A Question of Gender

By Emily Nussbaum

Photos by Lynn Johnson

Meet Emma, one of 65,000 babies born each year neither male nor female. Once surgeons made them into females. Now parents wonder if these infants shouldn't be left untouched.

When Emma McDonald was born on September 15, 1998, doctors whisked her away so quickly her family barely had a chance to look at her. For hours, they waited anxiously for news. Then Emma's grandmother, Anita Jones, overheard a doctor speaking to a group of medical students. Alarmed, she hurried back to her daughter. "Vicki," she said, "that doctor called Emma a hermaphrodite."

Vickie and Charles McDonald were shocked. They had turned to an open adoption after pursuing infertility treatments for 18 years, and the birth mother had agreed to relinquish custody at the hospital. Now they had to decide whether to take home a baby that was neither male nor female.

Months later, Vicki, 43, played happily with Emma. "Hey, punkadoodle," she said as she changed Emma's diaper in the kitchen of the McDonald's Gainesville, Florida, home, revealing the child's ambiguous genitals. Her phallus was larger than a clitoris, but it lacked many of the structures of a penis and was bound down by foreskin. Emma had one testicle, no vaginal opening, and an enlarged urethral hole located where a vagina would have been. The structures looked lumpy and unfamiliar, but not startling. Within Emma's abdomen are a rudimentary uterus, one fallopian horn, and an undeveloped gonad. Her chromosomes, Vicki explained, are "mosaic," a pattern called XY/XO.

Intersexuality, in a variety of forms, occurs in about one of every 2,000 births – about the same proportion as cystic fibrosis. Sex, in reality, is more than the simple blueprint learned in high-school biology – XX for female, XY for male. All embryos are identical for the first eight weeks of gestation, and then several factors nudge the infant toward male or female development. But some embryos step off track.

The cause can be chromosomal or hormonal. Infants with androgen insensitivity syndrome, for example, have XY cells but cannot process testosterone and they look like females. An inherited condition called 5-alpha-reductase deficiency triggers an apparent female-to-male sex change at puberty. Congenital adrenal hyperplasia – the most common intersexual condition – results from hormonal imbalances that masculinize the genitals of XX children. Scientists speculate that such an imbalance may also masculinize the brain, establishing gender and sexual orientation.

Intersexual infants range from hard-to-classify children like Emma to those with much subtler anomalies. To some degree, intersexuality is in the eye of the medical beholder: A large clitoris may be considered normal by one doctor, ambiguous by another. One thing all intersexual children have in common, however, if that modern medicine regards them, in the words of the surgical training videotape *Surgical Reconstruction of Ambiguous Genitalia in Female Children*, as a "social and psychological emergency." Surgeons typically perform plastic surgery early on to protect the child – and, not incidentally, the parents – from any sense of ambiguity.

Nearly all intersexual babies are assigned to be female, because the surgical techniques are better. In Emma's case, doctors planned to remove her testicle, shift her urethral hole, and carve a

labia and clitoris from existing tissue. The medical team said that Emma would look as much as possible like a normal girl after the operation. Nonetheless, on the eve of the surgery, Vicki remained in deep conflict – especially about the clitoroplasty, a procedure she feared would harm her daughter's future sexual sensitivity. "This child," she said quietly, "is perfect as she is. She was sent to us by God."

If Emma had been born only a few years ago, Vicki might not have been so tormented. She might not have been told many details about Emma before the infant went into surgery. American medical protocols were established in the 1950s: Assign a sex, operate, and shield the family from the notion that their baby's gender is in question. Typically, a medical team – made up of a surgeon, an endocrinologist, a geneticist, and more recently, a psychologist – reached a committee decision on sex assignment, then informed the parents that their child had a correctable deformity.

That approach was given credence by psychologist John Money of John Hopkins University. Money maintained that gender was plastic until age 2. In 1972 he published a book called Man & Woman, Boy & Girl on the John/Joan case, which appeared to prove his theory. John, a male twin, lost his penis in a circumcision accident, and his sex was changed to female. Money claimed Joan grew up happily feminine. But in 1994, biologist Milton Diamond tracked down Joan and discovered that in her early 20s she had chosen a sex-change operation back to male. Gender, it seemed, was not so easily manipulated.

