Knowledge, stress and adopted coping strategies of parents of children having congenital adrenal hyperplasia: An exploratory survey

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ABSTRACT

Background: Congenital adrenal hyperplasia (CAH) is an inherited disorder of adrenal glands, the most common type being due to a deficiency of the enzyme 21-α-hydroxylase. Girls affected with it are virilized to a variable extent. The taboo associated with ambiguous genitalia in female babies poses an immense societal pressure on parents for keeping the disease a secret. **Objective:** The aim of the study was to generate the evidence on the knowledge, stress, and coping strategies used by the parents of CAH children. **Methodology:** In a cross-sectional descriptive survey using mixed methods approach, 30 parents of children with CAH attending the outdoor department of pediatric medicine and surgery in a tertiary care hospital were enrolled. The CAH knowledge questionnaire, parental stress scale (PSS), and COPE inventory were the standardized tools used along with demographic profile sheet to assess the knowledge of parents related to CAH, stress, and coping strategies used by them. **Results:** Majority of the parents (67%) had poor knowledge (<7) and moderate stress (42-65) scores. Knowledge and stress were negatively correlated (r=−0.39, p=0.035). Coping strategies used by the parents in dealing with stress in moderate amount were mainly problem-focused (active coping [60%] and planning [60%], instrumental social support [53%], suppression of competing activities [50%]), and emotion-focused (positive reinterpretation [60%], emotional social support [67%], acceptance [53%], and religious coping [50%]). Less useful coping strategies such as focus on and venting of emotions, behavioral disengagement, mental disengagement, humor, and substance abuse were not used by the majority parents. **Conclusion:** Parents of children with CAH had poor knowledge and moderate stress related to the disease condition of their children and were using problem focused and emotion focused coping strategies to reduce their stress.

Key words: *Congenital adrenal hyperplasia, Coping strategies, Knowledge, Stress*

Congenital adrenal hyperplasia (CAH), the most common endocrine disorder of the genetic origin, usually diagnosed at birth is caused by the deficiency of enzymes involved in adrenal steroidogenesis [1]. In 95% of the cases, mutation occurs in the CYP21A2 gene, causing varying degrees of inactivity of the 21-α-hydroxylase enzyme, with consequent defects in the final products of the glucocorticoid (GC) and mineralocorticoid (MC) pathways, and excessive production of androgens in the body. Depending on the degree of enzymatic blockage, the clinical presentation of the child can be classical or nonclassical. The classical form can further be salt-wasting (SW) and simple virilizing forms [2] while nonclassical form, a milder one is diagnosed in later part of the life.

The classic form of the disease is evident at the time of birth or in the early months of life. In this group, 75% of the patients may have combined deficiency in the formation of GC and MC, called SW form the most severe presentation of the disease can be predicted by genotype "null" and "I2splice" [3]. These children may land up with dehydration, shock, and hyponatremia [4]. The remaining 25% are simple virilizers, where the reduced enzymatic activity is adequate enough to prevent deficiency of MC, thus preventing adrenal crisis [5].

Normal growth and development in CAH children are major concerns of the parents. Knowledge deficit of parents related to the disease and uncertainties concerning gender assignment of the female child may lead to the development of stress among parents. Moreover, CAH is a lifelong disorder having no definite cure available, and these patients have to receive cortisol and aldosterone hormones lifelong. In addition, girls with CAH may require surgery to restore the genitalia appearance to the normal.

Parents are usually confused about the sex of their child because of abnormal appearance of external genetalia. Moreover, a lack of information about the disease condition, misconception and stigma associated with CAH and the parental concerns about the future of the child, what he/she would look like, how would be their lives in adulthood are some of the questions might bother parents resulting to stress [6]. Some studies [4,7,8] have explored the emotional reactions of the parents, not the knowledge related to CAH, stress and the coping strategies used by them.There is a felt need to explore the knowledge of parents about the disease as

knowledge influences the mental status, reduces parental stress and helps them in adopting healthy coping strategies. Therefore, this study was conceptualized.

METHODOLOGY

In a descriptive cross-sectional survey using mixed methods approach, we used convenience sampling to enroll 30 parents (out of 80 registered) of children with CAH (0-18 years) from the outdoor patient Departments of Pediatric Medicine and Pediatric Surgery of a selected tertiary care facility. Ethical permission was obtained from the Institutional Ethics Committee. Written consent was obtained from the parents before their enrolment. Parents having children with CAH with chronic (HIV, asthma, cancer, cerebral palsy, epilepsy, cystic fibrosis, diabetes, and sickle cell disease) or critical illness or siblings affected either with CAH, or any other chronic illnesses were excluded. Both quantitative and qualitative data were collected. 10 parents were randomly selected using draw of lots for in-depth interview to collect the qualitative data.

