Congenital absence of the vagina — results of conservative treatment

Richard E. Lappöhn

Department of Obstetrics and Gynecology, Academisch Ziekenhuis Groningen, Oostersingel 59, 9713 EZ Groningen, The Netherlands

Accepted 14 December 1994

Abstract

Objective: To assess the efficacy of a combination of Frank’s mold therapy with intercourse as a treatment for congenital vaginal aplasia. Study design: From 1973–1993, thirty-three patients with congenital aplasia of vagina and uterus were seen by one gynecologist. Patients with a partner were instructed how to create a functional vagina by a modified form of coitus (‘interfemoral intercourse’). Mold therapy (using modified molds) was used as a primary treatment when patients had no partner (n = 9) or added to the treatment of nine patients in whom interfemoral intercourse alone had failed. In case of failure of conservative treatment, the surgical creation of a neovagina (McIndoe procedure) was offered. Results: Conservative therapy in patients with a stable sexual relationship (n = 24) gave significantly better (p = 0.0104) results than when patients were single. In the former, a vaginal depth of > 8 cm was reached within six months at most with intercourse alone and four months if combined with mold therapy. With molds alone, 3 out of 9 obtained a satisfactory vagina. Treatment failures occurred (with one exception) only in patients who had no or unstable sexual relationships. Conclusion: Intercourse as a treatment of vaginal atresia can be very successful, especially if practised together with modified mold therapy. A stable sexual relationship — implying a healthy self-concept — appears to be an important determinant of treatment outcome.

Keywords: Congenital absence of vagina and uterus; Mold therapy; Intercourse

1. Introduction

Congenital aplasia of vagina and uterus, the Mayer-Rokitansky-Küster syndrome, is said to occur in 1 in 5000 live-born female infants and explains up to 9% of the cases of primary amenorrhea [1,2]. Girls with this form of primary amenorrhea have normal ovaries and go through puberty at a normal age. In some, the absence of menarche causes no concern and medical advice is only sought when problems with sexual intercourse are first encountered. Even then, consultation may be delayed: patients have been described in whom satisfactory vaginal intercourse had evolved through practice [1,3]. This is not surprising as congenital absence of the vagina usually leaves pliable perineal skin between the urethra and anus. Frank treated six patients by means of persistent pressure with pyrex glass molds on this spot, thus obtaining a functional vagina without resorting to surgery [4]. Modifications of dilators using a specially designed stool can often be of help [5]. However, this conservative treatment does take a commitment from both the patient and the doctor. Failing this, one of the many types of surgical procedures is usually offered to the patient.

This paper presents the results of 20 years’ experience with a treatment procedure in which a modified Frank’s procedure is used as a complement to intercourse. They demonstrate that surgery can be avoided in many patients with congenital absence of the vagina.

2. Patients

From 1973 to 1993, 33 patients with congenital absence of the vagina were seen. Two young girls, aged 9 and 14 years, had abdominal symptoms due, respectively, to an ovarian dysgerminoma (with 46,XX chromosomal pattern) and severe varicosities of the ovarian
veins with ectopic ovaries. Twenty-seven patients were between 16 and 25 years old, four were older. Fifteen complained of primary amenorrhea, 11 had been unable to achieve vaginal intercourse. Three had abdominal complaints (not due to concealed shedding of endometrium), one woman had incurred a vesico-vaginal fistula due to forceful insertion of a Hegar dilator. The oldest patient was seen in consultation while hospitalized in the end-stage of renal insufficiency.

Twelve patients had associated renal anomalies, nine in the form of unilateral agenesis. From these nine, three were disfigured by the Klippel-Feil anomaly, a congenital malformation of the cervical vertebrae resulting in a very short neck. Three suffered from congenital deafness. One patient was severely handicapped by the absence of both legs and her right arm.

3. Methods

If a diagnosis had not been made elsewhere, patients were investigated with standard gynecological procedures. Kidneys and ureters were visualized by i.v. urography or ultrasound. X-rays of the neck region were made for documentation. Exams for hearing loss were done if necessary.

