

Sleeping problems in mothers and fathers of patients suffering from congenital central hypoventilation syndrome

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Abstract

Purpose Advanced medical technology has resulted in an increased survival rate of children suffering from congenital central hypoventilation syndrome. After hospitalization, these technology-dependent patients require special home care for assuring ventilator support and the monitoring of vital parameters mainly during sleep. The daily challenges associated with caring for these children can place primary caregivers under significant stress, especially at night. Our study aimed at investigating how this condition affects mothers and fathers by producing poor sleep quality, high-level diurnal sleepiness, anxiety, and depression.

Methods The study included parents of 23 subjects with congenital central hypoventilation syndrome and 23 healthy subjects. All parents filled out the Pittsburgh Sleep Quality Index (PSQI) questionnaire, Epworth Sleepiness Scale (ESS), Beck Depression Inventory (BDI-II), and Beck Anxiety Inventory (BAI).

Results A comparison between the two groups showed that parents of patients had poorer sleep quality, greater sleepiness, and higher BDI-II scores compared to that of parents of healthy subjects (respectively, PSQI score 6.5 vs 3.8, ESS score 6.2 vs 4.3, BDI-II score 8.4 vs 5.7). Specifically, mothers of patients showed poorer sleep quality and higher

BDI-II scores compared to that of mothers of controls (respectively, PSQI score 7.5 vs 3.8, BDI-II score 9.3 vs 5.9), whereas fathers of patients showed greater levels of sleepiness with respect to fathers of healthy children (respectively, ESS score 6.8 vs 4.0). These differences emerged in parents of younger children.

Conclusions Congenital central hypoventilation syndrome impacts the family with different consequences for mothers and fathers. Indeed, while the patients' sleep is safeguarded, sleeping problems may occur in primary caregivers often associated with other psychological disorders. Specifically, this disease affects sleep quality and mood in the mothers and sleepiness levels in the fathers.

Keywords Sleep quality · Sleepiness · Depression · Anxiety · Congenital central hypoventilation syndrome

Introduction

Over the last decade, medical advances have led to increasing numbers of families living with technology-dependent children in their homes. Several studies have evaluated the experiences of these families [1–4], but very few have examined the stressful condition in parents of patients suffering from congenital central hypoventilation syndrome (CCHS) [5, 6].

CCHS, also known as Ondine's curse, represents a paradigm of this unusual situation. An extremely rare disease and a lifelong condition first described in the literature by Mellins in 1970 [7], it involves a genetic multisystem disorder characterized by a complex autonomic nervous system dysfunction, the main feature of which is represented by failure to sustain the ventilatory drive during sleep. CCHS patients therefore require lifelong mechanical ventilation during sleep and also during wakefulness in the most severe cases. The complex picture of the illness may also include Hirschsprung's disease

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(HD), tumors of neural-crest origin, and dysregulation of the autonomic nervous system which governs thermoregulation, cardiac rhythm, blood pressure, and digestive motility [8]. In 2003, genetic research revealed that CCHS is caused by a mutation in the paired-like homeobox PHOX2B gene. Thus, the discovery of the genetic link between PHOX2B gene mutations and CCHS represents a breakthrough and a new step forward in early diagnosis of CCHS [9, 10].

In France, the incidence of this disease is estimated around 1 per 200,000 live births [11]. Furthermore, a higher incidence can be expected if we take into account later-onset cases of CCHS, which are characterized by a less severe genetic alteration and hypoventilation only during sedation or pulmonary illness [12]. Although no official data exist in Italy for the incidence of CCHS, it is plausible to expect a similar rate to that reported in other European countries.

CCHS implies lifelong ventilator assistance, and this dependence may affect the quality of life of patients and families alike. However, good parent compliance and vigilant, ongoing care is likely to improve the long-term outcome [13].

