Archives of Sexual Behavior, Vol. 27, No. 2, 1998

Long-Term Psychological Evaluation of Intersex Children

Froukje M. E. Slijper, Ph.D.,[1,4] Stenvert L. S. Drop, M.D.,[2] Jan C. Molenaar, M.D.,[3] and Sabine M. P. F. de Muinck Keizer-Schrama, M.D.[2]

Treatment of psychological problems of 59 children with a physical intersex condition is described. The group consisted of 18 female pseudohermaphrodites with congenital adrenal hyperplasia (CAH), 20 male pseudohermaphrodites and 2 true hermaphrodites born with ambiguous external genitalia assigned the female sex (ambiguous girls), 14 male pseudohermaphrodites born with completely female external genitalia and assigned the female sex (completely female group), and 5 male pseudohermaphrodites born with ambiguous external genitalia and assigned the male sex. Despite the sex assignment, genital organ correction soon after birth, psychological counseling of parents and intensive psychotherapy of the children, general psychopathology developed equally in all 4 groups (39% of total group). Although 87% of the girls with a physical intersex condition developed in line with the assigned sex, 13% developed a gender identity disorder though only 1 girl (2%) failed to accept the assigned sex. Gender identity disorder and deviant gender role were in evidence only in girls with CAH and girls of the ambiguous group. Biological and social factors seem responsible for the development of gender identity disorder, such as pre- and postnatal hormonal influences on the brain enabling deviant gender role behavior to develop, and an inability on the part of parents to accept the sex assignment. A reconsideration of the sex assignment in male pseudohermaphrodites and true hermaphrodites born with ambiguous external genitalia is discussed.

[1]Department of Child and Adolescent Psychiatry, Sophia Children's Hospital, Erasmus University, Dr. Molewaterplein 60, 3015 GJ Rotterdam, The Netherlands.
[2]Department of Pediatrics, Division of Endocrinology, Sophia Children's Hospital, Erasmus University, Rotterdam, The Netherlands.
[3]Department of Pediatric Surgery, Sophia Children's Hospital, Erasmus University, Rotterdam, The Netherlands.
[4]To whom correspondence should be addressed.

0004-0002/98/0400-0125$15.00/0

KEY WORDS: gender identity disorder; congenital adrenal hyperplasia; male pseudohermaphroditism; true hermaphroditism; physical intersex condition.

INTRODUCTION

Sex assignment problems can emerge in male pseudohermaphrodites and true hermaphrodites born with ambiguous genitalia. Male pseudohermaphroditism occurs in individuals born with an XY karyotype or a mosaic form with disorders in the production, synthesis, or receptor sensitivity to androgens, so that their male genitalia are not sufficiently developed. In these children, the external genitalia are inadequately virilized, the gonads are located in the abdomen or inguinal canal, the uterus and ovaries are not in evidence, and the vagina has been inadequately formed, if at all. If the insensitivity to androgens is complete, as in the complete androgen insensitivity syndrome (CAIS), or the defect in the production of androgens is complete, as in Leydig cell hypoplasia, the child is born with female external genitalia and gonads that may be positioned in the abdomen or inguinal canal.

There is no doubt about the management of patients with CAIS or with a complete defect in the production or synthesis of androgens: The sex these patients are reared in is always female. However, sex assignment in the group of male pseudohermaphrodites and true hermaphrodites born with ambiguous genitalia is a difficult matter, since there are still no reliable criteria for their sex assignment. The debate about what is more important for gender identity development, the biological sex or the sex a child is reared in, is still going on. Money *et al.* (1955) were the first to emphasize the importance of upbringing. They felt that unambiguously raising a child with a physical intersex condition as a member of the assigned sex would be more important for the child's gender identity development than his or her chromosomal sex. Almost three decades later, Diamond (1982) demonstrated the controversial nature of this opinion with the well-known case history of the monozygotic twins, both of whom were genetic males at birth. Due to the accidental burning of his penis at the age of 7 months, one of the boys was assigned the female sex (Money and Tucker, 1975). Although raised as a girl, the patient could not identify with the female sex, and as an adult asked for sex reassignment. He now lives as a married man with his wife and adopted children (Diamond and Sigmundson, 1997).

Studies on female pseudohermaphrodites with congenital adrenal hyperplasia (CAH) (Money and Ehrhardt, 1968) have demonstrated that prenatal and postnatal hormones can cause sex-dimorphic behavior. CAH is an autosomal recessive disorder caused by a defect of one of the enzymes (in 90% the 21-hydroxylase) necessary for the formation of cortisone. This

results in increased levels of adrenal androgens, causing various degrees of virilization of the genitalia, such as labial fusion and clitoral enlargement. Several studies (Berenbaum and Hines, 1992; Dittmann *et al.*, 1990a, 1990b; Ehrhardt and Baker, 1974; Hines and Kaufman, 1994; Slijper, 1984) have demonstrated that girls with CAH exhibit more boyish conduct than other girls. Although there is more knowledge about the influence of prenatal hormones on gender role behavior, it is still unclear if and how this behavior can influence gender identity development and sexual orientation. In a study by Slijper (1984), CAH girls appeared to be significantly more boyish on a gender test than their sisters and healthy female peers. Girls with the salt-wasting form of CAH (SW) appeared to be significantly more boyish in this study than girls with the "simple" virilizing form of CAH (SV). The conclusion that only prenatal androgens were responsible for the deviant gender role behavior seems obvious, since girls with SW are more virilized at birth than girls with SV. However, the degree of boyish behavior in the group of girls with SW did not correlate with the degree of virilization of their external genitalia (Prader's degree). However girls whose sex was reassigned tended to exhibit more boyish conduct than girls whose female sex was immediately assigned at birth. This might indicate that the sex parents feel their child has influences how they deal with the child, thus enabling the child to develop gender models appropriate to that sex. So in addition to pre- and postnatal androgens, social factors also can influence the development of gender role and even gender identity development. As regards psychosexual development in women with CAH, some authors found it was delayed (Slijper *et al.,* 1992), whereas others noted an increased percentage of bi- and homosexuality (Dittmann *et al.,* 1992; Money *et al.*, 1984) or lower rates of exclusive heterosexual fantasy and fewer sexual experiences with men (Zucker *et al.*, 1996).

