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CONGENITAL MALFORMATIONS OF FEMALE GENITAL TRACT

BY

S. N. UPADHYAY, B.S., M.D., (Pat.), M.R.C.O.G. (Lond.)

and

SUKUMAR MITRA, M.B., B.S. (Pat.)

Department of Obstetrics and Gynaecology,

P.W. Medical College, Patna.

Variations from complete absence of development of the female genital organs to its milder deviations in development (like the uterus arcuatus) have been recorded. Certain group of cases develop clinical symptoms and signs to warrant attention, while the remaining may not be recognised during life time. The problems presented to gynaecologists in management of such cases are often embarrassing as no relief may be possible in some of them. Surgical relief to the extent of providing successful marital relations can probably be offered to almost all of them, but provision for conception may not be possible in a very great majority of instances. Till further knowledge of embryology enables us to understand the aetiology of such aberrations, and uniform opinion regarding surgical treatment is crystallized to bring about maximum utility out of minimum presented in individual cases, interest in the subject will continue.

Case Material

A review of the records of cases admitted to the in-patient department of Hospital for Women, Patna, from 1942, presents interesting contrast in the type of abnormalities taken notice of till 1948 and between 1949 and 1952.

In the earlier years, cases belonged mostly to the category of minor degrees of developmental anomalies. Eleven cases of imperforate hymen with haematocolpos were operated by classical crucial incision technique. One interesting case of pinpoint hymen with full time pregnancy was ad-

mitted in labour. She was ultimately delivered by incision of the hymen, followed by episiotomy and low forceps extraction.

During this very period, only 4 cases of gross abnormalities are on record. There were three cases of bicornuate uterus, in one of them (uterus biocorporis bicornis) subtotal hysterectomy of one cornu was performed, the other two cases were noted during caesarean section. The fourth case was the most interesting of the lot, a case of vaginal occlusion with haematometra. The haematometra was drained by hysterotomy and the vagina dissected from the uterine end till a communication at the stretched urogenital part was made. She subsequently became pregnant and delivered normally.

A striking difference will be noted in the types of cases managed from 1949 to 1952. Factual data of cases which came under our care during this period have been presented in a tabular form (Table III). Failure to get records of such cases in earlier years can only be accounted for by the adoption of an attitude of indifference and resignation on the part of the clinicians attending to them, because in most of the cases the possibility of conception in future could be ruled out.

It would be convenient to present our cases by discussing them under the heading of different abnormalities.

I. *Vestibular anus*

Two cases of vestibular anus were observed.

The first case (Case No. 3, Table III) was a married lady who came with the following symptoms:

(i) Passage of faeces per vaginam following sexual intercourse after marriage, (ii) primary amenorrhoea. She had vaginal bleeding once following sexual intercourse after marriage.

The abnormalities noted were a short perineum, vestibular anus, absence of vagina except for a distance of half an inch from below, absence of fused portions of Mullerian ducts, the broad ligaments on either side being continuous in midline, and absence of uterine vessels.

Closure of vestibular anus by flap-splitting and reconstruction of vagina by dissection of the rectovesical space and subsequent packing with BIPP was done, with an exploratory laparotomy at the same sitting. McClellan and Williams, however, in a case of congenital recto-perineal fistula with absence of vagina, uterus and fallopian tubes, performed a two stage operation of repair of the fistula and vaginal reconstruction by Wharton technique. This two stage procedure, unless indicated by poor health or other complications, seems avoidable to us from the experience in our case in which we also performed an exploratory laparotomy.

The second case (Case No. 19, Table III) was an unmarried girl who came with the following symptoms:

(i) Severe dysmenorrhoea, menstrual and post-menstrual for three years.

(ii) Lower abdominal swelling, duration two years.

The patient's attendant gave history of operation on anus performed

immediately after her birth to establish patency.

The abnormalities noted were a vestibular anus of small size, a very small cervix difficult to visualize, uterus bicorporis bicornis with chocolate cysts of both ovaries.

The enlarged right horn, with fallopian tube and ovary of that side, was removed after laparotomy. Left ovary was resected. Cut section of the removed uterine horn showed evidences of adenomyosis.

The vestibular anus was not repaired as the patient was not inconvenienced by it and she resented, being an unmarried girl, to get herself operated in that area.

The embryology of cloacal duct was described by Reichel who also cited a case observed by him. The history was similar to our first case (Case No. 3, Table III), but unlike ours there was no other abnormality of genital tract. After discussing the possibility of mechanical injury, he concluded that the abnormal communication between anus and vestibule was present as a slender fistula from birth and that it was dilated following coitus. Such a condition would arise in an embryo of 12 m.m. provided that the perineal tissue encircle the cloacal duct instead of obliterating it. Nevertheless, the probability of rupture of a persistent weakened urorectal membrane in its lower part following coitus could not be ruled out in a case like ours where the anal canal was defined and communicated with the rectum.

