CONGENITAL GYNATRESIA

A clinical report on nine cases

by

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In the development of the vagina, the müllerian ducts and the uro-genital sinus take part. The lower ends of the fused müllerian ducts lie in close association with the posterior part of the uro-genital sinus. Here they give rise to the müllerian tubercles. Where the müllerian tubercles invaginate the uro-genital sinus, the sinovaginal bulbs are formed. The upper $\frac{3}{4}$th of the vagina is formed from the müllerian tubercles by its progressive growth and canalisation. The lower $\frac{1}{4}$th is formed from the sinovaginal bulbs. The failure of the distal parts of the müllerian ducts to develop or to canalize, seems to be rather rare. Hence a report on nine personal cases may be of interest.

Case 1

L., aged 15 years, was admitted on 3-11-1956 with a history of primary amenorrhoea, periodical lower abdominal pain of two years' duration and the presence of a mass in the lower abdomen for six months. She had no urinary complaints. She was unmarried.

Physical findings:- Patient was well developed. Secondary sex characters and external genitalia were normal. Vagina was $\frac{3}{4}$th of an inch deep and ended blindly.

Rectal examination revealed a firm, rounded, mobile and tender mass arising from the pelvis in the midline and rising to an inch above the pubic symphysis.

Intra-venous pyelography showed no abnormality in the urinary tract.

Treatment

Drainage of the haematometra was done from blew the next day after admission. The space between the bladder and the rectum was opened into through the vault of the shallow vagina. Blunt dissection in this space for about two inches brought into reach the wall of the distended uterus and an opening was made in the most dependent part. After the thick black blood was drained away, the edges of the opening were stitched to the upper part of the vaginal epithelium to avoid closure of the opening. This opening could admit the index finger.

This patient left the hospital against medical advice two weeks after operation and did not come again for check up.

Case 2

S., aged 17 years, was admitted on 21-11-1956 with a history of primary amenorrhoea, severe pain in abdomen for three days and a previous history of severe pain at monthly intervals for $3\frac{1}{2}$ years. She was married and complained of dyspareunia. There was no urinary complaint.

Physical findings:- She was well developed with normal secondary sex characters and external genitalia. Vagina was only $\frac{3}{4}$th of an inch deep and ended blindly.

Per abdomen two masses were visible and palpable. One arose from the pelvis (as confirmed by rectal examination) and extended to an inch above the symphysis pubis. Attached to this towards the right

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Received for publication on 18-6-68.

side was another mobile mass which was the size of a big orange.

Intravenous pyelography showed no abnormality.

Treatment

The abdomen was opened because the upper mass was mistaken for an ovarian cyst. However, it was found that the midline tumour was the dilated cervical canal filled with blood and the tumour above was the haematometra. Abdomen was closed without proceeding any further. The haemato-cervix and-metra were then evacuated from below by dissecting the space between the bladder and rectum, through the vault of the short vagina. In this case a dimple could be identified marking the external os and an opening was made through this by thrusting the knife point, After the evacuation of collected blood, by introducing the index finger through the opening, the dilated cervical canal and internal os could be identified. Six weeks later (after a normal menstrual period) an artificial vagina was created by turning in the skin flaps from the labia minora. But the vagina gradually contracted down to about 2 inches depth. This patient was kept in hospital for 4 months and had three normal periods. She did not come for further check up.

Case 3

M., aged 25 years, was admitted on 25-10-1960 with a history of pain in lower abdomen off and on for ten years and a tumour in the lower abdomen for seven years. She was a child widow and so gave no history of dyspareunia. She had not menstruated at any time.

Physical findings:- She was well nourished with well developed secondary sex characters. The thumb of the left hand was rudimentary and was arising from the index finger.

Vagina was ¾th inch deep and was split into two equal parts by a longitudinal septum. Vagina ended blindly and no cervix was felt.

Rectal examination and abdominal examination revealed a mass arising from the pelvis and extending to the right iliac fossa, globular, fixed and of the size of 20 weeks pregnancy.

Intravenous pyelography was done and it showed no abnormality in the urinary tract.

Treatment

Abdominal route was chosen because the long history of ten years made the diagnosis difficult. On opening the abdomen, numerous vascular adhesions between the pelvic organs, omentum and intestines were found. The uterus was about the size of 18 weeks pregnancy. The left tube was greatly enlarged and densely adherent to the uterus and the pelvic wall. It rose to an inch above the fundus of the uterus. The left ovary was very big and was adherent to the pelvic colon and the fallopian tube. On the right side there was an ill-developed uterine horn, 1½″ x ¾″ x ½″ in size. A normal ovary and a thin fallopian tube were attached to this horn. The left tube, left ovary and the uterus with the rudimentary horn were removed. No cervix could be identified either macroscopically or microscopically in the specimen. The tube and ovary showed evidence of endometriosis.

