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Caregiver's burden and psychosocial issues in alternating hemiplegia of childhood

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ABSTRACT

Alternating hemiplegia of childhood (AHC) is rare disorder characterised by recurrent attacks of hemiplegia followed by developmental delay. We investigated the parental perceptions and psychosocial issues of AHC. Using a questionnaire sent to the French AHC association, we investigated families' concerns and needs of support. Additionally, we evaluated the impact of this disease.

Results: We analysed 47 questionnaires from 30 families. At time of diagnosis, the concern of the parents was highest for the hemiplegic spells and abnormal eye movements. These concerns decreased over time. The highest concern at the time of the study was the outcome of the patients with an emphasis on cognitive consequences and the level of autonomy. The results showed that AHC has a significant impact on families.

Interpretation: Our data enhance how the explanation of the disease by healthcare professional is important. This study also highlights the need for family support over time.

1. Introduction

Alternating hemiplegia of childhood (AHC) is a rare disorder characterised by recurrent attacks of hemiplegia affecting either sides of the body, abnormalities of ocular movement, movement disorders and progressive developmental delay.[1,2] Recently, it appeared that de novo mutation of ATP1A3 gene cause AHC.[3] Children with AHC often have a delay in diagnosis, therefore a broad differential diagnosis is necessary when this condition is considered. AHC was first described more than three decades ago but its cause remains unknown,[1,2] and there are few treatments available.[2,4,5] Usually the severity of clinical presentation and neurological disability remain constant with age, with the exception of abnormal ocular movements and hypotonia that appear to regress, but not disappear, into adulthood.[2]

The delay of diagnosis can cause extreme psychosocial stress for parents.[6] When parents are informed of a definitive diagnosis they often experience a wide spectrum of emotions including relief, guilt, shock or anger.[7] Family members also experience stress from the lack of information available about the illness from the medical providers who may be unfamiliar with the intricacies of the disorder.[7]

Children with chronic disease are, as a group, vulnerable to behavioural, emotional, and medical problems that can interfere with daily activities and functioning. These challenges

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also impact the family.[8,9] Chronic disease frequently affects the psychological functioning of parents and siblings, the employment activities and the economic situation of parents.[6] Parents have an intense burden of providing daily care, and the illness often involves coping with ambiguities including uncertain outcome and misdiagnosis.[9]

Compared with the wealth of knowledge that exists regarding the impact of chronic illnesses on family functioning, there is a lack of research on psychosocial issues faced by families of children with rare developmental disorders. We conducted a study on the parental perception and the burden of AHC. Our aim was to ascertain the parental perception of diagnosis and clinical symptoms in cases of AHC. We also attempt to identify parental needs and concerns at time of diagnosis and after diagnosis. Finally, we evaluate the psychosocial adjustment and the burden of AHC on the family.

2. Population and methods

We conducted this survey by an anonymous, self-administered questionnaire (Table 1). The study was conducted among 50 families from the French alternating hemiplegia association (Association Française de l'hémiplégie alternante). Of these, 47 questionnaires were returned (17 fathers and 30 mothers) from 30 families (62%). We examined their responses with their informed consent.

The survey consisted of 12 closed and nine open-ended questions designed to elicit parental opinions about the impact of AHC in their lives. It included demographic variables and information concerning medical history. It also investigated families' concerns (at time of diagnosis and at time of the study). We also investigated their experiences and their needs at time of diagnosis. Finally, we studied, using

Table 1 – Questionnaire used in our study.

1. Identification of the responder:
2. Number of siblings:
3. Current age of your child:
4. Age at the onset of symptoms:
5. Age at diagnosis:
6. Did your child experience seizure? If yes: the current frequency of seizure
7. Who is the doctor of your child? Child neurologist, neurologist, physician of the rehabilitation centre or your GP
8. What is for you the most important question regarding your child or the disease?
9. Please score from 1 to 10 the cause of current concern

Abnormal eye movement, movement disorders, seizure, hemiplegic spells, dystonia, pain, behaviour, cognitive outcome, autonomy

10. Please score from 1 to 10 the cause of concern before the diagnosis

Abnormal eye movement, movement disorders, seizure, hemiplegic spells, dystonia, pain, behaviour, cognitive outcome, autonomy

11. Please score from 1 to 10 the cause of concern during early childhood (2–5 years)

Abnormal eye movement, movement disorders, seizure, hemiplegic spells, dystonia, pain, behaviour, cognitive outcome, autonomy

12. Before the diagnosis of AHC, did the physician discuss with you other diagnosis

13. If yes, which diagnosis?

14. What did you feel when the diagnosis was tell to you? Concern, misdiagnosis, distresses, sadness, disbelief, confusion, culpability, anger, relief, need to understand the disease, need to know long-term consequences of the disease, other

15. After the explanation of the diagnosis, what was a cause of concern? (Please report the three main causes of concern).

