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Clinical Case Report Series

"Any Decision is Better Than None" Decision-Making About Sex of Rearing for Siblings with 17β-Hydroxysteroid-dehydrogenase-3 Deficiency

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Children with 17β-hydroxysteroid-dehydrogenase-3 (17β-HSD-3) deficiency have a defect of testosterone biosynthesis with subsequent diminished virilization in XY individuals. Some are raised as girls and some as boys. There were two purposes of this case report: First, it analyzed the process of decision-making in a family with a pair of siblings with identical mutations leading to 17β-HSD-3 deficiency whose parents chose to raise one child as a boy and one as a girl. This analysis was based on narrative interviews with the parents. Second, we assessed the gender role behavior and gender identity in the children to examine if the psychosexual development of these children correspond with the sex of rearing their parents chose. When participating in the study, the children were 7 (boy) and 5 (girl) years old. Parents described a difficult process of decision-making and voiced concerns about lack of appropriate and understandable information, and anticipated decision regret. However, they did not feel that the decision to "normalize" the external genitalia should have been deferred. Both children appeared to show age-typical gender-related behavior and did not show any signs of physical or mental distress.

KEY WORDS: disorders of sex development; 17β-hydroxysteroid dehydrogenase; gender role behavior; gender identity; decision-making.

INTRODUCTION

Disorders of somatosexual differentiation may be associated with an atypical development of the external genitalia. Among these are deficiencies in androgen biosynthesis or defects of the androgen receptor, so that in children with XY karyotype a virilization deficit will occur. This introduction will delineate three issues involved in deciding on sex of rearing in a child with an XY intersex condition: (1) specific aspects of the condition itself; (2) professional tradition and concepts related to corrective surgery in children with intersex conditions; and (3) conceptual considerations in decision-making processes.

The Condition

17β-Hydroxysteroid dehydrogenase type 3 (17β-HSD-3) converts androstenedione to the more potent androgen testosterone in the testes and is the major determinant of testosterone synthesis in males. Mutations in the encoding autosomal gene termed HSD17∃3 lead to defective testosterone biosynthesis and have been described in patients with 46,XY karyotype and moderate to severe diminished virilization. Affected children usually have female-looking external genitals at birth, but lack internal female genitalia, such as a uterus and ovaries. At the time of puberty, virilization of the external genitals is likely to occur. Many children, mostly raised as girls, develop a deep voice, acne, and enlargement of the penis/clitoris. This is probably due to expression of other

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17β-HSD isoenzymes that create bypass production of testosterone from the elevated androstenedione at this time (Boehmer et al., 1999).

Individuals with 17β-HSD-3 deficiency are often raised as girls. This policy has been challenged because of evidence that, in puberty or adolescence, many of these individuals develop a male gender identity and choose to live as men (Hiort et al., 2003; Imperato-McGinley, Peterson, Stoller, & Goodwin, 1979; Mendonca et al., 2000). Cohen-Kettenis (2005) reviewed all published cases in the literature and identified reports about psychosexual development in 30 individuals with 17β-HSD-3 deficiency. Among 28 individuals who were raised as females, 18 changed to the male gender in adolescence. Neither of the two individuals who were raised as males changed to female gender. In one report, one of the two young adults with 17β-HSD-3 deficiency raised as female identified as "intersex" (Warne et al., 2005). There is evidence that this uncommon high rate of sex change can be explained by prenatal androgen effects on the developing brain, causing male typical behavior and perhaps a male gender identity (Hines et al., 2002; Hines & Collaer, 1993; Lephart et al., 2001).

17β-HSD-3 deficiency has to be differentiated from other enzyme defects of androgen biosynthesis as well as from partial defects of the androgen receptor leading to partial androgen insensitivity syndrome (pAIS). In pAIS, a general insensitivity toward androgens is presumed, but the pubertal surge of testosterone may also lead to partial virilization. In contrast to 17β-HSD-3 deficiency, patients with pAIS also show partial female characteristics with breast development because of aromatization of testosterone and subsequent estrogenic effects.

Corrective Surgery in Children with Intersex Conditions

The birth of a child with ambiguous genitalia does not in most cases constitute a medical emergency, but is often interpreted as a "psychosocial emergency" (Izquierdo & Glassberg, 1993; Meyers Seifer & Charest, 1992; Reinecke, Hampel, Richter-Appelt, Hiort, & Thyen, 2005). Parents and medical personnel feel pressed to assign a gender to the child as soon as possible. This impression of urgency is supported by the commonly used "optimal gender policy," developed by Money (1985, 1994), and Money, Hampson, and Hampson (1955). On the assumption that gender development is based predominantly on social influences, Money advised to assign a gender to children with intersex conditions as soon as possible and to surgically "correct" the genitalia in early childhood, so that it will look as "adequate" as possible. Money was convinced that the child will develop gender role behavior and gender identity according to the chosen sex of rearing, if the child is raised without ambiguity causing no reason to question its own gender (e.g., because of ambiguous external genitals).

The decision which sex of rearing will be the best for a special child depends on functional aspects: the goal of treatment is to reach optimal outcomes in terms of sexual and reproductive functioning and psychological, physical, and social well-being. Following this policy, many children with 17β-HSD-3 deficiency and ambiguous genitalia were raised as girls but accurate molecular diagnosis was not available at the time.

