CONGENITAL ATRESIA OF VAGINA, WITH INCOMPLETE DEVELOPMENT AND FUSION OF MUELLERIAN DUCTS WITH SPECIAL REFERENCE TO THE ROLE OF RECONSTRUCTIVE SURGERY

by


and

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Introduction

Subjects of atresia vagina are typically feminine in appearance with well developed secondary sex characters. Some of these cases are chromatin negative; these are cases of Testicular Feminisation. In these cases, gonads are testes, but the target organs are insensitive to androgen produced by testes. This condition has already been described in a previous communication, Gun et al (1973).

The other group of cases of atresia vagina are chromatin positive. Mullerian fusion, in these cases is practically absent. Though tubes are well developed, the uteri are represented by two rudimentary knob-like structures attached to medial ends of each fallopian tube. Gonads are as a rule ovaries; histologically and functionally they are normal.

The present study is based on the record of 15 such cases emphasising specially on the scope of establishing menstruation by reconstructive surgery utilising tubal graft. These cases were selected from a group of cases of primary amenorrhoea attending the outpatients' department of Gynaecology, N.R.S. Medical College Calcutta, during the period of 4 years from 1.6.69 to 31.5.73.

Methods of Study

After clinical examination, each case had a buccal smear tested for detection of chromatin dot in the nuclei of the squamous epithelial cells. Detailed chromosome analysis could be done in two cases. For lack of adequate facilities, this investigation could not be performed in others. A few were selected for pelvic pneumogram and intravenous pyelogram. All cases had exploratory laparotomy and ovarian biopsy was performed in 12. In 4 cases, reconstructive surgery was attempted with a view to establish menstruation by uniting the two rudimentary uteri and creating an artificial utero-vaginal canal on a polythene tube. Tubal graft was used with the hope that this would provide a source for generation of endometrium in the newly constructed uterine cavity.

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Clinical, Laboratory and Laparotomy Findings:

TABLE I
Findings on Clinical Examination

(a) Age:

<table>
<thead>
<tr>
<th>Age in years</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>15-20</td>
<td>13</td>
</tr>
<tr>
<td>21-30</td>
<td>1</td>
</tr>
<tr>
<td>Above 31</td>
<td>1</td>
</tr>
</tbody>
</table>

(b) Marital Status:

<table>
<thead>
<tr>
<th>Status</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Married</td>
<td>9</td>
</tr>
<tr>
<td>Single</td>
<td>5</td>
</tr>
<tr>
<td>Divorced</td>
<td>1</td>
</tr>
</tbody>
</table>

(c) Secondary Sex Characters, including external genitals were well developed in all cases. Patients looked feminine in all respects.

(d) Length of Vagina:

- Almost absent, represented by a dimple: 7
- 1 inch: 1
- 1½ inches: 2
- 2 inches: 5

(e) Pelvic Findings (Rectal Examination)

- Nodular uterus felt: 8
- Uterus not felt: 7

The diagnosis of absence of vagina is usually delayed till the age of 15 when the girl or her mother seeks medical advice for failure of onset of menstruation. Detection in few cases may be further delayed till the woman is married and she comes for help not only for primary amenorrhoea but also for difficulties of marital relationship. Thirteen cases in the present series were diagnosed between the ages of 15 and 20 and 9 were already married.

In married women with atresia vagina, dyspareunia is an expected but not an invariable symptom. Five women in this series who were married did not complain of dyspareunia and the length of vagina in these cases measured about two inches. Perhaps, vagina acquired some length due to adjusted marital relationship.

Rectal examination is imperative not only to detect presence or absence of uterus but also to exclude haematocolpos and haematometra. To ascertain correct pelvic findings rectal examination is not enough. Help of the laparoscopic examination or laparotomy is essential. This is evident from the findings of the present series.

