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Psychological Problem in Children with a Disorder of Sex Development and Their Relatives: A Narrative Literature Review

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ABSTRACT

Disorder of sexual development (DSD) is a challenging disorder that must be done clinically and physiologically. The patients and relatives may experience various psychosocial problems that impact their lives. This study aimed to describe psychological problems in children with DSD and their relatives. Psychosocial and social-environmental problems and management of DSD that are identified in earlier times can reemerge during childhood. Diagnosing a child with a rare disorder, such as mosaic sex chromosome DSD, is often stressful for parents and families. Parents' stress levels and coping strategies are influenced by the diagnosis, type of disease, severity, cause, and presence of non-invasive, invasive, surgical, mental, and behavioral changes and treatment or therapy. In conclusion, psychosocial care for patients and relatives affected by a disorder of sex development is currently limited. Families struggle with challenges such as genetic information, medical complexity, anatomical differences (whether or not surgery is done), surgical complications, lack of clarity in some gonadal tumor risks, doubt about the stability of the child's gender identity, and fertility potential.

1. Introduction

A pediatric disorder of sexual development (DSD) happens when a child is born with sex organs that did not develop properly due to a chromosomal abnormality or the production of hormones.[1,2] The disorder can range from mild to severe, affecting the development of genitalia and reproductive organs at different ages. A child with DSD is often associated with stigma and kept hidden from society, especially in developing countries. That can be happened because of the poor understanding and awareness about DSD.[3,4] The molecular analysis needed to diagnose DSD is limited to research centers in big cities, whereas early diagnosis and treatment are crucial for DSD patients.[5] These limitations made the patients had not received proper medical care and were still in doubt about their gender identity.

Individuals with DSD have several manifestations, from genital ambiguity to phenotypes, that can go unnoticed or appear normal.[6] Genetic, hormonal, and environmental factors during prenatal and postnatal development are likely the factors that are responsible for DSD manifestations.[5] The incidence of DSD is estimated to be 1 in every 4,500-5,500 births.[7] Diagnosis and containment procedures in DSD condition as soon as possible is essential, so it can be done to minimize the complications, not just medical but psychological and social.[8] This study aimed to describe the psychological problems in children with DSD and their relatives.

Psychological problems in patients with DSD

Psychosocial and social-environmental problems and management of DSD that are identified in earlier times can reemerge during childhood. The pediatric and child life psychology specialties are highly useful in cases, especially when urogenital surgery is done beyond infancy. However, the genital examination should be kept as minimum as possible because the child will recall these experiences as traumatic or contributing to a negative body image. Compared to children who remain uninformed or misinformed about their condition, children with adequate education will have better opportunities to develop adaptive coping skills, including positive self-image and expectations for fulfilling their future adult life.[9]

In puberty, they may develop some anxieties. Repeated genital examinations, medical photography, repeated medical treatments, and atypical genital appearance are particularly anxiety provoking. Some will develop uncertain feelings about their masculinity or femininity, sexual adequacy, or sexual orientation.[10] They often postpone intimate relationships due to insecurities and fear of rejection. All of these events can lead to behavioral problems.

Psychological problems in relatives

The birth of a child with mosaic sex chromosome DSD with ambiguous genitalia often creates confusion for parents. They are faced with their children being born without a "clear gender" or with "two genitals." This condition will undoubtedly be challenging to accept by parents, especially when there is a lack of information about children's disorders. Parents often show different expressions, such as surprise, anger, sadness, shame, and, especially for mothers, feelings of guilt. Parents with DSD in their children have often isolated their selves from the community.[11,12]

Diagnosing a child with a rare disorder, such as mosaic sex chromosome DSD, is often stressful for parents and families. The level of stress and coping strategies parents use are influenced by the time of diagnosis, type of disease, severity, cause, presence of non-invasive, invasive, or surgical, mental and behavioral changes, and treatment or therapy.[7] Empirically, a coping mechanism is a process by which an individual can manage a problem through emotionally acceptable behavior. Coping can also be interpreted as an individual's acceptance of demand, the ability to reduce or tolerate pressure, and management to control emotions to reduce stressful situations. Psychological support for parents must be provided continuously and continues to extend to the whole family to accept the child's condition and use appropriate coping to overcome it.[13] Support from family, neighbors, work environment, and the environment around the house is an essential factor for parents who have children with DSD.[13,14]

Children diagnosed with DSD have a heavy psychological impact on them or their parents. Depression, anxiety, post-traumatic stress, and uncertainty of cure are some things that might happen to them. Another problem is that the patient will be treated for a long time. This condition is exacerbated by the culture, which is considered taboo to talk or discuss abnormality in sex and genitalia, so the parents often keep their child's condition secret and do not seek help.[9] Detection of a DSD patient at birth is commonly an acute problem. Some parents explain that high emotional distress and cognitive confusion often occur at that time.[10] The delayed intervention may reduce the quality of life in patients, implying the significance of early detection and diagnosis.

DSD is not widely known among health practitioners. Clinicians specialized in DSD treatment are challenged with patients' and parents' difficulties in coping with their atypicality's unusual physical development and depreciative reactions. Diagnostic and treatment facilities also limit it. Treatments necessary for survival aim to reduce physical atypicality and repair sexual function. These interventions have been criticized for impacting the child's life and are often performed without the child's consent.[15]

2. Conclusion

Psychosocial care for patients and relatives affected by a disorder of sex development is currently limited. Families struggle with challenges such as genetic information, medical complexity, anatomical differences (whether or not surgery is done), surgical complications, lack of clarity in some gonadal tumor risks, doubt about the stability of the child's gender identity, and fertility potential.

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