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AUTHOR'S NOTE

Hopkins psychiatrist William Reiner is among those calling for dramatic change in the way doctors treat babies with intersex conditions.

HEALTH AND MEDICINE

Into the Hands of Babes

By Melissa Hendricks
Illustration by Kim Barnes

Hopkins's William Reiner vividly remembers the child whose case made him rethink his approach to medicine.

"It was a little 7-year-old," says Reiner, "who just about tore everybody's guts out."

A urologist turned psychiatrist, Reiner had devoted his career to treating young patients born with irregular genitals. Some had ambiguous genitals--a scrotum along with a phallus that resembled a clitoris, for example. Others, though genetic males, had no penis or had an extremely rare condition called micropenis.

Physicians for several decades had recommended that children in this latter group be raised as girls--thinking based largely on pioneering work done in the 1950s and '60s at Hopkins, one of the few medical centers in the world that specialized in treating children with such disorders. Common practice in a case of a boy who lacked a penis or had a micropenis was to surgically remove the testes, construct female external genitals, and prescribe female hormones--then send the child home to be brought up as Sally or Susie. To do otherwise, it seemed, was to subject these babies to an unimaginably cruel childhood of locker room taunts and psychological pain, followed by a frustrating adulthood of sexual inadequacy.

Gender reassignment occurred in other cases too, depending on the diagnosis. But in all cases, the standard was to decide promptly whether the infant ought to be a boy or girl and then to be consistent in rearing the child as that gender (a process described in my story "Is It a Boy or a Girl?" in the November 1993 Johns Hopkins Magazine).

Reiner had wholeheartedly subscribed to this model throughout the 1970s and '80s, first as a urology resident at Hopkins and then as a urologist in private practice in central California. But over the course of his career, he had become troubled by a dearth of information on the psychological and sexual outcome of children with urogenital conditions.

In 1992, he returned to Hopkins to train as a psychiatrist and to specialize in treating patients with urogenital disorders. He now directs Hopkins's Gender Identity and Psychosexual Disorders Clinic. Recently, he concluded the first phase of an outcomes study that included 36 genetic males who had been born with a complicated birth defect that included the lack of a

So the next day, Reiner explained to Kayla that she had been born a boy who had no penis, so her doctors and parents had decided to raise her as a girl.

"His eyes opened about as wide as eyes could open," recalls Reiner. "He climbed into my lap and wrapped his arms around me and stayed like that."

As Reiner cradled the child in his arms, he felt as though an enormous weight had been lifted, and he himself was overcome with emotion. The child remained in his arms without moving for half an hour.

Reiner: "The sense of who one is--[boy or girl]--is powerful and inborn."
Photo by Keith Weller.

Reiner now believes children are born either boys or girls, and that no matter what happens to them, be it surgery or rearing, they remain that way.

"When you work with these kids, you see that they're not making a decision," he says. "They have always known. The sense of who one is--[boy or girl]--is a crucial existential aspect of humanity. It is powerful and inborn." The absence or presence of a penis is incidental. "The most important sex organ is the brain."

Reiner now says that surgeons ought to hold off on surgically castrating patients like Kayla. Further, he suggests that the same cautious approach should perhaps apply to patients with other urogenital conditions. "One can draw inferences, but one has to do it with caution," he notes, given that having ambiguous genitals stems from myriad causes. "But if it's not life threatening, I would favor prudence." If a child has ambiguous genitals, he says, families and physicians might decide using the best medical information available whether to call them boys or girls, but wait for the child to decide whether to opt for genital surgery.

In the past year, he has reported his findings at several medical meetings including the Lawson Wilkins Pediatric Endocrinology annual meeting, which was held in Boston this past May.

A number of other physicians and psychologists who have treated patients with ambiguous genitals and other urogenital conditions (now often referred to collectively as intersex disorders) say they, too, have changed their views. One of them is Philip Gruppuso, a pediatric endocrinologist at Brown University School of Medicine. "In the absence of definitive data [on outcome], the most compelling thing to do is the

In trying to make a patient like herself conform to standards of normalcy, Chase contended, physicians sacrificed the patient's sexuality. "The constant invasion of the child's body is emotionally traumatizing, and the surgeries create scarring and destroy sensation," she wrote to me, back in February 1994. "In the absence of a painful or life-threatening condition, genital surgery must never be imposed on a person who cannot weigh the trade-off of erotic function and make his or her own decision."

Chase signed her letter as director of the Intersex Society of North America (ISNA), a patient support and advocacy group that she had founded. Since that time she has mobilized other intersexuals who were dissatisfied with the treatment they had been given as children, begun publishing a newsletter, and started a website. Recently, Chase and I exchanged emails and spoke by phone. "There's been tremendous movement since we last spoke," she said. She was encouraged that a number of physicians were advocating reform, and had been invited to speak at the pediatric endocrinology meeting in Boston. ISNA had helped reduce the shame and secrecy that has been the intersexual's lot, she said, but still had work to do.

