Sex Reassignment at Birth: Long-term Review and Clinical Implications
[Review]

Diamond, Milton PhD; Sigmundson, H. Keith MD

From the Department of Anatomy and Reproductive Biology, Pacific Center for Sex and Society, University of Hawaii-Manoa, John A. Burns School of Medicine, Honolulu (Dr Diamond); and the Department of Psychiatric Services, Ministry of Health, Victoria, British Columbia (Dr Sigmundson).

Abstract

This article is a long-term follow-up to a classic case reported in pediatric, psychiatric, and sexological literature. The penis of an XY individual was accidentally ablated and he was subsequently raised as a female. Initially this individual was described as developing into a normally functioning female. The individual, however, was later found to reject this sex of rearing, switched at puberty to living as a male, and has successfully lived as such from that time to the present. The standard in instances of extensive penile damage to infants is to recommend rearing the male as a female. Subsequent cases should, however, be managed in light of this new evidence.

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Among the more difficult decisions physicians have to make involve cases of ambiguous genitalia or markedly traumatized genitalia. The decision as to how to proceed typically follows this contemporary advice: "The decision to raise the child as a male centers around the potential for the phallus to function adequately in later sexual relations" [1] (p580) and "Because it is simpler to construct a vagina than a satisfactory penis, only the infant with a phallus of adequate size should be considered for a male gender assignment." [2] (p1955) These management proposals depend on a theory that says it is easier to make a good vagina than a good penis and because the identity of the child will reflect upbringing and the absence of an adequate penis would be psychosexually devastating, fashion the perineum into a normal looking vulva and
vagina, and raise the individual as a girl. Such clinical advice, concerned primarily with surgical potentials, is relatively standard in medical texts [3-6] and reflects the current thinking of many physicians. [7]

This management philosophy is based on 2 beliefs held strongly enough by pediatricians and other physicians to be considered postulates: (1) individuals are psychosexually neutral at birth and (2) healthy psychosexual development is dependent on the appearance of the genitals. These ideas arise most strongly from the original work of Money and colleagues. [8-11] (pp46-51), [12] Typical pronouncements from that research include that "erotic outlook and orientation is an autonomous psychological phenomenon independent of genes and hormones and a permanent ineradicable one as well" [9] (p1397) and "[it] is more reasonable to suppose simply that, like hermaphrodites, all the human race follow the same pattern, namely, of psychological undifferentiation at birth." [10] The first postulate was derived not from normal individuals, but from hermaphrodites and pseudohermaphrodites. The second had only anecdotal support. Money [13] no longer holds such extreme views, but his involvement in this particular case and acceptance of the thesis was notable enough that it became a totem in the lay press and a classic for the academic and medical community. And, as already noted, the textbooks have not kept abreast of the new thinking.

REPORT OF A PATIENT

The case involved a set of normal XY twins, one of whom, at 8 months of age, had his penis accidentally burned to ablation during phimosis repair by cautery. [11] After a great deal of debate, the child was seen for consultation at The Johns Hopkins Hospital, Baltimore, Md, and, following the beliefs mentioned earlier, the recommendation was made to raise the child as a girl. The pseudonym John will be used when referring to this individual when living as a male and the pseudonym Joan when living as a female. Orchiectomy and preliminary surgery followed within the year to facilitate feminization. Further surgery to fashion a full vagina was to wait until Joan was older. This management was monitored and reinforced with yearly visits to The Johns Hopkins Hospital. The treatment was described as developing successfully with John accepting life as Joan. [11]

"Although the girl is not yet a woman, her record to date offers convincing evidence that the gender identity gate is open at birth for a normal child no less than for one born with unfinished sex organs or one who was prenatally over or underexposed to androgen, and that it stays open at least for something over a year after birth." [12] (p98)

A follow-up stated: "The girl's subsequent history proves how well all three of them (parents and child) succeeded in adjusting to that decision." [12]

The effects of such reports were widespread. Sociology, psychology, and women's study texts were rewritten to argue that, as Time magazine (January 8, 1973) reported,

"This dramatic case... provides strong support... that conventional patterns of masculine and feminine behavior can be altered. It also casts doubt on the theory that major sex differences, psychological as well as anatomical, are immutably set by the genes at conception."

