“Sex is like an inside joke that everyone gets but me:”

Controversies in feminizing genitoplasty for classical congenital hyperplasia

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Introduction

The most common surgery performed in children with the difference or disorder of sex development (DSD) known as 46,XX classical congenital adrenal hyperplasia (CCAH) is feminizing genitoplasty (FG) for genital difference. FG, often performed in early childhood before children can assent or consent, consists of procedures to change the appearance of the external genitals to be more “female,” and to separate the urethra and vagina (vaginoplasty). In salt-wasting (SW) CCAH, genetic alteration of 21-hydroxylase enzyme function causes life-threatening decreases in both cortisol and aldosterone; in a milder form, “simple virilizing” (SV) CCAH, only cortisol is diminished. In both forms, reductions in enzyme activity lead to diversion of steroid precursors into pathways causing more androgens than usual to be produced, leading to variable genital differences. Differences range from a clitoris that is slightly larger than usual to development of a penis, scrotum, and urogenital sinus with a single external opening from the vagina and bladder. This interactive visual aid demonstrates how atypia occurs along a continuum (Prader 1-5).

There are ongoing controversies regarding the necessity and timing of surgery for genital difference in CCAH. In July 2017, Human Rights Watch released a report, "*I Want to be Like Nature Made Me,"* finding that current surgical practices in intersex/DSD lack evidence-based scientific justification and violate children's rights to self-determination and bodily autonomy.[1] Critics of the report point out that absence of evidence is not evidence of absence. Since there are many studies on CCAH, specific arguments have been made in favor of early FG including:

1. Advocates just want to ban all surgery
2. Intersex arguments don't apply to CCAH because parents reject DSD and intersex terminology.
3. Early FG promotes parent bonding, prevents stigma and has psychosocial benefits.
4. The consequences of growing up with intact genital difference are unknown.
5. Feminizing genitalia before gender identity is apparent is safe because the rate of gender dysphoria in CCAH is insignificant.
6. Data support satisfactory results of early vs. late FG
7. Adults and parents prefer early FG over later consensual FG
8. The complications of early FG observed in older children and adults are a result of outdated procedures; outcomes will be better when expert surgeons use today's neurovascular-sparing procedures
9. Parents are fully informed regarding the risks and benefits of FG.
10. Ethicists agree that parents act in their children's best interest and have the right to consent to early FG

In response to these specific points:

1. Advocates just want to ban all surgery

While surgery may be helpful in some specific situations, criteria to define those who will benefit have not been defined. Portrayal of activists by groups such as the Societies for Pediatric Urology (SPU) as demanding a "complete moratorium" on all childhood genitourinary surgery [2] eclipse advocates' primary mission: the development and implementation of effective psychosocial interventions, including routine inclusion of peer

support prior to irreversible decisions when the contemplated procedures violate human rights standards. [3] Despite a 2006 international Consensus Statement on Management of Intersex Conditions that stressed psychosocial support, and a recent practice survey of US multidisciplinary teams reporting psychologists/psychiatrists as always available or available by consult, 1/3 of families lack routine access to psychological services. [4-6] A limited number of behavioral health providers have specialized training and experience, reimbursement is lacking, and few centers provide services promoting acceptance of differences. [3]

2. Intersex arguments don't apply to CCAH because parents reject DSD and intersex terminology.

Language used to refer to DSD/intersex is a controversial area. Clinicians sometimes criticize the use of intersex/DSD terminology because a physician survey of Congenital Adrenal Hyperplasia Support Research Education and Research (CARES) Foundation members found that a majority of respondents disliked the term DSD and found it harmful. The significance of the study is questionable, since only 8% had heard the term previous to being asked about it for the study. [7] Clinicians, researchers, and people living with CCAH may use the terms DSD and intersex to refer to CCAH, and both will be used here interchangeably. [8-11]

3. Early FG promotes parent bonding, prevents stigma, and has psychosocial benefits.

According to proponents of early FG, its purpose is **"**to avoid stigmatization related to atypical anatomy, to respond to the parents' desire to bring up a child in the best possible conditions, [and] to restore functional genital anatomy to allow future penetrative intercourse (as a male or female)."[12] Other "benefits" of early FG include promotion of parental bonding and prevention of future psychosocial issues.