Indeed, many studies [3, 14–16] indicate that care delivery to a technology-dependent child entails several repercussions that permeate the social, emotional, and financial dimensions. The presence of technical devices necessary for the patient's survival may transform the home environment into a miniature intensive care unit [17]. The usual connotation of "home" is modified by the presence of medical technology, and the sense of comfort and familiarity usually associated with "home" is diminished [18]. Negative sibling relations, behavioral changes in siblings, marital problems, and discord may occur [18]. In addition, physical and emotional burnout, a loss of privacy, and the financial burden can deteriorate family functioning [18]. The effects of this technological dependence on the neuropsychological, behavioral, and adaptive functioning of children with this rare syndrome have been evaluated by several studies [19, 20], but to our knowledge, very little data exist concerning the impact of this extremely rare disease on the caregivers' quality of life and particularly on their sleep quality considering mothers and fathers separately, since they may be different with respect to their psychological and physiological reactions to the child's disability [21–23].

A recent trial [5] performed on 26 Italian patients aged between 1.5 and 17.5 years reported that CCHS is associated with reduced attention capacity, reduced learning and school performance, language problems, and inadequate social interactions which could lead to a worsening in the quality of life of the children themselves and their parents. Furthermore, Carnevale and colleagues [6] reported in families with ventilator-assisted children at home, "a dense sense of isolation, suggesting that in the light of the complex medical needs of these patients, neither the extended families nor the medical system could support the families' respite needs". More recently, Meltzer and colleagues [24] exploring the relationship

between home care nursing support and daytime functioning in primary caregivers (for the majority, mothers) of ventilator-assisted children found that caregivers without regular night nursing had a reduced amount of total sleep time and clinically significant symptoms of depression and sleepiness compared to that of caregivers with regular support. Certainly, a chronic illness has a significant impact on diurnal family functioning with sleep disruptions for patients and caregivers during the night [25]. In particular, parent sleep can be impaired by heightened vigilance (mainly with patients who are technology dependent), frequent night awakenings, worries about the child's health, and changes in sleeping arrangements [26], but these impairments may be different between mothers and fathers. Significant sleep shortening can lead to decreased alertness and cognitive impairment [27], and the sleep curtailment experienced by parents may also interfere with their ability to provide the best medical care [28, 29]. Many studies have highlighted the relationship between sleep disruptions and symptoms of depression, anxiety, poor parent health, and a high level of marital discord [19, 30–32]. In fact, sleep disturbances can impair daytime functioning, increase the risk of psychiatric disorders, and also worsen the course of chronic conditions [33–35]. Furthermore, poor sleep quality and a decreased amount of sleep can also negatively affect mood and diurnal vigilance.

In view of the abovementioned aspects, to be parents of a technology-dependent child is expected to induce significant discomfort leading to poor sleep quality, diurnal sleepiness, high level of anxiety, and depression. Nevertheless, poor attention has been devoted to explore whether these effects differ between fathers and mothers. The aim of this study was to evaluate sleep quality, diurnal sleepiness, anxiety, and depression in primary caregivers of CCHS patients comparing mothers with fathers.

Material and methods

Participants

Participants provided written informed consent upon enrollment in the study.

The study was approved by the Ethics Committee of the Meyer Children's Hospital, University of Florence, in April 2013.

Twenty-three families attending the annual meeting of the Italian CCHS Association were enrolled for the study. All CCHS patients had a polyalanine expansion mutation in the PHOX2B gene. The patients were ventilated during sleep via a nasal mask or via tracheostomy, with the following types of ventilators: Philips Respironics® Trilogy 100 and Puritan Bennett® 560. Two of them also occasionally used the diaphragma pacemakers mod. Mark IV (Avery Biomedical®).

The monitoring devices were pulse oximeters (Masimo® - Radical 7, Nellcor® N-560), capnograph/oximeters (Nellcor® NPB-70), and capnographs (Novamatrix® Capnograd).

The alarms were set as follows: oxygen saturation 90 % and carbon dioxide 50 mmHg.

The heart rates were set depending on the age: first month of life 80/bpm, second month of life 70/bpm, from third to sixth month of life 60/bpm, and after the sixth month of life 50/bpm.

Twenty-three age-matched families of healthy subjects carrying out checkups in outpatient centers were recruited as a control group. All control subjects were in good health and without any history of neurological or other central nervous system-related disorders or referral for sleep complaints Table 1.

