

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

LWPES

Baltimore, Maryland

Psychological Management of DSD Education

5/2/09

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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

PRINCIPLES OF EDUCATION

- 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)

AGE RANGE

TOPIC

5-8

Naming of body parts including genitals

9-12

Pubertal development

Normal variation in pubertal development

Reproductive system

Pregnancy

Birth defects

15-18

Increased knowledge of anatomy,
reproduction and gender

MODIFIED SIECUS GUIDELINES FOR DSD

AGE RANGE

TOPIC

5-12

Genital differences

Pubertal variation/development

8+

Variations of development

Your development

Pre-adolescence/
Adolescence

Diagrams of sexual system

Diagnosis (CAIS)

Review

Post adolescence

Recycling: job not over (CAIS/MRKH)

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.

PHOTO
REMOVED

(from S. Waxman, 1976)

But Lucas has long hair.
And he's a boy.

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.

**PHOTO
REMOVED**

(from S. Waxman, 1976)

But Noah is taking
care of his doll.
And he's a boy.

Some people say a boy
is someone who doesn't cry.

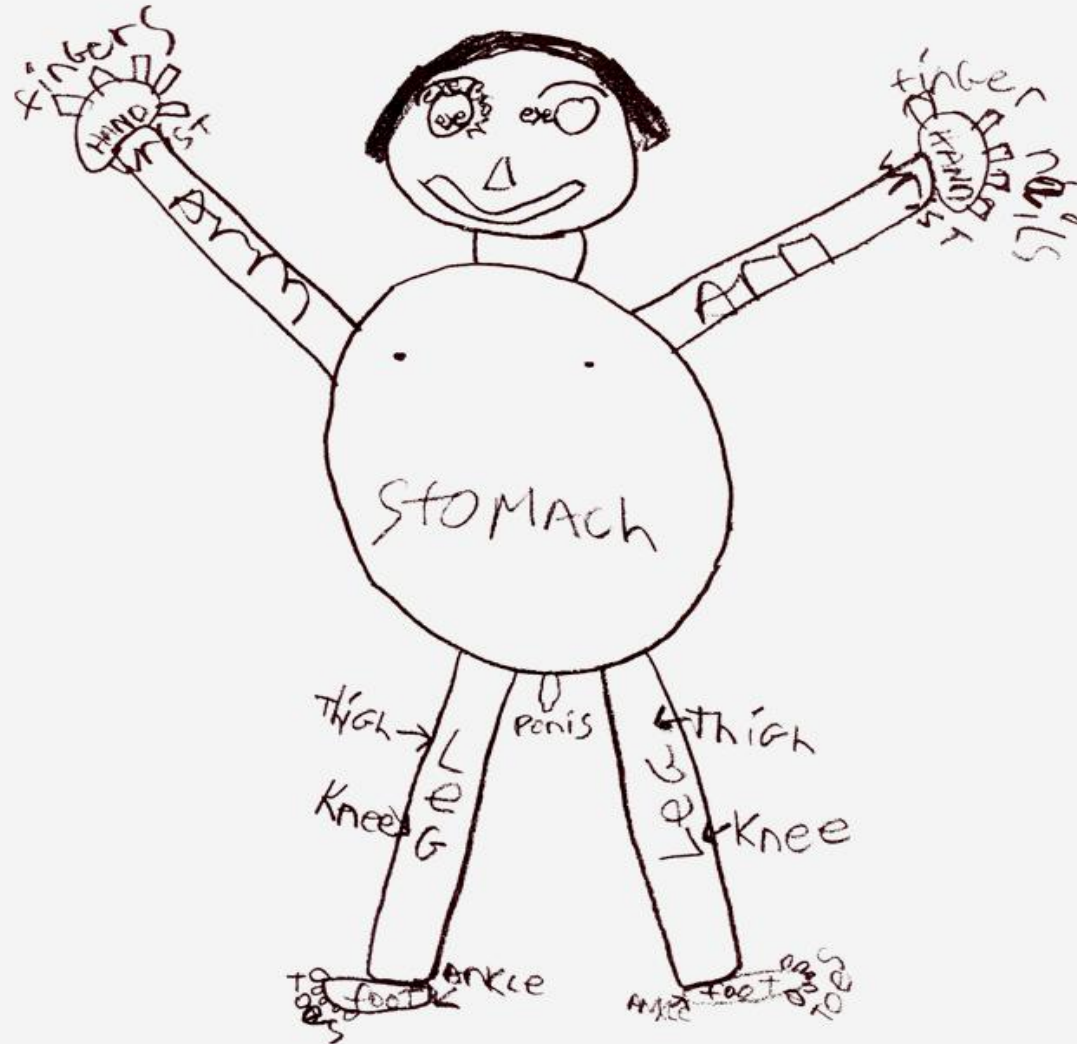
**PHOTO
REMOVED**

(from S. Waxman, 1976)

But Eric is crying.
And he's a boy.