Although intersex infanticide is routine in some cultures outside of the United States,[13] adverse effects of genital difference on parent-child bonding in the United States are not documented. In a small study of French families who deferred early surgery in favor of endocrine treatment, genital difference did not impair bonding.[14]

Several recent studies documented that stigma was experienced by adult women with CCAH in a variety of settings, but it should be noted that this stigma was not primarily due to genital difference. Stigma experienced by nearly 2/3 of adult in the general social environment was related to obvious physical differences, such as hirsutism or a deep voice, rather than genital difference.[10] 25% of the same women reported that doctors' actions frequently resulted in stigma, mostly via frequent genital exams in teaching settings.[9] This practice has not been abandoned; it continued in contemporary multidisciplinary clinics whose practices were recently surveyed by the NIH-funded Translational Research Network (TRN), where 30% still performed genital exams for teaching.[5] Sexual and romantic stigma were experienced by 40% of the women studied, whether they had surgery (the majority) or not, but nearly all women described maladaptive coping in interviews, including secrecy, hiding genitalia, sex avoidance or abstinence, and substance abuse.[11] Rather than being a result of genital difference, shame can result from the mere fact of having genitals that "required surgery," suggesting significant iatrogenic moderators of the relationship between genital difference and sexual/romantic stigma.[11]

There are few studies using DSM criteria for psychiatric diagnoses. In a study of children with CAH, girls (surgical status unspecified) had twice the control rate of DSM-diagnosed anxiety disorders, while boys without genital difference had increased rates of ADHD, disruptive behavioral disorders and anxiety disorders, indicating that genital difference alone does not explain psychological issues. [15] Data on psychological outcomes in adults are complex and confusing. In Sweden, ICD-9 codes for anxiety disorders were found in twice as many women with CCAH as in the general population,[16] while adults in the UK and Turkey lacked substantially increased risk of psychiatric disorder.[17] Increased anxiety, which is common in chronic illness, is not unexpected in children and adults living with the steroid disturbances of CCAH, and has not been directly associated with genital difference.

Reports of stigma, psychiatric diagnoses and psychometric test results don't capture the full spectrum of challenges to wellbeing in adults, including barriers to intimacy, dynamic identities, and poor education about their medical and surgical histories. [18] Even proponents of FG acknowledge "the need for some form of intersex-related psychosocial intervention [in] some women," [11] but there is little evidence for effectiveness of specific interventions in CCAH. [17] A needs assessment

study of another DSD group, men with congenital hypogonadotropic hypogonadism (Kallmann syndrome), found that patients are receptive to online interventions aimed at addressing their unmet needs, including peer support to enhance coping and promote health.[6] Replacing irreversible surgery with an approach prioritizing psychosocial interventions could address potential consequences of genital difference that are expected to vary with life stages.

4. The consequences of growing up with intact genital difference are unknown.

Claims that there is no evidence on mental health outcomes of children living with discordant genitals ignore well-known research that children with gender dysphoria who have family support for their identities do well psychologically.[19] Outcomes of unoperated children with CCAH are rarely published, but in the French study on families deferring early FG reported "so far girls and their parents have not experienced significant concerns regarding genital ambiguity''.[14]

Although some centers provide anecdotal evidence of adult CCAH patients desiring primary or revision surgery, there are also anecdotes of unoperated intersex adults who are grateful to have been spared infant surgery, such as 60-year-old Jim Costich: "I did not have any genital surgery to make me look any different and… my love life, my social life, my gym life, even my life as a nudist has not been adversely affected!"

Surgery on older children and adolescents must also be approached carefully. As the American College of Obstetrics and Gynecology states in regard to cosmetic labial surgery, psychosocial services and counseling are essential:

"Although reconstructive procedures aimed at correction of abnormalities (caused by congenital defects, trauma, infection, or disease) or cosmetic procedures performed to reshape normal structures may improve function, appearance, and self-esteem, not all adolescents are suited for surgical intervention. Appropriate counseling and guidance of adolescents with these concerns require a comprehensive and thoughtful approach, special knowledge of normal physical and psychosocial growth and development,

and assessment of the physical maturity and emotional readiness of the patient."[20]

5. Feminizing genitalia before gender identity is apparent is safe because the rate of gender dysphoria in CCAH is insignificant.