In addition to problems with the sex assignment, gender identity, and gender role behavior, children with a physical intersex condition and their parents can also have difficulty dealing with the diagnosis and accepting reconstructive surgery of the genitalia.

In this paper the psychological results of 10 years' work with children exhibiting a physical intersex condition are discussed. The team working with these children and their parents at Sophia Children's Hospital consisted of a child psychologist, a pediatric endocrinologist, a pediatric surgeon, and a pediatric urologist. The aim of the team was to prevent the development of cross-gender identification in children born with a physical intersex condition, especially in neonates born with ambiguous genitalia. This meant early sex assignment and early correction of their genitalia, and providing intensive and long-term counseling for the parents and later the child. The parents were openly informed about the physical and psychological nature

of their child's disorder, and thus had full knowledge of the condition. The children were informed in stages, depending upon their mental and emotional development. This program is described in Slijper *et al.* (1994) and is based on Money's (1992) guidelines for treating intersex patients.

METHODS

Psychological interventions could occur as follows: (i) as a component of the treatment by the entire team in decision-making and information-giving situations with reference to the diagnosis, sex assignment, or surgery on the genitalia; (ii) as a component of the treatment by part of the team (the psychologist in conjunction with the pediatric endocrinologist or the pediatric surgeon) in situations where further information was called for pertaining to the diagnosis, sex assignment, or surgery on the genitalia; and (iii) as a treatment by the psychologist in situations where coping with the physical intersex condition was called for.

Psychological treatment given by the psychologist (the first author) could consist of (i) individual psychotherapy: at least once every 2 weeks for a minimum of 1 year; (ii) short frequent psychotherapy: at least once every 3 weeks for anywhere from 3 months to 1 year; or (iii) infrequent long-term counseling: at least four times a year for longer than 1 year.

RESULTS

Description of the Patient Group

The patient group comprised 59 children referred to the Sophia Children's Hospital between 1984 and 1994. Female sex was assigned to 54 of the children and male sex to 5. Details on the diagnosis, karyotype, sex assignment, and reassignment are given in Table I.

To make it possible to compare the various groups, the total patient group was divided into four main groups: 18 female hermaphrodites (girls with CAH), 14 male pseudohermaphrodites born with completely female external genitalia assigned the female sex at birth (completely female), 20 male pseudohermaphrodites born with ambiguous external genitalia and 2 true hermaphrodites (ambiguous girls), and 5 male pseudohermaphrodites born with ambiguous external genitalia (ambiguous boys). The mean ages of the patients in the four groups are presented in Table II.

Table I. Diagnosis, Karyotype, Sex Assignment, and Sex Reassignment

Diagnosis[a]	*n*	Karyotype	Assigned sex	Sex reassignment and age of sex reassignment
CAH	18	46 XX	18 female	8 yes 10 no (6 at 0–3 months, 2 at 6 months)
CAIS	12	46 XY	12 female	12 no
Leydig cell hypoplasia	2	46 XY	2 female	2 no
PAIS	8	46 XY	7 female 1 male	7 yes, 1 no (12 months)
Extrophia cloacae	4	46 XY	4 female	4 no
Rudimentary gonadal syndrome	1	46 XY	1 female	1 no
17-keto-reductase deficiency	2	46 XY	2 female	2 no
Transversal penis	1	46 XY	1 female	1 no
Gonadal dysgenesis	9			
		2 46 XY	1 female 1 male	1 yes, 1 no (12 months)
		4 45XO 46XY	3 female 1 male	4 no
		1 46XO 46Xi, Yq 46X+ m	1 male	1 no
		1 45XO 46Xyfragment	1 male	1 no
		1 XY XX XXY	1 female	1 no
True hermaphroditism	2			
		1 XX	1 female	1 no
		1 XY	1 female	1 no

[a]CAH = congenital adrenal hyperplasia; CAIS = complete androgen insensitivity syndrome; PAIS = partial androgen insensitivity syndrome.

Table II. Mean Age at the Onset of General Psychopathology in Four Diagnostic Categories

Category	*n*	$\bar{x}$ age (years)	SD	Range
CAH girls	18	13.5	7.46	2–27
Completely female	14	12.4	8.52	3–26
Ambiguous girls	22	14.3	7.1	4–26
Ambiguous boys	5	12.4	5.8	6–18

Psychological Treatment

Neither all the children nor their parents could be treated by a psychologist starting from the time the diagnosis of the physical intersex con-

dition was made. Since many of the children had been diagnosed and treated elsewhere before being referred to the Sophia Children's Hospital, and 16 girls with CAH were not referred to the psychologist upon referral to the Sophia Children's Hospital, it was only in 39 of the 59 cases (53%) that a psychologist was involved with the children from the time of diagnosis. From the time the psychologist was first consulted, the development of all the children—the ones who needed psychological treatment as well as those who did not (42%)—was followed via counseling of the parents and incidental contact with the children. A total of 97% of the parents received psychological counseling (1 child was too old for the parents to be counseled, 1 child was at an institution where the parents were receiving counseling). Fifteen children received psychotherapy (26%), 7 (12%) received frequent psychotherapy for a short period, and 11 (19%) infrequent psychotherapy for a long period. For 1 child, family therapy was indicated.

In all types of treatment with the exception of family therapy, parents and children were treated separately. The purpose of treatment was to help parents cope with the sex assignment or sex reassignment, to help patients and their parents cope with the diagnosis and genital surgery, and with the behavioral and emotional problems of the child including gender role and gender identity.

