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# Rectovestibular fistula—rarely recognized associated gynecologic anomalies

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**Key words:**
Vestibular fistula;
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Vaginal anomaly;
Vaginoscopy

**Abstract**

**Introduction:** Vestibular fistulas are the most common anorectal malformations (ARMs) in females. Associated gynecologic defects are rarely mentioned in the literature but may have serious clinical implications if undetected. The definitive repair of the ARM offers an opportunity for diagnosis and treatment of these conditions.

**Methods:** Two hundred seventy-two patients with vestibular fistula were retrospectively reviewed, with emphasis on gynecologic defects.

**Results:** Forty-eight patients (17%) had 83 gynecologic anomalies. Fourteen patients had a vaginal septum, all with 2 uterine cervices. All septa were resected at the main repair. Twenty-six patients had no vaginal opening. Twenty of them had absent vagina. Eighteen of those had an absent uterus. Patients with absent vagina underwent vaginal replacement with distal rectum (12), sigmoid (6), and ileum (2). Six patients had a patent upper vagina; 3 reached the perineum after mobilization and 3 required replacement; 2 with sigmoid and 1 with rectum.

**Conclusion:** Vaginal septa are easily diagnosed and can be resected during the repair of the vestibular fistula. The presence of 2 cervices has important obstetric implications. Absent vagina requires a technically demanding repair, with special preoperative planning. Vaginoscopy or careful inspection should precede surgical reconstruction.



Rectovestibular fistula is the most common anorectal malformation seen in females. When treated appropriately, these patients have an excellent functional prognosis for bowel and urinary control [1]. In a review of the literature, we found that associated gynecologic defects are rarely mentioned [2-11].

In this series, we identified a significant number of gynecologic anomalies associated with this anorectal malformation. Early recognition is very important to offer these patients timely and optimal treatment, as well as adequate follow-up into young adulthood. The definitive repair of the anorectal malformation offers an ideal opportunity for adequate diagnosis and management of most of these gynecologic anomalies. A delayed or

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**Table 1** Summary of gynecologic abnormalities found in 272 patients born with vestibular fistula

| No. of patients | Type of gynecologic anomaly | | Associated gynecologic anomaly |
|---|---|---|---|
| 26 | No vaginal opening | Total vaginal atresia = 20 | 18 Absent uterus<br>1 Absent cervix<br>1 Hemiuterus |
| | | Partial vaginal atresia = 6 | 5 Normal uterus<br>1 Bicornuate uterus |
| 14 | Vaginal septum | | 14 Hemiuterus |
| 3 | Uterine anomalies only (2 bicornuate uterus, 1 hemiuteri) | | |
| 3 | Vulva anomaly only (2 lipomas, 1 hemangioma) | | |
| 1 | Ectopic vaginal opening | | Hemiuterus |
| 1 | Clitoromegaly only | | |
| Total = 48 patients | Total = 83 gynecologic anomalies [a] | | |

[a] Many patients had more than one anomaly.

missed diagnosis may result in adverse late consequences for the patient.

The purpose of this report is to increase the index of suspicion of pediatric surgeons in relation to these important associated gynecologic defects.

## 1. Materials and methods

A retrospective review was performed of the medical records of 272 patients with rectovestibular fistula, operated on by us during the last 28 years. Special emphasis was placed on evaluating for malformations of the external and internal genitalia, as well as the treatment used to repair them, and the results obtained.

At the time of the main repair of the anorectal malformation, all patients had a vaginoscopy, which allowed detection of the gynecologic abnormalities of the external genitalia. In those patients who had a colostomy performed before the main repair, the anatomy of the internal genitalia was evaluated at the time of the colostomy closure.

We obtained institutional review board approval (no. 2008-1011) for this study.

## 2. Results

Of the 272 patients reviewed, 48 (17%) had a total of 83 gynecologic anomalies (Table 1). Fourteen patients had a vaginal septum, with 2 cervices; each one of them communicated with a hemiuterus. All vaginal septa were resected during the main repair of the anorectal malformation. The septum resection was a simple, straightforward procedure, performed with a needle cautery (Fig. 1), and there were no postoperative complications.