Publications favoring early FG in CCAH contain statements such as "female assignment is suggested for those with 46,XX and CAH, since 95% develop female gender identity,"[21] or "there is usually no gender issue in this group."[12] In children with CCAH, as in all children, gender identity is a result of "complex, multiple and interactive developmental processes." [22]Liao 2012 It is not fixed at birth, nor is it confirmed by "fixing" genitals with gender-conforming surgery.

A 2015 literature review discredited results of studies on gender identity purportedly supporting surgical reinforcement of female gender assignment because of their limited methodologies; Pasterski et al also prospectively assessed gender ID of 4- to 11-year old children with CAH using mixed methods, including DSM criteria for gender dysphoria (GD).[23] They found that 12% of girls with CCAH met all 5 DSM GD criteria. [23] Pasterski cross gen A 12% rate is equivalent to 1 out of every 8, the same proportion commonly used to describe women's lifetime incidence of breast cancer, which is not considered unusual. It is especially concerning that 30% of female-assigned children with CCAH who had GD actually transitioned genders to live as men in adulthood.[24] More expansive understandings of gender as dynamic and non-binary are needed, as shown by a mixed methods study in adults finding that 26% of adults with diverse DSD, including CCAH, had identities not encompassed in current terminologies. [25] With errors in early childhood gender assignment a significant possibility, deferring surgery in children preserves options for later transition. Irreversible surgery compounds the magnitude of harm from misassignment to catastrophic proportions, as in the removal of a healthy penis from a child subsequently identifying as male.

6. Data support satisfactory results of early vs. late FG

Unsupported assertions of superiority of early FG distract from the glaring truth that pediatric specialists themselves prefer it because they are not trained to perform surgery in older, consenting individuals.[2] Supporters of

early FG agree with detractors that there are currently insufficient data to support assertions that adult women are satisfied with the results of early surgery.[11] Since there is no research directly comparing outcomes of early and late FG, we cannot know which is better, although some gynecologists who perform both primary FG and surgery to treat subsequent complications in older patients advocate for deferral.[26] Six-year follow up of successful single-stage adult genitoplasty with preservation of orgasm was reported by Tjalma in 2016; corpora cavernosa-preserving surgery in a previously-orgasmic woman with CCAH also eliminated the need for revision vaginoplasty because the woman was already sexually active.[27] Failure to acknowledge the concept embodied in comments like “sex is like an inside joke that everyone gets but me” in the design of research undermines the validity of data on sexual satisfaction: without a preoperative basis for comparison, questioning women who had early FG about orgasm may be like asking someone with undiagnosed color blindness about the color red. Using questionnaires that do not collect descriptive accounts of sexual experiences, how do we expect women to respond truthfully about something they may have never experienced? Furthermore, the limited research that has purported to measure the functional outcome of genital surgery is notable for largely neglecting to use validated instruments to measure female sexual dysfunction. Rationales for early FG consistently prioritize heteronormative penetrative intercourse, and quantitative data derived from questioning adults about such intercourse may reflect compliance with medical and social expectations rather than actual desire or enjoyment. The “coital imperative” [28] conflates heterosexual intercourse with sex, devalues other expressions of intimacy and sexuality, and creates additional anxiety.[29] The shame of having genitals that required surgery [11] and negative emotions related to lack of choice may impair sexual wellbeing beyond physical sequelae. [30]

7. Adults and parents prefer early FG over later consensual FG

The oft-repeated contention that women “clearly” prefer early surgery relies on uncritical acceptance of the conclusions of very few studies,[31] and is contradicted by closer scrutiny of the actual study methods and data.[32] For example, participants in studies who “preferred” early FG were not told that not having surgery at all might be an option; that rates of reoperation for stenosis could have been lower if surgery were performed later; or that

significant technical innovations anticipated to improve outcomes had occurred in the years since their early childhood surgery.[32] As for parents, families of young children choosing early FG to whom it was presented as helpful and necessary would be expected to wish it had been done even earlier.[33, 34]

8. The complications of early FG observed in older children and adults are a result of outdated procedures; outcomes will be better when expert surgeons use today's neurovascular-sparing procedures

