Understanding Intersex Conditions

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Welcome

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Goals for Today

- Review historical and current practices and thinking about infants born with intersex conditions
- Increase understanding of psychosocial impacts on patients and families
- Differentiate gender identity from chromosomal or anatomical sex
- Increase understanding of patient-centered and multidisciplinary care models
Meet: Kayla
- Age 4, African American
- Born with external female genitals, XX chromosomes
- Raised as female
- Begins to assert stereotypic male preferences

Thoughts on Kayla??

Meet: Jackie
- Age 16, Latina
- Born with CAH
- Wants surgery for enlarged clitoris
- Family has not managed medications well due to resource issues

Thoughts on Jackie??
Meet: Tag

- Age 15, South Asian
- Born with Y chromosome and penis over 1 inch
- Penis was “reconstructed” shortly after birth
- Vineet raised as male
- Tag begins to say “I’m both”
- Parents did not tell Tag about intersex condition

Thoughts on Tag??

Intersex

- “Intersex” is a general term used for a variety of conditions in which a person is born with a reproductive or sexual anatomy that doesn’t seem to fit the typical definitions of female or male
Intersex vs Disorders of Sexual Difference (DSD)

- Term “Intersex” more embraced by advocacy groups as more than a condition (historical language)
- DSD embraced by medical community to discuss variance in chromosome/anatomy
- Mixed views on terms as pathologizing/↑ stigma
- No set position on language, varies among communities and providers

Intersex Conditions and Prevalence

- Congenital Adrenal Hyperplasia (CAH) — One in 13,000 births
- Testosterone Biosynthetic Defects — One in 13,000 births
- Androgen Insensitivity Syndrome (AIS) — One in 13,000 births
- Gonadal Dysgenesis — One in 150,000 births
- Micropenis — No estimate available
- Klinefelter Syndrome — One in 1,000 births
- Turner Syndrome — No estimate available
Prevalence (cont.)

- 1 in 100 bodies different from standard
- 1-2 in 1000 receiving surgery to “normalize” genital appearance
- **1.7 percent** of human births might be intersex, including variations that may not become apparent until, for example, puberty, or until attempting to conceive — NOTE: Late onset CAH may be an outlier in the numbers that offer 1.7% of population (Blackless, Fausto-Sterling et al.)

Intersex (cont.)

- Anatomy doesn’t always reveal variations/conditions at birth (puberty, infertility or autopsy after death)
- Some people live and die with intersex anatomy without anyone (including themselves) ever knowing
- There is a wide spectrum of sex categories — in order to simplify social interactions, express what we know and feel, and maintain order, sex categories are presented as male, female and sometimes intersex
Anatomy and Physiology

- External genitals that cannot be easily classified as male or female (historical: ambiguous genitalia)
- Incomplete or unusual development of the internal reproductive organs
- Inconsistency between the external genitals and the internal reproductive organs
- Abnormalities of the sex chromosomes
- Abnormal development of the testes or ovaries
- Over- or under-production of sex-related hormones
- Inability of the body to respond normally to sex-related hormones

Intersex (cont.)

- Born appearing to be female on the outside, but having mostly male-typical anatomy on the inside
- May be born with genitals that seem to be in-between the usual male and female types — for example, a baby may be born with a noticeably large clitoris, or lacking a vaginal opening, or be born with a notably small penis, or with a scrotum that is divided so that it has formed more like labia
- Person may be born with mosaic genetics, so that some cells have XX chromosomes and some of them have XY
Intersex (cont.)

Some intersex conditions signal underlying metabolic concerns, a person who thinks they might be intersex should seek a diagnosis and find out if they need medical intervention.

Historic Practice: Concealment

- Abnormality — must be normalized
- Can make a child male or female by creating convincing genitals
- Focus on genitals over genes, brain, hormones, prenatal life
- Untreated can lead to poor mental health outcomes (depression, suicidal ideation, etc.)
Concealment (cont.)

- Viewed as a psychosocial emergency
- Surgery is recommended over parental support/counseling
- Parent bond with child can be impacted
- Suggested child is told very little or never told even when older

Historic: Determination of Gender Assignment Based on Sex

- If the child has a Y chromosome and an adequate or "reconstructable" penis, the child will be assigned a male gender. (Newborns must have penises of 1 inch or larger if they are to be assigned the male gender.)
- If the child has a Y chromosome and an inadequate or " unreconstructable " penis according to doctors, the child will be assigned a female gender and surgically "reconstructed."
- If the child has no Y chromosome, it will be assigned the female gender. The genitals will be surgically altered to look more like what doctors think female genitals should look like. This may include clitoral reduction surgeries and construction of a "vagina" (a hole).
Cultural Considerations