In 26 patients, a vaginal opening was not identified (Fig. 2). Further intervention, including a laparotomy or laparoscopy, demonstrated that 20 of these patients had a completely absent vagina; 18 of them also had an absent

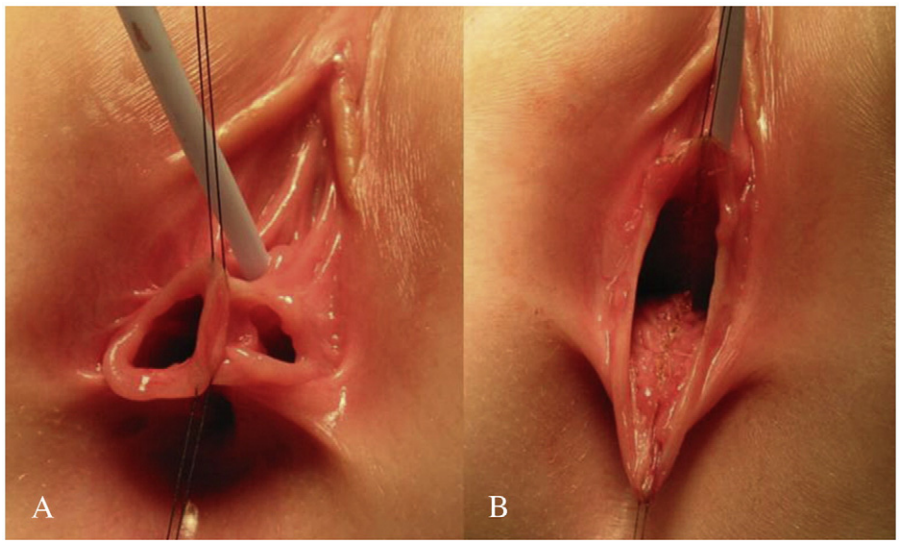


**Fig. 1** A, Vaginal septum. B, Final appearance after resection.

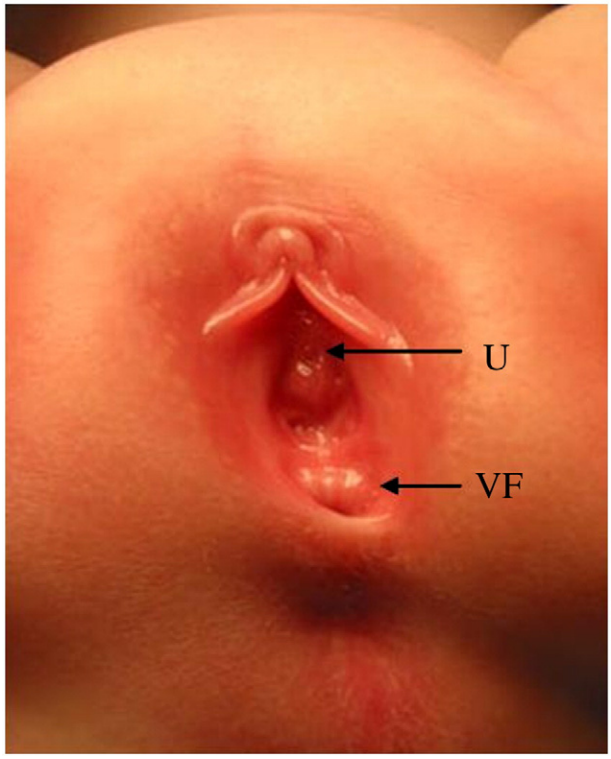


**Fig. 2** Absent vaginal orifice. U indicates urethra; VF, vestibular fistula.

uterus (Fig. 3); one had a well developed uterus with an absent cervix (Fig. 4) and another only had one hemiuterus. In 6 patients, a patent upper vagina was identified; in 3 of them, the patent upper vagina was long enough to allow its mobilization down to the perineum (Fig. 5A). The other 3 patients had a very short but patent upper vagina (Fig. 5B) and required a partial vaginal replacement, with sigmoid (2 cases) and with distal rectum (1 case). Of the 6 patients with a patent upper vagina, 5 had a normal uterus, and the remaining case had a bicornuate uterus (Fig. 6).