Tools used for data collection were standardized; knowledge questionnaire (CAHKAQ, 22 items, α=0.67), parental stress scale (PSS, 18 items, α=0.83), COPE inventory (60 items) and in-depth interview guide along with subject data sheet were used. Permission to use the standardized tools was obtained. Overall, possible knowledge scores related to CAH ranged from 0 to 22, subcategorized as excellent (16-22), good (8-15), and poor knowledge (<7). Parents' stress on PSS ranged from "strongly disagree" to "strongly agree" (ranged from 1 to 5) on 5 point Likert scale with overall possible PSS ranging from 18 to 90, and categorized as mild (18-41), moderate (42-65), and severe stress (66-90). COPE inventory, a multidimensional coping inventory on 5 scales (of 4 items each) (α=0.37-0.93) measures conceptually distinct aspects of problem-focused coping (active coping, planning, suppression of competing activities, restraints coping, and seeking of instrumental social support); emotion-focused coping (seeking of emotional social support, positive reinterpretation, acceptance, denial, and turning to religion); and less useful coping (focus on and venting of emotions, behavioral disengagement, mental disengagement, humor, and substance abuse); responses included "I usually do not do this at all, I usually do this a little bit, I usually do this a medium amount, and I usually do this a lot" and was scored from 1 to 4, respectively. In-depth interview guide (developed in Hindi) included some trigger questions for exploring parental knowledge, and stress arising from fear, personal perceptions, views of people in society about the disease, and the concerns about the future of their child. All the interviews were audio recorded using intex mobile (model number i6). Active listening, nonverbal encouragement, reinforcing, and support were used to enable parents to give the information. At the end of the interview, if parents had any myths they were corrected by the researcher.

Data Analysis

Collected quantitative data were entered in Excel sheet and analyzed using SPSS 18.0 software. Appropriate descriptive and inferential statistics were used for data analysis. Demographics related to child and parent, knowledge and stress were described in mean, frequency, percentage, and standard deviations. The correlation between knowledge and stress was analyzed using Spearman's correlation coefficient test. The p=0.05 was taken as significant. Qualitative data were analyzed by generating themes and validated by a psychologist.

RESULTS

Study participants consisted of an equal number of mothers and fathers (50%), with most having age more than 35 years of age (50%). The level of education of fathers was more than mothers. The majority of the parents were Hindu (66.7%), and were from joint family (80%). Most of the fathers (60%) were engaged in a private job, while majority of the mothers (86.7%) were housewives (Table 1). Monthly family income was between Rs. 20,001 and 40,000 (80%). All the parents had an informal support group of either friends or family members, and most of them (60%) were satisfied with the support of the friends and relatives. Majority of the children were female (73.3%), below 5 years of age (60%), and diagnosed at birth (73.4%). The majority of the enrolled parents of children with

Table 1: Sociodemographic profile of parents of children with CAH (n=30)

Variables	Frequency (%)
Age (years)	
20-25	2 (6.7)
25-35	13 (43.3)
More than 35	15 (50.0)
Relationship of the informant with child	
Father	15 (50)
Mother	15 (50)
Marital status	
Married	30 (100)
Education qualification (mother)	
Illiterate	9 (30)
Primary	3 (10)
Senior secondary	12 (40.0)
Graduate and above	6 (20.0)
Education qualification (father)	
Illiterate	3 (10)
Primary	2 (6.7)
Secondary	6 (20)
Senior secondary	9 (30)
Graduate and above	10 (33.3)
Religion	
Hindu	20 (66.7)
Muslim	9 (30)
Sikh	1 (3.3)
Type of family	
Joint	24 (80)

(Contd...)

Table 1: ***(Continued)***

Variables	Frequency (%)
Single	6 (20)
Occupation (father)	
Pvt. service	18 (60)
Business	3 (10)
Farmer	2 (6.7)
Government job	7 (23.3)
Occupation (mother)	
Pvt. service	3 (10)
Government service	1 (3.3)
Housewife	26 (86.7)
Family income (Rs.)	
20,000-40,000	24 (80)
40,001-60,000	6 (20)
Number of children	
One	9 (30)
Two	10 (33.3)
Three or more	14 (36.7)
Age of child (years)	
0-4	18 (60)
4-9	7 (23.3)
>9	5 (16.7)
Sex of child	
Male	7 (23.4)
Female	22 (73.3)
Not assigned yet	1 (3.3)
Age at diagnosis	
At birth	22 (73.4)
Up to 2 years	4 (13.3)
3 years and above	4 (13.3)
Type of CAH	
Classical	1 (3.3)
Nonclassical	2 (6.7)
Do not know	27 (90)

CAH: Congenital adrenal hyperplasia

CAH (90%) did not know the type of CAH the child was suffering from.

