IN THE COURT OF APPEALS OF THE STATE OF OREGON

In the Matter of: JONES DAVID HOLLISTER, Petitioner-Appellant.	CA No. A171609 Lane County Circuit Case No. 19CV20980

BRIEF OF AMICUS CURIAE TRANSGENDER LAW CENTER, INTERACT, AND BEYOND BINARY LEGAL

Appeal from the Order and General Judgment of the Circuit Court of Lane Count

The Honorable Charles D. Carlson, Circuit Court Judge

John C. Clarke, OSB No. 153245
john.clarke@millernash.com
Bruce L. Campbell, OSB No. 925377
bruce.campbell@millernash.com
MILLER NASH GRAHAM & DUNN LLP
3400 U.S. Bancorp Tower
111 S.W. Fifth Avenue
Portland, Oregon 97204
Telephone: 503.224.5858

Attorneys for Amicus Curiae Transgender Law Center, interACT, and Beyond Binary Legal

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INTEREST OF AMICUS CURIAE

Appellant Jones David Hollister's appeal raises issues central to amici's missions.

Transgender Law Center is the largest national trans-led organization advocating self-determination for all people. Grounded in legal expertise and committed to racial justice, Transgender Law Center employs a variety of community-driven strategies to keep transgender and gender-nonconforming people alive, thriving, and fighting for liberation.

interACT: Advocates for Intersex Youth is a nonprofit organization that employs legal and policy advocacy to protect the rights of children born with variations in their sex characteristics, often called "intersex." interACT is the largest and oldest continuously operating organization in the country that is exclusively dedicated to this purpose. Founded in 2006 as Advocates for Informed Choice, interACT's mission is to end harmful, nonconsensual medical interventions on intersex children. Since its inception, interACT has continued to work to protect youth populations from harmful procedures such as clitoral reductions and sterilizations, while expanding its scope

of work to include raising awareness to end the shame and stigma faced by intersex youth. In addition, interACT oversees the largest cohort of intersex young people advocating on their own behalf, interACT Youth.

Beyond Binary Legal is an organization that seeks to uplift everyone who does not identify exclusively as a woman or a man, using law as a tool for survival and restorative change. Beyond Binary Legal provides legal services and other support by and for nonbinary people as well as information for allies. Throughout all of its work, Beyond Binary Legal values nonbinary people as experts of their own experiences, and seeks to bridge gaps in communication and resources.

SUMMARY OF ARGUMENT

This court should reverse the circuit court's decision to deny Appellant Jones David Hollister's request for a change of legal sex designation to nonbinary.

The circuit court believed that sex is a choice exclusively between male and female and distinct from gender. But that interpretation is inconsistent with the law and the reality of what it means to be transgender, nonbinary, intersex, or gender-nonconforming. Rather than grounding its decision in medical,

scientific, and academic literature, the circuit court relied on an unsupported notion of "tradition" when making this sex/gender distinction and limiting Appellant to one of two binary categories. Yet, cultural traditions around the world and throughout history have long recognized nonbinary people as integral parts of their communities.

Moreover, medical, scientific, and academic communities have long recognized intersex variations, or naturally occurring differences in physical sex traits that may cause people not to fit typical definitions of "male" or "female"; these communities have also recognized that self-attested gender identity is the most reliable determinant of an individual's sex when other traits seem to conflict. If self-attestation is definitive in the case of people with intersex variations, there is no reason why it should not be so for all individuals.

In addition, other states have granted changes to nonbinary designations under statutes similar to Oregon's statute, including when nonbinary designation was not a stated option. The circuit court's assertion that Oregon's statute applies only to people who medically transition is dangerous and unsupported. Accordingly, amici urge this

court to reverse the circuit court's order and general judgment and remand the matter for a correct application of the law.

ARGUMENT

I. Sex Traits Vary Beyond the Stereotypical Male/Female Binary

The circuit court was unpersuaded by cases involving nonbinary people who are intersex, suggesting that this case might come out differently if there were evidence in the record showing Appellant is intersex. Apart from imposing an inappropriate burden that would invade Appellant's privacy, the circuit court's rationale misses the mark. Intersex lives demonstrate that the circuit court's binary notions of sex and gender are simply wrong. Because self-attestation is definitive of gender regardless of individual sex traits, the circuit court should be reversed.

A. Background on Intersex People

"Intersex" is an umbrella term describing a wide range of natural variations of physical traits—in external genitals, internal sex organs, chromosomes, and hormones—that do not fit typical binary

notions of "male" and "female" bodies. Each year, as many as 2 percent of all babies are born with these variations.[1]

In general, intersex traits originate from variations in the embryonic sexual development process. A fertilized egg typically has two sex chromosomes: XX or XY. For the first few weeks of gestation, XX and XY embryos look the same, but they later develop differently depending on genetic and hormonal factors. Along one sex development path, the gonads become testes; the genital tubercle becomes a penis; and the labioscrotal folds fuse and form a scrotum. Along another, the gonads become ovaries; the genital tubercle becomes a clitoris; and the labioscrotal folds develop into the outer labia. Later, at puberty, hormones cause expression of secondary sex characteristics, such as breast development, body hair, musculature, and depth of voice.[2]

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[1] Anne Fausto-Sterling, *Sexing the Body: Gender Politics and the Construction of Sexuality* 51 (2000); Melanie Blackless et al., *How Sexually Dimorphic Are We? Review and Synthesis*, 12 Am J Hum Biol 151 (2000).

[2] I. A. Hughes et al., *Consensus Statement on Management of Intersex Disorders*, 118 Pediatrics 488, 491 (2006); Bruce E. Wilson & William G. Reiner, *Management of Intersex: A Shifting Paradigm in Intersex in the Age of Ethics* 119 (1999); SRY Gene, National Institutes of Health, https://ghr.nlm.nih.gov/gene/SRY (last visited Oct. 22, 2019).

This "typical" sex development process, however, can vary in many ways.[3] The variations present at different ages. For example, external genitalia that appear noticeably different from stereotypical expectations may mean that a child is deemed intersex at birth, but other times, genital differences or differences in secondary sex characteristics do not develop until later. In addition, variations in internal organs or sex chromosomes may not be discovered until puberty, an attempt to conceive children, or may never be discovered. At least 20 different intersex traits exist, the most common of which include Congenital Adrenal Hyperplasia ("CAH"), hypospadias, and Androgen Insensitivity Syndrome ("AIS").

Intersex children—like other children—are typically "assigned" a binary (male or female) sex at birth based on some combination of their genitalia, gonads and other internal organs, and chromosomes.[4] For the most part, this is a subjective process, and

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[3] Hughes, *supra* note 2, at 488; Laura Hermer, *Paradigms Revised: Intersex Children, Bioethics & the Law*, 11 Ann Health L 195, 204 (2002); Carla Murphy et al., *Ambiguous Genitalia in the Newborn: An Overview and Teaching Tool*, 24 J Pediatric Adolescent Gynecology 236, 236-37 (2011).

[4] Hughes, *supra* note 2, at 491.

experts may disagree on the "correct" sex to assign to an intersex child.[5] Unlike other children, intersex infants and children are often subjected to harmful, nonconsensual "normalizing" surgical procedures to erase their intersex differences—a severe violation of their bodily integrity and autonomy.

B. Intersex and Other Communities Face Harm Based on Their Differences

Since the 1960s, intersex children have faced this paradigm of nonconsensual surgical intervention, including the mutilation and removal of genital and gonadal tissue (e.g., clitoral reductions and orchiectomies).[6] These surgeries are typically performed not for any

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[5] *See, e.g.*, Anne Tamar-Mattis, *Report to the Inter-American Commission on Human Rights: Medical Treatment of People With Intersex Conditions as a Human Rights Violation*, Advocates for Informed Choice (Mar. 2013), https://goo.gl/Nf7Xt7, at 5 ("There is still controversy and uncertainty about gender assignment in [cases of partial AIS], and it can go either way, depending largely on the doctor's judgment."); David A. Diamond et al., *Gender Assignment for Newborns With 46XY Cloacal Exstrophy: A 6-Year Followup Survey of Pediatric Urologists*, 186 J Urol 1642, 1643 (2011) (reporting that only 79 percent of surveyed clinicians agreed on a male gender assignment in 46,XY cloacal exstrophy).

[6] Jeremy Toler, *Medical and Surgical Intervention of Patients with Differences in Sex Development* 1, Gay & Lesbian Med Ass'n (Oct. 3, 2016); Katrina Karkazis, *Fixing Sex: Intersex, Medical Authority, and Lived Experience* 57-58, 60-61 (2008); Martin Kaefer & Richard C. Rink, *Treatment of the Enlarged Clitoris*, Frontiers in Pediatrics (Aug. 2017);

valid medical reason, but for cosmetic purposes or to ease parents' or doctors' discomfort with the child's difference.[7] Medical professionals commonly perform these surgeries before the age of two, when the child is too young to understand what is taking place, let alone provide informed consent.[8]

The consequences of these surgeries are dire and permanent. The child may be left unable to have biological children; may suffer a lifelong diminution or loss of sexual sensation and function; and may experience scarring, incontinence, and chronic infections.[9] The long-

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Jennifer Yang et al., *Nerve Sparing Ventral Clitoroplasty: Analysis of Clitoral Sensitivity and Viability*, 178 J Urol 1598-1601 (Oct. 2007); Sarah Creighton et al., *Timing and Nature of Reconstructive Surgery for Disorders of Sex Development–Introduction*, J Pediatric Urol (2012).

[7] Toler, *supra* note 36, at 1; Tamar-Mattis, *supra* note 14, at 2-3, 9; Hermer, *supra* note 3, at 207.

[8] Karkazis, *supra* note 36, at 57–58; Tamar-Mattis, *supra* note 14, at 2; Daniela Truffer, *It's a Human Rights Issue! in Voices: Personal Stories from the Pages of Nib–Normalizing Intersex* 26-29 (James M. DuBois & Ana S. Iltis eds., 2016) (describing a gonadectomy performed at two months of age); Lily C. Wang & Dix P. Poppas, *Surgical Outcomes and Complications of Reconstructive Surgery in the Female Congenital Adrenal Hyperplasia Patient: What Every Endocrinologist Should Know*, J Steroid Biochem & Molecular Biol (2017); Natalie Nokoff et al., *Prospective Assessment of Cosmesis Before and After Genital Surgery*, 13 J Pediatric Urol 28.e1 to 28.e6 (2017).

[9] Toler, *supra* note 36, at 1; *Recommendations From interACT: Advocates for Intersex Youth Regarding the List of Issues for the United*

lasting emotional distress and trauma experienced by children subjected to these procedures can be comparable to that experienced by survivors of child rape or sexual abuse.[10] For all the harm that these surgeries entail, there is no persuasive evidence that they provide any benefit to the child when performed without individual consent.[11]

States for the 59th Session of the Committee Against Torture at 2, interACT (June 2016), https://tbinternet.ohchr.org/Treaties/CAT/Shared%20Documents/USA/INT_CAT_ICS_USA_24552_E.pdf; Tamar-Mattis, *supra* note 14, at 3-5; Peter Lee et al., *Review of Recent Outcome Data of Disorders of Sex Development (DSD): Emphasis on Surgical and Sexual Outcomes*, 8 J Pediatric Urol 611 (Dec. 2012); Sarah Creighton et al., *Objective Cosmetic and Anatomical Outcomes at Adolescence of Feminising Surgery for Ambiguous Genitalia Done in Childhood*, 358 Lancet 124 (2001); *"I Want to Be Like Nature Made Me": Medically Unnecessary Surgeries on Intersex Children in the U.S.* 58, Human Rights Watch & interACT (2017), https://bit.ly/2Y1N6DZ.

[10] *A Human Rights Investigation Into the Medical "Normalization" of Intersex People* 17-18, S.F. Human Rights Comm'n (2005), https://goo.gl/trBnGT; Tamara Alexander, *The Medical Management of Intersexed Children: An Analogue for Childhood Sexual Abuse*, ISNA (1997), https://goo.gl/fy9jae; Karsten Schützmann et al., *Psychological Distress, Self-Harming Behavior, and Suicidal Tendencies in Adults With Disorders of Sex Development*, Arch Sex Behav (2007).

[11] Sarah Creighton et al., *Timing and Nature of Reconstructive Surgery for Disorders of Sex Development—Introduction*, 8 J Pediatric Urol 602 (2012); Hughes, *supra* note 2, at 493; S.F. Human Rights Comm'n, *supra* note 40, at 19; Toler, *supra* note 36, at 1; Tamar-Mattis, *supra* note 14, at 3.

Today, these surgeries are widely condemned by intersex communities, and human-rights groups including the United Nations, the World Health Organization, and Amnesty International decry them.[12] Fortunately, parents are increasingly choosing to forgo invasive, medically unnecessary procedures until the child can consent. Nonetheless, families across the United States continue to report that unnecessary genital surgery has been encouraged by medical professionals as a "solution" to address differences in sex characteristics.[13]

The mistreatment of intersex people does not end with childhood surgery. Physicians who are unfamiliar with or who stigmatize intersex variations may deny intersex people medical

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[12] Juan E. Méndez, *Report of the Special Rapporteur on Torture and Other Cruel, Inhuman or Degrading Treatment or Punishment* ¶ 77, UN Doc. A/HRC/22/53 (Feb. 1, 2013); Toler, *supra* note 36, at 1; *Eliminating Forced, Coercive and Otherwise Involuntary Sterilization: An Interagency Statement*, OHCHR, UN Women, UNAIDS, UNDP, UNFPA, UNICEF & WHO (2014), https://goo.gl/nzXm6f; *Policy Statement on the Rights of Intersex Individuals*, Amnesty International (2013); *Recommendations From interACT*, *supra* note 39, at 1; Tamar-Mattis, supra note 14, at 7-9.

[13] Toler, *supra* note 36, at 1; Eric Lohman & Stephani Lohman, *Raising Rosie: Our Story of Parenting an Intersex Child* (UBCPress 2018).

treatment in adulthood,[14] and some intersex people report trauma and fear of doctors due to earlier mistreatment that leaves them unable to obtain necessary medical care.[15]

Intersex people also experience discrimination in education, public services, sports, and employment.[16] These struggles are shared with transgender and nonbinary communities as well, who also experience mistreatment in these realms related to societal ignorance or fear of any perceived sex or gender differences. Greater awareness of these communities' existence, which can be promoted by legal recognition, can help protect transgender, nonbinary, and intersex

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[14] Tamar-Mattis, *supra* note 14, at 2, 7; *Fact Sheet: Intersex* at 2, Free & Equal: UN for LGBT Equality (2015), https://www.unfe.org/system/unfe-65-Intersex_Factsheet_ENGLISH.pdf.

[15] S.F. Human Rights Comm'n, *supra* note 40, at 23; Tamar-Mattis, *supra* note 14, at 12; Davis, *supra* note 7, at 109-10 (quoting an intersex adult: "I don't like doctors. I don't go to the doctor very often. I don't trust doctors. That's a very triggering environment for me.").

[16] *Fact Sheet: Intersex*, *supra* note 44, at 1; United Nations Office of the High Commissioner for Human Rights, *Tackling Discrimination Against Lesbian, Gay, Bi, Trans, & Intersex People: Standards of Conduct for Business* (2017), https://www.unfe.org/wp-content/uploads/2017/09/UN-Standards-of-Conduct.pdf.

individuals' ability to participate in society free of discrimination or violence.

C. Gender Identity is the Primary Determinant of Sex

As set forth above, the circuit court's interpretation of sex—as a choice exclusively between male and female and different from gender identity—does not account for the multifaceted nature of sex. The evidence shows that gender identity is a deep-seated, persistent trait that is fundamental to a person's sense of self, evidenced by how they live their life every day.

For the estimated 1.7 percent of people with intersex traits, the anatomical and physiological factors that encompass what is traditionally thought of as "sex," differ from the presumptive male/female binary and often point in different directions. Such cases illustrate the inadequacy of the circuit court's interpretation of sex.

For example, an individual with complete AIS may have typically female external genitalia and secondary sex characteristics, such as breasts, with internal testes and an XY chromosome pattern. Another with CAH may have masculinized external genitalia and secondary sex characteristics, such as a deep voice and body hair, with

ovaries, a uterus, and an XX chromosomal pattern. A third may have external genitalia that do not appear either typically "male" or typically "female," internal ovotestes, and mosaicism (i.e., sex chromosomes that differ from cell to cell). For each of these individuals, what is their sex under the circuit court's interpretation of sex, which limits designation to male or female and does not rely on the individual's self-reported gender?

The circuit court advanced a misconception of sex that is contrary to the scientific consensus, as recognized by courts in other jurisdictions.[17] A definition of sex that encompasses an individual's identity and lived reality would avoid arbitrary designations in the instances discussed above as well as the arbitrary denial of Appellant's effort to change their legal sex designation to nonbinary. It would also comport with medical practice. Physicians agree that the goal in

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[17] *See, e.g.*, *Schroer v. Billington*, 424 F Supp 2d 203, 211-13 (DDC 2006) (scientific observation confirms that "sex is not a cut-and-dried matter of chromosomes" but rather consists of "different components") (internal quotation marks and citation omitted); *In re Heilig*, 816 A2d 68, 73 (Md 2003) (gender is determined by seven factors); *Maffei v. Kolaeton Indus., Inc.*, 164 Misc 2d 547, 551-52, 626 NYS2d 391 (Sup Ct NY Cty 1995) (explaining that "at least seven variables * * * interact to determine the ultimate sex of an individual," including gender identity); *F.V. v. Barron*, 286 F Supp 3d 1131 (D Idaho 2018).

assigning a sex to an intersex child is to predict how the child will self-identify as an adult, and that when an intersex person's gender identity differs from the binary sex they were assigned as an infant, that identity should prevail.[18] While some intersex people continue to identify with their originally assigned sex throughout their lives, others do not.[19] Within most intersex variations, studies show that between 5 and 29 percent of intersex people do not identify with their originally assigned sex.[20] In some circumstances, the rate of sex assignment rejection can exceed 60 percent.[21]

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[18] Peter A. Lee et al., *Global Disorders of Sex Development Update Since 2006: Perceptions, Approach and Care*, Horm. Res. Paediatr. (2016), doi: 10.1159/000442975; *Clinical Guidelines for the Management of Disorders of Sexual Development in Childhood* 25-31 (2006), Consortium on the Management of Disorders of Sex Development, https://goo.gl/bKQcES.

[19] *Understanding Intersex and Transgender Communities* at 1, interACT, https://goo.gl/CY53ZZ.

[20] Julie A. Greenberg, *Intersexuality and the Law* 20 (2012); Hughes et al., *supra* note 2, at 491; P. S. Furtado et al., *Gender Dysphoria Associated With Disorders of Sex Development*, 9 Nat Rev Urol 620 (Nov. 2012) (reporting average rates of gender dysphoria at 5 percent for Complete Androgen Insensitivity Syndrome, 10 percent for CAH, 12.5 percent for Ovotesticular DSD, 20 percent for Partial Androgen Insensitivity Syndrome, and 29 percent for Mixed Gonadal Dysgenesis).

[21] Furtado et al., *supra* note 16 (reporting average rates of gender dysphoria at 57 percent for 17-beta-HSD3 deficiency and 63 percent for 5-alpha-RD2 deficiency).

In the case of intersex people, medicine's attempt to point to one unitary trait such as karyotype or genital configuration as evidence of an individual's "true" sex proved futile because none of these traits reliably predicted how an individual would identify as they reached adolescence and adulthood.[22] Thus, medical experts regard individual gender identity as the most important factor in determining an intersex individual's sex. If one's deeply felt identity and lived reality are relevant in the case of people who happen to be born with intersex traits, there is no reason why they should not be considered for all people who may have a gender identity that is different from what was presumed based on visible characteristics at birth.

As gender and biological fluidity show us, self-attested gender identity is the reliable predictor of how people will navigate sex categories in their lives. Legal recognition of their gender identity is essential to the ability of all people to enjoy rights and opportunities equal to those available to others, i.e., to have full rights to education,

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[22] The emphasis on which characteristic should prevail in determining a person's sex has changed over time. For a history of intersex management, *see generally* Elizabeth Reis, *Bodies in Doubt: An American History of Intersex* (2009).

employment, and health care; to travel; to navigate everyday transactions; and live in safety. Transgender, intersex, and gender-nonconforming people, regardless of how they identify or appear, should receive the legal recognition that all persons expect and deserve.[23]

II. The Law and Medicine Have Recognized Intersex and Nonbinary People for Millennia

The law and medicine have recognized intersex and nonbinary people for millennia. Indigenous communities recognize a number of traditional genders that are not strictly male or female. These include, among others, Native American Two-Spirit people,[24] the

[23] The World Professional Association for Transgender Health, Identity Recognition Statement (Nov. 15, 2017), https://www.wpath.org/media/cms/Documents/Web%20Transfer/Policies/WPATH%20Identity%20Recognition%20Statement%2011.15.17.pdf.

[24] Indian Health Service, Two Spirit, https://www.ihs.gov/lgbt/health/twospirit/ (accessed Nov. 4, 2019) ("Traditionally, Native American two spirit people were male, female, and sometimes intersexed individuals who combined activities of both men and women with traits unique to their status as two spirit people. In most tribes, they were considered neither men nor women; they occupied a distinct, alternative gender status.").

Hawai'ian Māhū and the Fa'afafine of Samoa, [25] and the Hijra in South Asia, recognized in India[26] and Pakistan's legal systems.[27]

Classical Jewish writings identify six sex categories—male, female, and four that would be recognized today as nonbinary or intersex. The Jewish Mishnah, Talmud, and legal codes mention these variations hundreds of times.[28] Greco-Roman culture also recognized intersex variations.[29] The Roman emperor Justinian permitted children

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[25] Eleanor Kleiber, University of Hawai'i at Mānoa Library, *Gender Identity and Sexual Identity in the Pacific and Hawai'i: Introduction* (Sept. 10, 2019), https://guides.library.manoa.hawaii.edu/c.php?g=105466&p=686754 (defining Māhū as "in Native Hawaiian culture this refers to an individual who may be considered third-gendered with characteristics of both sexes, usually a male to female. In contemporary Hawai'i the word is also used to describe people who are transgender, or gay"; and Fa'afafine as "in Samoan culture a third-gendered individual. A recognized and integral part of traditional Samoan culture, Fa'afafine, born biologically male, embody both male and female gender traits.").

[26] Francesca Trianni, Time Magazine, *Men, Women and 'Hijras': India Recognizes Third Gender* (April 15, 2014), https://time.com/63801/men-women-and-hijras-india-recognizes-third-gender/.

[27] Rabail Baig, Foreign Policy, *A First For Pakistan's Third Gender* (March 30, 2012), https://foreignpolicy.com/2012/03/30/a-first-for-pakistans-third-gender/.

[28] *More Than Just Male and Female: The Six Genders in Classical Judaism*, Sojourn Blog (June 1, 2015), https://goo.gl/5BsHzS; 1 Julia M. O'Brien, ed., *Oxford Encyclopedia of the Bible and Gender Studies* 311-12 (2014).

[29] Pliny, *Natural History* 7:3 (John Bostock trans., 1855),

with genitals that were not clearly male or female to choose their own sex category before marriage.[30]

Over the last few decades, intersex, transgender, and gender-nonconforming people have gained greater legal recognition. An increasing number of nations have adopted a third, neutral sex category on passports and other official documents. In the United States, states and municipalities are beginning to recognize a third and neutral sex category, including Oregon (in certain counties),[31] California,[32] the

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https://goo.gl/nHahlm (referring to "those who belong to both sexes, [whom] we call by the name of hermaphrodites * * * [or] Androgyni.") The term "hermaphrodite" is considered highly pejorative by the intersex community and should not be used outside historical reference or an intersex individual's own preference.

[30] 1 *Enactments of Justinian: The Digest or Pandects* tit. 5, ¶ 10 (S. P. Scott ed., 1932), https://bit.ly/2LecBPy; Michaela Koch, *Discursive Intersexions: Daring Bodies Between Myth, Medicine, and Memoir* 31 (2017).

[31] Benton County Circuit Court, Case No. 17CV01994; Multnomah County Circuit Court, Case No.16CV13991; Polk County Circuit Court, Case No. 16CV31356.

[32] Cal Health & Safety Code § 103425.

District of Columbia,[33] New Jersey,[34] Washington,[35] and several others.[36]

III. The Circuit Court Misinterpreted California's and Oregon's Standards

As noted above, other states have granted changes to nonbinary gender markers under statutes similar to Oregon's statute, including California.

The circuit court, however, misconstrued California's standard in denying Appellant's petition for a change of legal sex designation. The circuit court distinguished California and Oregon law on the ground that California law specifically recognizes gender changes to "nonbinary," whereas Oregon law does not. The circuit court was incorrect. As early as 2016, California followed Oregon's lead in granting petitions recognizing people as nonbinary. At that time, California's statute permitted a person to "file a petition with the

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[33] DC Code Ann § 50-1401.01.

[34] NJ Stat Ann § 26:8-40.12.

[35] Wash Admin Code 246-490-075.

[36] *Changing Birth Certificate Sex Designations: State-by-State Guidelines*, Lambda Legal, https://www.lambdalegal.org/know-your-rights/article/trans-changing-birth-certificate-sex-designations (last updated Sept. 17, 2018).

superior court in any county seeking a judgment recognizing the change of gender."[37] Like Oregon, California did not include limiting language, nor did it expressly define gender in terms of different choices (e.g., male, female, or nonbinary). Thus, California courts properly allowed "nonbinary" as an available choice, issuing orders for nonbinary gender changes.[38]

More recently, California took additional steps to address legislative and administrative challenges associated with gender marker changes. Beginning in 2017, California legislators introduced and passed the Gender Recognition Act (SB 179), effectively affirming trial court rulings that permitted "nonbinary" as an option for gender marker changes and official state documents. The law also reduced administrative burdens for gender marker changes. In other words, California streamlined the process for Californians to change their legal gender marker to nonbinary.

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[37] Cal Health & Safety Code § 103425.

[38] *See* Petition of Sara M. Keenan for Change of Name & Gender, Superior Court of California, Santa Cruz, Case No. 16CV02024.

Further, in misinterpreting the California and Oregon statutes, the circuit court applied outdated and incorrect conceptions of "sexual reassignment procedures and treatments," presuming that Oregon's statute is only for transgender men and women who are medically transitioning. In doing so, the circuit court misstated Oregon's standard, which does not refer to "sexual reassignment," but rather requires appropriate treatment "for the individual for the purpose of affirming gender identity."[39]

In addition, both California's prior standard[40] and Oregon's current standard[41] recognize that treatment affirming gender identity is individualized, and may or may not involve "sex reassignment," as stereotypically conceived. Rather, the Oregon standard recognizes, as does the World Professional Association for Transgender Health, that "Treatment is individualized: What helps one person alleviate gender dysphoria might be very different from what helps another person.

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[39] ORS 33.460(1).

[40] Former Cal Health & Safety Code § 103425(a) (2016) ("clinically appropriate treatment for the purpose of gender transition").

[41] ORS 33.460(1) ("undergone surgical, hormonal or other treatment appropriate for the individual for the purpose of affirming gender identity * * *.").

This process may or may not involve a change in gender expression or body modifications."[42] As California courts did under California's previous standard, Oregon courts should refuse to interpret medical language in gender change statutes from requiring any particular course of treatment, or from excluding nonbinary people from legal recognition.

Because California courts were already granting nonbinary gender changes before adopting the Gender Recognition Act, the circuit court's reliance on California law to distinguish it from Oregon law was erroneous.

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[42] The World Professional Association for Transgender Health, *Standards of Care for the Health of Transsexual, Transgender, and Gender-Nonconforming People*, Version 7 at 5 (2012), https://wpath.org/media/cms/Documents/SOC%20v7/Standards%20of%20Care_V7%20Full%20Book_English.pdf.

CONCLUSION

For the foregoing reasons and the reasons set forth in Appellant's brief, the court should reverse the circuit court's order and general judgment and should remand the matter for a correct application of ORS 33.460.

DATED this 8th day of November, 2019.

