Ambiguous Genitalia Management: It Takes a Team

BY SHERRY BOSCHERT
San Francisco Bureau

It's one of the first things that parents want to know when a baby is born: girl or boy?

When the answer isn't clear, physicians walk into a mine field of choices that could have lifelong repercussions for the child and the parents.

Psychiatrists, psychologists, and social workers today play a much bigger role—and surgeons less of a role—in the care of people with disorders of sex development, compared with past management practices.

"I think the field has revolutionized in the last 10-15 years. The impetus for that largely came from patients themselves," said Dr. Joel Frader, a pediatrician at Children's Memorial Hospital and Northwestern University, Chicago.

The revolution is still in progress, with a bit of chaos in the streets. A dearth of research data to inform guidelines leaves clinicians with little to follow besides opinion-based consensus statements and their instincts.

"Because the ultimate outcomes are unpredictable from our standpoint, there is no standard of care. We don't really know what to do clinically. We're not sure what to do psychologically. Yet mothers and fathers have to go ahead and raise their child. It's a very difficult situation," said Dr. William G. Reiner, a psychiatrist and urologist who is director of the psychosocial development clinic at the University of Oklahoma Health Sciences Center, Oklahoma City.

Some key points of agreement, however, inform current practices: Don't rush to surgery. Integrate psychological care for the family and the child. Consult a multidisciplinary team (preferably at an experienced center) that may include a pediatric endocrinologist, pediatric urologist or surgeon, gynecologist, child psychiatrist or psychologist, geneticist, neonatologist, social worker, nurses, and medical ethicist if needed. Be honest with parents and patients. Emphasize functional rather than cosmetic results in any treatment. Make management patient-centered and consider the long-term physical, psychological, and sexual well-being of the patient.

"The single biggest change is our recognition that the infant born with anomalous genitalia is a real, live human being, not a blank slate," Dr. Reiner said. "We have no idea what that child is going to be like at age 5, or 15, or 50."

A decade ago, Dr. Frader recalled, training manuals and textbooks for general pediatricians or pediatric endocrinologists referred to the problems now called disorders of sex development (DSD) as psychological emergencies.

"That sets a very inappropriate tone. There's nothing emergent in 99.9% of these cases," he said. A small minority of babies with congenital adrenal hyperplasia will have life-threatening endocrinologic disturbances that have nothing to do with the appearance of the genitalia. "So there's never a surgical emergency," he stressed.

Dr. William Byne, a psychiatrist at Mount Sinai School of Medicine in New York, added, "If there's a psychosocial emergency in the delivery room, a mental health professional should be brought in. The birth of an intersex child is rarely a medical emergency. We should not rush to make irreversible medical decisions at a time of crisis."

Unfortunately, he and other experts agreed, there are not enough mental health providers trained to handle DSD.

Dr. Byne, Dr. Frader, and Dr. Reiner contributed to the 2006 Clinical Guidelines for the Management of Disorders of Sex Development in Childhood, which was produced by a consortium of clinicians, patients, and parents and published by the Intersex Society of North America (www.dsdguidelines.org).

Dr. Reiner also was 1 of 50 U.S. and European experts who produced the separate Consensus Statement on Management of Intersex Disorders from the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatric Endocrinology around the same time (Pediatrics 2006;118:e488-500).

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DR. BYNE

In the past—and sometimes today—intersex newborns would be separated from mothers while a paternalistic cadre of physicians kept parents uninformed until the physicians declared how they would "fix" the child's problem, usually by surgically altering the genitalia. Most intersex children were never told of their condition.

"The medical model has been that sexual ambiguity is incompatible with their psychological health," said Cheryl Chase, executive director of the Intersex Society of North America, Rohnert Park, Calif., while speaking at the annual meeting of the American Psychological Association in San Francisco.

Parents, in consultation with their clinical team, can assign the child a gender without resorting to surgery, the recent guidelines note.

The veil of secrecy that physicians once placed around the parents and the child does not conform with today's ethics of informed consent, Dr. Byne said. Clinicians should stress what is good about the child when discussing the DSD with parents, encourage them to give the child a gender-neutral name, and promote bonding with the child.