Rudimentary double uterus was present in all cases, connected by a fibromuscular band. These uteri were found at the medial ends of the fallopian tubes. The sizes of the rudimentary knob varied in different cases, invariably rudimentary uterus of one side was relatively bigger than the rudimentary uterus of the other side. In two cases the tubes and the round ligaments were hypoplastic. In these cases the knob-like uteri were also extremely rudimentary. There was, however, no relationship between the length of vagina and the condition of the fallopian tubes. Ovaries looked normal in all cases.

Ovarian Biopsy

This was done in 12 cases and evidence of corpus luteum was found in 11.

Reconstructive Operation With Result:

In 4 cases reconstructive surgery was attempted.

(a) Incision: After opening the abdomen, incision was made on the medial aspect of the rudimentary uterus of either side and was extended up to the adjoining fibromuscular band. The incision was carried half way through the thickness of these structures. In 3 out of 4 cases a small canal representing miniature uterine cavity was detected in the rudimentary uterus, but there was no
TABLE II
Findings of Special Investigations

<table>
<thead>
<tr>
<th>Investigation</th>
<th>No. of cases investigated</th>
<th>Observation</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Buccal smear</td>
<td>15</td>
<td>All were chromatin positive.</td>
<td></td>
</tr>
<tr>
<td>Karyotype</td>
<td>2</td>
<td>46, xx</td>
<td>This was done by leucocyte culture. For lack of adequate facilities, the test could not be done in others.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>46, xo/xx</td>
<td></td>
</tr>
<tr>
<td>Pelvic pneumogram</td>
<td>4</td>
<td>Shadow of ovaries of reasonable size in all cases.</td>
<td>Cystoscopy examination is not possible in atresia vagina. For pelvic findings, we preferred laparotomy than laparoscopic examination.</td>
</tr>
<tr>
<td>Pyelogram</td>
<td>9</td>
<td>Normal</td>
<td>Urinary tract abnormality is fairly common with genital tract malformation.</td>
</tr>
</tbody>
</table>

TABLE III
Findings at Laparotomy

FINDINGS
(a) Uterus
  Rudimentary double uterus
  One rudimentary and one moderately developed uterus

(b) Fallopian Tubes and Round Ligaments
  Normal
  Hypoplastic

(c) Ovaries
  Normal looking
  Recent or old corpus luteum
  No evidence of corpus luteum

<table>
<thead>
<tr>
<th></th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rudimentary double uterus</td>
<td>14</td>
</tr>
<tr>
<td>One rudimentary and one moderately developed uterus</td>
<td>1</td>
</tr>
<tr>
<td>Normal</td>
<td>13</td>
</tr>
<tr>
<td>Hypoplastic</td>
<td>2</td>
</tr>
<tr>
<td>Normal looking</td>
<td>15</td>
</tr>
<tr>
<td>Recent or old corpus luteum</td>
<td>14</td>
</tr>
<tr>
<td>No evidence of corpus luteum</td>
<td>1</td>
</tr>
</tbody>
</table>

...
Abdominally the vault was incised and the tip of the forceps was pushed into the pelvis.

(d) Fashioning the Tubal graft: Incision on one rudimentary uterus was extended laterally up to the uterotubal junction. The medial end of the fallopian tube was cut and was separated at its medial part from the mesosalpinx. The tube was then split longitudinally for about one inch.

(e) Unification of the Rudimentary Uteri: This was done on a polythene tube. Two rows of continuous suture, using ‘0’ catgut on atraumatic needle, apposed the raw areas of uteri on either side. The sutures started posteriorly from the fibromuscular band, went up to the fundus of the rudimentary uteri. Before closing the anterior aspect, the split fallopian tube was spread and fixed to the inner wall of the united rudimentary uteri. The anterior raw edges were then apposed together using the same continuous suture, ending anteriorly at the fibromuscular band. The two fibromuscular bands united on the polythene tube, thus formed a potential tube to represent the cervical canal.

