THE
Journal of Obstetrics & Gynaecology of India

VOLUME XXIX No. 3 JUNE 1979

CONGENITAL UROLOGIC DEFECTS WITH MULLERIAN DUCT MALFORMATION

by

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Introduction

While investigating a series of cases of Mullerian duct malformation, it was observed that a significant number of cases exhibited simultaneous urologic defect. From the study it further appeared that these urologic defects remaining undetected, may occassionally lead to diagnostic as well as therapeutic problems. The objective of the present communication, therefore, is to evaluate the nature and clinical significance of the different types of urologic defects which often multiply the problems of these women who are already congenitally handicapped.

Material

Fifteen cases of urinary tract abnormalities associated with Mullerian duct malformation have been studied. Fourteen of them were recorded during prospective study of 98 cases of vaginal agenesis with various grades of uterine malformation. Only 1 case of urologic defect was detected during retrospective investigation of a case of habitual abortion who had a bicornuate uterus and intravenous pyelogram revealed gross abnormality of the urinary tract. The study was mainly concentrated at N.R.S. Medical College, Calcutta but cases referred from other institutions have also been included. These cases were recorded from January, 1969 through April, 1978, Out of 99, complete urological investigation could be carried out only on 63 cases (Table I).

Types of Urologic defect in relation to the specific nature of Mullerian Duct Malformation

Table II reveals that not only with gross abnormalities but even with minor developmental anomalies of the Mullerian

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Accepted for publication on 12-11-1978.

TABLE I

Number of Cases with Urologic Defect in Relation to Mullerian Duct Malformation

Total Number of cases with Mullerian duct malformation studied	99
Number of cases investigated urologically	63
Urologic defects detected	15
Incidence	23.8 Per cent

TABLE II

Types of Urologic Defect in Relation to Specific Mullerian Duct Malformation

	Types of urologic defect	Specific Mullerian duct malformation	No. of cases
1.	Unilateral absence of the kidney and ureter	Complete absence of the vagina and uterus	4
2.	Single pelvic kidney with short ureter	Complete absence of the vagina and uterus	3
3.	Blodder neck obstruction with hydroureter and mild degree of hydronephrosis	Complete absence of the vagina and uterus	1
4.	Abnormal size and situation of external urethral meatus	Complete absence of the vagina and uterus	5
5.	Congenital vesicouterine fistula leading to menouria	Complete absence of the vagina with haematometra	1
6.	Congenital band at the pelviureteral junction leading to hydronephrosis on the right side and non-functioning kidney on the left	Bicornuate uterus	1

duct the incidences of urologic defect is not uncommon. In 8 cases, gross urologic deftct was associated with complete vaginal agenesis of gross urologic defects, unilateral renal agenesis was detected in 7 cases. Of these, in 3 a single kidney with a short ureter was present in the pelvis (Fig. 1). Though they were asymptomatic urologically, clinical examination revealed a pelvic lump in 3 cases who had pelvic kidneys. Clinical diagnosis was initially mistaken as haematometra becauses this condition is commonly associated with vaginal atresia. Unilateral renal agenesis was noted in 4 cases and the single kidney present was in normal position. In 2 of them, there was duplication of kidney on the same side with a single ureter (Fig. 2 and 3). The last case in this group was a girl aged 16 who had bladder neck obstruction with chronic retention of urine leading to bilateral hydroureter and hydronephrosis (Fig. 4).

The opening of the external uretheral meatus was situated at the centre of the blind vagina in 5 cases and in 1, this was

reasonably big appearing more or less like a patent vagina (Fig. 5).

One case of partial absence of vagina with haematometra had congenital vesico-uterine fistula (Fig. 6).

The only case who had a bicornuate uterus without vaginal defect had history of recurrent abortion, intravenous pyelogram revealed nonfunctioning kidney on the left side and hydronephrosis due to congenital fibrous band pressing at the pelviureteral junction on the right side (Fig. 7).