MILLER NASH GRAHAM & DUNN LLP

s/ John C. Clarke

John C. Clarke, OSB No. 153245
john.clarke@millernash.com
Bruce L. Campbell, OSB No. 925377
bruce.campbell@millernash.com

Attorneys for Amicus Curiae Transgender Law Center, interACT, and Beyond Binary Legal

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s/ John C. Clarke

John C. Clarke, OSB No. 153245
john.clarke@millernash.com
Bruce L. Campbell, OSB No. 925377
bruce.campbell@millernash.com

Attorneys for Amicus Curiae
Transgender Law Center, interACT,
and Beyond Binary Legal

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How Sexually Dimorphic Are We? Review and Synthesis

MELANIE BLACKLESS, ANTHONY CHARUVASTRA,
AMANDA DERRYCK, ANNE FAUSTO-STERLING,*
KARL LAUZANNE, AND ELLEN LEE
Department of Molecular and Cell Biology and Biochemistry, Brown University, Providence, Rhode Island

ABSTRACT The belief that *Homo sapiens* is absolutely dimorphic with the respect to sex chromosome composition, gonadal structure, hormone levels, and the structure of the internal genital duct systems and external genitalia, derives from the platonic ideal that for each sex there is a single, universally correct developmental pathway and outcome. We surveyed the medical literature from 1955 to the present for studies of the frequency of deviation from the ideal male or female. We conclude that this frequency may be as high as 2% of live births. The frequency of individuals receiving "corrective" genital surgery, however, probably runs between 1 and 2 per 1,000 live births (0.1–0.2%). Am. J. Hum. Biol. 12:151–166, 2000.

Among primates, humans exhibit a modest sexual dimorphism with regard to characters such as body size or voice timbre (Fedigan, 1982). With respect to sex chromosome composition, gonadal structure, hormone levels, and the structure of the internal genital duct systems and external genitalia, however, we generally consider *Homo sapiens* to be absolutely dimorphic. Biologists and medical scientists recognize, of course, that absolute dimorphism is a Platonic ideal not actually achieved in the natural world. Nonetheless, the normative nature of medical science uses as an assumption, the proposition that for each sex there is a single, correct developmental pathway. Medical scientists, therefore, define as abnormal any deviation from bimodally distributed genitalia or chromosomal composition (Conte and Grumbach, 1989). If, however, one relinquishes an a priori belief in complete genital dimorphism, one can examine sexual development with an eye toward variability rather than bimodality.

Conte and Grumbach (1989) list more than 25 diagnoses affecting sexual differentiation. While the incidence of some individual medical syndromes is fairly well established, the overall frequency of intersexuality is a matter of dispute. Fausto-Sterling (1993a,b) cited a figure attributed to John Money that the frequency of intersexuality might be as high as 4% of live births, but Money (1993) responded that he never made such a claim. In fact, no well-documented overview of the frequency of intersex exists at present, and it is this lacuna that we address in the present article. The question is of interest to students of human development, medical practitioners, and human biologists, among others. Recently, the practice of surgically altering the genitals of intersexual infants to conform to assumptions about absolute dimorphism has been questioned (Fausto-Sterling, 1995/1996; Post, 1995/1996; Sandberg, 1995/1996; Walcutt, 1995/1996; Diamond, 1996; Zucker, 1996; Diamond and Sigmundsen, 1997; Kessler, 1998). Thus, both because of a theoretical interest in human sexual dimorphism and medical questions about the treatment of intersexuals, it is important to provide a frequency baseline for the varied events which lead to intersexuality.

METHODS

We surveyed the medical literature from 1955 to the present for studies of the fre-

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*Correspondence to: Anne Fausto-Sterling, Department of Molecular and Cell Biology and Biochemistry, Brown University, Providence, RI 02912.
E-mail: Anne_Fausto-Sterling@brown.edu

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quency of deviation from the ideal male or female. For a few rare syndromes, we considered the literature which predates 1955. Our sources included population surveys, genetic studies, case surveys from individual medical practitioners, and environmental population studies. In addition to Medline as a starting point, government documents, bibliographies in textbooks, previously located review articles, and specific articles provided additional sources. We did not exclude any articles which contained frequency estimates derived from an unselected population. Rather, where appropriate we indicate the limitations of particular publications.

We define the typical male as someone with an XY chromosomal composition, and testes located within the scrotal sac. The testes produce sperm which, via the vas deferens, may be transported to the urethra and ejaculated outside the body. Penis length at birth ranges from 2.5 to 4.5 cm (Flatau et al., 1975); an idealized penis has a completely enclosed urethra which opens at the tip of the glans. During fetal development, the testes produce the Mullerian inhibiting factor, testosterone, and dihydrotestosterone, while juvenile testicular activity ensures a masculinizing puberty. The typical female has two X chromosomes, functional ovaries which ensure a feminizing puberty, oviducts connecting to a uterus, cervix and vaginal canal, inner and outer vaginal lips, and a clitoris, which at birth ranges in size from 0.20 to 0.85 cm (Oberfield et al., 1989). In this article, we ask how often development meets these exacting criteria for males and females.

The literature which reports the frequencies of syndromes that produce intersexuality varies in quality and quantity. In some cases there are multiple surveys with large numbers replicated over many years and in many different geographical locations. In others no data exist with which to estimate frequency, while in still others the lack of better data dictated reliance on one or a small number of reports of uncertain quality. In each case the available data are presented. The strength or weakness of an estimate is also indicated.

"SEX" CHROMOSOME COMPOSITION

Individuals with XXY, XO, XYY, XXYY, XX males, and 47,XXX females comprise the most frequently encountered deviations from an XX (female) or XY (male) chromosomal make-up. An XO condition produces individuals with female external genitalia and streak gonads which are incapable of fetal or pubertal gonadal hormone synthesis and a variety of somatic alterations, while 47,XXX girls develop secondary sex characteristics at puberty and are sometimes fertile (Buckton, 1983). XXY individuals diagnosed with Klinefelter syndrome have external male genitalia, small testes, impaired spermatogenesis, and frequent gynecomastia. XXYY individuals are considered karyotypic variants of Klinefelter syndrome (Conte and Grumbach, 1989; Zinn et al., 1993). XYY males are taller, on average, than XY males, and commonly exhibit underdeveloped testes (Vogel and Motulsky, 1979). Recent work, however, suggests that many 47,XXY and 47,XYY males are undiagnosed because they present no symptoms which prompt a chromosomal analysis (Abramsky and Chapple, 1997). The category of XX males, which involves the translocation or deletion of a submicroscopic section of the sex determining region of the Y chromosome, is morphologically and genetically heterogeneous (López et al., 1995).

Table 1 summarizes the results of 17 studies of the frequencies of XXYY, XX (male), 47,XXX, and XYY individuals at birth. The total frequency ranges from 0.002 to 2.15/1,000 live births, with a mean of 0.639/1,000 and a standard deviation of 0.665. The mean/1,000 live births and standard deviations for each of the non-XX, non-XY chromosome compositions listed in Table 1 are 0.155 (0.185) for XXYY, 0.05 (0.019) for XX males, 0.47 (0.364) for 47, XXX, and 0.639 (0.665) for XYY.

In 24 different estimates of the frequency of Klinefelter syndrome (XXY), the incidence ranges from 0 (out of 3,890 births) to 2.13/1,000 births. The mean incidence for all 24 studies is 0.922/1,000 live births with a standard error of 0.102 (Table 2). The 18 different estimates for the population frequency of XO chromosome constitution are shown in Table 3. The incidence ranges from 0.0 for five small studies (sample size of less than 3,993) to 1.67/1,000 live births. The mean is 0.369/1,000 live births, with a standard error of 0.111. Recent data showing that not all Turner patients present with XO identifiable by traditional karyology suggest that the incidence calculated

*TABLE 1. Incidence of XXYY, XX(male), 47,XXX and XYY births in 17 published surveys**

Location	Year	Total # surveyed	XXYY	XX male	47,XXX	XYY	Incidence/1,000 live births	Method	Reference
Edinburgh	1964	20,725	1		11		0.575	Bs[a]	Maclean et al., 1964
Geneva	1968	8,184			1		0.122	Bs	Mikamo, 1968
London	1969	2,081				4	1.922	K	Sergovich et al., 1969
New Haven	1970	4,366			3	3	1.374	K	Lubs, 1970
Toronto	1974	73,229	1	2	2		0.068	Bs/K	Bell, 1974
Moscow	1974	2,500			1		0.400	K	Bochkov et al., 1974
Winnipeg	1975	13,939			5	7[b]	0.861	K	Hamerton et al., 1975
Ontario	1976	930				2	2.150	K	Lin et al., 1976
Denver	1976	40,371			12		0.297	Bs/K	Goad et al., 1976
USA	1977	13,751		1			0.073	K	Walzer et al., 1977
Tokyo	1978	12,319				3	0.244	K	Higurasi et al., 1979
Edinburgh	1979	23,196		1	11		0.517	Bs/K	Ratcliffe et al., 1979
Edinburgh	1980	3,993			3	4	0.002	K	Buckton et al., 1980
USA	1982	19,675			25		0.001	K	Schreinemacher et al., 1982
Telemark	1982	1,830				1	0.546	K	Hansteen et al., 1982
Belgium	1988	77,000	32[c]				0.416	K	Kleczkowska et al., 1988
Denmark	1991	34,910	5[d]	2	18	20[b]	1.289	K	Nielsen, 1991
Average total/1,000 live births (SD)			0.155 (0.185)	0.05 (0.019)	0.470 (0.364)	0.865 (0.740)	0.639 (0.665)		

*K, karyotype. B_5, buccal smear.
[a]Excludes perinatal deaths.
[b]Includes mosaic and nonmosaic.
[c]Includes 25 people with more than two X chromosomes.
[d]Includes four xx/xy mosaics.

from previously published studies may be an underestimate (Zinn et al., 1993).

Androgen insensitivity in XY individuals

Disruption of fetal hormonal metabolism in XY fetuses results most commonly from defects in androgen receptors. Clinical features range from a fully female external phenotype, with a blind-ending vagina and little axillary hair development, to a masculine phenotype with azoospermia and elevated levels of luteinizing hormone (Griffin and Wilson, 1989). Estimates of the frequency of complete androgen insensitivity (AIS) range from 0.049 to 0.016/1,000 male (Bangsbøll et al., 1992; Griffin and Wilson, 1989). In addition, 1–2% of girls with inguinal hernias may have androgen insensitivity (Griffin and Wilson, 1989). Jagiello and Atwell (1962) estimate the frequency of inguinal hernias in girls at 8/1,000 female births. Hence, the frequency of complete AIS may be 0.12/1,000 female births. Averaging the estimates for male births and combining them with the estimate for female births yields a figure of 0.076/1,000 live births. There are no solid estimates of the frequency of partial AIS, but Griffin and Wilson (1989) suggest that it is one-tenth as common as complete AIS. Using these estimates the rate would be 0.0076/1,000 live births.

Griffin and Wilson (1989) cite three other forms of AIS: 5α-reductase deficiency, Reifenstein syndrome and Infertile Male syndrome. The first two produce externally visible intersexuality, while those with Infertile Male syndrome are phenotypically male. 5-α-Reductase deficiency is quite common in a number of populations, ranging from Central America to Vietnam (Mendoca et al., 1996). Indeed, more than 50 families with over 100 affected individuals have been reported. However, no population or gene frequencies are available (Conte and Grumbach, 1989; Mendoca et al., 1996; Al-

*TABLE 2. Incidence of XXY births in 24 published surveys**

Location	Year	Total # surveyed	Total XXY[a]	Incidence/1,000 live births	Method	Reference
Winnipeg	1959	3,715	5	0.135	Bs	Moore, 1959
Bombay, India	1962	3,890	0	0	?	Subray and Prabhaker, 1962
Seattle[b]	1964	1,954	2	1.02	?	Paulsen et al., 1964
Edinburgh	1964	20,725	20	0.97	?	Maclean et al., 1964
Geneva	1968	8,184	6	0.73	?	Mikamo, 1968
London, England	1969	2,081	1	0.481	K	Sergovich et al., 1969
USA	1970	3,543	5	1.411	K	Gerald, 1970
New Haven	1970	4,366	4	0.916	K	Lubs, 1970
Germany	1973	1,000	2	2.00	?	Golob, 1973
Dehli	1973	3,100	3	0.97	?	Verma et al., 1973
Toronto	1974	73,229	43	0.59	?	Bell, 1974
Moscow	1974	2,500	2	0.800	K	Bochkov et al., 1974
Winnipeg	1975	13,939	6	0.430	K	Hamerton et al., 1975
Ontario	1976	930	1	1.075	K	Lin et al., 1976
Denver	1976	40,371	23	0.57	?	Goad et al., 1976
USA	1977	13,751	10	0.727	K	Walzer, 1977
Edinburgh	1979	23,196	21	0.91	?	Ratcliffe et al., 1979
Tokyo	1979	12,319	2	0.162	K	Higurasi et al., 1979
Edinburgh	1980	3,993	6	1.503	K	Buckton et al., 1980
Telemark	1982	1,830	1	0.546	K	Hansteen et al., 1982
Northeast (USA)	1982	19,675	20	1.017	K	Schreinemacher et al., 1982
Germany	1984	13,168	28	2.126	K	Murken, 1984
Denmark	1991	34,910	27	0.773	K	Nielsen, 1991

*?, Not specified; K, karyotype; Bs, buccal smear.
[a]Mosaic and nonmosaic.
[b]Study reported only male births; we doubled the denominator to estimate total births.

*TABLE 3. Incidence of XO births in 18 published surveys**

Location	Year	Total # surveyed	Total[a]	Incidence/1,000 live births	Method	Reference
Edinburgh	1964	20,725	4	0.400	Bs	Maclean et al., 1964
Geneva	1968	8,184	2	0.509	Bs	Mikamo, 1968
London	1969	2,081	0	0	K	Sergovich et al., 1969
?	1970	3,543	0	0	K	Gerald, 1970
New Haven	1970	4,366	1	0.458	K	Lubs, 1970
Dehli	1973	3,100	1	0.644	Bs	Verma et al., 1973
Toronto	1974	73,229	5	0.141	Bs	Bell, 1974
Moscow	1974	2,500	2	1.671	K	Bochkov et al., 1974
Winnipeg	1975	13,939	2	0.296	K	Hamerton et al., 1975
Denver	1976	40,371	27	1.370	Bs	Goad et al., 1976
Ontario	1976	930	0	0	K	Lin et al., 1976
Edinburgh	1979	23,196	1	0.105	Bs	Ratcliffe et al., 1979
Tokyo	1979	12,319	1	0.168	K	Higurasi et al., 1979
Edinburgh	1980	3,993	0	0	K	Buckton et al., 1980
Telemark	1982	1,830	0	0	K	Hansteen et al., 1982
New York State	1983	76,474	9	0.118	K	Hook, 1983
Denmark	1991	34,910	8	0.229	K	Nielsen, 1991

[a]Mosaic and nonmosaic.
*Bs, buccal smear; K, karyotype.

Attia, 1996). Similarly, no such estimates exist for Reifenstein or Infertile Male syndrome. Aiman and Griffin (1982) report evidence of androgen resistance in more than 40% of men with no other obvious cause for severe oligo- or azoospermia.

Congenital adrenal hyperplasia

The most common cause of intersexuality in XX females is congenital adrenal hyperplasia (CAH), a label which covers a heterogeneous set of genetically inherited alterations in steroid biosynthesis. The work on CAH has been thoroughly reviewed (Laue and Rennert, 1995; New et al., 1989; Newfield and New, 1997; Pang and Clark, 1990, 1993). Table 4 presents a summary. Although not all forms of CAH result in ambiguity at birth, the table includes estimates for any manifestations which alter sexually dimorphic presentation at any time during the life cycle. The classic form of 21-

TABLE 4. Incidence of 21-OHase (classic) in 36 published surveys

Location and subpopulation	Sample size	Date	Frequency/1,000 live births	Reference
Part A: Case surveys				
Switzerland	1,516,299	1980	0.0646	Werder et al., 1980
France	41,073,409	1985	0.031	Bois et al., 1985
France	13,921,803	1985	0.0356	Bois et al., 1985
Switzerland		1988	0.198	Pang et al., 1988
Canada		1988	0.038	Pang et al., 1988
Austria		1988	0.111	Pang et al., 1988
US (Wisconsin)		1988	0.067	Pang et al., 1988
Switzerland		1988	0.0542	Pang et al., 1988
Hungary	27,497,606	1989	0.0023	Thompson et al., 1989
Canada	346,000	1989	0.0689	Thompson et al., 1989
Hungary	2,119,727	1988	0.033	Sólyom, 1989
Japan	585,000	1990	0.0228	Pang and Clark, 1990
Kuwait	540,000	1990	0.001	Lubani et al., 1990
Sweden	1,727,928	1990	0.0827	Thilén and Larsson, 1990
Israel (Jewish population)		1986–1991	0.0333	Sack et al., 1997
Israel (Arab population)		1986–1991	0.1250	Sack et al., 1997
US		1993	0.025	Pang and Clark, 1993
Average (SD)	0.0584 (0.0495)			
Range	0.001–0.198			
Part B: Mass screening				
Alaska-Caucasian	13,733	1982	0.0728	Pang et al., 1982
Hungary	968,303	1989	0.0537	Sólyom, 1989
France (La Reunion)	31,472	1990	0.315	Pang and Clark, 1990
US (Illinois)	357,825	1990	0.0838	Pang and Clark, 1990
Portugal	100,000	1990	0.070	Pang and Clark, 1990
Scotland	119,960	1990	0.0583	Pang and Clark, 1990
Washington	255,527	1990	0.0547	Pang and Clark, 1990
Japan	2,523,948	1990	0.0223	Pang and Clark, 1990
Switzerland	65,823	1993	0.0911	Pang and Clark, 1993
Germany	12,500	1993	0.08	Pang and Clark, 1993
Italy	133,198	1993	0.09	Pang and Clark, 1993
Sweden	660,000	1993	0.0848	Pang and Clark, 1993
France (regional)	270,060	1993	0.0778	Pang and Clark, 1993
Canada	50,000	1993	0.06	Pang and Clark, 1993
Brazil	82,870	1993	0.1327	Pang and Clark, 1993
Spain	206,875	1993	0.058	Pang and Clark, 1993
Alaska-Yupik	3,740	1993	3.47	Pang and Clark, 1993
Native Alaskan	12,131	1993	1.236	Pang and Clark, 1993
New Zealand	536,915	1995	0.0428	Cutfield, 1995
Texas (white)	872,648	1989–1995	0.064	Therrell et al., 1998
Texas (Hispanic)	764,101	1989–1995	0.069	Therrell et al., 1998
Texas (African American)	253,854	1989–1995	0.0236	Therrell et al., 1998
Texas (other)	46,410	1989–1995	0.1293	Therrell et al., 1998
Average for screening (SD)	0.280 (0.738)			
Without Yupik or Native Alaskan	0.083 (0.06)			
Without Yupik, Native Alaskan, or La Reunion	0.0709 (0.0288)			

hydroxylase deficiency is the most common, and the frequency estimates are the most reliable. Pang and Clark (1990, 1993; Pang et al., 1988) have published detailed comparisons of estimates obtained from case reports in large population databases, as well as from more recently available mass screening programs. The direct screening programs have resulted in higher estimates for the condition than have the more traditional population surveys. In both instances, however, one striking fact emerges:

TABLE 5. Incidence of 11-B-hydroxylase (classic) in six published surveys

Location and subpopulation	Sample size	Method	Frequency/1,000 live births	Reference
Switzerland	1,516,299	Case	0.0032	Werder et al., 1980
France	13,921,803	Case	0.0071	Bois et al., 1985
Hungary	2,119,727	Case	0.0018	Sólyom, 1989
Kuwait	540,000	Case	0.0055	Lubani et al., 1990
Sweden	1,727,928	Case	0.0005	Thilén and Larsson, 1990
Moroccan Jews		Gene frequency estimates	0.1667	Rösler et al., 1992
		Mean (SD)	0.031 (0.067)	
		Range	0.0005–0.1667	
		Mean (SD) and range without Moroccan Jews	0.00362 (0.00267) 0.0005–0.0071	

the gene frequency for 21-hydroxylase deficiency varies significantly among populations. Worldwide and national estimates must be considered in this light.

Table 4 presents data obtained from original sources and from reviews by Pang and Clark (1990, 1993). The average frequency for 21-hydroxylase deficiency using case surveys is 0.0584 ± 0.0495 and a range of 0.001–0.198/1,000 live births. The corresponding statistics from mass screening (and minus the very high incidence among Yupik Eskimos) are 0.083 ± 0.060, and a range of 0.0428–0.315 per 1,000 live births. The worldwide frequencies do not include the Yupik data (3.47/1,000 live births) and the incidence in La Reunion, France (0.315/1,000 live births). It does not seem obvious, however, that it was appropriate to eliminate the La Reunion data; even though the number is high, it is in the same order of magnitude as other data used in the calculation (e.g., Brazil). Therefore, we averaged the numbers arrived at with and without the La Reunion data, arriving at a worldwide frequency of classic CAH due to 21-hydroxylase deficiency of 0.0770/1,000 live births.

Tables 5 and 6 contain data concerning the rarer enzyme deficiencies leading to CAH. The average incidence for 3-β hydroxysteroid dehydrogenase is 0.00068/1,000 live births, while that for 11-β-hydroxylase (minus the very high frequency found in Moroccan Jews) is 0.00362/1,000 live births (0.00267) with a range of 0.0005–0.0071/1,000. The one available frequency estimate for 17-alpha hydroxylase places the number at 7×10^{-5}/1,000 live births.

Finally, the unusual case of nonclassic or late onset 21-hydroxylase deficiency needs consideration. Nonclassic CAH is defined as a deficiency that arises anytime after the first 5 years of life. In childhood, such cases may come to medical attention because of premature signs of puberty, hirsutism, and clitoral growth. In adults, the signs can include hirsutism, menstrual disorders, and clitoral enlargement (Eldar-Geva et al., 1990; Pollack et al., 1981). Speiser et al. (1985) used estimates of gene frequencies for late onset 21-hydroxylase deficiencies to calculate the incidence of affected individuals in several different localities and ethnic groups. While the incidence of late-onset 21-hydroxylase varies widely among different ethnic groups, its overall frequency is extremely high. The calculated frequencies are 37/1,000 among Ashkenazi, 19/1,000 among Hispanics, 16/1,000 among Yugoslavs, 3/1,000 among Italians, and 0.01/1,000 among a mixed Caucasian population. Although the estimates are widely accepted and cited (Arnaut, 1992; Eldar-Geva et al., 1990; White et al., 1987; Newfield and New, 1997). We could not locate articles confirming the reported frequencies of Speiser et al. (1985). Thus, while we use this estimate in the final calculation of nondimorphism, future reports may contain modifications of the estimates.

Vaginal and penile agenesis

XY babies born with testes, but complete absence of a penis, are extremely rare, probably occurring only once in a million births (Bansal and Singh, 1990; Kumar et al., 1986; Rupprecht et al., 1989). In contrast, complete or partial vaginal agenesis is fairly common. Harkins et al. (1981) report that 6 of 26 patients with vaginal agenesis had AIS, while the remainder had Meyer-Rokitansky-Küster-Hauser (MRKH) syndrome, which is characterized by aplasia of

TABLE 6. Incidence of rare forms of classic CAH published surveys

Location and subpopulation	Type	Sample size	Method	Frequency/1,000 live births	Reference
Switzerland	3-Beta Ohase (classic)	1,516,299	Case	0.0019	Werder et al., 1980
France	3-Beta Ohase (classic)	13,921,803	Case	0.0007	Bois et al., 1985
France	17-alpha-hydroxylase	13,921,803	Case	0.00007	Bois et al., 1985
Kuwait	3-Beta Ohase (classic)	540,000	Case	0.0055	Lubani et al., 1990
Average for 3-Beta	0.00068/1000				

the vagina, typical female secondary sexual characteristics, attenuated fallopian tubes, and typical ovaries and female karyotype (Chervenak et al., 1982). To avoid "double counting" of AIS patients, we report complete estimates of vaginal agenesis incidence, but in the final calculations, assume that only 77% of these comprise an otherwise unmeasured deviation from the usual pathway of female development.

Unfortunately, the literature on the incidence of vaginal agenesis appears more promising than it actually is. Most articles quote an incidence of 1/4,000 to 1/5,000 births. When one traces the citations, however, they all lead to two publications. Engstad (1917, p. 330), reporting on cases encountered in a private practice, writes: "From my own experience I should judge that we might expect to find one case in about five thousand." Bryan et al. (1949) note the Engstad report and cite a paper by Owens, who reported six cases in 125,000 hospital admissions (0.048/1,000). From their own clinical experience, Bryan et al. (1949) estimate a frequency of 1/4,000 female patients (0.25/1,000). Currie (1974) reported that between July 1969 and 1973 there were 5,189 deliveries and 2,988 gynecological admissions at a USAF Medical Center. Of the total of 8,177, there were two records of complete or partial vaginal agenesis. In the most recent independent estimate, Willemsen and Dony (1988) estimated that 1/30,000 living Dutch-born women have Mayer-Rokitansky syndrome. Recognizing the limited basis of the present knowledge of the frequency of vaginal agenesis, we use the figure of 1/4,500. Assuming that 77% of these are due to unique causes, the final incidence is 0.1694/1,000 live births. Since congenital absence of the vagina can be asymptomatic, this may be an underestimate.

Hormone-producing tumors and exogenous sex hormones

Hormone-producing tumors are relatively rare, and no population-level estimates of incidence exist. They can, however, cause virilization of adult women, including voice changes, clitoral growth, and hirsutism. In addition, they have been known to cause fetal masculinization during pregnancy (Hensleigh and Woodruff, 1978; Ireland and Woodruff, 1976; Verhoeven et al., 1973). It is also difficult to ascertain the frequency of genital alterations caused by treatment with progestins during pregnancy. A recent meta-analysis of studies done on births following first trimester exposure to low doses of sex hormones, especially from oral contraceptives, suggests little or no danger to genital development (Raman-Wilms et al., 1995). However, earlier studies focused on much higher doses of progestin, used in efforts to avoid miscarriage. Not only were the doses greater in these cases, but treatment occurred well into the second trimester of pregnancy, a time when one might especially expect an effect on the development of external genitalia (Steinberger and Odell, 1989). Not all progestin-treated pregnancies result in fetal masculinization. However, the rate for high-dose, second-trimester treatments is probably quite high (Burstein and Wasserman, 1974; Ishizuka et al., 1962; Jacobson, 1962). Unfortunately, no good estimates exist of the number of individuals currently living with iatrogenically induced genital alterations. There is also no presently reliable way to know whether the practice of treating threatened miscarriages with progestins continues with any frequency today, although the data presented in Table 7 (section on True Hermaphrodites) suggest that the practice has significantly declined.

True hermaphrodites and idiopathic mixed genitalia

There are no published population-wide estimates of the frequency of true hermaphrodites (individuals born with both testicular and ovarian tissue). However, a number of surgeons and endocrinologists, who specialize in the treatment of nonconforming

TABLE 7. Comparative numbers of true hermaphrodites and other forms of intersexuality

Year	Hypospadias	CAH	Gonadal dysgenesis (mixed and pure)	AIS: complete and partial	True herma-phrodites	Exo-genous	Vaginal agenesis or atresia	Penile agenesis	Cloacal malforma-tions	Idio-pathic	XY: micro-penis	Virilizing adenoma	Reference
1960		87	71	47	3	22							Wilkins, 1960
1965		100	116	66	4	18				20			Wilkins, 1965
1967		39	5	7	1	16							Jones and Wilkins, 1967
1974		28			6							1	Kumar et al., 1974
1986	19	29	7	1	1	3	11		8				Currarino, 1986
1987	0	24	8	5	6	1				14			Lobe et al., 1987
1987		4	1	2	1						1		Oesterling et al., 1987
1989		21		10	2								Lorge et al., 1989
1990	4	6	6	5	2			1					Pinter and Koxatolanyi, 1990
1991		14	1	7						3			Abdullah et al., 1991
1991	6	32	10	10	3		4		4	5			Coran and Z., 1991
1992	6	46	8		4	9	1	2	6	7			Newman et al., 1992
1992		12	5	7	38	9							Danso and Nkrumah, 1992
1993		38	23	12									Ramani et al., 1993
1994	39(total)	4		1									Greenfield et al., 1994
Total		442			84								
% of CAH (all entries)					19%					11%			
% of CAH minus rows 7 (possible duplicate) and 13 (African data)					13%								

TABLE 8. Frequencies of various causes of nondimorphic sexual development

Cause	Estimated frequency/100 live births
Non-XX or non-XY (except Turner or Klinefelter)	0.0639
Turner	0.0369
Klinefelter	0.0922
Subtotal for chromosomal difference	0.193
Androgen Insensitivity syndrome	0.00760
Partial Androgen Insensitivity syndrome	0.000760
Classic CAH (omitting very high frequency population)	0.00770
Late-onset CAH	1.5
Subtotal of known hormonal causes	1.516
Vaginal agenesis	0.0169
True hermaphrodites	0.0012
Idiopathic	0.0009
Total	1.728

physical sex types, report on the distribution of patients. Data extracted from 14 such reports are summarized in Table 7. Although different reports come from different specialties, rendering the referral bias for any one report great, by analyzing a large number of such reports and making use of the fact that the frequency of CAH is well established, an estimate of the order of magnitude of the occurrence of true hermaphroditism can be obtained.

