



DSD Symposium

October 13-14 2006

Renaissance Parc 55 Hotel, San Francisco

Presented by [Intersex Society of North America](#) and [Gay and Lesbian Medical Association](#)

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Introduction

The Intersex Society of North America hosted the First DSD Symposium, a gathering of intersex adults, parents, and allied healthcare professionals, October 13-14, 2006, at the Renaissance Parc 55 Hotel in San Francisco. This was a chance to meet and learn from others working to improve the quality of healthcare for families with children born with Disorders of Sex Development, and for adults dealing with the many ongoing healthcare concerns that result from DSDs.

The DSD Symposium was a mini-conference, held within the Gay and Lesbian Medical Association's annual conference. Registrants to the GLMA conference (about 400 people) were welcome to attend all DSD Symposium presentations. There were about 40 people registered for the DSD Symposium alone. Spot counts of the room showed a maximum of 97 people attending at one time.

Background

A new standard of care has been established for medical management of disorders of sex development ("Consensus Statement on the Management of Intersex Disorders"¹). While certainly not perfect, the new standard incorporates many of the practices and ideas that ISNA has been recommending for over a decade.

In addition to many valuable changes in the standard of care, the Intersex Consensus Group agreed to update the medical nomenclature, bringing intersex conditions into line with other genetic and endocrine disorders, and with a hope and expectation that these conditions will be brought into the mainstream of evidence-based medicine.

The National Institutes for Health has published its "Strategic Plan for Urology."² The Plan recognizes that clinical management of congenital anomalies of sexual differentiation is "in crisis," and calls for efforts to build the skills of qualified mental health professionals; a patient registry; and a broad range of research covering issues of genital surgery; patient and family adaptation with specialist mental health care; decision-making around sex assignment and genital surgery; relationship of gender identity, sexual and reproductive function, and quality of life to early medical, surgical, and psychosocial factors; and molecular markers for rapid diagnosis.

¹ Lee, P. A., C. P. Houk, et al. (2006). "Consensus statement on management of intersex disorders." *Pediatrics* 118(2): e488-500. Available at <http://pediatrics.aappublications.org/cgi/reprint/118/2/e488>.

² National Institute of Diabetes & Digestive & Kidney Diseases (2006). NIDDK Research Progress Report and Strategic Plan for Pediatric Urology. Available from <http://www.niddk.nih.gov/federal/planning/Pediatric-Urology/>.

ISNA has recently published a pair of handbooks, one focused on the clinician and the other on parents, which offer detailed directions for providing patient-centered care for disorders of sex development. These handbooks are facilitating new collaborations between support organizations on the one hand, and clinicians and parents on the other.³

Program

The DSD Symposium offered a wealth of educational programming, including the following highlights. Our featured speaker was Lih-Mei Liao. Dr. Liao is the recipient of a Gay and Lesbian Medical Association Achievement Award for 2006.

Conversation Rounds

facilitated by Bec Kageyama and Lindsey Rourk

Affinity Groups

facilitated by multiple volunteers

Conceptualizing Competent Care for Intersex Adults

facilitated by Anne Tamar-Mattis JD

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William Byne MD, Nina Williams PhD

Clinical Guidelines for the Management of Disorders of Sex Development in Childhood

Charmian Quigley MD

How to Build a Team

Barbara Neilson PhD, Melissa Parisi PhD

³ Consortium on the Management of Disorders of Sex Differentiation (2006). *Handbook for Parents*. Rohnert Park, Intersex Society of North America. Available from www.dsguidelines.org.

Consortium on the Management of Disorders of Sex Differentiation (2006). *Clinical Guidelines for the Management of Disorders of Sex Development in Childhood*. Rohnert Park, Intersex Society of North America. Available from www.dsguidelines.org.

Handbook for Parents

Arlene Baratz MD

Counseling Parents

David Sandberg PhD, Arlene Baratz MD, Ellen Feder PhD

Setting the Research Agenda

Lih-Mei Liao PhD

Speakers

We are thrilled to introduce you to our featured speaker, **Lih-Mei Liao, BSc Hons, MSc, PhD**. Dr. Liao's research has repeatedly broken new ground in posing nuanced questions about the effects of DSDs and medical interventions on psychosocial functioning and adaptation.

As a Consultant Clinical Psychologist, Dr. Liao has played a part in shaping the development of the Middlesex Centre in the UK. The Centre provides clinical services and also carries out research to contribute to the evidence base in DSDs. She is interested in models of sexuality in DSD management and has a broader interest in social constructions in medical procedures for asymptomatic patients in general.

Arlene Baratz MD is a physician and mother. Arlene works with a support group of adults with AIS and parents with AIS children. She participated in creating the *Clinical Guidelines for the Management of Disorders of Sex Development in Childhood* and companion *Parent Handbook* as models of patient-centered care for DSDs. Arlene has long experience in the comprehensive care model through her career in the specialty of Mammography and Breast Imaging Radiology.

Katharine Baratz is a senior at Haverford College in Philadelphia. She is currently studying Classical Culture and Society, with an emphasis on gender dynamics in ancient literature. Katharine plans to enroll in medical school in the Fall of 2007 and hopes to practice Pediatric Endocrinology. Through her mother, Arlene Baratz, MD, Katharine has become involved with the Androgen Insensitivity Support Group and looks to continue to advocate for this and other DSD support groups.

William Byne MD is a board certified psychiatrist whose clinical practice has included adults with disorders of sex development. He is currently an associate professor of psychiatry at the Mount Sinai School of Medicine in New York where, in addition to directing a brain research laboratory, he teaches medical students and residents about sexual minority issues. His research has included investigations of sexual differentiation of the brain in both animals and humans. He has published several critiques and scholarly reviews of data pertaining to biological contributions to sexual orientation and gender identity. He has been active with ISNA since 1995 and is currently a member of ISNA's medical advisory board.

Cheryl Chase is the founder and executive director of Intersex Society of North America. Her work has been recognized in diverse publications and broadcasts, including *Newsweek*, the *New York Times*, NPR's *Fresh Air*, and NBC *Dateline*. Her presentation "Sexual Ambiguity: The Patient-Centered Approach" at the 2000 meeting of the Lawson

Wilkins Pediatric Endocrine Society is considered an unprecedented patient-led breakthrough in medical reform. She produced the videos "Hermaphrodites Speak!" (1997) and "The Child with an Intersex Condition: Total Patient Care" (2003). Chase was one of two intersexed adults (with Barbara Thomas, from Germany) who participated with fifty healthcare professionals in creating the 2006 "Consensus Statement on Intersex Disorders."

Ellen Feder PhD teaches philosophy at American University. She contributed to the Hastings Center project, "Surgically Shaping Children," and is a member of ISNA's Medical Advisory Board and Speakers' Bureau. Her book, Family Bonds: Genealogies of Race and Gender, is forthcoming from Oxford University Press, and she is currently working on a new project in ethics and the management of DSDs, Fixing Sex.

Lih-Mei Liao, BSc Hons, MSc, PhD has worked as a consultant clinical psychologist in the National Health Service in the UK for 20 years. For the past 8 years or so, she has played a key role in developing the Middlesex Centre in London UK, a main research and treatment centre for DSDs. She works with newly diagnosed women as well as those transferred from pediatric services. DSD management is political and dialogue is crucial for progress. She liaises with peer support groups in the UK and also co-founded Critical Sexology Seminar to facilitate inter-disciplinary debate. An important aspect of my work is to contribute to the evidence base in DSD management. In collaboration with like-minded colleagues and students, she has supervised psychological and multi-disciplinary clinical research. Two key themes that capture her interests in research and practice are: 1) Social constructions in medical complaints and decisions to undergo complex treatments; 2) Models of sexuality in clinical/social practices.

Barbara Neilson MSW, Res Dip SW, RSW has been a social worker with Toronto's Hospital for Sick Children for 16 years. Her clinical work has been with the pediatric urology population, and includes patients with rare congenital conditions as well as patients with ambiguous genitalia and gender re-assignment.

Melissa Parisi PhD received her PhD degree in Developmental Biology and MD degree from Stanford University. She is a pediatric geneticist at the University of Washington and has been a member of the multidisciplinary Gender Assessment Team at Children's Hospital in Seattle since 1997, serving as lead physician for the team for the past 2 years. The team provides evaluation, management, and counseling for families, infants, children, and adolescents with Disorders of Sex Development.

Charmian Quigley MBBS is a Senior Clinical Research Physician in pediatric endocrinology at Lilly Research Laboratories in Indianapolis, Indiana, and also holds a teaching position in pediatric endocrinology at Indiana University. She is a graduate of the University of New South Wales in Sydney, Australia. After completing her pediatric residency and pediatric endocrinology clinical fellowship in Sydney, she moved to the US and undertook a fellowship in molecular endocrinology at the Laboratories for Reproductive Biology at the University of North Carolina at Chapel Hill, North Carolina, where she focused on the molecular biology of the androgen receptor. It was this work that stimulated Dr. Quigley's interest in the area of sex differentiation and development. Following 3 years as a full-time member of the clinical faculty at Indiana University, she moved to her current position at Eli Lilly and Company in 1997, where she directs Lilly's research studies in pediatric patients in the United States.

David Sandberg PhD is Associate Professor and Director of the Division of Child Behavioral Health in the Department of Pediatrics at the University of Michigan in Ann Arbor. He obtained his doctorate from Concordia University in Montreal, Canada and completed postdoctoral fellowships at the University of Miami and at the College of Physicians & Surgeons of Columbia University. His research activities include the study of psychosocial aspects of short stature and the psychosocial management of individuals born with disorders of sex development (DSD). He recently served as co-investigator of an (NICHD-sponsored) interdisciplinary research network concerned with biological and socialization factors in sexual differentiation, is developing a psychoeducational treatment manual for clinicians caring for newborns with congenital adrenal hyperplasia identified by newborn screen, and is designing health-related quality of life measures for individuals with DSD and their families.

Anne Tamar-Mattis JD is a recent graduate of Boalt Hall School of Law, is the founder and Executive Director of the Institute for Intersex Children and the Law. Before attending law school, Ms. Tamar-Mattis served for many years as an organizer in the LGBTQI communities. She is the former Director of the LYRIC Youth Talkline, and former Program Director of the San Francisco LGBT Community Center. She and her partner, Suegee Tamar-Mattis MD, are the parents of two children.

Eric Vilain MD PhD was born in Paris, France and is currently an Associate Professor of Human Genetics, Pediatrics and Urology at UCLA. Dr. Vilain received his Ph.D. at the Pasteur Institute in 1994. In 1995, Dr. Vilain received his M.D. at the Faculte de Medecine Necker Enfants Malades. He is currently achieved the Chief of Medical Genetics at UCLA, the Director of Research on Sexual Medicine in the Department of Urology, the Graduate Advisor in the Department of Human Genetics and an attending physician in the Department of Pediatrics at UCLA.

In addition to his clinical responsibilities, Dr. Vilain is the Principal Investigator of a laboratory working at discovering the genetic bases of sex determination and sexual orientation. He has received numerous awards, notably from the NIH and the March of Dimes. Dr. Vilain is an expert in the field of the genetics of sexual development. He has deciphered a large number of molecular mechanisms responsible for intersexuality in humans, such as mutations in the sex-determining genes SRY and SOX9. His laboratory continues to work on the mechanisms of early gonadal development and brain sexual differentiation.

Nina Williams PhD is a clinical psychologist in private practice in Somerset, NJ. She specializes in the psychodynamic treatment of sexual disorders and trauma. She is a member of the Medical Advisory Board of the Intersex Society of North America, and has written and presented many workshops on the treatment of individuals with disorders of sexual development. Dr. Williams is an adjunct associate professor in the Department of Psychiatry at Robert Wood Johnson Medical School, a supervisor and lecturer at the Graduate School of Applied and Professional Psychology, Rutgers University, and on the faculty of the Institute of Psychoanalysis and Psychotherapy of New Jersey.

Sponsors

We are grateful to all of ISNA's individual donors for helping to make the DSD Symposium possible. Would you like to become a donor? Please visit <http://www.isna.org>.

We are particularly grateful to Arcus Foundation, for a \$150,000 grant for general operating expenses in the years 2006-2009.

Schedule

Friday October 13, 2006

Friday 10-12—Conversation Rounds and Affinity Groups

Friday 1:30—Conceptualizing Competent Care for Intersex Adults

Friday 3:00—Agenda for Change: Psychology and Clinical Management of Disorders of Sex Development in Adulthood

Saturday October 14, 2006

Saturday 8:45—Welcome to the DSD Symposium

Saturday 9:00—Report on Chicago Consensus Conference

Saturday 9:45—Nomenclature change

Saturday 10:30—Adult Counseling

Saturday 11:30—Lunch

Saturday 1:15—Clinical Handbook

Saturday 2:00—How to Make a Team

Saturday 3:00—Parents Handbook

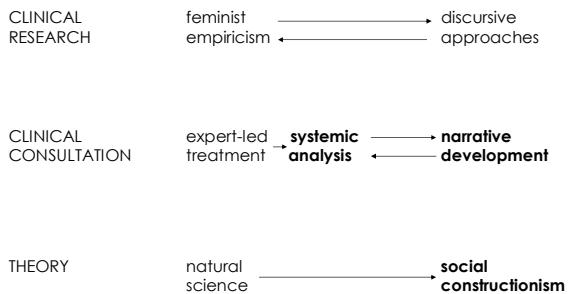
Saturday 3:45—Counseling Parents

Saturday 5:00—Setting the Research Agenda

Saturday 5:45—Closing

Agenda for change:
Psychology and clinical management of
disorders of sex development

Lih-Mei Liao



Middlesex Centre UK

Maligaye Bikoo
Clinical Nurse Specialist

Gerard Conway
Consultant Endocrinologist

Sarah Creighton
Consultant Gynaecologist

Lih-Mei Liao
Consultant Clinical Psychologist

Affiliated specialists:
Alfred Cutner **Melanie Davies** **Christopher Woodhouse**

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AHN

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Melissa Davies

Ida Ismail-Pratt

Cathy Minto

Aims

Through a focus on sexual and related issues in DSDs, to:

- 1) To increase awareness of problems associated with a corrective model of DSD management;
- 2) To demonstrate both the importance of psychological perspectives and the current limitations of psychological contributions to the health and well being of people with DSDs
- 3) To share ideas about the future

Disorders of sex development

"Congenital conditions in which development of chromosomal, gonadal or anatomical sex is atypical"

(Hughes et al., 2006)

Middlesex Centre: Main presentation scenarios

Ambiguous genitalia
(patients usually transferred from paediatrics)

Congenital adrenal hyperplasia [CAH]
Partial androgen insensitivity syndrome [PAIS]
Cloacal syndromes

Unambiguous external genitalia
(patients usually diagnosed in adolescence and adulthood)

Mayer Rokitansky Kusser Hausen syndrome [MRKH]
Complete androgen insensitivity syndrome [CAIS]
Swyer syndrome

Clinical conundrums...

Has physical capacity and wishes for intercourse but has never engaged in it

Reports to be physically and psychologically capable of intercourse but experiences no sexual pleasure

Willingly engages in intercourse but experiences pain and/or other symptoms that remains medically unexplained

Able to fully insert the largest vaginal dilator during examination but complains that partner's penis can't "go all the way in"

.....

Sexual problems and non-problems of currently heterosexually active samples of intersexed women [Percentage reporting normative sexual functioning (N) and sexual difficulty (D)]
(Combined data from Minto, Liao, Woodhouse et al., 2003 & Minto, Liao, Conway et al., 2003)

	CAH (SURGERY) (N=18)	CAH (NO SURGERY) (N=10)	CAIS (N=59)			
sexual problem dimensions	N	D	N	D	N	D
Infrequency	27.8	72.2	30.0	70.0	33.9	66.1
Non-communication	27.8	72.2	20.0	80.0	49.2	50.8
Avoidance	27.8	72.2	20.0	80.0	50.8	49.2
Vaginal penetration difficulties	33.3	66.7	33.3	66.7	41.4	58.6
Anorgasmia	38.9	61.1	60.0	40.0	66.1	33.9
Non-sensuality	22.2	77.8	80.0	20.0	57.6	42.4
Dissatisfaction	61.1	38.9	80.0	20.0	86.4	13.6

Sexual difficulties: Possible contributory factors

Physical

Psychological

Psycho-physiological

Sexual difficulties: Possible physical factors

- Insufficiently large introitus
- Insufficient vaginal volume
- Numbing of genital areas
- Scarring (reduced stretch, obstruction by scarred tissue)
- Vaginal dryness due to oestrogen deficiency
- Absence of libido due to androgen deficiency?

Ogilvie et al., 2006^a

Reference	Number of subjects	Era of surgery	Surgical Outcome	Sexual Function Outcome
Azziz (1984)	78 sw/iv	1934-1984	42% comfortable coitus, 2 nd procedure most successful if done at an older age	42 attempted coitus
Prabhavardhana (1987)	15	1964-1997	95% required 2 nd procedure 77% (10) acceptable introitus	5 sw and 2 sr regularly sexually active of 10 with adequate introitus
Reiley (1992)	28	Mean follow-up 16 years	22 required repeat surgery	11 sexually active
Kishihara (1994)	57	1961-1995	7 married, 4 had children	No details of sexual activity
Al-Bassam (1999)	13	1975-1985	2 complicated procedures, 5 ectopic cervices, 13 introital stenosis	No details of sexual activity
Krebs (2000)	27	1972 - 1988	36% (9) vaginal stenosis	No details of sexual activity
Creighton (2001)	21	1979-1995	24% acceptable coitus 65% unable to use tampons	80% introitus inadequate for penetration
Ford (2001)	49	1991-98	All but 2 operated on as infants	No details of sexual activity
Graziano (2002)	28	81970-1990	No complications described	18 sexually active
Lee (2002)	56	20 yr follow up	Details of surgery given	No details of sexual activity
Stikkelbroeck (2003)	6 sw	1973-1985	Small citoris in 3, 1 some incontinence Vaginal stenosis in 3	2 regular comfortable coitus (3 had attempted coitus), 4 reported ability to orgasm
Crouch (2004)	6	1985-2002	6 abnormal clitoral sensation none had an introitus adequate for coitus	5 attempted intercourse, 4 difficulty achieving orgasm, 3 difficulty with lubrication
Al-Bassam (2004)	45	1985-2001	78% satisfactory appearance of clitoris 84% satisfactory vaginoplasty	No sexually active

Summary of literature involving physical outcomes of feminizing genitoplasty in women with congenital adrenal hyperplasia
Key: sw – salt wasting, sv – simple virilism

Problems of surgical studies

- Absence of evaluation from patients' perspectives
- No explicit definition(s) of 'successful outcome'
- Where evaluation exists, there is inadequate theoretical formulation of psychological questions and methodology

Genital sensitivity

Pilot data based on first 6 patients (Crouch et al, 2004)-
• Impaired sensitivity to warmth, cold and vibration in areas operated on
• Normative sensitivity in area not operated on
• 5 sexually active
• 5 reported significant problem of coital infrequency
• 4/5 reported significant problem in orgasmia
• 3/5 reported significant global sexual dysfunction
• 2/5 reported significant dissatisfaction with sexual relationship

Key pilot findings repeated in main study (Crouch et al., in preparation)

Sexual difficulties: Possible emotional and behavioural factors

- Unexplored effects of repeat intimate examinations
- Unexplored effects of lack of control in clinical situations
- Embarrassment about genital appearance
- Beliefs that sexual lives can only be of the surgeons' making – absence of problem solving
- Feelings of sexual non-entitlement
- Internalised cultural preoccupation with intercourse
- Idealisation of 'normal'
- Sexual anxiety, behavioural avoidance

Intimate examinations

"I remember seeing the locum [GP] [for an arm injury] , he made me take all my clothes off..." (Woman with Swyer syndrome, late 40s)

"She [daughter] was ever so good [cooperative], she'd just jump up on the bed and lifted her skirt." (Mother of adult daughter with CAH)

"repeated psychological insult caused by frequent genital examinations and operations" (Jaaskelainen et al., 2001: p73)

Key:
... = text omitted; [] = significant pause; text = said with emphasis; {text} = explanation inserted by author(s)

Intimate examinations

"...not in a way that I could say no...they did ask me and I suppose I did say yes...I, got quite alarmed when I [] found myself saying, like [] comparing what happened [serialised examinations] with being with the SS...the mind games...all the questions [] like, when did you start growing pubic hair and, ...not telling you anything...all that mind games..."