Presenting Symptoms and Methods of Detection

It is apparent from Table III and IV that out of 15 cases only 6 had definite clinical evidence suggestive of some urologic defect. In 5 out of 6 cases, clinical examination revealed abnormal size and situation of external urethral meatus with complete absence of the vagina. The remaining case presented with the symptom of recurrent haematuria in addition to primary amenorrhoea. The exact diagnosis of vesicouterine fistula in

TABLE III
Methods of Detection

Methods	No. of cases	Nature of Urologic defect
Radiological	7	Unilateral absence of kidney including single pelvic kidney.
Clinical	5	Abnormal size and situation of external urethral meatus.
Clinical, cystoscopic and radiological	1	Bladder neck obstruction with mild hydroureter and hydronephrosis.
Clinical and operative	1	Congenital vesico-uterine fistula.
Clinical and radiological	1	Congenital band at pelviureteral junction with non-excretion of dye through left kidney and mild hydronephrosis of right kidney.

TABLE IV
Presenting Symptoms

Primary problem	Secondary Problem	No. of cases
1. Primary amenorrhoea	Nil	12
2. Primary amenorrhoea	Dysuria, frequency, pain in loin and occasional rise of temperature	1
3. Pain lower abdomen with recurrent haematuria	Primary amenorrhoea	1
4. Repeated abortion	Recurrent urinary tract infection	1

this case was confirmed at laparotomy. Prior to laparotomy she had examination of urine in between haematuria which did not reveal any evidence of urinary tract infection. It is because of an abdominal lump and complete vaginal atresia, laparotomy was performed. Haematometra with cord like solid cervix connected with blind vagina behind the bladder was detected. The tubes and ovaries were normal. While the bladder was being separated a fistulous tract was seen communicating between the uterus on one side and the bladder on the other. The opening in the uterus was at a higher level than the opening in the bladder (Fig. 7).

There were 2 more cases who had urinary symptoms but for them the urinary symptoms were of secondary importance. In 1 case, the primary problem was recurrent abortion and a scrutinising enquiry revealed that she had recurrent urinary tract infection during successive pregnancies. Renal radiological investigation finally proved that the basic etiological factor of her consecutive miscarriage was a congenital urological defect. The other case, who also initially had urinary symptom, presented with the primary problem of amenorrhoea and pain in lower abdomen. Clinical examination revealed congenital absence of the vagina and a lower abdominal lump. Rectal examination suggested absence of haematocolpos and the lower abdominal lump was presumed to be due to haematometra. Examination under anaesthesia revealed that lower abdominal lump was full bladder, but catheterisation was difficult. Careful dilatation of urethra followed by cystoscopic examination proved that she had congenital bladder neck obstruction. Radiological examination demonstrated mild ureteral and renal changes. On subsequent enquiry, history of difficulty and frequency of micturition with occasional rise of temperature could be elicited. The urologic defect in the remaining 7 cases could be detected by routine radiological examination and they had no urinary symptom whatsoever.

Discussion

From embryological consideration it appears that congenital defect of one system is likely to be associated with similar defect of other systems. With genetic or chromosomal background, the the deformities are usually unselective and widespread, affecting organs situated far apart in the body. In other words, the organs affected do not have any embryological link or proximity. On the other hand, when the congenital deformity is due to some environmental factor it is likely that the neighbouring organs are usually involved. Mullerian duct anomalies are frequently associated with urologic and osseous defects. They bony abnormalities are also restricted specifically to the lumbar spine and sacrum (Bryan *et al* 1949). These considerations suggest that the specific Mullerian duct abnormalities discussed in this study are possibly due to environmental factors and perhaps not due to genetic disorder. This view has been substantiated by similar observation of Selezneva *et al* (1976) and Tolmachevskaya (1976).

It has been estimated that 40 per cent of cases of congenital abnormalities of the reproductive system are associated with simultaneous malformation of other systems. Of these, renal and urinary tract defects exist in 60 per cent of cases (Tolmachevskaya, 1976). Fore *et al* (1975) reported 47 per cent urologic abnormalities with congenital absence of the

vagina. The incidence of urologic abnormalities recorded in the present series was 23.8 per cent.