Particularly striking in Table 7 are the data of Danso and Nkrumah (1992), who reported 38 true hermaphrodites in a database of 71 patients with ambiguous genitalia. Although the reported numbers seem inordinately high, the data are consistent with other reports suggesting high frequencies of true hermaphroditism in southern Africa (Krob et al., 1994; Ramsay et al., 1988). In addition, some forms of true hermaphroditism are familial (Kuhnle et al., 1993; Skordis et al., 1987; Slaney et al., 1998), which opens the possibility that, as with other inherited forms of sexual ambiguity, there may be pockets, perhaps even large geographical regions, with relatively high frequencies of true hermaphroditism.

The data in Table 7 were used to estimate the relative frequency of true hermaphroditism. First we compared the number of true hermaphrodites summed from all 14 reports with the number of cases of CAH (19%). Then we considered the percentage after eliminating the report from southern Africa, as well as the older of two reports which may contain duplicate data. The latter ratio is 11% and, splitting the difference, the estimate of true hermaphroditism equals 15% of the frequency of classic 21-OHase CAH. Using a figure of 0.0779/1,000 live births for classic CAH, the average frequency of true hermaphroditism is on the order of 0.0117/1,000 live births, or one in 100,000.

The data in Table 7 also allow the calculation of the frequency of idiopathic sexual ambiguity. Using the same reasoning as for true hermaphrodites, the birth of a sexually ambiguous child from unknown causes is about 0.009/1,000 live births.

Overview

This article began by asking how frequently members of the human population deviate from a Platonic ideal of sexual dimorphism. A summary of the frequencies of known causes of sexual ambiguity based on Tables 1–7 appears in Table 8. The grand total is 1.728% of live births. Because there are no general population-level frequency estimates for iatrogenic variations in genital anatomy, penile agenesis, and disorders of 5-α-reductase biosynthesis and some of the rarer forms of CAH, the data in Table 8 provide a minimal estimate. However, except for certain restricted populations the frequencies of such events are quite rare and would probably not greatly influence the overall estimate. The two most frequent deviations from complete sexual dimorphism arise from nondimorphic sex chromosome conditions and from alterations in steroid hormone metabolism. Although this generalization holds for a generic Euro-American, Caucasian population, it is inappropriate in certain geographical settings. Thus, there is strong evidence that CAH is very frequent among native Alaskans and that true hermaphroditism is surprisingly common in southern Africa. Because of the Eurocentric nature of most medical data, there may well be other large population groups worldwide which exhibit substantial frequencies of intersexuality.

TABLE 9. Incidence of hypospadias

Location	Date	Total sample	Incidence/1,000 live births	Reference
Rochester, MN	1954	8,716	3.901	Harris and Steinberg, 1954
Brooklyn	1958	30,398	0.921	Shapiro et al., 1958
International	1966	416,695	0.586	Stevenson et al., 1966
Liverpool, UK	1968	91,176	1.228	Smithells, 1968
US, multiregion	1968	35,680	2.41	Chung and Myrianthopoulos, 1968
South Wales, UK	1972	92,982	1.097	Roberts et al., 1972
Sweden (general)	1973	550,000	0.949	Kallen, 1973
Sweden (Uppsala)	1973	96,733	1.158	Pettersson, 1973
Jerusalem	1973	59,261	3.004	Harlap, 1973
Athens	1973	74,390	1.949	Trichopoulos et al., 1973
Atlanta, GA	1974	137,179	2.012	CDC statistics cited in (Avellan, 1975)
Jerusalem	1975	11,036	3.625	Harlap, 1973
Sweden	1975	480,607	1.386	Avellan, 1975
Latin American	1981	432,839	0.764	Neto and Paz, 1981
Emilia-Romagna, Italy	1986	42,156	3.985	Calzolari et al., 1986
Denmark	1986	801,241	0.975	Kallen et al., 1986
Hungary	1986	1,992,773	1.827	Kallen et al., 1986
Italy	1986	303,674	1.798	Kallen et al., 1986
Mexico	1986	162,105	0.352	Kallen et al., 1986
South America	1986	902,984	0.693	Kallen et al., 1986
Spain	1986	334,970	1.797	Kallen et al., 1986
Sweden	1986	896,954	2.085	Kallen et al., 1986
Alsace, France	1990	118,265	1.488	Stoll et al., 1990
Atlanta, GA	1993	?	3.0	Paulozzi et al., 1997
USA	1993	?	3.8	Paulozzi et al., 1997
Mean	1.87	SD	1.105	Range: 0.352–3.985

Estimates that combine categories

The approach used to estimate intersexuality, at all levels, from the chromosomal and hormonal to the anatomical, is plagued by the uncertainties inherent in the medical literature. Therefore, we derived a second type of estimate from statistics on the frequencies of cryptorchidism (undescended testes) and hypospadias (the incomplete closure around the urethra of the embryonic genital folds). These estimates serve as an order of magnitude check of the preceding calculations. Hypospadias and cryptorchidism both result from a variety of underlying causes of intersexuality (Aaronson et al., 1997; Aarskog, 1971; Gearhart et al., 1990; Gill et al., 1989; Gill and Kogan, 1997; Rajfer and Walsh, 1976). A U.S. Army survey in the 1940s (Rajfer and Walsh, 1976) found that 0.7% of the adult male population had cryptorchidism. Scorer and Farrington (1971) noted that the incidence of cryptorchidism is higher at birth and declines throughout the first year of life to 0.8% of male births (or roughly 0.4% of all births).

Table 9 contains an overview of the data for hypospadias. The mean incidence of hypospadias, averaged from over 20 studies over four decades, is 1.87/1,000 live births, with a standard deviation of 1.105 and a range of 0.352–1.043. The data on hypospadias may be further subdivided into the incidence of severe and medium types (urethral opening in the perineal region or along the shaft of the penis) and minimal types (urethral opening between the corona and the tip of the glans penis) (Sweet et al., 1974). Indeed Fichtner et al. (1995) have recently shown widespread variation in the meatal opening along the length of the glans penis. The authors suggested that such variation, sometimes classified as minimal or mild hypospadias, is normal and surgery in such cases unwarranted. It is difficult to obtain absolute estimates of the rates of minimal, medium, and severe hypospadias, because not all publications record the data in the same manner, and some contain numerical discrepancies. Nevertheless, the ratio of minimal to medium and severe hypospadias is about 3:1. If, as Fichtner et al. (1995) suggest, only medium and severe hypospadias represent deviations from a dimorphic ideal, then the incidence calculated from cases of hypospadias would be 0.5797/1,000, or 0.05%.

Several studies suggest that the rate of hypospadias has increased significantly in

the past two decades (Paulozzi et al., 1997; Toppari et al., 1996; Kristensen et al., 1997). Furthermore, severe hypospadias seems to have increased at a more rapid rate than the mild form, and there are both regional (within the U.S.) and national differences in overall rates of hypospadias, as well as in the rate of increase. While the cause of such increases is currently unknown, such trends mean that current estimates of the rate of intersexual births may require revision with time.

DISCUSSION

Adding the estimates of all known causes of nondimorphic sexual development suggests that approximately 1.7% of all live births do not conform to a Platonic ideal of absolute sex chromosome, gonadal, genital, and hormonal dimorphism. The incidence of hypospadias (0.05%) and cryptorchidism (0.4%), conditions of mixed origin affecting the apparently male population, are lower than the present estimate. However, the calculation includes categories which result in neither hypospadias nor cryptorchidism. The single largest contribution to the higher figure comes from late-onset CAH. If this cause of nondimorphism is deleted, the frequency estimates obtained from population surveys would come to 0.228%, the same order of magnitude found after combining the incidences of severe and medium hypospadias and cryptorchism (0.05 + 0.4 = 0.45%). Alternatively, if mild hypospadias and late-onset CAH in the final calculations are included the combined figure is 2.27% for hypospadias and cryptorchidism, compared with 1.728% obtained from summing the incidence of all known causes for which available data exist. These data, obtained using independent methods, are in general agreement. Which number one chooses to use depends on the specific population under study, and the assumptions as to what should count as true dimorphism. It would appear, however, that earlier estimates that intersexual births might run as high as 4% are unwarranted, except in populations in which a particular genetic condition occurs with high frequency (Fausto-Sterling, 1993a,b; Money, 1993).

Recently, a nascent social movement to recognize intersexuality as a legitimate state of nature has criticized medical approaches to the management of intersexuality (Nevada and Chase, 1995; Kessler, 1998) and these criticisms have begun to appear in more traditional medical settings (Diamond and Sigmundsen, 1997; Phornphutkul et al., 1999). Members of the Intersex Society of North America oppose the use of genital surgery to "normalize" children who are too young to decide for themselves whether to modify their genital structures. We define the intersexual as an individual who deviates from the Platonic ideal of physical dimorphism at the chromosomal, genital, gonadal, or hormonal levels. Not all intersexuals would be candidates for genital surgery. Lilford and Dear (1987) suggest that 0.05% (1 in 2,000) newborns have some ambiguity of the external genitalia, although they cite no medical or scientific literature to back up their claim. As one might expect, even given the data in Table 8, estimating the number of children subject to controversial genital surgery is an uncertain business. Turner and Klinefelter syndromes do not usually call for surgical intervention. However, many, although probably not all, of the other chromosomal alterations do, since they often result in intermediate genital development (Mittwoch, 1992). All of the hormonal disruptions potentially cause conditions which have been treated surgically. The highest frequency of intersexuality comes from late-onset CAH. When late-onset CAH occurs in childhood or adolescence and causes significant clitoral growth, it is quite possible that surgical intervention will ensue (Moreno and Goodwin, 1998). However, there is no way to estimate what proportion of late-onset CAH patients fall into this subcategory. Combining chromosomal deviations other than Turner or Klinefelter, all hormonal alterations, vaginal agenesis, true hermaphrodites, and idiopathic genital intersex, produces an estimate that 1.62% of the population may be subject to genital surgery as a treatment for intersexuality. Without late-onset CAH in this calculation, the estimate falls to 0.08%, or between one and two in a thousand. The true frequency of such surgeries probably lies somewhere in between.

Our culture acknowledges the wide variety of body shapes and sizes characteristic of males and females. Most sexual dimorphisms involve quantitative traits, such as height, build, and voice timbre, for which considerable overlap exists between males and females. Many cultures use dress code, hair style, and cultural conventions, e.g.,

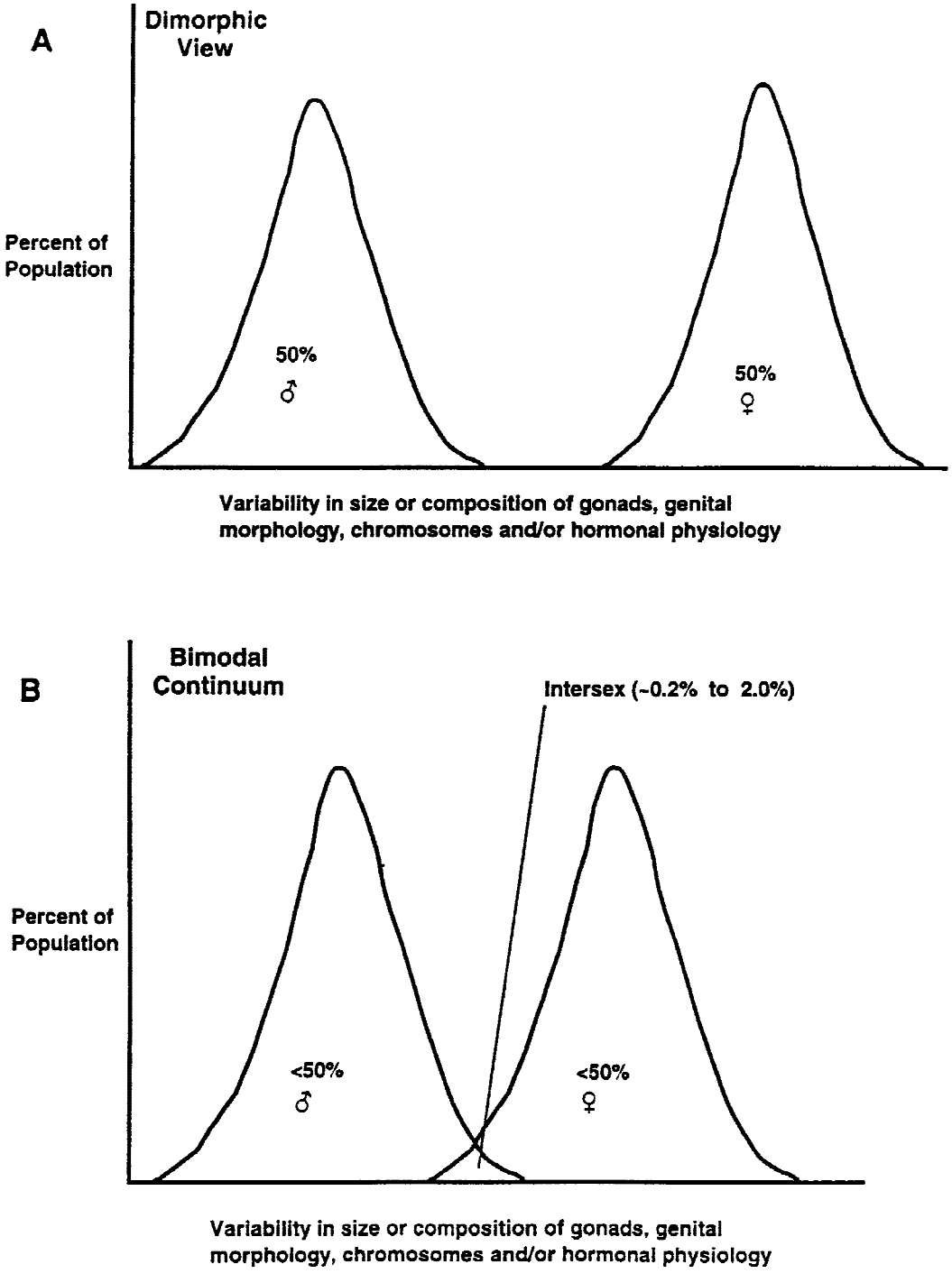

Fig. 1. (**A**) Absolute dimorphism; (**B**) incomplete dimorphism.

the view that in couples the male should be taller than the female, to accentuate awareness of such difference (see Unger and Crawford, 1992). But most consider that at the level of chromosomes, hormones, and genitals, dimorphism is absolute and, by implication, such traits are discrete rather than quantitative. Clearly, as a generalization, such a viewpoint makes some sense. However, developmental biology suggests that a belief in absolute sexual dimorphism is wrong. Instead, two overlapping bell-shaped curves can be used to conceptualize sexual variation across the population (Fig. 1). Within each major bell, genital morphology varies quantitatively, as shown, for example, by Fichtner et al. (1995). In the region of overlap, qualitative variation in chromosomal and genital morphology and in hormonal activity exists. If the view of the human population schematically illustrated in Figure 1B is accepted, the requirement for medical intervention in cases of intersexuality needs to be carefully reexamined. It seems likely that changing cultural norms concerning sex roles and gender-related behaviors may encourage a willingness to engage in such a reexamination.

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SPECIAL ARTICLE

Consensus Statement on Management of Intersex Disorders

Peter A. Lee, MD, PhD[a,b], Christopher P. Houk, MD[c], S. Faisal Ahmed, FRCPCH[d], Ieuan A. Hughes, FMedSci, FRCPCH[b,e], in collaboration with the participants in the International Consensus Conference on Intersex organized by the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology

[a]Department of Pediatrics, Penn State College of Medicine, Hershey, Pennsylvania; [c]Department of Pediatrics, Backus Children's Hospital, Mercer University School of Medicine, Savannah, Georgia; [d]Department of Child Health, Royal Hospital for Sick Children, Glasgow, Scotland, United Kingdom; [e]Department of Paediatrics, University of Cambridge, Addenbrooke's Hospital, Cambridge, United Kingdom

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THE BIRTH of an intersex child prompts a long-term management strategy that involves myriad professionals working with the family. There has been progress in diagnosis, surgical techniques, understanding psychosocial issues, and recognizing and accepting the place of patient advocacy. The Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology considered it timely to review the management of intersex disorders from a broad perspective, review data on longer-term outcome, and formulate proposals for future studies. The methodology comprised establishing a number of working groups, the membership of which was drawn from 50 international experts in the field. The groups prepared previous written responses to a defined set of questions resulting from evidence-based review of the literature. At a subsequent gathering of participants, a framework for a consensus document was agreed. This article constitutes its final form.

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[b]Co-chairs.

Key Words
intersex, sexual differentiation, ambiguous genitalia, genital surgery

Abbreviations
DSD— disorder(s) of sex development
CAH— congenital adrenal hyperplasia
CAIS— complete androgen insensitivity syndrome
5αRD2—5-α-reductase
PAIS—partial androgen insensitivity syndrome
MGD—mixed gonadal dysgenesis

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Address correspondence to Peter A. Lee, MD, PhD, Department of Pediatrics, MC-085, Penn State College of Medicine, Milton S. Hershey Medical Center, Box 850, 500 University Dr, Hershey, PA 17033-0850. E-mail: plee@psu.edu

NOMENCLATURE AND DEFINITIONS

Advances in identification of molecular genetic causes of abnormal sex with heightened awareness of ethical issues and patient advocacy concerns necessitate a reexamination of nomenclature.[1] Terms such as "intersex," "pseudohermaphroditism," "hermaphroditism," "sex reversal," and gender-based diagnostic labels are particularly controversial. These terms are perceived as potentially pejorative by patients[2] and can be confusing to practitioners and parents alike. We propose the term "disorders of sex development" (DSD), as defined by congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical.

The proposed changes in terminology are summarized in Table 1. A modern lexicon is needed to integrate progress in molecular genetic aspects of sex development. Because outcome data in individuals with DSD are limited, it is essential to use precision when applying definitions and diagnostic labels.[3,4] It is also appropriate to use terminology that is sensitive to the concerns of patients. The ideal nomenclature should be sufficiently flexible to incorporate new information yet robust enough to maintain a consistent framework. Terms should be descrip-

TABLE 1 **Proposed Revised Nomenclature**

Previous	Proposed
Intersex	DSD
Male pseudohermaphrodite, undervirilization of an XY male, and undermasculinization of an XY male	46,XY DSD
Female pseudohermaphrodite, overvirilization of an XX female, and masculinization of an XX female	46,XX DSD
True hermaphrodite	Ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

tive and reflect genetic etiology when available and accommodate the spectrum of phenotypic variation. Clinicians and scientists must value the nomenclature's use, and it must be understandable to patients and their families. An example of how the proposed nomenclature could be applied in a classification of DSD is shown in Table 2.

Psychosexual development is traditionally conceptualized as 3 components: "gender identity" refers to a person's self-representation as male or female (with the caveat that some individuals may not identify exclusively with either); "gender role" (sex-typical behaviors) describes the psychological characteristics that are sexually dimorphic within the general population, such as toy preferences and physical aggression; and "sexual orientation" refers to the direction(s) of erotic interest (heterosexual, bisexual, homosexual) and includes behavior, fantasies, and attractions. Psychosexual development is influenced by multiple factors such as exposure to androgens, sex chromosome genes, and brain structure, as well as social circumstance and family dynamics.

Gender dissatisfaction denotes unhappiness with assigned sex. Causes of gender dissatisfaction, even among individuals without DSD, are poorly understood. Gender dissatisfaction occurs more frequently in individuals with DSD than in the general population but is difficult to predict from karyotype, prenatal androgen exposure, degree of genital virilization, or assigned gender.[5–7] Prenatal androgen exposure is clearly associated with other aspects of psychosexual development.[8,9] There are dose-related effects on childhood play behavior in girls with congenital adrenal hyperplasia (CAH), whereby those with the more severe mutations and marked genital virilization play more with boys' toys.[10] Prenatal androgen exposure is also associated with other psychological characteristics such as maternal interest and sexual orientation. It is important to emphasize the separability of sex-typical behavior, sexual orientation, and gender identity. Thus, homosexual orientation (relative to sex of rearing) or strong cross-sex interest in an individual with DSD is not an indication of incorrect gender assignment. Understanding variations in psychosexual development in individuals with DSD requires reference to studies in nonhuman species that show marked but complex effects of androgens on sex differentiation of the brain and on behavior. Outcomes can be influenced by the timing, dose, and type of androgen exposure, receptor availability, and modification by the social environment.[11–14]

Data from rodent studies suggest that sex chromosome genes may also influence brain structure and behavior directly.[15,16] However, studies in individuals with complete androgen insensitivity syndrome (CAIS) do not indicate a behavioral role for Y-chromosome genes, although data are limited.[17] Sex differences in brain structures have been identified across species, some of which coincide with pubertal onset, perhaps suggesting hormonal responsivity.[18–20] The limbic system and hypothalamus, both of which play a role in reproduction, show sex differences in specific nuclei, but it is not clear

TABLE 2 **An Example of a DSD Classification**

Sex Chromosome DSD	46,XY DSD	46,XX DSD
45,X (Turner syndrome and variants)	Disorders of gonadal (testicular) development: (1) complete gonadal dysgenesis (Swyer syndrome); (2) partial gonadal dysgenesis; (3) gonadal regression; and (4) ovotesticular DSD	Disorders of gonadal (ovarian) development: (1) ovotesticular DSD; (2) testicular DSD (eg, SRY^+, duplicate SOX9); and (3) gonadal dysgenesis
47,XXY (Klinefelter syndrome and variants)	Disorders in androgen synthesis or action: (1) androgen biosynthesis defect (eg, 17-hydroxysteroid dehydrogenase deficiency, 5αRD2 deficiency, StAR mutations); (2) defect in androgen action (eg, CAIS, PAIS); (3) luteinizing hormone receptor defects (eg, Leydig cell hypoplasia, aplasia); and (4) disorders of anti-Müllerian hormone and anti-Müllerian hormone receptor (persistent Müllerian duct syndrome)	Androgen excess: (1) fetal (eg, 21-hydroxylase deficiency, 11-hydroxylase deficiency); (2) fetoplacental (aromatase deficiency, POR [P450 oxidoreductase]); and (3) maternal (luteoma, exogenous, etc)
45,X/46,XY (MGD, ovotesticular DSD)		Other (eg, cloacal exstrophy, vaginal atresia, MURCS [Müllerian, renal, cervicothoracic somite abnormalities], other syndromes)
46,XX/46,XY (chimeric, ovotesticular DSD)		

Although consideration of karyotype is useful for classification, unnecessary reference to karyotype should be avoided; ideally, a system based on descriptive terms (eg, androgen insensitivity syndrome) should be used wherever possible. StAR indicates steroidogenic acute regulatory protein.

when these differences emerge. Interpretation of sex differences is complicated by the effect of cell death and synaptic pruning on normal maturation and by effects of experience on the brain. Structure of the brain is not currently useful for gender assignment.

INVESTIGATION AND MANAGEMENT OF DSD

General Concepts of Care

Optimal clinical management of individuals with DSD[21] should comprise the following: (1) gender assignment must be avoided before expert evaluation in newborns; (2) evaluation and long-term management must be performed at a center with an experienced multidisciplinary team; (3) all individuals should receive a gender assignment; (4) open communication with patients and families is essential, and participation in decision-making is encouraged; and (5) patient and family concerns should be respected and addressed in strict confidence.

The initial contact with the parents of a child with a DSD is important, because first impressions from these encounters often persist. A key point to emphasize is that the child with a DSD has the potential to become a well-adjusted, functional member of society. Although privacy needs to be respected, a DSD is not shameful. It should be explained to the parents that the best course of action may not be clear initially, but the health care team will work with the family to reach the best possible set of decisions in the circumstances. The health care team should discuss with the parents what information to share in the early stages with family members and friends. Parents need to be informed about sexual development, and Web-based information may be helpful, provided the content and focus of the information is balanced and sound.

Ample time and opportunity should be made for continued discussion with review of information previously provided.[1]

The Multidisciplinary Team

Optimal care for children with DSD requires an experienced multidisciplinary team that is generally found in tertiary care centers. Ideally, the team includes pediatric subspecialists in endocrinology, surgery, and/or urology, psychology/psychiatry, gynecology, genetics, neonatology, and, if available, social work, nursing, and medical ethics.[22] Core composition will vary according to DSD type, local resources, developmental context, and location. Ongoing communication with the family's primary care physician is essential.[23]

The team has a responsibility to educate other health care staff in the appropriate initial management of affected newborns and their families. For new patients with DSD, the team should develop a plan for clinical management with respect to diagnosis, gender assignment, and treatment options before making any recommendations. Ideally, discussions with the family are conducted by one professional with appropriate communication skills.[24] Transitional care should be organized with the multidisciplinary team operating in an environment that includes specialists with experience in both pediatric and adult practice. Support groups can have an important role in the delivery of care to patients with DSD and their families[25] (see Appendix 1).

Clinical Evaluation

A family and prenatal history, a general physical examination with attention to any associated dysmorphic features, and an assessment of the genital anatomy in comparison to published norms need to be recorded (Table 3). Criteria that suggest DSD include (1) overt genital ambiguity (eg, cloacal exstrophy), (2) apparent female genitalia with an enlarged clitoris, posterior labial fusion, or an inguinal/labial mass, (3) apparent male genitalia with bilateral undescended testes, micropenis, isolated perineal hypospadias, or mild hypospadias with unde-

TABLE 3 **Anthropometric Measurements of the External Genitalia**

Sex	Population	Age	Stretched Penile Length, Mean ± SD, cm (Males), or Clitoral Length, Mean ± SD, mm (Females)	Penile Width, Mean ± SD, cm (Males), or Clitoral Width, Mean ± SD, mm (Females)	Mean Testicular Volume, mL (Males), or Perineum Length, Mean ± SD, mm (Females)	Ref No.
M	United States	30 wk GA	2.5 ± 0.4			26
M	United States	Term	3.5 ± 0.4	1.1 ± 0.1	0.52 (median)	26 and 27
M	Japan	Term to 14 y	2.9 ± 0.4 − 8.3 ± 0.8			28
M	Australia	24–36 wk GA	2.27 + (0.16 GA)			29
M	China	Term	3.1 ± 0.3	1.07 ± 0.09		30
M	India	Term	3.6 ± 0.4	1.14 ± 0.07		30
M	North America	Term	3.4 ± 0.3	1.13 ± 0.08		30
M	Europe	10 years	6.4 ± 0.4		0.95–1.20	27 and 31
M	Europe	Adult	13.3 ± 1.6		16.5–18.2	27 and 31
F	United States	Term	4.0 ± 1.24	3.32 ± 0.78		32
F	United States	Adult nulliparous	15.4 ± 4.3			33
F	United States	Adult	19.1 ± 8.7	5.5 ± 1.7	31.3 ± 8.5	34

GA indicates gestational age.

scended testis, (4) a family history of DSD such as CAIS, and (5) a discordance between genital appearance and a prenatal karyotype. Most causes of DSD are recognized in the neonatal period; later presentations in older children and young adults include (1) previously unrecognized genital ambiguity, (2) inguinal hernia in a female, (3) delayed or incomplete puberty, (4) virilization in a female, (5) primary amenorrhea, (6) breast development in a male, and (7) gross and occasionally cyclic hematuria in a male.

Diagnostic Evaluation

Considerable progress has been made with understanding the genetic basis of human sexual development,[35] yet a specific molecular diagnosis is identified in only ~20% of cases of DSD. The majority of virilized 46,XX infants will have CAH. In contrast, only 50% of 46,XY children with DSD will receive a definitive diagnosis.[36,37] Diagnostic algorithms do exist, but with the spectrum of findings and diagnoses, no single evaluation protocol can be recommended in all circumstances. Some tests, such as imaging by ultrasound, are operator dependent. Hormone measurements need to be interpreted in relation to the specific assay characteristics and to normal values for gestational and chronological age. In some cases, serial measurements may be needed.

