19a. MEGAMEATUS AND INTACT PREPUCE

Megameatus and intact prepuce (MIP) is a rare variant of hypospadias. MIP is caused by failure of ventral midline fusion of the distal end of the penile urethra in the coronal sulcus, and failure of ventral midline fusion of the fossa navicularis or glanular urethra.
The result is
a) wide urethral sulcus, on the ventral aspect of the coronal sulcus and glans penis,
b) absent frenulum,
c) intact prepuce.

Understanding the abnormalities in MIP.

Development of fossa navicularis navicular raphe and external urethral meatus:

a) mucosa of the vestibule/urethra, (endoderm) folds ventrally, and fuses in the midline to form the *fossa navicularis* or glanular urethra.
b) Simultaneously, the *wings of the glans penis,* covered by genital fold (ectoderm), grow ventrally and drape around the lateral and ventral aspects of the *fossa navicularis*, except in the ventral midline.
c) Ventral to fossa navicularis, the *vestibular/urethral folds* fuse to form the *navicular raphe.*
d) Distal to the navicular raphe, the *vestibula/urethral folds, remain unfused. The unfused vestibular/urethral folds join in the anterior commissure* of the vestibular/urethral folds. The unfused vestibular/urethral folds, tilt dorsally and line up with the vertically oriented elliptical opening of the fossa navicularis, to form the *labia of the glans penis*, joined by the *dorsal commissure* of the labia of the glans penis. The labia of the glans penis and dorsal commissure of the labia, form the *external urethral meatus.*

The frenulum, navicular raphe and labia of the glans penis:

The *quadrilaminar frenulum* is formed by midline fusion of the *vestibular/urethral folds ventral to the coronal sulcus*, proximal to the *corona* of the glans penis.
The *quadrilinear navicular raphe* is formed by midline fusion of the *vestibular/urethral folds ventral to the fossa navicularis*.
The (dorsal end of the free) *anterior border of the quadrilaminar frenulum* joins the *posterior end of the quadrilinear navicular raphe*.
The distal end of the quadrilinear navicular raphe (fused vestibular/urethral folds), joins the ventral ends of the labia of the glans penis (unfused vestibular/urethral folds), united at the dorsal commissure of the labia of the glans penis.

Dorsal plate of the fossa navicularis:

In the glans penis the corpus spongiosum of the penile urethra, is reduced to a narrow curved dorsal plate, that is adherent to the mucosal roof of the fossa navicularis. The dorsal plate of the fossa navicularis is homologous with *pars intermedia* adherent to the flat roof of the anterior section of the vestibule, in the clitoris.

The dorsal plate of fossa navicularis (m) and pars intermedia of the anterior section of the vestibule (f), covered on its upper surface by a layer of tunica albuginea, is attached to the groove on the ventral aspect of the (tunica albuginea) of the bicavernosal body.

Development of the glans penis, covered by the genital fold:

The glans penis is generated by spongy tissue of the dorsal plate of the fossa navicularis. The distal end of the dorsal plate generates the crescentic cap of the glans penis, over the conical end of the bicavernosal body of the penis. The (upper layers) of the lateral borders of the dorsal plate, generate the roof section of the glans penis, that covers the dorsal and lateral aspects of the bicavernosal body of the penis. The cap and roof of the glans penis are covered by an intimate layer of the genital fold (ectoderm). The (lower layer) of the lateral borders of the dorsal plate of fossa navicularis, generates the wings of the glans penis, covered by an intimate layer of genital fold (ectoderm).

In megameatus with intact prepuce, development of the glans penis and fossa navicularis is arrested, prior to the

Epithelial connections of the genital fold and vestibular/urethral folds:

a) Inferior borders of the genital fold reflect on the outer borders of the fused vestibular/urethral folds of the navicular raphe
b) Anterior borders of the genital fold reflect on the outer borders of the unfused vestibular/ urethral folds of the labia of the glans penis (Hart's line).
c) Ventral border of the genital fold over the pointed tip of the crescentic cap of the glans penis, reflects on the outer border of the dorsal commissure of the labia of the glans penis. (Hart's line)

Development of the prepuce:

The prepuce develops as *circumferential two layered fold of ectoderm* (genital fold) of the shaft of the penis. The two layered fold extends over the glans penis.
The ventral borders of the outer and inner layers of the prepuce derived from the *genital fold*, *reflect on the lateral borders of the preputial raphe* (fused vestibular/urethral folds).
On the outer layer of the prepuce, the posterior end of the preputial raphe, joins the penile raphe, which is ventral to the penile urethra (proximal to the coronal sulcus).
In the recess of the prepuce and coronal sulcus, the (proximal) end of the (quadrilinear) *preputial raphe,* on the *inner layer of the prepuce*, joins the posterior end of the (quadrilaminar) *frenulum,* which is *ventral to the penile urethra in the coronal sulcus.*

In the condition of megameatus with intact prepuce, the roof and cap of the glans penis develop normally, as does the prepuce.