Since the early 1990s, the optimal gender policy was increasingly criticized (Androgen Insensitivity Syndrome Support Group [AISSG], 2006; Beh & Diamond, 2000; Diamond & Sigmundson, 1997; Intersex Society of North America [ISNA], 2006). The argument follows evidence that biological effects, such as hormones, play an important role in psychosexual development of humans. Another cause is advocacy for patients' (and children's) rights for bodily integrity and self-determination. The "full consent policy" is based on the informed consent doctrine (Faden & Beauchamp, 1986), arguing for the patient's right to make medical decisions on his/her own behalf. To do so, he/she needs unrestricted information about his/her diagnosis, treatment options, and possible outcomes.

With regard to children with intersex disorders, advocates of the full consent policy point out that most of the genital surgeries carried out in childhood are done for "cosmetic" and not for medical reasons and should be postponed until the child is able to take part in the decision and give its consent (Beh & Diamond, 2000; Chase, 1998; Diamond & Sigmundson, 1997; Kipnis & Diamond, 1998). This policy demands complete information of the child about its medical condition, prognosis, and possible ways of coping with it, information that is, under the optimal gender policy, often withheld from children and adolescents.

Neither the "optimal gender policy" nor the "full consent policy" relies on evidence-based clinical outcome studies (Hiort, Reinecke, Richter-Appelt, Holterhus, & Thyen, 2001/2002; Meyer-Bahlburg, 1998). Decision-makers (health professionals as well as parents) have to act in a situation of extensive uncertainty about expected outcomes.

Decision-Making

Classical rational normative decision theories assume that people responsible for a decision collect all information accessible to them, make a diagnosis that reflects uncertainties, develop alternative action plans, know and evaluate chances, risks, cost, benefit, and probability of occurrence (Jungermann, Pfister, & Fischer, 1998; von Neumann & Morgenstern, 1947). Finally, they choose the best of the possible alternatives having a high chance of success and low risk of loss. Such a deductive procedure does not fit situations where little is known about diagnosis, prognosis, and the consequences of decisions. Therefore, in this case, the best approach seems to be to reconstruct the parents' decision-making process by means of descriptive decision-making theories, allowing to describe and analyze the decision-makers' beliefs and preferences as they really are, not as they should be (Kahneman & Tversky, 2000).

The aim of this case study was to help reconstruct and analyze the influences that affected the decision-making process and describe the various problems and sense of urgency the parents (as well as the doctors) had to face. It was intended to clarify the factors that aggravate, ease or enable the initial decision-making and to show the consequences that can result from a decision, once it is made. The analysis was also expected to give an idea about how the parental decision-making process can be facilitated.

CASE REPORT

The subject of this case study was a couple whose two children were both born with XY karyotype and ambiguous genitalia and who decided to raise one of them as a boy and the other as a girl. In the decision phase, parents and physicians assumed the diagnosis of a partial androgen receptor defect (androgen insensitivity, pAIS). Later molecular-genetic analysis substantiated identical mutations in the 17β-HSD type-3 gene in both children. All material concerning physicians' management and recommendations came from parents' report.

Narrative Interviews

Because personal orientation, state of knowledge, and interpretation of situations play a central role in decision-making processes, we conducted narrative interviews (Bauer, 1996; Schütze, 1977, 1983) with both parents. By means of a suitable introductory question, the parents were invited to give an extempore narration of the child's "story." Supported by the inherent narrative constraints (Riemann, 2003; Schütze, 1977), this form of interviewing produces rather complex and comprehensive versions of the subjective views of the interviewees. They include personal experiences and orientations, and social state of knowledge, in which the interviewees' lives are embedded (e.g., Chamberlayne, Bomat, & Wengraf, 2002). This method rests upon everyday knowledge of the interviewees and identifies themes and aspects that might remain unmentioned in normal conversations and conventional interviews (Schütze, 1976).

A narrative-retrospective reconstruction of experiences enables the parents to relate from their current perspective to the situation they were living through at the time and render and interpret the associated feelings and judgments that led to the decision in question. The evaluation of the interviews in this report is restricted to those aspects of the interview that are meaningful for their decision-making. It cannot be presumed that the parents' decision-making behavior is shown in its entirety.

When participating in the study, the children were 7 (boy) and 5 (girl) years old. The interviews were conducted by an experienced staff member. With consent of the interviewees, the interviews were tape recorded. The parents were informed that they could ask to stop the interview or the recording at any time. At the end of the interview, after switching off the recorder, the parents were offered the opportunity to debrief.

The interviews were transcribed verbatim. To maintain anonymity, all personal data were modified or left out. We simplified the original transcription, omitting pauses or other paralinguistic features and phonetic expressions.

The analysis was undertaken along the lines of a case reconstruction study (Hildenbrand, 1994). We used the method of text reduction (e.g., Mayring, 1983), which allows one to develop analytical categories. Because the circumstances of this case of decision-making does not offer the opportunity to contrast with other interviews, the analysis remains descriptive.

Results

The Decision-Making Process: The First Child

The parents were unrelated and at the time of the first pregnancy in their early 30s and were healthy. Both worked as medical professionals. The pregnancy passed without any complications.

Mrs. A reported that she had always wanted her first child to be a boy "and during the pregnancy I always had the feeling it might be a boy. So that was somehow quite deep inside me." The parents had routine ultrasound

check-ups done but they did not want to know the gender of their child before the birth; they wanted it to be a "surprise."

When the baby was a few days beyond the expected date, birth was induced and eventually ended in a caesarian section. This was perceived as very stressful by the parents. Immediately after the birth, the child was noted to have ambiguous genitalia.

When Mr. A later visited his wife in the recovery room, he said to her, "Something is wrong with the child. Well, it looks rather like a girl. But it's not OK." Mrs. A remembered her own feelings when being told this, "I was pretty shocked at that and naturally done in, and then I saw him for the first time and, well, my feeling was: it's a boy. Because I wanted it to be." Mr. A reacted to the ambiguous genitalia of his child with "amazement, uncertainty," but told himself, "Well, this is something rare and it's not so good. But you always tell yourself: Well, there are worse things."

