

## MALE PSEUDOHERMAPHRODITE WITH UTERUS IN SCROTUM

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The determination of sex in patients having controversial or ambiguous external genitalia is still a problem, even in developed countries. In India, where illiteracy is there and sex of an infant is declared by untrained 'Dais' who conduct the home deliveries. The delayed sex determination results great psychological trauma to the child as well as to the parents. In hermaphrodites or in their variants, mostly gonads and genital organs are situated at their normal positions or may be undescended and lying in the pelvis. But here we had an unusual case, where an apparently male looking individual had right sided undescended testes and presence of uterus with tubes and both the testes in left scrotum along with left sided oblique inguinal hernia.

### Case History

A 32 year old, morphologically male individual attended our general surgery service for the treatment of left side oblique inguinal hernia. He was born in his village and was brought up as male. He got married, when he was 18 years, but remained issueless till date. He had normal sexual functions and had normal development of secondary sex character of a male.

On physical examination he was an young male with average built and average nutrition, having a height of 5'6" with well developed beard, pubic hairs and normal male breast (Fig. 1). The systemic examination of cardiovascular, respiratory and nervous system did not reveal any abnormality.

On local examination of external genitalia and inguinal region it was found that the both sides of scrotum were well formed and right side testes was undescended. He had leftsided reducible inguinal hernia, even after complete reduction of hernial contents, the left side scrotum was bulky (Fig. 2), having a soft tissue mass and testicle like two swellings.

The semen study revealed azospermia. The kidneys, ureters and urinary bladders along with urethra were normally demonstrated in intravenous pyelogram and micturating cystourethrogram respectively. The sex chromatin was negative for Barr bodies. The 17-Ketosteroid also was found to be in normal range in 24 hour sample of urine.

He had his hernial repair under general anaesthesia. On exploration of the left inguinal canal it was found that he was having a mass probably consisting of two gonads with vas deference and uterus with fallopian tubes besides the hernial sac (Fig. 3 and 4). An attempt was made to ascertain and assign the structures and their relative positions. The gonads were placed on either side of the uterus. There was an appreciable gap between the proximal end of vas and the epididymis. A small opening was made in uterine fundus. It was empty. A catheter could be passed into the uterine cavity and its canal onwards i.e. towards the pelvis.

A middle suprapubic exploration was done at the same stage, which revealed that

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right side, gonad was not present any where and the right deep inguinal ring was closed. The uterine cavity was extending towards the posterior surface of posterior urethra in form of a canal, alongwith two vas deference over its outer surface. Nothing further was done at this stage except that the biopsy pieces were taken from the gonadal masses, vas deference and the uterine wall. Histology confirmed the gonadal masses to be testes with atropic seminiferous tubules and endometrium of the uterus was found to be non secretory.

A re-exploration was done after a gap of 15 days. The uterus alongwith fallopian tubes were removed. During its removal a penile catheter was passed in the uterine canal. On pushing it further, it came out through the glans penis (Fig. 5) indicating its continuity with the urethra. The uterine canal was excised flush with the urethra deep down in the pelvic cavity. The continuity of the epididymis was restored with the vas deference and orchipaxy was performed. Then the hernial repair was done as usual.

The post operative phase was uneventful, and he left the hospital on 12th day of the second operation.

### Discussion

The existence of uterus and testes, together is an embryological possibility (Fig. 6). The female ductal system upto upper 1/3rd of vagina was formed and was opening in posterior part of urethra in this case. The male ductal system as well as female ductal system both opens in urogenital sinus in an embryo as is shown in figure 6.

The gonad develops from medulla and cortex at 6th week of development and has identical structure in both sexes. During 7th and 8th week the medulla of genetic male forms testes and cortex regresses. But

in genetic females, the cortex develops into an ovary and the medulla regresses.

The presence of uterus and both the tubes on one side and that too, in scrotum could be due to presence of hernia, which dragged down these structures also.

The patient was advised to continue as a male, which he happily accepted.

### Summary :

An unusual case of a male pseudohermaphrodite where uterus with its fallopian tubes and both the testes with their vas deference were present on one side only, has been presented. The gonadal abnormality was found to be associated with oblique inguinal hernia on the same side.

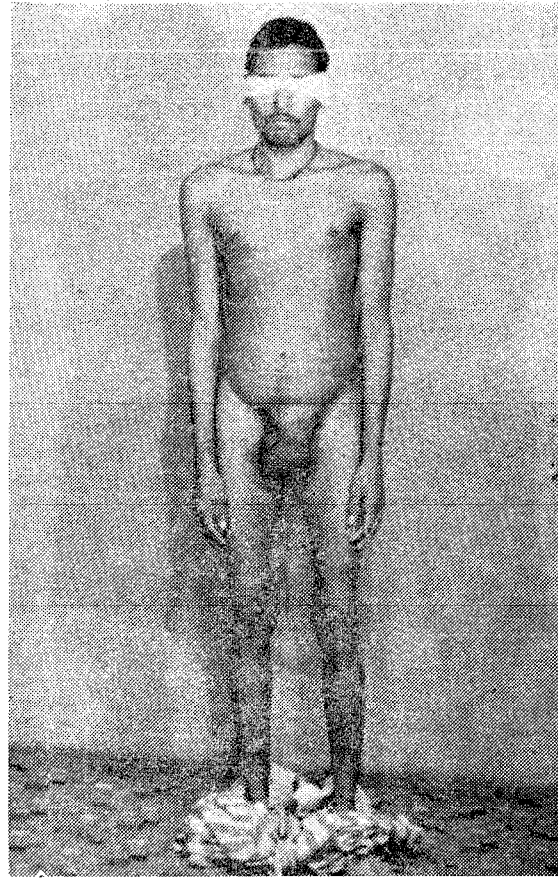


Fig. 1. Photograph showing patient in full size.



