

Alternating Hemiplegia

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Clinical Picture

Alternating hemiplegia (AH) (also called alternating hemiplegia of childhood) is a rare neurological disorder characterised by frequent, temporary episodes of paralysis on one side of the body (hemiplegia). Symptoms usually begin before 4 years of age and can range from mild, including episodes of paralysis occurring only at night and no neurological impairment, to severe, including paralysis of the legs, arms and eye muscles, mental impairment, problems with gait (step) and balance, excessive sweating, changes in body temperature, seizures and movement disorders. The cause of AH is unknown, although some cases may be inherited as an autosomal dominant trait.



Mike,
alternating hemiplegia
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The prevalence of AH is also unknown. Children with mild forms of AH have a good prognosis. Drug therapy may help to reduce the severity and duration of attacks of paralysis. However, those who experience more severe forms have a poor prognosis as intellectual and mental capacities do not respond to drug therapy, and balance and gait problems continue. Neurological problems may also be associated and can become increasingly severe

with age. While AH is not a progressive disease, episodes of weakness may get worse over time, recovery may become slower and walking unassisted can become difficult or impossible.

Living With Alternating Hemiplegia

Living with AH means permanent adjustment to changing and unpredictable situations for the patient, the family and anyone involved in the patient's life. A child's development is inhibited from birth, depending upon the severity of the neurological seizures and other accompanying

symptoms (e.g. epilepsy). The uncontrolled appearance of paralysis hinders the body's development, requiring a great deal of energy to continue, thus taking energy away from other developmental processes, such as learning. Skills autonomously learnt by the child, such as walking, are constantly disrupted by paralysis. As children with AH cannot rely on their own bodies, psychological problems are often experienced. The unpredictability of the intensity and the duration of the attacks are a burden for all caregivers and require constant attentive monitoring and flexible adaptation to sudden changes in the patient's condition. Activities must be managed differently depending on the presence or lack of paralysis. Not only are daily aspects

'Proper schooling has been a great challenge since the start. We have changed schools three times. We sought help from the local authorities in order to accommodate our daughter's needs, but eventually each time we found the school through friends and neighbours who had experiences with the schools. Even though our daughter has no hearing problems, she attends a school specialised for children with hearing problems — it simply offers small classes and accepted our daughter.'
Parent of child with AH, Austria

that require mobility affected, such as walking, self-sufficiency, continence, speech, eating, swallowing and danger of aspiration (accidental sucking in of food particles or fluids into the lungs), but also activities that require mental functions, such as the ability to concentrate on tasks, which is important in progressing in school. The ability to follow through with these daily functions fluctuates frequently depending upon the child's daily condition and requires a great deal of patience and flexibility from caregivers and teachers. Teachers may require some time to become acquainted with all the possible needs of a student with AH. Healthcare professionals must also take the time to develop appropriate care with patients who have very diverse and individual needs. The best source of information for these individual needs is often the parents of a patient.

For these reasons, it is particularly valuable for patients to take part in support groups that can allow them to learn from the experiences of, and obtain the necessary encouragement from other families with children with AH. Sometimes the best experts on the needs of their child, parents have become indispensable partners for doctors, teachers and caregivers of patients of AH.

Access to Medical and Social Services

PARTICIPANTS IN THE SURVEY

Responses from 79 families of AH patients from six countries were analysed in the survey (*Figure 1*).

Respondents were almost equally female and male (52% and 48%, respectively).

The mean age of patients was 15 years (mean age at diagnosis: 3.2 years).

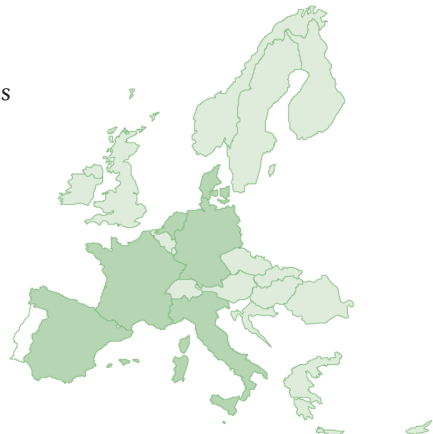


Figure 1
Survey participants affected by AH

NEED FOR MEDICAL SERVICES

Overall, each patient with AH needed an average of nine different kinds of medical services related to their disease (the same as the average number of services required for the 16 rare diseases surveyed). In addition to consultations mentioned in Figure 2, consultations of paediatrics (39%), rehabilitation medicine (30%), emergency services (30%) and orthopaedics (29%) were often needed. The most frequently required explorations were biological testing (56%), electroencephalogram (47%) and radiology (27%), as well as specialised imagery (20%). Other types of care included dental care (52%), physiotherapy (51%), glasses (39%), psychomotility therapy (32%), speech therapy (29%) and nursing care (27%). Hospitalisation occurred in 49% of patients for an average total duration of 15 days.

