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# PSYCHOSOCIAL CONSIDERATIONS IN THE MANAGEMENT OF LATE - DIAGNOSED MALE PSEUDOHERMAPHRODITISM*

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**Male pseudohermaphroditism (MPH), which causes ambiguous genitalia, rarely presents during adolescence. Herein we report two siblings diagnosed with MPH at the ages of 16 and 12 years and raised unambiguously as girls. Individuals with MPH provide caregivers the opportunity to observe the complex interaction between the biologic and psychosocial forces which shape gender identity. Factors which contribute to gender identity and the management of sex assignment may differ between cultures with regard to dominating sociocultural attitudes towards masculinity and femininity in a given society. The decision to perform feminizing surgery on both patients was made in consideration of these issues.** *Key words: pseudohermaphroditism, gender identity, adolescence, pseudohermaphroditism-psychology.*

Male pseudohermaphroditism (MPH) is the result of incomplete virilization of the 46, XY individual with testes, and patients with this condition have ambiguous genitalia[1]. The diagnosis of MPH is generally made at birth, and treatment must be initiated immediately in order to prevent physical and psychosocial consequences[2, 3]. If the diagnosis is not made until puberty, virilization occurs as a result of stimulated hormonal activity[1, 3, 4]. This may lead to severe sexual identity confusion. Given the fact that body awareness increases at puberty, at this time the choice of gender must be considered very carefully. Herein we report two siblings diagnosed with MPH at the ages of 16 and 12 years and raised unambiguously as girls.

## Case Reports

The patients were the second and third children in a family. The other two sisters were healthy. The mother and father were close relatives. The patients' maternal uncle also had MPH and was reassigned as a male at the age of 12 years.

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*Case 1*

A. was 16 years old and had been raised as a girl. Her complaints were having "double genitalia", deep voice and no breast development. Two years earlier she had discovered by herself that her genitalia were different from her sisters'. Preoccupied with this matter, she had withdrawn and become depressed with an overwhelming shame. After a year, her apathetic and morose behavior as well as voice were noticed by her parents. She reluctantly agreed to seek medical help.

On physical examination, stage I breast development and stage III public hair growth were found. There was incomplete labioscrotal fusion and urogenital sinus opening.

Bilaterally palpable testes were present in the inguinal canal under the labiosacral folds. This finding was confirmed with ultrasonography (USG), which also revealed that there was no uterus or ovaries. X-ray examination of the internal genitalia showed a rudimentary vagina with no uterus. Chromosomal analysis proved a 46, XY karyotype. Laboratory data revealed a plasma-free testosterone of 8.6 pg/ml, total testosterone of 283.9 ng / dl and dihydrotestosterone level of 1.5 ng/dl. Follicle stimulating hormone (FSH) was 64 IU/ml, luteinizing hormone (LH) 10.3 IU / ml, estradiol 48 pg /ml, dehydroepiandrosterone sulfate (DHEAS) 185 mg / ml and 17-OH progesterone was 1.4 ng/ml. All of these values were within the normal range for boys. Human chorionic gonadotropin (HCG) stimulation test denoted a normal response of testosterone and dihydrotestosterone.

In conjunction with these diagnostic procedures, psychiatric assessments were made. During the first interview, A. was almost whispering in order to conceal her masculine voice. Her head was covered by a scarf indicating a strong religious belief. Being fully aware of what was happening to her body, she was severely depressed at the time of evaluation and cried throughout the interview. The pressure of growing pains related to her sexual identity confusion lowered her self-esteem tremendously. As a result, she had become detached and fairly passive in her interpersonal relations. As expected, she took a considerably long time to think about the issue. Considering the sex reassignment as intolerable, A. decided not to change her female gender. In a determined manner, she stated that if she could not be a female, she would commit suicide. She had plans for suicide in the case of a masculinizating operation against her consent.

A battery of psychological tests consisting of the Sentence Completion test, Minnesota Multiphasic Personality Inventory (MMPI), Beck Depression Inventory (BDI) and the Draw-a-person test were performed. According to the results, the

patient was unusually sensitive, rigid and pessimistic. She was also expressing fears concerning her future sexual life. Her identification with the female sex was apparent in her drawings and wishes of becoming an aunt, a grandmother and a nurse. Subtle implications of negative identification with men were present. Her BDI score was 28, indicating a depressive episode.

A. had grown up in a village located in a rather conservative, rural area of Turkey. She had completed elementary school in the village and had not continued her education. Shortly thereafter, her family migrated to a metropolitan area and resided in a peripheral part of the city. She was treated unambiguously as a girl within the family. She was quiet, unaggressive, obedient and restricted in fancifulness. Traditional sex role expectations and stereotypes were adopted very rigidly by the family. Her parents consistently reinforced sex-appropriate behaviors. The patient busied herself with domestic chores. Under these sociocultural upbringing conditions, she identified completely with the female gender identity. A had strong sexual inhibitions and had not experienced any cross-sex friendship.

In a six-month follow-up period after surgical reinforcement of female genitalia, A. was relieved of her depression, gradually became more sociable and began to work in a factory. It was obvious that she had overcome her sexual-identity confusion.

*Case 2*

B., sister of A., was a 12-year-old girl with a four-month history of a deepening voice. When she was a baby, the family questioned whether her genitalia were normal, but the physician who examined her had confirmed that she was normal.

At the age of nine years, on second examination, it was said that she had a right-sided inguinal hernia and a repair operation was performed. Following A.'s diagnosis, B. was examined once again in our hospital.