The same day, Mr. A took the child to the nearby pediatric hospital where first diagnostic measures were undertaken. The child presented with a microphallus and bifid scrotum, corresponding to intersex Grade 4 (Quigley et al., 1995). The gonads were palpable in the labioscrotal folds. Ultrasound demonstrated a lack of Mullerian structures. Karyotype was 46,XY.

The parents decided to delay naming the child because they were uncertain about the child's sex. About the first days after birth, Mrs. A remembered "that I was forever lying awake at night, crying, and totally disappointed." She also found seeing family members, friends, and, even more so, people she knew less well very stressful. "Well, the most difficult people to meet are neighbors and the cleaning lady and people like that. Those that have seen you pregnant for weeks and then you're walking along with your pram, and that's really tough. Just going to the butcher's. Well, it's really exhausting keeping up appearances and they notice anyway: something's not quite right here."

When the result of the chromosome analysis was available, the parents felt very relieved. They considered this information to be the basis of their decision about their child's sex. It helped them to escape the ambiguous situation they felt to be truly unbearable. Mrs. A, "But I just find you have to decide as quickly as possible. This state really stresses you out." For further diagnoses and to make them certain in their decision, the parents contacted an endocrinologist. He expressed the hypothesis that this was a case of androgen insensitivity and advised the parents to raise the child as a girl, "He always thought it simply easier and more practicable and experience shows." Mr. A tended to agree with the endocrinologist's views. He pointed out that the endocrinologist was the one who has "the experience," and he worried that they "might regret it later during puberty." Mrs. A objected to this option. "To this, I always said: It's been clear to me from the start of the pregnancy. And I always said then: That's not for me: XY and girl. I won't do that." The parents decided to bring up the child as a boy and gave him a male name. They were conscious of the fact that this decision was not based on a concrete diagnosis or on any appreciable prognosis for the further development of the child. However, the feeling prevailed that it was better to make some kind of decision and maybe to revise it at some later date rather than carrying on leaving the question of the child's gender open. "We always used to say OK, let's go in the direction of 'boy' and see what happens. We would possibly have revised our opinion if a receptor resistance." Mrs. A stressed repeatedly that, in her view, "any decision is better than none."

Further diagnostic procedures aimed at checking the suspected diagnosis of androgen insensitivity. Laboratory analysis showed low normal testosterone at the age of 3 weeks with 0.4 ng/ml (reference range, 0.25–3.12). Androstenedione was measured at 0.47 ng/ml (reference range, 0.26–1.63). As these values were considered non-conclusive, further diagnostic testing was suggested for the age of 3–6 months. However, the parents decided against a further endocrinological evaluation because Mrs. A had little hope of gaining any further insight to help their decision-making. She rated the stress resulting from painful procedures (taking blood samples) greater than their potential benefit. Apart from that, she feared that new findings might re-open the question of the decision they had just made to raise the child as a boy.

When the child was 9 months old, the parents decided to ask for a third opinion and consulted an endocrinologist in another town. Although there was no further definitive diagnosis at this point, the final decision was made in the course of the conversation with this physician: The child was to be raised as a boy. One of the main criteria for this decision was the rather male appearance of the genitalia. Mrs. A also supposed that the doctor had noticed that the child had been raised as a boy up to that point and that the parents would find it hard to change. "He said our personal feelings about the children were very important." The parents found this decision a great relief. "And then everything was alright. After that we said, OK! And then we had the baptism and we said, We're sticking to it."

The step of the child having an operation to help along the intended gender characteristics was for them "quite clearly medically indicated." They were also afraid the atypical appearance of the child's genitalia might lead to other people "suspecting," and that there might be

rejection or teasing. "You go to the swimming pool with the child, don't you, and you always think: Has anybody noticed? Is anybody looking?" Mr. and Mrs. A agreed with the physicians caring for their child and decided that the respective operations should be carried out as soon as possible, "Firstly, it heals more easily at that earlier age and also a child should have it done when it doesn't yet really know what's going on."

The child had the operations between the ages of 14 and 21 months (hypospadia repair, penis enlargement, orchidopexy). Because of complications (fistula, urolithiasis), a third operation at the age of 2 years became necessary. Even though the intervening period was a very difficult experience both parents were altogether very satisfied with the result of the corrective surgery.

The Decision-Making Process: The Second Child

Looking back, Mrs. A summed up the time with her first child as exhausting and very frustrating. She and her husband decided, therefore, to have another child as soon as possible. Mrs. A became pregnant again, as desired. During this pregnancy, she repeatedly entertained "thoughts that something like this might happen again." She consulted a specialist in prenatal ultrasound when she was about 15 weeks into the pregnancy. The consultant expressed himself carefully and did not want to commit himself: the fetus was female, but he was not able to detect a uterus. At a repeat check-up 6 weeks later, "you could tell then: somehow he was not 100% certain, but he stuck to his guns: It is a girl. And I just know." In retrospect, Mrs. A wondered how this false diagnosis could have come about and went over the situation at the ultrasound check-up: "I think we're also at least partially capable of interpreting things but none of us could really see the uterus. Of course, we also failed to spot the testicles. But we didn't expect to see them, and he didn't want to see them either and so you then don't see it."

After the ultrasound check-up, another consultation took place. When Mrs. A asked for an amniocentesis, the doctor was against it: "It isn't necessary. That was a one off. Something like that doesn't happen again. And he tells me: Now just look forward to a girl and it definitely won't happen again." Mrs. A allowed herself to be convinced and did not go for an amniocentesis.

