

Epidemiology and Management of ALS

ALS is an invariably fatal disease. It is the most frequent motor neurone disorder and the annual incidence in most countries ranges from 1 to 2 per 100,000 inhabitants. The disease is considered to be more frequent in men than women and usually it starts by pareses and atrophy of one or more extremities often in association with the presence of fasciculations representing involvement of the lower motor neurone. Very early in the course of the disease a generalised lower motor neurone abnormality may be detected by EMG and not clinically. The term Progressive bulbar palsy or "bulbar ALS" refers to pareses starting as speech or swallowing difficulties and occurs in 10-40% of all ALS cases. It is more frequent in women and in late onset cases. Generally the prognosis in bulbar cases has been considered worse than in spinal cases, but this may have changed with increasing use of non-invasive assisted ventilation.

Increasing incidence of ALS?

During the last few years convincing evidence has been presented that the incidence of ALS has been increasing over the last decades.^{1,2} Initially these data were interpreted as being due to the generally increasing age of the population as a whole, but this seems unlikely to be the whole explanation and indeed environmental factors may be contributing to this observed increase in ALS incidence. We have also reported that not only is there an increase in the annual incidence of ALS from 1.5 to 2.5 per 100,000 of the population, but there has been a change in gender affected by the disease during the period from 1960 to the mid nineties such that by the end of the observation period the risk was similar in men and women³ (see Fig. 1). This could indicate that environmental factors contributed to the increased incidence and in this respect exposure to heavy metals and electric magnetic fields has been associated with the disease.⁴ Moreover, studies of Gulf war veterans have indicated increased risk in veterans, especially in air-force and army personnel.⁵ Overall it can be concluded that there is increasing incidence of ALS and whilst this is in part explained by the increasing age of the general population, it may also be related to environmental factors that affect the integrity of motor neurons.

Care of the ALS patient

Although great progress has been made to better understand the pathophysiology of ALS, especially in genetic variants of the disease, the possibility of treatments that truly affect survival-time is still very limited. The glutamate antagonist Riluzole has been shown to prolong survival by an average of 3 months, and whilst great efforts have been made to study molecules affecting glutamate metabolism and trophic factors (inc. positive reports in transgenic mouse models of ALS), no other treatments than riluzole have been demonstrated to prolong survival in human ALS. In contrast to the paucity of disease modifying treatments in ALS, during the last decade there has been a substantial change in the management of the disease.

In the past, ALS patients were carefully diagnosed, but offered little support to manage their life as they developed increasing pareses. Most frequently the patients were hospitalised when the disease had progressed to a stage where the patient or family could not manage, possibly with the insertion of naso-gastric tubes in patients with bulbar palsy and swallowing difficulties. Today it is accepted that ALS patients are best treated in multidisciplinary ALS clinics with a team that includes a neurolo-

gist, specialised nurses, physical, occupational and speech therapists, social worker as well as pulmonologist and nutritionist. It has been demonstrated that such care is not only good for the well-being of the patient, but in fact increases survival at least as much as with the administration of riluzole.⁶

The ALS care at Haukeland University Hospital in Norway was organised in such a way so that it has had an ALS clinic from 1990, inspired from that which exists in the Department of Neurology at the University of Chicago (see Fig. 2). The diagnosis of ALS is not usually established in the out-patient clinic but during a stay at the Department of Neurology. This makes it easier to have all the necessary investigations performed before the diagnostic discussion is undertaken with the patient. Usually within a month the patient is seen in the multi-disciplinary ALS clinic, and a week before this consultation the patient receives a description of the clinic and the professions they may meet there. The patients will usually see most professions on their first consultation but later only when specific problems arise and need sorting out. Usually the first consultation in the clinic is used to ascertain the diagnosis and to inform the patients about the possible health outlook with this condition. The patients are given a card for the ALS nurse with all the possibilities of how to get in touch with the clinic as and when they need to do this. The ALS nurse is the primary contact point for the patient and will transfer requisites to actual professions, again as and when required. One of the most important aspects of management in the ALS clinic is to take the initiative for ensuring that the primary health care system engages and supports the patient in the community. As the disease is rare, most community based services have no experience on the management of ALS and so the resource group with the general practitioner, physical and occupational therapist and nurse need to be organised in the community. This group has regular meetings with the patient and his family and can at short notice, supply the patient with aids required for them to remain independent.

As the disease progresses the patients will need more sophisticated aids. This is organised through the ALS clinic, but the patients are frequently admitted for short periods to the ward in the Department. This could be for establishing PEG feeding or evaluation with regard to them developing respiratory failure. Usually establishment of PEG requires a 2-day stay in the ward and is established in collaboration with the gastroenterology section of the Department of Internal Medicine. After insertion of the PEG, the patient may stay another day in the Neurology ward to learn how to use the PEG and to

