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REHABILITATION PROBLEMS AFTER ACUTE DISSEMINATED ENCEPHALOMYELITIS: FOUR CASES

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The aim of this study is to describe rehabilitation problems in patients with acute disseminated encephalomyelitis. The study examines retrospective clinical data. Data are reported from 4 patients, who were consecutively admitted and examined with the Functional Independence Measure and magnetic resonance imaging. It was found that the lesions in the brains affected by acute disseminated encephalomyelitis are widespread, but become smaller with time. Motor symptoms dominate at first, but recovery is quite good. Social and cognitive functions are also affected, however, and require a much longer recovery time. These symptoms are, thus, the dominating problem in the rehabilitation ward. At clinical follow-up after 3 years the cognitive problems still influenced the lives of the patients. It is concluded that social and cognitive problems seem to be common in patients with acute disseminated encephalomyelitis. Recovery in these areas takes a long time and this is the main rehabilitation problem, since it affects the capability of the person to reintegrate into society.

Key words: ADEM, MRI, Functional Independence Measure, psychosocial, cognitive

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INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an uncommon condition that affects the central nervous system with demyelization. It is thought to be immune-mediated (1) and most patients have a viral prodrome followed by the development of focal neurological deficits. There are sometimes problems in differentiating between ADEM and multiple sclerosis, although the former is usually monophasic and the latter, by definition, a multiphasic disease (2). Magnetic resonance imaging (MRI) has demonstrated differences between the 2 disorders (3).

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ADEM may develop within a period of days to weeks after a viral illness or vaccination, and a rapid progression to coma or death can occur (4). The long-term prognosis is not well described and focuses on motor deficits (5). Most reports are based on a small number of patients, except that of Miller & Evans (5) who followed 27 patients. However, their study was carried out when the diagnosis was not as exact as it is now. Few publications deal with cognitive and neuropsychological aspects. ADEM can present itself as a psychosis (6). Weir et al. (7) published a case history in which communication difficulties and cognitive deficits were mentioned along with hemiplegia. A more recent report focused on the neuropsychological problems associated with ADEM (8).

To our knowledge, aspects of rehabilitation among these patients have not been described. On the rehabilitation medicine ward, every patient was evaluated within 2 weeks of entry as an in-patient as well as during the rehabilitation period and on discharge from the rehabilitation ward using the Functional Independence Measure (FIM) (8–10). Four patients with diagnosed ADEM, for whom changes in the brain were described with MRI and the rehabilitation period was documented with FIM, were evaluated.

PATIENTS AND METHODS

The 4 patients in this study were seen first at the Department of Neurology, Sahlgrenska University Hospital, Göteborg, Sweden, and were later transferred to the Department of Rehabilitation Medicine at the same hospital. They were consecutively admitted; the first patient was seen in 1991–92, the second in 1994–95, the third in 1995 and the fourth in 1996. ADEM was diagnosed by the development of a central nervous system (CNS) white matter syndrome occurring in close temporal relationship to a systemic disease, which had the clinical characteristics of a viral infection.

The FIM consists of 18 items which can be separated into 2 domains (11), in the present paper called physical (13 items) and social and cognitive (5 items). The FIM assesses self-care, sphincter management, transfer, locomotion (physical items), communication, social interaction and cognition (social and cognitive) using 7-level scales anchored by extreme ratings of total assistance as 1 and complete independence as 7. FIM data were assessed during the first week of the patient's stay (admission) on the rehabilitation ward and at discharge during the last week. Members of the rehabilitation team (attending physician, nurse, occupational therapist and physiotherapist) made the assessment at a short team conference on the basis of their previous observations of the patient on the ward.

MRI examinations of the patients were performed as part of the clinical routine.

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RESULTS

Case 1

Male, 43 years of age. Status post-viral infection. Acute MRI showed multiple, bilateral, asymmetrical lesions with high signal intensity on T2-weighted images and low intensity on T1-weighted images in the deep and periventricular white matter, thalami, basal ganglia and brainstem. Several lesions show gadolinium enhancement as a sign of a disrupted blood-brain barrier.

On follow-up study: the lesions were partly recovered with remaining confluent hyper-intensity in the periventricular white matter.