Instruments

Pittsburgh Sleep Quality Index (PSQI)

The Pittsburgh Sleep Quality Index (PSQI) is an instrument used to measure the quality and patterns of sleep in adults. It consists of 19 self-rated questions grouped in seven sections (sleep latency, sleep duration, sleep disturbances, subjective sleep quality, habitual sleep efficiency, use of sleeping medication, and daytime dysfunction) each weighted equally on a 0–3 scale [36]. The global sleep quality score of “5” or more indicates a poor sleeper. The Italian version was used [37], Cronbach’s alpha 0.83.

Epworth Sleepiness Scale (ESS)

This is a self-administered scale intended to measure daytime sleepiness in daily life which measures the probability of falling asleep in different real-life situations [38]. Subjects were asked to indicate on a 4-point Likert scale (0=never, 3=high chance) the likelihood of “dozing off or falling asleep” in 8 different situations. The scores range from 0 to

24, and the proposed range for normal subjects is 0–10 [39]. The Italian version was used [40], Cronbach’s alpha 0.88.

Beck Depression Inventory II (BDI-II)

The BDI-II consists of 21 questions for assessing the intensity of depression in clinical and normal adolescents and adults. Each question has a set of four statements arranged in increasing severity about a particular symptom of depression and the total score ranges from 0 to 63 [41]. The manual states that higher scores indicate more severe depressive symptoms with scores 0–13 indicating minimal depression, scores 14–19 indicating mild depression, scores 20–28 indicating moderate depression, and scores 29–63 indicating severe depression. The Italian version was used [42], Cronbach’s alpha 0.80.

Beck Anxiety Inventory (BAI)

BAI is a 21-question self-report instrument for measuring the severity of anxiety in adolescents and adults. It distinguishes anxious diagnostic groups (panic disorder, generalized anxiety disorder, etc.) from non-anxious diagnostic groups (major depression, dysthymic disorder, etc.). Each symptom item has four possible answers from no symptom to severe symptom. For each item, the patient is asked to report how he or she has felt during the last week. The items are scored as 0, 1, 2, or 3. The score range is from 0 to 63. A total score of 0–7 is considered minimal range, 8–15 is mild, 16–25 is moderate, and 26–63 is severe. So, higher scores correspond to higher levels of anxiety [43]. The Italian version was used [44], Cronbach’s alpha 0.89.

Procedure

All mothers and fathers of healthy subjects and patients completed, independently from each other, a series of self-reported measures including the Pittsburgh Sleep Quality Index (PSQI), Epworth Sleepiness Scale (ESS), Beck Depression

Table 1 Characteristics of CCHS and healthy subjects

	CCHS group	Control group
Gender	M=11; F=12	M=10; F=13
Age	9.2±6.9	8.2±6.4
Type of ventilation		
Nasal mask ventilation	11	Nothing
Ventilation via tracheostomy	12	
Other children in the home, No.		
0	11 (47.8)	10 (43.5)
1	11 (47.8)	12 (52.1)
2	1 (4.4)	1 (4.4)
≥3	0	0
Age of other children in the home	11.9±7.5	11±7.8

Table 2 Demographic variables for parents^a

Characteristics	CCHS group (n=46)	CONTROL group (n=46)	χ^2	p
Age	43.2±6.9	41.9±7.5		
Education				
Primary school	1 (2.2)	0	4.1	0.53
Secondary school	10 (21.7)	13 (28.2)		
High school	27 (58.7)	26 (56.5)		
Bachelor's degree	0	2 (4.4)		
Master's degree	5 (10.9)	3 (6.5)		
Ph.D.	3 (6.5)	2 (4.4)		
Marital status				
Married	42 (91.3)	40 (87)	10.0	0.007
Not married	0	6 (13)		
Divorced	4 (8.7)	0		
Employment				
Full time	20 (43.5)	30 (65.2)	9.3	0.009
Part time	10 (21.7)	12 (26.1)		
Unemployed	16 (34.8)	4 (8.7)		
Kind of job				
Worker	7 (23.3)	9 (21.4)	0.7	0.85
Office worker	13 (43.3)	22 (52.4)		
Freelancer	8 (26.7)	8 (19)		
Manager	0	0		
Other	2 (6.7)	3 (7.2)		
Psychotropic medications	7 (15.2)	6 (13.0)	0.08	0.76

