Psychiatric Manifestations in Young Females with Congenital Adrenal Hyperplasia in Taiwan

Hsin-Yi Liang, MD; Hsueh-Ling Chang, MD; Ching-Yen Chen[1], MD; Pei-Yeh Chang[2], MD; Fu-Sung Lo[3], MD; Li-Wei Lee[4], MS

Background: Congenital adrenal hyperplasia (CAH) is a syndrome of prenatal and/or postnatal androgen excess secondary to genetic deficits in the cytochrome p450 enzymes of the cortisol synthesis pathway. Women with CAH may suffer from different degrees of androgenization. This study documented psychiatric manifestations in young women born with CAH in Taiwan.

Methods: From July 1, 2005 to February 31, 2006 the psychiatric morbidity, gender identity and sex-typed behavior during childhood of 11 young women with CAH were assessed either using questionnaire-based semi-structured interviews or self-reported questionnaires.

Results: Of the 11 young women (mean age 15.3 ± 5.6 years), 3 (27.3%) had salt-wasting CAH and 8 (72.7%) had simple-virilizing CAH. Two patients without prior gender assignment (delayed diagnosis) were evaluated for primary amenorrhea at age 19 and 24 years, respectively. Four of the eleven participants (36.4%) had had an axis I psychiatric diagnosis within the past year. The subjects with more atypical sex-typed behavior in childhood tended to have a higher risk of minor psychiatric illness ($r = -0.706$, $p < 0.05$).

Conclusions: This study examined the impact of CAH on the patients' psychological well-being. Psychiatric needs were found to be unmet. Earlier psychiatric evaluation and intervention for these patients is suggested.

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Key words: congenital adrenal hyperplasia (CAH), psychiatric comorbidity

Congenital adrenal hyperplasia (CAH), an autosomal recessive disorder, is one of the intersex disorders. It is primarily a family syndrome of prenatal and/or postnatal androgen excess secondary to genetic deficits in the cytochrome p450 enzymes of the cortisol synthesis pathway. The incidence is about 1 in 5,000 to 15,000 live births in Caucasians[1] and about 1 in 20,000 in Taiwan.[2] When the deficiency is severe, the result is the salt-wasting (SW) form, and when the deficiency is partial, the result is the simple-virilizing (SV) form of CAH. Females with ambiguous genitalia, and males with sexual precocity and accelerated growth rates with no history of SW crises are classed as SV. Girls with CAH are exposed to excess fetus androgens from the adrenal gland, which causes androgenization of the external genitalia. The androgenization can range from mild clitoral enlargement to external genitals that look

From the Department of Child Psychiatry; [1]Department of Psychiatry; [2]Department of Pediatric Surgery; [3]Department of Pediatrics, Chang Gung Memorial Hospital, Taipei, Chang Gung University College of Medicine, Taoyuan, Taiwan; [4]Center of General Education, Chang Gung University, Taoyuan, Taiwan.

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Correspondence to: Dr. Hsueh-Ling Chang, Department of Child Psychiatry, Chang Gung Memorial Hospital. 5, Fusing St., Gueishan Township, Taoyuan County 333, Taiwan (R.O.C.) Tel.: 886-3-3281200 ext. 8540; Fax: 886-3-3280267; E-mail: chang0687@cgmh.org.tw

like a normal scrotal sac but hidden behind these external genitals are a vagina and a uterus.[1]

The optimal medical treatment of intersex conditions to maximize psychological functioning in children born with ambiguous genitalia remains controversial.[3,4] Traditional medical practices - immediate sex assignment prior to 18 months of age, early surgery to create the appearance of normal external genitalia and instructions to parents to provide unwavering sex-typical socialization - have been thought to maximize psychological adjustment, including acceptance of assigned sex.[5] Recent challenges have focused on the need to examine the outcome of traditional practices, particularly in terms of quality of life, and the possibility that early surgical interventions cause, rather than prevent, emotional problems.[3-5] Pediatric endocrinologists, pediatric surgeons, neonatologists and genetic specialists, but not psychiatrists and social workers, are usually part of the case management team.