On admission: tetra-paresis with the most pronounced weakness in the right leg, somnolent. Orientation to person, but not to time or place. The length of stay at the neurological ward was 18 weeks. He was then transferred to the rehabilitation ward but was psychotic and had to be referred to a psychiatric ward. The actual rehabilitation period started after this period (8 months after becoming ill).

On admission to the rehabilitation ward: continued tetraparesis with dominance in the right leg, orientation to person but not to time or place and cognitive problems. The length of stay at the Rehabilitation ward was 22 weeks. On discharge, the summary stated that the patient had improved in his walking capacity but lacked insight into his problems and required someone to look after him.

FIM: physical dimension at admission was 69/84 and at discharge 78/84. Social and cognitive dimension at admission was 24/35 and at discharge 29/35.

Follow-up after 3 years: the patient still had cognitive difficulties with memory problems, sometimes hallucinosis, and he lacked insight into his problems. He was dependent on others to take care of him.

Case 2

Female, 19 years of age. Status post-viral infection. Acute MRI showed multiple, relatively symmetric zones of high signal intensity in the brainstem, thalami, both internal capsules, cerebellar peduncles and right cerebellar hemisphere (Figs 1a and b).

Follow-up after 7 months showed nearly complete recovery with discrete hyper-intensity in the brainstem and thalami (Figs 2a and b).

On admission: unconscious, tetraparetic, need of assisted breathing. The length of stay at the neurological ward was 10 weeks.

When referred to the rehabilitation ward she had double vision, feeling of discomfort, anxiety, dysdiadochokinesis right side, dysmetria, positive Romberg sign with a tendency to fall towards the right. The length of stay at the rehabilitation ward was 6 weeks. On discharge, the summary stated that she showed great improvement physically and could compensate better for balance problems. She was improved cognitively but often experienced feelings of things being unreal and was restless.

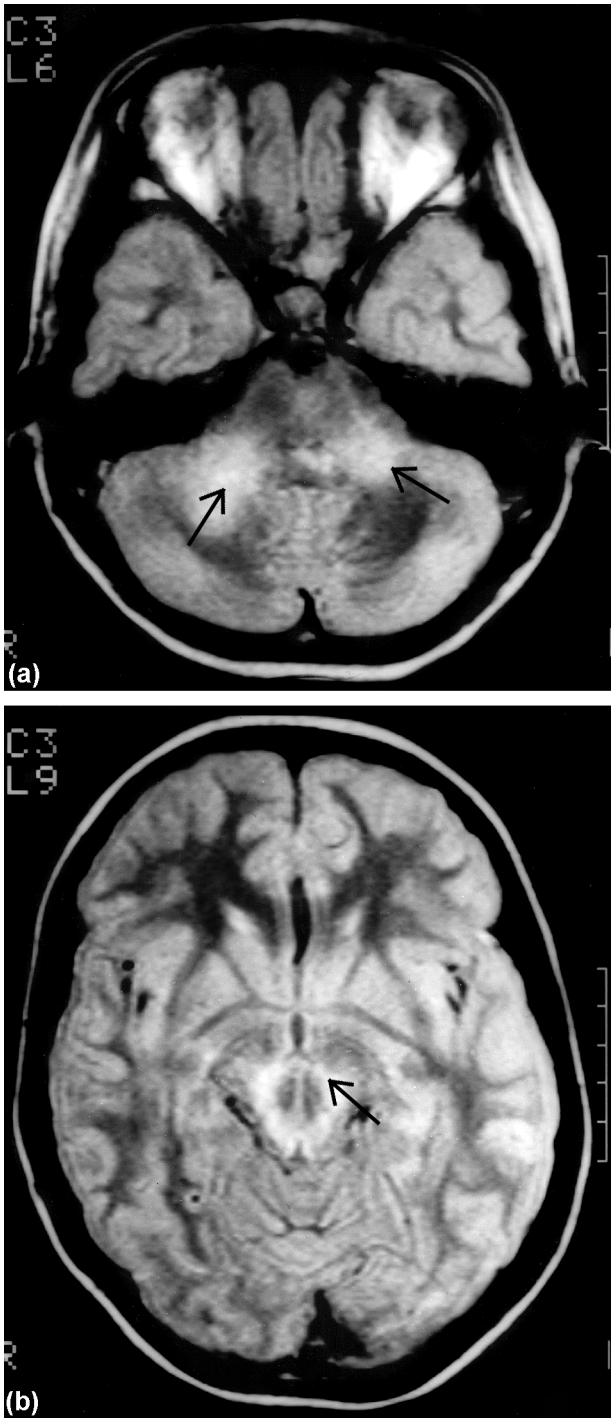

Fig. 1. Young female (case 2). The first magnetic resonance image examination, proton density-weighted obtained at her admission. (a) High signal lesions bilaterally in the middle cerebellar peduncles extending posteriorly on the right side (arrows). Minor changes also in the pons. (b) Similar, symmetric changes also in the mesencephalon.