Sex Assignment and Sex Reassignment

Girls with CAH

Of the 18 female pseudohermaphrodites, 2 had ambiguous genitalia at birth and were assigned the female sex on the grounds of a CAH diagnosis. In 8 children, the external genitalia were so severely virilized that the male sex was not doubted. Of these 8, 4 had their sex reassigned when they were younger than 1 month, 2 had theirs reassigned when they were 3 months old, and 2 when they were 6 months old. In the female pseudohermaphrodites with CAH, the advice of the team was always to assign the female sex, even if a child was so severely virilized as to have been assigned the male sex at birth and had already been reared as a boy for several months. The female karyotype and the female internal genitalia and the knowledge their child would be fertile as a female convinced the parents that their child was not a boy but a girl. Despite this clarity as to sex reassignment, dealing with it was still a difficult process and several parents, especially fathers, mourned the loss of their son. Four of the eight couples who were confronted with sex reassignment and four of the couples who were confronted with doubt about the gender of their child at birth received

immediate psychological help. Ten couples did not receive help until later. Although all parents continued to be concerned about the gender of their child, 89% were able to accept it to a reasonable degree.

The Completely Female Group

In 14 of the 39 male pseudohermaphrodites (12 with CAIS, 1 with Leydig cell hypoplasia, and 1 with gonadal dysgenesis), female sex was not doubted at birth because the external genitalia were completely female. The diagnosis was made in 3 girls at the age of 16 or 17, in 2 girls at the age of 8, and in 10 girls before they were 2 years old. In none of these girls was sex reassignment advised by the team and in none was sex reassignment considered by the parents. Eleven (79%) of the 14 couples who had a child with this condition received psychological counseling starting from the time the diagnosis was made. Although all 14 couples continued to be concerned about their daughters' XY chromosome pattern, 11 of the couples (79%) were able to satisfactorily start dealing with it.

Ambiguous Girls and Ambiguous Boys

Sex assignment was questioned at birth in 27 of the 39 male pseudohermaphrodites and 2 true hermaphrodites (69%), because they either had ambiguous external genitalia (22) or incomplete or improperly constructed male genitalia (4 with extrophia cloacae and 1 with transposition of the penis). Male sex was assigned to 5 of them (ambiguous boys) and female sex to the other 22 (ambiguous girls). The sex of 2 of the children (1 with partial androgen insensitivity syndrome and 1 with gonadal dysgenesis) who were first considered female was revised twice. Both of these children, who were assigned the female sex for good when they were about 1-year-old, were born about 20 years ago, at a time when experience in diagnosing and counseling children with a physical intersex condition was very limited.

Parents of babies with male pseudohermaphroditism or true hermaphroditism who were born with ambiguous genitalia had a hard time making the sex assignment decision. The function of the team was to help them make this decision by providing information about the somatic and psychological consequences of sex reassignment for the development of their child and by making recommendations in this connection (Slijper *et al.*, 1994, Money *et al.*, 1986, Money and Norman, 1987). However, the final decisions were always made by the parents. The sex assignment criteria the team used were the length of the phallus, its capacity for reacting to testosterone stimulation (occasionally a trial of androgen treatment could be performed), the

extent to which the male external genitalia would be functional in adulthood, and the capacity of the body to virilize at puberty. On the basis of these criteria, the parents of four of the children decided to have male sex assigned. One couple, whose child had a partial insensitivity syndrome opted for male sex despite the penoscrotal hypospadias with a phallus of 2 cm and the fact that the gonads had not descended into the scrotum. There were other patients with the same disorder in the family, who had developed into socially and psychologically healthy male adults despite repeated surgical correction of their genitalia. At the moment, the patient is at nursery school and has not developed any signs of gender pathology or psychopathology. The parents are well aware of the risks of stigmatization and deal with their son in a way that is adequate and supportive.

Of the 27 couples (59%) with a child in the ambiguous group, 16 received immediate help in dealing with the lack of gender clarity and 11 couples (41%) did not receive help until much later because they had been referred at a late stage to Sophia Children's Hospital. Despite the intensive counseling the parents received, 50% were not able to work through the trials and tribulations their child's lack of gender clarity entailed. Two mothers and 1 father openly rejected their child as a result. The following factors played a role in the acceptance process: (i) the time when assistance was offered for 5 couples who had problems dealing with their child's lack of gender clarity, help came too late, since it had already been several years since the trauma; (ii) the instability of the marriage for 5 couples, the child's anomaly played an important role in their divorce; (iii) the number of times the sex assignment was revised the 2 couples whose child's sex assignment was revised twice continued to doubt whether they had made the right decision regarding the sex assignment; and (iv) the personality structure of the parents, particularly as regards rigidity and the inability to cope with setbacks and tolerate embarrassment.

As regards the children themselves, successful coping with the physical intersex condition seemed to be determined by the flexibility or resilience of their personality structure and the immediate psychological help they received.

Genital Surgery

Reconstruction of External Genitalia

The team policy was to correct the virilization of the external genitalia immediately after birth or as soon as possible after the diagnosis was made so as to avoid cross-gender identification. In cases of severe virilization,

the correction of the genitalia was always recommended before the child's first birthday. The stronger the virilization of the genitalia, the less difficult the decision to have the, correction performed immediately appeared to be for the parents. If there was only an enlarged clitoris, the correction was often not such an obvious choice for the parents. In these cases, the operation was postponed until the child was old enough to make his or her own decision. From the age of 4, the children were able to express their own opinion in interviews or in play with the psychologist about clitoris reduction by stating their serious concerns about the length of the clitoris or its erectile function. These conversations were helpful for the children and their parents in deciding to perform the operation and preparing the child for it, as well as in refraining from surgery in cases where the child was proud of her large clitoris and enjoyed the erections. Gender identity problems appeared to contraindicate clitoris reduction.

Feelings of shame and guilt sometimes made parents deny their child's genital anomaly, which remained visible even after the correction, thus depriving the child of an opportunity to deal with it. This denial was often reinforced by the overly positive evaluation of the surgeon, who viewed the results of the operation from the angle of what was technically feasible. Working through the rage and sadness about the genital anomaly in psychotherapy appeared to have a positive effect on the acceptance process.

