Benign diseases of the vulva and vagina

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Preface

The chief impetus for writing this book was the realization that benign diseases of the vulva and vagina, although constituting a large percentage of all gynecologic disease, are only sketchily covered in most of the general gynecologic texts. We believe that a single comprehensive volume devoted especially to the clinical and pathologic features of these diseases will prove useful to the physician whose practice, in whole or in part, consists of gynecology.

Many of the affections discussed come first to the attention of the dermatologist or urologist, a fact pointing up the overlapping interests of these specialties with gynecology. We hope that the subjects are presented in such a manner as to be of interest also to the dermatologist, urologist, and perhaps the pathologist.

We have endeavored to include all benign diseases involving the vulva and vagina that are likely to be encountered by clinicians practicing in the temperate zones of the world. Some lesions only rarely observed in the United States are included, with the thought that a ready reference source might preclude their complete disregard in differential diagnosis should they be presented unexpectedly. Several conditions are discussed that should have been omitted, while others have been omitted that probably should have been included. In view of the many voluminous treatises on the subject of invasive malignant lesions, these have been purposely omitted. Intraepithelial carcinoma of the vulva is included, however, because of its position between benign disease and invasive cancer. Such lesions as fistulas and relaxations, although intimately related to the vulva and vagina, do not fall within the scope of the planned text. The length of the discussions of most lesions is somewhat in keeping with their incidence and importance, although the length of the discussion of others is, perhaps, in accord with our own special interests. If the space allotted to histopathologic descriptions seems excessive in comparison to that in most nonpathology textbooks, it is because of our conviction that a knowledge of pathology is necessary for a thorough understanding of disease processes and for the optimal care of patients.

Despite every effort to give proper credit for significant contributions, such an accomplishment may have proved unattainable. Any failure in this regard is far from intentional. We are indebted to the many authors whose ideas have been incorporated. Some of the opinions expressed may be in conflict with published reports; in every such instance, however, we have attempted to weigh personal experiences with these opinions. In no instance has a contradictory opinion been advanced solely for the purpose of disagreement.

Most of the illustrations are from our personal files, the majority of those of gross lesions having been made from colored slides. Many authors and friends have graciously contributed other illustrations that seemed particularly instructive. A glossary of selected terms frequently used by the pathologist and

the dermatologist is included for the use of students and clinicians to whom they may not be entirely familiar.

We were fortunate in being able to persuade Doctor Arnold A. Zimmermann to write the chapter on embryology. His many years of intimate contact with the subject as Professor of Anatomy at Baylor College of Medicine make him eminently qualified for the task. Doctors Robert R. Franklin and L. Russell Malinak, Department of Obstetrics and Gynecology at Baylor College of Medicine, because of their special knowledge and interest, were asked to contribute the chapter on developmental anomalies.

We wish to acknowledge gratefully the invaluable help and advice of our many colleagues and friends of the Houston medical community. These include Doctors John M. Knox, Marvin E. Chernosky, Elizabeth W. Rauschkolb, Joseph M. Glicksman, and Robert G. Freeman, all of the Department of Dermatology, Baylor College of Medicine; Doctor Willson J. Fahlberg, of the Department of Microbiology, Baylor College of Medicine; Doctor C. Dean Dukes, of the University of Arizona at Tuscon; and Mr. Reuben D. Wende, Director of Laboratories for the Department of Public Health for the City of Houston.

We wish to thank also Mrs. Allene Jefferson for her extensive editorial counsel, Miss Mary Smith for accumulating the bibliographic data and for editorial work, Miss Juanita Wells, M.T., for her many years of laboratory assistance and data processing, and Miss Joyce Lessard for her secretarial help. Particular indebtedness is expressed to Mrs. Velma Heiser, a long-time employee, for her dedicated efforts in every phase of the preparation and typing of the manuscript. The visual education departments of Baylor College of Medicine and the Methodist Hospital have been most cooperative in the preparation of the illustrations.

Finally, we wish to express apologies to our wives, LeNan and Patricia, for the deprivations that they experienced while this work was in progress, as well as appreciation of their tolerance of our occasional altered moods, which we only too willingly attributed to the undertaking.