Lay and social science writings still echo this case as do medical texts. [3-6,14] The following quote is typical:

"The choice of gender should be based on the infant's anatomy, not the chromosomal karyotype. Often it is wiser to rear a genetic male as a female. It is relatively easy to create a vagina if one is absent, but it is not possible to create a really satisfactory penis if the phallus is absent or rudimentary. Only those males with a phallus of adequate size that will respond to testosterone at adolescence should be considered for
Our current article challenges those reports and advice. It is based on a review of the medical clinical notes and impressions of therapists originally involved with the case and on contemporary interviews. One of us (H.K.S.) was head of the psychiatric management team to which the case was referred in the patient's home area. Although the patient was assigned to the immediate care of female psychiatrists to foster female identification and role modeling, H.K.S. maintained direct supervisory control of the case. The unique character of this case attracted the attention of the British Broadcasting Co and they invited M.D. as a consultant. In 1994 and 1995, we collaboratively reinterviewed and recorded John, his mother, and his wife to provide updated accounts of his progress. Findings are listed in chronological order under the appropriate postulate for pediatric sexual assignment. John himself, while desiring to remain anonymous, strongly desires his case history be made available to the medical community to reduce the likelihood of others suffering his psychic trauma.

**POSTULATE 1: INDIVIDUALS ARE PSYCHOSEXUALLY NEUTRAL AT BIRTH**

Mother recalls:

"As soon as he had the surgery, the doctor said I should now start treating him as a girl, doing girl things, and putting him in girl's clothes. But that was a disaster. I put this beautiful little dress on him... and he [immediately tried] to rip it off; I think he knew it was a dress and that it was for girls and he wasn't a girl."

On the other hand, Joan could act quite feminine when she wanted to and at approximately 6 years old was described as doing so, eg, his mother was quoted as saying: "One thing that really amazes me is that she is so feminine. I've never seen a little girl so neat and tidy as she can be when she wants to be...." More often, however, Joan rejected such behavior. More commonly she, much more so than the twin brother, would mimic the father. One incident the mother related was typical. When the twins were 4 or 5 years old, they were watching their parents. Father was shaving and mother was applying makeup. Joan applied shaving cream and pretended to shave. When Joan was corrected and told to put on lipstick and makeup like mother, Joan said: "No, I don't want no makeup, I want to shave."

Girl's toys, clothes, and activities were repeatedly proffered to Joan and most often rejected. Throughout childhood Joan preferred boy's activities and games; she had little interest in dolls, sewing, or girl's activities. Ignoring the toys she was given, she would play with her brother's toys. She preferred to tinker with gadgets and tools, dress up in men's clothing, and take things apart to see what made them tick. She was regarded as a tomboy with an interest in playing soldier. Joan did not shun rough and tumble sports or avoid fights.

John recalls when the Joan of age 8 or 9 years wanted an umbrella:

"I had a couple of bucks and went to the store to take a look at the umbrellas, and right beside the umbrellas was the toy section. I started to eyeball a machine gun. I said to myself, 'Do I have enough money for that?'... I put the gun on the counter and asked the clerk if I had enough money. She had that look like 'You don't have enough but we'll let you go anyway.' I used it to play army with my brother."

Mother recalls Joan was in a dress at the time. The brother often refused to let Joan play with his toys, so she saved her allowance money and bought a truck of her own.