Vaginal Plastic Surgery and Vaginal Dilatation

The team policy was to start preparing a child for vaginal plastic surgery at the age of 11, and to consult them about when to operate. Vaginal surgery was performed on 9 children, 7 of whom could be prepared for it in advance; one of the 7 girls was 12 at the time, 4 were 14, and 2 were 16 years old. In the case of the 12-year-old girl, the fact that she was already menstruating and could not discharge the menstrual blood was the reason for surgery. At present, 10 children are being prepared for vaginal plastic surgery in the future. Two children with severe psychological complaints who had first been treated elsewhere came for help after operation. Dilating the vagina at a younger age appeared to lead to severe psychological problems because it was experienced as a violation of the body integrity.

General Psychopathology

Psychiatric diagnoses were based on a semistructured psychiatric interview with the parents and patients separately. A diagnosis of psychopathology, whether general psychopathology or gender identity disorder

(GID), was based on the diagnostic classification system of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV, American Psychiatric Association , 1994).

Of the total group of 59 patients, 25 children (42%) had no psychological problems. Mild psychological problems were observed in 11 children (19%). They had problems with their gender role, the operation on their genitalia, and with the diagnosis itself. General psychopathology (GID not included) that met the diagnostic criteria of DSM-IV was observed in 23 children (39%). General psychopathology occurred twice as often among children and parents who had not received counseling starting at the time of the initial diagnosis. There was a wide range of general psychopathology according to DSM-IV criteria, as is shown in Table III.

Two of the boys, both with gonadal dysgenesis, had a DSM-IV diagnosis, one an obsessive compulsive disorder and the other a conduct disorder.

In the total group of 54 girls, 21 had at least one DSM-IV diagnosis of general psychopathology. Eight children had more than one DSM-IV diagnosis. The mean age at the onset of general psychopathology (mental retardation not included) in girls was 9.8, SD = 4.4. If we look at the general psychopathology in the girls according to the diagnostic categories, 22% of the CAH girls exhibited general psychopathology, as did 50% of the completely female group and 46% of the ambiguous girls. The relationship between the diagnostic category and DSM-IV diagnosis was not significant , $\chi^2(2) = 3.23$, $p = 0.2$. The prognosis of treatment was positive in 4 of the 5 CAH girls, in 4 of the 6 girls from the completely female group, and in 6 of the 8 girls in the ambiguous girl group. The relationship between the diagnostic category and the prognosis was not significant, $\chi^2(2) = 0.26$, $p = 0.88$. The mean age of all the girls at the onset of the general psychopathology was 9.84, SD = 4.4. The classification of the DSM-IV diagnosis of internalizing disorders such as depressive neurosis, anxiety disorder, selective mutism, and sexual disorder not otherwise specified (NOS) and externalizing disorders such as oppositional defiant disorder, attention deficit hyperactivity disorder, and conduct disorder results in the following distribution of internalizing and externalizing disorders in patients of the three diagnostic categories: 75 and 25% in CAH girls, 67 and 33% in the completely female group, 56 and 44% in the ambiguous group of girls.

Gender Identity Disorder

None of the 5 boys had a gender identity disorder. However 7 of the 54 girls (13%) exhibited gender identity disorder of childhood, as defined

Table III. Patients with General Psychopathology (DSM-IV Diagnosis) and/or GID, Deviant Gender Role, and Physical Intersex Diagnosis[a]

Category	Intersex diagnosis	DSM diagnosis	Age at onset of DSM diag.	GID	Age at onset of GID	Deviant gender role
CAH girls	CAH sw	Selective mutism	10	No	—	Yes
	CAH sv	Ment. retard+ anxiety dis.	16	No	—	No
	CAH 11beta	Ment. retard+ ODD	6	Yes	4	Yes
	CAH sw	Sexual problems	16	No	—	Yes
	CAH sw	—	—	Yes	3	Yes
Complete female	CAIS	ODD	10	No	—	No
	CAIS	Depressive neur.	17	No	—	No
	CAIS	ODD	5	No	—	No
	CAIS	Ment.retard	—	No	—	No
	CAIS	Anxiety dis.	6	No	—	No
	CAIS	Depressive neur.+ sex. probl.	7/16	No	—	No
	Leydig Cell Hyp.	Depressive neur.+ sex. probl.	17/17	No	—	No
Ambiguous girls	PAIS	Depressive neur.+ sex. probl.	13/16	Yes	10	Yes
	Leydig Cell Hyp.	Ment.retard+ ADHD	6	No	—	No
	Extr. cloacae	ODD	6	Yes	4	Yes
	Extr. cloacae	Ment.retard+ sex. probl.	—/15	No	—	Yes
	Rud. gonal. synd.	Mental. retard	—	No	—	Too young
	17-Keto-red.def.	Depressive neur.+ sex. probl.	10/18	No	—	Yes
	Trans. penis	Conduct disorder	7	Yes	4	Yes
	Gonad. dys.	Depressive neur.+ sex. probl.	7/12	Yes	4	Yes
	Gonad. dys.	Anxiety disorder	8	No	—	Yes
	True herm.XY	ODD	5	Yes	5	Yes
Ambiguous boys	Gonad. dys.	OCD	10	No	—	No
	Gonad. dys.	Conduct disorder	9	No	—	No

[a]CAH = congenital adrenal hyperplasia, sw = 21-OH deficiency, salt wasting, sv = 21-OH deficiency, simple virilizing, 11 beta = 11 beta-OH deficiency, CAIS = Complete androgen insensitivity syndrome, PAIS = partial insensitivity syndrome, Leydig Cell Hyp = Leydig Cell hypoplasia, 17-Keto-red. def. = 17-keto-reductase deficiency, Trans. penis = transversely constructed penis, Rud. gonal. synd. = Rudimentary gonadal syndrome, True herm = True hermaphroditism, Extr. cloacae = Extrophia cloacae, Gonad. dys = Gonadal dysgenesis, Ment. retard = mental retardation, Depressive neur. = depressive neurosis, sex. probl. = sexual problems-NOS, ODD = oppositional defiant disorder, OCD = obsessional compulsive disorder.

in the DSM-IV, with intense sadness and dissatisfaction with the assigned sex and a preference for behavior appropriate to the other sex. Two of these girls had CAH, 1 had a partial androgen insensitivity syndrome, 1 was a true hermaphrodite with an XY karyotype, 1 had extrophia cloacae, 1 had a transversely constructed penis, and 1 had gonadal dysgenesis. Of the 7 girls with GID, 4 had experienced sex reassignment. Six of the 7 children exhibited general psychopathology in addition to GID. Three girls developed an oppositional defiant disorder, 1 a conduct disorder, and 2 a depressive neurosis later followed by a sexual disorder-NOS. Problems in the family due to divorce or the death of one of the parents affected 6 of the 7 patients (86%). This was true in 24% of the total group. In addition, 3 of the 7 children were openly rejected by one or both of their parents as a consequence of their condition.