The John/Joan revelations dumped fuel on an ethical fire that flared up in 1993, when an adult intersexual named Cheryl Chase, now 43, founded the Intersex Society of North America, an organization that has grown to 1,500 members. The group lobbies against unnecessary genital surgery on infants and maintains that current medical protocols are based on stereotypes: Maleness is equated with penis size, femaleness with fertility. Surgery on intersexual infants, they say, sacrifices the sexual sensitivity of babies assigned female. They also claim that medical secrecy intensifies the stigma it is intended to protect against. Therefore, the Intersex Society advocated a non-interventionist approach: No surgery unless medically necessary and full disclosure to parents and ultimately the child. An intersexual child, they say, could be raised with a regular gender identity – sex-stereotypical clothing, name and hair – without altering the genitals. With therapy and a supportive family, an intersexual could make his or her own decision whether to choose cosmetic procedures – or not.

Although only a dozen doctors have contacted Chase and sided with the group, the treatment of intersexual infants remains essentially the same as it was 50 years ago, last year several journals – including the *Journal of Urology* and the *Journal of Clinical Ethics* – published articles on the debate, examining ethical concerns and the lack of follow-up data on patients. The *Journal of Clinical Ethics* included essays written by adult intersexuals in its special issue on the subject.

"I was born and raised in the South, and you respect authority," says Vicki. "A doctor is an authority figure. I've had to fight against just blindly saying, 'OK, OK, OK.'" During the first confusing days after the diagnosis, Vicki and her mother immersed themselves in research, eventually contacting Chase through the Intersex Society's Web site. At their first meeting with Emma's doctors, Vicki suspected the worst. "I was like, 'You're not touching my baby!'" she recalls. But she found herself wavering. The doctors openly acknowledged the baby was intersexual. They discussed the debate about gender imprinting – the concern that Emma might feel male because hormonal exposure had already masculinized her brain. The surgeon, Dixon Walker, impressed Vicki as being open to new ideas: "He said, 'There's a group of people who believe that we're doing the wrong thing. In 30 years we may find out they're right, but for now, this is the best we know how.'"

Walker said surgical procedures had improved, and Emma would have an easier life as a female. Current techniques couldn't give her a working penis, and she wouldn't require an operation to create a vagina until she was sexually active. Vicki was relieved, because she had read that the dilations required to keep such a vagina open were traumatic for children. Psychologist Suzanne Johnson warned the family that without cosmetic surgery Emma might suffer from gender confusion and reassured Vicki that she knew well-adjusted girls who had received such operations. Finally, the doctors suggested doing all the procedures – medical and cosmetic – at one time, thereby sparing Emma multiple surgeries.

Vicki scheduled the surgery for April, telling herself she could still cancel. Perhaps, she thought, Emma might have the best of both worlds: an operation to protect her from scorn, coupled with family honesty. Emma's knowledge of her adoption might be beneficial: She could know she had been chosen despite her unusual body and would understand it was possible to be a mother without giving birth. Still, as the date of the surgery neared, Vicki's conflict grew. She had spoken to a half-dozen adult intersexuals who had been operated on as children. Many were deeply unhappy with the results, and most wished they could have made their own decision about surgery. One intersexual she spoke to chose surgery as an adult and did not regret it – but even she had urethral scarring. Emma's doctor had not put Vicki in contact with any of the content postoperative intersexuals they had told her about, saying their former patients were unwilling to talk or that they had lost track of them. Vicki wanted reassurance from at least one adult intersexual who was happy with his or her childhood surgery. She would never get it.

In early April, Vicki sat on the porch swing of her mother's house, ticking off the procedures in Emma's upcoming operation. First, the surgeon would remove the gonad in Emma's abdomen and then her descended left testicle. Emma's enlarged urinary hole would be made smaller and moved upward, leaving space for a future vagina. Finally, Walker would perform the clitoroplasty, an operation to cut down her phallus to the size of a typical clitoris.

Vicki learned all she could about the operations. Chase, the Intersex Society founder, had sent her a 1990 medical training video of the clitoroplasty; Vicky wept all the way through it. She now knew that in the three-hour operation, Emma's phallus would be split open so that doctors could remove the spongy tissue of the shaft. The tip, still attached to its blood supply and nerve endings, would be trimmed, a wedge of skin removed, and the sides pulled together. This structure would be sewn into place as a clitoris and be surrounded by a labia minora, made of foreskin, and a labia majora, made from Emma's hair-bearing scrotal skin.

