

## King's ALS Staging SOP

***This procedure takes the form of a semi-structured interview. It is hierarchical and is not therefore a functional scale. In all cases, if involvement of a region is obvious to an untrained observer it should be counted as involved. Only findings related to ALS should be scored.***

1. Stage 4 has been reached if there is explicit evidence of **feeding failure secondary to ALS** (defined as **weight loss 10% or more** of premorbid weight in the context of swallowing difficulty, or there is a **stated recommendation of gastrostomy** by the multidisciplinary team) or **respiratory failure secondary to ALS** (which **clearly meets the definition** according to the NICE NIV Guideline, summarized below in notes parts A and B). If gastrostomy or NIV are recommended but the intervention is *refused* by the patient, Stage 4 has still been reached.

2. To assess earlier stages the El Escorial regions are considered. The number of regions involved defines the stage. Abnormal reflexes may suggest involvement of a region (relevant reflexes are detailed below with each region subheading).

### *Bulbar:*

Involvement defined as slurred speech, dysphonia, problems swallowing liquids, choking. Examination showing tongue atrophy, fasciculation, slowness of movement or a pathologically brisk jaw jerk reflex is acceptable as an alternative if no symptoms are reported.

### *Upper limbs:*

Involvement defined as trouble with keys, doors, buttons, zips or carrying bags, reported by the patient. Examination showing wasting of the first dorsal interosseus, pectoral reflexes or Hoffman's sign is acceptable as an alternative if no symptoms are reported. (A clinician may examine any reflexes and draw their own conclusion).

### *Lower limbs:*

Involvement defined as stiffness, spasm or cramping, falls, or the foot catching on walking. Examination showing gait stiffness or foot drop, crossed adductor reflexes, pathologically brisk patellar reflexes or ankle clonus is acceptable as an alternative if no symptoms are reported. (A clinician may examine any reflexes and draw their own conclusion). Extensor plantar responses are *not* acceptable as suggesting of involvement.

## **Notes**

### *A. Definition of respiratory failure*

SNP:

In the absence of symptoms, respiratory failure is defined as sniff nasal inspiratory pressure (SNP) <40cm H<sub>2</sub>O or decrease > 10cm H<sub>2</sub>O per 3 months. If there are symptoms of orthopnoea or excessive daytime sleepiness, respiratory failure is defined as SNP <65cm H<sub>2</sub>O for men and 55cm H<sub>2</sub>O for women.

### *B. Other acceptable definitions of respiratory failure*

FVC:

In the absence of symptoms, respiratory failure is defined as FVC <50% predicted. If there are symptoms of orthopnoea or excessive daytime sleepiness, respiratory failure is defined as FVC <80% predicted.

Oximetry measures:

Respiratory failure is defined as SpO<sub>2</sub> at 94% or lower, with either pCO<sub>2</sub> >6kPa, or overnight oximetry showing >5 dips per hour to SpO<sub>2</sub> <80%.

Derived from the NICE NIV Guideline at

<http://www.nice.org.uk/nicemedia/live/13057/49885/49885.pdf>.

*C. Any deep tendon reflexes* may be examined at the time of interview if the interviewer is clinically trained, or they can be taken from the most recent clinical examination. If an appropriate recent clinical examination is not available, the following abnormal signs can be tested by a suitably trained non-clinical examiner: a pathologically brisk jaw jerk, pectoral reflexes, Hoffman's sign, crossed adductor reflexes, pathologically brisk patellar reflexes or ankle clonus.

*D. Please note the following examples of clinical findings which do not meet staging criteria for involvement:*

1. Extensor plantar reflexes with no other lower limb involvement do not satisfy criteria for involvement of the lower limbs.
2. Dysphagia not secondary to ALS or without weight loss greater than the 10% threshold and respiratory symptoms not fulfilling NICE respiratory failure guidelines do not meet Stage 4 criteria.
3. Fasciculation without wasting, weakness or reflex changes does not constitute involvement of a limb. It is acceptable as indicating involvement in the tongue.