The birth of the second child also took place at term. Again, it was a difficult birth, but this time a caesarean section was not necessary. Instead, Mrs. A witnessed the reaction in the delivery room, "Well, she came out and there was embarrassed silence. It was really just like it shouldn't be. Nobody said a thing. I mean, as a mother you really get a scare and you think it's something life threatening, and I say: What's the matter then? And the midwife doesn't give a peep; she was shocked in some way. They hold the child in the hand and look at it and don't say anything, you see? I mean, well the baby had cried, so I knew she isn't dead or something, but you can't do that." When it was clear that it was "the same problem as with the first child," the baby was removed from the room. Mrs. A was very clearly aware of the uncertainty around her and got the impression that the medical personnel were trying to avoid her, "I found that with both births, well, you compare with other mothers, but I can't remember any midwife. Later on, no one came to me."

The parents realized that the baby had the same disorder as the first, even though the appearance of the external genitalia was more on the female side. This baby also had a micropenis: the penile length was extremely small (0.5 cm). Both gonads were again palpable in the labioscrotal folds. Again, no Mullerian structures were found on ultrasound. Karyotype was 46,XY. Laboratory analysis revealed a testosterone of 0.19 ng/ml and androstenedione of 0.75 ng/ml.

Mrs. A thought it obvious to proceed in the same fashion as with the first child. "I want it to be a boy. I'm not doing it any other way. It's the same thing." Her main argument for this desire at this moment was that she did not want to be unfair to one of the children. She wanted them to be treated in the same way, "because you can't be unfair to one of them and I wanted it like that then, and I want the same thing again now." The parents thought about boys' names for the child and tried to think of it as a boy but found it difficult. Mrs. A reported, "then I tried to force the issue and said: it's a boy. But B [her husband] was becoming quite withdrawn."

The parents talked about the chromosome analysis with the endocrinologist. He considered himself confirmed in his hypothesis that in both children's cases it was a matter of partial androgen insensitivity (pAIS) and drew the conclusion that it would be better to bring the child up as a girl. A surgeon who had moved into the area stated that he could do an operation to make the child into a girl or a boy, but that it was easier "to make it into a girl." Mrs. A described her feelings after this talk, "and we somehow noticed at this point that I'm fighting against something that doesn't work somehow."

In the first week of the child's life, the family consulted the doctor who helped them make the decision for the first child. He, too, tended to the view to let the child grow up as a girl because of the child's external appearance. The experience of the lengthy decision-making process, the operations, and the connected complications for their son confirmed the parents in the wish to protect at

least this child from surgery: "I won't let anybody interfere with this child." Apart from that, they were afraid that the operations for the second child would be more extensive and more difficult than for the first one. "Too much would need to be done and X [the son] had already had his first operations at this point and they hadn't been without complications. So we said, We can spare the child that."

The decision to let the second child grow up as a girl was again strongly influenced by Mrs. A's prenatal feeling that the child would be a girl. At this consultation, a possible "feminizing" operation was also discussed. The endocrinologist saw no immediate need for action and advised "to do as little as possible now but to wait until she herself can decide and know what's going on, and also that research is making progress and everything is going to be better, in terms of operation techniques and everything else."

In spite of that, the parents gathered information on possible operations and contacted several pediatric gynecologists to whom they introduced the child. The help they had hoped for, however, was not forthcoming. The doctors informed them about the different surgical options, but the decision what would be best for the child had to be made by the parents alone. The conclusion of this consultation was "Let's wait for puberty."

In the immediate aftermath of the decision not to let the child undergo any surgical procedures, something happened that, in retrospect, was perceived as a "hint of fate" by Mrs. A. At 6 weeks, the child had a hernia. At this juncture, the parents thought "some kind of surgery is going to be done in that area anyway; well, then let's have it done, once and for all." Mrs. A backed up her decision with the statement that she had given up waiting for a final diagnosis for her children at this point. "That was so far away that I thought there's no point in doing any more tests." Looking back, she considered her ability to come to a reasonable decision at that time as clearly restricted: "I mean, you're really in shock with something like that, aren't you?"

The child's gonads were removed and a diagnostic biopsy taken. Mr. A stressed in the interview, "We just took the hernia as an opportunity. They've got to come out, they had both been inguinal testicles, so simply from the risk of malignancy and also the hormones which they don't need in this case it was absolutely clear: they had to be removed."

The Diagnosis

Over half a year later, the parents were given the results of the molecular–genetic findings: 17β-HSD-defect. For molecular–genetic investigations, DNA had been extracted from peripheral blood leukocytes. The whole coding sequence of the HSD17B3 gene was amplified with polymerase chain reaction and subsequently analyzed for variations as previously described (Twesten et al., 2000). In both children, two abnormalities were found. In exon 3, a guanine to adenosine exchange induces an amino acid substitution of the normally present arginine to glutamine in codon position 80, whereas an adenosine to guanine exchange in exon 5 will introduce the mutation asparagine to serine in codon position 130. Both mutations have been described earlier (Moghrabi, Hughes, Dunaif, & Andersson, 1998; Rösler, Silverstein, & Abeliovich, 1996). Especially the mutation affecting codon 80 with a substitution of arginine by glutamine will leave considerable function of the partially defective 17β-HSD-3 enzyme. This may be the reason for the measurable testosterone levels in both children and, hence, also the considerable virilization seen. Both parents were found to be heterozygotes for one of the mutations, proving the autosomal-recessive inheritance of the mutations.