(Woman with SARD, mid 40s, speaking of experiences at age 14)

Intimate examinations: Related clinical contexts

Intimate assessment procedures could in themselves be psychologically damaging (Solyom et al., 1980)

Some children may even experience physical examinations or medical photography of the genitalia as abusive (Money & Lamacz, 1987)

Pain of humiliation in medical encounters greater for the younger and less psychologically robust children, and for those who could access less external support and were less well prepared for the medical procedures (Shopper, 1995)

Intimate examinations: General population studies

- Negative experiences of pelvic examinations highly prevalent (Areskog-Wijma, 1987)
- Psychological distress most prevalent amongst young women and nulligravid women (Fiddes et al., 2003)
- Health professionals often not adept at predicting women's feelings and expectations (Fiddes et al., 2003)
- Health professionals often not adept at dealing with women's discomfort (Lang, 1990)

Non-entitlement

"And if you've lived your whole life knowing that [] you know, that you - that the vagina's, like, one centimetre or whatever, um [] you know, you just, you just get this sort of fixation in your mind that you're not [] entitled, really, to have that sort of relationship."

"You want some instant resolution [] so you're kind of prepared to put up with [] the sort of side effects, because you think, you think the overwhelming benefits are going to be [] overwhelming [laughs]."

(Boyle et al., 2005)

Preoccupation with intercourse

"If I could have sexual intercourse with a man inside my vagina."

"As soon as I manage to have sex. Oh my God, I'm 19 and still a virgin."

(May et al., 1996)

How imperative is coitus?

"I have been very conditioned to see um sex as sort of you know, intercourse, whatever. And like politically I don't think that [] and I don't think that but um certainly very much kind of conditioned that way to react that way."

(McPhillips et al., 2001:234)

Anxiety

On anticipation

"The whole scenario [sex] is so filled, filled with anxiety... the whole thing has become associated with [] something being wrong [] not being adequately equipped []"

(Boyle et al., 2005)

"...if I was to have sexual relations with a woman the secret would get out. My cover would be blown. Mixed up with 'what's she gonna think of me, am I gonna be tomorrow's gossip?'... That alone would be enough to put me off."

(Chadwick et al, 2005)

Anxiety

On avoidance

"Initially, you know, it probably took four years, three to four years, post-operatively, before I was really able to put it to the test."

(Boyle et al., 2005)

May et al. (1996) -

- Delay in sexual milestones
- Less sexually experienced
- (Compared to women with Type I Diabetes, women with CAH reported fewer initiated attempts at problem solving)

Anxiety

On sexual experience

"It's like, everything is all a test you know, it's [] like with this second guy, I regarded the whole thing as, like, a test, you know, I was testing if all out sort of thing, to see if part A fitted into part B and stuff like that."

"I've only tried it out on someone four or five times, but even then, when I did try it [] I think I was just too conscious about [] um, is it long enough, and have I dilated enough in the last two days to make it, make it the right size?"

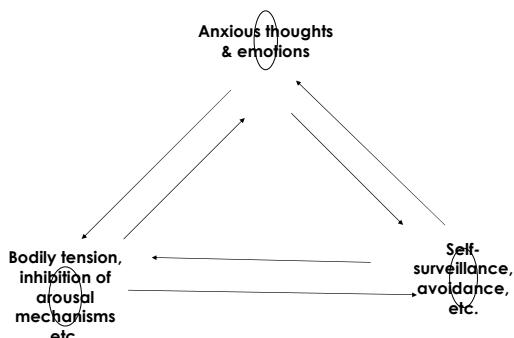
"[Intercourse] was this [] big [] kind of thing that I had to cross [] that I was working to understand... I still thought that the big [] test will be intercourse."

(Boyle et al., 2005)

On disappointment

"...it was like 'Is that it?' And the best thing to come out of the whole thing (vaginal intercourse) was 'Yeah, I can actually do it.' "

(Chadwick et al, 2005)



Surgery is not a cure

"But then I still, even though that [] part of the problem had been taken away, that, you know, penetrative sex could be possible, I still had all the other hang-ups, the shit that I was still carrying around"

"Having to have [] having to go to a doctor every year for a physical, rather than [] the assumption that, OK, when the surgery's over that's all you get for a lifetime, you know? [] And I didn't know that I'd...have to dilate."

(Boyle et al., 2005)

Pre-surgery decision counselling

- What's led up to the decision to have (more surgery)? What outcomes are expected?
- What changes will these have on patient and anyone else?
- What information does patient have, e.g. procedural information, or what risk perceptions?
- What sacrifices might the patient be making, what values are attached to these potential losses?
- Who/what else can assist in decision making?

Difficulties with post-surgical dilation

"It's, it's, I'm sure they explained those things to me, it's just that I was [] so desperate at the time that explanations just went right on by."

"I haven't been dilating, it's just painful...The first year I was very good at it [] but then, after that [] um, I think maybe, I got more sick about doing it...I just worry that I'll progressively go worse and worse and eventually have to have an[other] operation."

(Boyle et al., 2005)

Non-surgical vaginal reconstruction

Rationale

- Virtually all heterosexual women and many lesbian women with DSDs request vaginal reconstruction
- Less invasive
- Fewer risks
- Not irreversible
- Options for most women not operated on are: dilation with surgery or dilation without surgery

Obstacles

- Compliance with post-vaginoplasty dilation is poor and patient satisfaction low (Minto Liao, Conway et al., 2003)
- Significant barriers to self management (Liao et al., 2006)



Dilation as 1st line approach to vaginal agenesis: A multi-disciplinary development

Liao et al. (2006) –

- Psychology-led protocol development
- Drew on motivational and behavioural interventions to enhance 'self management' in other chronic disease contexts
- Informed by critical sexological perspectives
- Designed to centralise the role of the nurse specialist
- Opportunities for psycho-sexual education and counselling
- Multiple outcomes and patient satisfaction assessed

Dilation as 1st line approach to vaginal agenesis: A multi-disciplinary care delivery

Ismail et al. (in submission) –

- 1st prospective evaluation
- 21/26 completed programme (17 sexually active; 4 maintenance therapy) (MRKH & CAIS)
- 3/26 opted out temporarily; 2/26 referred to surgery (Vecchietti)
- Changes in vaginal perceptions correspond with changes in vaginal dimensions
- Significant reduction in sexual anxiety and depression

In contrast...

Bowel vaginoplasties 1980-2004 (Hensle et al., 2006)

- N=36/57 mixed diagnoses (majority MRKH & CAIS)
- Sigmoid colon, ileum & cecum
- 2/36 home dilation + oestrogen suppositories
- 20/36 require pads for mucus production
- 34/36 home douching
- No pre-treatment measure available for comparison but:
 - High rate of reported orgasms, any sexual satisfaction and any sexual desire
 - 33% (any) sexual arousal
 - 33% (any) sexual confidence

Importance of sexual knowledge and confidence

"I felt...like [...] I hadn't learned all the social sort of skills that were needed to [...] you know to-to establish a relationship and that maybe that was the main problem, and having a vagina wouldn't really help...there's more going on than just vaginal length."

The (UK) NHS context

Shift in ethos
Facilitate change in patients from passive recipients of care to Expert Patient...

Health care...
"...not simply about educating or instructing patients about their condition..."

Rather...
"developing the confidence and motivation to use their own skills and knowledge to take effective control over life with a chronic illness."

Expert Patient, DoH 2001

Psychological interventions

- Sense making of DSD with patients and close others
- Increasing control of choices in self disclosure
- Increasing knowledge and skills in social and sexual situations
- Facilitate least invasive treatment options
- Decision counselling for (more) surgery

Critiques of DSM narratives

"Sexual dysfunction or heterosexual dysfunction?"
(Boyle, 1993)

"Full genital performance during heterosexual intercourse is the essence of sexual functioning, which excludes and demotes nongenital possibilities for pleasure and expression. Involvement or non-involvement of the nongenital body parts becomes incidental, of interest only as it impacts on genital responses identified in the nosology."

(Tiefer, 1995:53, emphasis original)

Critiques of clinical psychology

- Pathologization and categorisation of human distress
- Individualization of suffering
- Self-positioning as scientific wizards 'able to identify and expose the processes leading to the patient's disorder and manipulate them such that the abnormalities are repaired'

(Smail, 2001: 57)

**An alternative approach:
Assessing sexuality dimensions**

Developmental trajectories of:

- Gender positionings of self
- Gender(s) of preferred partners
- Body perceptions
- Relationship aspirations
- Sexual experiences and fantasies
- Knowledge and attitude relating to sexual practices

**An alternative approach:
Moving the therapeutic goal post**

- Expanding on understanding of past and present influences on the identified problem(s)
- Increasing control over social and sexual situations
- Increasing awareness of variations in male and female sexualities
- De-centralising vaginal intercourse
- Self permission to explore a range of sexual activities – alone or partnered
- The sexual imperative? Self permission not to pursue sexual activities – as appropriate to clients (and partners)

	CORRECTION	ADAPTATION
V A L U E S	<ul style="list-style-type: none"> Difference as intolerable Acceptance as unimaginable Expert-led clinical management Normal sex is heterosex 	<ul style="list-style-type: none"> Difference can be tolerable Acceptance is possible Collaborative clinical care Challenging heteronormativity
S U R G E R Y	<ul style="list-style-type: none"> No! an option Observations evident Observations obvious Intimate examinations transcede questioning 	<ul style="list-style-type: none"> An option Subject to interrogation Evidence is crucial Observations and intimate examinations strictly protocolised
P S Y C H O L O G Y	<ul style="list-style-type: none"> Sooth distress Absorb problems from suboptimal care Offer service for those who 'fail to' respond to medical interventions 	<ul style="list-style-type: none"> Work with patients (and families): increase expertise, explore meanings, co-create alternative narratives Psycho-sexual work: question gender performance model of sex, emphasise personal choice, counselling over treatment decisions 'Holding' team: acknowledgement of moral dilemmas, increase tolerance of emotional communication Sharing tasks with team: optimal information delivery, research designs and interpretation of data
O U T C O M E	<ul style="list-style-type: none"> Normal gendered identity and sexual practices Absence of psychiatric morbidity 	<ul style="list-style-type: none"> Perceived control of clinical care Optimal physical health and psychological well being Positive self evaluation Satisfaction with gender positioning Optimal sexual life as defined by self

Adaptation v Correction: Challenges to service providers

- Clinicians will need to be more aware of the cultural frames that govern relationship ideals and within which we, our patients and their families make judgments of the 'rights' and 'wrongs' of aspects of their sexuality
- A barrier that we all face is that under strong social pressure, patients are reluctant to relinquish gendered expectations – forcing clinicians to offer more of the same
- Strong barriers exist in building evidence for alternative approaches (e.g. rarity of some conditions)
- Cost implications of multi-disciplinary approaches

Benefits to service users...

Sexuality not dependent on experts' making:

A model of sexual satisfaction based on sensual enjoyment rather than performance is more likely to enable people to (re-)learn to take pleasure in their body parts, to become more flexible towards opportunities for sexual and non-sexual relating;

In letting go of 'normal' – if only to a greater extent, individuals are more likely to transform their relationship to their diagnosis in positive ways...

SUCCESSES

at the
Chicago
Conference

Consensus on Nomenclature Change

Eliminate
“pseudo-
hermaphrodite”

New
nomenclature
rapidly entering
medical usage

Will promote
better use of
evidence

Atypical
gender role
more common,
not indicator for
reassignment

life-long condition;
behavioral support
as needed long term

Disclose facts
about karyotype,
gonadal status,
prospects for
future fertility

Psychological Care Integral in Management

Disclosure
is associated with
enhanced
psychosocial
adaptation

Quality of life:
intimate
relationships

Progress in Patient-centered Care

Limit
genital exams,
medical
photography

More focus
on the stigma,
not solely gender
and genital
appearance

Promote open
communication
with
patients & family:
essential in
decision making

Support
gender change
as needed
in accord with
patient wishes

More Cautious Approach To Surgery

Surgeon has
responsibility to
outline the surgical
consequences from
infancy to
adulthood

Consider
clitoral surgery
only in
severe cases

No
vaginal dilatation
before
puberty

No vaginoplasty
in infants with
short or absent
vaginas

“Google” search
produces more
relevant results

*Consensus Statement On Management Of Intersex Disorders **

LWPES/ESPE Consensus Group
Chicago, October, 2005

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Pediatrics, University of Michigan

Eric Vilain, MD, PhD
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Cheryl Chase
Intersex Society of North America

*Peter A. Lee, Christopher P. Houk, S. Faizal Ahmed, Julian A. Hughes in collaboration with the participants in the International Consensus Conference on Intersex organized by the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology. *Consensus Statement on Management of Intersex Disorders*. Pediatrics, Aug 2006; 118: e488 -e500.

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Background

- Progress in diagnosis, surgical techniques, understanding psychosocial issues, and ascendance of patient advocacy.
- 50 international experts organized into multiple work groups focusing on range of topics
- Work groups reviewed published research
- Participants gathered to review reports and arrived at a consensus

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Terminology & Definitions



- Intersex
- Male/Female Pseudohermaphroditism
- Hermaphroditism
- Sex Reversal
- Disorders of Sex Development i.e., "congenital conditions in which development of chromosomal, gonadal, or anatomical sex is atypical"

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Proposed Revised Nomenclature

Previous	Proposed
Intersex	Disorders of sex development (DSD)
Male pseudohermaphrodite Undervirilisation of an XY male Undemasculation of an XY male	46,XY DSD
Female pseudohermaphrodite Overvirilisation of an XX female Masculinisation of an XX female	46,XX DSD
True hermaphrodite	Ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

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Investigation and Management of DSD General concepts of care

- Evaluation and long-term management must be carried out at a center with an experienced multidisciplinary team;
- All individuals should receive a gender assignment ("third gender" not an option);
- Open communication with patients and families is essential and participation in decision making is encouraged;

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Investigation and Management of DSD The multidisciplinary team

- Optimal care for children with DSD requires...
- Experienced multidisciplinary team generally found in tertiary care centers.
 - Ideally, the team includes pediatric subspecialists in ...
... endocrinology, surgery (+/-), psychology/psychiatry, gynecology, genetics, neonatology, social work, nursing, medical ethics.
 - Ongoing communication with the family primary care physician is essential.

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Investigation and Management of DSD The multidisciplinary team

Team has the responsibility to ...

- educate other health care staff in initial management of affected newborns and families.
- For new patients with DSD, team develops plan for clinical management (diagnosis, gender assignment, and treatment options) *before* making any recommendations.
- Ideally, discussions with the family are conducted by one professional with appropriate communication skills.

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Investigation and Management of DSD The multidisciplinary team

Team has the responsibility to ...

- Transitional care should be organized with the multidisciplinary team operating in environment comprising specialists with both pediatric & adult practice experience.
- Support groups have an important role in the delivery of care to DSD patients and their families.

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Investigation and Management of DSD Gender assignment in newborn infants

- Gender assignment recommendations should reflect the diagnosis, genital appearance, surgical options, need for life-long replacement therapy, potential for fertility, and family views and cultural background.

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Investigation and Management of DSD Surgical management

- Only surgeons with expertise in the care of children and specific training in the surgery of DSD should undertake these procedures
- Surgical management should consider options facilitating the chances of fertility.
- Surgery in 46,XX (CAH) should only be considered in cases of severe virilization (Prader III to V)
- Emphasis is on *functional* outcome rather than a strictly *cosmetic* appearance
- **Evidence is lacking that surgery carried out for cosmetic reasons in first year relieves parent distress and improves parent-child bond**

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Investigation and Management of DSD Surgical management

- The rationale for *early reconstruction* is based on
 - guidelines on the timing of genital surgery from the American Academy of Pediatrics (AAP), the beneficial effects of estrogen on tissue in early infancy
 - the avoidance of potential complications from the connection between the urinary tract and peritoneum through the Fallopian tubes.
 - It is anticipated that surgical reconstruction in infancy will need to be refined at the time of puberty
 - vaginal dilatation should not be undertaken before puberty.

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Investigation and Management of DSD Surgical management

- In DSD associated with *hypospadias*, magnitude and complexity of phalloplasty in adulthood should be taken into account in initial counseling if successful gender assignment is dependent on this procedure.
- Patients must not be given unrealistic expectations about penile reconstruction, including the use of tissue engineering.

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Investigation and Management of DSD

Psychosocial management

- Mental health services integral to patient care
- Atypical gender role behavior more common in DSD, but not indicator for gender reassignment

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Investigation and Management of DSD

Psychosocial management

- The focus should be on interpersonal relationships and not solely on sexual function and activity
- Repeated genital examinations, including medical photography, may be experienced as deeply shaming
- Long-term outcome in DSD should include the following: external and internal genital phenotype, physical health including fertility, sexual function, social and psychosexual adjustment, mental health, quality of life, and social participation.

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Investigation and Management of DSD

Future studies

- Establishing a precise diagnosis in DSD is important with lifelong consequences
 - developments facilitated by progress in molecular research
 - international collaborative approach should be promoted
- Determinants of gender identity in DSD
 - need for studies with representative samples

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Investigation and Management of DSD

Future studies

- Psychosocial management:
 - evaluate the effectiveness of information disclosure with regard to timing and content
- Surgery:
 - evaluate the effects of early versus later surgery in an holistic manner
- Long-term outcome studies:
 - use psychometrically-sound instruments and incorporate guidelines relevant to all chronic conditions (WHO)
 - preferably prospective and designed to avoid selection bias.
 - create patient registries

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Investigation and Management of DSD

Future studies

- Continuing education:
 - for healthcare professionals caring for those with DSD and their families

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Additional Components to Consensus Statement

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Psychosexual Differentiation

- *Gender identity:* "person's self representation as male or female"
- *Gender role:* "psychological characteristics that are sexually dimorphic within the general population, such as toy preferences and physical aggression"
- *Sexual orientation:* direction(s) of erotic interest (heterosexual, bisexual, homosexual) and includes behaviors, fantasies, and attractions.

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Psychosexual Differentiation

Influenced by multiple factors such as...

- Androgens in development
- Sex chromosome genes (?)
- Brain structure and function
- Social/cultural and family background

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Psychosexual Differentiation

- Gender role behavior influenced by androgens (e.g., toy play in girls with CAH)
- However, gender-atypical play behavior is NOT an indication of incorrect sex assignment or later gender dissatisfaction

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Investigation and Management of DSD General concepts of care

- The health care team should discuss with the parents what information to share in the early stages with family members and friends.
- Parents need to be informed about sexual development, and web-based information may be helpful.
<http://www.sickkids.ca/childphysiology/cpwp/genital/intro.htm>

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Investigation and Management of DSD Gender assignment in newborn infants

Initial gender uncertainty is unsettling and stressful for families.

