



EUROPEAN
SOCIETY FOR
CLINICAL
NUTRITION AND
METABOLISM

ESPEN Congress Madrid 2018

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

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ALS case

ESPEN congress Madrid 2018



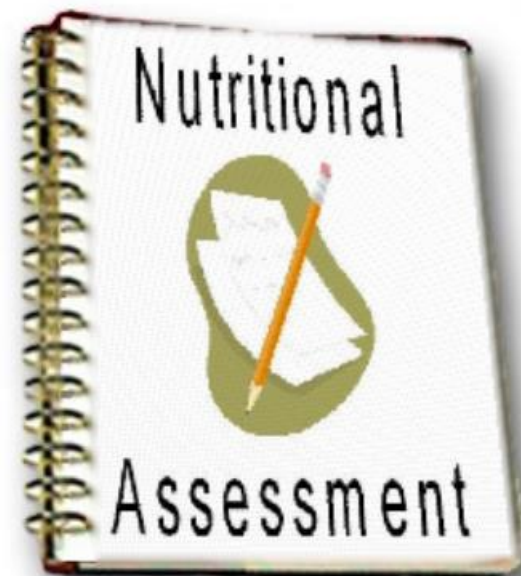
Presenter: Pierre Jesus (FR)

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.

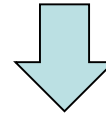


Live Content Slide

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Poll: Is this patient malnourished according to the ESPEN diagnostic criteria for BMI or weight loss?

MALNUTRITION?

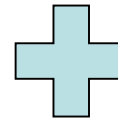


BMI < 18,5

Weight loss > 10%
(indefinite of time)

Weight loss > 5%
(last 3 months)

or



BMI < 20
(age < 70)

or

BMI < 22
(age ≥ 70)

or

FFMI
♀ < 15
♂ < 17

Clin Nutr. 2015 Jun;34(3):335-40. doi: 10.1016/j.clnu.2015.03.001. Epub 2015 Mar 9.

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%.



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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°



Biological assessment

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



Vit-D

- Supplementation usefull?
- Influence on the disease course?

Funct Neurol. 2017 Jan/Mar;32(1):35-40.

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
- Referral to a dietician
- Food enrichment and oral nutritional supplements (ONS) to try to stop/reduce weight loss.
- Reducing a little bit his physical activity.

**Dietary
Advice**

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?

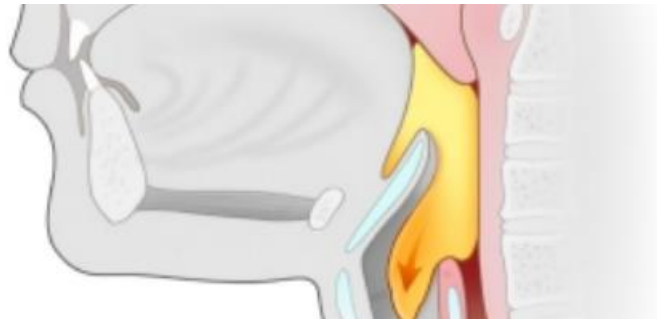


Dietary advice

- 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?)

Hypersalivation

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 kg (-14.5% compared to his usual weight)
- BIA: an important decrease of FFM and FM.

