

MEDICINE AND SOCIETY: PEER-REVIEWED ARTICLE

A Call to Update Standard of Care for Children With Differences in Sex Development

Nat Mulkey, MD, Carl G. Streed Jr, MD, MPH, and Barbara M. Chubak, MD

Abstract

For years, physicians have debated how best to care for children with differences in sex development (DSD, also termed *intersex*). Stories of suffering of adults who underwent early surgical intervention for DSD have led many health organizations to call for deferral of unnecessary procedures. While some have instituted full deferral of cosmetic procedures, standard of care remains an interdisciplinary team approach informed by parents' wishes. As the medical community hesitates to institute full deferral, citing absence of long-term data, legislation restricting early procedures is mounting. This article highlights recent data from the DSD-LIFE Study and considers whether and to what extent they support deferral.

To claim one AMA PRA Category 1 Credit™ for the CME activity associated with this article, you must do the following: (1) read this article in its entirety, (2) answer at least 80 percent of the quiz questions correctly, and (3) complete an evaluation. The quiz, evaluation, and form for claiming AMA PRA Category 1 Credit™ are available through the [AMA Ed Hub™](#).

Sex Development and Best Interests

Differences in sex development (DSD, also termed *intersex*) comprises multiple diagnoses in which there is a congenital condition with inconsistent chromosomal, gonadal, or anatomic sex development.¹ These conditions provoke many questions: What are the potential health risks and **future fertility options** for children with DSD? What surgeries should be offered to parents of children with DSD? What gender should parents raise their child, and what will be the gender identity of the child? Grounded in the historical notion that gender and anatomy are linked, surgical interventions have been performed on infants to align their anatomy with their “optimal gender,” often chosen for them based on potential for heteronormative sexual relationships.^{2,3,4} Today, intersex children continue to receive early cosmetic genital surgery at medical institutions across the United States and worldwide.^{5,6,7} As we discuss here, whether or not to perform early surgical intervention has been framed as a bioethical conflict between upholding the bodily autonomy of the child vs acting on behalf of what are perceived to be the child's best interests. Yet there is little evidence to show surgical intervention supports these children's best interests.

Are Early Surgeries Best?

In 2016, the Global DSD Update Consortium consensus statement reiterated its 2006 recommendation of a multidisciplinary team approach for children with DSD to help ensure that the patient and family were being provided optimal and thorough counseling. This recommendation focuses on **shared decision making** with the parents.⁷ Some have argued that when patients are too young to make their own decisions, the most ethical thing to do is to honor the preferences of parents—who are legally entitled to make health decisions for their minor children—when it comes to surgical interventions.⁸ With this position, however, there is the danger of prioritizing parental wishes and anxieties over supporting the child’s autonomy.⁹

Many individuals upon whom these procedures were performed have come forward as adults to share their dissatisfaction and health complications.^{5,10} National intersex organizations, such as InterACT, have formed worldwide and advocate for the cessation of early surgical intervention on intersex children.¹¹ Members of this community have produced movies and books sharing how the medical interventions performed on them resulted in both physical and psychological harm.^{12,13} Some of these interventions include clitoral reduction, vaginoplasty, and gonadectomy and are accompanied by multiple genital examinations.^{10,12,13} Supporting these stakeholder claims, many prominent national and international health organizations and legislative bodies have issued recommendations arguing for deferral of elective procedures until the child can actively participate in the decision-making process (see Table).

