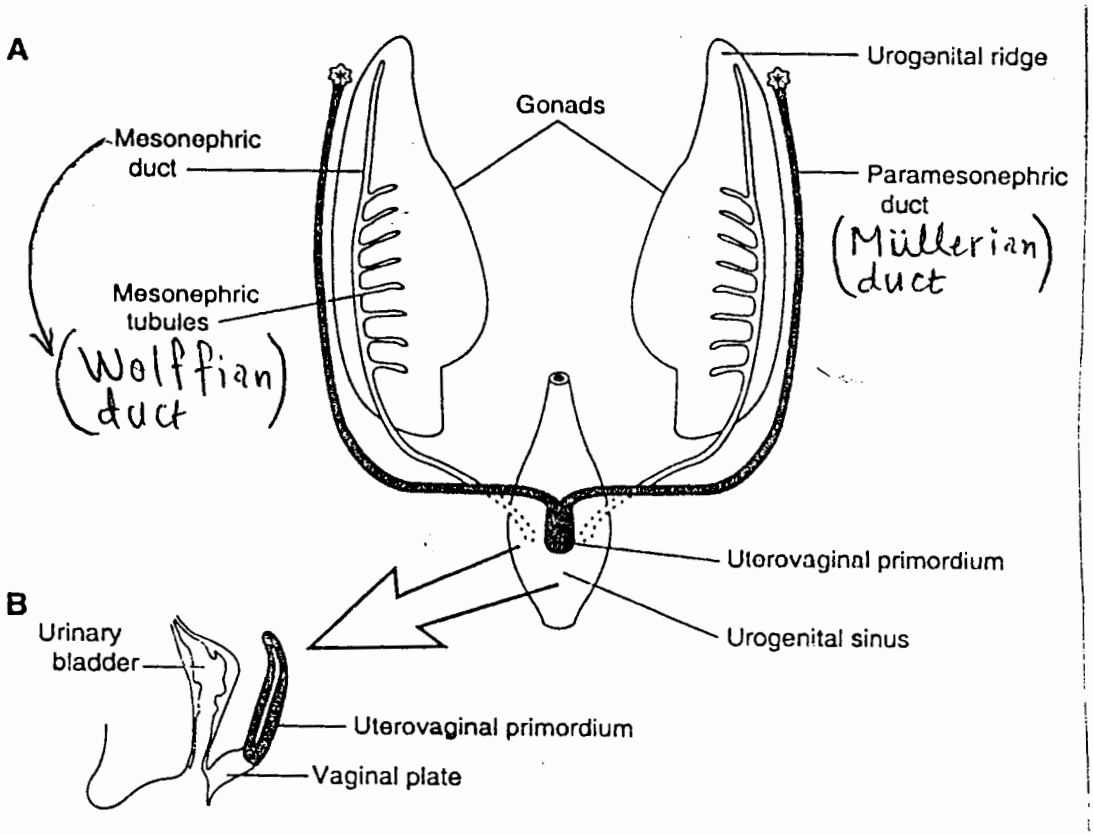
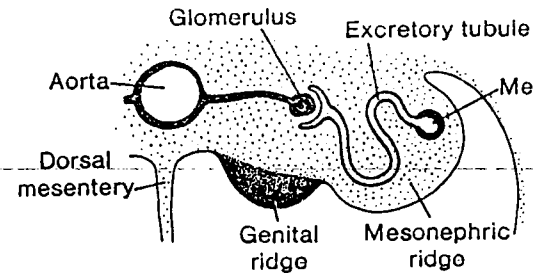
Bustami (87) B

Genital System

Gen. (1)

I. Although the genetic sex of the embryo is determined at the time of fertilization, there is no morphological indication of sex until the 12th week.
II. The immature gonads are identical in both sexes.
III. The beginning of the external genitalia is simiiar.
IV. Two separate duct systems, one capable of forming the male genital ducts and the other capable of giving rise to the female genital tract, also develop in all embryos.
V. Thus the young embryo is sexually "indifferent" and is capable of developing along either male or female lines.
VI. Depending upon the genetic sex of the embryo, one duct system develops and the other degenerates.
VII. Differentiation occurs first in the gonads, later in internal genital organs, and finally in the external genitalia.





GONADS

I. Indifferent stage
   A. Gonads are derived from three sources:
      1. Coelomic epithelium (the germinal epithelium)
      2. Underlying mesenchyme
      3. Primordial germ cells
   B. Proliferation of the coelomic epithelium and the condensation of underlying mesenchyme produce a longitudinal gonadal ridge between the mesonephros and the dorsal mesentery → genital ridge (gonadal ridge)
   C. The coelomic epithelium proliferates, sending finger-like projections, the primary sex cords, into the underlying mesenchyme.

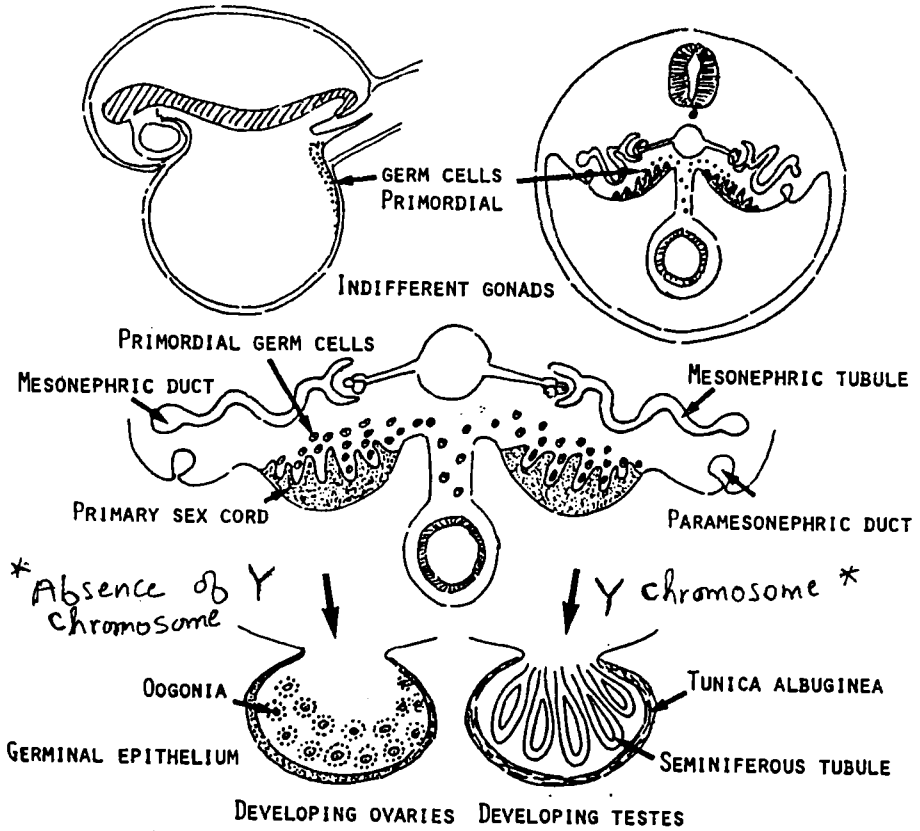


Fig. 18-1. Development of gonads.

   D. Large primordial germ cells appear among the endodermal cells of the yolk sac (Fig. 18-1).
   E. The primordial germ cells migrate along the dorsal mesentery to the gonadal ridge.
   F. The primary sex cords gradually surround the invading primordial germ cells.

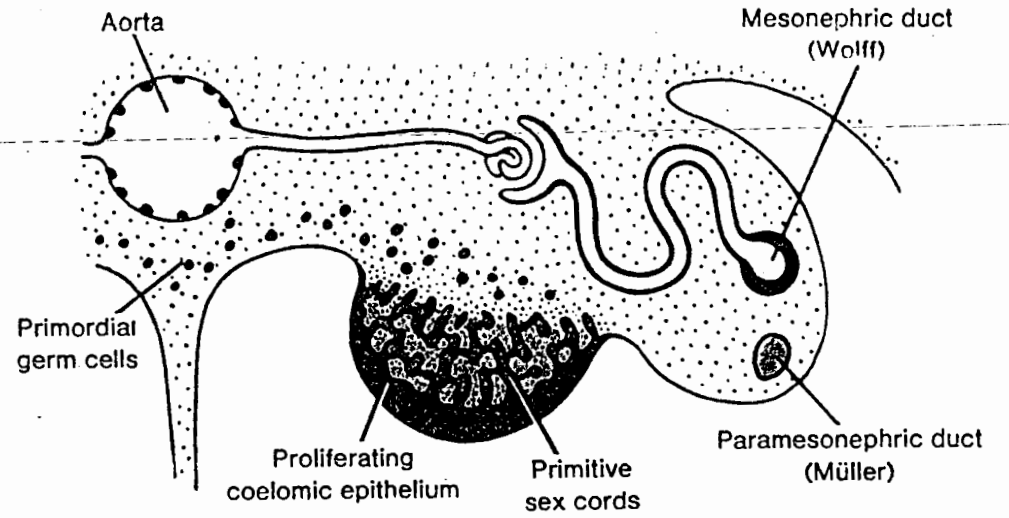


Figure 15-16. Schematic transverse section through the lumbar region of a 6-week embryo, showing the indifferent gonad with the primitive sex cords. Some of the primordial germ cells are surrounded by cells of the primitive sex cords.

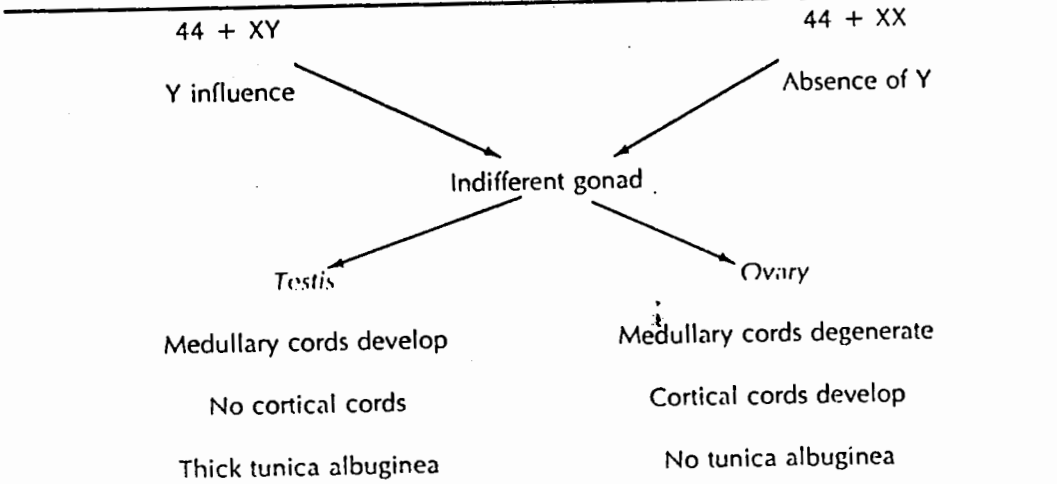
G. As the gonad grows, a deep groove appears which separates the gonad from the mesonephros and the dorsal body wall. The gonad becomes suspended by its own narrow mesentery.

H. This indifferent gonad consists of outer cortex and inner medulla.

1. If the embryo contains the XX sex chromosomal complex, its cortical region differentiates into ovary, and its medulla regresses.

2. If the embryo has the XY sex chromosomal complex, its medullary region differentiates into testes and its cortex regresses.
3. It is the presence of the Y chromosome which influences the medulla to differentiate into the testes.
4. Absence of the Y chromosome results in development of ovaries (Fig. 18-1).

Influence of Primordial Germ Cells on Indifferent Gonad



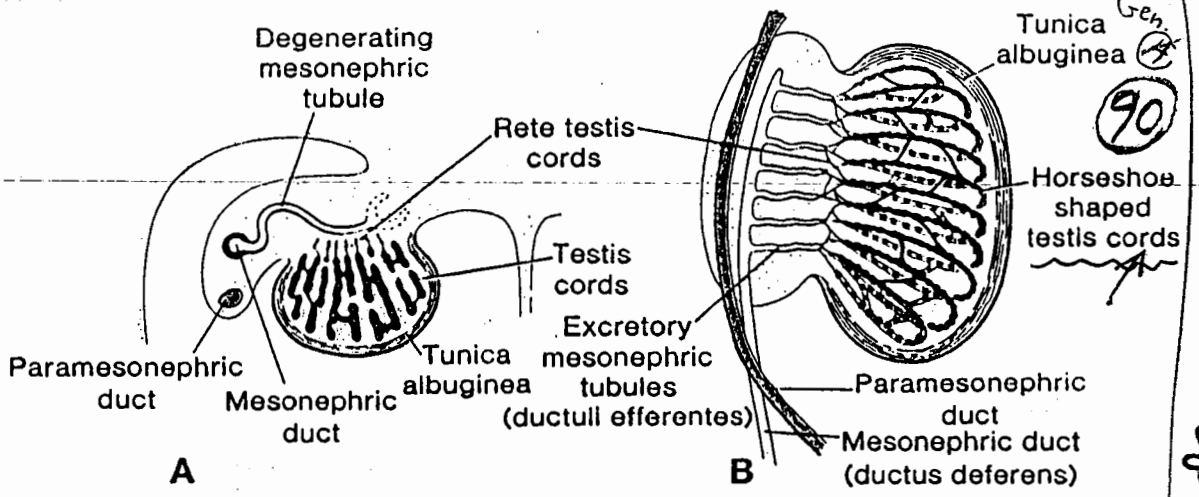


Figure 15-17. A, Transverse section through the testis in the 8th week of development. Note the tunica albuginea, the testis cords, the rete testis, and the primordial germ cells. The glomerulus and Bowman's capsule of the mesonephric excretory tubule are in regression. B, Schematic representation of the testis and the genital duct in the 4th month of development. The horseshoe-shaped testis cords are continuous with the rete testis cords. Note the ductuli efferentes (excretory mesonephric tubules) which enter the mesonephric duct.

