Surgeries for Genital Tract malformations

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Summary: **The objective was to evaluate scope and limitations of surgical correction of genital tract defects in adolescent girls.**

Two hundred and seventy three women with different grades of Mullerian dysgenesis were categorised into two groups:

(a) Those without a functioning uterus (n = 249) and

(b) Those with a functioning uterus (n= 24).

In group A, Vaginoplasty was performed in 85 while Hystero-vaginoplasty was attempted in 164 women. Amniotic membrane graft was used for vaginoplasty. In Group B, 16 women had cervico-vaginoplasty, 4 had resection of vaginal septum and 2 had retrograde cervical canalisation.

In group A, 17 women (10.3%) were menstruating for more than 10 years. Vaginal length was satisfactory in 155 (62.2%) women, though no pregnancy was recorded in this group. In group B, cyclic menstruation could be restored in 100% women, satisfactory vaginal length could be achieved in 66.6% and 5 women delivered viable babies.

In conclusion, it appears that surgical treatment for genital tract malformations in adolescents can avoid psychological impact and may help in the onset of cyclic menstruation and offer some potential for fertility restoration in women with a functioning uterus.

Introduction:

An adolescent girl usually presents with problems related to lower genital tract defects. Lower genital tract anomalies are frequently associated with similar abnormalities of upper genital tract. These defects invariably lead to either true amenorrhoea or cryptomenorrhoea. The present communication will deal with our personal experience of the scope and limitations of reconstructive surgery for genital malformations involving both lower and upper genital tract.

Materials & Methods Of Diagnosis:

From Jan 1973 to Dec 1997, 273 women in the age group ranging from 12 to 20 years were referred to our clinic with different grades of utero-cervico-vaginal atresia. The exact nature of Mullerian malformation was primarily assessed by clinical evaluation and finally confirmed by laparotomy.

Currently ultrasonography, laparoscopy and even MRI are being used to identify the precise nature of the defect, but cervical atresia and its degree can perhaps be exactly diagnosed only on laparotomy

A definite correlation almost always exists between the type of anatomic defect of the vagina and the nature of abnormalities found in the internal pelvic organs. We have classified vaginal malformations in four grades (Table I). The classification has been made on the basis of their prognosis after surgical correction. For example restoration of menstrual and reproductive functions is possible in Grade I (a), Grade II, Grade III and Grade IV malformations while these are not possible in Grade I malformations.

Imperforate hymen and "short blind vagina" associated with testicular feminising syndrome have not been included in this classification.

Vertical septum of vagina when unassociated with cervical or uterine septum do not create problem and hence surgical treatment is not essential. However, rarely a vertical vaginal septum attached to lateral vaginal wall with or without a similar septum in the cervix or uterus may create the problem of lateral haematocolpos and haematometra. This group of women should be treated

Table I. Grading of genital tract malformations

Gr.I :	Complete agenesis of vagina with absence of functioning uterus (typical Mayer Rokitansky Kuster Hauser Syndrome (Fig: 1,2,3)	249
Gr.I(a) :	Complete atresia of vagina and cervix with a functioning uterus (Fig:4)	2
Gr.II :	Agenesis of upper one third of vagina and cervical atresia with a functioning uterus (Fig:5)	16
Gr.III :	Transverse septum or agenesis of lower third of vagina, normal cervix and functioning uterus (Fig:6).	4
Gr.IV :	Isolated cervical atresia with normal uterus and normal vagina with or without constriction of upper part of vagina (Fig:7).	2

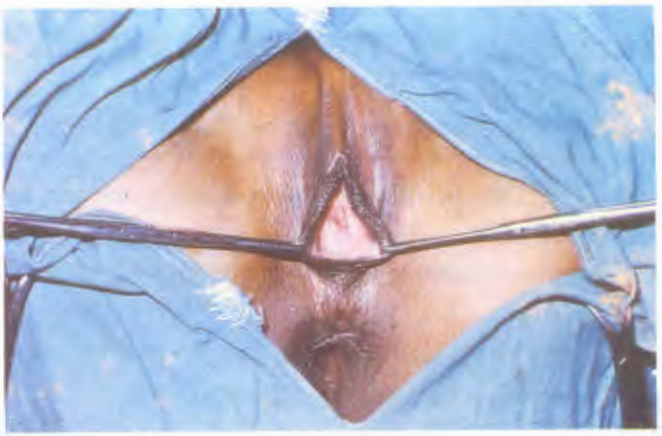

Fig 1. Complete absence of Vagina (R.K.H. Syndrome)