Loss of recklessness, loss of friends; changes in relationship with the partner; financial consequences; emotional impact; how to maintain professional life and manage the child's disease? How to organise adult care with our child? How to organise the necessary rehabilitation? Who will take care of our child when we are no longer able?

16. Did you look at information about this disease by yourself? How did you proceed?

17. According to you, what should a physician say when they announce the diagnosis?

18. What did you want to know or hear, and you didn't, when the physician explained the diagnosis?

19. Is there a change of medical team at the time of your child become adult (more than 18-years-old)

20. How has having a child with AHC affected your relationship with your partner? Brought us closer together, no substantial effect, caused conflict, better communicators, caused us to separate

21. What were your most important sources of support? Spouse, family, medical personnel, rehabilitation personnel, AHC support group, friends

22. What are the consequences of the occurrence of AHC in your child on: the family, your social life, your professional life?

open-ended items, the impact of this chronic disease on the family (parental couple, siblings) (Table 1).

2.1. Parents and patients

Demographic data of the patients are summarised in Table 2. The mean age of patients was 19.8 [2–56 years]. The families of three deceased patients also answered the questionnaire.

The diagnosis of a neurological disorder was usually discussed very early in their medical history at the time the patients were newborn or young infants. Most of the parents have had to face the diagnosis of a neurological disorder before age of two (68%), between two and nine years of age (30%) or beyond 10-years-old (2%). Sixty percent of the parents have had to face another diagnosis before the diagnosis of AHC. Differential diagnoses are usually discussed before the diagnosis of AHC: epilepsy (23%), opsoclonus-myoclonus syndrome (4%), Leigh's syndrome (4%) or cerebral palsy (4%).

Fifty-five percent of the cases had epileptic seizures in addition to their hemiplegic episodes based on parental report. The first seizures were observed between one-month-old and fourteen-years-old (median 3.3 years) Of the children with epileptic seizures 8% had events weekly, 31% monthly and 61% <5 events a year.

Anonymous data were recorded. Percentages were rounded to a whole. We report mean ± standard deviation of continuous data. The range of minimal and maximal values of continuous variables is also reported [Min–Max]. Statistical analysis was performed using Kruskal–Wallis test and Dunn post-hoc analysis using GraphPad Prism 5.02 (San Diego, CA, USA). Statistical significance was considered to be $p < 0.05$.

3. Results

3.1. Parental concerns over time

Using questions about various symptoms (Fig. 1) (Table 1, questions 9–11), we evaluated the cause of concern before the diagnosis, after the diagnosis and currently. We observed a change of parental concerns over time. At the time of diagnosis, three symptoms were the most important worries for parents: abnormal eye movements, movement disorders and hemiplegic spells. We noted a significant decrease of these preoccupations over time. Conversely, autonomy and behaviour become more recurring preoccupations over time (Fig. 1). Dystonia, pain and cognitive outcome remain constant worrying themes throughout childhood.

Table 2 – Sample socio-demographic and clinical characteristics ($n = 47$).

Characteristic of the AHC patients	
Mean age at time of the study (years)	19.8 ± 10.3 [2–56]
Mean age of the first symptoms of AHC (months)	3.7 ± 5 [0–24]
Mean age at diagnosis of AHC (years)	3.1 ± 4 [0.2–16]
Mean age at first hemiplegic attacks (years)	2.8 ± 0.5 [1.6–3.1]
Epilepsy history	
Mean age of first seizure (years)	3.3 ± 4.3 [0.08–14]
Current seizure frequency (number by year)	15.2 ± 46.7 [1–216]
Medical follow-up	
Done by a child neurologist	53%
Done by a neurologist	26%
Done by a physician within their institution	11%
Done by the GP	53%

3.2. Parental feelings and needs at the time of diagnosis

Looking at the score given by the parents at the questions 14–15 (Table 1), we observe that they experienced a wide spectrum of emotions (Table 3). Indeed, among the subjects who succeeded in identifying their feelings at the time of diagnosis, we observe a majority of parents which reported initial disorientation, followed by concern and the need to understand. Thereafter, parents reported sadness and feelings of fear.

