REPORT OF THE BOARD OF TRUSTEES

B of T Report 7-I-16

Subject: Supporting Autonomy for Patients with Differences of Sex Development (DSD)
(Resolution 3-A-16)

Presented by: Patrice M. Harris, MD, MA, Chair

Referred to: Reference Committee on Amendments to Constitution and Bylaws
(John P. Abenstein, MD, Chair)

At the 2016 Annual Meeting, the American Medical Association (AMA) House of Delegates referred to the Board of Trustees Resolution 3-A-16, "Supporting Autonomy for Patients with Differences of Sex Development (DSD)," introduced by the Medical Student Section. Resolution 3 asked:

> That our AMA affirm that medically unnecessary surgeries in individuals born with differences of sex development are unethical and should be avoided until the patient can actively participate in decision-making.

Testimony was largely in favor of referral. Those offering testimony understood the key developmental issues surrounding individuals born with DSD. However, testimony revealed gaps in understanding about how to address appropriately surgical and medical options in providing care, necessitating a call for further study.

BACKGROUND

The term "differences of sex development" (DSD) refers to congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical [1]. The frequency of DSDs varies with etiology [2], but overall incidence of DSD is estimated to be one in 5,500 births; some 60 percent of affected children are now diagnosed prenatally [3]. Diagnosis of DSD is complex, encompassing family and prenatal history, physical examination (particularly of genital anatomy), and various laboratory tests, including determination of chromosomal sex. Diagnosis may also involve ultrasound or other imaging studies, hormonal stimulation tests (eg, human chorionic gonadotropin or adrenocorticotropin stimulation), and, in rare cases, laparotomy or laparoscopy [3]. Not all cases of DSD are diagnosed perinatally.

DSD include potentially life-threatening developmental anomalies that may require immediate intervention, for example, hypotension resulting from salt-wasting nephropathy, which occurs in 75 percent of infants born with congenital adrenal hyperplasia. DSD also includes "cosmetic" abnormalities for which elective interventions to normalize appearance can be undertaken at various stages in the child's life [2,4].

Historically, assigning gender in a newborn with ambiguous genitalia has been viewed as a "medical emergency," with immediate surgery recommended to match genitalia to the assigned gender, on the rationale that uncertain gender is distressing for the family, may adversely affect the child's mental health, and can lead to stigmatization [3,5]. This view has been increasingly challenged [2,4,6]. DSD communities and a growing number of health care professionals have condemned such genital "normalizing," arguing that except in the rare cases in which DSD presents as life-threatening anomalies, genital modification should be postponed until the patient can meaningfully participate in decision making [4,7,8].

In 2006, the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) observed the lack of sufficient data to guide decisions about gender assignment and absence of clear guidelines for clinical practice [9]. The NIDDK also noted that there are only limited long-term outcome data on early surgical reconstruction, despite concern about irreversibility and possible sensory damage to the genitalia. Finally, the NIDDK cited a lack of "systematic outcome data about sexual function in individuals with disorders of sexual differentiation [sic]" and of data "pertaining to the association of sexual function with genital appearance and types of genital surgery." It therefore called for prospective studies of gender identity, reproductive function, and quality of life for patients with DSD "to guide clinicians and families in making decisions about gender assignment and surgical reconstruction."

Also in 2006, the Intersex Society of North America (ISNA) released its "Clinical Guidelines for the Management of Disorders of Sex Development in Childhood," gathering perspectives of treating physicians, past patients, and parents who have been involved in the management of DSD [1]. The guidelines address appropriate treatment options for common genital anomalies, focusing on patient- and family-centered care provided by a well-trained multidisciplinary team. The guidelines acknowledge that each patient requires unique attention and resources. Importantly, ISNA guidelines note that gender assignment "is a social and legal process *not requiring medical or surgical intervention*" (original emphasis) [1].

A small study carried out in 2011-2012 among medical students in Zurich found that how physicians discussed treatment for a child with DSD influenced the choice for or against surgery, despite respondents' belief that their personal attitudes governed decision making [10]. Participants watched brief counseling videos that offered either a "medicalized" or "demedicalized" approach. That is, the video described DSD as a condition that is static, has an inherent psychosocial component, and requires treatment, and for which predetermined treatment regimens focus on biological function, or as a dynamic disorder characterized by context-dependent impairment for which coping strategies should be fostered, with treatment geared to the individual's interests and capabilities. Sixty-six percent of participants who viewed the medicalized video said they would choose early surgery for their child, compared to 23 percent of those who viewed the demedicalized video.

