Affirming Care for People with Intersex Traits:
Everything You Ever Wanted to Know, But Were Afraid to Ask

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Goals

By the end of this hour, you will be able to:

▪ Appreciate the diversity of intersex traits, and the conditions associated with them

▪ Describe the traditional approach to people with intersex traits and its impact on health

▪ Implement an affirming approach to physical and behavioral health care for people with intersex traits
What are intersex traits?

Group of *congenital* variations relative to endosex traits

- Sex chromosomes, hormones, and/or internal or external genitalia
- May also see variations in secondary sex traits
- Included among sexual and gender diverse/minority populations
- Present at any time across the lifespan
About Language…

That is complicated

▪ Hermaphroditism

▪ Intersex/uality

▪ Differences/Disorders of Sex Development

▪ Intersex (traits/conditions), DSD

▪ Endosex

ADVANCING EXCELLENCE IN SEXUAL AND GENDER MINORITY HEALTH
Why Learn About Intersex?

People with intersex traits...

- Are common (1 in 100 - 2000)
- Benefit from quality medical care
- May receive care in SGM health settings
- Are rarely intentionally included in SGM health
Review of Sex Development
Sex Chromosomes

- Eggs: X, XX, XO
- Sperm: X, Y, O, XX, YY
- Sex chromosomes initiate gonad development
- Gonads produce hormones and gametes
Prenatal Development

Undifferentiated gonads

Wolffian Duct

Müllerian Duct

5 weeks gestation
Prenatal Development: Gonads

- **Testes**
  - Y chromosome
  - SRY gene

- **Undifferentiated**
  - 

- **Ovaries**
  - X chromosome
  - WNT4 gene
Prenatal Development: Internal Genitalia

Wolffian duct

Müllerian duct

MIF/AMH

testosterone

no testosterone
Prenatal Development: External Genitalia

- Glans
- Urogenital folds
- Labioscrotal swelling
- Urogenital sinus
Prenatal Development: External Genitalia

- Penis and scrotum: DHT
- Labia and vagina: No DHT, estrogen?
<table>
<thead>
<tr>
<th>Endosex female</th>
<th>Endosex male</th>
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<td><strong>Karyotype</strong></td>
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<td><strong>Hormones</strong></td>
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<td><strong>Internal genitalia</strong></td>
<td>Ovaries</td>
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<td>(Gonads, urogenital sinus, Wolffian &amp; Mullerian ducts)</td>
<td>Fallopian tubes</td>
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<td>Uterus &amp; cervix</td>
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<td><strong>Secondary sex traits</strong></td>
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<td>Pubic &amp; axillary hair</td>
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Overview of Intersex Traits by Developmental Process
Classification of Intersex Variations

- Karyotype
- Gonads
- Hormonal milieu
- Genitalia
Classification of Intersex Variations

- Karyotype
  - Klinefelter Syndrome (XXY)
  - Turner Syndrome (XO)
  - Mosaicism (XX/XY)

- Gonads

- Hormonal milieu

- Genitalia
Classification of Intersex Variations

- Karyotype
- Gonads
  - Gonadal Dysgenesis (Swyer’s Syndrome)
  - Ovotesticular Syndrome
- Hormonal milieu
- Genitalia
Classification of Intersex Variations

- Karyotype
- Gonads
- Hormonal milieu
- Genitalia
  - Mülllerian agenesis (MRKH)
  - Hypospadias
  - Penile agenesis or microphallus
Classification of Intersex Variations

- Karyotype
- Gonads
- Hormonal milieu
  - Androgen Insensitivity Syndrome (AIS)
  - Congenital Adrenal Hyperplasia (CAH)
  - 5-alpha Reductase Deficiency (5-ARD)
- Genitalia
Classification of Intersex Variations

