The Surgery Controversy Continues...

In the July 2003 edition of the Journal of Clinical Endocrinology and Metabolism, two Letters to the Editor debated the recommendations on early reconstructive surgery for virilized CAH girls contained in the 2002 CAH Consensus Statement. We obtained permission from the Journal to reprint these letters for you to read in their entirety. Please note that the CAH Consensus Statement is now available online for free at http://jcem.endojournals.org/cgi/content/full/87/9/4048 We thank the Journal of Clinical Endocrinology and Metabolism for the opportunity to reprint these letters in our newsletter.

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Regarding the Consensus Statement on 21-Hydroxylase Deficiency from the Lawson Wilkins Pediatric Endocrine Society and The European Society for Paediatric Endocrinology

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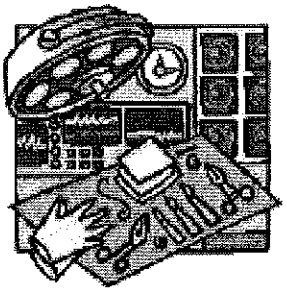

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To the Editor:

This consensus statement is a comprehensive review of a range of issues involving the management of 21-hydroxylase deficiency from before birth until adulthood. Many sections of this article are constructive and helpful, but the section on "surgical management and psychology" may be misleading and potentially detrimental to patient care.

The surgical management of ambiguous genitalia is controversial because few long-term follow-up data are available on the effects of surgery on sexual function and psychological outcome. There is increasing concern from intersex consumer groups about possible detrimental effects of genital surgery. Adult patients and parents of affected children should have a central role in this debate.

The authors list three goals of surgery (page 4050, first paragraph): 1) genital appearance compatible with gender; 2) unobstructed urinary emptying without incontinence or infections; and 3) good adult sexual and reproductive function.

1) The authors use the word gender but presumably mean sex of rearing as decided by the clinicians and parents. There is, to date, no evidence that surgery to render the genital appearance compatible with sex of rearing improves psychological or psychosexual outcome or promotes a stable gender identity 2) Unobstructed urinary emptying without incontinence or infection is an important quality of life issue for the child. Until now clinical instinct that surgery is beneficial has led to early surgery, although there are no data to support this. Surgery itself can result in urinary infections and fistulae (1).

3) The final goal is good adult sexual and reproductive function. There is no evidence that reconstructive surgery gives a better outcome if performed in an infant rather than an adolescent. Clitoral surgery may not promote good sexual and reproductive function, and some studies suggest there may be damage to sexual function (2). Surgery performed in an infant may require revision in adolescence in a significant number of patients (3). An additional major advantage of surgery in an adolescent or adult is that informed consent can be obtained.

Early surgery may be the appropriate course of action and may or may not be supported by long-term outcome data in due course. For the moment, the apparently rigid guidelines in the consensus statement remove flexibility and potentially prejudice the possibility of constructive debate between

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specialties and with patient groups.

The only consensus attainable at the present time is that of a dedicated multidisciplinary team addressing an individual case including full participation of the affected family who will be responsible for the nurture of the child in the modern world.

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LETTER to the EDITOR

Authors' Response: Regarding the Consensus Statement on 21-Hydroxylase Deficiency from the Lawson Wilkins Pediatric Endocrine Society and The European Society for Paediatric Endocrinology

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To the Editor:

We thank Dr. Sarah Creighton and her colleagues for their views concerning the consensus statement on 21-hydroxylase deficiency from the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology, which was published in the September 2002 issue of the *JCEM* (1). The consensus statement was prepared by a group of 40 endocrinologists, psychologists, and surgeons concerned with the management of congenital adrenal hyperplasia (CAH), representing 35 institutions in 12 countries on four continents. This highly qualified and diverse group, which included one of the signatories to Dr. Creighton's letter, represented a broad range of views and experience.

We disagree with the assertion that the guidelines concerning surgical management and psychology are misleading, and we strongly disagree that they might be detrimental to patient care. Dr. Creighton's letter refers to "increasing concern from Intersex consumer groups." These groups primarily represent the experience of women with disorders other than CAH, primarily androgen insensitivity; many of these women were subjected to ill-advised mutilating surgery by inexperienced surgeons. By contrast, the consensus statement deals only with 21-hydroxylase deficiency and emphasizes focusing surgical care in the hands of a small number of highly experienced surgeons. Whereas the care of patients with ambiguous genitalia who do not have CAH may be controversial, there should be little controversy regarding CAH patients who are 46,XX with normal female internal structures and variable masculinization of the external genitalia. Only a small number of these patients are extremely masculinized.

Although there are few studies of long-term outcome showing that improvement of genital appearance improves psychological or pschosexual outcome or promotes a stable gender identity, competent surgery can provide an excellent outcome in the most severely affected children (2) and is compatible with normal reproductive function (3). Therefore, we believe that it is cruel to leave children in a state of gender uncertainty until "they can participate in an informed consent," particularly in this disorder. While a third sex may be acceptable in some cultures, we believe that it is not so in either North America or Western Europe. Body image, while growing up, particularly through stormy adolescence, is very important to confidence and identity of self.

It is also true that surgery can result in urinary infections and fistula and that clitoral surgery may damage sexual function. The decision to reduce clitoral size is not taken lightly, and every precaution is made

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