

EPISPADIAS IN FEMALES

by

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Introduction

Epispadias is a very rare congenital anomaly in which the anterior wall of urethra is partially or completely absent. Dees (1949) found its incidence to be 1:117,604 in males and 1:481,110 in females. Other workers have found its incidence to be higher in males, but in females it continues to be rare.

The anomaly is a regular accompaniment of vesical extrophy and, even though the bladder is closed epispadias should always be considered as the early stage in the gradation of the extrophy.

In females, the degrees of epispadias are designated as clitoric, subsymphyseal and complete. The clitoric form is the mildest degree of deformity and is characterised by a bifid clitoris; the urethra is intact and the patient is continent. In the subsymphyseal form, the urethra is slit dorsally further up to the level of bladder sphincters which are intact. In the complete variety which is the commonest (incidence 90%), the

cleft involves the bladder sphincters as well and these children never gain continence; the urethra is short and has a very wide opening a little anterior to the bifid clitoris through which the examining finger can be readily admitted into the bladder and often there is prolapse of the bladder mucosa. There is constant dribbling of urine and the bladder is contracted. There is separation of labia and pubic bones, the extent of which is proportional to the degree of the epispadias. The gap between the pubic bones is bridged by a band of fibrous tissue. Usually the other genital organs are normally developed and there are no reproductive problems. Coitus and pregnancy are possible.

The high incidence of the complete variety may partly be apparent; since the clitoric and subsymphyseal varieties are asymptomatic and the deformity is of mild degree, it may be inconspicuous and likely to be missed unless a very careful examination is made.

The embryogenesis of epispadias is the same as that of vesical extrophy, the difference being that of degree. Amongst many theories advanced, that of Gilles and Harrison (1948) is widely accepted. In this theory a forward displacement of the cloacal

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Received for publication on 4-9-1969.

membrane (also called anal plate) is postulated. Since the mesoderm fails to develop locally in the cloacal membrane, the mesodermal structures comprising the lower abdominal wall here fail to develop. The cloacal membrane ultimately ruptures leaving an opening which extends in the mid-line to the umbilicus to a degree proportional to the size and displacement of the cloacal membrane. The details of the embryological mechanism involved are well documented by Patten and Marry (1952) in their work.

Treatment

The operations for epispadias in males aim at correction of incontinence, if present, and reconstruction and repair of urethra and penis, so that they will be cosmetically acceptable and will function as intended by nature. However, in females the problem is much simpler and exclusively that of incontinence. Hence in clitoric and subsymphyseal varieties, where incontinence is absent, surgical correction is not required. But in the complete type of epispadias, surgical repair for incontinence has to be done. Different operative procedures have been devised by Young (1922), Gross and Cresson (1952), Campbell (1952) and others. All these procedures aim at mobilisation and approximation of the muscle tissue in the vicinity of vesical neck so as to establish a functioning sphincter. However, because of the rarity of this condition, each series consists of a few isolated cases, treatment does not always bring about cure. Quite a few of these patients ultimately need some sort of urinary diversion, commonly a ureterosigmoidostomy.

Millin (1947) introduced a sling operation originally for stress incontinence in females. This was later successfully tried for post-prostatectomy urinary incontinence following trans-urethral resections. Isolated instances of success of this procedure in complete epispadias in the female have been reported by Everett and Williams (1963). The operation is carried out by the suprapubic approach through a curved incision and employs the use of two transverse strips of the anterior rectus sheath, with one end attached to and continuous with the external rectus sheath of the other side. These strips after being passed through the rectus muscles are brought underneath the bladder neck and their ends stitched to the strips of the opposite side (Fig. 1).

STEPS OF MILLIN'S RETROPUBIC SLING OPERATION

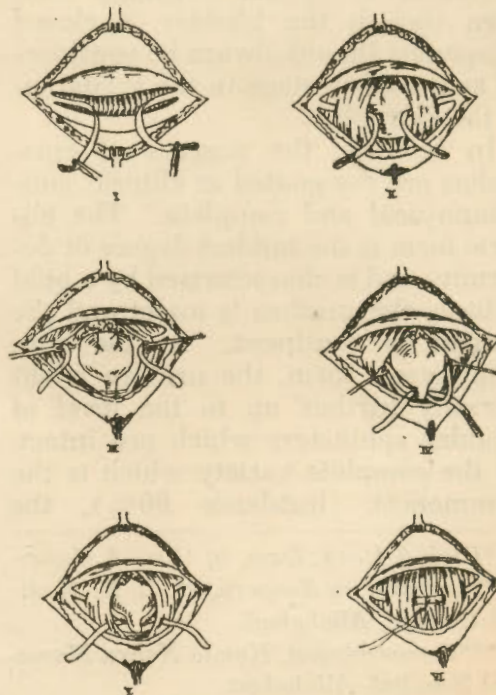


Fig. 1

This firm hammock-like suspension made of crossed fascial flaps gives strong support to the urethra and also forms an efficient obliterating angle at the vesicourethral junction. During coughing, sneezing or exercise, contraction of the rectus and other abdominal muscles elevates the sling and obliterates the urethra at the vesico-urethral junction. During the act of micturition on the other hand, the muscles are relaxed and consequently the sling comes down and the vesical neck is now opened. Fig. II Thus after a short period of post-

operative bladder re-education with sphincter exercises the patient can develop continence satisfactorily.

