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## **Title: Management of Intersexuality: Guidelines for dealing with individuals with ambiguous genitalia.**

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Following publication of our paper on a classic case of sex reassignment<sup>1</sup> the media attention was rapid and widespread [e.g.<sup>2-4</sup>] and so too the reaction of many clinicians.

Some wanted to comment or ask questions but many contacted us directly or indirectly [e.g.<sup>5</sup>] asking for specific guidelines on how to manage cases of traumatized or ambiguous genitalia.

Below we offer our suggestions. We first, however, add this caveat: These recommendations are based on our experiences, the input of some trusted colleagues, the comments of intersexed persons of various etiologies and the best interpretation of our reading of the literature. Some of these suggestions are contrary to today's common management procedures. We believe, however, that many of those procedures should be modified. These guidelines are not offered lightly. We anticipate that time and experience will dictate that some aspects be changed and such revisions will improve the next set of guidelines to be offered. Underlying our presentation is the key belief that the patients themselves must be involved in any decision as to something so crucial to their lives. We accept that not every one will welcome this opportunity or these suggestions.

## **GUIDELINES**

Foremost, we advocate use of the terms "typical," "usual," or "most frequent" where it is more common to use the term "normal." When possible avoid expressions like maldeveloped or undeveloped, errors of development, defective genitals, abnormal, or mistakes of nature. Emphasize that all of these conditions are biologically understandable while they are statistically uncommon. It helps in discussion with parents and child that they come to accept the genital condition as normal although atypical. Individuals with these genitalia are not freaks but biological varieties commonly referred to as

intersexes. Indeed, it is our understanding of natural diversity that a wide offering of sex types and associated etiologies should be anticipated (see e.g.<sup>6,7</sup>). Our overall theme is to destigmatize the conditions.

1. In all cases of ambiguous genitalia, to establish most probable cause, do a complete history and physical. The physical must include careful evaluation of the gonads and the internal as well as external genital structures. Genetic and endocrine evaluations are usually needed and interpretation can require the assistance of a pediatric endocrinologist, radiologist and urologist. Pelvic ultrasonography and genitography may be required. Do not hesitate to seek expert help; a team effort is best. The history must include assessment of the immediate and extended family.

Be rapid in this decision making but take as much time as needed. Hospitals should have established House Staff Operating Procedures to be followed in such cases. Many consider this a medical emergency (and in cases of electrolyte imbalance this may be immediately so) nevertheless, we believe that most doubt should be resolved before a final determination is made. We simultaneously advise that all births be accompanied by a full genital inspection. Many cases of intersex go undetected.

2. Immediately, and simultaneously with the above, advise parents of the reasons for the delay. Full and honest disclosure is best and counseling must start directly. Insure that the parents understand this condition is a natural variety of intersex that is uncommon or rare but not unheard of. Convey strongly to the parents that they are not at fault for the development and the child can have a full, productive and happy life. Repeat this counseling at the next opportunity and as often as needed.
3. The child's condition is nothing to be ashamed of but it is also nothing to be broadcast as a hospital curiosity. The child and family confidentiality must be respected.
4. In the most common cases, those of hypospadias and congenital adrenal hyperplasia (C.A.H.) diagnosis should be rapid and clear. In other situations, with a known diagnosis, declare sex based on the most likely outcome for the child involved. Encourage the parents to accept this as best; their desire as to sex of assignment is secondary. The child remains the patient. When assignment is based on the most likely outcome, most children will adapt and accept their gender assignment and it will coincide with their sexual identity.
5. The sex of assignment, when based on the nature of the diagnosis rather than only considering the size or functionality of the phallus, respects the idea that the nervous system involved in adult sexuality has been influenced by genetic and endocrine events that will most likely become manifest with or after puberty. In the majority of cases this sex of assignment will indeed be in concert with the appearance of the genitalia

(e.g., in A.I.S.<sup>8</sup>). In certain childhood situations, however, such assignment will be counter to the genital appearance (e.g., for reductase deficiency<sup>9</sup>). Our concern is primarily how the individual will develop and prefer to live post puberty when he or she becomes most sexually active.

