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Gender Change in 46,XY Persons with 5α-Reductase-2 Deficiency and 17β-Hydroxysteroid Dehydrogenase-3 Deficiency

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Individuals with 5α-reductase-2 deficiency (5α-RD-2) and 17β-hydroxysteroid dehydrogenase-3 deficiency (17β-HSD-3) are often raised as girls. Over the past number of years, this policy has been challenged because many individuals with these conditions develop a male gender identity and make a gender role change after puberty. The findings also raised doubts regarding the hypothesis that children are psychosexually neutral at birth and emphasized the potential role of prenatal brain exposure to androgens in gender development. If prenatal exposure to androgens is a major contributor to gender identity development, one would expect that all, or nearly all, affected individuals, even when raised as girls, would develop a male gender identity and make a gender role switch later in life. However, an estimation of the prevalence of gender role changes, based on the current literature, shows that gender role changes occur frequently, but not invariably. Gender role changes were reported in 56–63% of cases with 5α-RD-2 and 39–64% of cases with 17β-HSD-3 who were raised as girls. The changes were usually made in adolescence and early adulthood. In these two syndromes, the degree of external genital masculinization at birth does not seem to be related to gender role changes in a systematic way.

KEY WORDS: 5α-reductase deficiency; 17β-hydroxysteroid dehydrogenase deficiency; gender dysphoria; gender identity.

INTRODUCTION

Prenatal exposure of the brain to androgens has increasingly been put forward as a critical factor in gender identity development. Reports on a seemingly sudden gender identity and role change in persons with a certain intersex condition (Imperato-McGinley, Guerrero, Gautier, & Peterson, 1974; Imperato-McGinley, Peterson, Gautier, & Sturla, 1979) were interpreted by some as an indication that androgens may play a role in gender identity development. The condition, 5α-reductase-2 deficiency (5α-RD-2), is an enzyme deficiency secondary to a deletion or mutation of the underlying gene. During fetal development, the isoenzyme 5α-reductase-2 is required for the conversion of testosterone into dihydrotestosterone which, in turn, is responsible for the development of the external male genitals and prostate. When the enzyme is missing, the external genitals are not, or not completely, masculinized. An enzyme deficiency that has similar effects in terms of genital appearance at birth is 17β-hydroxysteroid dehydrogenase-3 deficiency (17β-HSD-3). During fetal life, the isoenzyme 17β-hydroxysteroid dehydrogenase-3 is required for the biosynthesis of testosterone, which has to be converted into dihydrotestosterone in order for the male external genitals to develop. In 5α-RD-2, the brain is prenatally exposed to normal male testosterone levels. In 17β-HSD-3, testosterone production is deficient; however, some testosterone production may still be possible due to other 17β-HSD isoenzymes.

In both conditions, genital ambiguity or the presence of descended testicles in female-looking genitals may indicate that the child has a sexual differentiation disorder. In more severe cases, atypical characteristics may be

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missed and the child is assigned and raised as a girl. Especially if the children live in a community where these conditions are rare, no one will suspect that they have XY chromosomes and internal male genital structures. They will, however, develop male secondary sex characteristics during puberty, if the condition is not discovered and treated timely (Sobel & Imperato-McGinley, 2004). Their clitoris will enlarge, their voice will coarsen, they will have a masculine muscular development, and their body and facial hair growth will increase. They may also have noticeable erections in their enlarging clitoris. Pubertal virilization occurs in 5α-RD-2, probably because at this age there is increased production of 5α-RD-1, which converts part of the pubertal testosterone into dihydrotestosterone (Wilson, 2001). In 17β-HSD-3, virilization is possible because other 17β-HSD isoenzymes in peripheral tissues convert androstenedione to testosterone. Because prostate and seminal vesicles are underdeveloped and testicles often undescended, infertility may occur. The pubertal virilization of individuals who were assigned and raised as girls but change gender after puberty makes it conceivable that androgens also influence gender development postnatally.

Both conditions are rare autosomal recessive disorders. The prevalence in the general population is unknown. In isolated communities where consanguinity is rather common, the prevalence can be very high. Kohn et al. (1985) estimated the prevalence of affected 46,XY individuals in the Gaza strip to be 1 in 100–150. The largest groups that have been studied are from the Dominican Republic, Papua New Guinea, Mexico, and Brazil, and certain areas in the Middle East (Gaza strip), but individual cases from other countries have also been reported.