Then, what *is* a boy?

BOY



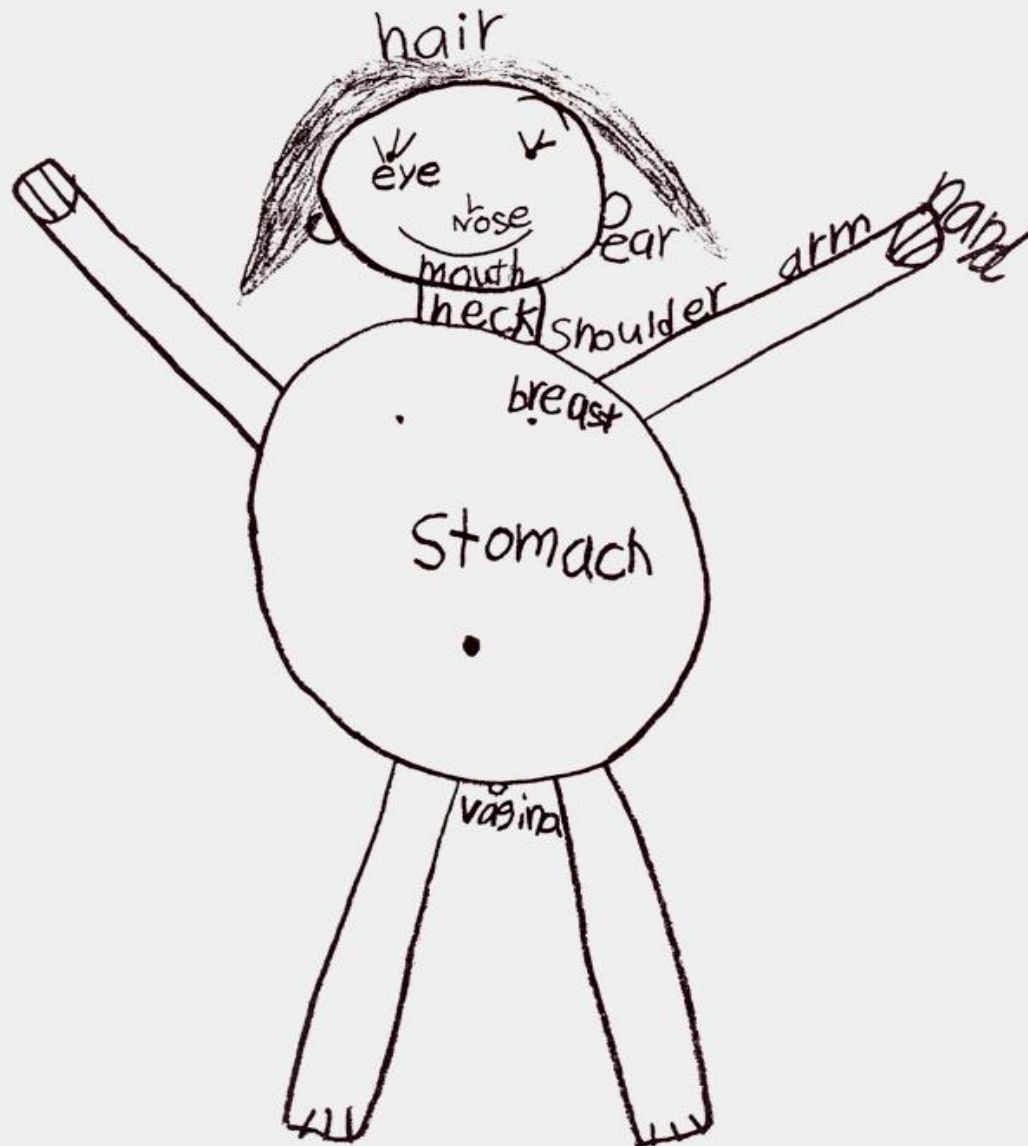
A boy is someone with a penis.

Every boy has a penis.

**PHOTO
REMOVED**
(from S. Waxman, 1976)

And, what *is* a girl?

GIRL



A girl is someone with a vagina.

Every girl has a vagina.

