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"PENILE DUPLICATION" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT
Penile duplication is a very rare common congenital anomaly. It occurs alone, but the more prominent examples are usually associated with anomalies of the remnants of the genito-urinary tract and the lower gastrointestinal tract. The quantity of the anomaly ranges from a partial duplication of the glans to two separate penis situated at some distance from each other. A demonstrated classification is presented. The cause is unidentified but is best explained by an early disturbance in the embryologic development of the hindgut and ventral abdomen. Treatment must be personalized and consists of various processes to restore normal appearance and function.

Introduction

Duplication of the penis or diphallus is a rare common congenital anomaly that occurs once in every five to six million Living births. Nearly hundreds of cases has been reported since the first case reported by Weckerin 1609.[1] Neugebauer in 1898 and Nesbit and Bromme in 1933 studied cases in the literature. The amount of duplication and the number of associated anomalies vary significantly, extending from a double glans rising from a common shaft with no other abnormality to widespread duplication of the phallus accompanied by the several anomalies, such as bifid scrotum, ectopic scortum, hypospadias, imperforate anus, bladder exstrophy, double bladder, colon duplication, and vertebral deformities.[2] Embryo logically the duplication of penis malformation arises from each separation" of the pubic tubercle, where each penis will have only one corporal body and urethra, or "cleavages" of the pubic tubercle where each penis will have two corporalcavernous bodies and urethras. Diphallus has been categorized in different ways, such as glandular, concealed, bifid and complete, hemidiphallus and triple penis. Schneider classified diphallus in three different groups: diphallia of glans alone,[3] complete diphallia and bifid diphallus recently a fourth category of pseudodiphallia has been added. The majority has single corpus cavernosum in each organ. Duplicated urethras usually may be related with diphallus.[4] Children's born with this disorder are at an improved risk of child death because of the deficiencies and infections that are accompanying with it. [5] Duplication of penis develops around 23–25 days of gestation because the genital tubercle fails to fuse properly. ·Treatment should always be individualized. The deformities that are potentially dangerous should be resolved first.

Incidence

It is estimated to occur in one out of five million lives births[6]. It is usually accompanied by other congenital anomalies such as renal, vertebral, hindgut, or anorectal duplication also there is a high risk in spina bifida. Penile duplication occur nearly 1 in 1000 peoples

Ontogenesis of the normal fate of Mullerian duct

By the eighth week of the gestational age, the Leydig cells of the developing testis are capable of manufacturing [7] testosterone under the stimulations of human chorionic gonadotropin. Circulating testosterone causes the development of tissues with testosterone receptors. [8]Normally, testosterone is taken into the cell and bound by the androgen receptor. In the cytoplasm, it is converted to dihydrotestosterone by this enzyme five -alpha-reductase[9]. Dihydrotestosterone is at least four times more effective than testosterone. The genital prominence, an exterior mound arising between the umbilicus and the tail, and it is prepared up of the genital tubercle and by the genital swellings[10]. The urogenital sinus orifice lies at the base of the genital tubercle, between the genital swellings. [11]And these structures forms identically in males and females embryos up to seven weeks gestational age[12]. At nine weeks of gestational age, and under the stimulus of testosterone, the genital tubercle starts to elongate. [13]In addition, the genital swellings also called the labio-scrotal folds expand and rotate posteriorly.[14] As they meet, they originate to fuse from posterior to anterior. For example the genital tubercle becomes longer, two sets of tissue folds develop on its ventral surface on either side of a developing gutter, the urethral groove.[15] The additional medial endodermal folds will fuse in the ventral midline to form the male urethra. The extra lateral ectodermal folds will fuse over the developing urethra to form the penile shaft skin and the prepuce. [16]As these two layers fused from posterior to anterior, they leave behind a skin line: the median raphe. By thirteen weeks, the urethra is almost developed. A disc of ectoderm forms just proximal to the developing glans penis. [17]This skin progresses over the corona glandis and finally covers the glans completely as the prepuce or foreskin.

