

MEDICAL, PSYCHOLOGICAL AND LEGAL ISSUES IN THE CLINICAL MANAGEMENT OF THE COMPLETE ANDROGEN INSENSITIVITY SYNDROME PATIENT INTO ADULTHOOD

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Introduction

The author writes from the vantage point of a patient in considering the needs of patients and their families. This paper addresses the physician who is confronted with the diagnosis of complete androgen insensitivity syndrome (CAIS) (**Ref?**). It is a perspective which is missing from medical literature: of the 398 journal articles [*this was early '96; figure higher now?*] concerning androgen insensitivity syndrome (AIS) written between 1953, when Morris provided the first classic comprehensive report defining the syndrome,¹ and the present, there have been only two brief letters in a British journal² and four in a Canadian journal,³ describing the patient's experience of being managed for the condition. Psychologist Suzanne Kessler, in an analysis⁴ of the way intersexed infants are treated, asserts that there is virtually no reported data in the medical literature concerning how adolescent/adult intersexuals experience their treatment (medical and psychological) or their genital appearance and functioning.

This article is based, in part, on in-depth personal communications between the author and twenty-four other women (19 to 62 yrs.) who have represented to the author that they have CAIS. These women describe a genital phenotype which would seem to fall within Quigley et al's androgen insensitivity Grade 7 (normal exterior female genital phenotype with *absence* of pubic and axillary hair post-puberty) or Grade 6 (normal exterior female genital phenotype *with* androgen-dependent pubic and/or axillary hair).⁵ The author has also communicated with fourteen parents (unrelated to either the adult CAIS women or each other) who have advised the author that they have at least one child with CAIS (2 to 18 yrs.). No independent confirmation of such diagnoses has been made. The author is not a physician, has not reviewed any medical records to confirm any diagnosis, and is not trained in making any evaluation of the information provided to her by such individuals. It is the sole intent of this article to offer the clinician insight into certain medical, psychological and legal considerations which are often overlooked in the management of the patient.

1. Morris J.M., Mahesh VB: Further observations on the syndrome, "testicular feminization". Am J Obstet Gynecol 87:731-748, 1963. [**This is his 2nd AIS paper - need ref. to the first one**]

2. Anonymous: 'Once a Dark Secret' BMJ, 308, 19 Feb 1994, p 542 and Anonymous: 'Be Honest and Open with Sufferers'. Letter to the BMJ, 308, 16 April 1994, p1042.

3. Can. Med. Assoc. J., June 15, 1996; 154 (12): 1829-1833.

4. Kessler S: 'The Medical Construction of Gender: Case Management of Intersexed Infants' published in Signs, the Journal of Women in Culture and Society, Autumn 1990.

5. Quigley et al: Androgen Receptor Defects: Historical, Clinical and Molecular Perspectives. Endocrine Reviews, Vol. 16, No. 3, pp271-321 (1995).

Paradigms of Patient Management

One model for managing AIS would be to counsel parents, tell the truth to the patient, prepare for some emotional trauma and arrange psychological counselling, allow the patient to decide on how and when to lengthen the vagina (if necessary), delay gonadectomy until the patient is post-puberty, commence hormone replacement therapy (HRT) post-surgery, discuss the patient's concerns during annual visits, and communicate with the patient in a dignified, sensitive manner.

Unfortunately, the author's communications with both AIS women and parents of AIS children suggest that the current paradigm of treatment is to tell the *parents* the truth, offer only lip service to psychological counselling (**Ref?**), lie to the patient (**Ref?**), parade interns and residents past her while she is naked (communicating to her that she is an oddity) (**Ref?**), and pretend that the physician has 'cured' her of her intersexuality by removing her testes and surgically correcting her vagina, often in infancy and without her informed consent.

Counselling/Support for Parents

Communications with the fourteen parents of AIS children reveal that in only two cases, can the parents recall being offered any psychological support. At a symposium of the Royal Society of Medicine, London, entitled *The Management of Intersex into Adult Life*,⁶ AIS genetic expert, Professor Ieuan Hughes⁷ stated that a survey by his department in conjunction with the British Paediatric Endocrine Society showed that no trained psychological counsellor was involved in discussions with parents about the management of their AIS children (**Ref?**).