All 20 cases with a totally absent vagina underwent a vaginal replacement. The distal rectum was used in 12 of them (Fig. 7), sigmoid colon in 6 (Fig. 8), and terminal ileum in 2 (Fig. 9). One patient had an ectopic vaginal opening next

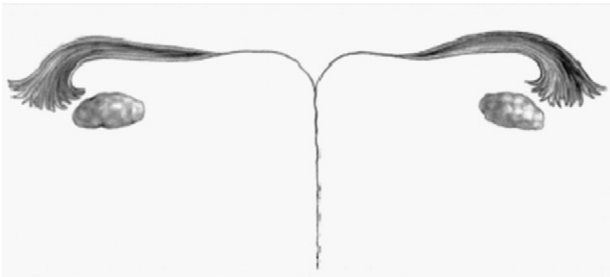

**Fig. 3** Internal anatomy of 18 patients with absent vagina, absent uterus, normal ovaries, and fallopian tubes.

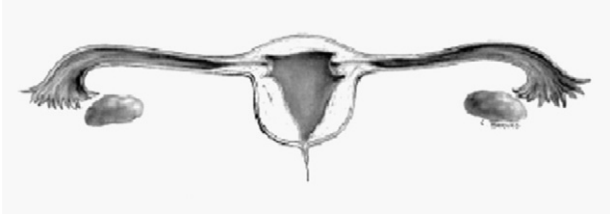

**Fig. 4** Internal anatomy of one patient with absent vagina, absent uterine cervix, normal ovaries, and fallopian tubes.

to the rectum, both structures were located posterior to the urethra, and the vagina was connected to a hemiuterus. In 3 patients with a normal vagina, a uterine anomaly was found, including 2 bicornuate uteri and 1 case with a hemiuterus. Three cases had an abnormality of the vulva: 2 lipomas, 1 hemangioma. One patient had a hypertrophic clitoris with normal endocrinologic workup.

Concerning the function of the rectum left as a neovagina, one of our patients complained that the piece of rectum was too small to have satisfactory intercourse and required an enlargement with an extra piece of colon. A second patient reports sexual activity with no problems. The remaining 10 patients have not yet reported sexual activity. In the group of patients that had an anastamosis between the neovagina and the internal genitalia, one patient is menstruating normally through the neovagina (rectum).

## 3. Discussion

Associated gynecologic anomalies in patients with rectovestibular fistulae are more common than previously thought. Vaginal septum occurred in 5% of our cases, thereby justifying the search for this defect, at the time of the main repair of the anorectal malformation. This can be accomplished by direct examination or preferably by vaginoscopy using the pediatric cystoscope because it allows for visualization and inspection of the cervices. The resection of the septum takes a few minutes, and it is technically easy

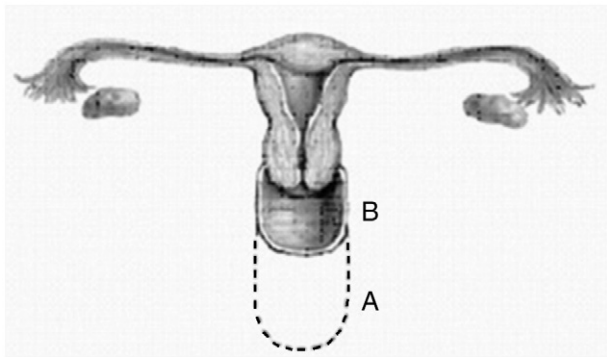


**Fig. 5** Internal anatomy of 6 patients with patent upper vaginas. A, Long patent upper vagina. B, Short patent upper vagina.

