A background image of a Sheffield Supertram, specifically a Citylink CL 202, in red, blue, and yellow livery. The tram is stopped at a station platform. The text 'Citylink' and 'CL' are visible on the front display, and '202' is on the side. A 'Theo' logo for The Children's Hospital Charity is on the side. The front of the tram shows the number '399 202' and 'SUPERTRAM' branding.

ALS/MND Nutritional Guidelines: What do...and don't they tell us?

Dr Sean White

Honorary Senior Lecturer

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Dietitian and (soon to be MND Coordinator)

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Trust

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Guidelines not tramlines



ALS and me...



European Academy of Neurology (EAN) guideline on the management of amyotrophic lateral sclerosis in collaboration with European Reference Network for Neuromuscular Diseases (ERN EURO-NMD)

Philip Van Damme , Ammar Al-Chalabi, Peter M. Andersen, Adriano Chiò, Philippe Couratier, Mamede De Carvalho, Orla Hardiman, Magdalena Kuźma-Kozakiewicz, Albert Ludolph, Christopher J. McDermott, Jesus S. Mora, Susanne Petri, Katrin Probyn, Evelyne Reviers, François Salachas, Vincenzo Silani, Ole-Bjørn Tysnes, Leonard H. van den Berg, Gemma Villanueva, Markus Weber

First published: 12 March 2024 | <https://doi.org/10.1111/ene.16264> | Citations: 4

Canadian best practice recommendations for the management of amyotrophic lateral sclerosis

Christen Shoesmith ¹, Agessandro Abrahao ², Tim Benstead ², Marvin Chum ², Nicolas Dupre ², Aaron Izenberg ², Wendy Johnston ², Sanjay Kalra ², Desmond Leddin ², Colleen O'Connell ², Kerri Schellenberg ², Anu Tandon ², Lorne Zinman ²

Affiliations + expand

PMID: 33199452 PMID: [PMC7683000](#) DOI: [10.1503/cmaj.191721](#)

Motor neurone disease: assessment and management

NICE guideline | NG42 | Published: 24 February 2016 | Last updated: 23 July 2019

Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review)

Report of the Quality Standards Subcommittee of the American Academy of Neurology



ESPEN guideline clinical nutrition in neurology

Rosa Burgos ^{a,*}, Irene Bretón ^b, Emanuele Cereda ^{c,d}, Jean Claude Desport ^e, Rainer Dziewas ^f, Laurence Genton ^g, Filomena Gomes ^h, Pierre Jésus ^e, Andreas Leischker ⁱ, Maurizio Muscaritoli ^j, Kalliopi-Anna Poulia ^k, Jean Charles Preiser ^l, Marjolein Van der Marck ^m, Rainer Wirth ⁿ, Pierre Singer ^o, Stephan C. Bischoff ^p

Guideline "Motor neuron diseases" of the German Society of Neurology (Deutsche Gesellschaft für Neurologie)

Susanne Petri , Torsten Grehl, Julian Grosskreutz, Martin Hecht, Andreas Hermann, Sarah Jesse, Paul Lingor, Wolfgang Löscher, André Maier, Benedikt Schoser, Marcus Weber & Albert C. Ludolph

Neurological Research and Practice 5, Article number: 25 (2023) | [Cite this article](#)

Beware of 'BOGSAT'





ELSEVIER

Contents lists available at [ScienceDirect](#)

Clinical Nutrition ESPEN

journal homepage: <http://www.clinicalnutritionespen.com>

Original article

Understanding the current nutritional management for people with amyotrophic lateral sclerosis - A mapping review

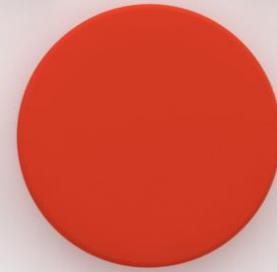
Munira Essat^a, Elizabeth Coates^a, Mark Clowes^a, Daniel Beever^a, Gemma Hackney^a, Sean White^b, Theocharis Stavroulakis^c, Vanessa Halliday^{a,*}, Christopher McDermott^c, on behalf of the HighCALS group



- 109 docs and 13 guidelines reviewed
- Top line recommendations
- Focus on dysphagia and gastrostomy
- Lack detail required to guide nutritional management