Of all urologic defects, unilateral renal absence was observed in 7 out of 15 cases (47%) in the present series. Of these, 3 (about 50%) had a single pelvic kidney. Unilateral renal agenesis associated with congenital absence of the vagina has been reported by Wiersma *et al* (1976); Hingorani (1976); Dougherty (1968) and Hauser (1976). Fore *et al* (1975) studied 36 cases of congenital absence of vagina urologically and recorded 2 cases who had a single pelvic kidney.

Unilateral renal agenesis is a significant observation particularly when the kidney is situated in the pelvis. Pelvic kidney with Mullerian duct malformation is usually solitary. Unless this fact has been predetermined, the kidney by mistake may be removed considering it to be a "Pelvic Mass". Besides this, there is also the risk of traumatising the kidney when an attempt is made for reconstruction of vagina by blind approach from below unless the position of the kidney in the pelvis has been known by preoperative radiological investigation.

Fore *et al* (1975) performed laparoscopic examination in cases of congenital absence of the vagina with pelvic kidney and reported that though these patients had normal ovaries they did not have significant Mullerian tissues. This finding is not consistent with our observation and we have found existence of same type of Mullerian tissue in these subjects as is found in other cases of Rokitansky-Kuster-Hauser Syndrome. Due to the presence of pelvic kidney the Mullerian knobs are displaced far laterally in the pelvis—sometimes on the iliac vessels and the connecting fibromuscular duct is stretched out on the top of the pelvic kidney. This precise pelvic finding could be observed in our cases because we got a better view of pelvic structures by laparotomy (Fig. 8).

Haematuria in association with vaginal atresia is a significant symptom suggestive of some urologic defect. One such case has been recorded in this series who had vaginal atresia with haematometra. The uterus, distended with blood, communicated with the bladder. Richter (1976) reported a similar case. But in his case the orifice of the portio vaginalis along with the right ureter opened into the proximal urethra. These cases apparently have a valvular mechanism which allows blood to enter into the urinary tract but prevents urine to escape into the genital organs.

In complete vaginal atresia the position and size of external urethral meatus may be abnormal. Unless this is appreciated, there remains a greater risk of injury to the urethra during dissection of space for new vagina between the bladder and rectum. Wharton (1946) has reported a similar case in which the urethra was so much dilated that this was used as a regular copulatory organs.

Majority of the congenital urologic defects in association with Mullerian duct abnormalities have been reported when there was a specific developmental anomaly of the vagina. Wiersma *et al* (1977) reported one case of bicrnuate uterus with congenital absence of the right kidney. She had practically no vaginal deformity. Herlyn (1976) described a syndrome characterised by double uterus, homolateral aplasia of the kidney and open Gartner duct cyst draining into the normal vagina. In the present series one who had recurrent mid-trimester abortion had bicornuate uterus and a congenital fibrous band pressing at

the pelvi-urethral junction leading to moderate degree of hydronephrosis on the right side and absent renal shadow on the left. Recurrent urinary tract infection due to this congenital urologic defect rather than the uterine anomaly was the contributory factor for her obstetric failures. Pitkin (1977) has commended that whenever an obstetric hazard is believed to be due to some uterine malformation, the urinary tract should be thoroughly studied. Conversely, in young girls with urinary tract abnormalities the normalcy of the reproductive organs should also be investigated.

Summary

Congenital urological defect was recorded in 15 cases in association with Mullerian duct malformation. The urologic defects were detected during prospective study of 98 cases of vaginal atresia and retrospective study of one case who had bicornuate uterus.

The specific urologic defects detected were; unilateral renal agenesis in 4 of which 2 had contralateral duplication, single pelvic kidney in 3, congenital band at pelviureteral junction, congenital vesicouterine fistula leading to menouria, congenital bladder neck obstruction—one each and abnormal size and situation of external urethral meatus in 5 cases.

The significance of urologic defects associated with Mullerian duct malformation has been discussed.

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See Figs. on Art Paper I-II