Several studies have evaluated the psychiatric/psychological issues associated with CAH.[5-9] According to Riepe et al., 71% of female CAH patients suffer from psychosexual problems. Of these, only 17% undertook routine psychiatric diagnosis and counseling.[9] Berenbaum et al. found that adult females with CAH as a result of 21-hydroxylase (21-OH) deficiency had good overall psychological adjustment, similar to that of the control group. In addition, their study showed that psychological adjustment was not significantly associated with the degree of genital virilization, the appearance of external genitalia or the age at genital surgery.[7] However, specific problems, such as gender identity, sexual orientation and sex-typed behavior,[10-14] psychosexual function,[10,13,15-17] body images,[5,18] psychiatric adjustment[18,19] and quality of life,[20] have been evaluated and found to be associated with the illness when using different assessment instruments. With regard to childhood psychiatric comorbidity, few studies have revealed that intersex people have an increased prevalence of mental disorders, except that some individuals with CAH struggled to adjust to their condition.[7]

Apart from psychosocial factors and the endocrine influence on psychiatric manifestations, there may be other considerations contributing to psychiatric distress. Charmandari et al. reported that carriers of CAH due to 21-OH deficiency appear to have chronic mild hypocortisolism and compensatory changes of corticotropin-releasing hormone (CRH) secretion secondary to lower cortisol concentration. These changes might predict a mild predisposing of these subjects to physical and psychological pathways, and may have important clinical implications for psychological problems and overall vulnerability to stress.[21]

There are few studies related to CAH cases found in Taiwan.[2,22] The motivation for this study was to help fill the gap relating to the lack of information regarding people with CAH in Taiwan. Specifically, we wanted to focus on the psychiatric aspects of CAH in young females. Therefore, this study aims to investigate psychiatric manifestations, including psychiatric morbidity, gender identity and recalled sex-typed behavior, of young females with CAH in Taiwan.

METHODS

Subjects

The Medical Ethics and the Human Clinical Trials Committee of Chang Gung Memorial Hospital approved the study protocol. After providing a description of the procedures and purpose of the study, written informed consent was obtained from each patient's parent or guardian, and written assent or oral assent was obtained from patients.

To identify potential genetic female participants, a computer search was conducted for all patients with a diagnosis of CAH registered at Chang Gung Children's Hospital at Chang Gung Memorial Hospital. The hospital was selected as the source of subject recruitment because it is one of the largest pediatric hospitals in northern Taiwan. The computer search used the code numbers of the International Statistical Classification of Diseases and Related Health Problems (ICD-9), with 7527 (indeterminate sex and pseudohermaphroditism) and 2552 (adrenogenital disorders) as keywords. Eighty-one subjects with ICD-9 7527 and 135 with ICD-9 2552 were identified. Chart reviews were conducted and cases with incomplete medical records, those lost to follow-up and those with loss of contact were excluded. In addition, non-CAH cases were excluded.

Fifty-five patients with CAH were included for further analysis (mean age: 9.3 ± 6.6 years, range

from 3 to 35 years, legal female sex: n = 35 (63.6%), legal male sex: n = 20 (36.4%)). The patients or their parents or caregivers were contacted and invited to participate in the study. Of the females, 11 participated in the study, 18 patients could not be contacted, 4 refused to participate in the study and 2 had moved too far away to be able to take part in the study.

Assessment

The research protocol of this cross-sectional study included a half-day series of systematic interviews regarding demographic data, information of sex assignment at birth, sex rearing by parents, gender identity, recalled sex-typed behavior, sexual orientation and psychiatric morbidity, as well as self-reported inventories (oral explanation if necessary) and psychometric tests.