Currently, parents do not seem to be encouraged to confront their own fears and concerns about having a 'pseudohermaphrodite' child. Instead, they are offered 'damage control' in which the doctor tries to put the best face on the condition and tells the parents how much like a normal little girl their XY child is. In a private communication to the author (**Ref?**), the parents of a two-year old with AIS reported that their pediatrician told them that their child was "100% anatomically female." The problem with this is that the AIS child will some day develop into a young woman who will have to confront the real issues of looking different (i.e. no pubic hair and a scar across her lower abdomen), of not having a menstrual cycle when all her friends are discussing their periods, of having XY chromosomes and a pair of testes, and of being sterile. It is only the last of these issues which physicians seem to focus on, perhaps because the other issues seem 'unpleasant' or trivial. But the physician must address *all* issues squarely with parents and refer them to competent, informed psychological counsellors so that the parents can work through their own feelings. Then, when their daughter becomes an adolescent, the parents will be in a position to create an open and comfortable environment in which to help their child come to terms with all aspects of having the condition.

The physician should also not ignore that when a diagnosis is made in infancy there is the unarticulated question "What is my child going to grow up to look and act like?" It is disorienting for parents to know that their child, who appears to all the world as a baby girl

6. *The Management of Intersex into Adult Life: Especially Androgen Insensitivity/Testicular Feminization Syndrome*. A one-day symposium at the Royal Society of Medicine, 26 April 1995.

7. Prof. of Paediatrics, Addenbrookes Hospital, Cambridge, UK.

actually has male chromosomes and testes. But if, immediately after diagnosis, the parents are put in contact with a successful adult CAIS woman, such fears can be quelled. A^LIAS (Looking At AIS), the journal/newsletter of the UK Androgen Insensitivity Support Group (<http://www.medhelp.org/www/ais>), reports that of 8 parents assembled at the group's first meeting "[t]he parents said that the single most valuable benefit of having attended the meeting, was to have met two adults with CAIS who were not freaks, but who were leading successful lives in spite of their problems in certain areas."⁸ Being able to talk to an adult with CAIS, and other parents who have AIS children, may be more reassuring than any words of encouragement the physician might offer. And seeing that an adult is able to thrive despite knowing the truth about the condition will place the parents on a more sure footing as they raise their child.

The Office Examination

During office visits, the physician should be sensitive to the impact on the patient of questions about menstrual cycles, an unfortunate routine question asked by intake nurses. The psychological trauma of amenorrhea is often overlooked in the CAIS patient. In a letter to the *British Medical Journal*⁹ an adult with AIS writes "[t]he issue of childlessness, is, I consider, no greater an issue in my case than in any other; however, the absence of menarche as a confirmation of womanhood is." It is a sentiment the author has heard repeatedly from CAIS women who contact the support group. This is not surprising: the issue of infertility may seem remote to an 11- 15 year old, while the inability to participate in the coming-of-age rite of menstruation (which the patient's friends, mother, sisters and/or hygiene teacher all communicate as being the 'threshold to womanhood') is both immediate and devastating.

The doctor should meet with the patient while she is still clothed and make her feel comfortable before asking her to undress for an examination. Preferably she should not be seen, as many AIS women are in an 'infertility clinic' replete with bulletin board photographs of IVF newborns and a waiting room chock full of pregnant women.

Unfortunately some clinicians have allowed the 'viewing', of their AIS patients by junior medical personnel. Employing the justification of physician education, these clinicians have offered teenage AIS girls as a 'sacrifice' on the altar of the examining room table, to interns and residents. Money et al report incidents where genital examinations which are handled insensitively may be perceived as tantamount to sexual abuse.¹⁰ It is disingenuous to seek the underage patient's consent to being examined by interns and residents who are viewing her solely for their education and not in furtherance of the patient's treatment. Fletcher et al (**Ref?**) have demonstrated that subjects of nontherapeutic research often confuse research and treatment. Even if this is not the case, consent is often 'engineered' because of the stresses and intimidation presented by any interaction between the under-age patient and her physician. (**Ref?**)

8. A^LIAS No. 2, Summer 1995. Journal/newsletter of the AIS Support Group (UK).

9. Anonymous: 'Once a Dark Secret' *BMJ*, 308, 19 Feb 1994, p 542. Anonymous author, not known to the support group.

10. Money J., Lamacz M: Genital examination and exposure experienced as nosocomial sexual abuse in childhood. *J. Nervous and Mental Diseases*, Vol 175, No 12, pp713-720, (1987 ?)

