Anomalies of the Female Genitalia*

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With 40 Figures

I. Anomalies of the Vulva and Vagina

1. Reduplication of the Vulva

Reduplication of the vulva in whole or in part is an extremely rare but striking anomaly. The few duplications which have been reported are uniformly lateral and in the most extreme form have consisted of complete vulvar duplication with two clitorides, four labia majora, and four labia minora. Such duplications are associated with other anomalies, such as reduplication of the internal structures, i.e. vagina and uterus, bladder, rectum and colon, as well as an occasional anomaly of the bony structures where this has been investigated.

This anomaly seems much more profound than the problems of lateral fusion of the müllerian ducts, in spite of the fact that failure of fusion is often a part of the anomaly of reduplication. Embryologically, as is well known, the external genitalia arise from the so-called genital tubercle and scrotolabial folds, external structures which are quite distinct from the internal müllerian ducts which form the uterus and part of the vagina. Thus, the disorder which causes reduplication involves much more than a single organ system.

BREEN and WEINBERG (1965) reported a remarkable example of reduplication and reviewed the literature up until that time. They reported a 20 year old Mexican Indian female whose history revealed that puberty had occurred at the normal age of 12 and that her menstrual periods were quite regular with a 30 day interval and a duration of 8 days. Her first pregnancy terminated prematurely at 28 weeks and her second pregnancy resulted in a stillborn delivery at home. She was seen when her third pregnancy was at term. All pregnancies had occurred through the right vagina and the patient apparently felt that psychologically she would be more normal if only one of her genital tracts was utilized for coitus, conception and delivery. The third pregnancy terminated normally after a 10 hour spontaneous delivery.

On further examination before discharge from the hospital, the complete double vulva was noted with two vaginas, each of which had a uterus at the vault. Catheters in each urethra produced independent drainage from two separate bladders (Fig. 1). There were two anal orifices. An intravenous pyelogram revealed normal kidneys but each ureter opened into a separate bladder (Fig. 2). A barium enema revealed two normal colons which ran parallel from the anus to the cecum (Fig. 3).

Unfortunately, further x-ray studies of the gastro-intestinal tract and of the generative tract were not performed.

* Based in part on material from Pediatric and Adolescent Gynecology by Howard W. Jones, Jr., and Richard H. Heller, Williams and Wilkins Company, Baltimore, 1966.
Eight other cases, very similar to this, all of which have been reported previously are shown in Table 1 which is modified from the paper by Breen and Weinberg.

Our experience with reduplication of the external genitalia is limited to a single patient who had a single clitoris but reduplication of the urethra with two ani, double vagina, double uterus and partial reduplication of the colon which branched in the region of the sigmoid (Fig. 4). An intravenous urogram disclosed normal kidneys which entered a single bladder which could be emptied through either urethra. However, there was a peculiar duplication of the sacrum (Fig. 5). This patient was severely handicapped by a large extra buttock and in this respect differed from most patients with reduplication of the vulva who seem to have no large bulky masses of tissue.

2. Masculinization of the Vulva

See chapter on the intersex states.

3. Imperforate Hymen

The hymen is located at the site where the embryonic vagina buds from the urogenital sinus. The hymenal area is, therefore, composed entirely of urogenital