After the diagnosis of AHC (Table 3), most of the parents wonder whether the disease will improve or worsen (or even whether their child may die). The majority (72%) also wonder who will take care of their child when they won't be able. Anxiety is also frequently reported, often related to specific concerns such as how to maintain their professional life and manage their child's disease (50%), or how to organise the rehabilitation necessary for the care of their child (50%). Conversely, loss of the friends (2%) and the financial consequences (2%) were infrequently reported by the parents.

After the diagnosis, only 28% of the parents did not seek additional information concerning AHC. Some of them report that they feared additional information. The majority of families used the internet research information on the disease (56%). The two other ways to find additional information about the disease were to ask their physicians (35%) and to contact the French AHC association (38%).

Parental suggestions of what information should be provided by the physician at the time of diagnosis include: (1) disease outcome (47%), (2) role of early rehabilitation (23%), (3) treatment options (21%), (4) management of AHC in daily life (13%), and (5) existence of a support group (11%).

Parents provided multiple responses when they were asked to express what they would like to hear from the physician at time of diagnosis. Most of them expressed the fact that the physician should have an empathic attitude. They also would like further information including any possible treatment and the evolution of the disease.

3.3. Impact on the family

In most cases, this neurological disease impacts negatively on the family. Table 4 reports changes to the relationship between the parents. The occurrence of AHC in their child has had extremely different consequences in the relationship of the parents. Many declare having a better relationship because of the need to face the disease together (33%), while 27% evoke conflicts caused by the illness' consequences. However, 20% of parents did not observe any effect in their partnership. It seems also that the occurrence of AHC in their