First-line testing in newborns includes karyotyping with X- and Y-specific probe detection (even when prenatal karyotype is available), imaging (abdominopelvic ultrasound), measurement of 17-hydroxyprogesterone, testosterone, gonadotropins, anti-Müllerian Hormone, and serum electrolytes, and urinalysis. The results of these investigations are generally available within 48 hours and will be sufficient for making a working diagnosis. Decision-making algorithms are available to guide additional investigation.[38] These assessments include human chorionic gonadotropin– and adrenocorticotropin-stimulation tests to assess testicular and adrenal steroid biosynthesis, urinary steroid analysis by gas chromatography mass spectroscopy, imaging studies, and biopsies of gonadal material. Some gene analyses are performed in clinical service laboratories. However, current molecular diagnosis is limited by cost, accessibility, and quality control.[39] Research laboratories provide genetic testing, including functional analysis, but may face restrictions on communicating results.[40]

Gender Assignment in Newborns

Initial gender uncertainty is unsettling and stressful for families. Expediting a thorough assessment and decision is required. Factors that influence gender assignment include diagnosis, genital appearance, surgical options, need for lifelong replacement therapy, potential for fertility, views of the family, and, sometimes, circumstances relating to cultural practices. More than 90% of patients with 46,XX CAH[41] and all patients with 46,XY CAIS assigned female in infancy[42] identify as females. Evidence supports the current recommendation to raise markedly virilized 46,XX infants with CAH as female.[43] Approximately 60% of 5-α-reductase (5αRD2)-deficient patients assigned female in infancy and virilizing at puberty (and all assigned male) live as males.[5] In 5αRD2 and possibly 17β-hydroxysteroid dehydrogenase deficiencies, for which the diagnosis is made in infancy, the combination of a male gender identity in the majority and the potential for fertility (documented in 5αRD2 but unknown in 17β-hydroxysteroid dehydrogenase deficiencies) should be discussed when providing evidence for gender assignment.[5,44,45] Among patients with partial androgen insensitivity syndrome (PAIS), androgen biosynthetic defects, and incomplete gonadal dysgenesis, there is dissatisfaction with the sex of rearing in ~25% of individuals whether raised male or female.[46] Available data support male rearing in all patients with micropenis, taking into account equal satisfaction with assigned gender in those raised male or female but no need for surgery and the potential for fertility in patients reared male.[42] Those making the decision on sex of rearing for those with ovotesticular DSD should consider the potential for fertility on the basis of gonadal differentiation and genital development and assuming that the genitalia are, or can be made, consistent with the chosen sex. In the case of mixed gonadal dysgenesis (MGD), factors to consider include prenatal androgen exposure, testicular function at and after puberty, phallic development, and gonadal location. Individuals with cloacal exstrophy reared female show variability in gender identity outcome, but >65% seem to live as female.[6]

Surgical Management

The surgeon has a responsibility to outline the surgical sequence and subsequent consequences from infancy to adulthood. Only surgeons with expertise in the care of children and specific training in the surgery of DSD should perform these procedures. Parents now seem to be less inclined to choose surgery for less severe clitoromegaly.[47] Surgery should only be considered in cases of severe virilization (Prader III–V) and be performed in conjunction, when appropriate, with repair of the common urogenital sinus. Because orgasmic function and erectile sensation may be disturbed by clitoral surgery, the surgical procedure should be anatomically based to preserve erectile function and the innervation of the clitoris. Emphasis is on functional outcome rather than a strictly cosmetic appearance. It is generally felt that surgery that is performed for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parents[48–51]; the systematic evidence for this belief is lacking.

Currently, there is inadequate evidence in relation to establishment of functional anatomy to abandon the practice of early separation of the vagina and urethra.[52]

The rationale for early reconstruction is based on guidelines on the timing of genital surgery from the American Academy of Pediatrics,[53] the beneficial effects of estrogen on tissue in early infancy, and the avoidance of potential complications from the connection between the urinary tract and peritoneum via the Fallopian tubes. It is anticipated that surgical reconstruction in infancy will need to be refined at the time of puberty.[54–56] Vaginal dilatation should not be undertaken before puberty. The surgeon must be familiar with a number of operative techniques to reconstruct the spectrum of urogenital sinus disorders. An absent or inadequate vagina (with rare exceptions) requires a vaginoplasty performed in adolescence when the patient is psychologically motivated and a full partner in the procedure. No one technique has been universally successful; self-dilatation, skin substitution, and bowel vaginoplasty each have specific advantages and disadvantages.

In the case of a DSD associated with hypospadias,[57] standard techniques for surgical repair such as chordee correction, urethral reconstruction, and the judicious use of testosterone supplementation apply. The magnitude and complexity of phalloplasty in adulthood should be taken into account during the initial counseling period if successful gender assignment depends on this procedure.[58] At times, this may affect the balance of gender assignment. Patients must not be given unrealistic expectations about penile reconstruction, including the use of tissue engineering. There is no evidence that prophylactic removal of asymptomatic discordant structures, such as a utriculus or Müllerian remnants, is required, although symptoms in the future may indicate surgical removal. For the male who has a successful neophalloplasty in adulthood, an erectile prosthesis may be inserted but has a high morbidity.

The testes in patients with CAIS[35] and those with PAIS, raised female, should be removed to prevent malignancy in adulthood. The availability of estrogen-replacement therapy allows for the option of early removal at the time of diagnosis that also takes care of the associated hernia, psychological problems with the presence of testes, and the malignancy risk. Parental choice allows deferment until adolescence, recognizing that the earliest reported malignancy in CAIS is at 14 years of age.[59] The streak gonad in a patient with MGD raised male should be removed laparoscopically (or by laparotomy) in early childhood.[35] Bilateral gonadectomy is performed in early childhood in females (bilateral streak gonads) with gonadal dysgenesis and Y-chromosome material. In patients with androgen biosynthetic defects raised female, gonadectomy should be performed before puberty. A scrotal testis in patients with gonadal dysgenesis is at risk for malignancy. Current recommendations are testicular biopsy at puberty seeking signs of the premalignant lesion termed carcinoma in situ or undifferentiated intratubular germ cell neoplasia. If positive, the option is sperm banking before treatment with local low-dose radiotherapy that is curative.[60]

Surgical management in DSD should also consider options that will facilitate the chances of fertility. In patients with a symptomatic utriculus, removal is best performed laparoscopically to increase the chance of preserving continuity of the vas deferens. Patients with bilateral ovotestes are potentially fertile from functional ovarian tissue.[35,61] Separation of ovarian and testicular tissue can be technically difficult and should be undertaken, if possible, in early life.

Sex-Steroid Replacement

Hypogonadism is common in patients with dysgenetic gonads, defects in sex-steroid biosynthesis, and resistance to androgens. The timing of initiation of puberty may vary, but this is an occasion that provides an opportunity to discuss the condition and set a foundation for long-term adherence to therapy. Hormonal induction of puberty stimulates replication of normal pubertal maturation to induce secondary sexual characteristics, a pubertal growth spurt, and optimal bone mineral accumulation, together with psychosocial support for psychosexual maturation.[62] Intramuscular depot injections of testosterone esters are commonly used in males; another option is oral testosterone undecanoate, and transdermal preparations are also available.[63–65] Patients with PAIS may require supraphysiologic doses of testosterone for optimal effect.[66] Females with hypogonadism require estrogen supplementation to induce pubertal changes and menses. A progestin is usually added after breakthrough bleeding develops or within 1 to 2 years of continuous estrogen. There is no evidence that the addition of cyclic progesterone is beneficial in women without a uterus.

Psychosocial Management

Psychosocial care provided by mental health staff with expertise in DSD should be an integral part of management to promote positive adaptation. This expertise can facilitate team decisions about gender assignment/reassignment, timing of surgery, and sex-hormone replacement. Psychosocial screening tools that identify families at risk for maladaptive coping with a child's medical condition are available.[67] Once the child is sufficiently developed for a psychological assessment of gender identity, such an evaluation must be included in discussions about gender reassignment. Gender identity development begins before the age of 3 years,[68] but the earliest age at which it can be reliably assessed remains unclear. The generalization that the age of 18 months is the upper limit of imposed gender reassignment should be treated with caution and viewed conservatively. Atypical gender-role behavior is more common in children with DSD than in the general population but should not be taken as an indicator for gender reassignment. In

affected children and adolescents who report significant gender dysphoria, a comprehensive psychological evaluation[69] and an opportunity to explore feelings about gender with a qualified clinician is required over a period of time. If the desire to change gender persists, the patient's wish should be supported and may require the input of a specialist skilled in the management of gender change.

The process of disclosure concerning facts about karyotype, gonadal status, and prospects for future fertility is a collaborative, ongoing action that requires a flexible individual-based approach. It should be planned with the parents from the time of diagnosis.[70] Studies in other chronic medical disorders and of adoptees indicate that disclosure is associated with enhanced psychosocial adaptation.[71] Medical education and counseling for children is a recurrent gradual process of increasing sophistication that is commensurate with changing cognitive and psychological development.[72]

Quality of life encompasses falling in love, dating, attraction, ability to develop intimate relationships, sexual functioning, and the opportunity to marry and raise children, regardless of biological indicators of sex. The most frequent problems encountered in DSD patients are sexual aversion and lack of arousability, which are often misinterpreted as low libido.[73] Health care staff should offer adolescent patients opportunities to talk confidentially without their parents and encourage the participation in condition-specific support groups that enhance the ability of the patient to discuss their concerns comfortably. Some patients avoid intimate relationships, and it is important to address fears of rejection and advise them on the process of building a relationship with a partner. The focus should be on interpersonal relationships and not solely on sexual function and activity. Referral for sex therapy may be needed. Repeated examination of the genitalia, including medical photography, may be experienced as deeply shaming.[74] Medical photography has its place for record keeping and education but should be undertaken, whenever possible, when the patient is under anesthesia for a procedure. Medical interventions and negative sexual experiences may have fostered symptoms of posttraumatic stress disorder, and referral to a qualified mental health professional may be indicated.[75]

OUTCOME IN DSD

As a general statement, information across a range of assessments is insufficient in DSD. The following is based on those disorders for which some evidence base is available. They include CAH, CAIS, and PAIS, disorders of androgen biosynthesis, gonadal dysgenesis syndromes (complete and partial), and micropenis. Long-term outcome in DSD should include external and internal genital phenotype, physical health including fertility, sexual function, and social and psychosexual adjustment, mental health, quality of life, and social participation. There are additional health problems in individuals with DSD, including the consequences of associated problems such as other malformations, developmental delay and intellectual impairment, delayed growth and development, and unwanted effects of hormones on libido and body image.[76]

Surgical Outcome

Some studies suggest satisfactory outcomes from early surgery.[43,46,47,77] Nevertheless, outcomes from clitoroplasty identify problems related to decreased sexual sensitivity, loss of clitoral tissue, and cosmetic issues.[78] Techniques for vaginoplasty carry the potential for scarring at the introitus necessitating repeated modification before sexual function can be reliable. Surgery to construct a neovagina carries a risk of neoplasia.[79] The risks from vaginoplasty are different for high and low confluence of the urethra and vagina. Analysis of long-term outcomes is complicated by a mixture of surgical techniques and

TABLE 4 **Risk of Germ Cell Malignancy According to Diagnosis**

Risk Group	Disorder	Malignancy Risk, %	Recommended Action	Patients, *n*	Studies, *n*
High	GD[a] (+Y)[b] intraabdominal	15–35	Gonadectomy[c]	12	>350
	PAIS nonscrotal	50	Gonadectomy[c]	2	24
	Frasier	60	Gonadectomy[c]	1	15
	Denys-Drash (+Y)	40	Gonadectomy[c]	1	5
Intermediate	Turner (+Y)	12	Gonadectomy[c]	11	43
	17β-hydroxysteroid	28	Watchful waiting	2	7
	GD (+Y)[b] scrotal	Unknown	Biopsy[d] and irradiation?	0	0
	PAIS scrotal gonad	Unknown	Biopsy[d] and irradiation?	0	0
Low	CAIS	2	Biopsy[d] and ???	2	55
	Ovotesticular DSD	3	Testicular tissue removal?	3	426
	Turner (−Y)	1	None	11	557
No (?)	5αRD2	0	Unresolved	1	3
	Leydig cell hypoplasia	0	Unresolved	1	2

[a] Gonadal dysgenesis (including not further specified, 46,XY, 46,X/46,XY, mixed, partial, and complete).
[b] GBY region positive, including the TSPY (testis-specific protein Y encoded) gene.
[c] At time of diagnosis.
[d] At puberty, allowing investigation of at least 30 seminiferous tubules, preferentially diagnosis on the basis of OCT3/4 immunohistochemistry.

TABLE 5 **Genes Known to be Involved in DSD**

Gene	Protein	OMIM No.	Locus	Inheritance	Gonad	Müllerian Structures	External Genitalia	Associated Features/Variant Phenotypes
46,XY DSD								
Disorders of gonadal (testicular) development: single-gene disorders								
WT1	TF	607102	11p13	AD	Dysgenetic testis	+/−	Female or ambiguous	Wilms' tumor, renal abnormalities, gonadal tumors (WAGR, Denys-Drash and Frasier syndromes)
SF1 (NR5A1)	Nuclear receptor TF	184757	9q33	AD/AR	Dysgenetic testis	+/−	Female or ambiguous	More severe phenotypes include primary adrenal failure; milder phenotypes have isolated partial gonadal dysgenesis
SRY	TF	480000	Yp11.3	Y	Dysgenetic testis or ovotestis	+/−	Female or ambiguous	
SOX9	TF	608160	17q24-25	AD	Dysgenetic testis or ovotestis	+/−	Female or ambiguous	Camptomelic dysplasia (17q24 rearrangements; milder phenotype than point mutations)
DHH	Signaling molecule	605423	12q13.1	AR	Dysgenetic testis	+	Female	The severe phenotype of 1 patient included minifascicular neuropathy; other patients have isolated gonadal dysgenesis
ATRX	Helicase (? chromatin remodeling)	300032	Xq13.3	X	Dysgenetic testis	−	Female, ambiguous, or male	α-Thalassemia, mental retardation
ARX	TF	300382	Xp22.13	X	Dysgenetic testis	−	Ambiguous	X-linked lissencephaly, epilepsy, temperature instability
Disorders of gonadal (testicular) development: chromosomal changes involving key candidate genes								
DMRT1	TF	602424	9p24.3	Monosomic deletion	Dysgenetic testis	+/−	Female or ambiguous	Mental retardation
DAX1 (NR0B1)	Nuclear receptor TF	300018	Xp21.3	dupXp21	Dysgenetic testis or ovary	+/−	Female or ambiguous	
WNT4	Signaling molecule	603490	1p35	dup1p35	Dysgenetic testis	+	Ambiguous	Mental retardation
Disorders in hormone synthesis or action								
LHGCR	G-protein receptor	152790	2p21	AR	Testis	−	Female, ambiguous, or micropenis	Leydig cell hypoplasia
DHCR7	Enzyme	602858	11q12-13	AR	Testis	−	Variable	Smith-Lemli-Opitz syndrome: coarse facies, second-third toe syndactyly, failure to thrive, developmental delay, cardiac and visceral abnormalities
StAR (steroidogenic acute regulatory protein)	Mitochondrial membrane protein	600617	8p11.2	AR	Testis	−	Female	Congenital lipoid adrenal hyperplasia (primary adrenal failure), pubertal failure
CYP11A1	Enzyme	118485	15q23-24	AR	Testis	−	Female or Ambiguous	CAH (primary adrenal failure), pubertal failure
HSD3B2	Enzyme	201810	1p13.1	AR	Testis	−	Ambiguous	CAH, primary adrenal failure, partial androgenization caused by dehydroepiandrosterone sulfate

TABLE 5 **Continued**

CYP17	Enzyme	202110	10q24.3	AR	Testis	−	Female, ambiguous, or micropenis	CAH, hypertension caused by corticosterone and 11-deoxycorticosterone (except in isolated 17,20-lyase deficiency)
POR (P450 oxidoreductase)	CYP enzyme electron donor	124015	7q11.2	AR	Testis	−	Male or ambiguous	Mixed features of 21-hydroxylase deficiency, 17α-hydroxylase/17,20-lyase deficiency, and aromatase deficiency; sometimes associated with Antley Bixler craniosynostosis
HSD17B3	Enzyme	605573	9q22	AR	Testis	−	Female or ambiguous	Partial androgenization at puberty, androstenedione/testosterone ratio
SRD5A2	Enzyme	607306	2p23	AR	Testis	−	Ambiguous or micropenis	Partial androgenization at puberty, testosterone/dihydrotestosterone ratio
Anti-Müllerian hormone	Signaling molecule	600957	19p13.3-13.2	AR	Testis	+	Normal male	Persistent Müllerian duct syndrome; male external genitalia, bilateral cryptorchidism
Anti-Müllerian hormone receptor	Serine-threonine kinase transmembrane receptor	600956	12q13	AR	Testis	+	Normal male	
Androgen receptor	Nuclear receptor TF	313700	Xq11-12	X	Testis	−	Female, ambiguous, micropenis, or normal male	Phenotypic spectrum from CAIS (female external genitalia) and PAIS (ambiguous) to normal male genitalia/infertility
46,XX DSD								
Disorders of gonadal (ovarian) development								
SRY	TF	480000	Yp11.3	Translocation	Testis or ovotestis	−	Male or ambiguous	
SOX9	TF	608160	17q24	dup17q24	Not determined	−	Male or ambiguous	
Androgen excess								
HSD3B2	Enzyme	201810	1p13	AR	Ovary	+	Clitoromegaly	CAH, primary adrenal failure, partial androgenization caused by dehydroepiandrosterone sulfate
CYP21A2	Enzyme	201910	6p21-23	AR	Ovary	+	Ambiguous	CAH, phenotypic spectrum from severe salt-losing forms associated with adrenal failure to simple virilizing forms with compensated adrenal function, 17-hydroxyprogesterone
CYP11B1	Enzyme	202010	8q21-22	AR	Ovary	+	Ambiguous	CAH, hypertension caused by 11-deoxycortisol and 11-deoxycorticosterone
POR (P450 oxidoreductase)	CYP enzyme electron donor	124015	7q11.2	AR	Ovary	+	Ambiguous	Mixed features of 21-hydroxylase deficiency, 17α-hydroxylase/17,20-lyase deficiency, and aromatase deficiency; associated with Antley Bixler craniosynostosis
CYP19	Enzyme	107910	15q21	AR	Ovary	+	Ambiguous	Maternal androgenization during pregnancy, absent breast development at puberty, except in partial cases
Glucocorticoid receptor	Nuclear receptor TF	138040	5q31	AR	Ovary	+	Ambiguous	Adrenocorticotropin, 17-hydroxyprogesterone and cortisol; failure of dexamethasone suppression (patient heterozygous for a mutation in CYP21)

OMIM indicates Online Mendelian Inheritance in Man; TF, transcription factor; AD, autosomal dominant (often do novo mutation); AR indicates autosomal recessive; Y, Y-chromosomal; X, X-chromosomal. Chromosomal rearrangements likely to include key genes are included. Modified from Achermann JC, Ozisik G, Meeks JJ, Jameson JL. Genetic causes of human reproductive disease. *J Clin Endocrinol Metab*. 2002;87:2447-2454.

diagnostic categories.[80] Few women with CAIS need surgery to lengthen the vagina.[81]

The outcome in undermasculinized males with a phallus depends on the degree of hypospadias and the amount of erectile tissue. Feminizing genitoplasty as opposed to masculinizing genitoplasty requires less surgery to achieve an acceptable outcome and results in fewer urologic difficulties.[46] Long-term data regarding sexual function and quality of life among those assigned female as well as male show great variability. There are no controlled clinical trials of the efficacy of early (<12 months of age) versus late (in adolescence and adulthood) surgery or of the efficacy of different techniques.

Risk of Gonadal Tumors

Interpretation of the literature is hampered by unclear terminology and effects of normal cell-maturation delay.[82–84] The highest tumor risk is found in TSPY (testis-specific protein Y encoded) positive gonadal dysgenesis and PAIS with intraabdominal gonads, whereas the lowest risk (<5%) is found in ovotestis[85] and CAIS.[83,86] Table 4 provides a summary of the risk of tumor development according to diagnosis and recommendations for management.

Cultural and Social Factors

DSD may carry a stigma. Social and cultural factors, as well as hormonal effects, seem to influence gender role in 5αRD2 deficiency. Gender-role change occurs at different rates in different societies, suggesting that social factors may also be important modifiers of gender-role change.

In some societies, female infertility precludes marriage, which also affects employment prospects and creates economic dependence. Religious and philosophical views may influence how parents respond to the birth of an infant with a medical condition. Fatalism and guilt feelings in relation to congenital malformations or genetic conditions have an influence, whereas poverty and illiteracy negatively affect access to health care.[87]

FUTURE STUDIES

Establishing a precise diagnosis in DSD is just as important as in other chronic medical conditions that have lifelong consequences. Considerable progress has been achieved with molecular studies, as illustrated in Table 5, which summarizes the genes known to be involved in DSD. Use of tissue-specific animal knock-out models, comparative genomic hybridization, and microarray screens of the mouse urogenital ridge will provide benefits in identifying new genes causing DSD.[88] It is essential that the momentum for an international collaborative approach to this task be maintained.

Much remains to be clarified about the determinants of gender identity in DSD. Future studies require representative sampling to carefully conceptualize and measure gender identity, recognizing that there are multiple determinants to consider, and gender identity may change into adulthood. In terms of psychological management, studies are needed to evaluate the effectiveness of information management with regard to timing and content. The pattern of surgical practice in DSD is changing with respect to the timing of surgery and the techniques used. It is essential to evaluate the effects of early versus later surgery in a holistic manner, recognizing the difficulties posed by an ever-evolving clinical practice.

The consensus has clearly identified a major shortfall in information about long-term outcome. Future studies should use appropriate instruments that assess outcomes in a standard manner[68,69] and take cognizance of guidelines relevant to all chronic conditions (see www.who.int/classifications/icf/en). These studies would preferably be prospective in nature and designed to avoid selection bias. A number of countries already have registers of DSD cases, but there could be added benefit from pooling such resources to enable prospective, multicenter studies to be undertaken on a larger number of cases that are clearly defined. Allied to this should be an educational program to ensure that multiprofessionals tasked with providing care to families with a child with DSD are suitably trained to discharge their responsibilities.

APPENDIX 1: ROLE OF SUPPORT GROUPS

The value of peer and parent support for many chronic medical conditions is widely accepted, and DSD, being lifelong conditions that affect developmental tasks at many stages of life, are no exception.

Those affected by DSD and parent members value the following:

- Peer support ends isolation and stigma, providing a context in which conditions are put into perspective and intimate issues of concern can be discussed safely with someone who has "been there."
- Children who form relationships with peers and affected adults early in their lives benefit from a feeling of normalcy early on, with support in place well before adolescence. Adolescents often resist attempts to introduce them to peer support.
- Support groups can help families and consumers find the best quality care.

Although clinical practice may focus on gender and genital appearance as key outcomes, stigma and experiences associated with having a DSD (both within and outside the medical environment) are more salient issues for many affected people.

Support groups complement the work of the health care team and, together, can help improve services. Initiatives by support groups have led to improvements in

management of DSD and research directed toward clinically relevant issues. Dialogue between health care professionals and support groups and collaboration as partners is to be encouraged.

APPENDIX 2: LEGAL ISSUES

Basic principles of medical law will remain even as research and clinical experience evolve in etiology, diagnosis, and treatment. This Appendix draws on practice in 3 countries on standards of medical negligence and patient informed consent. In the United States, the medical profession sets standards of care on the basis of prevailing medical custom.[89] However, a treatment may also be that used by a respected minority of practitioners.

Informed consent in the United States was founded on the principle of battery, whereby it is an offense to violate another person's bodily integrity without consent. Nowadays, most states are concerned with negligent nondisclosure to the patient. The standard of adequate disclosure may be physician based, requiring conduct of a reasonable practitioner, or it may be patient based, asking what a reasonable patient would find material. Physician-based disclosure must include information about risks, alternatives, outcomes, and prognosis, with or without treatment.

US courts assume that parents know what is best for their child when parental authority applies to consent for the child (substituted judgment). Parental decisions are deferred to except in situations in which potentially life-saving treatment is withheld. Consent to treatment by a child depends on an understanding of its nature and consequences.

Medical negligence in the United Kingdom defines treatment that falls below the standard expected of a reasonably competent practitioner. The standard of proof in court is whether negligence is demonstrated on the balance of probabilities. It is incumbent on the practitioner to demonstrate that treatment was consistent with a rationally defensible body of medical opinion. A shift in parental prerogative to consent to treatment was reflected in the Children Act 1989 in which parental rights were replaced by parental responsibilities. United Kingdom courts can intervene with orders made requiring or preventing a specific action related to the child. Age is not a barrier to informed consent, providing that a minor demonstrates an understanding of the issues sufficient to have the capacity to consent.

Colombian law is noted for a reasoned set of guidelines advanced by the highest court in cases of DSD.[90] A protocol was formulated for parental and physician intervention. The process of consent requires "qualified and persistent informed consent" over an extended period of time. Authorization is given in stages to allow time for the parents to come to terms with their child's condition. The court aimed to strike a balance between parental autonomy for those who did and those who did not want early surgery for their child until there was clear evidence of harm in deferring surgery until the child was competent to decide. Parents cannot consent for children over 5 years of age, because by then, children are deemed to have identified with a gender and, thus, are considered to be autonomous.

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Paradigms Revised: Intersex Children, Bioethics & the Law

Laura Hermer
UTMB-Galveston

Paradigms Revised: Intersex Children, Bioethics & The Law

*Laura Hermer, J.D.**

INTRODUCTION

In November 2000, a four day-old infant lay supine on the operating table at a children's hospital in the United States, draped and prepped for surgery. The child's problem lay exposed to the view of the urology residents who crowded the operating room: rather than male or female genitalia, the child had a tiny phallus with a urethral opening at its base, two bifurcated scrota which appeared to form a labia minora, and a vagina which ended blindly, rather than leading to a uterus. The child's gonads were nowhere to be found. The surgical task was to find the gonads and take a sample of them to determine, among other matters, whether they were comprised of testicular or ovarian material. Upon analysis of the specimen, the laboratory found both types of tissue. This infant was neither male nor female, but "intersex."

Intersex individuals are classically defined as having both male and female sexual characteristics.[1] Researchers estimate that intersex conditions may affect up to one out of every 2,000 children born.[2] The notion of an intersex individual may bring to mind an image of Hermaphroditus, the fabled child of Aphrodite and Hermes who had the complete external genitalia of both a man and a woman. However, such cases are purely

* Laura Hermer is a Postdoctoral Research Fellow at the Institute for Medical Humanities, UTMB-Galveston. Ms. Hermer received her Juris Doctor from Northeastern University, and is an L.L.M. Candidate in Health Law at the University of Houston. I would like to thank my husband, Lars Cisek, M.D., Ph.D., for first bringing the medical issue to my attention. This essay was awarded first place in the 2002 Robert M. Toth Health Law Essay Contest.

1. Julia S. Barthold & Ricardo Gonzalez, *Intersex States*, *in* PEDIATRIC UROLOGY PRACTICE 547 (E. Gonzales ed. 1999).

2. Melanie Blackless et al., *How Sexually Dimorphic Are We?*, 12 AM. J. HUM. BIO. 151-66 (2000); Intersex Society of North America *at* http://www.isna.org/faq/frequency.html (last visited May 14, 2002) (providing estimates of the frequency of specific intersex conditions among births, viewed on November 20, 2001). A review of the medical literature performed by Anne Fausto-Sterling suggests that children with intersex conditions comprise 1.7 percent of all births. ANNE FAUSTO-STERLING, SEXING THE BODY 51-53 (2000).

mythological. Intersex individuals instead have "ambiguous" genitalia. While the external genitalia may appear male, for example, the person also possesses ovaries rather than testes, and a functional uterus. Or a person may have a bifurcated scrotal sac/vulva and a urethral opening at the base of what appears to be a small, curved phallus, but, upon surgical exploration, has undescended testes and no female reproductive organs. While identification of a child as intersex can often be made by physical examination alone, in some cases normal-appearing external genitalia can hide an internal ambiguity or an anomalous chromosomal sex. As a result, families and physicians face a dilemma concerning how to treat such infants. Prior to the advent of modern surgery, such individuals were left as they were born. Some did not survive, depending on their respective medical conditions and urogenital structures. Enough lived to present perplexities in law and society; for example, the *Talmud* and *Tosefta* contain regulations for people of mixed sex,[3] and in 16th century England, Lord Coke declared with respect to the law of inheritance that "a hermaphrodite may be either male or female, and it shall succeed according to the kind of sex that doth prevail."[4]

Starting in the 1950's, once surgical practice became sufficiently sophisticated, physicians commonly believed the best practice with respect to such individuals was to assign them surgically to an "appropriate" sex prior to the age of two, if not earlier.[5] In many cases, physicians believed it was so important for parents to be able to identify a child as male or female at birth, based on the appearance of the child's sex organs, that they would suggest immediate surgical reassignment.[6] In conjunction with the surgery, parents were counseled to raise their child in strict adherence to convention in accordance with the

3. *See* FAUSTO-STERLING, *supra* note 2, at 33.