Fig 3. Small mullerian knobs, in RKH Syndrome; (unsuitable for hysteroplasty).

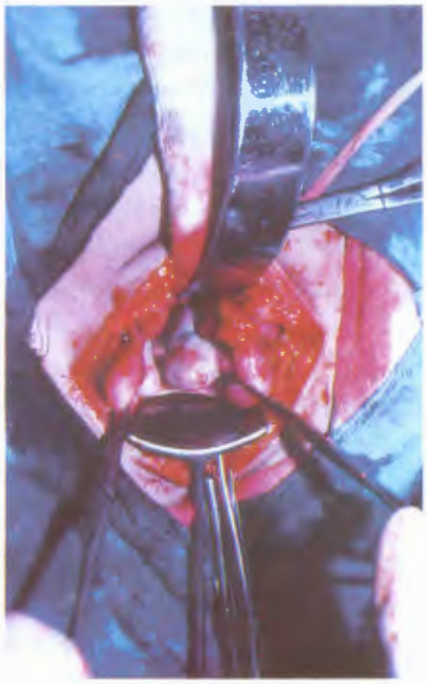

Fig 2. Internal pelvic organs in RKH syndrome. Big mullerian bulbs, (grasped with Allis Tissue Forceps) with Fibromuscular cord connecting the bulbs; normal ovary with corpus luteun, suitable for hysteroplasty.

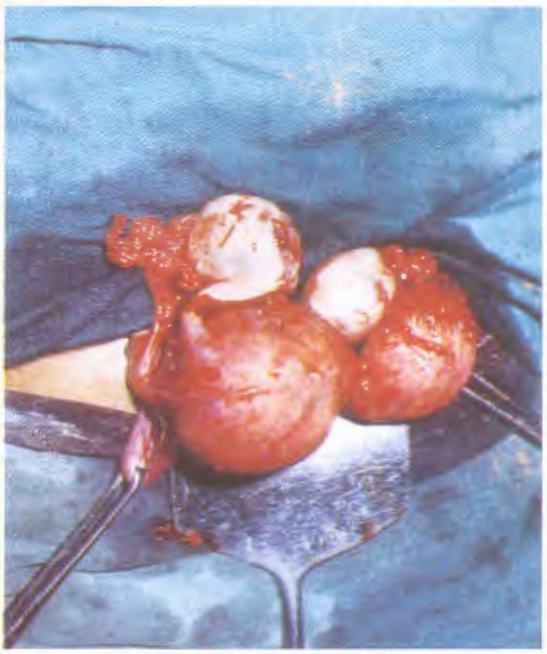

Fig 4. Functioning uterus (bicornuate in the picture with haematometra in one horn; rare association with RKH syndrome, less than 2%; common with cervico-vaginal or cervial atresia.

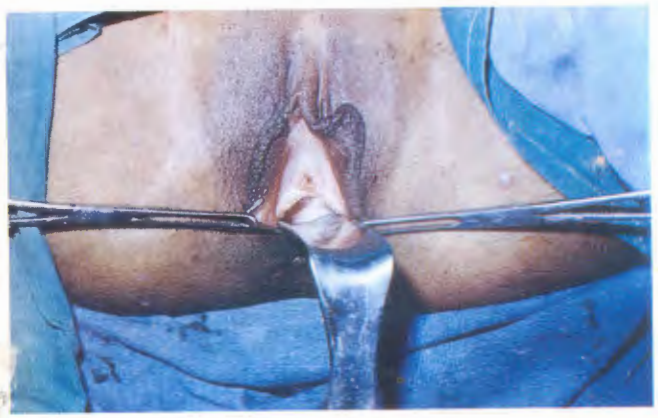

Fig 5. Upper third vaginal agenesis. Diagnosis may be confused with transverse vaginal septum. U.S.G. or needle puncture test may help.