For 3 of the children, intensive psychotherapy had a positive effect in that they could accept their female gender. Three of the children are still being treated. In the case of 1 child, the treatment had no effect; she is a young adult today, and became extremely unhappy with her female sex after the start of her puberty. That was when she discontinued the psychotherapy she had been having since she was 10. The parents of this child, whose sex was changed twice after birth without any counseling, had serious doubts about whether they had been right to have the female sex assigned.

In the group of 6 girls with GID and general psychopathology, the mean age at the onset of GID (4.86, SD = 2.34) was significantly lower, $t(5) = 4.54$, $p = 0.006$, than the mean age at the onset of general pathology (7.33, SD = 2.9). Although this group was small, the mean age at the onset of general pathology in the total group of girls (9.8) suggests that GID developed at a younger age than the general psychopathology.

If we look at the relationship between diagnostic categories and GID, there are significantly more girls with GID in the ambiguous group than in the completely female group in which GID was absent, $\chi^2(1) = 3.89$, $p = 0.05$, but not more in the CAH group, $\chi^2(2) = 4.26$, $p = 12$.

In the treatment of the GID, making the patient aware of the conflict between the fantasy of being a boy and the reality of having the female gender occupied a central position. The child often had to be informed about the doubts there had been about her sex right when she was born. Working through the sadness and anger about not being able to be a boy was an important aspect of the treatment. For the parents as well, treatment was necessary to dispel their doubts about whether they had been right in having the female sex assigned to their child. Doubts on the part of the parents unwittingly promoted the child's gender fantasies.

Gender Role Behavior

Although deviant gender role behavior was not in evidence in the boys, their behavior was not very assertive. The boys were fearful and bothered about the smallness of their penis.

Deviant gender role behavior was not only exhibited by the girls with a gender identity disorder, it was noted in 25 (46%) of the total group of girls. Two of the children were still too young to have their gender role behavior evaluated. Of the 25 girls with boyish conduct, 12 had CAH, 12 were male pseudohermaphrodites born with an ambiguous external genital, and 1 was a true hermaphrodite born with an XY karyotype. Deviant gender role behavior was not in evidence in the completely female group. Deviant gender role behavior was significantly more frequent in the ambiguous girls compared to the completely female group, $\chi^2(4) = 24.06, p = 0.00008$, but not significantly more frequent in the CAH girls compared to the ambiguous girls. In the ambiguous girls, the deviant gender role behavior was more frequently a source of concern for parents regarding their daughter's gender identity, than in the CAH girls. Apparently the XY chromosome pattern remained a source of uncertainty for parents as regards their daughters' gender identity development. The boyish conduct was perceived as an indication that the decision to assign the female sex had been wrong. In particular, the wild, rough play of these children was difficult for their parents to regulate, so that secondary behavior problems developed in the form of oppositional conduct.

DISCUSSION

Despite early sex assignment (93% within the first 4 weeks of their lives and all before they were 1-year-old) and correction of the genitalia at a young age, which was the policy of the team to avoid the development of cross-gender behavior and psychological problems, 39% of the children developed severe general psychopathology. Since a psychologist was consulted immediately in only 53% of the cases to help the patients and their parents cope with the anomaly, one might wonder whether this did not account in part for the high percentage of severe psychopathology. Psychological problems were twice as prevalent: among the children who did not receive help starting as soon as the diagnosis was made as among those who did. Early counseling thus seems to have had a preventive effect, although it could not keep all the children from exhibiting signs of psychopathology.

General psychopathology was in evidence in the girls of all three diagnostic categories, as well as in boys. Since gender identity disorder developed significantly earlier than general psychopathology in girls, GID can be an explanation for the development of general psychopathology in girls with GID, but it does not explain the general psychopathology in girls without GID. Deviant gender role development can add to the development of general psychopathology in girls with CAH and in girls in the ambiguous group, but it cannot explain general psychopathology in the completely female group because all had normal female gender role behavior. If we look at the type of general psychopathology in the three diagnostic categories, there is no difference between these groups in the proportion of internalizing and externalizing disorders. CAH girls and male pseudohermaphrodites born with ambiguous genitalia and assigned the female sex thus did not develop more aggressive behavior that could be classified as externalizing behavior problems than male pseudohermaphrodites born with completely female external genitalia and assigned the female sex. This means that there is no indication that general psychopathology is related to pre- and postnatal androgenization of the brain. Although general psychopathology is not uncommon in children with a chronic disease (Wallander and Thompson, 1995), only girls with CAH can be viewed as having such a condition. However, compared with healthy children, children with a physical intersex condition are confronted more with developmental interferences, such as sex reassignment, hospitalization, sex operations, lifelong dependence on hormone substitutes, and infertility. Other factors in the children themselves could have also played a role, such as greater congenital vulnerability (6 of the children were mentally retarded, 5 of them mild and 1 severely) and a reduced tolerance for stress.

From these results, it seems obvious that there are also parental influences. The anomaly was particularly difficult for parents to cope with if their daughter was born with ambiguous external genitalia and an XY karyogram, since this was a diagnosis 50% of these parents did not know how to deal with. Although parents with a daughter in the completely female group were more able to cope with the condition of their child, they continued to be concerned about their daughters' XY chromosome pattern. It is possible that this inability on their part was one of the causes of the psychopathology.