Only the clitoroplasty, Vicki emphasized, would be cosmetic. Emma's testicle and gonad might have precancerous tissue and must be removed. The size and position of her urethral hole put her at risk for urinary infections. Were that procedure solely cosmetic, she added, she would not have given consent. But Vicki recognized that the clitoroplasty was her decision – one which put her in the peculiar position of safeguarding her daughter's future sex life: "The doctor said that the surgery preserves the blood supply and nerves, so she will be able to have an orgasm." Anita wondered whether, even if Emma's responsiveness is reduced, she would be able to tell the difference. Wouldn't it be more difficult to have a sensitive but "weird" set of genitals, which might make it too embarrassing to wear a bathing suit, let alone be naked with another person?

Still, the lack of follow-up data on sexual sensitivity worried Vicki: "As parents, we're often forced to make decisions for our children that are hard. And this is one of 'em, as far as I'm concerned." If Emma has regrets, Vicki said, she'll be able to blame her family instead of herself.

But Vicki's worst nightmare was that Emma might grow up and identify as male. In that case, it would be too late.

The day before the operation, Vicki's extended family gathered for Chinese takeout. The mood was raucous: on-going debates have rattled each family member's assumptions about gender: "What makes me female anyway?" asked Anita. "I don't feel like a woman every minute of the day." Cecil, Vicki's once-traditionalist father, talked about "third-gender identity" and blurted out, "My God, wouldn't it be fantastic if Emma wound up being a lesbian!" Instead of seeing Emma's condition as something to be fixed and overcome, the McDonald's had a positive interpretation: Emma is spiritually "whole," they said, sent for a purpose.

But they doubted others would be as accepting. Charles, Vicki's husband, has searing memories of adolescence. "I never had hair under my arms in the locker room. I was uncircumcised. I was the littlest of 4,000 students. There was constant teasing." How much worse would things be for his child? "I'm realistic," said Vicki. "Sexuality under the best of circumstances is difficult for adolescents." She adjusts the baby in her arms. "But my hope is that by the time she gets there, she'll have such a secure faith in her worth that it won't have the devastating effect on her that it had on some of the older hermaphrodites who are just so angry and bitter."

On the day of surgery, while Emma lay on the operating table, Vicki met with Helena Harmon-Smith, the head of a support group for the families of intersexual children and the mother of Patrick, a child born with a condition much like Emma's. Unlike Vicki, she disagreed with her doctors sex assignment and is suing the medical team for removing Patrick's one testicle without consent. As the two mothers talked, Patrick crashed around the room, shouting, "You're under arrest," and pretending to handcuff people. Vicki is hoping Emma and Patrick will be friends.

Immediately after the operation, Vicki and Charles were called to the recovery room. The surgery had gone beautifully, Walker said. Although Emma would have to catheterized for several days, there were no complications. For the next 48 hours, the family took shifts watching the baby. Vicki rocked her daughter as Emma shuddered with urinary spasms, her belly quaking. Morphine helped, but when it kicked in, Emma scratched wildly and her eyes rolled from side to side. Vicky stroked Emma's palm, crooning, "Yes, you can tell me about it. It's terrible, it hurts."

That night, when the doctors removed Emma's diaper, the area was raw and inflamed. "Everybody says it looks great," Vicky said doubtfully, "but it looks awful to me." But she was relieved that it was all over. The nurses reassured her that each day Emma would get a little better, and after midnight, Vicky climbed into the metal crib to curl up with her. She sang a song she'd made up just after Emma was born, to the tune of an old Jerome Kern standard: "Emma, be you girl or boy, you're my pride and joy – no need to be coy, because I love you. Just the way you are tonight."

The day after the surgery, Walker, 60-ish and cowboy-sinewy, with a folksy manner, said he had never read any materials criticizing current intersex protocols. He said he had seen protests at pediatric conventions: "I've always liked the idea of somebody who's the devil's advocate."

Pleased with the surgery, he characterized the cosmetic result of the clitoroplasty as "really excellent" and didn't foresee future operations. One by one, he discussed each procedure. Asked if the urinary hole had to be moved to prevent infection, he said, "No, no. If you didn't do anything to it, then basically it was not that much different from a normal female." The procedure, he noted, was cosmetic, intended to make room for a future vagina.

