CONGENITAL VAGINAL MALFORMATION: CLINICAL EXPERIENCES ON VAGINAL AGENESIA

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Summary: After having described, with brief allusions to the causes which determine vaginal malformations and the different surgical techniques which are generally used for treatment of such forms, the case series is examined of 53 patients who came under our observation in the Gynaecologic and Obstetric Clinics of Padua between 1965 and 1987, and of Verona, limited to the period from 1969 to 1974.

We consider that the choice of the most suitable treatment for such pathology cannot be separated from the analyses of such diverse parameters as, for example: the age of the patient at the time of diagnosis, the type of malformation present, and other eventual associated malformations.

This induces us to support the personalisation of the treatment of such pathology in each individual case.

The problem of genital malformations is very ancient in the history of human pathology: hermaphroditism, for instance, before being studied as a morphological anomaly, was always presented in the form of a myth ([1]).

The congenital absence of the vagina was described first by Avincena (980 - 1026). Later, other observations were reported by Albucasis (1013 - 1106), then by Realdus Columbus (1572), and up to Dupuytren (1817).

It is not easy to check the frequency of genital malformations statistically: first of all because they are malformations which sometimes escape notice at the time of birth, above all when the newborn is examined superficially in order to determine the sex; secondly because later these malformations are often hidden, from understandable motives of a social order, such, for example, as the fear of losing personal consideration, or from modesty and other emotive problems.

From data in literature it appears that the congenital lack of the vagina is met with in a frequency varying from 1 : 4,000 to 1 : 15,000 women born ([2, 3, 4, 5, 6, 7, 8]), even if some case series have reported minor frequencies, up to one case in 83,000 ([9, 10, 11]).

The aim of this study is to make a clinical contribution to such malformative pathology by examining the clinical cases brought to our notice between 1965 and 1987 at the Gynaecologic and Obstetric Clinic of the University of Padua, with particular reference to the type of surgical therapy adopted and the results obtained.

CAUSES OF VAGINAL AGENESIA
(indications)

The causes of this malformation are not dissimilar to those responsable for other types of congenital malformations. Such causes may act either in determining genico-chromosomic damage or an embryofetal dysmorphism.

The principal causes of preconceptional genico-chromosomic damage are to be attributed to:

a) advanced age of the parents;

b) states of intoxication: alcohol, drugs, endogenous intoxication;

c) infectious diseases; rubella, viral hepatitis;

d) ionised corpusculate and electromagnetic radiation;

e) consanguinity of the parents;

f) chemical substances (drugs, toxic exogenous variables, etc.;

g) genetic predisposition;

h) chromosomic interaction;

i) autoimmune diseases.

With regard to embryo-fetal dysmorphisms, it must be remembered that the relationship between the teratogenic agent and the damage which derives from it is not always specific, but often conditioned by some factors among which we recall:

1) the gestational age at which the teratogenic action is verified;

2) the duration of action;

3) the type of teratogenic agent;

4) its dosage.

Therefore, summarising briefly, we can classify the various teratogenic agents:

1) *infective agents*: viruses, schizomycetes and protozoa;

2) *chemical agents*: toxins, endocrine-metabolites, alimentary and respiratory (embryo-fetal hypoxemia);

3) *physical agents*: ionised radiation and traumas;

4) *abnormal conditions of pregnancy*: pathological alterations of the amniotic fluid, umbilical cord or placenta from which may derive "contriction rings", "intra-uterine amputation", "cutaneous furrows", etc.

THERAPY OF VAGINAL MALFORMATIONS

From the beginning of last century, the techniques described for the correction of vaginal malformations have been innumerable. In classifying them, a fundamental distinction is made between the operative and non-operative methods.

A) *Non-operative techniques*

Frank's technique (1938).

B) *Operative techniques*

1) Vecchietti technique (1965);

2) Dupuytren-Warthon technique (1817);

3) Neovagina by transposition of the intestinal loop:

a) Ruge-Novak method (1913);

b) Baldwin method (1907);

c) Mori method (1908);

d) Schubert method (1911);

4) Vulvovagina plastic technique;

5) McIndoe technique (1938);

6) Lining techniques with various withdrawals:

a) amniotic membrane;

b) fetal tissue

c) pelvic peritoneum;

d) pedunculous border of the bladder;

e) pedunculous border of the skin.

OUTLINES OF SURGICAL TECHNIQUES

1) *The technique of Vecchietti* [1, 6, 19, 43]

By this method the neovagina is obtained through the traction exercised by an "olive" of acrylic material which is drawn by two threads inserted (following laparotomy) into the recto-vaginal space and inserted into the "olive" itself. The threads are drawn from the other and by a special drawthread placed on the abdominal wall. The method is simple and yields good long term functional results.

