IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 5 Ver. VI (May. 2015), PP 07-09
www.iosrjournals.org

Ectopia Vesicae with Deep Clefting of Toes

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***Abstract*:** *Bladder exstrophy (also known as Ectopia vesicae) is a congenital anomaly that exists along the spectrum of the exstrophy epispadias complex and most notably involves protrusion of the urinary bladder through a defect in the abdominal wall. A part of the wall of the bladder in front is absent and the inner part of the bladder is exposed. It is a rare condition affecting one in 50,000 people. Cleft foot also a rare congenital anomaly in which the foot didn't develop properly during fetal development. This causes the affected foot to have missing toes, a V-shaped cleft and other anatomical differences. A deep congenital cleft between the 1std and 2nd digits of the feet, due to an absent central digital 'ray'. Congenital malformation have ever since been throwing a challenge to both scientists and to the parents as they are neither totally curable nor the babies are mercilessly abandonable. In ancient days such anomalies babies were considered to be curse by nature and were mostly abandoned. These deformities were even described in ancient mythology. The present study is about the incidence of gross abnormalities in the growing fetuses in relation to environmental causative factor, with a hope to arrive at reasonable conclusions regarding the influence of environmental factors on the fetuses, though mindful of the influence of the genetic predisposition. This study may help to know the probable cause and to prevent and treat such malformations.*

Key words: *Absence of the anterior wall of the bladder, Ectrodactyly, Epispadias of the penis, Fibrous band, a flattened pubo rectal sling.*

I. Introduction

Exstrophy of the bladder (ectopia vesicae) is a rare congenital anomaly with an incidence of about 1 per 50000. It is usually treated in the neonatal period thus presentation in the adult period is a rarity. The malignant potential is considerable and most cases are adenocarcinomas but squamous carcinomas do occur. Ectopia vesicae is no new disease and it is interesting to consider the methods of treatment adopted by our predecessors. On April 4th Ectopia vesicae occurs about once in every 50,000 births; males are affected seven or eight times as often as females. 1849, at the Medico Chirurgical Society of Edinburgh "Dr.Mackay exhibited a child a few weeks old with congenital exstrophy of urinary bladder.

In complete ectopia the absence of the pubic symphysis and linea alba leaves a gap in the infra-umbilical part of the abdominal wall through which the extroverted bladder bulges. The pubic tubercles, to which the recti abdominis are attached, are widely separated, and the inner margins of the recti can be felt or seen to form the boundaries of a triangular opening whose base is at the bladder and whose apex reaches above the umbilicus. This latter structure may appear to be absent, but its upper half can usually be distinguished as a scar at the apex of the bladder swelling. A scarred margin, as of a hernial ring, can be felt to extend from it around the bladder to the pubic tubercles which are united by a fibrous band.

Incomplete absence of the anterior wall of the bladder leaves the posterior wall to project as a soft red swelling below the umbilicus. It is tender to touch; it becomes tense when the child strains or cries, and it can be reduced by pressure. When the upper half is lifted up the trigone can be observed, the ureteric orifices can be picked out and spurts of urine seen from them. Below the trigone the urethra lies open, there is no urinary control and the whole lower area is constantly wet.

In the male the penis is cleft and is usually, but not always, rudimentary, it may be hidden by the overhanging bladder. The scrotum is small and sometimes bifid and the testes are often maldescended. Other congenital abnormalities may be present such as umbilical or inguinal herniae and there may be congenital anomalies of the upper urinary tract, the symptoms are only too apparent, there is a constant leakage of urine from the ureters, it runs down the legs and perineum and soaks the clothes so that the unfortunate patient is continually wet and frequently smelly. He becomes a social outcast and if he can undertake any work at all it is usually out of doors.

II. Materials & Methods

In the present study 530 Infants and fetuses obtained from Rajiv Gandhi Institute of medical sciences general Hospital, Srikakulam from department of Obstetrics and Gynaecology, and private hospitals during the year April 2013 to March 2015.

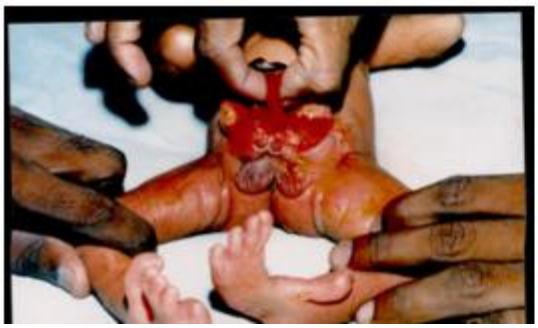
Weight of the foetus, Head circumference, crown - rump length, both right and left arm forearm length, groin to tip of the first hallux and right and left foot lengths were taken with help of Thread, vernier calipers & Scale for the present study.

