# EXSTROPHY OF THE URINARY BLADDER



ESSAY

Presented by

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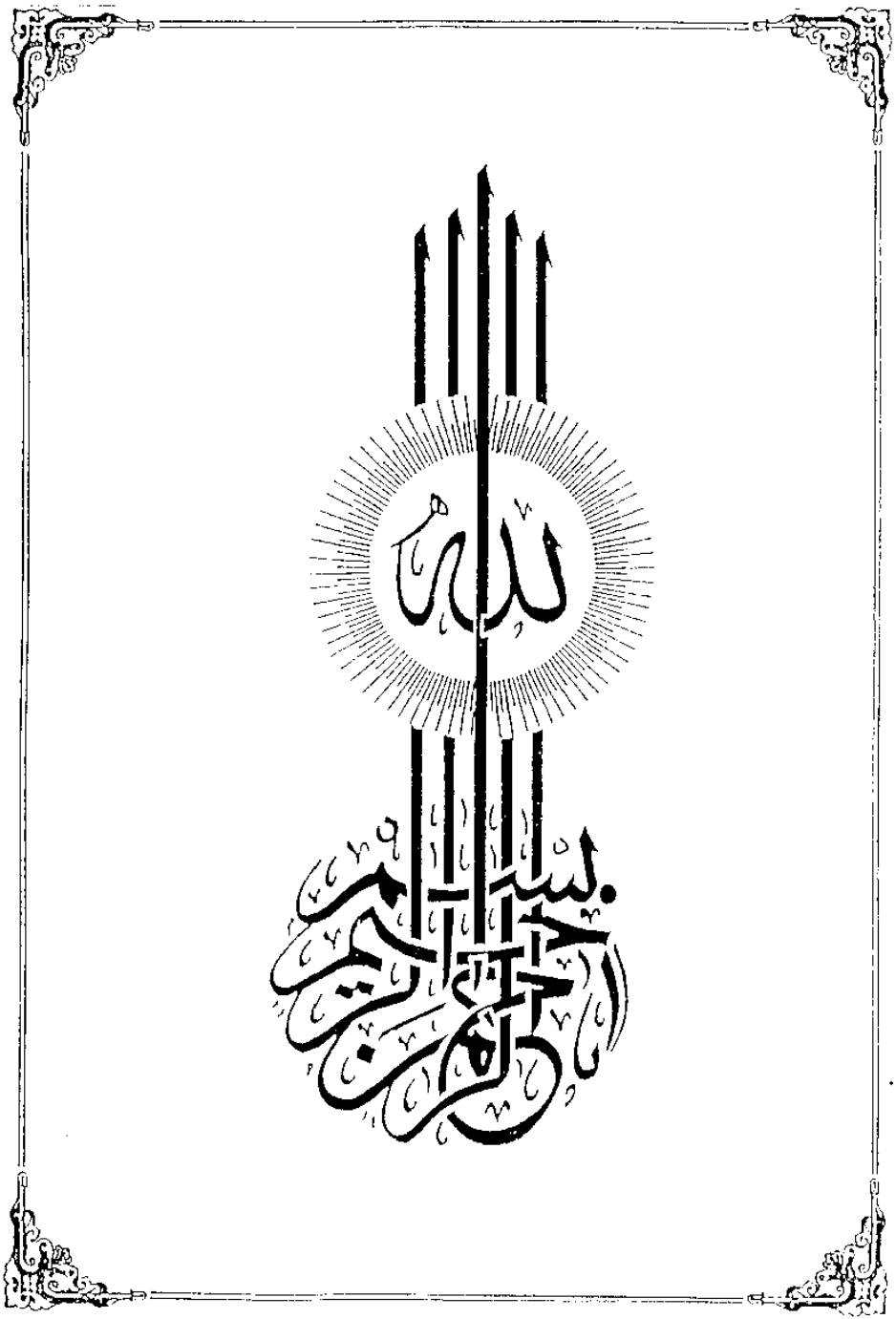

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CONTENTS

# I N T R O D U C T I O N

Exstrophy of the bladder (Ectopia vesica) is one of the common problems in Pediatric surgery. Apart from its social problems reflected on the parents, early detection and management is mandatory to avoid probable complications.