In this article, I will give an overview of studies on the gender outcome of affected individuals. I will focus on the prevalence of changes of the social gender from female to male because this is the most reliably reported variable. As very little information is given about gender-related behavior, those data have been omitted from consideration.

METHOD

Relevant literature was collected by performing a computer-based search (Medline, PsycINFO) using the following key words: 5α-reductase deficiency (5α-RD) and 17β-hydroxysteroid dehydrogenase deficiency (17β-HSD), 17-ketosteroid reductase deficiency (17-KRD), male pseudohermaphroditism, gender identity, gender role, gender development, and psychosexual development. References cited by Zucker (1999, pp. 18–27) on 5α-RD were also examined. Studies that only reported on children younger than 12 years and studies giving no information regarding the ages of subjects were not included. If studies included both pre- and post-pubertal subjects, only the data on subjects who were 12 years or older at the time of the study were included in the tables. As it is likely that, in early childhood (<5 years), physicians rather than the children induced a gender role change, these very early reassigned children, who had primarily been raised as boys, were excluded from our analyses. The gender role changes that were relevant to our overview, because they probably resulted from gender dysphoria, were in 46,XY individuals of 12 years and older, who had a female sex assignment at birth and were raised as girls. We therefore also did not include 46,XY individuals who had a male assignment at birth and were raised as boys in our tables. If authors explicitly mentioned that they had reported on certain subjects in another study, the subjects were only counted once.

Reports that gave explicit information about gender identity and/or gender role are summarized in Tables I and III. In some articles, the gender identity or gender role could only be inferred from treatment information (e.g., feminizing hormones or surgery). These reports are summarized in Tables II and IV.

RESULTS

From Tables I and II, a gender role change from female to male was reported in 63% of the patients with 5α-RD-2 ($N = 99$). Two additional patients with a male gender identity presented themselves as males when visiting the clinic, but primarily lived in the female social role. They feared the social consequences of disclosing their male gender identity. Both had sexual relationships with women as males (they did not have genital surgery) (Al-Attia, 1996; Imperato-McGinley et al., 1980). Information about one other patient was less explicit, but it was reported that this person had a male gender identity and had sexual relations with women, but dressed himself as a woman (Imperato-McGinley, Peterson, Gautier, et al., 1979). When the articles with less clear information about gender outcome (Table II) are included, the percentage was somewhat lower (56%; $N = 110$). The proportion is similar for the reports on 17β-HSD-3 that gave explicit psychosexual information (64%; $N = 28$) (Table III), but somewhat lower when studies with less clear psychosexual information are included (39%; $N = 49$) (Table IV).

Sixteen (48%) out of 33 female-raised individuals with atypical looking genitals at birth ("clitoromegaly,"

Table I. Explicitly Described Gender History, Gender Identity, and Gender Role in Adolescent and Adult 46,XY Individuals with 5α-RD-2

General information			Gender history				Latest report			
Reference	Country	No. of subjects 12 years or older	Genital appearance at birth	GA[a,b]	No. of Pat GRA[b]	Pat GRA at age	Age at time of study	Lives as M/F[c]	Identifies as M/F/other	Gender dysphoria
Cantú et al. (1976)	Mexico	1	Ambiguous	1 F	1	15 years	23 years	1 M	1 M	
Fisher et al. (1978)	USA	2	Clitoromegaly at birth	2 F			12 years 13 years	2 F	2 F	
Imperato-McGinley et al. (1979)	Dominican Republic	33	n.i.[c]	18 F (+1 F[d]; 9 M; 5 raised ambiguously)	17	17: 14–24 years (average 16 years)	17: >14 years	16 M 1 F 1 F/M	1 F 17 M	
Money (1979)	USA	1	n.i.	1 F			Teenager	1 F	1 F	
Cantú et al. (1980)	Mexico	1	Ambiguous	1 F			18 years	1 F	1 F	
Imperato-McGinley et al. (1980)	Italy/USA	1	n.i.	1 F			65 years	1 F/M	1 M	Yes
Mauvais-Jarvis, Kuttenn, Mowszowicz, and Wright (1981) (including 1 patient from Kuttenn et al., 1979)	France	2	Clitoromegaly	2 F	1?[c]	n.i.	25 years 20 years	1 F 1 M?	1 F 1 M	
Corrall et al. (1984)	UK	1	n.i.	1 F			24 years	1 F	1 F	
Price et al. (1984) (including 2 subjects of Savage et al., 1980)	2 Cyprus 1 Pakistan 1 Malta	4	4: perineoscrotal hypospadias and bifid scrotum	4 F	4	2: 14 years, 16 years, 24 years	17 years 19 years 23 years 26 years	4 M	4 M	
Deslypere, Coucke, Robbe, and Vermeulen (1985)	Belgium (patient from France)	1	Clitoral hypertrophy; bilateral labia	1 F	1	Post puberty	30 years	1 M	1 M	
Bartsch, Decristoforo, and Schweigert (1987)	Austria	1	'Normal'	1 F			Adult	1 F	1 F	
Herdt and Davidson (1988)	New Guinea	12[e]	n.i.	5 F[f] (+1 F reassigned at 2; 6 "hermaphroditic")	5	Post puberty	Adults	4 M (including 3 who died) 1 F?	4 M (including 3 who died) 1 F?	