#### Rear as male:

XY individuals with Androgen Insensitivity Syndrome (A.I.S.) (Grades 1-3)

XX individuals with Congenital Adrenal Hyperplasia (C.A.H.) with extensively fused labia and a penile clitoris

XY individuals with Hypospadias

Persons with Klinefelter syndrome

XY individuals with Micropenis

XY individuals with 5-alpha or 17-beta reductase deficiency

#### Rear as female:

XY individuals with Androgen Insensitivity Syndrome (A.I.S.) (Grades 4-7)

XX individuals with Congenital Adrenal Hyperplasia (C.A.H.) with hypertrophied clitoris

XX individuals with Gonadal dysgenesis

XY individuals with Gonadal dysgenesis

Persons with Turner's syndrome

For those individuals with mixed gonadal dysgenesis (MGD) assign male or female dependent upon the size of the phallus and extent of the labia/scrotum fusion. The genital appearance of individuals with MGD can range from that of a typical Turner's syndrome to that of a typical male. Evaluation of high male-like testosterone levels in these cases is also rationale for male assignment.

True hermaphrodites should be assigned male or female dependent upon the size of the phallus and extent of the labia/scrotum fusion. If there is a micropenis, assign as male.

Admittedly, in some cases a clear diagnosis is not possible, the genital appearance will seem equally male as female and prediction as to future development and gender preference is difficult. There is little evidence a poorly functioning clitoris and vagina is any

better than a poorly functioning penis and there is no higher reason to save the reproductive capacity of ovaries over testes. In such difficult cases, whichever decision is made, the likelihood of the individual independently switching gender remains. The medical team in such cases will be taxed to make the best management decision.

6. While sex determination is ongoing, the hospital administration can wait for a final diagnosis before entering a sex of record and Staff can refer to the child as "Infant Jones" or "Baby Brown." After a sex designation has been made, naming and registration can occur. In those cases mentioned above, where prediction of future outcome is in doubt, parents might consider that a name be used that is appropriate for either males or females (e.g., Lee, Terry, Kim, Francis, Lynn, etc.).
7. Perform no major surgery for cosmetic reasons alone; only for conditions related to physical/medical health. This will entail a great deal of explanation needed for the parents who will want their children to "look normal." Explain to them that appearances during childhood, while not typical of other children, may be of less importance than functionality and post pubertal erotic sensitivity of the genitalia. Surgery can potentially impair sexual/erotic function. Therefore such surgery, which includes all clitoral surgery and any sex reassignment, should typically wait until puberty or after when the patient is able to give truly informed consent.

Major prolonged steroid hormone administration (other than for management of C.A.H.) too should require informed consent. Many intersex or sex reassigned individual's have felt they were not consulted about their use and effects and regretted the results.

8. In individuals with A.I.S, do not remove gonads for fear of potential tumor growth; such tumors have not been reported to occur in prepubertal children. Retention of the gonads will forestall the need for hormone replacement therapy and possibly help reduce osteoporosis.

Furthermore, delaying gonadectomy until after puberty will allow the young woman to come to terms with her diagnosis, understand the reason for her surgery and participate in the decision.

Advice regarding gonad removal from true hermaphrodites, individuals with streak gonads and others where malignancies can potentially occur is not so clear. Prophylactically it is common to remove these early; particularly in cases of gonadal dysgenesis<sup>10, 11</sup>.

Watchful waiting with frequent checks is always prudent<sup>12</sup>. Our suggestion, whenever the gonads are removed, is to explain as best as possible why the procedure is needed and attempt to get consent. If the child is too young to understand the reason for the surgery, its necessity should be explained as early as possible.