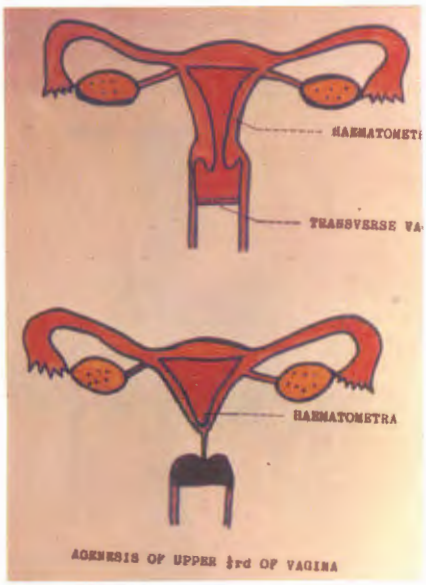

Fig 6. Diagrammatic representation of upper third vaginal agenesis and transverse vaginal septum. Cervical atresia with only haematometra is the invariable finding in upper third agenesis whereas colpohaematometra is clinical sequele of tansverse vaginal septum

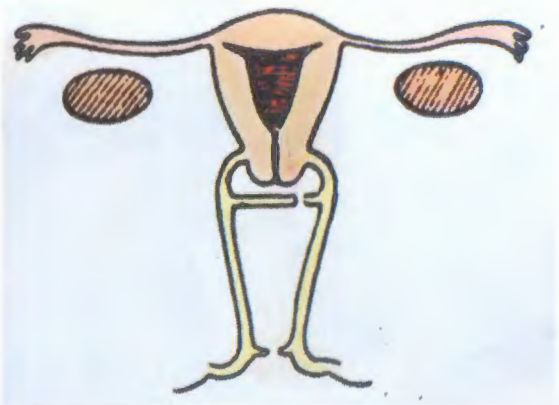

Fig. 7. Isolated cervical atresia with annular constriction of upper part of vagina.

in the line similar to those advocated for transverse vaginal septum.

Surgical Approach For Different Types Of Utero-Cervico Vaginal Atresia

Basis of Surgical Approach:

The objective of surgical approach is to restore sexual and if possible, the menstrual and reproductive function. The surgical approach, however has to be tailored depending upon the existing anatomical abnormality and the dexterity of the surgical team.

Grouping of cases for Surgical Approach on the basis of Anatomical Deformity

Surgical treatment may be classified into two groups:

(a) Cervico-vaginal atresia without a functioning uterus (Classic Rokitansky-Kuster-Hauser Syndrome)

(b) Cervico-vaginal or Isolated Cervical atresia with a functioning uterus.

A. Surgical Treatment For Cervico-Vaginal Atresia Without A Functioning Uterus

Vaginoplasty is the accepted form of treatment. Vaginoplasty means creation of a functioning vagina. This can be performed in one of two ways : dilatation or surgery. In the dilatation procedure, Frank's technique involves creation of a vaginal vault. This is accomplished by applying pressure to the center of the dome of blind vagina with progressively larger Lucite dilators. If dilatation procedure is not possible or acceptable, surgical reconstruction should be performed.

Various operations with a variety of grafts were developed in the past and most of them have been discarded. Modified Mc Indoe using split thickness skin graft is the currently practised procedure for vaginoplasty. The author uses amniotic membrane instead of skin with encouraging results.

B. Surgical Approach For Cervico-Vaginal Atresia And Transverse Vaginal Septum With A Functioning Uterus

Two types of surgical treatment have been suggested for this group of women viz, Conservative approach by surgical canalisation of genital tract or hysterectomy with vaginoplasty.

Steps of surgical canalisation in both cervico-vaginal and cervical atresia are similar except that vaginal reconstruction is not essential in "Isolated Cervical Atresia".

