

## **Position Statement on Genital Surgery in Individuals with Differences of Sex Development (DSD)/Intersex Traits**

The mission of the Pediatric Endocrine Society (PES), which has over 1300 members in pediatric endocrinology and a variety of other disciplines, is to advance and promote the endocrine health and well-being of children and adolescents. This position statement addresses surgical management for patients with differences of sex development (DSD)\* and intersex traits and advocates for a process that 1) includes providing patients and families with full evidence-based information about all potential options, including not performing any genital surgery, and potentially associated risks and benefits, 2) involves shared decision-making between providers, parents and patients, and 3) is not restricted by government- or advocacy group-set limitations.

DSD are defined as conditions in which the development of chromosomal, gonadal, and/or anatomic sex is atypical.<sup>1</sup> In this statement, we specifically address the subset of DSD conditions that affect the development of the genitals and reproductive organs. These encompass a large variety of diagnoses, including androgen insensitivity syndrome (AIS), congenital adrenal hyperplasia (CAH), 5 $\alpha$ -reductase deficiency, gonadal dysgenesis, ovotesticular DSD, and many others. Care of a patient with a DSD condition must take many individual factors into consideration, including the patient's age and cognitive development, the specific condition and the resulting physical features, and implications of the condition for present and future health and gender development (see Appendix 1 for a more extensive list of considerations).

In this context, important concerns have been raised about the appropriateness of performing surgical procedures on the genitals or reproductive organs, many of which are irreversible, in children who are too young to participate in the decision. When preparing for joint decisions on surgery, the parents should be provided with detailed information not only on the general risks of anesthesia and surgery (such as blood loss), but also on the experience of the surgeon and team, diversity of potential surgical options, the goals and specific risks for each of these options, and the potential outcomes of either performing or deferring surgery. They should also be made aware that individuals with DSD are more likely than the general population to identify with a gender different from the sex designated at birth and/or the gender in which they are raised. When surgery is planned at later stages of development, the same should be done with the patients themselves, on a level commensurate with their cognitive development, so that they can participate fully in the decision process. Potential surgical options for individuals with DSD conditions and associated goals and risks are listed in Appendix 2.

The available data are too limited to allow a fully evidence-based balancing of the above risks and benefits. Some published surveys indicate that adult patients support genital surgery before the age of consent, but such opinions may differ based on the type of procedure and diagnosis.<sup>2-5</sup> Furthermore there is no robust outcome data that individuals with DSD who have

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\* This document uses the term "differences of sex development", but acknowledges that this term is embraced by some but alienating to others.

undergone surgical revision of their genitalia are any more or less psychologically healthy than those who have not. Personal and anecdotal reports from families and patients should not be discounted when considering the potential risks of these procedures. There remains a need for further research that will help teams provide the best care for each individual, including input from patients and family members who have experienced similar decisions. However, even if ample data were available, optimal care for a given individual must still take into account that patient's unique diagnosis, biology and anatomy, personal goals, family values, and social and cultural context.

Given the deeply personal nature of genitalia, gender identity, sexual function, and fertility, it is not surprising that opinions about surgery vary among patients, their parents and other caretakers, support organizations, advocacy groups, clinicians, medical societies, and other parties (such as former US Surgeons General and the United Nations Office of the High Commissioner for Human Rights). Discussions that focus solely on potential benefits or solely on potential harms of these procedures, or on anecdotal experiences, can result in oversimplification of the issues.

Therefore, the PES believes that there can be no single approach to individuals with DSD conditions. The PES opposes government bans on genital surgery for DSD because legislation cannot integrate the myriad of factors that determine the choices for any specific individual. While deferring surgery may be the right decision for one person, a deferral may result in adverse mental health outcomes or suboptimal surgical outcomes in another individual. Thus, in an effort to protect some individuals, a ban may deny benefit to other individuals.

Instead, the PES believes that individuals with DSD deserve a comprehensive, individualized approach to care, ideally provided by a multidisciplinary team of expert and experienced providers that includes not only endocrinologists, but also surgeons (urologists, gynecologists, pediatric surgeons), mental health providers, geneticists, radiologists, clinical and anatomic pathologists, ethicists, and patient advocates.<sup>6,7</sup> The PES presents the following general principles:

- Caregivers and patients (when cognitively capable of providing informed consent/assent) should be presented with all surgical options, with discussion of:
  - Each type (or component) of surgical procedure as a separate procedure and choice
  - The options not to perform or to defer each procedure
  - The potential risks and benefits of each option, including the risks of surgical complications and the expected requirement for revision surgeries
  - The possibility that the future gender identity and expression, desire for sexual intercourse, and/or desire for fertility for the individual may be different from that anticipated at the time of the discussion
  - The influence of surgical timing, understanding that for some procedures, deferral of surgery to an older age may result in a surgery that is more technically difficult and a recovery phase that is prolonged and/or more painful
  - Decisions about surgery are rarely urgent and discussions leading to a decision should take place over many visits and over time
  - Awareness of different viewpoints and debates surrounding decisions (and decision-makers) for surgical management of individuals with DSD

- When surgery is considered at later stages of development and deferral does not introduce technical challenges or other risks (e.g., stigma), similar informed consent must be undertaken with the patients themselves, on a level commensurate with their cognitive development, so that they can participate fully in the decision-making process.<sup>†</sup>
- Decisions about surgery may be facilitated by a shared-decision making model (i.e., a model utilizing an open discussion of evidence-based knowledge with providers, parents and patients to come to joint treatment decisions). This approach can be particularly valuable for clinical decisions marked by considerable uncertainty. This allows clinicians and caretakers to begin these discussions early and continue to have these discussions over time to consider the best available evidence, the specific clinical diagnosis and features of the patient, and the values of the patient, family, and surrounding knowledgeable community to achieve an informed decision.
- Discussions about whether or when to perform surgery should include multiple members of the expert team in addition to the surgeon (e.g., the endocrinologist to discuss gonadal function and the mental health professional to assess understanding of options and individual social/cultural context).

