

## LETTERS TO THE EDITOR

**Re: Editorial: It is (Sort of) a Boy and (Sort of) a Girl. You have (Sort of) a Say and You (Sort of) Don't? The Uneasiness of Genital Restoration Surgery [JPEM 2006(11); 19: 1285-1289]**

**To the Editor:**

As a physician and mother of two young adult daughters with complete androgen insensitivity syndrome (AIS), I have counseled a group of 50 parents of children with AIS and related disorders of sex development (DSDs) for 8 years. These parents consistently express frustration over the lack of evidence from which to make choices for the treatment of their children. Decisions like the advisability of genitoplasty are fraught with anxiety because reliable outcome data are scant. The recent international consensus statement detailing a new patient-centered standard of care for DSDs laments this lack of research<sup>1,2</sup>.

In his editorial, Dr. Rivkees advocates for urogenital sinus repair as “necessary to open up a covered vaginal orifice for menstruation and sexual intercourse”. Genitoplasty is a separate, elective procedure that is not part of routine urogenital sinus repair. Dr. Rivkees describes operations performed on children with cleft palate and clubfoot as analogous to genitoplasty. Ambiguous genitalia, unlike facial and orthopedic anomalies, are not constantly visible differences. While craniofacial and orthopedic surgery can improve feeding and ambulation, Dr. Rivkees does not reveal what function is restored by genitoplasty.

In supporting the performance of genitoplasty, Dr. Rivkees cites the development of neurovascular-sparing techniques, based on principles identical to those used to preserve potency following prostatectomy. The sexual function of men who have undergone nerve-sparing prostatectomy is well-studied. Despite continued surgical refinements, results consistently fail to meet expectations. Recent data show that even when skilled surgeons at a large tertiary-care academic center performed anatomically-based, nerve-

sparing prostatectomy, only 62% of patients regained full potency at 2 years of follow-up<sup>3</sup>. Similar scientific evidence regarding the retention of adult sexual function after genitoplasty is absent.

Conversely, techniques purporting to preserve the neurovascular structures during genitoplasty have unknown effects on at least two components of sexual function, genital sensitivity and erectile tissues. While genital sensitivity is subjectively evaluated during follow-up of genitoplasty, function of the erectile tissue is not. Reduced vascularity of erectile tissue is a cause of disorders of arousal and orgasm in women. Recent functional MRI studies show a previously unsuspected interconnectedness among the clitoris, labia minora, urethra, and clitoral bulb, including the trabecular tissue, during sexual arousal<sup>4</sup>. Feminizing genitoplasty inevitably disrupts the connections between these sexually-responsive erectile tissues. The functional consequence of these disruptions can be assessed using MRI of patients post-genitoplasty to determine whether actual blood flow in the genital structures remains intact after surgery. Findings during sexual arousal of adults with DSDs who did and did not have surgery should be compared with those in typical women to further illuminate the costs and benefits of genitoplasty. Objective results like these will be welcome to affected families during decision-making and follow-up.

As part of the choice of treatment, physicians and families should consider the effectiveness of counseling. Psychosocial counseling addresses many of the same issues that genitoplasty aims to fix, including the promotion of healthy sexual relationships and positive body image. Counseling develops long-term skills and strategies for children with DSDs to use in these and many other situations, and is complementary to physical treatments.

I applaud Dr. Rivkees' call for federally-funded patient-centered research. Physicians must collaborate with parents and patient organizations to demand long-term study of treatment outcomes for children with DSDs, emphasizing valid comparison

of various treatments, both psychological and surgical. Parents' discomfort with ambiguous choices will never fade away until proposed treatments have been thoroughly investigated and accurately compared.

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### **To the Editor: Managing Variations in Sex Development**

The November 2006 editorial about treatment of children born with ambiguous genitalia proceeds from a problematic premise, and goes on to imbue questions about elective genitoplasty with a rhetoric of optimism that could be misleading to distraught parents.

Dr. Rivkees argues that surgical 'gender reversal' no longer occurs because 46,XX infants with CAH are typically assigned as female regardless of virilization, and 46,XY infants with testes and microphallus are typically assigned as male. However, gender identity development in children born with atypical genitalia is poorly understood<sup>1</sup>. It is certain that neither chromosomes nor gonads decisively determine gender. Patients with complete or partial androgen insensitivity syndrome (AIS), for example, commonly develop a female gender identity; but not always<sup>2</sup>. Parents who are considering irreversibly altering their child's genitals need to understand this uncertainty in order to make an informed decision.

Whether or not gender assignment carries a degree of certainty, however, the larger issue is whether it is acceptable, given the lack of evidence of any benefit, to subject infants to the loss of autonomy and degree of risk involved in any imposed surgeries. A significant number of people born with genital anomalies say it is not. This

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persistent critique distinguishes infant genitoplasty from surgeries to correct clubfoot or cleft palate.

While acknowledging that "we do not have long-term data related to contemporary genital restorative surgical techniques", Dr. Rivkees asserts that "we are now in a bright era" in which it is "possible to achieve both favorable functional and favorable cosmetic outcome". This assessment is not uniformly accepted, and cannot be taken for granted. As recently as 2006 the NIH stated that "there is currently a crisis in clinical management" of children with ambiguous genitalia due to uncertainty about both sex assignment and surgical outcomes<sup>1</sup>. Principles of informed consent require that parents know if the outcome of treatment is uncertain, yet many parents who consent to infant genitoplasty later report feeling that they were under-informed. Parents need to know that questions about the effects of surgery are more than 'lingering'; that many experts now recommend against elective infant genitoplasty due to concerns that any incisions in the genital region may damage sensitivity; that modern surgical techniques were not developed in adherence to principles of evidence-based medicine; and that evidence of the likelihood of complications necessitating repeat surgeries is plentiful<sup>3</sup>. Particularly troubling is orchiectomy that often accompanies the management of those with AIS. While it is true that there is a potential for gonadal tumors the likelihood is less than that of breast cancer for woman, yet no one

recommends prophylactic mastectomy. Watchful waiting is preferred<sup>4</sup>.