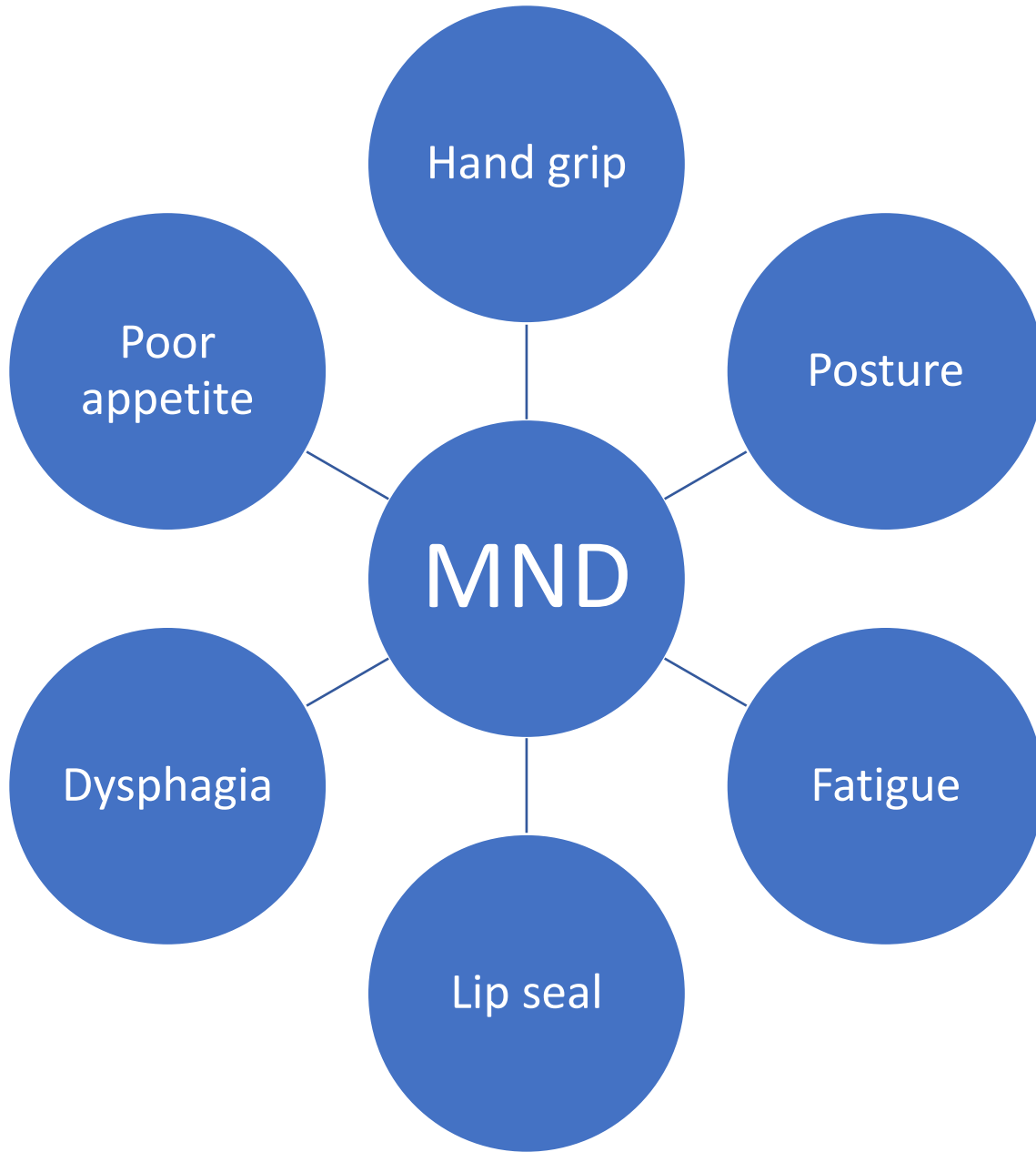
Aims

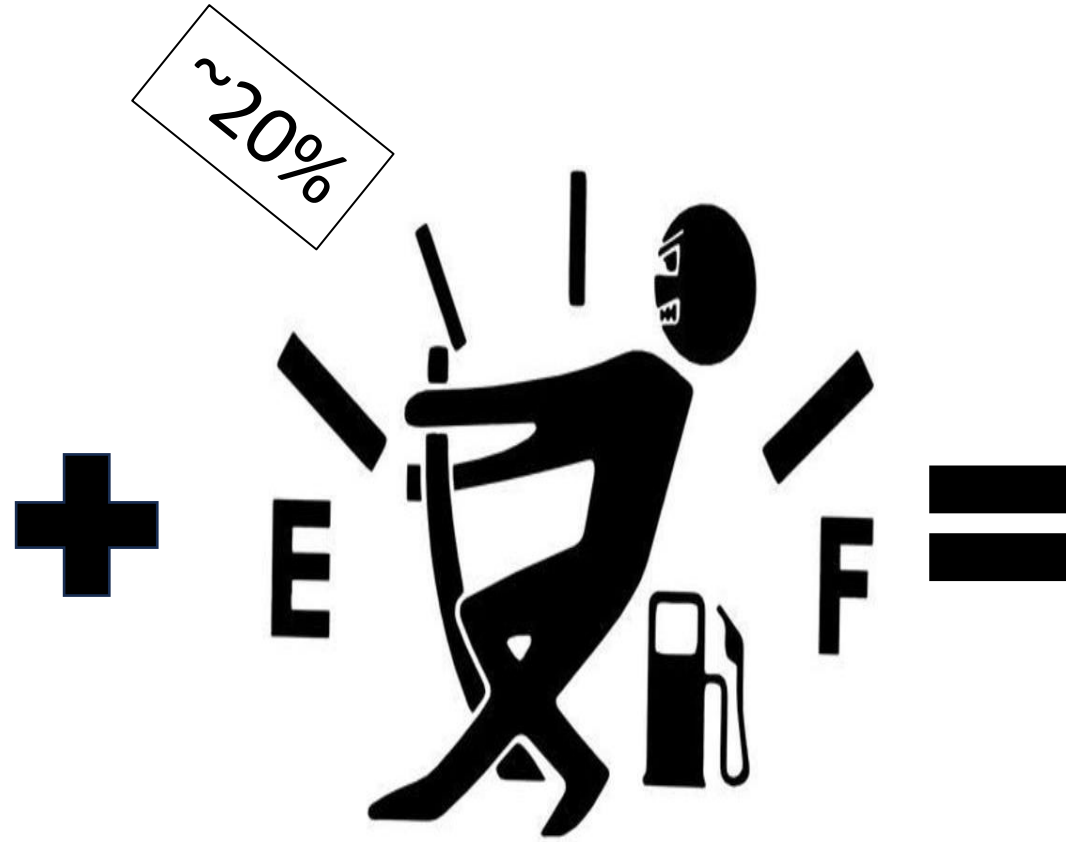
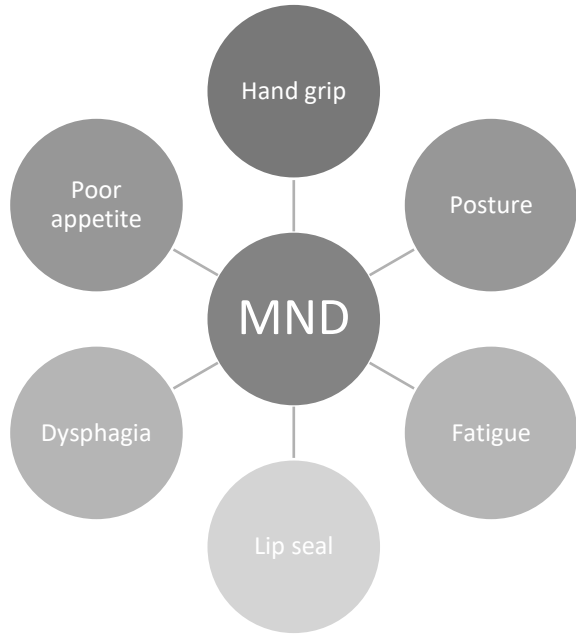
- What is the problem?
- Current guidance
- What are the gaps or uncertainties?
- Where next?



What is the dietetic
problem?

Hypermetabolism

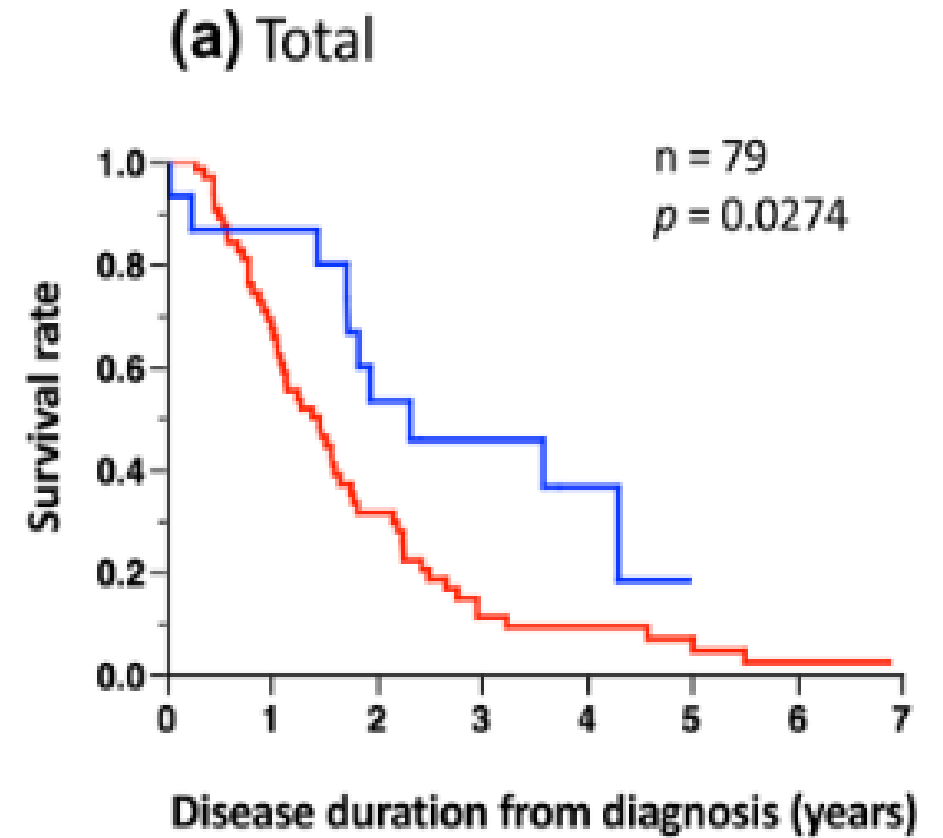




Weight matters...



After
diagnosis



MALNUTRITION SCREENING

‘Screening for malnutrition (BMI, weight loss) is recommended at diagnosis and during the follow-up every 3 months.’

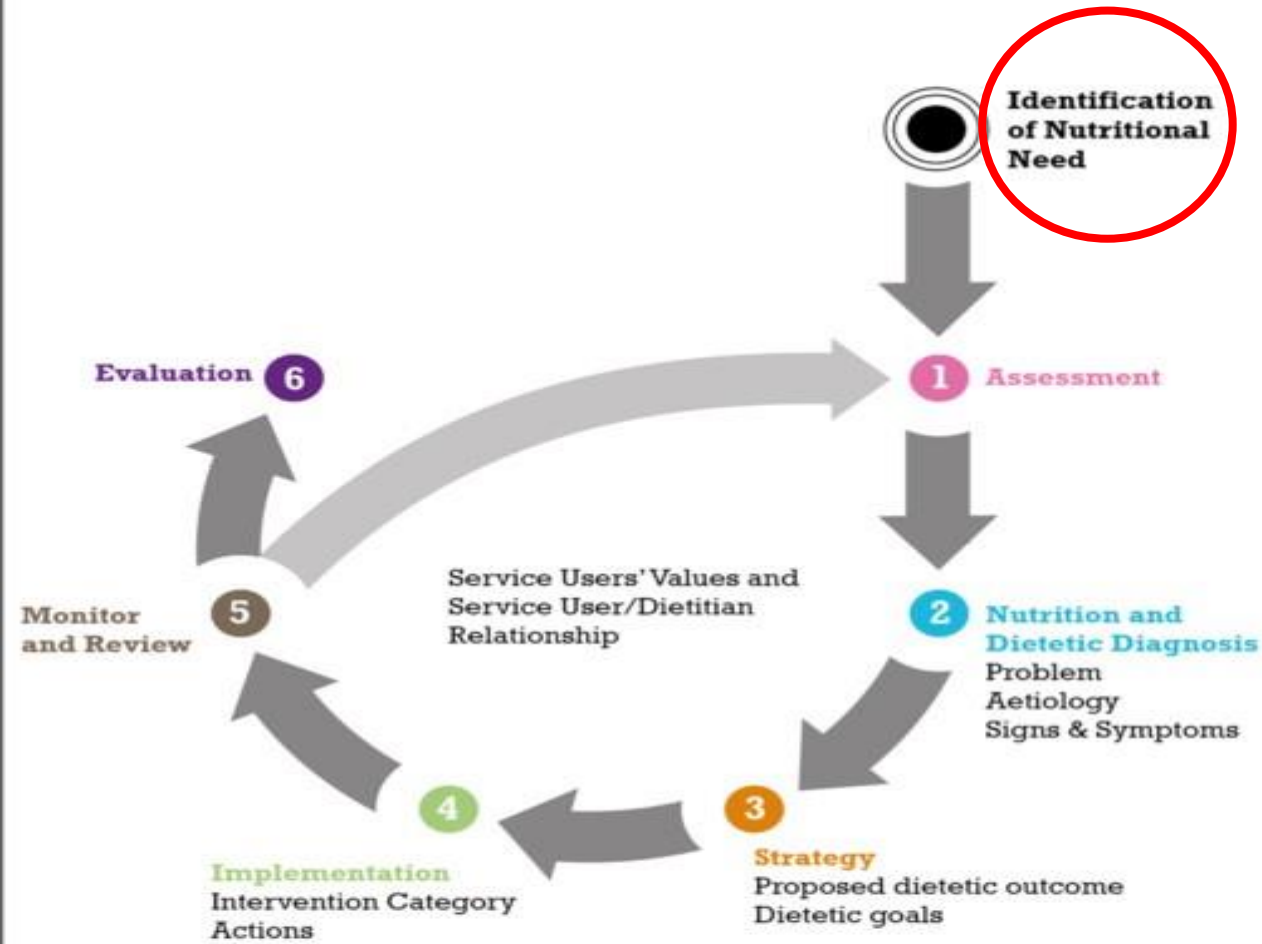
(Burgos et al 2018)