II. *Congenital anomalies of vagina*

Twelve cases of congenital anomalies of vagina have been presented in the series.

It was observed that gross abnormality of vagina, viz. absence in its entire length or in a segment, is usually associated with varying degrees of malformations of uterus and its appendages. Only one case of duplication of vagina and uterus was observed which indicated its relative rarity and also suggested that duplication of vagina was probably more commonly associated with duplication of uterus. Normal functioning uterus is perhaps compatible with milder varieties of vaginal anomalies like imperforate hymen. In case No. 2 (Table III) development of uterus was more or less normal, cervical glands were functioning but function of endometrium was in abeyance. In most of the cases tags of hymenal tissue were observed.

Out of the twelve cases, we attempted to reconstruct vagina in eight cases. The method adopted in seven out of eight cases was after McIndoe's technique with certain modifications. The results were extremely gratifying and we have extended it to six cases of acquired vaginal atresia of severe degree.

The dissection of vesico-rectal space is started near urethral meatus and is completed by keeping a rubber catheter in the urethra as a guide. Brisk haemorrhage was not encountered in any of the cases. A priapos shaped mould was prepared from dental stent with a lumen in the centre. The size of the mould was adjusted according to the size of the space dissected, making due allowance for certain amount of contrac-

tion during healing. Due to absence of skilled hands we had to contend with small pieces of Thiersch skin grafts stitched on mould. In some of the cases the whole of the mould could not be covered with grafts. The introitus was narrowed by splitting the epithelio-cutaneous junction of perineum and approximating the raw surface.

We have advised keeping the mould for at least three months. In some cases the mould was spontaneously expelled by that time while in others the mould had to be removed by coaxing it out with a finger in rectum or by a perineotomy incision.

The epithelization achieved, considering the small pieces of grafts utilized, was highly satisfactory. Subsequently we advised these patients to use a silver mould anointed with a bland ointment for three months. Frequent coitus was encouraged during this period.

The vaginae, so constructed, were of adequate length and of good elasticity. They were in no way inferior to the reconstructed vaginae by McIndoe as seen by one of us (S.N.U.). It is interesting to note that even cyclic cellular changes have been observed in reconstructed vaginae (Ayre).

In the eighth case simple dissection of the space followed by gauze packing (Kanter) was done. The space was epithelized but contracture of lumen and unsatisfactory resilience of walls resulted.

We have been able to trace only three cases of pregnancy following reconstruction of vagina in the literature available to us. Its rarity is due to the frequent association of uterine anomaly with gross malformation of vagina as has been pointed out by us. The case reported by Wagner, who had reconstruction of vagina by Baldwin's technique, delivered spontaneously. The cases reported by Whittemore and Baer and DeCosta were delivered by caesarean section. To this may be added our case mentioned earlier. Haematometra was drained per abdomen and vagina was dissected from above by Professor Duncan Murdoch. The patient subsequently became pregnant and one of us conducted the delivery, which was normal (S.N.U.).

The earliest attempts at reconstruction of vagina were made by Dupuytren by simple method of dissection and by Heppner by skin grafts.

Numerous methods have subsequently been advocated, e.g. utilizing rectal transplant (Sneguireff), double (Baldwin), or single loops (Mori, cited by Miller et al,) of ileum, skin flaps from labia (Jewett, Graves) pedicle graft from thigh in single (Beck) or multiple (Frank and Geist) stages, simple dissection of the space and introduction of a vaginal mould (Wharton), free skin grafts on sponge prosthesis (Kirschner and Wagner) and Thiersch skin grafts on vulcanite mould (McIndoe and Banister).

Non-operative method by repeated dilatation has been adopted by Frank in co-operative patients. Reconstruction with amniotic membrane (Marsalck, Snoeck and Rocman) has been attempted with success.

Although our experience is limited and does not extend to all the methods reviewed in literature we are of opinion that technique utilising

Thiersch skin-grafts on vaginal mould is the most effective method of reconstruction of vagina due to its simplicity, safety and its ability to restore the organ to its natural form. It should always be undertaken even when there is no possibility of restoration of function, menstruation or conception. The psychological improvement observed after the marital function is restored is sufficient justification to undertake the operative procedure.