4. *See* JOHN MONEY, SEX ERRORS OF THE BODY AND RELATED SYNDROMES 3 (2d ed. 1994).

5. *See, e.g.*, Hazel Glenn Beh & Milton Diamond, *An Emerging Ethical and Medical Dilemma: Should Physicians Perform Sex Assignment Surgery on Infants with Ambiguous Genitalia?*, 7 MICH. J. GENDER & L. 1, 2-3 (2000).

6. *Cf.* Evan Kass et al., *Timing of Elective Surgery on the Genitalia of Male Children with Particular Reference to the Risks, Benefits, and Psychological Effects of Surgery and Anesthesia*, 97 PEDIATRICS 590 (1996) available at LEXIS, Medical & Healthcare Journals Library, Pediatrics File (noting that "opportunities for establishing a strong and stable mother-father-infant relationship must be fostered" during the first year of life, and that surgery is indicated at as early as six weeks of age).

chosen sex assignment, in order to ensure that the child's gender identity matched its assigned sex.[7]

For decades, very few researchers studied the psychosocial and psychosexual outcomes of these children as they matured to adulthood.[8] On the basis of several limited follow-up studies performed in the 1960's, it was assumed that gender was primarily a function of societal conditioning, rather than biological determination.[9] As such, most physicians believed that an individual assigned to a given sex, if raised appropriately and sufficiently surgically modified, would be assured a reasonable outcome; i.e., the individual would identify with the assigned sex and would become a heterosexual within that assignment.[10]

In the late 1990's, however, this assumption was knocked askew by the revelation that the most prominent research subject from the sex reassignment studies had definitively rejected his female assignment and was now married to a woman and living as a male in Canada, despite the fact that his penis had been ablated in a surgical accident in infancy, and his testes had been surgically removed in the subsequent effort to make him a female.[11] Contemporaneously, a small but vocal group of other individuals who had undergone cosmetic genital or sex assignment surgery in infancy and childhood came forward to demand a moratorium on such surgeries. Some of these individuals had rejected their assigned sex; others protested their loss of sexual sensation and function. These individuals found the surgeries to be unreasonable invasions of their bodily integrity and psychosexual and psychosocial identities.

The management of intersex infants and children is presently enmeshed in controversy. While the American Academy of Pediatrics still recommends sex assignment surgery and certain

7. *See, e.g.*, Kass et al., *supra* note 6. For example, Beh and Diamond note that the family of one boy who had been surgically reassigned as a girl were instructed not only to strictly raise the child as a girl, but also to move to another city in order to help keep the child's birth sex a secret. Beh & Diamond, *supra* note 5, at 7.

8. John Money, whose research is discussed in Part I, *infra*, performed one of the only such studies prior to the 1960's. *See* John Money et al., *Imprinting and the Establishment of Gender Role*, 77 ARCH. NEUROL. PSYCHIATRY 333-36 (1957).

9. Dr. Money published his results in the early 1970's. *See* JOHN MONEY & A. EHRHARDT, MAN AND WOMAN, BOY AND GIRL (1972).

10. This belief was held as recently as 1996. *See* Kass et al., *supra* note 6.

11. *See* Milton Diamond & H. Keith Sigmundson, *Sex Reassignment at Birth: Long-Term Review and Clinical Implications,* 151 ARCH. PEDIATR. ADOLESC. MED. 298, 298 (1997).

cosmetic genital surgeries in infancy,[12] a growing number of physicians and other health professionals are suggesting that, in many cases, surgical revision should wait until the child comes of age and can decide for itself whether to undergo surgery directed towards achieving male or female appearance and "function" or remain as it is.[13]

The controversy branches into several legal arenas. This paper will focus on two in particular, both of which may impact future medical practice concerning cosmetic genital and/or sex assignment surgeries as performed on intersex infant and children.[14] The first area is that of medical malpractice. What right, if any, do intersex individuals have to recover for surgeries performed upon them? If no such right exists, should they have any such right? The second area is that of informed consent. Some commentators argue that surgeries on intersex children were (and may still be) regularly performed with serious defects in informed consent. Given this history, should the practice of early cosmetic genital and sex assignment surgeries be allowed to continue? If so, then under what circumstances? Also, given that the surgery has such potentially major ramifications on the child's social and sexual identity, should parents be permitted to consent for non-emergent surgery on behalf of an intersex child?

In one of the few legal articles addressing the issue of intersex surgeries, Hazel Beh and Milton Diamond (the latter being the Honolulu professor of anatomy and reproductive biology who first uncovered and publicized the rejection of one research subject's sex reassignment) evaluate medical malpractice law and the law of informed consent as a means of controlling and/or ceasing the practice of cosmetic genital and sex assignment surgeries.[15] They ultimately conclude that deficiencies in informed consent warrant a moratorium on such surgeries.[16]

12. *See* American Academy of Pediatrics, *Evaluation of the Newborn with Developmental Anomalies of the External Genitalia*, 106 PEDIATRICS 138, 138 (2000) *available at* LEXIS, Medical & Healthcare Journals Library, Pediatrics File.

13. See, e.g., Kenneth Kipnis & Milton Diamond, *Pediatric Ethics and the Surgical Assignment of Sex*, 9 J. CLINICAL ETHICS 398 (1998); Bruce E. Wilson & William G. Reiner, *Management of Intersex: A Shifting Paradigm*, 9 J. CLINICAL ETHICS 360 (1998).

14. For a discussion of other legal issues, such as the right to marry and alter identifying legal documents, *see* Julie A. Greenberg, *Defining Male and Female: Intersexuality and the Collision Between Law and Biology*, 41 ARIZ. L. Rev. 265 (1999).

15. Beh & Diamond, *supra* note 5, at 2-3.

16. *Id.* at 56–59.

This extreme prescription, however, is likely as shortsighted as the prior practice of reflexively performing cosmetic surgeries on most intersex children in infancy. On the one hand, use of the legal system to interdict these surgeries may be necessary if one desires a rapid end to them. The other two alternatives, waiting for change in medical practice and using political agitation, will not likely achieve such a result in a short period of time. And political agitation, while useful in quickly bringing an issue to the attention of the public, will likely have only limited success in bringing the practice to an end without some assistance from the legal arena. On the other hand, however, the legal system is ill-equipped to deal with the cultural and social issues underlying intersex surgeries. Failing to come to terms with these issues threatens to leave in place the norms which brought about the surgical practices in the first place.

While certain changes in the management and treatment of intersex individuals are undoubtedly warranted, an absolute moratorium on childhood cosmetic genital and sex assignment surgeries cannot be justified. This paper advocates a middle approach to the treatment of intersex individuals, one which takes account not only of the concerns of intersex activists, but also of issues concerning medical research, family dynamics, and social and cultural considerations. Part I of this paper provides an overview of sex and gender. It briefly sketches part of the outline of the debate over what constitutes sex versus gender, and places intersex individuals within its context. Part II discusses intersex conditions, their present treatment and outcomes. Part III evaluates proposed methods of enlisting the legal system in modifying the present treatment regimen of intersex individuals. It concludes that the proposed legal solutions are inadequate to resolve the matter, as they do not address the social and cultural issues which underlie the present management and treatment of intersexuals. Without squarely facing and treating such issues, any alteration to the current treatment paradigm will be cosmetic at best. Thus, as developed in Part IV, the alterations proposed in this paper take into account not merely deficiencies in present data concerning treatment outcomes and legal deficiencies with prevailing treatment protocols, but also the mores which influenced the adoption of current treatment regimens and the social and psychological needs of intersex children and their families.

I. Overview of Sex and Gender

At least in recent centuries in the western world, sex traditionally has been considered to be biologically determined.[17] Conventionally, one's sex is determined by being born with a certain set of reproductive organs. This test has the ease of simplicity: what one finds between the legs determines one's sex. Physicians, midwives and farmers have used it for millennia. Chromosomal analysis has only recently entered into the picture, but now also has a standardized place; rather than using the visual test, one can instead do a chromosomal analysis and see whether one's chromosomal makeup, or "karyotype," is 46,XX, which usually yields a person with the reproductive organs and secondary sex characteristics of a female, or 46,XY, which usually yields a person with the reproductive organs and secondary sex characteristics of a male.[18] As for gender, under the traditional analysis, gender is simply the sociocultural manifestation of one's sex. The traditional definitions result in a binary system, in which a person is destined to be a man or a woman – socially, biologically and experientially – based on the sexual organs which he or she possesses.

In recent decades, however, certain theorists have questioned the traditional concepts of both sex and gender. Some posit that our definition of sex, rather than being scientifically objective, has a culturally-constructed component.[19] From this perspective, one might think of the body as "the point of intersection, as the interface between the biological and the social."[20] Sex is not merely a biological given; rather, it is conditioned by our perception of it within our historical and cultural context. This point becomes clearer when one considers how to apply the traditional definition of sex to an individual whose gonads do

17. For a detailed discussion of changes over time in the medical definition of sex in England and France, *see* Alice Domurat Dreger, Hermaphrodites and the Medical Invention of Sex (1998).

18. *See, e.g.*, Keith L. Moore, The Developing Human: Clinically Oriented Embryology 272 (3rd Ed. 1982).

19. *See, e.g.*, Michel Foucault, The History of Sexuality: An Introduction 154-55 (R. Hurley, trans.) (1978); Fausto-Sterling, *supra* note 2, at 23 ("To talk about human sexuality requires a notion of the material. Yet the idea of the material comes to us already tainted, containing within it preexisting ideas about sexual difference").

20. Rosi Braidotti, *The Politics of Ontological Difference*, *cited in* Lois McNay, Foucault & Feminism 24 (1992). *See also, e.g.*, Susan Bordo, The Male Body (1999) (noting, in her discussion of clothing, nakedness and masculinity, "we need to think about the body not only as a physical entity – which it assuredly is – but also as a cultural form that carries meaning with it").

not match his or her external genitalia, or who has a non-standard set of chromosomes. Can sex be determined solely by one's gonads, or by the external appearance of one's sex organs, or by one's chromosomes? How does one choose which criteria to use? Is it perhaps defined by some combination of the above? Who determines what sex is, and the contexts in which it is appropriate to define it?

Correspondingly, some philosophers and cultural theorists posit that gender is created by sociocultural and other factors, rather than being an outgrowth of sex.[21] Gender does not merely denote the manner in which one manifests whether one is male or female. Such a definition would limit gender to a description of the range of appearances, behaviors and interactions which a given society deems to be "male" or "female." Rather, gender encompasses the entire means by which these appearances, behaviors and interactions come into being in a given sociocultural setting.[22] Additionally, some theorists argue that, because gender is socioculturally produced, there is no necessary relation between sex and gender. In this context, "[g]ender becomes a free-floating entity with the consequence that 'man and masculine might as easily signify a female body as a male, and women and feminine a male body as easily as a female one.'"[23]

Against these theoretical backdrops, what does one make of intersex individuals? If gender is based on one's sex, and – a

21. *See, e.g.*, JUDITH BUTLER, GENDER TROUBLE: FEMINISM AND THE SUBVERSION OF IDENTITY 7 (1990).

22. *See, e.g., id.* Whether or not one agrees with the foregoing proposition, the following brief survey of bygone gender norms helps demonstrate the fluid, even arbitrary nature of gender:

> Female inverts were described in the literature as possessing "masculine straightforwardness and sense of honor" (Ellis, 1942, p. 250), having "a dislike and sometimes incapacity for needlework" as well as "an inclination and taste for the sciences" (Krafft-Ebing, 1893, p. 280), being demanding of voting rights, and skillful at whistling (Browne, 1923; Claiborne, 1914; Ellis, 1942). Accounts of male inverts include such descriptors as, "sentimental," "something of a chatterbox" (Carpenter, 1911, p. 132), "never smoked "entirely averse to outdoor games," and having a "fondness for cats" (Rivers, 1920, p. 22). Krafft-Ebing (1893) noted that this "abnormality of feeling and of development of the character [was] often apparent in childhood" (p. 279). On one such case, he wrote that "the boy likes to spend his time with girls, play with dolls, and help his mother around the house" (Krafft-Ebing, 1893, p. 279).

Nancy H. Bartlett et al., *Is Gender Identity Disorder in Children a Mental Disorder?*, 43 SEX ROLES: A JOURNAL OF RESEARCH (2000).

23. MCNAY, *supra* note 20, at 23.

crucial assumption – if one's sex is determined by the appearance of one's external genitalia, then under the traditional theory, sex assignment surgery should yield healthy individuals who identify appropriately with their assigned sex and gender of rearing. If, at the opposite extreme, gender is solely a sociocultural construct and has no necessary relation to sex, it would seem that an individual who was reared in an ideal social setting (another crucial postulate) to become a particular gender would, notwithstanding his or her physical or chromosomal sex, identify more with his or her gender of rearing rather than with the gender traditionally corresponding to the person's physiological or chromosomal sex. In such an idealized case, sex assignment surgery, which is supposed to make a person's genital appearance correspond with their assigned sex and gender, would be icing on the cake, a non-essential trapping to help reinforce the gender of rearing.[24]

Yet the problem is by no means this simple. Children who were surgically assigned to one sex in infancy and raised according to the reassignment have rejected it in adulthood. Most notoriously, this occurred to the subject of the most famous sex reassignment case in the medical literature. Until the individual's actual outcome had been reported, this case had formerly been the cornerstone on which the theory that a child could be successfully reared as either a boy or girl following sex assignment surgery was based. In this case, a surgeon accidentally burned "John's" penis so badly during a circumcision at the age of seven months that the majority of it was completely destroyed.[25] Before this time, John had been an otherwise normal boy. On the advice of John Money, then a psychologist at Johns Hopkins University, his parents agreed to have John surgically reassigned as a girl ("Joan").[26] Following the accident and reassignment, Dr. Money counseled the child's parents as to Joan's upbringing as a girl, and advised them never to disclose to Joan that s/he had been a boy.[27]

24. Due to the myriad different pressures society puts on individuals to conform with various gender norms based on the person's perceived sex, however, and given multiple other factors at play (such as the child's own preferences and the family's role in reinforcing or problematizing the child's assigned gender), it is unlikely that any social constructivist theorist would postulate that the outcome of rearing a child according to one gender norm or another could be guaranteed.

25. John Colapinto, *The True Story of John/Joan*, ROLLING STONE 54-97 (Dec. 11, 1998).

26. *Id.*

27. Beh & Diamond, *supra* note 5, at 7; *see also* Colapinto, *supra* note 25.

According to Money, other than displaying some tomboyishness, Joan had accepted his/her reassignment.[28] Money cited the case as a success, and, largely on its basis, recommended sex assignment surgery or surgery to "normalize" the genitals for intersex individuals in infancy.[29] If an otherwise normal boy could be successfully reared as a girl following the appropriate surgery, then – the theory went – such a strategy should surely make sense with respect to intersex children, whose sexual appearance is frequently ambiguous. Physicians and families could feel secure, based on this case, that their decision to assign an intersex child to a given sex in infancy would ultimately be in the child's best interest, and that the child would most likely grow up to be a sexually and psychosocially well-adjusted individual.

However, Money failed to publish signs of trouble in his reports on Joan. Joan eventually refused to participate in his/her counseling sessions.[30] S/he insisted in standing to urinate.[31] When s/he was given estrogen at the age of twelve in order to stimulate development of breasts, widened hips and other female secondary sex traits, Joan refused to take the hormone.[32] S/he thought s/he was a "freak," and began contemplating suicide.[33] At the age of fourteen, Joan finally learned the truth about his/her sex at birth from his/her father.[34] Immediately after hearing this news, Joan began living as a boy.[35] When Dr. Diamond, a critic of Dr. Money, found him again in 1994, John was living as a man, had married a woman, and had adopted the woman's three children.[36] John's true outcome did not become published until 1997.[37]

Notwithstanding the frank failure of John's sex reassignment, John's case had, up until that date, profoundly influenced the standard of care for treating intersex children since the 1960's in favor of surgical assignment in infancy.[38] Based on the case, which Dr. Money reported in 1972, a standard of care developed in which infants with ambiguous genitalia were surgically as-

28. Diamond & Sigmundson, *supra* note 11.
29. *See, e.g.*, Beh & Diamond, *supra* note 5, at 9.
30. *See* Diamond & Sigmundson, *supra* note 11.
31. *Id.*
32. *Id.*
33. *Id.*
34. *Id.*
35. *Id.*
36. Diamond & Sigmundson, *supra* note 11, at 300.
37. Beh & Diamond, *supra* note 5, at 9-10.
38. *See id.* at 12.

signed as a boy or girl as soon as possible after birth.[39] As late as 1996, the American Academy of Pediatrics Action Committee on Surgery stated that "children whose genetic sexes are not clearly reflected in external genitalia (i.e., hermaphroditism) can be raised successfully as members of either sex if the process begins before the age of two and one-half years. Therefore, a person's sexual body image is largely a function of socialization."[40] All the works to which the committee cited in support of the proposition were co-authored by Dr. Money. Thus, so as "[t]o prevent the development of cross-gender identification in children born with a physical intersex condition . . . early sex assignment and early correction of their genitalia" was typically considered necessary.[41]

II. Intersex Conditions: Current Treatments and Outcomes

Intersex conditions are myriad in number and type; virtually all develop in utero.[42] Around the age of six weeks, an embryo develops undifferentiated gonadal tissue, which may become male or female, depending on the presence or absence of certain genetic and hormonal factors. In the absence of these factors, an embryo will develop into a female, but in their presence, it will develop into a male. Intersex conditions can develop when an abnormality develops with respect to the fetus' sex chromosomes and/or hormones.[43]

Intersex conditions may be classified in a number of different ways. Most frequently, the medical literature – as a residual of the former primacy of gonads in defining sex – groups intersex individuals into "true hermaphrodites" and "pseudohermaphrodites."[44] True hermaphrodites are characterized as having both ovarian and testicular tissue. Pseudohermaphrodites, on the other hand, have gonadal tissue of only one type. The literature may also group those with mixed gonadal dysgenesis separately from other pseudohermaphrodites. Individuals with mixed gon-

39. *Id.* at 16.
40. Kass et al., *supra* note 6.
41. Froukje M E Slijper et al., *Long-Term Psychological Evaluation of Intersex Children*, 27 Arch. Sex. Behavior 125, 127 (1998).
42. The only exceptions are those created by surgical mistake, such as the circumcision disaster which befell John.
43. *See* American Academy of Pediatrics, *supra* note 12.
44. *See* Dreger, supra note 17, at 145-50; Fausto-Sterling, *supra* note 2, at 37-39.

adal dysgenesis have at least one immature or undifferentiated ("streak") gonad. The following provides examples of a number of the more common causes of intersex conditions potentially leading to ambiguous genitalia.

A. True Hermaphroditism

True hermaphrodites may be of a number of different karyotypes: 46,XX, 46,XX(or XO)/46XY mosaic, or 46XY karyotype, among others.[45] Such children usually have ambiguous genitalia. A constant or near-constant (eighty percent to one hundred percent of all cases) feature is the possession of a (frequently abnormal) uterus and vagina.[46] The gonads of a true hermaphrodite are either ovotestes or a combination of ovary, testis and/or ovotestis.[47] Almost all are infertile as males, and most – although not all – are infertile as females.[48] Presently, true hermaphroditic children are raised either as male or female (with more than seventy-five percent presently raised as male).[49] Recently, some researchers have advocated rearing them as females, given the slightly increased possibility for childbearing.[50] Depending on choice of gender, the incompatible portions of the child's gonads are usually removed, in order to avoid potential malignancies, as well as complications at puberty such as gynecomastia (development of female post-pubertal breasts) in males.[51]

B. Pseudohermaphroditism

Children with pseudohermaphroditism may have ambiguous genitalia, although, unlike true hermaphrodites, they possess only testicular or ovarian gonadal tissue, rather than a mixture. Pseudohermaphroditic conditions are usually linked to either chromosomal or endocrinological issues.

The most common pseudohermaphroditic (and intersex) condition is congenital adrenal hyperplasia (CAH), when it appears in chromosomal females. One study suggests that individuals

45. Barthold & Gonzalez, *supra* note 1, at 556.
46. *Id.*
47. *Id.*
48. *Id.* (noting that 21 pregnancies have been reported in true hermaphrodites, almost all of whom have had at least one normal ovary and a normal uterus).
49. *Id.*
50. Barthold & Gonzalez, *supra* note 1, at 556.
51. *See id.*

with CAH comprise over 1.5 percent of all births.[52] Females with this condition generally have a normal female karyotype, and a normal uterus and ovaries. However, due to a congenital defect in the adrenal gland which causes it to produce high amounts of androgens, they develop partially or completely masculinized external genitalia (i.e., they possess a phallus which is longer than the average clitoris and may even be "penis-sized," and may have no externally-apparent vagina). Most karyotypically female children are presently raised as females in America, although those with fully masculinized external genitalia are occasionally raised as males.[53]

Another relatively common condition is androgen insensitivity syndrome. Children with this condition are karyotypically 46,XY, and thus chromosomally male, but are either partially or totally insensitive to androgens.[54] Because of the insensitivity, these children appear to the observer to be partially or completely feminized. Those with complete androgen insensitivity syndrome (CAIS) have the external genitalia and, after puberty, secondary sex characteristics of a woman.[55] However, they usually also have inter-abdominal testes, a blind-ending vagina, and lack a uterus and ovaries.[56] Virtually all such individuals are raised as girls, and in fact are usually not identified as having CAIS until puberty, when they fail to menstruate.[57] Children with partial androgen insensitivity syndrome (PAIS) vary in their genital development in a spectrum from complete external feminization to virilization producing hypospadias (a condition in which the penis is very short, squat and/or curved and the urethra exits from the shaft or base rather than from the glans,

52. FAUSTO-STERLING, *supra* note 2, at 53.

53. Barthold & Gonzalez, *supra* note 1, at 559-60; *see also* Jaime Frias et al., *Technical Report: Congenital Adrenal Hyperplasia*, 106 PEDIATRICS 1511 (2000) *available at* LEXIS, Medical & Healthcare Journals Library, Pediatrics File. One form of CAH can cause a life-threatening metabolic crisis within day or weeks of birth. Thus, a diagnosis of CAH can be a true *medical* – not surgical – emergency. The crisis is forestalled by cortisol injections. Cosmetic genital surgery plays no role in this treatment. *Id.*

54. *See* Amy B. Wisniewski et al., *Complete Androgen Insensitivity Syndrome: Long-Term Medical, Surgical, and Psychosexual Outcome*, 85 J. CLINICAL ENDOCRIN. & METAB. 2664 (2000).

55. *Id.*

56. Barthold. & Gonzalez, *supra* note 1, at 564.

57. *See, e.g.*, Wisniewski et al., *supra* note 54 (noting additionally that 100 percent of their adult CAIS study participants expressed satisfaction with their female sex of rearing).

among other abnormalities) and partial scrotal development.[58] PAIS children may experience further virilization at puberty, or may develop breasts, depending on the degree of their androgen insensitivity.[59] They may be raised as either boys or girls.[60]

Another endocrine disorder is 5-alpha reductase deficiency. This hereditary deficiency, seen in chromosomal males, causes moderate to severe genital ambiguity *in utero*, often including a significantly small penis with severe hypospadias, variable degrees of scrotal development, and undescended testes.[61] They are frequently raised as females, prior to puberty. If untreated, these children become masculinized at puberty, with moderate penile growth, testicular descent and, generally, a change in gender identity from female to male (although a minority retain their female gender).[62] They are usually infertile.[63]

Exstrophy, a major but rare congenital anomaly, may affect both chromosomal males and females.[64] In children with exstrophy, the abdominal wall over the bladder as well as the bladder itself, the urethra, and the penis or clitoris are split in two.[65] In males, the interior of the bladder and of the penis and urethra are open and visible. In females, the clitoris is duplicated and there may be other minor genital anomalies. Early surgery is necessary in order for the child's urinary tract to function.[66] Because of the particularly small penile size occurring in certain forms of the anomaly, males with exstrophy may be surgically reassigned as females.[67]

Rarely, males who are otherwise karyotypically and endocrinologically normal are born with a micropenis or without any penis at all. The condition can be caused by the torsion and death of the children's testes during their descent *in utero*, prior

58. Barthold. & Gonzalez, *supra* note 1, at 564.

59. *Id.*

60. *Id.* at 564-65.

61. *See, e.g.*, Berenice B. Mendonca et al., *Male Pseudohermaphroditism Due to Steroid 5-Alpha-Reductase 2 Deficiency: Diagnosis, Psychological Evaluation, and Management*, 75 MEDICINE 64 (1996).

62. *Id.*

63. *Id.*

64. *See* William G. Reiner et al., *Psychosexual Dysfunction in Males with Genital Anomalies: Late Adolescence, Tanner Stages IV to VI*, 38 J. AM. ACAD. CHILD & ADOLES. PSYCHIATRY 865 (1999), *available at* 1999 WL 11376171.

