

AAP: Androgen Insensitivity Does Not Mean Immediate Surgery

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SAN FRANCISCO, Nov. 2 -- Many patients with the intersex syndrome of complete androgen insensitivity can safely delay gonadectomy and vaginal reconstruction at least until late adolescence, suggests a long-term study.

Of 27 patients who underwent gonadectomy, 20 had the procedure in late adolescence or early adulthood, and seven had surgery in childhood, Todd Purves, M.D., of Johns Hopkins, reported at the American Academy of Pediatrics meeting here. None of the surgical specimens demonstrated evidence of malignancy.



Audio: Charles Bankhead speaks with Todd Purves, M.D., Johns Hopkins University, Baltimore

Action Points

- Explain to interested patients that this study found that people with the intersex syndrome of complete androgen insensitivity can safely delay decisions about orchiectomy and vaginal reconstruction.
- This study was published as an abstract and presented orally at a conference. These data and conclusions should be considered to be preliminary as they have not yet been reviewed and published in a peer-reviewed publication.

Additionally, 11 patients have had vaginal reconstruction, 10 procedures performed after puberty. Seven of the 10 postpubertal patients who had vaginoplasty are sexually active, as are 12 of 15 who decided not to have the surgery.

"A woman who has a vaginal depth of two or four centimeters won't be able to have sexual intercourse, but that finding and that decision [about surgery] can be made at age 19," Dr. Purves said in an interview. "The decision can't be made at age two or three or four."

"One of the bottom-line findings of this study is that if a physician sees a two- or three-year-old child with this condition, it would be inaccurate, inappropriate, and wrong to tell the parents 'Your child is going to need vaginal surgery,'" he added. "That is incorrect. Not all of these patients need surgery."

Much of the debate about caring for patients with complete androgen insensitivity syndrome centers on the need for, and the timing of, gonadectomy and vaginal reconstruction or dilation, Dr. Purves noted. For patients who have surgery, the principal issue becomes timing: Should the surgery be done before or after puberty?

The testes are not necessary for development after puberty, but patients with the syndrome face a risk of malignant transformation of 2% to 5% per year after age 25. Additionally, some patients and parents are advised that surgery will be required for normal sexual functioning.

Complete androgen insensitivity syndrome occurs in two to five of every 100,000 live male births, according to the National Institutes of Health. Those with the condition have XY sex-determination chromosomes of males, but because their body does not respond to androgen, they may develop female characteristics, including sexual characteristics.

The syndrome typically is diagnosed in one of two ways. A female child may develop an inguinal hernia and testes are discovered during the examination. Or evaluation of primary amenorrhea during adolescence leads to the finding.

Dr. Purves reviewed the history of 29 patients followed at Johns Hopkins, including 14 described in an earlier study by Hopkins investigators (*J Clin Endocrinol Metab* 2000; 85: 2664-2669). All 29 patients have undergone orchiectomy, and the exact date of surgery is known in 27 cases.

Of the 11 patients who have undergone vaginal reconstruction, the preoperative vaginal depth was 2 to 4 cm. In contrast, vaginal depth averaged 6.6 cm in the 18 patients who have not had vaginoplasty. Of the 25 patients who are older than 18, a total of 19 are sexually active, and the proportion is similar among those with and without vaginal reconstruction.

In summarizing the findings, Dr. Purves said the argument for delayed gonadectomy has at least three lines of support. Delayed gonadectomy:

- Is associated with a low risk of malignancy before puberty (three cases in the literature).
- Delays hormone replacement therapy until late adolescence.
- May enhance breast development and bone mineralization better than does exogenous hormones.

The argument for delaying vaginal reconstruction rests on several observations.

- Most affected patients elect not to have surgery.
- Sexual function does not always require surgery.
- Sexually functional genitalia are not required in childhood.
- Older, more mature patients are better prepared to face complications and psychological issues associated with surgery.
- 80% of patients in the earlier Hopkins study cited late adolescence or early adulthood as the optimal time for surgery.

"A delayed approach to gonadectomy and vaginal reconstruction prevents unnecessary surgery, respects patient autonomy, and allows for a more mature patient to handle the psychological and physiological trauma of surgery and rehabilitation," Dr. Purves concluded.

Neither Dr. Purves nor his colleagues had relevant disclosures.

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peggy s - Nov 05, 2007

As a patient with AIS and a past president of the AIS support group USA, I am glad to see this article which will help prevent unnecessary and premature surgery. Here are some comments: • This article's recommendations were conventional wisdom about AIS 30 years ago. Only in the 1990s was there a trend towards early gonadectomy and sometimes early vaginoplasty, which, ideally, this article will reverse. • The article's main omission is that it does not mention non-surgical dilation as a treatment for vaginal hypoplasia. In nearly all women and girls with complete AIS, and most with high-grade PAIS, surgery to lengthen the vagina is not needed at all. (Some with PAIS may need minor surgery to divide fusion of the labia majora.) The results of non-surgical vaginal dilation are superior by all criteria to the results of surgical vaginoplasty. Those criteria include sexual function after the procedure, as the non-surgically dilated vagina is lined with self-lubricating vaginal mucosa with tactile sensation. The non-surgical procedure also involves less pain and discomfort than surgical vaginoplasty and less expense and inconvenience. In most cases surgery would not even save time because the wait for surgery and the extended convalescence, even without complications, often take more time than the non-surgical procedure would. For more information, see: http://www.aissg.org/31_HPLASIA.HTM#Pressure I would be glad to answer any questions by e-mail. Yours sincerely, Peggy

german cais - Nov 07, 2007

As a patient with CAIS I do generally agree with the conclusions on vaginal dilation. it is absolutely reasonable not to perform surgery before late adolescence. The surgery itself is a very individual decision and every autonome decision, surgery or not, is OK. REGARDING the question on gonadectomy I am a little confused. If there is "only" a risk of 2-5% for degeneration, the question should at least be "Is it then necessary to remove them?" and not so much the timing. EVEN in cases with CAIS testosterone from the testes does have effects on the body, e.g. the libido, sexual functioning, muscles, bones etc. Many CAIS women in the German group "XY-Frauen" have experienced loss of energy and libido, depressive episodes, bone demineralisation etc. as a result of gonadectomy. In most of those patients symptoms disappeared as they started to substitute with testosterone in a self-experiment. IT HAS to be mentioned, that in Germany there is no legal concession to prescribe estrogene for CAIS patients, because they are permitted for xx-women only. Furthermore it should be considered, that there are no long term results available for contrachromosomal hormone treatment. At last, changing the hormonal status does not change sexual identity or the fact, that CAIS patients are intersexed. It won't change identity, because there are still xy-chromosomes. THE most important question is, why would you want the testes to be removed and hormones to be replaced? The testes are working very well, they can be monitored very easily and patients do not need to have surgery and lifelong dependence on hormone replacement. Just think about it. No one would think about prophylactically castrating a man because of the possible risk of degeneration of his testes. No one would remove a working pancreas and replace insulin. It does not make sense.