All the girls who developed a GID also exhibited boyish conduct. Studies with CAH girls have shown that hormonal influencing can be the cause (Berenbaum and Hines, 1992; Dittmann *et al.*, 1990a, 1990b; Ehrhardt and Baker, 1974; Hines and Kaufman, 1994; Slijper, 1984). All 7 female children with a physical intersex condition had been exposed prenatally to the influence of an excess of male hormones and were susceptible to them. Gen-

der identity disorder and deviant gender role were equally present in the girls with CAH and the girls of the ambiguous group, but absent in the girls of the completely female group. Girls with GID developed more externalizing behavioral problems (3 had an oppositional defiant disorder and 1 a conduct disorder) than internalizing behavioral problems (2 had a depressive neurosis and later a sexual problem-NOS). In 3 of the 7 girls with GID, the associated psychopathology was oppositional defiant disorder and in 1 a conduct disorder, both disorders of the externalizing type where aggression plays a major role. These results suggest that GID is associated with pre- and postnatal androgenization of the brain. However, pre- and postnatal virilization of the brain cannot be the only cause of a GID, since boyish conduct was also evident in children in our group who did not develop GID, and most of the girls with very virilized external genitalia developed in line with their assigned sex. In the group of 59 children, the 3 whose parents openly rejected them all developed GID, as did the 2 children whose sex assignment was altered twice. Of the 7 children with GID, 6 also exhibited other severe psychopathology, and 6 of the 7 children were confronted with family problems due to a divorce or a death in the family. The combination of boyish conduct and problems in the parent–child relationship appears to disturb the gender identification process. Zucker *et al.* (1987) also noted a combination of hormonal and psychosocial factors in a girl with true hermaphroditism as an explanation for her gender dysphoria.

Although data are not available on the prevalence of GIDs in the normal population, Verhulst *et al.* (1985) noted on the Child Behavior Checklist that 0–1% of the boys and 2–5% of the girls of the Dutch normal population of 4–11 years of age would rather be of the other sex (as reported by their parents). This sex ratio can be explained by the greater tolerance in our society for cross-gender behavior in girls than boys. However, in clinical samples of physically normal children with gender identity conflicts, boys are seen 6 to 8 times as frequently as girls (Meyer-Bahlburg, 1994, Zucker and Bradley, 1995). As explanations, Zucker and Bradley cited biological factors (male fetal development is more complex than female fetal development) as well as social factors (the peer group is less tolerant of cross-gender behavior in boys than in girls and parents appear much more concerned with cross-gender behavior in boys than girls). In our study, the situation seems to be reversed, since gender identity problems were more common in the girls than in the boys. However, first our girls were genetic males who had had an atypical fetal development, and second, their parents appeared to be constantly alert to signs of gender identity disturbances and were very open to psychological treatment, which

was easily available for them since they had regular contacts with the psychologist.

Why are girls with an intersex condition at a greater risk of developing GID than boys? Maybe it is too much of a burden for these girls to try to develop gender role behavior and gender identity that goes against the pre- and postnatal forces of the androgens. It is possible that the conflict between biological and psychological forces can produce stress which, in a genetically vulnerable child who grows up in a family unable to raise the child unambiguously in the assigned sex, results in GID and general psychopathology. Although the external genitalia of these girls are always corrected at an early age to give them an opportunity to live with completely female genitalia, the result of the surgery in severely virilized girls is not always optimal. This means these girls are often aware of their ambiguity and are inclined to ask their parents about their gender. Since the wish to be of the opposite sex is greater in girls than in boys, it is possible that girls with an intersex condition mourn the loss of their male sex. This mourning was observed in the psychotherapy of the girls with a GID. Meyer-Bahlburg *et al.* (1996) concluded from a study of gender change from female to male in four adult CAH 46,XX individuals (3 with poor adherence to glucocorticoid treatment, and 1 with late treatment), that gender atypical behavioral self-image and gender atypical body image are more important factors in the development of gender change than sex-typing bias on the part of the parents, a particular genotype or endrocrinotype.

In most of the girls, gender identity appeared to develop in line with their assigned sex, even if their gender role behavior was atypical. Some of the girls developed gender identity problems that did not meet DSM-IV criteria for GID. But 13%, a substantial percentage, of our patients developed, GID that met DSM-IV criteria. Meyer-Bahlburg (1994) noted "that patients with intersexuality or similar medical conditions should be excluded from the GID diagnosis," because they differ from nonintersex patients in the age at onset, the presentation, and the sex ratio of their GID. The age at the onset of GID was very young in our group, just as in nonintersex children: 6 of the 7 girls were referred to the psychologist for GID before they were 5 years old and retrospectively the parents reported that the gender identity problems had been observed from the moment ($1\ ^1/_2$–2 years of age) the children became aware of their gender.

Since 5 of the 7 girls were genetic males, we can rationalize that the sex ratio was not different from biological normal boys with a GID. But in the presentation of their GID, our patients were different from nonintersex patients; the obsessive preoccupation with gender dysphoria was clearly absent, since they consciously knew—having been informed by their parents about their condition—or unconsciously knew—since they felt boy-

ish and knew their genitalia were not completely female—that their gender wish was genetically right. Also 4 of the 7 patients with GID exhibited externalizing general psychopathology, a finding that is not consistent with Zucker and Bradley (1995) in biologically normal boys and girls with GID. They found a predominance of internalizing symptomatology in boys as well as in girls with GID.