Different methods have been introduced to treat exstrophy of the bladder. Successful urinary diversion began with Maydle in 1894, who transplanted the trigone into the rectum. Ureterosigmoidostomy began with Coffey's, but it was faced by many complications (Coffey's,1911). The introduction of mucosa-to-mucosa anastomosis by Nesbit in 1949, and the combined tunnel and mucosa technique of Leadbetter in 1955, had improved the results of Coffey's method, and helped to avoid its complications (mainly infection and acidosis).

Anatomical reconstruction of the bony pelvis in exstrophy of the bladder was attempted first by Trendelenburg in 1906, but the first sporadic successes were reported by Burns in 1924 and Janssen in 1933.

Although since 1950, many methods have been applied both in reconstruction and diversion, other newer techniques are still expected in coming years for management of exstrophy of the bladder.

# H I S T O R Y

The first description of exstrophy of the bladder is on an Assyrian tablet from 2000 B.C. preserved in the British Museum in London. No other written reference occur until Seheuke von Grafenberg described it in 1597, which may indicate a rather drastic treatment of newborn anomalies in ancient times. In 1849, Mackay and Syme advocated the application of an external urinary receptacle. Syme performed the first successful ureterosigmoid anastomosis in 1852, but 9 months later the patient died of ascending pyelonephritis. Carl Thiersch (1869) covered the bladder with lateral flaps obtaining a bladder capacity of 100 ml. Successful urinary diversion began with Karl Maydl (1894), who transplanted the trigone into the rectum. Ureterosigmoidostomy began with Coffey's (1911) method, but infection and acidosis caused severe complications until the introduction of mucosa-to-mucosa anastomosis by Nesbit in 1949 and the combined tunnel and mucosa technique of Leadbetter(1955).

The anatomical reconstruction of the bony pelvis in exstrophy was first attempted by Trendelenburg in 1906, but the first sporadic successes were reported by Burns in 1924 and Janssen in 1933. H.H. Young in 1942 reported the first female with urinary continence after closure of exstrophy and, Michon 1948 reported success in a male .

Since 1950, many methods have been applied both in reconstruction and diversion. In diversion, emphasis has been on preventing reflux of infected urine to the kidneys. In reconstruction, increasing success has been achieved and can be attributed to staging of the repair, employing separate procedures for osteotomy, closure of bladder, repair of genitalia, and correction of continence at the bladder neck, which includes relocation of the ureters.

# E M B R Y O L O G Y

Exstrophy of the bladder is part of a spectrum of conditions resul-
ng from abnormal development of the cloacal membrane. The spectrum ra-
es from glandular epispadias to cloacal exstrophy, but exstrophy acco-
ts for 50% of the patients in the group (Jeffs, 1982).

The variance of the exstrophy-epispadias complex results from abnor-
l development during the 4th to the 10th week of gestation. Initially,
e common cloaca is separated from the amniotic space by the bilayered
oacal membrane, which occupies the infraumbilical abdominal wall. Mes-
chymal ingrowth between the ectodermal and endodermal layers of the
oacal membrane results in formation of the lower abdominal muscles and
e pelvis bones. The simultaneous downgrowth of the urorectal septum
vides the cloaca into a bladder anteriorly and a rectum posteriorly .
stally, this septum meets the posterior remnant of the cloacal membra-
, which eventually perforates, forming anal and urogenital openings.
e paired genital tubercles migrate medially and fuse in the midline
phalad to the cloacal membrane before perforation.(Fig .1)

The present theory of embryologic maldevelopment in exstrophy, held
Marshall and Muecke (1968), is that the basic defect is an abnormal
erdevelopment of the cloacal membrane, preventing migration of mesenc-
nal tissue and proper lower abdominal wall development. The position
d timing of the rupture in this cloacal membrane would determine the
riant of the epispadias-exstrophy complex that would result. This the-
y is supported by Muecke's (1964) work in the chick embryo and by the
pected high incidence of central perforation resulting in the prepond-
ance of classic exstrophy variants. Pohlman's (1911) observation

of an abnormal embryo in which the cloacal membrane was usually large lends support to the Marshall-Muecke theory of overdevelopment of this membrane.