Fig. 2. Photograph showing External genitalia of the patient.

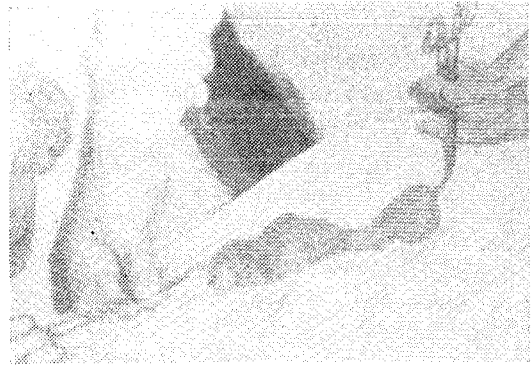


Fig. 3. Operative photograph showing Internal genital organs.

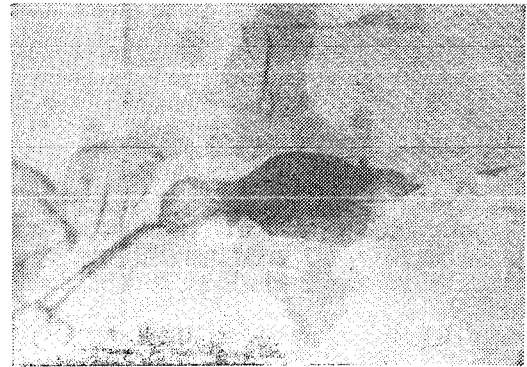


Fig. 4. Operative photograph showing internal genitalia.

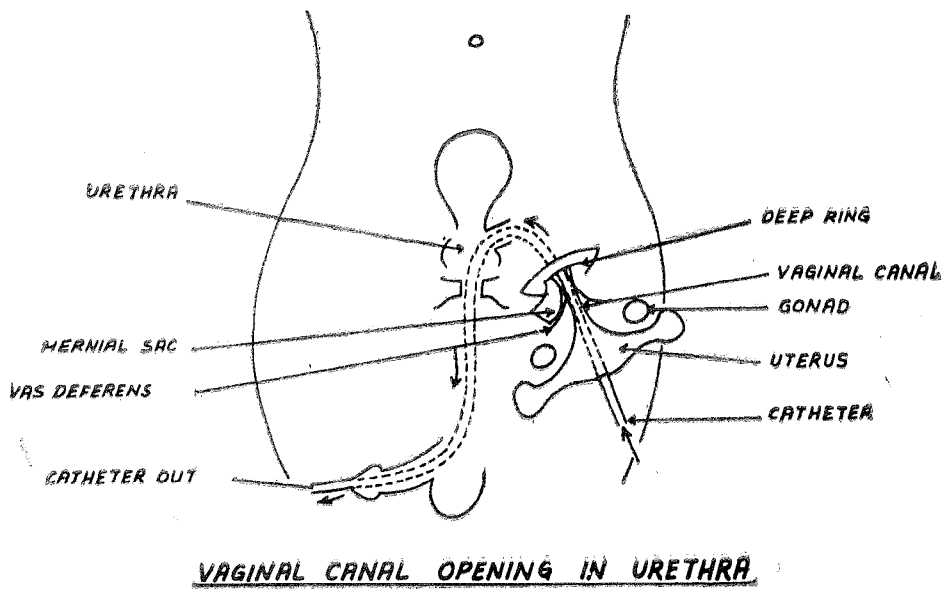


Fig. 5. Diagram showing pathway of catheter from uterus to glans penis.

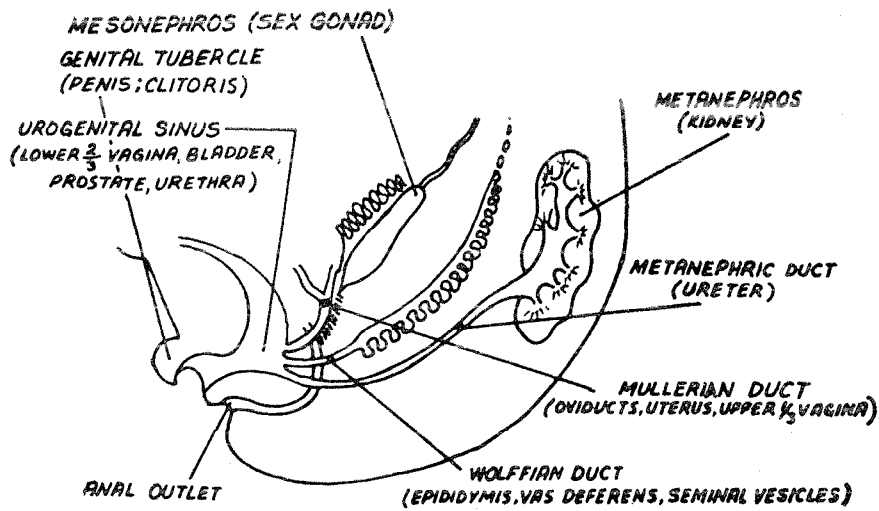


Fig. 6. Diagram showing Embryological stage of development of gonad and ductal system.

#### References

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