Joan's realization that she was not a girl jelled between ages 9 and 11 years. John relates:
"There were little things from early on. I began to see how different I felt and was, from what I was supposed to be. But I didn't know what it meant. I thought I was a freak or something.... I looked at myself and said I don't like this type of clothing, I don't like the types of toys I was always being given. I like hanging around with the guys and climbing trees and stuff like that and girls don't like any of that stuff. I looked in the mirror and [saw] my shoulders [were] so wide, I mean there [was] nothing feminine about me. I [was] skinny, but other than that, nothing. But that [was] how I figured it out. [I figured I was a guy] but I didn't want to admit it. I figured I didn't want to wind up opening a can of worms.""

Joan knew she already had thoughts of suicide caused by this sort of cognitive dissonance and did not want additional stress.

Joan fought the boys and the girls who were always "razzing" her about her boy looks and her girl clothes. She had no friends; no one would play with her. "Every day I was picked on, every day I was teased, every day I was threatened. I said enough is enough.... " Mother relates that Joan was good looking as a girl. But, "When he started moving or talking, that gave him away, and the awkwardness and incongruities became apparent."

The other girls teased Joan so aggressively that she often retaliated forcefully. At age 14 years one girl sat behind Joan and continued to hit her. The adult John demonstrated: "I grabbed her like that, by the shirt, and rammed her round the wall like this, threw her on the ground... until the teacher grabbed me." This resulted in Joan being expelled from school.

Despite the absence of a penis, Joan often tried to stand to urinate. This made a mess as it was difficult to direct the urine stream. Although she learned to sit and void, she nevertheless continued to occasionally stand and urinate. Despite admonitions against the behavior and its untidiness, Joan persisted. At school, at age 14 years, she was caught standing to urinate in the girls' bathroom so often that the other girls refused to allow her entrance. Mother recalls the other girls threatening to "kill" her if she persisted. Joan would also sometimes go to the boy's lavatory to urinate.

Joan was put on an estrogen regimen at the age of 12 years but rebelled against taking the hormones. They made her "feel funny" and she did not want to feminize. She would often dispose of her daily dose. She was unhappy at developing breasts and would not wear a bra. Things came to a critical point at age 14 years. In discussing her breast development with her endocrinologist she confessed, "I suspected I was a boy since the second grade." The physician, who personally believed Joan should continue her medication and proceed as a girl, used that opening to explore the possible male and female paths available and what either one would mean. The local psychiatric team had noticed Joan's preference for boy's activities and refusal to accept female status so they already had discussed among themselves the possibility of accepting Joan's change back to a male. The endocrinologist explored Joan's options with her. Shortly thereafter, at age 14 years, Joan decided to switch to living as a male.

Joan was the daily butt of her peers' jibes and the local therapists, having knowledge of her previous suicidal thoughts, went along with the idea of sex re-reassignment. In a tearful episode following John's prodding, his father told him of the history of what had transpired when he was an infant and why. John recalls: "All of a sudden everything clicked. For the first time things made sense and I understood who and what I was."

John requested male hormone shots and gladly took these. He also requested a mastectomy and phalloplasty. The mastectomy was completed at the age of 14 years; surgical procedures for phallus construction were at ages 15 and 16 years. After the surgical procedures, John adjusted well. As a boy he
was relatively well accepted and popular with boys and girls. At 16 years, to attract girls, John obtained a windowless van with a bed and bar. Girls, who as a group had been teasing Joan, now began to have a crush on John. When occasions for sexual encounters arose, however, he was reluctant to move erotically. When he told 1 girlfriend why he was hesitant, that he was insecure about his penis, she gossiped at school and this hurt John very much. Nevertheless, his peers quickly rallied around him and he was accepted and the girl rejected.

John's life subsequently was not unlike that of other boys with an occult physical handicap. After his return to male living he felt his attitudes, behaviors, and body were in concert in a way they had not been when living as a girl. At age 25 years he married a women several years his senior and adopted her children.