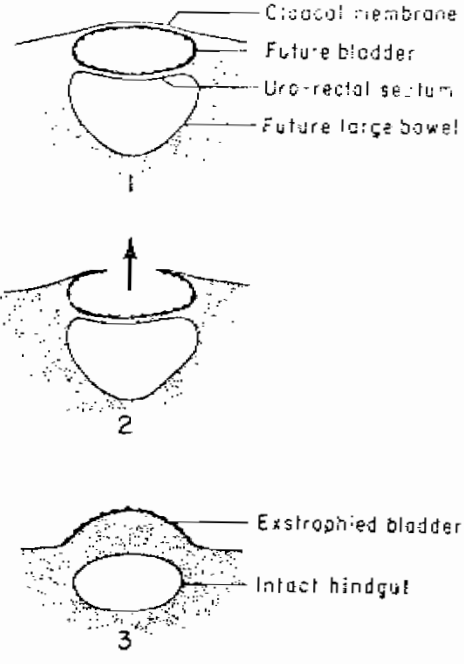



Fig -1.—Diagram of embryologic events leading to classic exstrophy.

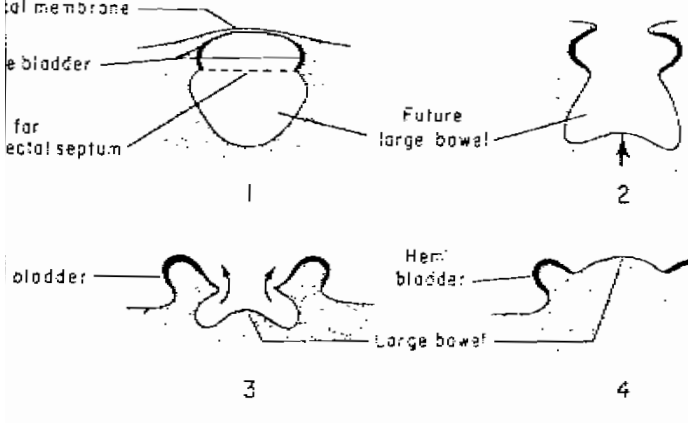



Fig -2.—Diagram of event ation of cloaca to form cloacal exstrophy.

## INCIDENCE AND INHERITANCE

The incidence of exstrophy of the bladder has been estimated at between 1 : 10,000 (Rickham, 1960) to 1 : 50,000 (Lattimer Smith 1966) live births. The male-to-female sex ratio of exstrophy of the bladder derived from the combined series of Higgins, Gross and Cresson, Jeffs et al., Bennett and Harvard and Thompson is 2.3 : 1.0. (1951).

The risk of recurrence of the exstrophy of the bladder in a given family is approximately 1 : 100 (Ives, et al., 1980) . Shapiro et al., (1984), surveyed pediatric urologists and surgeons in North America and Europe by questionnaire and identified recurrence of exstrophy and epispadias in only nine of approximately 2,500 indexed cases. Lattimer and Smith (1966) cited a set of identical twins with exstrophy of the bladder and another set of twins in which only one child had the condition. Higgins (1962) observed two sets of twins and two sets of siblings in the same family with exstrophy of the bladder. From Shapiro's data (1984), five sets of male and female nonidentical twins were identified in which only one twin was affected; five sets of male identical twins were identified in which both twins were affected; one set of identical male twins was identified in which only one twin was affected; and three sets of female identical twins were identified in which only one twin had the exstrophy anomaly.

The inheritance pattern of exstrophy of the bladder has not been established. Clemetson's (1958) literature review identified 45 females with exstrophy of the bladder who produced 49 offspring, and in no instance did any of their offspring demonstrate features of the exstrophy epispadias complex. Until recently, exstrophy of the bladder or epispadias had not been reported in offspring of parents with exstrophy-

pispadias complex. Shapiro et al., (1984) reported two females with omplete epispadias. Each gave birth to a son with exstrophy of the bl- dder, and another female with exstrophy of the bladder. Each daughter roduced a son with exstrophy of the bladder. The inheritance of these hree cases of exstrophy of the bladder was identified in a total of 25 offspring (75 males and 150 females) produced by individuals with xstrophy of the bladder and epispadias. Shapiro (1984) determined that he risk of the bladder in offspring of individuals with exstrophy of he bladder and epispadias is 1 : 70 live birth, a 500+ -fold greater ncidence than that in the general population.

In Pediatric Surgery Unit, Ain Shams University, Egypt ,twenty-four tients with bladder exstrophy were treated over the last ten years ( e; 2.4 per year). There were eighteen males and six females (i.e;3:I) ahmasy,et al,I985).