9. In rearing, parents must be consistent in seeing their child as either a boy or girl; not neuter. In our society intersex is a designation of medical fact but not yet a commonly accepted social designation. With age and experience, however, an increasing number of hermaphroditic and pseudohermaphroditic individuals are adopting this identification. In any case, advise parents to allow their child free expression as to choices in toy selection, game preference, friend association, future aspirations and so forth.
10. Offer advice and tips on how to meet anticipated situations, e.g., how to deal with grandparents, siblings, baby sitters and others that might question the child's genital appearance (e.g., "He/she is different but normal. When the child is older he/she and the doctors will do what seems best.") Parents should minimize the opportunities for such questioning by strangers.
11. Be clear that the child is special and, in some cases might, before or after puberty, accept life as a tomboy or a sissy or even switch gender altogether. The individual may demonstrate androphilic, gynecophilic or ambiphilic orientation. These behaviors are not due to poor parental supervision but will be related to an interaction of the biological, psychological, social and cultural forces to which a child with intersexuality is subject. Some individuals will be quite sexually active and others will be altogether reserved and have little or no interest in sexual relationships.
12. The patient's special situation will require guidance as to how to meet potential challenges from parents, peers and strangers. He or she will need love and friendly support.

Not all parents will be helpful, understanding, or benign and childhood, adolescent, and adult peers can be cruel. Positive peer interaction should be facilitated and encouraged.

13. Maintain contact with family so that counsel is available particularly at crucial times.

Counseling should be multi-staged (at birth, and at least again at age two, at school entry, prior to and during pubertal changes, and yearly during adolescence) and it should be detailed and honest. Counseling should be straight-forward, neither patronizing or paternalistic, to parents and to the child as he or she develops with as much full disclosure as the parents and child can absorb. The counseling should ideally be by those trained in sexual/gender/intersex matters.

14. As the child matures there must be opportunity for private counseling sessions and it is essential the door remains open for additional consultation as needed. On the one hand, the full impact of the situation will not always be immediately apparent to the parents or child. On the other hand, they might magnify the developmental potential of the

genital ambiguity. As above, the counseling should ideally be by those trained in sexual/gender/intersex matters.

15. Counseling must include developmental sequelae to be anticipated. This should be along medical/biological lines and along social/psychological lines. Do not avoid honest and frank talk of sexual and erotic matters. Discuss the probabilities of puberty such as the presence or absence of menses and the potential for fertility or infertility. Contraception advice may be needed and safe-sex advice is always warranted. Certainly the full gamut of heterosexual, homosexual, bisexual and even celibate options --however these are interpreted by the patient-- must be offered and candidly discussed. Adoption possibilities can be broached for those that will be infertile. It is better to discuss these issues early rather than late. Do not obfuscate; knowledge is power enabling the individuals to structure their lives accordingly.
16. The family should be encouraged to openly discuss the situation among themselves, with and without a counselor present, so the child and parents can honestly come to terms with whatever the future holds. Parents have to understand their child's needs and feelings and the child has to understand the concerns of the parents.
17. As early as possible put the family in touch with a support group. There are such groups for individuals with Androgen Insensitivity Syndrome, Congenital Adrenal Hyperplasia, Klinefelter Syndrome, and Turner's Syndrome. Intersexed individuals as a whole (hermaphrodites and pseudohermaphrodites of all etiologies) have a support group, the Intersex Society of North America [addresses for these groups are listed below]. It is emphasized that one on one contact with another person having similar experiences can be the most uplifting factor in an intersexed person's healthy development!

Individual groups or chapters might be more inclined toward parental concerns while others might be tilted toward the intersexed person's concerns. Both perspectives are needed and separate meetings for each faction are useful. Parents need to talk about their feelings in an environment free of intersexed children and adults and the intersexed children and adults similarly need to be able to discuss their feelings and concerns free of their parents. There are times when it is appropriate for physicians to be present and times when it is not.