The communication of the diagnosis confronted the parents with the question as to the correctness of their decisions and renewed all their doubts. Although their initial bad conscience toward the child that grew up as a boy was replaced with relief about having taken the right decision in his case, the decision to let the second child grow up as a girl now seemed questionable. "When at some point the call came, the whole thing was quite hard to cope with in your mind. You say: With X [the son] it was right to stay hard. But, in retrospect, you have to admit that you'd have taken a different decision with T [the daughter] if you'd known."

After learning about the diagnostic findings, Mrs. A blamed herself for not having persisted with the diagnostic process for the first child, but she comforted herself with the assumption that the tests planned at that point would not have produced any results. Her conclusion was rather fatalistic: "It was meant to be like that. It was fate."

About a year later, at the age of 22 months, the parents decided for their second child to have a surgical reduction of the external genitalia and a correction of the labia. This decision was strongly influenced by the worry that the unusual appearance of the genitalia might lead to teasing or rejection by the child's peers. The parents agreed with the view of the endocrinologist and the widespread conviction in the literature that having atypical external genitalia would affect the child's psychosexual development. However, the surgical creation of a vagina was not undertaken for the time being. The operation went without complications, "and it went without any great to do. It was really only 3 days and he [the surgeon] explained

it, although he isn't really good at explaining and he did it and we were back home in no time. In no time it was forgotten."

At present, Mrs. A is sometimes of two minds about whether the decision about her daughter's sex of rearing and the operations were correct. The feeling remained that she might have taken some possibilities away from her daughter, especially the one of having children. She also fears the surgery might have resulted in reduced sexual sensitivity.

In spite of her doubts, Mrs. A stressed that she was very pleased with the surgery on both her children, even if the boy sometimes had nocturnal enuresis. She thought the children look mostly "inconspicuously," so that the parents would have no hesitations in letting their children run around naked (e.g., on the beach). Mrs. A also wondered how she would eventually explain to her children how they decided differently for each of them. With respect to her daughter, whom she takes to be "the less introverted of the two," Mrs. A was quite optimistic that she will be able to understand the decisions of her parents. The possibility that her daughter might decide later on to live as a boy/man did occur to Mrs. A and she said she was mentally prepared for it. She did not think she will have a problem with it: "I think I could accept that. Well, if I have the feeling she would be happy, of course." In bringing up the children, they were guided by the principle of strengthening the children by a "happy and unburdened childhood" to a degree that later on they can cope well with problems that might occur. This means that up to now the children have received only scanty information about their development, in order not to confuse or unsettle them.

Satisfaction with Medical Advice, Care, and Social Support

Looking back, the parents emphasized that the lack of information and social support was a great strain. Particularly, the fact that they themselves were working in the medical field seems to have resulted in the fact that "of course, nobody wanted to tell us anything because they think: They'll know that themselves. And they don't have the courage to say anything because they think we've probably read more about it than they have." When information was given, it was of a purely "medico-technical" nature. The parents said they were never given practical advice and help with actual decision-making. Physicians approached them predominantly as "colleagues" and not as concerned parents needing help and support. Mrs. A stated that she felt very alone in these decisions and that she did not feel up to making them. She would, for instance, have been happy to have regular follow-up appointments with a doctor who was monitoring the children's development and who would have shared the responsibility. The lack of support from the doctors in charge, their lack of guidance about the adequate diagnostic procedure, and the way to deal with a child with unusual gender development, as well as mistakes in prenatal diagnoses, confirmed the parental impression that they were the only ones responsible for their children's lives: "Of course, you want to spare the children, you've got pretty big problems and you somehow have to cope with that."

The feeling of being left alone was reinforced by the fact that the subject of intersexuality was not discussed with family or friends. What transpired clearly in the interviews was that the parents were ambivalent about telling the people they knew: On the one hand, they wanted to exchange views about this problem area with other people; on the other hand, they only opened up to those who, by nature of their close contact to the children, could not be left uninformed (and even those received only very limited information). With this policy of discretion, they tried to prevent the children and themselves from being stigmatized. They did not want to give rise to any "moments of suspicion." This ambivalence kept Mrs. A from joining a self-help group, although quite a few times she felt the desire to exchange views with other people. Mr. A was not interested in this as he did not think it would help to talk to uninvolved people about the specific development of his children.

Some relief was obtained through few talks with close friends and relatives that did take place and by the relationship itself, which was strengthened by the jointly experienced problems and expectations of those yet to come. "For our marriage, it was probably rather positive. It really cemented our bond, having to deal with this on a permanent basis and to offer an unending supply of future. Particularly nowadays, when lots of people give up too easily and say: Let's have a divorce. Of course, I think something like this is rather a reason for saying: No, with children like these you've got to stay married."

Psychological Outcomes

The 5- and 7 year-old siblings and their parents were interviewed as part of a larger study of gender-role behavior and behavior phenotypes in children with XY karyotype and intersex conditions between 2002 and 2004. Written consent was obtained from all participating

parents and verbal assent from the children. The study was approved by the Ethical Committee on Human Studies of the Medical Faculty of the University of Lübeck.