Of the 7 children with GID, 6 were effectively treatable. All 6 were 6 years old or younger when they began treatment. Four of the 6 experienced sex reassignment in the neonatal period, one at 3 months, and one child was sex-reassigned twice (in the neonatal period and at the age of 1). The one child whose treatment was not successful did not start until the age of 10 and was also sex-reassigned twice (in the neonatal period and at the age of 1). It is possible that there is a critical period for sex identification changes, after which treatment is no longer effective. Money *et al.* (1957) also mentioned a critical period for sex identification changes. They reported that 1 out of 4 of the children with an intersex condition who experienced a sex reassignment after age 27 months was not able to adjust to the change. However, 11 of the 14 children who were sex reassigned prior to the age of 27 months adjusted to the change without complications. This sensitive-period hypothesis has been refuted, however, by reports of successful reversal in gender identity even after toddlerhood. The best known examples of such a reversal are the patients with 5-α-reductase deficiency described by Imperato-McGinley *et al.* (1979). But there are also more recent examples, such as the four 46XX CAH individuals who changed gradually from female to male (Meyer-Bahlburg *et al.*, 1996), a patient with partial androgen insensitivity syndrome who was raised as a girl but applied for sex reassignment at the age of 33 (Gooren and Cohen-Kettenis, 1991), or the teenage girl who was unambiguously raised as a girl but declared himself a male at the age of 14, when the diagnosis of mixed gonadal dysgenesis was made (Reiner, 1996). Gender change in hermaphroditic children are almost always from female to male with the exception of CAH 46, XX individuals.

Studies on sexual behavior of both sexes in the normal Dutch population (Mureau *et al.*, 1995) have shown that the average age for the first French kiss is 12.6 years and for the first sexual intercourse 14.8. On the basis of these figures, one might conclude that 12 is a proper age for vaginal plastic surgery, giving the child ample time to integrate her feelings about her vagina into her body image before she is ready for sexual intercourse. The children in our study did not appear to be ready for vaginal plastic surgery until somewhere between the ages of 14 and 16. The strain of vaginal dilatation certainly played a role in postponing the operation, but it is also possible that children with a physical intersex condition disorder tend to be late psychosexual developers (Slijper *et al.*, 1992).

Recommendations

Although 87% of the girls with a physical intersex condition developed in line with the assigned sex, 13% developed a GID, but only one (2%) failed to accept the assigned sex. Although there is no hard evidence of a relationship between early assignment to the sex other than the biological one, the general psychopathology percentage of 35% in girls suggests that a physical intersex condition constitutes a high risk for the development of psychopathology. On the grounds of these results, we should consider the possibility that sex assignment to the sex opposite the chromosomal sex can be psychologically harmful to genetic males or strongly virilized genetic females. Diamond (1996) is of the opinion that sex assignment and genital surgery should be delayed until the child can decide for itself. This means the child should neither be raised as a boy nor a girl, but as an intersex person. Our study shows that children can say at a young age whether or not they want to have their genital organ corrected, but we feel it is beyond the capability of a child to develop an intersex identity. The two patients in our group who were sex-reassigned twice both developed a GID. The period of uncertainty as to the gender assignment was traumatic for the parents and the child and remained a constant source of concern. Children born with an intersex condition should be assigned either male or female sex and be raised unambiguously in the assigned sex; from this safe psychological and social position, judging from their gender feelings, they can decide whether the assigned sex is right or wrong. This means parents have to decide about the gender of their child at birth. Postponement does not seem to be a reasonable option, but a reconsideration of the sex assignment criteria especially in genetic males born with ambiguous genitalia, is inevitable.

Construction of a neophallus is feasible although still at an experimental stage (Horton, 1995); it can be promising for future genetic males born with ambiguous external genitals. The assignment of the biological sex of a neonate with partial insensitivity syndrome remains problematic, even with phallic reconstruction, since these boys virilize poorly at puberty in spite of hormone substitution (Berkovitz, 1995). But assignment to the biological sex seems more reasonable for neonates with cloacal exstrophy, with a severe translocation of the penis, or with a partial disturbance in the production or synthesis of androgens since these patients virilize normally in puberty, if necessary with the help of hormones. In puberty they could weigh the advantages and disadvantages of phallic construction against those of living with a micropenis. Female sex assignment in girls with CAH, even if they have to be reassigned as a girl after living for several months as a boy, is the best option since these girls feminize normally in puberty

and have normal fertility. Meyer-Bahlburg *et al.* (1996) stated that early corrective genital surgery may risk functional impairment while improving the maintenance of gender consistency.

Since our study shows that children with a physical intersex condition constitute a high-risk group as regards the development of psychopathology including GID, the long-term treatment of parents and children should start as soon as the diagnosis is made, so that psychopathology can be detected and treated at an early stage.

ACKNOWLEDGMENTS

We thank Dr. J. M. Nijman and Dr. J. C. Scholtmeijer, pediatric urologists, for their efforts on our team, and Dr. J. M. Koot and Dr. H. J. M. Versluis-den Bieman for their help.

REFERENCES

American Psychiatric Association. (1994). *Diagnostic and Statistical Manual of Mental Disorders*, 4th ed., rev., APA, Washington, DC.

Berenbaum, S. A., and Hines, M. (1992). Early androgens are related to childhood sex-typed toy preferences. *Psychol. Sci.* 3: 203-206.

Berkovitz, G. D. (1995). Gender assignment in newborns with ambiguous genitalia. *Dialog. Pediat. Urol.* 18: 7-8.

Diamond, M. (1982). Sexual identity, monozygotic twins reared in discordant sex roles, and a BBC follow-up. *Arch. Sex. Behav.* 11: 181-185.

Diamond, M. (1996). Sex assignment considerations. *J. Sex Res.* 22: 161-174.

Diamond, M., and Sigmundson, K. (1997). Sex reassignment at birth. *Arch. Pediat. Adolescent Med.* 151: 298-304.

Dittmann, R. W., Kappes, M., and Kappes, M. H. (1992). Sexual behavior in adolescent and adult females with congenital adrenal hyperplasia. *Psychoneuroendoc rinology* 17: 153-170.

Dittmann, R. W., Kappes, M. H., Kappes, M. E., Börger, D., Stegner, H., Willig, R. H., and Wallis, H. (1990a). Congenital adrenal hyperplasia: II Gender-related behavior and attitudes in female saltwasting and simple virilizing patients. *Psychoneuroendoc rinology* 15: 421-434.

