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Gender identity outcomes in children with disorders/differences of sex development: Predictive factors

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ABSTRACT

Disorders/differences of sex development (DSD) comprise multiple congenital conditions in which chromosomal, gonadal, and/or anatomical sex are discordant. The prediction of future *gender identity* (i.e., self-identifying as male, female, or other) in children with DSD can be imprecise, and current knowledge about the development of gender identity in people with, and without DSD, is limited. However, sex of rearing is the strongest predictor of gender identity for the majority of individuals with various DSD conditions. When making decisions regarding sex of rearing biological factors (e.g., possession of a Y chromosome, degree and duration of pre- and postnatal androgen exposure, phenotypic presentation of the external genitalia, and fertility potential), social and cultural factors, as well as quality of life should be considered. Information on gender identity outcomes across a range of DSD diagnoses is presented to aid in sex of rearing assignment.

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Disorders/differences of sex development (DSD) comprise multiple congenital conditions in which chromosomal, gonadal, and/or anatomical sex are discordant.^{1,2} The presence of ambiguous external genitalia can add to the complexity of DSD care, leaving parents and physicians to predict a child's *gender identity* (i.e., self-identifying as male, female, or other) and decide sex of rearing for a young child based on a limited understanding of how that child may develop physically and behaviorally over time.³ This is particularly challenging as predicting gender identity is imprecise. However, research suggests that the strongest predictor of adult gender identity in the majority of DSD diagnoses is sex of rearing.⁴

Multiple factors are considered when assigning a sex of rearing to young infants with DSD, with the goal of having the child be satisfied with that assignment.⁵ These include biological factors, such as possession of a Y chromosome, degree and duration of pre- and postnatal androgen exposure, and, for both sexes, fertility potential, as well as social and cultural factors such as the presence of traditional beliefs about gender roles, parent attitudes about sex of rearing, and quality-of-life considerations.⁶

The review that follows presents data on the development of gender identity across a range of DSD diagnoses for which sufficient research on gender development is available. *Gender roles*, or the overt behaviors a person exhibits that are

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classified by society as either masculine or feminine,^{7,8} will not be discussed in this article, as little evidence supports that a gender role incongruent with one's sex of rearing negatively impacts gender identity or quality of life in individuals with or without DSD.⁹⁻¹¹

Gender identity outcomes in 46,XX DSD

Congenital adrenal hyperplasia due to 21-OH deficiency

46,XX DSD is most often due to congenital adrenal hyperplasia (CAH) resulting from 21-OH deficiency. For newborns with this type of CAH who possess a female chromosomal complement, sex of rearing is a strong predictor of gender identity outcomes, as the majority of newborns reared female develop a female gender identity.^{4,12} Specifically, it is estimated that 95% of individuals with CAH develop a female gender identity.³ The other 5% of individuals include a portion who are the most masculinized at birth, including those with male genitalia. Although recent literature proposes male rearing as a consideration in newborns with fully masculinized external genitalia,^{13,14} fertility potential could be compromised and data on psychosexual outcomes are limited for this scenario.^{13,15} Current evidence supports female rearing in the vast majority 46,XX newborns with CAH due to 21-OH deficiency, though future research may provide further evidence of the other option.³

Although 46,XX individuals with CAH experience elevated prenatal (and often postnatal) androgen exposure that is associated with masculinized gender role behaviors such as preference for rough-and-tumble play, this does not negatively impact female gender identity outcomes.⁹⁻¹¹ Additionally, the minority of 46,XX individuals with CAH who identify as male, despite their female rearing, were not exposed to greater amounts of prenatal androgens, as suggested by their Prader rating at birth, compared to girls who identify as female.¹⁶ Thus, while androgen exposure plays a part in masculinized gender role development, it does not appear to be a major predictor of gender identity development in 46,XX CAH.

Gender identity outcomes in 46,XY DSD

The term 46,XY DSD refers to a wide range of DSDs. Biological influences, such as early androgen exposure and diagnosis, are clearer predictors of gender identity outcomes for some of these conditions, while social influences appear to be stronger predictors for others.^{17,18} In contrast to 46,XX CAH for whom female rearing is recommended despite their Prader rating at birth, genital phenotype and androgen responsiveness is an important determinant of sex of rearing for newborns with certain types of 46,XY DSD, such as partial androgen insensitivity syndrome.¹⁹ However, this is based on the fact that no single assignment is highly successful.

46,XY DSD and female external genitalia at birth

Complete androgen insensitivity syndrome (CAIS) is defined by the complete resistance of target tissues to androgenic effects. Complete gonadal dysgenesis, also known as Swyer syndrome, occurs when precursor gonadal tissue fails to develop or differentiate.¹ Both of these conditions occur in people with a 46,XY chromosomal complement. People with CAIS are unable to respond to the androgens produced by their testes, and individuals with complete gonadal dysgenesis are not exposed to testicular hormones because their bipotential gonadal tissue does not differentiate into testes. The external genitalia of newborns with 46,XY DSD due to CAIS or Swyer syndrome are female at birth.

Newborns with CAIS and Swyer syndrome are reared female, and female gender identity develops in nearly all of these children.^{20,21} Because newborns with CAIS and Swyer syndrome exhibit female external genitalia at birth, they may not be diagnosed until adolescence when menses fails to occur (for both conditions) pubic hair is minimal or absent (CAIS), and/or breasts fail to develop (Swyer syndrome). Regardless of their possession of a Y chromosome, the combination of female-typical socialization and the absence of androgenic effects may explain the preponderance of female gender identity development for individuals with these conditions. However, there are a small number of individuals with these conditions who report male gender identity.²²⁻²⁴

46,XY DSD and ambiguous external genitalia at birth

Partial androgen insensitivity syndrome (PAIS) refers to the partial inability to respond to androgens by target tissues. Partial gonadal dysgenesis refers to the incomplete differentiation of bipotential gonadal tissue into testes during fetal development. Newborns with PAIS or partial gonadal dysgenesis may possess ambiguous external genitalia at birth because they are exposed to more androgens than an unaffected female, but less androgens than an unaffected male, during fetal development.¹

It is increasingly common for parents to rear newborns with PAIS as male.^{19,25} However, individuals with PAIS or partial gonadal dysgenesis are more likely than those with CAH, CAIS or Swyer syndrome to report a gender identity that is discordant with their sex of rearing, regardless of whether they are reared male or female.^{1,4,26} Although dissatisfaction with sex of rearing is more likely in individuals with 46,XY DSD including ambiguous genitalia at birth, rearing and socialization continue to be the best predictors of long-term gender identity outcomes.^{17,26} However, social factors may more greatly influence the development of gender identity for this population compared to people with other types of DSD,²⁷ contributing to dissatisfaction with sex of rearing.

Research has shown that mothers of children with 46,XY DSD including ambiguous genitalia, are more distressed by their child's condition than mothers of children with other types of DSD.²⁸ For these mothers, greater degree of genital malformation is associated with greater degree of distress.²⁹ More importantly, evidence indicates that the distress reported by parents of children with 46,XY DSD including

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