65. *Id.*

66. *Id.*

67. *Id.*

to full penile development.[68] While some boys with a micropenis are presently raised as a male, others, as well as boys born without any penis at all, are often surgically reassigned to the female sex in infancy.[69]

C. *Gonadal Dysgenesis*

Children with gonadal dysgenesis generally have some combination of streak or absent gonad and dysgenetic testis or ovary.[70] The condition may manifest in a number of different ways. In Swyer's Syndrome, the child is chromosomally 46,XY, however, due to problems with the Y chromosome, the child fails to develop testes.[71] Such a child, although karyotypically male, appears phenotypically female, and usually is raised as such.[72] As with CAIS, the child's condition may not be discovered until adolescence, when the child fails to start menstruating.[73]

In children with partial gonadal dysgenesis, one most often finds hypospadias with cryptorchidism (missing testes), or another form of ambiguous genitalia.[74] Occasionally, one finds fully masculinized external genitalia, however, such children also have a uterus and at least one fallopian tube.[75] These individuals are, under present management, often raised as females.[76]

D. *Treatment*

The treatment of intersex conditions is currently undergoing revision. Previously, the birth of a child with an intersex condition was considered an emergency requiring the immediate determination of a sex of rearing and the first surgery to establish the child in that sex. A quote from a 1969 treatise on the subject is instructive as to the attitude taken towards such children:

68. Interview with Dr. Lars J. Cisek, Assistant Professor of Pediatric Urology, Baylor University, Houston, Tex. (Nov. 27, 2001) [hereinafter Cisek Interview].
69. Barthold & Gonzalez, *supra* note 1, at 566-67. Incidentally, there apparently is significant variation in the results of measurement of stretched penile length from one clinician to another. *See, e.g.*, Michael L. Ritchey & David Bloom, *Summary of the Urology Section*, 96 PEDIATRICS 138 (1995), *available at* LEXIS, Medical & Healthcare Journals Library, Pediatrics File.
70. Barthold & Gonzalez, *supra* note 1, at 550.
71. *Id.*
72. *Id.* at 550-52.
73. *Id.*
74. *Id.* at 555
75. *Id.*
76. *Id.*

> [The] normal functioning [of sex] is vital to the survival of our race, essential for our full assimilation as individuals into society, and pervades every aspect of our lives. To visualize individuals who properly belong neither to one sex nor to the other is to imagine freaks, misfits, curiosities, rejected by society and condemned to a solitary existence of neglect and frustration. Few of these unfortunate people meet with tolerance and understanding from their fellows and fewer still find even limited acceptance in a small section of society: all are constantly confronted with reminders of their unhappy situation. The tragedy of their lives is the greater since it may be remediable; with suitable management and treatment, especially if this is begun soon after birth, many of these people can be helped to live happy well-adjusted lives, and some may even be fertile and be enabled to enjoy a normal family life.[77]

As recently as 1996, the American Academy of Pediatrics (AAP) espoused a treatment regimen arguably deriving from this perspective, in which sex assignment or genital normalizing surgery was recommended to be undertaken as early as possible, ideally between six weeks and fifteen months of age.[78]

The AAP recently issued new guidelines for the evaluation and treatment of intersex conditions.[79] According to these guidelines, the birth of a child with an intersex condition constitutes a "social emergency" – but notably not a surgical one.[80] Contrary to their prior recommendations, the new AAP guidelines suggest that physicians refrain from suggesting a diagnosis or gender assignment at birth, and that parents refrain from registering the birth until a sex of rearing is established.[81] The guidelines suggest that the following factors should be used in determining the sex of rearing: (1) fertility potential; (2) capacity for normal sexual function; (3) endocrine function; (4) potential for malignant gonadal change; and (5) testosterone imprinting.[82] Fertility is usually only an issue with respect to girls with CAH (although assisted reproduction techniques may be able to broaden this).[83] Despite the presence of externally-male genitalia, chromosomally-female children with CAH usu-

77. CHRISTOPHER J. DEWHURST & RONALD R. GORDON, THE INTERSEXUAL DISORDERS vii (1969).
78. Kass et al., *supra* note 6.
79. *See* American Academy of Pediatrics, *supra* note 12.
80. *Id.*
81. *Id.*
82. *Id.*
83. *Id.*; Cisek Interview, *supra* note 68.

ally have fully-formed and fully-functional uteruses and ovaries, and can be fertile. Thus, the AAP recommends raising these individuals as girls, notwithstanding the condition of their external genitalia and any androgenizing effects on the brain.[84] With respect to most other intersex conditions, fertility is significantly diminished or absent, and thus plays a less significant role or no role at all.[85]

In the early decades of sex reassignment surgery, individuals undergoing "feminizing" surgery – usually females with CAH – frequently had their phallus completely resected, if it was deemed to be too large to be considered feminine.[86] Although this does not represent the present dogma, capacity for normal (i.e., conventional heterosexual) sexual function, valuing male capacity for penetration and female capacity for receptivity, remains a strong determinant of the sex of rearing. In this connection, the AAP recommends evaluating the size of the infant's penis and its likelihood for increasing in size at puberty.[87] Those with a phallus too small for conventional heterosexual intercourse as a male (or who are chromosomally female yet whose phallus is so large as to shock the sensibilities of family and/or physicians) may be surgically assigned as females.[88] Such individuals will undergo surgery to reduce their phallus in size, if deemed excessively large in appearance for a clitoris, will have their testes removed, if any, and will eventually undergo vaginoplasty, or the construction of a blind-ending hole into which an average-sized penis can fit.[89]

Proper endocrine function is important, not only for the development and support of secondary sex characteristics at puberty, but also for development and maintenance of bone density.[90] The sex hormones produced by gonads are essential for proper endocrine function. Gonads need to be removed, however, if they conflict with the sex of assignment, as they may lead to development of inconsistent secondary sex characteristics at puberty. An individual whose gonads are removed must

84. American Academy of Pediatrics, *supra* note 12, at 141.

85. *Id.*

86. *See, e.g.*, DEWHURST & GORDON, *supra* note 77, at 41 (noting that, although "in theory preservation of the glans has something to recommend it, the results of amputation appear satisfactory," and is the "simplest" method for clitoral reduction).

87. American Academy of Pediatrics, *supra* note 12, at 141.

88. *Id.*

89. *Id.*

90. *See, e.g., id.*

remain on hormone therapy for the duration of his or her life.[91] The AAP therefore recommends, whenever possible, retaining gonads appropriate to the sex to which an infant is assigned.[92] This is particularly important for an individual with ovaries or a partial ovary, as they may produce adequate levels of estrogen throughout the individual's life for these purposes.[93] Conversely, the testes of intersex individuals are less likely to produce sufficient amounts of testosterone throughout an individual's life for the maintenance of sufficient bone density and development of secondary sex characteristics, particularly if the individual is a true hermaphrodite or has mixed gonadal dysgenesis. For this reason, it is considered less problematic to remove testes if they are undescended.[94]

Potential for malignant gonadal change (i.e., cancer) is a less significant factor in determining the sex of assignment. Testes and immature gonads with a Y chromosome are at risk for developing cancer, particularly if they remain in the individual's abdomen rather than descend into a scrotum.[95] The AAP recommends attempting to retain such gonads where the individual is to be raised as a male, however, provided they contain normal testicular tissue, and can be brought down into a scrotum at a later date.[96]

The final factor considered by the AAP in their guidelines is testosterone imprinting. Over the past decade, the impact of testosterone imprinting on the brain has become an increasingly important factor to consider in choosing the sex of assignment. Studies have shown that exposure to significant amounts of testosterone *in utero* has a masculinizing effect on the individual's behavior.[97] Thus, for example, some studies have shown that CAH girls, who are exposed to high levels of androgens as a result of their condition, exhibit more "masculine" behavior than other girls, and may be more likely to have a lesbian sexual orientation.[98] The AAP therefore recommends "caution" in recommending a sex of rearing different than an individual's

91. *Id.*
92. *Id.*
93. American Academy of Pediatrics, *supra* note 12, at 141.
94. *Id.*
95. *Id.*
96. *Id.*
97. *Id.*
98. American Academy of Pediatrics, *supra* note 12, at 141; Barthold & Gonzalez, *supra* note 1, at 560.

chromosomal sex (particularly as the majority of intersex individuals are assigned to the female sex).[99]

E. Outcomes

There are alarmingly few studies reported in the literature evaluating the sexual and psychological success or failure of sex assignment surgeries, even though such surgeries have been performed long enough for a substantial cohort to have reached adulthood. One of the largest published studies evaluated fifty-nine intersex individuals, ninety-three percent of whom had been assigned to a sex within the first four weeks of life and underwent early genital surgery. Nearly forty percent of the cohort exhibited "general psychopathology."[100] Seven of the fifty-nine exhibited frank gender identity disorder.[101] These children exhibited "intense sadness and dissatisfaction with the assigned sex and a preference for behavior appropriate to the other sex."[102] Two had CAH, one had PAIS, one was a true hermaphrodite with an XY karyotype, one had cloacal exstrophy, one had a transversely constructed penis, and one had gonadal dysgenesis.[103] Twenty-five out of the forty-seven other individuals assigned as females exhibited "deviant" gender role behavior, or "boyish" behavior.[104] The authors of the study did not define what they meant by "deviant" gender role behavior or "boyish" behavior, but noted the latter included "wild, rough play."[105] CAH girls were most likely to exhibit "boyish" behavior, followed by formerly "male" pseudohermaphrodites and true hermaphrodites.[106] Individuals with CAIS were least likely to exhibit such behavior. "Deviant" gender role behavior was most often a source of concern for the parents of the formerly "male" pseudohermaphrodites, as it caused them to question the wisdom of the sex assignment.[107] None of the five individuals assigned as boys experienced gender identity disorder or "deviant" gender role behavior, although they were "not assertive" and were "fearful and bothered about the smallness of

99. American Academy of Pediatrics, *supra* note 12, at 141.
100. Slijper et al., *supra* note 41, at 134.
101. *Id.*
102. *Id.* at 136.
103. *Id.*
104. *Id.* at 137.
105. Slipjper et al., *supra* note 41, at 137.
106. *Id.*
107. *Id.*

their penis."[108] The study concluded that both hormonal and psychosocial influences led to the gender disturbances experienced by a significant number of the study participants.[109]

Another recent study evaluated cosmetic and anatomical outcomes in adolescence of feminizing surgery performed in infancy and childhood.[110] The cohort of 44 intersex individuals had a variety of diagnoses, including congenital adrenal hyperplasia, XXY and ambiguous genitalia, true hermaphroditism, XY females, mixed gonadal dysgenesis, and extrophy.[111] More than half (fifty-nine percent) had a good or acceptable cosmetic result (i.e., no surgery or only minor surgery or elective deferment of major surgery until after puberty was recommended).[112] However, forty-one percent of the cohort had a poor cosmetic result (i.e., further major surgery was recommended), and sixty-six percent had a poor overall outcome.[113] Moreover, ninety-eight percent needed further treatment to improve cosmetic appearance or to facilitate tampon use or sexual intercourse.[114] The authors note that the children's outcomes were poorer than previously reported.[115] They recommend delaying most surgeries until the child is old enough to be involved in the decision, and note that clinicians and parents must understand that "for most individuals further treatment will be necessary in adolescence and the long-term impact of such treatment on adult sexual function is still unknown."[116]

Anecdotally, one can find numerous histories told by intersex individuals who rejected both their sex and gender assignment, or who rejected their gender of rearing.[117] Many others resent having had operations performed upon them without adequate informed consent, and before they personally could choose what

108. *Id.*
109. *Id.* at 138.
110. Sarah M. Creighton et al., *Objective Cosmetic and Anatomical Outcomes at Adolescence of Feminising Surgery for Ambiguous Genitalia Done in Childhood*, 358 LANCET 124 (2001) *available at* 2001 WL 10159197.
111. *Id.*
112. *Id.*
113. *Id.*
114. *Id.*
115. Creighton et al., *supra* note 110.
116. *Id.*
117. *See, e.g.*, the Intersex Society of North America website *at* http://www.isna.org (last visited Apr. 22, 2002); the United Kingdom Intersex Association website *at* http://www.ukia.co.uk (last visited Apr. 22, 2002); and the Androgen Insensitivity Syndrome Support Group *at* http://www.medhelp.org/www/ais (last visited Apr. 22, 2002); *see also* DREGER, *supra* note 17, at 167–80.

they felt was best for them. The surgeries frequently result in the removal of tissue which otherwise could have been used for sex assignment surgery, if desired in the individual's adulthood. They also result in scarring and, often, decreased sensitivity.[118] As one intersex individual put it, a "very special form of sexuality, arousal, and all of that that was uniquely hermaphroditic was taken [by the sex assignment surgeries]. That is the crime."[119] The worst effect, however, appears to have been the secrecy with which many intersex individuals' conditions were treated. "[B]eing encouraged to keep silent about their differences and surgical alterations only served to enforce feelings of isolation, stigma and shame – the very feelings that such procedures are attempting to alleviate."[120]

III. The Court System As a Means of Modifying The Standard of Care

A. Medical Malpractice

Medical malpractice actions are arguably one means of policing the medical profession. Tort claims are thought to have deterrence value: the threat of liability allegedly helps prevent negligent conduct.[121] If this theory is correct, a large judgment or settlement against a physician for malpractice, along with the adverse publicity accompanying it, may prompt prudent health care providers to refrain from similar conduct. Some studies indicate that medical malpractice suits do have a significant role in spurring physicians to practice defensive medicine.[122] While some forms of defensive medicine appear to consist of inappropriate precautions which, at best, waste resources, others presumably comprise "intelligent precautions that tort law seeks to

118. *See, e.g.*, Cheryl Chase, Letter to the Editor, 28 Arch. Sex. Behavior (1999).

119. Videotape: Hermaphrodites Speak! (Intersex Society of North America 2000).

120. Sharon E. Preves, *For the Sake of the Children: Destigmatizing Intersexuality*, 9 J. Clinical Ethics 411, 414 (1998).

121. *See, e.g.*, Gary T. Schwartz, *Reality in the Economic Analysis of Tort Law: Does Tort Law Really Deter?*, 42 U.C.L.A. L. Rev. 377, 381 (1994).

122. *Id.* at 401-02 *citing* Paul C. Weiler et al., A Measure of Malpractice 127 (1993); Ann G. Lawthers et al., *Physicians' Perceptions of the Risk of Being Sued*, 17 J. Health Pol. Pol'y & L. 463, 470 (1992); Roger A. Reynolds et al., *The Cost of Medical Professional Liability*, 257 JAMA 2776, 2777-78 (1987); Stephen Zuckerman, *Medical Malpractice Claims, Legal Costs, and the Practice of Defensive Medicine*, 3 Health Affairs 128, 132 (1984)).

encourage."[123] Given the foregoing, might intersex individuals who underwent sex assignment surgery in infancy or childhood bring negligence claims against their surgeons as one means of altering the present standard of care in treating infants with ambiguous genitalia?

The answer is: not likely. Broadly speaking, all U.S. jurisdictions generally adhere to some form of the "professional custom" standard of care in medical malpractice actions. Unlike general tort claims, in which a defendant is held to the standard of care which a "reasonable person" would exercise under the circumstances, physicians alone determine the standard of care to which the members of their profession are legally held.[124] This poses a significant problem for intersex individuals who underwent sex assignment surgery in infancy and who wish to sue their surgeons for malpractice as a result of the surgery since, in short, it was the professional standard of care to treat such individuals with sex assignment surgery at the time that the surgery was performed. Thus, as there were no violations of the professional standard of care, the physician in question cannot be found negligent in most cases.

There have, however, been a handful of cases in which a court has refused to hold a physician merely to the standard set by his or her own profession, but instead has appeared to opt in part or whole for the traditional "reasonable care" standard used in most tort cases. The most famous is *Helling v. Carey*, a Washington case in which the court held the defendant ophthalmologist negligent for failing over a number of years to perform a glaucoma test on a young woman, who later lost much of her vision to the condition.[125] At the trial level, the physician's experts testified that it was not the standard of care for ophthalmologists to regularly perform glaucoma tests on individuals under the age of forty, as glaucoma is rare in younger individuals. The plaintiff's expert concurred in this testimony, and the defendant prevailed.[126]

123. *Id.* at 402; *but see, e.g.*, Michelle M. Mello, *Of Swords And Shields: The Role Of Clinical Practice Guidelines In Medical Malpractice Litigation*, 149 U. PA. L. REV. 645, 646 (2001) (noting some inefficiencies of defensive medicine engendered by medical malpractice litigation, and that the costs of such defensive medicine total nearly $7 billion annually).

124. *See, e.g.*, Theodore Silver, *One Hundred Years of Harmful Error: The Historical Jurisprudence of Medical Malpractice*, 1992 WIS. L. REV. 1193, 1194, 1201 [hereinafter Silver].

125. 519 P.2d 981, 983 (Wash. 1974).

126. *Id.* at 982.

On appeal, the plaintiff argued that the trial court improperly prevented her from arguing that the standard of care was inadequate to prevent her from harm.[127] The Supreme Court of Washington agreed with the plaintiff and reversed the trial court.[128] Quoting Judge Hand, it noted that:

> (I)n most cases reasonable prudence is in fact common prudence; but strictly it is never its measure; a whole calling may have unduly lagged in the adoption of new and available devices. It never may set its own tests, however persuasive be its usages. Courts must in the end say what is required; there are precautions so imperative that even their universal disregard will not excuse their omission.[129]

Using this rationale, it found that "reasonable prudence required" that the defendant give the glaucoma test to the plaintiff, even though it was not the standard of care for ophthalmologists to do so at the time.[130]

The standard set in *Helling* is a minority view. In fact, virtually no other court has adopted it since its inception.[131] Even Washington courts, while never expressly overruling it, do not generally follow it as written. Rather, the state's supreme court later backpedaled from the case's holding, noting that "[w]hile it is a reasonably prudent health care provider, rather than any reasonably prudent person, against which the defendant's conduct is to be measured, this qualification was also implicit in the standard established by *Helling* and *Gates*."[132]

As one California court of appeals noted in declining to follow *Helling*:

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127. *Id.*

128. *Id.* at 983.

129. *Id.* (citing *The T. J. Hooper*, 60 F.2d 737, 740 (2d Cir. 1932)).

130. *Helling*, 519 P.2d at 983.

131. *See, e.g.*, Osborn v. Irwin Mem'l Blood Bank, 7 Cal. Rptr. 2d 101, 126-27 (1992) (noting that most of the commentary on *Helling* has been "unfavorable," and that only one California case has followed it, notwithstanding established California law holding that "the professional standard of care is a function of custom and practice"). A case in apparent accord with *Helling* is Townsend v. Kiracoff, 545 F. Supp. 465, 468 (D. Colo. 1982) (citing *The T.J. Hooper*, 60 F.2d 737 (2d Cir. 1932) ("even if the defendant's affidavits and evidentiary materials could establish that the hospital acted in accordance with the standard of care and custom of the community of Colorado hospitals, the plaintiff would still be entitled to prove at trial that the entire community's custom is negligent"). This case, however, does not appear to be widely followed.

132. Harris v. Groth, 663 P.2d 113, 116 (Wash. 1983). The Washington Supreme Court held in *Gates v. Jensen*, 595 P.2d 919, 924 (Wash. 1979), a negligence suit against an ophthalmologist for failure to perform a glaucoma test, that the "reasonable prudence" standard articulated in *Helling* still applied.

> A contemporary observer wrote that the *Helling* court had 'unwisely . . . arrogated to itself medical decisions, superimposing its medical judgment upon the collective experience of the medical profession. Can it really be said that medical judgments of the courts will be 'right' more often than those guided by approved medical practices?'[133]

This is a significant problem. The "professional custom" standard in medical malpractice cases exists largely because of the technical and scientific complexity perceived to be involved in most areas of medical practice.[134] If one accepts this premise, should one expect a court to do a better job than a jury of evaluating medical judgments based solely on common sense?

Hazel Glenn Beh and Milton Diamond, in their discussion of potential remedies for intersex individuals who believe they were harmed by sex assignment surgeries in their infancies and childhood, argue that allowing physicians to set the standards of care by which they will be judged in medical malpractice actions promotes professional inertia.[135] They state that "[b]y allowing the medical community to set the standard by which negligence is determined and by protecting the divided medical community, tort law renders itself impotent to promote positive changes within the medical community."[136] This concern has been sounded elsewhere: "[w]ith professional custom as the standard, the nation's physicians may lawfully adopt and follow practices that are patently negligent and unreasonable under the standard of ordinary care to which all others are held. The medical community is answerable not for want of care but for want of conformity."[137] Beh and Diamond's discussion implies, without directly so stating, that the standard set in *Helling* may be a more appropriate one in select cases, such as those – like the intersex cases – in which the standard of care developed without reference to sound and thorough scientific research.[138]

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133. Osborn, 7 Cal. Rptr. 2d at 126 (citing Joseph H. King, *In Search of a Standard of Care for the Medical Profession: The "Accepted Practice" Formula*, 28 VAND. L. REV. 1213, 1250 (1975)).

134. *See, e.g.*, Silver, *supra* note 124, at 1215 (citing the premise that "medical practice, being highly complex, is not susceptible to evaluation through ordinary common sense and must instead be assessed pursuant to the customs of those with experience" as one reason for the development of the professional custom standard).

135. *See* Beh & Diamond, *supra* note 5, at 33.

136. *Id.* at 33-34.

137. Silver, *supra* note 124, at 1213.

138. Beh & Diamond, *supra* note 5, at 33-34.

This is not a feasible suggestion, because it would expose practitioners to liability for failing to foresee, for example, that the studies on which they based their practice would later be exposed as fraudulent or faulty. Practitioners ought not to be held responsible for independently verifying the correctness of each and every piece of research on which they base their practice. Yet this is what would be required to find surgeons liable for the otherwise acceptably-executed sex assignment surgeries they performed on intersex children at infancy. Unless the surgeons themselves were responsible for the research in question, and knew of the fraudulent or faulty nature of their research yet nevertheless propounded it as correct, there should be no basis for finding them negligent for otherwise reasonably relying on research to inform and shape their practice.

It may be more prudent, instead, to impose a rule of negligence holding that physicians may be held not merely to the prevailing custom or practice of similar physicians, but also to that practice which is reasonable to expect, given the state of medical knowledge at the time of treatment.[139] As the Supreme Court of Wisconsin noted in *Nowatske v. Osterloh*, altering the usual standard of care to which physicians are held to include those practices which a reasonable physician would use, given current medical knowledge, would not frequently yield a difference between current practice and "reasonable" practice.[140] Nonetheless, it could make a significant difference in a small minority of cases in which prevailing practice lagged behind what the reasonable practice would have been, had prevailing practice reasonably kept up with notable and firm advances in medical knowledge.

In the case of intersex individuals, a revised standard such as this might yield some positive changes. First, given the doubt cast on current practice, such a revised standard would likely require health care professionals, at minimum, to reevaluate the current practice of sex assignment and cosmetic genital surgeries. It is now apparent that the management of intersex individuals developed on the basis of anecdotal case reports, including one which was later found to have omitted key information which would have significantly changed the conclusion to be drawn from it. Moreover, there are no large-scale studies of long-term outcomes of such surgeries for the children who

139. *See, e.g.,* Nowatske v. Osterloh, 543 N.W.2d 265, 272 (Wis. 1996).
140. *Id.*

underwent them.[141] Recent years have nevertheless yielded enough small studies, along with anecdotal evidence, to strongly suggest that many classes of cosmetic genital and sex assignment surgeries have been at least as detrimental as beneficial to those on whom they were performed.[142] The conclusions of the latter studies are by no means sufficiently certain to warrant a finding of malpractice for those physicians who now fail to heed them, even under the proposed revised standard. Nevertheless, they provide enough evidence, in conjunction with the flawed standard on which the prevailing practice is based, to problematize the prevailing practice and suggest that further study is necessary.

For the same reasons, the revised medical malpractice standard may also help contraindicate surgeries which would assign an individual to a sex which matched neither his/her chromosomal sex nor a sex congruent with significant androgen imprinting *in utero*.[143] While the requisite large-scale studies have not yet been performed, there is enough evidence on a smaller scale to suggest that certain sorts of sex assignments ought not to be frequently performed, as they may carry a significant risk that the individual will ultimately reject the assignment. Nonetheless, such a revised standard would not require a moratorium on all sex assignment surgeries, and would not impose any prohibitions at all on cosmetic genital surgeries. In both cases, there is presently an insufficient amount of research to determine with reasonable certainty which, if any, surgeries tend to be beneficial for the recipients (as opposed to, for example, the parents of the child),[144] and whether any of the surgeries tend to have more detrimental than beneficial effects for the recipients, to warrant

141. *See, e.g.*, Kenneth Kipnis & Milton Diamond, *Pediatric Ethics and the Surgical Assignment of Sex*, 9 J. CLINICAL ETHICS 398, 401 (1998).

142. Anne Fausto-Sterling, for example, reviewed with a colleague the (often scarce and anecdotal) literature on reduction clitoroplasties and vaginoplasties, among other surgeries. They found mentions of scarring, multiple surgeries (leading to increased scarring), and residual pain and/or hypersensitivity at the clitoris or clitoral stump in the review of reduction clitoroplasties. Frequently, the only criterion listed for the success of a reduction clitoroplasty was cosmetic appearance, not later sexual function. The literature on vaginoplasties revealed frequent multiple surgeries, scarring, and vaginal stenosis. Where specific criteria for evaluating the operation's success were given, it was frequently the ability to have vaginal intercourse. *See* FAUSTO-STERLING, *supra* note 2, at 80-87.

143. *See, e.g.*, American Academy of Pediatrics, *supra* note 12.

144. *See, e.g.*, Wilson & Reiner, *supra* note 13, at 363 (noting physicians often recommend early surgeries "to spare parents the trauma of seeing their child as intersexed each time they change the infant's diaper").

any changes in the practice based on a revised rule of negligence.[145]

The adoption of such a revised general standard of care for medical malpractice cases might thus add impetus towards a scientific evaluation of the long-term effects of sex assignment and genital normalizing surgeries and would prohibit some of the most controversial surgeries. It would not, however, have any appreciable effect on the majority of treatment protocols for intersex children. The proposed rule would therefore have little short-term impact. It would also leave the impetus for change largely in the hands of physicians and researchers, both of whom were responsible for the old treatment paradigms, and whose trustworthiness has been compromised in the eyes of some intersex activists as a result. Thus, altering the medical malpractice standard is inadequate to revise some of the more problematic aspects of current practices in treating intersex children.

B. Informed Consent

Informed consent is another area intersex activists have investigated as a potential source of legal action against physicians performing genital normalizing and sex assignment surgeries. Under the doctrine of informed consent, a competent patient (or his or her authorized representative) must decide whether to undergo medical treatment or surgery after his or her physician explains the risks and benefits of treatment, as well as treatment alternatives in some cases. Activists and others who have examined the issue note that cosmetic and sex assignment surgeries on intersex children are frequently performed without adequate disclosure, and under rushed circumstances which are not conducive to careful or thoughtful deliberation.[146]

145. Again, while there is anecdotal evidence that certain sex assignment and normalizing surgeries are more detrimental than beneficial for the recipients, few actual studies have been performed, and, apparently, none with a sufficiently large cohort and control group from which one could make reasonably certain conclusions.

146. *See, e.g.*, FAUSTO-STERLING; *supra* note 2, at 80-85; Beh & Diamond, *supra* note 5, *passim*. Recent research on gender identity and patient satisfaction by John Gearhart, M.D., et al. with respect to surgery performed for a variety of intersex conditions revealed that patients most often wished they had been given more information, even when they were otherwise satisfied with the outcome. *See* J.P. Gearhart et al., *CAIS: Long-Term Medical, Surgical and Psychosexual Outcome*; *PAIS and Partial Gonadal Dysgenesis: Long-Term Medical, Surgical and Psychosexual Outcome of Patients Reared Male or Female*; and *Micropenis, presented at* 2001 American Academy of Pediatrics National Conference and Exhibition, Section on Urology.

The doctrine of informed consent has only recently appeared on the scene of medicine. Hippocrates, the ancient Greek philosopher of medicine, noted that "physicians should conceal 'most things from the patient while you are attending to him . . . revealing nothing of the patient's future or present condition.'"[147] Little changed in America from that time until the middle of the 20th century. "Three beliefs dominated pre-mid-20th century physician-patient relationships: patients must (1) honor physicians; (2) have faith in them; and (3) 'promise obedience.'"[148] These tenets were woven into physicians' codes of conduct. Until just a few decades ago, the American Medical Association (AMA) Code of Medical Ethics asserted in one form or another that a patient should obey the prescriptions of his or her physician, without heed to the patient's own opinion about the matter.[149]

The roots of the informed consent doctrine stem from the turn of the century, with cases such as *Schloendorff v. Society of the New York Hosp.* establishing the right of a patient to sue his or her physician for battery in the event of an unconsensual surgery.[150] The true birth of the doctrine, however, did not come until 1957, with the case of *Salgo v. Leland Stanford Jr. Univ. Board of Trustees*.[151] This case – decided less than fifty years ago – first recognized the doctrine of informed consent as an element of the physician-patient relationship, and permitted a negligence action on this basis.[152]

In the ensuing decades, the doctrine of informed consent has evolved to focus on protecting "the right of every individual to

147. Sheldon F. Kurtz, *The Law of Informed Consent: from 'Doctor is Right' to 'Patient Has Rights,'* 50 SYRACUSE L. REV. 1243, 1243 (2000) (quoting 2 HIPPOCRATES, DECORUM 297 (W. Jones trans., Cambridge: Harvard University Press 1967)).

148. *Id.* (citing JAY KATZ, THE SILENT WORLD OF DOCTOR AND PATIENT (1984)).

149. *Cf. id.* In contrast, the AMA Code of Medical Ethics states, in relevant part, that:

> The patient has the right to receive information from physicians and to discuss the benefits, risks, and costs of appropriate treatment alternatives. Patients should receive guidance from their physicians as to the optimal course of action . . . The patient has the right to make decisions regarding the health care that is recommended by his or her physician. Accordingly, patients may accept or refuse any recommended medical treatment.

AMERICAN MEDICAL ASSOCIATION, FUNDAMENTAL ELEMENTS OF THE PHYSICIAN-PATIENT RELATIONSHIP (last revised 1994).

150. 105 N.E. 92 (N.Y. 1914); *see also, e.g.,* State v. Housekeeper, 16 A. 382 (Md. 1889).

151. 317 P.2d 170 (Cal. Ct. App. 1957).

152. *See, e.g.*, Kurtz, *supra* note 147, at 1244-45.

the possession and control of his own person, free from all restraint or interference of others, unless by clear and unquestionable authority of law."[153] The doctrine generally requires that physicians share decision-making power with their patients.[154] It further requires that physicians give patients the information necessary for patients to meaningfully exercise such power.[155] For a consent to be valid, it generally must be informed, voluntary, and given by an individual who is both authorized and competent to give consent.[156] As minors are generally incapable of giving valid informed consent, their parents must do so for them.[157]

Beh and Diamond, among others, argue that sex assignment and cosmetic genital surgeries on intersex infants and children are deficient with respect to informed consent.[158] According to Beh and Diamond, parents frequently are not in a position to provide valid informed consent, as health care providers often fail to provide sufficient information concerning the proposed surgeries.[159] They note that health care providers often convey an aura of urgency regarding sex assignment and cosmetic genital surgeries that is not medically or surgically justified.[160] Information concerning the surgeries is frequently incomplete,

153. *See, e.g.*, Beh & Diamond, *supra* note 5, at 34 (*citing* Cruzan v. Director, Mo. Dep't of Health, 497 U.S. 261, 269 (1990)).

