Case Discussion: Amyotrophic Lateral Sclerosis

Case Discussion 1: ALS Patient Journey

P. Jesus (FR)
VOTING SYSTEM

Using any mobile device (smartphone, tablet, laptop, etc.) to easily participate to the voting system

Please access this website: espen18.cnf.io
EVERY 90 MINUTES
SOMEONE IS DIAGNOSED WITH ALS
AND EVERY 90 MINUTES
SOMEONE LOSES THEIR LIFE TO ALS

Presenter: Pierre Jesus (FR)
Expert Panel

- Kurt Boeykens: Nutrition Nurse (BE)/moderator
- Laurence Genton: Physician (CH)/moderator
- Irene Breton: Physician (ES)
- Arved Weimann: Physician (DE)
- Liana Poulia: Dietician (GR)
- Peter Austin: Pharmacist (UK)
- Chloé Chalmin: Speech Therapist (FR)
• Mr. D, 56 years old, is married with 2 children. He is a company manager.
• He is also an active person and runs twice a week and runs marathon four times a year.
• Since 8 months, he presents walking difficulties with muscle weakness associated with asthenia.
• After neurological assessment (electromyogram, MRI, lumbar puncture) a motor neuron disease: amyotrophic lateral sclerosis (ALS) is diagnosed according to Airlie House criteria.
• It is a spinal onset form.
• A treatment by riluzole is initiated by the Neurologist of ALS expert center.
• **Nutritional Assessment:** 3 month after diagnosis:
  – BW = 71kg
  – Height = 1.8 m
  – UBW (12 months ago) = 77 kg
  – Weight loss = 6.5%
  – BMI = 21.9 kg/m².
Poll: Is this patient malnourished according to the ESPEN diagnostic criteria for BMI or weight loss?
MALNUTRITION?

BMI < 18.5

Weight loss > 10% (indefinite of time)

Weight loss > 5% (last 3 months)

BMI < 20 (age < 70)

BMI < 22 (age ≥ 70)

FFMI
♀ < 15
♂ < 17

Diagnostic criteria for malnutrition - An ESPEN Consensus Statement.

Cederholm T1, Bosaeus I2, Barazzoni R3, Bauer J4, Van Gossum A5, Klek S6, Muscaritoli M7, Nyulasi I8, Ockenga J9, Schneider SM10, de van der Schueren MA11, Singer P12.

Indirect Calorimetry

- Resting energy expenditure (mREE) = 1853 kcal/d.
- REE calculated (cREE) by Harris & Benedict 1919 formula is 1565 kcal/d
- REE variation between mREE and cREE = +18.4%.
Poll: Which formula would you use to calculate energy needs?
Energy Expenditure

• When to repeat mREE?
• ALS patients higher EE?
BIA

- Bioelectrical impedance analysis (BIA) is also realized according to a valid formula.
- His fat-free mass (FFM) and fat mass (FM) are normal and the phase angle is $3.5^\circ$. 
Biological assessment

- No particular anomaly except for vitamin D deficiency at 23 nmol/L
- Albumin is at 37 g/L.
Vit-D

• Supplementation useful?
• Influence on the disease course?


**Vitamin D in amyotrophic lateral sclerosis.**

Libonati L, Onesti E, Gori MC, Ceccanti M, Cambieri C, Fabbri A, Frasca V, Inghilleri M.
Dietary advice

• Weight loss and hypermetabolism
• Referal to a dietician
• Food enrichment and oral nutritional supplements (ONS) to try to stop/reduce weight loss.
• Reducing a little bit his physical activity.
Specific Nutrients

Questions from the patient:

• omega-3 polyunsaturated fatty acids?
• Antioxidant supplementation?
• Some foods forbidden or favorable?