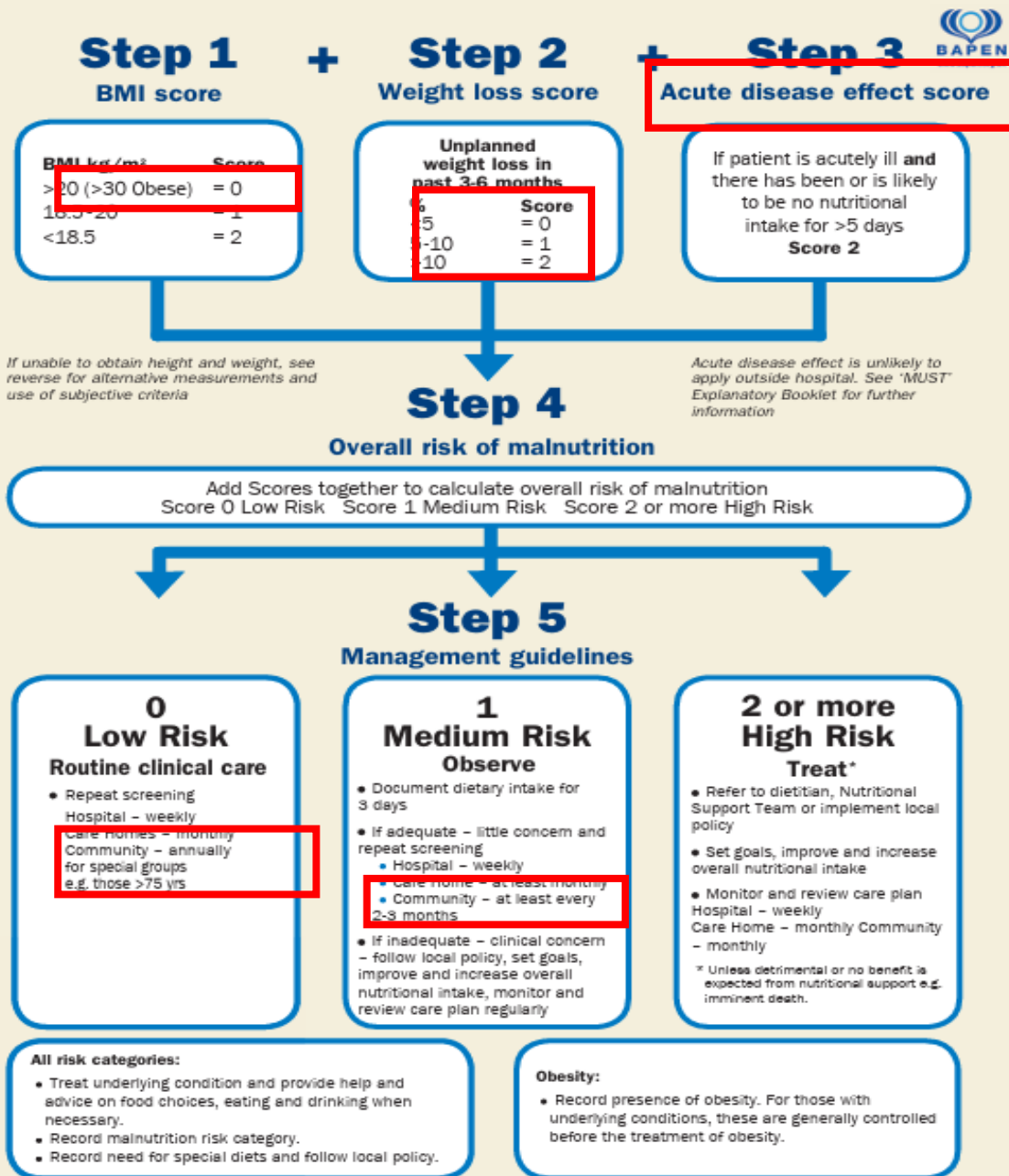
Screening is our 'precog'



Model and Process for Nutrition and Dietetic Practice

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Quality ALS nutritional screening should be...

Proactive

Quick

Repeated appropriately

Everyone's responsibility

ENERGY REQUIREMENTS

‘Energy requirements in non-ventilated ALS patients should be estimated ~30 kcal/kg’

‘NIV [...] requirements should be estimated as 25-30 kcal/kg body weight or using the Harris-Benedict equation’

(Burgos et al 2018)

Estimating energy requirements – 65kg male



PENG guideline :

- BMI 18.5-30kg/m²: <65 years: 22kcal/kg; >65 years: 24kcal/kg
- Add additional kcals for PAL (can range from 10-60% depending on activity)
- TDEE=2184kcal (24kcal/kg; PAL 40%)

FULL LENGTH ARTICLE · Volume 37, Issue 1, P354-396, February 2018



ESPEN guideline clinical nutrition in neurology

Rosa Burgos^a · Irene Bretón^b · Emanuele Cereda^{c,d} · ... · Rainer Wirth^e · Pierre Singer^a · Stephan C. Bischoff^f ... Show more

Affiliations & Notes ▾ Article Info ▾

ESPEN guideline :

- Indirect calorimetry or,
- 25-30 kcal/kg body weight (depending on ventilation status) or,
- Harris-Benedict equation and then adapt based on monitoring
- TDEE=1950 kcal (30kcal/kg); 1933kcal (H-B + 40% PAL)

Estimating daily energy expenditure in individuals with amyotrophic lateral sclerosis^{1 2 3}

Edward J Kasarskis, Marta S Mendiondo, Dwight E Matthews, Hiroshi Mitumoto, Rup Tandan, Zachary Simmons, Mark B Bromberg, Richard J Kryscio, for the ALS Nutrition/NIPPV Study Group

Kasarskis (2014) :

- Model 6 TDEE recommended for practice kcal/d (men):
[66+ (13.7 x weight(kg))+(5 x height (cm))- (6.76 x age (years))]+ (55.96 x ALSFRS-6 score)-168
- TDEE: 2216kcal (65yrs; 172cm tall; 65 years; ALSFRS-6: 18)
- Note: Modelled – measured TDEE(kcal/d): 11 +/- 521kcal

Predicting energy requirements – can we get better?



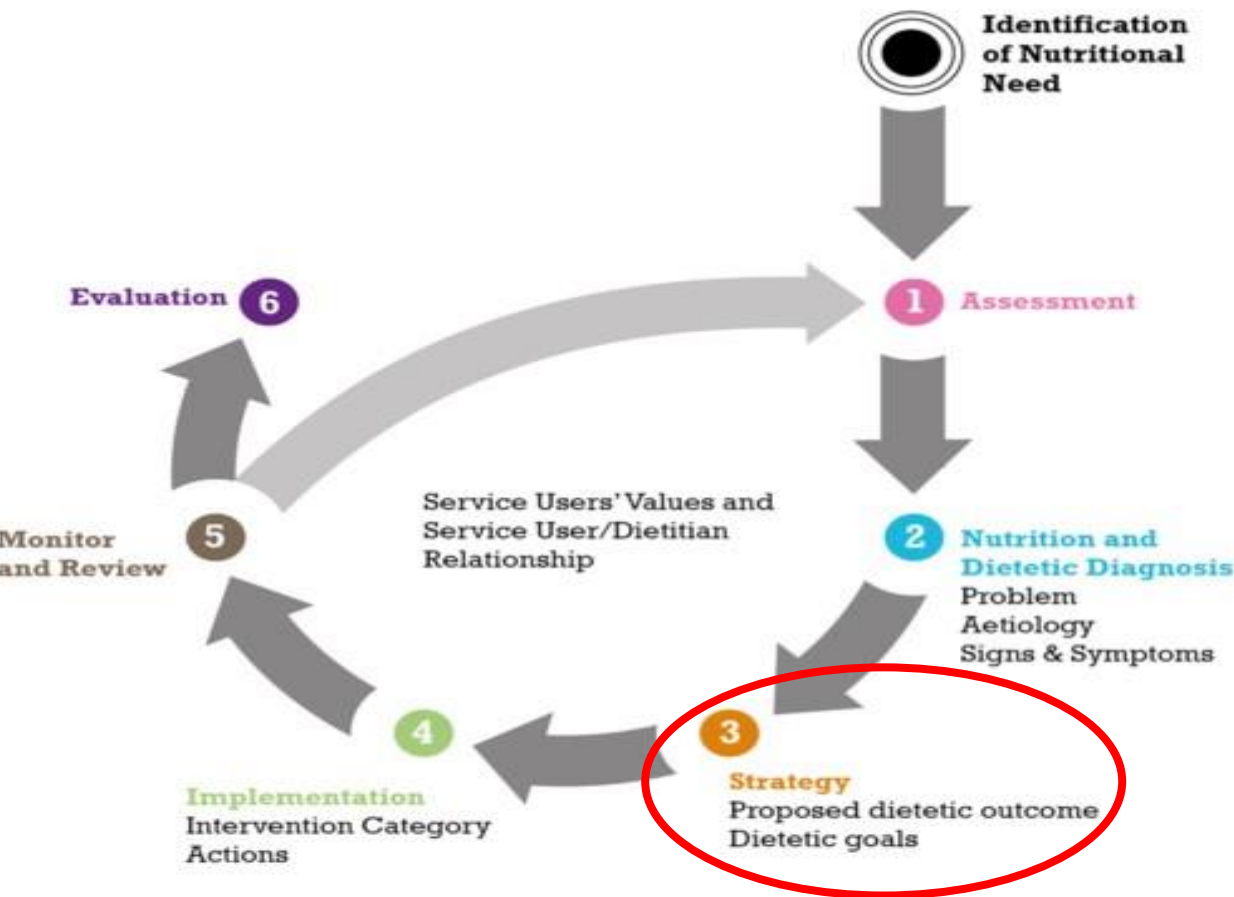
DIETETIC GOALS

‘Weight gain should be recommended in patients with a BMI <25.0kg/m², weight stabilization in those with a BMI 25-35kg/m², and weight loss in patients with a BMI>35kg/m²’

(Burgos et al 2018)

Model and Process for Nutrition and Dietetic Practice

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Recommendation 5:

In ALS patients, weight loss is detrimental for survival, but whether oral or EN should aim at weight stabilization or weight gain has not been clarified and may depend on baseline nutritional state. Weight gain should be recommended in patients with a baseline body mass index (BMI <25.0 kg/m²), weight stabilization in those with a BMI between 25 and 35 kg/m², and

BMI (kg m ⁻²)	Weight goal set for people with ALS, n (%)		
	Weight loss	Weight maintenance	Weight gain
< 18.5 (N = 130)	0 (0)	12 (9)	115 (91)
18.5–25 (N = 130)	0 (0)	92 (72)	35 (28)
25–30 (N = 130)	2 (2)	125 (98)	0 (0)
> 30 (N = 127)	27 (21)	100 (79)	0 (0)

ORAL NUTRITION SUPPORT

‘Nutritional supplementation is recommended for ALS patients who do not cover their nutritional requirements with an enriched diet.’

