PALLIATIVE CARE IN ALS

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EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis….

European Federation of Neurological Societies

- Patients with symptoms suggestive of ALS should be assessed as soon as possible by an experienced neurologist.
- Early diagnosis should be pursued.
- The patient should be informed of the diagnosis by a consultant with a good knowledge of the patient and the disease.
- Following the diagnosis, the patient and relatives/carers should receive regular support from a multidisciplinary team.
- Medication with riluzole should be initiated as early as possible.
- Control of symptoms such as sialorrhea, thick mucus, emotional lability, cramps, spasticity and pain should be attempted.
- Gastrostomy tubes should be placed before respiratory insufficiency develops.
- Non-invasive positive-pressure ventilation also improves quality of life.
- Maintaining the patient's ability to communicate is essential.
- During the entire course of the disease, every effort should be made to maintain patient autonomy.
- Advance directive for palliative end-of-life care should be discussed early with the patient and carers.

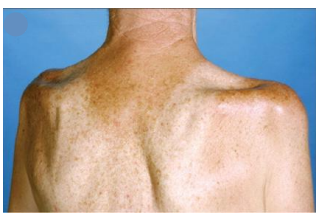
European Journal of Neurology 2012, 19: 360–375

AMYOTROPHIC LATERAL SCLEROSIS

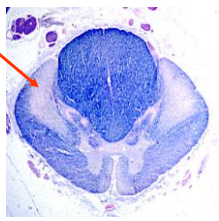
Amyotrophy = muscle atrophy

Lateral = laterally in the spinal cord

Sclerosis = scar like lesion



1869 – Jean-Martin Charcot, French neurologist, first describes and publishes his findings on ALS

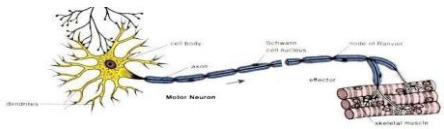


ALS SYMPTOMS

ALS often starts focally with muscle weakness and wasting:

- Foot/leg/hand/arm (limb onset) - about 70%
- Mouth, throat (bulbar onset) - about 25%
- Trunc/respiration (thoracic onset) - around 5 %

The muscle weakness gradually spreads to surrounding areas.
Finally, respiratory failure will occur.



ALS is a motor neurone disease.

It affects neurones to striated (volontary) muscles.

Smooth (non-voluntary) muscles + cardiac muscles are not involved:

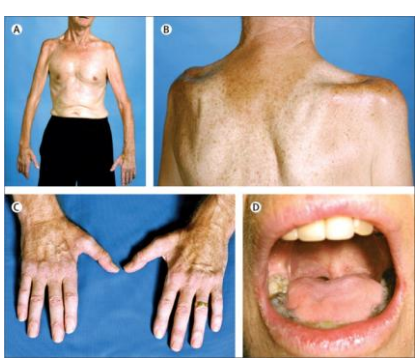
- heart, urinary bladder, gastro-intestinal tract

Oculomotor and sphincter functions are however spared.

Cognitive dysfunction is seen in up to 50% (3-5 % develop dementia, often of the frontotemporal type).

SYMPTOMS:

- progressive weakness
- cramps
- …twitchings (fasciculations)

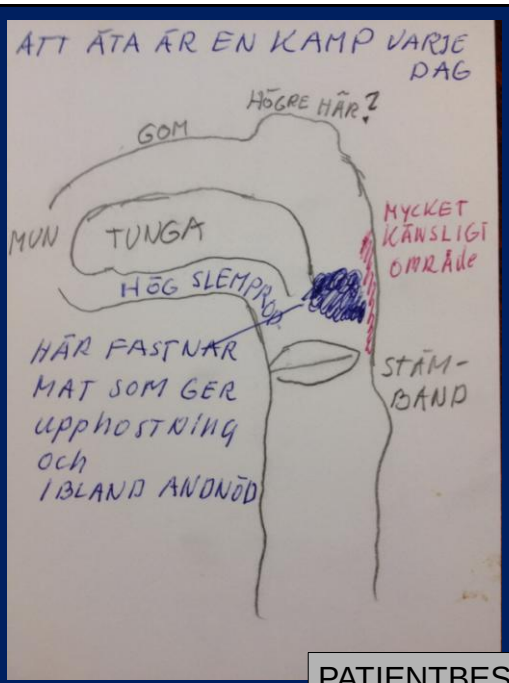


NEUROLOGIC EXAMINATION:

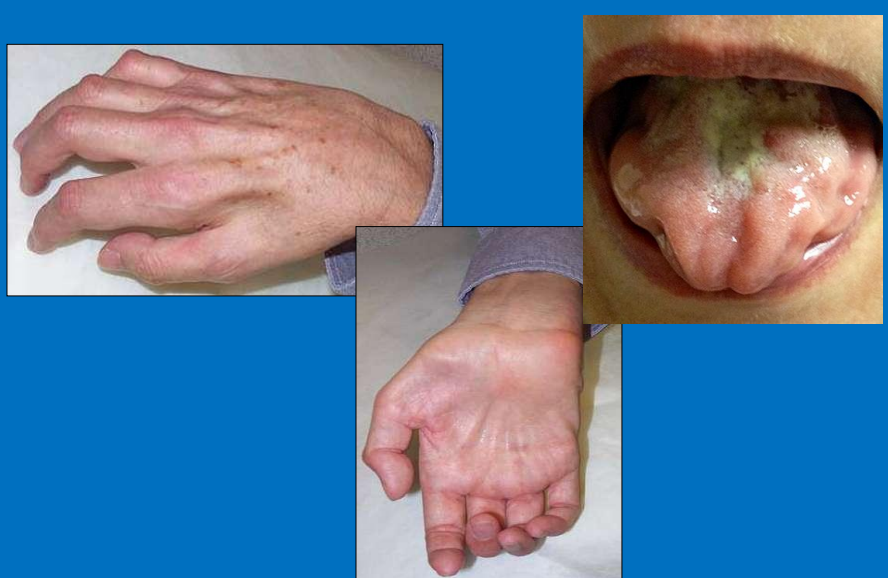
- weakness
- muscle atrophies
- fasciculationer
- brisk/absent tendon reflexes

NEUROLOGIC INVESTIGATION

- history
- examination
- neurophysiology (EMG/NCV)
- MR
- cerebrospinal fluid analysis



PATIENTBESKRIVNING

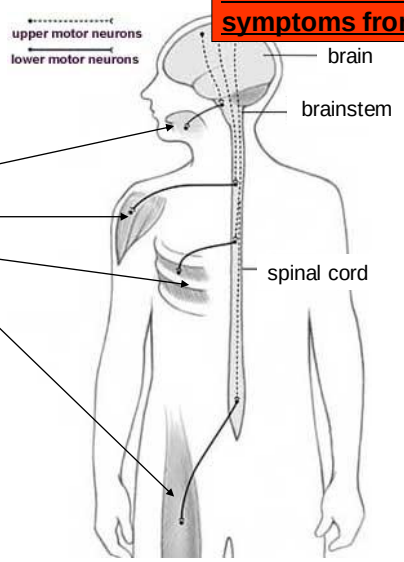


MUSCLE ATROPHY IN ALS

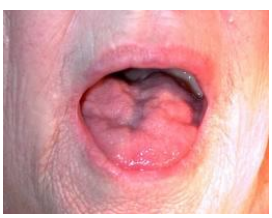
A definitive diagnosis requires symptoms from at least 3 areas

4 areas:

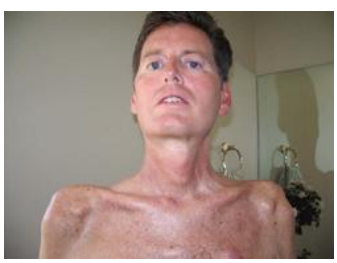
- Bulbar
- Cervical
- Thorcic
- Lumbosacral

