Rights and responsibilities Description of the situations encountered

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Personal Rights
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Intersex – DSD

- Definitions
  - Chicago Consensus Statement: disorders of sex development are “congenital conditions within which the development of chromosomal, gonadal, and anatomic sex is atypical”
  - include at least 50 congenital conditions / variations affecting human sex determination and differentiation, resulting in atypical or ambiguous (external) genitalia
- [terminology:
  - intersex
  - differences /divergence / variations of sex development: biological variations in sexual development instead of pathological condition
  - identification with individual condition]
- incidence: between 1 in 1,500 and 1 in 2,000 children

Examples

A. 46,XX (1 in 14,000 - 15,000 live births)
- congenital adrenal hyperplasia from 21-hydroxylase deficiency
  - genetically female; internal genitalia female
  - severe degree of virilisation of external genitalia (genital tubercle sometimes constituting normal-looking phallus)
  - no vaginal connection to perineum; enlarged and merged genital fold (sometimes scrotal-like appearance)

B. 46,XY (1 in 20,000 live births)
- 5-alpha reductase deficiency
- 17-beta hydroxysteroid dehydrogenase III deficiency
  - genetically male
  - varying degrees of undervirilisation of external genitalia
    - often appear female
    - sometimes ambiguous genitalia
    - sometimes predominantly male, often with an unusually small penis (micropenis) and urethra opening on underside of penis (hypospadias)
  - with or without undescended gonads
  - with or without feminine remnants

Examples

- complete androgen insensitivity syndrome
  - genetically male; internal genitalia male
  - external genitalia appear female
  - with or without undescended gonads
  - with or without feminine remnants

C. Sex chromosome

- [45,X: Turner Syndrome; 47,XXY Klinefelter Syndrome]
- 45,X/46,XY: mixed gonadal dysgenesis (1 in 10,000 live births)
  - may present with ambiguous or asymmetrical genitalia

D. Disorders of gonadal development

- ovotesticular DSD (1 in 100,000 live births)
  - both ovarian and testicular tissues; abnormally differentiated genital structures

E. Non-hormonal/non-chromosomal DSD

- cloacal exstrophy (1 in 200,000 - 400,000 live births)
  - non-closure of bladder; exposed abdominal organs
  - genital tracts may not be completely formed

Surgical intervention

- dominant treatment method: surgical reconstruction of genitalia = estimated to be 0.1-0.2 % of all live births
- no consensus regarding indications, timing, and procedure

A. 46,XX
   - congenital adrenal hyperplasia

B. 46,XY
   - 5-alpha reductase deficiency
   - 17-beta hydroxysteroid dehydrogenase III deficiency
   - complete androgen insensitivity syndrome

C. Sex chromosome
   - 45,X/46,XY: mixed gonadal dysgenesis

D. Disorders of gonadal development
   - ovotesticular DSD

E. Non-hormonal/non-chromosomal DSD
   - cloacal exstrophy

Early surgical intervention

1. Emergent surgery (imminent threat to patient's health)
    a) creating a urinary opening where none exists
    b) correcting exstrophic conditions in which bladder/organs protrude from abdominal wall
    c) removing malignant tissue (gonadal dysgenesis)
2. Early genitical surgery to reinforce initial gender assigment
    - ("reconstructive"/"corrective") / during early infancy (before 24 months)
    - normalise appearance of genitals corresponding with initial gender assignment + removal of gonadal tissue at odds with initial gender assignment

    a) feminising genitoplasty
        - removal of tissue from enlarged clitoris or small penis (clitoroplasty)
        - create or enlarge vagina (vaginoplasty) and create/refashion labia minora
        - removal of (healthy) testes

    b) masculinising genitoplasty
        - creation of penis (phalloplasty)
        - creation of scrotum / correction of penoscrotal transposition
        - move down testicle that has not descended (orchidopexy)
        - removal of feminine remnants (Müllerian structures:) vagina, uterus and fallopian tubes

Early surgical intervention

Arguments provided for early intervention

1. Avoiding potential health hazards related to altered anatomy and function of urogenital tracts:
   a) Improve urinary functioning (avoid incontinence and urinary tract infection).
   b) Remove malignancy risk of retained gonads.
   c) Avoid blood of fluid retention in vaginal or uterine cavities.
2. Early surgical intervention is relatively easier:
   a) Pubertal or post-pubertal surgery has a much greater risk for morbidity (blood loss and infection).
   b) Much less surgical experience with late feminisation.
   c) Psychological impact of late genital surgery likely to be more significant.
3. Helping persons to achieve future satisfactory sexual function, consistent with their gender identity:
   a) Restore functional genital anatomy to allow future penile-vaginal intercourse.
   b) Surgical intervention to allow menstruation.
   c) Facilitate future reproduction if possible.
   d) Avoid late virilisation at puberty in persons raised as girls, or breast development in persons raised as boys.