Case 4

J., aged 24 years, was admitted on 5-1-1960 in obstructed labour. When she was 14 years old, she started getting periodic pain in lower abdomen. After 8 months, she had a vaginal operation and old blood was drained away. She was amenorrhoeic before the operation, but started regular monthly periods after the operation.

Vaginal examination showed normal external genitalia. Urethral orifice was grossly dilated. Vagina was one inch deep. No cervix or os could be identified.

Per abdomen, uterus was at term and was tonically contracted. The foetal head was engaged in the pelvis.

Treatment

A lower segment caesarean section was done and a live child was delivered. Soon after the operation it was found that no lochia was coming into the vagina and an attempt was made to locate the os. An opening admitting a thin probe was found. This was gradually dilated up to No. 10 Hegar and lochia began to flow freely into the vagina.

Case 5

N., aged 15 years, was admitted on 7-11-1962 complaining of irregular pain in the lower abdomen for one year and swelling in the lower abdomen for 8 months. She had not started menstruation. She was married for two years, but had no sexual relations.

Physical findings:- Patient had normal secondary sex characters and external genitalia. Vagina was ¾th of an inch deep and ended blindly. Per rectum, a firm, round, tender mass was felt arising from the pelvis and reaching to an inch above symphysis pubis.

Treatment

Next day after admission, the haematometra was evacuated from below. The dilated cervical canal and internal os could be felt by putting the index finger through the opening made in the cervix. After the thick blood was drained away, the edges of the artificial opening were stitched to the upper part of the vaginal epithelium. However, the cervical opening tended to close down in this patient and had to be dilated off and on. She stayed for 2 months but had no menstruation during this time. Mucoid material used to drain through the vagina for few days after each dilatation. She was very young and probably had not started regular periods. She could not be persuaded to stay longer or to come for check up.

Case 6

D., aged 18 years, was admitted on 27-12-1962 for intermittant attacks of pain in lower abdomen for eight months and a mass for two months. She complained of dysuria and difficulty in defaecation for two months and also of dyspareunia.

Physical findings:- She had well developed secondary sex characters and external genitalia. Vagina was only 1″ deep and ended blindly. Cervix was not seen per abdomen, two masses were visible and palpable. One arose from the pelvis and extended to just above the pubic symphysis. Attached to this there was another mass on the right side. This was mobile and about 4″ x 3″ in size.

Treatment

Because of the experience in case No. 2, the two masses were diagnosed as haematocervix and haematometra. So, abdomen was not opened. By dissecting in the space between the bladder and rectum through the vault of the short vagina, a small dimple could be identified marking the external os and an opening was made through this. By introducing the index finger through this opening the greatly dilated cervical canal and the constriction above this (internal os) could be identified. After drainage of the pent up old blood, the edges of the external os were stitched to the vault of the short vagina. The patient had 2 monthly periods in the hospital but did not come for further check up.

Case 7

L., aged 16 years, was admitted on 31-7-1963 with history of primary amenorrhoea, periodic pain in lower abdomen for two years, constipation and dysuria for ten days.

Abdominal examination showed a firm mass rising to a finger's breadth above the symphysis pubis. External genitalia looked normal. Vaginal and anal orifices appeared normal. Digital examination, however, showed the lower rectum, anal canal and vagina to be one. Except for the skin and subcutaneous tissues of the perineum, there was nothing else to separate the vagina from the anal canal and the rectum. The vaginal epithelium was visible for about 2 inches from the introitus anteriorly, and at the sides it merged with the rectal and anal mucosa.

Treatment

The haematometra was drained from below as in the previous cases. An attempt was made at the same time to separate the vaginal and ano-rectal canals as in the case of a complete perineal tear. But, because of the lack of tissues of the recto-vaginal septum especially the muscle layer, this attempt failed. After a fortnight, the opening made in the cervix closed up and the uterine cavity started to enlarge again and the patient was febrile. A further attempt to evacuate the uterus from below failed. So, a laparotomy was done and the

uterus and the cervix with some infected material inside were removed. Due to the infection from the ano-rectal canal, the procedure performed in the previous cases could not be adopted in this particular case. The patient made an uneventful recovery after the hysterectomy.