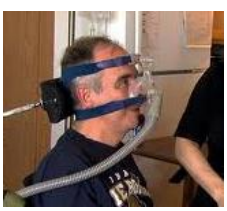


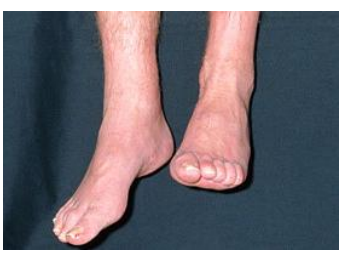
BULBAR



CERVICAL

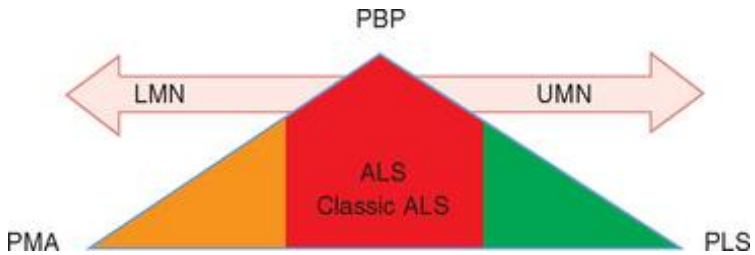






THORACIC

LUMBAR

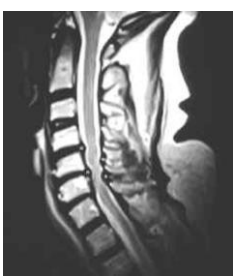


ALS = involvement of both upper and lower motor neurons

PLS (primary lateral sclerosis) = involvement of upper motor neurons

PSMA (progressive spinal muscle atrophy) = involvement of lower motor neurons

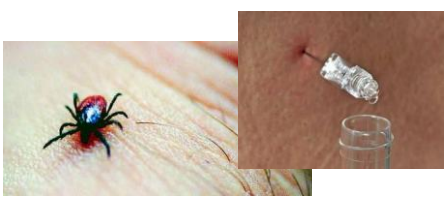
PBP = progressive bulbar palsy

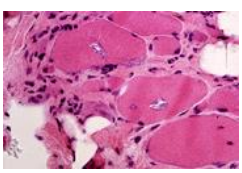


Differential diagnoses:

Ex:

- spinal cord lesion (eg cervical stenosis)
- multifocal motoric neuropathy (MMN)
- Kennedy´s syndrom (spinobulbar muscle atrophy)
- myasthenia gravis
- myopathy (inclusion body myositis)
- neuroborreliosis





How to communicate the diagnosis…..??

1 The diagnosis should be communicated by a consultant with a good knowledge of the patient.

2 The physician should start the consultation by asking what the patient already knows or suspects.

3 The diagnosis should be given in person, ensuring enough time for discussion (suggest at least 45–60 min). Provide printed materials about the disease, about support and advocacy organizations and informative websites.

4 Assure patients that they will not be 'abandoned' by healthcare services and will be supported by a professional ALS care team (where available), with regular follow-up visits to a neurologist. Make arrangements for a first follow-up visit, ideally within 2–4 weeks.

5 Avoid the following: withholding the diagnosis, providing insufficient information, imposing unwanted information, delivering information callously, taking away or not providing hope .

EFNS GUIDELINES

WHO GETS ALS ???

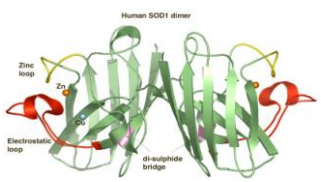
Richard Olney, renowned ALS researcher, dies at 64 of the disease

Incidence = around 2 /100,000 (fairly uniform in Europe).

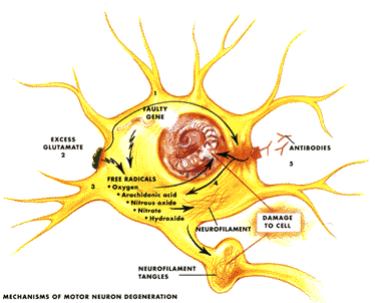
Peak age at onset for sporadic ALS = 58-63 years.
For familial ALS = 47-52 years.

Male:female = 1.6:1.

Familial form (FALS) around 5-10 %.

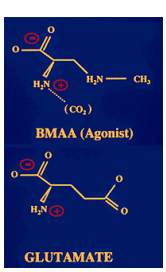


CAUSES OF ALS ?



It´s still unknown why the motorneurons die in ALS.

