case study

Culture Clash Involving Intersex

Parents from a Middle Eastern country bring their thirteen-year-old son to the hospital seeking treatment for a minor abnormality of the penis (hypospadias) and for breast development. The child has had two episodes of bleeding through the penis. The physician determines that the boy had a 46XX karyotype, both a uterus and ovaries, and severe congenital adrenal hyperplasia, which caused the child to virilize in utero. The bleeding was actually menstruation.

The physician tells the parents that a hysterectomy and an oophorectomy are necessary to prevent further bleeding. Also, one of the child's kidneys is not functioning and needs to be removed. The parents ask the pediatric urologist to perform hypospadias surgery, a bilateral mastectomy, hysterectomy, oopherectomy, and nephrectomy. Further, they want them performed all at one time (because they cannot remain in the country very long) and without involving the child in the decisionmaking or informing him of his medical condition or of his potential female reproductive capacity. The child expresses a desire to have the mastectomies performed in order to avoid teasing.

While in the United States 46XX babies with severe masculinization have traditionally been raised as females in order to preserve fertility, there has been a shift to male sex assignment for two reasons: evidence of early androgen imprinting on behavior and identity and a desire to minimize the trauma of surgery on external genitalia. Further, given that for thirteen years the child has been reared as a male and that, according to his parents, his behavior has been characteristic of his society's male gender role, he probably could not now develop a female gender identity.

The culture favors males, and the parents assert that they would have difficulty accepting their child if his gender were reassigned. They also indicate that in their society, if he turns out to be homosexual, someone would probably kill him. The physician tells the parents that the child may or may not, regardless of gender identity, develop a homosexual orientation, but that the child is too young to have a reliable opinion of his sexual orientation, especially given his ignorance of his medical condition and his culture's views about homosexuality.

The urologist consults with a psychologist knowledgeable about intersex, and both feel uncomfortable about doing the surgeries without the child's consent. The parents insist that the decision is the father's and that the father knows what is best for the child. Should the urologist comply with their wishes?

commentary

by David Diamond

The dilemma confronting the treating physicians is remarkably difficult under ordinary circumstances. In this situation it is further complicated by cultural differences.

The thirteen-year-old, pubertal patient is a chromosomal and gonadal female with a male appearance and male gender role. Following a thorough evaluation of his situation, it is apparent that his problems are far more complex than originally anticipated. There appear to be three management options for this boy. The first is to maintain (and enhance) his male appearance by repairing the penile abnormality, performing bilateral mastectomy, and removing all discordant female reproductive organs. Given the patient's age, exogenous male hormones should be started to enhance secondary sexual characteristics. This course of treatment would most closely approximate what the family had in all likelihood anticipated before the diagnosis of congenital adrenal hyperplasia with a 46XX karyotype. The second option is to assign the patient to a female gender role, convert the external genitalia to a female phenotype, and maintain the female reproductive organs. This approach would preserve the patient's fertility, whereas the first would certainly sacrifice it. A third option is to acknowledge that the patient's situation has turned out to be far more complex than anticipated and for the parents to defer treatment until the child can make the decision personally on the preferred management.

The father, assuming the responsibility of decisionmaker, has selected the first option. In so doing, he has preserved the child's gender role, thereby preserving his established status within the family and within his community. The father thereby rejects the consider-

able uncertainty of a gender reassignment, with its attendant risks for his child's role within the family and community. While we can appreciate such a risk in our own culture, the extent of the risk for this boy within his own culture is impossible for us to comprehend.

One might argue that the father's decision is supported by a "family-centered model" of autonomous decisionmaking. In this context, a higher priority is placed on harmonious functioning of the family than on autonomy of its individual members. One could imagine that a son's gender reassignment to female might well make it impossible for the family to return to its original community, making the price of such a decision for the family prohibitive.