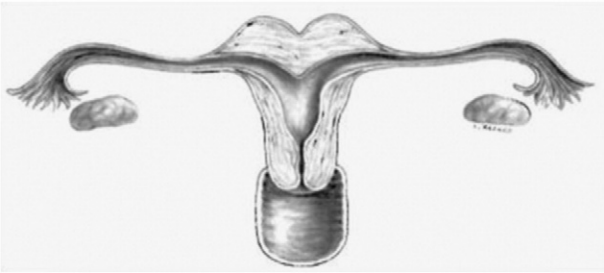

**Fig. 6** Internal anatomy of one of the patients with patent upper vagina and a bifid uterus.

(Fig. 1). However, in very young patients, the surgeon should be very careful not to damage the cervices while resecting its upper most portion because the cervices are located very close to the midline.

The largest reported series related to longitudinal vaginal septum included 202 adult patients [12]. Of the patients, 56.4% were asymptomatic. In the symptomatic group of patients (43.6%), dyspareunia was the major complaint, followed by problems with tampon use and significant difficulty or bleeding during intercourse. Another article reported intermittent mucopurulent discharge, which was relieved after the septum was resected [13]. Because of those findings and the possibility of laceration during a vaginal delivery, these authors recommend prophylactic surgical treatment of the vaginal septum [12]. Although these reports were not related to patients with anorectal malformations, we consider them relevant to the current study. Their findings support our recommendation for removal of the septum at the time of the main repair of the anorectal malformation. However, if the septum was left untouched, the removal may be performed electively in combination with other surgical procedures or deferred until after puberty, if asymptomatic.

The most important aspect of the vaginal septum is the fact that, in our series, it was always associated with the presence of 2 hemiuteri (uterus didelphys). Similar findings have been reported by others [12,13], although, again, not in cases with anorectal malformation. Early identification of such uterine anomalies allows important counseling in reproductive and obstetric prognosis. Interestingly, we found, in 2 previous publications, important information concerning the long-term gynecologic and obstetric implications of a didelphus uterus. Eighteen percent of these patients had an obstructed hemivagina with hematocolpos [13]. Between 21% [13] and 32.9% [14] of these cases had a miscarriage. In addition, between 24% [13] and 28.9% [14] had a preterm delivery, with associated fetal growth retardation.

These data strongly support our concept of the importance of an early diagnosis, as well as the need for long-term follow-up, and for specialized gynecologic and obstetric care for these patients.

The condition of absent vagina in association with rectovestibular fistula deserves special consideration. We believe that the frequency of this association has been underestimated. Excluding our previous publication [5], we found 10 references [3,6-9,11,15-18] that in total reported only 22 cases (Table 2), during the last 52 years. An interesting couple of articles [19,20], written by the same author, with the same data, reported on 13 vaginal septum, 5 vaginal agenesis, and 1 absent lower third of the vagina out of 162 cases of anorectal malformations. Unfortunately, the authors did not describe the specific type of anorectal malformation related to the gynecologic defect. In other words, included in those 162 cases were cloacas and other noncharacterized defects called “high rectovaginal fistulae” and “high rectovestibular fistula.” We believe that cloacas deserve a special, separate analysis because most of them have an associated a form of gynecologic anomaly [21-23].

The treatment modalities used in the 22 cases of vestibular fistula with absent vagina, reported in the literature [3,6-9,11,15-18], included 8 vaginal replacements with rectum, 3 replacements with sigmoid, 3 successful vaginal mobilization in cases of distal vaginal atresia, 2 replacements with small bowel, and 1 with skin flaps. No information was found related to the vaginal reconstruction in the remaining 5 cases.

