Long-Term Psychological Evaluation of Intersex Children

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Treatment of psychological problems of 59 children with a physical intersex condition is described. The group consisted of 18 female pseudohermaphrodites with congenital adrenal hyperplasia (CAH), 20 male pseudohermaphrodites and 2 true hermaphrodites born with ambiguous external genitalia assigned the female sex (ambiguous girls), 14 male pseudohermaphrodites born with completely female external genitalia and assigned the female sex (completely female group), and 5 male pseudohermaphrodites born with ambiguous external genitalia and assigned the male sex. Despite the sex assignment, genital organ correction soon after birth, psychological counseling of parents and intensive psychotherapy of the children, general psychopathology developed equally in all 4 groups (39% of total group). Although 87% of the girls with a physical intersex condition developed in line with the assigned sex, 13% developed a gender identity disorder though only 1 girl (2%) failed to accept the assigned sex. Gender identity disorder and deviant gender role were in evidence only in girls with CAH and girls of the ambiguous group. Biological and social factors seem responsible for the development of gender identity disorder, such as pre- and postnatal hormonal influences on the brain enabling deviant gender role behavior to develop, and an inability on the part of parents to accept the sex assignment. A reconsideration of the sex assignment in male pseudohermaphrodites and true hermaphrodites born with ambiguous external genitalia is discussed.

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INTRODUCTION

Sex assignment problems can emerge in male pseudohermaphrodites and true hermaphrodites born with ambiguous genitalia. Male pseudohermaphroditism occurs in individuals born with an XY karyotype or a mosaic form with disorders in the production, synthesis, or receptor sensitivity to androgens, so that their male genitalia are not sufficiently developed. In these children, the external genitalia are inadequately virilized, the gonads are located in the abdomen or inguinal canal, the uterus and ovaries are not in evidence, and the vagina has been inadequately formed, if at all. If the insensitivity to androgens is complete, as in the complete androgen insensitivity syndrome (CAIS), or the defect in the production of androgens is complete, as in Leydig cell hypoplasia, the child is born with female external genitalia and gonads that may be positioned in the abdomen or inguinal canal.

There is no doubt about the management of patients with CAIS or with a complete defect in the production or synthesis of androgens: The sex these patients are reared in is always female. However, sex assignment in the group of male pseudohermaphrodites and true hermaphrodites born with ambiguous genitalia is a difficult matter, since there are still no reliable criteria for their sex assignment. The debate about what is more important for gender identity development, the biological sex or the sex a child is reared in, is still going on. Money et al. (1955) were the first to emphasize the importance of upbringing. They felt that unambiguously raising a child with a physical intersex condition as a member of the assigned sex would be more important for the child’s gender identity development than his or her chromosomal sex. Almost three decades later, Diamond (1982) demonstrated the controversial nature of this opinion with the well-known case history of the monozygotic twins, both of whom were genetic males at birth. Due to the accidental burning of his penis at the age of 7 months, one of the boys was assigned the female sex (Money and Tucker, 1975). Although raised as a girl, the patient could not identify with the female sex, and as an adult asked for sex reassignment. He now lives as a married man with his wife and adopted children (Diamond and Sigmundson, 1997).

Studies on female pseudohermaphrodites with congenital adrenal hyperplasia (CAH) (Money and Ehrhardt, 1968) have demonstrated that prenatal and postnatal hormones can cause sex-dimorphic behavior. CAH is an autosomal recessive disorder caused by a defect of one of the enzymes (in 90% the 21-hydroxylase) necessary for the formation of cortisone. This