Medical data, including age at diagnosis, age at feminizing genitoplasty, medical treatment, medical compliance and physical illness, were obtained from hospital charts and clinical interviews. CAH subtype information was reassessed by pediatric endocrinologists for confirmation, and physical examinations were conducted by one child endocrinologist and one pediatric surgeon to identify current genital virilization as part of the study protocol after psychiatric interviews. The interviews and diagnoses were conducted by two board-certified child psychiatrists (H-L Chang and H-Y Liang).

Psychiatric morbidity within the past 12 months was assessed by child psychiatrists who conducted the Chinese version of the Kiddie Schedule for Affective Disorders and Schizophrenia for School-Age Children - Epidemiological Version (K-SADS-E) if the participant's age was from 6 to 16 years,[23,24] and the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV) for those 16 years old or above.[25] The K-SADS-E is a widely used semi-structured clinical interview for systematic assessment of both past and current episodes of mental disorders in children and adolescents.[23] Development of the Chinese version of the K-SADS-E was carried out by the Child Psychiatry Research Group in Taiwan, which included a two-stage translation and modification of several items with psycholinguistic equivalents relevant to Taiwanese culture.[24]

Minor psychiatric morbidity of participants within the current month was assessed by the self-rated Chinese Health Questionnaire (CHQ), which has 12 items rated on a four-point scale, with a higher score indicating more severe psychiatric symptoms. The Cronbach's α in community and clinical settings were 0.84 and 0.83, and 0.85 in this study. Persons with CHQ scores of 4 and over are considered to have minor psychiatric disorders.[26,27] Dysphoria, or gender identity uncertainty, was assessed by a semi-structured Gender Identity Interview developed for the identification of gender identity disorder. The interview provides three gender confusion scores including affective, cognitive and total aspects (a low/high score indicates a female/male gender identity).[28,29] A sample item is: "3. When you grow up, will you be a Mommy or a Daddy? (Mommy/Daddy)."

The clinical rating for the core behavioral features of gender identity disorder of children and adolescents was assessed with parents by the Recalled Sex-Typed Behavior Questionnaire, female version,[30] for participants displaying sex-typed behaviors during the past 6 months, based on the DSM-IV diagnostic criteria for gender identity disorder developed by Zucker et al. The scale consists of 7 items including: (i) identity statements, (ii) anatomic dysphoria, (iii) cross-dressing, (iv) toy and role play, (v) peer relations, (vi) mannerism, and (vii) rough-and-tumble play. Individual items were rated on a 3-point (item vii), 4-point (items i, ii, iii, v and vi) and 5-point (item iv) response scale. For instance, item (i): child states the wish to be of the opposite sex, or that he or she is a member of the opposite sex (e.g. "I want to be a boy, I am a boy or I want to grow up to be a Daddy not a Mommy"). A lower score indicates more sex-atypical behavior.

Data analysis

Descriptive statistics were used to present the demographic data, the mean score and the standard deviation of the questionnaires. A one-tailed Pearson correlation with significance at the 0.05 level was conducted. SPSS 11.5 for Windows was used for the data analysis.

RESULTS

This cross-sectional study of clinical interviews was based on a sample of young females with CAH. The mean age was 15.3 $\pm$ 5.6 years (range, 8-25

years) and the mean age at diagnosis was 5.6 ± 5.8 years (range, 0.7-19). Diagnosis was delayed to 14 and 19 years of age in two participants (no. 4 and no. 10) due to primary amenorrhea and their having secondary male sex characteristics, such as a beard, low voice tone and Adam's apple. After excluding these two, the mean age at diagnosis was 3.2 ± 1.9 years (range, 0.7-6 years). The mean age at the time of feminizing genitoplasty was 4.2 ± 2.2 years (range, 1-8 years).

All participants were born with female genitals with varying degrees of virilization. All were raised as females. Two subjects (no. 2 and no. 3) were assigned to the male sex at birth and then reassigned to the female sex within one month of birth. The attitude of the parents of one participant (no. 10) was ambivalent and inconsistent toward the child's gender identity (either male or female). They tended to neglect the child's ambiguous genitalia, which was noted at birth, and avoided dealing with the fact that the patient had secondary male sex characteristics (Table 1).