Table. Organization Statement Timeline

Year	Organization Statement
2013	WHO issues a statement that called for the cessation of medically unnecessary surgeries and sterilizations on individuals born with DSD. ¹⁴
2014	Provisional section on LGBT health and wellness of the AAP states that medically unnecessary irreversible procedures can be postponed until a child “is old enough to agree to the procedure.” ⁵
2015	Six UN committees call for regulation of medical interventions of nonconsenting intersex individuals. ¹⁵
2016	GLMA: Health Professionals Advancing LGBT Equality issues a recommendation to delay all medically unnecessary surgeries on intersex children until they “can provide informed consent/assent” (excepting procedures addressing emergent medical need). ¹⁶
2017	Physicians for Human Rights issues a statement calling for deferral of unnecessary surgical procedures on intersex children before they can “give meaningful consent.” ¹⁷
2017	Former US Surgeons General Joycelyn Elders, David Satcher, and Richard Carmona determined that current research does not support performing cosmetic genitoplasty on infants. ¹⁸
2018	California Senate passes SCR-110, which recommended delaying any procedure on children with DSD until the “child is able to participate in decisionmaking.” ¹⁹

Abbreviations: AAP, American Academy of Pediatrics; DSD, differences in sex development; LGBT, lesbian, gay, bisexual, transgender; UN, United Nations; WHO, World Health Organization.

Despite the stated positions of reputable health organizations and legislative bodies and anecdotal reports of suffering, a battle has engulfed the medical community regarding deferral of elective surgical interventions. Arguments for deferral focus on protecting the bodily autonomy of the child and preventing **potential harm** that can come from an irreversible intervention.²⁰ Harms cited that can carry significant implications as a child matures include a sex assigned at birth that does not align with the individual's eventual gender identity and surgical complications that affect sexual functioning.²⁰ In favor of early surgical intervention is the argument that a child with ambiguous genitalia could face psychosocial distress because of this difference as well as the belief that younger children heal more easily and will not remember undergoing surgical correction.^{21,22,23} However, these arguments have little supporting data, while data supporting the opposite position continues to mount. There is an accretion of data that early interventions, their inevitable medical follow-up, and frequent need for surgical revisions later have led to the development of psychological distress.^{5,10,18} Supporters of early surgery counter with claims that these reports are anecdotal and that there is insufficient high-quality outcomes data to support deferral.⁹ However, this argument from inadequate evidence is equally applicable to the outcomes data referenced to support continuation of early, elective surgical interventions.

Evidence and Deferral

Organizations have repeatedly found that the data are insufficient to support DSD procedures as necessary, beneficial, or safe. In 2012, the Working Party on DSD concluded that more rigorous outcomes studies are needed to evaluate the success of early genitoplasty, noting that surgery for some conditions is associated with impaired sexual function and quality of life.²⁴ The Global DSD Update Consortium 2016 consensus statement, updated from the 2005 International Consensus Conference on Intersex, similarly concluded that current data is inadequate regarding key aspects of DSD interventions, including sex assignment and surgical outcomes.⁷

The latest and most rigorous study that assesses long-term outcomes of patients with DSD appropriately compares large samples of individuals who did and did not have surgical interventions. In the DSD-LIFE study, participants who had Turner syndrome, Klinefelter syndrome, congenital adrenal hyperplasia (CAH), or XY-DSD were recruited from 14 European clinics.^{25,26} Researchers examined rates of gender change and dysphoria as well as components of sexuality in this population. For those with CAH, rates of anorgasmia and genital anesthesia were higher among those who had undergone surgery than among those who had not.²⁵ Those who had undergone surgery also reported less intercourse and experienced more difficulties with vaginal penetration.²⁵ Across DSD conditions, having had genital surgery was negatively associated with satisfaction with sex life.²⁵ For those with DSD conditions other than Turner and Klinefelter syndromes, rates of gender dysphoria and subsequent gender transition after puberty were higher than in the general population.²⁶ This study is one of the first to examine quality-of-life outcomes in a large adolescent and adult sample with DSD. These results support deferral of intervention, as DSD individuals were more likely to change assigned gender than the general population.²⁶ The study's concerning findings that those who had undergone surgical intervention had worse sexual health outcomes add to the growing anecdotal reports of harm shared by members of DSD communities.