The median (range) knowledge and stress scores of the parents related to CAH were 1 (0-19) and 47 (33-66), respectively. Majority of the parents (67%) had poor knowledge, while 13% and 20% of the parents had good and excellent knowledge scores related to CAH. Most of the parents' (67%) poor knowledge and moderate stress related to the disease, were negatively correlated ($r=-0.386$, $p<0.05$) (Table 2 and Fig. 1). Qualitative data also revealed the same results. Some of the verbatim responses of the participants are described in Box 1.

The coping strategies used by parents in dealing with stress were mainly problem focused (active coping [60%] and planning [60%], instrumental social support [53%], suppression of competing activities [50%]), and emotion-focused (positive reinterpretation [60%], emotional social support [67%], acceptance [53%], and religious coping [50%]). The coping

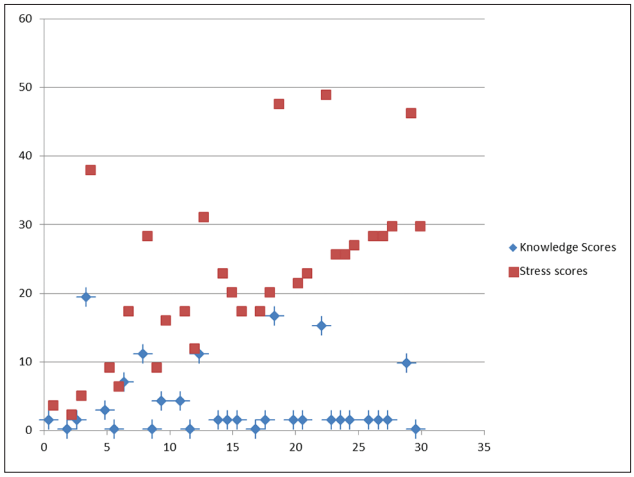

Figure 1: Correlation between knowledge and stress scores of parents related to congenital adrenal hyperplasia

strategies not used or used very infrequently in small measure were humor (100%), substance abuse (100%), behavioral disengagement (87%), and mental disengagement (80%), while less useful coping strategies used in medium amount were 'focus on and ventilate emotions' by 57% of parents (Table 3 and Fig. 2).

DISCUSSION

Main findings of the study were that most of the parents had poor knowledge and experienced moderate stress due to child's disease condition. Knowledge of the parents was negatively correlated with the stress related to CAH. Most of the parents reported using a wide range of problem or emotion-focused coping strategies. Less commonly used coping strategies were mental disengagement, humor, denial, and substance abuse, while behavioral disengagement coping strategy was used by more than half of the parents.

Knowledge makes a lot of difference in parents' perspective toward the disease and help to reduce their stress. In this study, the educational status of the interviewed mothers was lower than the interviewed fathers. However, we could not establish any association of knowledge and stress with the educational status of the parents due to small sample size. The majority of the parents did not know about the type of CAH child was suffering from. Both quantitative and qualitative data revealed that majority of the interviewed parents had poor knowledge and moderate stress related to the disease; however, no myths or misconceptions related to disease condition could be found. Findings of this study are in line with the findings of Barg et al. [9] who reported deficient knowledge among the parents of children with CAH. However, these findings are in contrast to the findings given by Bhakhri and Jain [10] in which myths and misconceptions were found highly prevalent among the parents. In this study, most of the parents had poor knowledge scores related to CAH suggesting the need for educating parents to improve their knowledge parents' knowledge, similar to Duguid et al. [11] who advocated for educating the parents about the genital surgeries.