Although no cervical canal could be felt by the finger in the primary operative procedure, during hysterectomy it was seen that she had a well developed cervix and the opening made from below was in the place of the external os.

Case 8

G., aged 20 years, was admitted on 30-8-1963 for pain in lower abdomen for three years and a mass in the same region for two years. She was married for five years, but her husband had left her after two years.

Patient was well developed with normal external genitalia and secondary sex characters. Vagina was only ¾th of an inch deep and ended blindly.

Abdominal examination showed a firm, rounded, mobile mass in the hypogastrium rising to 4 fingers breadth above the symphysis pubis. Rectal examination confirmed that this mass was arising from the pelvis. The anal orifice was very unhealthy probably due to anal coitus.

Treatment

The day after admission, the haematometra was drained from below and the edges of the opening were stitched to the vault of the short vagina. Cervical canal could not be made out by the exploring finger.

This patient stayed in the hospital for 3 months and had one scanty period. She was discharged with a patent opening from the short vagina into the uterus. She did not turn up for further check up.

Case 9

G., aged 15 years, was admitted on 3-3-1964 with pain in lower abdomen for 1½ months, dysuria and vomiting off and on. She was unmarried and had never menstruated.

Physical examination showed normal development and normal secondary sex characters. External genitalia were normal. Vagina was short and ended blindly about ¾th of an inch from the introitus. There was a firm, rounded, mobile, tender mass arising from the pelvis in the midline and rising to an inch above the pubic symphysis.

Treatment

Haematometra was drained from below as described in the previous cases. The cervical canal could be identified by inserting the finger through the artificial opening.

She stayed in the hospital for 7 weeks and had one menstrual period. On discharge she had a patent opening through the vault of the short vagina through which mucoid secretion was seen.

Discussion

The failure of the distal parts of the müllerian ducts to develop or to canalize, seems to be rather rare. Case 3 had in addition, an ill-developed and non-functioning uterine horn. All these patients came from villages 30 to 60 miles away from the hospital. They belonged to the low socio-economic group and were illiterate. All of them had primary amenorrhoea though the immediate reason for seeking medical aid was lower abdominal pain.

All these patients had the lower ¼th of the vagina. The symptamatology of this condition depends on the presence of a well-developed and functioning uterus. If the uterus is not present or if rudimentary, the only symptoms will be those of amenorrhoea and dyspareunia. When a normal uterus is present, the symptoms of concealed menstruation take predominance. Since there is no haematocolpos, urinary symptoms are not present except in a minority. An intravenous pyelography is a useful investigation in these cases. It

was done in cases 1, 2 and 3 and there was no abnormality.

A search through the literature shows that whereas a number of cases have been reported in which all the tissues of müllerian origin were absent and different techniques to create an artificial vagina have been discussed again and again, reports involving absence of vagina and/or in the presence of a functioning uterus, have been sparse. Miller and Stout described 71 patients with congenital absence of vagina and of these, in 25% the uterus was present 'invariably' in rudimentary form. Only one patient in these series had a functioning uterus leading to haematometra and in her the vagina was partially present. This patient was treated by the non-graft technique of channel dissection only. She had satisfactory coitus and was the only one in the whole series to become pregnant.

Baer and Decosta state that more or less complete absence of vagina in the presence of a functioning uterus is not unusual. They reported a case similar in all respects to the present series and the treatment was also similar, namely, dissecting a channel between the bladder and the rectum, incising the lower pole of the haematometra and anchoring the edges of this incision to the shallow vaginal pouch. This patient conceived after nine years and was delivered at term by caesarean section. (compare case 4).

Solomons reported a similar case diagnosed as congenital atresia at the level of the external os with absence of all müllerian tissue below this level. The treatment was different in that the dissection between the rectum and the bladder was done 24 days prior to draining the haematometra which was done per abdomen by opening into the cervical canal and inserting a rubber tube through it into the previously prepared channel. Twenty-six days later, skin grafting was done as in McIndoe technique. This artificial vagina became very narrow, but the patient got married, became pregnant next year and was delivered by caesarean section of a term infant.

Carpenter reported a similar case of a 14 year old girl who did not even have the lower $\frac{1}{4}$th of the vagina. Treatment consisted of creating an artificial vagina using excess tissue from the labia minora and insertion from the uterus (after laparotomy) into the vagina of a polythene tube to drain the haematometra. The vagina contracted down to a length of 2 inches and there was scarring of the uterine outlet for which reinsertion of polythene tube was done $2\frac{1}{2}$ years after the first operation.