Steps of operation:

The operation is performed in stages but all in the same sitting.

Stage I: Vaginal Approach

An avascular space between the bladder and the rectum is created for the neovagina by sharp and blunt dissection. Dissection is carried upwards till the shiny peritoneum of the Pouch of Douglas is visible. This space is temporarily plugged with a roller gauge.

Stage II: Abdomino-Vaginal Approach

Abdomen is opened by low transverse incision. Bladder peritoneum is incised. Bladder with bladder peritoneum is pushed down with sharp scissor dissection as low as possible so as to expose the vault of blind vagina (dissection like abdominal radical hysterectomy). The cord like cervical structure is exposed. The lower blind end of this cordlike cervical tissue is pulled up by means of a "stay" suture. Another stay suture is fixed at the lower end of the corpus just above the junction of the cervical tissue and the body of the uterus. A vertical incision is made in between these two stay sutures dividing the cervical tissue halfway through the thickness. The incision opens up the lower part of the body of the uterus through which the collected blood (haematometra) escapes and is aspirated. The lower ends of the bisected cervical cord are now anchored by two stay sutures. The previous stay sutures fixed at either end of the solid cervix are now removed.

A long artery forceps is then negotiated from below through the neovagina (after removing the roller gauge) which will stretch the vault of exposed vagina below the lower incised edges of the neocervix. This can be seen

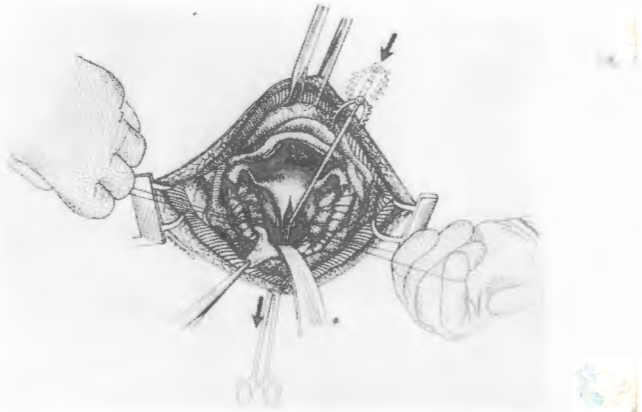

Fig 8. Abdominovaginal approach in cervico-vaginplasty. Bladder has been retracted. Solid hypoplastic cervix has been incised half way through its thickness. Tip of artery forceps can be seen protruding through incised vault of vagina. Artery forceps will pull down the 'Stay Sutures' and the barrel of loop introducer fabricated to IUCD inside the neovagina. Barrel will act as cervical stent.

Fig 9. Incised cervical tissue with cervical stent have been pulled inside neovagina.

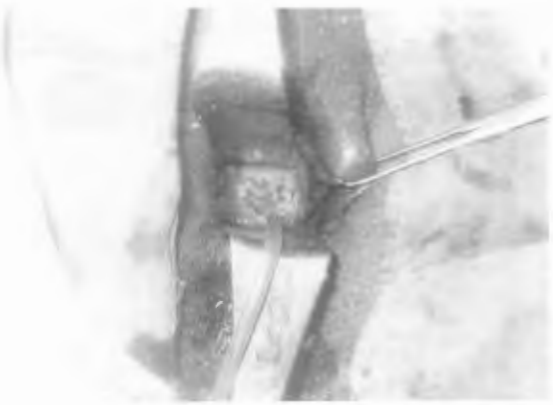

Fig 10. Cervical lips have been everted and fixed to the vault to act as ectocervix.