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Contents

Color plates

Chapter 1

Anatomy of the vulva and vagina

THE VULVA

The vulva (Fig. 1-1) is that part of the female anatomy between the genitocrural folds laterally, the mons pubis anteriorly, and the anus posteriorly. It is composed of the labia majora, labia minora, mons pubis, clitoris, vestibule, urinary meatus, vaginal orifice, hymen, Bartholin's glands, Skene's ducts, and the vestibulovaginal bulbs.

Labia majora

Gross anatomy

The lateral boundaries of the vulva are formed by the labia majora, which consist of two large folds of adipose and fibrous tissue. Anteriorly, the labia majora fuse into the mons pubis; posteriorly, they become narrower and flatter and terminate 3 to 4 cm. anterior to the anus, where they are united by the posterior commissure or fourchette. The lateral aspects of the labia majora are covered by a considerable amount of coarse hair, and the inner aspects by little or no hair. Prior to puberty, the labia majora are inconspicuous and the labia minora protrude between them in a conspicuous fashion. At puberty, the labia majora develop as one of the secondary sexual characteristics, with their lateral aspects and the mons becoming covered by coarse hairs. In most women, the upper border of this hair is sharply defined. The skin of the labia majora is usually somewhat darker than the adjacent skin.

Histology

The labia majora are covered by skin composed of an outer lining of stratified squamous epithelium (Fig. 1-2) consisting of a basal layer of cells, the stratum malpighii, a thin granular layer (occasionally absent),

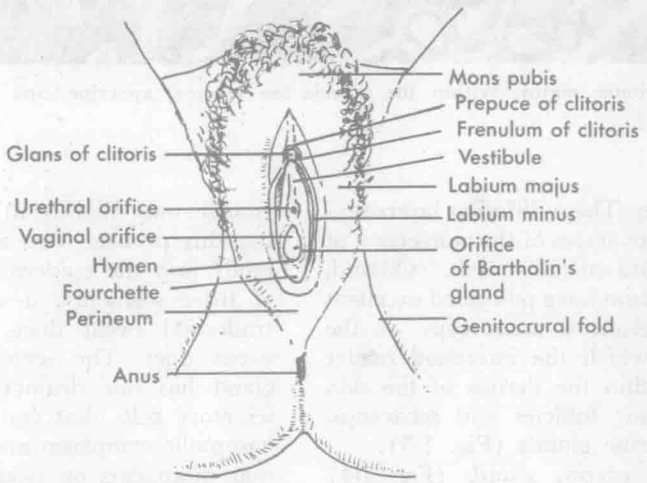

Fig. 1-1. Diagram of the vulva, showing its principal parts.

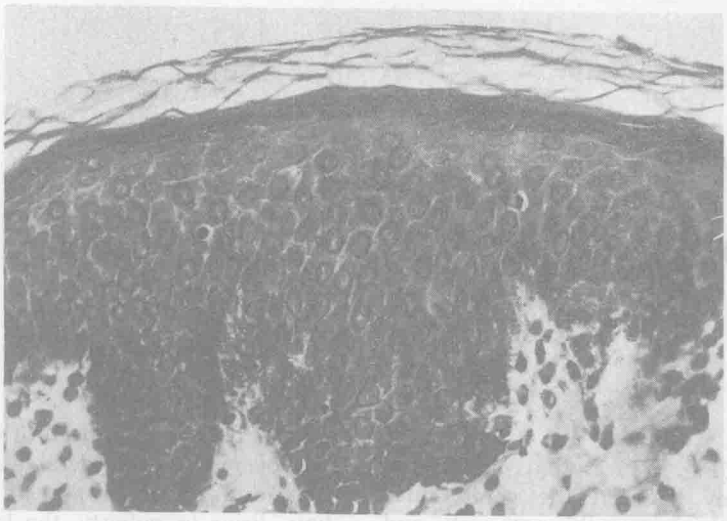

Fig. 1-2. Labium majus. Squamous epithelium; a thin granular layer and a slight degree of keratinization. (H & E × 495)