- excitatotory aminoacids (glutamate)?
- free radicals?
- lack of grow factors (ex VEGF, IGF-1)?
- virusinfection?
- toxins?
- cyanobacteria/BMAA…..?
- too much physical activities?
- genetics….



PHYSICAL ACTIVITES??

Prognosis?

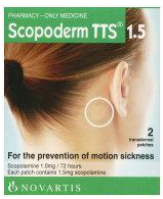
- 50 % live at least three or more years after diagnosis.
- About 20% survive between 5 years and 10 years.
- Older age, bulbar onset, early respirator muscle dysfunction are associated with reduced survival.
- Early weight loss is an indicator of a poor prognosis.
- Patients with predominantly lower/upper motor neuron forms have a better prognosis.

TREATMENT

DISEASE SPECIFIC MEDICATION

Riluzole 50mg 1x2 (glutamate antagonist)
= not a cure, but may prolong survival

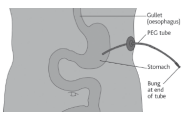
SYMPTOMATIC TREATMENT



Cramps/spasticity
- physical therapy
- diazepam, baclofen

Sialorrhea (drooling)
- anticholinergicum, – dermal scopolamine patch
- amitriptyline

Bronchial secretion
- N-acetylcysteine
- physical therapy
- portable home suction device
- cough machine



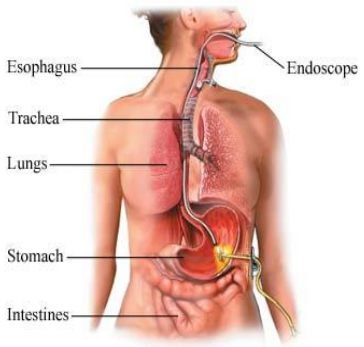
Dysphagia
- dietary counseling
- gastrostomy

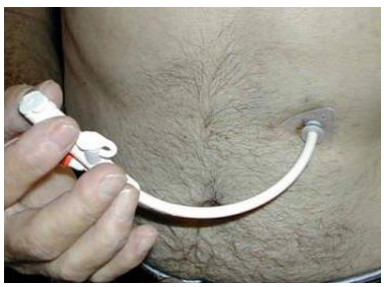
Dysarthria
- speech therapist
- communicator (Ipad?)

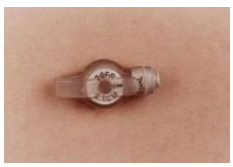
Pseudobulbar emotional lability, depression
- antidepressant, e.g. citalopram 10-20mgx1
- *Nuedexta has a beneficial effect on all bulbar symptoms in addition to pseudobulbar affect*,

Insomnia
- t ex zopiclone, zolpidem, amitriptyline….

Constipation
- dietary advice
- ….