Not informing patients of their DSD as they grow up can lead to psychiatric crises when they discover during puberty or adulthood what their parents and physicians haven't told them. Dr. Byne has treated a number of suicidal adults whose problems included a huge sense of betrayal after discovering their DSD history.

Although it is commonly believed that cosmetic surgery on intersex children in the first year of life relieves parental distress and improves the attachment between the child and the parents, there is scant evidence for this. Almost anything a surgeon does to "fix" a young child with DSD will interfere in the long run with the patient's ability to experience sexual pleasure, said Dr. Frader.

The "vast majority" of patients with DSD do have reconstructive surgery in the first year of life, said Dr. John P. Gearhart, professor and chief of pediatric urology at Johns Hopkins University, Baltimore.

Some surgeries are done for functional reasons (like ensuring proper urinary drainage to avoid infections), and others are done to make the baby "look normal," he said. Dr. Gearhart called the birth of an intersex child "a true emergency situation" in an article he coauthored that has been criticized by other DSD experts (Urol. Int. 2005;75:291-7).

Dr. Reiner said, "I think most of the children are going to want surgery, but what surgery isn't clear." The consortium's clinical guidelines recommend delaying surgery or hormonal treatment until the patient can participate in the decision, which usually means until puberty.

Others take a middle ground, saying evaluation must include assessment of the parents' ability to cope with the stress of genital ambiguity in their child. Some parents will demand surgery even when it is not being recommended. "I've seen it happen," Dr. Reiner said. ■

Anger Management for Boys Increases Emotional Vocabulary

BY SHERRY BOSCHERT
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San Francisco — An anger management intervention that was designed specifically for boys significantly improved their emotional vocabulary, according to preliminary results reported by Aimee Coonerty-Femiano at the annual meeting of the American Psychological Association.

Fourteen eighth-grade boys in a rural school district who were referred by a psychologist or guidance counselor met weekly for 44-minute group sessions with an anger management counselor for 8 weeks. One group of seven boys pursued the anger management program, and the other seven waited 8 weeks before starting the program, serving as a control group.

The intervention teaches that anger is secondary to other emotions, and uses concepts from cognitive-behavioral therapy to teach about connections between thoughts, feelings, and behavior. The program also raises awareness about differences between boys and girls related to emotions, said Ms. Coonerty-Femiano, a doctoral student at Boston College, and her associates.

The first treatment group significantly improved its emotional vocabulary score from four to eight emotions at the end of 8 weeks, she said. When the delayed-treatment (control) group went through the program, its emotional vocabulary score increased significantly from five to eight emotions.

One of the most successful strategies employed by the intervention was a questionnaire at the start that asked what media the boys consumed, and those media were incorporated into the intervention, she said.

Topics and exercises were introduced using examples of emotional experiences and reactions from the "South Park" television cartoon show and movies starring Adam Sandler. These references to the popular culture engaged the boys' attention and participation.

In one exercise, boys in the intervention group sorted "feeling cards" representing different emotions by categorizing them as emotions experienced by boys or girls or "not sure."

In another exercise, cards representing events that triggered anger generated discussion of the boys' thoughts and feelings at the time of the event, how they reacted, and how they felt about their reaction.

Participants in the program had a variety of diagnoses, including depression, attention-deficit/hyperactivity disorder, bipolar disorder, and oppositional-defiant disorder. Half the boys already were in individual therapy, and half were taking medications for a disorder.

The program raises awareness about differences between boys and girls related to emotions.

MS. COONERTY-FEMIANO

Anecdotally, after the study, the names of boys in the intervention group came up less often in weekly meetings of school guidance counselors, where the boys initially had been flagged for intervention. "I'm not sure if that's due to the group, but we would like to think so," she said.

Besides increasing emotional vocabulary, the intervention changed awareness of other feelings that accompany anger, and awareness of obstacles faced specifically by boys when expressing emotions. Themes that emerged from participants' comments were that boys don't like being vulnerable, and that strategies are needed to make a safe environment for them to express feelings, she said. ■

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