Septate Vagina

We have observed septate vagina in a case of uterus didelphys with primary sterility and oligomenorrhoea. Duplication of hymen was not noted in this case (Anderson). It appears from the literature that this condition has been often missed even in adult life and normal pregnancies and deliveries have also occurred. After the compilation of the article we have come across four cases of double uterus and vagina. None complained of dyspareunia, while all of them had some menstrual irregularity and complained of sterility (primary). In two of them dilatation of cervix and excision of the vaginal septum has been done while endocrine therapy has been advised in the other two.

Hydrocolpos

One case of hydrocolpos was noted in the present series. There were no presenting symptoms, the dull white bulging membrane drew the attention of the parents. Urinary obstruction may, however, be a feature of such cases due to pressure over the urethra. Some interesting cases of hydrocolpos (Maliphant and Sen) are on record in the literature.

III. *Congenital anomalies of uterus*

The following table (Table I) based on material collected from 19 cases in the present series aims at explaining the anomalous nature of association of a normal uterus with abnormalities in development of vagina and other parts of the genital tract, and also, the extent to which a normally developed vagina and other parts of genital tract could be found in association with an abnormal uterus.

To summarise, four groups of cases emerge from the data presented in Table I.

1. Normal uterus associated with abnormalities in vagina and other parts of the genital tract.

Two cases are included in this group, one with normal uterus associated with imperforate hymen, the other with pregnancy in a normal uterus with partial atresia of upper vagina.

2. Abnormal uterus associated with normal vagina and other parts of the genital tract.

Six cases are included in this group:

Uterus bicorporis unicollis with vagina	1
Uterus bicornis with normal vagina	1
Uterus subseptus with normal vagina	2
Rudimentary uterus with normal vagina (with ovarian tumour)	1
Uterus represented by nodule with normal vagina (secondary sex characters ill developed)	1

TABLE I

Uterine abnormality	Vaginal abnormality
1. Absent	Lower 1″ developed
2. Infantile	Lower ½″ developed
3. Cervix absent, portion of Mullerian ducts destined to form body of uterus represented as cords	Lower ½″ developed
4. Cervix absent, uterus represented as a nodule	Lower ½″ developed
5. Uterus bicorporis unicollis	No abnormality
6. Uterus subseptus	No abnormality
7. Cervix absent, uterus represented as a nodule	Lower ½″ developed
8. Absent	Lower ¼″ developed
9. Uterus duplex	Vagina duplex
10. No abnormality	Imperforate hymen (haematocolpos)
11. Rudimentary uterus	No abnormality
12. Absent	Lower ½″ developed
13. Uterus represented as nodule	No abnormality
14. Not noted	Imperforate hymen (Hydrocolpos)
15. Uterus subseptus	No abnormality
16. Normal	Partial atresia of upper ¾″ vagina
17. Uterus bicornis	No abnormality
18. Absent	Lower ½″ developed
19. Uterus bicorporis unicollis	No abnormality

3. Abnormal uterus associated with abnormality in vagina and other parts of the genital tract.

Ten cases have been included in this group:

Absence of uterus and vagina (except lower ½″—1″) ..	5
Uterus represented as nodule with absence of vagina (except lower ½″—1″)	2
Infantile uterus with absence of vagina (except lower ½″) ..	1
Uterus duplex with vagina duplex	1
Uterus bicorporis unicollis with vestibular anus	1

4. Imperforate hymen with hydrocolpos, state of vagina and uterus not known.

Abnormality of vagina is usually associated with abnormality of uterus varying from its absence to its being of infantile type. One case has not followed the rule in this series, the case with partial atresia of upper vagina with normal uterine pregnancy.

When uterine abnormality is associated with normal vaginal development, it is more often the result of failure of expansion of the horizontal part of the Mullerian ducts. In

the majority of instances this is the rule, though in a few cases arrested development of uterus as a whole may be encountered. In the two cases observed in the latter group uterine development was poor. In one (Case No. 11, Table III) the rudimentary uterus failed to respond to heavy dosage of oestrogen and anterior pituitary-like hormone after ovariotomy. The ovary of the other side was well developed and functioning. In the second case (Case No. 13, Table III) the uterus was represented by a nodule, whereas secondary sex characters were poorly developed. General body growth was satisfactory so that ovarian aplasia with failure of development of Mullerian ducts could be reasonably speculated.

Congenitally malformed uterus can be the seat of pathological state, like any other normal uterus. We have come across one case (Case No. 19 Table III) where one horn of a uterus bicornis bicorporis was the seat of adenomyosis. Both ovaries showed evidence of endometriosis. Endometriosis with uterus didelphys (McDonald) has been reported in the literature.

