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Factors influencing illness uncertainty in parents of children with congenital adrenal hyperplasia in a developing country: A cross-sectional study

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Abstract

Background: Illness uncertainty in parents of children with congenital adrenal hyperplasia (CAH) refers to parents' inability to create meaning in events related to their children having CAH. This may influence their role in caring for children with CAH.
Objective: The study aimed to determine factors associated with illness uncertainty experienced by parents of children with CAH in a developing country.
Methods: A cross-sectional study was conducted on 80 parents (43 mothers and 37 fathers) of children with CAH, selected using consecutive sampling methods. The Parent's Perception of Uncertainty Scale (PPUS) was used to measure the illness uncertainty levels. Data were collected from March 2020 to October 2020. Independent *t*-test and chi-square test were used to determine factors (parent's gender, age, educational level, monthly household income, number of children with CAH, history of child death due to CAH, child's age when first diagnosed with CAH, duration of therapy, gender change, type of CAH (salt wasting/SW or simple virilizing/SV), current gender, and genitoplasty) influencing illness uncertainty in parents.
Results: The mean scores of PPUS were 42.3 ± 12.91, and the majority of parents had a low PPUS score (49; 61%). Parents of children with SW-CAH showed higher uncertainty (44.2 ± 12.77) than those with SV-CAH (32.6 ± 8.86; $p = 0.003$). Parents who lost their children due to CAH were more likely to report a moderate illness uncertainty than parents who never experienced child mortality due to CAH ($\chi^2(1, 80) = 4.893$; $p = 0.027$).
Conclusion: The factors significantly affecting uncertainty in parents of children with CAH determined in this study might help healthcare professionals, including nurses, to play a pivotal role in giving pertinent information regarding their children's health, disease, and therapy to help manage parental uncertainty.

Keywords

congenital adrenal hyperplasia; uncertainty in illness; parents; developing country; Indonesia

Background

Congenital adrenal hyperplasia (CAH) is the most common cause of ambiguous genitalia in 46,XX disorders (differences) of sex development (DSD) in a newborn characterized by enzyme deficiency involved in steroidogenesis in the adrenal gland, in which 90% of cases are caused by 21-hydroxylase deficiency (Idris et al., 2014; Juniarto et al., 2018; Pezzuti et al., 2014; Tsuji et al., 2015; Zainuddin et al., 2019). This will result in cortisol and aldosterone deficiency and excessive androgen hormone production (Speiser et al., 2018; Szulczewski et al., 2017), with the incidence ranging from ~1:14,000 to 1:18,000 (Speiser et al., 2018). Data from the Indonesian Pediatrics Society registry in 2009 showed there were 69 children with CAH [56 (81%) girls and 13 (19%) boys]. However, this number surged to 439 children (303 (69%) girls and 136 (31%) boys) in 2020 (Armstrong et al., 2020). In addition, the Center for Biomedical Research (CEBIOR) recorded 84 patients with CAH were evaluated from 2004 to 2016 jointly by a multidisciplinary team from the Dr. Kariadi Hospital and Faculty of Medicine Diponegoro University, Semarang (Juniarto et al., 2018).

Unlike many Western countries, a newborn screening program to detect CAH is unavailable in Indonesia (Gidlöf et al., 2014; Held et al., 2015; Odenwald et al., 2015; Speiser et al., 2018; Tsuji et al., 2015). This causes many late-diagnosed CAH cases (Ediati et al., 2015), which leads to delayed treatment (Juniarto et al., 2018), virilization, masculinization (Bizic et al., 2018), psychological problems like gender dysphoria (Juniarto et al., 2018), stigmatization (Ediati et al., 2017), emotional and behavior problem (Ediati et al., 2015), cross-gender role behavior (Ediati et al., 2017; Idris et al., 2014), and fertility issues (Witchel, 2017). Because of these, parents may develop illness uncertainty, leading to developing psychological problems, such as stress (Hullmann et al., 2011; Wisniewski & Sandberg, 2015), depression (De Silva et al., 2014; Perez et al., 2019), and anxiety (McConkie-Rosell et al., 2018; Perez et al., 2019; Wisniewski & Sandberg, 2015). Furthermore, parents of children with CAH in Vietnam reported that the scarcity of CAH medicines and low incomes, particularly from remote and rural families, caused many parents to be unable to afford the drugs their children need (Armstrong et al., 2020). In addition, the diagnosis and treatment delay due to ambiguous genitalia uncovered cost of laboratory workup by National Health Assurance (or called JKN – Jaminan Kesehatan Nasional), and inaccessible fludrocortisone brings complexities in the management of CAH in a developing country like Indonesia (Armstrong et al., 2020).