Table 1. Clinical Features of Eleven Participants with Congenital Adrenal Hyperplasia

No	Age (yr)	Dx (yr)	OP (yr)	Type of CAH	Psychiatric diagnosis*	RSBQ	GCS
1	18	4	5	SV	N.D.†	24	15
2	15	0.7‡	2	SW	Social phobia	27	–
3	10	0.8‡	2	SW	N.D.	23	11
4	17	14	-	SV	Adjustment disorder	26	13
5	15	6	8	SV	N.D.	24	11
6	8	5	6	SV	N.D.	28	11
7	10	4	5	SV	N.D.	26	11
8	16	3	4	SW	Dysthymic disorder	12	15
9	10	4	5	SV	N.D.	26	12
10	24	19	–	SV	Gender identity disorder, not otherwise specified†	18	20
11	25	1	1	SV	N.D.	24	11

Abbreviations: Dx: age at diagnosis for CAH; OP: age at feminizing genitoplasty; CAH: congenital adrenal hyperplasia; yr: years old; RSBQ: recalled sex-typed behavior questionnaire, a lower score indicated more sex-atypical behavior; GCS: total gender confusion score, a high score indicated more gender dysphoria and gender confusion; SV: simple-virilizing type of CAH; SW: salt-wasting type of CAH; N.D.: no diagnosis.
*: psychiatric comorbidities of nos. 1, 4, 8, 10 and 11 were assessed by the DSM-IV, and those of nos. 2, 3, 5, 6, 7 and 9 were assessed by the K-SADS-E; †: no. 1 was sexually attracted to both males and females, and no. 10 was sexually attracted to females only; ‡: sex assignment as male at birth.

Two subjects had feminizing genitoplasty before the age of 18 months and five after the age of 5 years. SW type was diagnosed in three (27.3%) subjects and SV type in eight (72.7%). Six subjects, with ages between 6 and 16 years old, were assessed using the K-SADS-E. Two were diagnosed with major psychiatric illnesses, including dysthymic disorder (no. 8) and social phobia (no. 2). Five participants older than 16 years were assessed using the DSM-IV. One was diagnosed with gender identity disorder, not otherwise specified, and was sexually attracted to females (no. 10), while another was found to have adjustment disorder (no. 4). The latter subject suffered from peer conflicts and an adjustment problem toward her masculine secondary sex characteristics. However, the distress had subsided at the time of assessment.

The CHQ revealed minor psychiatric disturbances in four (36.4%) subjects. Only one participant (no. 11, 9.1%) had ever received counseling. This was given at university by a teacher due to the stigma and adjustment problems that arose as a result of this chronic illness. She specifically suffered from the fear of intimacy and interpersonal relationships due to being afraid of revealing this secret. None of the parents had ever received psychiatric intervention, either at initial diagnosis or subsequently (Table 2).

Table 2. Demographic Data, Psychiatric Comorbidities and Recalled Sex-Role Behavior in Two Subtypes of Congenital Adrenal Hyperplasia

	Simple-virilizing type		Salt-wasting type
	Early diagnosis (n = 6) Mean ± SD	Delayed diagnosis (n = 2) Mean ± SD	(n = 3) mean ± SD
Age (years)	14.3 ± 6.4	20.5 ± 5.0	13.7 ± 3.2
Age at diagnosis (years)	4.0 ± 1.7	16.5 ± 3.5	1.5 ± 1.3
Age at surgery (years)	5.0 ± 2.3	-	2.7 ± 1.2
Psychiatric comorbidity			
CHQ	2.0 ± 1.1	4.5 ± 2.1	4.7 ± 2.5
CHQ (+)	n = 1	n = 1	n = 2
Psychiatric morbidity	n = 0	n = 2	n = 2

Abbreviations: SD: standard deviation; CHQ: subjects' self-reporting scores from the Chinese Health Questionnaire; CHQ (+): number of subjects with minor psychiatric illnesses as assessed by the CHQ.

