Bringing Back the Term “Intersex”

Johanna Viau-Colindres, MD, a Marni Axelrad, PhD, b Lefkothea P. Karaviti, MD, PhD, a on behalf of the Texas Children’s Hospital Gender Medicine Clinic a

The birth of a child with a disorder of sexual development (DSD) is challenging from multiple perspectives, particularly that of sex assignment. Patients, parents, and health care professionals who care for patients with DSDs understand that the physical, biochemical, and mental characteristics of these patients better fit a model of fluidity. Yet, sex continues to be predominantly binary, and the decision that families make early on often has medical, social, and legal implications.

Until recently, all US birth certificates listed either male or female sex. However, in December 2016, the state of New York issued the nation’s first intersex birth certificate to a 55-year-old individual who was born with a DSD consisting of male chromosomes, female external genitalia, and mixed internal genitalia.1 This legal landmark challenges the traditional legal framework of the binary division of sex. Therefore, we are taking this opportunity to discuss the fluidity of sex assignment and how this unprecedented event may provide an opportunity to improve the care of patients with a DSD.

HISTORICAL PERSPECTIVE

Multiple terms and classification systems have been used to describe patients with DSDs. The classification systems were initially based on anatomy and were modified over time as the chromosomal basis of sex, gonadal characteristics, and sexual differentiation were better understood. However, the subclassifications had the qualifiers of male or female, implying the sex to be assigned.2 It was not until 2006 that the Pediatric Endocrine Society and the European Society of Paediatric Endocrinology revised the nomenclature into a classification system in which the diagnosis did not determine gender assignment, acknowledging that gender is complex and not determined by a single criterion. The resulting international Consensus Statement on the Management of Intersex Disorders, also known as the Chicago Consensus Statement, recommended the use of the term DSD to describe any condition in which sex-related chromosomes, molecules, and/or internal organs do not develop in a way that could be classified as male or female. It also recommended against the use of terms such as male/female, hermaphrodite, and pseudohermaphrodite because these terms can be confusing and may carry negative connotations (Table 1).3,4

Bringing Back the Term “Intersex”

According to the 2006 Consensus Statement on Management of Intersex Disorders.

Psychosexual development is a developmental process. However, the intersex option may be considered a permanent sex assignment that acknowledges the fluidity of gender. Alternatively, the intersex assignment could be used as an acceptable transitional phase until gender identity is developed. However, the intersex option may not be relevant to all DSDs. For example, in 46,XX infants who are virilized because of congenital adrenal hyperplasia and 46,XY infants with absolute absence of androgen insensitivity, available data show that a female sex assignment is associated with gender satisfaction in adulthood in 90% of the time. Therefore, the benefit of an intersex birth certificate in these individuals would be negligible. Conversely, other DSDs such as 5α-reductase deficiency, mixed gonadal dysgenesis, partial androgen insensitivity, and other less common conditions have higher rates of gender dissatisfaction or are missing good long-term data. Therefore, the intersex option could be highly beneficial. Finally, it should be clarified that intersex is a designation or qualifier (similar to male or female) rather than a diagnosis.

As the intersex option becomes available, the health care paradigm can become more society-centered rather than making a patient “fit” the societal binary sex norm. Additionally, clinicians could more appropriately focus on the patient’s individual needs as a person rather than as a male or female because their health care needs may not be applicable to either sex. In any case, clinicians must learn and gain experience in supporting these families, particularly regarding the child’s development and adjustments associated with an intersex designation.

**CURRENT APPROACH**

In addition to using the DSD nomenclature, the majority of providers who care for neonates with DSDs follow the Chicago Consensus Statement management recommendations. These include an extensive evaluation by a multidisciplinary group of experts in DSDs, discussion of the findings with the caregivers, and, finally, sex assignment on the basis of the findings in concert with the family’s values and preferences. This is done as early as possible after all essential information for sex assignment is available. In most patients, sex assignment occurs within the first days or weeks of life, whereas surgeries and irreversible interventions are delayed. Early male or female sex assignment has many advantages. It facilitates conforming to societal norms and reduces parental stress by providing a sense of certainty. However, this practice fails to recognize our inadequate understanding of the development of gender identity. Psychosexual development is a complex process that is influenced by multiple factors, many of which cannot be measured objectively and cannot be altered in patients with DSDs. Therefore, it is not surprising that gender dysphoria is common within the DSD population, with rates up to 50% for certain conditions regardless of the sex assigned. Nevertheless, once a sex has been assigned, sex assignment is seldom revisited because of the sensitivity of the topic, lack of experience discussing it, lack of time, challenges in assessing psychosexual development during early childhood, irreversibility of surgical interventions already completed, and, potentially, provider and/or parental fears that the wrong sex was assigned.

**THE FUTURE: ARE WE READY FOR A NEW APPROACH?**

Society has become more open to gender fluidity or nonconformity, although the legalities have fallen behind. Birth certificates that include the intersex option, such as the one issued in New York, might be advantageous to certain patients with DSDs and should be encouraged. The choice of intersex could be considered a permanent sex assignment that acknowledges the fluidity of gender. Alternatively, the intersex assignment could be used as an acceptable transitional phase until gender identity is developed. However, the intersex option may not be relevant to all DSDs. For example, in 46,XX infants who are virilized because of congenital adrenal hyperplasia and 46,XY infants with absolute absence of virilization because of complete androgen insensitivity, available data show that a female sex assignment is associated with gender satisfaction in adulthood in 90% of the time. Therefore, the benefit of an intersex birth certificate in these individuals would be negligible. Conversely, other DSDs such as 5α-reductase deficiency, mixed gonadal dysgenesis, partial androgen insensitivity, and other less common conditions have higher rates of gender dissatisfaction or are missing good long-term data. Therefore, the intersex option could be highly beneficial. Finally, it should be clarified that intersex is a designation or qualifier (similar to male or female) rather than a diagnosis.