We report here, two cases of complete epispadias in females, who were successfully treated by Millin's sling procedure.

Case Reports

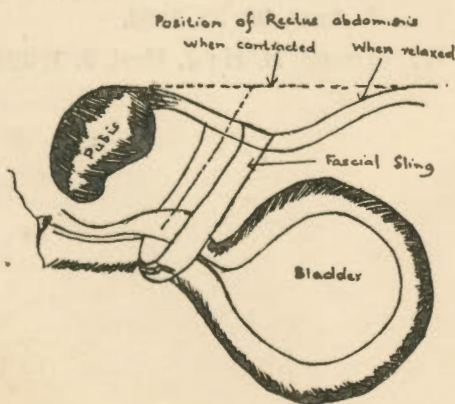
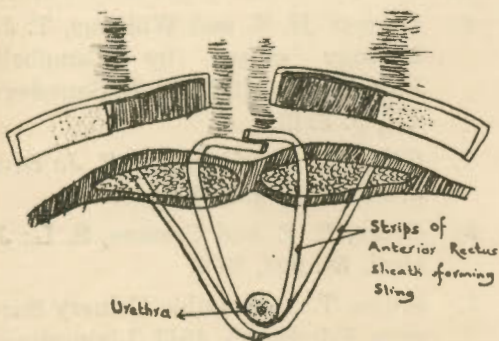
Case 1

A.B., aged 17 years—Hindu female, resident of a village near Allahabad, was admitted in Kamla Nehru Memorial Hospital in February 1965 with the complaints of constant dribbling of urine ever since she could remember. The patient said she had never passed urine in a stream, and had been constantly soiling her clothes.

Her parents, two sisters and one brother were all healthy. There was no history of any infectious fever in the mother while she (the mother) was expecting the patient. She was married at the age of 14 but because of her ailment, the husband deserted her one year later. Her periods were regular.

The patient was a shy girl of weak build. Her temperature, pulse and blood pressure were within normal limits. Examination of the genito-urinary system revealed a bifid widely separated clitoris with external urinary meatus in front of it, and wide enough to allow passage of the index finger. The urine constantly dribbled from the urethra. The pubic symphysis, though short in length, was otherwise well developed. The bladder was empty. The vagina was well developed, and vaginal examination revealed normally developed anteflexed and anteverted uterus. The breasts were developed normally. No other clinical abnormality could be detected.

Millin's retropubic sling operation was done from which she made an uninterrupted recovery. Five days later, when the foley's catheter was removed, she could experience some control over her micturition. Sphincter exercises brought further and quick improvement and fifteen days later she was allowed home. She was last seen four months later in a very happy state, since she had never felt so well before and had now returned to her husband.



SCHEMATIC REPRESENTATION OF ACTION OF SLING.

Fig. 2

Case 2

P.D., aged 18 years, was admitted in Kamla Nehru Memorial Hospital, Allahabad, with a history of dribbling of urine and strangury since birth. She was the second child of her parents, her two brothers being healthy and normal. She was married one year earlier and had no conjugal problems. Her menarche started at the age of 15 years and her menstrual cycle was regular and normal.

She was of average build, with temperature, pulse rate and blood pressure within normal limits. The breasts were well developed and examination of the vulva revealed a bifid clitoris with a more anterior and wider external urethral opening through which the urine constantly dribbled. The interoitus was tight but the vagina and uterus were well developed and normal. The bladder was empty. The other systems were clinically normal. Hb. was 12.4 gm %. X-ray of the pelvis revealed increased separation of pubic bones in the region of the symphysis pubis; other parts of the skeleton were normal.

Millin's retropubic sling operation was undertaken with uneventful post-operative recovery. The foley's catheter was removed from the urethra on the 4th post-operative day, when the patient reported continence and passed urine in a stream. This gradually improved with sphincter exercises. Cystography was done on the 15th post-operative day and showed that the patient could retain six ounces of fluid without leakage. She was allowed home 3 weeks after the operation and reported well six months later.

Summary

Two very rare cases of complete epispadias in females, who were successfully treated by Millin's retropubic sling operation are reported. The literature on the subject has been reviewed.

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