One categorized herself as homosexual (no. 10) and one as bisexual (no. 1). None of them currently had partners or intimate relationships, nor did they have experience of intercourse. Two of them had more recalled atypical sex-typed behavior in childhood (no. 8 and no. 10). Three of them had higher scores for gender confusion (no. 1, no. 8 and no. 10). Those participants with more atypical sex-typed behavior in childhood had more gender confusion in the affective aspect and tended to have more severe psychiatric symptoms ($r = –0.654$, $p < 0.05$; $r = –0.706$, $p < 0.05$) (Table 3).

DISCUSSION

Of the participants in the study, 36.4% (n = 4) had received an axis I psychiatric diagnosis within the past year, including dysthymic disorder (with past self-injury behavior), adjustment disorder (to the masculine secondary sex characteristics), social phobia and gender identity disorder not otherwise specified. This appears high in comparison to the epidemiological study conducted by Gau et al., which found the prevalence rates across 3 consecutive years for dysthymic disorder, adjustment disorder, social phobia and overall psychiatric disorder in Taiwanese adolescents were 0.2-0.6%, 0.3-1.3%, 1.8-3.4% and 14.8%-22.7%, respectively.[31] Furthermore, compared to the 11.5% of minor psychiatric illnesses in the normal population in Taiwan,[27] participants with CAH (33.3%) in the study had a higher risk of minor psychiatric illnesses, such as anxiety (psychological and physical symptoms), depressed mood and sleep problems, especially participants with SW subtype and SV subtype with delayed diagnosis. Although this study revealed that a high proportion of subjects suffered from psychiatric illnesses, only one of them had received psychiatric intervention and none had been referred to a psychiatric clinic. The low rate of psychiatric intervention was consistent with the report by Riepe et al., which found that only 17% of the 71% of female CAH patients with psychosexual problems received routine psychiatric diagnosis and counseling.[9]

The study revealed that the more atypical the sex-role behavior noted in childhood, the more severe the psychiatric illness of the subjects. The study by Ogilvie et al. on the psychosexual development of women with CAH suggested that excessive prenatal androgen exposure predisposed subjects to more cross-gender role behavior and less comfort with femininity, shifting typical female behavior patterns toward male behavior patterns.[12] Prenatal androgen levels may also influence sexual orientation and core gender identity. Several studies have been published regarding this area. Women with CAH have been found to be more likely to say they are bisexual or homosexual than female controls with other endocrine disorders,[17] and to score higher on a measure of homosexual interest and lower on a measure of heterosexual interest than their unaffected sisters.[15] Past research shows that females with CAH display increased male-typical play behavior in childhood, and may exhibit an increased risk of gender dysphoria and reduced heterosexual orientation as adults.[12,32] In this study, most of the young women with CAH identified themselves as females but they may be at increased risk of gender identity problems, such as gender dysphoria or gender confusion. Subjects with more gender confusion and gender

Table 3. Correlation between Gender Confusion, Recalled Sex-Typed Behavior and Psychiatric Morbidity (n = 11)

Variable	RSBQ			CHQ		
	r	*p* value	95% CI	r	*p* value	95% CI
CHQ	–0.706	0.015	–2.815 to –0.390			
GCS	–0.608	0.062	–2.010 to 0.062	0.625	0.054	–0.009 to 0.880
GCS-C	–0.508	0.133	–4.559 to 0.729	0.605	0.064	–0.073 to 2.059
GCS-A	–0.654	0.040	–3.390 to –0.098	0.612	0.060	–0.038 to 1.462
RSBQ				–0.706	0.015	–0.546 to –0.076

Abbreviations: CHQ: score from the Chinese Health Questionnaire by subject; CI: confidence interval; GCS: total gender confusion score (a higher score indicates more gender confusion); GCS-C: cognitive gender confusion; GCS-A: affective gender confusion; RSBQ: recalled sex-typed behavior questionnaire (a lower score indicates more sex-atypical behavior).

dysphoria were found to be more at risk of having psychiatric morbidity. In Meyer-Bahlburg et al's study, one out of the 53 women with CAH seen at a single clinic requested a change to the male sex in adulthood and, in another study, four women with CAH living in the New York City area were found to be gender dysphoric as adults.[16] Other studies have found the vast majority of girls with CAH have a typical female gender identity.[11] How and to what degree gender confusion influences psychiatric distress needs to be further evaluated.