^aData are reported as mean±SD values or number (percentage) of subjects

Table 3 Demographic variables for mothers in CCHS group and control group^a

Characteristics	Mothers CCHS group (n=23)	Mothers control group (n=23)	χ^2	p
Age	42.1±6.2	41.08±7.2		
Education				
Primary school	0	0	4.2	0.36
Secondary school	2 (8.7)	4 (17.4)		
High school	17 (73.9)	14 (60.8)		
Bachelor's degree	0	2 (8.7)		
Master's degree	3 (13)	1 (4.4)		
Ph.D.	1 (4.4)	2 (8.7)		
Marital status				
Married	21 (91.3)	20 (87)	5.0	0.08
Not married	0	3 (13)		
Divorced	2 (8.7)	0		
Employment				
Full time	1 (4.3)	11 (47.8)	17.8	0.000
Part time	6 (26.1)	9 (39.1)		
Unemployed	16 (69.6)	3 (13.1)		
Kind of job				
Worker	0	3 (15)	2.9	0.22
Office worker	4 (57.1)	14 (70)		
Freelancer	3 (42.9)	3 (15)		
Manager	0	0		
Other	0	0		
Psychotropic medications	5 (21.7)	4 (17.4)	0.13	0.71

^aData are reported as mean±SD values or number (percentage) of subjects

Table 4 Demographic variables for fathers and fathers in CONTROL group^a

Characteristics	Fathers CCHS group (n=23)	Fathers control group (n=23)	χ^2	<i>p</i>
Age	44.1±7.5	42.6±7.9		
Education				
Primary school	1 (4.4)	0	3.2	0.51
Secondary school	8 (34.8)	9 (39.1)		
High school	10 (43.4)	12 (52.2)		
Bachelor’s degree	0	0		
Master’s degree	2 (8.7)	2 (8.7)		
Ph.D.	2 (8.7)	0		
Marital status				
Married	21 (91.3)	20 (87)	5.0	0.08
Not married	0	3 (13)		
Divorced	2 (8.7)	0		
Employment				
Full time	19 (82.6)	19 (82.6)	1.1	0.56
Part time	4 (17.4)	3 (13)		
Unemployed	0	1 (4.4)		
Kind of job				
Worker	7 (30.4)	6 (27.3)	0.3	0.95
Office worker	9 (39.1)	8 (36.4)		
Freelancer	5 (21.8)	5 (22.7)		
Manager	0	0		
Other	2 (8.7)	3 (13.6)		
Psychotropic medications	2 (8.7)	2 (8.7)	0	1

^aData are reported as mean±SD values or number (percentage) of subjects

Inventory II (BDI-II), Beck Anxiety Inventory (BAI), and a questionnaire about socio-demographic characteristics (age, years of education, marital status, employment, psychotropic medication). Other information regarding the characteristics of families (ventilation modality for CCHS patients, presence and age of other children in the home) and sleeping habits (role of mother and father assumed during the night and techniques used to help child fall asleep) were obtained from the mother. The parents of CCHS patients filled the questionnaires during the annual meeting of the Italian CCHS Association. The parents of healthy subjects completed the

questionnaires while waiting for the sons’ check-up in outpatient centers.

Data analysis

Preliminary analyses were performed in order to evaluate whether there were any socio-demographic and sleep habit differences between CCHS and control parents, between the

Table 5 Sleep habit variables for parents^a

Children	CCHS group (n=23)	Control group (n=23)	χ^2	<i>p</i>
Nighttime caregiver				
Mother	4 (17.4)	0	5.5	0.06
Father	0	0		
Mother and father	5 (21.7)	3 (13)		
No necessity	14 (60.9)	20 (87)		
Techniques to help child sleep ^b				
Drink	6 (26.1)	1 (4.4)	11.6	0.02
Holding	0	2 (8.7)		
Staying close	2 (8.7)	0		
Bringing into bed	0	4 (17.4)		
No necessity	15 (65.2)	16 (69.5)		

^aData are reported as number (percentage) of subjects

^bMothers reported only the most used technique

mothers of both groups and between the fathers of both groups, using chi-squared test for all variables.