1. Development of the testes

–The **intermediate mesoderm** forms a longitudinal elevation along the dorsal body wall, the **urogenital ridge.**

–The coelomic epithelium and underlying mesoderm of the urogenital ridge proliferate to form the **gonadal ridge.**

–**Primary sex cords** develop from the gonadal ridge and incorporate **primordial germ cells (XY genotype),** which migrate into the gonad from the wall of the yolk sac.

–The Y chromosome carries a gene on its short arm that codes for **testes-determining factor (TDF),** which is crucial to testes differentiation.

–The primary sex cords extend into the medulla of the gonad and lose their connection with the surface epithelium as the thick **tunica albuginea** forms.

–The primary sex cords form the **seminiferous cords, tubuli recti,** and **rete testes.**

a. Seminiferous cords are made up of **spermatogonia** and **sustentacular (Sertoli) cells,** which secrete **müllerian-inhibiting factor (MIF).**

b. The mesoderm between the seminiferous cords gives rise to the **interstitial (Leydig) cells,** which secrete **testosterone.**

c. The spermatogonia, sustentacular (Sertoli) cells, interstitial (Leydig) cells, and connective tissue stroma of the testes are derived from mesoderm.

d. The seminiferous cords remain as solid cords until puberty when they acquire a lumen and are then called **seminiferous tubules.**

e. As the seminiferous tubules become canalized, the rete testis becomes continuous with 15 to 20 persistent mesonephric tubules, which become the efferent ductules.

f. The efferent ductules are connected to the mesonephric duct which forms the ductus epididymis, the vas deferens, the seminal vesicle, and the ejaculatory duct.

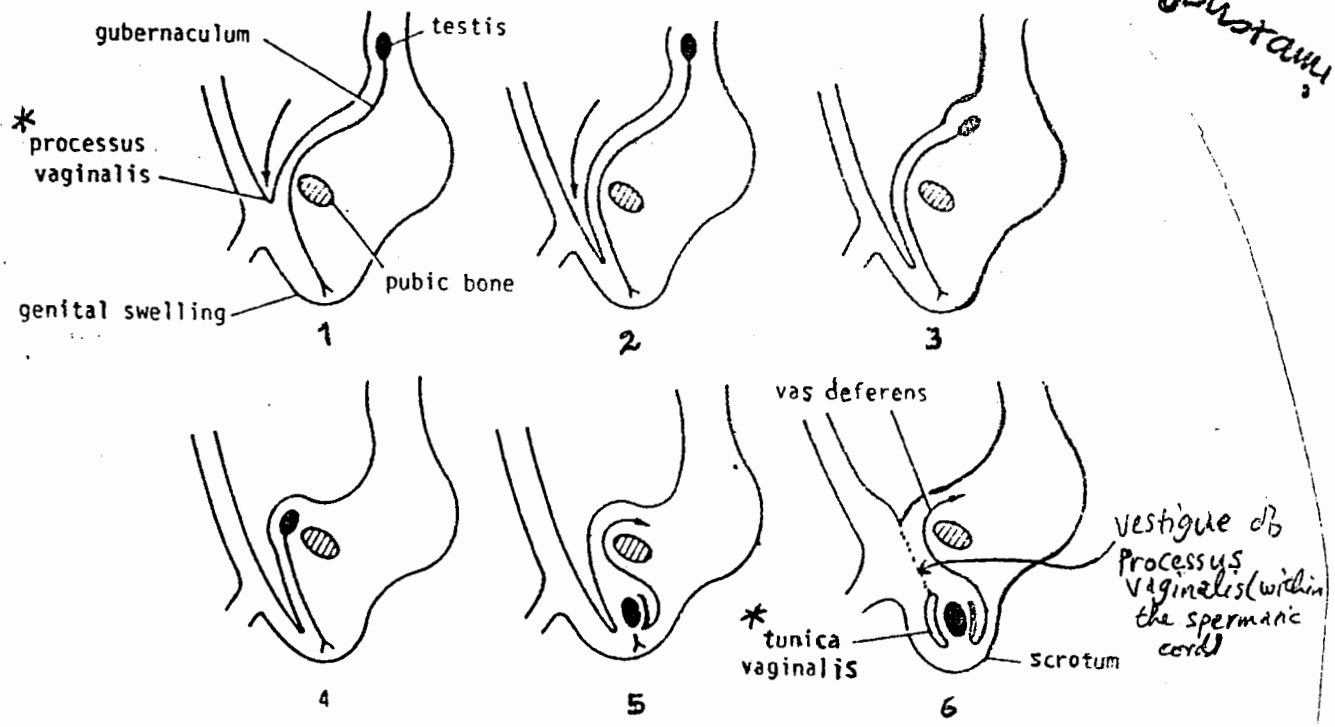
The **interstitial cells of Leydig** are derived from the original mesenchyme of the gonadal ridge. They lie between the ~~testicular~~ testis cords and begin development shortly after the onset of differentiation of these cords. By the 8th week of gestation, testosterone production by the Leydig cells begins and the testis is now able to influence the sexual differentiation of the genital ducts and external genitalia (16).

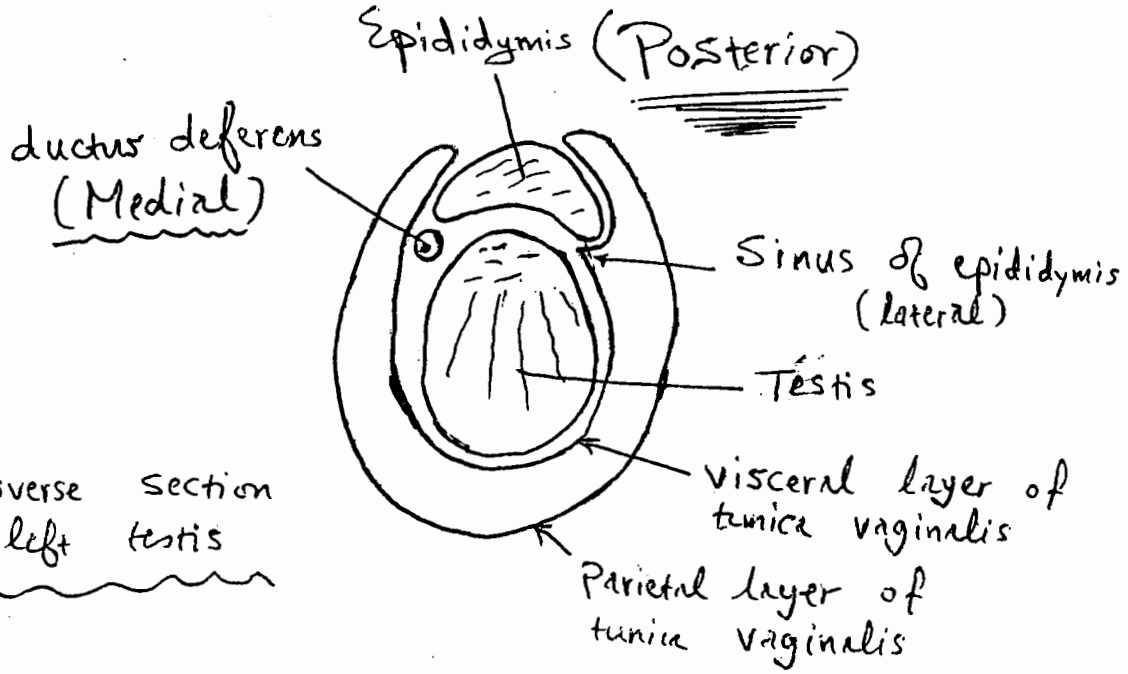
Relative descent of the testes

–The testes originally develop within the abdomen but later descend into the scrotum. The relative descent of the testes to the inguinal canal is a result of disproportionate growth of the upper abdominal region away from the pelvic region.

Descent of testis

Brostami





Transverse section of left testis

I. Descent of the testes

A. As the mesonephros regresses, its remnants become ligamentous and are known as the gubernaculum (Fig. 18-5).

B. The gubernaculum extends from the lower pole of the testes along the posterior body wall, passes through the developing abdominal wall at the site of future inguinal canal, and attaches to the genital swelling.

C. Because the gubernacula do not grow in proportion to the growth of the body, the testes are held back. Thus, the change in position (descent) of testes is only relative to the growing body wall.

D. On each side, a sac of peritoneum, the vaginal process, extends through the abdominal wall into the scrotum ventral to the gubernaculum, following the path formed by gubernaculum.

E. The final descent of the testes through the inguinal canal into the scrotum is regulated by gonadotropic and androgenic hormones.

F. The gubernaculum becomes short, causing the testes to migrate into the scrotum posterior to the vaginal process.

G. As the testes enter the scrotum, the proximal part of the vaginal process becomes obliterated.

H. The distal part of the vaginal process surrounds the anterior part of the testes and persists as the tunica vaginalis.

I. A remnant of gubernaculum anchors the testes within the scrotum, preventing it from twisting on its spermatic cord.

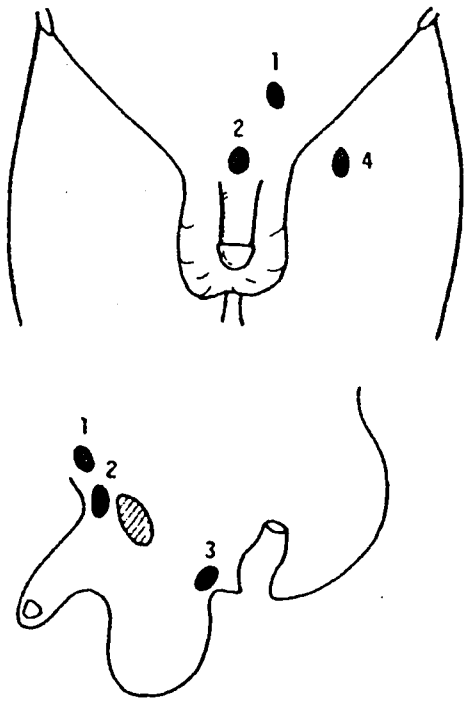


Fig. 16-12. The common sites of maldescent of the testis: (1) abdominal, (2) root of penis, (3) perineum, and (4) femoral triangle.

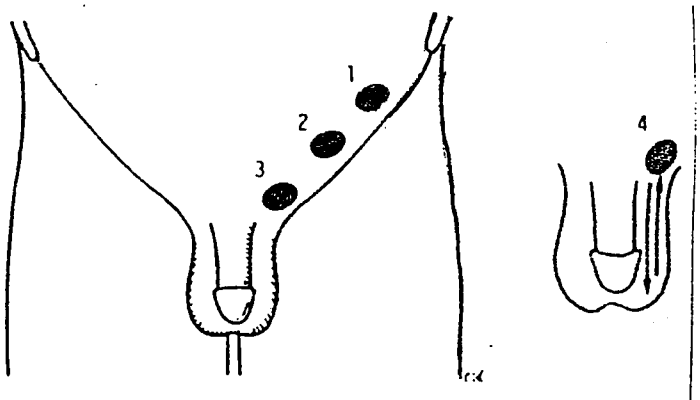


Fig. 16-9. The common sites of incomplete descent of the testis: (1) abdominal, (2) inguinal canal, (3) superficial inguinal ring, and (4) retractile testis.

VI. Cryptorchidism: Failure of the testes to descend normally into the scrotum.

A. The condition may be unilateral or bilateral.

B. An abnormal androgen production and failure of the gubernaculum to become short could be the causing factors.

C. One or both testes may remain in the following areas:

1. In the abdomen, just above the deep inguinal ring
2. In the inguinal canal
3. In the superficial inguinal ring

D. Spermatogenesis is impaired in undescended testis and may eventually lead to its atrophy.

E. There is an increased possibility of development of malignant tumor in undescended testes. For these reasons it is necessary to position the testes surgically within the scrotum.

VII. Ectopic testes result when they deviate from the usual path of descent and they may come to lie in the lower abdominal wall, the medial aspect of the thigh, the base of the penis, the perineum, or other unusual places.

VIII. Congenital inguinal hernia results from failure of the vaginal process to close normally. The persistent peritoneal sac permits an intestinal loop to descend into the scrotum. Cryptorchidism is often associated with a congenital hernia on the affected side (Fig. 18-7).

