



:: Amyotrophic lateral sclerosis



- This document is a translation of the French recommendations drafted by Prof. Vincent Meininger, Dr, Pierre-François Pradat, and Dr Philippe Corcia, reviewed and published by Orphanet in 2007.
- Some of the procedures mentioned, particularly drug treatments, may not be validated in the country where you practice.

Synonyms:

Charcot's disease, ALS, Lou Gherig's disease, Motor neuron disease

Definition:

ALS is the commonest motor neuron disease of adults. It is linked to the progressive death of motor neurones involving both central (which run from the cortex to the spinal cord or cerebral trunk) and peripheral neurones (from the spinal cord or cerebral trunk to the muscles).

Further information:

[See the Orphanet abstract](#)

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Pre-hospital emergency care recommendations

Call for a patient suffering from amyotrophic lateral sclerosis

Synonyms

- ▶ ALS (Amyotrophic Lateral Sclerosis), Charcot's disease, Lou Gherig's disease, Motor neuron disease

Aetiology

- ▶ progressive destruction of central and peripheral motor neurones

Special risks in an emergency

- ▶ respiratory distress (respiratory muscles affected; infectious broncho-pneumonia, pulmonary embolism, atelectasis)
- ▶ choking (swallowing disorders)
- ▶ severe GIT passage problems (blockage)
- ▶ problems related to the gastrostomy tube

Frequently used long term treatments

- ▶ riluzole (neuroprotector)

Complications



- GIT passage problems aggravating respiratory problems (abdominal distension)
- any intubation decision must be made with the approval of the patient or his/her immediate family (written or clear instructions).

Specific medical care prior to hospitalisation

- ▶ special medical transport where necessary in half-sitting position with oxygen or NIV (non-invasive ventilation) depending on the respiratory status
- ▶ suction of bronchial fluids
- ▶ aerosols, respiratory physiotherapy, broad spectrum antibiotics
- ▶ scopolamine SC or beta blocker for excess secretions
- ▶ send the patient to a specialised centre once stabilised

Recommendations for hospital emergency departments

Emergency situations

1. Respiratory problems

Respiratory involvement is usually the direct result of the respiratory muscles being affected by the condition but the possibility of a linked triggering cause must be considered every time: pulmonary embolism, broncho-pneumonia, bronchial obstruction, atelectasis.

▶ Emergency diagnostic measures

■ Emergency respiratory evaluation:

- transcutaneous oxygen saturation.
- arterial blood gases (to be done as soon as possible)
- thoracic radiograph (to be done as soon as possible)
- measuring the vital capacity is important in emergency care.

■ Look for indicators of severity:

- supra-sternal indrawing
- paradoxical respiration
- cyanosis
- signs of hypercapnoea (consciousness disorder, sweating, headaches).

■ Diagnosing the trigger factor:

- D-dimer assay
- thoracic scan (which may be impossible if serious orthopnoea)
- ultrasound of lower limbs

▶ Immediate therapeutic measures

■ In cases of acute respiratory insufficiency:

- Place the patient in a **half-sitting position**
- **Reduce bronchial obstruction** related to swallowing or salivary secretions
- **Treat the trigger factor:** swallowing down the wrong way, pulmonary embolism, bronchial secondary infection
- If nasal oxygen therapy has been started, **adjust the oxygen flow rate based on the pCO₂** using arterial blood gases and saturation assays.
- Consider using **non-invasive ventilation if it is possible**
- Use small doses of morphine to regulate the respiratory rhythm if necessary
- If the situation is serious, **any intubation decision must be made with the approval of the patient or his/her immediate family** (written clear instructions).

