EXSTROPHY OF THE BLADDER IN TWINS

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Exstrophy of the bladder, or ectopia vesicae, one of the most unfortunate congenital anomalies, occurs once in approximately 40,000 births, the ratio in the male and female being 7 to 1. A portion of the lower anterior abdominal wall and a portion of the anterior bladder wall are completely lacking, so that the posterior wall, with the trigon, presents itself in the suprapubic region, partially everted, as a result of intra-abdominal pressure. In a review of the literature I have been unable to find a report of the occurrence of this anomaly in twins.

ETIOLOGY

The various theories advanced to explain the etiology fall into 3 groups: (1) mechanical, (2) pathologic, and (3) embryologic.

Von Geldern[1] states that the **mechanical theory** may be divided into: "1. The 'berstungs theorie' (rupture theory), which considers the cause of exstrophy of the bladder and epispadias as a rupture of the anterior wall of the bladder and the adjacent abdominal wall due to a retention of fluids in the bladder. This retention is believed to be due to constrictions at various points along the genito-urinary tract." This implies obstruction of the urethra or the vesical neck producing distention of the bladder, which by direct pressure separates the ends of the pubic bones and recti abdominis. Eventually, the anterior wall of the bladder and the abdominal wall contiguous to it rupture, and healing occurs along the edge of the exposed bladder. The obstructive lesion responsible for the distention of the bladder has never been demonstrated. "2. The mechanical theory, which considers a short or absent umbilical cord as the causative factor. 3. A theory which is not based entirely on a mechanical conception, but depends on a transposition of the embryonal anlagen, and considers that the complicated exstrophies (ectopia cloacae) are due to change in position of the vitelline duct."

The **pathologic** theory advanced by Keith[2] states that toxins from an endometritis, or other forms of infection or irritation, produce a separation of the lateral walls of the caudal portion of the embryo.

Velpeau[3] and Phillips[4] believe that ulceration of the anterior abdominal wall occurs and that this also occurs at the symphysis pubis, with subsequent necrosis and separation. Such ulceration, however, is not recognizable, and necrosis of the ends of the pubic bones has not been observed in postmorten study.

Embryologic theories. Forster[5] contends that arrest in development is the causative factor in the production of simple exstrophy of the bladder and epispadias.

Von Geldern[1] states that "the arrest has affected those parts which had not completed their embryonal development, and that when development recommenced, certain primitive embryonal structures were maintained and developed as such, and those which should have undergone differentiation, had no arrest occurred, failed to make these changes."

Johnston[6] states that any explanation of this anomaly must consider the normal development of the bladder and lower end of the alimentary tract. Complete exstrophy of the bladder complicated by intestinal openings on the extroverted area is caused by rupture of the cloacal membrane. The rupture may occur at any time between the first appearance of the membrane and the subdivision of the cloaca.

Keibel[7] believes that the anomaly may result from a persistent open blastopore.

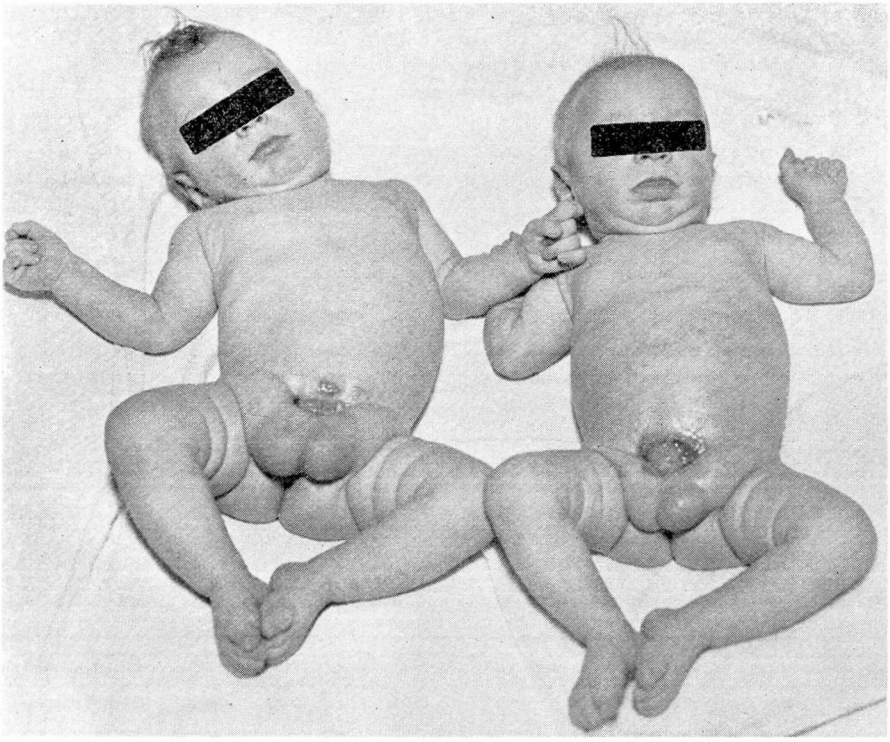

FIGURE 1

Wyburn[8] concludes: "(5) At any early stage the cloacal membrane is a relatively large area of contact of ectoderm and endoderm extending some distance along the allantoic diverticulum. (6) The allantoic cloacal membrane is later obliterated by the mesoderm pressing in towards the midventral line between the ectoderm and endoderm. (7) Extroversion of the bladder is due to mesodermal deficiency, particularly of the processes of secondary mesoderm arising from the hindend of the primitive streak, following an impaired development of the muscular coat of the bladder, of the symphysis pubis, and the formation of external genitals and the infra-umbilical portion of the anterior abdominal wall. (8) Epispadias is a similar mesodermal error in a minor form."

The following case reports are preliminary observations on the occurrence of exstrophy of the bladder in identical twins.

CASE REPORTS

Twin boys were admitted to the Clinic on June 23, 1943 for surgical correction of exstrophy of the bladder. The boys had been born March 23, 1943, there being one placenta and two cords. M. T. had weighed 5 pounds and 4 ounces at birth, and L. T. had weighed 4 pounds and 12 ounces. Two other children in the family, one girl aged 6 and another aged 5, were normal in all respects. Likewise, there was no evidence of a similar anomaly in the family history.

The children were 3 months old when seen. Their gain in weight had been retarded by frequent stools since birth. M. T. weighed 10 pounds and 12 ounces, and L. T. weighed 8 pounds and 14 ounces.

On physical examination both children appeared malnourished and dehydrated. M. T. had complete exstrophy of the bladder with an associated hypospadias and a bilateral indirect inguinal hernia. The testicles were descended and normal. L. T. likewise had a complete exstrophy of the bladder and associated epispadias. A right indirect inguinal hernia was also present. There was a well healed incision where a left indirect inguinal hernia, which had been strangulated, had been operated upon when the child was 6 weeks of age. The testicles were descended and normal.

The laboratory studies were normal. Complete urologic survey revealed no pathology in addition to the previously mentioned congenital anomalies.

SUMMARY

A preliminary report of identical twins born with exstrophy of the bladder is presented. The occurrence of the anomaly in twins would seem to give credence to the embryologic theory of the development of exstrophy of the bladder rather than to the mechanical and pathologic theories.

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