FIM: physical dimension at admission was 67/84 and at discharge 79/84. Social and cognitive dimension at admission was 16/35 and at discharge 24 of 35 (Fig. 3).

Follow-up after 3 years: she still had some problems with

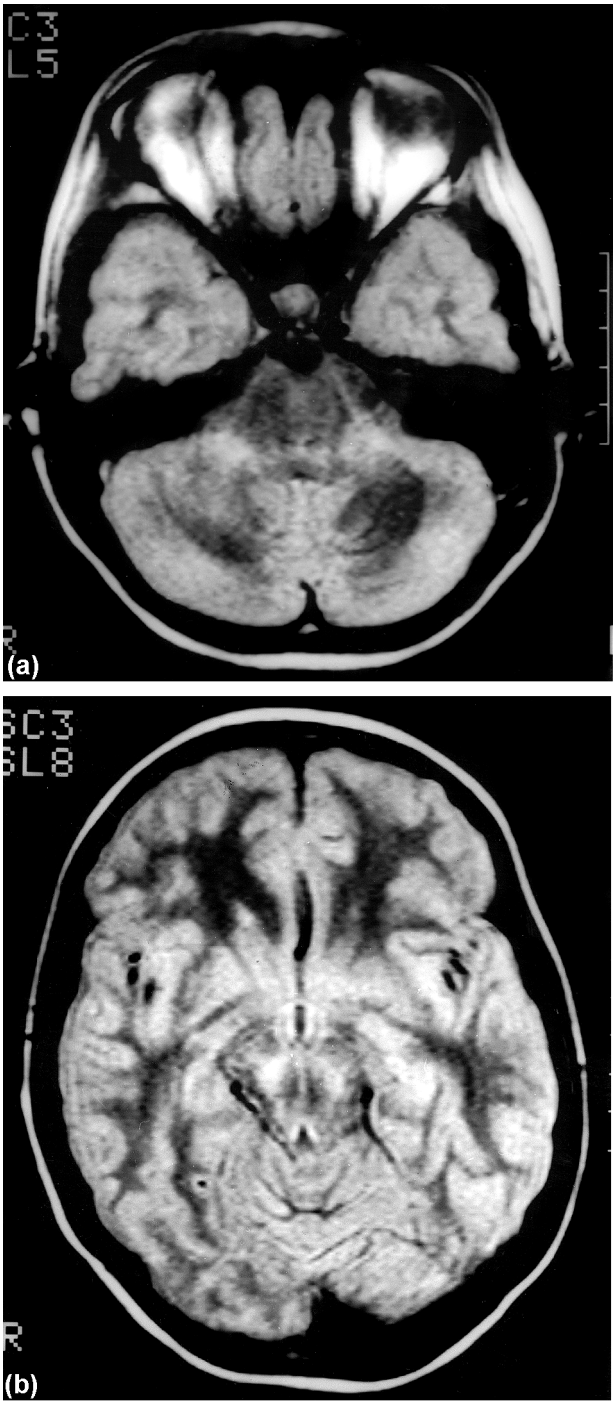

Fig. 2. (a, b) Magnetic resonance image examination of the same patient as in Fig. 1, 1 month later. Lesions are still present, but they show a clear regression when compared with the primary examination.

energy and was more easily tired after intellectual work. She was studying at the university, but had learned how to cope with this tiredness. She has had contact with a clinical psychologist for 2 years in order to learn how to cope with her problems that have become reduced with time.