Two cases of uterus bicorporis were observed in the present series (Cases No. 5, 19, Table III). One horn was removed in each case. The guiding factor in removal of a horn differed. In case No. 5 (Table III) the left horn was supplied with a thin attenuated uterine artery. The development of both horns was, however, uniform. The left horn was removed along with the rectovesical ligament. In case No. 19 (Table III) the right horn was the seat of adenomyosis and was removed. Both uterine vessels were of equal size.

Plastic repair of uterus by joining the two uteri has been recommended (MacArthur, Campbell cited by Curtis and Moore) and report of conception taking place after such treatment has been recorded (Eisaman; Rheman). Berkeley, Bonney and McLeod, however, reported their experience in several cases of this group where conception failed to occur after plastic repair of uteri. Opinions in the two extremes of noninterference (Smith; Sweet; Gransberry and Faust) or sterilization (Snyder) have been expressed. We have put forward our arguments in favour of hemihysterectomy elsewhere (Upadhyay and Mitra). Subsequent search of literature revealed reports of three cases where hemihysterectomy was followed by conception and delivery of a live child.

The obstetric history of Acosta-Sison's case was as follows:

1st pregnancy—marginal placenta praevia;
2nd pregnancy—obstructed labour with threatened rupture of uterus—Porro's caesarean section was performed;
3rd pregnancy—spontaneous full-time normal delivery.

Smith's case was one of uterus bicornis unicollis with haematometra in the left horn. The haematometra was wrongly diagnosed as rectal abcess and incised per rectum. Subsequently hemihysterectomy alone was performed on the left side in view of the poor health of the patient and presence of severe infection. The patient later conceived and was delivered of a live child by caesarean sec-

tion. The scar on the left side was much thinned out.

Cowle's case was one of pregnancy following caesarean section and subtotal hysterectomy in an unrecognized uterus duplex.

In no case the hemihysterectomy was planned; nevertheless they are instructive and support the stand we have taken in this group of cases.

Congenital malformation of uterus in obstetric practice:—

Five cases, presented in the series, were of obstetric interest.

Partial atresia of upper vagina with normal uterine pregnancy (Case No. 16, Table III) was noted in one case who was subsequently delivered by caesarean section.

One case with uterus bicorporis and rectovesical ligament (Case No. 5, Table III) gave history of three consecutive breech deliveries with intranatal foetal death every time. The horn with attenuated uterine vessel was removed.

Another case (Case No. 17, Table III) gave history of breech delivery with intranatal foetal death in her first confinement. She consulted us in her second pregnancy. It was a breech presentation again, and repeated attempts at external cephalic version failed. Skiagram of abdomen showed breech presentation with extended legs. She was delivered by lower segment caesarean section of a live female child. It was a uterus bicornis.

There were two cases of uterus subseptus. One (Case No. 15, Table III) gave history of three repeated full time pregnancies with hand prolapse delivered by decapitation in a district hospital. She consulted us in her fourth pregnancy at the thirty-eighth week. The lie was transverse. Classical caesarean section was done and a live female child was delivered. Incomplete uterine septum was observed during operation.

The other case (Case No. 6, Table III) gave history of four full-time normal deliveries and two abortions. All the previous labours were prolonged and conducted in hospital. She consulted us in her seventh pregnancy during labour. She delivered normally; the placenta was retained and had to be removed manually. During manual removal an incomplete uterine septum reaching fairly low to the level of internal os was observed. A constriction ring was formed and placenta was retained in one of the horns.

After the article was compiled we performed lower segment caesarean section in an elderly primigravida, who conceived after fifteen years of marital life. The lie was transverse and repeated attempts at external cephalic version failed. It was a uterus subseptus.

Abnormal presentation is very common in malformed uteri (Way). Prolongation of labour partly as the result of malpresentation and partly due to irregular uterine action is apt to occur. Placenta may be retained (Way) and may need manual removal. We failed to note high incidence of abortion, premature labour, placenta praevia (Way) and postpartum haemorrhage in the few cases encountered.

Fenton and Singh and Way have

recently reviewed the obstetric abnormalities associated with malformed uteri. The following table reveals the differences with our cases.

TABLE II

	Transverse Lie	Breech	Prolapsed cord	Abortion	Premature labour	Retained placenta	Placenta praevia
Incidence in thirty-two pregnancies in twelve patients (Way) ..	12	4	2	12	3	3	2
Incidence in seventeen pregnancies in five patients (Authors)	4	5	—	2	—	1	—

IV. *Congenital abnormality of Fallopian Tubes and Ovaries*

One case of congenital abnormality of fallopian tubes was noted in the series (Case No. 1, Table III). Mullerian ducts on both sides except at the cephalic end were suppressed. Only the fimbriated extremities of the fallopian tubes were present. Gross abnormality of ovarian development is a rare clinical entity and we can speculate aplasia of ovaries in one of our cases only (Case No. 13, Table III). It could not, however, be confirmed by laparotomy.