PATHOLOGY

The pathological changes are also discussed under the heading "Urinary Defect", used by Williams(1974).

The size, distensibility, and neuromuscular function of the exstrophied bladder, and the size of the triangular fascial defect to which the bladder muscle is attached, affect the decision to attempt functional closure. When the bladder is small, fibrosed, and inelastic, functional closure may be impossible. The more normal bladder may be invaginated or may bulge through a small fascial defect, indicating the potential for satisfactory capacity.

At birth, the bladder mucosa may appear normal; however, ectopic bowel mucosa or an isolated bowel loop may be present on the bladder surface. Abnormal histologic indications of the bladder were observed in each of 23 bladder specimens obtained from individuals with exstrophy of the bladder between the ages of 1 month to 52 years (Jeffs, 1982). Squamous metaplasia, cystitis cystica, cystitis glandularis and acute and chronic inflammation were commonly identified in these exstrophic bladder specimens (Culp, 1964). Scanning and transmission electron microscopy of human exstrophic bladders have demonstrated microvilli and the absence of surface ridges on the uroepithelial cells (Clark, O'connell, 1973). The abnormal histologic features demonstrated by both light and electron microscopy may represent chronic mucosal changes secondary to persistent infection.

Also, normal detrusor function was achieved in only 22% of closed exstrophies (Nisonson, Lattimer, 1972). When bladder function was assessed in continent closed exstrophy patients, normally refluxive bladders and normal plug electromyograms were demonstrated in 70% and 90% of cases respectively (Toguri, et al., 1978).

## EVALUATION OF THE PATIENT

### CLINICAL PICTURE

Abnormal development and clinical picture will be discussed under the headings used by Williams (1974).

(I) Musculoskeletal Defects : (Fig.3)

1- All cases of exstrophy have the characteristic widening of the symphysis pubis caused by outward rotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints.

2- There is an outward rotation or eversion of the pubic rami at their junction with the ischial and iliac bones.

3- A lateral separation of the innominate bones inferiorly with the fulcrum at the ilial sacral joint. This is only present in more severe cases of exstrophy.

The bony defect causes little or no long-term orthopedic problem. Attempted reapproximation of the pubic rami anteriorly may, however have advantages in initial bladder closure, in;

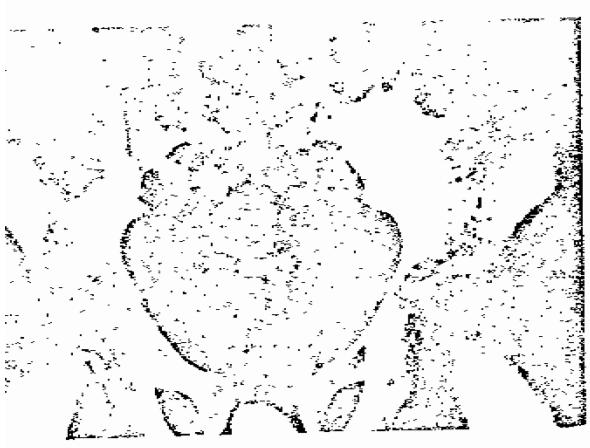
a) Molding the pelvis to more normal dimensions.

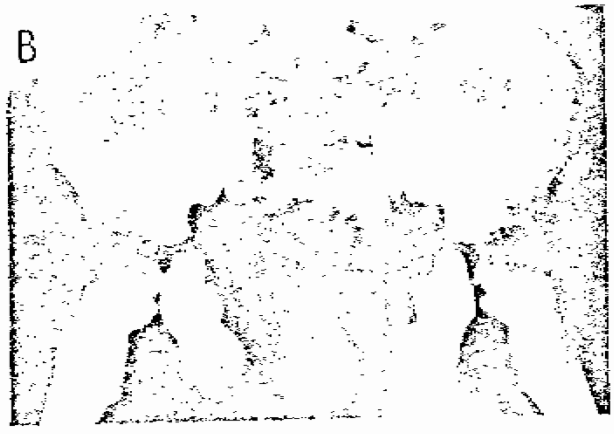
b) Bringing penile attachments closer together.