Other physicians have placed patients naked before a camera to photograph them for medical journal articles or textbook chapters. The author's survey of medical literature between 1953 and 1996 reveals approximately -#- [enter no.] young AIS women <18 yrs. whose naked photographs appear in journal articles. These young women will feel no less 'freakish' because this takes place in the sterile confines of an examining room as opposed to the tawdry venue of a circus sideshow.

The evolving standard of informed consent, discussed in greater detail below under 'Gonadectomy and Informed Consent', may also impinge on such examinations and photographs. in Kus v. Sherman Hospital, 644 N.E.2d 1214 (1995) and Moore v. Regents of the University of California, 271 Cal. Rptr. 146 (Cal. 1990) ("Moore"), the courts recognized causes of action for battery and lack of informed consent when a physician did not disclose that the patient would be participating in clinical research studies or that the physician had personal interests (research or otherwise) unrelated to the patient's health. By analogy, a young woman who is examined by interns and residents for educational purposes, or photographed for a clinician's forthcoming journal article, should be informed that this is unrelated to her treatment and may need to be told the *complete* facts about her medical condition to ensure that proper informed consent is obtained.

The author has spoken to more than one dozen CAIS women who have residual trauma stemming from incidents as teenagers of being naked as residents were paraded past them, or being photographed by their doctors. Four women, including the author, have described that the experience of being viewed in this manner made them feel that they had been raped. Photographs can be 're-cycled' so that new generations of CAIS patients do not have to endure this experience. All of the women the author describes have acknowledged the importance of educating young physicians; consequently, the author is among many adult CAIS women who would gladly travel at their own expense to any medical school or teaching hospital in the country, be viewed and prodded and photographed, to save a teenager from this fate.

Truth Disclosure

The author believes that the most crucial recommendation of this article is for the physician to encourage the parents to tell the patient the truth. Regrettably this seems to be contrary to the sentiment expressed in most medical literature. Speroff et al state in their 1994 treatise (**Ref?**) that "Conventional wisdom warns against unthinking and needless disclosure of the gonadal and chromosomal sex to a patient with androgen insensitivity syndrome." The result has been dissimulation to the AIS patient by both physicians and, in turn, parents. It is the author's belief, based on communications with AIS women who were told lies and half-truths during adolescence, that the patient will experience deep feelings of shame resulting from an aura of secrecy which accompanies such prevarication.

Quattrin et al¹¹ and Goodall¹² each report positive outcomes when the truth was told in 'stages' so that an adolescent was told certain facts at the time hormone replacement therapy

11. 'Management of male pseudohermaphroditism; a case report spanning twenty-one years.' by Quattrin T., Aronica S., Mazur T: J. Paediatr. Psychol. Dec 1990, 15 (6) p699-709.

12. Goodall J: Helping a child understand her testicular feminization. The Lancet, 5, Vol 337, 33-35, 4 Jan 1991.

commences and eventually, in her later teen years, learned the remaining facts. But nothing that is told, at any stage, should ever be a lie.

Why do this when it is bound to cause the CAIS patient emotional pain? The author has identified eleven key risks/benefits based on her personal experience, as well as discussions with both CAIS women and parents of CAIS children:

- 1) The truth satisfies in a way that lies and half-truths never can; the patient will only be left with confusion if you tell her that she had 'twisted ovaries' and she later discovers that she inexplicably also has a blind-ending vagina and no cervix, uterus or fallopian tubes.
- 2) By knowing the truth the patient can seek out the AIS Support Group and find encouragement and support from her peers.
- 3) Truthful disclosure avoids the risk that the patient will learn the truth in a library or by glancing at her medical chart, and not have appropriate emotional support when she does.
- 4) If the patient discovers the truth on her own and realizes that her doctors and parents have lied she will feel a breach of trust, may become estranged from her family, might avoid needed medical treatment because she does not trust doctors, and may have difficulty ever again trusting anyone.
- 5) Early truth disclosure enables the patient to work through her trauma and come to terms with the condition in her late teens/early twenties so that by her late twenties/early thirties she is ready to invest her emotional energy in adoption/surrogacy if this is of interest to her. It is overwhelming for the patient to discover she has AIS in her late twenties and have to face the emotional challenge of surrogacy/adoption at the same time.
- 6) If the patient senses that her parents are accepting and comfortable with the truth about her diagnosis, her long-term prospects for emotional healing are greatly increased.
- 7) Siblings and other relatives may need to know about the condition in order to determine carrier status or to care for the AIS child in the event of the parents' untimely demise.
- 8) There is always a risk that in the heat of an argument, or a momentary lapse of judgment, a secret will 'spill out' in an insensitive and damaging way.
- 9) There is a risk of malpractice or other legal action if the physician removes the testes or lengthens the vagina surgically without the patient's informed consent.
- 10) The patient may well construe any talk about removing the 'twisted ovaries' to prevent them from becoming cancerous as a message that she has cancer and is going to die.
- 11) In an age of sophisticated technology for infertility, the lie of 'twisted ovaries' offers false hope since the patient may think that a new procedure will cure her infertility.

