

AMBIGUOUS GENITALIA: AN OVERVIEW OF 27 YEARS EXPERIENCE

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Aim: The newborn with abnormal genital development presents a difficult diagnostic and treatment challenge for the pediatric surgeon providing care. The purpose of this study was to evaluate the results of surgical treatment for children with ambiguous genitalia.

Patients and Methods: The records of 79 patients managed surgically for ambiguous genitalia in our unit from 1988 to 2005 were reviewed retrospectively. Age at surgery, operative procedures, and outcome were recorded. The mean age at surgery was 4.5 years, and the follow-up period averaged 7.2 years.

Results: There were 37 (46%) genotypic girls with congenital adrenal hyperplasia (CAH) in female pseudohermaphroditism group. Among the 24 (30%) genotypic boys in male pseudohermaphroditism group 9 (37%) had either testosterone biosynthetic defects or hCG unresponsiveness, 11 (45%) had either complete or partial androgen insensitivity syndrome or 5 alpha-reductase deficiency, 2 (8%) had dysgenetic male pseudohermaphroditism and 2 (8%) had isolated MIF deficiency. There were 7 (8%) patients were in the true hermaphroditism group and 6 (7%) patients in gonadal dysgenesis group and 3 (3%) patients with Mayer-Rokitansky syndrome. All patients but 3 with female pseudohermaphroditism underwent clitoroplasty and vaginoplasty. 12 patients with male pseudohermaphroditism were given a female sex assignment and underwent gonadectomy and vaginoplasty and the remaining 12 patients were given male sex assignment and underwent orchiopexy and hypospadias repair. All patients with gonadal dysgenesis and 4 patients with true hermaphroditism were raised as girls and 3 patients with true hermaphroditism were raised as boys. Ovaries were removed and gonads and Müllerian structures were either remained or removed due to given sex assignment. Of the 58 patients reared as females, 32 (55%) required perineal vaginoplasty, 14 (24%) had pull-through vaginoplasty, and 12 (20%) had colovaginoplasty. 18 (32%) patients with feminization procedures and 8 (38%) of 21 patients with masculinization procedures experienced complications and required redo operations. Vaginal stenosis was the most common complication.

Conclusion: It is important that a definitive diagnosis be determined as quickly as possible so that the appropriate treatment plan can be established to minimize medical, psychological and social complications in patients with ambiguous genitalia.

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