Part of the reason it has taken so long for changes to be made in the care of children with DSD can be attributed to how the biomedical community ranks the quality of

information according to the hierarchy set forth by the evidence-based medicine (EBM) model.²⁷ In medicine, anecdotes and case reports are considered the lowest quality of data on which to base standards of care. Relegating anecdotal information to a lower tier has been called into question, as it can hinder communication between doctors and patients and delay updates in care models.²⁸ Anecdotes can be interpreted through the lens of autonomy, as bioethicists recognize that to truly uphold patient autonomy, one must respect the individual experiences that motivate a patient's decision making and ownership of their narrative.²⁹ The EBM model is a step forward in clinical practice, but we must recognize that it can unintentionally devalue patient experiences and thus autonomy.

Defining Normal and Optimal

The parental role in a pediatric surgical intervention decision is critical, and a shared decision-making approach is the current standard of care. Critiques of this model highlight that gaps in parental knowledge of DSD hinder parents' perceived and actual participation in these decisions.²⁰ Lack of common understanding of DSD elevates the importance of what information is provided to parents during the decision-making process. One study analyzing parent-clinician interactions found that discussion of surgery to "normalize" the child's anatomy prevails over discussion of the controversies that surround early surgery and the child's autonomy.³⁰ True **informed consent** requires an increased emphasis on the clinician's responsibility to protect the child's autonomy in these discussions. This responsibility includes describing new outcomes data and mention of the multiple organizational calls for deferral of surgical intervention.

More recently, Boston Children's Hospital and the Ann and Robert H. Lurie Children's Hospital of Chicago have stated they would stop performing certain cosmetic genital surgeries on children born with DSD.^{31,32} However, in our experience, clinicians and professional societies hesitate to follow suit or update what constitutes standard of care. Given the growing frustration of health organizations and organizations representing the interest of DSD communities, legislative bodies have begun to recommend deferral of nonessential surgical interventions on children with DSD. Passed in August 2018 in California, Senate Concurrent Resolution 110 recommends delaying any procedure on children with DSD until the individual "child is able to participate in decision making."⁴⁹ The associated bill did not advance out of committee, but efforts toward its passage are ongoing in California, with similar legislation being developed in other states.^{33,34}

Medical experts and patient advocates alike continue to call for more research to determine the optimal treatment for children born with DSD. Meanwhile, reports of suffering by those in the intersex community continue to be devalued, as such anecdotal reports do not take the form of privileged evidence. The new data highlighted in this article and in health care organization statements reinforce the multitude of anecdotes that support deferral. We call on clinicians to heed these repetitive calls to action and to recognize that when an individual is born with atypical genitalia that poses no physical risk, treatment should focus not on surgical intervention but on psychosocial and educational support for the family and child. Cosmetic genitoplasty should be deferred until children are old enough to voice their own views and meaningfully assent to undergoing surgery.