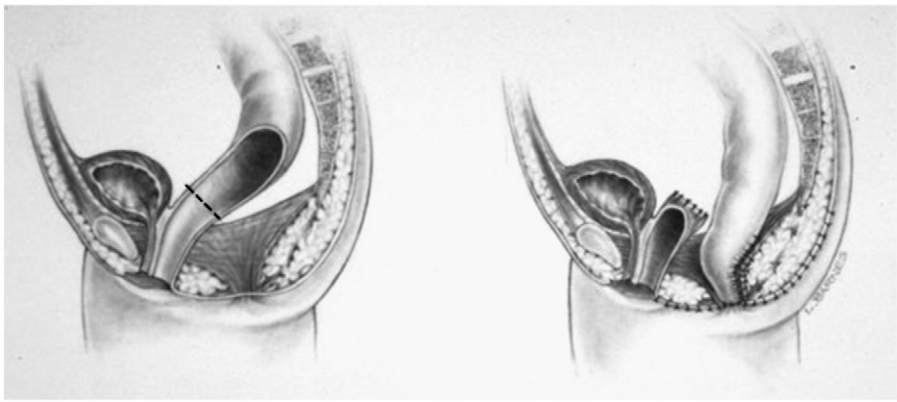

**Fig. 7** Vaginal replacement with distal rectum. Reprinted from [5] with permission from Elsevier.

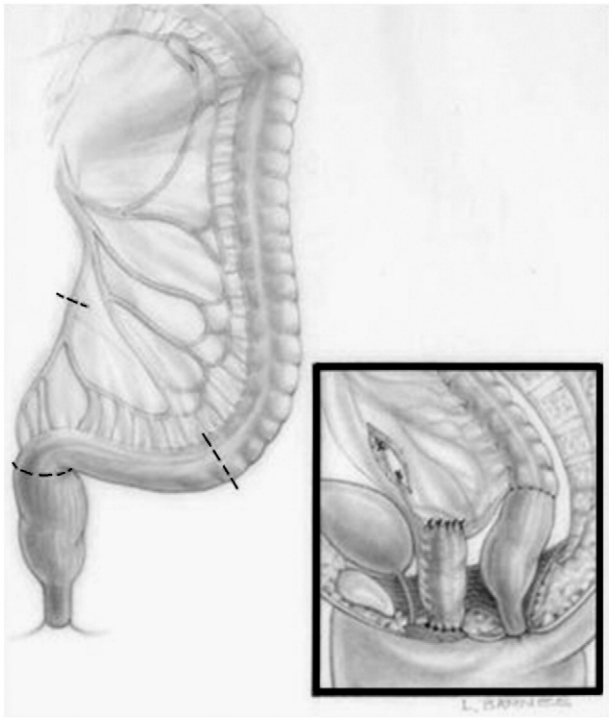

**Fig. 8** Vaginal replacement with sigmoid colon. Reprinted from [23] with permission from Elsevier.

In our series, there were 4 modalities of treatment (Table 3) that included (*a*) rectum left as a neovagina (13 patients), (*b*) sigmoid replacement (8 patients), (*c*) mobilization of the native upper vagina (3 patients), and (*d*) replacement with terminal ileum (2 patients). In 18 patients, the neovagina was created only for sexual purposes because those patients did not have patent well-developed internal genitalia (uterus). The upper part of the neovagina was left blind. In 5 cases, the neovagina was anastomosed to patent internal genitalia. Three of them had a small patent upper vagina and uterus with a normal cervix (Fig. 5). In one case, the neovagina was anastomosed to a bicornuate uterus, and in the last one, the anastamosis was created between the neovagina and the uterus, lacking the cervix.

In the most common treatment modality of our series (rectum left as neovagina), a more proximal portion of rectum was pulled down and placed within the limits of the sphincter mechanism as a new rectum (Fig. 7). This procedure was performed via a posterior sagittal approach. The operation proved to be reproducible, and the patients recovered uneventfully. However, the patients experience a tendency to have very frequent bowel movements, which made the toilet training process more difficult [22]. We felt this was because of having removed their natural reservoir (rectum) from their fecal stream to be used as a neovagina.

Eight patients underwent vaginal replacement with sigmoid and 2 with ileum. We decided on that treatment modality in these later cases to preserve the rectum as a rectum, trying to disturb bowel control as little as possible, hoping that the toilet training process would be easier. Our first choice for vaginal replacement is colon because of its thick wall and the anatomy of its blood supply, which allows for a straight forward pull-through. In 2 cases we used small bowel because the colon was not available because of the presence and location of the colostomy. Our patients are still too young to be evaluated for bowel and sexual function.