Long-term follow-up studies

- 46,XX DSD assigned **female**:
 - > 90% of CAH identify as females
 - Rx: assign female

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Investigation and Management of DSD Gender assignment in newborn infants

- 46,XY DSD assigned **female**:
 - 100% *complete androgen insensitivity (AIS) syndrome* identify as females
 - ~60% *5α-reductase* identify as males
 - Rx: assign male
 - 25% *partial AIS*, androgen biosynthesis defects, incomplete gonadal dysgenesis express dissatisfaction
 - Rx: ?
 - 100% *micropenis* identify as female
 - Rx: assign male

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Investigation and Management of DSD Gender assignment in newborn infants

- 46,XY DSD assigned **male**:
 - 100% *5α-reductase* (and 17β-HSD3) identify as males
 - Rx: assign male
 - 25% partial AIS, androgen biosynthesis defects, incomplete gonadal dysgenesis express dissatisfaction
 - Rx: ?
 - 100% micropenis identify as males
 - Rx: assign male

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Investigation and Management of DSD Sex steroid replacement

- Hypogonadism is common in patients with dysgenetic gonads, sex steroid biosynthesis defects, and androgen resistance.
- Timing of pubertal initiation may vary and provides opportunities to discuss the condition and set a foundation for long-term treatment adherence.

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Investigation and Management of DSD Psychosocial management

- Quality of life encompasses..
 - ... falling in love, dating, attraction, ability to develop intimate relationships
 - ... sexual functioning
 - ... the opportunity to marry and to raise children (regardless of biological indicators of sex)
- Frequent problems encountered are sexual aversion and lack of arousability

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Intersex Consensus Meeting

Successes of the Chicago Conference (Oct 2006)

Cheryl Chase (with input from Barbara Thomas)

Historic inclusion of patient perspectives

Since medical management of intersex conditions came under criticism over a decade ago, there have been a number of attempts by the medical community to respond to the criticism. Before Chicago, none of these attempts had been very successful.

The Chicago Consensus conference did a better job of including and listening to intersexed adults: this is the first time that many of our criticisms have actually been addressed. Past conferences tended to focus exclusively on gender identity as the only outcome of interest, and early surgery as the only means to assign a sex or to produce a gender identity.

The Chicago conference also benefited from much more diversity of discipline and geography than previous attempts.

1. Nomenclature change

The group reached remarkable consensus on nomenclature change, agreeing to replace the taxonomies based on “hermaphrodite” as well as the term “intersex” with “disorders of sex development.” This nomenclature is already entering medical usage, as can be seen by performing a google search on the phrase.

The new nomenclature helps doctors understand that intersex is about much more than gender identity, and it is clear that it includes many conditions (such as hypospadias, Klinefelter, MRKH) where gender identity is not usually an issue.

2. Elements of patient-centered care

DSDs recognized as **lifelong conditions**. Call for experienced multidisciplinary teams (including behavioral health) to provide long term care.

“**Open communication** with patients and families is essential and participation in decision making is encouraged.” Confidentiality to be respected.

Link to <http://www.dsrguidelines.org> (*Clinical Guidelines and Handbook for Parents* viewable online).

Statement on the usefulness of **support groups** included as appendix. “The value of peer and parent support for many chronic medical conditions is widely accepted, and DSDs, being lifelong conditions which affect developmental tasks at many stages of life, are no exception.”

“While clinical practice may focus on gender and genital appearance as key outcomes, stigma and experiences associated with having a DSD (both within and outside the medical environment) are more salient issues for many affected people.”

“The **initial contact** with the parents of a child with a DSD is important, as first impressions from these encounters often persist. A key point to emphasise is that the DSD child has the potential to become a

well adjusted, functional member of society. While privacy needs to be respected, DSD is **not shameful.**"

"Repeated examination of the genitalia, including **medical photography**, may be experienced as deeply shaming."

"Medical interventions and negative sexual experiences may have fostered symptoms of **post-traumatic stress disorder**."

"**Medical photography** should be undertaken whenever possible if the patient is under anaesthesia for a procedure."

3. Surgery

A more conservative approach to surgery is called for, and it is acknowledged that there is no data supporting the need for or effectiveness of early genital surgeries.

Surgeon has responsibility to outline the surgical consequences from infancy to adulthood, including repeated surgeries.

"Clitoral surgery should only be considered in cases of severe virilisation (Prader III, IV, and V). Parents now appear to be less inclined to choose surgery for less severe clitoromegaly."

"It is generally felt that surgery that is carried out for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parents. The systematic **evidence for this belief is lacking.**"

No vaginal dilatation before puberty. No vaginoplasty in infants born with short or absent vagina. (But early vaginoplasty still recommended for urogenital sinus.)

"Only surgeons with expertise in the care of children and specific training in the surgery of DSD should undertake these procedures."

New data on gonadal malignancies shows that risk depends strongly on diagnosis. Option for parents to defer gonadectomy until adolescence in AIS represents progress (though the article still recommends early gonadectomy for both cAIS and pAIS).

Data shows that surgery to construct vagina carries a risk of cancer (thus more reason not to perform in infancy.)

4. Psychosocial management

"**Psychosocial care** provided by mental health staff with expertise in DSD should be an integral part of management in order to promote positive adaptation. This expertise can facilitate team decisions about gender assignment/reassignment, timing of surgery, and sex hormone replacement."

"The process of **disclosure** concerning facts about karyotype, gonadal status, and prospects for future fertility is a collaborative ongoing action which requires a flexible individual based approach. It should be planned with the parents from the time of diagnosis. It should be planned with the parents from the time of diagnosis. Studies in other chronic medical disorders and of adoptees indicate that disclosure is associated with enhanced psychosocial adaptation."

“Atypical **gender role** behaviour is more common in children with DSD than in the general population but should not be taken as an indicator for gender reassignment. In affected children and adolescents who report significant gender dysphoria, a comprehensive psychological evaluation and an opportunity to explore feelings about gender with a qualified clinician is required over a period of time. If the desire to change gender persists, the patient’s wish should be supported and may require the input of a specialist skilled in the management of gender change.”

“**Quality of life** encompasses falling in love, dating, attraction, ability to develop intimate relationships, sexual functioning, and the opportunity to marry and to raise children, regardless of biological indicators of sex.”

“The most frequent problems encountered in DSD patients are sexual aversion and lack of arousability, which are often misinterpreted as low libido.”

5. Other

“only 50% of 46,XY children with DSD will receive a definitive diagnosis.” That is, all XY children with partial virilization do not have pAIS! Many of them have other disorders, which we won’t be able to diagnose until we understand better the genetics of sex development.

Participants, Working Groups, and Questions

Organizers	
Ieuan A Hughes Department of Paediatrics University of Cambridge Cambridge, UK	Peter A Lee Professor of Pediatrics Penn State College of Medicine The Milton S Hershey Medical Center Hershey, PA, US
Administrators	
Pauline Bertrand ESPE Secretariat BioScientifica Ltd Bristol, UK	Alan and Joanne Rogol Secretary, LWPES University of VA Department of Pediatrics Charlottesville VA, US
Rapporteurs	
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Genetics

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<p>John C Achermann Paediatric & Adolescent Endocrinology Institute of Child Health London UK</p>	<p>Jean D. Wilson Department of Internal Medicine UT Southwestern Medical Center Dallas TX US</p>
<p>Vincent R Harley Prince Henry's Institute of Medical Research Monash Medical Centre Victoria Australia</p>	<p>Erica Eugster Department of Pediatric Endocrinology, James Whitcomb Riley Hospital for Children Indianapolis, IN US</p>
<p>Yves Morel Laboratoire de Biologie Endocrinienne et Moléculaire Hôpital Debrousse Lyon France</p>	

1. What is the current state of knowledge of molecular mechanisms of sexual development in humans?
Are there "activational" and/or "organisational" effects?
2. What lessons can be learned from animal models?
3. Should there be a new nomenclature for the classification of intersexuality based on genetic etiology?
4. What is the current availability of genetic testing for intersexuality, including prenatal testing? Should there be "Centres of Excellence" for testing?
5. Is there any genotype/phenotype correlation that would provide clinically useful information?
6. Are there ethnic or geographic influences on the prevalence of specific intersex conditions?

Brain Programming	
Melissa Hines City University London UK	Sheri Berenbaum Department of Psychology The Pennsylvania State University University Park PA US
Peggy Cohen-Kettenis Vrije Universiteit Medisch Centrum, Genderteam, Amsterdam The Netherlands	Jay Giedd Child Psychiatry Branch National Institute of Mental Health Bethesda MD US
William G. Reiner Departments of Urology and Psychiatry, University of Oklahoma Health Sciences Center Oklahoma City OK US	Emilie F. Rissman Department of Biochemistry and Molecular Genetics University of Virginia Charlottesville VA US
Anna Nordenström Karolinska Institutet, Huddinge University Hospital Stockholm Sweden	

1. Causes of gender identity disorder in patients without intersex conditions
2. What are the causes of gender dysphoria and gender change in individuals with intersex conditions, i.e., what are the genetic, environmental, and hormonal contributions to gender identity, including effects of prenatal, early neonatal, and pubertal hormones?
3. What are the human behavioral effects (excluding gender identity) of prenatal androgens and what characteristics of hormone exposure account for variations across individuals and across behaviors, e.g., timing and dose? How are behavioral effects of androgens separable from physical (especially genital) effects?
4. How are neural and behavioral effects of early hormones in rodents and primates dependent on characteristics of hormone exposure (especially timing and dose), and social context? How are behavioral effects of androgens separable from physical (especially genital) effects?
5. What is the role of the sex chromosomes in neural and behavioral masculinization and feminization in human and nonhuman species?
6. How does brain structure differ in human males and females at different stages of development? What do we know about the causes of the neural sex differences, particularly effects of genes, gonadal hormones and experience?

Medical Management

<p>S Faisal Ahmed Consultant in Paediatric Endocrinology & Bone Metabolism Royal Hospital For Sick Children Glasgow, UK</p>	<p>Patricia A. Donohoue, MD Division of Pediatric Endocrinology and Diabetes University of Iowa Hospitals and Clinics Iowa City, IA US</p>
<p>Silvano Bertelloni Endocrine Unit, Department of Paediatrics University of Pisa Pisa, Italy</p>	<p>Cheryl Chase Intersex Society of North America Rohnert Park, CA US</p>
<p>Kenji Fujieda Dept of Paediatrics Asahikawa Medical College Asahikawa Hokkaido Japan</p>	<p>Claude Migeon Johns Hopkins Medical Hospital, Pediatric Endocrinology Baltimore, MD US</p>
<p>Felix Conte San Rafael, CA US</p>	<p>Mr Chris Driver Department of Surgical Paediatrics Royal Aberdeen Childrens Hospital Aberdeen Scotland</p>

1. Definition of normal and ambiguous genitalia
2. Description of the evaluation of newborns with ambiguous genitalia
3. What are the factors influencing the choice of sex of rearing?
4. Patient Support Groups views of medical care (of patients with intersex)
5. What counselling should be offered to the parents of newborns with ambiguous genitalia?
6. Management of the adolescent intersex patient

Surgical Management

Pierre D.E. Mouriquand Department of Paediatric Ur Debrousse Hospital Lyon Cedex France	Laurence Baskin M.D Pediatric Urology UCSF Children's Hospital San Francisco CA US
Melvin M Grumbach Department of Pediatrics University of California, San Francisco San Francisco CA US	Philip G. Ransley Urology Consulting Rooms London W1N 5PH UK
Richard Rink Pediatric Urology James Whitecomb Riley Hospital for Children Indiana University Medical Center Indianapolis, IN US	John Brock, III, M.D. Pediatric Urology Vanderbilt Children's Hospital Nashville, TN US
Christopher Woodhouse The Institute of Urology and Nephrology, University College London, London UK	

1. Should surgery be performed on the clitoris?
2. Vagina: surgical management of the common urogenital sinus. Timing and techniques
3. vaginal replacement
4. Penis: Reconstruction, Penile size, Durability, Hormonal stimulation, Tissue engineering, Timing for surgery
5. Gonads, Wolffian and Müllerian Structures: Cancer Risk, Removal, and Timing
6. Surgical Factors Influencing the Gender Assignment in the “Y Deficient” Patient

Psychosocial Management

Heino F. L. Meyer-Bahlburg Department of Psychiatry Columbia University New York, NY US	Polly Carmichael Department of Psychological Medicine Great Ormond Street Hospital London UK
David E. Sandberg Pediatric Psychiatry & Psychology Women & Children's Hospital of Buffalo Buffalo, NY US	Norman Spack Division of Endocrinology Children's Hospital Boston MA US
Hertha Richter-Appelt Institute for Sex Research Department of Psychiatry and Psychotherapy University Medical Center Hamburg-Eppendorf Hamburg Germany	Barbara J. Thomas Rottenburg am Neckar Germany
Kenneth J. Zucker Child and Adolescent Gender Identity Clinic Dept. Child, Youth and Family Centre for Addiction and Mental Health Toronto, Ontario	

1. Gender assignment: How should gender assignment be psychosocially managed in newborns
2. Gender reassignment: How should gender reassignment be psychosocially managed in children, adolescents, and adults?
3. Genital surgery and sex-hormone treatment: How should the psychosocial aspects of genital surgery and sex-hormone treatment be managed?
4. Information Management: How should disclosure of sensitive personal information be handled?
5. Sexuality: How should intersex-related problems in romantic and sexual (including reproductive) functioning and orientation be handled in adolescents and adults?
6. Structural issues: How can the need for clinical collaboration of multiple disciplines be accommodated?

Outcomes	
S Drop Sophia Children's Hospital Rotterdam The Netherlands	Garry L. Warne Royal Children's Hospital International Melbourne Australia
Ute Thyen Klinik für Kinder- und Jugendmedizin Universitätsklinikum Schleswig-Holstein, Campus Lübeck Lübeck Germany	Berenice Mendonca Division of Clinical Medicine and Endocrinology University of São Paulo School of Medicine Hospital São Paulo Brazil
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L Looijenga Department of Pathology Erasmus Medical Center/Daniel den Hoed Rotterdam The Netherlands	Amy Wisniewski Drake University Des Moines, IA US

1. What constitutes long-term outcome?
2. Long-term outcome per diagnostic category
3. What is the role of culture and social circumstances on the long- term outcome?
4. What is the incidence and what are the diagnostic criteria regarding gonadal tumors in various diagnostic categories?
5. What is the impact of repeated medical examination, photography and surgery on psychological condition?
6. Additional health problems in intersex.

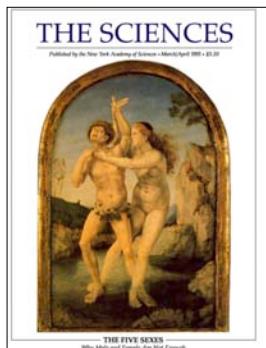
Advocacy Perspective

Chicago Meeting

Traditional Practice Challenged

(a small selection)

1993



1993



1994



1993-present

- ISNA
- AISSG (many countries)
- Bodies Like Ours
- CARES
- Hypospadias & Epispadias Association
- MRKH
- etc etc

AAP picketed 1996



1997

Science Times

The New York Times

New Debate Over Surgery on
Gentails

By Natalie Angier

For those who think that genital cutting and surgery of a girl's clitoris are nothing more than largely irrelevant to the essentials of safe health care, consider the case of Michaela Crowley, a 10-year-old, prepubescent Miami girl.

While playing with her, Ms. Crowley's mother took pleasure in an offhand remark:

"Michaela, you know, she has a clitoris."

Well, we inch long, two or three times the average, and we have a clitoris," Michaela responded, looking well beyond the reporter.

Michaela was worried about how the

clitoris would affect her growing up," said Ms. Crowley's mother, Kristin Crowley.

They just snipped it right off," Ms. Crowley was told only later that she had had "something cut off her," she said.

She was 11, she pressed her father for details, he

Married and overwhelmed, Ms. Crowley

did not seek further guidance for the next three

years until the doctor who had performed the



1997



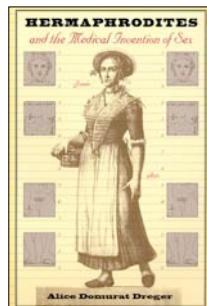
Video: Hermaphrodites Speak!

1997

David Reimer

- Rolling Stone 1997
- book, television 2000

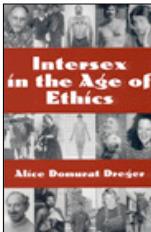
1998



1998



1998



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London Group

first publications 2001

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Medical Response

Denial

- advocates are “zealots” (Gearhart, 1996)
- support groups are not representative
- exclusive focus on surgery and gender identity
- Migeon et al pubs (2000-2004)

2000

- Dallas Urology Meeting 2000

Zderic, S.A., D.A. Canning, et al., Eds. (2002). Pediatric Assignment of Sex: A Critical Reappraisal. New York, Kluwer Academic/Plenum.

2000

- North American Task Force on Intersex (2000-2001)

2001

- British Association of Paediatric Surgeons statement on surgical management

2002

- CAH Consensus Statement

2002

- “Gubbio” Conference, Tempe AZ 2002
- NIH Meeting in Tempe AZ 2002

2004-2005

- Chicago Consensus Conference
- historic inclusion of patient perspectives
- nuanced response to criticisms
- concedes many points to critics

Consensus Conference

- LWPES/ESPE joint sponsorship
- six work groups
- 50 participants from five continents

Consensus Conference diverse constituents

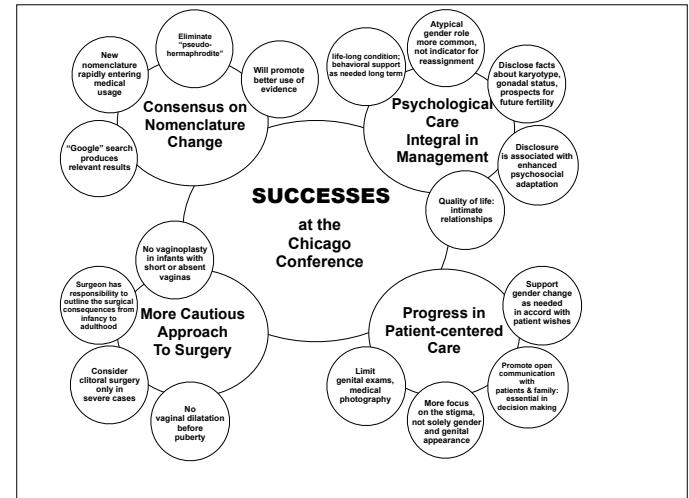
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- psychology
- ped urology
- genetics
- psychiatry
- patient advocacy