- In Western nations, there is increasing consensus about ethical approaches to clinical intersex management. At the same time, as Western-trained physicians increasingly encounter intersex patients in other parts of the world, new ethical tensions arise. Which cultural values are fair parameters for gender assignment decision-making, particularly in cultural milieus where there is social and economic inequality between the sexes? How can physicians uphold universal bioethical principles, while remaining culturally sensitive? Physicians have a primary commitment to patient beneficence and universal human rights, requiring physicians to promote concordance between the child’s assigned gender and his/her likely future gender identity. Ultimately, the potential patient distress posed by gender dysphoria fundamentally outweighs the influence of local cultural factors such as economics, gender politics, and homophobia.


Cultural Considerations (cont.)

- Culture values one sex over the other in terms of human value
- Worry of response from community or family
- No “one face” to a cultural group, still must dialogue with family
- Caregivers may not share equal voice in decision making
- Clash with Western value of image, correction, intervention
- How to determine if issue is beyond only cultural position and reflective of other issues (mental health, etc.)
Intersex Definition Expanded

Intersex is a socially constructed category that reflects real biological variation.

Distinction: Gender Identity — Prevalence

Individuals with transgender and gender nonconforming (TG/GNC) identities continue to become more visible, along with increased understanding of their unique needs. At least 0.6% of the population, or 1.7 million Americans, TG/GNC people present across the spectrum of age, sexual orientation, race, geographic area, culture, socioeconomic status, and health status.

**Assumptions on Gender**

- There are only two genders
- You can tell a person’s gender when (or before) they are born
- Gender determines what we can/cannot do
- Thinking or stating a person is “born in the wrong body”; “born a boy and became a girl”; “girl with a boy’s body”
- All people who are gender expansive are unhappy or have “dysphoria”
- All people want to change the body they have medically
- Cisgender is “normal”
“Transgender” and “gender nonconforming” are umbrella terms that refer to someone whose gender identity (i.e., sense of male-ness or female-ness) does not align with their assigned birth sex based on genitals and chromosomes. TG/GNC individuals may identify specifically as “transgender” or simply as male or female, without the transgender label. In addition, some TG/GNC people are non-binary, meaning they do not identify with a male or female label.
MYTH: Gender affirming care PROMOTES gender transition or transgender identity

FACT: rather listens to child and carefully considers all variables
Gender-Affirmative Care

- Gender variations are not disorders and vary across cultures
- Gender is the complex interplay of biology, psychology, and culture/society
- Acceptance by family/others significantly improves mental health outcomes
- Multidisciplinary team approach (child/youth, family, gender specialist, mental health as indicated, medicine as indicated, legal, school, clergy, etc.)

Gender-Affirmative Care

- Listens to child
- Opposed to conversion tactics/therapies
- Dysphoria varies by type and level of severity
- Wishes vary for each person (clothing, hair, name, pronoun)
- Not all transgender/gender nonconforming people want medical intervention
- Medical intervention can include hormone replacement therapy and/or surgery when indicated
Gender Expression

Gender Identity
GLB…

Orientation

Gender Expanding
Intersex vs Gender Identity

- Distinct categories
- Share perspective while anatomy can be one variable, it is not whole picture
- Share notion that variations are not, onto themselves, disorders
- Gender expression may or may not indicate gender identity
- Language examined and evolving — AFAB, AMAB (historical MTF, FTM)

Intersex vs Gender Identity (cont.)

- Both use multidisciplinary team approaches, including family in decision making as much as possible
- Both support concept of patient self determination
- In intersex “watch-and-wait” is prevention. In gender care it can increase distress
Discussion/Self Reflection:
- What did you learn about gender growing up?
- What messages did you get about what was/ was not okay based on gender?
- How did race/culture or other experiences influence your experience of gender?
- Could these constructs prematurely promote medical intervention?

Current Practice on Intersex
- Partner with family to be part of decision making process
- Triage any concerns related to metabolic issues
- Help family understand distinction between cosmetic and medically necessary
- Link family with emotional and social supports (other families, grief counseling, accurate information, support groups)
- Consider own bias as a provider to ensure decisions are made from ethical and medically necessary place
- In most cases, wait on surgical intervention so person develops sense of gender identity and care wishes
Multidisciplinary Care Model

- Family/patient
- Pediatrics
- Labor and Delivery
- Eurology
- Pediatric Surgery
- Social Work
- Clergy, as indicated
- Cultural ambassadors, as indicated