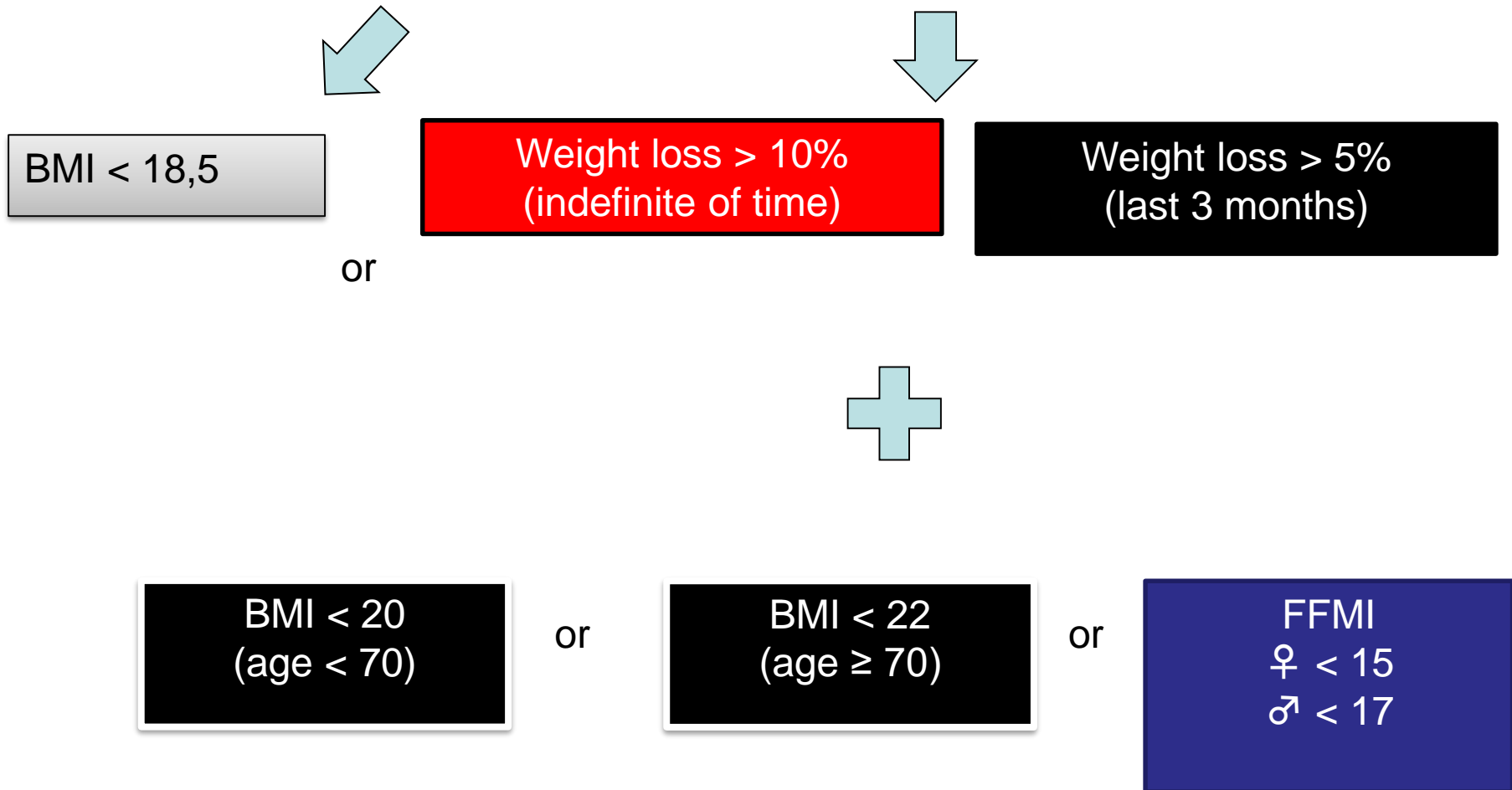


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Poll: Is this patient now malnourished according to the ESPEN diagnostic criteria for BMI or weight loss?

MALNUTRITION?



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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 CO₂ of 50mmHg.
- A non-invasive ventilation (NIV) is indicated.

Artificial Nutrition

- Starting EN: 500ml of an isocaloric EN product (1kcal/ ml)
- Gradually increased up to 1250 ml of an hypercaloric EN product (1.5kcal/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?
 - Vitamins? Trace-elements? Which form? Dose? Necessary?
- 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.

Live Content Slide

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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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Marjolein Van der Marck ^m, Rainer Wirth ⁿ, Pierre Singer ^o, Stephan C. Bischoff ^p