesentery, lesser post-operative problem of mucous secretion and wider caliber. We however didn’t find much difference in our preliminary experience.

We prefer bowel segment for vaginoplasty, because it is associated with number of advantages like (1) minimal likelihood of “poor take” or later contraction, because a well vascularized epithelial-lined tube is used; (2) initial length is no problem and depth can be managed without a mold and with minimum dilatation; (3) spontaneous mucus production matches that of normal vagina; (4) dyspareunia is avoided by the greater resistance of the sigmoid segment to local trauma; (5) unassociated with external scarring, unlike free skin grafts; (6) minimal long-term care is required, although surveillance must be maintained at regular intervals to watch for adenocarcinoma in the transplanted bowel segment.[7]

It is important to discuss here, that in patients of anorectovestibular fistula with uterovaginal agenesis, the surgery for neovagina formation using any of the aforementioned procedure could be avoided. A simple procedure of leaving the distal most part of the ARVF undissected and cutting and closing it about 8-10 cms proximally, will leave the distal most part of ARVF as future vagina and the bowel just proximal could be pulled through as neoanus [Figure 5]. This procedure utilizes the native tissue as vagina, leaves a relatively scar-less perineum and does not require bowel anastomosis. We have done this procedure in two of our patients of ARVF with MRKH syndrome. The time consumed for surgery was significantly reduced and the final cosmetic appearance was extra-ordinary. This surgery does sacrifice the possible internal sphincter tissue as well as the rectum and may theoretically have deleterious effects on fecal continence. But the short-term results (ranging over 5 months to one year) in the two patients where we used such a technique, had been satisfactory.

The patients of cloaca are another group of patients that require substitution for vagina. Total genital reconstruction by posterior sagittal urethropaginoanorectoplasty (PSVUARP) is not possible in patients of cloaca with long common channel (Raffensperger’s classification type 1)[17] and these patients require replacement vaginoplasty. Bowel vaginoplasty is also indicated in patients having severe breakdowns following PSVUARP.

The other indications for vaginal reconstruction include intersex conditions and post exenterative surgery done for vaginal malignancy.[11] In our experience, reduction of the phallus and minimal cutback procedure to expose the orifice of the pseudovagina, was sufficient for the male pseudohermaphrodites, that were raised as girls. Advancement in chemotherapy has replaced the exenterative surgery done for vaginal neoplasms and virtually eliminated one of the indication of vaginal reconstruction.[18]

Counseling of the parents regarding the disease process, the management options available and the need for long-term follow-up, is one issue that is very important, but is often neglected. The management requires careful counseling of the parents by the primary care physician, pediatric endocrinologist, pediatric urologist and psychiatrist. In addition to the surgical and medical options, other relevant issues for discussion include future fertility, capacity for future sexual intercourse and genital appearance.

REFERENCES

1. Frank RT. The formation of an artificial vagina without operation.
Bowel vaginoplasty in children