EXSTROPHY OF THE BLADDER DURING PREGNANCY

(A Case Report with Review of Literature)

by

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Introduction

Exstrophy of the bladder, as such, is a rare congenital anomaly found only in the human species. Morris (1949) stated the incidence as 1:50,000. It was found to be five times more frequent in males. This condition is rare in adults, because 50 per cent of the untreated children die before the age of ten years from renal complications; only one-third live up to the age of 20 years. Frank & Morris (1949) stated that pregnancy complicated by exstrophy of the bladder should be an extremely rare occurrence.

Review of Literature

Bonnet (1722) reported the first delivery in a case of exstrophy of the bladder. Miller and King (1918) found 13 cases in a review of literature and added one of their own.

Lotimer and Hamilton (1954) recorded 37 cases of pregnancy complicated by exstrophy of the bladder. These 37 women delivered 49 children. There were 38 vaginal deliveries including one set of twins and 10 caesarean sections. Forty-one children were born alive with no congenital anomaly. At least 18 of these patients had not had ureteral transplantations. Randall and Hardwick (1934) observed only 20 women who lived long enough to become pregnant along with the exstrophy of the bladder and hence had received little consideration. Additional cases were reported by Damm (1937) Schumann (1042) and Osterguard (1943).

The case reported by Dawson (1933) was a 27 years old woman on whom repeated bladder plastics had failed. Patient had a classical caesarean section at term followed by tubal ligation.

Damm's case had congenital prolapse along with the exstrophy of the bladder, plastic operation in childhood had failed. She had a term stillborn baby. Version and perforation of after-coming head was done.

Osterguard reported a case of a 39 years old female, who had no previous surgery done on her. She delivered a term baby vaginally, complicated by a post-partum prolapse.

Dawson (1923) stated that all patients with exstrophy of the blad-

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ho become pregnant should be vered by elective caesarean section, the reason being to avoid genital prolapse after vaginal labour. Certainly a large percentage of the reported cases who delivered vaginally developed prolapse. But it is of interest to note that three cases of exstrophy of bladder had prolapse when they were nulliparous. One of these three became pregnant later on.

Another report of interest was made by McKenzie (1935). A patient had a split pelvis but no exstrophy of the bladder. For obstetrical reasons caesarean section was performed. During laparotomy bladder was found under the skin, incorporated into anterior abdominal wall. This indicates that this embryonic maldevelopment may be as mild as not to cause any symptoms, to a more severe form to become incompatible with life.

Ledfors and Lansing (1966) reported a case, who had exstrophy of the bladder. Transplantation of ureters into the colon was done at an early age and a repair of the lower abdominal wall at a later date. She was pregnant twice and aborted both times. She then had prolapse of the uterus and some renal disease.

Case Report

The patient, D.C., 25 years primigravida, was admitted on 13th July 1965. Her complaint was dribbling of urine since birth. She was 36 weeks pregnant. Her menstrual history revealed that she had her menarche at the age of 13 years. Her past menstrual cycles were menorrhagic. She used to have profuse flow for 7 days with an interval of 15 days. During these days she used to have spasmodic dysmenorrhoea. Her present menstrual cycles are comparatively normal. She has moderate flow for 4-5 days with interval of 28 days without any dysmenorrhoea. Her last menstrual period was on 28th October 1964, expected date of delivery, 4th August 1965. She was married one year ago.

On examination of the patient it was found that she was an average built woman, 5 feet 2 inches tall, weighing 120 pounds. Her conjunctivae were pink, tongue moist, gums healthy and there was no oedema. Systemic examination of gastrointestinal, respiratory, cardiovascular as well as central nervous system did not reveal any abnormal finding. Her blood pressure was 120/80 min of Hg on admission and remained constant till her labour.

Examination of the abdomen revealed that uterus was enlarged to 36 weeks' size, with a live foetus as breech presentation. There was no umbilicus and posterior wall of the bladder was seen in the infraumbilical region with ureteral opening on both the sides. There was continuous dribbling of urine and at times spurts of urine came out. The bladder mucosa was congested and red. There was no mons pubis. Labia majora were normal. Labia minora were seen on both sides, they were thick, oedematous and nodular. In between labia minora there was a transverse slit which was the opening of the vagina. Radiologically a split pelvis was found. (Figs. 1 & 2).

Her past history did not reveal any urinary disease. Elaborate interrogation showed that she had poor sense of sanitation. She utilised domestically used cloth pieces during menses. It used to traumatise the bladder area and so movements were very painful. Hence she used to remain recumbent during menstruation. Her husband was a bus conductor. He knew about her condition prior to their marriage. They had frequent sexual intercourse. Penetration was never complete and the act was always painful. Coitus occurred with the husband in the dorsal position. Her mother had experienced no difficulty in her labour, which was conducted by a dai. She has 5 brothers and 3 sisters but there was no history of any congenital abnormality in the family.

She was kept under observation. Time passed uneventfully till 18th August 65,

when an elective classical caesarean section was done. A female baby of 5 pounds and 10 ozs. was born; she was given 300 c.c. of blood in immediate post-operative period. Post-operatively she developed a faecal fistula at the lower end of the abdominal incision on the 8th day, which healed spontaneously.

In October 66, the patient again presented herself with the history of amenorrhoea of 3 months. Last menstrual period was on 5th July 1966. Her expected due date would be 12th April 1967. Her history revealed that her first baby died at the age of 10 months due to diarrhoea. On examination, it was found out that she was 12 weeks pregnant. In the antenatal period, on a number of occasions the breech was the presenting part. Haemoglobin readings were around 60 per cent. No symptoms of toxaemia developed.