One of our patients presented with cervical agenesis, which is a very rare anomaly occurring in 1:80,000 to 100,000 cases [9]. In the past, hysterectomy was the treatment of choice. Currently, the recommended options are suppression of menses with preservation of the uterus for pregnancy with reproductive assistance, or utero vaginal anastamosis [9]. A report of 18 cases of uterovaginal anastamosis in patients with cervical atresia reported 6 spontaneous pregnancies in 4 patients, and only one required cervical cerclage [24]. Because we operate on our patients at a very young age, we believe it is worth giving them the possibility of a future pregnancy. It is still too early to comment on the follow-up of our one case.

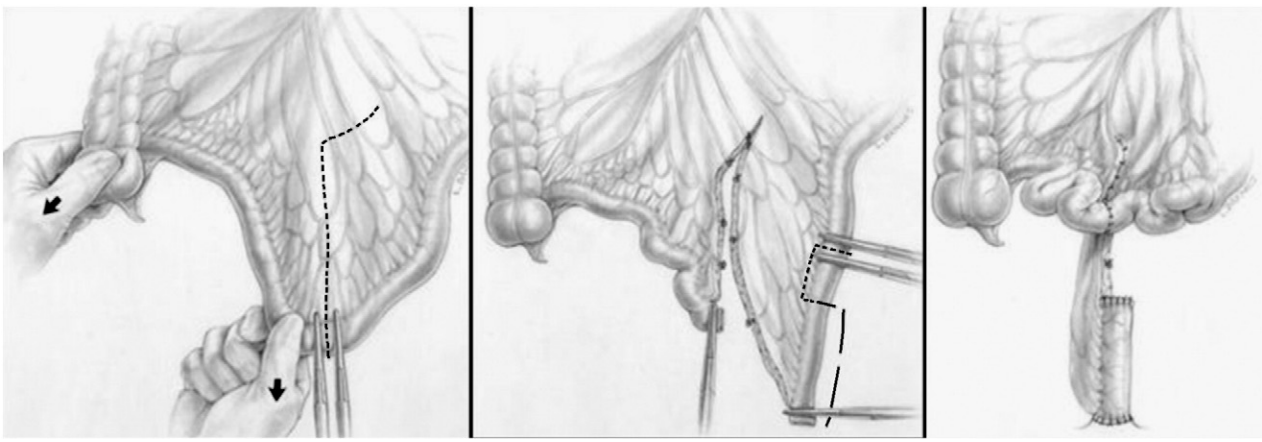

**Fig. 9** Vaginal reconstruction with terminal ileum. Reprinted from [23] with permission from Elsevier.

**Table 2** Summary of vestibular fistula and absent vagina cases published in the literature [3,6-9,11,15-18]

| Vestibular fistula with | Type of vaginal reconstruction | Follow-up | |
|---|---|---|---|
| | | Bowel function | Sexual function |
| Totally absent vagina = 18 | Distal rectum = 7 | One patient incontinent | |
| | Colon = 3 | | |
| | Small bowel = 2 | | |
| | Skin flaps = 1 | | Active = 1 |
| | No information = 5 | | |
| Distal (partial) absent vagina = 4 | Mobilization of upper vagina = 3 | One patient continent | Active, got pregnant and delivered through C section = 1 |
| | Partial replacement with distal rectum = 1 | | |

**Table 3** Type of vaginal reconstruction in our series

| | | |
|---|---|---|
| 26 Patients with vestibular fistula | 20 With total absent vagina | 12 Distal rectum |
| | | 6 Sigmoid |
| | | 2 Terminal ileum |
| | 6 With distal (partial) absent vagina | 3 Mobilized upper native vagina |
| | | 2 Sigmoid |
| | | 1 Distal rectum |

We believe that an absent vagina should not be an unexpected surprise during the repair of a vestibular fistula. As shown in this series, the repair of the absent vagina frequently requires the use of colon or small bowel for the replacement and is ideally performed at the time of the rectal pull-through.

