PubMed Included

ISSN 0971-9261

July-August 2022 | Volume 27 | Issue 4

Journal of Indian Association of Pediatric Surgeons

JIAPS

Official Publication of
Indian Association of Pediatric Surgeons

Online at
www.jiaps.com

Wolters Kluwer

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Changing Paradigms in Intersex Management: Legal, Ethical, and Medical Implications



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Background

The term "intersex" covers a variety of conditions also known as disorders/differences of sex development (DSD). The older terminology, with phrasing such as "hermaphrodite" and "pseudohermaphrodite," was felt to be pejorative, confusing, and stigmatizing. The new classification proposed is sensitive to individuals and families, and also more reflective of molecular understanding in sex development research.[1,2] There is still considerable controversy in the field regarding proper and respectful nomenclature. The consensus statement of the year 2018[3] renamed all the conditions known erstwhile as intersex, pseudohermaphroditism, or disorders of sex differentiation proposed in 2006[2] into a more inclusive terminology, DSD. While the term "intersex" has been replaced by DSD in medical literature, patient support groups still prefer this term.

The terms "gender," "sex," and "sexual" have discordant interpretations. "Gender" is a social concept, which is the way the society mirrors the "individual identity." It does not take into account the "individual identity" ("inside identity") and the future "gender role" ("behavioral identity"), which are invisible at birth and the modalities of which are mostly unknown.[4] Gender identity refers to a fundamental sense of belonging and self-identification of one's gender as male, female, or an alternative gender.[5] The term gender role describes the behaviors, attitudes, and personality traits that a society designates, in a given culture, as masculine or feminine. Gender role should be considered distinct from core gender identity.[6]

The term "sex," on the other hand, refers to anatomical characteristics (phenotypic sex, genotypic sex – karyotype, gonadal sex, and internal anatomy) and sex of rearing – sex assigned by parents or caregivers. Sexual orientation, a totally different entity from gender, is defined by a person's responsiveness to another person, the salient dimension being the sex of the person to whom one is sexually attracted (heterosexual, bisexual, etc.).[2] Thus, when we discuss further, we would be talking of sex assignment and not gender assignment, as gender is a self-identity.

Several studies[7,8] have disputed the "optimal gender policy" for infants with DSD. While the notion that "nurture overrules nature" in gender identity prevailed in the past, studies have shown that DSD children do not always conform to the sex of rearing as they grow into adults.[9] Gender dysphoria refers to a conflict between a person's physical or assigned sex and the gender with which they identify. The more recent term for gender dysphoria is gender identity disorder (GID). People with GID[10] have a strong and persistent cross-gender identification manifested in adolescents and adults by symptoms such as a stated desire to be the other sex, desire to live or be treated as the other sex, or the conviction that he or she has the typical feelings and reactions of the other sex.

Legal Implications

The term transgender refers to a person whose sense of personal identity and gender does not correspond with their birth sex. They are totally different from DSD in the sense that they are born with unambiguous sexual anatomy. They seek sex reassignment surgery in adulthood due to gender dysphoria, and it is performed by adult surgeons with their personal consent. On the other hand, intersex surgery is often performed in children with ambiguous genitalia, essentially for medical indications by pediatric surgeons after obtaining parental consent.

Access this article online

Quick Response Code:

Website: www.jiaps.com

DOI: 10.4103/jiaps.jiaps_41_22

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How to cite this article: Babu R. Changing paradigms in intersex management: Legal, ethical, and medical implications. J Indian Assoc Pediatr Surg 2022;27:372-5.

However, the LGBTQIA+ activist groups consider intersex children's rights along with transgender rights. The 2019 case in Madurai bench of Madras High Court started with rights of a transgender marriage. When the intersex activists pleaded to be heard on intersex surgeries, the judge[11] asked the director of medical education to formulate guidelines on intersex surgeries. The judge quoted from Khalil Gibran: "Your children are not your children. They are the sons and daughters of Life's longing for itself. They come through you but not from you, And though they are with you yet they belong not to you. You may give them your love but not your thoughts, For they have their own thoughts. You may house their bodies but not their souls, For their souls dwell in the house of tomorrow, which you cannot visit, not even in your dreams. You may strive to be like them, but seek not to make them like you."

A different hearing on Madras High Court in 2022 on a transgender marriage grievance (they sought police protection from their parents!) took up difficulties faced by LGBTQIA+ community[12] and issued directions to the National Medical Council (NMC) and State Medical Councils to incorporate medical and legal updates in their curricula: avoid pathologization and criminalization of nonheterosexual relations, gender nonconformity, gender incongruence, etc., The NMC and State Medical Council were asked to appoint a committee and submit a compliance report on the recommendations. In this regard, the 2020 guidelines already formed by the Tamil Nadu government (following the earlier court directive) to regulate DSD management gains relevance.

Ethical Implications

Human Rights Watch, USA, Congenital Adrenal Hyperplasia Research/Education/Support Foundation, and Advocates for Intersex Youth (InterACT) along with Dr. Dix Poppas, chief of pediatric urology at Cornell-Weill Medical Center in New York City, produced an extensive document after interviewing several intersex adults.[13] They felt that assigning a sex of rearing to a child never required surgery and suggested society of pediatric urology to issue guidance on surgeries in individuals born with DSD. They also felt that intersex surgeries are unethical and should be avoided until the patient can actively participate in decision-making.