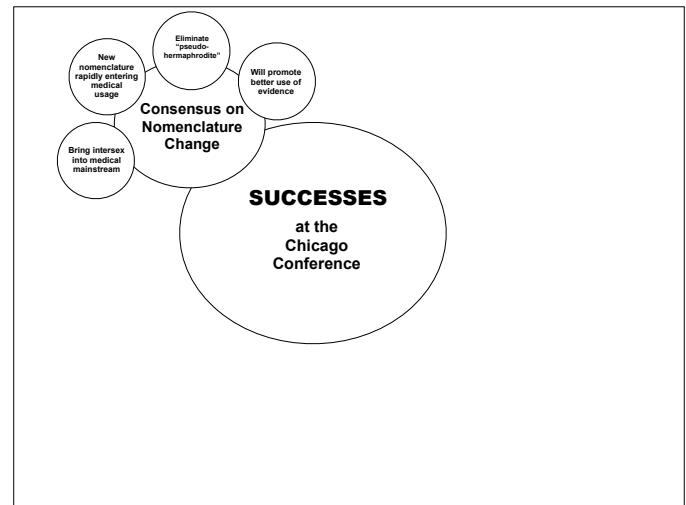
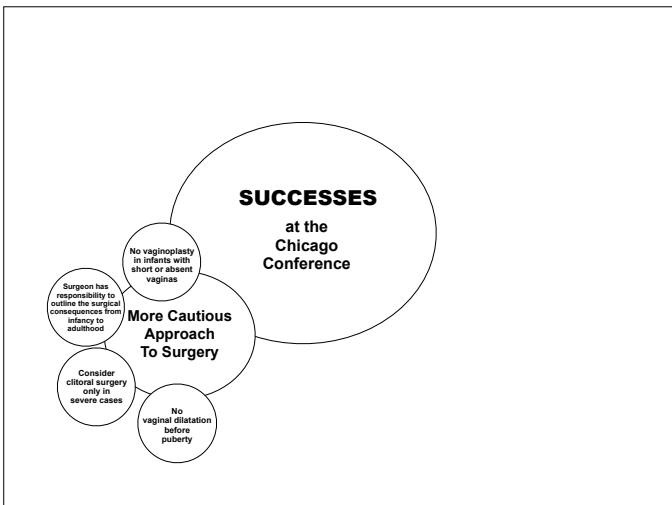
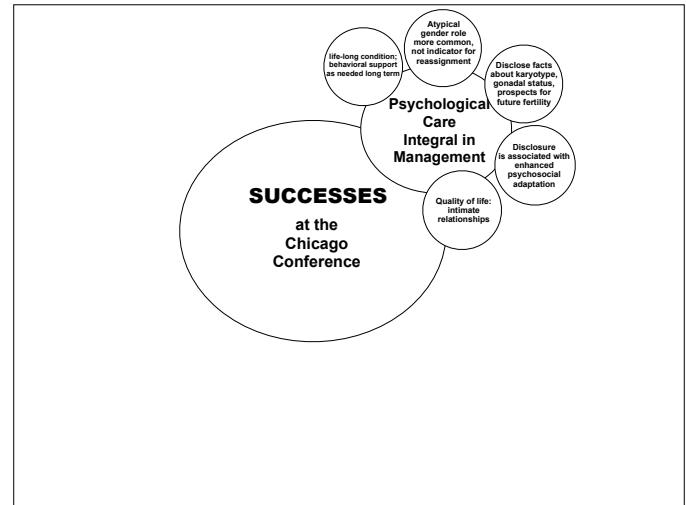
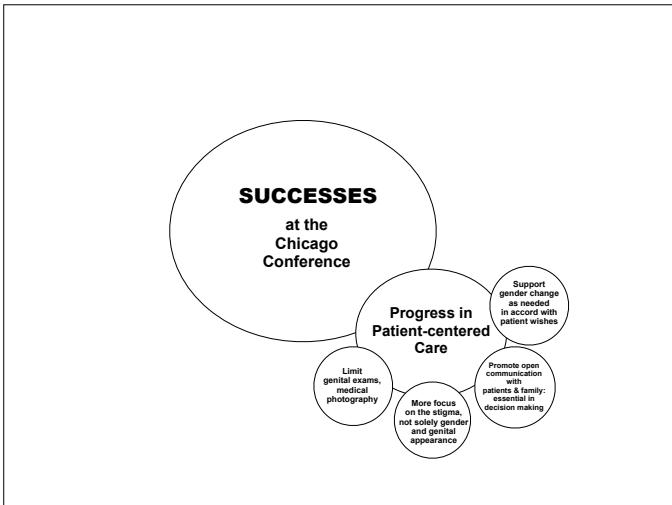
Work Groups

- Genetics
- Brain programming
- Medical management
- Surgical management
- Psychosocial management
- Outcomes

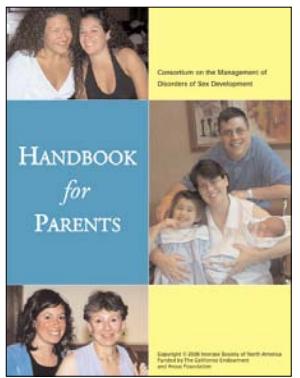
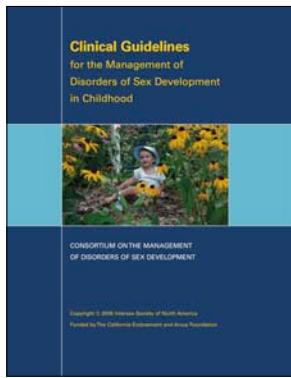
Less than full inclusion

and yet ...





Resources



Nomenclature Overview

Old Language

Intersex Disorders

male pseudohermaphrodite

undervirilization of an XY male

female pseudohermaphrodite

overvirilization of an XX female

true hermaphrodite

XX male

XX sex reversal

XY sex reversal

New Language

Disorders of Sex Development (DSD)

46,XY DSD

46,XY DSD

46,XX DSD

46,XX DSD

ovotesticular DSD

46,XX testicular DSD

46,XX testicular DSD

46,XY complete gonadal dysgenesis DSD

Nomenclature Example 1

Old Language

“A novel frameshift mutation in the 5alpha-reductase type 2 gene in Korean **sisters with male pseudohermaphroditism**.”

Kim SH et al., *Fertil Steril.* 85:750,
2006

New Language

“A novel frameshift mutation in the SRD5A2 gene in Korean **sisters with 46, XY DSD**.”



Nomenclature Example 2

Old Language

“Neonatal outcome of a
prenatally detected 46,XX/46,XY
true hermaphrodite.”

Chen CP et al., *Prenat Diagn.*
26:185, 2006

New Language

“Neonatal outcome of a
prenatally detected case of
46,XX/46,XY ovotesticular DSD”



Nomenclature Example 3

Old Language

“Only one patient with cytochrome b5 deficiency has been reported and studied at a molecular genetic level. That patient was a **male pseudhermaphrodite** who had female genitalia at birth...”



New Language

“Only one patient with cytochrome b5 deficiency has been reported and studied at a molecular genetic level. That was **a patient with 46, XY DSD** who had female genitalia at birth...”

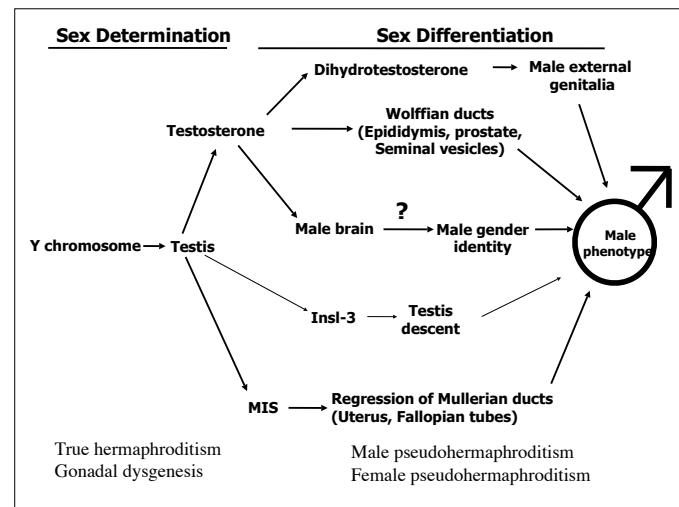
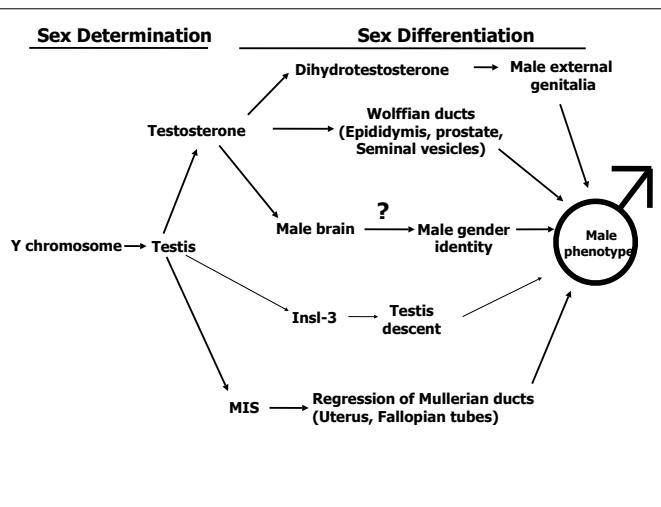
Miller WL and Auchus RJ, *J Clin Endo Metab*, 85:1346, 2000

Nomenclature Change: I am not a disorder

Eric Vilain, MD, PhD
UCLA
Los Angeles, CA

Human Intersexuality

- Wide spectrum of phenotypes
- Overall frequency: 1/5,000 to 1%
- Controversial clinical management
- Questions:
 - Precise diagnosis? Molecular mechanisms?
 - Gender assignment?
 - Clinical management?



Pathologies of Sex Determination

- Abnormal development of the gonads: gonadal dysgenesis
- Klinefelter syndrome
- Turner syndrome
- XX males
- XY females with gonadal dysgenesis
- XX true hermaphrodites

Genes of Sex Determination in Humans

Gene	Function	Localization	Mutation	Duplication	Genetic Test
SRY	transcription	Yp11.3	XY female	normal	clinical
DAX1	transcription	Xp21.3	AHC	XY female	clinical (deletion)
SOX9	transcription	17q24	XY female, CD	XX male	clinical
DMRT1	transcription	9p24.3	XY female	?	research
SF1	transcription	9q33	XY female, AHC	?	research
WT1	transcription	11p13	XY female, DDS	?	clinical
Wnt-4	signalling	1p35	masculinized XX	XY female	research

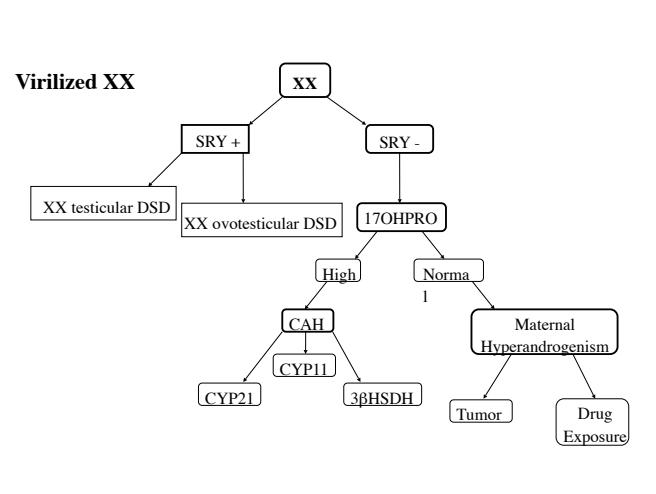
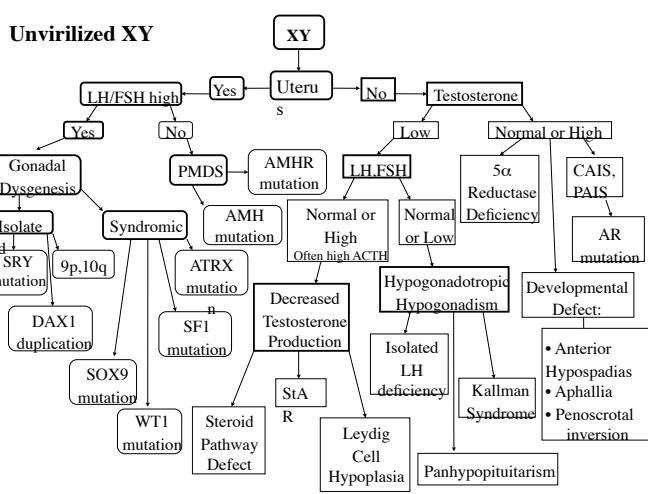
Only about 20% of disorders of sex determination are explained genetically
Most known mutations affect the development of the testis

Pathologies of Sex Differentiation

- Normal gonad development, but abnormal development of the genitalia
- Feminized XY: defect of synthesis of testosterone, androgen resistance
- Masculinized XX: excess of androgens (medications, tumor, congenital adrenal hyperplasia)

Sex Differentiation Genes

- Steroidogenic genes
 - Testosterone biosynthesis
 - 5 alpha reductase
- Androgen receptor genes
- Anti-Müllerian genes
 - MIS
 - MIS receptor



Should advances in genetics of sex determination be reflected in diagnostic classifications?

- Current nomenclature
 - Emphasizes gonadal anatomy above other parameters
 - Is vague, not useful for outcome studies
 - Labels patients in a socially harmful way (e.g., AIS as "male" pseudohermaphrodites)
- New nomenclature
 - Should recognize the complex genetic categorization
 - Should recognize that diagnosis inform but do not determine gender assignment (avoid "male" and "female")

International Consensus Conference on Intersexuality

- October 27–30, 2005 in Chicago
- Organized by LWPES and ESPE
- Multidisciplinary
- 6 groups (genetics, brain programming, medical management, surgical management, psychosocial management, outcome data)

Disorders of Sex Development (DSD)

Congenital conditions in which the development of chromosomal, gonadal or anatomical sex is atypical

Proposed Changes in Nomenclature

Previous	Suggested
intersex	Disorders of Sex Development (DSD)
male pseudohermaphrodite	46,XY DSD
undervirilization of an XY male	46,XY DSD
female pseudohermaphrodite	46,XX DSD
overvirilization of an XX female	46,XX DSD
true hermaphrodite	ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

Sex Chromosome DSD	46,XY DSD	46,XX DSD
A. 45,X (Turner Syndrome and variants)	A. Disorders of gonadal (testicular) development	A. Disorders of gonadal (ovarian) development
B. 47,XXX (Klinefelter Syndrome and variants)	1. complete gonadal dysgenesis (Swyer syndrome)	1. ovotesticular DSD
C. 45,X/46,XY (mixed gonadal dysgenesis)	2. partial gonadal dysgenesis	2. testicular DSD (eg, SRY+, dup SOX9)
D. 46,XX/46,XY (e.g., ovotesticular DSD)	3. gonadal regression	3. gonadal dysgenesis
	4. ovotesticular DSD	
	B. Disorders in hormone synthesis or action	A. androgen excess
	1. androgen biosynthesis defect (eg, 17 hydroxysteroid dehydrogenase deficiency, 5α reductase deficiency, StAR mutations)	1. fetal (eg, 21 hydroxylase deficiency, 11 hydroxylase deficiency)
	2. defect in androgen action (eg, CAIS, PAIS)	2. fetoplacental (aromatase deficiency, POR)
	3. disorders of AMH and AMH receptor (persistent Mullerian duct syndrome)	3. maternal (luteoma, exogenous, etc)
	C. Other (eg, severe hypospadias, cloacal extrophy)	C. Other (eg, cloacal extrophy, vaginal atresia, MURCS, other syndromes)

15

Examples

"A novel frameshift mutation in the 5alpha-reductase type 2 gene in Korean sisters with male pseudohermaphroditism."

Kim SH et al., Fertil Steril. 85:750, 2006

"A novel frameshift mutation in the SRD5A2 gene in Korean sisters with 46, XY DSD"

Examples

Neonatal outcome of a prenatally detected 46,XX/46,XY true hermaphrodite.
Chen CP et al., Prenat Diagn. 26:185, 2006.

Examples

"Only one patient with cytochrome b5 deficiency has been reported and studied at a molecular genetic level. That patient was a male pseudohermaphrodite who had female genitalia at birth..."

Miller WL and Auchus RJ, J Clin Endo Metab, 85:1346, 2000.

"Neonatal outcome of a prenatally detected case of 46,XX/46,XY ovotesticular DSD"

"Only one patient with cytochrome b5 deficiency has been reported and studied at a molecular genetic level. That patient, with 46, XY DSD, who had female genitalia at birth..."

Change in Nomenclature

- Wide consensus from all specialties
 - Supported by the American Academy of Pediatrics
 - Supported by authors of major endocrinology, pediatrics and urology textbooks
 - Goal: modification of ICD

Intersex versus DSD

- “I am not a disorder”
 - “Words can wound”
 - “Why not VSD–Variations of Sex Development?”
 - “People are free to identify as Intersex”

Intersex versus DSD

- Intersex is a vague term. Does it include Turner? CAIS? DSD clearly include conditions that do not necessarily appear with ambiguous genitals
 - “Intersex” has a political meaning and history. DSD does not. Intersex could refer to a social identity, and DSD to a medical diagnosis
 - Intersex labels a person. DSD labels a condition
 - DSD labels a medical condition, not an identity
 - DSD is a diagnosis, therefore knowledge from all medical fields apply. Evidence-based medicine should be used for all medical/surgical decisions on patients with DSD

Acknowledgements

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Pascal Bernard	Maria Alaniz
Saunders Ching	Charleston Chiang
Sven Bocklandt	Susan Berkovitz
Arnaud Lacombe	



Prince Henry's Institute Vince Harley

Nomenclature Change

Arlene Baratz

I am a physician and the mother of 2 girls with complete Androgen Insensitivity Syndrome. As a member of and advisor to the [Androgen Insensitivity Syndrome Support Group's](#) 54-member parent support group, I have counseled these parents for 7 years.

When the “[Consensus Statement on Intersex Disorders](#)” published in the highly-regarded medical journal *Pediatrics* in Summer 2006, AIS parents were overjoyed by its primary message, the proposal of a new paradigm of care in which the emotional and physical health of children and families is of central importance. In the past, AIS parents have completely rejected the word intersex for its many implications outside the strictly medical. As one mother wrote, “When I first saw the term intersex, quite frankly it made me sick to even read it. I think it sounds like ‘it’ or like the gender is in limbo or undecided, and I hate that.”

Parents who unconditionally love their new baby are traumatized when they are “confronted with marginalizing, insensitive and stigmatizing ‘hermaphrodite’ based terms.” It is likely that fear of damaging a vulnerable child who is neither this nor that lies behind many decisions made to hide the condition or to accept “normalizing” surgery. Parenting these special children involves a constant balancing of truth-telling and developmental readiness during the stages of infancy, early childhood, adolescence and young adulthood.

There is a constant desire to cultivate a positive self-image in the child combined with extra vigilance to avoid potentially negative situations. Parents whose children were exposed casually to the term “intersex” found the word disturbing, damaging, and “brutal” in one mother’s words.

They believe, as another mother asserts, that “the new term DSD (disorders of sex development) accomplishes describing that the child’s development has deviated from the norm without placing a socially stigmatizing label on the child which may drive the parents to rightfully seek to protect the child even at the forfeiture of truth and appropriate medical care.” AIS parents are able to assimilate the idea of a *DSD* much more easily than the notion of *intersex*.

At the August meeting of the Androgen Insensitivity Syndrome Support Group USA, the DSD nomenclature and paradigm of care proposed in the “[Consensus Statement on Intersex Disorders](#)” were presented to the AIS parents group. Fifty copies of the [Handbook for Parents](#) using the DSD language were distributed. The DSD terminology is working for the families who are its intended beneficiaries. We parents deeply appreciate the openness and honesty of intersex adults. We believe that children raised in a healthy and loving environment will develop a strong foundation of self-esteem that will eventually enable them to choose their own unique personal identities. Careful and thoughtful consideration must be given to the use of any term that is used in the care of those who are powerless to speak for themselves.

I Am Neither “Pseudo” nor “Disordered”

Peter Trinkl

My name is Peter Trinkl, and I am the Board President of [Bodies Like Ours](#), an intersex educational and peer support organization. We have an active on-line support forum for intersex people and people seeking information about intersex issues. Our Executive Director, Betsy Driver, is not here today, but many of you are aware of her intersex work.

The term “hermaphrodite” goes back to ancient times to describe a person who is sexually seen as being both male and female. With the rise of the medical study of intersex conditions, there arose what the historian Alice Dreger has called the Age of Gonads, roughly dating from the end of the nineteenth century to the early twentieth century. During this time, it was thought that the true nature of a person’s sex could be determined by studying gonadal tissue. Ovarian tissue with associated with being female. Testicular tissue was associated with being male. The presence of both ovarian and testicular tissues in various configurations was a sign of a being a true hermaphrodite. This gave rise to the notion that there were three types of hermaphrodites: “true hermaphrodites”, “female pseudo- hermaphrodites”, and “male pseudo-hermaphrodites”. In the 1920’s, the term “intersex” arose partly in response to problems of sex and gender that were not addressed by the traditional hermaphrodite nomenclature. But the idea that there are three types of hermaphrodites has had a surprisingly long life, and can still be found in many current reference materials.

In a recent article, entitled “Nomenclature/Taxonomy for Intersex: A scientific and Clinical Rationale” published in the *Journal of Pediatric Endocrinology and Metabolism*, ISNA staff members and members of the ISNA Medical Advisory Board recommended that the term “hermaphrodite” and the associated terms of “Pseudo-hermaphrodite” and “True-hermaphrodite” not be used in future medical literature. I also recommend that the term “hermaphrodite” not be used in a medical context.