(f) Fixation of the Fibromuscular Band and Polythene tube: The ends of the catgut with needle and the polythene tube were then brought down into the vagina with the help of sponge forceps. The united fibromuscular band thus brought down was fixed on to the walls of vagina. The polythene tube was also sutured to the side wall of the vagina by a nylon stitch. The end of the tube was cut flushed with labia minora so that it did not project outside.

(g) Putting a Mould and a Graft: After ensuring complete haemostasis, skin graft on a mould was placed in the space created for artificial vagina. Opposite margins of labia minora were sutured to keep the mould in place. A Foley catheter was placed inside the bladder for continuous drainage.

Follow-up

These cases are getting cyclical oestrogen therapy.

Result

In 2 cases the mould and the polythene tube came out on the 10th and 12th postoperative days. In 2 cases mould has been removed after 6 months but the polythene tube is still in position. Two of these cases have menstruated and menstruation is continuing with cyclic oestrogen therapy.

Discussion

While the condition of atresia or absence of vagina is well recognised, the other abnormalities of pelvic organs associated with such malformation have not been very clearly authenticated. Te Linde (1962), quoting Counseller has stated that in vast majority of women born without vagina, there is also congenital absence of the uterus. Macleod & Hawkins (1964) hold the same view. Hawkins & Bourne (1971) believe that with congenital absence of vagina, the uterus, if present, is either duplicated or rudimentary. Though reports of anatomically and functionally normal uterus with complete absence of vagina is rare, Counseller (quoted by Te Linde, 1962) on laparotomy, found normal uterus in 4 cases, endometriosis in 3 and haematosalpinx in 3. One of us found haematosalpinx in a single uterus. This case has not been included in this series. Unilateral partial vaginal atresia with functioning double uterus has been reported by Hingorani (1972) and Heera (1973).
Hence the theory of mullerian agenesis along with atresia vagina cannot be fully accepted. Our study has shown that in phenotypic females, mullerian agenesis is present in cases of testicular feminisation syndrome. In a patient, who is chromatin positive and has absence of vagina, we have never found complete agenesis of mullerian ducts. In most of the cases, we have found normal ovaries, normal fallopian tubes and rudimentary double uteri connected by a fibromuscular band.

At the present stage, we cannot offer a satisfactory explanation for this abnormality. Probably, the factors responsible for normal development of the upper part of mullerian ducts (e.g. fallopian tubes) is different from that controlling the middle (uterus) and the lowest part (vagina) of the duct. Chromosomal abnormality may be a factor responsible for this particular abnormality. In one of our cases, definite mosaicism was noted. Detailed chromosomal analysis might show mosaicism in many of these cases.

The treatment that can reasonably be offered to these women, is to create an artificial vagina, by which it is hoped that they would be capable of adjusting to normal marital relationship. The possibility of establishing menstruation in at least some of these women has never been considered. While performing routine laparotomy in the present study, it was observed that in some cases the two rudimentary uteri on either side, if connected together would assume the shape of a single hyoplastic uterus. This was done in 4 cases and the medial end of one fallopian tube was grafted within the potential space of the newly constructed uterus. Reifenstuhl and Kroemer (quoted by Basu Mullik, 1971) have used tubal graft in cases of uterine synechia with encouraging result. At least two cases in this series have menstruated one to one and a half year after the operation just before writing this article.

That the mucous membrane of the fallopian tube may behave like endometrium is a presumption that is yet to be established. But it may be possible that the endosalpinx, by changing its environment within the uterine cavity may be transformed into mucous membrane resembling endometrium, both anatomically and functionally. Final opinion is reserved till this is proved experimentally.

Summary

(i) Fifteen cases of congenital atresia of vagina have been presented.
(ii) All cases were chromatin positive and had well developed secondary sex characters.
(iii) Ovaries were normal in each case, normal fallopian tube was found in most of the cases, while rudimentary double uteri were present in all cases. In one, both the kidneys were found on right side.
(iv) Unification of two rudimentary uterii with tubal graft and restoration of the uterovaginal canal performed in 4, yielded satisfactory result in two cases only.

Acknowledgement

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References


See Figs. on Art Paper III