III. Observations

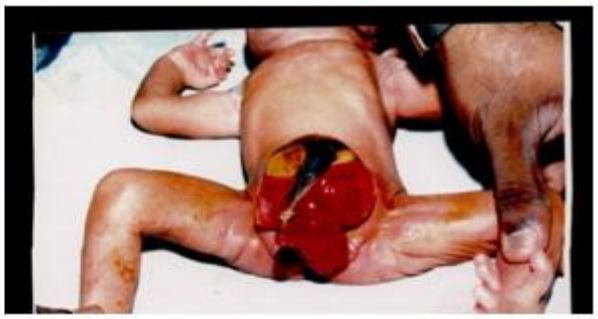
Among the 530 new born 17 were having congenital abnormalities out of which, only one new born is with Ectopic Vesicae, with Deep Clefting of toes and eventration of anterior abdominal wall, and the measurements of foetus are as follows:

Weight of the foetus: 2500 grams
Head circumference: 40cms
Crown rump length: 42 cms
Arm length:
Right: 10cms
Left: 10cms
Forearm length:
Right: 9cm
Left: 9cm
Femur length: 12cms
Foot length
Right: 8.45cms
Left: 8.45cms

IV. Figures& Tables



Eventration of anterior abdominal wall



Ectopia vesicae with deep clefting of toes

V. Discussion

Its presentation is variable, often including abnormalities of the bony pelvis, pelvic floor and genitalia. The underlying embryologic mechanism leading to bladder exstrophy is unknown, though it is thought to be in part due to failed reinforcement of the cloacal membrane by underlying mesoderm.

Complete exstrophy - In this case, the ends of the pubic bones in front of the pelvis are widely separated (normally they fuse before birth). The abdominal muscles in front also separate, the umbilicus may be absent, and there may be an umbilical or inguinal hernia associated with it. In males, the urethral opening at the tip of the penis is placed on the upper part of the shaft of the penis and the penis curves upwards; this is known as epispadias and almost always accompanies this condition.

Incomplete exstrophy- Here, the pubic bones are fused while the external genital organs may be normal or there may be a minor defect where the urethra opens at the tip of the penis.

Cleft foot is a rare congenital (meaning your baby was born with it) anomaly in which the foot didn't develop properly during fetal development. This causes the affected foot to have missing toes, a V-shaped cleft and other anatomical differences. Left foot (lobster foot) characterized by the absence of 2 or 3 central digital rays of the foot. One or more toes and parts of their metatarsals are absent, and often the tarsals are abnormal. Human and mouse mutations for lobster foot will reveal apical ectodermal ridge (AER) functions and the pathogenesis of this entity. Amongst the congenital anomalies, central deficiency or cleft hand is relatively rare. The association of cleft foot with cleft hand is an even rarer occurrence. A deep congenital cleft between the

1std and 2nd digits of the feet, due to an absent central digital ray the lobster claw deformity is more common in the foot, and is often accompanied by syndactyly, it classically occurs in ectodermal dysplasia—EEC syndrome—ectrodactyly, ectodermal dysplasia, and cleft palate, and is associated with organ defects. Cleft foot is very rare, affecting fewer than 1 in 1,000,000 babies.

The classic manifestation of bladder exstrophy presents with:

- A defect in the abdominal wall occupied by both the exstrophied bladder as well as a portion of the urethra.
- A flattened puborectal sling.
- Separation of the pubic symphysis.
- Shortening of the pubic ramii.
- External rotation of the pelvis.

Females frequently have a displaced and narrowed vaginal orifice, a bifid clitoris, and divergent labia. The cause is congenital due to failure of development of the front wall of the urinary bladder and along with it failure of the pubic bones to fuse. The abdominal muscles in front also fail to develop causing a defect in the abdominal wall. It is seen more often in males.

Symptoms and signs of ectopia vesicae:

- The sides of the bladder are attached to the skin of the abdomen in front.
- The inner lining of the bladder has a tendency to bleed easily and get infected as it is exposed to the air.
- Urine which enters the bladder from the kidneys via the ureters will leak outside the body causing further infection and unhygienic conditions.
- The umbilicus may be absent, and there may an umbilical hernia (some tissue protruding through the umbilical opening) or there may be an inguinal hernia (tissue protruding through a defect or weakness in the lower abdominal wall near the groin.
- Epispadias of the penis is seen. The urethral opening which is normally at the tip of the penis is somewhere on the upper side of the shaft of the penis. This causes difficulty in urinating. The penis is usually shorter and broader than normal.
- The testes may be in another area, not in the normal position in the scrotum (ectopic testes)and the scrotum may not be completely developed.
- In females, the clitoris may be cleft (as if divided into two)and the two labia minora are separated. The uterus and vagina may be abnormal.
- The anal opening is loose or lax.

Complications of ectopia vesicae:

- Hydronephrosis - The kidneys may enlarge and have multiple bubbles in it called hydronephrosis; this happens when there is an obstruction to the passage of urine in the ureters.
- The inner lining of the urinary bladder may change causing cancer.
- Infection in the bladder spreads upwards to the ureters and kidneys. Recurrent kidney infection may occur.
- The person always smells of urine, and it is socially embarrassing.
- The child goes into renal failure which may be fatal.

VI. Conclusion

Fundamentally it is difficult to know the actual incidence of congenital malformations as an unknown number of abortions may also have the deformed fetuses. So manifestations of deformations miss the records since the manifestations of deformities may take place at a later age.

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