In this study, majority of the parents reported experiencing a moderate amount of stress, despite having adequate self-reported

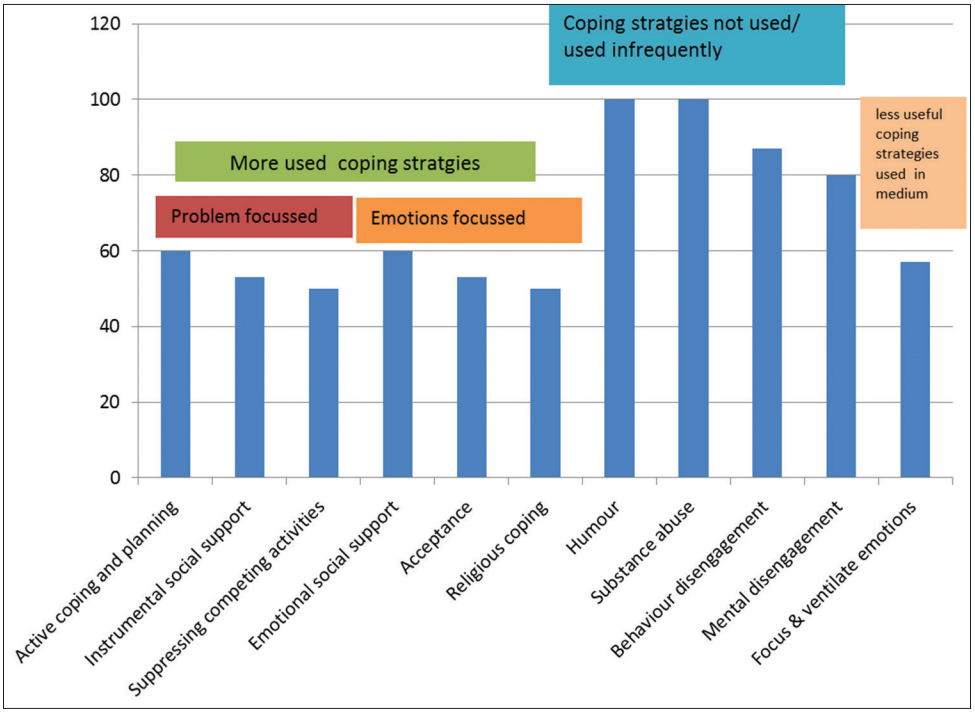

Figure 2: Coping strategies used by parents of congenital adrenal hyperplasia children

Table 2: Correlation between knowledge and stress of parents of children with CAH (n=30)

Variable	Category (scores)	f (%)	Scores (mean±SD), Min-max	r value	p value
Knowledge	Excellent (16-22)	6 (20)	3.5±5.31 (0-19)	-0.386	0.035*
	Good (8-15)	4 (13)			
	Poor (<7)	20 (67)			
Stress	Mild (18-41)	9 (30)	47.6±10.4		
	Moderate (42-65)	20 (67)			
	Severe (66-90)	1 (3)			

CAH: Congenital adrenal hyperplasia

Box 1: Verbatim

Knowledge:

- They (doctors) have told us that for her (child) everything is alright. Like other ladies, like us. She is also having all the internal organs alright. Just only. It is enlarged (referring to clitoris)…….so, by operation this would be corrected….we got her operated. I do not know much about the disease, but I know that ….medicine will be given to her until she is alive. (I-1)
- "...That I do not know. 'Yes I came to know that it is due to deficiency of hormone. Like there is deficiency in my husband as well as in me, so it happened." (I-5)
- "...We are informed that he is having a problem of salt in his body, but the reason for it I do not know." (I-6)
- "Many of us do not know about this disease. Those who understand, they do not think badly about it, and those who are from this generation. But. ...who are from the old generation, If we tell them in detail about the disease, they think badly about it." (I-3)
- "....Means. People from village have a wrong view about it. Doctor informed us that she is a girl, but ladies think she is that (referring hijras)." (I-7)

Causes of stress:

- **Worry and fear**
  When I came to know., I was very much disturbed. Then...like in our house and near ones told us that she looks like female, no need to worry about. everything will be alright, ...But still we worry and have fear in our heart, about the future of child. (I-2)
  I. am very much afraid off, when people talk about it, very afraid off that if somebody will take her away. (crying). (I-3)
  Means everyone will laugh after seeing her, which is the only fear I have whether my child will be alright. (I-5)
- **Concern about the future of the child**
  In her life she will not conceive then her husband will not accept her, and she will be in great trouble, might be deserted by her husband…. For how long parents will take care of her? We will be satisfied only when she conceive and deliver. Then only we will be satisfied. (I-1)
  Future.after marriage life? . "Will she have a normal married life?" (I-4)
  To me ..."I fear that in future she will become a girl . how she will look, all this I think." (I-5)

informal support in the form of family or friends, which could be directly related to the inadequate knowledge of parents related to CAH. Analyzed qualitative data also suggested that parents did not feel free to talk about their child's disease and wanted