(Burgos et al 2018)



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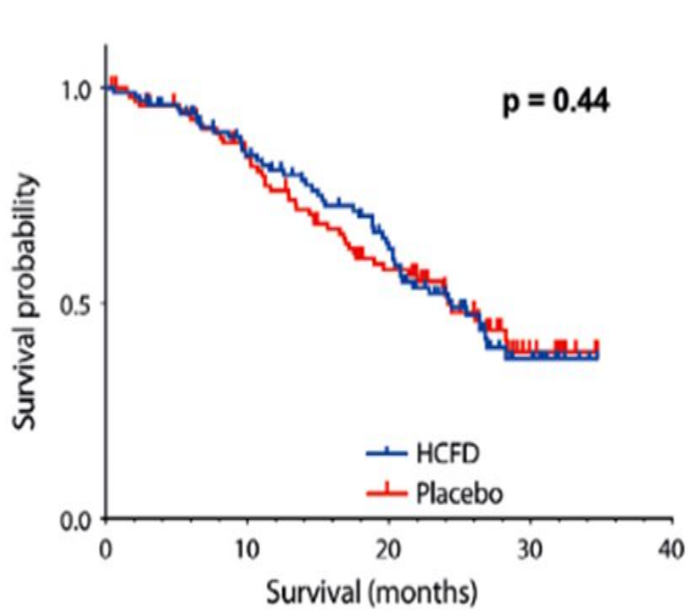
Clinical Nutrition

journal homepage: <http://www.elsevier.com/locate/clnu>

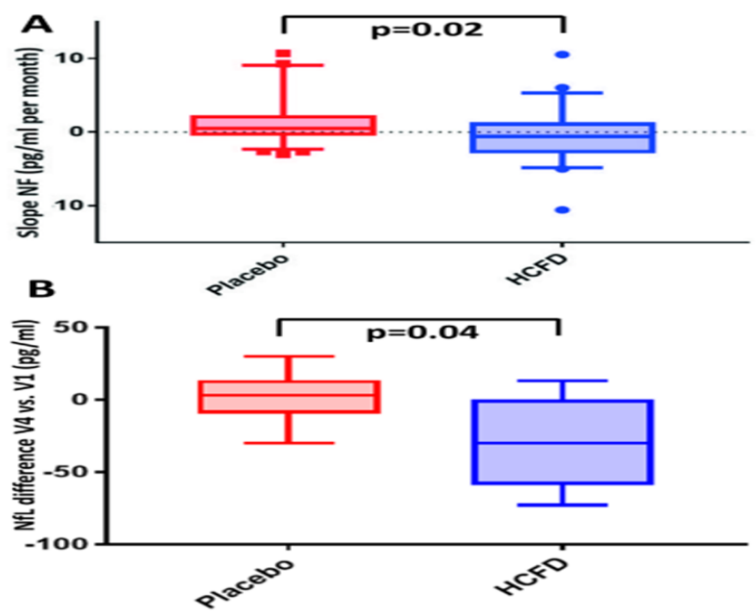
Original article

Interventions to promote oral nutritional behaviours in people living with neurodegenerative disorders of the motor system: A systematic review

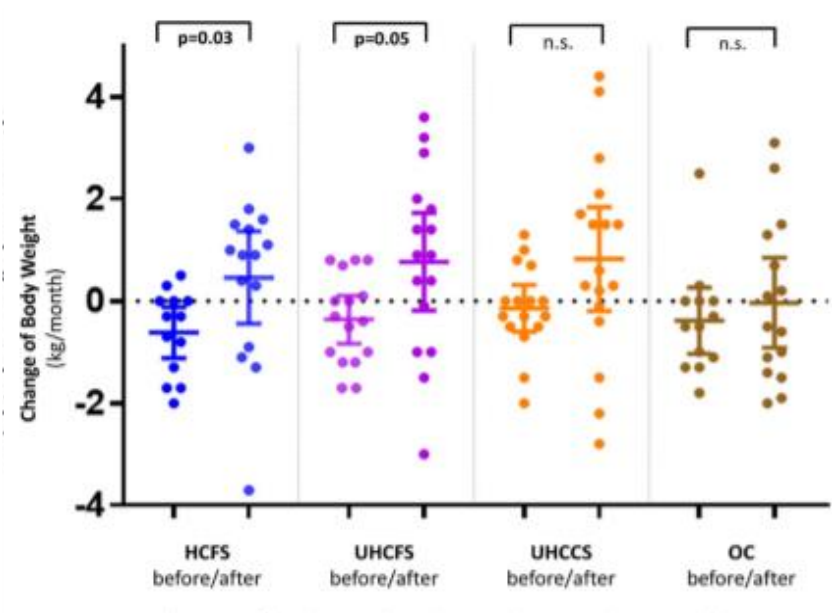
Munira Essat ^a, Rachel Archer ^a, Isobel Williams ^b, Nicolò Zarotti ^c, Elizabeth Coates ^a, Mark Clowes ^a, Daniel Beever ^a, Gemma Hackney ^a, Sean White ^d, Theocharis Stavroulakis ^c, David White ^a, Paul Norman ^{b, *}, Christopher McDermott ^c, on behalf of the HighCALS group



Ludolph et al 2020



Dorst et al 2020



Dorst et al 2022



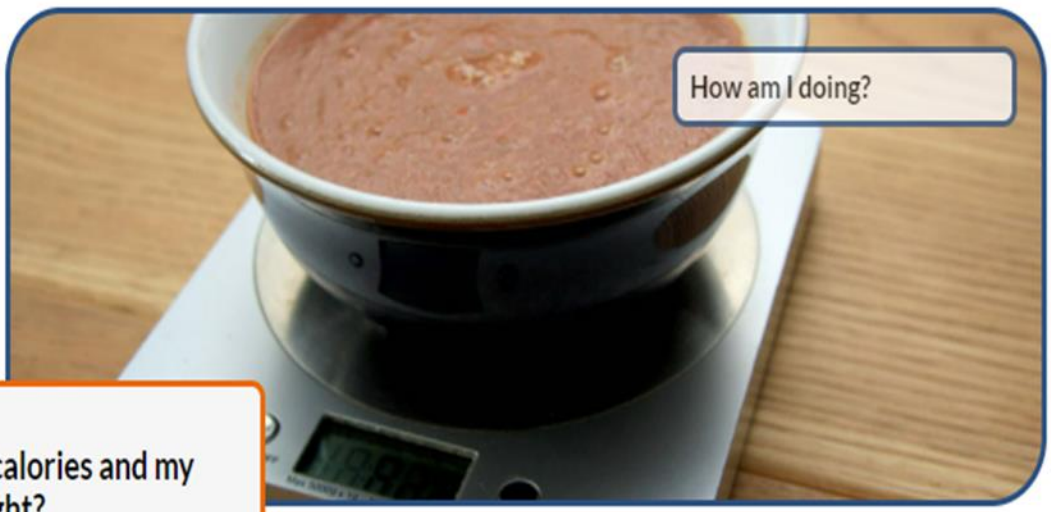
Aim:

To develop, implement and evaluate a complex nutritional intervention for people with MND to improve survival and quality of life.



- My Food Diary
- How am I doing?
- All Resources
- My Resources
- Why boost my calories and my weight?

Welcome to OptiCALS Joe Bloggs



How am I doing?

Why boost my calories and my weight?

[Start Here >>](#)



All Resources



My Resources

GASTROSTOMY PLACEMENT (ENTERAL FEEDING)

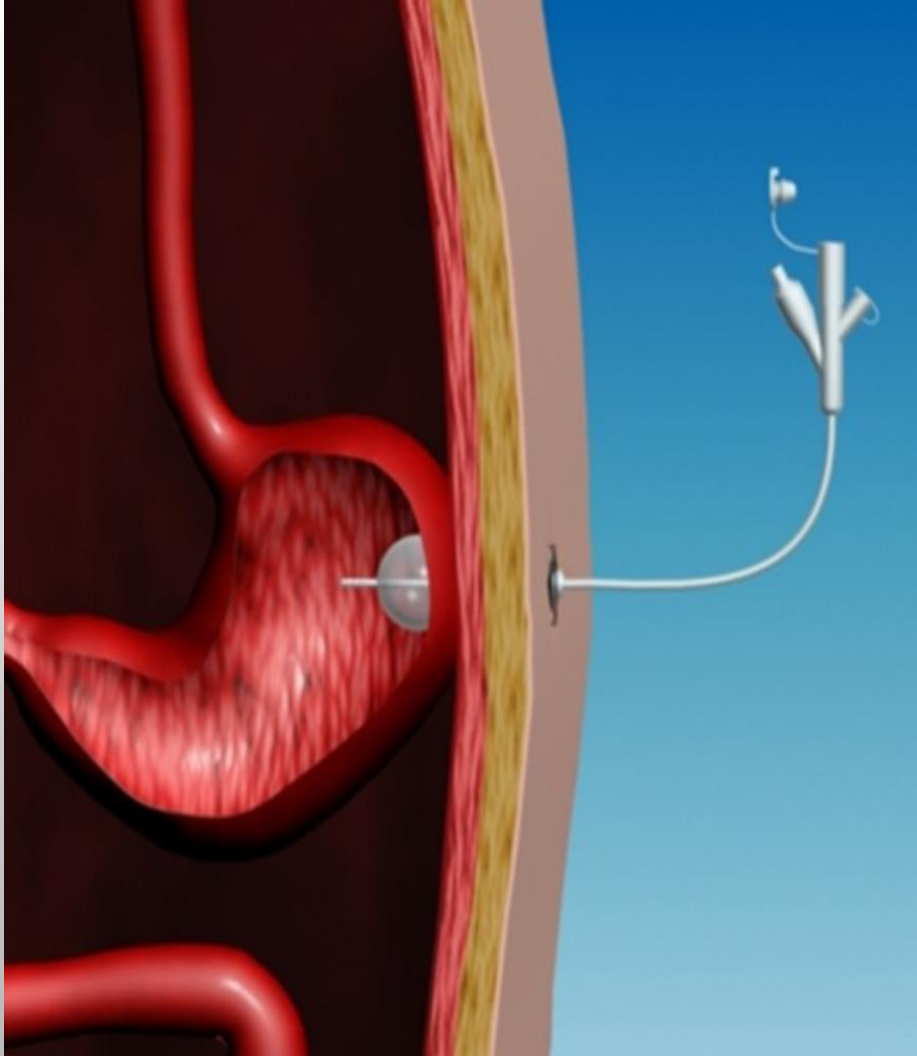
‘Percutaneous endoscopic gastrostomy (PEG) [...] is recommended in cases of more advanced dysphagia and weight loss.’