I quote from the ISNA website “The authors end by suggesting that it would be better to use specific etiology-based diagnosis (like AIS, 5-alpha reductase deficiency, etc.) along with an umbrella term. Which umbrella term? In our travels in the last few months, we notice there seems to be an emerging consensus around using disorders of sex development, abbreviated DSD’s This seems to be a term that doctors and many patient advocates can agree encompasses all of the conditions we at ISNA have traditionally labeled intersex.” (Note: You can visit the ISNA website for more information.)

One group of people that was not widely consulted on the proposed usage of the DSD nomenclature was the intersex community itself. The proposed use of the term “disorders of sex development” hit a raw nerve in the intersex community and continues to be a painful issue. Condition specific support groups have received many complaints from their membership about the DSD nomenclature. For example, Esther Morris Leiholf, President and founder of [MRKH.org.](#), and Sherri Groveman Morris, founder of the United States branch of the [Androgen Insensitivity Syndrome Support Group](#), report receiving numerous complaints about the DSD nomenclature. They both oppose the DSD nomenclature.

The [Organization Intersex International](#), led by Curtis Hinkle, has been very vocal in its opposition to the use of the term Disorders of Sex Development. In their analysis of the DSD nomenclature, they focus on what OII sees as the homo-phobic and trans-phobic nature of the DSD nomenclature. You can visit the OII website for more information.

I am opposed to labeling intersex bodies as “disordered.” I believe that the nomenclature “Disorders of Sex Development” further stigmatizes and pathologizes our lives. The DSD nomenclature strongly suggests that the ends of sexual development are either a normal female body or a normal male body. It ignores the vast diversity of the biological world as documented by Stanford biologist Joan Roughgarden in her book “Evolution’s Rainbow”. I find it interesting, that whereas both the hermaphrodite nomenclature and the intersex nomenclature implicitly accepted biological diversity, the DSD nomenclature is the least accepting of biological diversity. I believe that any move to have the DSD nomenclature officially recognized within the medical community would be wrong.

Where do we go from here? I hope that open discussion of the issues raised by the controversy over the DSD nomenclature will continue. The move to adopt DSD nomenclature has caused much pain in the intersex community. As the DSD nomenclature is hurtful to many intersex people, I believe that it would be a healing move not to use the DSD nomenclature.

Working Therapeutically With Adults With Atypical Sex Development

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Multiple Modes of Therapy Need Consumers' Guide For Intersexes

- Supportive
- Insight Oriented
 - Psychoanalysis
 - Psychodynamic psychotherapy
- Cognitive
 - Schema-based cognitive
- Cognitive Behavioral
- Eclectic
- Couples
- Family
- Group
- Sex
- Many others

Essentials

- Listen empathically
- Identify *patient's* problems and conflicts
 - Particular themes common with intersexes
 - Avoid assumptions
 - Don't lead the patient
- Goals
 - Not to solve problems for patient
 - Enable patients to deal effectively with their problems and conflicts
 - Facilitate self discovery and self acceptance

Essentials

- Assess social, family, relationship, sexual functioning
- Assess and treat any underlying psychiatric condition (e.g., depression)
 - Medication will not resolve problems but will enable patient to address them in therapy
 - Belief that problems are insurmountable may be symptom of depression

Essentials

- Clarify biological/surgical/hormonal status as relevant
 - Always relevant if therapist has medical license
 - Facilitate compliance with necessary medical care
 - Facilitate continuity of care
- Psychoeducation as indicated
 - Sexual differentiation
 - Sex vs gender
 - Sexual orientation vs gender identity
- Assess your limitations and know when to refer

Intersexes Need Empathic Validation

- History of deception by doctors
- Sense of having been betrayed by parents
- History of lack of validation
 - Concerns about genitalia, scars, medical/surgical treatments
 - Temperament/gender-stereotyped behavioral propensities

Patients will expect you to be like their other doctors (Transference)

- Many intersexes approach medical professionals with dread, mistrust, hostility and anger
- Medical professionals must analyze this transference and not respond with defensiveness and anger
 - E.g., labeling patient
 - "difficult"
 - "personality disordered"
 - "zealot"
 - "gender radical"

- Empathizing with distress will defuse anger and make a trusting therapeutic alliance a possibility
- Gay/gender affirmation begins in the waiting room
 - GLBT Magazines
 - Rainbow flag, etc.

Common Failures of Empathy

- Assurance that doctors and parents were well-meaning
- Assurance that treatment was state-of-the art
- Assurance that parents were following doctors' advice

The above may be true, but prior to empathic validation of patient's concerns such interpretations will damage therapeutic alliance.

Understanding Origin of Common Themes in Therapy

- Anger toward medical professionals
- Strained relationships with parents
- Guilt
- Shame
- Defectiveness
 - Low expectations for success in intimate relationships

Surgical Perspective On Intersex

"The birth of an intersexed child is a psychosocial emergency in the delivery room. The parents will be in a state of shock and unable to process the information necessary to make decisions. They will rely on you. You must be confident that you know the right thing to do and assure parents that you can make that little problem go away."

Surgeon addressing infant genital surgery at CME surgical conference at major NYC medical center (1995)

- Doctors cannot make intersex go away
 - Belief sets up false expectations, repeated disappointments
 - Belief responsible for patients being abandoned by adulthood
- Intersex is fixed by destigmatization and acceptance

Role of Mental Health Professionals

- Psychiatric emergency requires psychiatric intervention, not irreversible genital surgery
- Discourage irreversible decisions in atmosphere of crisis
- Minimize shame and secrecy
- Involve parents in decision making processes
 - Informed consent requires realistic expectations
 - Doctors cannot make intersex go away
- Distinguish between gender assignment and genital surgery
- Foster parent/infant bonding

Some Current Guidelines Impede Rather Than Foster Parent/Infant Bonding

- Frimberger and Gearhart (2005) *Urol Int* 75:291-97.
 - "For the inexperienced physician it is often best not to discuss the issue with the parents but to transfer the child to a center."
 - "Until a diagnosis has been made and a sex of rearing decided parents should be discouraged from naming the child"

Foster bonding between infant and parents from the moment of birth

- Do not convey sense of disgust or undue alarm to parents
- Allow parents to hold infant
- Transfer to intensive care nursery only if necessary (e.g., salt wasting CAH)
- Comment on positive attributes of the infant
- Encourage gender-neutral name rather than delaying naming until gender assignment is made

Intersex Does Not Go Away

- Maintain rapport and an ongoing discussion with the child about his or her intersex status in an age appropriate way
 - Distinguish between child's desires for his or her body, gender role and gender identity as opposed to child's desire to please parents
- No one should have to discover their intersex status in an uncontrolled setting

BODY LANGUAGE: WHAT THERAPISTS CAN LEARN FROM CLIENTS WITH DSDS

Nina Williams, Psy.D.

*Delivered at DSD Symposium (ISNA and GLMA), October 14, 2006,
Renaissance Parc 55 Hotel, San Francisco*

I am honored to be among people whose lives and work have taught me so much. Thank you. I am a psychologist who does sex therapy and psychoanalysis, trains other clinicians, and teaches people in health care about DSDs.

I'd like to give you a handout of suggestions for ways you can help clients with disorders of sexual development who are struggling with some aspects of their condition (see Appendix). You will notice the interventions in this handout are nothing new, and I'm not alone in wondering why it is that intersexed adults have so much trouble getting competent help in psychotherapy when nothing about their psychology is unique.

But although I can't say that I'm doing now is so different from what I was doing twelve years ago when I first worked with someone who was intersexed, I am sure that I am better at it now. What I believe has made my work more effective is finding a level of self-awareness where I can—more often than not —think clearly and respond empathically. I'd like to start with a vignette from a long-term treatment with a woman with atypical genitals. Then I'd like to talk about what this experience has taught me, which is not just about DSDs, but about our shared personal and societal reactions to these individuals.

What is different about how I handled my work with these clients from how I responded a few years ago? First, I know now that a client's reluctance to be specific about their medical treatment isn't a resistance to the therapy but a way of remembering her experience with doctors, where she was frequently exposed against her will. Second, I understand that my clients have seen me in various guises over the years, from a dangerous, pain-inducing doctor, to an idealized loving mother. Sometimes they wish had the power to take the medical history out of the mind so they never have to think about it again.

What do we make of such a client's desire to destroy her or his inner life? The analyst Christopher Bollas describe families in which the production of a 'normal' child necessarily made the messy, inner life of the child a dangerous threat. These are the kinds of circumstances in which families raising a child with DSDs are placed by recommendations of secrecy. In such a family, the parents do not "facilitate the creative expression of the inner core of the self." (1987, p. 144) What they respond to is "the child's adaptation to convention, with praise and material reward." By adolescence, when these children have learned not to complain, they may appear to have everything they should want except vitality and enthusiasm.

Rogers (2006) wrote that the first loss of a trauma is the realization that words cannot entirely express the experience. Trauma “is unsayable,” writes Rodgers (2006). My work with adults who have DSDs reminds me that the opposite is also true: that the unsayable becomes traumatic. My opposition to resolving parent anxiety with surgery is that it supports the illusion that damage that cannot be seen will not need to be talked about. For most of my patients, this pretense is deeply disconnecting.

Just as there’s a difference of opinion among physicians about how to help people with DSDs, there’s an identical difference among therapists. Psychotherapy can be seen as helping the client fit in with other people by reducing the behaviors, thoughts, and feelings that separate the individual from the world of ordinary life. Or psychotherapy can be seen as helping the patient accept his or her individuality, the ways we all sometimes fail to meet our own or other people’s expectations, and occasionally rise above them. Psychological health is defined as the freedom to make conscious choices between fitting in or standing alone in each situation.

This work has taught me how much psychological power is wielded by the body, particularly what write Susan Wendell calls “the rejected body” the one our culture regards as troublesome because it doesn’t function the way we want it to (1996). This psychological power expands exponentially when the part of the body that is rejected is the one between our legs.

For someone with ordinary anatomy, there is something uncanny about meeting an adult with a DSD. I want to apologize for this, and to be honest about it. The idea that people don’t neatly fall into two categories is on one level exciting because it revives the childhood wish that biological sex is fluid. But it’s also shameful for therapists because it makes you, even momentarily, think about some else’s privates. That’s an inevitable human curiosity, but given the harm this uncontrolled curiosity has caused, it can temporarily pull you out of your professional identity as a helper.

Thus, life for someone with a DSD is governed by our culture’s discomfort about their bodies. Our higher reasoning buckles under the weight of that preoccupation, and intersexed people suffer the results. Fifty years ago, in the same era in which John Money proposed his theory of psychosexual neutrality at birth, another author wrote, “having to associate … with people … with different physical attributes is a severe attack on the feelings of identity, and thus missionaries are always sent in pairs even to non-dangerous areas” (Greenacre in DeLevita, 1966).

Two years ago, I put together a DSD training group for other therapists. The group met one afternoon a month for a year to read, talk to experts, and process the impact of the material. There was a selfish motive to this; I wanted a group of peers with whom I could talk about this material.

All ten members of the group were female, one pregnant and nine in menopause or beyond. One idea we have about this was that having this small but personally and

socially disorientating experience at the mercy of our biological weather gave the members a taste of how the body governs one's sense of personal integrity and belonging.

Over time, the thinking got deeper. Members reported wondering privately whether their own biological sex was reliable, questioning the meaning of minor physical differences, a childhood preference for rough and tumble play, a mother who seemed particularly strict about her daughter's assumption of traditional gender role and sexual orientation. One member remarked, "maybe nobody feels female for the same reason; it's just that we aren't ever asked to prove it." Gradually, the group moved away from a fascination with gender. I was relieved, because I feel too much gets made of this topic anyway.

Instead, members began to relate to the experience of secrecy, stigma, and shame through their memories of rejection. For one, it was being teased as a child for wearing a leg brace. Another described being Jewish in a largely Christian school. One woman told about being so ashamed of her chubby body that she changed her clothes under her nightgown at a slumber party until the other girls pulled the nightgown off and pushed her, naked, into the hallway.

For me, my lifelong experience of serious depression gave me a connection to these feelings. Depression yields the same two social effects of making people sorry for you and a little afraid of you, too. Although I've become outwardly at ease in disclosing this, inside there is always the sinking feeling I've just destroyed my chance for equal standing. All those years in therapy, being the only person who is exposed in the room, so hoping for a cure, that I came to feel that being looked over by the pros was about as close to being accepted and understood as I was going to get. I think making this connection and feeling my way through it in my own therapy is the single factor most relevant to my developing competence with clients who have intersex conditions.

Although the group ultimately spawned several activists, who are busy giving talks and correcting misconceptions about gender and sexuality, one of the experiences the members reported was most helpful to them was the chance to share their own secret fears about not fitting into their sex. How universal was this concern, I wondered, and what would happen if I began to ask patients about it?

Quite a bit, as it turns out. Because many of my clients go to therapy for a sexual problem, the topic is easier to introduce than I imagined. One patient with vaginismus told me she secretly believed that unless she could enjoy intercourse, she would never truly be a woman. Clients with histories of other childhood illness as well as those with various problems in relationships have also disclosed fears that their difficulties in intimate relationships are because their bodies have been damaged or are not normal. Without the experience of working with intersexed people, it would not have occurred to me that having the chance to name this fear and see that the ceiling does not collapse is both a relief and enlightening.

If I were really blunt about what I think would help therapists do better work with DSDs, I'd say, ponder your most secret fear, your deepest shame, your worst moment of humiliation. Then print it on a t-shirt and wear it to your high school reunion.

I know I sound angry, and I am. What psychotherapists have to offer anyone is an understanding of human experience, including the parts that are hardest to think about. I believe that if we professionals could seriously analyze how fascinated and anxious DSDs can make us, we might leave you and your bodies alone. We might appreciate the potential of a culture that appreciates variety and start to learn from you. For the time being, our ignorance gets in the way of empathic practice, and that's a dangerous weapon when you have a license to stare.

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COMMON CONCERNS, HELPFUL STRATEGIES, AND HAZARDS IN CLINICAL WORK WITH ADULTS WHO HAVE DSDS

Compiled by Nina Williams, Psy.D.

CONCERNS

- Self-disclosure to sexual partners, friends, family (what, when, and how)
- Lack of pleasure in sex, participation in sex to achieve normalcy or acceptance, and rigidity about acceptable acts.
- Getting full information about condition
- Loss of control over disclosures made by health workers or parents
- Medical trauma (public exams, painful or humiliating procedures)
- Avoidance or idealization of relationships
- Sense of damaged self, compromised gender
- Fears of devaluation and rejection, passivity
- Avoidance of or difficulties with medical care
- Loneliness and social isolation
- Changes in expectations of life following discovery of diagnosis.

STRATEGIES

- Explore and question assumptions about ‘normal’ sexual activities and gender
- Support search for medical information, family history
- Encourage social contact and identify resources
- Be transparent, humble, and empathic, but realistic about limits of treatment.
- Deliver information sensitively
- Let patient set agenda
- Make time to tell history, grieve losses, and notice repetitions
- Encourage behavioral change
- Suggest reading books, keeping journal
- Challenge self-destructive treatment of body and health
- Start a group
- Become part of a multi-disciplinary team

HAZARDS

- Lack of knowledge of conditions
- Not consulting with peers or getting supervision about counter-transference
- Focusing on anatomy or gender rather than emotional experience of patient
- Not knowing one’s feelings about intersex conditions
- Idealizing/devaluing of patient, treatment, self

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A team approach to coordinated care for individuals with DSD

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Outline

- Why a team?
- Discussion of two teams' approaches
- How does a team work?
- Benefits of team approach
- Keys to success
- Challenges to effective teams
- Questions and comments

Purpose of Team Approach

- Diagnosis
 - Clinical
 - Laboratory
 - Molecular
 - Surgical
- Management
 - Medical
 - Surgical
- Research
 - Longitudinal data
 - Outcomes
- Prenatal consultations
- Genetic counseling
 - Recurrence Risks
 - Other family members at risk
- Psychosocial/support
 - Parents/families
 - Patients
- Education
 - Best practices for patient care
 - Journal club/invited speakers
 - Patient advocacy groups

Definitions

- Terms we avoid:
 - Intersex
 - Sex reversal
 - Pseudo-hermaphroditism
 - True hermaphrodite
 - Testicular feminization syndrome
- Terms we prefer:
 - Disorders of sex development (DSD)
 - Precise definition (when known)

Composition of the Ideal Team

- Medical Genetics/Cytogenetics
- Pediatric Endocrinology
- Pediatric Urology
- *Child Psychology
- *Child Psychiatry
- *Social Work
- Genetic Counselor
- Gynecology/Reproductive endocrinology
- Child Life
- Nursing
- Chaplaincy and Bioethics-ad hoc

*Need at least one from this group, but ideal to have all 3

The Seattle approach: urgent situations (newborns with ambiguous genitalia)

- If stable, bring to home institution for evaluation
- Specialists meet with family within 2 days
- No discussion regarding gender assignment with family before team meets
- Group conference is ideal, with family present
- Gender assignment made as soon as possible
- Hormonal profile at 2–4 months of age is important in newborns ("mini-puberty")
- Close followup at 2–4 weeks, 8 weeks, and beyond
- Provide resources and support group information throughout (www.isna.org)

The Seattle approach: non-urgent situations

- Triage system with point person–Genetic counselor
- Consults are seen by all appropriate specialists at one time or within a few weeks in outpatient setting
- Discussion with family held after healthcare team has gathered information and conferred
- On-going care: regular followup with referrals to appropriate subspecialists
- Primary care physicians involved
- Team conference every 2 months:
 - Short-term issues: diagnosis and management
 - Long-term issues: longitudinal followup of patients

The Seattle experience over 25 years: 1981-2005

250 patients evaluated

On average, 10-15 new patients/year

- Infants 76%
- Children/Adolescents 17%
- Known multi-system genetic syndromes 7%

The Seattle experience: 6 most common diagnoses

- Congenital adrenal hyperplasia 12%
- Androgen insensitivity syndrome 10%
- Clitoromegaly / labial anomalies 8%
- Mixed gonadal dysgenesis 8%
- Hypogonadotropic hypogonadism 8%
- 46,XY SGA males with hypospadias 7%
- TOTAL 53%

The Toronto approach: urgent situations

- Social Worker– point person
- Either patient brought to HSC or we go to them (also for prenatal consultations)
- If team can't go together, SW stays for all consults
- Family given recorder to tape all discussion (helps to clarify and for digital record to help patient later)

- Crucial from the beginning to stress developing a narrative, for the child to understand
- Normalize from day one, telling patient, and developing a plan
- Consultations with others as requested
(i.e. siblings, grandparents, cousins, etc.)
- Clarifying information the family is getting, helping them to understand

The Toronto approach:

- MUG team clinic 2nd Friday of the month
- 3-10 patients seen
- Group rounds, discussion of results and updates, decision who needs to see
- Follow-up on a developmental timeline
- Initial workup, then at age 2-3, school entry, latency, adolescence,

Clinical Issues that arise in the MUG Clinic

- Non compliance
- Telling
- Psychological testing
- Post Traumatic Stress
- Ongoing Education
- Mutual Support Groups
- Connecting families from the start
- Culturally sensitive care

Gender Assessment Team

Diagnostic Approach

Multiple malformations

Isolated genital anomaly

Syndrome identification:

- Multiple congenital anomaly syndrome
- Autosomal chromosomal abnormality
- Exstrophy of the cloaca

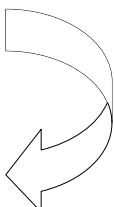
Determination of:

- Chromosomal sex
- Gonadal sex
- Phenotypic sex

GOAL

- Chromosomal sex
- Gonadal sex Parental input
- Phenotypic sex

Sex of Rearing



Timing: As soon as possible based on best information

Basic Evaluation: Abnormal genitalia or DSD

- Perinatal history
- Past medical history
- Family history
- Psychological assessment of parents
- Complete physical examination
- Genital examination
- Pelvic imaging – US/MRI
- Blood karyotype with FISH- X and Y probes
- Endocrine evaluation: 17OHP, Testosterone, LH, FSH, etc

Additional factors to consider in Gender Assignment

- Fertility Potential
- Capacity for Normal Sexual Function
- Endocrine Function
- Risks of Malignancy
- Prenatal Testosterone Effects
- Timing of Surgery

Approach-Psychological Evaluation

Family's concerns and wishes:
Cultural/religious/family values
Desired gender of child
Gender identity in adulthood
Sexual orientation
Sexual function
Potential for fertility

Psychosocial Implications: Working with Families

- Assign sex with understanding that child may make a different decision later in life
- Postpone surgical interventions if possible until child is old enough to participate in decision making
- Importance of team approach, full disclosure, ongoing education and support for patients and families

Recommendations for talking to parents

- In the delivery room: "The sex of the baby cannot be determined yet."
- Refrain from assigning gender: admit the infant as "Baby" Lastname
- Refer to the infant as "the baby" or by name if parents have chosen one
- Let them know that the baby has a difference in the formation of the genitalia, similar to babies born with a heart defect or cleft lip
- Assure them that a team of doctors familiar with these issues will be meeting with them and that they will be involved in the process

<http://www.dsguidelines.org>

Recommendations for talking to parents

- Avoid repeated exams with multiple trainees and limit photography
 - Do not make the infant a spectacle
- Suggestions for parents regarding disclosure:
 - Name a friend/family member to serve as contact to field calls
 - "We are pleased to announce the birth of a healthy baby."
 - "Our baby was born with a urogenital difference and is undergoing medical evaluation."
- Explore with them their biggest concerns:
 - Talking to their other children, who to bring in, etc.
- Remind them that although this is hard time for them, the baby will not remember the newborn period
- Encourage them to keep mementos and share this story with their child in an age-appropriate, honest manner

<http://www.dsguidelines.org>

Benefits of the Team

- A multidisciplinary approach aids in timely diagnosis, management, and counseling
- Adequate experience is acquired to identify and manage under-recognized conditions
- New conditions may be identified leading to familiarity with their treatment
- Stay current with evolving recommendations

"Neilson and Parisi's" Top 6 Keys to a Team's Success

#6. Broad representation by multiple specialties

- Having at least one strong member in each key specialty is important
- Having pediatric urologists who are sensitive to these issues is important

#5. Strong leadership

- People want to work with dynamic leaders
- A good leader can corral the renegade specialists

#4. Good communication

- The key to almost everything in life!
- Knowing the email address, back-line, clinic coordinator, and pager number of team members is essential

#3. Mutual respect and trust

- You might not be best friends with your coworkers, but you have to trust their judgment
- When you call them on a Friday night at 10 pm, they will respond positively

#2. Efficient coordination

- Analogous to “herding cats” !
- The Coordination can be a team effort:
Toronto: Endocrinology (Medical Director) and Social Work (Coordinator)
Seattle: Genetics (Medical Director)

And the #1 key to team success: Humility

- There is inadequate longitudinal data on outcomes for DSDs
- As healthcare professionals, we cannot know everything about these complex DSDs
- We must respect the experiences of our patients and their families

Special challenges to effective teamwork

- High turnover of medical staff
- Multiple specialists with complicated schedules
- Increasing numbers and complexity of patients with DSD
- More testing/evaluation options
- Lack of consensus regarding treatment:
 - “That’s not the way we did it at Institution X...”
- Medical model that emphasizes efficiency and expediency over coordinated care

Creating a smooth transition to adult care

- Providing complete medical records to patients and families
 - Clinic notes/operative reports
 - Chromosome reports
 - Specialized testing
- Gynecologist who takes female patients to the adult hospital
- Pediatric urologists who gain privileges at adult hospitals
- Geneticists often can see adults in their practice
- Celebration of transition to adult care

If you want to put together a team....

- Consider enlisting your colleagues!
- Resources available:

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- Put these issues on agendas for discussion at future medical and scientific meetings
- Put the word out—advertise your services as a team, and deliver

Additional Resources: Information for families

- Hospital for Sick Children genital development web page: www.sickkids.ca/childphysiology/cpwp/Genital/GenitalIntro.htm
- Clinical guidelines and parent handbook for management of disorders of sex development in childhood: www.dsrguidelines.org
- Intersex Society of North America: www.isna.org
- AIS support groups: www.aissgusa.org, www.medhelp.org/www/ais/

Acknowledgments

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A Psychoeducational Treatment Manual for Families of Newborns with CAH Broadening the Goals of our Interventions

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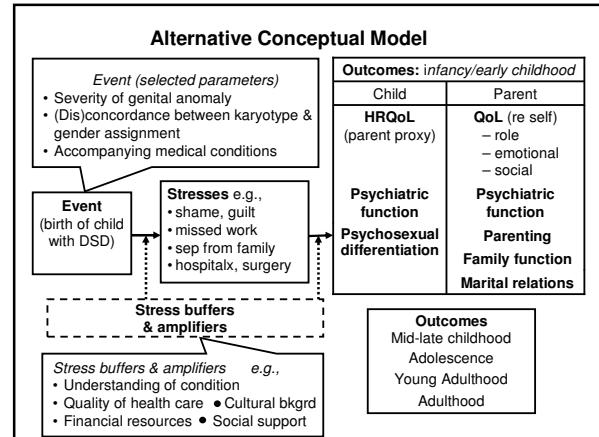
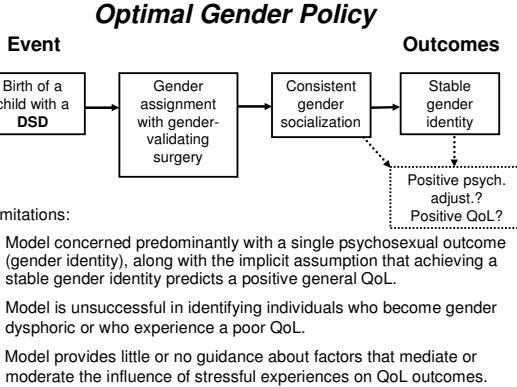
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CAH and Newborn Screening (NBS)

- Because CAH is life-threatening if not detected early, it is screened for at birth
- U.S. moving toward a uniform panel across states, including 29 conditions
- Many conditions identified by NBS are rare; health care providers may lack adequate knowledge of the condition and associated management
- For families, diagnosis confirmation can precipitate crisis

- Inundated with complicated information/complex medical terminology
- Difficulty in processing information compounded by emotional distress; disbelief that apparently healthy child has a chronic, life-threatening condition, and (unjustified) feelings of guilt
- Emotional distress/limited understanding of medical information interfere with clinical decision-making and adherence to recommended treatments

- Parental emotional difficulties can place child at risk for behavioral and emotional problems
- All reactions exacerbated in conditions associated with stigma (eg, physical appearance)



Alternative Conceptual Model

Advantages:

1. Expands domains of study beyond formation and stability of gender identity.
2. Elucidates the domains of child and family experience influenced by *disorders of sex development* and highlights areas where clinical interventions could be focused to maximize positive QoL for patient and family across the lifespan.
3. Provides information that expands the theoretical model explaining psychosexual differentiation and generates testable hypotheses combining biological and social factors.

Psychoeducational Treatment Manual

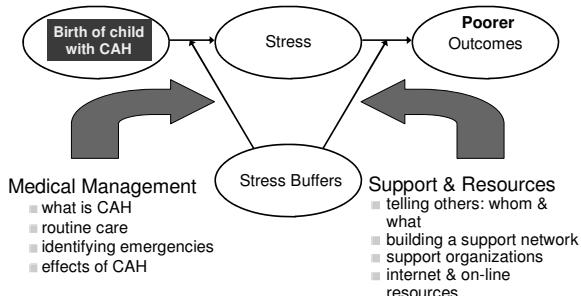
Objective:

- Develop a comprehensive psychoeducational treatment manual *for clinicians* to reduce emotional stress and enable families to be informed participants in clinical care and decision-making

Goal:

- Promote a positive *health-related quality of life* in affected children (and their families)

Conceptual Model



CAH: A Family Guide Format

Treatment manual (clinician administered):

- 12 to 16 sessions (~1 hr duration)
- Presented to the family over the course of child's first year
- Modules presented to family by members of the healthcare team (peds endo, nurse, psychology, urology, etc)
- Manual for clinicians includes handouts for families and model letters for ED, school, child care, etc

CAH: A Family Guide Format

Interactive DVD for family:

- Reviews all information covered by healthcare professionals
- DVD provides simulated discussions (employing actors) among parents and affected adults
- Each module includes a multiple choice test to assess knowledge

CAH: A Family Guide Format

Benefits of DVD for families:

- Opportunity to review content between return visits to hospital
- Educate family members unable to attend clinic visits
- Share information with family, friends, and others
- DVD is 100% reliable, ie material is never forgotten/skipped

A Family Guide to CAH: CAH-Specific Modules

Medical Management	Behavior & Psychological Development
<ul style="list-style-type: none"> ❖ What is CAH ❖ Routine care ❖ Identifying emergencies ❖ Effects of CAH 	<ul style="list-style-type: none"> ❖ Cognitive & academic ❖ Emotional & behavioral ❖ Gender identity ❖ Gender role ❖ Sexual orientation
Genetics	Surgical Considerations for Daughters
<ul style="list-style-type: none"> ❖ Genetic counseling ❖ Addressing <i>guilt & shame</i> ❖ Heritability of CAH ❖ Family planning ❖ Informing others 	<ul style="list-style-type: none"> ❖ Informed "permission" ❖ Balanced discussion of benefits/risks

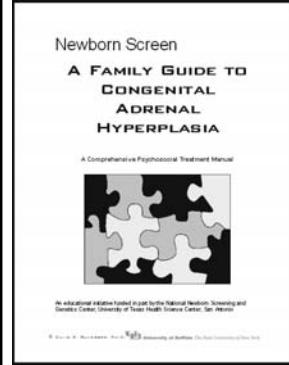
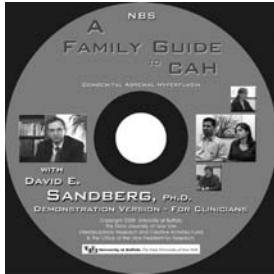
A Family Guide to CAH: Common Modules

Support & Resources	Taking Care of Yourself
<ul style="list-style-type: none"> ❖ Telling others: whom & what ❖ Building a support network ❖ Support organizations ❖ Internet & on-line resources 	<ul style="list-style-type: none"> ❖ Importance of parental emotional health ❖ Problem-solving skills ❖ Coping strategies ❖ Sharing responsibilities of caring for child & family
The Health Care Team	
<ul style="list-style-type: none"> ❖ Members of the health care team ❖ Roles & relationships of each member ❖ Effective interactions with healthcare professionals 	

A Family Guide to CAH: Common Modules

Parenting a Child with Chronic Health Condition
<ul style="list-style-type: none"> ❖ Effects of child's condition on the family, parenting, siblings ❖ Parenting strategies ❖ Sharing parenting responsibilities
Money Matters
<ul style="list-style-type: none"> ❖ Health insurance ❖ In-hospital resources ❖ Foundations providing assistance ❖ State & federal assistance programs

A Family Guide to CAH: Manual & DVDs

Also a DVD for families

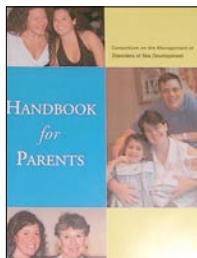
Next Steps

- Complete interactive DVD for parents
- Pilot/feasibility test the manual in different healthcare settings
- Revise the manual and develop training program for healthcare professionals
- Conduct a randomized clinical trial

Acknowledgements

- The manual, *A Family Guide to CAH*, developed under contract to National Newborn Screening and Genetics Resource Center (119525/118815)
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Handbook for Parents



Arlene B. Baratz MD

free download: www.dsguidelines.org

Parents Handbook

- What is the handbook?
- Usefulness for parents
- Usefulness for physicians and counselors



"There is no instruction manual!"

- Parents Handbook addresses parenting issues
- Information and support for parents
- Strategies for communication with children
- Ideas for interacting with others

Health care professionals

- Understand and anticipate needs of parents
- Provide advice and information
- Source for contacting support groups

Need for Handbook

- Little parenting guidance from physicians
- Scarce information on parent needs for counselors
- Few resources on how to deal with the whole child
- Rare access to trained counselors

Previously available

- Terminology from the 1800's
- Gender philosophy from the 1950's
- Treatment in a vacuum
- No data on outcomes

Prior generations of parents

- Guilt for keeping secrets
- Shame for not questioning doctors
- Grief for damage to adult child's well-being
- Regret for alienation of child



Parent stress raising DSD child

- "There is this 'breath' I hold, I can't let it out right now no matter how hard I try, and it's waiting for something terrible to happen, for something to go wrong, for AIS to destroy us, for me to handle something wrong."

• AIS parent

Nomenclature and Handbook

- Rejection of "intersex" by AIS parents
 - "Brutal"
 - "Damaging"
 - "Stigmatizing"

Parents and DSD

- Disorder of sex development explains child's condition with neutral medical terms
- Fewer political and social implications

DSD terminology

- Acknowledges parents' concerns
- Goals of care
 - Functionality of genitals
 - Physical and emotional health of child
 - Integrating care with a team

Acceptance of Handbook

- Fifty-four parents in AISSG-USA parents' circle
- Acceptance of "DSD" with new paradigm of care
- Not one parent objected to "DSD"

Do parents want this information?

- Fifty copies of Handbook distributed in August 2006 at national AISSG meeting
- Enthusiastic reviews in AIS parents email circle

Parents and Clinical Guidelines

- Expectations for child's care
- Participation in a team approach
- Discussion with healthcare providers

Previous role of physicians

- Help with medical issues
- Lack of referral to support groups or therapists

Modern role of physician

- Part of a team including family
- Work with multidisciplinary approach
- Track outcomes
- Help determine "best practices" with data



Chapter one: Welcome to Parents

- Understanding facts of the condition
- Assimilating condition into
 - Parents' lives
 - Life of the child and family
 - Wider circle of people



Acknowledging parents' feelings

- Isolation
- Grief
- Fear of damaging a fragile child



Cycle of emotions

- Loss
- Grief
- Acceptance



Helping parents help their children

- Support system
 - Family
 - Support groups
- Asynchrony of family emotions
- Acceptance of own feelings



Secrecy and isolation

- Hide intense feelings from child
- Difficulty discussing with family
- Result: no one to empathize

Development

- Age 12-36 months
- Age 3-5 years
- Age 6-11 years
- Adolescence



Evolving strategies

- Context of mental and physical development
- Unique nature of each child



Stages of development

- Understanding emotional maturity
- Age-appropriate information
- Suggestions for conversations with child
- Gradual independence of child



Chapter 3: How to Talk with Others

- General information about condition
- Delayed gender assignment
- Religious considerations

Talking with medical providers

- Describe child
- Long-term goals
 - Healthy child
 - Positive body image
 - Child is loved and lovable



Preparing for a medical appointment

- Questions
- Comments
- Notes

Diagnosis, please!

- Explain tests and procedures
- Discuss differential diagnosis
- Insist on the truth

Urgency of medical issues

- Most are not life-threatening
- Understand and treat real medical problems

Medical records

- HIPAA gives legal right to have copies
- Right to correct the records
 - Case of incorrect gender on birth certificate
- Medical notes and test results

Parents' underlying fears

- Team to address:
 - Picking the "wrong" gender
 - Masculinization of a girl
 - "Causing" homosexuality

Privacy of child

- Avoid the "parade"
- Decline repeated genital exams
- No pictures!
- Understand young doctors must learn-set parameters



Psychological support

- Need to communicate when there is an emergency
- Professionals- psychiatrist, trained therapist
- Physicians who care for DSD adults

Support

- Request names of individuals and groups
- Offer to support others

Gender assignment

- Sex of rearing
- Which gender will child feel?
- Support for child to explore identity

Genital surgeries

- Evidence, evidence, evidence!!!

Delaying genital surgery

- Child understands and can consent
- Outcome of childhood surgery in adulthood
- American Academy of Pediatrics policy
 - Informed consent
 - Right to have all information
 - Rights of child

Questions for surgeon

- How many have you done?
- Data on outcomes
 - Physical
 - Emotional
 - Individual satisfaction

Questions for endocrinologist

- Purpose of gonadectomy and hormone therapy
- Growth pattern
- Understanding puberty

Before any treatment

- Discuss with people who have undergone the treatment
- Support group resources

Gender and sexuality

- Play behavior not gender-specific
- Influence of prenatal hormones
- Unusual to have discordance with gender assignment
- Help child on journey of gender discovery

Sex, gender and sexuality

- Gender
 - Identity as boy or girl
- Sex
 - Physical aspects
- Sexual orientation
 - Straight
 - Gay



Sexuality

- Expression of sexual feelings
- Component of intimate relationships



Parents and sexuality

- Experiencing that part of life
- Desire for child to have intimate, loving partnership with another
- No parent can predict or assign blame
- Groundwork for acceptance

Adult relationships

- No inherent obstacles
- Family formation
 - Adoption
 - Surrogacy



Future assisted reproduction techniques

- Possibility of preserving potential "gametes"



DSDs and Cancer: Caring for Intersex Patients

Presented by Katharine Baratz
13 October 2006

Objectives

- How a DSD patient's medical history impacts necessary and routine screenings
- How a patient's diagnosis elevates risk of malignancy in gonadal and genital tissues
- How best to develop an effective doctor-patient relationship with DSD patients

Survey

- Issued to AISSG-USA
- Questions
 - Do these patients feel that they have been maltreated by the medical community?
 - If so, does this mean that they are less likely to put themselves into medical settings in which they feel uncomfortable?
 - Does this put them at higher risk for undetected, non-intersex specific, cancers?

Background Information

- 33 participants
- Mean age: 46 years old
 - Range 21 - 75
- Diagnoses
 - 67% CAIS
 - 15% PAIS
 - 6% AIS (degree unknown)
 - 9% GD
 - 3% Other (Ovotesticular DSD)

Medical Habits (Non-DSD)

- Asked for frequency of screenings of three common cancers:
 - Breast
 - Skin
 - Colorectal

Breast Cancer

- 75% have ever had a mammogram
 - 63% within the last year (ACS recommended)
 - 37% more than one year ago
- Only 40% have regular mammograms (once per year)
- 87.5% have ever practiced self-examination
 - 62.5% at least every few months

Breast Cancer (cont.)

- 75% of women surveyed over 40
- Family History
 - 13% in immediate family (can double risk)
 - 19% in extended family
 - 68% no family history
- 10% have had breast cancer

Breast Cancer Conclusions

- While women over 40 do undergo mammograms as advised, only a small majority go as frequently as recommended.
- Many women have practiced BSE.
- Majority of women no family history.
- Thus these women are aware of the risks and are willing to exam themselves but are perhaps unwilling to undergo mammograms.

Vaginal Cancer

- 40% see a gynecologist annually
- 70% have had a vaginal exam within the last 2 years
- No reported incidence of vaginal cancer

Vaginal Cancer Risk Factors

- Human papillomavirus (HPV)
 - 65% - 85% of vaginal cancers contain HPV
 - HPV infection increases with sexual activity
 - 75% sexually active within last 6 months
 - Mean number of partners: 12 (Median: 5)
 - 13% have been diagnosed with and treated for STI

Vaginal Cancer: Conclusions and Questions

- The vast majority of women are currently sexually active.
- A relatively high number of women have engaged in high-risk behavior
 - Multiple partners
 - Potentially less likely to use protection?
- Only 40% are seeing a gynecologist regularly, and are thus at risk for undetected vaginal cancers.

What about Vaginoplasty and Vaginal Cancer?