B. also had ambiguous genitalia. Stage I-II pubic hair growth, stage I breast development and an absence of axillary hair were noted. A palpable gonad was present only in the left inguinal canal. Instead of a penis she had a small clitoris. There was also a urogenital sinus opening. There was no genital pigmentation. Neither uterus nor ovaries were seen on pelvic USG examination. Genitography visualized a rudimentary vagina. Her hormonal profile was completely normal for boys. According to laboratory data, free testosterone was 1.5 pg / ml, total testosterone 30.3 ng / dl, basal dihydrotestosterone 12 ng / dl, FSH 17.6 IU / ml, LH 2.9 IU / ml, estradiol 32 pg / ml, DHEA-S 251 mg / ml and 17-OH progesterone was 1.1 ng / ml. Following HCG stimulation, normal responses of testosterone and dihydrotestosterone were elicited.

On psychiatric evaluation, B. seemed to be an intelligent, stable and conforming school girl. She was popular within her peer group. Night fears were the only psychological complaint that she reported. She was somewhat uninterested in and less affected by the existing problem. Her mood was euthymic; however, she regretted that her problem saddened her parents. Psychological testing data provided evidence that B. had a sense of being female and no interest in sex. She was well adjusted to the "cheerful girl" image which was a part of her "self." Highly influenced by her sister's gender choice, B. was also determined to live as a female. Six months after the feminizing surgery, she was continuing to function well psychosocially.

## Discussion

Both cases were diagnosed as incomplete testicular feminization. Experiencing shame and guilt feelings, the parents left the decision of gender choice to the physicians and patients.

After a multidisciplinary team approach, the decision was made to perform feminizing surgery on both patients. In a final meeting with the participation of pediatricians, endocrinologists, radiologists, pediatric surgeons, medical geneticists and psychiatrists, the issues of gender role, gender identity, sex of rearing and the anatomy of the genitalia were carefully evaluated. Nevertheless the most influential factor in making the final decision was the gender identity which was tenaciously defended by both patients. Gender identity is acquired by postnatal psychosocial experiences as well as prenatal hormonal influences[1, 3-5]. Individuals with ambiguous genitalia, such as MPH, provide health care providers the opportunity to observe the complex interaction between these two forces.

Gender role change during adolescence has been noted in MPH individuals[4,6,7]. Imperato-McGinley et al.[4] described 18 MPH patients with 5-alpha-reductase deficiency who were raised as girls; at puberty, after virilization had occurred, 17 out of 18 changed to the male gender. These data have been interpreted as the effect of testosterone, which predominantly influences sexual identity and overrides the effects of rearing as girls. In another study, MPH individuals with 17-beta-hydroxysteroid dehydrogenase enzyme deficiency, contrary to having been raised as girls, mostly preferred to become male after puberty[7]. Nonetheless, subtle sociocultural factors may have been involved in both of these studies[1, 5, 7].

Presumably, sex reversal at puberty may create medical, psychological, social and moral problems. Surgical interventions, such as reinforcement of gender or reassignment, is difficult and multistage[8]. Even early surgical interventions

were not able to prevent psychosocial injuries to personality and gender role, which were the results of parent-child and environment-child interactions[9]. As shown by Money et al.[10], childhood stigmatization originating from either family or peers was the most important factor significantly correlated to later gender transposition such as bisexualism, lesbianism or sex reassignment to live as male in a group of MPHs who were assigned as female at birth. This implies the critical importance of socio-cultural factors, whether subtle or overt.

Cultural generalizations and expectations about sex-typed behaviors shape an individual's sexual orientation and sex roles[5, 11]. To complete the discussion, it is crucial to take into account our patients' culture-bond status. In Turkey, strictly defined sex roles and separate social networks exist[12, 13]. Dealing with domestic chores, child care and the home are the women's duties. Thus, in contrast to men, women are restricted to a home-bound and inferior status. As Kandiyoti[12] noted, "This status difference is reflected in a stereotypic definition of sex roles as well as the custom of physical and social segregation." Because of well-differentiated sex roles, socialization into a female world occurs and same-sex social networks develop[12, 13]. Together with the strict social control, these networks are the sources of submissive role identification of women.

Noting that these values also dominated in our patients' patriarchal nuclear family, gender role change was a real threat for their already established sexual and social identity. It would have necessitated changing all of their networks and losing their most intimate relationships. Neither was ready for the traditional "bread-winner" role of men. In both patients, the preference for surgical reinforcement after considering all the implications of management was the result of fears related to these future uncertainties and challenges. Although fertility is very important for both sexes and gives an opportunity to increase women's status in the Turkish family[14], and being a male has advantages in the community[12, 13], both patients prefered to be sterile females rather than become dysfunctional males. Such a sacrifice is the product of the culture and is contrary to certain western social values[7]. In addition, our report defies oversimplifications such as, "In underdeveloped countries, males enjoy a higher status than females, even if imperfect"[3].

As suggested by previous researchers[1-3], early diagnosis of ambiguous genitalia before gender identity establishment is imperative, and prompt surgical treatment should be initiated. In both cases, gender identity and sex role identifications had been developed rigidly by the time of diagnosis. Under the aforementioned circumstances, the future risks associated with sex-reassignment were greater than the price the patients had already paid. In conclusion, these cases suggest the relative importance of sociocultural forces–at least in some societies–on the classic "nature versus nurture" debate. In different societies, depending on the power of cultural values and norms, individuals may differ in their choices of gender identity.

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