Due to restriction in availability of skiagram films we were not able to submit any of our cases to a routine intravenous pyelography, in order to exclude renal abnormalities, but in nine cases where abdomen was opened we failed to note any gross abnormality in the urinary tract.

Discussion

A careful study of the cases presented in previous pages reveals most strikingly a "selective" arrest in the development of a primitive structure like the Mullerian duct. An abnormality involving the entire length of the ducts could probably be explained on theories current in literature, but a case with normal vagina in association with a grossly malformed or absent uterus is paradoxical, particularly when four-fifths of the vagina is admitted to be of Mullerian origin. It is this type which predominates in daily practice. Our observations on absent or attenuated uterine vessels in a few cases is particularly interesting. We feel justified in stating that the disturbances in blood supply from non-development of pelvic vessels, particularly the uterine vessels, does at least represent a suppression of mesenchymal element related to Mullerian ducts in its different segments. Relative avascularity of rectovesical space during dissection for reconstruction of vagina where none existed lends further support to our contention. However, before we proceed further in justification of our impressions let us consider some popular theories.

TABLE III

Case No.	Year.	Age.	Presenting symptom.	General examination and secondary sex characters.
1.	1949	17 years 6 months.	Primary amenorrhoea and sterility.	Ht—5 ft. 2 in. Wt—112 lbs. Secondary sex characteristics developed.
2.	1949	20 years.	Primary amenorrhoea and dyspareunia	Ht—4 ft. 5 in. Wt—90 lbs. Secondary sex characteristics developed.
3.	1950	16 years 3 months.	Passage of faeces per vaginam and per anus since her marriage ten months ago. After her marriage and following sexual intercourse she had vaginal bleeding—menstruation as she said for the first time. The bleeding lasted for 15 days and then she started having the present symptoms.	Ht—4 ft. 8 in. Wt—102 lbs. Secondary sex characteristics well developed.
4.	1950	17 years.	Primary amenorrhoea, dysmenorrhoea and sterility. An unsuccessful attempt at operation a week before admission.	Ht—4 ft. 6 in. Wt—98 lbs. Secondary sex characteristics well developed.
5.	1950	27 years.	3rd para: all full time breech deliveries—with loss of child in each case.	Ht—4 ft. 7½ in. Wt—114 lbs. W.R.—Negative (Serum) Secondary sex characteristics well developed.

Clinical findings.	Treatment.	Developmental defects
Occlusio vaginae—lower ¼″ developed. Hymenal tags noted. Uterus could not be palpated per rectum.	Dissection and skin grafting on mould after McIndoe. Laparotomy.	Fimbriated extremities of fallopian tubes developed. Rest of the fallopian tubes, uterus and upper ¾th of vagina absent and replaced by a transverse fold of peritoneum (plica transversalis) in coronal plane of pelvis. In the fold at the proximal ends of ligaments small nubbin (1.5 cm. x .5 cm) on either side seen to which ovarian ligaments were attached. Uterine vessels could not be traced. Ovaries and their vessels well developed.
Occlusio vaginae—lower ½″ developed. Hymenal tags noted. Uterus anteverted, anteflexed, infantile (per rectum).	Dissection and skin grafting on mould after McIndoe.	Uterus—infantile, size of uterine cavity—2″. Thin drops of mucus noted when cervix was approached.
Short perineum, anal canal gaping indicating absence of sphincteric arrangement. Small anovestibular fistula. Hymenal tags noted. Occlusio vanae—lower ½″ of vagina developed. Uterus could not be palpated (per rectum).	Anovestibular fistula repaired by flap splitting technique. Dissection and packing the canal by gauze soaked with B.I.P.P. Laparotomy.	Multiple cystic structures resembling hydatids of Morgagni attached to both tubes. The part represented by fused portion of Mullerian ducts absent and replaced by fold of peritoneum. Uterine vessels could not be traced. Ovaries and their vessels well developed.
Occlusio vaginae—lower ½″ of vagina developed. Hymenal tags noted. Uterus could not be palpated —small nodule in middle felt per rectum.	Dissection and skin grafting on mould after McIndoe. Larotomy.	Fallopian tubes well developed. An unsuccessful attempt at development of fused portion of Mullerian ducts noted—a small nobbin instead of uterus present. Uterine vessels absent. Ovaries and their vessels developed.
Uterus bicorporis unicollis. Vagina well developed.	Laparotomy. Uterus bicorporis unicollis with rectovesical ligament. Left cornu (with attenuated uterine vessel) removed — hemihysterectomy.	Uterus bicorporis unicollis with rectovesical ligament. Left cornu with attenuated uterine vessel.