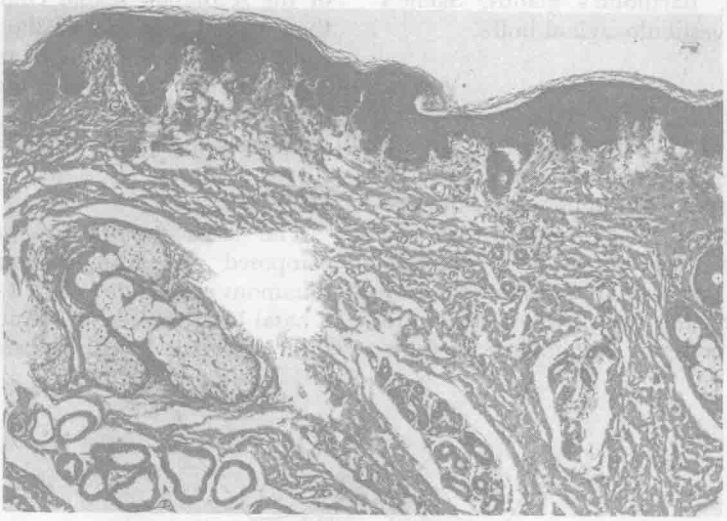

Fig. 1-3. Labium majus. Within the dermis are eccrine, apocrine, and sebaceous glands. (H & E × 77)

and a horny layer. These different layers represent the different stages of the conversion of the basal cells into cornified cells. (Odland, Selby, and Zellickson have published excellent works on the electron microscopy of the human skin to which the interested reader is referred.) Within the dermis of the skin are numerous hair follicles and sebaceous, sweat, and apocrine glands (Fig. 1-3).

The *sweat* or *eccrine glands* (Fig. 1-4) are tubular structures whose cells secrete and change only slightly in size and shape during this process. The sweat glands lead directly into the epidermis and are composed of three segments: a secretory portion, intradermal sweat duct, and intraepidermal sweat duct. The secretory portion of the gland has one distinct layer, consisting of secretory cells that contain a clear, slightly basophilic cytoplasm and abundant glycogen that disappears on sweating. Small myoepithelial cells are scattered between the bases

Fig. 1-4. Labium majus. Eccrine sweat glands and ducts. Arrow points to a myoepithelial cell. (H & E × 495)

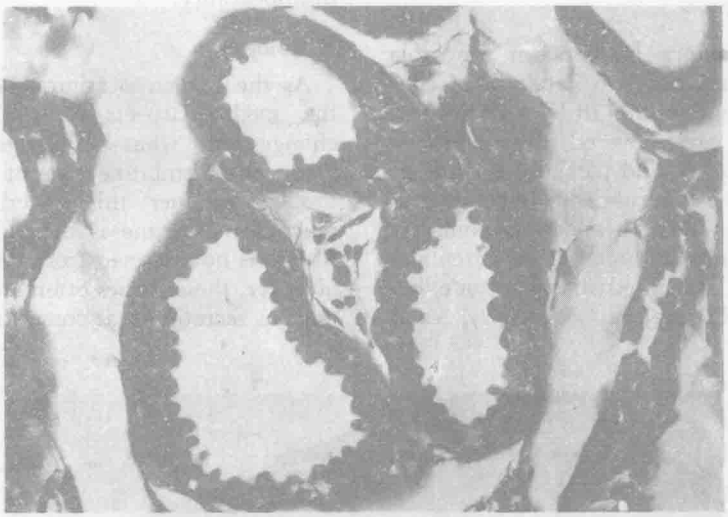

Fig. 1-5. Labium majus. Apocrine glands. Note variations in height of the secretory cells. The lumina of the glands are large as compared to those of the eccrine sweat glands. (H & E × 495)

of the secretory cells; contraction of the myoepithelial cells forces out the secretion of the latter. The intradermal portion of the sweat duct is composed of two layers of cuboidal, deeply basophilic cells. The intraepidermal duct consists of a single layer of cuboidal cells surrounded by a sheath of epidermal cells.

The *sebaceous glands* are alveolar, holocrine glands that have no lumen. Their secretion, which is formed by decomposition of the cells, is evacuated through the sebaceous ducts into pilosebaceous follicles. These follicles may or may not contain hair. Each gland is composed of several lobules, and at the periphery of each lobule one layer of deeply basophilic cuboidal cells, referred to as the *generative cells,* is present. The cells within the central portion of each lobule have a cytoplasm arranged in a delicate network and are filled by fat.

The *apocrine glands* (Fig. 1-5) of the

labia majora are identical to those found in the axilla, breasts, and perianal regions. Sometimes referred to as the scent glands, the apocrine glands are tubular structures whose cells pass through a full cycle of secretory stages. At the beginning of the cycle the cells are of a low cuboidal type. Gradually, they increase in height until they protrude into the lumen; then, upon releasing their secretion, they again become low cuboidal. Recent studies by Montes, Baker, and Curtis suggest that, contrary to prior opinion, the lining cells in these glands do not release part of their cytoplasm into the lumen during secretion. The secretory portion of the apocrine gland is lined by a distinct layer of epithelial cells that varies in height, depending upon the stage of the secretory cycle. The duct is lined by two layers of cuboidal epithelial cells.