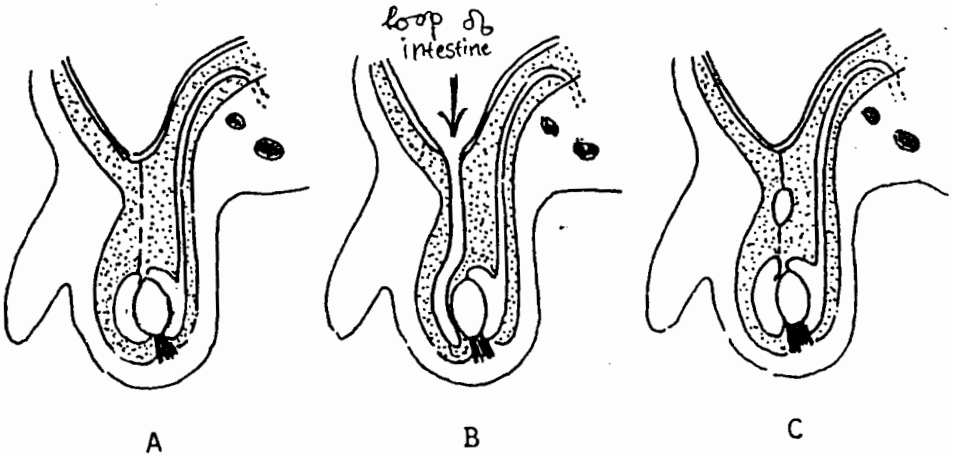


Fig. 18-7. Malformations associated with descent of testes: (A) normal, for comparison; (B) congenital inguinal hernia; (C) hydrocele (of the cord)

IX. Hydrocele results from accumulation of fluid in an unobliterated portion of the vaginal process.

A. Occasionally the abdominal end of the vaginal process remains open and, although it is too small to permit herniation, peritoneal fluid passes into the passageway.

B. If the middle portion of the canal persists, it gives rise to hydrocele of the spermatic cord (Fig. 18-7).

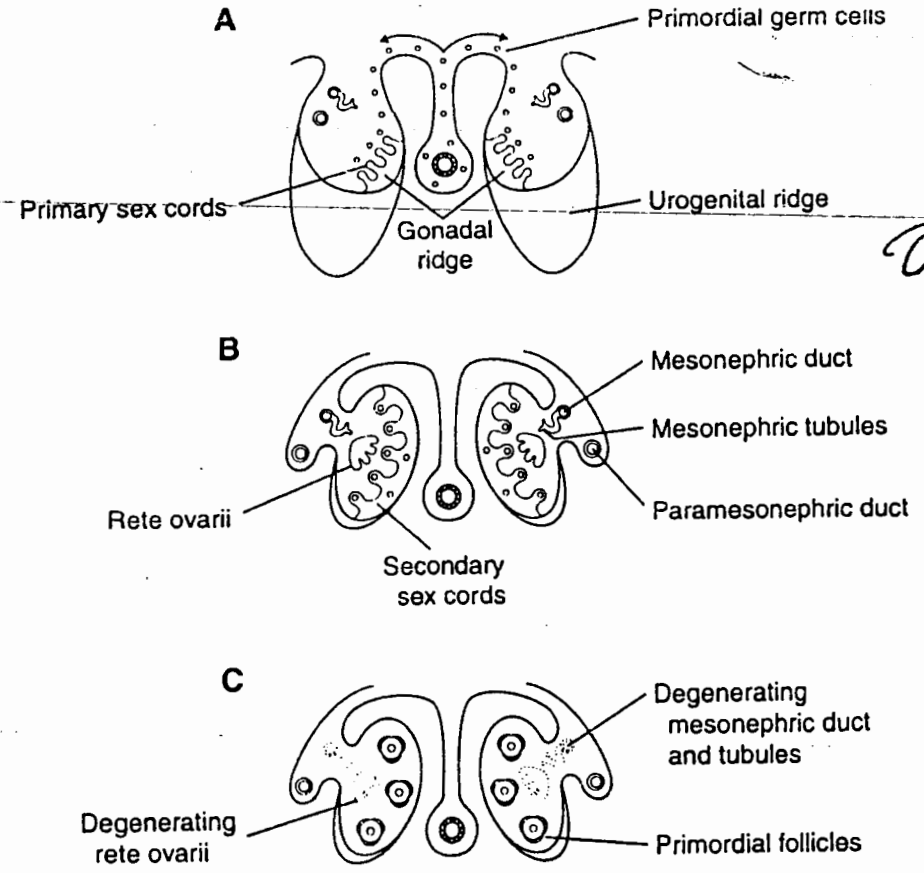


Figure 14-1. Diagram indicating the differentiation of the gonad in the female. (*A*) Gonad in the indifferent embryo. (*B*) Ovary at week 12 of development. (*C*) Ovary at week 20.

1. Development of the ovaries

- –The **intermediate mesoderm** forms a longitudinal elevation along the dorsal body wall, the **urogenital ridge.**
- –The coelomic epithelium and underlying mesoderm of the urogenital ridge proliferate to form the **gonadal ridge.**
- –**Primary sex cords** develop from the gonadal ridge and incorporate **primordial germ cells (XX genotype),** which migrate into the gonad from the wall of the yolk sac.
- –Primary sex cords develop into the **rete ovarii,** which degenerate.
- –Later, **secondary sex cords** develop and incorporate primordial germ cells as a thin **tunica albuginea** forms.
- –The secondary sex cords break apart and form isolated cell clusters, **primordial follicles,** which contain **primary oocytes** surrounded by a layer of **follicular (granulosa) cells.**
- –Primary oocytes, follicular cells, and connective tissue stroma of the ovary are derived from mesoderm.

II. Descent of the ovaries

A. The gubernaculum pulls the ovaries into the pelvis.

B. The gubernaculum becomes attached to the developing uterus near the site of entry of the uterine tube. This attachment changes the direction of pull of the gubernaculum, causing the ovary to be directed medially into the pelvis.

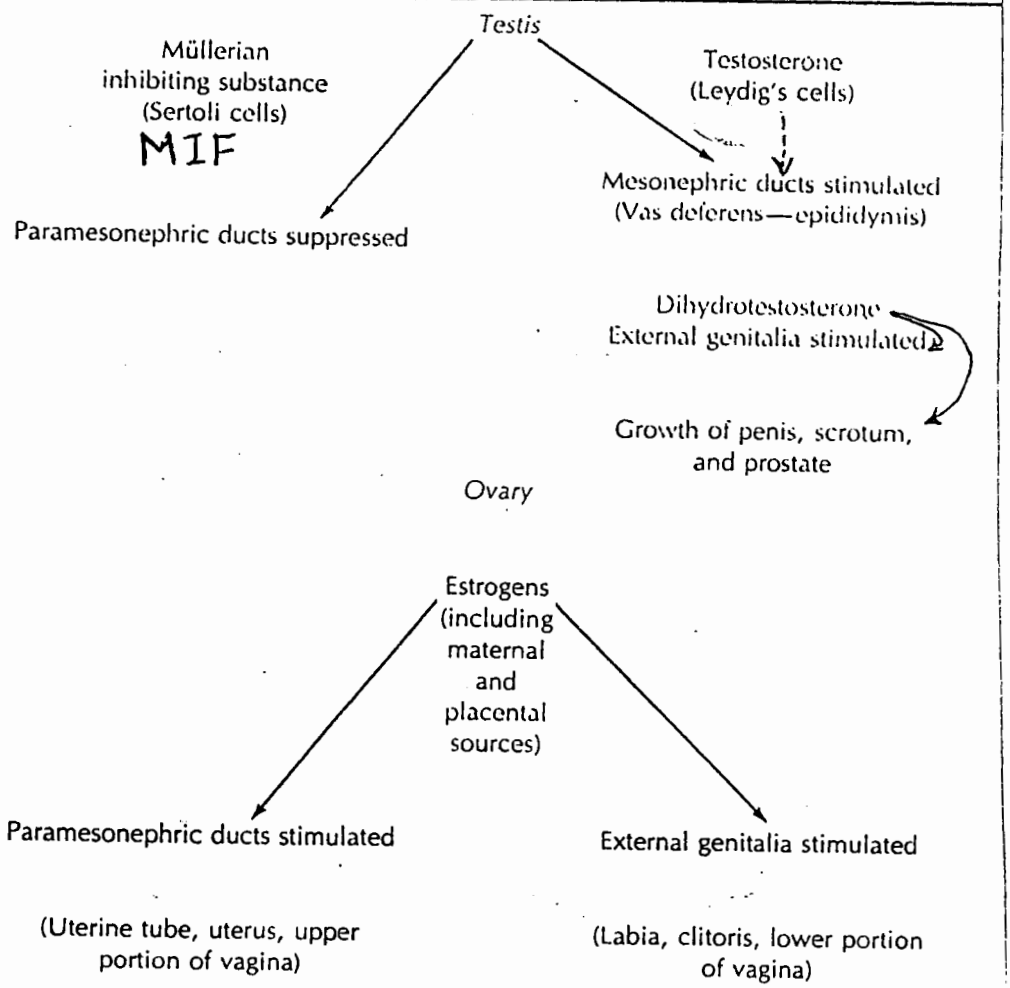
C. The part of the gubernaculum which extends from the medial pole of the ovary to the uterus forms the ovarian ligament. The part of the gubernaculum extending from the uterus to the labia majora becomes the round ligament of the uterus

Genital duct systems

–The indifferent embryo has two separate genital duct systems.

1. **Paramesonephric (müllerian) ducts** play a major role in the female.
2. **Mesonephric tubules** and **mesonephric (wolffian) ducts** play a major role in the male.

Influence of Sex Gland on Further Sex Differentiation



DIFFERENTIATION OF THE DUCT SYSTEM

Development of the genital duct system and the external genitalia occurs under influence of hormones circulating in the fetus during intrauterine life (16). Also, Sertoli cells in the fetal testes produce a nonsteroidal substance, known as **Müllerian inhibiting substance** (MIS), which causes regression of the paramesonephric duct (17, 18). In addition to this inhibiting substance, the testes produce **testosterone** (the major **androgen** produced by the testes), which enters the cells of target tissues. Here it may be converted by a 5α-reductase enzyme to **dihydrotestosterone**. Testosterone and dihydrotestosterone bind to a specific high-affinity intracellular receptor protein, and ultimately this hormone-receptor complex binds to DNA to regulate transcription of tissue-specific genes and their protein products (Fig. 15-20). Testosterone receptor complexes mediate virulization of the mesonephric ducts, whereas dihydrotestosterone receptor complexes modulate differentiation of the male external genitalia (Table

In the female, the paramesonephric duct system develops into the uterine tubes and uterus. Controlling factors for this process are not clear, but may involve estrogens produced by the maternal system, placenta, and fetal ovaries (16). Since the male inducer substance is absent, the mesonephric duct system regresses. In the absence of androgens, the indifferent external genitalia are stimulated by estrogens and differentiate into labia majora, labia minora, clitoris, and part of the vagina (Table 15-2).

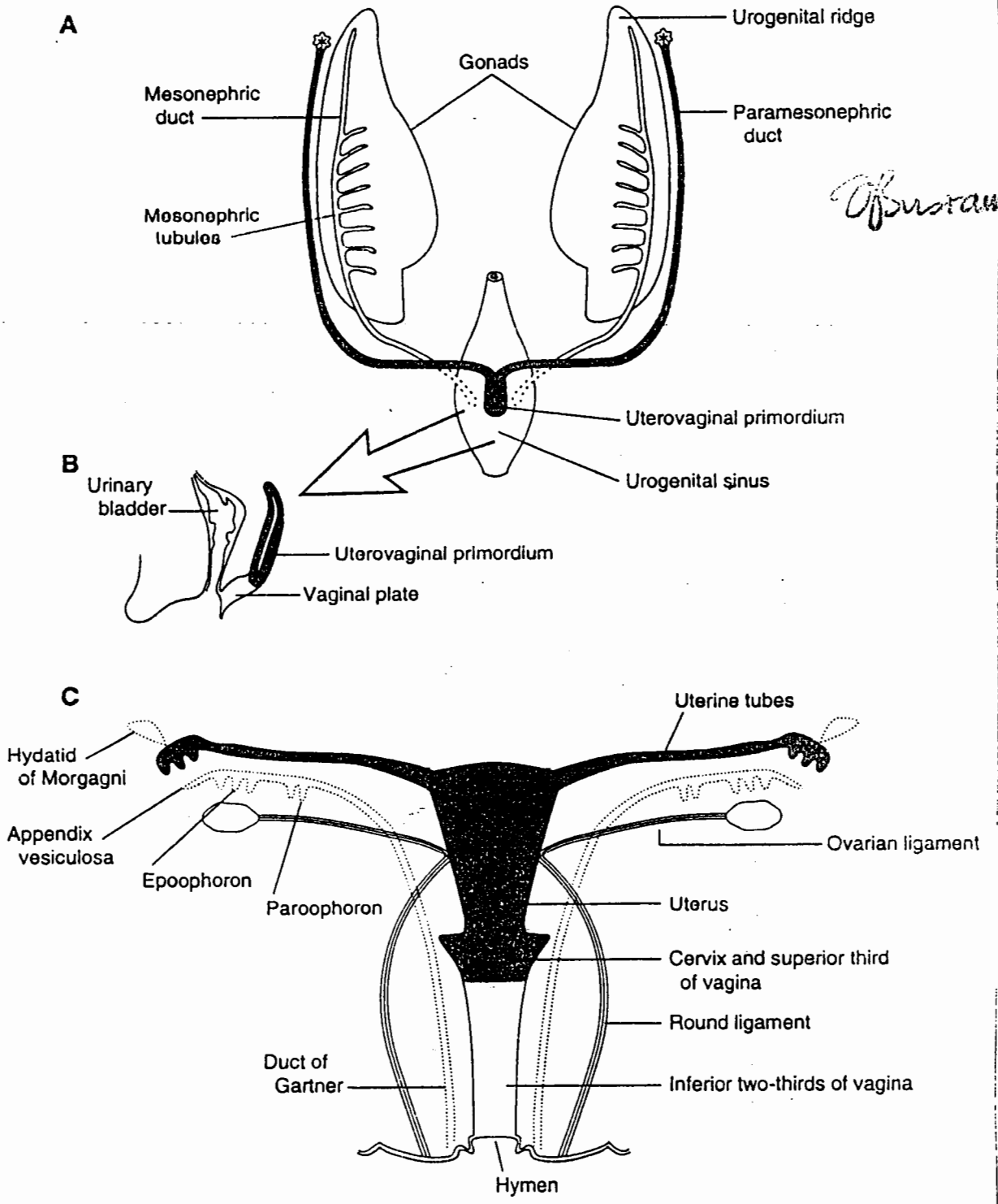


Figure 14-2. Diagram indicating the differentiation of the genital duct systems in the female. (*A*) Genital duct systems in the indifferent embryo. (*B*) Side view showing the dual origin of the vagina. (*C*) Female components and vestigial remnants (*dotted lines*) at birth. The paramesonephric duct and its derivatives are *shaded*.

