10
Congenital anomalies of the vagina

C. A. SALVATORE

Congenital anomalies of the vagina are represented by vaginal agenesis or atresia associated with uterine agenesis (Rokitansky–Kuster–Hauser Syndrome), vaginal agenesis with functioning uterus, septate vagina, double vagina and Wolffian cystic formation (Gartner’s cyst). Agenesis is the term used for complete absence of the vagina and atresia when a fibrous cord exists at the site of the vagina.

The most common anomalies are represented by vaginal agenesis which influences sexual relations. Congenital agenesis exerts a greater influence on a woman’s personality, sexuality and acceptance by her partner. It is a legal, social and medical problem requiring treatment at the proper time in a woman’s sexual evolution.

The vagina is the end portion of the internal female genital organs, the birth canal, and is the passage for the flow of cervical and menstrual secretion.

DEVELOPMENT OF THE VAGINA

It is essential to review here the embryology and morphology of the vagina. In a 42 mm female fetus, when the posterior ends of the Müllerian ducts reach the urogenital sinus, there is contact with the Wolffian ducts. The two Müllerian ducts also fuse into a uterovaginal canal. This is prolonged by a solid cord of clear cells, the ‘vaginal cord’, up to the vestibule. As the Wolffian ducts regress, the vaginal cord shows a central lumen. Several studies\textsuperscript{1,2} have shown that the participation of the Müllerian ducts in the vaginal cord occurs only in the upper part of the vagina. At least the major part of the vagina is derived from the urogenital sinus\textsuperscript{3}.

The epithelium of the urogenital sinus progressively extends around the posterior part of the Wolffian ducts which enlarge and will constitute the vaginal plate. The outgrowths from the sinus are double in origin—one around each Wolffian duct—but posteriorly they fuse under the Müllerian ends. It seems that the urogenital sinus is induced to proliferate within the lumen and outside of it at the area of contact with the Müllerian ducts.
The major part of the vaginal cord first increases in diameter and acquires a lumen near the urogenital sinus. The fornices and the lumen of the whole genital tract appear at the 200 mm stage of the fetus.

The development of the uterus precedes that of the vagina. For this reason, most cases of vaginal agenesis are usually found in association with uterine agenesis. However, the vagina can be partially formed in the absence of the uterus and vice versa, although this is rare. Other congenital anomalies, especially of the urinary tract, are commonly found in association with those of the genital system.

After birth, the vagina together with the uterus continues developing slowly. Later, during adolescence, this development is more rapid.

Normally, the vagina is found between the urethra—bladder and the rectum posteriorly. The vagina is attached to the lateral pelvic wall by connective tissue condensation and smooth muscle adherent to the adventitia of the vaginal blood vessels. There is a large amount of elastic tissue which permits great distensibility with subsequent return to the previous state, as is observed during and after labour.

Radiographic colpography done by Nichols and Randall shows a distinct superiorly convex perineal curve in the lower vagina. The upper vagina is parallel to the levator ani muscles. The rectum, vagina and urethra pass through the hiatus of the pelvic diaphragm. Under normal conditions the cardinal and uterosacral ligaments are important for maintaining not only the uterus but also the upper vagina.

**PATHOGENESIS**

The aetiology of the congenital anomalies of the vagina is not clearly understood. In general, there are no chromosomal anomalies in relation to vaginal agenesis (Rokitansky’s syndrome). However, three cases of vaginal agenesis have been reported in three monozygotic twins with normal twin sisters. They have been reported in relation to genetic aetiology by Linscke et al. Le Duc et al. described only of 14 cases of vaginal anomaly without karyotype. In the Rokitansky–Kuster–Hauser syndrome, normal 46,XX karyotypes occur. It seems that the congenital malformations depend on environmental factors in the presence of changes in DNA. The DNA mutation can be spontaneous or induced by ionizing radiations, viral or chemical agents which affect the DNA code.

Pathogenesis of vaginal anomalies is related to the development of the Müllerian ducts during the fourth week of embryonic development. During the fifth month, the definitive lumen of the vagina is partially formed. A process of excavation and cellular breakdown begins caudally, giving rise to partial or total vaginal and uterine agenesis. According to the studies of Jost, Müller’s ducts begin to regress or develop when the primitive gonad differentiates into either the fetal testicle or ovary, respectively. On the differentiation of the primitive gonad into an ovary, the wolffian ducts, in the absence of stimulation, will regress, but sometimes persist as small ducts which later give rise to the wolffian vaginal cysts.

Cessation of the development of the entire uterovaginal canal causes