Table 3: Coping strategies used by the parents of CAH children (n=30)

Domain	Category	f (%)
Positive reinterpretation and growth	Never do	1 (3)
	Do a little bit	3 (10)
	Do in medium amount	18 (60)
	Do a lot	8 (27)
Mental disengagement	Never do	11 (37)
	Do a little bit	13 (43)
	Do in medium amount	5 (17)
	Do a lot	1 (3)
Focus on and ventilate emotions	Never do	1 (3)
	Do a little bit	5 (16)
	Do in medium amount	17 (57)
	Do a lot	7 (24)
Use instrumental social support	Do a little bit	2 (7)
	Do in medium amount	16 (53)
	Do a lot	12 (40)
Active coping	Never do	1 (3)
	Do a little bit	4 (13)
	Do in medium amount	18 (60)
	Do a lot	7 (24)
Denial	Never do	10 (33)
	Do a little bit	17 (57)
	Do in medium amount	3 (10)
Religious coping	Never do	1 (3)
	Do a little bit	2 (7)
	Do in medium amount	12 (40)
	Do a lot	15 (50)
Humor	Never do	17 (57)
	Do a little bit	13 (43)
Behavioral disengagement	Never do	9 (30)
	Do a little bit	17 (57)
	Do in medium amount	4 (13)
Restraint	Never do	2 (7)
	Do a little bit	6 (20)
	Do in medium amount	14 (46)
	Do a lot	8 (27)
Use emotional social support	Do a little bit	4 (13)
	Do in medium amount	20 (67)
	Do a lot	6 (20)
Substance abuse	Never do	19 (63)
	Do a little bit	11 (37)
Acceptance	Do a little bit	4 (13)
	Do in medium amount	16 (53)
	Do a lot	10 (34)
Suppression of competing	Never do	2 (7)
	Do a little bit	6 (20)
	Do in medium amount	15 (50)
	Do a lot	7 (23)
Planning	Do a little bit	3 (10)
	Do in medium amount	18 (60)
	Do a lot	9 (30)

CAH: Congenital adrenal hyperplasia

to hide it from others. There were no studies available on stress among the parents of children with CAH to compare our findings with. In our study, majority of the parents were young, having female children below 5 years of age. Older children could not be enrolled due to time constraints. All the children were diagnosed at birth, therefore started early on treatment, and had controlled symptoms, not showing the features of severe illness or virilization, therefore, may not truly reflect the feelings of all the parents of children with CAH. There is also the need to explore the feelings and reaction of older children with CAH. Small sample size, cross-sectional study design and convenient enrolment of parents further limit the generalizability of the study. Longitudinal study may help in real exploration of stress and the coping strategies used by the parents at different points of time.

Ignorance about the child's disease condition can be the source of stress for the parents resulting to anxiety and depression. In our study, parents having low knowledge scores had more stress. Similar to our finding, de Silva et al. [12], Barg et al. [9], Bhakhri and Jain [10], and Duguid et al. [11] reported of parents' having depression, clinical levels of stress, anxiety, worry, and helplessness as emotional reactions toward the future sexual life of their child. In this study, majority of the parents had expressed concern about the future of the child as revealed by the qualitative data. There is a need to establish a formal multidisciplinary forum that can act as a platform to address to knowledge deficit needs of the parents, where parents could express and share their concerns. It was lacking in this setting, but it could be existed informally. However, a small number of parents with severe stress due to their children with CAH were referred to clinical psychologists for counseling.

Coping strategiesaims at changing the way parents think or feel about a stressful situation arising due to child's condition. Type of coping effort used by the parents can result in adaptation or maladaptation. Most of the parents, in this study used had reported of using a wide range of problem or emotion-focused coping strategies. Little bit used coping strategies by the parents were mental disengagement, humor, denial, and substance abuse, while behavioral disengagement (withdrawing before reaching the goal, quitting, etc.) coping strategy was used by more than half of the parents.

This shows that majority of the parents were using positive coping strategies to overcome their stress, which is in agreement with the study findings of Duguid et al. [11] where the majority of parents did not display abnormal level of coping. As health-care personnel, we should encourage parents to use healthy positive and useful coping strategies rather than negative and less useful ones.

CONCLUSION

Parents of children with CAH were having negatively correlated poor knowledge and moderate stress. Most of the parents used problem focused or emotion-focused coping strategies in medium amount.

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