DISORDERS OF SEX DEVELOPMENT (DSD)
When To Tell the Patient?

LWPES
Baltimore, Maryland

Psychological Management of DSD Education
5/2/09
Tom Mazur, Psy.D.
Women & Children’s Hospital of Buffalo
Questions Other Than “When”

Who?

How?

What?
“I wish I would have had the opportunity to have them (parents) explain as much as they knew. Certainly I realized they could not have explained it in professional terms to a youngster, but they could have tried at least to make it have some sense or some semblance as to the reason why I was going rather than rely upon the doctors per se to spell it all out for me. I think perhaps hearing it from my parents might have softened the blow, not that I actually considered there was much of a blow, but I mean, made it more comfortable and more willing to be accepted by me, if it seemed to be okay with them.”

Quattrin, Aronica and Mazur 1990
EDUCATION OF PARENTS

Creating a template

- Internal sexual/reproductive system
- External sexual/reproductive system
- Gender identity development
LESSONS FROM TEMPLATE

Get the big picture

- Not a freak of nature and understandable explanation
- Gender identity equals complex interaction of many factors
PURPOSE OF DIAGRAMS

- A template upon which their child’s condition (diagnosis) can be placed and understood
- Remove from the realm of myths, freaky, weird
- Allay fears, not create fears (MGD)
EDUCATION OF PARENTS

Goals

- Understanding why gender not yet assigned (infant with severe hypospadias)
- Supplant fear, anxiety with confidence
- Answer questions
- Support: family siblings
CHROMOSOMES: VARIATIONS ON A THEME

- 46,XX (females/males)
- 46,XY (males/females)
- 45,X
- 47,XXY
- 45,X/46,XY
“I’ve been raised as a female, and my perception of myself is female. So relating to it is difficult. I mean, if I was a male, I could relate to it. But as far as I’m concerned, I’m female. Even though my genes say otherwise. So, meaning I couldn’t relate to it since it was alien because I consider myself female and not male.”

Quattrin, Aronica and Mazur 1990
“Sounds like a Y to me”

Complete Androgen Insensitivity Syndrome
“Does that mean I’m a male”
(then smiles)

Complete Androgen Insensitivity Syndrome
“Does that mean I’m a female?...No way!”

46,XX male
Not a question of “to tell or not to tell.” It is rather a question of “when and how to tell”

Tell gradually to prevent mental indigestion

Be flexible

Patient focused

Gender does not equal biology
## GUIDELINES FOR COMPREHENSIVE SEX EDUCATION

**Sexual Information and Education Council of the United States (SIECUS)**

<table>
<thead>
<tr>
<th>AGE RANGE</th>
<th>TOPIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-8</td>
<td>Naming of body parts including genitals</td>
</tr>
<tr>
<td>9-12</td>
<td>Pubertal development</td>
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<td></td>
<td>Normal variation in pubertal development</td>
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<tr>
<td></td>
<td>Reproductive system</td>
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<tr>
<td></td>
<td>Pregnancy</td>
</tr>
<tr>
<td></td>
<td>Birth defects</td>
</tr>
<tr>
<td>15-18</td>
<td>Increased knowledge of anatomy, reproduction and gender</td>
</tr>
</tbody>
</table>
## MODIFIED SIECUS GUIDELINES FOR DSD

<table>
<thead>
<tr>
<th>AGE RANGE</th>
<th>TOPIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-12</td>
<td>Genital differences</td>
</tr>
<tr>
<td></td>
<td>Pubertal variation/development</td>
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<tr>
<td>8+</td>
<td>Variations of development</td>
</tr>
<tr>
<td></td>
<td>Your development</td>
</tr>
<tr>
<td>Pre-adolescence/</td>
<td>Diagrams of sexual system</td>
</tr>
<tr>
<td>Adolescence</td>
<td>Diagnosis (CAIS)</td>
</tr>
<tr>
<td></td>
<td>Review</td>
</tr>
<tr>
<td>Post adolescence</td>
<td>Recycling: job not over (CAIS/MRKH)</td>
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</tbody>
</table>
EDUCATION OF CHILD

Creating a template

- Genital differences
- Pubertal development
- Making a baby
EDUCATION OF CHILD

Genital differences
“What is a Girl? What is a Boy?”

Stephanie Waxman
1976
PHOTO REMOVED
(from S. Waxman, 1976)
What is a girl?

Some people say a girl is someone with long hair.
But Lucas has long hair. And he’s a boy.

(from S. Waxman, 1976)
What is a boy?

Some people say a boy is someone with short hair.
PHOTO REMOVED (from S. Waxman, 1976)

But Mimi has short hair. And she’s a girl.
Some people say a girl is someone who plays with dolls.
But Noah is taking care of his doll. And he’s a boy.
Some people say a boy is someone who doesn’t cry.
But Eric is crying.
And he’s a boy.
Then, what *is* a boy?
A boy is someone with a penis.
Every boy has a penis.

PHOTO REMOVED
(from S. Waxman, 1976)
And, what *is* a girl?
A girl is someone with a vagina.
Every girl has a vagina.

PHOTO REMOVED
(from S. Waxman, 1976)
Are you a girl?
or
Are you a boy?
Can you finish this drawing?
EDUCATION OF CHILD

Pubertal development
If you are a boy, you will grow up to be a man.
Every man has a penis.
When Dennis became a man his voice got deeper and hair grew on parts of his body.

(From S. Waxman, 1976)
If you are a girl, you will grow up to be a woman. Every woman has a vagina.
When Natalie became a woman her breasts got bigger and hair grew on parts of her body.

PHOTO REMOVED
(from S. Waxman, 1976)
EDUCATION OF CHILD

What do you need to make a baby?
PHOTOS AND ILLUSTRATIONS REMOVED
(from S. Waxman, 1976)
GENITAL DIFFERENCES
(Lesson Two)

- Different shapes/sizes
- Different places (testes, urinary opening)
- Different looks
- Different number
PHOTO REMOVED
(from S. Waxman, 1976)
PUBERTAL DIFFERENCES
(Lesson Two)

- Different shapes/sizes
- Different rate of growth
- Different parts
- No parts
Sometimes people can have different parts; boys can have girl parts and girls can have boy parts
Different is just different
MAKING A BABY
(Lesson Two)

- No tadpole (sperm) factory
- No egg factory
- No baby nest (uterus)
- No baby tunnel (vagina)
- Sometimes parts are there, but we don’t know why no baby
EDUCATION OF CHILD

- Adoption
- An instant family
- Deciding not to be a mommy/daddy
- Assisted Reproductive Technology (ART)
“The sharing of information accurately described here not only helped me understand my medical condition at a rate I could grasp, but it served to reduce significantly my anxieties and fears…They encouraged me to ask questions… I came to understand my condition and accept it as a fact of nature rather than a defect of the individual.”

Quattrin, Aronica and Mazur 1990
HOW DO YOU GET THERE?

- Psychologic management
- Empower parents
  - Provide accurate medical context
  - Nature abhors a vacuum
- Honest information over time
- Answer all questions
- No information withheld
- Non-judgmental spirit
RESOURCES


- Hospital for Sick Children, Toronto. Interactive website that helps explain the development of external genitalia and internal reproductive glands as well as some of the variations that can occur. [http://www.aboutkidshealth.ca/HowtheBodyWorks/Sex-Development](http://www.aboutkidshealth.ca/HowtheBodyWorks/Sex-Development)
RESOURCES