(Petri et al 2023)

‘PEG should be considered for prolonging survival in patients with ALS’

(Miller et al 2009)

Gastrostomy: an effective intervention?



‘There are no RCTs or quasi-RCTs to indicate whether enteral tube feeding is effective compared to continuation of oral feeding for any of the outcome measures.’
(Sulistyo et al 2023)



‘Discuss gastrostomy at an early stage, and at regular intervals as MND progresses.’
(NICE 2019)

REFERRAL-PLACEMENT DELAY

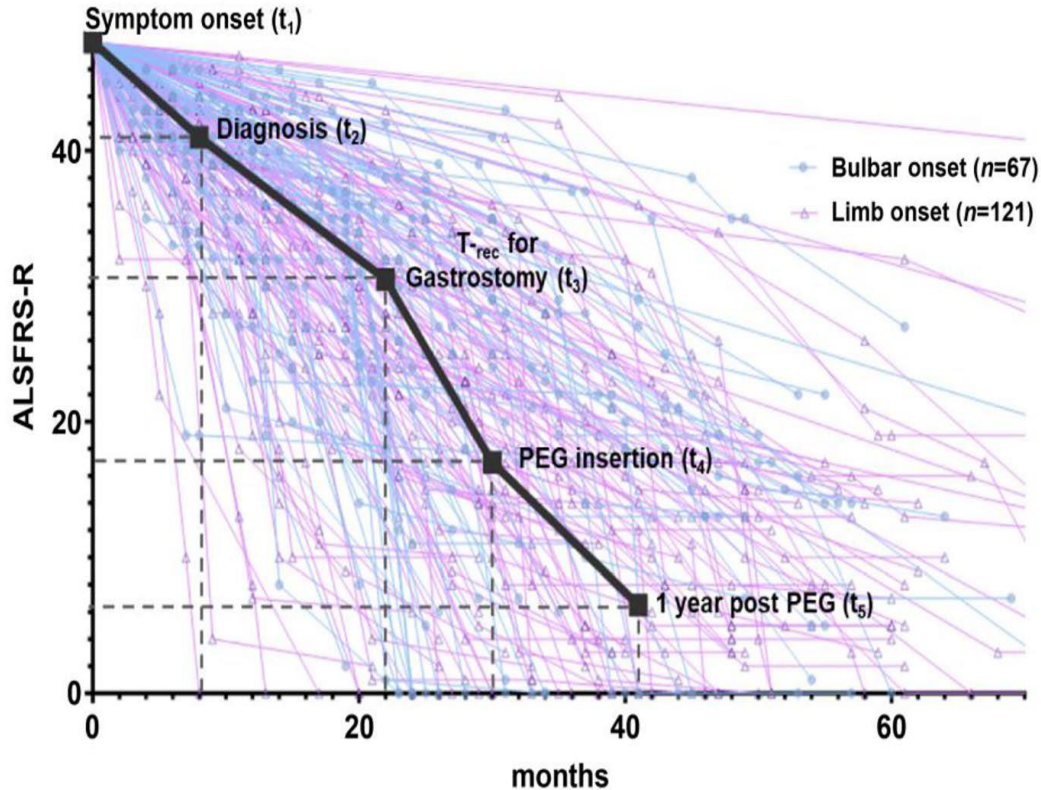
‘If a person is referred for a gastrostomy it should take place with-out delay.’

(NICE 2019; Van Damme et al 2023)

‘Once a decision is made to insert an enteral feeding tube, insertion should be performed within 4 weeks.’

(Shoesmith et al 2020)

Indication-Decision-Referral-placement delays



Son et al 2024

- Malnutrition, aspiration and respiratory risks progress post indication and referral
- Information about delay informs decision-making
- Aim: more efficient gastrostomy placement pathways

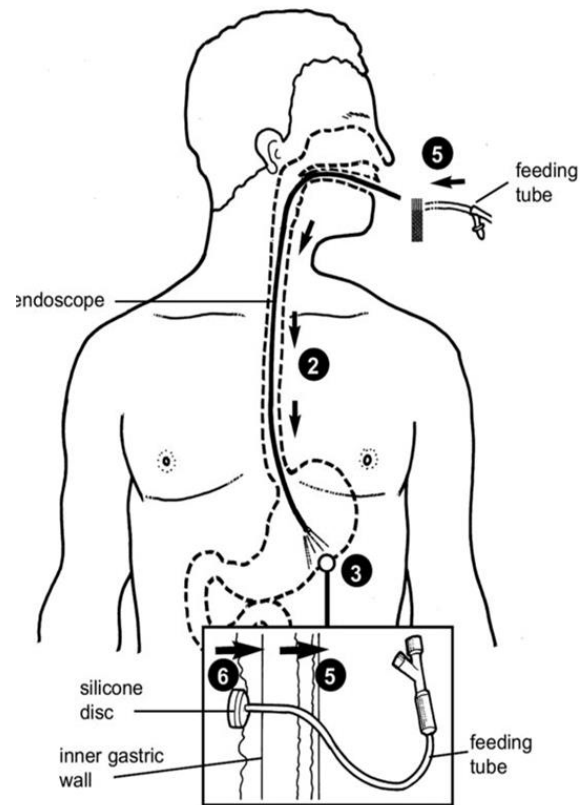
GASTROSTOMY PLACEMENT PROCEDURE

‘We recommend PEG as the preferred approach for gastrostomy. When available, in more frail patients, RIG positioning by expert team maybe indicated.’
(Burgos et al 2018)

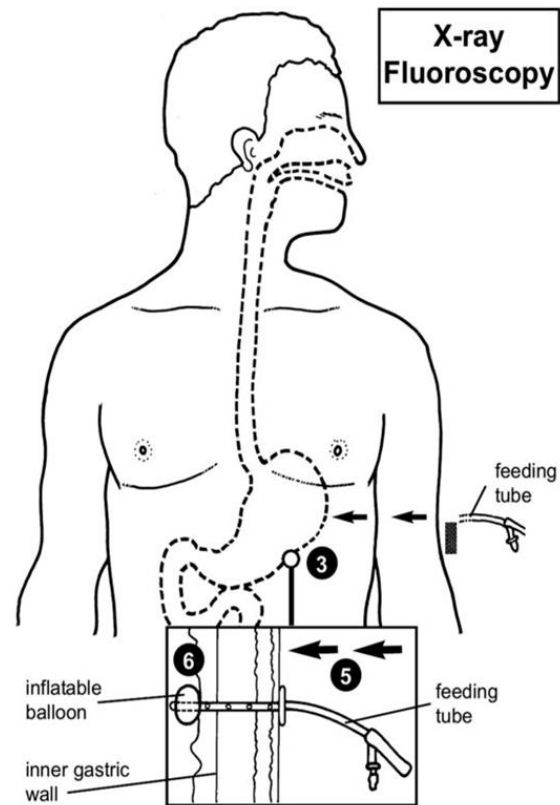
‘There is insufficient evidence to recommend PEG or RIG as the usual procedure for gastrostomy insertion.’
(Shoesmith et al 2020)

Gastrostomy in MND

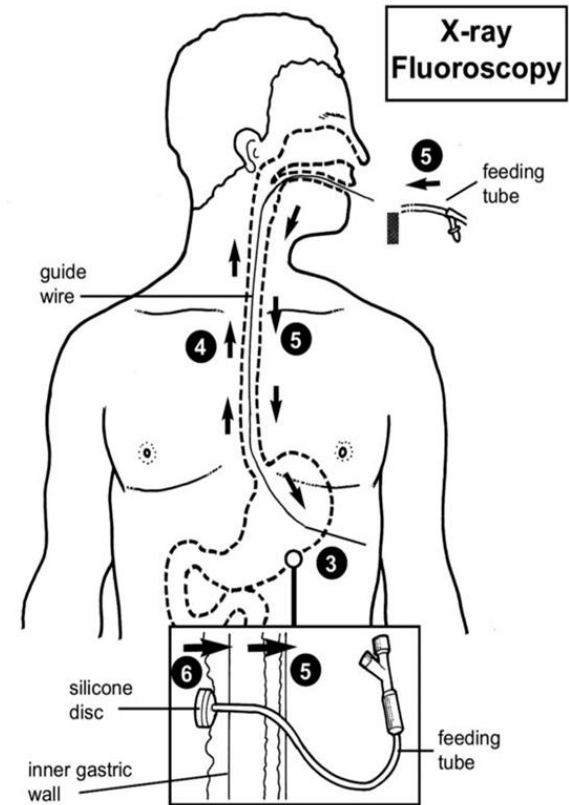
Percutaneous Endoscopic Gastrostomy (PEG)