Patient-Centered Model

- Relatively common anatomical variation from the “standard” male and female types
- Intersex genitals are not considered a medical problem
- Sexual and reproductive anatomy vary along a wide spectrum
- Intersex is neither a medical, nor a social pathology
- May signal an underlying metabolic concern and should be assessed and treated as indicated (not intersex genitals, but rather the metabolic condition, if diagnosed)
- Suggested full disclosure and information to facilitate respect for their autonomy and self-determination, truth about their bodies and their lives, and freedom from discrimination
- Gender determined by an interplay between biology, society and psychology
Developmental Family Approach

- Support families in finding language to talk to their children about their condition using language appropriate at each age/stage
- Later onset or awareness may increase psychological distress for patient and family
- Families that knew from birth may have increased distress at later stages of child’s development

Impact on Patient and Families

- Secrecy vs privacy
- Isolated (never met anyone else like them or their child)
- Fearful to tell family/friends
- Grief reactions
- Others sexualizing anatomy and gender in children
- Compliance issues of medication (hormones) without adequate information
- Feel betrayed by medical provider
Paradigm Shift

- From “normal looking” to “there are variations”
- From focus on permitted by law to patient-lived experience
- From focus on parental distress and physician subjective to “watch-and-wait” or in older children, patient wish
- From focus on acute preventative (infancy) to long-term preventative over time (sensation, fertility, continence, etc.)

Questions to Consider:

- Would greater societal acceptance enable a more holistic approach to medical care?
- Would increased awareness of culture increase understanding of how care recommendations may be interpreted?
- When should the role include advocacy?
- How is “do no harm” defined in these cases?
Tips from Intersex Advocates and Patients

- Educate yourself about the specific intersex condition the person has
- Be aware of your own attitudes about issues of sex, gender and disability
- Learn how to talk about issues of sex and sexuality in an age-appropriate manner
- Remember that most persons with intersex conditions are happy with the sex to which they have been assigned
- Do not assume that gender-atypical behavior by an intersex person reflects an incorrect sex assignment

Tips from Intersex Advocates and Patients (cont.)

- Work to ensure that people with intersex conditions are not teased, harassed or subjected to discrimination
- Get support, if necessary, to help deal with your feelings — intersex persons and their families, friends and partners often benefit from talking with mental health professionals about their feelings concerning intersex conditions and their implications
- Consider attending support groups, which are available in many areas for intersex persons and their families, friends and partners
CASE: Kayla

- Age 4, African American
- Born with external female genitals, XX chromosomes
- Raised as female
- Toy and clothing preference “male” (stereotype)
- Tells parents “I’m a boy. I have a penis like the other boys at school”
- Parents feel regretful and think they made a mistake not intervening at birth
- Kayla requests to be called KYLE and he/him
- KYLE gets distressed at any mention of “girl” or “she”
CASE: Jackie

- Age 16, Latina/Caucasian mixed race
- Born with CAH
- Family has not managed medications well
- Jackie has always preferred male type clothing, male friends
- Jackie identifies as a heterosexual female
- Jackie and parents want surgery to correct the enlarged clitoris
- Referred to a gender clinic for assessment of gender identity prior to approving surgery
- Care team also concerned clitoris will enlarge again if patient is not medication compliant
- Family felt referral was unnecessary and were distressed

Case: Tag

- Age 15, South Asian
- Born with Y chromosome and penis over 1 inch
- Penis was “reconstructed” shortly after birth
- Tag raised as male
- Parents did not tell Tag about being intersex
- Tag begins at age 13 to tell parents they have never felt quite like a boy and they don’t feel like a girl either
- Parents take Tag to a therapist who tells the parents to “help him accept he is male”
- The family is open to gender fluid ideas, yet feel worried Tag will suffer persecution and for that reason living as male is best
- Tag becomes distraught and starts to self injure and use alcohol
In Sum

- Optimal care includes a team approach including family input
- Watch-and-wait approach reduces the later poor outcomes about gender or distress or issues with sensation and fertility
- Rule outs for metabolic issues or conditions requiring medical care should be considered
- Linkage to resources for families to manage grief, questions and connections with other families with shared experience
- Gender identity is independent and may or may not overlap related to anatomy or chromosomes

Resources

- Intersex Society of North America: http://www.isna.org/
- Family support: Gender Spectrum: www.genderspectrum.org
- Legal: Transgender Law Center: https://transgenderlawcenter.org
- Sexual Medicine Society: http://www.sexhealthmatters.org/sex-health-blog/for-parents-when-your-child-is-intersex
- https://www.youtube.com/watch?v=KeAVdOJOfKk&t=1s
- https://www.youtube.com/watch?v=cAUDKEI4QKI
- https://www.youtube.com/watch?v=rINtjntqZE
Thank You

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Questions

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