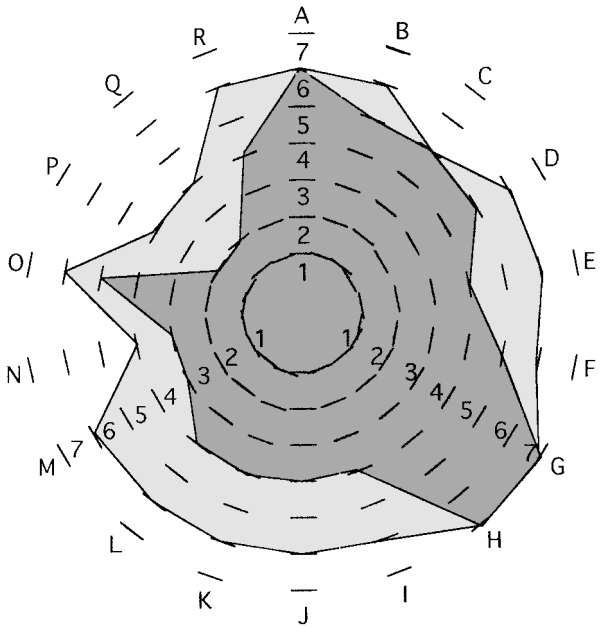

Fig. 3. FIM profile for case 2. The inner dark gray area shows levels of independence on admittance to the rehabilitation ward. The outer lighter area shows the improvement in independence during the stay. There are less motor deficits (A–M) than social and cognitive (N–R) and the patient is more independent in these areas on discharge.

Case 3

Female, 58 years of age. Status post-viral infection. Acute MRI showed a hyper-intense central lesion in the mesencephalon.

Control study 10 days later showed essentially no change.

On admission: vertigo, problems with vision, speech problems. The length of stay at the neurological ward was 5 weeks.

Clinical presentation on admission to the rehabilitation ward: dysarthria, problems with balance and ataxia. The length of stay at the rehabilitation ward was 8 weeks. At discharge, the summary stated that she had improved physically but had a tendency to take risks and overestimate her capacity. Some spatial problems.

FIM: physical dimension at admission was 57/84 and at discharge 78/84. Social and cognitive dimension at admission was 26/35 and at discharge 31/35.

Follow up after 3 years: memory problems, did not take initiatives, reduced capacity in simultaneous activities. Was not capable of going back to work, but had been granted early retirement.

Case 4

Male, 36 years of age. Status post-viral infection. Acute MRI showed extensive hyper-intense lesions in the right hemisphere (mostly white matter), splenium of corpus callosum, left centrum semiovale and dorsal part of the left frontal lobe. Slightly increased signal in the right part of the brainstem and both cerebellar hemispheres (Fig. 4).

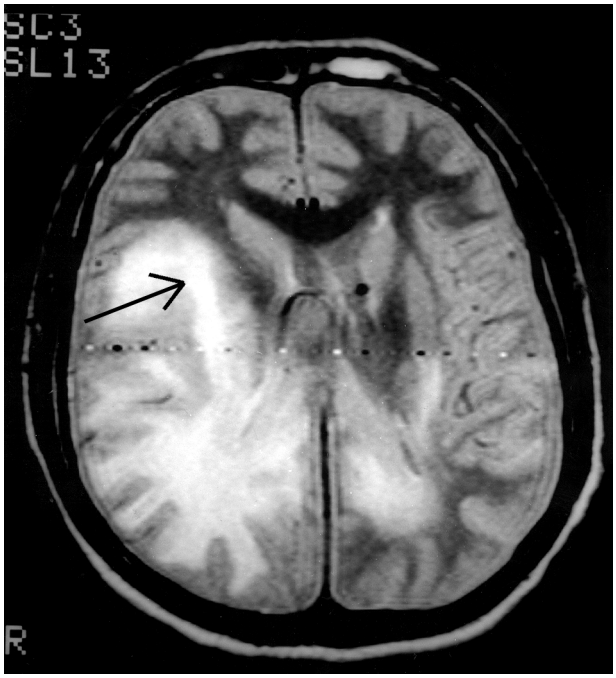

Fig. 4. Male (case 4). The first, proton density-weighted magnetic resonance image examination on admission. Widespread asymmetric high signal changes in the deep white matter (arrow). The right hemisphere is more severely affected.