Illness uncertainty, described as the inability to determine the meaning of illness-related events, has several factors that may contribute to its development: symptom pattern, event familiarity, event congruence, cognitive capacity, education, social support, and credible authority (Mishel, 1988). Understanding what causes uncertainty may help nurses and other healthcare professionals with patients' experiences regarding their illnesses. In addition, a patient should develop a coping mechanism to cope with the uncertainty, and nurses play a critical role in helping patients to comprehend illness uncertainty and cope. This could help patients and their families increase their quality of life (Smith & Liehr, 2018).

A previous study reported that mothers and fathers of DSD children scored higher uncertainty but lower than parents of children with chronic illness (Perez et al., 2019). Unfortunately, the study was conducted not only on parents of children with CAH but also on parents of children with 46, XX DSD, 46,XY DSD, and sex chromosomal DSD. To our knowledge, this is the first study of illness uncertainty in parents of children with CAH in a developing country. This study aimed to identify factors contributing to illness uncertainty in parents to promote early detection, prevention, and better management of children with CAH.

Methods

Study Design

A cross-sectional study was conducted from March 2020 to October 2020 at the pediatric endocrinology clinic, National Diponegoro Hospital (RSND) in Semarang, Central Java, Indonesia, among parents who had children with CAH.

Samples/Participants

Consecutive sampling was used by inviting all parents of children with CAH registered in the pediatric endocrinology clinic. The sample size was calculated using a 95% of confidence interval, 5% of margin error, and 5% of the population proportion. The sample size calculation was done using a sample size calculator (https://www.calculator.net/sample-size-calculator.html) based on an estimate of 90 young patients with CAH and resulted in a minimum sample of 41 participants. Of 90 young patients with CAH, the parents of 44 patients participated in the study. The participants of this study consisted of 37 fathers and 43 mothers who had children with CAH. Parents aged 18 years or older who have children diagnosed with CAH aged 0-18 years were included in this study. Parents of children with other severe congenital disorders were excluded.

Instruments

A semi-structured questionnaire was utilized to assess sociodemographic data, such as parents' gender, age, educational level, and monthly household income. In addition, data regarding the affected child were obtained from the medical records consisting of the date of birth, current gender, age at the start of treatment, duration of therapy, type of CAH (salt wasting/SW or simple virilizing/SV), genitoplasty, gender change status, sibling with CAH, and deceased CAH sibling.

In this study, parents were divided into two age groups using mean age as a cut-off (below or above mean age). Upper secondary education is defined as someone whose latest education is a secondary school, including high school and vocational school, or below. Bachelor's or equivalent education refers to someone who attained tertiary school, including a diploma, undergraduate, graduate, and postgraduate. Only parents of female children with CAH were included in the genitoplasty analysis.

The original version of the Parent Perception of Uncertainty Scale (PPUS) was translated and validated into Bahasa Indonesia by Ediati et al. (2020) on 70 parents of children with CAH and diabetes mellitus (DM). Originally, PPUS comprised four factors (ambiguity, lack of information, lack of clarity, and unpredictability) that yielded 31 items. The construct validity of the Indonesian version of PPUS (PPUS-Indonesia) was explored using confirmatory factor analysis and revealed a one-factor model with satisfactory reliability (Cronbach's alpha 0.903).

The PPUS-Indonesia consisted of a 20-item parent-reported measure that assesses the level of illness uncertainty in parents. The following are sample items in the Indonesian version of PPUS: "Saya tidak tahu apa yang tidak beres dengan anak saya." (I don't know what is wrong with my child); "Saya tidak yakin apakah anak saya membaik atau makin parah." (I am unsure if my child's illness is getting better or worse); "Tujuan setiap pengobatan jelas bagi saya." (The purpose of each treatment is clear to me). Item responses are rated on a 5-point Likert scale ranging from "strongly agree" to "strongly disagree" and scored 1 to 5, resulting in a possible total score ranging from 20 to 100. A higher PPUS score indicates greater levels of perceived uncertainty. The total score of PPUS is categorized into three groups: low (20-46), moderate (47-73), and high (74-100).