References

1. Beh HG, Diamond M. An emerging ethical and medical dilemma: should physicians perform sex assignment surgery on infants with ambiguous genitalia? *Mich J Gen Law*. 2000;7(1):1-63.
2. Money J, Hampson JG, Hampson JL. An examination of some basic sexual concepts: the evidence of human hermaphroditism. *Bull Johns Hopkins Hosp*. 1955a;97(4):301-319.
3. Money J, Hampson JG, Hampson JL. Hermaphroditism: recommendations concerning assignment of sex, change of sex and psychologic management. *Bull Johns Hopkins Hosp*. 1955b;97(4):284-300.
4. Money J, Hampson JG, Hampson JL. Imprinting and the establishment of gender role. *AMA Arch Neurol Psychiatry*. 1957;77(3):333-336.
5. Knight K. *A Changing Paradigm: US Medical Provider Discomfort With Intersex Care Practices*. Human Rights Watch; 2017. Accessed March 15, 2020. <https://www.hrw.org/report/2017/10/26/changing-paradigm/us-medical-provider-discomfort-intersex-care-practices>
6. Diamond DA, Swartz J, Tishelman A, Johnson J, Chan YM. Management of pediatric patients with DSD and ambiguous genitalia: balancing the child's moral claims to self-determination with parental values and preferences. *J Pediatr Urol*. 2018;14(5):416.e1-416.e5.
7. Lee PA, Nordenström A, Houk CP, et al; Global DSD Update Consortium. Global disorders of sex development update since 2006: perceptions, approach and care. *Horm Res Paediatr*. 2016;85(3):158-180.
8. Daaboul J, Frader J. Ethics and the management of the patient with intersex: a middle way. *J Pediatr Endocrinol Metab*. 2001;14(9):1575-1583.
9. Diamond M, Beh HG. David Reimer's legacy: limiting parental discretion. *Cardozo J Law Gen*. 2005;12(5):5-30.
10. Davis G. Normalizing intersex: the transformative power of stories. *Narrat Inq Bioeth*. 2015;5(2):87-89.
11. Shipenberg Z. InterACT educates about intersex. *Hofstra Chronicle*. October 30, 2019. Accessed March 15, 2020. <https://www.thehofstrachronicle.com/category/news/2019/10/30/interact-educates-about-intersex>
12. Knight K. Film shows surge in support for intersex autonomy. *Human Rights Watch News*. June 17, 2019. Accessed March 15, 2020. <https://www.hrw.org/news/2019/06/17/film-shows-surge-support-intersex-autonomy>
13. Zieselman K. *XOXY: A Memoir (Intersex Women, Mother, Activist)*. Jessica Kingsley Publishers; 2020.
14. OHCHR; UN Women; UNAIDS, et al. Eliminating forced, coercive and otherwise involuntary sterilization: an interagency statement. World Health Organization; 2014. Accessed March 15, 2020. http://www.unaids.org/sites/default/files/media_asset/201405_sterilization_en.pdf
15. Garland J, Slokenberga S. Protecting the rights of children with intersex conditions from nonconsensual gender-conforming medical interventions: the view from Europe. *Med Law Rev*. 2019;27(3):482-508.
16. Toler J; GLMA Policy and Government Affairs Committee. Medical and surgical intervention of patients with differences in sex development. GLMA. October 3, 2016. Accessed February 11, 2020. <http://glma.org/index.cfm?fuseaction=document.viewdocument&ID=CEB9FEE4>