PSQI, ESS, BDI, and BAI scores between the CCHS and control parents, between the mothers of both groups, and between the fathers of both groups and among parents as function of children age (early childhood, and middle and late childhood and adolescence) [45] were compared. Specifically, three age groups were identified: 1–5 years (9 mothers and 9 fathers of CCHS patients, 10 mothers and 10 fathers of healthy children), 6–11 years (6 mothers and 6 fathers of CCHS patients, 7 mothers and 7 fathers of healthy children), and 12–22 years (8 mothers and 8 fathers of CCHS patients, 6 mothers and 6 fathers of healthy children). All the statistical analyses were performed through Kruskal-Wallis test. Significance was set at $p \leq 0.05$.

Results

All the parents of both groups completed every item of the questionnaires; no data were missing.

Socio-demographic characteristics

The socio-demographic characteristics were similar in both groups, with the exception of the marital and the occupational status (see Tables 2, 3, and 4). More specifically, a greater number of parents were not married in the control group, and a greater number of parents were divorced in the CCHS group. Moreover, a greater number of mothers were unemployed in the CCHS group with respect to the control group.

Sleep habits

The majority of parents in both groups reported no necessity of intervention during the night (see Table 5). When intervention was needed, presumably among parents of younger children, both mothers and fathers were available during the night without difference between the two parents' groups. The parents of CCHS patients reported using drink to help child

Table 6 Pittsburgh Sleep Quality Index, Epworth Sleepiness Scale, Beck Depression Inventory, and Beck Anxiety Inventory scores in parents of CCHS patients and healthy subjects (mean±SD)

	CCHS group	Control group	χ^2	p
Total PSQI score	6.5±3.7	3.8±2.5	11.8	0.0006
Total ESS score	6.2±4.4	4.3±3.3	3.8	0.04
Total BDI-II score	8.4±6.1	5.7±5.2	4.8	0.02
Total BAI score	6.6±6.5	5.0±5.3	1.1	0.29

Table 7 Pittsburgh Sleep Quality Index, Epworth Sleepiness Scale, Beck Depression Inventory, and Beck Anxiety Inventory scores in mothers of children with CCHS and healthy subjects (mean±SD)

	Mothers CCHS group	Mothers control group	χ^2	p
Total PSQI score	7.5±3.3	3.8±2.4	12.2	0.0005
Total ESS score	5.5±3.5	4.6±3.2	0.5	0.47
Total BDI-II score	9.3±6	5.9±4.6	3.9	0.04
Total BAI score	7.3±6.9	6.2±5.9	0.2	0.59

sleep, whereas the parents of healthy subjects were more prone to hold children or bring into bed.

Quality of sleep, sleepiness, depression, and anxiety

The parents of CCHS patients showed higher PSQI, ESS, and BDI-II scores than the parents of healthy subjects (see Table 6).

The comparison between mothers of subjects affected by CCHS and mothers of healthy subjects showed that mothers of patients had higher PSQI and BDI-II scores than that of mothers of healthy subjects (see Table 7), whereas fathers of the CCHS group had higher ESS scores than that of fathers of the control group (see Table 8). Only the parents of CCHS patients ranging between 1 and 5 years showed higher PSQI, ESS, and BDI-II scores than that of the parents of healthy subjects (respectively, PSQI score (mean±SD): 7.3±3.4 vs 3.4±2.0, $\chi^2=12.2$, $p=0.0005$; ESS score: 6.3±5.0 vs 3.2±2.8, $\chi^2=5.3$, $p=0.02$; BDI-II score: 10.5±6.1 vs 4.2±3.5, $\chi^2=10.1$, $p=0.001$). More specifically, only mothers of CCHS patients ranging between 1 and 5 years showed higher PSQI and BDI-II scores than that of the mothers of healthy subjects (respectively, PSQI score: 8.1±2.2 vs 3.6±2.3, $\chi^2=9.1$, $p=0.002$; BDI-II score: 10.1±3.9 vs 4±3.6, $\chi^2=8.1$, $p=0.004$), and only fathers of CCHS patients ranging between 1 and 5 years showed higher ESS scores than that of the fathers of healthy subjects (respectively, ESS score: 7.8±6.7 vs 2.9±3.1, $\chi^2=3.8$, $p=0.05$).