Data Collection

Data were collected by the researchers themselves. The researchers conducted a personal interview using the

questionnaire for the 23 illiterate participants. In contrast, for the well-educated participants, the researchers gave two options for participants to fill out the questionnaire while being accompanied or interviewed.

Data Analysis

Data were analyzed using IBM SPSS (Statistical Package for the Social Sciences) version 26 (IBM, New York). Comparisons between numerical variables were analyzed using an independent *t*-test, while categorical variables were analyzed using the chi-square test. Fisher's exact test was applied for variables not aligned with the chi-square assumption, i.e., type of CAH and genitoplasty. Both bivariate analyses were conducted to identify the significant factors of parental uncertainty. The significant level was set at <0.05. Sensitivity analysis using Fisher's exact test was performed by splitting the analysis based on the parent's gender to compare the variables found to be significant in the chi-square test, i.e., type of CAH and child death due to CAH, and hypothetically categorized PPUS score. In addition, multivariate analysis using logistic regression was conveyed for the parents' gender, number of children with CAH, child death due to CAH, and type of CAH.

Ethical Considerations

The ethical committee of the Faculty of Medicine, Universitas Diponegoro, Semarang, approved the study protocol (No. 483/EC/KEPK/FK UNDIP/XI/2019). Prior to the study, all participants received the study information and were free to decide on their participation in the study. Participants who joined the study voluntarily signed written informed consent.

Results

Participants consisted of 37 (46%) fathers and 43 (54%) mothers of children with CAH. The mean age of the participants was 35 ± 7.7 years, with most of them [44 (55%)] aged ≥ 35 years. The majority of parents (74%) had an upper-secondary education. Overall, 84% of parents had children with SW-CAH. Data from two parents were excluded from bivariate analysis by genitoplasty and the child's current gender because they were parents of two children with CAH raised as a boy and a girl. Therefore, their response on PPUS-Indonesia might be biased as it is difficult to specify whether their response on uncertainty was referring to the son or daughter conditions or might not have represented both genders equally. Among 56 parents of girls with CAH, only 14% of their children had undergone genitoplasty (**Table 1**).

Table 1 Characteristics of participants

	Fathers (*n* = 37)	Mothers (*n* = 43)	Total (*n* = 80)
	n (%)	*n* (%)	*n* (%)
Parent-Related Factors			
Age			
< 35 years	14 (35)	23 (54)	36 (45)
≥ 35 years	24 (65)	20 (47)	44 (55)
Educational level			
Upper secondary	29 (78)	30 (70)	59 (74)
Bachelor's or equivalent	8 (22)	13 (30)	21 (26)
Household monthly income (IDR)			
< 2.000.000 (< ±$140)	15 (41)	17 (40)	32 (40)
≥ 2.000.000 (≥ ±$140)	22 (60)	26 (61)	49 (60)
Child-Related Factors			
Number of children with CAH			
More than one	11 (30)	13 (30)	24 (30)
One	26 (70)	30 (70)	56 (70)
Child death due to CAH			
Yes	8 (22)	10 (23)	18 (23)
No	29 (78)	33 (77)	62 (78)
Type of CAH			
Salt wasting	30 (81)	37 (86)	67 (84)
Simple virilizing	7 (19)	6 (14)	13 (16)
Child's gender change due to CAH			
Yes	9 (24)	10 (23)	19 (24)
No	28 (76)	33 (77)	61 (76)
Child's age when first diagnosed with CAH			
< 3 months	18 (49)	23 (53)	41 (51)
≥ 3 months	19 (51)	20 (47)	39 (49)
Duration of therapy			
< 3 years	16 (43)	20 (47)	36 (45)
≥ 3 years	21 (57)	23 (53)	44 (55)
Child's current gender (*n* = 78)			
Male	10 (28)	12 (29)	22 (28)
Female	26 (72)	30 (71)	56 (72)
Genitoplasty (*n* = 56)‡			
Yes	4 (15)	4 (13)	8 (14)
No	22 (85)	26 (87)	48 (86)

‡Genitoplasty analysis was performed only on the parents of girls with CAH

The majority of parents in the study showed a low level of uncertainty, as indicated by an average score of PPUS at 42.3 ± 12.91. Although none reported a high level of uncertainty, 39% of parents reported a moderate level of uncertainty (**Table 2**).

