Profile of Siblings with Disorders of Sexual Differentiation and Gender Dysphoria

To the Editor,

Parents are always curious about the sex of the child immediately after the delivery. The obvious answer they expect is whether it is a "boy" or a "girl." Ambiguous genitalia in the child disrupt the social expectations of the parents entirely.[1] 46XY karyotype and normal testes that produce testosterone lead to the development of external male genitalia. 46XX karyotype and the presence of ovaries that produce estradiol lead to the development of external female genitalia.[2] Ambiguous genitalia result due to increased androgen in females and decreased androgen in males. It is an urgent condition as the sex of rearing needs to be determined, and physicians, psychiatrists, sociologists, and surgeons need to give their valuable perspectives to help parents make a decision. The parents need to assign a gender to the child keeping in mind the different social and psychological issues.[3] Predicting the future gender identity of the child is a complex task.[4,5] This is due to the uncertainty about the exact influence of biological versus psychosocial factors.[3] "Nature" versus "Nurture" has been a longstanding enigma for the health care professionals.

Case Report

We present a report of two siblings with disorders of sexual differentiation (DSD) who were 18 and 20 years of age. They belonged to a very conservative and orthodox male-dominated family and culture. Because they had ambiguous genitalia at birth, their parents chose to rear them as females, due to lack of awareness and the significant stigma attached to getting a medical opinion. The elder sibling had a male gender identity due to his strong sexual attraction towards females and a strong repulsion towards performing female social roles. The younger had a female gender identity due to minimal sexual attraction towards females and a liking to perform female social roles.

They were referred by the Department of Urology, Christian Medical College, Vellore, after a detailed medical examination. Chromosomal analysis revealed 46XY karyotype. The referral was primarily for the elder sibling who had significant gender dysphoria.

Detailed psychiatric assessment was performed on both the siblings using a questionnaire developed by the Dept. of Adolescent Psychiatry, which had two parts. The first part was a cross-cultural assessment of the family and the issues related to rearing and gender identity. The second part was with the siblings, where a gender identity interview was conducted. Though both the siblings were reared as girls, there was a significant difference in the gender identity and the sexual orientation in them. The gender identity was compatible with the sex of rearing in the younger sibling and caused dysphoria in the elder sibling who was more comfortable identifying with the biological gender. Parents, knowing that the siblings were phenotypically male, came for an opinion regarding the appropriate course of action. This was reflected onto both the siblings in several sessions within a period of one year.

The elder sibling was fully convinced from the outset that he wants to perform male sexual roles and had never been comfortable being reared as a female. He had a heterosexual orientation and was sexually attracted to the female sex. On serial interviewing, his ideas and conviction did not change, and he expressed a strong desire to undergo surgery for changing ambiguous genitalia to those of a male.

The younger sibling, in the initial interview, was happy with the female gender he was reared as and was happy to perform the female sexual roles. He did not express any sexual interest or attraction to either sex. He was given feedback about genotype and phenotype. He was willing to reconsider the pros and cons of continuing to have a female gender identity. Six months later, he changed his mind to live as a male and not as a female. He decided to undergo a surgery to change the ambiguous genitalia to those of a male. He attributed it significantly to his elder brother's attitude and parents' suggestions of the social advantages of being a male.

During a joint session, after taking a verbal consent from the siblings and parents, they were advised to perform male roles as part of the "real-life test." [6] They had been living and dressing in the same manner as the conservative women of their community. They were advised to change their clothing and appearance to match with the typical males of their community. This was advised because we needed both the siblings to understand and experience the male social roles. They were emphasized about improving their personality as a whole rather than primarily focusing on sexuality. They were able to understand the limitations of the sex reassignment surgery.

The next review was six months later, and both the siblings were satisfied with living as a "male." Though the Transgender Persons (Protection of Rights) Act, 2019, was in force from December 5, 2019, its legal implications were not applicable to any of the siblings, because the first stage of the surgery was already done prior to that. They had even changed their names legally to what is appropriate for males in their culture. Finally, both the siblings were mentally prepared to undergo sex reassignment surgery, which was planned in two stages.

Both the siblings underwent the first stage of the surgery without any significant physical or emotional setbacks. They continued to have a male identity and were satisfied with performing the social roles as males.

Discussion

This case report highlights the interesting presentation where both the siblings who had similar biological phenotype and rearing had different gender identities. This clearly reinforces that both "nature" and "nurture" are equally important in determining gender identity. The challenges, in this case, were particularly with the younger sibling. He preferred a female social and sexual role, which was in contrast to his male phenotype. Secondly, the surgeons had explained to him that sex reassignment to a female would have a lot of deleterious effects in terms of hormone therapies and challenges in surgical procedures. During the psychotherapy sessions, these points were clearly explained to him. The point that his body is incompatible with performing female sexual functions was reinforced to him as well as his parents. Since enough time was given to the younger sibling along with good family support, he could align to a male gender identity. This was also strengthened by performing the "real-life test."[6]

This report highlights the importance of early psychological intervention and education prior to sex reassignment

surgery to prevent psychological problems.[7] It also signifies the importance of the society in assigning the social roles appropriate to the individual's gender identity rather than the sex determined by the external genitalia.[8] The significant amount of confusion and dysphoria faced by these individuals can be appropriately addressed by timely identification, referral, and expert psychological management.[9] This also results in favorable post-surgical outcomes in terms of patient satisfaction and improved quality of life.[10]

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Pleural Effusion In A Patient With Injection Heroin Use: An Unusual Presentation With An Unusual Pathogen

Sir,

According to a recently conducted nation-wide survey, in India, about 8.5 lakh people use injection drugs. Punjab, with nearly 88,000 people who inject drugs (PWID), has the second-largest drug-using population.[1] Injection drug use (IDU) can cause a wide array of infective and non-infective pulmonary complications: pneumonia, cardiogenic edema, acute lung injury, pulmonary hemorrhage, and aspiration pneumonia.[2,3] We report a rare pulmonary complication of injection drug use: septic emboli caused by an unusual agent, *Burkholderia cepacia*, which resulted in pleural effusion.

Case Report

A 22-year-old unmarried, unemployed male, resident of urban Chandigarh, was admitted in the surgical emergency in mid-December 2019 with complaints of acute onset of chest pain (which worsened during inspiration), breathlessness, and intermittent pain in the right lower abdomen for the last one week and fever for two days. History revealed regular use of heroin for the last five years and use was predominantly via injections for the last two years; if unable to procure heroin, he would chase *Smack* once or twice a month. He would share injection equipment and use tap water as a reconstituent. The place of injection would largely be empty and dirty parking lots and public toilets. Urine drug screen revealed the presence of morphine and cannabis. Initial assessment revealed a pulse rate of 94/min, blood pressure 120/76 mmHg, and an axillary temperature of 101° F. The chest auscultation was unremarkable. His past medical record revealed hepatitis C seropositivity.

Initial blood investigations revealed the following: hemoglobin 12 g/dl, total leucocyte count 7300/mm^3 (differential count: neutrophil 67%, lymphocyte 32%, monocyte 7%, and eosinophil 4%), platelet 2,73,000/mm^3, bilirubin (total: 0.7 mg/dl; conjugated: 0.2 mg/dl), and total protein 7.1 gm/dl. He was seronegative for both hepatitis B and human immunodeficiency virus (HIV1&2). Electrocardiogram was unremarkable.