CURRENT AMA POLICY

Current AMA policy does not address treatment for patients with DSD directly. Rather, a limited number of ethics and House policies speak to decisions for minors more broadly, as well as to issues pertaining to gender identity, sexual orientation, transgender health, and discrimination toward sexual minority communities:

- Opinion 2.2.1, "Pediatric Decision Making," encourages involving minor patients in decision making at a developmentally appropriate level, including decisions that involve life-sustaining interventions, and recommends that clinicians work with parents or guardians to simplify complex treatment regimens for children with chronic health conditions.
- Opinion 2.2.4, "Treatment Decisions for Seriously Ill Newborns," articulates the considerations that must be taken into account when addressing emotionally and ethically challenging cases involving newborns, including: the medical needs of the child; the interests, needs, and

resources of the family; available treatment options; and respect for the child's right to an "open future." It calls on physicians to inform parents about available therapeutic options and the nature of those options and to discuss the child's expected prognosis with and without intervention.

- Opinion 2.2.5, "Genetic Testing of Children," identifies conditions under which physicians may ethically offer genetic testing for minor patients. It observes that testing implicates important concerns about the autonomy and best interests of the minor patient and holds that medical decisions made on behalf of a child should not abrogate the opportunity to choose to know his or her genetic status as an adult.
- H-525.987, "Surgical Modification of Female Genitalia," opposes medically unnecessary surgical modification of female genitalia and encourages the development of educational programs to address complications and corrective procedures.
- H-475.992, "Definitions of 'Cosmetic' and 'Reconstructive' Surgery," distinguishes cosmetic surgery, performed on normal bodily structures to improve patient appearance, from reconstructive surgery, performed on abnormal bodily structures to improve function or approximate normal appearance.

DECISIONS FOR PEDIATRIC PATIENTS

Parents (or guardians) are granted the authority to make health care decisions for their minor children when the child lacks the ability to act independently or does not have the capacity to make medical decisions [11]. Parents are deemed to be in a better position than others to understand their child's unique needs and interests, as well as their families', and thus to be able to make appropriate decisions regarding their child's health care. Historically, the best interest standard has predominated as the appropriate decision-making standard for medical decisions for minors. Current consensus rests on a more nuanced view that encompasses not only the patient's medical interests, but psychosocial and familial concerns as well [11].

The "harm principle" has been suggested as a further refinement on the decision-making standard, requiring not only that decision makers consider the patient's best interests, broadly understood, but also that a threshold of harm be identified, below which decisions should not be tolerated [11]. Parents (or guardians) are also recognized to have a responsibility to foster their children's autonomy and moral growth, a responsibility clinicians share. Providing information in a developmentally appropriate way that respects the minor patient's cognitive ability, engaging the child in decision making to the extent possible, and seeking the child's assent to proposed interventions helps to fulfill that responsibility [11].

With respect to DSD specifically, it has been suggested that decisions should seek to foster the well-being both of the current child and the adult he or she will become; respect the rights of patients to participate or make decisions that affect them; and foster family and parent-child relationships [4].

In cases of DSD, decisions about a child's best interests and appropriate interventions involve sensitive issues of sex, gender, and sexuality, and interventions that may be irreversible. Parents are often concerned about the future well-being of their child with regard to self-identity, relationships, and reproductive capacity [7]. Because of these concerns, they may be quick to want to establish sex and gender identity for their child in order to promote "normalcy" and reduce stigmatization. Moreover, when physicians perceive early intervention to be urgently needed or wholly beneficial, they may not fully recognize that there is a decision to be made, or the complexity of that decision for the family and patient.

A 2013 lawsuit, though unsuccessful, raised constitutional issues with respect to early surgical intervention and sex assignment. In 2013, the adoptive parents of a South Carolina child, MC, born with “ovotesticular DSD” filed suit in the US District Court for the District of South Carolina against physicians who had performed feminizing genitoplasty on the child at age 16 months. At the time of surgery, MC was under the legal custody of the South Carolina Department of Social Services, which authorized the intervention. Despite initially being raised as a girl by his adoptive parents, consistent with his surgically assigned sex, MC identified as a boy and at the time the lawsuit was filed was living as a boy. Because of the surgery, MC is now sterile. Although the action was dismissed on appeal by the US Court of Appeals for the Fourth Circuit (in January 2015) [12], the lower court had denied the defendants’ request for dismissal on the grounds that the defendants may have violated MC’s constitutional right to procreate [13].

RECOMMENDATION

The Board of Trustees recommends that the following be adopted in lieu of Resolution 3-A-16 and the remainder of this report be filed:

> That our American Medical Association support optimal management of DSD through individualized, multidisciplinary care that: (1) seeks to foster the well-being of the child and the adult he or she will become; (2) respects the rights of the patient to participate in decisions and, except when life-threatening circumstances require emergency intervention, defers medical or surgical intervention until the child is able to participate in decision making; and (3) provides psychosocial support to promote patient and family well-being. (New HOD Policy)

Fiscal Note: Less than $500.

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13. M.C. v. Aaronson, No. 2:13-cv-01303-DCN (D. S.C. Aug. 29, 2013) (order denying motion to dismiss).