Table I. Continued

General information			Gender history				Latest report			
Reference	Country	No. of subjects 12 years or older	Genital appearance at birth	GA[a,b]	No. of Pat GRA[b]	Pat GRA at age	Age at time of study	Lives as M/F[c]	Identifies as M/F/other	Gender dysphoria
Imperato-McGinley et al. (1991) (including 2 patients from Gajdusek, 1977)	New Guinea	5	n.i.	1 F (4 M)	1	Post puberty	35 years	1 M	1 M	
Hurtig (1992)	USA	2	n.i.	2 F	1	Probably post-puberty	n.i.	1 F	1 F	
								1 M	1 M	
Aguilar-Diosdado, Gavilán-Villarejo, Escobar-Jiménez, Beltrán, and Girón (1995)	Spain	2	Ambiguous	2 F	2	Post puberty	17 years	2 M	2 M	
Boudon et al. (1995)	2 France 2 Poland	4	n.i.	4 F			14 years 4: 13–16 years	4 F	4 F	
Méndez et al. (1995)	Mexico	6	n.i.	6 F	4 (1 not yet treated)	4: 18–28 years	6: 14–36 years	5 M	5 M	Perhaps in the subject with F gender role
								1 F (under external pressure)	1 F	
Al-Attia (1996)	Oman	4	n.i.	4 F	2	13 years	4: 16–28 years	1 M	1 M	
						15 years		1 F	1 F	Yes
								1 F despite physical reassign-ment	1 M	Yes
								1 F/M	1 M	
Hochberg et al. (1996)	Lebanon	8	Unambiguous female external genitals	8 F	8	Around 16	Adults	8 M	2 M 6 M?	
Canto et al. (1997)	Mexico	3 (+5[g])	n.i. about the 2 F	2 F (1 M)	1	22 years	24 years 17 years	1 F 1 M	1 F 1 M	

Ferraz, Guerra, Baptista, Maciel-Guerra, and Hackel (1998)	Brazil	1	n.i.	1 F			16 years	1 F	1 F
Nordenskjöld, Magnus, Aagenaes, and Knudtzon (1998)	Norway (patient from Pakistan)	1	n.i.	1 F	1	14 years	19 years	1 M	1 M
Ferraz et al. (1999)	Brazil	1	Palpable gonads	1 F	1	18 years	25 years	1 M	1 M
Chávez, Valdez, and Vilchis (2000)	Mexico	2	n.i.	1 F (1 M)			14 years	1 F	1 F
Hedia et al. (2001)	Tunesia	1	n.i.	1 F	1	15 years	15 years	1 M	1 M
Mendonca et al. (2003) (including 16 Mendonca et al., 1996 subjects)	Brazil	26	'Female-like appearance' in the 25 F	25 F (1 M)	11	2 pre-puberty 9 post-puberty; 2 in adulthood	25: 14–47 years	13 M 11 F 1 F (14 years) " still has to decide"	13 M 11 F 1 F?
Mazen, Gad, Hafez, Sultan, and Lumbroso (2003)	Egypt	2	n.i.	1 F (1 M)			14 years	1 F	1 n.i.
Yücel and Polat (2003)	Turkey	1	n.i.	1 F			25 years	1 F	1 F
Total		129		99 F (2 F not in analyses; 17 M; 11 other)				62 M 34 F 3 F/M	66 M 32 F 1 n.i.

