CASE REPORT

Sigmoid vaginoplasty in testicular feminising syndrome: surgical technique, outcome and review of the literature

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SUMMARY

Vaginal agenesis occurs in approximately 1:5000 live female births. It results from failure of the sinovaginal bulbs to develop and form the vaginal plate. Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is the most common cause of vaginal absence followed by complete or partial androgen insensitivity syndrome. Treatment of these patients encompasses a spectrum from simple non-operative dilation to the more complicated surgical creation of a neovagina. We present a case of a patient with testicular feminising syndrome who was reared as a female and underwent bilateral gonadal excision and sigmoid vaginoplasty.

BACKGROUND

Most of the studies on vaginoplasty focus on Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome or disorder of sexual differentiation. Few studies have reported outcomes of vaginoplasty in patients with testicular feminising syndrome because it is uncommon. We present the surgical technique and outcome in a patient of complete androgen insensitivity syndrome (CAIS) who underwent sigmoid vaginoplasty at our centre.

Figure 2 Bilateral incompletely descended gonads.

CASE PRESENTATION

An 18-year-old married woman presented to us with history of primary amenorrhoea and inability to consummate her marriage. On evaluation, she was found to have normal secondary sexual characteristics. Local examination revealed well developed labia and clitoris with shallow vaginal pit (figure 1). Pubic and axillary hair was present; breast development was Tanner stage 4. Two soft swellings were palpable in the groin, one in the

Figure 1 Shallow vagina.

Figure 3 Karyotyping showing XY genotype.

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Figure 4 Left gonadectomy.

labial fold on the right and the other in the inguinal region on the left; both were assumed to be gonads ([figure 2](#)). A provisional diagnosis of CAIS was made; karyotyping was performed, which confirmed the diagnosis (46 XY) ([figure 3](#)). After

Figure 5 Right gonadectomy.

Figure 6 Approximately 12 cm of sigmoid colon isolated on its vascular pedicle.

prolonged discussion with the patient and her parents, she underwent sigmoid vaginoplasty under general anaesthesia following written consent and proper counselling.

INVESTIGATIONS

MRI of the pelvis and inguinal region showed incompletely descended gonads with absence of ovaries and uterus.

Serum testosterone levels were markedly elevated: 1339 ng/dL. LH levels were also raised: 23 mIU/mL.

DIFFERENTIAL DIAGNOSIS

The primary differential diagnosis was MRKH syndrome. In MRKH syndrome, the karyotype is XX. In patients with testicular feminising syndrome (as in our patient), the karyotype is XY with high androgen levels.

Figure 7 Edges of the vaginal pit sutured to the distal end of sigmoid segment.

Figure 8 Vaseline soaked vaginal mould kept in the neovagina for 48 h.

In MRKH syndrome, MRI of the pelvis may show ovaries along with a rudimentary uterus. Presence of gonads and absence of ovaries and uterus are suggestive of CAIS, as in our case.

TREATMENT: SURGICAL TECHNIQUE

The peritoneal cavity was entered via a Pfannenstiel incision. First, the left gonad was isolated and excised followed by the right gonad (figures 4 and 5). Approximately 12 cm of sigmoid colon was isolated on its vascular pedicle (figure 6). Continuity of colon was restored by single layer end to end anastomosis using 3-0 polyglactin (vicryl) sutures. The proximal end of the isolated sigmoid segment was closed in 2 layers and fixed to the sacral promontory. An H-shaped incision was made for

reconstruction at the proposed vaginal site. The distal end of the sigmoid segment was brought down to the perineum through a tunnel between the bladder anteriorly and rectum posteriorly. The edges of the vaginal incision were sutured to the distal end of the sigmoid segment (figure 7). A Vaseline soaked vaginal mould was inserted in the neovagina for 48 h (figure 8). A Foley catheter was kept in situ for 4 days. Postoperatively, the patient was taught neovagina self-care for cleaning of mucus by saline irrigation on a daily basis.

OUTCOME AND FOLLOW-UP

The patient has been on regular follow-up for the past 4 months. She has a cosmetically acceptable vagina, and is sexually active and doing well. She has not reported any complications to date.