On the other hand, the son's autonomy is sacrificed by the father's approach. He has been denied an explanation of his diagnosis and a discussion of the alternatives. The extent to which the father's approach is culturally driven is difficult to ascertain. In a previous era, such a secretive and paternalistic approach to intersex disorders was commonplace in this country, and it was justified on the basis of beneficence. However, long-term feedback from many patients has demonstrated that the veil of secrecy heightened anxiety and undermined the physician-patient relationship. Such practices would be regarded as improper today in the context of enlightened American medicine.

So what is the proper posture of the treating urologist? Given the significant cultural divide, the parents' own value system must be the guide. Such an approach would seem to combine the family-centered model of autonomous decisionmaking with a patient's best interest standard of surrogate decisionmaking by the father. As a result, a determination would be made of the net benefits among available options, incorporating quality-of-life criteria for the patient and the family. The urologist must believe that the selected treatment will benefit the child and justify the associated surgical and anesthetic risks. Without such conviction, surgical treatment by the practitioner would be inappropriate.

commentary

By Sharon Sytsma

The doctors in this case face a heartrending quandary, caught as they are in a culture clash that places their patient in a precarious situation. Acceding to the parents' wishes places the child at greater risk from the multiple surgeries performed, fails to respect the child's autonomy, irreversibly deprives the child of all procreative capacity, and puts the child at significant psychological risk. On the other hand, not performing the surgery means the child will continue to be taunted and suffer almost certain disenfranchisement and rejection, and that he quite possibly will be murdered.

Balancing the advantages against the disadvantages of performing the surgeries would seem to indicate that the physicians should agree to the parents' requests. We now know that those who have been assigned to a certain gender because of intersex conditions usually do not express a desire to change their gender as they mature. Because the child has maintained a firm male identity throughout his childhood and seems to enjoy participating in male pattern behavior, he is even less likely to assume a female gender identity and to resent the loss of his female reproductive capacity. Given the cultural bias toward males, the parental attitudes, and the apparently consistent male gender identity and behavior, the child would probably choose not only the mastectomies and kidney removal, but the other surgeries and treatment as well. Changing the boy is certainly more within our power than changing his culture, and the surgeries will make it easier for him to thrive in that culture.

However, we have learned that withholding information about intersex from children is more likely to be damaging than not. Children who are kept in the dark are made to feel freakish, alone, and fearful that they must be dying. They think of their parents as coconspirators with the doctors, causing a deep feeling of alienation. Children's trust in their parents and in the medical profession is thus often irretrievably lost. The experience of not being unconditionally loved causes lifelong psychological difficulties. Allowing the child to make the decisions to accept the greater risk of the combined surgeries is more respectful of the child's intrinsic worth; and should the child actually come to identify as female and regret the decision, at least he would not experience resentment toward his parents and the doctors. Nevertheless, this child has not been prepared for learning the truth about his condition and could very well be traumatized by it, and trauma increases the risks of any surgery. Because he must leave the country soon, he would not be able to receive sufficient counseling to enable him to recover psychologically.

Insisting that the child participate in the decision might seem to fail to recognize the right of parental autonomy and to display a lack of respect for the values of another culture. The concern is compelling, but problematic. Surely, not all cultural values are worthy of respect. We should not be morally required to set aside our own moral judgments, especially when they are backed by experience, scientific study, and ethical principles. Allowing the values of other cultures to override our own would be appropriate only when those values are morally or epistemically on par or superior to our own. In other cases, doing so would be a matter of moral abdication. A duty to respect the values of another culture cannot consist in simply deferring to those values, but only the duty to try to understand why a culture values what it values, to withhold wholesale condemnation of individuals belonging to that culture for holding such values, to be open-minded to the possibility that the values of another culture may be either equally tenable or morally superior to our own, and to refrain from imposing our own values on a culture

whose circumstances are such that doing so would lead to harm.