Labia minora

Gross anatomy

The labia minora lie between the labia majora and consist of two flat folds of connective tissue that contain little or no adipose tissue. They are covered by skin on their lateral aspects, and partially so on their medial aspects. As a rule, the labia minora are 4 to 5 cm. in length and approximately 0.5 cm. in thickness, although in some females they are more elongated and protrude between the labia majora. Anteriorly, each divides into two parts, one passing over the clitoris to form the prepuce, or foreskin, and the other passing behind the clitoris to form the frenulum. Posteriorly, they tend to become smaller and blend with the medial surfaces of the labia majora or to unite anterior to the posterior commissure to form the fourchette. The cleft between the two labia minora is referred to as the *vestibule*, and into this vestibule open the vagina, the urethra, Skene's ducts, and Bartholin's gland ducts. Between the posterior fourchette and hymeneal ring is a depression, the *fossa navicularis*. As already mentioned, the labia minora are relatively more prominent in children, since the labia majora are not well-developed at this time. In the postmenopausal woman the labia majora tend to atrophy, and the labia minora again become relatively prominent. Occasionally, the labia minora may atrophy almost to the point of disappearance.

Histology

As the hymen is approached, the skin on the medial aspects of the labia minora changes into what many morphologists call a mucous membrane type of epithelium, although whether this covering is skin or mucous membrane is debatable (Fig. 1-6). Mucus is not secreted from the labia minora; however, these tissues often are bathed in the mucous secretion that comes from Bartholin's

Fig. 1-6. Outer labium minus. The lining epithelium contains little keratin, and the granular layer is indistinct. (H & E × 495)

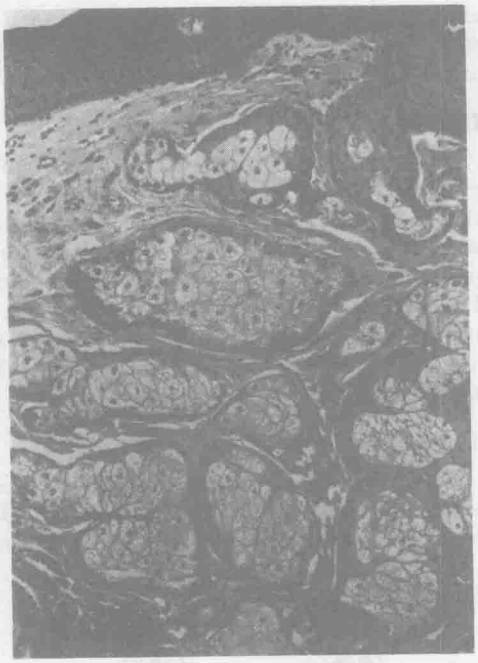

Fig. 1-7. Labium minus. Numerous sebaceous glands are apparent in the dermis. (H & E × 100)

Fig. 1-8. Labium minus. Prominent sebaceous glands within the dermis may result in the presence of pinpoint papular structures.

glands and the cervix. The epithelial covering is much less cornified than true skin; it has a scant or no granular layer, and the dermis contains no hair follicles. The skin and mucosa of the labia minora are extremely rich in sebaceous glands (Figs. 1-7 and 1-8). These glands enter directly into the skin through tunnels in the epithelium. Sweat glands are sparse or completely absent.

The deeper tissues are composed of dense connective tissue containing many veins and some smooth muscle elements. Numerous bundles of elastic tissue are mixed within the connective tissue, and a minimal amount of fatty tissue is present. During sexual excitement, the labia minora frequently become swollen and congested and take on the appearance of erectile tissue.