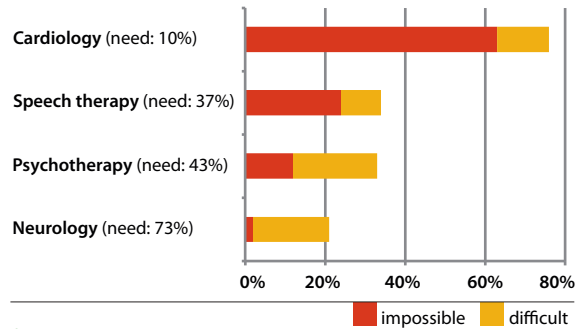


Figure 2
Need for and access to four representative medical services for AH

ACCESS TO MEDICAL SERVICES

Lack of access to medical services in 18% of situations overall for AH patients

Impossible access to services was most frequently reported for cardiology (63%) and speech therapy (24%) services, but also for psychotherapy (12%) and neurology (2%) (Figure 2). A lack of referral was the most frequent barrier to access for all services: neurology (100%), cardiology (80%), speech therapy (71%) and psychotherapy (50%). Unavailability was a hurdle for access to psychotherapy (25%) and speech therapy (14%) services. Personal cost (25%) and waiting time (25%) were also hurdles for accessing psychotherapy services.

Access to medical services was difficult in 14% of situations overall

Although possible, access was reported as difficult by patients for psychology (21%), neurology (19%), cardiology (13%) and speech therapy (10%). An insufficient number of appointments were reported for psychotherapy (28%), neurology (13%) and speech therapy (9%). Personal cost was considered excessive for psychotherapy (44%). Professional assistance for travelling to care centres was provided in 29% of cases for speech therapy services, in 17% of cases for psychotherapy and in only 13% of cases for neurology consultations, despite the fact that neurology consultations were most frequently in another region as compared to other services.

Satisfaction with medical services

Overall, 88% of patients considered that medical services, when obtained, responded fully or partially to their expectations (*Figure 3*). Patients were less frequently satisfied with neurology consultations (87%) and psychotherapy services (87%) compared to cardiology (100%) consultations and speech therapy services (100%).

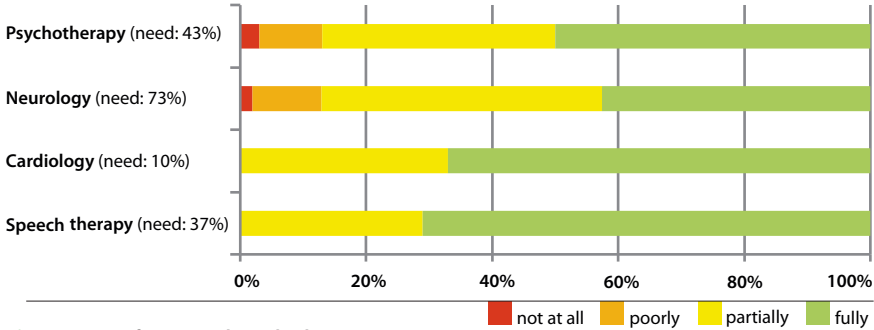


Figure 3 Satisfaction with medical services for AH patients

SOCIAL SERVICES

Amongst the 39% of families requiring social assistance, 3% failed to meet with a social worker and 21% met one with difficulty. When obtained, the level of satisfaction with social assistance was 57% overall, lowest for specialised technical support (22%) and highest for exceptional financial support (62%) (*Figure 4*).

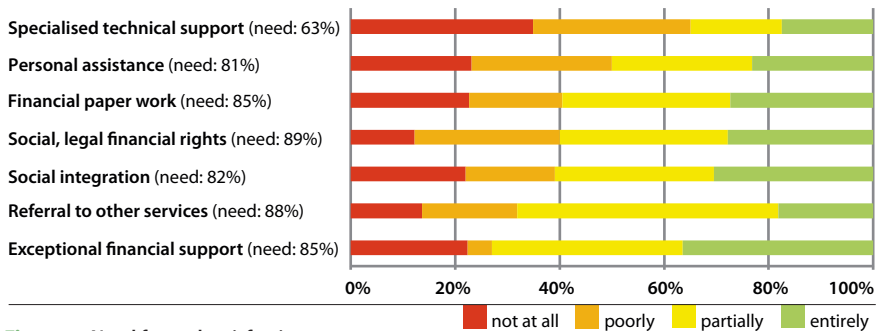


Figure 4 Need for and satisfaction with social services

REJECTION

Patients with AH experienced rejection by health professionals with similar frequency (19%) to the overall respondents for the 16 surveyed rare diseases (18%). The reluctance of health professionals due to the complexity of the disease was reported by 100% of rejected patients. Patients were also rejected for personal aspects including difficulties in communication (7%) and disease-related behaviour (7%). Even if the rejection was mainly linked to the disease rather than the patient, its extent was perceived as a refusal of health professional to treat AH patients.