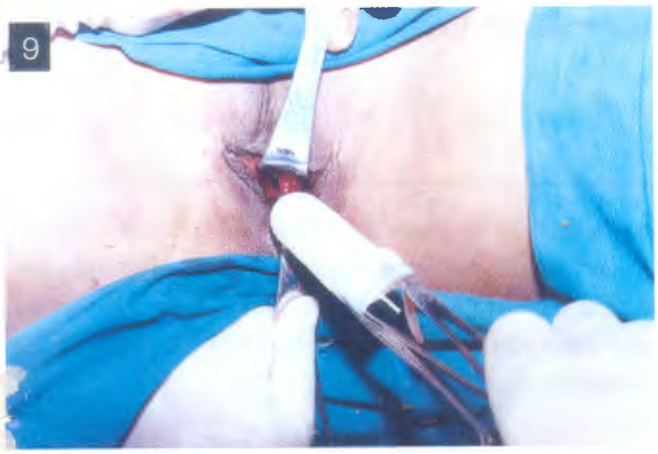

Fig 11. Vaginal mould covered with fresh amniotic membrane is about to be introduced inside the neovagina. The Cervical Stent is negotiated through a central hole inside the mould.

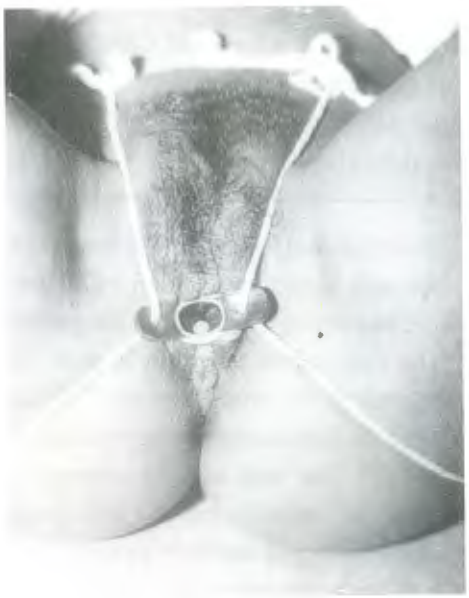

Fig 12. Patient changes the mould every day and reintroduces the way shown in the picture.

Table II : Complete absence of vagina with absence of a functioning uterus (typical R.K.H Syndrome Gr.I) Vaginoplasty and Hysterovaginoplasty.

Total No. of cases	249
Vaginoplasty	85
Vaginoplasty with Hysteroplasty	164
Number menstruating for more than 10 years	17 (10.3%)
Uterus reasonably adult size	14 (8.5%)
Satisfactory vaginal length	155(62.2%)
Pregnancy	NIL

Table III: Complete or upper one third vaginal agenesis with cervical atresia and a functioning uterus (Gr.Ia,GrII):

Cervico-Vaginoplasty

Number of cases operated	18
Number having regular periods	18
Satisfactory vaginal length and stretchability	12(66.6%)
Pregnancy and viable deliveries	2

Table IV: Transverse septum, lower third vaginal agenesis, normal cervix, functioning uterus (Gr.III) Resection of Septum-Vaginoplasty

Number of cases operated	4
Number having regular periods	4
Satisfactory vagina	4
Viable deliveries	2

Table V : Isolated Cervical Atresia (Gr.IV)-Retrograde Cervical Canalisation

No. of cases operated	2
Regular periods	2
Viable delivery	1

easily through abdominal route after the bladder has been retracted. Through a stab incision on the blind vaginal vault the tip of the artery forceps can be pushed inside the pelvis. The opening of the vault is now dilated by stretching the arms of the artery forceps. The barrel of a loop introducer is fabricated to the vertical bar of a Multiload Cu 250. This will act as a cervical stent and the loop inside the uterine cavity will prevent it's expulsion. The barrel and the stay sutures fixed in the lowermost ends of the neocervix are now grasped by the tips of the artery forceps which is still projecting inside the pelvis (Fig 8). These structures are now pulled down inside the neovagina. The Multiload Cu 250 is pushed into the uterine cavity. The cut edges of the neocervix are now united by rows of catgut sutures over the cervical stent, the lower stitch takes a bite on the vault of the vagina. The raw area is covered by bladder peritoneum and abdomen is closed in layers.