Anatomy of megameatus with intact prepuce:

In the coronal sulcus

a) the *bicavernosal body* passes through the coronal sulcus.
b) *corpus spongiosum and vestibule/penile urethra* do not fuse in the midline, to form the penile urethra.
c) The *vestibular/penile urethral folds* do not fuse to form the frenulum.
d) The *ventral border of the genital fold* (ectoderm of the coronal sulcus), reflects on the *lateral borders of the unfused vestibular/penile urethral folds*

Corpus spongiosum of the penile urethra in the coronal sulcus is reduced to a *dorsal plate over the fossa navicularis* (vestibule/glanular urethra).

The lateral borders, and distal end, of the dorsal plate of fossa navicularis, generate the roof, and cap, of the glans penis.

The lower borders of the dorsal plate do not generate wings of the glans penis.

In the glans penis:

a) the *bicavernosal body* is covered by the roof of the glans penis. The conical tip of the bicavernosal body is covered by the crescentic cap of the glans penis.

b) the *vestibule/glanular urethra* (fossa navicularis) does not fold and fuse in the ventral midline to form the tubular vestibule/glanular urethra (fossa navicularis). *Fossa navicularis* forms a *flat mucosal roof* without side walls or floor.

c) the *vestibular/glanular urethral (fossa navicularis) folds*, *do not* fuse to *form the navicular raphe.*

d) the *ventral border of the genital fold* (ectoderm of the roof of the glans penis) reflects on the *lateral borders of the unfused vestibular/glanular urethral (fossa navicularis) folds*. The ventral point of the genital fold (ectoderm of the ventral point of the crescentic cap of the glans penis) reflects on the outer border of the anterior commissure of the vestibular/glanular urethral (fossa navicularis) folds.

Megameatus:

Result of failure to form the penile urethra in the coronal sulcus and failure to form fossa navicularis in the glans penis. The penile urethra (in the coronal sulcus) and the fossa navicularis in the glans penis, form a wide horizontal groove on the ventral aspect of the coronal sulcus and glans penis.

The genital fold of the *coronal sulcus* encloses the dorsal and lateral aspects of the bicavernosal body of the penis and the flat mucosal roof of the penile urethra.

The genital fold covering the roof and cap of the glans penis, encloses the dorsal and lateral aspects, and conical end of the bicavernosal body. The genital fold encloses the flat mucosal roof of the glanular urethra (fossa navicularis).

The unfused vestibular/urethral folds (ventral to the penile urethra of the coronal sulcus and ventral to the glanular urethra/fossa navicularis of the glans penis) *are aligned with the ventral border of the genital fold and lateral borders of the penile* urethra (of the coronal sulcus) *and lateral borders of the glanular urethra* (fossa navicularis of the glans penis).

The ventral borders of the genital fold in the coronal sulcus and glans penis, reflect on the *outer borders* of the unfused vestibular/urethral folds, ventral to the penile urethra in the coronal sulcus, and ventral to the glanular urethra in the glans penis. The curved *anterior/distal end of the genital fold* (covering the crescentic cap of the glans penis) reflects on the outer border of the *anterior commissure of* the vestibular/urethral folds.
The *inner borders* (ectoderm), of the unfused vestibular/urethral folds, meet the lateral borders of the mucosal roof (endoderm), of the penile urethra (of the coronal sulcus) and mucosal roof (endoderm), of the glanular urethra (of the glans penis), at a much widened, Hart's line.

This arrangement is somewhat analogous to that of the female vestibule, ventral to the genital swelling.
However, females do not develop a *penile urethra*, but do develop a *dorsal hood of the clitoris*.
In MIP (megameatus with intact prepuce), the penile urethra does develop, except that tubularization of the penile urethra, is arrested in the *coronal sulcus.*
The *prepuce develops normally,* proximal to the failed tubularization, of the penile urethra, in the coronal sulcus.

***Intact prepuce*:**
In normal development, formation of the prepuce begins *proximal to the coronal sulcus*. Similarly, in MIP formation of the prepuce begins proximal to the coronal sulcus, and develops normally, independent of the megameatus.

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