'I stopped working mainly to look after my daughter, who has alternating hemiplegia.'
Parent of child with AH, Austria

The frequency and cause of rejection varied according to the patient's country of origin (Figure 5).

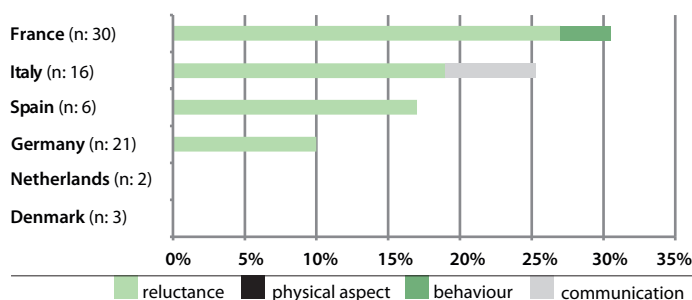


Figure 5
Cumulated frequencies of causes of rejection by country (n: total number of respondents) in AH. As patients may have been rejected more than once for more than one reason, the total number of rejections exceeds the number of rejected respondents.

CONSEQUENCES OF THE DISEASE

As a consequence of the disease, 26% of patients had to move house. Amongst these, 70% had to move to a more adapted house, 20% moved to be closer to a relative and 15% moved to be closer to a specially adapted care centre. As a consequence of the disease, 71% of family members had to reduce or to stop their professional activity to take care of a relative with the disease.

Expectations Regarding Centres of Expertise for Rare Diseases

Not differing from the overall opinion of survey participants, respondents with AH considered the following functions provided by a centre of expertise as the four most essential:

- Coordinating the sharing of medical information on the patient between all professionals who care for him/her in the specialised centre
- Communicating with other specialised centres and professional networks to harmonise treatments and research at the national and European levels
- Coordinating the sharing of medical information between professionals of the specialised centre and local professionals, to facilitate the continuity of the patients' follow-up
- Collaborating with research teams working on the rare disease (in particular for clinical studies)

As onset of AH is usually in early childhood, patients require medical attention throughout their lifetime. Sudden changes depending on the presence or lack of paralysis require constant attentive monitoring by all people involved in the child's life, including teachers.

These aspects of the disease were reflected in the fact that survey participants with AH considered 'facilitating the follow-up of patients at different stages of their life by easing the passage from paediatric care to adult care, or from adult care to geriatric care' and 'informing patients about their rights and guiding them toward social services, schools, leisure activities, or vocational

guidance, etc.’, as the fifth and sixth-highest most essential priorities. These findings also correspond with the greater need for social services reported by AH families as compared to the overall survey population. Respondents affected by AH more often expressed that ‘the main hurdles in travelling to a specialised centre are the time needed to get there and/or physical difficulties encountered by the patient (pain, fatigue and injuries)’ as compared to respondents overall. Respondents affected by AH also more frequently expressed the importance of the following statement regarding the implementation of a Centre of Expertise: ‘Rather than concentrating all the expertise and competences in a single, national centre, sharing them between several centres would be preferable because it is more accessible to patients.’

Reactions to Results

Alternating hemiplegia is a severe and very complex disease with paroxysmal and permanent motor sensory and psycho-intellective symptoms. Consequently the required medical and social healthcare services need to be varied, integrated, coordinated and continuous. Each patient needs scheduled routine check-ups (e.g. neurological, neurophysiologic, neuro-ophthalmologic, neuro-rehabilitative, neuropsychological, genetic, metabolic, orthopaedic, specialised technical support). It is clearly more comfortable but also necessary for the coordination of care for all possible explorations to take place in one unique multi-specialist centre.

In general, physicians are not very knowledgeable about this disease. Diagnostic, therapeutic and healthcare protocols do not exist. Each referral is dependent upon the physician’s evaluation. Patients often have to ‘test’ many specialists before finding one who is familiar with the disease and is available to take care of them in an adequate way. All patients have to be accompanied (usually by family members) to seek services.

In general, medical services are more available for AH patients during infancy. As patients get older, the availability of adequate services decreases so much that care for adults with AH is almost non-existent (in particular rehabilitation and the social-educational intervention).

Patients experience rejection often because of a lack of knowledge about the disease, as it is extremely rare. Patients most frequently experience rejection due to a lack of coordination between social and medical services. For example, patients are often rejected by rehabilitation centres (in which the services of physiotherapy, speech therapy, psychological aid, etc., are available) because of numerous and sudden absences due to the frequent and unexpected crises associated with AH, resulting in high-running costs for the centres themselves.

In addition to being scarce in general, social workers often restrict themselves to fulfilling the minimum specific needs of AH patients and often only after many incessant requests. Very often, family members have no choice but to stop professional activity. A better coordination between social and medical service providers would allow social workers to be more knowledgeable and therefore more proactive in suggesting social services to which AH patients have rights.