Stage III: Vaginal Approach

Neovagina is now retracted by two Landon's retractors. By traction on the stay sutures the lowermost cut edges of the cervix are pulled inside the neovagina (Fig 9). The pulled down part of the cervix is then fixed to the vault of the vagina which will act as an ectocervix (Fig 10). A plastic mould with a central hole for negotiation of projecting end of the cervical stent is covered with amniotic membrane and is placed inside the neovagina (Fig 11). Amniotic membrane is obtained from a placenta following caesarean section delivery performed on the same day. The membrane is washed several times with Ringer's solution and should be made blood free before it is placed on the vaginal mould. Labial skin is apposed to keep the mould in place. A self-retaining catheter is introduced for post-operative bladder drainage.

Post Operative Follow Up

The labial stitches are removed on the sixth post operative day. The patient or her mother is trained to wash and change the mould every other day. The mould can be fixed around the groin with tapes in the way as shown in Fig 12.

The mould can be gradually withdrawn between 6 months to one year. The cervical stent is removed after one year. Repeat cervical dilatation and incision on the cicatrix near the vault of the vagina had to be performed for retrograde cervical dilatation.

Transverse vaginal septum and lower third vaginal agenesis

In transverse vaginal septum, resection of septum is only necessary. This can be performed by vaginal approach. But if doubt exists about the exact nature of the defect abdomino-vaginal approach will be safer procedure. Introduction of a needle through the septum and aspirating dark coloured blood will settle the diagnosis in favour of vaginal septum.

Results

Results have been analysed with regard to the onset of cyclic menstruation, adequacy of reconstructed vaginal length and viable deliveries (Tables II, III, IV and V).

Discussion

In the absence of a functioning uterus, vaginal reconstruction should be performed in the late adolescent period, that is between the age of 16 to 20 years. It is expected that at this age the girl will become mature and will take care of the mould which she has to use for some time in the post-operative period.

There are reports (Jones, 1994) which suggest removal of rudimentary Mullerian bulbs either by laparotomy or through laparoscope, on the ground that they wer responsible for pain in these women. The author has performed metroplasty in addition to Vaginoplasty in this group of women (Chakravarty, 1977). Mullerian knobs in these women invariably contain islets of endometrial glands and stroma and occasionally small amount of blood (haematometra). Though pregnancy has not yet been reporded, about 10 percent of women are menstruating regularly for more than 10 years and the reconstructed uterus has reasonably increased in size. W

have observed in our preliminary studies (Chakravarty, et al, 1977) that patchy endometrium develops within the cavity of the newly created uterus formed by bisecting the solid Mullerian bulbs halfway through their thickness and then uniting them over a uterine stent. It may be possible that the unfused Mullerian bulbs, once united in the midline, may respond to the growth stimulus offered by normally existing endogenous ovarian steroids and finally assume the size of an adult uterus.

We have used amniotic membrane graft instead of skin for vaginal reconstruction. We believe that amniotic membrane acts as a scaffolding material preventing the growth of excess granulation tissue. It is possible that the epithelium from the dome of blind vagina grows upwards and covers the raw area of the newly created vagina. In fact, the vaginal epithelium covering that follows after amniotic membrane graft looks pinkish with normal vaginal mucous membrane rugosity and stretchability.

Isolated transverse vaginal septum or lower vaginal agenesis seldom creates therapeutic problems, if the diagnosis is made correctly and at the correct age. But cervical atresia with a functioning uterus with or without vaginal agenesis indeed creates a surgical dilemma.

There are two treatment options. These are conservation of uterus with restoration and maintenance of utero-cervico-vaginal patency or hysterectomy with vaginoplasty.

There are several reports in which authors (Mariculla et al, 1978; Geary & Weed, 1973; Farber and Merchant, 1976) primarily attempted to conserve the uterus by creating a fistulous tract between uterus and neovagina but subsequently hysterectomy had to be performed because of restenosis and consequent pelvic infection. Two deaths have been reported due to postoperative fulminating pelvic peritonitis following the conservative procedure (Geary & Weed, 1973; Niver et al, 1980). On the other hand, there are sporadic reports of achieving spontaneous pregnancies following successful canalisation of an atretic cervix with or without combining vaginoplasty (Zarau et al, 1973; Fraser 1989; Hampton et al, 1990). There are still others who have achieved pregnancies following creation of a fistulous tract between the uterus and vagina by the procedure of assisted reproductive technology (Thijssen et al, 1990; Nargund & Parsons, 1996).