Table 2 Parental uncertainty based on PPUS score hypothetical category

Score category[¶]	Low	Moderate	High	Mean ± SD
Father (*n* = 37)	20 (54%)	17 (46%)	0 (0%)	44.8 ± 13.22
Mother (*n* = 43)	29 (67%)	14 (33%)	0 (0%)	40.1 ± 12.37
Total (*n* = 80)	49 (61%)	31 (39%)	0 (0%)	42.3 ± 12.91

[¶]Low = 20-46; Moderate = 47-73; High = 74-100

Table 3 displays the comparison analysis results between parent-related and child-related factors and the PPUS score. Parents of children with SW-CAH showed a higher score of uncertainty (44.2 ± 12.77) than parents of children with SV-CAH (32.6 ± 8.86). In addition, the type of CAH significantly affected the uncertainty perceived by parents (p = 0.003).

Table 3 Comparison analysis between parent- child-related factors and PPUS score

	Mean (SD)	Mean difference (CI 95%)	Median (Minimum-Maximum)	*p*-value[§]
Parent-Related Factors				
Gender (*n* = 80)		4.7 (10-10.4)		0.101
Father (*n* = 37)	44.8 (13.22)			
Mother (*n* = 43)	40.1 (12.37)			
Educational level (n = 80)		3.2 (4.4-10.9)		0.394
Upper secondary (n = 59)	43.1 (11.86)			
Bachelor's or equivalent (*n* = 21)	39.9 (15.56)			
Age (*n* = 80)				0.230
< 35 years (*n* = 39)			42.5 (20-68)	
≥ 35 years (*n* = 41)			39.0 (20-63)	
Household monthly income (*n* = 80)				0.582
< IDR 2.000.000,00 (n = 32)			43.0 (20-67)	
≥ IDR 2.000.000,00 (n = 48)			39.5 (20-68)	
Child-Related Factors				
Number of children with CAH (*n* = 80)		1.7 (4.6-8.0)		0.599
One (*n* = 24)	43.5 (13.08)			
More than one (*n* = 56)	41.8 (12.92)			
Child death due to CAH (*n* = 80)		5.4 (1.4-123)		0.116
Yes (*n* = 18)	46.5 (12.48)			
No (*n* = 62)	41.1 (12.87)			
Type of CAH (*n* = 80)		11.6 (4.2-18.9)		0.003*
Salt wasting (*n* = 67)	44.2 (12.77)			
Simple virilizing (*n* = 13)	32.6 (8.86)			
Gender change due to CAH (*n* = 80)		0.1 (6.7-6.9)		0.975
Yes (*n* = 19)	42.4 (11.61)			
No (*n* = 61)	42.3 (13.38)			
Child's age when first diagnosed with CAH (*n* = 80)		1.1 (4.7-6.9)		0.703
< 3 months (*n* = 41)	42.8 (12.25)			
≥ 3 months (*n* = 39)	41.7 (13.70)			
Child's current gender (*n* = 78)		2.2 (4.3-8.6)		0.679
Male (*n* = 22)	44.1 (14.81)			
Female (*n* = 56)	41.9 (12.17)			
Genitoplasty (n = 56)		6.3 (15.5-3.0)		0.178
Yes (*n* = 8)	36.5 (12.56)			
No (*n* = 48)	42.8 (12.00)			
Duration of therapy (*n* = 80)				0.247
< 3 years (*n* = 55)			42.5 (20-68)	
≥ 3 years (*n* = 25)			39.0 (20-63)	

Note: IDR=Indonesian Rupiah | [§]The independent t-test was performed | *Significant at $p < 0.05$

Table 4 demonstrates the results of the comparison analysis of parent-child-related factors between low and moderate levels of uncertainty. Significant differences in illness uncertainty were found between groups of parents with SW-CAH and SV-CAH, as well as parents with and without experience of child mortality due to CAH. All parents of children with SV-CAH reported low uncertainty, whereas parents of SW-CAH reported lower to moderate uncertainty (p = 0.001). Parents who had lost their children due to CAH were more likely to have a moderate level of illness uncertainty, whereas parents who never experienced child mortality due to CAH were more likely to report a low level of illness uncertainty ($\chi^2(1, 80) = 4.893$; p = 0.027). No significant differences were found across different groups based on parent-related and

child-related factors between low and moderate levels of uncertainty. In our data, two factors are associated with illness uncertainty in parents of children with CAH. First, parents with a history of child death due to CAH. They were more likely to report moderate uncertainty than parents without experience of child death due to CAH. Second, CAH type. None of the parents from the simple virilizing CAH group reported a moderate level of uncertainty. In contrast, almost half of the parents from the salt-wasting group reported a moderate level of uncertainty.