The truth will inevitably hurt. The patient may perform poorly in school for a while. She may become depressed and withdrawn; she may need psychological counselling. But all during this time her mind is digesting and making sense of the challenge of having AIS. Depression and trauma are part of life. In an open environment the patient can parse through the constellation of emotions she has a right to feel. To pretend, instead, that nothing is wrong only causes delayed mourning and grieving. Studies of post-traumatic stress disorder document that when an individual is unable to express emotion about the incident, the result is an exponentially greater period of depression and trauma. **(2 Refs)** *[there will be additional studies referenced here]*

With AIS, there is a significant risk that the patient will ultimately discover the truth in her twenties or thirties. (Ref?) The support group journal/newsletter, A^LI^AS, is replete with narratives by adult AIS women who experienced severe trauma at learning of the diagnosis later in life and discovering that family doctors had lied about the condition.

Peer Group Support

With the advent of the AIS Support Group (<http://www.medhelp.org/www/ais>), and the resulting availability of support from other CAIS adolescents and adult women, the risk that a teenager (who is otherwise not clinically depressed) will become emotionally distraught is greatly reduced. The author's personal communications with 24 CAIS women reveal that the single most difficult aspect of having CAIS is not the issue of gonads or karyotype – it is feeling all alone in the world with a taboo and 'freakish' condition. One adult woman with CAIS who has a degree in biology, writes in A^LI^AS "[W]hat I really needed . . . was to speak to someone who also had the condition. Throughout my childhood I grew up knowing that I was different to every other girl and trying to keep it a secret. I thought I was unique and would have found it much easier to cope with if I had had the opportunity to meet with other girls with the condition."¹³ Another adult stated "Apart from counselling, probably the single most therapeutic action would be for a patient to be put in touch with another person with AIS. This is unlikely to happen if the condition remains an 'unspeakable monster' within the family."¹⁴

There is tremendous positive interaction at AIS Support Group meetings between parents of AIS children, adult AIS women, and partners and siblings. In a recent article, Camosy explains the benefits of support groups to both the patient and the physician. Regrettably Camosy does not include any such group for intersex conditions, such as the AIS support Group, in a listing of 131 support groups for various medical conditions (Ref?). Overall, the AIS Support Group provides a wonderful vehicle for psychological healing for both parents and adults. Conversely, if the patient senses the shame and secrecy which inevitably accompanies half-truths and lies, she may well become incurably anxious and depressed and is prevented from obtaining this invaluable support from her CAIS peers. As one adult wrote in A^LI^AS, "I can't help feeling that, above all, it is the encouragement of secrecy, leading to isolation, that is so damaging and not the nature of the diagnostic information itself."¹⁵

Vaginal Construction

Shah et. al. report that in a group of twenty-one patients with AIS, 11 had a vagina of 5 cm or less in length.¹⁶ A survey of 24 patients reported by Mobus et. al. in Germany found that 6 cm was the cut-off point below which the patients and their partners indicated problems in achieving penetrative intercourse.¹⁷

13. A^LI^AS No. 3, Winter 1995.

14. A^LI^AS No. 1, Spring 1995.

15. A^LI^AS No. 1, Spring 1995.

16. Shah R., Woolley M.M., Costin G: Testicular feminization: the androgen insensitivity syndrome. J. Paediatric Surgery, Vol 27, No 6 (June) 1992, pp 757-760.

