Differences/Disorders of Sex Development (DSD): A World of Uncertainty

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Winnie Mandela calls on South Africa to stand behind Caster Semenya after claims the gender-row runner is a hermaphrodite

By DAILY MAIL REPORTER
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The former wife of Nelson Mandela has called on all of South Africa to stand behind gender-row athlete Caster Semenya after claims the 800m champion runner is a hermaphrodite.

Winnie Mandela spoke out as Caster's mother Dorcas reacted with fury to the report that her daughter has both male and female sex organs, but no womb or ovaries.

The Sydney Daily Telegraph published the details yesterday after reportedly speaking to a source close to the International Association of Athletics Federation (IAAF).
Disorders of Sex Development

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

a world of uncertainties

Naming
Diagnosis
Gender
Genital surgery
Disclosure
Fertility
Outcomes
INTERSEX vs DSD

“I am not a disorder”

“Words can wound”

“Why not VSD (Variations of Sex Development?) Or Differences of Sex Development?”

“People are free to identify as Intersex”
Intersex is a vague term
Does one need to have ambiguous genitalia to be “intersex?”
Do men who are XX or women XY “qualify?”
What about transgender individuals identifying as “intersex?”

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

DSD is a diagnosis
Knowledge from all medical fields apply
Evidence-based medicine should be used for all medical/surgical decisions on patients with DSD
Medical labeling has clinical and social consequences

Should clinicians label members of a sexual minority?

Should there be an “intersex exception” when it comes to clinical labeling?

Could “Disorders/Differences in Sex Development” be a compromise?
DIAGNOSTIC UNCERTAINTY DSD
Questions from the real world

• Is it worth and useful to test « everything »?
• To know what exactly?
• When do we know « enough » to provide high quality patient-centered care?

• Tentative Answers:
  – We don’t know what we don’t know if we never capture it
  – It’s been a historically growing target and will continue to grow
  – Genome sequencing has allowed to increase dramatically the amount of information at a lesser effort for patients
## Gender Assignment

<table>
<thead>
<tr>
<th>GID DETERMINANT</th>
<th>TRADITIONAL</th>
<th>CHALLENGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender of rearing</td>
<td>Crucial to identity, adjustment</td>
<td>Reflects brain masculinization</td>
</tr>
<tr>
<td>Prenatal Androgen</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ROLE OF GENITALIA</td>
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</tbody>
</table>
ASSIGNING GENDER

Based on outcomes from medical literature

Considerations of hormones, fertility, genetics

What about external genitalia?
CONGENITAL ADRENAL HYPERPLASIA (CAH):

Excess of Androgens masculinizes the genitals of XX fetuses. Internal reproductive organs are female and fertile. Typical assignment has been female mostly based on preserving fertility.
CAH WITH SEVERE MASCULINIZATION

**REARED AS GIRLS**

- 95% (N=250) maintained gender identity as women
- 5% experienced gender dysphoria (not related to degree of genital virilization) (Dessens et al, 2005)

**REARED AS BOYS**

- Among those with severe masculinization; low/variable incidence of developing gender dysphoria
- N=35 initially assigned male, but not reassigned female after diagnosis (Can Cua et al, 1989; Woelfle et al., 2002, Gupta et al., 2006; Dessens et al., 2005)
- 18/35 adults
- 4/18 dysphoria
- N=10 no dysphoria (Lee, Houk & Husmann, 2010)
## UNCERTAINTY

### Genital Surgery

<table>
<thead>
<tr>
<th></th>
<th>TRADITIONAL</th>
<th>CHALLENGE</th>
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</thead>
<tbody>
<tr>
<td><strong>RATIONALE</strong></td>
<td>Anatomy to match rearing</td>
<td>Comfort of others</td>
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</table>
| **CONSEQUENCES** | + gender identity  
                 | + adjustment  
                 | + sex. intercourse                  | - gender change  
                 | - sex. function                          |
| **DECISION-MAKER** | Physician and Family                      | Patient (when old enough)          |
Genital Surgery

Based on literature?

*Little available, with no easy objective assessments*

Surgical techniques change

No evidence-based answers
QUESTIONS TO ASK BEFORE GENITAL SURGERY

Would anyone desire atypical genitals?
Will surgery create normal-looking genitals?
Will surgery preserve sexual sensation?
Will there be complications?
Will surgery solve psychological issues?

Slide inspired by Bo Laurent
CAH WITH SEVERE MASCULINIZATION

REARED AS GIRLS

• Genital surgery
• Multiple surgeries
• Complications (vaginal stenosis, hairy vagina, short vagina)

• Functional and cosmetic results (according to physician and patient) in general not optimal (e.g., pain during intercourse, cosmetic dissatisfaction) (Creighton et al., 2001, Gastaud et al., 2007)

REARED AS BOYS

• Little to no surgery needed, no loss of sensitive genital tissue
## UNCERTAINTY

### Disclosure

<table>
<thead>
<tr>
<th>STRATEGY</th>
<th>TRADITIONAL</th>
<th>CHALLENGE</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Limit disclosure</td>
<td>Full disclosure</td>
</tr>
<tr>
<td>RATIONALE</td>
<td>Prevent GID confusion</td>
<td>Prevent secrecy/shame</td>
</tr>
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</table>
“These are social interventions on your genitals — if we’re going to use that language about Africa, we’re going to have to use it here,” Alice Dreger, a historian of medicine and intersex patient advocate, told BuzzFeed News.

“This completely antagonizes the medical community. This is not progress, in my opinion,” Eric Vilain, professor of human genetics and pediatrics at UCLA, told BuzzFeed News.
The DSD-Translational Research Network
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“Disorders of Sex Development:
Platform for Basic and Translational Research”

For more information, please contact the DSD-TRN coordinator,
Dr. Emmanuèle Délot: edelot@childrensnational.org (202) 476 6011.