Dittmann, R. W., Kappes, M. H., Kappes, M. E., Börger, D., Stegner, H., Willig, R. H., and Wallis, H. (1990b). Congenital adrenal hyperplasia: I. Gender-related behavior and attitudes in female patients and sisters. *Psychoneuroend ocrinology* 15: 401-420.

Ehrhardt, A. A., and Baker, S. W. (1974). Fetal androgens, human central nervous system differentiation, and behavior sex differences. In Friedman, R. C., Richart, R. M., and van de Wiele, R. L. (eds.), *Sex Differences in Behavior*, Wiley, New York.

Gooren, L., and Cohen-Kettenis, P. T. (1991). Development of male gender identity/role and sexual orientation towards women in a 46,XY subject with an incomplete form of the androgen insensitivity syndrome. *Arch. Sex. Behav.* 20: 459-471.

Hines, M., and Kaufman, F. R. (1994). Androgen and the development of human sex-typical behavior: rough-and-tumble play and sex of preferred playmates in children with congenital adrenal hyperplasia (CAH). *Child Dev.* 65: 1042-1053.

Horton, C. E., Jr. (1995). Total phallic reconstruction. *Dialog. Pediat. Urol.* 18: 4-7.

Imperato-Mc Ginley, J., Peterson, R. E., Gautier, T. (1979). Androgens and the evolution of male-gender identity among male pseuodohermaphrodites with 5 alphareductase deficiency. *New Engl. J. Med.* 300: 1233-1237.

Meyer-Bahlburg, H. F. L. (1994). Intersexuality and the diagnosis of gender identity disorder. *Arch. Sex. Behav*. 23: 21-40.

Meyer-Bahlburg, H. F. L., Gruen, R. S. , New, M. I., Bell, J. J., Morishima, A., Shimshi, M., Bueno, Y., Vargas, I., Baker, S. W. (1996). Gender change from female to male in classical congenital adrenal hyperplasia. *Horm. Behav.* 30: 319-332.

Money, J. (1992). Psychological aspects of disorder of sexual differentiation. In Carpenter, S. E., and Rock, J. A. (Eds.), *Pediatric and Adolescent Gynecology*, Raven, New York.

Money, J., Devore, H., and Norman, B. P. (1986). Gender identity and gender transposition: longitudinal outcome study of 32 male hermaphrodites assigned as girls. *J. Sex Marital Ther.* 12: 165-178.

Money, J., and Ehrhardt, A. A. (1968). Prenatal hormonal exposure: Possible effects on behaviour in men. In Michael, R. P (ed.), *Endocrinology and Human Behavior*, Oxford Press, London.

Money, J., Hampson, J. G., and Hampson, J. L. (1955). Hermaphroditism: Recommendations concerning assignment of sex, change of sex and psychological management. *Bull. Johns Hopkins Hosp.* 97: 284-300.

Money, J., Hampson, J. G., and Hampson, J. L. (1957). Imprinting and the establishment of gender role. *Arch. Neurol. Psychiat.* 79: 333-337.

Money, J., and Norman, B. P. (1987). Gender identity and gender transposition: longitudinal outcome study of 24 male hermaphrodites assigned as boys. *J. Sex Marital Ther.* 13: 75-92.

Money, J., Schwartz, M., and Lewis, V. (1984). Adult erotosexual status and fetal hormonal masculinization and demasculinization: 46,XX congenital virilizing adrenal hyperplasia and 46,XY androgen-insensitivity syndrome compared. *Psychoneuroendocrinology* 9: 405-414.

Money, J., and Tucker, P. (1975). *Sexual Signatures: On Being a Man or Woman,* Little, Brown, Boston.

Mureau, M. A. M., Slijper, F. M. E., van der Meulen, J. C., Verhulst, F. C., and Slob, A. K. (1995). Psychosexual adjustment of men who underwent hypospadias repair: A norm-related study. *J. Urol.* 154: 1351-1355.

Reiner, W. G. (1996). Case study: sex reassignment in a teenage girl. *J. Am. Acad. Child Adolescent Psychiat.* 35: 799-804.

Slijper, F. M. E. (1984). Androgens and gender role behavior in girls with congenital adrenal hyperplasia (CAH). In de Vries, G. J, de Bruin, J. P. C, Uylings, H. B. M, and Corner, M. A. (eds.), *Progress in Brain Research*, Vol. 61, Elsevier, Amsterdam.

Slijper, F. M. E., Drop, S. L. S., Molenaar, J. C., and Scholtmeijer, R. J (1994). Neonates with abnormal genital development assigned the female sex: parent counselling. *J. Sex Educ. Ther.* 20: 9-17.

Slijper, F. M. E., van der Kamp, H. J., Brandenburg, H., de Muinck Keizer-Schrama, S. M. P. F., Drop, S. L. S., and Molenaar, J. C. (1992). Evaluation of psychosexual development of young women with congenital adrenal hyperplasia; a pilot study. *J. Sex Educ. Ther.* 18: 200-207.

Verhulst, F. C., Akkerhuis, G. W., and Althaus, M. (1985). Mental health in Dutch children: I. A cross-cultural comparison. *Acta Psychiat. Scand.* 72(Suppl. No. 323).

Wallander, J. L., and Thompson R. J. (1995). Psychosocial adjustment of children with chronic physical conditions. In Roberts, M. C. (Ed.), *Handbook of Pediatric Psychology*, Guilford, New York.

Zucker, K. J., and Bradley, S. J. (1995). *Gender Identity Disorder and Psychosexual Problems in Children and Adolescents*, Guilford, New York.

Zucker, K. J., Bradley, S. J., and Huges, H. E. (1987). Gender dysphoria in a child with true hermaphroditism. *Can. J. Psychiat.* 32: 602-609.

Zucker, K. J., Bradley, S. J., Oliver, G., Blake, J., Fleming, S., and Hood, J. (1996). Psychosexual development of women with congenital adrenal hyperplasia. *Horm. Behav.* 30: 300-318.