The construction of a neo-vagina, using pressure dilation or surgery, is of critical importance to the patient who is challenged by shortened vaginal length. The variety of available methods for creating a neovagina suggests that the methodology has not yet been perfected.

At any age, there is no sound reason to perform surgery unless pressure dilation has been tried and is unsuccessful. Moreover the author suggests that it is medically, legally and ethically unjustifiable to perform any surgery before the AIS patient is aged at least 15 or 16 and can make an informed choice about her treatment options. The doctor is doing her no favor to perform surgery in infancy in order to have the vagina 'ready' years before she needs it.

With many of the available procedures early surgery (to create a vagina which does not then get regular use) carries a significant risk of stenosis and prolapse. There is greater scarring, resulting in dyspareunia when the patient does become sexually active (**Ref?**). Bailez et al (**Ref?**), in a study of 28 vaginoplasties performed at Johns Hopkins on children with congenital adrenal hyperplasia <5 yrs. between 1970 and 1990, concludes that in 22 cases later surgeries were required to correct problems such as stenosis. Van der Kamp et al (**Ref?**) report an 80% stenosis rate in 10 patients who underwent vaginoplasty in childhood; more compelling, 9 of the 10 patients regretted that surgery had been performed prior to adolescence.

Both the Abbe-McIndoe and McIndoe-Reed techniques, which employ split-skin grafts to construct a vagina, cause the patient to be disfigured by the surgery because skin is taken from the thigh or buttocks; the patient may come to resent that more than she did the lack of a vagina. All forms of the McIndoe-based procedures create a vagina which tends to be dry regardless of exogenous hormone administration. (**Ref?**)

The use of intestinal transposition to create vagina, as developed by Baldwin in 1904 (**Ref?**), and later modified by Ober and Mainrenken (**Ref?**) using pedicled transplants of the sigmoid colon, carries with it the problems of unpleasant secretions and possible fistula.

All of these problems are avoided in patients who employ pressure dilation.

In a private communication to the AIS Support Group, Professor M.M. Woolley states that "The consensus of surgeons in the U.S. is that, with rare exceptions, patients are better off if they stretch their vaginas with a dilator and then continue the stretching via sexual intercourse. . . [t]hose who have been operated on tend to have dyspareunia secondary to scarring from the surgery." The so-called Frank method, named in honor of Dr. R. Frank who, in 1938, first reported¹⁸ creation of a neo-vagina using regular sustained pressure in the interlabial space, offers numerous advantages over surgery. Edmonds reports an 80% success rate with this method.¹⁹ Pressure dilation has been used successfully even in the most extreme cases where the vagina is only a Mullerian pit. (**Ref?**) All forms of dilation can be commenced in the teenage years when the adolescent is ready for a sexual relationship.

Because pressure dilation requires maturity and 'dedication', inasmuch as the patient will have to maintain pressure with the dilators for at least 15 minutes twice per day, the physician must have confidence that she is emotionally ready to commence dilation and is motivated to do so because of a desire for penetrative intercourse.

17. Mobus V., Sachweh K., Knapstein P.G., Kreienberg R: Women after surgically corrected vaginal aplasia: a follow up of psychosexual rehabilitation. (German with English abstract). *Geburtshilfe Frauenheilkd* (Germany) Feb 1993, 53 (2), pp125-31.

18. Frank R.T: The formation of an artificial vagina without operation. *Am. J. Obstet. Gynecol.*, 1938; 13: 1053-5.

19. Edmonds D. K: *Practical Paediatric and Adolescent Gynaecology*. London, Butterworths, 1989: 40.

There are a variety of different dilators available. Many of these have been created to treat gynecological cancers. It is important to understand that in AIS, unlike many forms of gynecological cancer requiring the surgical removal of the vagina, vaginal *length*, rather than width, will be the main problem. Thus, dilators which replicate penises, or are made of glass or even metal, may be intimidating and uncomfortable for a young woman (Fig. 1).