The clitoris

Gross anatomy

The clitoris is the homologue of the penis. It consists of two cylindrical erectile bodies, called the *corpora cavernosa*, that terminate in the vestibule as the glans. The body of the

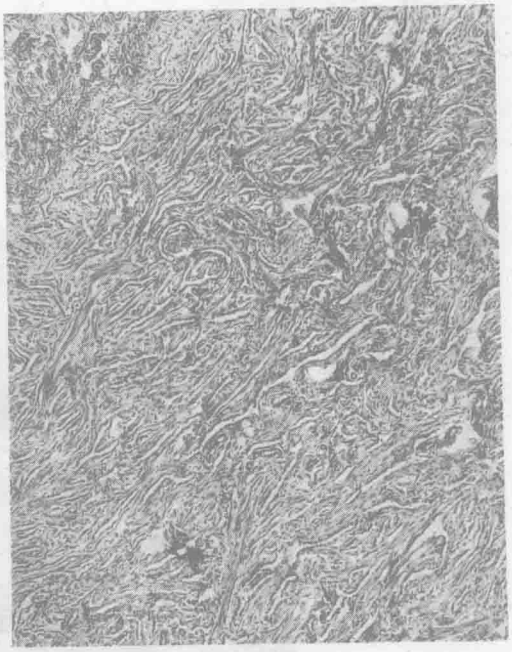

Fig. 1-9. Section through clitoris of 2-year-old child showing erectile tissue composed of numerous thin-walled vascular channels and nerve fibers. (H & E × 65)

clitoris, which is formed by fusion of the two corpora cavernosa, extends from the pubic arch anteriorly to the glans posteriorly and is approximately 2 cm. in length. At the inferior border of the pubic arch, the two corpora cavernosa separate and follow the inferior border of the inferior rami of the pubic bones, to which they are attached. These are the crura of the clitoris. They are covered by the ischiocavernosus muscles which, by contraction, trap blood within the corpora cavernosa to cause erection of the clitoris.

Histology

The glans clitoris is covered by a mucous membrane containing many specialized nerve end-organs. It is composed of erectile tissue (Fig 1-9) with many large and small venous channels surrounded by large amounts of smooth muscle tissue. This erectile tissue is arranged into the corpora cavernosa.

The vestibule

The vestibule is the portion of the vulva extending from the clitoris to the fourchette that is visible on separation of the labia minora. It is the remains of the urogenital sinus of the embryo. The vagina, ducts of Bartholin's glands, Skene's ducts, and urethra open into the vestibule. The vestibule is covered by a mucosa lined by stratified squamous epithelium.

The hymen

The hymen is a thin membrane of connective tissue over the entrance of the vagina into the vestibule. It has a central

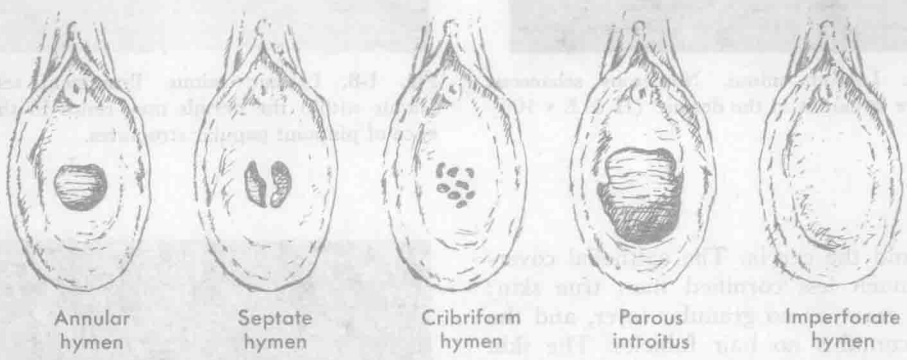

Fig. 1-10. Diagram of various types of hymens described in text.

Fig. 1-11. Section through hymen. A lining of squamous epithelium on both inner and outer surfaces is separated by a relatively thin layer of connective tissue. (H & E × 52)

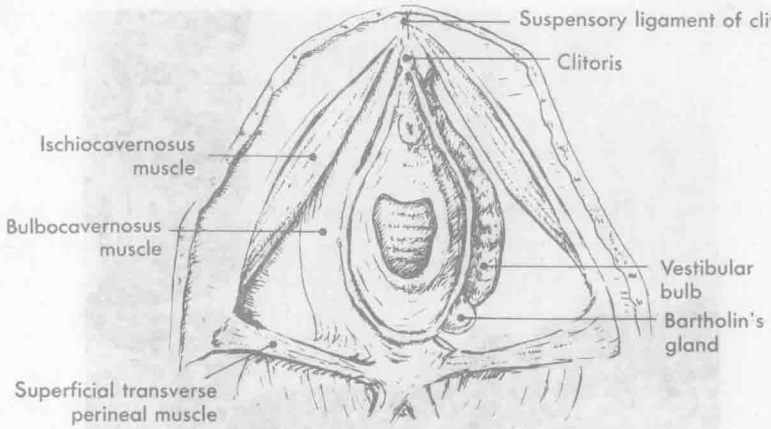

Fig. 1-12. Diagram of deeper structures of vulva, showing the location of Bartholin's glands and vestibular bulb.