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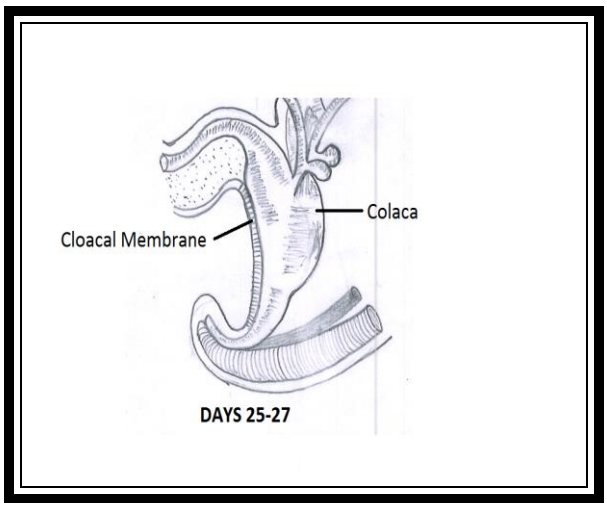


Fig 1. The schematic representation shows the normal development of glans penis on 25-27 days embryo.

In 3rd week development .the cloacal membrane, is gradually surrounded by mesenchyme from the primitive streak. This mesenchyme forms a pair of elevations, the cloacal folds, which fuse with each other in front of the cloacal membrane to form the cloacal eminence.

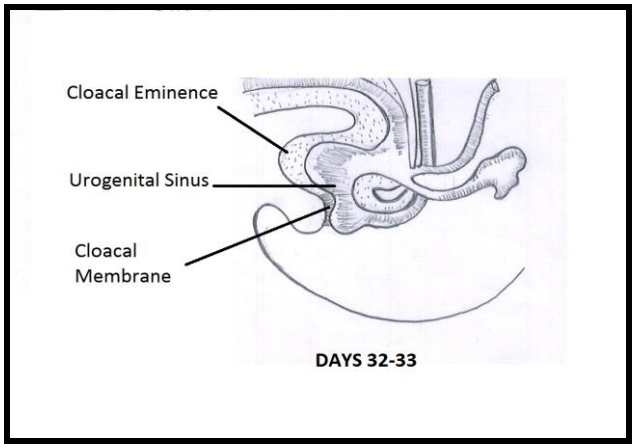


Fig 2. The schematic representation shows the normal development of glans penis on 32-33 days embryo.

In 4th week development the cloacal eminence elongates and forms the genital tubercle

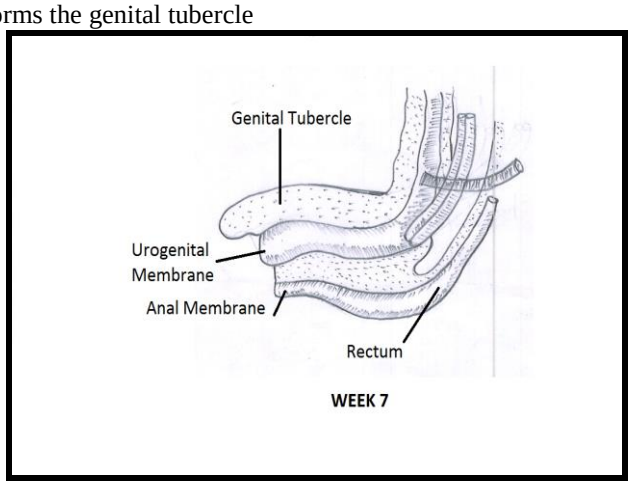


Fig 3. The schematic representation shows the normal development of glans penis on week 7.

In 7th week development urorectal septum into the urogenital and anal membranes; the cloacal swellings also are split into the genital or urethral folds. The membranes rupture a week later to form the urogenital and anal openings elongates by week 7 to form a *phallus*, which in turn will form the future penis.