- Karyotype

- Gonads

- Hormonal milieu
  - Androgen Insensitivity Syndrome (AIS)
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- Genitalia
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<th>XY/XX</th>
<th>Endosex male</th>
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<td><strong>Internal genitalia</strong></td>
<td>Ovaries</td>
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<td>Tetes</td>
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<td>Fallopian tubes</td>
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<td>Epididymis</td>
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<td>Uterus &amp; cervix</td>
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<td>ductus deferens</td>
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<td>Upper vagina</td>
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<td>seminal vesicle</td>
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<td>Clitoris</td>
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<td>Penis</td>
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<td>Scrotum</td>
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<td><strong>Secondary sex traits</strong></td>
<td>Breast development</td>
<td>Gynecomastia</td>
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<td>Voice change</td>
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<td>Menstruation</td>
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<td>Genital enlargement</td>
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<td>Pubic &amp; axillary hair</td>
<td>Primary amenorrhea</td>
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<td>Pubic, axillary, facial hair</td>
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<td><strong>Brain</strong></td>
<td>Girl</td>
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<td>Boy</td>
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<td></td>
<td>Endosex female</td>
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<td><strong>Hormones</strong></td>
<td>Estrogens</td>
<td>Androgens -&gt; Estrogens</td>
<td>Androgens</td>
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<tr>
<td><strong>Internal genitalia</strong></td>
<td>Ovaries, Fallopian tubes, Uterus &amp; cervix, Upper vagina</td>
<td>Testes, No/limited seminal structures, Shorter vagina, No uterus or cervix</td>
<td>Testes, Epididymis, ductus deferens, seminal vesicle, ejaculatory duct, Prostate</td>
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<tr>
<td><strong>External genitalia</strong></td>
<td>Clitoris, Vulva/Labia</td>
<td>Endosex vulva, Glans and labioscrotal variations, Urethral variations</td>
<td>Penis, Scrotum</td>
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<td><strong>Secondary sex traits</strong></td>
<td>Breast development, Menstruation, Pubic &amp; axillary hair</td>
<td>Breast development, Variable pubic, facial axillary hair</td>
<td>Voice change, Genital enlargement, Pubic, axillary, facial hair</td>
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<tr>
<td><strong>Brain</strong></td>
<td>Girl</td>
<td>Varies (often girl)</td>
<td>Boy</td>
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Traditional Medical Approaches to Intersex Traits
History of Intersex

- Before the 1960s
- From the 1960s to the 1990s
- From the 1990s to the Present
History of Intersex

- Prior to the 1960s
  - Limited diagnostic tools
  - Limited surgical options

- From the 1960s to the 1990s
  - Developments in genital surgery
  - Gender theory
  - “Concealment” model of care
Gender Theory

- Biological sex is not the same as social gender
- Biological sex informs social gender
- "Normal" gender aligns genitals, role, sexuality
- Nurture can override nature
  - Before the “window” closes at 4 years old
  - No ambivalence in rearing
Gender Theory

In practice:

- Gender assignment influenced by surgical technique and capacity for heterosexual intercourse
- Diagnostic and surgical information withheld from patients, and many parents
History of Intersex

- From the 1990s to the Present
  - Founding of activism and support groups
  - Surgical complications
  - Culture of shame, secrecy, and stigma

- 2006 Consensus Statement
  - Multidisciplinary teams, more conservative surgical management
  - Disclosure of diagnoses
The Clinical Needs of People with Intersex Traits
Legacy of the Old Model

Disclosure of Diagnoses:

- Information routinely withheld from patients and families up through early 2000s
- Propagated shame, stigma, and isolation
- Patients continue to grapple with these
Legacy of the Old Model

Interventions Continue *Today*:

- Gonadectomy
- Clitoral reduction
- Vaginoplasty
- Hypospadias surgery
- Hormonal interventions
Legacy of the Old Model

Interventions Continue *Today*:

- In infancy
- Before ability to assent
- With insufficient psychosocial support
- To address distress
Parental Stress

Western endo/cis/hetero-normativity → Intolerance of Uncertainty → “Do something!” → Depression and Anxiety

Distress

- False dichotomy: “Do surgery or do nothing.”
- No compelling evidence that:
  - Distress is unmanageable for parents
  - Genital surgery reduces psychosocial distress relative to no surgery
Legacy of the Old Model

Physical risks:
- Scarring and chronic pain
- Urinary and sexual dysfunction
- Sterilization
- Lifelong hormone replacement therapy
- Complications requiring multiple follow-up surgeries

Psychological risks:
- Depression, PTSD, suicidal thoughts
- Shame, isolation, and inadequacy
- Gender incongruence

Population level risks:
- *Negative health outcomes due to negative experiences accessing care*
Groups Calling for Delay

- US Bureau of Public Affairs for State Dept
- State legislatures
- German and Swiss ethics councils
- Australia, Chile, Argentina, Malta governments
- World Health Organization
- Several UN organizations, Special Rapporteur on Torture
- Amnesty International, Human Rights Watch
- Physicians for Human Rights
- GLMA Health Professionals Advancing LGBT Equality
- American Medical Student Association
- American Academy of Family Physicians
- Indian, Colombian, Kenyan courts
What do Patients Need?

A different model of care, that:

- Affirms sexual and gender diversity
- Celebrates strength of patients and families
- Repairs trauma
## Affirming Care

<table>
<thead>
<tr>
<th>Traditional Model</th>
<th>Affirming Model</th>
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<tbody>
<tr>
<td>Sex defined by single factor</td>
<td>Sex defined by balance of factors</td>
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<tr>
<td>Sex is binary</td>
<td>Sex exists on a spectrum</td>
</tr>
<tr>
<td>Intersex is a disorder</td>
<td>Intersex is a natural human variation</td>
</tr>
<tr>
<td>Gender is binary &amp; predictable</td>
<td>Gender is flexible &amp; exploratory</td>
</tr>
<tr>
<td>Genitals must be “normal”</td>
<td>Genital diversity can be affirmed</td>
</tr>
<tr>
<td>Children will be ostracized and distressed</td>
<td>Children can be prepared and supported</td>
</tr>
<tr>
<td>Only heterosexual, penovaginal intercourse is normal</td>
<td>A wide range of sexual activity is normal and enjoyable</td>
</tr>
</tbody>
</table>
What Do Patients Need?