The instruments used were

1. KINDL, to measure health-related quality of life in children (Ravens-Sieberer & Bullinger, 2000). This questionnaire consists of six scales (physical well-being, psychological well-being, self esteem, family, friends, and kindergarten/school).
2. To assess any possible behavior problems, we used the Child Behavior Checklist (CBCL; Arbeitsgruppe Deutsche Child Behavior Checklist, 1998).
3. In addition, we used the German shortened version of the Parenting Stress Index (PSI; Tröster, 1999).
4. To assess children's activities and interests, preferences in gender-typical games, and dressing-up in role play we constructed a parent report questionnaire. We adopted the methodology from the Child Game Participation Questionnaire (CGPQ; Meyer-Bahlburg, Sandberg, Dolezal, & Yager, 1994a1994b) and developed culturally adapted items suitable for German children. Parents were asked to indicate the frequency (0 = *never*; 1 = *seldom*; 2 = *often*; 3 = *very often*) of participation for each item. Female-typical items were summed to yield a score of female-typical activities and interests, male-typical items to a score of male-typical activities and interests. We calculated the percentage of girl-typical (AIQ-female) and boy-typical activities and interests (AIQ-male; range = 0–100).
5. Play behavior was observed in a structured free-play task (modified from Berenbaum & Snyder, 1995; Zucker, Doering, Bradley, & Finegan, 1982): Toys were arranged in a standard order on the floor or a suitable table while the child was absent. The selection of toys was based on interviews with parents on their children's toy preferences and on lists of bestsellers from toy-stores. For typical "Boys' Toys," we chose four play figures (science fiction figurines, warriors), six cars, a toy pistol, and a tool box. The typical "Girls' Toys" included play figures (horsewoman, horse), a Barbie doll with clothing, a baby doll with several care products, and cooking accessories. The child was brought into the room and asked to play freely for 10 min with the toys. The observer stayed in the background with a stopwatch, measuring the time the child played with toys typical for boys or for girls. For each child, we calculated a score reflecting the percent of total time of playing with male-typical toys (range, 0–100).
6. Toy preference of the children was assessed by a "toy to keep" task, modified from Berenbaum and Snyder (1995). After participating in the study, the child chose one "toy to keep" among five toys. These toys had been a priori assigned scores, ranging from 1 (*very typical girls' toy*) to 5 (*very typical boys' toy*). We used two different sets of toys: Children aged 3–6 years chose from a play figure (princess with horse and coach), a children's book "Flowers," a kaleidoscope, a children's book "Airplanes," and a truck with trailer. The set for children aged 7–12 years included a set of plastic beads, a board game, a kaleidoscope, playing cards featuring "Car Monsters," and a Lego building set "Star Wars."
7. To assess gender-typical behavior and attitudes, we developed a German version of the Child Behavior and Attitudes Questionnaire (CBAQ; Meyer-Bahlburg, Sandberg, Yager, & Ehrhardt, 1994a1994b). This German translation is a short version of the CBAQ and consists of 30 items for boys and 29 items for girls. The CBAQ includes two scales: The Femininity Scale measured the extent of typical feminine behavior (bipolar; 17 items; high scores = feminine). The Cross-Gender Scale measured the extent of cross-gender behavior which may indicate confusion or instability of gender identity (unipolar; 10 items; high scores = cross-gender).
8. Gender Identity was assessed by the Gender Identity Interview (Zucker et al., 1993).

If available, we used normative data for comparison and interpretation. For questionnaires Nos. 3, 4, and 7, comparative data were taken from questionnaires given to 166 kindergarten and school children (89 boys, and 77 girls) between 3 and 12 years (Naujoks, 2005). For the free-play task (No. 5), the toy to keep task (No. 6), and the Gender Identity Interview (No. 8), it was not possible to obtain comparative data.

Test Results and Clinical Impression of the Children

When participating in the study, the children showed age-appropriate psychomotor development. According to

the parents' (see Table I) and their own statements, there were no restrictions with respect to their health-related quality of life (KINDL). The results of the Child Behavior Checklist (Table I) showed no abnormalities in the individual scales for either of the children, but for the scale "Externalizing Behavior" (second-order scale), the children scored in the borderline clinical range (boy $T = 60$; girl $T = 63$). The Parenting Stress Index (PSI) did not indicate any abnormal stress levels related either child (Table I). On the Gender Identity Interview, neither of the children showed any signs of self-doubts about the gender assigned to them by their parents.

The evaluation of a list of play preferences, activities, and hobbies (Activities and Interests Questionnaire; see Table I) showed that the boy had a slightly higher value than the comparison group of boys when performing "activities typical for girls" (AIQ-female), but he was clearly below the average value for girls. On the scale "activities typical for boys" (AIQ-male), his value was slightly below the comparison group of boys but clearly above the comparison group of girls. Taken together, the results indicated that his behavior seemed to be typical of an average boy. In play situations, he played exclusively with the typical boys toys. When choosing a toy to keep, he chose the one typical for boys. Compared to boys in the control group, the boy had on the CBAQ a slightly raised value on the scale for cross-gender behavior and a slightly lower value on the femininity scale, but both values were within 1 *SD* and must be interpreted as normal variation.

In comparison with the control group, the girl showed a higher value for cross-gender behavior and a lower value on the femininity scale. On average, she displayed less "girlish" behavior than other girls. In play situations, she played a little longer with the typical boys toys (55% of the time) than with girl's toys. The evaluation of play preferences, activities, and hobbies showed a similar result: She had a score of 40.0 on the scale of "activities typical for girls" (AIQ-female) and therefore scored below the median reached by girls in the comparison group but also clearly above the median of the comparison group of boys. On the scale "activities typical for boys" (AIQ-male), the girl scored above the comparison group of girls and almost achieved the values of the comparison group of boys. Taken together, the girl had a slightly higher value for "activities typical for boys" than for "activities typical for girls." As a toy to keep, the girl chose one "typical for girls."