Table 4 Comparison analysis between parent-child-related factors and PPUS score hypothetical category

	Low	Moderate	*p*-value‡
Parent-Related Factors			
Gender (*n* = 80)			0.220
Father (*n* = 37)	20 (54.1%)	17 (45.9%)	
Mother (*n* = 43)	29 (67.4%)	14 (32.6%)	
Educational level (*n* = 80)			0.943
Upper secondary (*n* = 59)	36 (61.0%)	23 (39.0%)	
Bachelor's or equivalent (*n* = 21)	13 (61.9%)	8 (38.1%)	
Age (*n* = 80)			0.982
< 35 years (*n* = 39)	22 (61.1%)	14 (38.9%)	
≥ 35 years (*n* = 41)	27 (61.4%)	17 (38.6%)	
Household monthly income (*n* = 80)			0.779
< IDR 2.000.000,00 (*n* = 32)	19 (59.4%)	13 (40.6%)	
≥ IDR 2.000.000,00 (*n* = 48)	30 (62.5%)	18 (37.5%)	
Child-Related Factors			
Number of children with CAH (*n* = 80)			0.176
One (*n* = 24)	12 (50.0%)	12 (50.0%)	
More than one (*n* = 56)	37 (66.1%)	19 (33.9%)	
Child death due to CAH (*n* = 80)			0.027**
Yes (*n* = 18)	7 (38.9%)	11 (61.1%)	
No (*n* = 62)	42 (67.7%)	20 (32.3%)	
Type of CAH (*n* = 80)[1]			0.001**
Salt wasting (*n* = 67)	36 (53.7%)	31 (46.3%)	
Simple virilizing (*n* = 13)	13 (100.0%)	0 (0.0%)	
Gender change due to CAH (*n* = 80)			0.377
Yes (*n* = 19)	10 (52.6%)	9 (47.4%)	
No (*n* = 61)	39 (63.9%)	22 (36.1%)	
Child's age when first diagnosed with CAH (*n* = 80)			0.684
< 3 months	26 (63.4%)	15 (36.6%)	
≥ 3 months	23 (59.0%)	16 (41.0%)	
Child's current gender (*n* = 78)			0.702
Male (*n* = 22)	14 (63.6%)	8 (36.4%)	
Female (*n* = 56)	33 (58.9%)	23 (41.1%)	
Genitoplasty (*n* = 56)[1]			0.318
Yes (*n* = 8)	6 (75.0%)	2 (25.0%)	
No (*n* = 48)	27 (56.3%)	21 (43.8%)	
Duration of therapy (*n* = 80)			0.982
< 3 years (*n* = 55)	22 (61.1%)	14 (38.9%)	
≥ 3 years (*n* = 25)	27 (61.4%)	17 (38.6%)	

Note: IDR=Indonesian Rupiah | ‡The chi-square test was applied, with the exclusion of type of CAH and genitoplasty | [1]The Fisher's test was applied | **Significant at $p < 0.05$

Table 5 Sensitivity analysis, split by parent's gender, between child death due to CAH and type of CAH and PPUS score hypothetical category

		Low	Moderate	*p*-value‡
Child death due to CAH (*n* = 80)				
Father (*n* = 37)	Yes (*n* = 8)	3 (37.5%)	5 (62.5%)	0.428
	No (*n* = 29)	17 (58.6%)	12 (41.4%)	
Mother (*n* = 43)	Yes (*n* = 10)	4 (40.0%)	6 (60.0%)	0.055
	No (*n* = 33)	25 (75.8%)	8 (24.2%)	
Type of CAH (*n* = 80)				
Father (*n* = 37)	Salt wasting (*n* = 30)	13 (43.3%)	17 (56.7%)	0.009**
	Simple virilizing (*n* = 7)	7 (100.0%)	0 (0.0%)	
Mother (*n* = 43)	Salt wasting (*n* = 37)	23 (62.2%)	14 (37.8%)	0.155
	Simple virilizing (*n* = 6)	6 (100.0%)	0 (0.0%)	