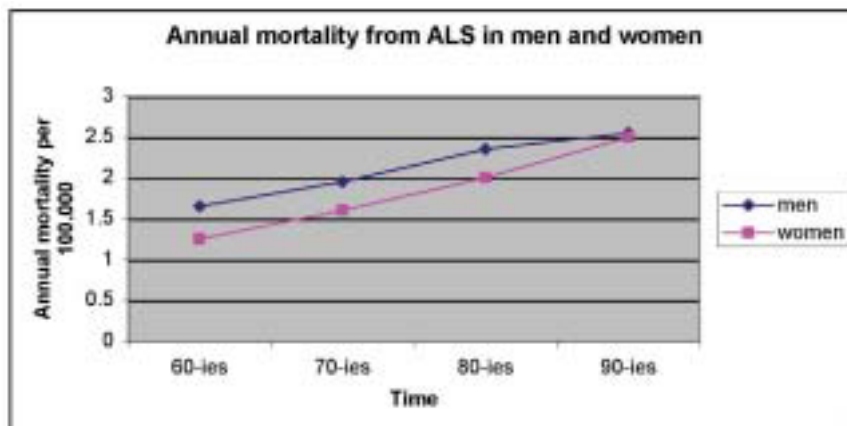


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Figure 1



start to get used to the nutrition given through it. After 3 months the tube is changed to a Mic which is usually easier to handle as the tube can be troublesome for the patient (especially during night/sleep). Some patients have problems with drooling. These are treated either by one shot radiotherapy to the parotid and submandibular glands done in collaboration with the Department of Oncology. More recently we have also tried Botulinum toxin A injections to the parotid glands with some success. From a respiratory perspective, an increasing number of patients are offered non-invasive ventilation support when required. The evaluation of respiratory function is performed by pulmologists and during this evaluation the patients may have a short stay in the Department of Pulmology. Follow up of the BIPAP (Bilevel Positive Airway Pressure) is also done by the pulmologist associated with the ALS clinic. We thought that the more abundant use of BIPAP would lead to an increasing number of ALS patients deciding to have tracheostomy and permanent ventilation support, but this has not proven to be the case. Most patients decide not to continue life when the disease has progressed to a stage where they are totally dependent on family and care-givers for all their activities.

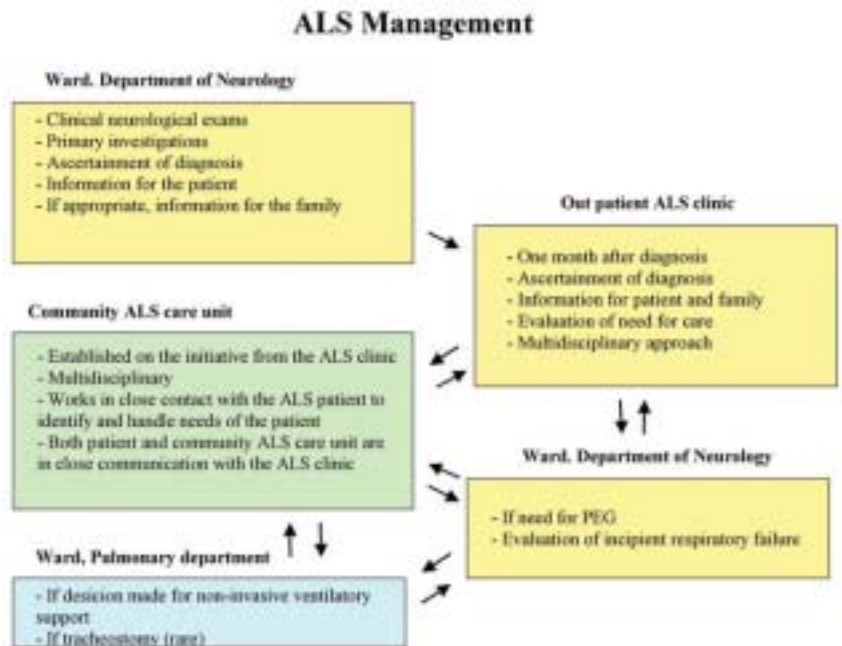
Conclusion

The incidence of ALS is increasing due in part to the increasing age of the general population together with an unknown factor that may be environmental. During the last decade there has been little development in the medical treatment of this disease but management has considerably changed from a passive attitude to a fatal disease to considerable work to help the ALS patient to live with his symptoms. Crucial for success in ALS management is a multidisciplinary approach tailored to the needs the patient as they develop and change during the course of the disease. Needs must be identified early and handled quickly. This requires close communication between an ALS clinic with the Neurological ward, the primary care unit and the Pulmonary Department.

References

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5. Horner RD, et al. *Occurrence of amyotrophic lateral sclerosis among Gulf War veterans.* Neurology 2003;61(6):742-9.
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Figure 2



Challenges in ALS Management

On time of diagnosis

- To have a good relationship with the patient
- Realistic information but not without hope
- Not all information is needed at once
- Make sure that the patient has a place to make contact if needed

Community ALS care unit

- Organise multidisciplinary meetings in the home of the patient
- Fulfill needs within a short period of time (days)
- The psychological stress of the close relation to an invalided ALS patient
- Take care of family members

At follow ups in the ALS clinic

- Information on current research in ALS
- Some patients want alternative treatments
- Deal with peruses and make aid items (walking aids, wheelchair, requirements to sit or for hygiene) available (occupational therapist)
- Deal with communication problems (speech therapist)
- Avoid delay in achievement of aid facilities
- Have options for the patient to make contact (telephone, E-mail)
- Short waiting time if consultation is needed
- Good organisation if PEG is needed
- Discussion of end-of-life issues. Use all experience to inform on expected life situation in case of permanent assisted ventilation.

2005 COURSES AND CONFERENCE PROGRAMME

11 Jan	SRR Winter Conference	£80/50	27 May	Psychological formulation of emotional consequences	£110
	Understanding Brain Injury	£30/50	15 June	Acquired Dyslexia	£110
	25 Feb 22 April 22 July 25 Nov		16 Sept	Behavioural experiments in the rehabilitation of acquired brain injury	£110
11 Mar	Introduction to Cognitive Behavioural Therapy in the Rehab of acquired brain injury	£110	20/21 Oct	Attention and executive skills	£200
7/8 April	Introduction to Neuropsychological Rehabilitation	£200	9 Dec	Fatigue after Brain Injury	£110

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