Data on short-term results of up-to-the-minute early FG techniques used by trained specialists do not support the belief that surgery performed by specialists in multidisciplinary team settings will have low complication rates.[2, 35] One recent study of 6-year follow which reported few complications in young children at a single center did not assess vaginal stenosis, a common major complication usually detected at puberty [36]. A prospective study of short-term cosmetic outcomes of surgery performed in multiple multidisciplinary centers reported a 10% major complication rate in CCAH at just 1 year (labial fusion, vaginal stenosis), with 1 child having already undergone a second operation.[37]

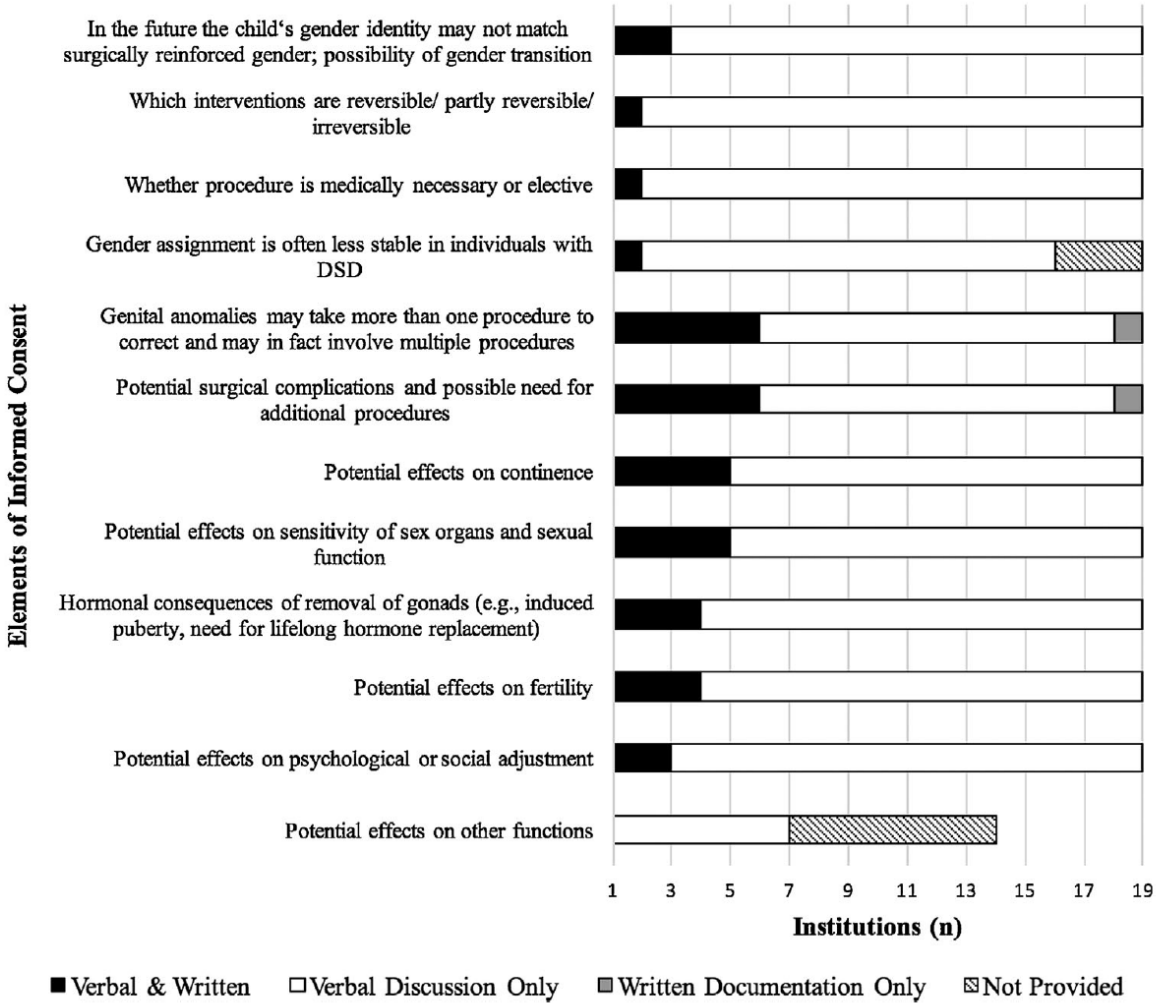
Although work in 1999 by Baskin et al [38, 39] on anatomy of the human clitoris is frequently cited as the foundation of current neurovascular-sparing surgical techniques,[35] it reported typical human anatomy and projected typical male anatomy to apply to "masculinized" atypical female anatomy. Recognizing this shortcoming, since actual specimens of "masculinized" female human fetuses were unavailable, they sought an animal model. The female spotted hyena has a long phallus-like clitoris through which it urinates, copulates and delivers young. Proof of concept was reported when female hyena anatomy matched predictions of how "masculinization" would affect the developing human clitoris.[40] Surgeons enthusiastically embraced the correlation of hyena and human anatomy, and extrapolated that preservation of anatomic structures would protect sexual sensation and function. In a subsequent study of 27 clitoral tissue specimens removed during nerve-sparing clitoroplasty that showed branches of the dorsal nerve in 23 specimens, these nerves were dismissed as insignificant even though there were no functional data for correlation.[41] Claims of intact postoperative clitoral sensation with