Genital duct systems (Figure 14-2)

1. Paramesonephric ducts

–develop as invaginations of the lateral surface of the urogenital ridge.

a. The cranial portions run parallel to the mesonephric ducts and develop into the **uterine tubes;** the **hydatid of Morgagni** is a vestigial remnant of the paramesonephric duct.

b. The caudal portions fuse in the midline to form the **uterovaginal primordium** and bring together two peritoneal folds, the **broad ligament.**

(1) The **uterovaginal primordium** develops into the **uterus, cervix,** and **superior third of the vagina.**

(2) The uterovaginal primordium projects into the dorsal wall of the urogenital sinus and induces the formation of the **sinovaginal bulbs.**

(3) The sinovaginal bulbs fuse to form the solid **vaginal plate,** which canalizes and develops into the **inferior two-thirds of the vagina.**

(4) Although the vagina has a dual origin, most authorities agree that the epithelial lining of the entire vagina is of **endodermal origin.**

(5) The lumen of the vagina is separated from the urogenital sinus by a membrane called the hymen. This membrane usually ruptures during the perinatal period.

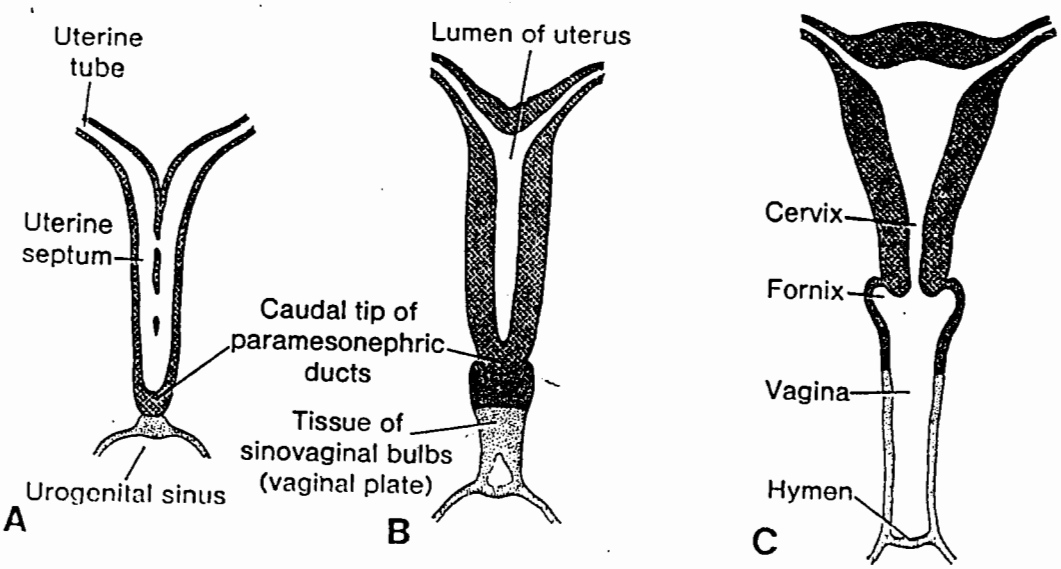
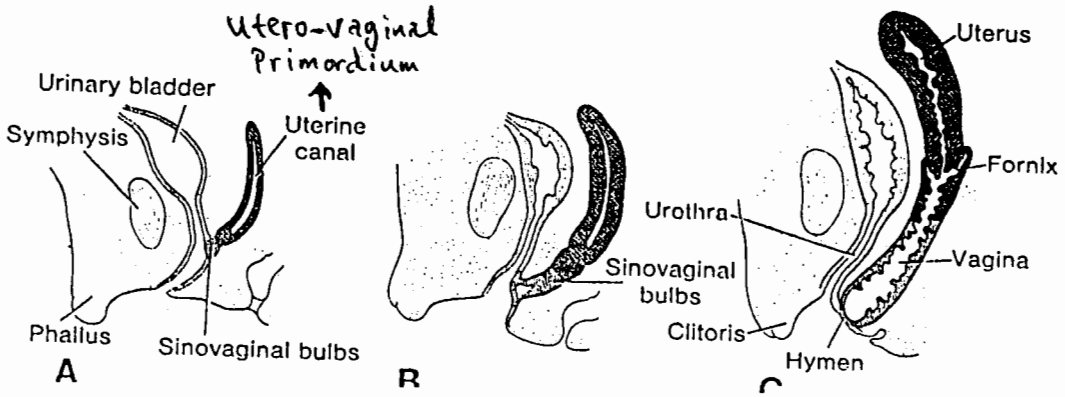


Figure 15-24. Schematic drawing showing the formation of the uterus and vagina. A, At 9 weeks. Note the disappearance of the uterine septum. B, At the end of the 3rd month. Note the tissue of the sinovaginal bulbs. C, Newborn. The upper portion of the vagina and the fornices are formed by vacuolization of the paramesonephric tissue and the lower portion by vacuolization of the sinovaginal bulbs.



2. Mesonephric tubules and ducts

- –develop as part of the urinary system; the ducts are critical in the formation of the definitive metanephric kidney.
- –regress completely in the female after formation of the metanephric kidney; vestigial remnants persist as the **appendix vesiculosa, epoophoron, paroophoron,** and **Gartner's duct.**

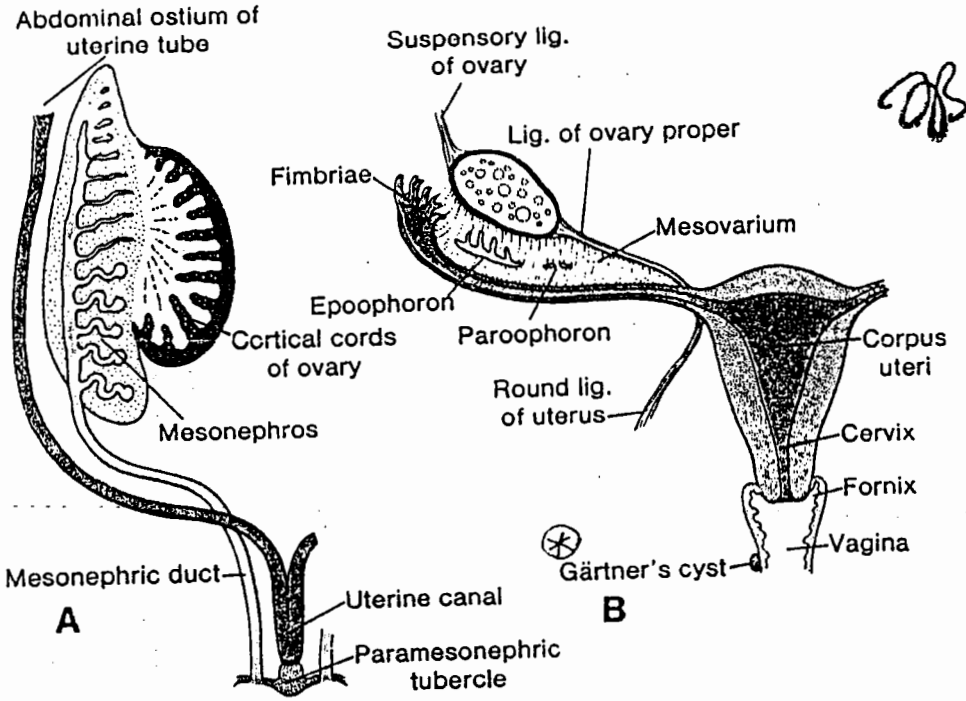


Figure 15-22. A, Schematic drawing of the genital ducts in the female at the end of the 2nd month of development. Note the paramesonephric or Müllerian tubercle and the formation of the uterine canal. B, The genital ducts after descent of the ovary. The only parts remaining of the mesonephric system are the epoophoron, paroophoron, and Gärtner's cyst. Note the suspensory ligament of the ovary, the ligament of the ovary proper, and the round ligament of the uterus.

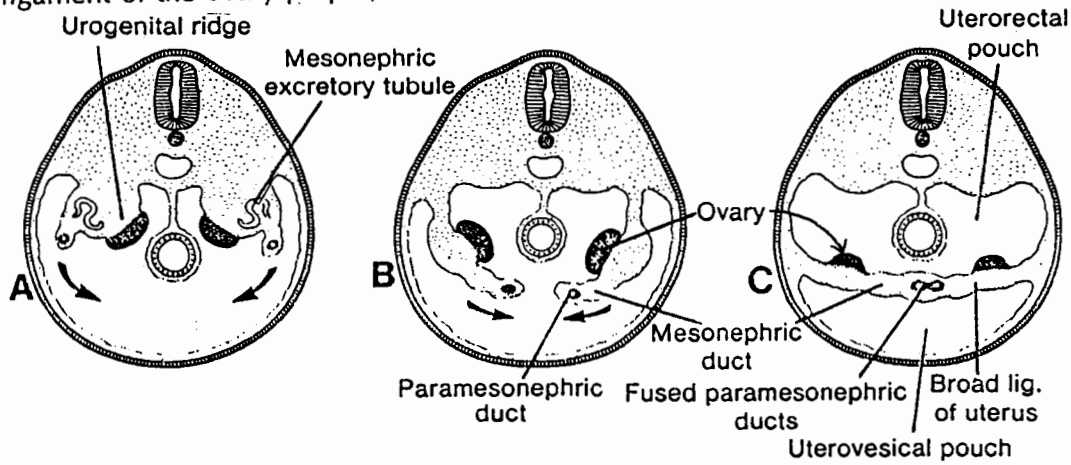


Figure 15-23. Transverse sections through the urogenital ridge at progressively lower levels. Note that the paramesonephric ducts approach each other in the midline to fuse. As a result of the fusion, a transverse fold, the broad ligament of the uterus, is formed in the pelvis. The gonads come to lie at the posterior aspect of the transverse fold.

As the paramesonephric ducts turn medially to fuse, they bring together the peritoneal folds which give rise to the following areas:

1. **The left and right broad ligaments.**
2. **Two peritoneal compartments of the pelvic cavity:**
   a) The uterorectal Pouch
   b) Uterovesical pouches

Uterovaginal malformations

A. Agenesis of the uterus results when the paramesonephric ducts fail to develop. This fails to induce a vaginal plate to form.

B. Underdevelopment of the caudal part of the paramesonephric ducts results in a rudimentary uterus.

C. Duplication and Atresia of Uterine Canal

Lack of fusion of the paramesonephric ducts in a localized area or throughout the length of the ducts may explain all different types of duplication of the uterus. In its extreme form, the uterus is entirely double (**uterus didelphys**) (Fig. 15-26A); in the least severe form, it is only slightly indented in the middle (**uterus arcuatus**) (Fig. 15-26B). One of the more common anomalies is the **uterus bicornis**, in which the uterus has two horns entering a common vagina (Fig. 15-26C). This condition is normal in many of the mammals below the primates.

In patients with complete or partial atresia of one of the paramesonephric ducts, the rudimentary part lies as an appendage to the well-developed side. However, since its lumen usually does not communicate with the

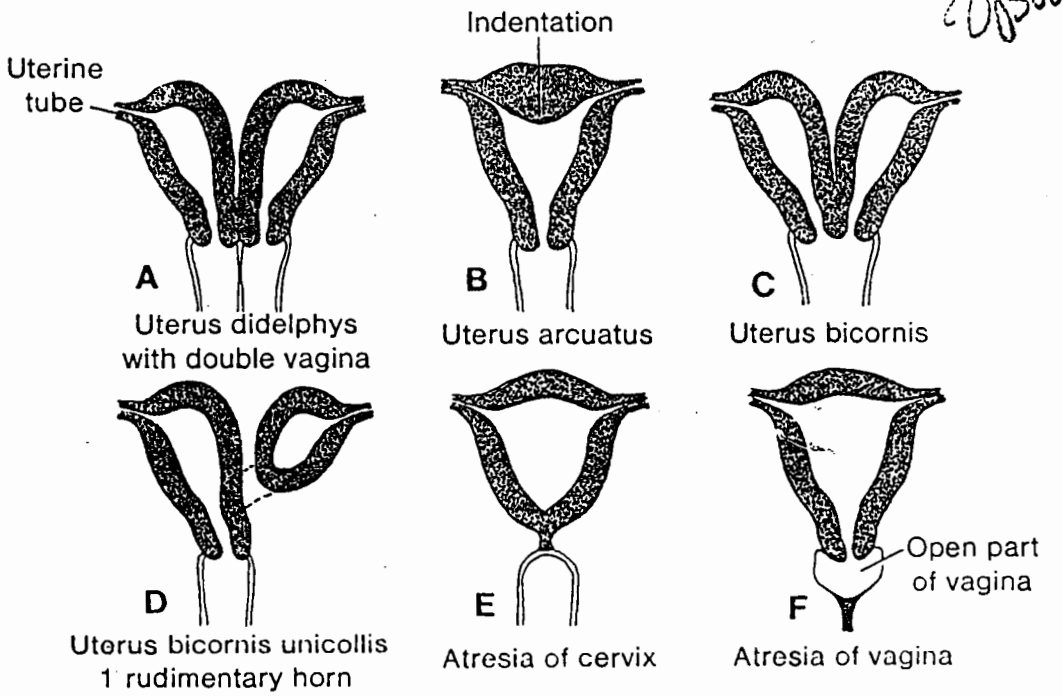


Figure 15-26. Schematic representation of the main abnormalities of the uterus and vagina, caused by persistence of the uterine septum or obliteration of the lumen of the uterine canal.

vagina, complications frequently ensue (**uterus bicornis unicollis with one rudimentary horn**) (Fig. 15-26D). If the atresia involves both sides partially, an **atresia of the cervix** may result (Fig. 15-26E). If the sinovaginal bulbs fail to fuse or do not develop at all, a **double vagina** or **atresia of the vagina**, respectively, results (Fig. 15-26A, F). In the latter case, a small vaginal pouch, originating from the paramesonephric ducts, usually surrounds the opening of the cervix.