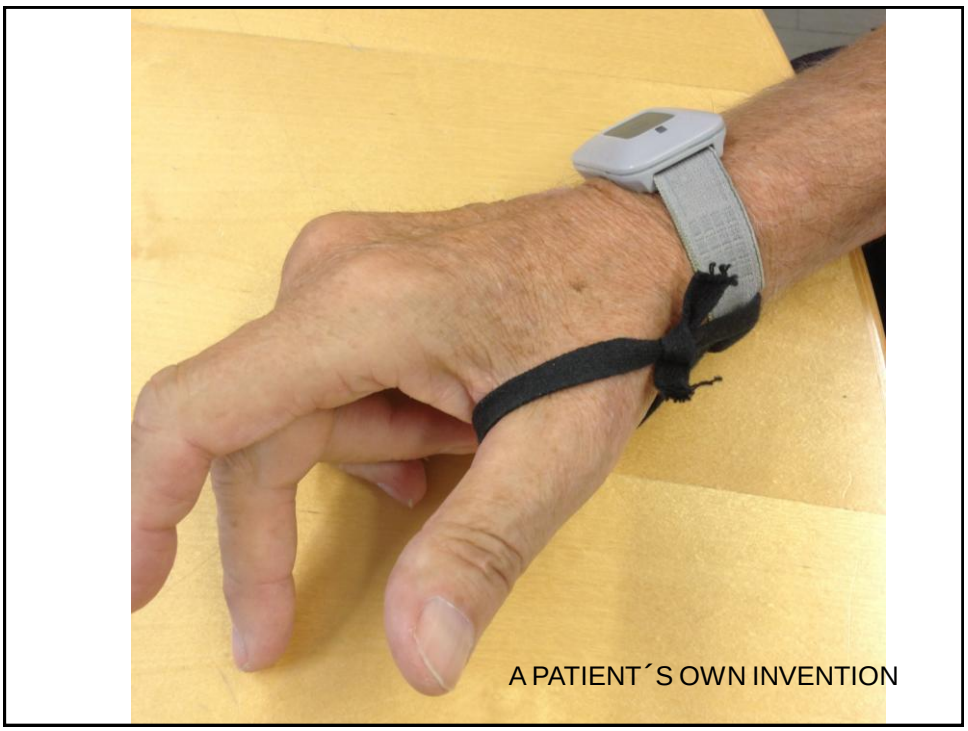
Percutaneous Endoscopic Gastrostomy
PEG

Weight loss/malnutrition in ALS :

- Secondary to reduced food intake

- About 50 % of ALS patients have a hypermetabolic state.
  This is associated with reduced survival.
  (Dysfunction of mitochondria?)

Insertion of a gastrostomy tube should therfore be offered to patients with substantial weight loss, even in the absence of dysphagia


A PATIENT´S OWN INVENTION

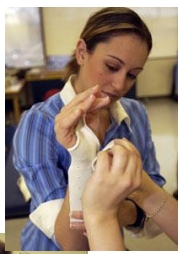

ABC
DEF
GHI
JKL
NO
PQR
STU
VWX
YZ
SPACE
I H G F E D C B A
K
M
O
R
JA Ö Ä Å X Y V U T NEJ

Dyspnea

- non-invasive ventilation
- invasive ventilation/tracheostomy ..???
- oxygen therapy alone should be avoided
- morphine

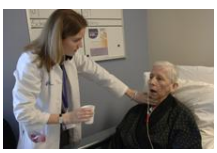
Anxiety

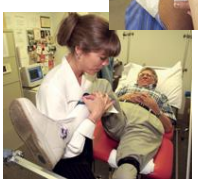
- benzodiazepines
- morphine



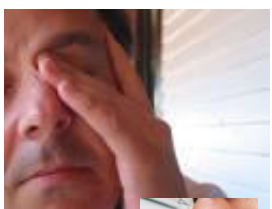
MND-TEAM

- occupational therapist
- physiotherapist
- dietitian
- social worker
- speech therapist
- neurologist
- nurse

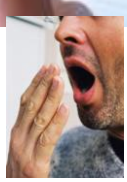




Symptoms/signs of respiratory insufficiency:



- Dyspnea on minor exertion or talking
- Orthopnea
- Daytime fatigue
- Frequent yawnings, daytime sighings
- Morning headache
- Frequent nocturnal awakenings

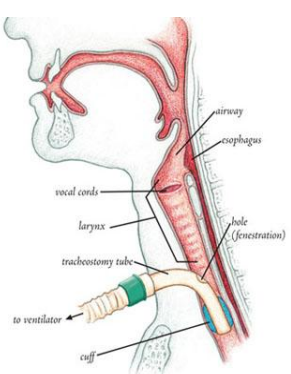


- Tachypnea
- Use of auxilliary muscles
- Weak cough
- Sweating
- Weight loss
- Morning confusion, hallucinations

Non-invasive positive-pressure ventilation

Invasive ventilation….??

For whom ???



INVASIVE MECHANICAL VENTILATION

Advantages:

- Increases survival time
- Prevents aspiration
- Ability to provide more effective ventilator pressures and better gas exchange

Drawbacks:

- Generates more bronchial secretions
- Increases risk of infection
- Introduces risk of tracheo-oesophageal fistula, tracheal stenosis or tracheomalacia
- Greatly increased costs
- Increased family and carer burden, including 24-h nursing requirement

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QUALTY OF LIFE?
The patient has a right to have the invasive ventilation turned off (in Sweden).
But…..has to be able to communicate this.

WHEN SHOULD A PALLIATIVE CARE TEAM BE INVOLVED ??