The details outlined in this statement and appendices demonstrate the complexity of the issues involved in decisions of whether and/or when to perform surgery on individuals with DSD. The PES believes that legislation cannot adequately synthesize these factors and rather may inadvertently harm some of the individuals it seeks to protect. Rather than address these issues through governmental restrictions, the PES believes in providing complete and evolving information to patients and families and engaging in a shared decision-making process with expert teams, supports ongoing reporting of results and outcomes no matter what surgical decisions are made, supports working together as interdisciplinary teams to share data to guide these difficult decisions, supports efforts to raise resilient individuals who have genital differences, advocates for advances in acceptance by the broader society of individuals with physical differences and/or non-binary gender expression and will continue to evaluate the role of surgery for individuals with DSD.

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<sup>†</sup> For example, the gonads of individuals with complete androgen insensitivity syndrome appear to have a higher risk for developing tumors, but this risk does not arise until the late teenage years. The practice of gonadectomy in infancy for patients with CAIS has, therefore, largely been replaced by gonadectomy in adolescence or later after discussion with the patient and caretakers.

## References

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## Appendix 1

### Important Considerations for an Individual with DSD

- Age (and degree of cognitive development) when the patient comes to medical attention, which could be before birth, at birth, or in infancy, childhood, adolescence, or adulthood
- Physical features resulting from the DSD condition, which may include:
  - typical genital appearance or genital difference
  - presence or absence of internal reproductive structures, such as a uterus
  - separation or fusion of the urinary and reproductive ducts (e.g., a single fused opening for the vagina and urinary tract or a urogenital sinus)
- Implications for present and future health, including:
  - inability (in some conditions) to produce hormones essential for survival
  - risks (in some conditions) for developing cancer of the gonads
  - inability (in some conditions) to produce hormones necessary for puberty
- Implications for fertility, which may be unimpaired, possible with assisted reproductive techniques, or impossible with currently available techniques
- Gender development, as individuals with DSD are more likely than the general population to identify with a gender different from the sex designated at birth and/or the gender in which they are raised.<sup>‡</sup> For some conditions, long term gender identity outcomes are largely consistent across affected individuals, while other conditions are associated with much more variability.

## Appendix 2

### Potential Surgical Options for Patients with DSD

- Gonadectomy or partial gonadectomy: removal of all or part of the gonads (gonads may be testes, ovaries, “streak gonads” that do not have an obvious testicular or ovarian appearance, and/or ovotestes that contain both testicular and ovarian tissue)
  - Potential positive effects may include:
    - reducing the risk of developing cancer of the gonad (elevated risk in some DSD conditions)
    - preventing the development of undesired secondary sex characteristics by preventing production of sex hormones (e.g., testosterone production in someone who identifies as female)
  - Potential negative effects may include:
    - infertility (if fertility potential is initially present)

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<sup>‡</sup> The terms below were used in this document as follows (but do not represent a formal definition):

-“Sex” or “designated sex” meaning biologic sex

-“Gender” meaning legal label or gender identity

- inability to produce sex hormones, necessitating sex-hormone replacement
  - reduction in maximal bone mineral density and challenges maintaining bone mineral density in adult life
- Reduction in size of the clitorophallus (clitoris, penis, or related structure)
  - Potential positive effects may include:
    - reducing social stigma and distress related to atypical genital appearance (which may be influenced by factors including the values of the family, their cultural context, and available psychosocial supports)
    - avoiding interference with sexual function
  - Potential negative effects may include:
    - irreversible reduction in size of the clitorophallus that may be considered undesirable by some individuals (e.g. a person later identifying as male)
    - loss of erectile tissue
    - reduction or loss of sexual sensation
    - scarring with associated pain
- Hypospadias repair, to bring the opening of the urethra to the tip of the penis
  - Potential positive effects may include:
    - reducing distress related to atypical genital appearance
    - allowing urination while standing
    - facilitate fertility by allowing deposition of ejaculate into the vaginal canal
  - Potential negative effects may include:
    - multiple staged operations are generally required
    - development of a fistula (undesired opening) or stenosis (blockage) that may require additional surgical correction
- Vaginoplasty, to construct a vagina with typical appearance and function, usually combined with additional procedures to create separate openings for the vagina and the urinary system if they are initially fused
  - Potential benefits may include:
    - preventing urine from entering the reproductive tract, which has the potential to cause incontinence and infections
    - allowing for receptive vaginal intercourse in the future
  - Potential negative effects may include:
    - vaginal strictures (narrowing), which may require manual dilations and/or additional surgical correction
    - urinary tract infections
    - urinary incontinence
    - scarring with pain during sexual activity