## 4. Conclusion

Gynecologic anomalies associated with rectovestibular fistulae are more common than previously thought. Our results suggest a high index of suspicion is appropriate in girls with this type of anorectal malformation. The definite repair of the anorectal malformation represents the best opportunity for the evaluation and treatment of most of these defects. Early diagnosis allows for adequate planning and preparation in cases of absent vagina. In addition, potential sexual and obstetric implications mandate specialized care and long-term follow-up.

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## Discussion

*Dr. Mary Brandt (Houston, Tex):* I had a question because for vaginal replacement there is also the skin lined vagina and I am not aware of a series this long that can look at the mucus secretion and whether or not that is a problem for the girls. Has that been something that you have addressed or noticed or have any comments on?

*Alberto Pena (response):* Yes, we don't have a perfect technique for vaginal replacement. With colon the patients complain, not all of them, but some of them, of leakage of mucus. Some patients are happy because they only use a little pad. Mucus usually means irritation and sometimes it is irritated because if you leave a very long portion of colon and very, very twisted, and then it accumulates mucus and produces colitis. Sometimes we can teach them how to irrigate the mucus with a little soft catheter twice a week and that takes care of the problem.

*Dr Steven Fishman (Boston, Mass):* I am curious as to the age at which you performed the vaginal replacement. When you used the native distal fistula you obviously noticed that at birth and did it then. And some of these, I suspect, were referral cases. What age do you recommend, if this is recognized early, that the vaginal reconstruction be performed?

*Dr Pena (response):* Yes, all the patients that I presented here had the reconstruction as soon as we detected the problem because when we repair the vestibular fistula we thought there was a great opportunity to do everything. There is a little controversy about the patient that comes to you that had a repair of the vestibular fistula, the surgeon was not aware of the vaginal septum or the absent vagina, is 5 years old, and the question is should we replace the vagina, yes or no? We don't have a straightforward answer for that. I can give you a list of arguments in favor or against doing that. In general, we prefer to do it earlier because the operation technically is more difficult in older patients. And you can argue which is more traumatic, the operation or the notion of not having a vagina. We don't have time for that but I would be happy to comment with you later.

*Dr Fishman:* Thank you.

*Dr Andrea Hayes-Jordan (response):* Thank you Dr Pena. That was a wonderful presentation. I have a question about the children with the vaginal septum and the hemiuterus. Do you have any data to look at whether taking out one uterus affects their miscarriages and ability to bear children or is there any difference?

*Dr Pena:* We don't have that data. The information about miscarriages and about premature labor comes from the adult literature and from fertility clinics when they study ladies that are not fertile and then they find that they have a hemiuterus, or incidental findings with the delivery by cesarean section. But we are not aware that resecting one hemiuterus will affect the function of the other. We have a long series of hemiuterus cases, but that is another presentation. But our patients are getting into that age. Some of them are sexually active and if you invite me again in 5 years I will be able to tell you a lot of interesting things about these patients.

*Ala Frey, MD (Response):* The question I have is for the patients in our practice that we have operated on with rectovestibular fistula that did not have a septum. Would you recommend because there is a higher incidence of gynecologic anomalies in these patients, to screen them with ultrasound and/or MRI to look for some of the things that you can't see just by examining and/or inspecting the perineum?

*Dr Pena:* If they don't have a septum, the possibility that these patients have other internal anomalies is extremely low. Because those patients have a colostomy, at the time of colostomy closure we always take the opportunity to look at the internal genitalia and we have not seen, in patients with no vaginal septum, anomalies inside.

*Dr Al Chahine (response):* Dr Pena, when you divide the septum do you put them on a regimen of vaginal dilatations as well as the anal dilatations?

*Dr Pena:* No. We divide the septum with the cautery and sometimes the septum is extremely thin. You don't have to do anything about that. We have never seen a synechia after that, but if the septum is very thick then you find a gap between the mucosa and the mucosa we put 6 or 5 sutures to bring together the mucosa to avoid the possibilities of a synechia. Be careful when you resect the septum when you get high because then sometimes the cervices are very close and you don't want to resect 100 percent. Perfection is enemy of good because you may hurt the cervices.