- 13% had undergone vaginoplasty
 - Only 1 had vaginoplasty with skin grafts (from her thigh)
 - Other 3 from surgical dilation
- Does vaginoplasty increase risk of vaginal cancer?

Malignancy of the Neovagina

- Schober J. "Long-Term Outcome of Feminization"
 - 14 cases of cancerous neoplasia: all from split-thickness skin graft or McIndoe variations (involving grafts)
 - Risk of development no greater than natural vagina
- Only small minority of this group have had vaginoplasty, surgical dilation favored

Looking Back

- Women in this group were found to be less likely to have breast and vaginal examinations.
 - 84% feel that they have exams about as frequently as recommended

Gonadal Cancers

TABLE 4 Risk of Germ Cell Malignancy According to Diagnosis

Risk Group	Disorder	Malignancy Risk, %	Recommended Action	Patients, n	Studies, n
High	GD (+Y) intraabdominal	15–35	Gonadectomy*	12	>350
	PAIS nonscrotal	50	Gonadectomy*	2	24
	Frasier	60	Gonadectomy*	1	15
	Denys-Drash (+Y)	40	Gonadectomy*	1	5
Intermediate	Turner (+Y)	12	Gonadectomy*	11	43
	17β-hydroxysteroid	28	Watchful waiting	2	7
	GD (+Y) scrotal	Unknown	Biopsy* and irradiation?	0	0
	PAIS scrotal/gonad	Unknown	Biopsy* and irradiation?	0	0
Low	CAIS	2	Biopsy* and ???	2	55
	Ovotesticular DSD	3	Testicular tissue removal?	3	426
	Turner (−Y)	1	None	11	557
	5αR2D2	0	Unresolved	1	3
No (?)	Leydig cell hypoplasia	0	Unresolved	1	2

*Gonadal dysgenesis (including not further specified, 46,XY, 46,X/46,XY, mixed, partial, and complete).

†GBY region positive, including the TSPY (testis-specific protein Y encoded) gene.

‡At time of diagnosis.

§At puberty, allowing investigation of at least 30 seminiferous tubules; preferentially diagnosis on the basis of OCT3/4 immunohistochemistry.

From Hughes, 2006

Malignancy: GD

- Cools M, et. al., J Clin Endocrinol Metab. 2006 June
 - Overall incidence of germ cell tumors = 35%
 - Evidence that carcinomas tend to arise in areas of differentiated testicular tissue
 - Gonadoblastoma in undifferentiated OCT3/4+ germ cells
- Pena-Alonso R, et. al. 2005 Mar
 - 4 develop gonadoblastomas by age 2
 - 1 presents bilateral dysgerminoma at 15
 - Advise prophylactic removal of gonads

Malignancy: General

- Fallat, et. al. Curr Opin Pediatr 2006
 - Divide patients into groups according to increasing risk:
 - Presence of Y chromosome with intra-abdominal gonads (AIS)
 - Dysgenetic gonads (Swyer's)
 - Dysgenetic syndromes with splice variants of the WT1 gene (Frasier and Denys-Drash)

Survey Group

- 2 Cases of Gonadal Cancer
 - 1 CAIS (of 22)
 - Age 20, gonadectomy
 - Unilateral tumor, unilateral precancerous
 - 1 GD (of 3)
 - Age 12, gonadectomy
 - Unilateral tumor
 - 2nd “streak” removed at 37

Gonadectomy?

- Vast majority (over 96%) of patients have undergone gonadectomy.
- Mean age: 16
- 20% believe that more than half of women will develop cancer
- Given low incidence of cancer, especially for CAIS, may be unnecessary
 - Investigate pathology reports from surgeries
 - Monitor gonads at yearly exam

(Dis)Honesty

- Both previously mentioned cases, neither patient informed of karyotype, tissue identity, or cancer until much later (ages 46 and 37, respectively).
- Over 20% of patients surveyed responded that doctors had informed either the patient or her parents that she had cancer when she did not.

(Dis)Honesty cont.

- Concerning their diagnoses, 55% of patients feel that they have been lied to, and only 38% feel that their doctors have been honest.
- 60% feel that their experiences make them less willing to go to the doctor.

What can we do?

- Patients who feel comfortable with their physicians describe them as:
 - Honest
 - Willing to learn if not already well-informed
 - “I don’t inherently distrust physicians themselves -- I just distrust the depth of their knowledge.”
 - Sensitive to issues (infertility, amenorrhea, innate distrust of medical community)
 - Willing to make time to talk with them

But above all...

- “I understand my body and it doesn’t scare me,” so it should not scare or fascinate physicians.
- Don’t treat DSD patients in a way that is any different than other patients. They are not worthy of time because they are “fascinating cases” (especially if they are seeing someone for care irrelevant to a DSD), but because they are, above all, a patient.

The women of the AISSG-USA

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TABLE 4 Risk of Germ Cell Malignancy According to Diagnosis

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^b GBY region positive, including the TSPY (testis-specific protein Y encoded) gene.

^c At time of diagnosis.

^d At puberty, allowing investigation of at least 30 seminiferous tubules, preferentially diagnosis on the basis of OCT3/4 immunohistochemistry.

“Love, too, has to be learned”: Reflecting on Love in Counseling Parents

Ellen K. Feder¹

American University

DSD Symposium, October 2006

In 2004, Archives of Pediatrics & Adolescent Medicine published a Hastings Center proposal for new guidelines for treatment of intersex conditions that recommended delaying cosmetic procedures until a child could consent. In an invited critique, pediatric endocrinologist Erica Eugster wrote that a delay in cosmetic surgery would be inadvisable, not for any medical reason, but because it would impede a “family’s ability to accept and unconditionally love their child.” Eugster’s position—one that appears to be a prevalent, if largely unspoken motivation for corrective genital surgery—raises a central issue in the management of DSDs, which is the imperative to facilitate the attachment between child and parents. This paper examines this issue of fostering attachment in the “extraordinary” case of DSDs, figured historically as “a disorder like no other,” and asks how the treatment of DSD’s as “disorders like many others,” could provide a basis for fostering the parent-child relationship.

Many of you will be familiar with the study conducted by psychologist Suzanne Kessler a few years ago where college students were asked to imagine that they had been born with “clitoromegaly,” a condition defined as having a clitoris larger than one centimeter at birth. In response to the question as to whether they would have wanted their parents to sanction clitoral surgery if the condition were not life-threatening, an overwhelming ninety-three percent of the students reported that they would not have wanted their parents to agree to surgery. Kessler reports that, and I quote,

[w]omen predicted that having a large clitoris would not have had much of an impact on their peer relations and almost no impact on their relations with their parents...they were more likely to want surgery to reduce a large nose, large ears, or large breasts than surgery to reduce a large clitoris (Kessler 1998, 101).²

These findings, Kessler reflects, are not surprising given that the respondents characterized genital sensation and the capacity for orgasm as “very important to the average woman, and the size of the clitoris as being not even ‘somewhat important’” (Kessler 1998, 101-2).

¹ Ellen K. Feder teaches philosophy at American University. She contributed to the Hastings Center project, “Surgically Shaping Children,” and is a member of ISNA’s Medical Advisory Board and Speakers’ Bureau. Her book, *Family Bonds: Genealogies of Race and Gender*, is forthcoming from Oxford University Press, and she is currently working on a new project in ethics and the management of DSDs, *Fixing Sex*.

² This prediction is borne out by the fact that there is no published evidence suggesting any “hazards, biological or otherwise, of having a large clitoris.” While men with small penises have suffered some indignity, published studies have found that, “[c]ontrary to conventional wisdom, it is not inevitable that such [men] must ‘recognize that [they] are incomplete, physically defective and... must live apart.’” (Kipnis Diamond 1999, 181).

Men in the study were faced with a different dilemma, the one facing parents of boys with “micropenis,” a penis smaller than the putative standard of 2.5 centimeters at birth. Their question was whether to stay as male with a small penis, or to be assigned female. More than half rejected the prospect of gender reassignment. But, according to Kessler,

That percentage increases to almost all men if the surgery was described as reducing pleasurable sensitivity or orgasmic capability. Contrary to beliefs about male sexuality, the college men in this study did not think that having a micropenis would have had a major impact on their sexual relations, peer or parental relations, or self-esteem (Kessler 1998, 103).

In a separate study, Kessler and her team asked students to imagine that their child was born with ambiguous genitalia. Students in this study indicated they would make what Kessler describes as “more traditional choices” to consent to “corrective” or cosmetic surgery. Their rationales mirrored those of parents which can now be found on internet bulletin boards devoted to parenting children with intersex conditions: Students reported that they did not want their child to feel “different,” and believed that early surgery would be less traumatizing than later surgery (Kessler 1998, 103). Like parents over the last forty years who have been faced with these difficult decisions, students did not reflect on the somatic experience of the child, and with it, the possibility of lost sensation that so concerned the students in the first study.

Kessler’s paired studies confirm a kind of common sense that individuals, as individuals, are disinclined to compromise their erotic response for the sake of cosmetic enhancement. At the same time, parents, as parents, want “what is best for their child,” and the promise of a “normal life” figures prominently in that conception. The juxtaposition of the two studies raises the obvious, if nonetheless vexing, question: Why would parents consent to procedures on behalf of their children that they would refuse for themselves?

This question goes to heart of what I want to address today. The stark differences between hypothetical choices individuals would make for themselves and those they would make for their children, raises some important questions about what motivates corrective genital surgeries. I think that this study suggests that we need to consider with the utmost care the assumptions—centered mostly on the value of “normalcy” promised by surgeries—that lead well-intentioned physicians and parents to see surgery as the answer to challenges children may face as a result of having anatomies that differ from the norm. More than anything else, it appears that doctors are motivated to recommend and perform surgery because they believe sincerely that surgery is the best way to help families deal with atypical genitals.³

In 2004, *Archives of Pediatrics & Adolescent Medicine* published a proposal written by a Hastings Center working group for new guidelines for treatment of intersex conditions that recommended delaying any cosmetic procedure until a child is old

³ In making this claim, I should add a caveat, however. The work of Katrina Karkazis, who was unable to attend these meetings, points to how our understanding of the “good intentions” of doctors may be, if not suspect, then a good deal more complicated than we generally like to believe (see e.g. Karkazis 2005).

enough to consent (Frader, et al. 2004). In an invited critique, pediatric endocrinologist Erica Eugster responded that a delay in cosmetic surgery would be inadvisable, not for any medical reason, but because it would impede, as she put it, a “family’s ability to accept and unconditionally love their child” (Eugster 2004, 428).

Apart from the significant fact that Dr. Eugster has such a grim view of parents’ capacity to love their children—one that I think would be readily challenged by any number of physicians who work with children and families faced with other congenital disorders—it does raise a central issue in the management of DSDs, which is the imperative to facilitate the attachment between child and parents.

Against the perspective offered by Dr. Eugster I think we should consider the reflections offered by bioethicist Adrienne Asch. Asch affirms that “the most compelling argument for early surgery is that by altering the child’s appearance, the parent will more easily, naturally, and wholeheartedly invest in the child who looks more like what he envisioned” (Asch 2006, 240). But, Asch cautions:

what seems like a ‘fix’ to the parents and professionals may not feel like a ‘fix’ to the child whose body has been changed. By undertaking surgery before children can voice feelings about their bodies and their lives, the most loving parent can unwittingly undermine the child’s confidence that she is lovable and loved. It is confidence in that love and loveableness that provides the foundation for dealing with what life brings (Asch 2006, 229).

Asch’s point here gets to yet another conflict in the motivations of parents with respect to the decisions to consent to cosmetic surgery. As Alice Dreger writes in “What to Expect When You Have the Child You Weren’t Expecting,” the imperative to “do what’s best” for a child introduces another kind of conflict in parents’ own motivations. While, Dreger writes, “doing what’s best” clearly speaks to the idea of doing everything you can for your child [,] it also seems to negate the idea of accepting your child just as she is...it seems like opting for surgery might be rejecting your child” (Dreger 2006, 254), or at least, this is the message that may be unintentionally expressed.

Dreger offers to parents the following advice:

You have to keep in mind the decision you make about your child will affect how she thinks, not only about herself, but about your relationship with her, and how she thinks about the meaning of appearance. As you think about who you want your child to become, think about whether you’re modeling that outcome in your methods (Dreger 2006, 262).

Asch echoes this sentiment, affirming that the alternative to surgery is (and indeed cannot be) to “do nothing” (as one urologist put it during a meeting I attended). Asch writes that during

their child’s early life, the adults need not be passive and feel that their options during this time are limited to railing at societal cruelty. Whether or not the children ever choose surgery, how their parents and doctors behave toward them can give children tools for dealing with decisions about surgery and life (Asch 2006, 229).

I think that these two views—on the one hand, the idea that a parent must “do everything [she] can to improve” a child’s prospects, on the other, “accepting and

loving the child you have,” capture, in another way, precisely that conflict demonstrated in Kessler’s study of college students’ responses. It may very well be that the birth of a child with an abnormality of any sort can present problems for parents with respect to forming attachments to that child. But there are a variety of ways—nonsurgical ways—to address that challenge which can also be responsive to the kind of warning implicit in Asch’s counsel regarding how parents’ behavior can provide—or fail to provide—tools for living.

In making this claim, I should emphasize that I by no means intend to minimize the distress parents may feel on the birth of a child with atypical genitalia. We know that parents often experience shame and confusion. It is no doubt for this reason that in the “Consensus Statement” issued by the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology, the authors state: “It is generally felt that surgery that is carried out for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and parents.” “But” the authors immediately note, “The systematic evidence for this belief is lacking” (Hughes, et al. 2006, 4).

On the one hand, a surgeon’s efforts to normalize a child’s body to alleviate the shame that can result from the birth of a child with atypical genitalia may seem reasonable, but it is also in the surgeon’s power to normalize the child’s condition for the parents, that is, to explain that:

“These are things that happen, that there are in fact many children like this, and importantly, if you’d like to speak to some parents of children like yours—those who opted for early surgery and those who did not, I can help facilitate these connections for you.”

It is absolutely imperative that parents be provided with information, resources, and behavioral support. Without such support, parents may feel not only isolation but shame, which they could project onto their child.

As one social worker who helps families with craniofacial deformities puts it:

In my experience, if time is not dedicated to talking about the almost automatic development of shame, and how to reframe this experience, then it continues to flourish. This then leaves everyone at risk for creating a plan that is directed by shame-based decisions even when we least expect it. It’s important to avoid the impression that all negative reactions can somehow be stopped with surgery or other medical care. Shame can best be dealt with when we talk about it. The idea is to talk about it out loud or directly. Shame can be an isolating and terrifying thing, especially when it is either all we talk about or something we never talk about. (Consortium 2006, 27).

Surgery, in other words, cannot prevent feelings of inadequacy, of being “damaged goods,” but can send the powerful message that the body isn’t normal, that it is in need of a “fixing.” Psychologist Silvan Tomkins, whose work on affect has recently been so influential, describes how shame is “felt as an inner torment, a sickness of the soul” (Sedgwick and Frank 1995, 133). “There is no feeling more painful” (Biddle 1997, 227), Jennifer Biddle writes in a landmark feminist article on “Shame.” Shame is damaging, certainly, to one’s sense of self, and to one’s relationships, particularly with parents and extended family, but I think we do not

consider some of the unexpected ways that shame can also be debilitating to adults.

I want to suggest here that a good part of the motivation for corrective surgeries for children with atypical genitalia, and for the secrecy and silence that has surrounded them is owing to the historical treatment of intersex as a “disorder like no other.” I think that understanding DSD’s as “disorders like *many* others,” could provide some basis for fostering the parent-child relationship. I can say that I came to this proposal, in part, through my recent discussions with one family whose story—or by now, many stories—provide important lessons with respect to the damage that the conventional treatment of atypical genitalia can bring to a parent-child bond, and also how, in this case, the family’s coping with treatment of the underlying disorder, which requires the same skills and assistance as so many diseases or chronic conditions, can provide models for the assistance of families.

I first became acquainted with the woman I call “Ruby” when I was working on a project on feminist ethics and intersex several years ago (see Feder 2002). She has two grown daughters, both diagnosed with salt-losing CAH after being announced as boys and going into adrenal crisis soon after their birth. I have maintained contact with Ruby in the interim, and met her daughters a few years ago. I contacted them again, knowing that if there were experts in adult care, they would be the ones to ask. Not only did their ambiguous genitalia signal a serious life-threatening condition, their CAH—much like diabetes—would require lifetime management.

Theirs is a long and complicated story, so I certainly won’t attempt to recount it here. Rather, what I want to relate very briefly today is how talking to Ruby and her two daughters about the challenges they face today made me understand, in a different way, why it is so important that parents think carefully about the messages they send to their children through the medical decisions they make on their behalf when they are young. Ruby’s decisions to consent to corrective genital surgeries have had lasting effects for her daughters, who have had to understand that her decisions were guided above all by doctors’ understanding of “the right decision,” that is, to make the external genitalia of the girls conform to their sex of assignment. But if Ruby did not have the resources or information to make different decisions about cosmetic surgeries at that time and in that place, her fierce attention to her daughters’ health, her willingness to challenge doctors’ conclusions when her experience indicated she must, and most importantly, her instruction of her children in the ways of providing information and interacting with doctors over the long years of their treatment, expressed unambiguously her commitment to their health and well-being.

What she did, over and over again, was to support and guide them in describing their symptoms clearly to doctors, and from an early age, she insisted that doctors address their questions to, and seek information from, her daughters themselves. That is to say, she instructed her children in the ordinary business of seeking medical care—for a disease or disorder like any other. What makes her story so heartbreakng, as I have detailed elsewhere, is the incredible lack of support she received throughout these years, the secrecy she maintained—and maintains still, and the isolation she continues to experience.

Let me turn to the young women she still affectionately calls “my girls.” Growing up with the assistance of specialists who were keenly interested in developing their expertise in an area of medicine that was only beginning to be understood, her daughters learned not only to recognize their own symptoms—and to trust

their mother who had herself learned to anticipate crises—it came as a surprise when, as adults, they were unable to find doctors with any expertise in, much less genuine knowledge about, managing adult CAH. This is a state of affairs that has at least once threatened the life of the younger daughter, who, having taken herself to an emergency room with a syringe of solu-cortef she was unable to administer at home, found herself alone on a gurney in the hallway of an ER. The attending physician ignored the instructions on her medic-alert bracelet and refused to contact the endocrinologist on call. Just when he was preparing to release her, visibly ill and incoherent, her sister arrived—having received an emergency call from her mother—to explain in plain terms that if the physician did release her, she would not be alive the next day, and that she was calling their endocrinologist who would explain to him her need for immediate care.

This is but one of the many stories that this family has shared. It's such a complicated tale—of medical incompetence, in this case, but also of a family's banding together to take care of one another. But it's a challenging story because it emblemizes what Ruby's daughters, with the stalwart support of their mother, have had to do to in order to secure the medical attention most of us—and most people with chronic conditions that receive more attention and are more common, are privileged to take for granted.

I have tried in this short time to do too many things. I think that one of the important lessons I have learned in talking to parents, and in my own experience as a parent, is that almost nothing comes "naturally." Everything has to be learned. This was a point that the doctors at that Children's hospital embraced when it came to learning about this disorder that was taking the lives of children shortly after birth, and was perhaps why Ruby was "permitted" to participate with the doctors as a partner in her children's care, and in the gathering of information necessary to understand this frightening disorder. And if Ruby understood early on that learning was essential to the well-being of her daughters, she embraces the ways that she must continue to learn as her daughters struggle, into adulthood, to negotiate the difficulties of finding medical care and with the challenges—both medical and social—of having unusual anatomies and gay identities. What Ruby understands so well, and what Dr. Eugster seems not to have grasped, is, as Nietzsche puts it in the *Gay Science* (§ 334), "One must learn to love" (Nietzsche may seem like a surprising source of insight on love, but I find this passage quite remarkable, and I wanted especially to share it with this group today.)