Criteria for using a ventilator with ALS [Consensus Conference, 2005]

Clinical criteria	Symptoms of nocturnal or diurnal alveolar hypoventilation: dyspnoea, orthopnoea, sleep disturbance, snoring, nocturnal apnoea, sudden wakening with feeling of suffocation, diurnal sleepiness, morning headaches, unexplained cognitive deterioration
Paraclinical criteria	One of the following: <ul style="list-style-type: none">▶ paCO₂ > 45 mmHg▶ CV < 50 % or maximum PI and SNIP < 60 %▶ anocturnal oxymetric anomalies<ul style="list-style-type: none">■ SpO₂ < 90 % for more than 5 % of nocturnal recording time when there is no obvious obstructive sleep apnoea syndrome■ and/or SpO₂ < 89 % for longer than 5 minutes

■ For bronchial obstruction:

- **Suction for bronchial secretions**
- Use **broad spectrum antibiotics**
- Start providing expectoration assistance using appropriate physiotherapy (assisted coughing)
- Use aerosol sessions and treat the swallowing problems
- Scopolamine sub-cutaneous in small doses (¼ - ½ ampoule SC tid or qid) which can help dry out the secretions.
- Beta blockers when the bronchial secretions are thick and when no contra-indications are present with a dose rate adjusted for the cardiac resistance.

2. Swallowing problems

▶ Immediate therapeutic measures

■ For swallowing down the wrong way:

- Free upper airways and use the **Heimlich technique in the acute phase**
- **Reassure the patient and his/her family**
- **Oxygen** therapy
- Administer **5mg morphine SC if there is anxiety**
- Then thoracic radiograph and broad spectrum antibiotics if there has been inhalation

■ If oral alimentation is impossible:

- **Keep well hydrated by SC or IV fluids** based on the clinical status and blood electrolytogram results
- **Nasogastric tube** as a temporary solution **before possibly inserting a gastrostomy catheter**. Avoid providing food too quickly (less than 500 cc per 24h during the first two days)

3. Problems related to the gastrostomy catheter

▶ Immediate therapeutic measures

■ If there is inflammation around the gastrostomy tube entry:

- **Local antiseptics**
- Parenteral antibiotics if there are systemic infection signs

- Surgical drainage if there is parietal collection
- **Blocked gastrostomy tube:**
 - Gently knead the tube to break up the blockage
 - Use a large sized syringe to inject water into the tube
 - If this fails, inject 2 ml hydrogen peroxide into the tube. If the blockage is not quickly removed, leave it to act for 30 minutes then try to unblock it again.
 - Change the tube, as a last resort.
- **Gastrostomy tube removed:**
This is an emergency as the stoma orifice will close in a few hours
 - **Insert a urinary catheter into the gastrostomy orifice without inflating the balloon**, attach it and **contact the gastroenterology service as quickly as possible.**

4. GIT passage problems

GIT passage problems can be the cause of a significant deterioration of the general status because abdominal distension can limit diaphragmatic movement and heighten respiratory problems.

- ▶ **Immediate therapeutic measures**
 - osmotic drugs, mild laxatives or enemas
 - surgical review in some cases.

Drug interactions

- ▶ No contra-indication or special precaution related to the use of riluzole.

Anaesthesia

- ▶ Conduct an **initial respiratory evaluation** (blood gases, vital capacity) in all cases.
- ▶ **Avoid curare-related products** and respiratory depressors.
- ▶ **Prefer local/regional anaesthesia.**
- ▶ If respiratory insufficiency is present, **partial general anaesthesia without intubation** (neuroleptanalgesia) **involves a risk of respiratory degradation** during or following the intervention.
- ▶ **The decision to intubate in an emergency must be discussed with the patient and his/her family.**
- ▶ An intervention under general anaesthetic with intubation in patient with respiratory insufficiency runs the **risk of not being able to extubate** or turn the ventilator off. **This possibility must be mentioned to the patient and the family.**

Additional actions and hospitalisation

- ▶ **Adjust the nurse call system** to the patient's verbal and motor capabilities (e.g. tactile or muscular contacts linked to a ball).
- ▶ **Use the patient's equipment** (vocal synthesisers, tables of letters or pictograms...)
- ▶ If there are **deglutition problems**
 - Deglutition advice must be given to the whole of the care team **to avoid choking**: sitting position, back as straight as possible, swallowing with the head bent forward with the chin touching the sternum.
 - **Adapting the food**: enrichments, texture changes, nutritional additives, thickened liquids, jellified water
- ▶ Maintain a regular GIT passage to **avoid constipation.**

Organ donation



Organ donation is **not contra-indicated** by this condition.

Documentary resources

- ▶ Verdon R, Dargère S. Complications infectieuses de la gastrostomie percutanée endoscopique. *Nutri Clin métabol* 2000 ;14 : 149-152.
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