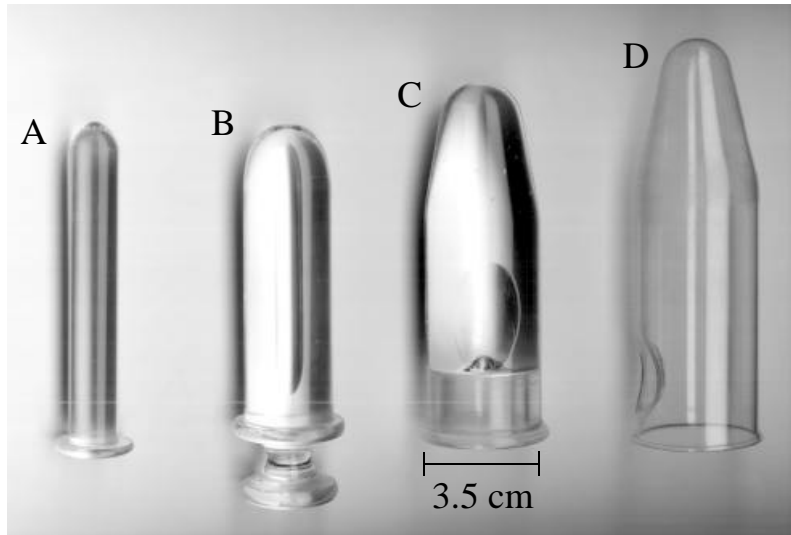


Fig. 1 – University of California dilator (A) and standard UK dilators (B, C and D)

The University of California at San Diego has engineered dilators constructed of lucite. Sets of these dilators, which have now been distributed worldwide by the author, have been met with sighs of relief by parents and AIS women who were emotionally distraught when confronted with more conventional dilators available from medical supply firms.

Patients with vaginal hypoplasia who are using pressure dilatation under the supervision of the Middlesex Hospital, London, are now being provided with a new type of ‘user-friendly’ dilator set (Fig. 2). Made of highly-polished, light-weight plastic, the four hollow cones are stored inside one another like Russian dolls and come with a handle that fits all sizes and a discreet carrying case like a make-up bag.²⁰

20. They are called ‘Amielle Trainers’ and are supplied by Owen Mumford, Brook Hill, Woodstock, OX20 1TU. Tel: 01993 812021, Fax: 01993 813466 (ask for Maria Simpson). US address: Owen Mumford Inc., 849 Pickens Industrial Drive, Suite 14, Marietta, GA 30062. German address: Owen Mumford GmbH, Runde Turmstrasse 8, D-63785, Oberngburg.



Fig 2 – Amielle Trainers

The Vecchietti procedure, an ingenious surgically-assisted and accelerated variant of the pressure dilation technique, has been a well-accepted method in continental Europe, Giuseppe Vecchietti having first described his technique in 1965 and having subsequently reported a 14-year cumulative experience with 307 consecutive cases in 1979 and 1980.²¹ Various authors²² have further developed the method but, for the most part, the technique remains unfamiliar in the English-speaking world, although experience with the method has recently been reported in the US by Veronikis et al. Dr. Veronikis also claims to have developed some high-tech vaginal dilators which will create a neovagina via the Frank method in one month without the need for other intervention, even the Vecchietti procedure.²³

The physician/surgeon must inquire about the patient's needs before settling on any technique. The patient may not care about penetrative intercourse, or may decide with a partner that she does not need a long vagina for sexual expression. She may choose to be celibate, or she may be a lesbian who does not care about the length of her vagina.

21. Vecchietti G: Neovagina nella sindrome di Rokitansky-Kuster-Hauser. *Attual. Ostet. Ginecol.* 1965; 11: 131-47. Vecchietti et al: Le neo-vagina dans le syndrome de Rokitansky-Kuster-Hauser. *Rev. Med. Suisse Romande* (1979); 99: p593-601 (In French). Vecchietti G: Die neovagina beim Rokitansky-Kuster-Hauser Syndrom. *Gynakologe*, 1980; 13: 112-5.

22. Gauwerky et al: An endoscopically assisted technique for construction of a neovagina. *Arch. Gynecol. Obstet.* (1992); 252: p59-63 (In English).

Fedele et al: Laparoscopic creation of a neovagina in Mayer-Rokitansky-Kuster-Hauser syndrome by modification of Vecchietti's operation. *Am. J. Obstet. Gynecol.* (1994); Vol. 171, No. 1. p268-269 (In English).

Keckstein et al: Laparoscopic creation of a neovagina: modified Vecchietti method. *End. Surg.* (1995) 3:93-95.

23. Veronikis D.K et al: The Vecchietti operation for constructing a neovagina: indications, instrumentation and techniques. *Obstet. and Gynecol.*, 90(2): 301-4, Aug 1997.