The patients were fully informed about their condition. It was carefully stipulated that treatment, possibly including surgery, might lead to a happy sexual relationship but not to the possibility of becoming pregnant. Those who had not attempted to engage in sexual intercourse and were confident enough were advised to contact us later, together with their partner. Mold therapy was offered as a primary treatment to the women who did not want to delay the creation of a vagina because they felt incomplete as a woman or because they were afraid to start a sexual relationship.

Interfemoral intercourse as a means to create a vagina was discussed with women with a partner. Thereby, the friction to the moving penis is provided by the adducted upper legs and the penile thrust impresses the female’s perineum. When earlier vaginal intercourse had failed, the couple was instructed in the same technique. The patients were seen after one month and two months thereafter. When the vaginal depth did not increase, supportive treatment with molds was offered. The molds were specially made from plexiglass and fitted to a plexiglass plate formed to rest on the labia majora without touching the urethral ostium or the clitoris. Depending on the depth and pliability of the initial vaginal pouch, the first molds had a minimal diameter of 1.5 cm; the shortest molds were 2.5 cm. In order to avoid dilatation of the urethra, the angle of the axis of the initial molds with the mounting plate was 135°. Thicker and longer (≥ 5 cm) molds were mounted at an angle of 90° to the mounting plate. An idea of the form and dimensions of a mold is given in Fig. 1.

The molds were to be worn daily for at least 30 min. Patients were instructed to wear tightly fitting briefs to keep the mold in place with the necessary (slight) pressure. The result was checked once a month and a larger mold was given as soon as possible.

Patients who did not want a trial of conservative treatment or in whom conservative treatment had failed were offered surgery: vaginoplasty according to McIndoe was performed by, or in co-operation with, a plastic surgeon. It is important to realize that post-operative mold therapy is essential to this procedure [6].

4. Results

An overview of the results is given in Table 1.

4.1. Interfemoral intercourse, without and with molds.

Five women had started intercourse without prior knowledge of, or disregarding, their condition. At intake, a vaginal pouch of normal depth (> 10 cm), easily admitting two fingers, was seen in two patients. One couple had no problems with vaginal intercourse despite a shallow (about 5 cm deep) vagina; the patient had abdominal complaints caused by ovarian cysts in ectopic ovaries.

In two, the vagina was < 3 cm deep; the urethral ostium was exceptionally wide and displaced dorsally. One of these women had stopped sexual intercourse because of intolerable dyspareunia, the other also complained of painful intercourse. She appeared to have intercourse in the urethra; this was probably also the case in the first patient. These two patients received instructions for interfemoral intercourse together with
Table 1
Overview of the results of the various stages in the treatment of congenital absence of the vagina (n = 33)

<table>
<thead>
<tr>
<th>Treatment type</th>
<th>N</th>
<th>Initial vaginal depth (cm) (range)</th>
<th>Duration of treatment (Months)</th>
<th>Resulting depth (n)</th>
<th>Transfer to treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. 'Naive' start with intercourse</td>
<td>5</td>
<td>?</td>
<td>?</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>2. Coitus I.F.</td>
<td>11</td>
<td>0.5-2</td>
<td>4-6</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>3. Coitus I.F. + molds</td>
<td>9</td>
<td>1-5</td>
<td>2-4</td>
<td>9</td>
<td>0</td>
</tr>
<tr>
<td>4. Molds alone</td>
<td>9</td>
<td>0.5-2</td>
<td>1-11</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>5. Melndoe</td>
<td>3</td>
<td>2-4</td>
<td>-</td>
<td>1</td>
<td>?</td>
</tr>
<tr>
<td>6. Melndoe delayed</td>
<td>2</td>
<td>0.5; 3</td>
<td>-</td>
<td>-</td>
<td>2 1</td>
</tr>
</tbody>
</table>

33 + 6

mold therapy, and a vaginal depth of 8 and 10 cm was obtained after 3 and 4 months, respectively.