TABLE III—(Contd.)

Case No.	Year.	Age.	Presenting symptom.	General examination and secondary sex characters.
6.	1950	28 yrs.	7th Gravida, 2 abortions. 4 labours prolonged and conducted in hospital. The present pregnancy full time, normal delivery. Retained placenta—P.P.H.—manual removal of placenta.	Average build Secondary sex characteristics well developed.
7.	1951	18 yrs. 2 months.	Primary amenorrhoea ? Cyclic pain.	Ht—4 ft. 8 in. Wt—105 lbs. Secondary sex characteristics well developed.
8.	1951	17 yrs. 3 mths.	Primary amenorrhoea. Primary sterility? Cyclic pain.	Ht—4 ft. 10 in. Wt—112 lbs. Secondary sex characteristics well developed.
9.	1951	19 yrs. 7 mths.	Oligomenorrhoea and primary sterility.	Ht—5 ft. 0 in. Wt—110 lbs. Secondary sex characteristics well developed.
10.	1951	16 yrs. 1 mth.	Cyclic pain—2 years.	Ht—4 ft. 7 in. Wt—100 lbs. Secondary sex characteristics well developed.
11.	1951	17 yrs. 2 mths.	Primary amenorrhoea. Pain and lump abdomen.	Ht—4 ft. 8 in. Wt—100 lbs. Secondary sex characteristics well developed.

Clinical findings.	Treatment.	Developmental defects
Full time normal pregnancy. Position—L.O.A.	Manual removal of placenta. Placenta separated but retained by a constriction ring in right cornu.	Uterus bicorporis unicollis.
Occlusio vaginae—lower ¾″ of vagina developed. Hymenal tags noted. Uterus could not be palpated. Small nodule in midline felt per rectum.	Dissection and skin grafting on mould after McIndoe. Laparotomy not done.	The anomaly, in all probability, was similar to S.N. 4.
Occlusio vaginae—lower ¼″ of vagina developed. Hymenal tags present. Uterus could not be palpated.	Dissection and skin grafting on mould after McIndoe. Laparotomy not done.	The anomaly is more severe in deggree than S.N. 4 and 7.
Uterus didelphys. The two uteri were smaller than normal in size. Hymenal duplication not noted.	Patient absconded.	
Imperforate hymen with typical bulging of the membrane. Suprapubic lump.	Hymen incised and drained. Silver mould to introduce in vagina for 2 months. Advised to come after 4 months for insufflation.	Haematocolpos and Haematometra. Imperforate hymen.
Small rudimentary uterus. Ovarian tumour.	Laparotomy: ovariotomy right side. Ovarian cystectomy left side. Salipingostomy left side. Exhibition of heavy dosage of oestrogen upto 6 months after operation did not succeed in initiating uterine bleeding.	Rudimentary uterus perhaps with aplasia of endometrial element. Congenital atresia of fallopian tube of left side. Ovarian tumour—serous cystadenoma.

TABLE III—(Contd.)

Case No.	Year.	Age.	Presenting symptom.	General examination and secondary sex characters.
12.	1951	20 yrs.	Primary amenorrhoea.	Ht—5 ft. 0 in. Wt—106 lbs. Secondary sex characteristics well developed.
13.	1951	19 yrs. 3 mths.	Primary amenorrhoea.	Ht—4 ft. 9 in. Wt—103 lbs. Sparse axillary hairs. Breasts not developed.
14.	1951	5 mths.	Bulging growth between labia minora.	Wt—12 lbs.
15.	1951	30 yrs.	4th Gravida. Previous 3 deliveries—Hand prolapse, treated with decapitation in district hospital.	Average build. Secondary sex characteristics well developed.
16.	1951	32 yrs.	O para. Amenorrhoea 20 weeks. Irregular menstruation.	Average build. Secondary sex characteristics well developed.
17.	1951	26 yrs.	2nd Gravida. 1st pregnancy—breech with foetal death intranatally.	Average build. Secondary sex characteristics well developed.
18.	1952	18 yrs.	Primary amenorrheoa. Primary sterility.	Average build. Secondary sex characteristics well developed.
19.	1952	25 yrs.	Primary sterility. Dysmenorrhoea.	Average build. Secondary sex characteristics well developed.