…to slow down the progression of the disease?
Dietary advice

• Frequency of follow-up
• Rescreening?
• Can oral nutritional supplementation or specific nutrients have an impact on survival?
• What about reducing physical activity?
  – Can resistance exercise not slow down the progression?
Follow-up

• At 3 months:
  – Weight stable
  – Bulbar disorders
  – Speech disorders
  – Swallowing disorders
  – Referral to a speech therapist
Speech therapist

• When to contact?
• Investigations? Maneuvers? Clinical evaluation?
• Modification of the diet?
• Follow-up? (frequency?)
The patient complains of hypersalivation:

- Scopolamine transdermal patch treatment is prescribed, which improves salivation disorder.
Three months later

**Videofluoroscopy:**

- Swallowing disorder with liquid but also with normal and chopped texture food. Meal time is over 45 min, with asthenia after meal.
- \( BW = 65 \text{ kg} \) (-14.5% compared to his usual weight)
- BIA: an important decrease of FFM and FM.
Poll: Is this patient now malnourished according to the ESPEN diagnostic criteria for BMI or weight loss?
**Diagnostic criteria for malnutrition - An ESPEN Consensus Statement.**

Actions?

• Serious alteration of nutritional status
• Important swallowing disorders
• Proposition of a gastrostomy tube for EN:
  – He refuses
• Dietary advice: smooth texture to avoid aspiration pneumonia.
Actions?

- Optimal timing to discuss the gastrostomy?
- Impact on survival/QoL?
- What about ethical considerations?
  - IC?
  - DNR-code?
Two months later

- Hospitalization for an aspiration pneumonia.
- Further weight loss: ABW = 60 kg
  - 21% compared to his usual weight
  - BMI: 18.5
- Once the acute episode is over, he accepts the insertion of gastrostomy feeding tube
- Respiratory tests find a forced vital capacity (FVC) to 40% of the theoretical value and blood gases finds an arterial pressure of CO$_2$ of 50mmHg.
- A non-invasive ventilation (NIV) is indicated.
Artificial Nutrition

• Starting EN: 500ml of an isocaloric EN product (1 kcal/ml)
• Gradually increased up to 1250 ml of an hypercaloric EN product (1.5 kcal/ml) = 1875 Kcal/day = 31 Kcal/kg/ABW
• Supplementation of vitamins and trace elements to avoid a refeeding syndrome
• EN is administered discontinuously twice in the day.
Artificial nutrition

• Feeding schedule to avoid refeeding syndrome?

• Which supplementation?
  – Thiamine? Dosis? Timing?

• Feeding regime? (twice a day?)
  – Why 1250 ml? (= 1000 + 250?)
  – Extra water?
Follow-up

• EN allowed a slight weight regain to 63kg but without further evolution.
• Increase of EN to 1500 ml of an hypercaloric EN product = 2250 Kcal/day = 35 Kcal/kg/ABW
• The patient described constipation with digestive discomfort after EN.
• You set up an EN with fibres and increase hydration on gastrostomy.
Follow-up

- Risk of overfeeding? (impact on respiratory function?)
- Not fibers from the start of EN?
- Changing the EN formula?
Detoriaration

- Despite your care, the patient has an alteration of his respiratory function and the NIV is set up continuously.
- EN is poorly tolerated with non-invasive ventilation
- The patient and his wife wants to stop EN but maintain a nutritional support
- A parenteral nutrition (PN) is started on peripherally inserted central catheter.
Poll: Would you, according to his poor condition, in your country or clinical setting, still go for the TPN option?
End stage

- Significant deterioration of his condition with respiratory failure
  - 18 months after the onset of his ALS
- Hospitalization in palliative care
- Decision in agreement with his wife to stop PN
End stage

• Everybody agrees?
  – From an ethical point of view?

• What about hydration?
Home care messages

• In most ALS patients malnutrition (which is detrimental for survival) will develop in the course of the disease so:
  – Complete assessment, follow-up and nutritional care plan are recommended on a regular basis and in a multidisciplinary way

• The pros and the cons of artificial nutrition have to be adequately discussed with the patient, family and caregivers.
ESPEN guideline clinical nutrition in neurology

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