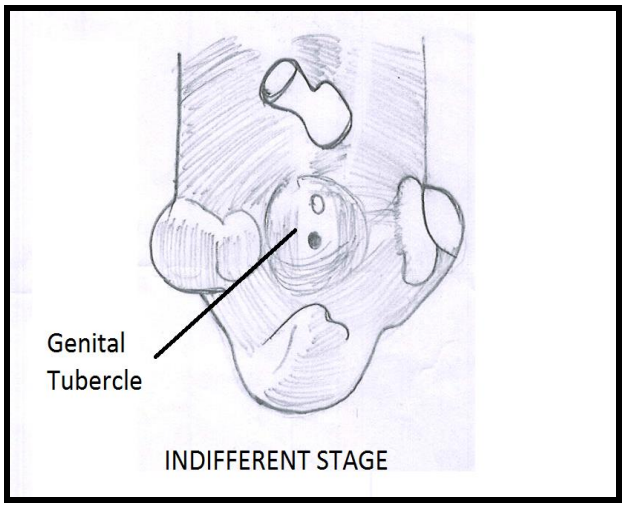


Fig 4. The schematic representation shows the normal development of glans penis during indifferent stage.

In the indifferent stage development the cloacal eminence elongates and forming the genital tubercle.

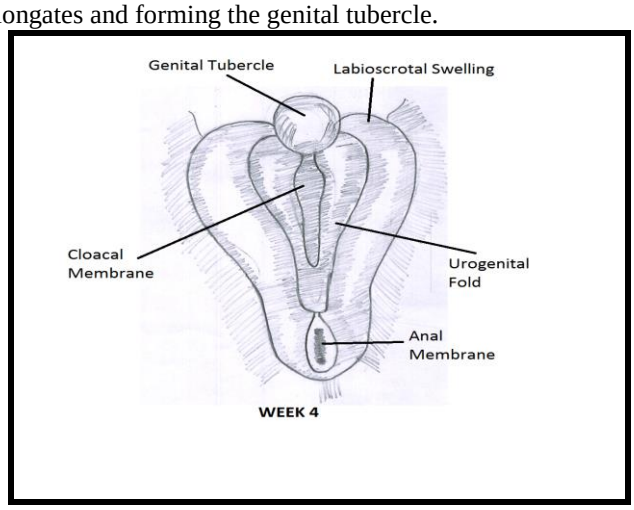
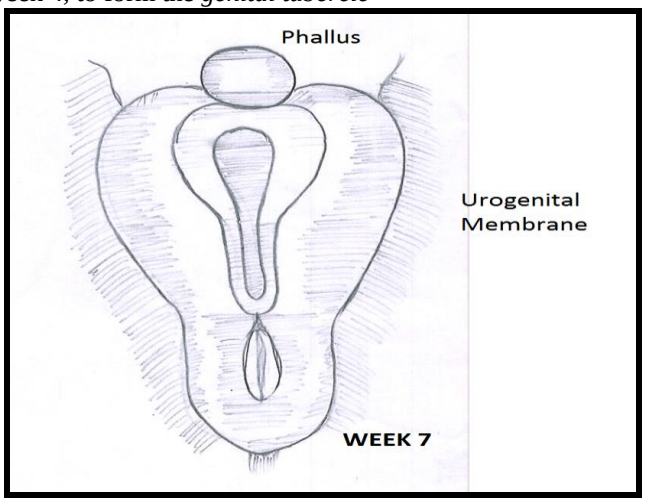


Fig 5. The schematic representation shows the normal development of glans penis on week 4.