In describing a new technique for constructing an artificial vagina, Shears mentions two cases which appear similar to the ones described in this paper. One of them had two vaginal deliveries and the other three caesarean sections.

In the case described by Williams the findings would seem similar except non-canalization of the cervix and the absence of menstruating endometrium. Cases 1 and 8 and perhaps 3 in the present series, where no cervical canal was palpable, could be considered as cases of cervical atresia. Williams states that 'atresia of the cervix appears to be among

the rarest of congenital anomalies'.

The cases described by Rotter and Zaron *et al* of congenital atresia of the cervix are not comparable to the present series since a normal vagina was present in these cases. Only the cervix was non-canalized. But the treatment was that of suturing the endometrium to the vaginal epithelium over a skingrafted polythene tube.

Conception and delivery following the construction of an artificial vagina are rare. The successful result in each of the six reported cases was achieved by means of different operations. Whittemore did channel dissection between bladder and rectum and grafted it with skin from the thigh and the labia minora. Wagner substituted a segment of sigmoid to join the uterus and the 2 c.m. long vaginal pouch. The technique of Baer and Decosta and that of Solomons are already described elsewhere.

The diagnosis of this condition should not be difficult. It should be suspected in cases of primary amenorrhoea with periodic abdominal pain. Routine probing of vagina in the female infant is unnecessary and dangerous. But the earlier the condition is recognised the better the prognosis for a functional uterus and tubes. Otherwise the chances of developing endometriosis is high as shown by Hanton. In the present series only case 3 where the condition was of ten years duration, showed evidence of endometriosis. Adolescents above the age of 14 years, complaining of primary amenorrhoea, should be examined for congenital anomalies before being prescribed hormone tablets. Several girls in Miller's series were placed on hormone therapy blindly. Commenting on the Miller and Stout series, the Editor of 'Survey' opines, "in these partial cases there is simply a failure of the down-growing müllerian tubercle to fuse with the upgrowing portion of the vagina derived from the urogenital sinus." However, none of the cases quoted above conform to this. The defect was not only a lack of fusion of the two sources of the vagina, but a lack of development of the müllerian part of the vagina.

In treating these patients the main aim should be to establish a connection between the uterus and the vulva so as to provide an outlet for the menstrual flow and provide a canal for coitus and impregnation. Hanton *et al* reported seven cases from the Mayo Clinic. The treatment in most was removal of the uterus and appendages. The child bearing function was preserved only in one. In another a serious attempt was made to preserve the reproductive organs. But she developed a recto-vaginal fistula, was subjected to a series of surgical procedures and ultimately underwent hysterectomy. Despite this, they believe that 'under proper conditions in a surgically repairable congenital obstruction to menstrual flow, every attempt should be made to preserve the child-bearing function'. They advocate using a segment of sigmoid to replace the atretic portion of the vagina. Skin grafting and drainage of the haematometra at the same sitting may not be successful due to the constant flow of old blood on to the graft. Solomon's treatment of channelling,

draining and grafting at three to four weeks' intervals should work provided that the patients are not admitted in acute and severe pain needing prompt drainage. Also the patients should be willing to stay long enough in the hospital and they should be intelligent enough to use a mould. Unless the cases are of long standing duration and associated abnormalities or diseases are suspected, there is no need to do a laparotomy. Good functional results can be achieved by a perineal approach. The treatment adopted in the present series of cases was very simple and though it cannot be considered as adequate in so far as the construction of a good functioning vagina was not achieved, it was suited for the conditions under which these patients were treated. The difficulties experienced by Frith in Arabia for skin grafting are encountered in the villages of India too. Most patients do not agree for a second operation nor do they stay long enough. It is futile to send them home with a mould as they do not come for check up unless troubled by further complaints to seek medical aid. In case 2 an artificial vagina was created six weeks after the first operation by turning in the skin flaps from the labia. But the vagina contracted down to about two inches depth.

Summary

Nine cases with congenital atresia of the müllerian part of the vagina resulting in haematometra, haematocervix and haematosalpinx are described. Three of these might have also had atresia of the cervix. One case had a congenital absence of the recto-vaginal septum and the perineal body. Another had a rudimentary horn of the uterus and septum in the lower ¼th of the vagina in addition to the vaginal atresia. One patient was admitted with a full term pregnancy and obstructed labour and was delivered by lower segment caesarean section.

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