Physical Necessity:

- Cortisol replacement
- Relief of obstructions
- Address malignancies
- Sex hormone replacement for hypogonadism
- Evaluation for a specific DSD
- Education on anatomy, medical history, and medical records
What do Patients Need?

Physical Considerations:

- Gonadectomy for cancer risk
- Puberty blockers for discordant development
- Relocation of urethral meatus
- Vaginal dilation
- Genital surgery for appearance or sexual function
- Fertility preservation
What Do Patients Need?

Psychosocial Necessity:

- Professional *and* peer support
- Sex and Gender 101
- Flexible sex assignment
- Education on sexual health and wellbeing
- Discussions about family formation
Clinical Management

Psychosocial Themes

- Intersections with other identities
- Parents:
  - Adaptation and normalizing narratives
  - Decision-making in uncertainty
- Youth:
  - Navigating difference
  - Gender, sexuality, and global identity development
Clinical Management

Psychosocial Themes

- Adults: Shame, secrecy, isolation
  - Withholding of information
- Trauma
  - Medical photography and exams
  - Nonconsensual surgery
- Minority stress? Stigma?
Intersex Inclusion

- Consider documents and history-taking: not everyone assigned female at birth has a uterus, or produces estrogen

- Ask patients what they understand about their bodies

- Minimize intrusive examinations and questions

- Mirror language chosen by the patient, including names and pronouns

- Medical language may be associated with trauma
Intersex Inclusion

- Promote patient-driven goals regarding gender-affirming care
- Utilize multidisciplinary teams to optimize care, including mental health
- Careful, comprehensive informed consent
- Ongoing education of families and patients
- Refer to support groups
Case Discussion

- Natalia is a 16 year-old assigned female with partial androgen insensitivity syndrome who presents to discuss vaginoplasty.

- At birth, Natalia had mid-range glans length, partial labioscrotal fusion, and bilateral inguinal testes.

- Natalia’s testes were removed at age 2 due to concern for malignancy risk, and laparoscopy confirmed lack of uterus.

- Natalia reports considering surgery “so I can have sex.”

- What else do you want to know?
Natalia

- Mother and grandparents “want me to be normal”
- Understanding of surgery: “I have no idea”
- Sexuality: “No one will be interested in me.”
  - Romantically attracted to multiple genders
  - No fantasies, masturbation, or sexual partners
- Gender: “I guess female?” Androgynous, femme-leaning expression
- Physical exam: narrow vaginal opening, separate urethral opening
- How do you talk with Natalia about surgery? What else does Natalia need?
Natalia

Traditional model of intersex care:

- Surgeon-led
- Intolerant of uncertainty in decision-making
- Recommend “normalizing” vaginoplasty
- Discuss options for neovagina
- Obtain informed consent
- Schedule patient, often “before college”

Affirming model of intersex care:

- Psychosocial or medicine-led
- Understand and offer education on spectrum of sexual and gender identities and behaviors
- Understand context of decision
- Allow time for processing of information and consent
- Offer dilation as alternative
Natalia: 6 month f/u

Now Natalia has a clearer understanding of identity:

- Gender identity: “nonbinary/femme,” they/them/theirs or she/her/hers
  - “I don’t need a vagina to feel like myself.”

- Sexuality:
  - Panromantic, reluctant to label sexual orientation
  - Likely interested in peno-vaginal intercourse, “but there are other ways.”

- Researched different options for dilation, vaginoplasty, post-op dilation

- “All together, I probably want the surgery, but I’m still trying to figure out if it’s to make me feel good, or to make it easier for me to date. I think I’ll try dilation first.”
Resources

Peer support groups

- AIS-DSD Support Group (AISDSD.org)
- OII - USA, UK, Australia

Legal support and advocacy

- Inter/Act (interactadvocates.org)

Intersex stories

- Inter/Act Youth (interactyouth.org)
- "Born Both: An Intersex Life," Hida Viloria
- "XOXY," Kimberly Zeiselman
- “Contesting Intersex: The Dubious Diagnosis,” Georgiann Davis, PhD
- “Gender Revolution,” Katie Couric and National Geographic

Medical Education Resources

- Diversity 3.0 Learning Series (www.aamc.org/initiatives/diversity)
References