Table 8 Pittsburgh Sleep Quality Index, Epworth Sleepiness Scale, Beck Depression Inventory, and Beck Anxiety Inventory scores in fathers of children with CCHS and healthy subjects (mean±SD)

	Fathers CCHS group	Fathers control group	χ^2	p
Total PSQI score	5.5±3.9	3.8±2.7	2.3	0.12
Total ESS score	6.8±5.2	4.0±3.4	3.6	0.05
Total BDI-II score	7.5±6.3	5.5±5.9	1.4	0.22
Total BAI score	5.8±6.1	3.8±4.5	1.0	0.30

Discussion

Our results showed that congenital central hypoventilation syndrome impacts the quality of life of primary caregivers, influencing both physiological and psychological aspects.

More specifically, we found that parents of patients had poorer sleep quality, greater sleepiness, and higher Beck Depression Index II scores compared to that of parents of healthy subjects. These effects can be due also to the absence of night nursing support in CCHS families as shown recently by Meltzer and colleagues [24].

The impact of the illness on caregivers is mainly evident among parents of younger patients, and it differs between mothers and fathers. In particular, the mothers of patients had higher Beck Depression Index II and Pittsburgh Quality Sleep Index scores than that of mothers of healthy subjects, whereas the fathers of patients showed greater daytime sleepiness than that of fathers of the control group.

In addition to worries about their children's health [27] and the impact of the illness on daily life [5], the occupational status of mothers of congenital central hypoventilation syndrome subjects could be one of the contributing factors to their higher Beck Depression Index II scores. In fact, while 94 % of these mothers were employed prior to the disease, at the time of the interview, they were the parental figures most heavily involved in congenital central hypoventilation syndrome patient care, and the majority (69.6 %) was not employed outside the home. Furthermore, as demonstrated by the Beck Depression Index II scores, the poorer sleep quality found in this group could derive not only from sleep disruption due to their children's illness but also from psychological suffering. Indeed, most of the CCHS patients sleep well and have no necessity of night care, but their mothers show sleep problems. Indeed, poor sleep quality might mediate the relationship between the child's health and the caregiver's depression [46]. Several studies [47–49] have highlighted the interrelation between depression and sleep complaints. The total scores of BAI revealed minimal level of anxiety in CCHS and the lack of significant difference between patients' and controls' parents could be explained with the fact that CCHS is a chronic illness, and these caregivers were probably adapted to this long-lasting condition.

The higher daytime sleepiness complained of by fathers of congenital central hypoventilation syndrome subjects can be explained by the fact that in the congenital central hypoventilation syndrome group, the alarm system can awaken or lighten the sleep of both parents to the same extent (the mean number of night-awakening reported by CCHS' mothers (3.6) was very similar to that (3.3) reported by CCHS' fathers). Nevertheless, only mothers could be able to recover from daytime sleepiness due to sleep fragmentation, since the majority of them had to leave their jobs to respond to

new needs related to their child's illness and have the opportunity to rest during the day (48 % of CCHS mothers reported that they sleep in the afternoon), whereas fathers who are employed outside the home are deprived of this possibility.

Although our study provides evidence of the impact of congenital central hypoventilation syndrome on parents, mainly mothers, some limitations should be kept in mind when evaluating our findings.

First, the sample size is small due to the low incidence of this extremely rare disease. Second, there is a lack of objective measures confirming what we found through the self-reported data.

In conclusion, congenital central hypoventilation syndrome impacts the family with different consequences for mothers and fathers. These results underscore the need for family-centered psychosocial intervention, mainly during the early years of patients, focusing more on parent assistance and not just on technical and short-term rehabilitation of the patients. Indeed, greater awareness of the consequences of congenital central hypoventilation syndrome on the parents' life may help health professionals to better understand and support the needs of these families.

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