The surgical technique conceived, suggested and nowadays used in many European Centers consists in the following parts:

1) laparotomic incision according to Pfannenstiel;

2) the vesico-uterine peritoneal fold opening, or, in absence of this one, the vesico-rectal peritoneal fold opening;

3) preparation of an intervesico-rectal tunnel;

4) perforation of the pseudo-hymenal septum by an apposite straight needle, conceived by the Author to allow the en-

dopelvic reascent of the two not reabsorble threads which derive from the two poles of the acrylic olive laying on the vulvo-vaginal introitus;

5) extraperitoneal passage of the two threads laterally at the straight abdominal muscles;

6) progressive traction of the threads by an apposite instrument which, at the operation's conclusion, will be positioned on the patient's laparotomic suture.

After a week of progressive traction, a vaginal canal about 8-10 cm long will be formed artificially. The use of vulvo-vaginal prothesis is absolutely recommended for at least 14 days.

2) *Frank's technique* ([1, 6, 13, 14])

This is the only non-operative technique which, re-proposing that of Amussat (1832) is based on the simple pressure exerted on the vestibular mucosa by means of special dilators of increasing caliber, so as to obtain, without any surgical incision, a neovagina wide enough to facilitate coitus. With daily applications of the dilator for 40 minutes at a time, a neovagina of satisfactory dimensions can be obtained in about 6 - 8 weeks ([15]).

A modification of this method ([16, 17]) provides for the application of the dilator on a bicycle saddle, so that during the application a sitting position can be maintained to allow for the continuation of any working activity.

The advantages of this method are: the almost total absence of complications, a percentage of success up to 90% ([6]), a neovagina covered by normal vaginal mucosa and a very short hospitalization. The disadvantages consist in its necessitating a vaginal outline of at least 2 cms depth which will allow for the application of the dilator ([17, 18]) or vaginal intrusor.

3) *Dupuytren-Warthon's technique* ([19])

This technique provides for the simple dissection of the rectovescical space, by applying a dilator, and awaiting for spontaneous epithelisation. The advantage of the technique lies in the limited difficulty of the operation. The disadvantages, on the other hand, are slow cicatrization with risk of infection and stenosis of the neovagina and possible bladder, urethral and recto-vaginal fistulas.

4) *Ruge-Novak's technique* ([20, 21])

With this technique the vesical-rectal space is lined through the transposition of the sigma. The advantages ([15]) lie in the excellent outcome of the operation from a functional point of view. The principal disadvantages are represented by the possible intra and post-operative complications related to the complexity of the operation itself, such as for example, rectal or vescical lesions, dehiscence of intestinal anastomosis, necrosis of the transplanted loop, disturbances in defecation ([10]), and excessive secretion of the mucosa.

Ample case series report 3 deaths out of 175 operations (1.7%) and 1 death out of 125 operations (0.57%), due to grave post-surgical complications.

5) *Baldwin's technique* ([19, 20])

Such a technique provides for the utilisation of an ileal loop which is transposed into the newly formed recto-vesical space bent back like a gun-barrel. The median section is then incised after two weeks, so as to obtain a single cavity.

6) *Mori's technique* ([23, 24])

With this variation the loop of the small intestine is used as a single tube, and it is not bent back like a gun-barrel.

7) *Schubert's technique* ([19, 25])

This is carried out with the isolation of the rectum by way of the removal of the coccyx and part of the sacrum; then, the

intestine is at the full width of the canal, while the rectum is transposed into the brim of the anus. The excellent results obtained from the functional point of view [25] are contrasted by the complexity of the operation required and by the possible residual disturbances in defecation, including fecal incontinence [26, 27] besides the post-operatory complications.

8) *The vulvovaginal plastic technique (Williams - Capraro)* [3]

This technique provides for the construction of an antero-vulvar sac apt for allowing coitus. The sac, obtained by means of a U incision on the median face of the labia major, besides allowing coitus, serves to guide and rationalise the natural force applied during copulation, so as to lead, in a short time, to the creation of a recto-vescical space by simple pressure [28]. The operation is simple, but the vagina obtained is far from natural, both from an anatomical and a functional point of view.