**POSTULATE 2: HEALTHY PSYCHOSEXUAL DEVELOPMENT IS INTIMATELY RELATED TO THE APPEARANCE OF THE GENITALS**

First in Baltimore and then with the local therapists prior to the sex re-reassignment, Joan's expressed feelings of not being a girl would draw ridicule. She would be told something such as: "All girls think such things when they're growing up." John recalls thinking: "You can't argue with a bunch of doctors in white coats; you're just a little kid and their minds are already made up. They didn't want to listen." To ease pressures to act as a girl, Joan would often not argue or fight the assignment and would "go along."

Beginning at age 7 years, Joan began to rebel against going for the consultations at The Johns Hopkins Hospital. Her reasons were discomfort and embarrassment with forced exposure of her genitals and constant attempts, particularly after the age of 8 years, to convince her to behave more like a girl and accept further vaginal repair. This was always strongly resisted and led to recurrent confrontations. To temper Joan's reluctance to travel to the consultants, her parents combined such visits with vacation trips.

In Baltimore the consultants enlisted male-to-female transsexuals to convince Joan of the advantages of being female and having a vagina constructed. She was so disturbed by this that in one instance Joan, at age 13 years, ran away from the hospital. She was found hiding on the roof of a nearby building. After age 14 years, Joan adamantly refused to return to the hospital. Joan then came fully under the care of local clinicians. This group consisted of several pediatricians, 2 pediatric surgeons, an endocrinologist, and a team of psychiatrists.

John recalls thinking, from preschool through elementary school, that physicians were more concerned with the appearance of Joan's genitals than was Joan. Her genitals were inspected at each visit to The Johns Hopkins Hospital. She thought they were making a big issue out of nothing and they gave her no reason to think otherwise. John recalls thinking: "Leave me be and then I'll be fine.... It's bizarre. My genitals are not bothering me; I don't know why it is bothering you guys so much."

When asked what Joan thought of her genitals as a youngster, John replied, "I didn't really have anything to compare myself against other than my brother when we were taking a bath." Mother confirmed that as a devout family in a very conservative religious community there would have been few opportunities for the twins to have seen anyone else's genitals. Nudity was never acceptable. At their yearly visit to The Johns Hopkins Hospital, the twins were made to stand naked for inspection by groups of clinicians and to inspect each other's genitalia. This experience, in itself, was recalled with strong negative emotions. John's brother, decades later, recalls the experience with tears.

John recalls frustration, which remains, at not having his feelings and desires recognized. Without consideration of genitals, despite the obvious absence of a penis, Joan nevertheless knew she was not a girl. When she tried to express such thoughts the physicians would change the subject. "[They] didn't want to
hear what I had to say but wanted to tell me how I should feel." Clinical notes from the time report Joan saying she felt "like a trapped animal."

In middle school Joan had difficulty making friends. Her clothes and demeanor did not jibe. Because of her behavior, her peers teased her with names like "caveman" and "gorilla." Few children would play with her. None of Joan's peers knew anything of her genitals.

At first, as suggested by the consultants from The Johns Hopkins Hospital, the local physicians and her parents continued to treat Joan as a girl, preparing her for vaginal reconstructive surgery and life as a female. Psychotherapy, primarily by female therapists, was aimed at reinforcing her female identity and redirecting her male ideation. This course of action became increasingly difficult because of Joan's growing conviction that she was not right as a girl and anger at being treated like one. Joan's reactions were not unlike those in posttraumatic stress disorder, where the cause of the stress is not remembered. John recalls, "They kept making me feel as if I was a freak."

John knew what the clinicians wanted and recognized it was not what he wanted. Beginning at age 14 years, against the recommendations of the clinicians and family and without yet knowing of the original XY status, Joan refused to live as a girl. Jeans and shirts, because of their gender-neutral status, became her preferred manner of dress; boy's games and pursuits her usual activities. Joan's daytime fantasies and night dreams during elementary school involved seeing herself "as this big guy, lots of muscles and a slick car and have[ing] all kinds of friends." She aspired to be a mechanic. She rejected requests to look at pictures of nude females, which she was supposed to emulate. Rorschach and Thematic Apperception Tests at the time elicited responses more typical of a boy than a girl. Her adamant rejection of female living and her improved demeanor and disposition when acting as a boy convinced the local therapists of the correctness of sexual re-reassignment.