DISCUSSION

Many methods of vaginal reconstruction have been reported in the literature. A non-operative technique, also known as a Frank procedure, can be used if the vaginal pouch is present. The Frank procedure involves progressive mechanical dilation using graduated hard dilators to create progressive invagination of the vaginal pouch.¹ This procedure may not have good results in patients with a vaginal dimple.²

Use of a bowel segment for vaginoplasty was first reported in 1904 by Baldwin.³ Goligher⁴ reported positively about his experience with a pedicled portion of the sigmoid colon for construction of a neovagina in the early 1980s. Hensle and Reiley reported their experience in 20 patients with MRKH syndrome and stated that the sigmoid colon is better suited for vaginal replacement because of its long mesentery. According to the authors, small bowel segments have thicker mesenteries, making it difficult for them to reach the perineum without tension² (table 1).

Rajimwale *et al*¹⁵ stated the following advantages of sigmoid vaginoplasty: (1) it is self-lubricating; (2) mucus production is less of a problem as compared to using small bowel; (3) it can grow with the child when used to create a neovagina before puberty; (4) there is minimal risk of stenosis; (5) it is close to the perineum; (6) the vascular pedicle can be easily mobilised; and (7) it does not require moulds or stenting. Sigmoid vaginoplasty thus provides a cosmetically acceptable neovagina with a good length and natural lubrication, obviating the need for stenting and/or dilation.

Table 1 Summarises the studies reported on sigmoid vaginoplasty for various indications

Series	Total patients	MRKH	CAIS	Other DSD/Pelvic surgery	Follow-up period total/mean (years)	Sigmoid segment used	Complications
Kisku <i>et al</i> ⁵ (2004–2014)	55	20	00	35	10	50	09
Nowier <i>et al</i> ⁶ (2003–2009)	31	16	12	03	06	31	05
Karateke <i>et al</i> ⁷ (2003–2009)	29	29	00	00	07	27	03
Rawat <i>et al</i> ⁸ (2002–2008)	08	08	00	00	07	08	01
Freitas Filho <i>et al</i> ⁹ (1997–2000)	10	00	07	03	03	10	01
Dunlop <i>et al</i> ¹⁰ (1990–2005)	26	26	00	00	11	23	07
Kapoor <i>et al</i> ¹¹ (1990–2004)	14	14	00	00	09	14	00
Ekinci <i>et al</i> ¹² (1990–2003)	06	03	02	01	13	06	01
Hensle <i>et al</i> ¹³ (1980–2004)	57	42	00	15	23	39	Not applicable
Hensley and Reiley ² (1980–1996)	31	20	05	06	16	20	08
Hanna <i>et al</i> ¹⁴ (1980–1996)	20	06	01	13	16	06	02

CAIS, complete androgen insensitivity syndrome; DSD, disorder of sexual differentiation; MRKH, Mayer-Rokitansky-Kuster-Hauser syndrome.

Intestinal complications are rare with sigmoid vaginoplasty; however, preoperative bowel preparation, expertise in bowel surgery and meticulous suturing are essential to avoid such complications. In our department, we regularly use different bowel segments for neobladder formation in patients diagnosed with muscle invasive bladder cancer and for other procedures. We are well versed with bowel surgery.¹⁶

Mucosal or entire neovaginal prolapse has been reported, with an incidence of 0–14%.¹³ In our patient, we fixed the proximal segment to sacral promontory to avoid this complication. Variable incidence of introital stenosis has been reported with sigmoid vaginoplasty, ranging from 8.1% to 19.3%.^{2 17}

The need for regular vaginal dilation after sigmoid vaginoplasty is debatable. We do not advise our patients to continue with routine vaginal dilation if they are sexually active. Even though excessive mucus production has been reported in the literature, we have noticed that the amount of discharge seems to decrease with time and is not a major problem for most of our patients.

Learning points

- Sigmoid neovagina is an excellent method of vagina reconstruction.
- The sigmoid neovagina is capacious, does not require prolonged dilation, can be easily mobilised on its vascular pedicle, is not associated with any major complications and has adequate natural secretion allowing lubrication for sexual intercourse.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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