18. Keep genital inspection to a minimum and request permission for inspection even from a child. Hold in mind that a child may not feel able to deny a physician's request even though that might be his/her wish. The individuals must come to realize that their genitals are their own and they, not the doctors, parents or anyone else, have control over them. Allow others to view the patient only with his or her permission. Often the genital inspections themselves become traumatic events.
19. Let the child grow and develop as normally as possible with a minimum

of interference other than needed for medical care and counseling. Let him/her know that help is available if needed. Listen to the patient; even when as a child. The physician should be seen as a friend.

With increasing maturity the designation of intersex may be acceptable to some and not to others. It should be offered as an optional identity along with male and female.

20. As puberty approaches be open and honest with the endocrine and surgical options and life choices available. Be candid at the sexual/erotic and other trade-offs involved with surgery or gender change and insure that any decision finally be that of the fully informed individual regardless of age. To have him/her discuss the treatment with someone who has undergone the procedure is ideal.
21. Most individuals are convinced by the age of 10-15 as to the direction that would be most suitable for them; male or female. Some decisions, however, should be stalled as long as possible to increase the likelihood that the individual has some experience with which to judge. For instance, a female with a phallic clitoris, sexually inexperienced with partner or masturbation, may not realize the loss in genital sensitivity and responsivity that can accompany cosmetic clitoral reduction. Insure that sufficient information is provided to aid in any decision.
22. Most intersex conditions can remain without any surgery at all. A woman with a phallus can enjoy her hypertrophied clitoris and so can her partner. Women with the androgen insensitivity syndrome or virilizing congenital adrenal hyperplasia who have smaller than usual vaginas can be advised to use pressure dilation to fashion one to facilitate coitus; a woman with partial A.I.S. likewise can enjoy a large clitoris. A male with hypospadias might have to sit to urinate without mishap but can function sexually without surgery. An individual with a micropenis can satisfy a partner and father children.

There is disagreement as to whether gonads that might prove masculinizing or feminizing at puberty should be removed early on to prevent such changes in a child that does not desire such changes. The disagreement involves the concept that the individual faced with such changes might actually come to prefer them to the habitus of rearing but will only become aware of them post hoc. Our bias is to leave them in so any genetic-endocrine predisposition imposed prenatally can come to be activated with puberty. We admit, however, there is no good body of clinical data from which the best prognosis can be made in such cases. There are some indications, however, that even without the gonads the adrenals might prod pubertal changes.

23. If a gender change is being considered, have the individual experience a real-life living test (see e.g.<sup>13, 14</sup>). In this way the individual will have first hand experience in how it actually is to live in the other role. Experience has shown that most indeed make the switch permanent but some return



to their original sex of rearing. Some, usually as adults, will accept an identity as an intersex and plot their own course.

24. Maintain accurate medical, surgical, and psychotherapy records of all aspects of each case. This will facilitate whatever treatment is needed and assist in future research to enhance management of subsequent intersex cases. These records should be available to the patient.

Whenever possible, long term follow-up evaluations, e.g., at 5, 10, 15, and even 20 years of age, should become part of the record.

25. Last, we believe we have to be "authorities" in providing information and advice to the best of our ability yet not be "authoritarian" in our actions. We must allow the postpubertal individual time to consider, reflect, discuss and evaluate and then, have the last word in his or her genital modification and gender role and final sex assignment.

## FINAL COMMENT

We are often asked about those intersexed individuals that have had early surgery of one sort or another, or even sex reassignment, and gone on to be happy and lead successful lives. Doesn't that demonstrate the wisdom of past practices? Our response: Humans can be immensely strong and adaptable. Certainly some intersexed individuals can, in dignity, maintain themselves in a manner that they neither would have chosen nor in which they feel comfortable -- as have others with a life condition from birth that cannot be changed (from cleft palate to meningomyelocele).

Many can adjust to surgery and reassignment for which they were not consulted and many have learned to accept secrecy, misrepresentations, white and black lies and loneliness.