154. *Id.*

155. Jurisdictions differ over whether the information necessary for patients to meaningfully exercise such power should be judged on a professional or patient basis. Those jurisdictions employing a professional standard require a physician to disclose those risks which similarly-situated physicians disclose, as established by expert medical testimony. *See* Culbertson v. Mernitz, 602 N.E.2d 98, 102-03 (Ind. 1992). Conversely, the patient standard is based on the theory that "[r]espect for the patient's right of self-determination on a particular therapy demands a standard set by law for physicians rather than one which physicians may or may not impose upon themselves," and requires that physicians disclose all risks which a prudent patient would consider material to his or her decision whether to undergo treatment. *See* Canterbury v. Spence, 464 F.2d 772, 784 (D.C. Cir. 1972).

156. *See, e.g.*, FLA. STAT. ANN. § 766.103 (West 2001); 24 ME. REV. STAT. ANN. §2905 (West 2001); TX. HEALTH & SAFETY CODE §462.009 (West 2001).

157. Although there are variations, the law generally evaluates a parent's right to consent to medical treatment on behalf of her child in light of the child's "best interest." *See* Jennifer Rosato, *Using Bioethics Discourse to Determine When Parents Should Make Health Care Decisions for Their Children: Is Deference Justified?*, 73 TEMPLE L. REV. 1, 7- 8 (2000).

158. *See, e.g.*, Beh & Diamond, *supra* note 5, at 34-60; Kishka-Kamari Ford, *"First, Do No Harm" – The Fiction of Legal Parental Consent to Genital-Normalizing Surgery on Intersexed Infants*, 19 YALE L. & POL'Y REV. 469, 474-88 (2001).

159. Beh & Diamond, *supra* note 5, at 34-60.

160. *Id.* at 43-46.

particularly issues concerning the cosmetic outcome and potential effects of scarring on future sexual sensation.[161] Secrecy concerning the surgeries has been fostered in the past, particularly with respect to what the intersex child does or does not learn about them.[162] Beh and Diamond also note that physicians have frequently failed to disclose the possibility that the child will ultimately reject the sex to which the surgery will assign him or her, and that surgical intervention in childhood forecloses that child's "right to an open future."[163]

Because of frequent deficiencies in information and misrepresentations, particularly concerning how the surgeries may affect the child once he or she reaches adulthood, Beh and Diamond conclude that a moratorium should be imposed on surgeries undertaken solely for cosmetic purposes on intersex children, and that such children and their families should instead be treated with counseling to manage the psychosocial issues.[164] They believe this strategy better protects the self-determination rights of intersex individuals by allowing them to decide for themselves, once they reach adulthood, whether they wish to undergo sex assignment or cosmetic genital surgery.[165]

There undoubtedly have been, and may still be, serious informed consent issues with many intersex surgeries on infants. This is not, however, a basis on which one can reasonably call for a moratorium on the surgeries, particularly when one can take the less drastic step of offering more complete information (e.g., indicating gaps in information, such as those concerning long-term outcomes). It also does nothing to expose *why* – es-

161. *Id.* at 47-50.

162. *Id.* at 50-55. Beh and Diamond's contentions are borne out by the literature. *See, e.g.*, FAUSTO-STERLING, *supra* note 2, at 63-66 (noting the explanations clinicians recommend giving to the parents of intersexuals, e.g.: "accurate patho-physiological explanations are not appropriate and medical honesty at any price is of no benefit ot the patient;" and "[E]very effort should be made to discourage the concept that the child is part male and part female. . . . This is often best handled by explaining that 'the gonads were incompletely developed . . . and therefore required removal'"). *See also* DEWHURST & GORDON, *supra* note 77, at 80 (in discussing how to counsel parents of an older intersex child who did not previously undergo genital surgery, noting that "[t]he idea which must be conveyed to them is that sex is being corrected not changed; that a mistake was made initially and this is now being put right; that the child was never male but always female or vice versa"); Susan Baker, *Psychological Management of Intersex Children*, 8 PEDIAT. ADOLESC. ENDOCR. 261 – 269 (1981) ("the first communication must include the information that the infant has a birth defect of *unfinished* genitalia") (emphasis added).

163. Beh & Diamond, *supra* note 5, at 56-59.

164. *Id.* at 59-60.

165. *Id.*; *see also* Ford, *supra* note 158, at 488.

pecially in light of informed consent law – physicians have frequently provided significantly incomplete or skewed information to parents with respect to their children's intersex conditions and surgeries. Or why the physicians employ euphemisms, gloss over poor information about long-term outcomes, and counsel secrecy. The historical problems with informed consent in this context suggest there are many hidden or obscured issues with intersex conditions and their treatment. Why was it once standard medical practice to openly advocate limiting and obfuscating information for parents and children concerning intersex conditions and surgeries in ways which likely violated informed consent norms?[166] Why did physicians regularly instruct the families of intersex individuals never to disclose the truth about their children's conditions, even though this secrecy may have had significant detrimental effects on the very individuals it was supposed to protect?[167] Without further investigation, merely criticizing defects in informed consent will not likely yield the desired effect of stopping the practice of cosmetic and sex assignment surgeries on intersex infants and children; rather, it may result simply in the proffer of more information to parents, while the surgeries themselves continue with little abatement.

Furthermore, while the desire to preserve the ability for patient self-determination in adulthood by refraining from surgery in childhood is laudable, there is no reason its citation alone should yield the necessary changes. Even if we decide that cosmetic genital and sex assignment surgeries should no longer be performed until the patient has reached the age of majority and can decide for him or herself whether to undergo them, we will not have done anything in the process to alter the social or cultural climate in which such decisions must be made. An exclusive dualism of "male" and "female" presently reigns in America, with significant exceptions only in small pockets of society. Although it has increased in the decades since the advent of the gay rights movement and the second advent of feminism, there is still little tolerance for gender ambiguity. A society which feels compelled to pass a Defense of Marriage Act,[168]

166. *See, e.g.*, FAUSTO-STERLING, *supra* note 2, at 63-66.

167. *See generally,* Preves, *supra* note 120, at *passim.*

168. 28 U.S.C.A. § 1738C (West 2002). The act also provided a federal definition of marriage: "In determining the meaning of any Act of Congress, or of any ruling, regulation, or interpretation of the various administrative bureaus and agencies of the United States, the word 'marriage' means only a legal union between one man and

which blamed a mother, who worked part-time, for the death of her infant at the hands of her au pair,[169] and which tacitly prevents men from taking advantage of their rights under the Family and Medical Leave Act[170] is not likely one which will, without some adjustment, openly embrace intersexuals who do not conform to current sex and gender norms. An empty right to self-determination carries little weight in the absence of an environment in which one can meaningfully exercise that right.

IV. Alternatives to Modifying the Standard of Care

It is therefore not enough to give intersexuals the right to decide for themselves, once they become adults, whether and how to alter the genitals with which they were born. Although there does not appear to be any evidence that the present methods of dealing surgically with intersex infants and children developed for sound scientific reasons, the present treatment protocols did not arise by accident or chance. Dewhurst and Gordon wrote in 1969 that society views intersexuals as "freak[s]" or "misfit[s] . . . condemned to a solitary existence of neglect and frustration."[171] More recently, another commentator noted that "the mystification of sex leaves no room for doubt, no place for ambiguity. The first thing asked of every new human being is whether it is a boy or a girl. It must be one or the other. There are no additional categories."[172] To such commentators, to think otherwise is to fall into the category of a "social constructionist . . . who maintain[s] that our concepts of man and woman are fictions dreamed up to keep everyone comfortably in their prescribed

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one woman as husband and wife, and the word 'spouse' refers only to a person of the opposite sex who is a husband or a wife." 1 U.S.C.A. § 7 (West 2002).

169. *See, e.g.*, Dave Howland, *Au Pair Trial: Public Scorns Parents*, Dayton Daily News, October 28, 1997, at 3A, *available at* 1997 WL 16061865 ("'It's almost chilling to gauge the reactions toward them,' said Court TV programming chief Erik Sorenson, who was struck by the number of callers who condemned Mrs. Eappen for choosing to work instead of caring for her children full time").

170. 29 U.S.C.A. § 2601 (West 2002); *see also, e.g.*, Martin Malin, *Fathers and Parental Leave*, 72 Tex. L. Rev. 1047, 1077-78 (1994) ("Large employers are least likely to experience negative financial effects from fathers taking parental leave. Yet . . . sixty-three percent of large employers considered it unreasonable for a man to take any parental leave, and another seventeen percent considered paternal leave reasonable only if limited to two weeks or less").

171. Dewhurst & Gordon, *supra* note 77, at vii.

172. Louis Gooren, *Forward to the Second Edition*, *in* Money, *supra* note 4, at ix.

place."[173] If one accepts such a schema, the need for surgical correction of intersex conditions is virtually a given. To fail to do so would presumably be to "ignore[ ] the very real pain and suffering experienced by individuals with sexual anomalies."[174]

Do statements such as those cited above truly represent general American societal views of intersexuals? According to Carl Elliott they likely may. Elliott argues that physicians who advocate early cosmetic genital surgeries for intersex children rather than suggesting restraint until the children are old enough to decide what to do for themselves are not making a "conscious effort to fend off threats to a cultural order."[175] Instead, "we treat these children the way we do because this is how we see the world."[176] We take for granted our concepts of male and female in our culture, and their exclusively binary nature. Yet there is nothing necessary in our conceptions of sex and gender, Elliott states. Rather, one can look at other cultures – for example, the Navajo in the 1930's, who had a third gender with a special social status – to see this. This is not because the Navajo (or any other society with different concepts of sex and gender) are more or less enlightened than ours. Rather, extending upon Ludwig Wittgenstein's philosophy,

> [t]he issue dividing us and the Navaho . . . is one of commonsense judgments, our untutored, no-nonsense, matter-of-fact attitudes towards the world. It isn't just that what the Navaho call *nadle* we call hermaphrodites or transvestites, or that what certain Dominican Republic villagers call *guevedoche* we call (some of us, anyway) 5-alpha-reductase deficiency syndrome. The difference lies in our basic apprehensions of the obvious, the way life is, once it is stripped of artifice and theory and intellectual pretensions: the things anyone knows (or at least anyone with a lick of sense).[177]

Given our conceptual framework, Elliott notes, the intersex child does not fit into our usual way of seeing the world, and therefore poses a problem. Presently, we usually deal with this problem by performing cosmetic genital surgery on intersex children in infancy or early childhood in order to make the individual conform to our dominant notion of gender. This of

173. *Id.* at x–xi.
174. *Id.* at xi.
175. *Id.* at 40.
176. *Id.*
177. CARL ELLIOTT, BIOETHICS, CULTURE AND IDENTITY: A PHILOSOPHICAL DISEASE 36 (1999).

course does not mean that practice of early cosmetic and sex assignment surgeries should therefore continue. Instead, it indicates that successfully changing the paradigm will require more than a mere alteration in medical practice.

As a number of commentators have noted, physicians' management of intersex infants and children is already undergoing some flux.[178] Diamond and Sigmundson's revelation of the actual outcome of the John/Joan case, in conjunction with vocal protests from intersex activists and increasing attention from academicians, is starting to lead to a more "rational and comprehensive" evaluation which recognizes that "it is more important that the [gender] assignment be right than that it be fast."[179] This shift has nothing to do with a change in beliefs about gender, however. On the one hand, physicians may increasingly refrain from making hasty pronouncements concerning a child's sex to the child's parents, and may wait longer to perform genital surgeries except where deemed to be medically necessary (e.g., due to cancer risk or significant urinary tract impairment). They may also increasingly involve the parents in the decision-making concerning the child's gender and management on a more equal basis. On the other hand, however, physicians will also likely continue to recommend infant and childhood cosmetic and sex assignment surgeries to parents on a number of grounds, including the specter of locker room humiliations for adolescent intersex children, as well as social shame and stigma with respect to dating and sexual relations during adolescence and early adulthood. Grounds such as these were also used to justify the older methods of surgical management. But because the social views of sex and gender remain virtually the same, there is no reason for them to have disappeared just because other aspects of intersex management are in the process of altering. As long as the current exclusive dualism prevails in the mainstream view of sex and gender in this society, justifications such as the above will likely remain compelling reasons to at least some parents for early surgery.[180]

178. *See, e.g.*, Alice Domurat Dreger, *A History of Intersexuality: From the Age of Gonads to the Age of Consent*, 9 J. CLINICAL ETHICS 345, 353 (1998); Wilson & Reiner, *supra* note 13, at 364-65.

179. Wilson & Reiner, *supra* note 13, at 365.

180. Note, for instance, that the most recent guidelines from the American Academy of Pediatrics still advocate early genital surgeries for intersex individuals, even though it also acknowledges that "some suggest[ ] that the current early surgical treatment should be abandoned in favor of allowing the affected person participate in

So how best to proceed? As Anne Fausto-Sterling notes, intersex individuals have been used as a "natural experiment" in the search for hormonal causes of behavioral differences between the (two) sexes, serving as nature's guinea pigs, so to speak.[181] Yet she also makes the following observation:

> [c]urrently, [intersexual] bodies are . . . 'unthinkable, abject, unlivable.' By their very existence, they call into question our system of gender. Surgeons, psychologists, and endocrinologists, through their surgical skills, try to make good facsimiles of culturally-intelligible bodies. If we choose to eliminate mixed-genital births through prenatal treatments . . . we are also choosing to go with our current system of cultural intelligibility. If we choose, over a period of time, to let mixed-gender bodies and altered patterns of gender-related behavior to become visible, we will have, willy-nilly, chosen to change the rules of cultural intelligibility.[182]

It must be recognized that, if we choose the latter course, we will again have used intersex individuals as guinea pigs of sorts, this time in a cultural experiment. However much any of us may wish to see Fausto-Sterling's latter course prevail, the outcome is not certain. Thus, a physician cannot, in good conscience, assure the parents of an intersex infant that they ought not to choose cosmetic or sex assignment surgery for their child, on the ground that the unaltered child, along with his/her intersexual forebears and brethren, will (over time) alter our present sex and gender systems to make space for those who do not conform to the present norms. Given our present state of knowledge concerning the long-term outcomes of intersex individuals (both those who have and have not had surgery), and given our current sex and gender systems and the fact that intersex individuals have no choice but to cope with them one way or another, there can presently be no reasonably certain "right" answer for physicians and parents of intersex children with respect to surgery.[183]

Nevertheless, there does appear to be a likely "wrong" answer with respect to the timing of surgery, if any is to occur at

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gender assignment at a later time." American Academy of Pediatrics, *supra* note 12, at 141.

181. *See* FAUSTO-STERLING, *supra* note 2, at 73.

182. *Id.* at 76.

183. Of course, allowing present norms to sway one's decisionmaking will only further cement those norms. *See, e.g.*, ELLIOTT, *supra* note 177, at 28 (discussing "'the ethics of complicity': the notion that by giving in to the[ ] pressures that you justifiably feel are oppressive, you are yourself reinforcing the very norms that produce them").

all. Contrary to the prevailing norm, there appears to be few good reasons to perform cosmetic genital and sex assignment surgeries early in an intersex child's life. Surgeries in infancy have been advocated on a number of grounds. The one proposed by Money – that gender identity is malleable in the first months and years of life – has largely fallen out of favor after the revelation of John/Joan's true outcome. A second major reason for early surgeries has been its ability to keep children from knowing about their original condition. This secrecy, however, has been branded as harmful or otherwise strongly problematic by most reports from adult intersexuals.[184] As numerous other recent commentators have noted, this secrecy must cease.[185] Thus, early surgery ought not to be justified on that ground.[186] Improved wound healing in infants is an additional reason set forth for early surgeries. Certainly, if there is inevitably going to be a surgery, it is better – all else being equal – to perform it when visible scars are less likely to form, as is the case in infancy.[187] However, with respect to surgeries which are not necessary to preserve the physical health of the child, all else

184. *See, e.g.*, Gearhart et al., *supra* note 146; Preves, *supra* note 120, at 414-15; FAUSTO-STERLING, *supra* note 2, at 80-85.

185. *See, e.g.*, Dreger, *supra* note 178, at 352 ("In no other realm in medicine do doctors regularly argue for active, nearly wholesale deception"); Sherri A. Groveman, *The Hanukkah Bush: Ethical Implications in the Clinical Management of Intersex*, 9 J. CLINICAL ETHICS 356, 358-59 (1998) ("of the more than 60 women with AIS whom I personally know, I have not heard of a single instance where someone has reported that it was worse to know the truth than to live with lies"); Edmund G. Howe, *Intersexuality: What Should Careproviders Do Now*, 9 J. CLINICAL ETHICS 337, 338 (1998) (reporting that the shame of genital surgeries "was further exacerbated by doctors withholding information, which implied that their condition was too shameful to discuss"); Kipnis & Diamond, *supra* note 141, at 407 ("Unless the entire profession is complicit . . . one must expect that the truth will emerge. And when it does, the patient will learn anyway what she or he was never supposed to have found out. If the patient is going to find out anyway, surely it is better for the physician to initiate disclosure"); Justine Marut Schober, *A Surgeon's Response to the Intersex Controversy*, 9 J. CLINICAL ETHICS 393, 395 (1998) ("We desire the easiest psychological adjustment for a patient. Though deception might allow an easier adjustment in some cases, the parent and patient have a right to know, as well as the right to make educated, prospective choices"); Wilson & Reiner, *supra* note 13, at 364 ("Ultimately, as with most attempts to keep diagnostic/prognostic information from a child . . . the truth is not as devastating as what the child imagines").

186. Note as well detrimental psychological effects of surgery cannot be avoided merely by operating at an early age. The 1996 American Academy of Pediatrics recommendations for genital surgeries on boys, for example, notes several studies evaluating the psychological risks of surgery on children, some of which conclude the risks are greatest for children between the age of one and three. *See* Kass et al., *supra* note 6.

187. *Cf.* Cisek Interview, *supra* note 68.

is not equal. It may be that the intersex individual, if given the opportunity to decide for him/herself once s/he is older, would choose not to undergo the surgery. If the intersex individual's preference is to be given any significant weight, the ability to achieve a better cosmetic result by timing the surgery earlier pales in comparison.

The remaining reason justifying earlier surgery cannot be so quickly dispelled. Prevailing social norms may significantly and adversely affect an intersex infant in two ways. First, parents may bond poorly with or even reject an intersex child, due to its malformed genitals. As Wilson and Reiner note in the context of difficulties in involving parents in decisionmaking concerning their intersex child, parents will likely grieve the loss of their "expected 'perfect' child."[188] Kass et al. recommend genital surgery between the ages of six weeks and fifteen months, as waiting longer "potentially prolong[s] the child's 'defective' status and crystallize[s] any disruption in family relationships that the child's condition may have produced."[189] Second, parents are not the only ones with whom relationships may be disrupted; given the large number of children in day care, a diapered intersex infant's genitals will likely be exposed regularly to other caretakers.

Data suggests, however, that cosmetic genital or sex assignment surgery may not alleviate these issues. Slijper et al. report that, out of a group of 27 couples with intersex children who underwent surgery for their conditions in infancy:

> [d]espite the intensive counseling . . . 50% [of the couples] were not able to work through the trials and tribulations their child's lack of gender clarity entailed. Two mothers and 1 father openly rejected their child as a result. The following factors played a role in the acceptance process: (i) the time when assistance was offered: for 5 couples who had problems dealing with their child's lack of gender clarity, help came too late, since it had already been several years since the trauma; (ii) the instability of the marriage for 5 couples, the child's anomaly played an important role in their divorce; (iii) the number of times the sex assignment was revised the 2 couples whose child's sex assignment was revised twice continued to doubt whether they had made the right decision regarding the sex

188. Wilson & Reiner, *supra* note 13, at 365; *see also* Dreger, *supra* note 178, at 353.

189. Kass et al., *supra* note 6, at 590. For data which may help support this contention, notwithstanding the small sample size, see Slijper, *supra* note 41.

assignment; and (iv) the personality structure of the parents, particularly as regards rigidity and the inability to cope with setbacks and tolerate embarrassment.[190]

The data suggest that, even where surgery had been performed, many parents of the children still did not perceive their infant as "normal," or otherwise had significant difficulties accepting them. Those with children whose gender and sex assignments differed from the child's chromosomal sex also experienced particular difficulties.[191] Given the sample size, the study is not conclusive, however, it suggests that, notwithstanding surgery, the very issues surgeons hope to avoid through early surgery may nevertheless arise for a sizable number of families. In the meantime, the intersex individual's ability to provide input into the decision as s/he matures will have been completely sidestepped.

The dearth of large, long-term studies makes it impossible to provide well-schooled suggestions for clinical and surgical practice. However, in light of the foregoing, several conclusions can nevertheless be drawn. First – and most importantly – physicians need to discuss the child's condition openly with the child's parents and, as the child matures, with the child him/herself. The old policy of withholding information both prevented intersex individuals from knowing significant medical facts about themselves which were pertinent to their health, and exacerbated feelings of stigma and shame rather than preventing them.[192] Candor and full disclosure should therefore replace the prior policy of secrecy. The child, who will face psychological and social problems regardless of his/her medical and/or surgical treatment, should also be referred for counseling and, once of sufficient maturity, to intersex support groups.[193]

Second, physicians need to present the options available to parents in a neutral and inclusive fashion. Early surgeries should be limited to those necessary to establish normal urinary tract function and correct conditions that could cause recurrent infections or other damaging physical problems.[194] With respect to surgeries performed for cosmetic or sex assignment purposes, physicians should emphasize to parents that hasty decisions are

190. Slijper et al., *supra* note 41, at132.
191. *Id.*
192. *See, generally, supra* note 132.
193. *See, e.g.,* Dreger, *supra* note 178, at 354; Wilson & Reiner, *supra* note 13, at 365-66.
194. *See* Wilson & Reiner, *supra* note 13, at 365.

neither warranted nor advisable. They should further make clear that, while early cosmetic or sex assignment surgeries used to be (and still are) the norm, there is no definitive scientific evidence to recommend them and that, moreover, there is evidence that they may ultimately cause more harm to their recipients' later sexual function than good. Where relevant, they should also note that, with respect to sex assignment surgeries, there is evidence that some people later reject their assigned sex and that, if surgery has already been performed, there may not be enough tissue remaining with which to fashion functional genitals, should the individual later desire such surgery.[195] Additionally, they should emphasize that, in many cases, surgery will not provide their child with "normal" genitals, but may instead merely bring the genitals' appearance more in line with the norm, potentially at the expense of their function and sensation and at the cost of further surgeries.[196] Also, inter-abdominal testes which pose minimal cancer risk in a child's first years should be left intact whenever feasible until shortly before puberty, even if they have a chance of becoming malignant thereafter, so that the child will have some time to determine his/her gender.[197] As Wilson and Reiner advocate, hormone therapy should also be avoided whenever possible until the child has a chance to come to some decision about his/her gender identity.[198]

Third, parents need early psychological counseling and support.[199] Parents of children whose anatomical appearance is outside the norm typically "grieve the loss of the anticipated 'normal' child."[200] Given this likely effect, in conjunction with

195. *Cf.* Kipnis & Diamond, *supra* note 141, at 405-06.

196. *See, e.g.*, Creighton et al., *supra* note 110.

197. *See* Wilson & Reiner, *supra* note 13, at 365-66.

198. *Id.* at 366. Wilson and Reiner note in this connection:

> a recent referral[,] involv[ing] a child diagnosed as a true hermaphrodite with a female sex assignment referred by her pediatric psychologist. Her endocrinologist wanted to use estrogen therapy early to quiet the child's feelings of perhaps being male. But such an attempt to deny the child's sense of identity and suppress the gender confusion would seem to risk greater gender confusion and conflict as an adult, similar to that experienced by adult transsexuals. In addition, there is no data that feminizing hormones affect the evolution of gender identity.

Id.

199. Dreger, *supra* note 178, at 353; *see also* Wilson & Reiner, *supra* note 13, at 365.

200. Dreger, *supra* note 178, at 353. *See also* Wilson & Reiner, *supra* note 13, at 365.

the medical and social issues which the parents will face, counseling is advisable. Alice Dreger notes that a professional counselor, rather than a surgeon, urologist, or other member of the intersex child's medical team at birth, should perform this role, likely due to the inculcation of most of the latter in the traditional surgical management of intersex conditions.[201] Parents should also be referred to peer support groups for parents of intersex children.

Fourth, the parents and physicians should determine a gender of rearing for the infant.[202] This is a gendered society. An intersex child should be raised as a boy or a girl, even though the child may later reject the chosen gender and forge his or her own way. Physicians can assist parents in determining which gender a child will most likely choose; as an easy example, most children with CAIS will likely identify with the female gender, rather than the male, notwithstanding their testes and 46,XY karyotype, and therefore should probably be raised as girls.

Fifth, as Dreger points out, physicians should provide parents (and later, their children) with non-pathologized images of intersex individuals. If provided only with pathologized images, parents, intersex individuals – and members of the medical community – "will inevitably see intersexuality as deeply pathological."[203] The intersex community, rather than the medical community, can provide a source for these images.[204]

Notably, these suggestions do not include a moratorium on all early surgeries other than those necessary for the physical health of the intersex child. As Wilson and Reiner observe, there is very little data suggesting that parents can (or, for that matter, cannot) raise children with ambiguous genitalia unambiguously in one gender.[205] There is similarly little data concerning how well intersex children interact with their peers during adolescence, when ambiguities are likely to come to light in gym class and elsewhere (if they have not already done so).[206] Children are particularly hard on those whom they perceive to be different. Despite the dearth of data, however, Wilson and Reiner, among others, recommend a moratorium on genital surgeries which are not necessary for the physical health of the intersex

201. Dreger, *supra* note 178, at 353.
202. *See, e.g., id.*; Wilson & Reiner, *supra* note 13, at 365.
203. Dreger, *supra* note 178, at 353.
204. *Id.*
205. Wilson & Reiner, *supra* note 13, at 366-67.
206. *Id.*

child.[207] They note that this recommendation (among others) is largely the product "of a relatively small number of very vocal former patients and of a pilot study of six adolescents sex-reassigned at birth."[208]

It is undisputed that there are intersex individuals who have been harmed physically and/or psychologically by childhood cosmetic and/or sex assignment surgeries. Intersexuals have also been harmed by their deception at the hands of physicians and family members concerning their condition, and by the stigma and shame they felt as a result. Some of these individuals argue forcefully and persuasively that the decision to operate during their childhood, rather than waiting until they could decide for themselves what to do, was wrong. This does not mean, however, that such decisions are wrong for all people with an intersex condition. As discussed above, parents generally have a legal right to consent for their child's surgical treatment. If contested, courts frequently use a best interest test to determine whether consent or lack of consent was appropriate. Until larger, long-term studies are performed which show that cosmetic genital and/or sex assignment surgeries are generally not in an intersex child's best interest, neither an outright ban on such procedures nor removing parents' general right to consent to such surgeries can be justified, either ethically or legally.

Moreover, it is not merely the intersex individual who is affected by his/her condition. Rather, the parents, who must rear the child, and the child's family are also affected, as well as other close members of the child's community. Given that the intersex child does not enter into the world as an autonomous and independent being, the ability of the parents and others in the child's life to rear and relate to the child must be taken into account in determining which treatment options to permit and which to foreclose. However close-minded or otherwise regrettable it may be, not all parents may be able to cope with their child's anatomy without surgical alteration. And without a reasonable amount of love and support from their parents, intersex children – like any other children – will be more likely to experience significant social and/or psychological problems, both as children and as adults.

207. *Id.* at 365; *see also, e.g.*, Cheryl Chase, *Surgical Progress is not the Answer to Intersexuality*, 9 J. CLINICAL ETHICS 385, 391 (1998); Dreger, *supra* note 178, at 353; Kipnis & Diamond, *supra* note 141, at 405-06.