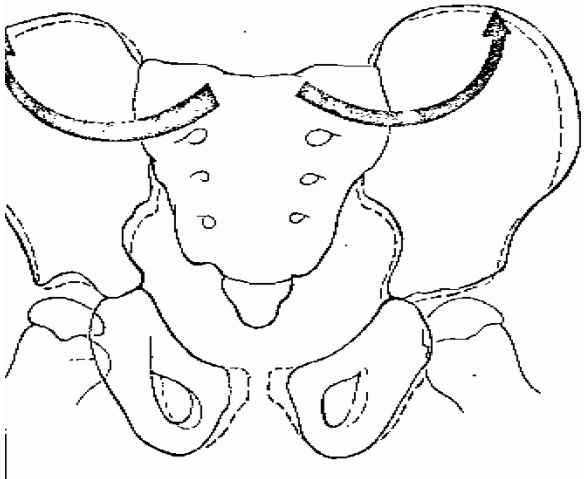
c) Making continence more easily achieved following reconstruction.

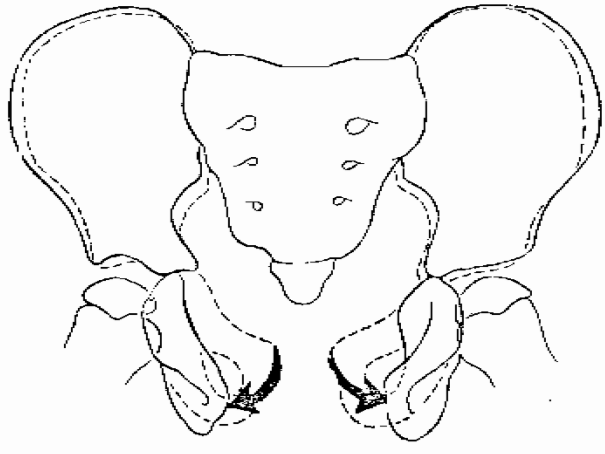
4- The distance between the umbilicus and the anus is always foreshortened because of the poor development adjacent to the abnormal cloacal membrane, leaving an unusual expanse of uninterrupted upper abdominal skin.

5- Rupture of the cloacal membrane allows bladder mucosa to fuse to adjacent abdominal skin through a "triangular fascial defect". This fascial defect is limited laterally by



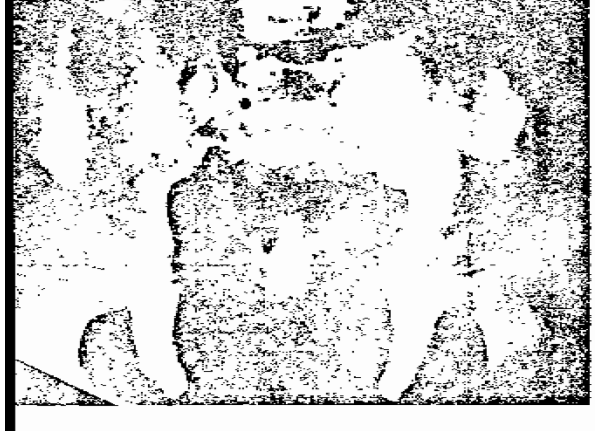


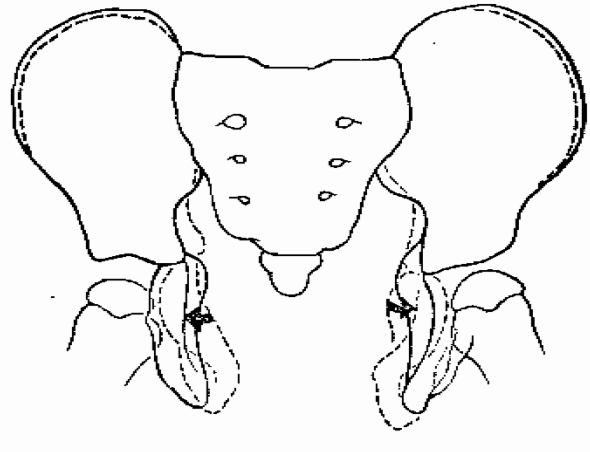





1 + 2

3 —Rotational and lateral deformities of the pelvic girdle in cases of phy. A, widening of the symphysis caused by outward rotation of the innate bones. This is usually the only skeletal abnormality present in epispadias. B, additional external rotation of the pubic bones; the characteristic skeletal changes in classic forms of exstrophy





1 + 2 + 3

Fig 3 Cont.—C, the final addition of lateral inferior separation of the innominate bones, present in the extreme manifestation of the complex, namely, cloacal exstrophy