In 4th week development The cloacal eminence elongates, by week 4, to form the *genital tubercle*



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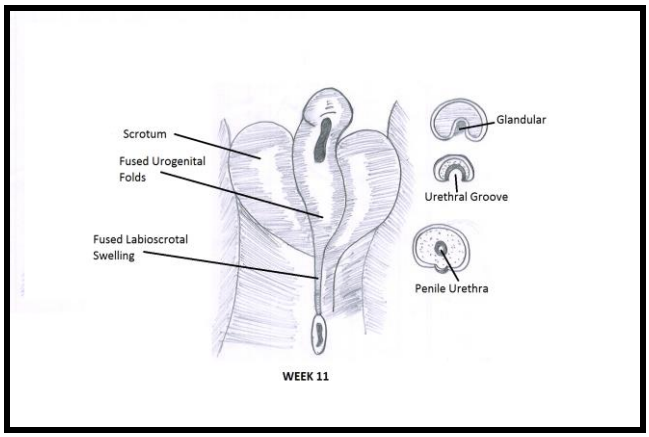

Fig 7. The schematic representation shows the normal development of glans penis on week 11.

In 11 week development .the labioscortal or genital swelling grow toward each other and fuse in the midline to form the *scrotum* Both the scrotum and penis bear the signs of their early formation through closure of the urogenital groove as is evidenced by the median raphae

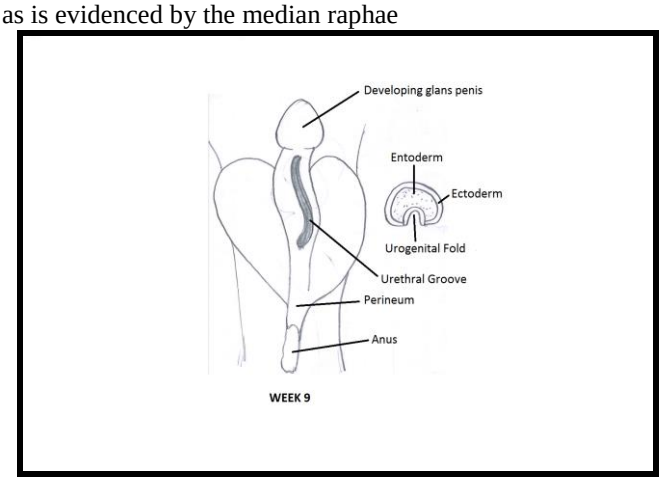

Fig 8. The schematic representation shows the normal development of glans penis on week 9.

In the 9th week development the urethral groove is lined by an extension of the entoderm from the phallic portion of the urogenital sinus and is continuous with the urogenital opening .At the base of the groove, the entoderm thickens into a urethral plate The posterior portion of the genital swellings thickens to form the scrotal swelling

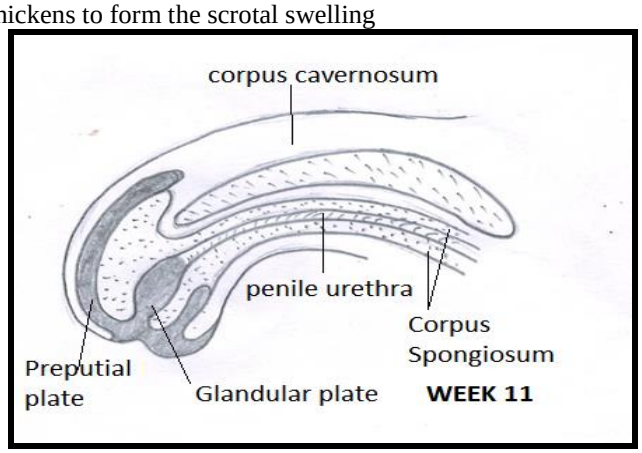

Fig 9. The schematic representation shows the normal development of glans penis and penile urethra on week 11.