**PHOTO
REMOVED**

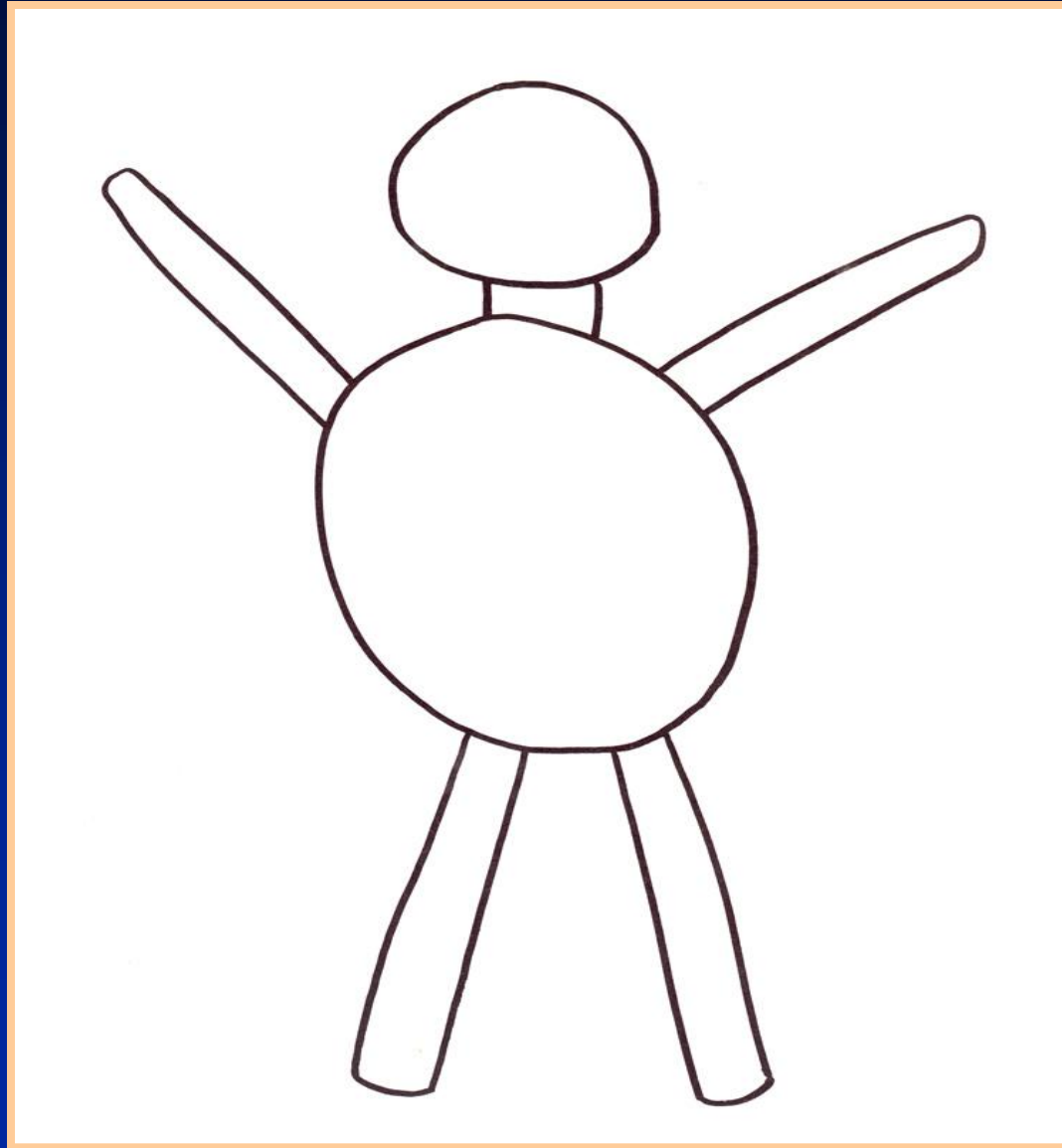
(from S. Waxman, 1976)

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.

**PHOTO
REMOVED**

(from S. Waxman,
1976)

When Dennis became a man his voice got deeper and hair grew on parts of his body.

If you are a girl, you will
grow up to be a woman.
Every woman has a vagina.

**PHOTO
REMOVED**

(from S. Waxman,
1976)

When Natalie became a woman her breasts got bigger and hair grew on parts of her body.

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

EDUCATION OF CHILD

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

- Carmichel, P.A. & Ransley, P.G. (2002). Telling children about a physical intersex condition. *Dialogues in Pediatric Urology*, June, 2002, 7-8.
- Consortium for the Management of Disorders of Sex Development. (2002) DSD Guidelines for Clinicians. May be downloaded from <http://www.dsdguidelines.org/files/clinical.pdf> (includes scripts for talking to patients)
- 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>

RESOURCES

- Mazur T. Ambiguous genitalia: Detection and counseling. *Pediatric Nursing* 1983; 9:417-422.
- Quattrin T, Aronica S, Mazur T. Management of male pseudohermaphroditism: a case report spanning 21 years. *Journal of Pediatric Psychology* 1990; 15:699-709.