As a pediatric urologist, Walker said he rarely saw adults, much less the grown patients he had assigned to be female. It's hard to get good data, he added, because little is known about female sexuality. Many women, he said, are anorgasmic despite having normal genitals. For males, he said, a too-small penis may "determine how they see themselves", making them "less likely to take leadership roles." He also described the process of diagnosis for infants who are "in between." "That's a difficult group of patients," he said. "We try to get the parents involved in that decision-making, and you get all sorts of responses. There are some parents, not very sophisticated parents, from poor families, who will say 'That's the way God made 'em, and we don't want you to change anything.'" Other parents, he explained, are "more sophisticated" and "you can talk to them about making such a change."

That afternoon, Emma was released from the hospital. Vicki held her gingerly. She asked if Walker had said anything surprising. Told that he had said that moving the urethral hole was a cosmetic procedure, Vicki was distraught. "If I'd known that," she said, "there would be no catheter in this child." But by evening, Vicki had changed her mind. Walker, she said, "put more importance on the cosmetic risk; I put more importance on the risk for infection."

Later, Walker was given a speech Cheryl Chase gave to a group of doctors about her experience. She was clitoridectomized and for years was suicidal, trying to ferret out details of her condition. "It's impossible not to sympathize with a story like that," he said.

Did Walker mislead Vicki about the risk of urinary infection? No, he said: "There probably is some lack of consistency. There's not hard data on something like that." Doctors, he noted, shift their presentation depending on the patient's needs. He estimated the need for the surgery as 80 percent cosmetic, 20 percent medical. He considered the decision a necessary one: Making space for a vagina, he argued, can't be construed as cosmetic.

Back at the McDonald's, Emma has thrived. There have been no complications – no scarring, no infections. "I'm tired, chasing, picking up, feeding, twenty-four/7, but I do love it!" writes Vicki. "I'm watching her discover something new every day." Except for her regular pediatric check-ups, Emma should have no more complications until puberty. At that point, she will begin taking hormones and can make her own decision about vaginoplasty.

Will Emma grow up to be one of the well-adjusted women the doctors have told Vicki about? Or will she regret the surgery – identifying as male or feeling sexually damaged? Vicki's deepest wish is that by the time Emma is a teenager, the culture will have shifted around her, and families like theirs won't be so isolated. Going public, Vicki hopes, may be one step in that process. Before Emma's operation, Vicki said: "In an ideal world, I would do what is medically necessary and leave her alone. And she would be absolutely accepted for who she is, the special person that she will be." Then she paused. "But that's not the way our world works."

END

Sidebar article:

Chromosome Chaos

Usually we can count on the consistency of chromosomes. Every cell in a man's body has an XY pattern, while every cell in a woman's has an XX. But Emma McDonald's chromosomes are mosaic, and thus unpredictable. A cell from one part of her body could be XY, but a cell from

another part could be XO – meaning that the cell carries a lone X chromosome lacking an X or a Y partner.

This mix-up starts at conception, when a sperm cell containing half of the father's chromosomes merges with an egg containing half of the mother's. All eggs carry an X chromosome, but sperm cells tote either a matching X or a smaller, less gene-dense Y. Usually when egg X meets sperm X, the result is a girl – XX. When it meets sperm Y, it's a boy – XY.

But there are exceptions. As one fetal cell divides, a sex chromosome may become detached from the other 45 chromosomes. One daughter cell ends up with too many sex chromosomes, and the other with not enough. And, in fact, these glitches are quire common. "Sometimes the sex chromosome just doesn't make it," says Debra Saxe, codirector of the Emory Genetics Laboratory. "It's a mistake that happens normally." If the malfunction occurs soon after conception, before fetal cells differentiate, the abnormal cells can spread throughout the body. If the mosaic split comes later, such cells may appear only in parts of the body. And if a mosaic split happens to cells destined to be sex organs, the baby may be born with ambiguous genitals.

But scientists still don't know how much influence mosaic chromosomes have on the development of the intersexual baby, partly because they aren't sure how often mosaic splits occur. Testing for the condition is rare, done only after the fact. And because so few people are tested, geneticists suspect mosaic chromosomes may be far more common than we know.