Following the surgery for penile construction there was difficulty with urethral closure. Despite repeated attempts at repair, the problem was never rectified. John now urinates through a fistula at the base of his penis while sitting down. Much of the penis is without sensation, as are the areas of scarring from where the grafts were taken.

John's first sexual partner was a girl. He was 18 years old. While living as a girl and afterward as a boy, John was approached sexually by males. He claims never to have been attracted to them and his responses to such questions are matter-of-fact and not homophobic. John thinks his first recognizable sexual interest occurred at about age 16 or 17 years, although he did recall wanting to go see the "sexy" Rockettes in New York on one of his trips to see the consultants.

Coitus is occasional with his wife. This frequency, he claims, is sufficient for his needs but is less than his wife would desire. They mostly pleasure each other with a great deal of physical affection and mutual masturbation. John can have coital orgasm with ejaculation.

John recalls thinking it was small-minded of others to think all his personality was summed up in the presence or absence of a penis. He expressed it thus:

"Doctor... said, 'it's gonna be tough, you're going to be picked on, you're gonna be very alone, you're not gonna find anybody [unless you have vaginal surgery and live as a female].' And I thought to myself, you know I wasn't very old at the time, but it dawned on me that these people gotta be pretty shallow if that's the only thing they think I've got going for me; that the only reason why people get married and have children and have a productive life is because of what they have between their legs.... If that's all they think of me, that they justify my worth by what I have between my legs, then I gotta be a complete loser.'"
GENERAL COMMENTS

As an adult John was asked why he did not accept being a female rather than fighting it. His answer was simple. Doing so did not feel right. He wanted to please his parents and placate the physicians so he often went along with their decisions, but the conflict between his feelings and theirs was mentally devastating and would have led to suicide if he had been forced to continue.

The most often voiced and deeply felt emotion expressed by Joan was always feeling different from what was expected or desired by others. At first, as a toddler, the feeling of being different was relatively amorphous. Even as a preschoo, it shifted to clearly being different from girls. And later, in elementary school, she began to feel not only different from girls but similar to boys. Having a twin might have made this comparison much easier for Joan than it might have been for a singleton. Such a progression in thinking is common for atypical individuals such as homosexual males and females, [17] intersexed individuals, or those with ambiguous genitalia. [18]

The transition was gradual. When Joan thought she might really be a boy instead of the girl her parents and physicians told her she was, the psychic discord frightened her even though she had suspected since the second grade that it was true. When finally told the truth, she was relieved because her feelings now made sense. John's anger at not having been told the truth from the beginning persists.

Following John's sex re-reassignment, the family decided to disregard the clinical recommendation to move from their family home. Instead they stayed and were open about the change. Aside from the financial concerns, the parents judged that the word would get out anyway. This strategy seemed to work and John was accepted in a way that Joan never was.

John was given testosterone treatment following his return to male status. As is typical of many teenage boys, John began to work out with weights. He blossomed into an attractive muscular young man.

According to John's wife, "Before he came along I was a lot tougher on the kids because I had to be. [Now] John is the real hard one and I am the soft one. There is no doubt who wears the pants in this family."

John is a mature and forward-looking man with a keen sense of humor and balance. Although still bitter over his experience, he accepts what happened and is trying to make the most of his life with support from his wife, parents, and family. He has job satisfaction and is generally self-assured.

COMMENT

Long-term follow-up of case reports are unusual but often crucial. This update to a case originally accepted as a "classic" in fields ranging from medicine to the humanities completely reverses the conclusions and theory behind the original reports. Cases of infant sex reassignment require inspection and review after puberty; 5- and 10-year postsex reassignment follow-ups are still insufficient.