On 24th March '67 she was admitted with the complaint of pain in abdomen. On examination there was a breech presentation. The scar of previous caesarean section was healthy with no overstreching or tenderness. Internal examination revealed that the cervix was one finger dilated, membranes intact, breech was presenting above the brim. She was kept under observation upto 8th April 67. On 9th April at 5 A.M. she complained of labour pains. Per abdomen it was found that the uterus was contracting and vertex was the presentation. Hence it was decided that she should have a vaginal labour.

At 8.30 A.M. internal examination revealed that the cervix was half dilated, taken up, well applied to the vertex, biparietal diameter was just at the level of the ischial spines, membranes were absent.

Left mediolateral episiotomy was done; patient delivered at 11-45 a.m. on 9th April, a male baby weighing 6 lb. 14 oz. Placenta came out spontaneously after 5 minutes. After the delivery of the placenta patient had profuse bleeding. The haemorrhage was both atonic and traumatic. There was a lateral cervical tear on the right side. Few superficial lacerations at the introitus were present; 300 c.c. of blood, group 'B' was given. The puerperium was uneventful.

Comments

Exstrophy of the bladder is characterised by absence of the anterior abdominal wall, infraumbilically. The anterior wall of the bladder is also absent. Posterior wall is everted. The ureters directly open on the surface, spurting out urine in a rhythmic manner. Because of urinary incontinence, continuous bathing of the genitals with urine occurs. Sexual activity is frequently distasteful to both the partners. However, in rare instances pregnancy has occurred.

Pubic rami are widely separated causing a spilt pelvis of a juxta-major variety. The two bellies of the recti muscles diverge in the lower portion there by producing a triangular defect. The perineum is shifted forwards. Along with the exstrophy of the bladder, following genital abnormalities are associated. There is a complete absence of urethra, the clitoris is cleft or is completely absent. There is a wide separation of labia with the exposure of vaginal orifice. The vagina is short, often double, the cervix protrudes up to the vaginal orifice. At times the uterus has been found to be of a bicornuate type. The degree of this anomaly varies from a small exposure of the trigone to a complete exposure of the posterior bladder wall.

There are three groups of hypothesis advanced as to the cause of extrophy of bladder by Otto (1948).

1. *Pathological hypothesis:* The pathological group claims that certain toxins from endometritis or any other irritation or infection cause a separation of the lateral walls of the caudal portion of the embryo. Keith

and Von Gildern (1924) advanced this theory and according to Von Gildern the arrest affects the parts which have not completed their embryonal development at the onset of infection. When the development recommences certain primitive embryonal structures are maintained and developed as such and those that should have undergone differentiation fail to do so, Velpeau and Phillips are of the opinion that ulcerations of the anterior abdominal wall involves the symphysis pubis with necrosis and its separation as an end result.

2. *Mechanical hypothesis:* The supporters of this group claim that exstrophy is caused by an intra-uterine rupture of a normally formed bladder. They are of the opinion that urethra is closed, not allowing any urine to come out of the bladder, thus leading to retention of urine. There is increased intravesical pressure in a greatly distended bladder. It is pushed forwards separating recti muscles and also the pubic bone until the bladder finally ruptures and forms adhesions to the torn abdominal wall. This "obstruction" which closes off the urethra has, however, never been anatomically demonstrated.

3. *Embryological:* Embryologists are at a greater variance in their views as to the cause of exstrophy of the bladder than are the other two groups. Wyburn (1937) in a study made on embryos came to the following conclusions. (1) At an early stage, the cloacal membrane is a relatively large area of contact, ectoderm and entoderm extending some distance along the allantoin diverticulum. (2) The allantoic cloacal membrane is later on obliterated by the mesoderm pressing in towards the midventral line between the ectoderm and entoderm. (3) Extravasation of the bladder is due to mesodermal deficiency, particularly the process of secondary mesoderm arising from the hind end of the primitive streak, following on which there is a persistence of the primary extensive cloacal membrane, impaired development of the muscular coat of bladder, of the symphysis pubis and of the formation of the external genitals and the infraumbilical portion of the anterior abdominal wall. (4) Epispadias is a mesodermal error in a minor form. Keibel believed that this condition resulted from persistent open blastopores.

Out of these three, the third one has some basic foundation to explain the exstrophy of the bladder. It is believed to be due to a mesodermal deficiency, particularly of the process of secondary mesoderm arising from the posterior end of the primitive streak. Many other malformations, particularly of reproductive systems, are associated with it, but these women usually retain their power of reproduction.

There are various gradations of the exstrophy—

(a) *Superior vesical fissure* in which union of the pubis is normal, but there is a defect in the upper part of the bladder.

(b) *Inferior vesical fissure* in which symphysis pubis is normal, but there is a rent in the lower part of the bladder.

(c) *Complete exstrophy* in which there is no symphysis pubis. Various

other malformations are also associated with it.

Symptomatology: The stellar symptom is urinary incontinence. Due to the absence of the anterior abdominal wall the posterior wall is seen and the ureteric openings are seen spurting out urine.

The mucosa of the bladder is infected, congested and ulcerated. These patients carry persistently an aromatic odour which causes considerable embarrassment and discomfort. They may exhibit the emotional disturbances associated with urinary incontinance. Their gait is duck-like due to separation of symphysis pubis and outward rotation of femora. Inguinal and femoral hernias are usually associated. Most of them lead a sexual life.

Conclusions

With increasing improvement in surgery and operations the complications of ectopia vesicae in pregnancy will be more frequently encountered.

The management of pregnancy complicated by ectopia vesicae has not received any special considerations. More of those who receive the benefit of modern surgery will live to bear children. The reproductive capacities are interfered with but little. Before allowing these patients to become pregnant, excretory urography to test renal function must be done.

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Figs. on Art Paper VI