Clearly, the physicians here face a conflict of duties. Refusal puts the child's well-being in greater jeopardy, but performing the surgeries involves going against what we have come to see as morally required and might lead to more such requests. The physicians should deliberate with the parents and explain the advantages of allowing the child to participate in decisions about his medical treatment. The physicians involved should also attempt to educate our own public—citizens and doctors—about the importance of open communication and informed consent. The importance of full disclosure should be included in pediatric urology textbooks and should be posted on the Internet. Doing so could have the effect of dissuading parents from other cultures seeking surgery for their children without their participation.

commentary

by Alice Dreger and Bruce Wilson

From our own extensive contact with people born with intersex conditions, and from emerging follow-up reports on the care of people with intersex conditions, this is what we know: The gender identities and sexual orientations of children with (or without) intersex conditions cannot be engineered with medicine, nor can they be easily diagnosed or predicted. Surgeries designed to make children with sexual ambiguity look "normal" carry with them substantial uncertainty: they often do not achieve the aim, and they frequently result in lifelong detrimental "side effects," physical and psychological.

We also know that for decades people with intersex conditions have been exempted from the moral rules employed to protect others. A standard practice in medicine has been to actively deceive people with intersex conditions about their diagnoses and medical histories. Many have been subjected to extensive cosmetic genital surgeries while being led to believe that the surgeries were necessary for their physical survival. This unjustified double standard has harmed many people with intersex. More to the point, it is just that—a double standard—where none ought to exist. Just because intersex makes most of us uncomfortable does not mean that people with intersex should be treated with care we would otherwise consider substandard. We therefore begin and end our commentary on this case of intersex with the assertion that people with intersex are entitled to the same decency in health care as others. Even as children, they are entitled to know the truth about what is happening to them, to whatever extent they are capable of understanding, and they are entitled to make critical decisions about their bodies when they are able, particularly when there is no medical urgency.

A fundamental but common mistake has been made in the treatment of this family. The case has been understood primarily as a surgical problem, and the question is, "Should the surgeries be done?" But that is the wrong way to approach intersex. Because intersex poses a multifaceted problem, when a case is uncovered, a team approach should be implemented, with the participation of a pediatric psychiatrist or psychologist and a social worker as the keystone. Ideally, the team would also include an endocrinologist, a surgeon, a geneticist, and a primary care physician.

In the case at hand, a team could help the child and parents explore the data, the risks, and the options—many of which are not presented here because the case has been framed by a surgical mindset. A psychiatrist or psychologist would already have seen the child repeatedly and would be able to advise about his competency to participate in making decisions about the non-urgent surgeries. If the child is not ready to decide whether he wants hypospadias surgery, bilateral mastectomy, hysterectomy, and oopherectomy, then the endocrinologist could prescribe leuprolide, a once-a-month injectable hormone which would essentially stall puberty, halting the menstruation (if that's what the bleeding is), preventing further breast development, and so on. Regardless of which surgeries are done, this patient is going to need regular medical care. The case description implies that, if the surgeries are done, the patient might go on in life "cured," never having been the wiser. But in fact, this patient will need lifelong endocrinological management, regardless of the surgeries. (An oopherectomy will only increase that need.) Now is the time to start enlisting the *knowing* cooperation of this patient in his lifelong medical care.

It is more important to get it right than to be fast. There is no compelling reason to override the right to self-determination of this child, and there are many reasons not to override it. Some might argue that the cultural differences justify following the father's wishes. Nevertheless, we are unsympathetic to the idea that children's sexual anatomies are an acceptable locale for cultural relativism. In 1996, federal legislation was enacted to protect girls from a set of cultural practices known as female genital mutilation, female circumcision, or female genital cutting. The central premise of that legislation was that every minor girl is entitled to legal protection from adults who seek to surgically alter her genitals for social reasons—regardless of her family's cultural background—because, according to the law, "the practice of female genital mutilation often results in the occurrence of physical and psychological health effects that harm the women involved." People with intersex bear the same sorts of risks and deserve the same protections.

In the case before us, if even with sensitive team care the parents refuse to allow the child to be consulted about his condition and treatment, the physicians should refuse to cooperate in the deception and should, if they feel the child's well-being is at serious risk, seek legal help in protecting this child from what might amount to neglect or abuse.