Clinical findings.	Treatment.	Developmental defects
Occlusio vaginae—lower ½" of vagina developed. Hymenal tags noted. Uterus could not be palpated.	Dissection and skin grafts on mould after McIndoe. Laparotomy not done.	The abnormality was like S.N. 8.
Hymen well developed and perforated. External genitalia ill - developed with sparse pubic hair. Vagina well developed and of average length. Uterus and cervix felt as a nodule of pea-size. External os could not be identified.	Ovarian hormones advised. Exploratory laparotomy contemplated but refused by the patient.	Involves the fallopian tubes and uterus. Probably ovarian development was also impaired.
White bulging membrane in vestibule. Hymen could not be identified. The bulge felt cystic.	Advised operative treatment. Patient has not turned up as yet.	Clinical diagnosis—Hydrocolpos.
Transverse lie. F.H.S. audible. External version was not attempted as patient came at 37th week.	Classical caesarean section. Live female child 6 lbs. 4 oz.	Uterus sub-septus.
Pregnancy 20 weeks. F.H.S. audible. Vaginal atresia in upper half of vagina.	Delivered by caesarean section at term.	(?) Congenital atresia of upper half of vagina.
Pregnancy—Breech presentation—Repeated attempts at external version failed.	Lower segment caesarean section. Live female child—5 lbs. ½ oz.	Uterus bicornis.
Vagina developed in lower fourth. Uterlus absent.	Dissection and skin grafts on mould after McIndoe.	Absence of uterus and vagina.
Uterus bicorporis bicornis unicollis. Vestibular anus.	Laparotomy. Removal of one horn which was seat of adenomyosis. Resection of ovaries —seat of endometriosis.	Uterus bicorporis unicollis vestibular anus.

Theory of hormonal failure in intra-uterine life (Falls).

This is perhaps untenable in view of the huge production of all types of sex hormones from chorionic elements, besides in none of the cases observed by us or reported in the literature, history of disturbances in pregnancy, in which the subject was born, could be noted. We are unable to explain the "selective failure" of development on this theory.

Theory of differential rate of growth of Mullerian ducts (Anderson)

The factors determining the uneven growth could probably be explained only on differential blood supply.

Theory of abnormal fusion of Mullerian ducts

Fusion takes place in 22-28 m.m. embryo. We shall revert to a discussion of this subject again.

Shattenberg and Ziskind attempted to establish chronological development of different uterine anomalies, viz. unilateral or bilateral failure in first month, uterus didelphys in second month, uterus bicornis or uterus arcuatus in third or fourth month. Unilateral or bilateral failure of Mullerian ducts occurring in first month is usually associated with gross malformation of other structures developing in meso-nephric fold, viz., urinary system and reproductive gland analage.

Comparative anatomists recognised similarity in uterine anomalies in women to normal uteri in animals lower in developmental scale. The conformity appeared so great that Blair Bell suggested the term "atavism" in preference to malformation. Reference may be made to the review of the subject made by Jarcho.

It has been difficult for us to reconcile the current theories and postulates with the facts observed by us and we feel a time for reorientation has come. It is realised that dogmatic assertions are not possible yet our inferences regarding the aetiological factors are worth a thought.

Maldevelopment of uterus falls into two broad groups. Firstly, errors in development of one or both Mullerian ducts and secondly, errors in fusion of the ducts.

Errors in development of one or both Mullerian ducts

Eight cases have been presented. The defective development is attributable either to:

(1) Complete or partial non-development of one or both ducts, or to

(2) Early development followed by regression as a result of adversely acting circumstances. We believe regression following early development to be more important a factor than non-development in part or whole of one or both ducts.

What then brings about the change?

It appears to us on close analysis of our cases that the development of female genital tract from the primitive Mullerian ducts to its adult form is accomplished through three distinct stages.

1. In the earliest stage of development when no vascular connections have been established, the

nourishment of the developing Mullerian ducts must necessarily be governed by factors determining the growth of the embryo in general. During this stage following changes may be noted.

Appearance of Mullerian ducts at 10 m.m. the approximation of their caudal thirds at 19.4 m.m. and their fusion at 22-28.5 m.m. to form the uterovaginal canal.