9) *McIndoe's technique* [3, 12]

With this technique the lining of the space obtained by recto-vescical dissection is carried out with a dermo-epidermic border removed by dermotomy of the thigh, from the abdomen or back of the patient. The border is sutured onto a hollow and perforated acrylic support and afterwards inserted into the neo-vagina. The disadvantages of the method consist in rectal and vesico-vaginal fistulas in 5-10% of cases [10], necrosis of the cutaneous border, stenosis of the neovagina [15, 30], scars on the site of the removal [12], also of the keloid type [31], and dystrophy of the vaginal introitus [29].

10) *The technique of relining with amniotic membrane* [31]

This technique too, like those that follow, provides for preventive dissection of the recto-vaginal space, but in this case the intrusion is lined with amniotic membrane removed from a placenta at not more than 12 hours after delivery.

The technique is simple and the amnios has excellent regenerative and metaplastic capacity [33]; the results are good, both from an anatomic and a functional point of view [26].

A serious inflammatory reaction has been reported, with the formation of a granuloma of the vaginal vault [33] on account of immunitary reaction to the transplant, besides necrosis of the membrane.

11) *The technique of lining with transplant of fetal cutaneous borders*

This variation [12, 35] provides that the space provided is lined with the skin of fetuses born alive after 5 - 5½ months of pregnancy and which then died. The results obtained are good, but it is necessary to select the patients and plan the intervention awaiting the event of the possibility of a fetal donor. It is, however, a heterologous transplant.

12) *The technique of lining by means of pelvic peritoneum* [36, 37, 38]

In this technique, the neovagina is lined with a "tube" of pelvic peritoneum prepared through an abdomthennal stage and stretched below so as to reach the ostium of the neovagina. Thus complete epithelialisation with satisfactory results is obtained in a brief period of time [36, 39]. Both pelvic and vaginal infections have been reported, besides functional occlusion of the neovagina [36].

13) *The technique of lining with pedunculate borders of the bladder* [40]

In this case, the neovagina is lined with a border of the vescical wall 5 - 6 cm wide, "overturned" into the vagina by means of an incision of the pelvic peritoneum. It is, however, a complex operation and

vesical-vaginal fistulas and necrosis of the transplanted border have been reported.

14) *The technique of lining with peduncular borders of skin* ([42])

With this technique, conceived originally by Graves (1921), the neovagina is lined up to a certain height with two rectangles of mucosa obtained through the incision of the internal implant of the labia minor with successive separation of the two small sheets.

The principal disadvantage consists in the brevity (3 - 5 cm) of the tract lined, with the risk that the remaining tract may obliterate.

MATERIAL AND METHODS

The clinical case series examined include cases coming to our observation at the Gynaecologic and Obstetric Clinic of Padua from January 1965 to June 1987, and in the period 1969-1974 in the Center of Verona.

A wide case series regards 56 cases of vaginal malformation, distributed as recorded in Table 1.

Table 1. – *Distribution of the clinical case series in relation to the type of malformation (total cases: 56).*

Syndrome	No. cases
Rokitansky-Kuster-Hauser	42
Morris	10
Pseudo-hermaphroditism	4
Tot.	56

It will be observed that the most numerous groups are those reported as Rokitansky-Kuster-Hauser syndrome and Morris syndrome (respectively, 42 and 10 cases).

The average age of the patients is reported in Table 2.

Table 2. – *Average age of the patients examined.*

Syndrome	Average age	Maximal age	Minimum age
R.K.H.	20.5	30	10
Morris	21	34	15

From the examination of the patients affected by the R.K.H. syndrome it was revealed that 36% of the cases presented other associated malformations, especially regarding the urinary apparatus: this is easily understandable considering that both the genital and urinary structures derive from common embryonal blueprints.

Besides the urinary apparatus, the associated malformations were encountered in the skeletal apparatus, both isolated and multiple. High laxity of the inguinal canals was also met, with consequent bilateral inguinal hernias. With regard to the skeletal anomalies the schistasis of the posterior arch of the last two sacral vertebrae was found in two cases, the sacralisation of L5 in one case, and a supernumery cervical rib in another case. The urinary anomalies included both simple ectopy such as monolateral renal agenesia and the presence of a solitry pelvic kidney. In some cases urinary and skeletal malformations were associated (table 3).

Table 3. – *Vaginal agenesia. Incidence of malformations associated with the Rokitansky K.H. syndrome: tot. cases examined 33/41.*

Rokitansky K.H. syndrome without associated malform.	cases	21 (64%)
Rokitansky K.H. syndrome associated with other malform.:		
– single	cases	9
– multiple	cases	4
– total	cases	13 (36%)

All the patients were submitted to a series of clinical controls, and in particular to:

- anamnesis and objective examination
- routine examination
- pelvic echography
- laparoscopy
- urography
- hormonal profile
- chromosomic map
- psychiatric examination (only for those cases of pseudohermaphroditism, tending to ascertain the sexual identity of the patient).