Radiologically Inserted Gastrostomy (RIG)



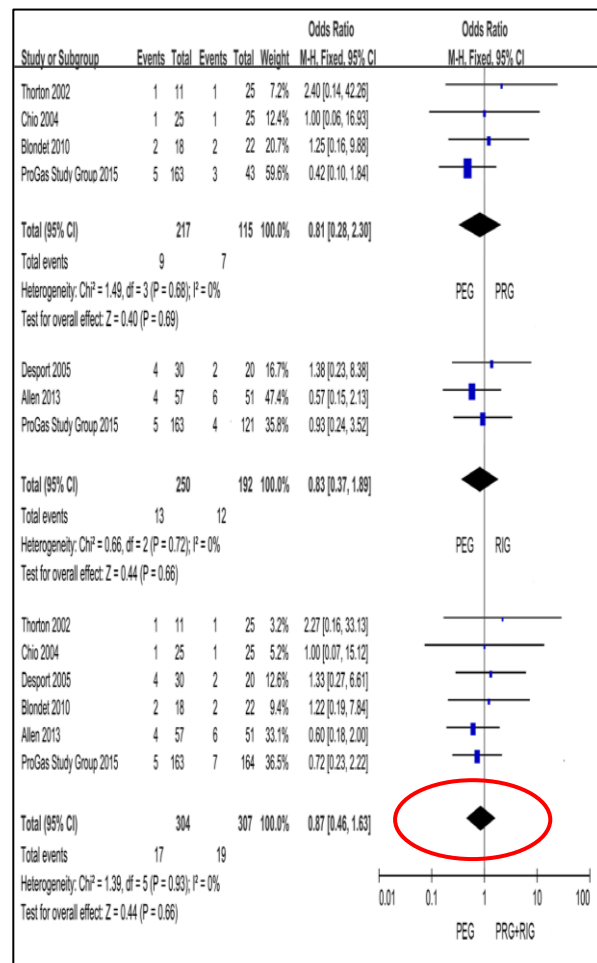
Per-oral Image-guided Gastrostomy (PIG)