Despite suggestions by Money et al (**Ref?**) that AIS women are invariably heterosexual, the author's personal communications with ten North American women with CAIS reveals that two (29 yrs. and 62 yrs.) describe themselves as presently exclusively lesbian (one living full-time with a partner) while three others (35 yrs., 36 yrs. and ? yrs.) report having had at least one lesbian experience. Thus, the sexual preference of the patient must not be ignored in suggesting options for treating vaginal aplasia. Otherwise, the patient may feel compelled to pursue either pressure dilation or surgery to accommodate the physician's recommendations rather than her own sexual needs.

Where decreased vaginal length is found the physician should raise the issue and not wait for the patient to reveal that she has discovered on her own that something is wrong. The patient may be too scared to ever inquire even though her mind is swirling in confusion and anguish. If the physician is silent about the issue it has the potential to communicate to the patient that she is such a 'freak' that no one would ever consider her as a sexual partner. This was the author's unfortunate experience and has resulted in significant challenges to her ability to develop mature sexual relationships as an adult.

Treatment for the issue of vaginal aplasia can be tailored to the needs of the patient by communicating the following kinds of reassuring statements: "Many young women with AIS have a problem with vaginal length. We have several options for helping you with this. I want to explain all of these techniques so that you can help us decide what's best for you. I can also put you in touch with other young women who have had these techniques so that you can hear their experiences with dilation, surgery and sexual expression. Learning about these options doesn't mean you have to choose any of them if you decide not to. Some young women feel they don't care about the length of their vagina because they are interested in forms of sexual expression other than intercourse, or might want to wait until they meet someone with whom they want to have a long-term relationship. But I think the important thing is for you to know about all the options so that together we can decide what feels right for you."

Gonadectomy and Informed Consent

It is widely recommended that at some point prior to adulthood the AIS patient undergo the surgical removal of the testes. (**Ref?**) In infancy, the decision will rest with the parents. Hopefully further studies will elucidate the benefits and drawbacks of early versus late gonadectomy, and physicians will inform parents about this. However, Quigley (**Ref?**), in a personal communication, relates that the testes are most often removed in infancy solely for 'psychological reasons' even though they would have produced estrogens, resulting in spontaneous breast development and feminization of body contours. Speroff et al (**Ref?**) also recommend later gonadectomy, stating "[G]onadectomy should be performed at approximately age 16 or 18, to allow endogenous hormonal changes and a smooth transition through puberty."

The author is concerned that the gonads may be removed in infancy not for therapeutic reasons, but simply as a way of avoiding issues of informed consent which arise in an older patient. This is expressed by Kaplan, in a treatise on Clinical Pediatric Endocrinology (**Ref?**), which states "The problem of deciding upon a pubertal, rather than a neonatal, time for gonadectomy involves the need for explaining to the patient the reason for the gonadectomy."

Absent special circumstances, the risk of testicular cancer prior to age 18 is so slight that the surgeon cannot justify surgery on an uninformed adolescent on these grounds alone. Muller et al (Ref?) report no intratubular germ cell neoplasia in the testes of four children with CAIS, while Lukus et al (Ref?) report no gonadal tumors in a recent series of 14 individuals with AIS. The risk of malignant germ cell tumors, reported by Manuel et al (Ref?) is <3% at 18 yr. Speroff et al (Ref?) state that "In contrast to dysgenetic gonads with a Y chromosome, the occurrence of gonadal tumors is relatively late, rarely before age 25, and the overall incidence is less, about 5%." Thus, gonadectomy can safely be delayed until the patient is of an appropriate age to offer her informed consent.

Assuming there is no hernia necessitating surgery before adolescence, the surgical removal of the testes should be accompanied by a complete disclosure of the truth. Otherwise the primary care physician and the surgeon may be at risk of a medical malpractice or battery (i.e. an unallowed touching of the plaintiff's body) action as and when the patient learns the truth in later years. (Ref?) Under the informed consent doctrine, as expressed in the Moore case, infra, any use of gonadal tissue retrieved from the patient to facilitate the physician's research interests would potentially expose both the referring physician, and the surgeon, to a battery action unless accompanied by complete disclosure about the specific nature of both the operation, and the research interests, prior thereto.