Limitations

The cross-sectional nature, the small sample size (low prevalence rate, stigma of intersex conditions) and bias in the selection of patients are limitations of this study. All participants in the study returned to the medical center for follow-up services, which implied they received more medical counseling and resources than those who did not. In addition, the lack of a control meant that the stigma of the illness for CAH patients could not be compared against another group. Additionally, there is no validity and reliability for using CHQ in adolescents. However, the internal consistency of CHQ in this study is higher than that for the community and clinical settings when administered to adults. During the interview, the interviewing psychiatrists provided assistance and explanations of the questionnaire where necessary. Furthermore, most of our patients were adolescents, which meant they were in the process of developing their identity. This study only assessed psychiatric morbidity within the past year. However, in the clinical interview, we found several participants suffered from a variety of adjustment problems relating to gender confusion, illness and body image. Finally, a survey of patients with CAH from the general population, i.e., untreated patients of different ages, might reveal very different experiences of dealing with CAH.

Clinical implications

In conclusion, this is one part of a series of studies to explore the psychiatric manifestations, gender identity and sexual orientation of young females with CAH in Taiwan. Interviews with 11 patients showed that the psychiatric needs of these patients were unmet and that psychiatric intervention was necessary. Earlier diagnosis, followed by adequate intervention and illness education for parents and patients, may result in better adjustment and mental health in later life. First-line clinicians should be alert not only to psychological problems associated with this disease but also to the need for early psychiatric evaluation and management. In line with Speiser et al.,[1] psychiatrists, along with pediatric endocrinologists, pediatric surgeons, neonatologists and genetic specialists, should be included as part of the team that cares for patients with CAH.

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患有先天性腎上腺素增生之年輕女性患者之精神症狀表現

梁歆宜 張學岑 陳景彥[1] 張北葉[2] 羅福松[3] 李立維[4]

背 景： 患有先天性腎上腺素增生之患者，在胎兒時期暴露在過多的雄性激素，導致外生殖器、性徵、幼年時期行爲，皆可能出現不同程度的雄性化表現。相關議題及病患心性適應等問題在國外已有不少的研究，但國內卻鮮少有此相關研究。因此，本實驗目的試圖探討台灣地區患有先天性腎上腺素增生之年輕女性患者之精神症狀表現。

方 法： 研究對象爲 11 名患有先天性腎上腺素增生之年輕女性患者。以半結構式之診斷性會談及自增問卷評估受試者之精神疾患共病症性別認同及幼兒時期性別類型行爲。

結 果： 受試者平均年齡爲 15.3 ± 5.6 歲。36.4% 之受試者患有精神科第一軸診斷。分別爲輕鬱症、適應障礙、社交恐懼症，及未分類之性別認同障礙。受試者在幼年期出現較雄性化之表現傾向有較高之精神科輕型疾患之罹病率 ($r = –0.706$, $p = 0.015$)。

結 論： 本研究發現，台灣地區患有先天性腎上腺素增生之患者，有較高的精神疾患共病症。建議精神科儘早診斷及介入，以協助病患適應。

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關鍵詞： 先天性腎上腺素增生，精神科共病症

長庚紀念醫院 台北院區 兒童心智科，[1]精神科，[2]兒童外科，[3]兒童內科；長庚大學 醫學院；[4]長庚大學 通識教育中心
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通訊作者：張學岑醫師，長庚紀念醫院 兒童心智科。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉8540; Fax: (03)3280267; E-mail: chang0687@cgmh.org.tw