opening that is usually round or concentric. In some hymens, a thin mucosal septa may create two or more openings; or the hymen may be cribiform, having numerous small openings through a solid mucosal plate. On rare occasions, the hymen may be imperforate; thus, no communication between the vagina and the vestibule is present (Fig. 1-10).

The hymen varies somewhat in thickness and elasticity. It may be quite elastic and easily stretched without laceration at the initial intercourse, or it may be so tough and rigid as to prevent intercourse. As a rule, the intact hymen is present only in the virgin; after coitus it is represented by small, membranous elevations referred to as the *carunculae myrtiformes.*

The hymen is lined on both sides by a thin mucous membrane composed of stratified squamous epithelium (Fig. 1-11). This epithelium covers a rather variable thickness of connective tissue containing many small blood vessels.

Bartholin's glands

Bartholin's glands, which are homologous to Cowper's glands in the male, lie deep beneath the fascia, one on each side of the vestibule, posterolateral to the vaginal orifice (Fig. 1-12). They are lobulated, racemose glandular structures about the size of small peas, and contain multiple acini grouped around the termination of each of their many branching ducts. The acini are lined by a cuboidal epithelium in which the cells

Fig. 1-13. Bartholin's gland. The acini are typical. In lower portion of the picture, a terminal duct enters several acinar structures. (H & E × 117)

have a clear cytoplasm and dark, basally situated nuclei (Fig. 1-13). The cells contain mucin, which is secreted during sexual excitation, and which serves to lubricate the vaginal orifice and canal. The main duct of a Bartholin's gland opens slightly posterior to the midportion of the lateral margin of the vaginal orifice, just outside the hymeneal ring. The duct is lined by stratified transi-

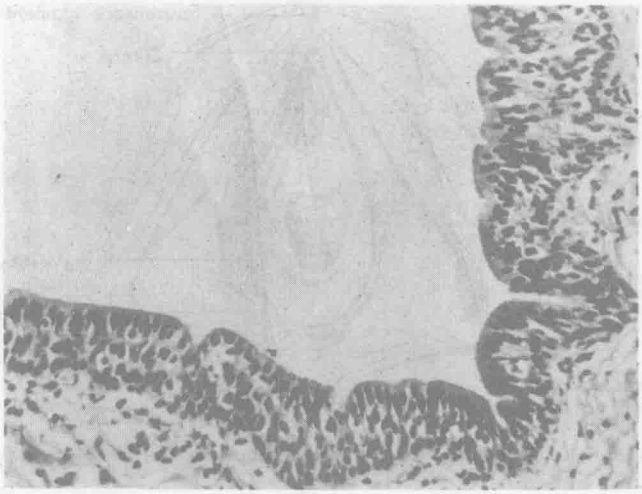

Fig. 1-14. Main Bartholin's duct. The lining is composed of transitional epithelium. (H & E × 286)

tional epithelium (Fig. 1-14) with the exception of the portion near the orifice; this portion is lined by stratified squamous epithelium. As the branching ducts approach the gland, they gradually become smaller and the epithelium becomes flatter, and in their finest, most terminal branches, they are lined by a single layer of cuboidal epithelium. As a rule, so-called Bartholin's gland cysts are actually cysts formed within the main duct of the gland following obstruction to the flow of secretion from this gland. Correctly, these cysts should be referred to as Bartholin's duct cysts.

The bulb of the vestibule

Two oval masses of erectile tissue, called the *bulbi vestibuli* (Fig. 1-12), are incorporated within the bulbocavernosus muscles beneath the floor of the vestibule on each side of the vaginal opening. The bulbi vestibuli are homologous to the bulbus penis of the male. A Bartholin's gland lies in the base of the bulb on each side. Unless affected by trauma, these structures are seldom related to vulvar disease.