In 11th week of development, the penile urethra ends blindly just before the end of the penis and is surrounded by a mass of erectile tissue of mesenchymal origin, the corpus cavernosum urethrae or spongiosum. This erectile tissue forms the end of the penis, the glans penis

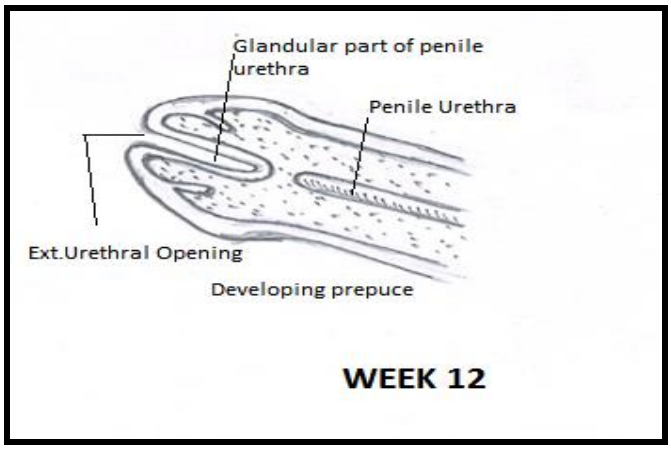

Fig 10. The schematic representation shows the normal development of glans penis on week 12.

In the 12th week development the *glandular* of the penis. Closure of the groove in the glans moves the urethral opening to the tip of the glans and joins the 2*urethra*, on the ventral part of the glans that is continuous with the urethral groove in the body

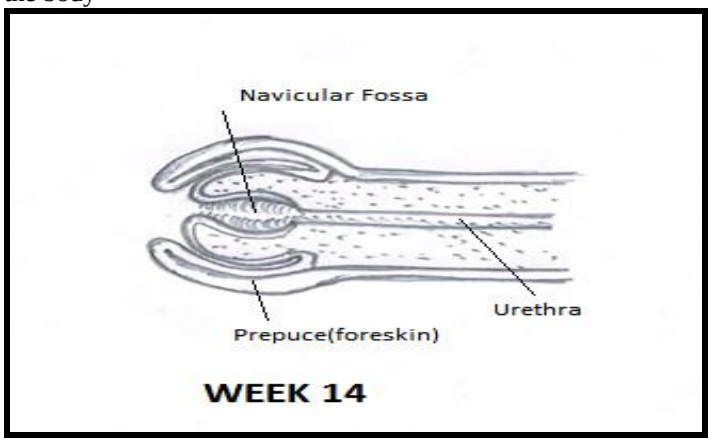

Fig11. The schematic representation shows the normal development of glans penis on week 14.

In 14th week of development, the second invagination is circular and is called the preputial epithelial plate. Cleavage of this plate before birth separates the glans penis from the prepuce or foreskin. The latter is a fold of skin at the tip of the penis which, during week 12, grows over the glans and surrounds it by week 1 It is fused to the glans and not retractable at birth, but breakdown of the fused surfaces normally occurs during infancy.

Ontogenesis of the abnormal fate of Mullerian duct

Hollow et al, reviewed the embryogenesis of penile duplication suggested that[18] the complete penile duplication could be resulting from the longitudinal duplication of infraumbilical cloacal membrane before by the fourth week of gestation [19], the subsequent of the mesodermal migration allowing two the separate , complete set of genital tubercle, genital folds and genital swellings to develops.[20] The combination of the genital folds and swelling may not. However, be entire normal, accounting for the finding that one of the two urethras may be blind pits or else be steno tic.[21] Rarely, one or both urethras also may be hypospadiac or epispadiac. A wide range of scrotal abnormalities may be present because of the duplicated clocal membrane is likely to be widened structure;[22] the" wedge" effect could result in the stigma of the covered exstrophy. In some patients, the abnormalities suggested from a partial caudal duplication involving the derivatives of the allantois, hind gut and neural tubes.[23].

It is thought diphalia occurs in the fetus between the twenty third and twenty five days of gestation when chemical stress, or injury or malfunctioning [24]homeoboxgenes obstruct proper function of the caudal cell mass of the fetal mesoderm as the urogenital sinus separate from the genital tubercle and rectum to form penis.