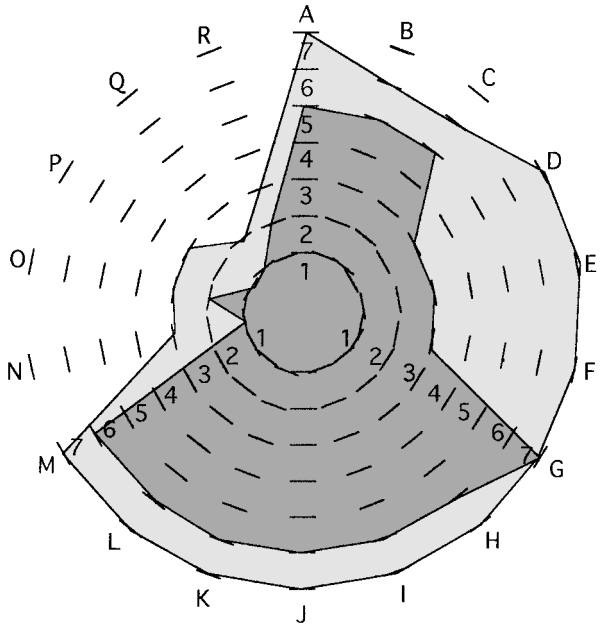

Fig. 6. FIM profile for case 4. The inner dark gray area shows levels of independence on admittance to the rehabilitation ward. The outer lighter area shows the improvement in independence during the stay. There is fast improvement in motor skills (A–M) which is not accompanied by a corresponding improvement in social and cognitive (N–R) items, where the patient is still very much dependent on discharge.

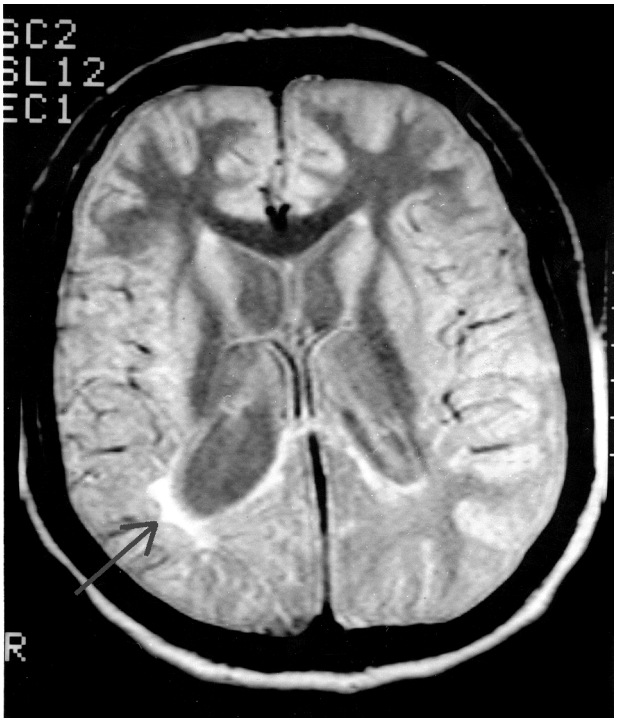

Fig. 5. The same patient (case 4) 2 years later. Most of the parenchymal high signal changes have disappeared. Only minor signal abnormalities along the posterior aspects of the lateral ventricle remain (arrow).

Follow-up: 1 year after the initial study showed evident recovery with remaining lesions in splenium of corpus callosum and both central semiovale, mostly on the right side (Fig. 5).

Clinical presentation on admission: headache, somnolent, left-sided hemiparesis. The length of stay at the neurological ward was 9 weeks.

On admission to the rehabilitation ward: weakness of the left side, neglect. Lack of emotional contact. The length of stay at the rehabilitation ward was 8 weeks. On discharge, the summary stated that he had improved in his hemiparetic side and was an independent walker. He had started to use a schedule with drawings of clocks (he was unable to follow a schedule with numbers). He could not travel on his own since he got lost. Could now find his way around his apartment.

FIM: physical at admission 57/ 84, at discharge 79/84. Social and cognitive dimensions at admission 7/35, and at discharge 14/35 (Fig. 6).

Follow-up after 3 years: he still had cognitive problems. Spatial problems dominated, but also lack of initiative and no real insight into his problems. Lack of emotional contact. Dependent on others to take care of him. Not capable of working, forced into early retirement.