B8DD8B7F4F7575376BD476C3A433379DD853BEA17913AFCCB8270299C0
731320B03D2F5E1022F1C15602FBEA

17. Unnecessary surgery on intersex children must stop. News release. Physicians for Human Rights; October 10, 2017. Accessed March 15, 2020.
<http://physiciansforhumanrights.org/press/press-releases/intersex-surgery-must-stop.html>
18. Elders MJ, Satcher D, Carmona R. Re-thinking genital surgeries on intersex infants. Palm Center; June 2017. Accessed May 11, 2021.
<https://www.palmcenter.org/wp-content/uploads/2017/06/Re-Thinking-Genital-Surgeries-1.pdf>
19. S Con Res 110, 2017-2018 Reg Sess (Ca 2018). Accessed June 17, 2021.
https://leginfo.ca.gov/faces/billTextClient.xhtml?bill_id=201720180SCR110
20. Gardner M, Sandberg DE. Navigating surgical decision making in disorders of sex development (DSD). *Front Pediatr*. 2018;6:339.
21. Hughes IA, Houk C, Ahmed SF, Lee PA; Lawson Wilkins Pediatric Endocrine Society/European Society for Paediatric Endocrinology Consensus Group. Consensus statement on management of intersex disorders. *J Pediatr Urol*. 2006;2(3):148-162.
22. Bougnères P, Bouvattier C, Cartigny M, Michala L. Deferring surgical treatment of ambiguous genitalia into adolescence in girls with 21-hydroxylase deficiency: a feasibility study. *Int J Pediatr Endocrinol*. 2017;2017:3.
23. Anderson S. Disorders of sexual differentiation: ethical considerations surrounding early cosmetic genital surgery. *Pediatr Nurs*. 2015;41(4):176-186.
24. Lee PA, Houk CP. Key discussions from the Working Party on Disorders of Sex Development (DSD) evaluation, Foundation Merieux, Annecy, France, March 14-17, 2012. *Int J Pediatr Endocrinol*. 2013;2013(1):12.
25. Kreukels BPC, Cohen-Kettenis PT, Roehle R, et al. Sexuality in adults with differences/disorders of sex development (DSD): findings from the DSD-LIFE Study. *J Sex Marital Ther*. 2019;45(8):688-705.
26. Kreukels BPC, Köhler B, Nordenström A, et al; DSD-LIFE Group. Gender dysphoria and gender change in disorders of sex development/intersex conditions: results from the DSD-LIFE Study. *J Sex Med*. 2018;15(5):777-785.
27. Borg HW. The evidence-based transformation of American medicine. *J Am Physicians Surg*. 2016;21(3):70-73.
28. Enkin MW, Jadad AR. Using anecdotal information in evidence-based health care: heresy or necessity? *Ann Oncol*. 1998;9(9):963-966.
29. Arrieta Valero I. Autonomies in interaction: dimensions of patient autonomy and non-adherence to treatment. *Front Psychol*. 2019;10:1857.
30. Timmermans S, Yang A, Gardner M, et al. Does patient-centered care change genital surgery decisions? The strategic use of clinical uncertainty in disorders of sex development clinics. *J Health Soc Behav*. 2018;59(4):520-535.
31. Luthra S. Boston Children's Hospital will no longer perform two types of intersex surgery on children. *USA Today*. October 22, 2020. Accessed Feb 12, 2021.
<https://www.usatoday.com/story/news/health/2020/10/22/intersex-surgery-boston-childrens-hospitals-decision-watershed-moment-rights/3721096001/>
32. Neus N. Major children's hospital apologizes for performing cosmetic genital surgeries on intersex infants. *CNN*. July 29, 2020. Accessed February 13, 2020.
<https://www.cnn.com/2020/07/29/health/intersex-surgeries-chicago-hospital/index.html>

33. Nora N. Ban on cosmetic surgeries on intersex infants fails key California vote. CNN. January 14, 2020. Accessed March 15, 2020. <https://www.cnn.com/2020/01/13/us/california-cosmetic-surgeries-intersex-infants-senate-vote/index.html>
34. Hoylman B. Senator Brad Hoylman announces new legislation to protect the rights of intersex New Yorkers. News release. New York State Senate; November 08, 2019. Accessed March 15, 2020. <https://www.nysenate.gov/newsroom/press-releases/brad-hoylman/senator-brad-hoylman-announces-new-legislation-protect-rights>

Nat Mulkey, MD received their MD from Boston University School of Medicine in Massachusetts and works closely with faculty on LGBTQI+ curricula in undergraduate medical education and on various LGBTQI+ advocacy initiatives. They participated in direct advocacy that contributed to the Massachusetts Medical Society issuing supportive intersex policy recommendations.

Carl G. Streed Jr, MD, MPH is an assistant professor at the Boston University School of Medicine and research lead in the Center for Transgender Medicine and Surgery at Boston Medical Center in Massachusetts. As a clinician-investigator, his research focuses on the health and well-being of sexual and gender minority populations, particularly transgender persons. He has served nationally as the chair of the Advisory Committee on Lesbian, Gay, Bisexual, Transgender, Queer Issues for the American Medical Association.

Barbara M. Chubak, MD is an assistant professor of urology at the Icahn School of Medicine at Mount Sinai in New York City, specializing in the treatment of sexual dysfunction for all patients, regardless of sex or gender. She has MA degrees in the history of medicine and bioethics, which inform her interdisciplinary research focused on improving physicians' understanding of controversial disease concepts and the patient centeredness of genital health outcomes assessment.

Citation

AMA J Ethics. 2021;23(7):E550-556.

DOI

10.1001/amajethics.2021.550.

Conflict of Interest Disclosure

The author(s) had no conflicts of interest to disclose.

The viewpoints expressed in this article are those of the author(s) and do not necessarily reflect the views and policies of the AMA.