Genital duct systems (Figure 14-5) in the male ♂ Gen (100)

1. Paramesonephric ducts

- –develop as invaginations of the lateral surface of the urogenital ridge.
- –The cranial portions run parallel to the mesonephric ducts.
- –The caudal portions fuse in the midline to form the **uterovaginal primordium.**
- –Under the influence of MIF, the cranial portions of the paramesonephric ducts and the uterovaginal primordium regress completely; a vestigial remnant, the **appendix testis,** may persist (Hydatid of Morgagni)

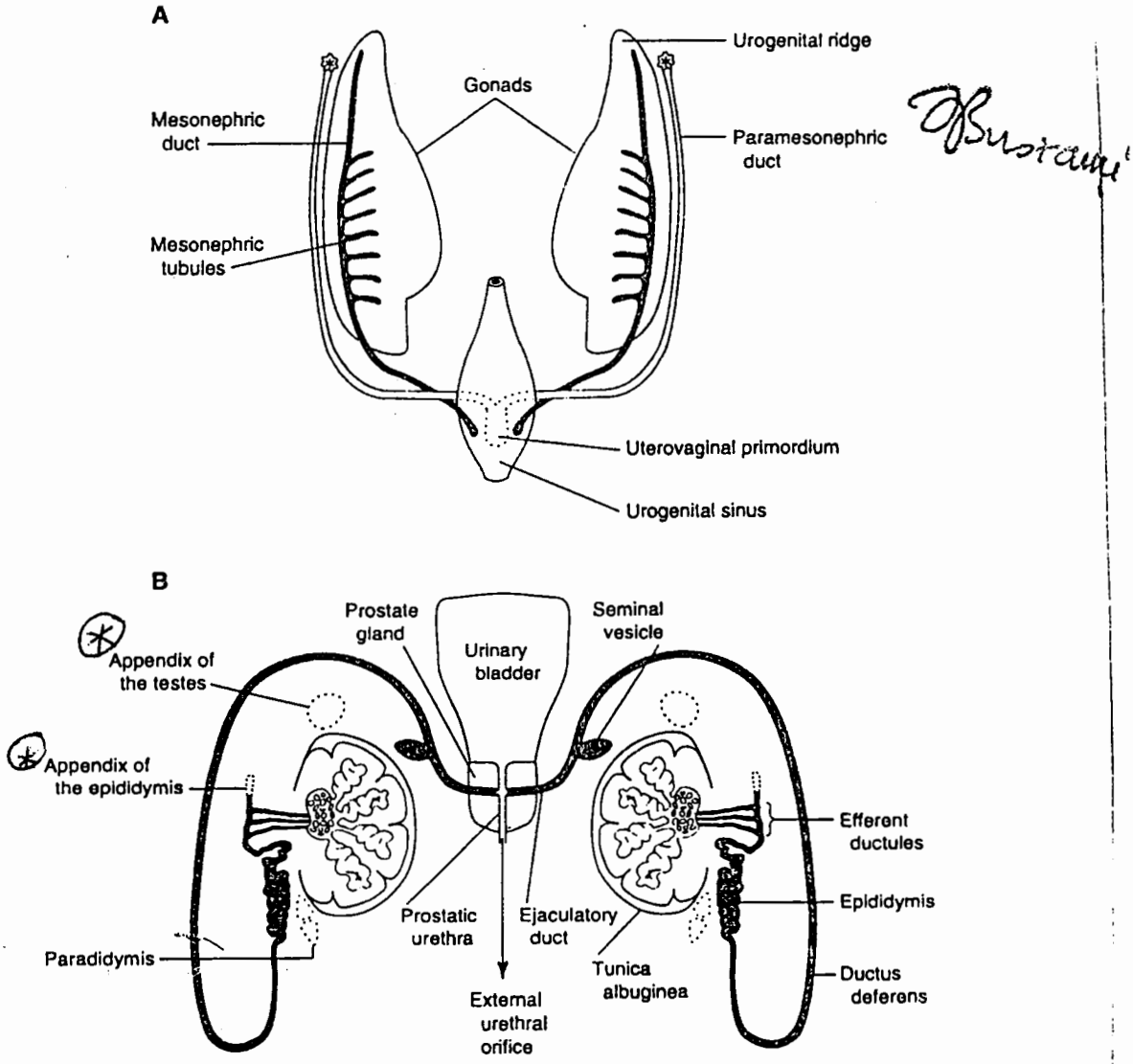


Figure 14-5. Diagram indicating the differentiation of the genital duct systems in the male. (*A*) Genital duct systems in the indifferent embryo. (*B*) Male components and vestigial remnants (*dotted lines*). The mesonephric tubules and mesonephric duct and their derivatives are *shaded*.

2. Mesonephric tubules and ducts

- –develop as part of the urinary system; the ducts are critical in the formation of the definitive metanephric kidney.

a. A few **mesonephric tubules** in the region of the testes form the **efferent ductules;** vestigial remnant of others is called the **paradidymis.**

b. The **mesonephric ducts** form the **epididymis, ductus deferens, seminal vesicle,** and **ejaculatory duct;** a vestigial remnant of the cranial end of the mesonephric duct, the **appendix epididymis,** may persist.

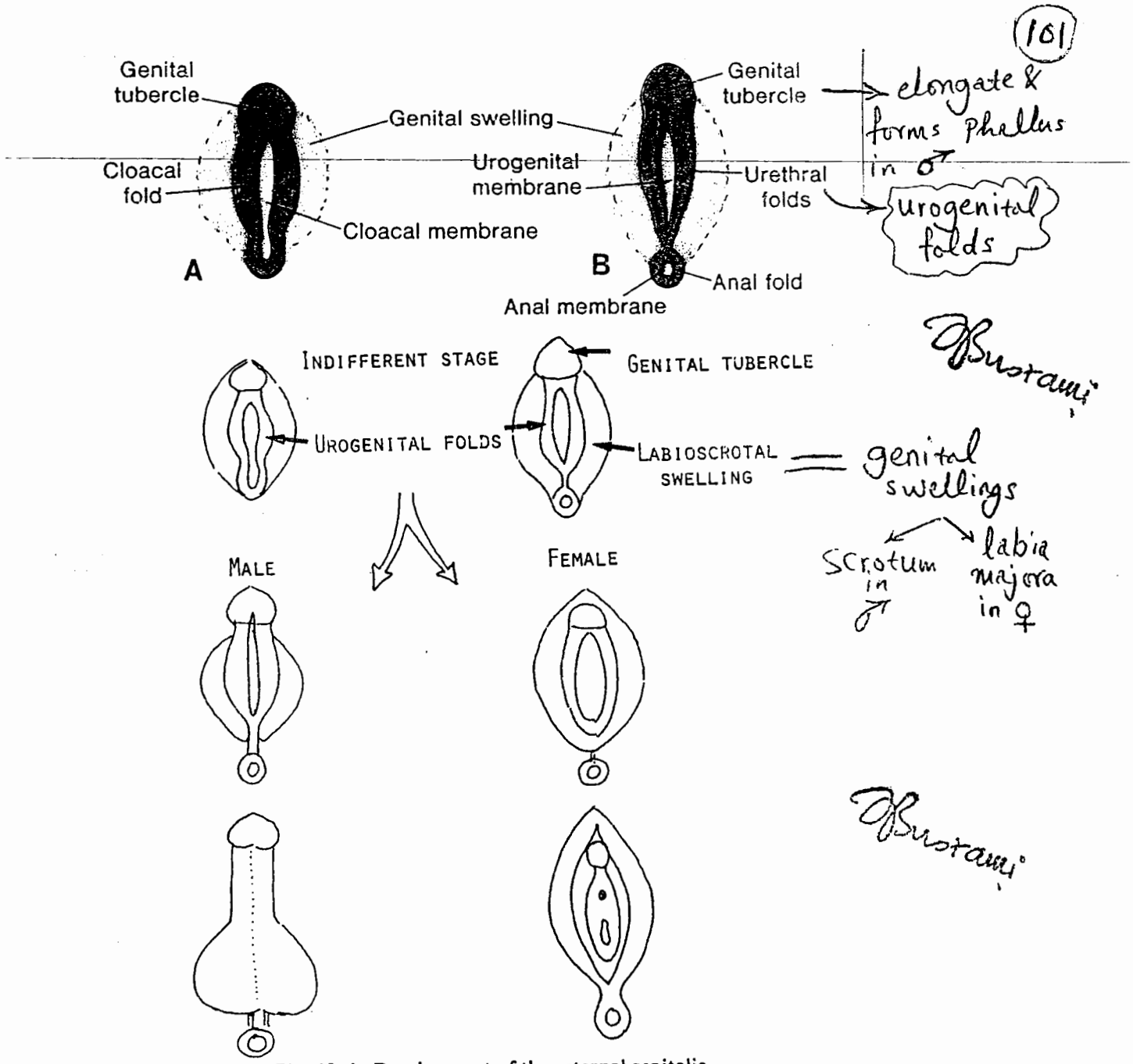


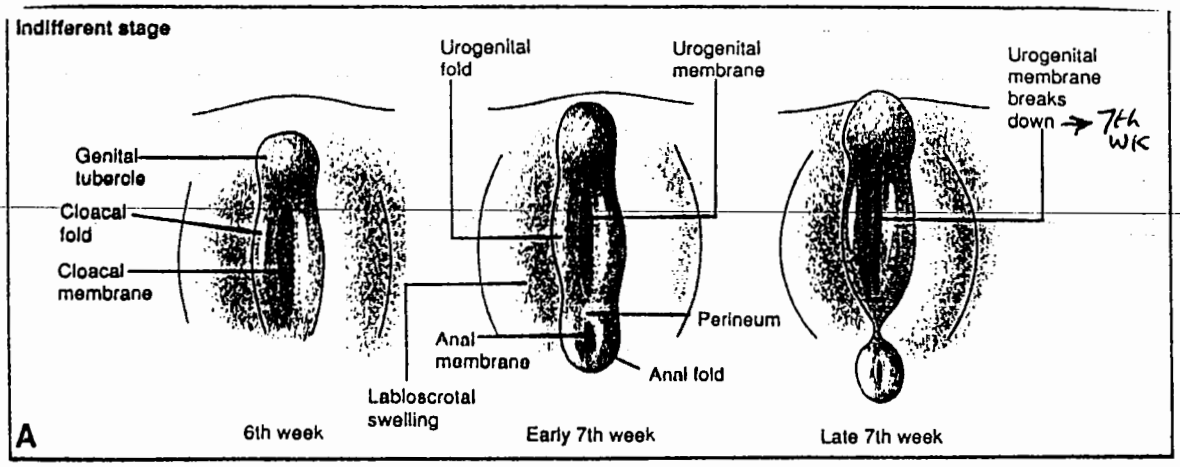
Fig. 18-4. Development of the external genitalia.