All of the patients showed some improvement in the long-term follow-up. Only 1 of the 4 had improved enough to allow her to study in a normal way, the other 3 were judged not capable of supporting themselves and were granted early retirement.

DISCUSSION

The weaknesses of this study are the small number of patients involoved and its retrospective design. However, patients with ADEM do exist and the limitation in their activities has an impact on their lives and those of their next-of-kin.

ADEM often produces widespread CNS disturbances with coma or drowsiness, seizures and multifocal neurological signs implicating involvement of the brain, spinal cord and/or optic nerve. MRI often shows multifocal involvement of the white matter in the brain. Kesselring et al. (3) followed 12 patients and noted persistent MRI changes for up to 18 months in spite of full recovery. However, what is full recovery? Since there are few notations of cognitive and social evaluations in the reports on ADEM, we do not know whether this has been taken into consideration. What we have found is that, as in many other disorders affecting the brain with diffuse involvement, such as traumatic brain injury (12) or anoxic brain damage (13), any motor or sensory involvement is restored more quickly and is of less importance for the outcome than social and cognitive problems. At a standard neurological examination, these matters are not taken into consideration. To recognize these problems the patient must be observed in his or her environment (for instance the ward). None of our cases was in such condition that a standard neuropsychological examination could be performed. However, the clinical follow-up shows that the cognitive problems are still present after a long period of time. These problems also affect the individuals' ability to function in society with effects on instrumental ADL and on their capacity to support themselves.

Case 1 was psychotic for a very long time, although at a short neuropsychological screening test his memory was normal. After he had stopped being psychotic, he continued to exhibit socially offensive behaviour, e.g. telling everyone the intimate details of his and his wife's sex life, etc. He also had a tendency to undress and walk naked around the ward. Case 2 showed little recognition of her problems. In the beginning, she was confused. At the end of the stay at the ward, her movements were slightly choreatic in her upper extremities, but she was persistent in her determination to return to a summer job in a shop selling china and crystal. Case 3 was ataxic in her movements and was told to use a walker. Nevertheless she climbed down the stairs in her house without requesting the assistance of her carers. She also took the walker outside on a very steep slope after rain. Case 4 was psychotic for some time. When he had become more oriented, cognitive deficits were noted. He could not follow the walls around the gymnasium; he could not complete a set of instructions, (such as "take this paper and fold it and put it on the table") but had to be given the instructions one at a time. It was very difficult to make him shower, and he refused for about 6 weeks to remove his clothing at night. He lacked insight into his own problems.

We have had more than usual problems with these patients to make them understand the consequences of their actions. They lack insight into their limitations. Their recovery was quite remarkable as far as motor skills were concerned. Since their family and the staff at the neurological wards noticed this, and were at first satisfied and pleased, they did not realize initially the eventual problems that arise with limited socials skills and judgement. We believe that since mortality has dropped (it used to be high, around 30%), we are now facing the same situation as for survivors of subarachnoidal bleedings or cardiac arrest, where the medical profession is making progress with saving lives but has not yet started to evaluate the subsequent quality of lives.

ADEM is a demyelinating disease with widespread lesions, where more lesions than before can be demonstrated with MRI. However, MRI, as every other imaging technique, is limited in terms of how small areas can be detected. Widespread small lesions in the cortex and sub-cortical areas may result in altered judgement and social skills. Earlier findings have demonstrated persistent MRI lesions (3), which have been said to be due to pathological changes of astrocytic hyperplasia and gliosis (14). With persistent small scars in the cortical areas, it is easy to understand that there can be persistent problems with higher functions.

CONCLUSION

In conclusion, the needs of a patient with ADEM vary over time. Firstly there is a question of maintaining vital functions, while later there is a dominance of motor problems. For those who survive the first period, recovery of the motor system seems to be quite good. However, there seem to be social and cognitive problems that are usually not identified on neurological wards and that are not explained by the lesions demonstrated by MRI. These problems seem to linger and have not disappeared by the time the patients leave the in-patient rehabilitation ward. It is of the greatest importance to be aware of these problems and identify them, since they will affect the rehabilitation process and its outcome.

Patients with ADEM often have cognitive deficits, which affect rehabilitation.

Cognitive deficits are more persistent than motor deficits and remain for years. The staff must be aware of the possible occurrence of cognitive problems in order to modify the rehabilitation process and goal setting for the patient.

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