People make life accommodations every day and try to better their lot for tomorrow.

We are aware of individuals that have come to terms with their life regardless of how stressed or painful. To them we offer our praise and admiration for their fortitude, strength and courage. Similarly we do the same for those that have rebelled against their circumstances and changed their lives with elective sex reassignment, surgery or whatever<sup>15</sup>.

However, unlike individuals who have been given neonatal surgery for cleft palate or meningomyelocele, many of those who have had genital surgery or been sex reassigned neonatally have complained bitterly of the treatment. Some have sex reassigned themselves. Others treated similarly have reasons not to make an issue of the matter but are living in silent despair but coping.

The suggestions and guidelines we present are an attempt to consider ways to better life and adjustment for those intersexed and genitally traumatized persons still battling with these issues and for those yet to come.



## REFERENCES

We have purposely kept our references limited to facilitate use of these guidelines and reduce complexity.

- <sup>1</sup> Diamond M, Sigmundson HK: Sex Reassignment at Birth: A Long Term Review and Clinical Implications. *Archives of Pediatrics and Adolescent Medicine* 1997; 151(March): 298-304.
- <sup>2</sup> Leo J: Boy, girl, boy again. *U.S. News & World Report*, 1997; 17.
- <sup>3</sup> Gorman C: A boy without a penis. *Time*, vol. 1997, 1997; 83.
- <sup>4</sup> Angier N: Sexual identity not pliable after all, report says. *New York Times* 1997 14 March 1997; A1, A18.
- <sup>5</sup> Benjamin JT: Letter -to-editor. *Archive of Pediatric and Adolescent Medicine* 1997; 151.
- <sup>6</sup> Fausto-Sterling A: The Five Sexes: Why Male and Female Are Not Enough. *The Sciences* 1993; 1993(March/April): 20-25.
- <sup>7</sup> Diamond M, Binstock T, Kohl JV: From fertilization to adult sexual behavior. *Hormones and Behavior* 1996; 30(December): 333-353.
- <sup>8</sup> Quigley C, De Bellis A, Merschke KB, El-Awady MK, Wilson EM, French FS: Androgen Receptor Defects: Historical, Clinical and Molecular Perspectives. *Endocrine Reviews* 1995; 16(3): 271-321.
- <sup>9</sup> Imperato-McGinley J: 5-alpha-reductase deficiency. In: Bardin CW, ed. *Current Therapy in Endocrinology and Metabolism*, 5th ed. St. Louis, Mo.: C. V. Mosby, 1994; 351-354.
- <sup>10</sup> Donahoe PK, Crawford JD, Hendren WH: Mixed gonadal dysgenesis, pathogenesis, and management. *Journal of Pediatric Surgery* 1979; 14: 287-300.
- <sup>11</sup> McGillivray BC: Genetic aspects of ambiguous genitalia. *Pediatric Clinics of North America* 1992; 39(2): 307-317.
- <sup>12</sup> Wright NB, Smith C, Rickwood AM, Carty HM: Imaging children with ambiguous genitalia and intersex states. *Clinical Radiology* 1995; 50(12): 823-829.
- <sup>13</sup> Clemmensen LH: The "Real-life Test" for Surgical Candidates. In: Blanchard R, Steiner BW, eds. *Clinical Management of Gender Identity Disorders in Children and Adults*, vol. 14. Washington, D.C.: American Psychiatric Press, 1990; 121-135.
- <sup>14</sup> Meyer JK, Hoopes JE: The Gender Dysphoria Syndromes: A Position Statement on So-Called Transsexualism. *Plastic and Reconstructive Surgery* 1974; 54: 444-451.

- <sup>15</sup> Diamond M: Sexual Identity and Sexual Orientation in Children With Traumatized or Ambiguous Genitalia. *Journal of Sex Research* 1997; 34(2): 199-222.