Possibly the initial impressions of the consultants [11,12] were appropriate at the time and Joan's behavior and thinking shifted with development. However, clinical notes and the impressions of the local physicians at the time, as well as John's contemporary recollections, indicate that he was at no time fully accepting of sex reassignment. The local physicians expressed their reservations early on. [16] Only when it became obvious to the local team that the original management program of maintaining this child as a girl proved no longer tenable and psychologically damaging, even life threatening, did they revise their thinking.
It is also possible that interpretations from the early years were mistaken. Results contrary to one's hypotheses and management plans are often difficult to see. Manifestations of typical boy behaviors would thus repeatedly be interpreted as tomboyish. This seems to have been the case for preferred activities, games, toys, and clothing. The conclusions that hermaphrodites and pseudohermaphrodites offer a model for normal development had been challenged before. [19-24] The implications of such challenges, however, do not seem to have been accepted or integrated by most pediatricians or surgeons. [7]

Joan was repeatedly admonished for behaving like a boy. Such management is in keeping with the belief that any agreement with doubt expressed by the patient will decrease the likelihood of a successful outcome. [25-28] To contend that Joan did not accept her imposed sex because of ambiguity in treatments, though, is circular reasoning. No evidence of such ambiguity exists and the initial reports held that the rearing was appropriate. [11,12] It is known, particularly from transsexuals, that casting doubt on an individual's sexual identity usually forces that person to introspection and eventual security regarding a preferred life direction, even if it is contrary to upbringing, parent's wishes, and social and cultural norms, and if it results in less than adequate genitalia. [29,30]

In the case under consideration, the initial management protocol was predicated on postulates that considered a male's self-image dependent on a functional penis. Although such adequacy is important, there is no body of data establishing its centrality.

Other considerations are in order. Gender reassignment as proposed for John, and the postulates on which it is based, assumes the individual will learn to accept rearing-appropriate, sex-typical behaviors, particularly when the genitals are at issue. These situations range from the urinary to erotic to narcissistic. Such behaviors, however important, are only one aspect of an individual's total sexuality. An individual's sexual profile comprises at least 5 levels: gender patterns, reproduction, sexual identity, arousal and physiological mechanisms, and sexual orientation, recalled by the acronym PRIMO. [31,32]

The sex reassignment of John to Joan addressed only the gender patterns and gender roles to which he would be subject. Expectations were that adjustments in his identity and other levels would follow. Joan did indeed become aware of the social expectations consistent with the female gender, but these were not in keeping with those with which he felt comfortable. Standing to urinate, despite its housekeeping and social consequences, is a dramatic display of preference. The sex reassignment obviously failed in the area in which it was designed most to succeed.

But it failed at the other 4 levels as well. The contrast between the female gender-typical behaviors the child was being asked to accept and her inner-directed behavior preferences presented a discordance that demanded resolution. Joan's analysis of the situation was that she best fit in not as a girl but as a boy. Despite her upbringing, Joan's sexual identity developed as a male. Sex reassignment also obviously went against Joan's or John's reproductive character. Castration removed any reproductive capacity. Certainly John was unaware of this as a child. He resents this now and decries the loss. Castration also removed the androgen source for sex-typical mechanisms of sexual arousal and other physiological processes. His ability to ejaculate returned with androgen treatment. The castration and surgical scarring, however, have reduced erotic sensitivity to the perineum and, consequently, reduced this pleasure. And notably, as many studies strongly indicate, sexual orientation is prenatally organized or at least predisposed. [33-40] The sex reassignment did nothing to affect sexual orientation. Joan remained totally gynecophilic despite being reared as a girl.

Comments from John's parents reveal another important consideration. With a sex reassignment they were asked to make a dramatic psychological adjustment in rearing an otherwise normal child. Mother
herself required psychiatric treatment to help manage her feelings. The penile ablation aside, the parents were more comfortable dealing with their child's original sex and the accident than with the reassigned sex. Although they had tried to make a success of the sex reassignment, they were supportive, while guilt ridden, when Joan decided to become John.