**TABLE 1 Nomenclature and Classification of Disorders of Sexual Differentiation**

<table>
<thead>
<tr>
<th>Preferred Nomenclature</th>
<th>46,XX DSD</th>
<th>46,XY DSD</th>
<th>Chromosomal DSD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Examples of conditions</td>
<td>Androgen excess (ie, congenital adrenal hyperplasia because of 21-hydroxylase deficiency, placental aromatase deficiency)</td>
<td>Defects in androgen synthesis (ie, 5α-reductase)</td>
<td>45,XX/46,XY mixed gonadal dysgenesis</td>
</tr>
<tr>
<td>Disorders of ovarian development</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Old nomenclature equivalents</td>
<td>Female pseudohermaphrodite</td>
<td>Male pseudohermaphrodite</td>
<td>Complete gonadal dysgenesis</td>
</tr>
<tr>
<td></td>
<td>Virilized female</td>
<td>Undervirilized male</td>
<td>Sex reversal</td>
</tr>
<tr>
<td></td>
<td>Masculinized female</td>
<td>Undermasculinized male</td>
<td>Hermaphrodite</td>
</tr>
<tr>
<td></td>
<td>Female sex reversal</td>
<td>Male sex reversal</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hermaphrodite (with 46,XX karyotype)</td>
<td>Hermaphrodite (with 46,XY karyotype)</td>
<td></td>
</tr>
</tbody>
</table>

*According to the 2006 Consensus Statement on Management of Intersex Disorders.*
Although this first legal step recognizes that sex does not always fit the male and female models, one must acknowledge that society remains mostly binary, and changing societal views may take years. In other words, individuals with an intersex birth certificate will have multiple challenges in situations in which the male versus female differentiation is integrated into a policy. For example, unless the current systems are updated, these individuals may have problems obtaining health insurance and other legal documentation such as passports. In addition, sex designations in schools, bathrooms, organized sports, and religious roles, among others, may continue to pose limitations to normalizing the child’s daily life.

It remains unknown whether other states will start issuing intersex birth certificates or whether New York will continue this practice. It will likely take years for this to become common practice. In fact, in the past, similar cases in other states have not resulted in a successful outcome. However, for the DSD community, the option of an intersex designation opens up a world of opportunities and should be encouraged. It offers a better way of managing uncertainty and avoiding an incorrect sex assignment, particularly in conditions in which there are high rates of gender dissatisfaction. Most importantly, it will shift the priorities of care from a series of interventions targeted toward making a patient fit into a male or female model to an acceptance that everyone is unique and differences are appreciated. Therefore, we urge the International Intersex Consensus working groups to support the issuance of birth certificates with an intersex designation, acknowledging that new challenges will arise with this approach but that it will be a move in the right direction.

ACKNOWLEDGMENTS
The Texas Children’s Hospital Gender Medicine Clinic is a multidisciplinary, tertiary referral center for children with DSDs. All of its members actively participated in writing and editing this manuscript. The Texas Children’s Gender Medicine team members are, in alphabetical order, Oluyemisi Adeyemi-Formode, MD (pediatric gynecology); James R. Banfield, JD, BS (legal department); Jennifer Bercaw-Pratt, MD (pediatric gynecology); Rebecca Butler, LMSW (social work); Jennifer E. Dietrich, MD (pediatric gynecology); Sheila K. Gunn, MD (pediatric endocrinology); David G. Mann, MD (ethics); Duong D. Tu, MD (pediatric urology); Vernon R. Sutton, MD (genetics); Jeffrey T. White, MD (pediatric urology); and Shae H. Wilson, JD, BS (legal department). We thank Dr Kimon Angelides for bringing to our attention the legal implications of the intersex sex assignment as an opportunity for better care of our patients. We also thank Dr Fernando Stein for encouraging us to challenge binary sex assignment norms. We are also grateful for the support to the Gender Medicine team provided by Dr Kushner.

ABBREVIATION
DSD: disorder of sexual development

REFERENCES
Bringing Back the Term "Intersex"
Johanna Viau-Colindres, Marni Axelrad, Lefkothea P. Karaviti and on behalf of the Texas Children's Hospital Gender Medicine Clinic

Pediatrics 2017;140;
DOI: 10.1542/peds.2017-0505 originally published online October 25, 2017;

Updated Information & Services
including high resolution figures, can be found at:
http://pediatrics.aappublications.org/content/140/5/e20170505

References
This article cites 6 articles, 1 of which you can access for free at:
http://pediatrics.aappublications.org/content/140/5/e20170505#BIBL

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Endocrinology
http://www.aappublications.org/cgi/collection/endocrinology_sub
Ethics/Bioethics
http://www.aappublications.org/cgi/collection/ethics:bioethics_sub
LGBTQ+
http://www.aappublications.org/cgi/collection/lgbtq

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.aappublications.org/site/misc/Permissions.xhtml

Reprints
Information about ordering reprints can be found online:
http://www.aappublications.org/site/misc/reprints.xhtml

American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN®
Bringing Back the Term "Intersex"
Johanna Viau-Colindres, Marni Axelrad, Lefkothea P. Karaviti and on behalf of the
Texas Children's Hospital Gender Medicine Clinic
Pediatrics 2017;140;
DOI: 10.1542/peds.2017-0505 originally published online October 25, 2017;

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://pediatrics.aappublications.org/content/140/5/e20170505