Early surgical intervention

Arguments provided for early intervention

4. Encourage psychosocial development and psychological well-being:
   a) Avoid shame and discomfort related to atypical anatomy.
   b) Avoid stigmatisation and social rejection related to atypical anatomy.
   c) Encourage stable body image, gender identity, and gender role behaviour.
   d) Encourage integration/relationships in social environment.
5. Encourage positive family dynamics:
   a) Encourage parent-child bonding.
   b) Avoid parental shame, grief and guilt.
   c) Avoid negative impact on family relationship and parent-parent relationship.
   d) Encourage social visibility of child among family and friends.
   e) Avoid feelings of having abandoned child in need of care and support.
   f) Align with cultural/religious/social context where it may be impossible to wait for adolescence to determine sex or gender identity.
   g) Respond to parents' ideas of raising the child in the best possible conditions.

A few, small studies among persons with congenital adrenal hyperplasia who who had undergone feminising genitoplasty at (very) young age indicate preference for early surgical intervention.

Early surgical intervention

Arguments provided contra early intervention

1. Significant complication rates and poor long-term physical outcomes:
   a) Chronic pain
   b) Scarring
   c) Incontinence
   d) Need for lasting hormone therapy
   e) Further genital surgeries will be required in puberty/adulthood
   f) Risk from anaesthesia
2. Impairing enjoyment of sexuality through nerve damage:
   a) Diminished sexual sensation; loss of orgasmic function
   b) Sexual dysfunction
3. Loss of of potential fertility and loss of benefits of endogenous hormones caused by removal of healthy gonadal tissue.
4. Traumatising effects of multiple operations and repeated genital examinations.
5. Most early surgery is (virtually) irreversible and restricts later options.
6. Most early surgery can be postponed until adolescence.

Early surgical intervention

Arguments provided contra early intervention

6. Most early surgery answers to cosmetic and social needs, and serves no legitimate medical purpose:
    a) Children raised with "ambiguous" sex anatomy are at no greater risk for psychosocial problems than general population
    b) Early surgery is prompted by anticipated distress in parents and community
7. Significant/high rates of gender dysphoria: incorrect gender assignment:
    a) Actual gender development/identity differs from early gender assignment
    b) May have non-traditional instead of binary gender identity
    c) Reassign as other gender

Gender dysphoria

A. 46,XX
- congenital adrenal hyperplasia

B. 46,XY
- 5-alpha reductase deficiency
- 17-beta hydroxysteroid dehydrogenase 3 deficiency
- complete androgen insensitivity syndrome

40-60%?

C. Sex chromosome
- 45,X/46,XY: mixed gonadal dysgenesis

D. Disorders of gonadal development
- ovotesticular DSD

E. Non-hormonal/non-chromosomal DSD
- cloacal exstrophy

Early surgical intervention

Arguments provided contra early intervention

Early surgery can cause significant harm to quality of life, and for some/many may have severely mutilising and traumatising effects.

Suggestions:

1. Delay surgical intervention until child is old enough to participate.
2. Parental distress should instead be addressed directly through peer support and competent mental health care.
3. Foster societal concept of gender diversity that includes persons whose gender assignment is provisional, and where binary sex and heteronormativity are no longer central.

Rights and obligations of healthcare professionals

Oviedo: **Article 4 – Professional standards**

Any intervention in the health field must be carried out in accordance with relevant professional obligations and standards.

- in accordance with law
  - prohibition?
  - allowed → - health law / law on patients' rights
    - code of medical deontology
    - codes of professional medical associations
- obligations:
  - provision of information
  - obtaining free and informed consent –> <u>representatives</u>
  - performance in accordance with due standard of care: medical state-of-the-art –> <u>aligned with best interests of child standard</u>

Rights and obligations of healthcare professionals

- standard of care redefined
    - some countries: prohibition of certain interventions
      (∼ recommendations from international human rights organisations)
    - other countries: much more strict standard of care due to consensus statements and recommendations from national and international professional medical associations
        1. obligations: organisation and approach
            - multidisciplinary teams
            - individualised patient- and family-centered care
            - holistic approach
                - ➢ all medical, psychological, social and cultural considerations of patient and parents
                - ➢ full disclosure of potential risks and benefits of options, empowering representatives to make decisions in best interest
                - ➢ long-term follow-up
                - ➢ peer support network