## **INTERSEX SUPPORT GROUPS**

For addresses or contact with groups outside the United States contact one of the groups below.

AIS (Androgen Insensitivity Syndrome) Support Group of the U.S.  
A support group for those with AIS, also their family and partners.  
4203 Genessee Ave. #103-436  
San Diego, CA 92117-4950  
Phone: (619) 569-5254  
email: [aissg@aol.com](mailto:aissg@aol.com)

Ambiguous Genitalia Support Network  
A support group for parents and others.  
428 East Elm St. #4D  
Lodi, CA 95240

## **CAH SUPPORT GROUPS**

For individuals or families with congenital adrenal hyperplasia

National Adrenal Diseases Foundation  
505 Northern Boulevard  
Great Neck, NY 11021  
Phone: (516) 487-4992  
web site: <http://medhlp.netusa.net/www/nadf.htm>

Congenital Adrenal Hyperplasia Support Association  
1302 County Road 4  
Wrenshall, MN 55797  
Phone: (218) 384-3863

H.E.L.P. (Hermaphrodite Education and Listening Post)  
A support group for parents and others affected by any sex differentiation disorder.  
PO Box 26292  
Jacksonville, FL 32226  
web site: <http://www.isna.org/faq.html#anchor643405>  
email: [help@southeast.net](mailto:help@southeast.net)

Intersex Society of North America  
A peer support and advocacy group of and for intersexuals.  
PO Box 31791  
San Francisco CA 94131

email: [info@isna.org](mailto:info@isna.org)  
web site: <http://www.isna.org>

K. S. & Associates (Klinefelter syndromes of all variety)  
A support and education group for families and professionals dealing with Klinefelter syndrome.

P.O. Box 119  
Roseville, CA 95661-0119  
web site: <http://www.genetic.org/>  
email: [ks47xxy@ix.netcom.com](mailto:ks47xxy@ix.netcom.com)

Turner's Syndrome Society of the U.S.  
A support group for those with Turner's syndrome, their family and friends.  
1313 Southeast 5th Street (Suite 327)  
Minneapolis MN 55414  
Phone: 1-(800) 365-9944  
Fax: (612) 379-3619  
web site: <http://www.turner-syndrome-us.org>

## **GENERAL SUPPORT GROUPS**

National Organization for Rare Disorders (NORD)  
Support and educational group for those concerned with any rare disorder:  
P.O. Box 8923  
New Fairfield, CT 06812-8923  
Phone: (800) 999-NORD  
Fax: (203) 746-6518  
<http://www.pcnet.com/~orphan/>

Our-kids  
Support group for parents of children with any sort of special need:  
web site: <http://wonder.mit.edu/our-kids.html>

PFLAG (Parents and Friends of Lesbians and Gays)  
A support group for parents and friends of lesbians and gays.  
1012-14th Street NW, Suite 700  
Washington, DC 20005  
Phone: (202) 638-4200  
email: [PFLAGNTL@aol.com](mailto:PFLAGNTL@aol.com)

## **SEXUALITY/GENDER/INTERSEX COUNSELORS**

Appropriate counselors might be contacted at one of these national organizations.

American Academy of Clinical Sexologists (AACS)  
P.O. Box 1166  
Winter Park, Florida 32790-1166  
Phone: (800) 533-3521  
Fax: (407) 628-5293

American Association of Sex Educators, Counselors and Therapists (AASECT)  
P.O. Box 238, Mount Vernon, Iowa 52314  
Phone (319) 895-8407  
Fax (319) 895-6203

Society for the Scientific Study of Sexuality (SSSS)  
P.O. Box 208, Mount Vernon, Iowa 52314  
Phone (319) 895-8407  
Fax (319) 895-6203

Society for Sex Therapy and Research (SSTAR)  
Secretary: Blanche Freund, Ph.D., R.N.  
419 Poinciana Island Drive  
N. Miami Beach FL 33160-4531  
Phone: 305 243-3113  
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