[a] Only patients reared as females are further analyzed.
[b] GA: Gender Assignment; Pat GRA: Patient-initiated gender reassignment.
[c] F: female; M: male; n.i.: no information; ?: text is suggestive but does not give explicit information.
[d] One female assigned individual was excluded form the analyses because no gender identity/gender role information was available.
[e] Diagnoses likely but not confirmed medically.
[f] One female assigned individual was excluded from the analyses because he was reassigned <3 years.
[g] Five patients from this study are already included in Mendez et al. (1995), so we only report on the remaining three.

Table II. Inferred Gender History and Assumed Gender Identity and Gender Role in Adolescent and Adult 46,XY Individuals with 5α-RD-2

General information			Gender history		Latest report		
Reference	Country	No. of subjects 12 years or older	Genital appearance at birth	GA[a,b]	Age at time of study	Lives as M/F[c]	Identifies as M/F/other
Hodgins, Clayton, and London (1971)	UK	1	n.i[c]	1 F	(post) puberty	1 F?[c]	1 F?
Walsh et al. (1974)	USA	1	1 F normal	1 F	13 years	1 F?	1 F?
Saenger et al. (1978)	USA	1	Ambiguous genitals	1 F	12 years	1 F?	1 F?
Okon et al. (1980)	Israel	2	n.i.	2 F	18 years 17 years	2 F?	2 F?
Wieacker, Flecken, and Breckwoldt (1992)	Germany	1	n.i.	1 F	17 years	1 F?	1 F?
Punnose, Mathew, and Ahmed (1995)	United Arab Emirates	1	Mild clitoromegaly	1 F	14 years	1 F?	1 F?
Sinnecker et al. (1996)	Germany/(patients from: 1 Germany 2 Turkey)	3	2 F predominantly female phenotype genitals 1 F ambiguous	3 F	3: 14–22 years	3 F?	3 F?
Walden, Rauch, Hiort, Sinnecker, and Dörr (1998)	Germany (patient from Turkey)	1	Ambiguous genitals with micropenis	1 F	14 years	1 F?	1 F?
Total		11		11 F		11 F	11 F

[a] Only patients reared as females are further analyzed.
[b] GA: Gender Assignment.
[c] F: female; M: male; n.i.: no information; ?: text is suggestive but does not give explicit information.

Table III. Explicitly Described Gender History, Gender Identity and Gender Role in Adolescent and Adult 46,XY Individuals with 17β-HSD-3

General information			Gender history				Latest report		
Reference	Country	No. of subjects 12 years or older	Genital appearance at birth	GA[a,b]	No. of Pat GRA[b]	Pat GRA at age	Age at time of study	Lives as M/F[c]	Identifies as M/F/other
Akesode, Meyer, and Migeon (1977)	USA (patient from Syria)	1	Ambiguous	1 F	1	17 years	28 years	1 M	1 M
Imperato-McGinley, Peterson, Stoller, and Goodwin (1979)	USA	1	Female	1 F	1	14 years	31 years	1 M	1 M
Millán et al. (1983)	Spain	1	n.i.	1 F			24 years	1 F	1 F
Lanes et al. (1983)	Venuzuela	3	Ambiguous	3 F	3	3: 11–13 years	22 years 19 years 16 years	3 M	3 M
Imperato-McGinley, Akgun, Ertel, Sayli, and Shackleton (1987)	Turkey	2	n.i.[c]	1 F (1 M?[c])	1	>15 years	25 years	1 M	1 M
Rösler, Silverstein, and Abeliovich (1996) (including patients from earlier studies: Rösler and Kohn, 1983; Kohn et al., 1985, Rösler, 1992)	Israel (patients of Arab origin)	15[d]	n.i.	9 F[e] (1 M)	7	1: 10 years 6: 12–18 years	9: 12–43 years	7 M; 2 F	7 M; 2 F
Mendonca et al. (2000)	Brazil	10 [+2[f]]	8 ambiguous genitals, 2 'normal' [2 n.i]	10 F [+2 F]	3[g] [+2]	3: 15–26 years [+14, 17]	10: 20–37 years [+2 n.i.])	3 M [+2 M] 7 F	3 M [+2 M] 7 F
Total		35[e]		28 F (2 M)				18 M 10 F	18 M 10 F

[a] Only patients reared as females are further analyzed.
[b] GA: Gender Assignment; Pat GRA: Patient-initiated gender reassignment.
[c] F: female; M: male; n.i.: no information; ?: text is suggestive but does not give explicit information.
[d] Three 46, XX individuals are not included in the table.
[e] Five female assigned individuals were excluded from the analyses because they were reassigned <3 years.
[f] These two individuals were not physically examined by Mendonca. They were a sib and a cousin of two of the examined patients. Their results are given separately in the table.
[g] All three had ambiguous genitals at birth.