Table I Health-Related Quality of Life, Behavioral Problems, Parenting Stress, and Gender-Related Behavior

	Comparison group			Case study	
Instrument	Boys & girls[a], M (*SD*)	Boys, M (*SD*)	Girls, M (*SD*)	Boy	Girl
KINDL (proxy: high score = high quality of life)					
Total	68.97 (9.31)			78.13	79.17
CBCL (high score = more behavior problems)					
Internalizing		4.8 (4.6)	4.6 (4.3)	7.0	3.0
Externalizing		7.8 (6.6)	5.9 (5.0)	13.0	14.0
Total		20.5 (4.8)	17.3 (2.8)	28.7	24.6
Parenting Stress Index (high score = more stress)					
Total	138.77 (36.20)	146.58 (39.20)	129.89 (30.34)	120.00	117.00
Activities and Interests Questionnaire (% of all activities and interests)					
AIQ–male		46.9 (8.1)	17.2 (8.1)	46.5	41.1
AIQ–female		26.9 (8.7)	55.8 (10.2)	32.4	40.0
Structured free-play task				100	55
Time spent with typical male toys (%)					
Toy to keep					
1: *very typical female toy*				4	2
5: *very typical male toy*					
Child Behavior and Attitudes Questionnaire (high score = more feminine resp. cross-gender behavior)					
Femininity scale		58.1 (6.72)	79.0 (7.63)	54.0	69.0
Cross-gender scale		22.3 (5.95)	21.6 (5.43)	25.0	31.0
Gender Identity Interview (high score = more signs of gender confusion)					
Total gender confusion scale	3.7 (3.2)			1.0	2.0

[a]KINDL scores of healthy children in this age group are not different for boys and girls.

DISCUSSION

In this case study, the parents decided to assign a gender to their children and to have the relevant corrective surgery done before a definitive diagnosis had been made. In their first child, they chose male sex of rearing; in the second, female. The decisions were made without advanced knowledge of their consequences and can therefore be seen as "risky choices" (Kahneman & Tversky, 2000). The reason why the parents were ready to take risky choices was their feeling of a great decision-making pressure, based on the parental need to be able to see, address, and treat the child as a boy or a girl as soon as possible; their feeling of chronic stress in their everyday lives through uncertainty and indecision; the insecurity they felt in dealing with their friends and relations and their worry about their lack of understanding; their fear of stigmatization/of social isolation of the child/the whole family; and their feeling of being left alone with their responsibility and decision-making imperative.

The decisive criteria for the choice of the gender seen as adequate for the respective child were the mother's prenatal intuition with respect to the sex of the child; the phenotype of the external genitalia; the suspected clinical diagnosis (pAIS); and surgical possibilities of "correcting" the genitalia (aim: good cosmetic and functional results).

In analyzing the process of decision-making, it became obvious that the parents faced a very complex situation, in which they had to choose between different options. To choose one option meant giving up others, which may lead to an internal struggle. In this perspective, the decision can be regarded as a source of conflict (Coombs, 1987; Keren & de Bruin, 2003) and the resolution of this conflict served as a reduction of stress. In almost all cases in "real life," the decision itself and the process of decision-making is influenced by emotions, which may override rational considerations. Therefore, to understand a decision, we have to take into account the emotional circumstances under which it occurred and we have to bear in mind that circumstances and related emotions may change over time so that decisions may be regretted by the decision-makers themselves later.

In this case study, the parents experienced the birth of their first child with ambiguous genitalia as a shock. Their first approach was to clarify what they thought was the "real sex" of the child by identifying the karyotype. The result, XY, was the motivation for the parents to decide: It's a boy. This initial decision, also supported by the mothers' prenatal intuition, was working as an "anchor" later on: Although the endocrinologist, who suspected AIS as a diagnosis for the child, and the father discussed rearing the child as a girl because of the expected lack of virilization in puberty, the mother rejected this proposal. Even results from hormonal testing showing poor reactions of the child to androgens were interpreted as "not that bad" by the mother.

In decision theory, this well-known phenomenon of over relying on first thoughts/first decisions by putting down a "mental anchor" and then paying less attention to alternatives is called the *primacy effect* (Einhorn & Hogarth, 1978; Tversky & Kahneman, 1974, 1981) or *anchoring trap* (Hammond, Keeney, & Raiffa, 1999). This effect is complemented by the *confirmation bias* (Einhorn & Hogarth, 1978) or *confirming-evidence trap* (Hammond et al., 1999), the tendency to seek out information and stimuli that support the chosen hypothesis. Contradictory evidence is not sought and relevant evidence ignored. When receiving ambiguous information, only the meaning that conforms to the existing hypothesis is stressed. Opposing information is often not actively sought because people want confirmation and they experience dealing with negative information as uncomfortable (Clark & Chase, 1974). Giving up a hypothesis and forming a new one is an energy-consuming undertaking. In our case, this effect was particularly in evidence at the prenatal investigations in the second pregnancy.

In this case study, it becomes obvious that not only the parents but also the physicians are affected by such psychological effects: The specialist in prenatal ultrasound, who was consulted early in the pregnancy with the second child, was not able to have an objective look on the fetus but saw what he wanted to see and what he thought would comfort the parents. Even though he knew about the ambiguous genitalia of the first child, he interpreted not seeing a phallus in ultrasound as an indisputable proof of fetus' sex.