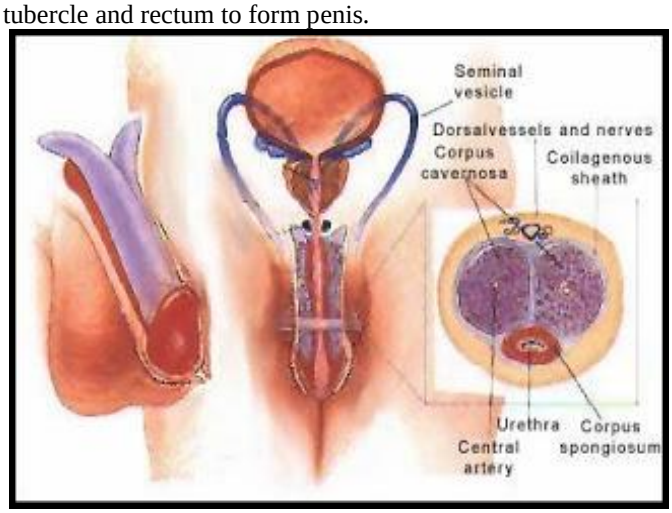

Fig12. Following diagram represents the abnormal development penis (duplication of penis).refer phrase 3 to 8.

Discussion

Embryologically diphallus is actually arise due to abnormal growth of pubic tubercle.[25] It could be due to either separation of pubic tubercles or cleavages of pubic tubercle.[26] In previous case each phallus will have only one corporal body and urethra but in later case each phallus will have two corporal cavernosus bodies and urethras[27]. Caudal duplication syndrome has been offered to explain the related duplication of hindgut, bladder and urethra. [28]Schneider categorised diphallus in three groups; diphallus of glans alone, complete diphallus and bifid phallus Vilanova and Raventos have additional a fourth category pseudodiphallus.[29] The urethra shows a great range of differences from functioning double urethra to complete absence of urethra in each penis. [30] Advanced classification currently widely accepted includes two main groups: true diphallia and bifid phallus. [31]These groups are additionally divided into partial and complete duplication. [32]True complete diphallus means each of the phallus has two corpora cavernosa and corpora spongiosum.[33] When this phallus is undeveloped or small it is called true partial diphallia. When there is only one corpora cavernosa is present in each phallus it is called bifid phallus.[34] When gradation of separation is to base of shaft it is called complete bifid phallus whereas if it is upto just glans, it is known as partial bifid phallus. Our circumstance was complete true diphallus.[35] True diphallia is more often associated with severe malformations as compared to bifid phallus.[36] Associated abnormalities include genitourinary anomalies which can be hypospadias, bifid scrotum, duplication of bladder and urethra, renal agenesis, [37]exstrophy alone or exstrophy with vesicointestinal fistula. Gastrointestinal anomalies can be imperforate anus with or without recto urinary fistula or duplication of colon.[38] Musculoskeletal abnormalities can be diastasis of pubis, club foot, polydactyl or lumbosacral anomalies. [39]Penile duplication postures a difficult treatment problem in terms of medical, ethical and aesthetic decision making. [40]Through examinations are mandatory to reveal associated congenital malformations that is potentionally life menacing and require immediate surgical corrections. [41].

The treatment of diphallia is by removal of the duplicated non-communicating penis[42]. The treatment primarily depends on the type of associated congenital abnormalities as well as preserving continence and erectile function which means individualizing each case. [43]Surgical correction is personalized with the aims of achieving proper urinary continence, urinary stream and production with adequate cosmesis. [44]Though the isolated diphallus has been described in the various literatures,[45] the penis ay associated with the hypospadias meatus, pseudo phallus or hypo plastic urethra. [46]Our circumstance has been unique that there was complete diphallia. Phallus have two each corpora cavernousa and well developed corpora spongiosum.[47] Although one phallus was smaller than the other, urethra was patent in both of them. [48]Complete removal without erectile dysfunction and urinary dysfunction may be possible.

Conclusions

Penile duplication is a rare common congenital anomaly. [49]Systematic investigations are mandatory in all cases to expose essential congenital malformations that is theoretically life threatening and require immediate surgical correction. [50]Treatment should always be personalized according to the amount of penile duplication and the degree of the associated anomalies.

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