Even in cases where the physician has no research interest in the gonadal tissue, the legal doctrine of informed consent requires that the nature of any medical procedure to be performed on the patient be disclosed in advance of treatment. There are only four possible exceptions to the doctrine: emergency, patient waiver, therapeutic privilege, and incompetency. (Ref?) With planned gonadectomy it is only the third of these – therapeutic privilege – which is apposite. *[there will be an inset re therapeutic privilege here]*

The endocrinologist and surgeon should not rely on therapeutic privilege as a safe haven given the number of patients who now report a desire for truth. As one 33 year old woman with AIS reported in the A^LI^AS journal/newsletter, "I experienced a feeling of joy and relief on receiving confirmation (at age 21) [of the diagnosis], because there was a definite reassurance in knowing that my unusual condition was after all, a 'known quantity,' that it had a name, was documented in the literature, that there were other sufferers and that I was not just a one-off freak of nature, after all."²⁴

There is much documented evidence that patients want truthful disclosure and that such disclosure does not result in any permanent harm to the patient's psyche. In a study of cancer patients dating back to the 1960's, Oken (Ref?) reports that of 100 patients with cancer, 89% wanted to know their true diagnoses. Interestingly, Oken also reported that only 3% of doctors always told patients the truth, while 69% either did not tell the truth or usually did not do so. More recently, Professor Theodore LeBlanc writes in a 1995 article (Ref?) that "Courts and legislatures have noted that the special relationship between physician and patient requires increasingly more candid and comprehensive disclosure of information to patients." Such trend is an outgrowth of the idea that the patient's medical information is not proprietary with the physician. As Grant Gillett, a physician and bioethicist, succinctly stated in a recent discussion of virtue and truth in clinical science (Ref?) "The knowledge about a patient (one who suffers) should be able to be owned equally by the sufferer and the agent who hopes to

24. A^LI^AS No. 1, Spring 1995.

intervene and alleviate that suffering." In short, the evolving relevant standard of care will be in favor of truth.

The clinician should also be aware of the risks attendant on waiting for the patient to make preliminary inquiries about her gonads before disclosing the truth. In a leading case on informed consent, Canterbury v. Spence, 464 F.2d 772, 783 n.36 (D.C. Cir.) cert. denied, 409 U.S. 1064 (1972) the Court stated, "We discard the thought that the patient should ask for information before the physician is required to disclose [the truth]. The patient may be ignorant, confused, overawed by the physician or frightened by the hospital, or even ashamed to inquire."

The physician should encourage that the truth be conveyed by the parents, at home and in advance of the meeting with the surgeon. But if the parents refuse to tell their AIS adolescent the truth, the physician should refuse to perform surgery until the patient has reached the age of majority and the parents' unfortunate decision to lie can be ignored.

Personal Anecdote

This article is written in the hope that the clinician will consider the psychological trauma inflicted on AIS patients when there is inadequate counselling for parents, and disregard of the patient's physical and emotional needs. I was motivated to write about the condition because I have spent the better part of thirty years feeling shame and secrecy about having AIS. Told standard lies about the condition at age 11, I was left to discern the truth myself using a medical school library. If you have always felt fairly certain of your female gender identity, it is emotionally painful to discover that you have XY chromosomes and once sported a pair of testes. But it is even more devastating to realize that those who were obligated to care about you (i.e. parents and doctors) have instead lied and left you to unearth the truth on your own. I construed my parents' duplicity to mean that they found the condition and, by extension, me, horribly unacceptable. This no doubt resulted in part because my family was offered no counselling but instead was left to flounder in a sea of confusion and concern. In turn, because I was offered no support by my parents, I dealt with the discovery by disengaging from family and diverting my energies into a career. I ignored all medical care, (including hormone replacement therapy) for the next fifteen years. All the while I lived in isolation, plagued by self-derision; I was afraid to disclose my knowledge to even my closest friends or simply utter the words 'androgen insensitivity'. It is the objective of this article to cause physicians to rethink how they manage AIS patients so that my unfortunate history is not repeated in other cases.

Conclusion

Truth and dignity are the handmaidens of quality medical care. A clinician who is sensitive to the emotional and physical needs of the patient has a wonderful opportunity to help her come to terms with the condition. While there is no cure for AIS, the author believes that with the proper physician, parent and counselling support there can, in fact, be tremendous healing.
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