Parents also need to know that assumptions that surgery will prevent psychological harm from childhood teasing are unsupported by evidence, and that adults who have experienced genitoplasty in childhood have complained that multiple surgeries and the frequent genital examinations they necessitate also cause psychological harm<sup>3</sup>. Parents who discover such matters after the fact are likely to be dissatisfied.

Some points in the editorial's legal analysis warrant clarification. While U.S. courts do generally "assume that parents know what is best for their children", this presumption is not the final word. Many legal experts now believe that elective genitoplasty is a decision that only the patient can make<sup>5,6</sup>. The Colombian cases referenced in the editorial did not turn on the genotype of the children, nor did they affirm the traditional rights of parents to decide in such cases. Rather, the cases turned on the children's *age*; and actually imposed new limitations on parental consent, intended to ensure that decisions are not made on the basis of parental discomfort or ignorance of alternate treatment models<sup>6</sup>. In America, too, parental discomfort is not a valid basis for surgery on a child. While a desire to relieve this discomfort is understandable, such relief should not come at the expense of giving complete information to the parents. Ultimately, this kind of surgical decision should

involve the individual most concerned: the child, when mature enough to express an informed opinion.

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#### To the Editor:

Dr. Scott Rivkees correctly points out in his editorial, "It is (Sort of) a Boy and (Sort of) a Girl ...." (November 2006), that it is now time to reframe the debate over reconstructive genital surgery in disorders of sexual development, such as congenital adrenal hyperplasia (CAH). The debate has, for the most of the last decade, been defined by a vocal few, and the voices of the quiet many have gone unheard. While the vocal 'intersex' advocates have brought about many needed changes in the realm of care of those with genital anomalies, Dr. Rivkees asks whether it is now time to treat genital

structural anomalies as we do other congenital birth defects. Rather than insisting that societal acceptance without surgical intervention is best for the child, as has been argued by some 'intersex' advocates, perhaps we should be looking at improving outcomes.

Overall, women with CAH, who make up more than 95% of individuals with disorders of sexual development, and their parents do not necessarily agree that unreconstructed societal acceptance is best for girls with CAH, and would support Dr. Rivkees argument. CARES Foundation, Inc., a support and advocacy organization for those affected by CAH, is now giving a voice to those with

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CAH and their families, and allowing this community to define its own needs and desires around the issues of surgical reconstruction of genital anomalies in CAH.

The issues involved are multifaceted and complex. Dr. Rivkees points out that little research has been done to study surgical techniques that improve outcomes. Moreover, those that oppose surgery have not shown that those individuals who have grown up without surgery have better outcomes. In fact, the definition of 'better outcome' is itself ill defined. Is it functional outcome? What is functional outcome? Is it the ability to engage in intercourse or achieve orgasm? Sexual dissatisfaction is common among those without reconstructed genitals, so this is a complicated measure to use. There are many barriers to doing outcome research but nevertheless, if we want answers, this research must be done.

So much has changed thanks, in part, to the attention placed on genital surgery by the 'intersex' advocates. Now, most surgeons and pediatric endocrinologists will agree that minimally virilized girls with CAH should not have surgical intervention. Clitoral reduction should be done conservatively, if at all. Many physicians are learning the importance of parental education about all options, meaningful informed consent, and referrals to surgeons with substantial experience with genital reconstructive surgery. However, significant problems still exist. The American Urological Association and The American Academy of Pediatrics, Urology Division must set standards and define centers of excellence for reconstructive genital surgery for each individual disorder of sexual development, including specifics for training of young surgeons in this specialty. Designating centers of excellence will allow families to get coverage for these complex surgeries out-of-network from managed healthcare and Medicaid.

All children deserve high quality surgical care, no matter their financial circumstances. Right now, it is not always available to those without financial means and the best insurance plans.

Parents of girls with CAH make decisions about surgery based upon a variety of factors. While cultural background may make some more likely to seek early surgical intervention, the degree of virilization seems to be the most critical determinant. Despite the best efforts of the vocal 'intersex' advocates over the last decade to change parental decision-making, very few parents of significantly virilized CAH girls, 4 or 5 on the Prader scale, have made the choice to defer surgery beyond early childhood. This speaks to an inherent desire on the part of these parents to help their girls live a normal life, minimizing the physical manifestations of their disease. Some adults with CAH whose parents waited to allow their children to undergo surgery until they were teens/pre-teens have expressed the sentiment that they wished their parents had done it earlier and saved them their perceived shame of being 'different'. They longed to be "like other girls". This is a common refrain.

As Dr. Rivkees postulates, discomfort with genital anomalies will not fade - not among the parents, those affected or society. With that said, we owe it to the children to help them adapt to the life they will live - today - and that may include surgical reconstruction. We owe them the best quality surgical reconstruction available, and we must not be overly distracted by the pull from the far-left. Pursuing outcome research and designation of centers of excellence will help us move closer to that goal.

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