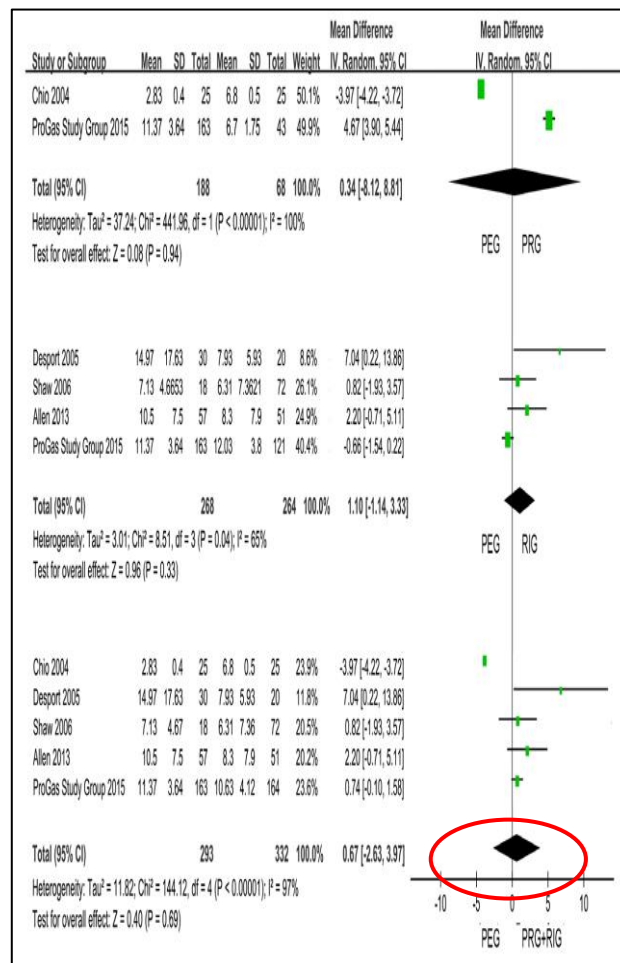
Yang et al 2017

- Systematic review of PEG v PRG v RIG in ALS
- 7 studies with 701 cases

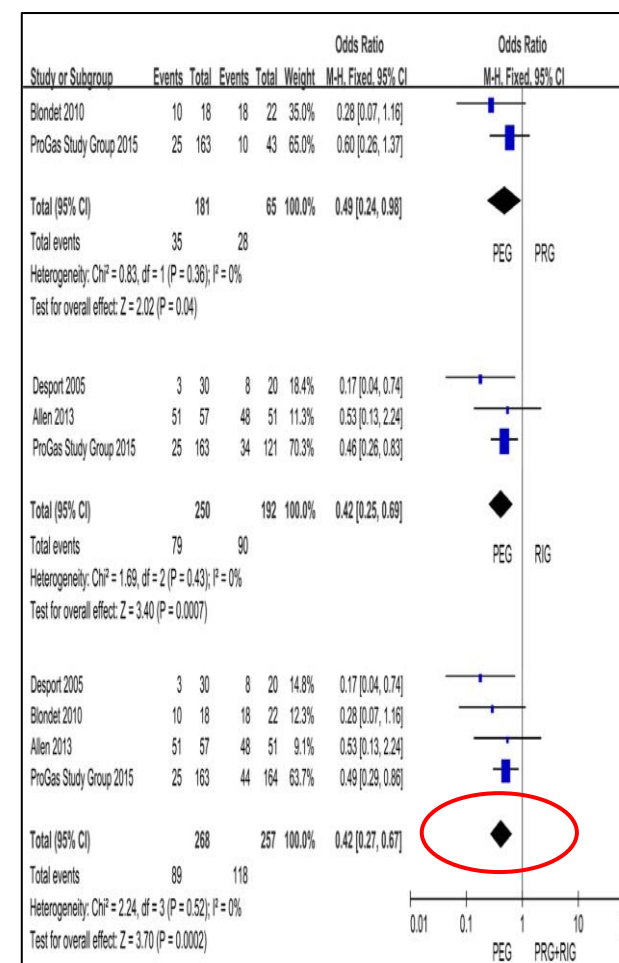
30-day mortality



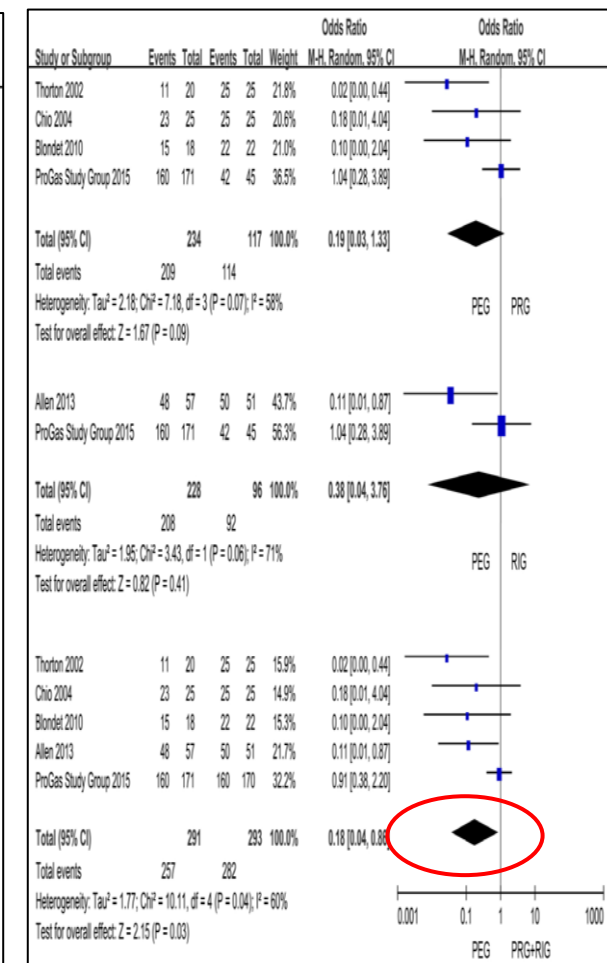
Survival length



Pain

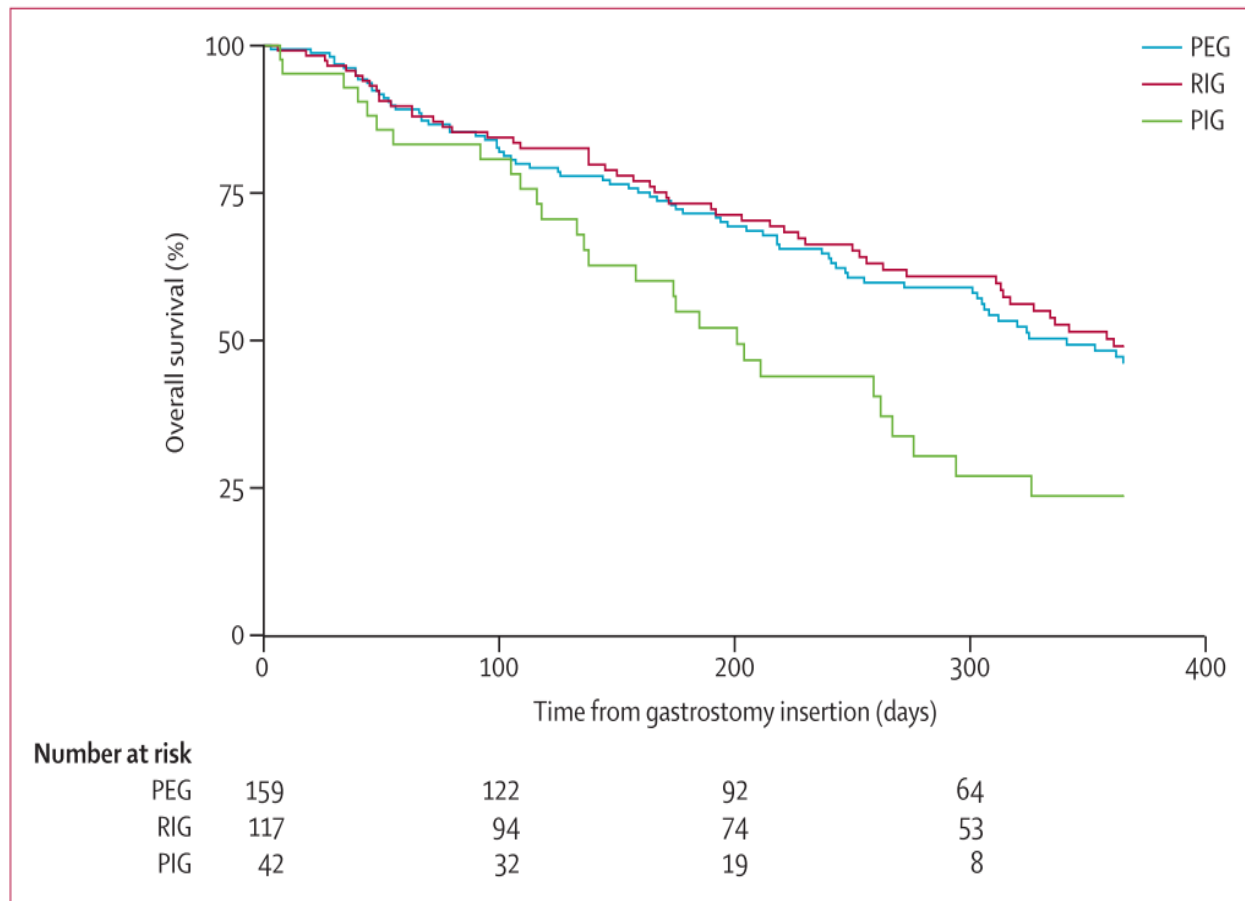


Success rate



Gastrostomy in patients with amyotrophic lateral sclerosis (ProGas): a prospective cohort study

ProGas Study Group*

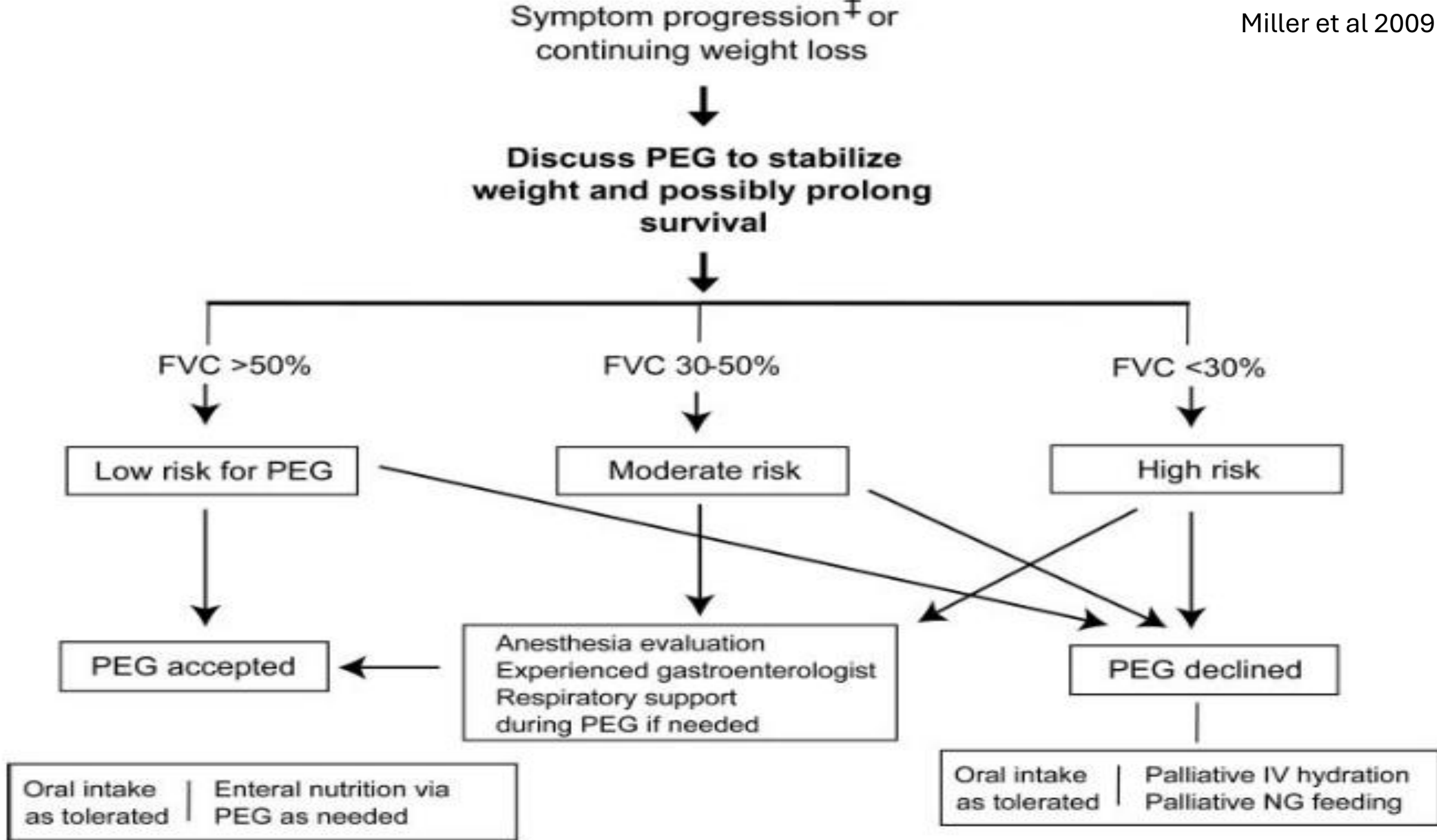


No difference in 30-day mortality or overall survival between methods after adjustment for case mix variables and treatment centre

RESPIRATORY PROCEDURAL RISK

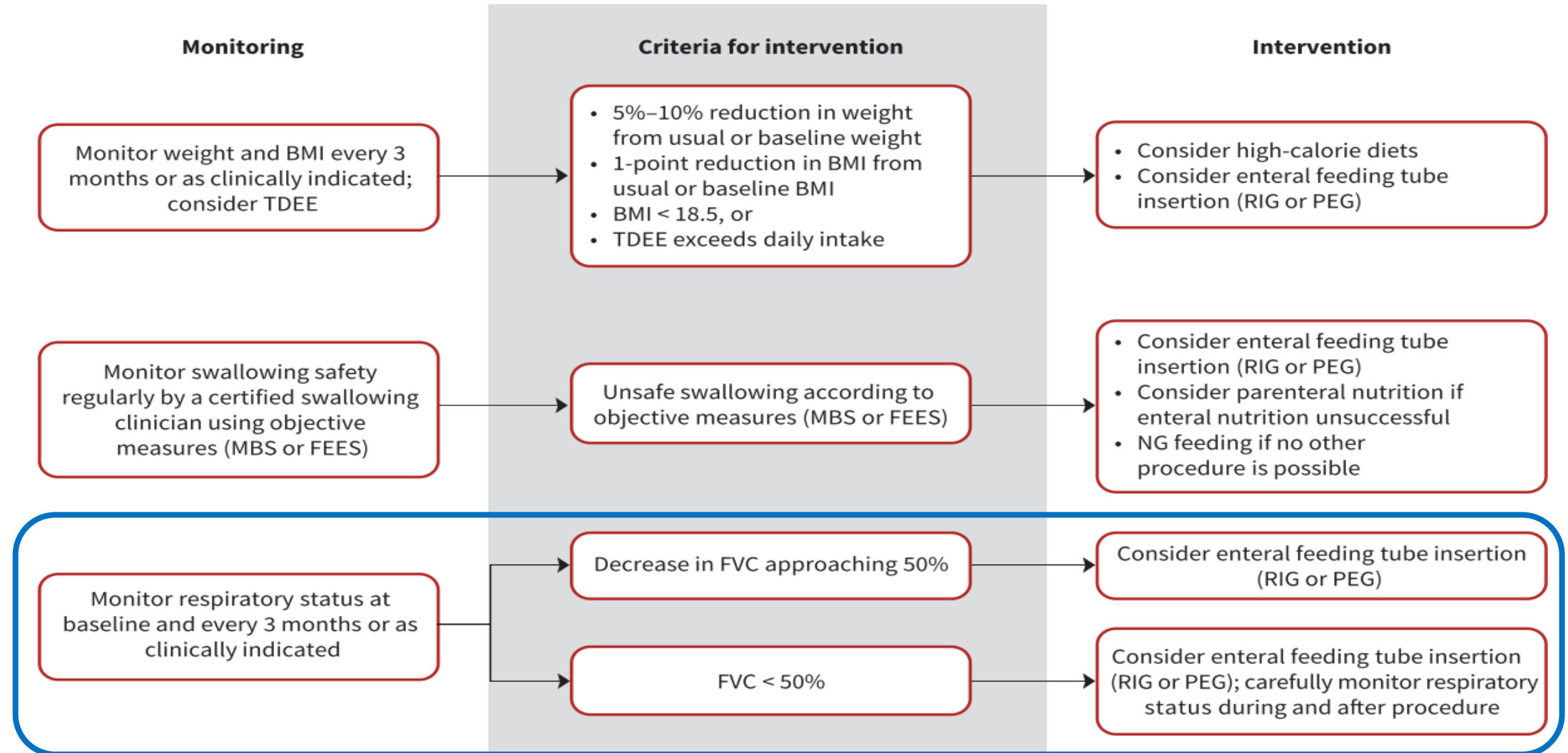
‘Patients with dysphagia will possibly be exposed to less risk if PEG is placed when FVC is above 50% of predicted’
(Miller et al 2009)

‘A decrease in FVC approaching 50% should prompt consideration of referral for enteral tube insertion, even in the absence of dysphagia. An FVC < 50% should not necessarily preclude the recommendation of enteral feeding tube insertion as long as respiratory status is carefully monitored during and after the procedure’
(Shoesmith et al 2020)



Canadian guidelines nutrition decision tree

GUIDELINE



Respiratory failure: a risk to manage but not a contraindication



FVC<50%?

