Vaginal Reconstruction for Ambiguous Genitalia and Congenital Absence of the Vagina: A Twenty-Seven Year Experience

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**Background**: Gender assignment to neonate with ambiguous genitalia is crucial. Patients with an absent vagina require the construction of an artificial vagina. In an effort to improve care, we have categorized our experience with this group of children.

**Methods**: Since 1974, we cared for 114 patients with anomalies for the genitalia. There were 53 genotypic females with congenital adrenal hyperplasia (CAH), 16 genotypic males with testicular feminization syndrome (TFS), 13 with mixed gonadal dysgenesis (MGD), 9 with Mayer-Rokitansky syndrome, and 4 true hermaphrodites. The remaining 19 had other genital abnormalities.

**Results**: After 1980, patients with CAH underwent clitoral recession and vaginoplasty. All patients with TFS were raised as females and underwent orchidectomy. Eleven of the MGD patients were given a female sex assignment and underwent gonadectomy. Twenty-eight patients underwent intestinal vaginoplasty including 8 of the TFS patients, 9 with Mayer-Rokitansky syndrome, 8 patients with cloacal anomalies, 2 patients for rhabdomyosarcoma, and 1 of the MGD patients.

**Conclusion**: 1) This review emphasizes the range of diagnoses the surgeon must be prepared to address in patients ambiguous genitalia. 2) Colovaginoplasty is an excellent procedure for replacement of a completely absent vagina. 3) Continued evaluation of this group will delineate appropriate timing and choice of procedure.

Index Words: Ambiguous genitalia, vaginal agenesis, colovaginoplasty, congenital adrenal hyperplasia, Mayer-Rokitansky syndrome