Considering the sporadic success and the published reports of devastating morbidity and mortality, one should be very critical in selecting patients for conservative surgical approach. In general, the chances of fertility will markedly be compromised by the presence of endometriosis, pelvic infection and the age of the woman at which operation is performed. In our experience, we have refused conservative surgery in women above the age of 20 years. Selection of girls at this adolescent age has allowed us time for repeated dilatation of neocervix and on a few occasions, retrograde dilatation of cervix by relaparotomy as well. While waiting for relaparotomy for retrograde dilatation consequent upon cervical restenosis, these girls were advised medroxyprogesterone acetate to suppress menstruation and prevent haematosalpinx and haemoperitoneum.

Possibility of spontaneous pregnancy following canalisation of cervico-vaginal atresia is very low. Four cases have been reported so far in the literature (Zarau et al, 1973; Hampton et al, 1990, Bates and Wiser, 1985). Though in these reports the type of cervical atresia has been stated as partial the type of vaginal agenesis has not been clearly mentioned. It appears from our experience as well as 7 cases reported by Fujimoto et al, (1997) that with complete vaginal agenesis, cervix is also totally hypoplastic and atretic. Only with lower third vaginal agenesis or transverse septum, where there is blind vaginal pouch above, cervix remains anatomically normal though rarely there may be conglutination of external os with an empty blind vaginal pouch.

Two cases in our series, one with complete cervico-vaginal atresia and the other with upper-third vaginal agenesis and total cervical occlusion achieved spontaneous pregnancy and had viable delivery following successful canalisation. Review of literature (Fujimoto et al, 1997; Rock et al, 1984) revealed that there is no documented evidence of pregnancy until now following

surgical correction of total cervical atresia combined with complete vaginal agenesis.

The rare incidence of pregnancy following successful canalisation of atretic cervix has been attributed to the lack of normal endocervical glandular function (Fujimoto et al, 1997). Though this may be one of the factors responsible for infertility in these women restenosis following canalisation is perhaps the more significant cause which prevents pregnancy. It has been reported (Fujimoto et al, 1997) that though 23 out of 39 patients experienced normal menstrual bleeding immediately following canalisation operation, only 4 eventually achieved patency after repeat canalisation procedures.

We have already reported that patchy endometrium develops in the newly created uterus reconstructed by unification of solid Mullerian bulbs in RKH Syndrome provided the cavity could be kept patent by placing an uterine stent. Since uterus and cervix are both Mullerian in origin, it may be possible that cervical epithelium may grow from mesodermally derived tissue if the patency of the cervical canal could be maintained for some length of time. Restenosis usually occurs at the junction of neocervix and neovagina. Therefore, our practice is to pull down portion of neocervix inside the neovagina and fix it to the vault of the newly created vagina. This will act as ectocervix. Most of the authors reported creation of fistulous tract between uterus and vagina. Attempt to create a new ectocervix (wherever possible) has been totally ignored. Our novel operative step, we believe, will reduce the risk of restenosis and improve the possibility of permanent patency. Of course, apart from technical accuracy the success will depend on the tissue available to create a neocervix and partly on the length and diameter of solid cervical cord Hampton et al, (1990) used a small piece of skin graft and sutured around the hub of a paediatric endotracheal tube which acted as a cervical stent. Whether this graft was able to assist cervical mucous secretion was only speculative.

In conclusion, it appears that with the lower genital tract defect, a functioning uterus may or may not exist. Vaginoplasty is the conventional treatment for those who do not have a functioning uterus. In selected cases, however, simultaneous hysteroplasty may be attempted. In the other group of women, who have a functioning uterus successful canalisation may be possible. This should be attempted while the patient is young otherwise the adverse consequences of outflow obstruction like endometriosis and recurrent pelvic infection may contraindicate a conservative surgical approach.

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