current early FG techniques have yet to be confirmed;[34] long-term outcomes of similar, frequently-updated techniques in nerve-sparing prostatectomy indicate a need for caution.

9. Parents are fully informed regarding the risks and benefits of FG.

In a recent online post, the SPU states that “if surgery is considered, complete informed consent with counseling and support should be provided prior to proceeding with any surgical intervention.” [2] In reality, the current lack of psychosocial support means families often consent to surgery in a state of emotional distress that impairs cognitive processing of information [42], and without a full understanding of the controversies surrounding these procedures including that they have been condemned by international human rights bodies such as the United Nations, the World Health Organization, and Physicians for Human Rights. Absence of a feasible psychoeducational care pathway leaves families “between a rock and a hard place,” with no meaningful alternative to surgery.[43] As one parent of a child with CCAH said, “It's close to no choice… we figured that it had to be done.”[44]

For the TRN study of clinical practice, intersex advocates created a list of key points of information to be discussed with families considering genital or gonadal surgery for their children which was used to survey centers on informed consent practices. While centers believed they had discussed most of these points, this chart summarizes how few actually documented what they told parents, especially regarding medical necessity, irreversibility, and gender uncertainty. [5] (Reproduced with permission from Aimee Rolston.)



In such retrospective surveys, the contents of discussions are subject to recall by families and clinicians. Without a formal process, guidelines, or documentation, there is a possibility that many parents may not have received important information. Even if they did, only half the centers imposed a thinking period before surgery to allow families to assimilate complex information.[5] In another prospective study of postoperative cosmesis that did not specify elements of informed consent, 40% of mothers and 50% of fathers who were invited to participate were satisfied with the preoperative appearance of their children's genitals, while 100% of surgeons were dissatisfied/very dissatisfied.[45] Despite the rate of parental satisfaction, 96% of families agreed to surgery. [45] Rates of consent that parallel surgeons' rather than parents' dissatisfaction with appearance may reflect surgeons' attitudes toward necessity, raising questions of how "informed" consent actually is in the face of surgeons' optimism regarding outcomes.

Parents should also be aware of the 2017 FDA safety warning regarding neurotoxicity recommending that “consideration should be given to delaying potentially elective surgery in young children where medically appropriate.”[46] As developed at Texas Children’s Hospital, a procedure including a discussion in which “the possibility that the procedure could be delayed until after 3 years of age” is suggested. [47]

10. Ethicists agree that parents act in their children’s best interest and have the right to consent to early FG

The ethics of FG are subject to fierce ongoing debate, and leading experts in the field condemn early FG.[48-50] 2010 articles by Wiesemann [51] and Gillam [52] are mentioned as supportive of early FG. Advocates of deferring surgery agree on basic ethical principles of treatment described by Wiesemann et al: fostering the wellbeing of the child and future adult; upholding the rights of children and adolescents to self-determination; and respecting family relationships.[51] Difficulties in actual management arise because these principles often conflict, particularly with respect to the value assigned to parental authority vs. children’s rights to autonomy. Wiesemann et al highlight this conflict in recommendations that “the family environment, the cultural context, and the preferred value system of the affected family must be given due consideration,” and that absent “a compelling medical indication… interventions that might have irreversible consequences for the person’s sex or negative consequences on their sexuality or reproductive capability… should be left up to the affected persons themselves.” [51]