Medical Implications

Assigning a sex of rearing is a great responsibility and should not be rushed. Many factors must be taken into account, and thus, it is essential to collect all relevant information while still being expeditious. When addressing the infant, it is important to initially be gender-neutral and avoid "he" or "she" pronouns that could inadvertently bias the family toward a gender that may later be reversed. Rather than referring to the child as "it," warmer terms like "your baby" may promote greater bonding. Staying neutral until the decision is finalized helps prevent misunderstandings and confusion. A recent meta-analysis[14] showed that GID is low in women with congenital adrenal hyperplasia (CAH), complete androgen insensitivity syndrome (CAIS), and complete gonadal dysgenesis favoring female sex of rearing in them. GID is high in women with 5-alpha reductase deficiency/17-hydroxysteroid dehydrogenase deficiency favoring male sex of rearing in these DSD. GID is variable in partial androgen insensitivity syndrome (PAIS) or mixed gonadal dysgenesis (MGD), and no recommendations on sex of rearing could be made in these conditions. In PAIS/MGD children, multiple factors such as local anatomy, hormonal profile, and genetics have to be considered. These parents have to be given the option of leaving the sex indeterminate until their children are able to develop gender identity as they go through adolescence.

Way Forward

In view of the growing concerns, it is imperative to understand the intricacies and come up uniform national & state guidelines. DSD is ideally managed only in specific centers with multidisciplinary committee. The medical director/dean of each tertiary hospital/medical college should act as a lead in the formation of such local multidisciplinary committee (LMDC). The committee should include at least three specialists: pediatrician/endocrinologist, pediatric surgeon, and psychologist/psychiatrist. Sex assignment should be done only at designated centers by LMDC after thorough evaluation, diagnosis, and discussion with all stakeholders. An Apex multidisciplinary committee should be formed at the national & state capital (the lead pediatric surgical center).

Decisions about nature and timing of any surgery are made with the family (or involving the adolescent child), by the LMDC acknowledging the considerable psychological impact. When a potentially "life-threatening" circumstance arises (urosepsis/cancer risk to the life) on the intersex child, the interest of the child is of paramount importance. Such decisions can be made after discussion with LMDC if urgent: DSD with holdup of menstrual or mucinous fluid or recurrent urine infection due to stasis of

urine in common channel – becoming potentially "life threatening" due to urosepsis (in these children, vaginoplasty/such procedures to clear excretory fluids are justified). In children with XY/MGD with streak gonads, the risk of cancer is high (40%–60%) making it potentially "life threatening."[15] Excision biopsy of streak gonad/gonad in PAIS/ovotestis with high risk of cancer is justified after getting informed consent. LMDC should keep a record of a signed document from all members and parents about the necessity for surgical intervention.

There are several other conditions like CAH, where the guidelines on timing of surgical intervention are still evolving. Clitoral surgery has been known to alter/impair sensation[16,17] although Canning felt that follow-up of older techniques is unlikely to represent outcomes of current nerve-sparing procedures.[18] In most cases of CAH, an adequate hormonal suppression is known to control clitorimegaly.[19] Several authors recommend delaying of vaginoplasty until puberty when it can be safely performed with informed consent of the grownup.[20,21] On the other hand, twothirds of caregivers of female infants with CAH reported not regretting their decisionmaking of early surgery during childhood.[22] Majority of females with CAH and parents believe that CAH should be excluded from the intersex designation, and should be considered separately in legislation pertaining to childhood genital surgery.[23] While the cry to delay cosmetic surgeries to a later date gets louder, unknown effects of having atypical genitalia on children and parents are unclear.[24] Only recently a small feasibility study on deferring surgery in CAH, has reported that girls and their parents did not express significant concerns regarding genital ambiguity.[25] Hence, when forming national/ state guidelines, one has to weigh the pros and cons of genital surgery in CAH. Another area of debate is Prader 5 CAH reared as male, in whom a late feminizing genitoplasty may be unwise as due to androgen effects they often do not have gender dysphoria as men.[26] These children may need hysterectomy/gonadectomy, but this should be delayed until puberty when an informed decision can be made.[27]

In children with CAIS, the risk of germ cell tumor is <1% and the risk is only after the second decade.[15] Therefore, gonadectomy can be postponed allowing spontaneous feminization at puberty while avoiding hormonal replacement.[19] The least controversial of all decisions would be hypospadias surgery in those with bilateral palpable testes and XY karyotype. Appropriate timing of staged hypospadias repair is around 9–18 months,[19] and these children do not need to go through LMDC/DSD pathway.

Summary

There is a growing concern among activists that sex assignment is often performed with binary notion (fitting them into male/female framework), discarding the option of third/neutral sex assignment. Although gender-neutral upbringing and a third sex of rearing are accepted by the Law, in the Indian context the parents prefer to have a sex assigned for the baby, in order to provide a safe and socially secure upbringing. Even in Western countries where there is social acceptance of third gender, the psychosocial ramifications of such approach (not choosing a gender) are unclear[28] and there is no law yet to regulate their medical management.

In an Indian setting, the children born with DSD are completely at the support of their parents, who in turn are dependent on their extended family and society (not social welfare/government support). Denying them the necessary medical attention may lead to child neglect, or casting their child away. While the court has directed school authorities to build toilets for third gender, in the Indian setting where open toilets are still prevalent, these children are likely to be bullied and teased by other children leading to depression or self-harm.

Until views of the society are changed via education or self-realization, imposing laws or rules to ban all DSD surgeries will make the parents turn to quackery to achieve their aims. A rather better way is to ensure that DSD children continue to receive required medical care from qualified pediatric surgeons. It is indeed prudent to regulate these surgeries via guidelines, regulations, consensus, and informed decisions involving LMDC and parents. Each DSD patient is unique, and they warrant multidisciplinary care and long-term psychosexual support.

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Submitted: 05-Mar-2022.
Accepted: 13-Mar-2022.
Published: 26-Jul-2022.

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