DISCUSSION

Regarding the surgical techniques adopted it will be seen from Table 4, that in practise almost all the principal groups of intervention are represented, with exception of the technique with transposition of the intestinal loop. The most nume-

Table 4. – *Surgical technique adopted in cases treated (no. 53). (Gyn. Obst. Clin. Univ. of Padua, Center of Verona: 5). (Gyn Obst. Clin. Univ. of Padua: 48).*

Rokitansky-Huster-Hauser syn.: Frank	
– neovagina acc. Frank+Warthon	20
– neovagina acc. Capraro	2
– neovagina acc. McIndoe	4
– neovagina acc. Vecchietti	14
Morris syndrome:	
– neovagina acc. Frank	7
– neovagina acc. Warthon	3
Pseudo-hermafroditism:	
– neovagina acc. Frank	2
– neovagina acc. Warthon	1

rous groups are those relating to the Frank's and Warthon's techniques ([28, 52]) and to Vecchietti's technique.

Of all the clinical case series only three cases were not treated, precisely two cases of pseudohermaphroditism and one case of Morris syndrome.

Vecchietti's operation has been the most adopted in our Institute, with Frank's operation. It allows the creation of a neovagina covered by mucosa which anatomical and receptorial characteristics are analogous to normal vagina. For this reason, such methodology has been also adopted by Anglo-Saxon and French Schools as the elective operation for vaginal agenesia correction, especially in Rokitansky-Kuster's syndrome.

This technique permits us to avoid the collateral effects, which, on the other hand, derive from other methodologies as, for examples, the vaginal hypersecretion (Baldwin's technique) or the lack of aestheticism (William's technique), which create some acceptance problems for the patient.

Frank's technique was also experimented for the treatment complete vaginal septa previous to surgical section of the septa themselves.

The dilators used in the Gynaecologic and Obstetric Clinic of Padua consist of a support in rigid plastic material (PVC), covered by a rubber tube which is turn coveder by foam rubber.

On the outside, it is moulded with latex film. Each dilator is mounted on a circular support of rubber cloth provided with four metal buttonholes which allow it to be anchored.

With regard to the complications which arose. and the unsuccessful cases we noticed, these include a recto-vaginal fistula in a case treated according to Vecchietti's technique and a case of hemorrhage of the recto-vesical space in a case treated according to Warthon's technique (table 5).

Table 5. – *Complications or failures.*

Failures:	
– neovagina acc. Vecchietti	1
– neovagina acc. Frank	1
Complications:	
– recto-vag. fistula (acc. Vecchietti)	1
– hemorrhage (acc. Frank+Warton)	1

Before vaginal malformations were surgically treated, they were usually associated with a general antibiotic therapy and a hormonal therapy. The cases of pseudohermaphroditism were obviously submitted to different hormonal treatment.

CONCLUSIONS

As will be evident from the description of the various surgical techniques, the number of operation proposed is multiple. From the analyses of Literature, it is likewise evident that every Author has experienced advantages and disadvantages related to every technique used without being able to arrive at a definition of a single type of conduct to be followed with these patients.

In our opinion the criterion to follow is that of personalising treatment, adapting

it as far as possible to the anatomical, psychological and clinical situation of every single patient.

In practise, in cases where the patient presents the sketch of a vagina at least 2 cm deep the most suitable method, in our opinion, reinforced by our experience, is Frank's.

In those cases where the depth is, instead insufficient, but in the presence of normal external genitals, it would be possible to practise Vecchietti's, Williams - Capraro's or Warthon's with Frank's technique later on.

It seems proper to underline how the moment chosen for the operation must immediately precede the initiation of regular sexual activity, both to give the patient time to complete her somatic development, given that the neovagina will not lengthen with a patient's development ([5]), and because regular sexual activity prevents stenosis of the neovagina itself.

Finally, even before the choice of the type of treatment, there remains the problem of studying the indication for surgical intervention, in as much as failure of the therapy cannot be excluded.

We may, therefore, conclude that the orientation of our School is to avoid complex surgical techniques which bring operative and post-operative high risks to the patients, and prefer others which are, less traumatic and personalized to individual cases, and understood in relation to the site in which they are presented. "Therapeutic personalisation" in every single case, also in relation to the age of the patient, remains a key-point in our School.

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