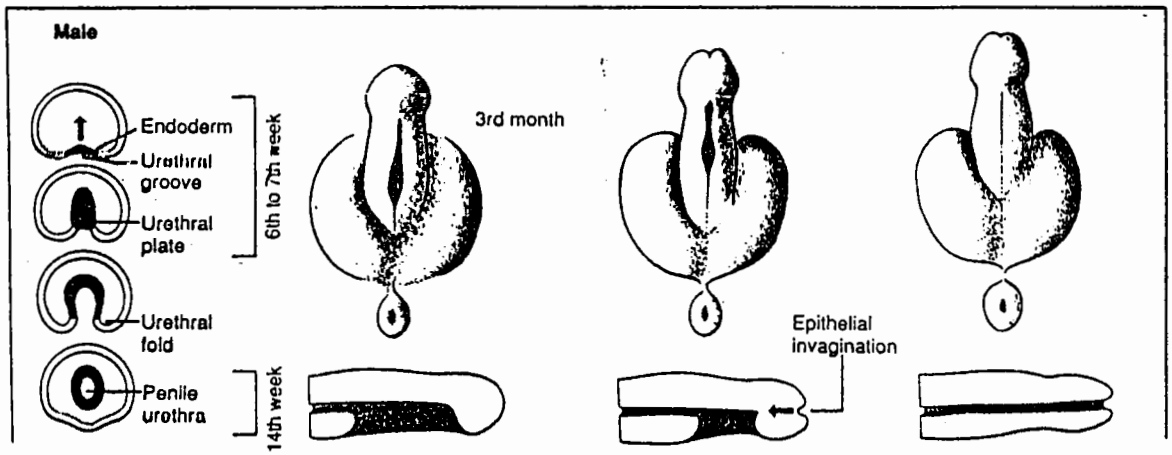
EXTERNAL GENITALIA

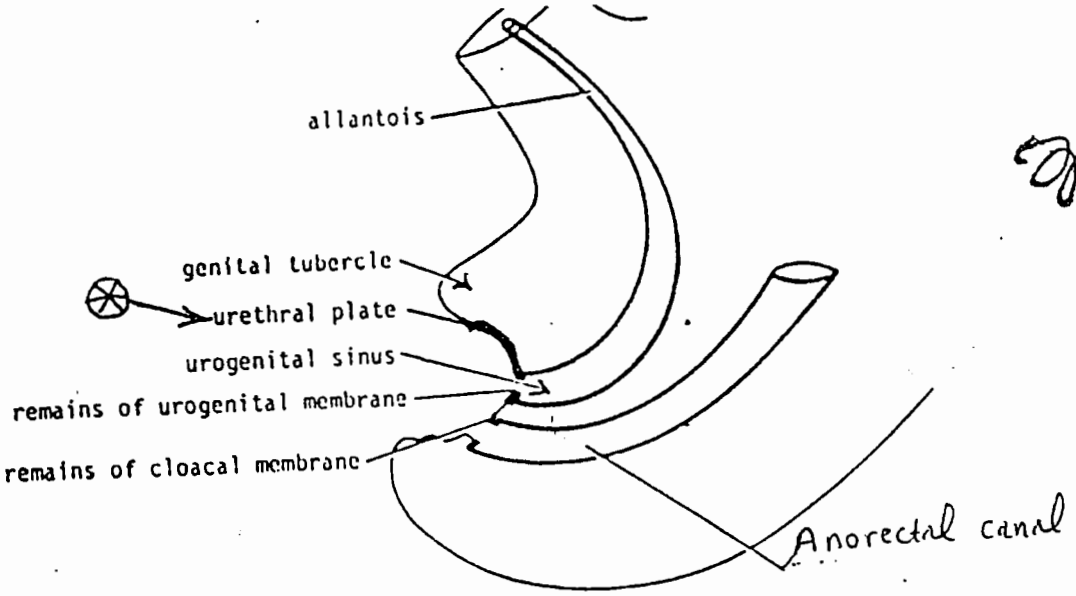
Indifferent Stage

In the 3rd week of development, mesenchyme cells, originating in the region of the primitive streak, migrate around the cloacal membrane to form a pair of slightly elevated folds, the **cloacal folds** (Fig. 15-27A). Directly cranial to the cloacal membrane, the folds unite to form the **genital tubercle**. When, in the 6th week, the cloacal membrane is subdivided into the urogenital and anal membranes, the cloacal folds are likewise subdivided into the **urethral folds** anteriorly and the **anal folds** posteriorly (Fig. 15-27B).

In the mean time, another pair of elevations, the **genital swellings**, become visible on each side of the urethral folds. In the male, these swellings later form the **scrotal swellings** (Fig. 15-28A), and in the female the **labia majora** (Fig. 15-31B). At the end of the 6th week, however, it is impossible to distinguish between the two sexes (Fig. 15-27B).





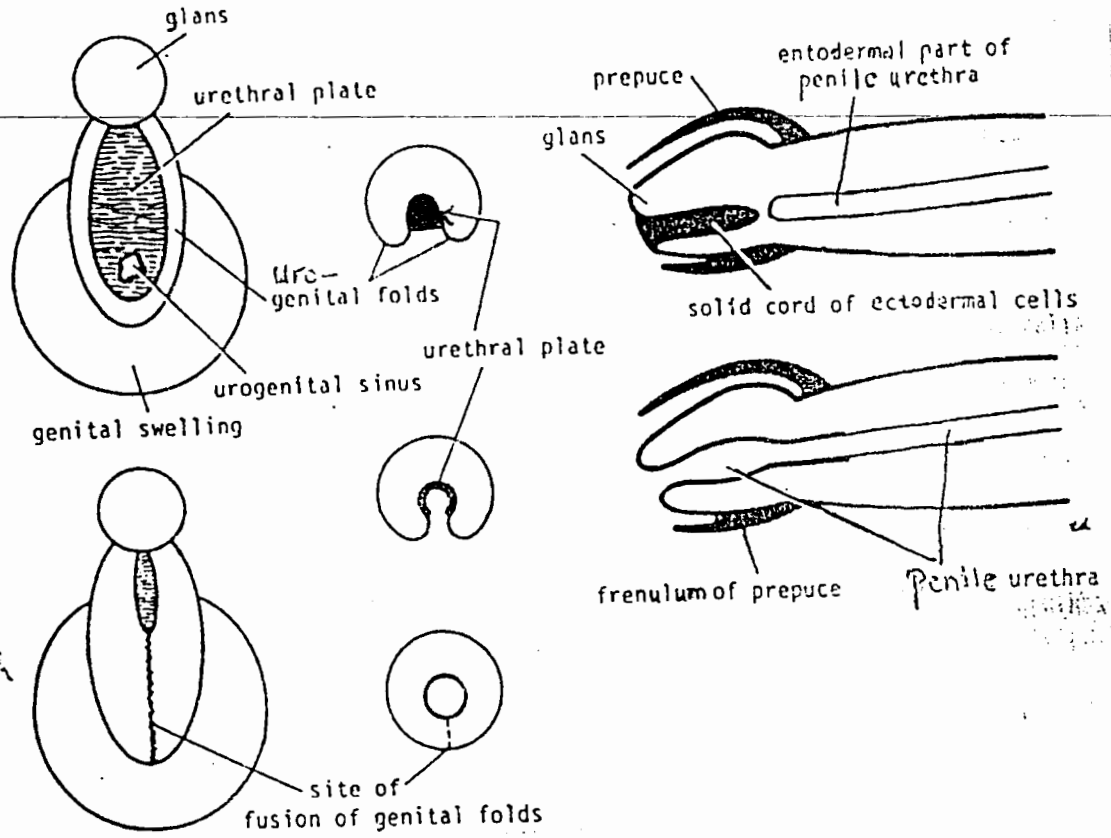


Development of the male external genitalia (Fig. 18-4)

A. The testes produce androgens which stimulate the growth and development of the male external genitalia.

B. As the phallus elongates rapidly to form the penis, it pulls the urogenital folds forward, forming a urethral groove which is continuous with the urogenital opening.

C. The cavity of the definitive urogenital sinus extends onto the surface of the enlarging genital tubercle in the form of an endoderm-lined **urethral groove** during the sixth week (Fig. 10-18B). This groove becomes temporarily filled by a solid endodermal **urethral plate**, but the urethral plate then recanalizes to form an even deeper groove. In males this groove is relatively long and broad, whereas in females it is shorter.



D. By the fusion of the urogenital folds, the urethral plate is transformed into a canal, the penile urethra. The penile urethra does not extend to the tip of the glans.

E. At the tip of the glans, an ectodermal cell cord grows inward, extending toward the lumen of the urethra. This cell cord later obtains a lumen and in this way a continuous channel is established from the base of the bladder to the end of the penis.

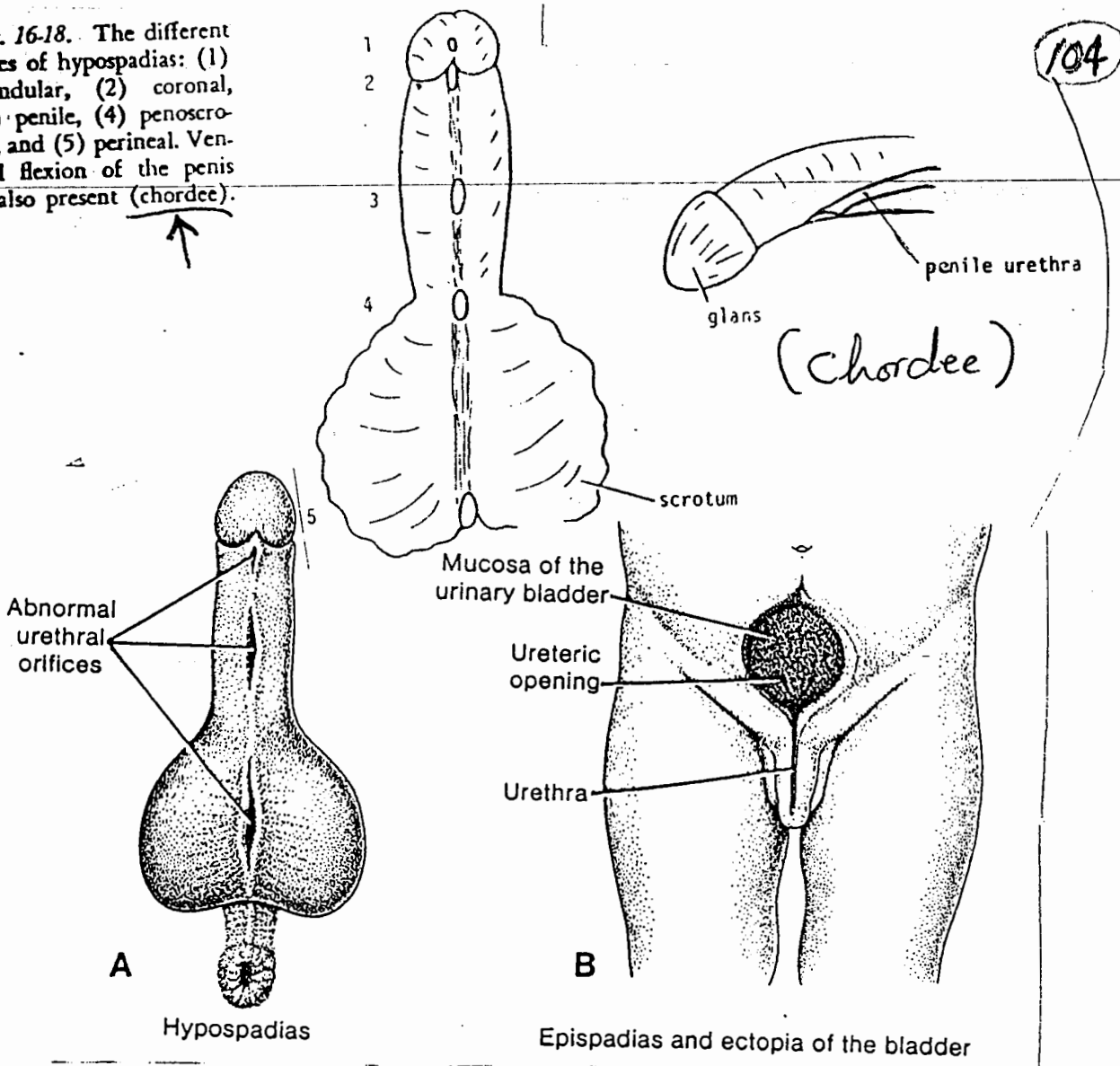
F. The skin at the base of the glans grows over the glans to form the prepuce (foreskin).

G. The labioscrotal swellings migrate caudally, grow toward each other, and fuse to form the scrotum.

Table 10-1. Development of Male and Female External Genitalia

PRESUMPTIVE ANLAGE	MALE STRUCTURE	FEMALE STRUCTURE
Genital tubercle	Glans and shaft of penis	Glans and shaft of clitoris
Definitive urogenital sinus	Penile urethra	Vestibule of vagina
Urethral fold	Penis surrounding penile urethra	Labia minora
Labioscrotal fold	Scrotum	Labia majora

Fig. 16-18. The different types of hypospadias: (1) glandular, (2) coronal, (3) penile, (4) penoscrotal, and (5) perineal. Ventral flexion of the penis is also present (chordee).



Malformations of the penile urethra

A. Hypospadias
   1. Results from incomplete fusion of the urogenital folds.
   2. Is also associated with failure of the ectodermal ingrowth from the tip of the penis.
   3. Abnormal openings of the urethra may be found along the inferior aspect of the penis.
   4. Usually the penis is underdeveloped.
   5. When fusion of the urethral folds fails entirely, there may be a sagittal slit along the entire length of the penis and the scrotum.
   6. This condition may result from inadequate production of androgens at the critical stage of fusion of urethral folds.

B. Epispadias results when the genital tubercle develops caudally in the region of the urorectal septum; and when the urogenital membrane ruptures, the urogenital sinus opens on the dorsal (upper) surface of the penis. Therefore, the urethral opening is found on the dorsum of the penis. It is often associated with exstrophy of the bladder.