Table IV. Inferred Gender History and Assumed Gender Identity and Gender Role in Adolescent and Adult 46,XY Individuals with 17β-HSD-3

General information			Gender history		Latest report		
Reference	Country	No. of subjects 12 years or older	Genital appearance at birth	GA[a,b]	Age at time of study	Lives as M/F[c]	Identifies as M/F/other
Saez, de Peretti, Morera, David, and Bertrand (1970)	France	2	n.i.[c]	2 F	15 years, 18 years	2 F	2 F?[c]
Virdis, Saenger, Senior, and New (1978)	USA (patient from Greece)	1	n.i.	1 F	12 years	1 F	1 F?
Leinonen, Dunkel, Perheentupa, and Vihko (1983)	Finland	1	Normal clitoris; at 7 weeks lumps in labia majora were discovered	1 F	12 years	1 F	1 F? ("tomboyish but well adjusted")
Balducci et al. (1985)	Italy	2	n.i.	2 F	15 years, 14 years	2 F	2 F?
Wilson, Hodgins, and Scott (1987)	UK	1	n.i.	1 F	12 years	1 F	1 F?
Andersson et al. (1996) (4 subjects were excluded because no relevant information was given)	USA	1	Normal	1 F	15 years	1 F?	1 F?
	USA	1	Normal	1 F	13 years	1 F	1 F
	Iran	1	n.i.	1 F	16 years	1 F?	1 F?
	Poland	1	Normal	1 F	15 years	1 F?	1 F?
	Brazil	1	n.i	1 F	21 years	1 F	1 F?
	Brazil	1	Ambiguous	1 F[d]	21 years	1 M	1 M
	USA	1	n.i.	1 F	17 years	1 F?	1 F?
	USA	1	n.i.	1 F	13 years	1 F?	1 F?
	USA	1	Enlarged clitoris	1 F	16 years	1 F?	1 F?
	Brazil	1	n.i.	1 F	15 years	1 F?	1 F?
	Italy/Germany/Ireland	1	n.i.	1 F	13 years	1 F	1 F
	USA	1	Penoscrotal hypospadias; single orifice; bifid scrotum	1 F	13 years	1 F?	1 F?
Wieacker and Von Mühlendahl (1996)	Germany	2	n.i.	2 F	16 years, 14 years	2 F?	2 F?
Total		21		21 F		20 F 1 M	20 F 1 M

[a] Only patients reared as females are further analyzed.
[b] GA: Gender Assignment; Pat GRA: Patient-initiated gender reassignment.
[c] F: female; M: male; n.i.: no information; ?: text is suggestive but does not give explicit information.
[d] He "began to function as a male" at 10; at 21, a male gender identity was confirmed by psychological testing.
[e] Most probably the same patient as reported by Park, Lee, and Witchel (1996); this study was therefore not included.

"clitoral hypertrophy," "perineoscrotal hypospadias and bifid scrotum," "palpable gonads," "ambiguous genitals with micropenis," "lumps in labia majora," "perineoscrotal hypospadias, single orifice and bifid scrotum") did not make a female-to-male gender role change. On the other hand, only 22 (54%) of 41 individuals who were born with female appearing external genitalia chose to live as males after puberty. Most of the gender role changes took place in late adolescence and early adulthood, rarely after the age of 30.

We identified 28 individuals, older than 12 years, 18 with 5α-RD-2 and 2 with 17β-HSD-3, who were assigned and raised as men. Nineteen subjects are included in the tables because the studies also reported on female-raised subjects. Six additional subjects with 5α-RD-2 were reported on by Akgun, Ertel, Imperato-McGinley, & Sayli, (1986) and two subjects with 5α-RD-2 were described in a study of Ivarsson, Nielsen, and Lindberg (1988). One subject, described by Forti et al. (1996), had a female assignment, but was reassigned at 3 years and raised as a male. None of the men appeared to have a wish to live as women or made a role change in the male-to-female direction.

DISCUSSION

Our review shows that the number of gender role changes reported on in the literature is considerable and certainly higher than in other intersex conditions. (The actual numbers may be even somewhat higher than we reported because we excluded children who were under 12 years of age. Some of these children may later still want to live as men.) However, as adults, a significant number of affected individuals still lived in the female role.