The urethra

The urethra opens into the vestibule just anterior to the vaginal introitus. Immediately within the meatus, the urethra is slit-like, with its long axis lying ventrodorsally. The edge of the external orifice is everted and may have two or three overhanging lips; because of these lips, the orifice is occasionally difficult to find. The most distal paraurethral ducts, *Skene's ducts,* open into the urethral canal just within or external to the meatus. According to Huffman, the majority of paraurethral ducts empty into the distal third of the urethral canal, then turn cephalad and divide into many smaller branches (Fig. 1-15). These smaller branches terminate as small bud-like outpocketings and tubular glands, chiefly in the lateral and inferior urethral walls. It is Huffman's opinion that the presence of two lateral ducts, as described by Skene, are the exception rather than the rule.

The *paraurethral glands* are branched tubular glands that empty into the paraurethral ducts. They are lined by epithelium that varies from the low columnar to the cuboidal type. Most of the smaller and many of the larger branches of the paraurethral ducts are lined by pseudostratified or true stratified columnar epithelium. Near the orifices the lining is identical to that of the urethra at that level. If infection develops within the ducts and glands, obstructing the outlet of the duct, abscesses of the anterior vaginal wall may form and rupture into the urethra, ultimately enlarging into diverticula.

The urethra, which measures from 3 to 5.5 cm. in length, extends from the neck of the

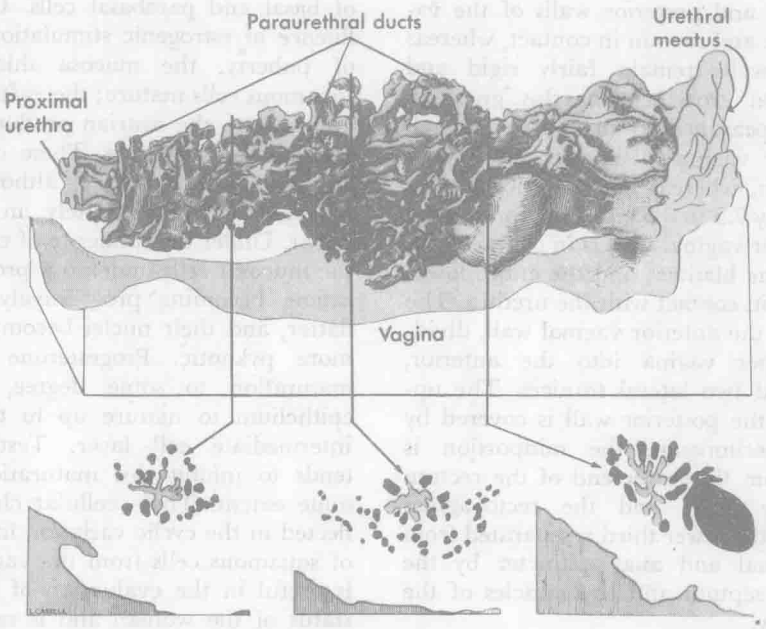

Fig. 1-15. Drawing of a wax model of an adult human female urethra with periurethral ducts and glands, as viewed from the right side. Tissues from which this model was reconstructed were obtained at necropsy of a 20-year-old virgin. The model represents the distal 2.4 cm. of a urethra having a total length of 2.8 cm. No periurethral ducts open at or immediately within the urethral meatus. Thirty-one ducts empty into the urethra, the majority into the distal third; others empty into the middle and proximal thirds. After leaving the urethra, the ducts turn cephalad and extend parallel to the urethral canal. One large duct on the right develops into a cyst of considerable size. At the midpoint in the urethra, many ducts and glands extend laterally far from the canal. At a more proximal level, the urethra is surrounded by many small tubules, and on the right, it is encompassed by a thin, compact, semicircular sheet of ducts and glands. (Huffman, J. W.: Amer. J. Obstet. Gynec. 55:86, 1948.)

bladder to the external urethral orifice. Its proximal two-thirds is usually lined by a stratified transitional epithelium and the outer third by a stratified squamous epithelium. The upper half of the urethra is separated from the anterior vaginal wall by connective tissue, whereas its lower half is firmly adherent to the musculature of the vaginal wall.

Blood, lymph, and nerve supply

The arterial blood supply to the vulva is abundant, coming from branches of the internal pudendal artery, which derives from the internal iliac artery (hypogastric), as well as from branches of the external pudendal artery, which derives from the femoral artery. The veins within the vulva form a large plexus, ultimately emptying into the internal pudendal and external pudendal veins.