Eleven women and their partners started with interfemoral intercourse after instruction. Within six months at most, a satisfactory vagina developed in nine women. One woman had problems with her partner and stopped treatment. In another, coitus took place in the urethra. With simultaneous mold treatment, a vagina of 8 cm was created within 4 months. Six women practised interfemoral intercourse together with mold application from the beginning of treatment. Within four months, satisfactory vaginal intercourse was possible in all of them.

4.2. Mold therapy alone
Nine patients without a partner, including two with Klippel-Feil, started therapy with molds alone. A good result was obtained in three patients. The two patients with Klippel-Feil and a third with an exceptionally sturdy perineum obtained less favourable results; when they gave up treatment (after 4, 6 and 11 months), vaginal depths of 5, 6 and 6 cm had been reached. They chose not to have surgery. We were unable to provide the patient with amelia with a fitting mold, after attempts at a modified form of intercourse had failed. Two patients were unable to cooperate and rejected mold therapy soon after they had started.

Patients with a stable sexual relationship enabling intercourse with or without the simultaneous use of molds had better results with conservative treatment (20 out of 24) than those who started with molds when single (3 out of 9); p = 0.0104 (Fisher's exact probability). The better result was reached in the remarkably short period of 6 months, at most.

4.3. Surgery
Five women (three of whom with a stable relation) were candidates for surgery. Repeated surgery was the only option for the patient with renal failure because a neovagina, created surgically when she was 18, had shrunk to a depth of 3 cm. She preferred to postpone treatment until after renal transplantation, which has not been performed yet. The girl with amelia also awaits surgery. The patient with a vesico 'vaginal' fistula underwent surgical repair together with a Melndoe operation. The two women who rejected mold therapy were operated upon in 1982 and 1991, respectively. The initial results were good.

4.4. Follow-up
Twenty-eight patients who had been treated before January 1, 1993 were contacted by students doing a special interest training course. Twenty-five responded; three of these had had surgery. Twenty of the conservatively treated patients were sexually active; 14 were completely happy with the situation, four reported minor problems such as frequent absence of orgasm or feeling tense at intercourse. Two had divorced and had not found a new partner. From the women who underwent surgery, the patient with a fistula reported satisfactory vaginal intercourse but some dyspareunia. Two had serious difficulties: one was severely vaginistic and the other had never attempted intercourse.

5. Discussion
Even though the simple conservative method has the advantage that the patient has created her vagina by herself, it is far less popular than surgical kallmannopoeisis. In most reports on surgery for congenital absence of the vagina, the authors state that earlier conservative treatment had not been successful; often a fibrotic vaginal top or a sturdy perineum are mentioned as the reason for failure. This was found only once in our group of patients. An important aspect of the surgical approach is the fact that patients, at least initially, can deny emotional handicaps in their body perception and (sexual) relationships. Possibly, many failures of conservative treatment are due to such emotional inhibitions.

Twenty-three out of our 30 treated patients (77%) had
excellent results, anatomically as well as in terms of satisfaction, with conservative treatment. It is safe to state that a failure rate of about 5% has to be expected after surgery, while 8–15% of the patients report major problems with sexual intercourse [7], resulting in a success rate of 80–87%. Our outcome is therefore comparable to the results of surgery but may have been influenced by selective referral.

Because of the significant difference between patients with and without a stable sexual relationship, it is reasonable to assume that psychological factors such as self-esteem and ability to establish stable relationships are among the main determinants of treatment outcome. Patients with a relatively undisturbed self-concept are the best candidates for conservative therapy. In others, it may be worthwhile to try to resolve psychological distress before the specific treatment. If successful, surgery might prove to be unnecessary in these patients as well.

Acknowledgements

The co-operation of Dr K.W. Marck, presently plastic surgeon in Leeuwarden, is gratefully acknowledged. The author thanks Dr W. Weijmar Schulz for critical comment.

References