However, the question of what is best for the intersex baby is a contentious one. Not everyone believes the model of care should change, and many specialists are reserving judgment. Even within Hopkins there is a range of opinions on what is most appropriate.

A recurring question in the discussion about the medical management of intersex children is how chromosomes, hormones, anatomy, neurons, family, and society blend together to determine gender.

Fifty years ago, in the flush of behaviorism, a young Hopkins psychologist named John Money proposed that nurture could take precedence over nature in the recipe for gender. The man who would go on to define the terms gender role and gender identity ran a clinic in which he counseled patients with a variety of intersexual conditions--he referred to them as hermaphrodites-- and he based his theory in part on psychological studies of those patients. In a paper published in the June 1955 Bulletin of the Johns Hopkins Hospital, Money concluded:

"Chromosomal, gonadal, hormonal, and assigned sex, each of them interlinked, have all come under review as indices which may be used to predict an

was not an intersexual per se, her case lent support to the practice of gender reassignment in those patients.

But in 1997, Milton Diamond, a biologist at the University of Hawaii; and Keith Sigmundson, a psychiatrist who had treated Brenda Reimer, dropped a bombshell. They reported in the Archives of Pediatrics and Adolescent Medicine that Brenda had rejected her assigned gender and exhibited boyish behavior throughout childhood and adolescence, even though she was not told the truth about her gender transformation until she was 14. After learning the truth, Brenda had reclaimed her male role, renaming herself David.

David Reimer eventually had surgery to reconstruct a penis and to remove breasts that he had developed as a result of prescribed estrogen therapy, according to the book As Nature Made Him, by journalist John Colapinto (HarperCollins, 2000). Reimer now works in a slaughterhouse, is married, and has adopted his wife's three children.

The fall-out for Money has been harsh. Several of his former colleagues told me that Money was a pioneer in the study of gender who advanced the science and clinical care of intersexuals, and helped to destigmatize their conditions. But they added that he also staked his career on a single experiment, and never acknowledged that he lost the gambit, with devastating consequences for Reimer. Money declined to be interviewed for this article, citing patient confidentiality.

A few years after William Reiner came to Hopkins to train in psychiatry, he began a collaboration with Hopkins's John P. Gearhart, who directs the Department of Pediatric Urology.

Gearhart's interest concerned patients with cloacal exstrophy, an extremely rare and severe birth defect that occurs once in only about 250,000 live births. Children with the condition are born with their bowel, bladder, and certain other organs exposed to the outside. Many suffer from serious spine problems and spend their lives in a wheelchair. Genetic males with the condition have testicles but no penis. Generally, these patients have been surgically castrated and raised as girls.

Genital Variations

Until relatively recently, many children with cloacal exstrophy did not survive past infancy. But their

fused labia. Treatment reduces androgen levels. The most severe form is fatal if not treated. CAH is the most common cause of ambiguous genitals, occurring in an estimated one in 10,000 to 15,000 births.

Androgen insensitivity: Babies have XY chromosomes and testes, but due to a genetic mutation, their cells cannot respond to androgens. The result is external female genitals. At puberty, androgen insensitive girls do not spontaneously menstruate. In partial androgen insensitivity, cells partially respond to androgens, causing the development of ambiguous genitals.

Gonadal dysgenesis: Baby has XY chromosomes but gonads are abnormally formed. External genitals are female. Internally, female structures develop. In partial gonadal dysgenesis, babies have XY chromosomes, and testes partially form. External genitals are ambiguous, and internal structures may be a combination of masculine and feminine.

Micropenis: External genitals are masculine, but penis is extremely underdeveloped, perhaps as a result of deficient androgens in later fetal development.

Klinefelter syndrome: Boys have an extra chromosome, or XXY karyotype. External genitals are masculine, but testes are often small. Breasts may develop at puberty. Masculine development, such as growth of the penis and facial hair, may be diminished. Sperm count may be

and development, and a parent interview, Reiner found that the children who had been raised as girls displayed male-typical behavior. "They don't wear dresses," says Reiner. "They engage in rough and tumble play. They tend to fight with boys, using their wheelchairs as a weapon. If they have Barbies, they use them as though they were GI Joes, put them in cars and bash them." When the children were asked what they wanted to be when they grew up, they chose male-dominated career roles such as an astronaut or race car driver.

Those raised as girls, says Reiner, "had terrible genital self-esteem." The older children were not dating. "If you pin them down, they say they're attracted to girls. But it's not acceptable to them to be homosexual."