The decision-making process about the sex of rearing in the second child differed in some respect from that of the first child: After birth, when it became obvious that the child had the same "problems" as its sibling, the parents decided to treat it like the first child and assign the male gender, particularly because they felt it would be unjust to proceed in a different fashion than with the first child. But in this case, according to the considered opinion of the ultrasound specialist, their "mental anchor" had been prenatally put down at the female sex of the child and they found it difficult to regard this child as a boy. In addition, the virilization of the external genitalia of the second child was less pronounced compared to the first child. The parents, who felt very stressed and worried about the surgeries their first child had to cope with, feared it would take even more surgeries to construct a more male looking genitalia in this child. In doing so, their decision was

strongly influenced by dramatic emotions and memories concerning their experiences with the surgeries the first child underwent. In this respect, their decision to rear this child as a girl was also based on their belief this would prevent their child from traumatic experiences they expected were very likely to occur. This common psychological mechanism, to focus on dramatic events and perhaps overestimate their chance of occurrence in future decisions, is called the *recallability trap* (Hammond et al., 1999).

Even though they did not cite it as a point of reference, the parents acted in accordance with the "optimal gender policy" guidelines (Money, 1985, 1994; Money et al., 1955). According to Money, it is necessary to assign the sex of rearing to the child as soon as possible and to bring it up in this gender role quite consistently. Money emphasized the primacy of social influences over biological factors on psychosexual development and regarded gender as an assignable trait. Therefore, he recommended to assign a sex of rearing in children with ambiguous genitalia as soon as possible and surgically adapt the child's genitalia to the chosen sex at the earliest opportunity.

In the case presented here, the parents found the option of keeping their children in an undefined status or assigning them to a certain sex without having adaptive measures performed as impractical. In their view, it was necessary for the development of the child to have a firm place in the existing gender system. After initial reluctance, the parents changed their personal views about telling the children about their condition and gender development. Their interest and participation in this study was influenced by the desire to obtain more help and advice with the problem of informing the children in a way that was appropriate to their age. In doing this, they were approaching the more recent recommendations of a "full consent policy" (Ahmed, Morrison, & Hughes, 2004; Berenbaum, 2003; Conte & Grumbach, 2003; Martin, 2003): They now wanted to talk to the children about the decisions they had made and to give them the chance to participate in future decisions (e.g., about surgical construction of a neo-vagina in the girl). However, the parents did not share the view that surgery should be delayed until the children reached the age of consent (Diamond, 1999). In this aspect, they were in accordance with studies showing that a majority of parents decide to let their children be surgically treated to look more "natural" (e.g., Dayner, Lee, & Houk, 2004).

In spite of persisting doubts as to the "correctness" of the sex assignment of their children, the parents saw themselves confirmed in their actions by their children's healthy development up to now. The parents emphasized that the girl's rather boyish behavior did not worry them because she showed no self-doubts about her gender identity, was accepted by her peers, and was otherwise developing completely normal. This assessment by the parents was verified by the results of our study. In the literature, three studies report on gender role behavior in some of their cases of prepubertal children with 17β-HSD-3 deficiency: some children raised as girls showed "tomboyish" behavior; some post-pubertal patients showed signs of gender dysphoria (Imperato-McGinley et al., 1979; Mendonca et al., 2000; Rösler & Kohn, 1983). In our case report, the siblings were too young to make an appraisal of future gender identity.

Knowing the diagnosis at an earlier date would have been helpful and a relief for the parents. They would, for example, have been able get information about their children's prognoses from the literature about the clinical course of events for boys and girls with 17β-HSD-3 deficiency. Early testosterone treatment, if the correct diagnosis had been made, might have been extremely helpful. Likewise, the decision to perform a gonadectomy on the second child would possibly have been postponed to a later date. In retrospect, the discontinuity of care and the lack of a coordinating physician also had created problems. Deficiencies in prenatal and postnatal medical care in this case report were apparent. Care for children with rare disorders are often characterized by lack of timely and efficient referral to specialized centers with collaborating disciplines. In many countries, lack of registries for rare diseases and of registered subspecialty experts impair access to appropriate care. Efficient case management, comforting guidance from an interdisciplinary team with expertise in the management of children with intersex conditions, and psychosocial support may have reduced feelings of pressure and urgency in decision-making on the part of the parents. The false assumption of parental autonomous self management because they were health care professionals contributed to a lack of support and emphasis to proceed with diagnostic measures.

The growing knowledge about the development of children with different variations in gender development makes it more possible to make well-informed decisions about sex of rearing and about the type, timing, and extent of any medical intervention. It remains questionable whether it will be possible, in view of the heterogeneous causes and effects and of the imperfect genotype–phenotype correlation, to design a concept of medical treatment that does justice to all people concerned. Existing research results make it clear that the psychosexual and psychosocial development of people with intersex conditions takes a variety of courses and is influenced by many factors. Certain tendencies can

be extracted from the literature (e.g., relatively frequent formation of male gender identity in youths/adults with 17β-HSD-3 deficiency; Cohen-Kettenis, 2005; Sobel & Imperato-McGinley, 2004) but it is impossible to deduce any "facts" valid for all people concerned. Today, it is pointed out that there are no generally established "right" or "wrong" decisions, but that every family has to find its own path when dealing with the specific nature of their child (Thyen, Richter-Appelt, Wiesemann, Holterhus, & Hiort, 2005). In the reported case, long-term follow up will be necessary to monitor health and psychological development in particular during adolescence. The complexity of medical care and decision-making requires assistance from an interdisciplinary team. The parents, as well as the children and young people concerned, should be accompanied and supported on their path. A more exact analysis of the specific conditions and requirements for the decision-making process, a better understanding of the significance of meta-cognition, and a supporting framework of medical practitioners, psychologists, and self-help groups for parents of children with intersex conditions can be of help to them in the future in giving direct and relevant support beginning at the birth of their child and continued into childhood and adolescence (Stein, Sandberg, Mazur, Eugster, & Daaboul, 2003).

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