The last decade has offered much support for a biological substrate for sexual behavior. In addition to the genetic research mentioned, there are many neurological and other reports that point in this direction. \[31,32,41-56\] The evidence seems overwhelming that normal humans are not psychosexually neutral at birth but are, in keeping with their mammalian heritage, predisposed and biased to interact with environmental, familial, and social forces in either a male or female mode. This classic case demonstrates this. And the fact that this predisposition was particularly expressed at puberty, a critical period, is logical and has been predicted. \[20,44\]

Although this article deals with a classic case of sex reassignment often cited in the literature, follow-up to related cases are available. Reilly and Woodhouse \[57\] described 20 patients with micropenises who were reared as boys. None of them had any doubt as to the correctness of the assignment as males. Other reports describe males originally reassigned as females who switched back and successfully lived as males, despite the absence of a normal penis. \[19,32,58-62\] Several of these cases offer findings similar to ours, including the ages at which various milestones were passed, feelings developed, and the reassignment challenged. \[59,60\] Another more recent case illustrates this.

Reiner \[63\] described an adolescent Hmong immigrant who precipitously dropped out of school at age 14. On subsequent interview she declared, though she was unequivocally raised as a girl from birth, "I am not a girl, I am a boy." Findings from a physical examination revealed a 46-chromosome, XY male with mixed gonadal dysgenesis with a female-appearing pelvis with clitoral hypertrophy. Her school friends had all been boys. She enjoyed rough and tumble play, avoided dolls and girl's activities, and dressed in a gender-neutral or boy's way. Her feelings of being different, being a boy, developed from the age of 8 years and came to a head at 14 years. Treatment involved surgery and endocrine therapy. This individual, after a period of some depression, progressively developed into a gynecophilic, sexually active male.

These cases of successful gender change, as well as the present one, also challenge the belief that such a switch after the age of 2 years will be devastating. Indeed, in these cases it was salutary.

It must be acknowledged that cases of males accepting life as females after the destruction of their penises has been reported. \[64\] These reports, however, do not detail the individuals' sexual or personal lives.

**CONCLUSIONS**

Considering this case follow-up, and as far as an extensive literature review can attest, there is no known case where a 46-chromosome, XY male, unequivocally so at birth, has ever easily and fully accepted an imposed life as an androphilic female regardless of physical and medical intervention. True, surgical reconstruction of traumatized male or ambiguous genitalia to those of a female is mechanically easier than constructing a penis. But the attendant sex reassignment might be an unacceptable psychic price to pay. Concomitantly, no support exists for the postulates that individuals are psychosexually neutral at birth or that healthy psychosexual development is dependent on the appearance of the genitals. Certainly long-term follow-up on other cases is needed.

In the interim, however, we offer new guidelines. We believe that any 46-chromosome, XY individual born normal and with a normal nervous system, in keeping with the psychosexual bias thus prenatally
imposed, should be raised as a male. Surgery to repair any genital problem, although difficult, should be conducted in keeping with this paradigm. This decision is not a simple one to make [7,13,18,63,65-67] and analysis should continue.

As parents will still want their children to be and look normal as soon after birth or injury as possible, physicians will have to provide the best advice and care consistent with current knowledge. We suggest referring the parents and child to appropriate and periodic long-term counseling rather than to immediate surgery and sex reassignment, which seems a simple and immediate solution to a complicated problem. With this management, a male's predisposition to act as a boy and his actual behavior will be reinforced in daily interactions and on all sexual levels and his fertility will be preserved. Social difficulties may reveal themselves as puberty is experienced. However, there is no evidence that with proper counseling and surgical repair when best indicated, adjustment will not be managed as well as teenagers manage other severe handicaps. Future reports will determine if we are correct.

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Corresponding author: Milton Diamond, PhD, University of Hawaii-Manoa, John A. Burns School of Medicine, 1951 East-West Rd, Honolulu, HI 96822.

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