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CHORDEE WITHOUT HYPOSPADIAS IS A RARE MORPHOLOGICAL ABNORMALITY-A CASE REPORT

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ABSTRACT

Ventral and lateral curve of penis are known as chordee and torsion respectively which can be associated with or without hypospadias. Chordee without hypospadias is a rare morphological abnormality of the penis capable of interfering with sexual function and observed either at birth or in older boys after retraction of prepuce or circumcision. Devine and Horton described three classes of underline defects. Type I has deficiency of corpus spongiosum, bucks and dartos fascia and urethra covered with only skin and considered to be severe type. In Type II bucks and dartos fascia is deficient whereas in type III only dartos fascia is deficient. Herewith we are reporting a case of 5 years old child with Type III chordee without hypospadias with review of relevant literature.

Key Words: Chordee without Hypospadias, Penile Torsion, Orthotopic Meatus

INTRODUCTION

Congenital chordee and penile torsion are commonly observed in the presence in hypospadias but may be associated with Orthotopic meatus. Surgical management of congenital curvature without hypospadias can present a challenge to the pediatric urologist (Montag and Palmer, 2011).

Many of these patients had straight erections as children but a ventral curvature developed as they achieved puberty. Surgical therapy begins with a circumcising incision and reflection of the skin (degloving of penis) to expose the shaft of the penis which can lead to satisfactory correction of Type II and Type III chordee (Devine et al., 1991).

After degloving of penis several surgical techniques (plication, excision, and graft insertion) are currently employed to repair penile curvature. Recent neuroanatomical studies of the developing fetal penis have shown that the dorsal nerve branches from the 11 and 1 o'clock positions to the 5 and 7 o'clock positions, being absent in the midline. Since the neuroanatomy is similar in both the hypospadiac and normal penis, we now recommend performing penile straightening in both hypospadiac and non hypospadiac patients with significant curvature by the placement of placation sutures at the 12 o'clock position. Placement of dorsal midline plication sutures corrects curvature without risk to the underlying nerve structures (Mingin and Baskin, 2002).Extensive plication of severe degree of chordee may result in significant shortening of penile length. Dorsal diamond shape wedged excision with transverse repair of bucks' fascia (Nesbit, 1966) or application of ventral dermal draft has been described in literature (Devine and Horton, 1975).

Etiology

While the causes of chordee are evident in boys with hypospadias, its precise etiology, as well as that of torsion, in the absence of hypospadias, remain incompletely understood. Recent studies have furthered our understanding of the possible etiology and previously proposed explanations have been revised, which largely resulted in changes in surgical techniques. The current surgical strategies are largely successful in correcting the penis with abnormal curvature. Mettauer, 1842 first defined its etiology in 1842 as "skin tethering implicating subcutaneous tissue for cause of penile curvature". Since chordee was first described in boys with hypospadias, the leading theories included: (1) abnormal development of the urethral plate, (2) presence of abnormal fibrotic mesenchymal tissue at the urethral meatus, and (3)

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ventral-dorsal corporal disproportion (Kaplam and Lamm, 1975). Recently, a large series of congenital chordee without hypospadias was evaluated and revealed that the etiology can be evenly divided among skin tethering, fibrotic dartos and Buck's fascia, and corporal disproportion

Devine and Horton, 1973 described three classes of underlying defects. In type I the corpus spongiosum, Buck's fascia and the dartos fascia are deficient, and the urethra lies immediately beneath the skin. In type II the Buck's and dartos fascial layers are deficient. Type III is characterized by a normal corpus spongiosum and Buck's fascia, with chordee being produced by an abnormal dartos fascial layer. The most severe defects are seen in patients with type I chordee without hypospadias.

Embryology

Recent studies show that ventral curvature is a normal stage of embryogenesis and, therefore, chordee without hypospadias may represent arrested penile development. The development of the penis and the urethra take place early in fetal development. The bilayered cloacal membrane (ectoderm and endoderm) becomes flanked by cloacal folds early in the 5th week that meet anteriorly to form the genital tubercle. The cloaca then divides into an anterior urogenital sinus and a posterior anorectal canal. The mesenchymal folds flanking the urogenital sinus become urogenital folds. The corporal bodies, connective tissue, and dermis of the penis are derived from mesodermal cells. The elongating phallus is covered with skin derived from ectoderm. The molecular mechanisms that regulate this mesenchymal differentiation likely depend on epithelial-mesenchymal interaction. Human fetal studies reveal that a ventral curvature is a normal state of penile development at the 16th week of gestation that resolves during the 20–25th week (Kaplam and Lamm, 1975).

CASES

5 year old child brought by parents in surgery OPD of BPS GMC for women Khanpur Kalan, Sonepat with complaining of wettings of leg and clothes due to downward and lateral deviation of urinary stream during micturition .On examination significant degree of chordee with left lateral torsion of glans was revealed (Fig. 1 and 2).



Figure 1: Preoperative picture Illustrating severe chordee

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Figure 2: Showing lateral torsion in same patient



Figure 3: Demonstrate the correction of chordee after degloving, dorsal plication was sufficient to correct this residual chordee

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After routine investigation patients was prepared for surgery. During surgery after catheterization of urethra a racket shaped incision given on corona and degloving of penis up to pubic tubercle was done. Up to 90% of chordee correction was noted that was checked by reproducing artificial erection by normal saline injection in corpus spongiosum (Fig. 3). Correction of this residual chordee we could achieved by two dorsal plication suture at the site of maximum curve and torsion was corrected by realignment of median raphe. During postoperative period catheter was kept for five days and after removing Catheter good results were achieved in the patient;

RESULTS

Excellent straightening of the phallus was achieved intraoperatively in the patient. These milder forms of chordee without hypospadias (Horton and Devine type III and lateral deviations of the penis) all benefited from Nesbit dorsal plication of the contralateral penile tunica albuginea and were corrected with a single operation.

Conclusion

Though chordee without hypospadias is a morphological abnormality and caused significant apprehension in parents regarding day to day activity and sexual development of child. Careful evaluation and management of abnormality may lead to good results with minimum complication.

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