The psychological analysis also revealed that eight of the children were clinically depressed. Two had attempted suicide. In contrast, neither of the two patients who had been raised as boys had experienced these psychological problems.

Twenty-two of the patients who were raised as girls have reassigned themselves as males.

"I'd advise that these kids should be assigned a gender. But they should be watched closely and seen regularly by medical professionals," says Reiner. "If at age 5, 6, or 7, they want to change gender, [they should be allowed to.]"

Continuing his study through a $780,000 grant from the National Institute of Mental Health, Reiner periodically counsels patients who have made the transition from girl to boy. He is sometimes called in to explain to schoolchildren how and why their classmate made that switch. The children, he has found, often tell him they already knew that their classmate was a boy. Says Reiner, "Kids often ask, 'How could the doctors ever make that mistake?'"

But not everyone within the medical community agrees that postponing cosmetic genital surgery should apply in all cases.

At Hopkins, some standards have changed, says Gearhart. Today a child with micropenis is unlikely to undergo gender conversion, for instance. Surgeons are also more reluctant to perform surgery to reduce the size of a baby's clitoris. "We tend to let them grow into their clitoris," says Gearhart.

phallus that resembled a penis into feminine genitals. "I put myself in my child's position and thought it would be easier socially for a child--going to the physician, taking gym class, whatever--to strive for as normal a life as any child could have," says the mother. "Someday she might come back and say, 'Why didn't you let me make the decision?'" But that was a risk she and her husband decided they had to take.

Migeon is currently conducting his own follow-up studies of patients with a number of manifestations of intersexuality. His study population includes 85 people ranging in age from 21 to the mid-60s. The patients have a variety of conditions including ambiguous genitals, micropenis, and androgen insensitivity, in which a woman has XY chromosomes but is genetically incapable of responding to male hormones. Some of the patients were assigned a gender that differed from their biological gender; for example, the group includes a half-dozen patients born with micropenis who were raised as women.

Preliminary analysis of the data, says Migeon, indicates that almost all of the patients are content with the gender in which they were raised. All but two showed a gender identity and gender role in accordance with their gender of rearing.

The results would seem to contradict Reiner's findings, acknowledges Migeon's collaborator, psychologist Amy Wisniewski. But the two groups of patients are very different. Reiner's patients at birth were boys in every way, except for the lack of a penis. But hers and Migeon's are more complicated and were probably exposed to lower levels of male hormones in utero, meaning the distinct line between male or female is murkier. Further, unlike the children in Reiner's studies, theirs are adults. "Possibly our people by adulthood have come to accept their gender," says Wisniewski.

Other experts, like Sheri Berenbaum, a psychologist and professor of physiology at Southern Illinois University School of Medicine, agree that the unfolding gender story is a complicated one.

Berenbaum has spent the last 15 years studying girls with congenital adrenal hyperplasia (CAH). This genetic disease causes the adrenal glands to produce excess androgens--the steroids that cause masculinization--a process that begins in the womb

Concludes Berenbaum, "I think the story on gender identity has yet to be told."

Over the past decade, the diagnosis and treatment of genital disorders has advanced dramatically. When Migeon first arrived at Hopkins, serving as a fellow under renowned pediatric endocrinologist Lawson Wilkins, scientists had not even identified the X and Y sex chromosomes. In the following decades, they have developed and improved hormone assays and imaging techniques, which can be used to diagnose intersex conditions; pinpointed genes involved in sex differentiation; and developed new reconstructive techniques for fashioning a penis out of transplanted skin and muscle and a plastic stiffening rod.

So now, debating what to do with a case like John/Joan's is a moot point, says Hopkins's Gearhart. Today, a child who tragically loses his penis would be raised as a boy, and would undergo penile reconstruction surgery at age 10 to 15. "Nobody in the U.S. is going to change his gender," says Gearhart.

Because so much has changed, clinicians who have built careers on treating intersexuals advise against judging past events through "year 2000" glasses. "We look through the retrospectoscope and say, 'My God! How did we do that?'" says Mel Grumbach, a pediatric endocrinologist who was a fellow at Hopkins in the early 1950s and is now a professor emeritus at the University of California at San Francisco. "It's not fair. A lot has changed since then. We must learn from the advances that have been made rather than point fingers."

Specialists who care for intersex children are clearly grappling with the model of care issue, says Ian Aaronson, a urologist at the Medical University of South Carolina and founder of NATFI.

"There are presently two points of view," says Aaronson. "The first--do nothing, let patients assign themselves, no surgery--is held by psychiatrists who have had to deal with patients who have, in retrospect, been badly handled--John/Joan, for example. This is countered by many physicians who believe, all in all, their patients are doing quite well."

NATFI plans to establish guidelines for the medical management of children born with ambiguous genitals but not until first completing multicenter follow-up studies of a range of intersex patients to determine