Rights and obligations of healthcare professionals

- standard of care redefined
  2. obligations: best medical practice
     - considerable improvement of surgical techniques
     - early surgery
       - ➢ emergency conditions
       - ➢ removal of malignant tissue
       - ➢ correct hypospadias
       - ➢ correct severe virilisation: clitoroplasty for CAH female infants with clitoromegaly
     - no removal of healthy, functioning gonadal tissue
     - nerve preservation wherever possible
     - postpone vaginoplasty or phalloplasty until adolescence

     → unclear to what extent this is followed in practice:
     - numbers of clitoral reduction operations remain high
     - vaginoplasty performed less frequently

Rights and obligations of representatives

Oviedo: **Article 6 – Protection of persons not able to consent**

- Intervention may only be carried out on person who does not have capacity to consent, for his or her direct benefit.
- Intervention may only be carried out with authorisation of his or her representative or authority or person or body provided for by law.
- Beforehand be given appropriate information as to purpose and nature of intervention as well as on its consequences and risks.

- rights:
  - direct upbringing and education of their children –> right to make medical decisions for their children
  - free and informed authorisation:
    - concern: not properly informed + may be nudged by physicians
    - full disclosure of risk of physical injuries from early surgical intervention and of risk of gender dysphoria
    - mention possibility of no intervention
    - no undue pressure; sufficient consultations and time to (re)consider

Rights and obligations of representatives

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- obligations:
  - act "for his or her direct benefit" –> best interests of the child
  - is there a right to authorise non-medically necessary irreversible procedures that may negatively effect child's identity and/or reproductive capacity?
    - ∾ sterilisation: authorisation by court or prohibited

Rights of the child: best interests

Consideration as to what constitutes best interests may be complex

- requires assessment of matters wider than purely medical benefits
- complexity of interaction between biological, psychological, social, and cultural factors as to physical and psychosocial benefits for child: essential to make decisions on case-by-case basis
- difficult to separate interests of child and parents when they are intimately connected, at least through infancy and early childhood: context of whole family might be important to determine child's best interests
- issue as to how much to take into account that child will grow up in culture strongly supportive of clear binary sex/gender and heteronormativity
- impossible to predict with confidence what gender child will eventually identify with

Rights of the child: best interests

Consideration as to what constitutes best interests may be complex

- systematic reviews of adult gender identity in persons who underwent early gender-enforcing surgery is lacking
  - some evidence that future well-being is improved for persons developing gender identity that corresponds to early gender assignment
  - little evidence regarding long-term impact/harm of non-corrected genitalia
  - strong evidence that for some disorders/variations significant to high percentage of individuals experience gender dysphoria: highly problematic

Rights of the child: best interests

Consideration as to what constitutes best interests may be complex

→ dissent within and between stakeholders regarding what constitutes best interests and optimal care:

1. Prohibiting early surgical intervention should be avoided:
   - "goes against right to highest attainable standard of health", considering that favourable physical, social, and emotional conditions are critical factors for optimal development
   - "option of inaction is harmful, as it commits to childhood of ambiguous genital anatomy, particularly difficult if parents' and society's values are not supportive"
   - "closes important window of opportunity for child"
   - "step backwards in evolution, rather than forwards"
   - "unnecessarily delaying interventions can be catastrophic in adulthood and is not in best interests of child"
   - "social experiment with unknown consequences"

Rights of the child: best interests

Consideration as to what constitutes best interests may be complex

2. Prohibiting early surgical intervention is crucial:
   - since it is difficult to find out whether early surgery would be in best interests of child, this decision can only be taken by individual concerned
   - since one's gender does not necessarily develop in conformity with one's assigned sex, early surgical intervention will predictably be very harmful to significant percentage of children

   –> major concern that for these persons human rights are severely infringed:
   - ➢ freedom from torture and inhuman or degrading treatment (Decision no 42821/18 *M v France*)
   - ➢ right to physical (and psychological) integrity
   - ➢ right to respect for private life: "right to an open future"
   - ➢ right to highest attainable standard of health
   - ➢ right to be heard in matters affecting the child
   - ➢ prohibition of discrimination

   –> wait until child attains capacity to participate in decision-making (10-14 y)

Thanks for your attention

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