‡The Fisher's test was applied. **Significant at $p < 0.05$

Sensitivity analysis using Fisher's exact test was performed for two variables found to be significant in the chi-square test, i.e., type of CAH and child death due to CAH. Each variable was split based on the parent's gender, and a

comparison was made between these two variables and PPUS score hypothetical category. Illness uncertainty was significantly different between fathers of SW-CAH and SV-CAH. All fathers of SV-CAH reported a low level of uncertainty (p = 0.009) (**Table 5**).

Logistic regression, which was conducted for parents' gender, the number of children with CAH, child death due to CAH, and type of CAH, found no significant impact on the PPUS score hypothetical category.

Discussion

The study aimed to investigate factors influencing illness uncertainty experienced by parents of children with CAH in a developing country. Our study findings showed that there were two factors affecting illness uncertainty in parents of children with CAH: the type of CAH and the presence of child mortality due to CAH. The chi-square test was performed to analyze the presence of child mortality, which was in line with the chi-square assumptions. Parents of children with the salt-wasting type of CAH and parents who experienced the loss of their children due to CAH are more likely to report higher illness uncertainty than other parents who have not. The study findings are important as higher uncertainty among parents of children with CAH reflects the need to seek appropriate information related to CAH and its treatment to reduce the uncertainty level of parents.

Parents of children with CAH in the present study reported somewhat low to moderate levels of uncertainty, and none reported a high level of uncertainty. A previous study about psychological distress experienced by parents of children with DSD showed a comparable level of uncertainty (Suorsa et al., 2015). This might indicate that their children with CAH have received medical treatment and regularly visit the pediatric endocrinology clinic to maintain their health. The role of healthcare professionals in delivering appropriate information regarding CAH to parents and the availability of guidelines about CAH that parents can refer to (Speiser et al., 2018) may play an essential role in reducing illness uncertainty in these parents of children with CAH.

In addition, social support is also available by using social media (WhatsApp group) of parents who have children with CAH (Mishel, 1988) may reduce uncertainty experienced by parents of children with CAH. But still, some parents experienced a moderate level of illness uncertainty. More efforts are needed to identify relevant information to reduce the illness uncertainty for these parents. Do they receive less or too much information regarding CAH, and what kind of CAH-related information do they need to reduce illness uncertainty on these parents should be investigated in the follow-up study because lack or too much information, as well as seeking or avoiding information, may influence uncertainty (Kerr & Haas, 2014).

Most participants were parents of children with salt-wasting type CAH (44.2 ±1 2.77), and they had a significantly higher mean score of PPUS than children with simple virilizing type CAH (32.6 ± 8.86). According to Utari et al. (2016), 10 out of 79 (13%) children registered with CAH in Central Java, Indonesia, had died because of adrenal crises since 2009. The lack of newborn screening for CAH leads to delayed diagnosis and treatment. In addition, the most severe form of CAH, the salt-wasting type, made the body unable to regulate salt (sodium) levels in the blood because of a lack of aldosterone, making it prone to salt-wasting crisis (Falhammar et al., 2014). This made salt-wasting CAH contribute to a higher level of uncertainty. Therefore, patients with CAH need hydrocortisone and fludrocortisone for cortisol and aldosterone replacement therapy, respectively. However, fludrocortisone is unavailable in Indonesia (Armstrong et al., 2020) and might further increase the illness uncertainty among parents.

A significant difference in illness uncertainty was discovered between fathers of SW-CAH and fathers of SV-CAH. A previous study on parents of children with DSD found contradictory results where mothers scored higher uncertainty despite being insignificant (Perez et al., 2019). Our result might be explained by the patriarchy that most Indonesians embraced. Most of the participants of this study came from rural areas. In addition, most mothers who participated in this study were housewives and unemployed; thus, the only source of income for the family came from the fathers. Therefore, even though mothers might be involved in the general decision-making process, as the head of the family, the final decision is for the fathers to decide (Ferdoos & Zahra, 2016; Sarwono, 2012).