Several factors may determine whether these individuals, who were raised as girls, will make a switch after puberty. A possible biological factor is the severity of the mutation in terms of the in vitro enzyme production deficiency, causing more or less (prenatal) exposure of the brain to androgens. Cultural or other environmental pressures have also been mentioned as potential determinants. Finally, the patient's psychological reaction (body image) and/or the reaction of the social environment to the patient's genital appearance (male/ambiguous/female) are a third possible explanation for the reported gender role changes.

In our overview, we did not take the severity of the genetic mutation into account, but it has been found that a gender role change is not simply related to the severity of the mutation (Wilson, 2001). The duration and extent of prenatal brain exposure to androgens is difficult to determine retrospectively at birth or later. The condition is always discovered long after the presumed hormone-sensitive prenatal phase in the sexual differentiation of the brain has passed and reliable indices of prenatal brain exposure to androgens are not yet known. Furthermore, no data are available on the relationship between the developing male gender identity and circulating androgens before, during, and after the gender role changes. This means that it is difficult to tell whether the observed variation in psychosexual outcome is related to the amount of androgen exposure.

In some of the studies, it is stated that the high percentage of gender role changes can be explained by the cultural advantages of the male role. The first studies reported on groups of affected individuals who lived in societies where the male role had a higher social status. However, our data show that a considerable number of affected persons who lived in societies that may pressure them to make a role change remained in the female role, whereas role changes did occur in societies where male and female roles are relatively equally valued. Unfortunately, the large majority of the reported studies give little information on more subtle external forces, such as parental desire (e.g., for a male or female child) or gender of rearing. It may well be that attitudes within the family are more important than more distant cultural influences. Moreover, even within families, subtle forces may work differently for different family members. This is in line with the finding that in a number of families, some, but not all, affected members changed gender role (e.g., Mendonca et al., 1996).

It is conceivable that genital appearance influences a person's sense of self and therefore his or her gender identity. The emerging gender identity may be reinforced by feedback from the environment. Family members who are uncertain whether the baby is a boy or a girl may convey their uncertainty to the child and thus enlarge an already existing "gender discomfort" in the child. This would mean that children with obvious genital ambiguity are likely candidates for a gender role change later in life. Our review does not support this explanation. About half of the persons with female appearing external genitalia developed a male gender identity (see also Hochberg et al., 1996; Mendonca et al., 2003) and about half of the persons with genital ambiguity developed a female gender identity, suggesting that genital appearance at birth does not determine psychosexual outcome in a consistent way. Our clinical impression is that the general appearance of the child, in combination with masculine behavior, is probably more important than the genital appearance for developing a male gender identity. Masculine-looking and -behaving girls may develop a different sense of self and evoke different responses from family members and

peers than feminine-looking and -behaving girls. Stoller (1985), a psychoanalyst with a special interest in gender development, described such a girl, Mary, who was later identified as having 17β-HSD-3. Mary was raised as a girl, "but," said Stoller, "the baby was difficult. She was too active, too forceful, too ungraceful. Later she refused to accept her mother's inducements to femininity. . . . I found 'her' grotesque as a girl; 'her' mother had attempted to dress 'her' properly for the doctor but it was a bad masquerade. . . . It was clear that this child was living an impossible existence as a girl. I acted on the clinical impression and told her to become a boy. 'She' did, and from that day on I was with a boy. He had immediately known how to be one. To the present, he has been unremarkably, unaffectedly masculine" (Stoller, 1985, p. 66). So Mary's gender role change appeared to be a very natural consequence of how she had been as a child. It is regrettable that virtually none of the studies included in the tables give the kind of information needed to test the impact of physical appearance, gender role behavior, interests, and gender dysphoria in childhood on gender identity.

It has been suggested that a fear of same-sex sexual attraction, making these persons who discover sexual feelings for other women decide that they are better off in the male role, would explain the role changes (Zucker, 1999). Though it seems doubtful that an internalized homophobia operated in all of the reported gender role changes, it may, in some cases, be an additional force in persons who are already gender dysphoric.

Summarizing the scarce literature, it seems that a masculine appearance in childhood, in association with masculine behavior (perhaps both partially caused by prenatal exposure to androgens), make a gender role change likely after the pubertal changes reinforce an already existing gender discomfort. These masculine-looking and -behaving girls will probably need very little additional external or internal pressure to let them decide that living a man's life is what they want.

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