208. Wilson & Reiner, *supra* note 13, at 366.

Again, physicians should strongly counsel parents against making hasty decisions. Also, parents should be counseled to respect and consider their child's future independence and decisionmaking power, and to recognize that their child's future desires may conflict with the parents' own present ones. Helena Harmon-Smith, founder of Hermaphrodite Education and Listening Post, a peer support group for parents of intersex children, counsels against scheduling the first surgery before the child leaves the hospital following birth, as it "foster[s] fear in the parents that this is life-threatening and they have an abnormal or damaged child."[209] She also counsels against taking any "drastic" steps in the first year, as they will need that time to adjust to their child, understand his/her condition, and learn his/her needs.[210] Nevertheless, it must be recognized that some parents – ideally only a very small minority, at most – may ultimately opt for cosmetic genital and/or sex assignment surgery after careful and lengthy consideration of all the choices at hand and their potential outcomes. It must be recognized that the parental or familial needs driving this choice may be just as intense as any the intersex individual him/herself may experience. One can criticize or even condemn those needs. However, if surgery permits those parents to better relate to their child, then both the parents and the child will have benefited from it, notwithstanding any ill effects the surgery may ultimately have on the child him/herself.

V. Conclusion

Intersex conditions pose a thorny set of problems for affected individuals, families, and medical practitioners. Parents who had hoped for a "normal" child must face coping with physiological differences which, in our present society, throw the child out of relation with others on the basis of sex and gender. Physicians, viewing the difference as pathological, seek to use their skills to "correct" the child's genitals. And the intersex individuals themselves must grapple not only with the fact that their physical bodies, and sometimes also gender identities, do not fit neatly into our concepts of either 'man' or 'woman,' 'male' or 'female,' but also with potential stigma in the way others have treated them and their condition throughout their lives.

209. Helena Harmon-Smith, *10 Commandments*, 9 J. Clinical Ethics 371, 371 (1998).
210. *Id.*

In recent years, activists and a small group of researchers have largely overthrown the theory that early cosmetic genital or sex assignment surgery, in conjunction with rearing that strictly reinforces the child's gender assignment, yields individuals who accept and are well-adjusted with respect to their assigned gender and sex. As many intersex individuals and commentators have noted, a new treatment paradigm is now necessary. Many steps can be taken to ameliorate present management and treatment of intersex conditions. Contrary to some recommendations, however, a moratorium on cosmetic genital and sex assignment surgeries for infants and children is not warranted. It would swing the pendulum to the other extreme: while such surgeries have previously been recommended and performed with scant, if any, data to support their beneficial effects, a moratorium would similarly cease all such surgeries on the basis of several small studies and some negative reports from a number of individuals who underwent the surgeries in infancy and childhood.

Instead, medical practitioners should focus on providing complete information to parents and, as they mature, to intersex children. Both parents and children should be timely referred to counselors and intersex peer support groups. When discussing surgical options, physicians should provide full disclosure to parents about prior practices and the dearth of data, decades after the practices began, to support them. They and/or the parents' counselors should also emphasize that there is no need to perform cosmetic genital and/or sex assignment surgery early, before the child can contribute to or direct the decision. While the parents must be able to accept their child, it is the child, him or herself, who must live most directly with the consequences of any decision the parents make on his or her behalf. Nevertheless, medical, social and familial considerations require that the option to operate in childhood should remain as one potential tool among many to be used in the management and treatment of intersex children. The gravity of such a decision counsels restraint, regardless of the path ultimately chosen.

GLMA
Health Professionals
Advancing LGBT Equality

Title:	**Medical and Surgical Intervention of Patients with Differences in Sex Development**
Author:	**Jeremy Toler, MD**
Introduced by:	**GLMA Policy and Government Affairs Committee**
Board Approval:	**October 3, 2016 (amended and reapproved November 2, 2016)**

Whereas, surgical intervention, including removal and reconstruction of sex organs, has been practiced for children born with atypical genitalia since the 1960s and has long been considered a mainstay of treatment in these individuals,[1] despite often being considered medically unnecessary and potentially resulting in unwanted effects such as sterilization, and

Whereas, GLMA: Health Professionals Advancing LGBT Equality passed Resolution 105-98-105 in 1998, "Call for Research and Disclosure Regarding Intersex Surgery," indicating the organization's support for further advancement in research regarding the methods of care and biopsychosocial outcomes for patients with Differences of Sex Development (DSD), as well as reinforcing the need for frank and involved discussions between providers, patients, and family regarding the risks and benefits of treatment for these patients,[2] and

Whereas, while further research has been conducted into the outcomes of medical, surgical, and psychological treatment for these individuals, there continues to be a need for further studies and a consensus on standard of care, and

Whereas, surgical interventions continue to represent a common plan of care for children with DSD but remain highly controversial among pediatric specialists[3] and are largely condemned by the Intersex community, and

Whereas, research has provided varying rates of patient satisfaction in genital appearance, as well as diminished sexual function/satisfaction in adults after childhood surgical procedures intended to treat DSD; and future reconstructive surgical interventions remain common outcomes after initial childhood surgery,[3] and

Whereas, justification for such treatments in infancy and childhood has included the presumption of better socialization and acceptance among peer-groups and psychological well-being throughout life; however, evidence to support this is lacking,[4] and

Whereas, several prominent human rights organizations including the World Health Organization, Amnesty International,[6,7] the United Nations Special Rapporteur on Torture, and the United Nations High Commissioner for Human Rights,[5] have expressed concern about the violation of the human rights of individuals when medically unnecessary surgeries are performed

without their consent/assent and recommend postponing any surgeries until consent/assent may be given; and

Whereas, the decision for genital surgery in infants and children, for whom informed consent or assent cannot be attained, is placed upon parents, relies heavily on the relationship of the caretakers and healthcare providers, and requires comprehensive disclosure of risks and benefits as well as alternatives for intervention, including postponing interventions that do not have medical necessity; and

Whereas, parents or caretakers may often rely on social norms, binary gender concepts, personal convictions, and/or influence among outside parties in their decision-making process without understanding all avenues of treatment, including postponing treatment, therefore be it

RESOLVED: that GLMA: Health Professionals Advancing LGBT Equality recommends that patients and parents/caretakers are provided a comprehensive explanation of risks and benefits to surgical/medical intervention for Differences of Sex Development (DSD), as well as all alternatives to treatment, including postponement of interventions; and be it further

RESOLVED: that GLMA recommends delay of any surgical interventions and gender-related medical interventions for DSD that are not deemed medically necessary until the patient can provide informed consent/assent to these interventions; and be it further

RESOLVED: that GLMA encourages additional comprehensive retrospective and prospective biopsychosocial research on the long-term outcomes of patients born with differences of sex development; and be it further

RESOLVED: that the development and execution of research should involve, where available, the input of community representatives, psychiatrists, and other mental health practitioners from the intersex and DSD communities, and be it further

RESOLVED: that facilities that provide genital surgical interventions and gender-related medical interventions to patients with DSD adopt a multidisciplinary model to patient care that includes input from mental health specialists, medical and surgical specialists, bioethicists, and community/peer support organizations to deliver comprehensive biopsychosocial treatments that support all patients, their families, and any other caretakers, and be it further

RESOLVED: that GLMA urges development of cultural competency education for health care professionals and development of best practice guidelines regarding treatment of individuals with DSD, and be it further

RESOLVED: that GLMA adopts this policy as replacement and update of Resolution 105-98-105 (1998).

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[1] Creighton SM, et al. Timing and nature of reconstructive surgery for disorders of sex development — introduction. *Journal of Pediatric Urology*. 2012; 8(6): 602-610
[2] GLMA: Health Professionals Advancing LGBT Equality, "Call for Research and Disclosure Regarding Intersex Surgery" Resolution 105-98-105, March 7, 1998
[3] Lee PA, et al. Global disorders of sex development update since 2006: perceptions, approach and care. *Hormone Research in Pediatrics*. 2016; 85(3): 158-180
[4] Karkazis K, Tamar-Mattis A, Kon AA. Genital surgery for disorders of sex development: implementing a shared decision-making approach. *Journal of Pediatric Endocrinology and Metabolism*. 2010; 23: 789-806
[5] World Health Organization. Eliminating forced, coercive or otherwise involuntary sterilization: an interagency statement. 2014 [Statement of OHCHR, UN Women, UNAIDS, UNDP, UNFPA, UNICEF and WHO];
[6] United Nations Office of the High Commissioner for Human Rights. Free & Equal,Fact Sheet: Intersex. 2015.
[7] Amnesty International. Policy statement on the rights of intersex individuals, 2013.

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[8] Liao LM, Wood D, Creighton SM. Parental choice on normalising cosmetic genital surgery: between a rock and a hard place. *British Medical Journal*. 2015; 351: h5124.
[9] Wang LC and Poppas DP. Surgical outcomes and complications of reconstructive surgery in the female congenital adrenal hyperplasia patient: What every endocrinologist should know. *Journal of Steroid Biochemistry and Molecular Biology*. 2016

InterACT
Advocates for Intersex Youth

365 Boston Post Road, Suite 163 Sudbury, MA

Recommendations from interACT: Advocates for Intersex Youth regarding the List of Issues for the United States for the 59th Session of the Committee Against Torture

June 2016

Despite continuous international condemnation *and their own explicit recognition that non-consensual genital surgeries on intersex children have been classified as torture by the United Nations*, physicians in the United States continue to perform intersex genital mutilation. Thus, interACT (formerly known as Advocates for Informed Choice) implores the Committee Against Torture to include the treatment of intersex children in its List of Issues for the United States in the 59th Session.

1. interACT, formerly known as Advocates for Informed Choice, is an independent human rights NGO based in the United States. It is the first and only organization in the country exclusively dedicated to advocacy on behalf of children born with intersex traits. The term "intersex" refers to variations in a person's sexual or reproductive anatomy such that their body does not fit typical definitions of male or female, and includes many different medical conditions including androgen insensitivity syndrome, virilizing congenital adrenal hyperplasia (CAH), Klinefelter's syndrome, Turner syndrome, hypospadias, bladder exstrophy, and others. Common estimates of the frequency of intersex births are between one in 1,000 and one in 2,000.[1]

2. Beginning in infancy and continuing throughout childhood, children with intersex traits in the United States have been, and continue to be, subjected to intersex genital mutilation (IGM). These children often experience irreversible sex assignment and sterilization, medical display and photography of the genitals, and medical experimentation. People with intersex traits may also be denied necessary medical treatment. Moreover, intersex individuals suffer life-long physical and emotional injury as a result of such treatment. These human rights violations often involve tremendous physical and psychological pain and constitute torture as recognized by multiple international human rights bodies.

3. In 2013, the Special Rapporteur on Torture "call[ed] upon all States to repeal any law allowing intrusive and irreversible treatments, including forced genital-normalizing surgery, involuntary sterilization, unethical experimentation, medical display, 'reparative therapies' or 'conversion therapies', when enforced or administered without the free and informed consent of the person concerned. He also calls upon the states to outlaw forced or coerced sterilization in all circumstances and provide special protection to individuals belonging to

[1] Blackless M, et al. 2000. How sexually dimorphic are we? Review and synthesis. American Journal of Human Biology 2000, 12:151-166.

marginalized groups."[2] The Special Rapporteur renewed this call in his 2016 report on gender perspectives on torture.[3] In addition, the High Commissioner for Human Rights acknowledged that the rights infringed by the genital-"normalizing" surgeries carried out on intersex children include "their rights to physical integrity, to be free from torture and ill treatment, and to live free from harmful practices."[4]

4. Following the actions of the SRT, in 2014 the Society for Pediatric Urology of the United States, the professional organization of physicians who perform IGM in this country, published a paper concerning their "standpoint on the surgical management" of intersex traits. They recognized that the practice has been classified as torture but nevertheless failed to call for a ban on such surgeries, instead stating that more information must be gathered and that surgery could be justified "to restore more normal visible anatomy, and avoid ambiguity which is often the parents' wish."[5] However, as we noted in our response, this cannot be an ethical justification for such surgery, and the paper "significantly understate[d] reported catastrophic outcomes including complete loss of sexual sensation, psychological trauma and PTSD, sterilization, and irreversible surgical restructuring of genitals not appropriate to the eventual gender identity."[6]

5. Thereafter, in 2015, the World Health Organization, UNICEF, OHCHR, UN Women, UNAIDS, UNDP and UNFPA explained, intersex children "are often subjected to cosmetic and other non-medically indicated surgeries performed on their reproductive organs, without their informed consent or that of their parents, and without taking into consideration the views of the children involved [...] As a result, such children are being subjected to irreversible interventions that have lifelong consequence for their physical and mental health."[7] The statement called for accountability, participation, and access to remedies for intersex people.

6. Yet the practice continues. Just this year, in 2016, a group of prominent physicians published a statement on the treatment of intersex children, "Global Disorders of Sex Development Update since 2006," and again failed to call for an end to these surgeries despite their recognition of "a number of agencies condemning or calling for a complete moratorium on elective genital surgery or

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[2] Report of the Special Rapporteur on Torture, Juan E. Mendez, UN Doc. A/HRC/22/53 (2013), available at http://www.ohchr.org/Documents/HRBodies/HRCouncil/RegularSession/Session22/A.HRC.22.53_English.pdf
[3] Report of the Special Rapporteur on the right of everyone to the enjoyment of the highest attainable standard of physical and mental health, UN Doc A/HRC/32/33 (April 4 2016).
[4] United Nations Office of the High Commissioner for Human Rights, Free & Equal Fact Sheet: Intersex (2015).
[5] Mouriquand P., Caldamone A., Malone P., Frank J.D., Hoebeke P. *The ESPU/SPU Standpoint on the Surgical Management of Disorders of Sex Development (DSD)*, 10 JOURNAL OF PEDIATRIC UROLOGY 8 (2014).
[6] Anne Tamar-Mattis, *Patient Advocate Responds to DSD Surgery Debate*, 10 JOURNAL OF PEDIATRIC UROLOGY 788 (2014).
[7] WORLD HEALTH ORGANIZATION, *Eliminating forced, coercive or otherwise involuntary sterilization: An interagency statement* (OHCHR, UN Women, UNAIDS, UNDP, UNFPA, UNICEF and WHO) (2014), available at: http://apps.who.int/iris/bitstream/10665/112848/1/9789241507325_eng.pdf?ua=1.

gonadectomy without the individual's informed consent" and that "many guidelines deem children's participation and input indispensable to decisions, especially those that will have a life-long deeply personal impact on their lives, with heightened awareness that young children, in particular, may not be able to vocalize adverse reactions to many interventions."[8] Though the physicians are aware of the human rights violations they perform, the paper instructed them merely to "be aware that the trend in recent years has been for legal and human rights bodies to increasingly emphasize preserving patient autonomy."[9]

7. Many other published papers have recognized the potential for harm against this population, yet intersex children continue to experience genital mutilation in the United States. Physicians argue there must be additional medical research prior to a change in practice, yet unbiased research including the input of the intersex community is nonexistent. This year the Journal of Pediatric Urology published an article proffering to address the aims of genital surgery yet failed to even mention the lack of informed consent when these procedures are performed in infancy, instead avoiding the issue altogether and asserting that while "surgery has been restrictively considered by some to be 'cosmetic surgery,' the cosmetic aspect of genitalia and the related stigma risk are also important issues for many patients."[10] Yet, as noted by a Swiss National Advisory Commission on Biomedical Ethics, "[a]n irreversible sex assignment intervention involving harmful physical and psychological consequences cannot be justified on the grounds that the family, school or social environment has difficulty in accepting the child's natural physical characteristics … If such interventions are performed solely with a view to integration of the child into its family and social environment, then they run counter to the child's welfare"[11]

8. Recently, doctors at a major United States conference presented information from one registry in the United States (that is currently unavailable to access from patient groups) confirming the frequency of these surgeries as performed on infants. Regarding *initial* surgical intervention for children with Congenital Adrenal Hyperplasia (CAH), one of the more common intersex conditions, they noted "544 patients underwent feminizing genitoplasty between 2004-2014, median age at initial surgery: 9.9 months."[12] If other conditions and surgeries

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[8] Lee PA, Nordenström A, Houk CP, *Global Disorders of Sex Development Update since 2006: Perceptions, Approach and Care*, Hormone Research in Pediatrics 158-180 (2016). Available at: http://www.karger.com/Article/FullText/442975.
[9] *Id.*
[10] Mouriquand PD1, Gorduza DB2, Gay CL, *Surgery in disorders of Sex Development (DSD)with a gender issue: If (why), when, and how?* Journal of Pediatric Urology (2016). Available at: http://www.jpurol.com/article/S1477-5131(16)30012-2/abstract
[11] Swiss National Advisory Commission on Biomedical Ethics. On the management of differences of sex development: Ethical issues relating to "intersexuality." Opinion No. 20/2012.
[12] The Society for Pediatric Urology Annual Meeting, *Cost analysis and clinical outcomes of feminizing genitoplasty on congenital adrenal hyperplasia using a large scale administrative database* (May 6, 2016). Abstract available at: http://spuonline.org/abstracts/2016/16.cgi.

were considered, such as hypospadias repair, gonadectomy, or follow-up surgeries, that number would increase significantly. This conference included discussions of how to ensure these surgeries continue to be cost-effective/profitable for health care institutions. Our organization receives continual inquiries from families explaining that surgery is or has been pressed upon them in respected hospitals in major cities across the United States.

9. The continued treatment of intersex individuals in the United States clearly meets the CAT's standards for torture: that the action be intentional and performed for discriminatory and non-medical purposes; performed with state control, custody or consent; cause severe physical and psychological pain or suffering; and involve those who are powerless to refuse. However, it is clear that more must be done—even the recognition of the classification of IGM as torture has failed to improve the treatment of intersex youth in the United States.

10. Much of the "treatment" performed by physicians in the United States has already been recognized as torture or CIDT, as we have explained in previous publications.[13] Coerced sterilization can constitute torture and CIDT, and states' obligations to protect persons from such treatment extends into the private sphere, including where such practices are committed by private individuals.[14] In the case of FGM, which encompasses the clitoral reduction surgeries carried out on many female-assigned intersex children,[15] the SRT has specifically pointed out that where this is performed in private clinics and physicians carrying out the procedure are not being prosecuted, the State de facto consents to the practice and is therefore accountable.[16] However, we are unaware of any jurisdiction in the U.S. that enforces its own FGM laws in cases where the girl undergoing clitoral cutting has an intersex trait. Further, as we have noted previously, the U.N. Committee on the Rights of the Child has addressed involuntary sterilization of persons with disabilities under the age of 18 as a form of violence, in violation of the child's right to physical integrity, causing life-long effects on physical and mental health.[17] The Committee has called upon States to prohibit by law the involuntary sterilization of children on grounds of disability. Again, no exception has been mentioned for children whose medical condition happens to cause atypical sex characteristics.

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[13] Tamar-Mattis, *Medical Treatment of People with Intersex Conditions as Torture and Cruel, Unhuman, or Degrading Treatment or Punishment* in Torture in Healthcare Settings: reflections on the Special rapporteur on Torture's 2013 Thematic report.
[14] UN Committee Against Torture, General Comment No. 2 (2007), CAT/C/GC/2.
[15] Fraser S. *Constructing the Female Body: Using Female Genital Mutilation Law to Address Genital-Normalizing Surgery on Intersex Children in the United States*, INTERNATIONAL JOURNAL OF HUMAN RIGHTS IN HEALTHCARE (2016) 9:1, pp. 62-72, available at http://dx.doi.org/10.1108/IJHRH-05-2015-0014
[16] Fraser S. *Constructing the Female Body: Using Female Genital Mutilation Law to Address Genital-Normalizing Surgery on Intersex Children in the United States*, INTERNATIONAL JOURNAL OF HUMAN RIGHTS IN HEALTHCARE (2016) 9:1, pp. 62-72, available at http://dx.doi.org/10.1108/IJHRH-05-2015-0014
[17] U.N. Committee on the Rights of the Child, General Comment No. 13: The Right of the Child to Freedom from All Forms of Violence (2011); U.N. Committee on the Rights of the Child, General Comment No. 9: The Rights of Children with Disabilities (2007).

InterACT
Advocates for Intersex Youth
365 Boston Post Road, Suite 163 Sudbury, MA

11. Despite international condemnation from bodies including the World Health Organization, Amnesty International, and multiple committees of the United Nations and the explicit classification of intersex surgery as torture under several frameworks of human rights abuse, in addition to United States physicians' own awareness of their actions, the surgeries inflicted on intersex individuals in the United States continue in flagrant violation of, most notably, the Convention Against Torture and the mandate of the Special Rapporteur on Torture (SRT 2013).

12. Thus, interACT files this submission to inform the List of Issues for the United States for the Committee against Torture's 59th Session, to occur from November 7 to December 7, 2016. We respectfully request that the Committee consider the following inquiries:

    - **Please provide information on what steps, if any, are being taken by federal and state government bodies to end non-consensual genital surgeries on intersex individuals;**
    - **Please provide information on what steps, if any, are being taken by federal and state government bodies to ensure full and free informed consent is provided in all cases where surgical intervention on an intersex individual is considered;**
    - **Please provide information on what steps, if any, are being taken to provide for full legal review of non-consensual genital surgeries, including FGM and sterilization on intersex individuals;**
    - **Please provide information on what steps, if any, are being taken to address the need for data collection and independent monitoring of births of intersex children and their medical treatment;**
    - **Please provide information on what steps, if any, are being taken to address the need for disinterested research on long-term patient satisfaction of surgical and other procedures on intersex children, in consultation with intersex individuals and their organizations**.

Sincerely,

Anne Tamar-Mattis
Legal Director

365 Boston Post Road, Suite 163 Sudbury, MA 01776

Understanding Intersex and Transgender Communities

Intersex and transgender people have a shared interest in autonomy -- and may have distinct legal needs while facing overlapping barriers to appropriate care. While intersex individuals are forced to undergo medically unnecessary surgeries in infancy, transgender individuals are often denied desired medical treatment in adolescence and beyond. Transgender people may conversely be unable to access gender-congruent documentation without undergoing surgeries that are, in some cases, unwanted. Both communities grapple with a loss of decision-making authority over their own bodies. By better understanding the similarities and differences between these two groups, both movements can implement better policy and educate the public about the shared structural barriers facing both communities.

Clarifying Terminology:

The two terms are often confused: while a person who is **transgender** has a gender that is different from the one traditionally associated with the sex they were assigned at birth, a person who is **intersex** was born with a variation in their sexual or reproductive anatomy such that their body does not fit typical definitions of male or female.

- Both intersex and transgender people can identify as men, women, gender-fluid, non-binary, or in a multitude of different ways.
- While transgender people may identify differently from how they were assigned, their biology at birth typically conforms to a binary understanding of sexual and reproductive anatomy.
- Intersex people are generally assigned male or female despite their anatomical atypicality, but may later identify differently and correspondingly identify as transgender.
- A person cannot transition to "become" intersex because having an intersex condition is defined as a variation in reproductive anatomy present at birth.

This document uses "intersex" to mean those who are at risk for non-consensual surgery in infancy on the basis of medically observable intersex traits (sometimes called Differences of Sex Development).

Consent and Autonomy:

In the United States, intersex children often suffer non-consensual surgery on their genitals and reproductive organs to make their body look more typical, even though these surgeries damage sexual function and fertility. International human rights entities have called for an end to these surgeries, including the World Health Organization, Amnesty International, and the United Nations. American physicians recognized the harm of these surgeries as early as 1998, when the Gay and Lesbian Medical Association issued a resolution noting the physical and psychological damage of early genital surgery and calling for physicians to adequately inform parents of the negative outcomes and the opportunity to delay or reject surgery altogether.[1] Pediatric endocrinologists at leading hospitals have consistently outlined the need for transparency when guiding parents' medical decision-making for their intersex children.

One noted physician, Dr. Jorge Daaboul, speaking of the previous medical treatment of intersex children, admitted: "Many of my colleagues do not believe we have been deceptive [about the impact of early genital surgeries], and they would resent my saying we have been deceptive . . . . But we have been deceptive."[2] He later articulated: "I hope we can make amends to the [intersex] individuals we have harmed over the years, and I think our profession should do that in a formal way."[3]

[1] GLMA Resolution 105-98-101, *available at* http://www.ifge.org/news/1998/march/nws3218b.htm.
[2] *Interview in* Louise Kiernan, *In Intersex Cases, Gender is a Complex Question*, CHI. TRIB., June 20, 1999, at 1.
[3] Jorge Daaboul, *interview in* Videotape: XXXY (Laleh Soomekh & Porter Gale 2000), *available at* http://www.planetout.com/popcornq/db/getfilm.html?63816.

365 Boston Post Road, Suite 163 Sudbury, MA 01776

While intersex children are forced to undergo surgery without their informed consent, before they can decide for themselves what surgeries, if any, are appropriate, transgender people are often denied life-saving medical treatment in the form of gender-affirming surgeries and hormone therapy despite not only informed consent but their strong desire for these treatments. Sometimes, on the other hand, transgender adults are required to undergo surgeries to conform their bodies to typical notions of male and female in order to obtain accurate identification and other documents, despite that these surgeries are unwanted in some cases.

While it may seem like transgender and intersex communities are at odds on the pros and cons of surgery, in reality this is not the case. Our communities are united by the principles of *consent* and *autonomy*. In addition, both transgender and intersex activists want our communities to be able to access care that is *medically necessary*—that a person needs to live a healthy, fulfilling life.

	Common transgender experience	**Common intersex experience**
Consent	When a transgender person seeks out a specific hormonal or surgical treatment or to live as a gender other than that which they were assigned, they **consent** to this medical care.	When an intersex child's parents or doctors decide a child's body should conform to typical notions of male or female, and the child is forced to undergo normalizing surgery without their own input, the child **does not consent** to the surgical alteration of their body.
Autonomy	Transgender people have the right to **autonomy**, which includes asserting and expressing their gender. They should be able to access the resources (including all forms of medical care) that they feel are necessary and appropriate to their individual process of transition. No one should force or pressure a transgender person to undergo procedures, such as hormonal treatment or surgery, that they do not wish for themselves.	Intersex children have the right to **autonomy**, which means the right to grow up and decide for themselves whether they want any procedures, such as hormonal treatment or surgery, performed on their bodies. No one, including the intersex child's parents or doctors, should be allowed to make that decision for them because they may choose an intervention that the intersex person would not wish for themselves. Preserving the intersex child's autonomy means avoiding making personal, irreversible decisions so that when the child is older, they can express their gender and change (or decide not to change) their body in just the ways they want.
Medical Necessity	For a transgender person, medical treatment related to their transition (such as hormones or surgery) is sometimes medically necessary. Not all transgender people want transition-related medical treatment, and if an individual does not want medical treatment, it is not necessary for that person. However, when a transgender person requests hormonal or surgical treatment from a doctor because treatment will alleviate their gender dysphoria, that treatment is **medically necessary** for them. Scientific studies have shown that providing transition-related treatment has health benefits for transgender people, and that denying such care causes harm.	For an intersex person, medical treatment related to their hormones, genitals, or reproductive organs is medically necessary *either when* (1) there will be adverse physical health effects on the intersex person if the procedure is not carried out, e.g., if a child born with no urinary opening needs one constructed so that urine can leave the body, or (2) an intersex person is able to provide informed consent and requests the treatment in order to alleviate gender dysphoria or feel more at home in their body. It is ***not* medically necessary** to perform genital or gonadal surgery on an intersex child without their consent when the goal is to make their body look more typically male or female and/or to make their medical providers or parents feel more comfortable. **No scientific studies have shown benefit to intersex people from medically unnecessary surgery, but such surgeries are known to cause harm.**

CERTIFICATE OF FILING AND SERVICE

I certify that on November 8, 2019, I filed the **BRIEF OF AMICUS CURIAE TRANSGENDER LAW CENTER, INTERACT, AND BEYOND BINARY LEGAL** with the State Court Administrator by the Court of Appeals' eFiling system.

CERTIFICATE OF SERVICE

I certify that on November 8, 2019, I served this **BRIEF OF AMICUS CURIAE TRANSGENDER LAW CENTER, INTERACT, AND BEYOND BINARY LEGAL** on the following parties via the Court of Appeals' eFiling system:

Lorena Reynolds The Reynolds Law Firm 555 NW Fifth Street Corvallis, Oregon 97330	Sara Kobak Schwabe Williams & Wyatt PC 1211 SW Fifth Avenue, Ste. 1900 Portland, Oregon 97204
Attorney for Petitioner-Appellant	*Attorney for Amicus Curiae Basic Rights Oregon, Inc., and American Civil Liberties Union of Oregon Inc.*

s/ John C. Clarke
John C. Clarke, OSB No. 153245
john.clarke@millernash.com

Attorney for Amicus Curiae
Transgender Law Center, interACT,
and Beyond Binary Legal