Child mortality due to CAH significantly affected illness uncertainty in parents. Parents of children with CAH who had lost their children due to CAH were more likely to report a higher level of uncertainty than parents who had never had such a grief loss. The result was similar to psychological distress experienced by parents of children who died from cancer (Sirkiä et al., 2000; Stroebe et al., 2006). Child loss was one of the deepest griefs experienced by parents (Davies, 2004). If a child died, the parent's hope about the future would die, too (Christ et al., 2003). Parents who have encountered child loss might have an intense fear of losing the other child or, in CAH cases. Facing conflicting information about caring for children with CAH, therefore, might need specific support during bereavement (Gijzen et al., 2016). Further investigation is needed to explore the experiences of 18 parents who lost their children due to CAH and whether they need specific support and knowledge related to CAH to reduce the illness uncertainty of these particular parents.

The findings reported two factors significantly affecting illness uncertainty in parents with CAH. However, illness uncertainty did not significantly differ in most factors investigated, i.e., parental gender, age, educational level, monthly household income, number of children with CAH, child's gender change due to CAH, child's age when first diagnosed with CAH, child's current gender, history of genitoplasty, and therapy duration. This might be because, nowadays, parents can easily search for their children's condition on the internet before a medical appointment (Molzon et al., 2014). On the contrary, a previous multicenter study on parents of children with DSD found that parents of boys reported a higher illness uncertainty compared to parents of girls, and parents who were unsure of the gender of their child also reported greater uncertainty (Suorsa et al., 2015).

Implications of the Study for Nursing Practice

Nurses' primary role was taking care of the patient, attending to their physical needs and holistically, including psychosocial, cultural, and social needs (Hodges & Tod, 2007). Children

were incapable of taking care of themselves hence needing parents or caregivers. Child's illness might cause uncertainty and psychological distress and uncertainty in parents (De Silva et al., 2014; Hullmann et al., 2011; Kerr & Haas, 2014; McConkie-Rosell et al., 2018; Perez et al., 2019; Suorsa et al., 2015; Szulczewski et al., 2017; Wisniewski & Sandberg, 2015). The uncertainty caused the parents to be unable to define the outcomes and assign a value to events relating to their child's illness. Therefore, nurses need to understand the circumstance of uncertainty and its related factors to help the patient and their family, particularly parents, to comprehend illness uncertainty and develop a healthy coping mechanism, thus enhancing their quality of life (Brimble, 2007).

Limitations of the Study

The sample size was moderate, and it would be better to find another sample size estimation using two mean difference sample tests or two proportional difference sample tests. This was a one-centered study, so the data collected might not be as diverse as the data retrieved from multiple centers; besides, collecting data from a multicenter was challenging during the COVID-19 pandemic. Therefore, conducting a multicenter study is recommended to collect more data and diverse results in future research. Our study was conducted on parents who had managed their children with CAH for years. Many factors might influence the degree of uncertainty. Some of them were support from other parents of children with CAH and information they got from health care professionals over time. Despite the uncertainty they face regarding their children's illness, parents of children with CAH can seek support from other parents with CAH and receive sufficient information and support from medical professionals.

Conclusion

Parents of children with CAH had a low level of uncertainty; however, some parents still experienced moderate uncertainty. Salt-wasting type CAH and a history of child death due to CAH significantly affected parental uncertainty because of the impendence of salt wasting crisis and limitation of drug availability for salt-wasting type CAH. Therefore, these groups of parents would particularly need more support and attention and thus benefit from getting social support from other parents of children with CAH and regularly going to the medical appointment to consult and receive appropriate information from health care professionals. Nurses' roles in caring for the patient extend beyond the physical need and providing holistic care, including psychosocial, cultural, and spiritual needs. In addition, nurses also played a role in educating the patient and their family so they could better understand the patient's health, illnesses, and treatments, thus reducing uncertainty.

Declaration of Conflicting Interest

The authors declare no conflict of interest in this study.

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Authors' Contributions

The authors confirm their contribution to the paper as follows: study conception and design: AU, FS, IAL; data collection: IAL, FS, AU; analysis and interpretation of results: AE, AU, FS, IA; draft manuscript preparation: IAL, FS, TIW, AE, AU. All authors reviewed the results and approved the final version of the manuscript.

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Data Availability

The datasets generated during and analyzed during the current study are available from the corresponding author upon reasonable request.

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