2. The second stage of development of Mullerian ducts is associated with development of the vessels related to them. Development of pelvic vessels is heralded by the appearance of the umbilical arteries at 7 m.m. from caudal portion of aorta. Subsequently the axial artery of the lower limb—a. ischiadica develops from each umbilical artery. Proximal to a. ischiadica a. femorales develop which in later life forms the external iliac and femoral arteries. The segment of umbilical artery from the origin of a. femorales to a. ischiadica may be taken as the anterior division of hypogastric artery which divides into two terminal branches—a. internal pudenda and a. inf. gluteales—in 15.5 m.m. embryo. From the literature available to us we could not trace further development of the pelvic vessels particularly those supplying the Mullerian ducts. However, it can well be conceived that differentiation of the embryo to female sex and consequent development of the Mullerian ducts and suppression of the Wolffian ducts, determine the disappearance of the inferior vesical artery and development of the uterine and vaginal arteries. The inferior vesical artery, in male (absent in human female), supplies the structures originating from the Wolffian ducts, viz. lower ureter, base of bladder, prostatic urethra, seminal vesicles, prostate, ejaculatory ducts and vas deferens. With retrogression of the Wolffian ducts the inferior vesical arteries either undergo repression or are diverted to adjacent Mullerian ducts and transformed to uterine and vaginal arteries. It is reasonable to deduce that any anomaly in vascular supply of Mullerian ducts and consequent anomaly of the ducts themselves, will take place later than 15.5 m.m. embryo stage but not later than 56 m.m. embryo stage when the Wolffian ducts start involuting in female.

It is clear from what has been said above that at a certain stage Mullerian ducts acquire definite segmental vascular supply, viz. cephalic vertical portion from ovarian artery, caudal vertical portion from hypogastric artery and middle horizontal portions from both, and it is probable that further development of the ducts depends, at least partly, if not entirely, on the efficiency of these vascular channels. For example, failure of development of fimbriated extremity of fallopian tube has not been observed in this series. This is probably due to its blood supply being derived from the ovarian blood vessels whose origin and development is on a firmer footing than those supplying Mullerian ducts elsewhere. Careful scrutiny of the Case No. 5 (Table III) revealed that the normal development of one half of uterus was possible with a thin attenuated uterine artery not anastomosing with ovarian artery of that side, if an additional source of blood supply could

be established via intercervical anastomosis.

This case also reveals that during the stage of fusion of Mullerian ducts, development of their vascular channels is initiated. If the mesenchymal elements fail on both sides to develop and form vascular channels of adequate calibre, arrest of further development of Mullerian ducts and their regression may take place. On the contrary if fusion and intercervical anastomosis of blood vessels take place, development of one uterine artery is sufficient for the growth of the two ducts.

It remains to be worked out whether it is the primary failure of the Mullerian ducts to stimulate specific response of connective tissue round them or it is the primary failure of mesenchymal element, which leads to non-development of vessels of the Mullerian ducts.

We cannot but over-emphasize the development of the Mullerian ducts in segments when their segmental blood supply is established. After this stage is reached maldevelopment of the duct in its entire length will be well nigh improbable; maldevelopment of a segment of the duct will be more common depending on the anomaly of the vascular apparatus of the corresponding segment. In our present knowledge this hypothesis alone can explain all sorts of paradoxical maldevelopment of the Mullerian ducts observed in clinical practice.

3. The final shaping of the Mullerian ducts in the formation of a normal human female uterus and vagina depends on the play of hormones on these structures. This process starts in intra-uterine life; occasional cases of vaginal bleeding in new-born human female testifies the responsiveness of the Mullerian structures to ovarian hormones in intra-uterine life. However the stage is completed after puberty when with the onset of cyclical uterine bleeding and better endocrinal influences the prepubertal uterus is changed to the adult form. The finer details of the development is not so simple as described above and it is not uncommon to note "dissociated effects" of endocrine system on the different parts of Mullerian ducts, viz. endometrium, endocervix, and vagina although they have the same origin.

II. *Errors in fusion of the Mullerian ducts*

The fused portions of the Mullerian ducts, utero-vaginal canal, give rise to cervix and vagina. The body of the uterus develops from the horizontal portion of the Mullerian ducts by the expansion of their walls (Keibel and Mall).

Forces which prevent the Mullerian ducts from fusing by pulling them apart, will cause uterus didelphys, while forces which appose the ducts too firmly and prevent the expansion of the walls in cranial direction will cause various anomalies ranging from uterus bicorporis bicornis to uterus arcuatus (Keibel and Mall).

Aegenesis of reproductive gland *analage* rarely takes place because its blood supply is ensured at an early stage from the mesonephric arteries. Absence of fallopian tubes is infrequent in comparison to the absence

of uterus or vagina, because of its blood supply being derived from the ovarian artery.

The reproductive gland thus forms an independent functional unit from intrauterine life. Development of secondary sex characters and functioning corpora lutea were frequently observed in association with absence of uterus and vagina and it supports the contention that presence of endometrium is not essential for proper functioning of ovary. When ovarian failure occurs after hysterectomy it is usually due to thrombosis of the ovarian vessels which ultimately performs a physiological castration, rather than due to absence of endometrium causing local hormonal imbalance, as is erroneously believed.

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