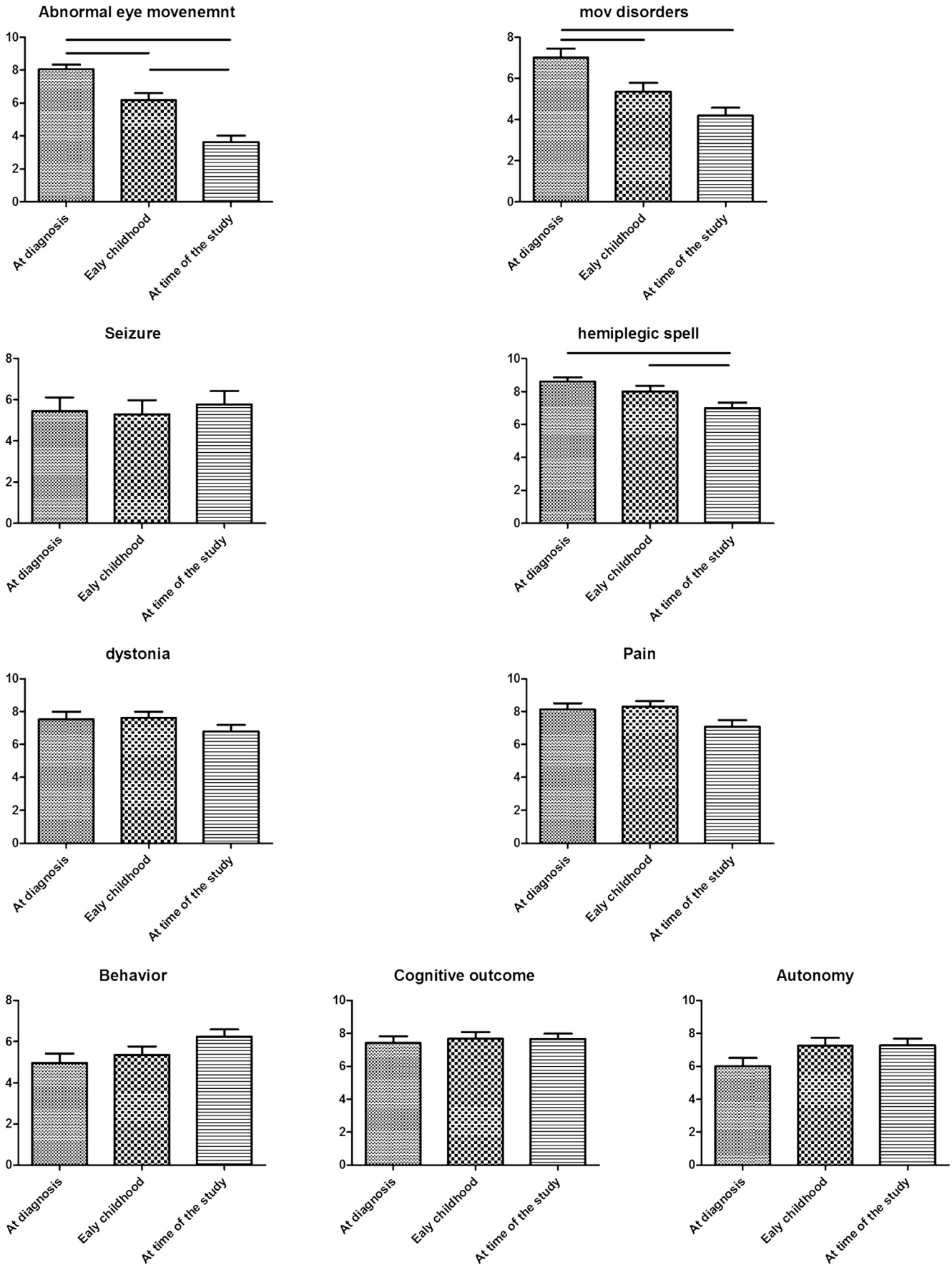

Fig. 1 – Graph of the mean scores (1–10) of various concerns of the parents regarding their child with AHC at time of diagnosis, in childhood and at time of the study.

Table 3 – Concerns following the diagnosis ($n = 32$).

Who will take care of our child when we are no longer able?	72%
Emotional impact	52%
How we maintain a professional career whilst coping with our child?	50%
How will we organize the therapies required for our child's care?	50%
Changes in relationship with partner	28%
Loss of recklessness	22%
How will we ensure that our child always has an adult care?	22%
Loss of friends	2%
Financial consequences	2%

Three maximum responses were possible.

child may have modified their initial life plan. For example, some parents report their doubts concerning the desire to have more children.

According to the parents, siblings are significantly impacted as well. The parents state that they are asked to assume more familial responsibility, and they may receive less attention because of the parental involvement in the care of the AHC patient. The parents report that they think that the siblings may develop anxiety, depressed mood and social isolation. Parents also have the feeling that the siblings frequently try to be perfect to avoid causing additional troubles in the family life. On the other hand, parents also state the fact that facing severe disease may help them to be open-minded and compassionate.

3.4. *Impact on the social and professional life*

All parents state that they have restricted or adapted their social life. Among parents, 21% reported a reduction in their previous social network and 19% report being socially isolated. Additionally, 38% also report a reduction in leisure activity. The parents report that this is also due to the symptoms of the disease, such as unpredictable hemiplegic attacks. Owing to the fact that all activities need to be anticipated, 34% report a reduction in their choice for vacation time due to the disease in their child. Some parents report the need to be close to a hospital.

The mothers have more frequently modified their professional life than the fathers. The parents report a reduction of working time in 19%, a break for 13% and a change of employment activity in 9%. Regarding their jobs, the parents reported fatigue and stress because of the fear of hemiplegic attack occurrence in their child. They also feel guilty to be less available for their child with AHC.

Table 4 – Impact of AHC on relationship with partner ($n = 45$).

Brought them closer together	33%
Caused conflict	27%
No substantial effect	20%
Better communication	7%
Caused them to separate	9%

3.5. *Supports*

When parents reported on the support received from their relatives (Table 5), we note that spouses have a major role in 74% of the cases, as well as the family for 51%. Indeed, the family represents the most important resource while the medical staff (20%), the rehabilitation team (18%) or friends (18%) are more rarely reported as supportive. The parents spontaneously report that their child with AHC brings them support by his/her behaviour and attitude in facing the disease. Family support, provided by the French AHC association was also reported by the parents.

4. Discussion

Families with a child with AHC have a number of psychosocial needs and concerns. Recurrent themes among participants were revealed. We noted that the families report that their concerns were modified over time. The initial worries are focused on the most impressive symptoms while later the concerns appear to focus on autonomy. Suffering constitutes a major theme of concern throughout childhood. This survey also highlights the familial and social burden of AHC.