- FVC<50% associated with increased 30-day mortality (Kasarskis et al 1999; Pena 2012))
- Risks do increase...but can be managed
- Outcomes and procedures are improving (Gorrie et al 2019)
- Need better risk communication
- Can we avoid people having gastrostomy before it is needed?

TIMING OF GASTROSTOMY

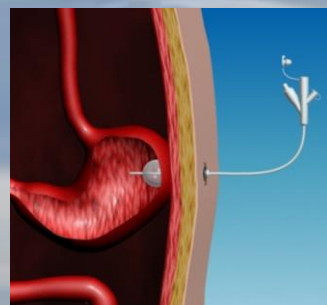
‘Explain the benefits of early placement of a gastrostomy, and the possible risks of a late gastrostomy’

(NICE 2022)

‘Information regarding potential benefits and risks of enteral feeding tubes...’

(Shoesmith et al 2020)

Compensating



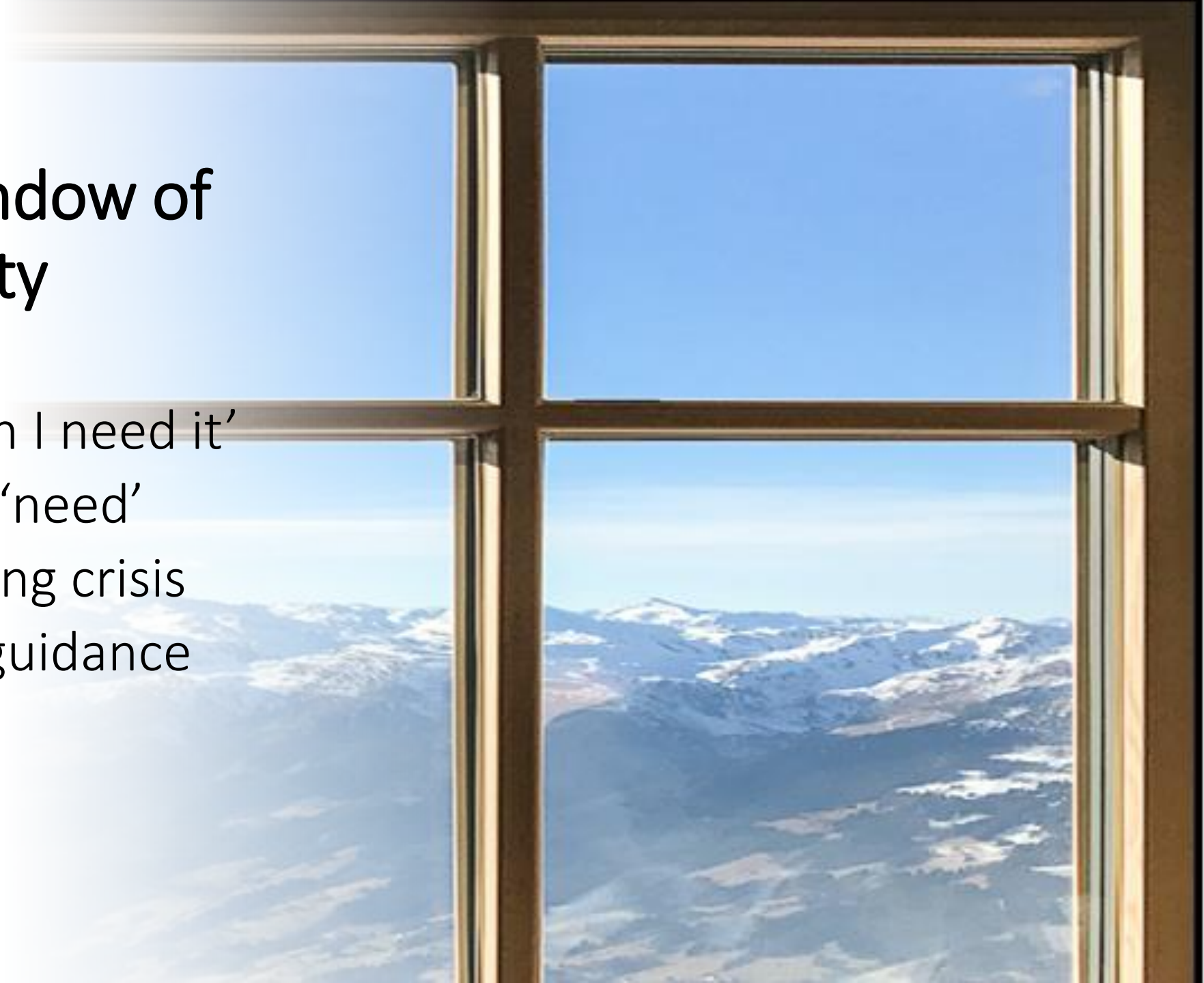
Decisional
cutoff

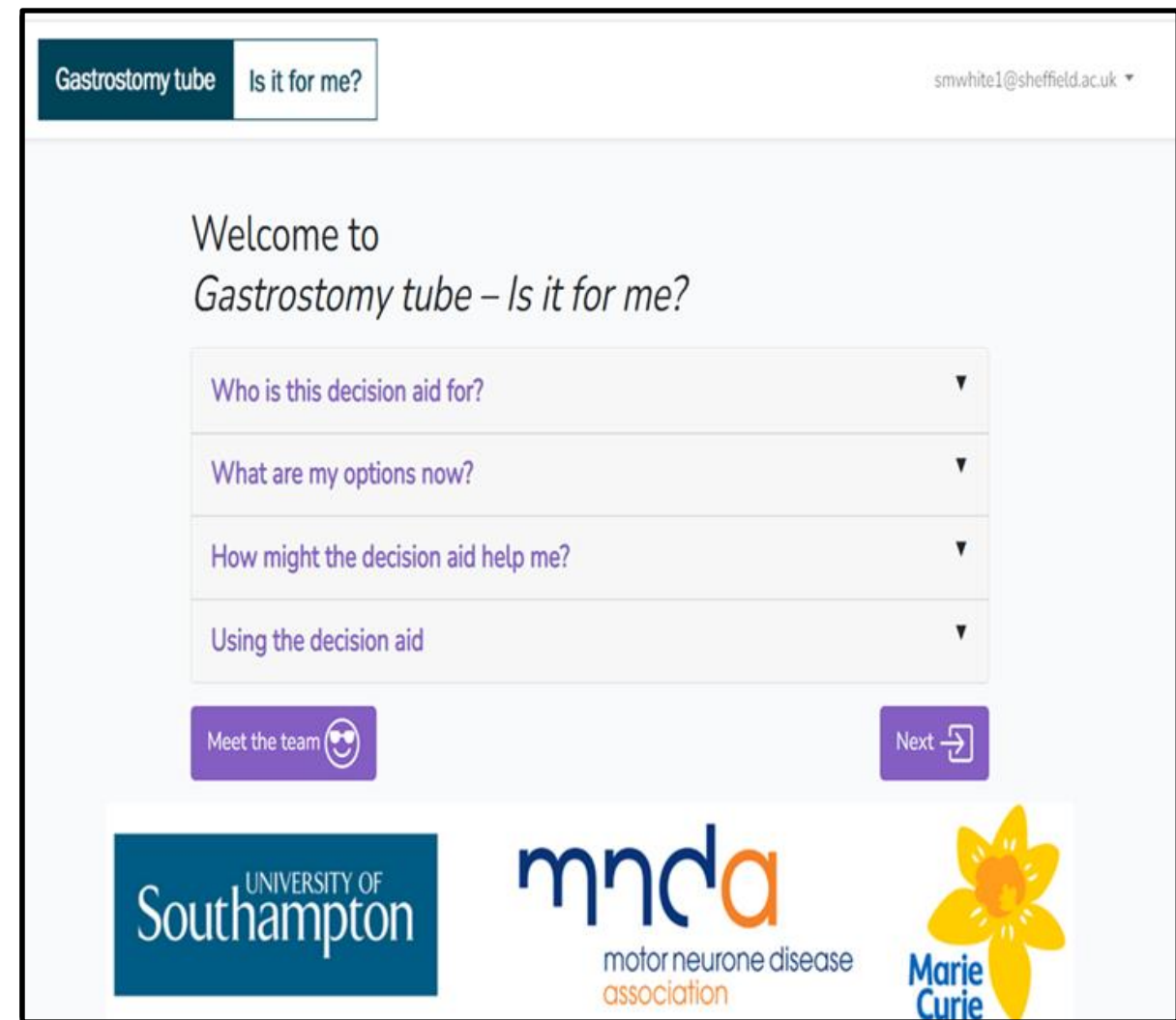


↑ Weight loss
Aspiration risk
Procedural risk
Mealtime burden

Defining the window of opportunity

- Uncertainty
- 'I will have it when I need it'
- HCP validation of 'need'
- Focus on preventing crisis
- Lack of concrete guidance





www.gastrostomychoice.co.uk

Summary

Guidelines guide practice not dictate it



Use your clinical judgement



More nuanced nutritional recommendations
required



Let's make sure dietitians are involved (leading)!!!