One must learn to love.—This is what happens to us in music: First one has to *learn to hear* a figure and melody at all, to detect and distinguish it, to isolate it and delimit it as a separate life. Then it requires some exertion and good will to *tolerate* it in spite of its strangeness, to be patient with its appearance and expression, and kindhearted about its oddity. Finally there comes a moment when we are used to it, when we wait for it, when we sense that we should miss it if it were missing; and now it continues to compel and enchant us relentlessly until we have become its humble and enraptured lovers who desire nothing better from the world than it and only it.

But that is what happens to us not only in music. That is how we have *learned to love* all things that we now love. In the end we are always rewarded for our good will, our patience, fairmindedness, and gentleness with what is strange; gradually, it sheds its veil and turns out to be a new and indescribable beauty.

That is its thanks for our hospitality. Even those who love themselves have learned it in this way; for there is no other way. Love, too, has to be learned.

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Psychological Research in Disorders of Sex Development: Current and Future

Lih-Mei Liao



Criticisms of psychological research in DSD and related fields

To date:

- Leanings towards academic interests (or interests of academics)
- Over focus on androgen based gender theories
- Few clinical applications (does not benefit patients)



Applied psychosocial research

- Too few
- Variable clarity in question formulation
- Retrospective
- Cross sectional designs
- Small sample sizes
- Equivocal choice of measures
- Absence of control for socio-economic status
- Over interpretation/conclusions
- Absence of data from close others



Research with adults: some agendas

- Proof for care protocol (e.g. adult adjustment)?
- Proof against care protocol (e.g. gender identity)?
- Critical analysis
- Evaluation of treatment
- Identify problems for service planning (e.g. prevalence of various difficulties)



Methodological considerations

CAIS studies-

"generally satisfactory medical, surgical and psychosocial outcomes"

Wisniewski et al., 2000

'fear of devaluation'

'compromised womanhood'

"many of their emotional needs had gone unmet and their questions left unanswered" (p.98)

Alderson et al., 2004



Interpretation and conclusion

Morgan et al., 2005-
18 women with CAH spread over 9 years

No report on response rate (i.e. 18 out of how many CAH women seen in 9 years?)

Assessed for eating disorders? No rationale provided (Note that authors are specialists in eating disorders)

Self esteem above average? No explanation

What study adds (based on opportunistic case series): "Most women with CAH have good long term psychological outcome..." but-

(fine print) "We conclude that the women with CAH in our small studies are psychologically robust."

Research aims

- Identify perceptions of and responses to DSDs, and experiences of living with diagnosis
- Identify differences in patterns and factors that could account for differences (i.e. develop psychological explanations)
- Test out assumptions
- Evaluate interventions - what works for whom

Testing assumptions

Study with women with Turner syndrome:

- Height and satisfaction with height: poor predictor of psychological distress
- Delayed puberty: highly predictive of psychological distress
(Chadwick et al., in submission)
- Women most frequently troubled by work related stress
(Potts, 2005)

Making use of a range of methods

Keegan et al., 2003-
Psychological effects of hirsutism

- 1) Standardised questionnaires:
Near normative psychological functioning
High self esteem
- 2) Qualitative analysis
Extreme distress and preoccupation with hair growth

Choice of measures

Explicit decisions, e.g.

- Standardised measures
V
Non-standardised assessments of pertinent variables (e.g. perceptions of diagnosis, use of clinical services, sexual debuts)
- Global (e.g. overall mental health)
V
Specific (e.g. sexual function)

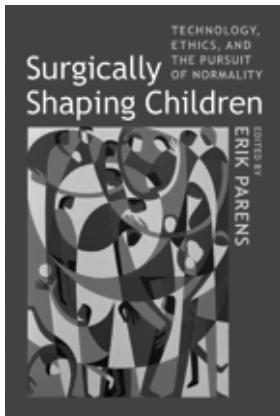
Raising standards for psychosocial research : Collaborative role of DSD pressure groups

- Formulate new questions
- Balanced focus between academic interests and clinical concerns
- Ethical guidelines (e.g. transparency, minimise intrusiveness)
- Discussion of methodology (e.g. as appropriate to small samples, or encourage multi-centre collaboration and longitudinal designs)
- Determine indices of "success" (e.g. presence of well being versus absence of mental illness, personal control over key aspects of life, quality of life..., not just gender outcomes)
- Development of measures (specifically for DSDs)
- Training in research
- Contribution to peer review processes



Surgically Shaping Children

Technology, Ethics, and the Pursuit of Normality



edited by Erik Parens

\$50.00 hardcover

0-8018-8305-9 (26 ctn qty)

2006 336 pp. 2 line drawings

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Description

At a time when medical technologies make it ever easier to enhance our minds and bodies, a debate has arisen about whether such efforts promote a process of "normalization," which makes it ever harder to tolerate the natural anatomical differences among us. The debate becomes especially complicated when it addresses the surgical alteration, or "shaping," of children. This volume explores the ethical and social issues raised by the recent proliferation of surgeries designed to make children born with physical differences look more normal.

Using three cases—surgeries to eliminate craniofacial abnormalities such as cleft lip and palate, surgeries to correct ambiguous genitalia, and surgeries to lengthen the limbs of children born with dwarfism—the contributors consider the tensions parents experience when making such life-altering decisions on behalf of or with their children.

The essays in this volume offer in-depth examinations of the significance and limits of surgical alteration through personal narratives, theoretical reflections, and concrete suggestions about how to improve the decision-making process. Written from the perspectives of affected children and their parents, health care providers, and leading scholars in philosophy, sociology, history, law, and medicine, this collection provides an integrated and comprehensive foundation from which to consider a complex and controversial issue. It takes the reader on a journey from reflections on the particulars of current medical practices to reflections on one of the deepest and most complex of human desires: the desire for normality.

Contributors

Priscilla Alderson, Adrienne Asch, Cassandra Aspinall, Alice Domurat Dreger, James C. Edwards, Todd C. Edwards, Ellen K. Feder, Arthur W. Frank, Lisa Abelow Hedley, Eva Fedder Kittay, Hilde Lindemann, Jeffery L. Marsh, Paul Steven Miller, Sherri G. Morris, Wendy E. Mouradian, Donald L. Patrick , Nichola Rumsey , Emily Sullivan Sanford , Tari D. Topolski

For further information and news in Bioethics, please visit [the Hastings Center](#) online.

Reviews

"A truly striking collection of voices that are largely absent from ordinary bioethics texts, and one of the finest anthologies I have read in years."—Carl Elliott, University of Minnesota Center for Bioethics, author of *Better than Well: American Medicine Meets the American Dream*

"*Surgically Shaping Children* is a must-read for anyone concerned about the cultural denial of differences in human embodiment and the desire for the 'surgical fix.' In a style that is the trademark of any conversation initiated by the Hastings Center, the contributors—philosophers, physicians, patients, and parents—tackle all the difficult questions without opting for easy answers. This is a book that will make you think."—Kathy Davis, Institute for History and Culture, University of Utrecht, The Netherlands, and author of *Reshaping the Female Body*

"In this thoughtful book, patients, parents, doctors, and distinguished philosophers speak to difficult questions of disability, technology, identity, and values."—Peter D. Kramer, Brown University, author of *Against Depression* and *Listening to Prozac*

"As medicine gains ever greater skill at 'correcting' the physical deficiencies of children, we are also acquiring the power to alter personal identity and change the meaning of normality. In *Surgically Shaping Children*, Erik Parens has collected a wonderful range of provocative and thoughtful essays that, while providing no easy answers, raise crucially important questions about when, why, and how we should 'fix' the appearance of our children. Doctors, patients, ethicists, and parents will all be enriched by its wisdom and empowered by its intelligent consideration of these thorny issues."—Stephen S. Hall, author of *Merchants of Immortality*

"It is extraordinary when a book manages to be both informative and critical. *Surgically Shaping Children* is an important book for parents who confront the reality of their children's appearing different from what they and society imagined. It is also a book for all readers interested in how norms of appearance affect the way we imagine ourselves and others and, equally important, how we employ medicine to rectify such differences."—Sander L. Gilman, Emory University

"This fascinating and disturbing collection explores the difficult question of when and how surgery might be used for children born with disabilities and other anomalies. It

speaks not just to every parent's desire to help his or her child, but also to concerns about the contested borders of health, normality, and difference, in an age when our biomedical powers may sometimes exceed our wisdom."—Tom Shakespeare,
University of Newcastle, UK

"It was a joy reading this brilliant collection of essays. This carefully conceived and well-written book will be welcomed by health care professionals and medical ethicists, but they are by no means its only potential audience. The challenging issues it raises would make it an excellent text for seminar courses on ethics and philosophy. But in my opinion its greatest and most lasting value will be as a resource for parents and other family members of affected patients."—Bruce J. Beckwith, Loma Linda University

Author Information

Erik Parens is a senior research scholar at The Hastings Center, adjunct professor in the Science, Technology, and Society Program at Vassar College, and the coeditor of *Wrestling with Behavioral Genetics: Science, Ethics, and Public Conversation* (Johns Hopkins Univ. Press, 2005). He is also editor of *Enhancing Human Traits: Ethical and Social Implications* (Georgetown Univ. Press, 1998) and *Prenatal Testing and Disability Rights* (Georgetown Univ. Press, 2000).



Qantas Television Awards 2004 Winner of New Zealand's prestigious Qantas Best Television Documentary Award

MANI'S STORY

Yellow For Hermaphrodites



**Intersex Society
of North America**

MANI'S STORY



Since filming "Hermaphrodites Speak!" (see info below) in 1995, Mani Bruce Mitchell has been on a remarkable healing journey. In this documentary Mani, an adult with an intersex condition, tells her poignant story of growing up in rural New Zealand. Subjected to genital surgeries at an early age, Mani shares her life – the difficult times when she considered suicide, and her path to healing, reconciliation, and service. She reaches out to other intersex individuals, giving viewers a chance to hear their stories as well.

Mani comments: "I agreed to do this [film] as a queer identified intersex person who believes with passion about creating change. Creating a world where it will be safe...that respects and enjoys ALL who are different. An easy process...

it was not. It became...the most challenging, powerful, unusual, and creative task I have ever done outside of a therapeutic setting." By making the choice to own her identity and her sense of herself as neither male nor female, Mani takes joy in life, inspiring family and friends with her strength.



"What I went through could have completely destroyed me, and yet, I was able to soar above it."
- Mani Mitchell



Time: 45 minutes
Written & Produced by John Keir
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Hermaphrodites Speak!



Imagine growing up different, not quite knowing why, feeling you were the only person in the world like you. Meet Angela, David, Heidi, Tom, Mani, Cheryl, Max and Hida sharing their stories of growing up intersexed & their joy in meeting each other. A milestone film in DSD awareness & public education.

Time: 30 minutes.
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Documentaries are Available for Purchase
For more information, Visit: www.isna.org



Since filming "Hermaphrodites Speak!" (see info below) in 1995, Mani Bruce Mitchell has been on a remarkable healing journey. In this documentary Mani, an adult with an intersex condition, tells her poignant story of growing up in rural New Zealand. Subjected to genital surgeries at an early age, Mani shares her life – the difficult times when she considered suicide, and her path to healing, reconciliation, and service. She reaches out to other intersex individuals, giving viewers a chance to hear their stories as well.

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Written & Produced by John Keir
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Imagine growing up different, not quite knowing why, feeling you were the only person in the world like you. Meet Angela, David, Heidi, Tom, Mani, Cheryl, Max and Hida sharing their stories of growing up intersexed & their joy in meeting each other. A milestone film in DSD awareness & public education.

Time: 30 minutes.
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Total Patient Care

the child with
an intersex
condition

Intersex is the general term used to denote a variety of conditions in which a person is born with mixed sex anatomy. It is important to note that intersex conditions sometime involve “ambiguous genitalia” but that intersexuality is not always evident from an external examination.

Intersexuality may be caused by congenital adrenal hyperplasia, mixed gonadal dysgenesis, androgen insensitivity syndrome (complete or partial), 5-alpha reductase deficiency, and many other factors.

The traditional standard of care held that normal gender identity could be assigned through genital plastic surgery and by limiting information given to parent and patient. Many patients were subjected to multiple surgeries.

Over the last ten years, new evidence and advances in medical ethics have led to the need for revision in the standard of care for the treatment of intersex. This film outlines the problems with many current practices and provides guidelines for a new standard of care that is more advanced scientifically and ethically.

We gathered a group of experts, including medical professionals, an adult with intersex, and a parent to talk about these issues.



Total Patient Care



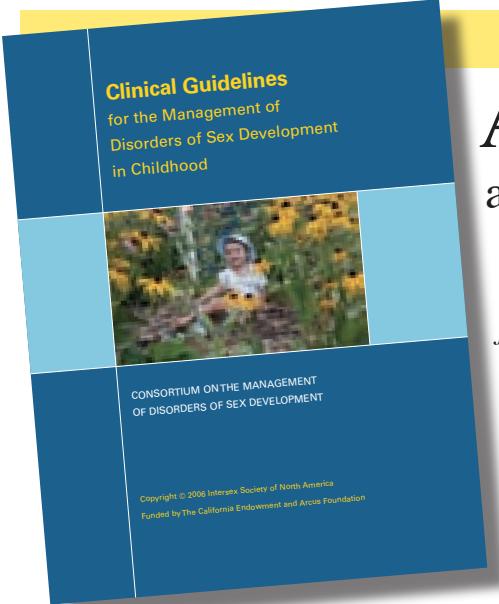
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now available from **Intersex Society of North America**

Announcing Two New Books about Disorders of Sex Development in Childhood



About the books

Clinical Guidelines for the Management of Disorders of Sex Development in Childhood is a handbook for health care professionals who provide care to pediatric patients with Disorders of Sex Development. It is also a valuable resource for health professional students, educators, parents of children with DSDs, and adults with DSDs.

The companion *Handbook for Parents* is a resource that physicians can give parents to aid in understanding and adjusting to the child's needs.

About the authors

The *Clinical Guidelines for the Management of Disorders of Sex Development in Childhood* and *Handbook for Parents* have been produced by a consortium consisting mainly of: (1) clinical specialists with experience helping patients with DSDs; (2) adults with DSDs; and (3) family members (especially parents) of children with DSDs.

From the experts

"As a social worker who has followed this population for 17 years, it is gratifying to finally have a tangible tool to use with and on behalf of patients and families."

— Barbara Neilson, MSW, RSW
Urology Social Worker
The Hospital for Sick Children

"The handbooks on Disorders of Sex Development are a real revolution. They focus on the most important person: the child."

— Eric Vilain MD, PhD
UCLA Department of Urology

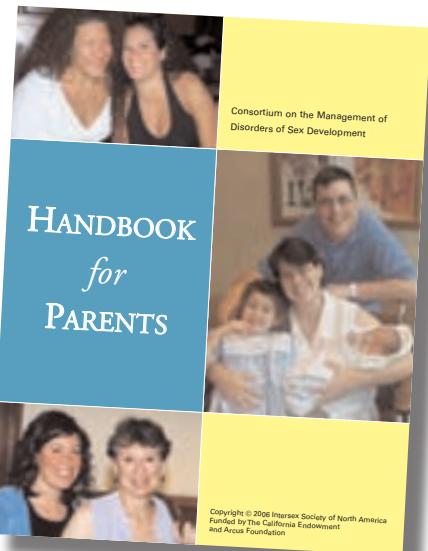
Clinical Guidelines for the Management of Disorders of Sex Development in Childhood and **Handbook for Parents**

Consortium on the Management of
Disorders of Sex Development

The *Clinical Guidelines* handbook was developed to assist health care professionals in the providing diagnosis, treatment, education, and support to children born with disorders of sex development (DSDs) and to their families.

While debates about the best way to care for patients with DSDs continue, this handbook offers individuals and institutions a model aimed at minimizing the potential for harm to patients and their families. It is therefore also designed to reduce the potential for liability and to improve patient follow-up.

These guidelines begin with the commonly-held assumption that the goal of DSD treatment is the long-term physical, psychological, and sexual well-being of the patient. This approach is therefore termed "patient-centered." (63 pages)



Handbook for Parents addresses the many questions and emotions parents have upon learning that their child has some kind of disorder of sex development (called a DSD for short).

Written in plain language, the book offers background information on child development, how to talk with the child and others about his or her DSD, and reproducible information pages to give family and others important in the child's world.

Common questions are answered, and the handbook provides guidance for preparing parents and the child for doctor's appointments and the value of keeping a journal.



Especially insightful are the generously shared thoughts and experiences of other parents and of adults with DSDs.

(133 pages)

More from the experts

"Brings a calm, reasoned, and honest approach to an emotional and upsetting condition—provides support and education for parents from professionals and other parents."

— Kaye Fichman, M.D.
Kaiser Permanente Medical Group



CLINICAL GUIDELINES Contents

1. Introduction

- Purpose of this Document
- Definition of DSDs
- Definition of Patient-Centered Care for DSDs
- Methodology

2. Treatment Guidelines

- Multidisciplinary Team Approach
- Team Composition
- Team Objectives
- Team Leader, Team Coordinator, and Team Liaison
- Treatment Protocols for the Management of Newborns with DSDs
- Treatment Protocols for the Management of Children Diagnosed with DSDs after the Newborn Period

3. Background and Elaboration

- Gender Assignment
- Psychosocial Support
- Timing of Surgeries
- Timing of Hormonal Therapy
- Reducing Stigma Related to Medical Care
- Telling the Truth
- The Importance of Sexual Well-Being

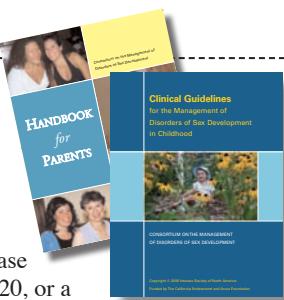
4. Scripts for Talking with Parents

5. Core References and Resources

- Videos and Animations
- Books
- Special Journal Issues (Devoted to DSD Management)
- Key Articles
- Support Group Information

References

Index



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You can purchase one book for \$20, or a set of both the *Clinical Guidelines* and the *Handbook for Parents* for \$30, plus shipping and sales tax as applicable.

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1. Welcome to Parents

- You Are Not Alone!
- What Are DSDs?
- What Causes DSDs?
- Acceptance Takes Time
- Speaking About Shame, to Try to Get Beyond It

- Helping Yourself to Help Your Child
- Secrecy=Shame, Honesty=Acceptance
- Take-Home Messages of This Chapter

2. Your Child's Development, and How to Talk with Your Child

- About the Language Used Here
- Key Background Points
- Ages 12-36 Months
- Ages 3-5 Years (Pre-Schoolers)
- Ages 6-11 Years
- Puberty
- Adolescence (11-18)
- Your Life Together
- Take-Home Messages of This Chapter

3. How to Talk with Others

- Introduction
- What to Tell Your Friends and Family in General
- If Your Newborn's Gender Assignment is Delayed
- What to Tell People Who May Think DSDs are Sinful
- Tips on Interacting with Teachers and Daycare Providers
- Talking with Your Child's Medical Care Providers
- Take-Home Messages of This Chapter

4. Answers to Common Questions

5. Helpful Handouts

- About Disorders of Sex Development (DSDs)
- How Genital Development Happens
- Things to Do and Things to Avoid
- Preparing for a Medical Appointment
- Record-Keeping and Journaling

6. Thoughts from Fellow Parents and from Adults with DSDs

- Introduction to This Material
- Letters from Fellow Parents
- Memories and Thoughts from Adults with DSDs

7. Other Resources (Where to Learn More)

- Websites
- Support Groups and Diagnosis-Specific Information
- Finding a Child Psychologist
- Books Devoted to DSD Issues
- Videos and Television Broadcasts
- Journal and Magazine Articles
- General Parenting
- Sex Education for Your Child

A. Key to Photographs

Glossary (Explanation of Terms)

References

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