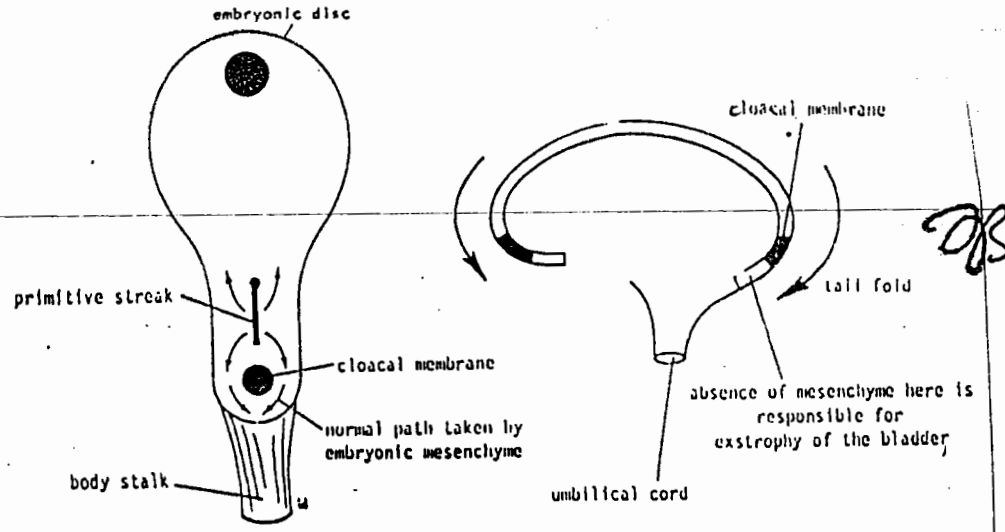


Fig. 15-16. Exstrophy of the bladder and the basis for its occurrence.

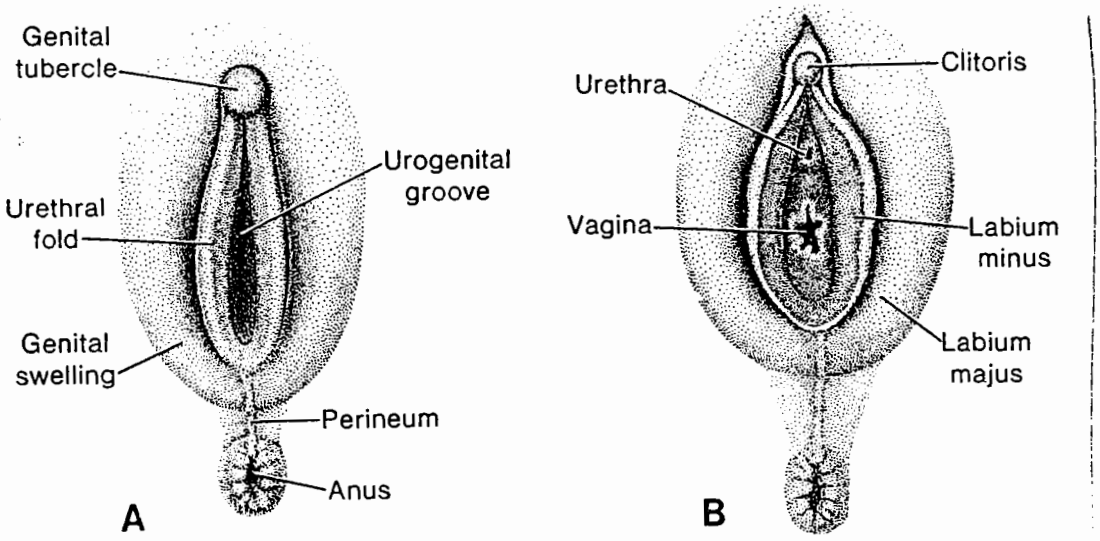


Figure 15-31. Development of the external genitalia in the female at 5 months (**A**), and in the newborn (**B**).

III. Development of the female external genitalia (Fig. 18-4)
   A. In the absence of androgens, the maternal and placental estrogens stimulate the external genitalia to become female.
   B. The phallus undergoes only slight enlargement and becomes the clitoris.
   C. The urogenital folds do not fuse; but develop into the labia minora.
   D. The genital swellings enlarge and form the labia majora. They fuse anteriorly to produce an elevation, the mons pubis, and their posterior margins unite to form the posterior labial commissure.
   E. The region between the labia minora represents the phallic part of the urogenital sinus that was not enclosed by urogenital folds. This becomes the vestibule of vagina into which both the urethra and the vagina open.

1. Epoophoron

It consists of 10-15 parallel tubules situated in the lateral part of mesosalpinx between the ovary and uterine tube. Inferiorly the tubles end blindly near the hilus of ovary. Their upper ends open into a rudimentary duct, the duct of epoophoron. These tubules represent the cranial mesonephric tubules which are attached to the mesonephric duct.

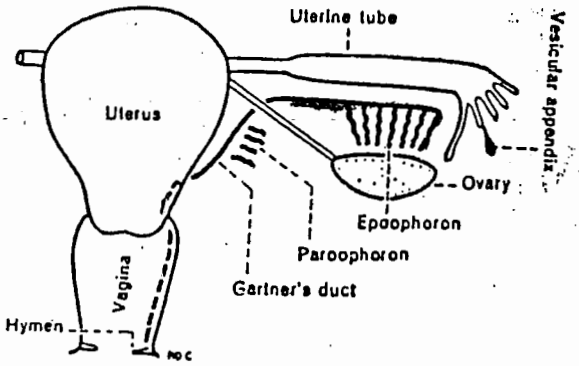


Fig. 332 Vestigeal remnants of the mesonepheric tubules and duct in a female, shown on right side.

2. Paroophoron

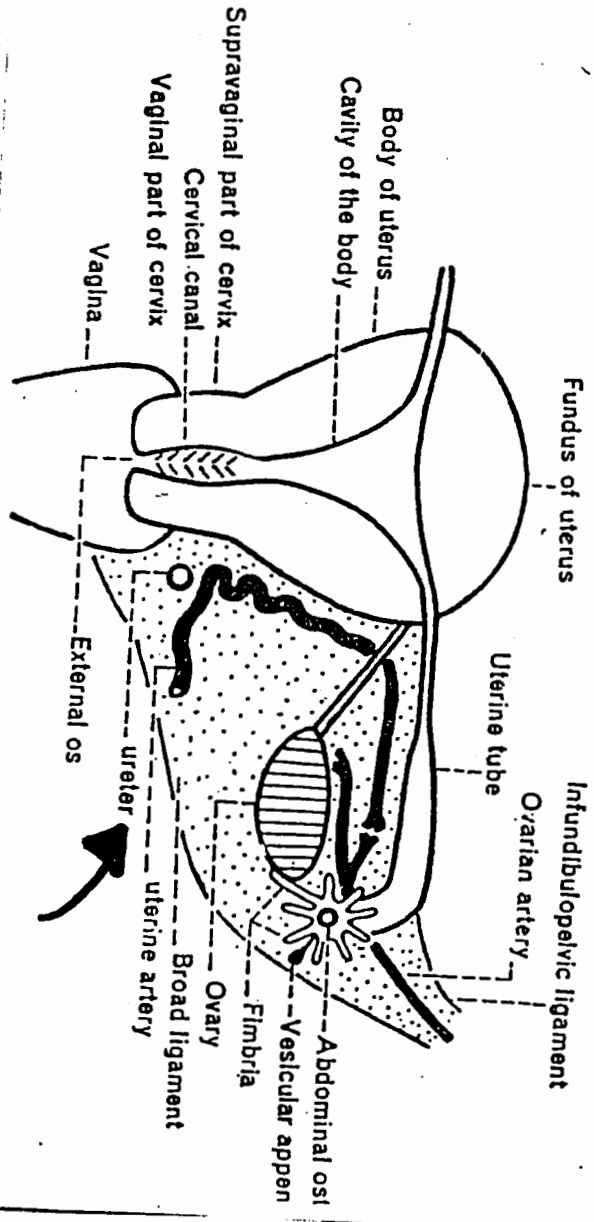
It consists of a few very short rudimentary tubules situated in the broad ligament between the ovary and uterus. Their both the ends are blind. They represent the caudal mesonephric tubules which are not attached to the mesonephric duct.

3. Duct of Epoophoron

When it persists it is called the *duct of Gartner*. It can be traced first along the uterine tube, and then along the lateral margin of uterus up to the level of internal os. Further down it runs through the cervix and lateral wall of vagina, and ends near the free margin of hymen. It represents the mesonephric duct. It may form a cyst in the anterior or lateral wall of vagina.

4. Vesicular Appendix

Occasionally, one or two pedunculated cysts are found attached to the fimbriated end of the tube. These are called the *vesicular appendices* or *paramesonephric appendices*. These are thought to develop from the cranial end of paramesonephric duct.



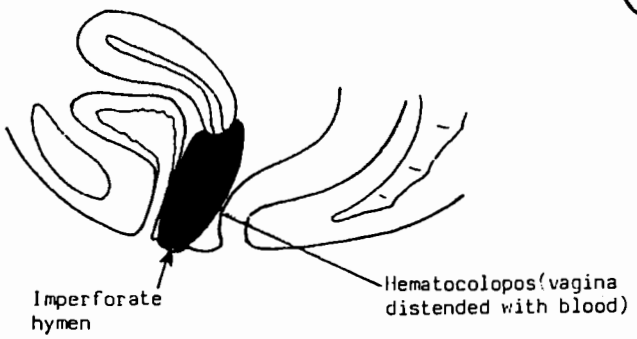
III. Intersexuality or hermaphroditism

A. A true hermaphrodite possesses the gonads and external genitalia of both sexes. Although patients who possess both testicular and ovarian tissue have been observed, true hermaphroditism has never been reported in humans.

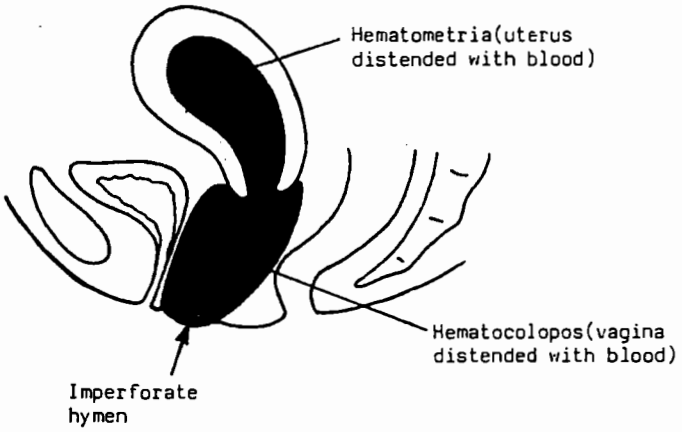
B. Pseudohermaphrodites are individuals who have either testes (male pseudohermaphrodite) or ovaries (female pseudohermaphrodite) and possess the external genitalia resembling the opposite sex. The sex of children born with this condition is often mistaken at birth. The mistake could be resolved by identification of a sex chromatin body in female cells.

1. Female pseudohermaphroditism (adrenogenital syndrome) results from congenital adrenal hyperplasia.
   a) The adrenals produce excessive amounts of androgens which cause the external genitalia to develop in a male direction.
   b) As these persons have a 44 + XX chromosomal complement, the ovaries develop normally.
   c) Frequently there is clitoris hypertrophy, partial fusion of the labia majora, and a persistent urogenital sinus.
   d) About half of all cases with ambiguous external genitalia are female pseudohermaphrodites.
   e) The administration of progestins during pregnancy may cause similar abnormalities.
   f) Early recognition and treatment of the associated adrenal disorder is very important.
2. Male pseudohermaphroditism results from either an inadequate amount of androgen production or androgens produced after the end of the period of maximum tissue sensitivity of the sexual structures. Patients have 44 + XY chromosomal complements but the internal and external genitalia are variable, resulting from varying degrees of development of the phallus and paramesonephric ducts.

Remember: The commonest enzymatic defect in Adrenogenital syndrome is deficiency of 21β-hydroxylase → the adrenal cortex will not be able to synthesize mineralocorticoids or glucocorticoids → steroid intermediates will accumulate above the enzyme block and be shunted towards the production of the adrenal androgens which then cause virilization of female (penis-like clitoris and scrotum-like labia).

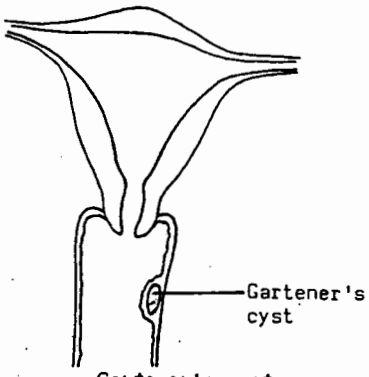


Hematocolopos



Hematocolopos and hematometria

14- Gartener's cyst: This is a cystic swelling which is sometimes seen in the wall of the vagina. The cyst represents a nonobliterated remnant of the mesonephric duct(duct of epoophoron or Gartener's duct).



Gartener's cyst

15- Congenital rectovaginal fistula: This condition may be explained by incomplete development of the urorectal septum. In addition, the uterine canal abnormally opens into the anorectal dorsal part of the cloaca.

16- Congenital vesicovaginal fistula: This is explained by an abnormal opening of the uterine canal into the vesicourethral canal instead of joining the definitive urogenital sinus.