SPU affirms that “societal norms do not dictate whether a child may be a candidate for surgery,” but doctors are an important repository of the beliefs and values that reflect societal norms [2] Overwhelmed families who may not have previously considered their feelings about genital difference are distressed, anxious, and protective toward their children [44, 53-55]. In a vacuum of previous experience with genital difference, parents are vulnerable to absorbing unfounded clinician beliefs such as the possibility Gillam et al describe of a “child… not [being] accepted by parents in the chosen sex of rearing, leading to impaired bonding; … of social or cultural disadvantage to child, for example, reduced opportunities for marriage or

intimate relationships or reduced opportunity for meaningful employment and capacity to earn an income; [and] of social isolation, restrictions or difficulties, for example caused by embarrassment or social stigma associated with having genitalia which do not match the gender in which the person lives."[52] Also conveying a vague sense of pessimism about a child's ability to live in the world are cryptic concerns recently expressed by international experts regarding the impact surgically untreated genital difference may have, not only on parents and children, but on "society."[12, 21] There has been no systematic investigation of the foundation of physician attitudes, but in *Fixing Sex*, Karkazis found that some physicians were disgusted by ambiguous genitalia.[48] Professional organizations might promote exploration of unconscious feelings and biases by clinicians who care for these children.

Since the papers discussed above were published, times have changed, with recent ethical and legal shifts including international bioethics institutions and human rights organizations such as the United Nations, World Health Organization, Amnesty International, and Physicians for Human Rights calling for recognition of intersex children's rights to bodily autonomy and postponement of genital surgery until they can decide for themselves.[56] NIH started an Office of Sex and Gender Minority Research in 2015 [57] and convened a workshop on DSD research. The ethicist NIH consulted for the workshop concluded that "[b]ecause children born with DSD have a right to an open future, and because the openness of their future is clearly enhanced by delaying cosmetic genitoplasty until they themselves can participate meaningfully in decision-making, early genitoplasty is ethically supportable only when medically indicated (e. g., when the child is unable to urinate without surgical intervention)."[50]

Human rights considerations provide a framework for understanding why parents who make all kinds of medical decisions for their children should not be able to approve genital surgery. Within the concept of self-determination, genital autonomy is a special kind of bodily autonomy because, unlike other body parts, genitals are personal, private, and central to sexual expression. Parents wishing to protect children from stigma cannot know what kind of sexual experiences their children will want to have when they are older; because it cannot be undone, genitoplasty on a child who is too young to consent also deprives the adult of genital autonomy.[30] Adult genital autonomy is central to adult rights to sexual and reproductive autonomy.

Female genital mutilation and FG are sometimes compared because, although they are performed for very different reasons, either can result in anger and resentment over their imposition at a time when children are too young to understand. Powerful negative emotions can impair sexuality beyond the purely physical sequelae of either intervention.[30] While noninvasive “pricking” of the clitoris of Muslim girls is prohibited because it is culturally motivated, surgeons admit that they sometimes perform FG because of “cultural concerns.” This suggests a troubling double standard of “acceptable” and “unacceptable” cultural motivations based on race, ethnicity or immigration status.[30]

Conclusion

Consensus on what constitutes safe and effective FG for genital difference in 46,XX CCAH is a controversial holy grail that has remained just beyond reach for decades. Recent studies of FG do not demonstrate better physical or psychosocial outcomes as a result of expert training or improvements in technique. A focus on surgery as a primary intervention has limited investigation of psychosocial interventions. While absence of evidence is not evidence of absence of benefit for surgery, surgery is irreversible, and psychosocial interventions are individualized and dynamic. Today’s surgery is an experiment whose results will not be known for 15-20 years, and the risk of disappointing results will be borne by those who will be harmed for a lifetime. That unconsented FG continues in young children is a disservice to the field of medicine and to the patients we serve.

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