At time of diagnosis, the families often experience disorientation, concern and a need to understand upon initial diagnosis of AHC. In addition, they experienced a wide spectrum of feelings. When the diagnostic phase is protracted, the long wait for diagnostic certainty can cause even more psychosocial stress for parents.[6] There is also sadness, as parents may wonder how much they can expect from their child, or feelings of grief over the loss of a healthy child.[6,10] These emotional reactions to diagnosis and particular needs are similar to those reported by other researchers in families affected by other chronic illness.[6] Looking at the literature, it seems that family members are wanting for an empathic attitude from the physician.[11]

Many demands and psychosocial issues faced by families of children with AHC are not unique to that specific condition. It seems that parental feelings in AHC are close to the feelings of parents with a disabled child regardless of its origin.[12–14] One of the main challenges for parents is the effective management of their child's chronic health problems while maintaining the requirements of everyday living. The understanding of the global impact of paediatric chronic illness such as AHC is an important issue. The healthcare professional should evaluate and manage the psychological consequences and social-emotional factors that are impacted by a childhood neurologic disorder such as AHC.

Table 5 – Sources of support (multiple responses were possible).

Spouse	74%
Family	51%
Medical personnel	20%
Rehabilitation personnel	18%
Friends	18%

Our study also highlights the impact of the disease on the family. Here, we noted that the relationship between the parents can be unpredictably affected. The parents reported that the disease of their child results in stronger links in 9% while an equal proportion reported conflicts or separation. It has been reported that family function plays a central role in both the physical and the psychological health of caregivers.[15] This construct affects health directly and also mediates the effects of self-perception, social support, and stress management. Some authors suggest that healthcare providers who work with families of children with long-term disabilities should develop interventions that support and nurture the family as a whole.[15] As described in other chronic diseases, the mothers most frequently change their professional life. More generally, it has been reported the differences in impact of child health on health-related quality of life for mothers and fathers.[16–18] Mothers are more inclined than fathers to experience depressive episodes. They also feel a deeper emotional distress. Fathers are more stressed about the financial implications in providing for their child's needs.[19] Some authors suggested that mothers are more often affected than the fathers because they are the most frequent direct caregiver.[18]

In our study, the parents also expressed their feelings regarding siblings of the affected child. They feel that they may exhibit psychological disorders such as anxiety and social isolation. On the other hand, the parents think that the siblings are able to manifest an early maturity and autonomy. The psychological adjustment of the unaffected siblings of a chronic neurologic illness has been documented to a limited extent. Many families experience feelings of guilt over the varied levels of attention they are able to dedicate to each child and anxiety about the effects this may have on the well-being and emotional growth of non-disabled siblings. The literature on siblings of children with other paediatric conditions suggests that they are at slightly increased risk of developing psychiatric problems, especially emotional disorders such as anxiety and depression.[20–22] It is unclear whether sibling age and gender influence adjustment, but some reports suggest increased psychological symptoms in female siblings, more specifically in older sisters of affected males.[23] There are also suggestions that conditions requiring daily treatment may be associated with more negative effects due to the differential treatment of siblings by parents, and/or the excessive domestic responsibilities or caretaking roles placed upon unaffected siblings. More recent research has found little difference in a range of behavioural and social constructs between siblings who do and do not have a brother or sister with an intellectual disability.[24] Current evidence suggests that it is the overall family functioning that is more likely to determine sibling adjustment, although financial hardship is one of the social and economic determinants impacting family functioning.[25]

In our survey, it seems that the mothers of the affected child constituted the major support. One way to cope with the illness, parents of a child with AHC also turn to support groups with parents who know and understand AHC. In the literature, many authors have analysed parental coping in neurologic disease. Other studies have reported that perception-focused coping strategies were used most often by parents. Mothers had a more extensive repertoire than fathers, and the relationship between maternal and paternal coping appeared to be complementary.

The ability of the physician to support the parents and to understand their feelings seems important. Child neurologists should be aware of the worries of parents of a child with AHC. At diagnosis, the parents would feel better if they know that they can contact medical staff in case of any questions. Listening to the parents' experiences and expectations might help the physician to handle the psychosocial aspects adequately. The way to provide information on the disease and its consequences should be done by a series of meetings with both parents, to better understand the illness' consequences and implications. Families expect accurate medical information as well as concrete explanations of their meanings for their future life. We suggest for the explanation of the diagnosis, as well as for the follow-up, to advance stepwise in an ethical approach involving mutual cooperation between the physician and the family. In case of AHC, a multidisciplinary approach may help the family in the care of the AHC patients. The health professional should be aware of the challenges that the parents of a child with AHC are facing. The findings from our study can be used to build stronger interdisciplinary healthcare teams to support families in their efforts to provide care to these children.

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