The anterior superior portion of the vulva is supplied by the cutaneous branches of the ilioinguinal nerve. The posterior inferior portion receives its nerve supply from the pudendal branches of the posterior femoral cutaneous nerve. Between these two groups of nerves, branches from the posterior labial and perineal branches of the pudendal nerve also extend into the vulva.

The primary lymph channels of the vulva empty into the superficial inguinal lymph nodes. Lymph vessels also pass from the posterior portion of the vulva to the perianal lymph nodes.

THE VAGINA

The vagina extends from the vestibule to the uterus and connects the two. It is directed obliquely upward and backward at an angle approximately 45 degrees to the horizontal; its long axis is parallel to the plane of the pelvic brim and at a right angle to the uterus.

The anterior and posterior walls of the vagina are slack and remain in contact, whereas the lateral walls remain fairly rigid and separated. On cross section, this gives an H-shaped appearance to the vaginal canal. The anterior vaginal wall averages 6 to 7 cm. in length, whereas the posterior wall is approximately 7.5 to 8.5 cm. The upper third of the anterior vaginal wall is in contact with the base of the bladder, and the entire lower two-thirds is in contact with the urethra. The cervix enters the anterior vaginal wall, dividing the upper vagina into the anterior, posterior, and two lateral fornices. The upper third of the posterior wall is covered by cul-de-sac peritoneum, the midportion is separated from the lower end of the rectum by fibrofatty tissue and the rectovaginal septum, and the lower third is separated from the anal canal and anal sphincter by the rectovaginal septum and the muscles of the perineal body.

The vagina consists of three principal layers: an outer fibrous layer, which derives from the pelvic fascia; a middle muscular layer; and an inner mucosal layer. The mucous membrane consists of wavy stratified squamous epithelium that is superimposed upon a tunica propria of fibrous tissue (Fig. 1-16). Small epithelial folds dip down into the tunica propria. In the prepubertal child, the vaginal mucosa is thin, perhaps consisting of only four to eight layers of basal and parabasal cells. Under the influence of estrogenic stimulation at the time of puberty, the mucosa thickens as the squamous cells mature; thereafter, it responds cyclically to the ovarian production of estrogen and progesterone. These cyclic changes are not evident grossly, although they are reflected fairly accurately in the vaginal smear. Under the influence of estrogen alone, the mucosal cells undergo a process of maturation, becoming progressively thinner and flatter, and their nuclei become smaller and more pyknotic. Progesterone inhibits this maturation to some degree, causing the epithelium to mature up to the superficial intermediate cell layer. Testosterone also tends to inhibit this maturation process to some extent. These cellular changes are reflected in the cyclic variation in the shedding of squamous cells from the vagina; this fact is useful in the evaluation of the hormonal status of the woman and is reflected in the vaginal cytogram. Immediately prior to ovulation, when the output of estrogen is at its peak, superficial cells are predominant in the vaginal smear. Following ovulation and throughout pregnancy, the intermediate cells predominate. As the menopause is approached and the output of hormones from the ovaries decreases, the vaginal epithelial cells undergo less maturation, as indicated by a shedding of cells largely from the deeper layers of the epithelium.

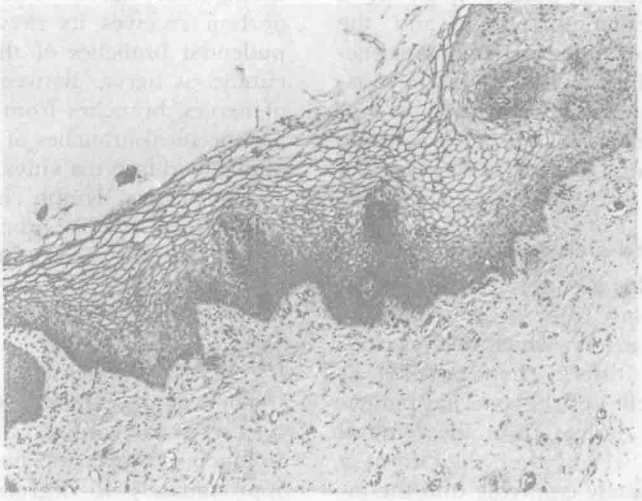

Fig. 1-16. Normal vagina. The mucosal lining consists of a wavy, stratified squamous epithelium overlying a tunica propria of fibrous tissue. (H & E × 88)