Sex abnormalities as Reflected on the Duct System & External genitalia (09)

Bustami

(1) Turner's Syndrome

→ found in Patients with 44 autosomes and one X chromosome → 45XO → "Absent Barr body"
→ After migration most of the germ cells degenerate
→ Since Y chromosome is absent → the placental & maternal oestrogens will influence development of the Paramesonephric duct system & the external genitalia as in the normal female however they stop development after birth (the gonads do not Produce any hormone after birth → gonadal dysgenesis) → sex characteristics remain infantile

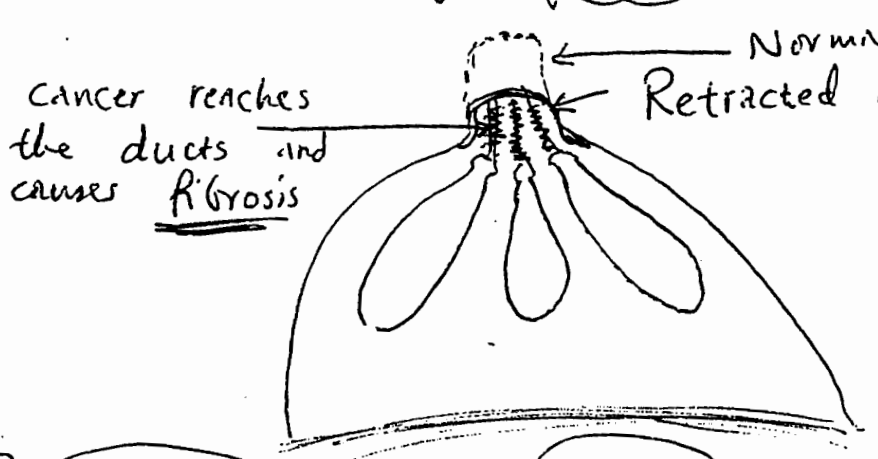
(2) Pure Gonadal dysgenesis

- No abnormalities in chromosomes in (No. or structure)
- Chromosomal complement may be

$$44 \oplus XX \quad \text{OR} \quad 44 \oplus XY$$

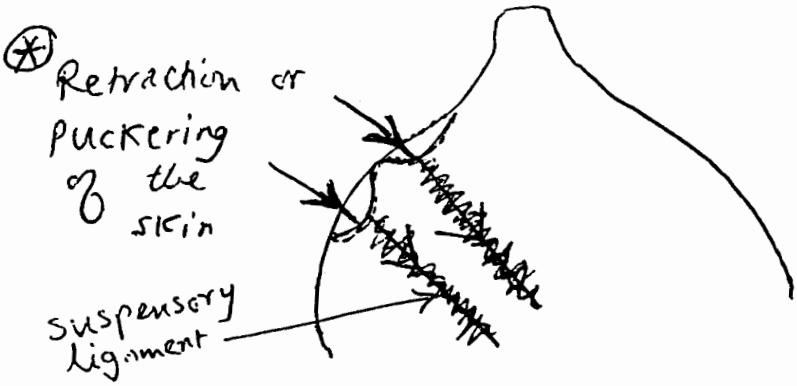
- Primordial germ cells do not form or do not migrate into gonadal area → Neither ovary Nor testis develop
- In the absence of androgens & MIF → Paramesonephric duct & external genitalia are influenced by maternal & placental estrogens But their differentiation stops after birth
- other symptoms of Turn's syndrome are absent (Chromosomes are Normal)

Carcinoma of the Breast may give rise to the following features:



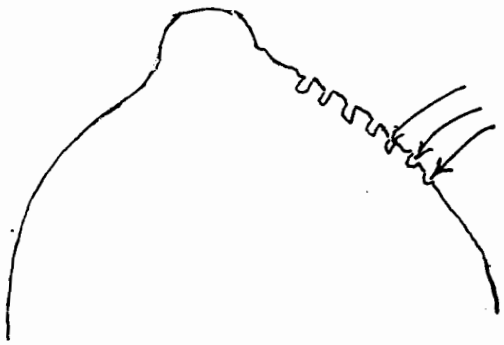
Bustami

(1) Retraction of the Nipple is due to extension of cancer along the lactiferous ducts which causes fibrosis of these ducts & their shortening



(2) Retraction or puckering of the skin due to invasion of the suspensory ligaments of Cooper

(3)



Peau d' orange
OR
oedema with pitting
↓
due to obstruction of cutaneous lymphatics by cancer cells

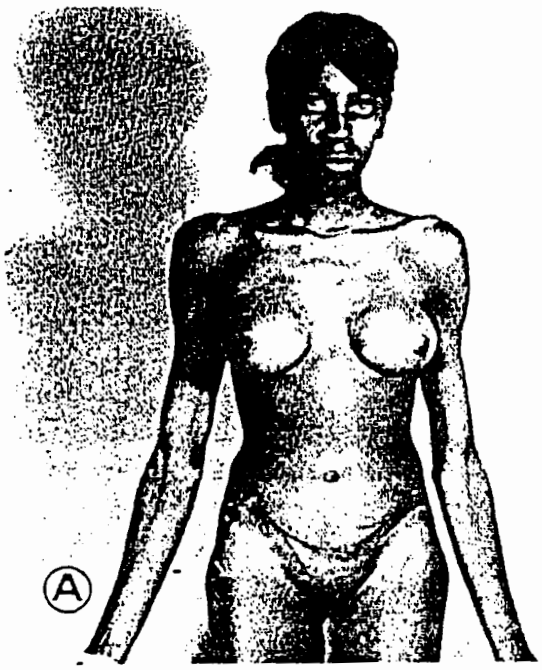
Androgen insensitivity Synd.

(Testicular feminization syndrome) 110

- Patients have (44 + XY) chromosome complement but have the external appearance of normal females

Bustami

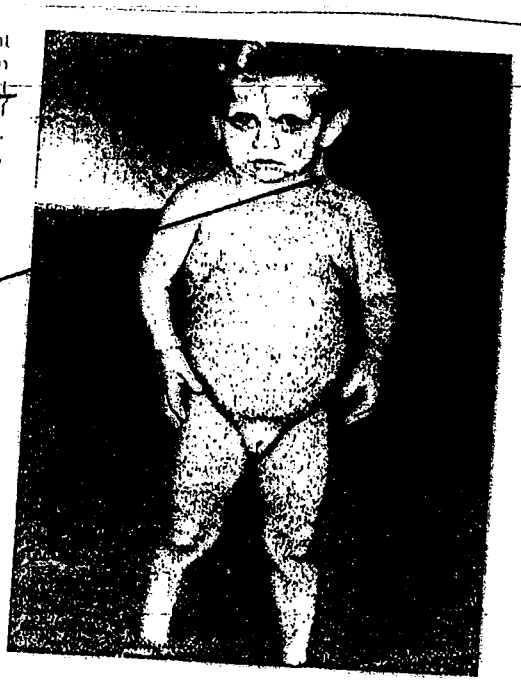
The tissues of the external genitalia are unresponsive to the androgens produced by the testis → they develop and differentiate as in the normal females under the influence of maternal & placental estrogens



→ Since the testes also produce MIF substance, the paramesonephric duct system is suppressed and the uterine tube and the uterus do not develop → vagina is short & ends blindly

→ The testes are found in the inguinal or labial region, but spermatogenesis does not occur.

Figure 8-5. Photograph of patient with Turner's syndrome. The main characteristics are webbed neck, short stature, broad chest, and absence of sexual maturation. (Courtesy Dr. J. Miller, Department of Neurology, University of Virginia.)



Webbed neck in Turner's Syndrome

Turner's Syndrome (e.g of Sex chromosome abnormalities)

↓

(A Single X chromosome)

- This condition, found in women is characterized by the absence of the ovaries (gonadal dysgenesis)
- the cells have only 45 chromosomes with an **XO** Chromosomal complement
- usually caused by nondisjunction in the male gamete during meiosis
- incidence: 2 in 3000
- features of Turner's syndrome:
  Short Stature webbed neck broad chest with wide-spaced nipples
- lymphoedema of the leg from birth
- 2ry sexual characteristics do not appear (uterus & vagina may be small)

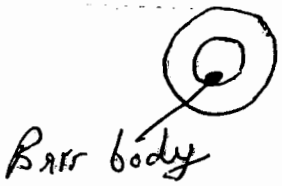
↓

If a buccal smear was examined for Barr bodies? → The normal ♀ has one (XX) → normal ♂ (XY) & patients with Turner's Syndromes (XO) have none

Barr Body

(150/C)

- inactivated X chromosome which can be seen by light microscopy near the nuclear membrane → sex chromatin body
- its presence indicates that the individual possesses 2 X chromosomes but does not necessarily mean that the person is genetically normal



The sex chromosome make up might be XXY instead of XX

↓

If the individual possesses an extra X i.e XXX → there will be two Barr bodies in some nuclei i.e there is always one Barr body less than the number of X chromosomes

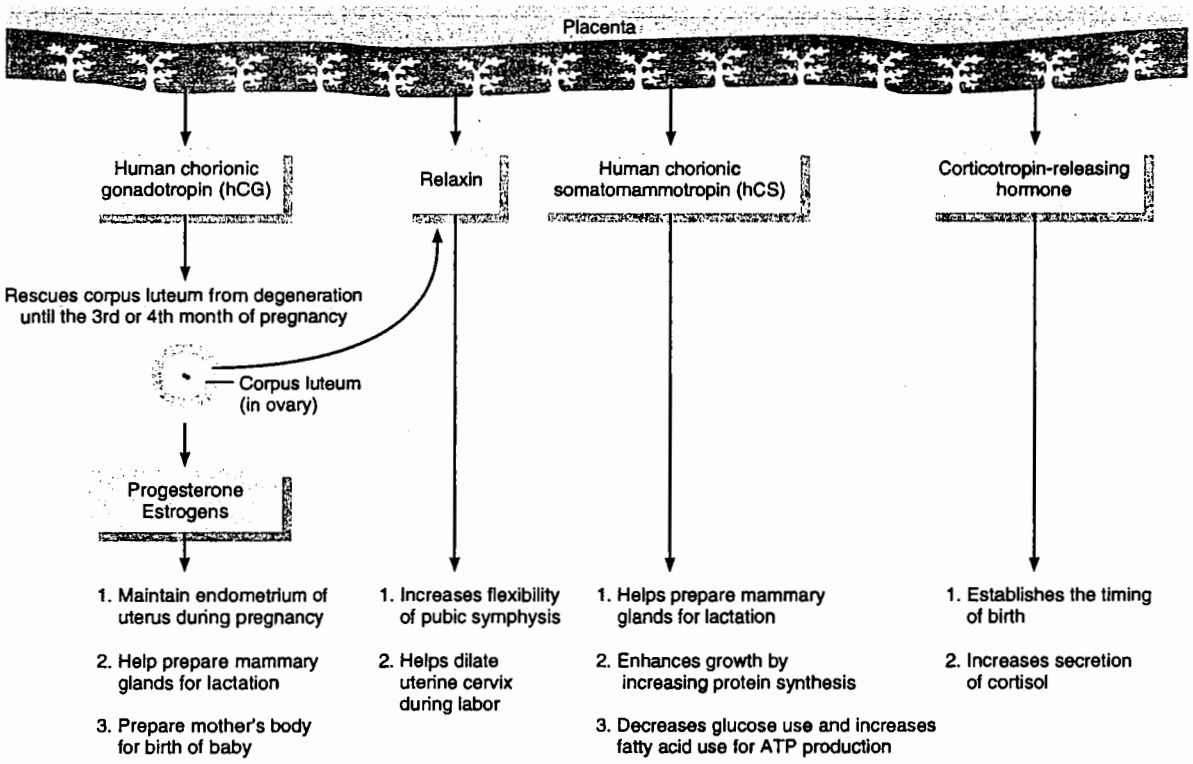
↓

Absence of Barr body ? indicates that the individual has one X chromosome i.e the subject is usually a male XY but may be incomplete female XO

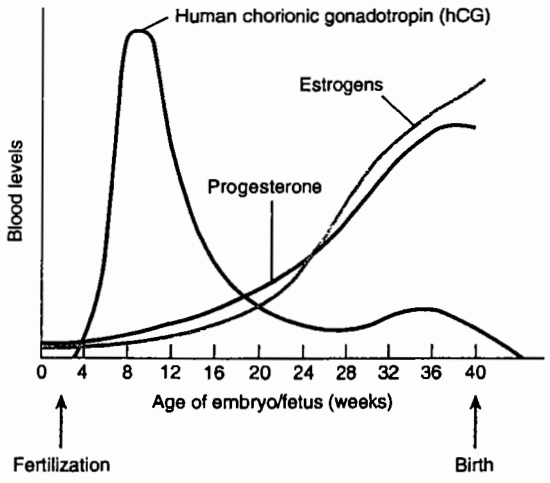
- Polymorphonuclear leucocytes in females commonly have a small mass of chromatin projecting from the nucleus i.e in the form of drumstick

Figure 29.16 Hormones during pregnancy.

Whereas the corpus luteum produces progesterone and estrogens during the first 3–4 months of pregnancy, the placenta assumes this function from the third month on.



(a) Sources and functions of hormones



(b) Blood levels of hormones during pregnancy

Which hormone is detected by early pregnancy tests?

alike. For example, hCS causes decreased use of glucose by the mother, thus making more available for the fetus. Additionally, hCS promotes the release of fatty acids from adipose tissue, providing an alternative to glucose for the mother's ATP production.

The hormone most recently found to be produced by the placenta is **corticotropin-releasing hormone (CRH),** which in nonpregnant people is secreted only by neurosecretory cells in the hypothalamus. CRH is now thought to be part of the "clock" that establishes the timing of birth. Secretion of CRH by the placenta begins at about 12 weeks and increases enormously toward the end of pregnancy. Women who have higher levels of CRH earlier in pregnancy are more likely to deliver prematurely, whereas those who have low levels are more likely to deliver after their due date. CRH from the placenta has a second important effect: It increases secretion of cortisol, which is needed for maturation of the fetal lungs and the production of surfactant.

Early Pregnancy Tests

Early pregnancy tests detect the tiny amounts of human chorionic gonadotropin (hCG) in the urine that begin to be excreted