Comments on Recommendations/Manuscript from CALS

XXXXXX

VTE. Should it be mentioned that it be treated according to guidelines applicable to patients with other chronic conditions? Type of VTE and length of Tx

XXXXXX

Re- FVC of 65% for initiation of BIPAP: "I don't agree with isolated FVC cutoff without Sx as an indication for NIV. Don't forget that bulbar patients often have unreliable FVC values due to mouth weakness and poor seal."

Re – option of tracheostomy for secretion management: "I personally do not agree with that statement. In the rare cases where we did it, it really did not help with secretion management. I understand that it is a weak suggestion but I think we should just remove it from the recommendations and let physicians use their judgment."

Re – insertion of gastrostomy when FVC is approaching 50%: "I am not sure why we should consider gastrostomy if FVC < 50% without dysphagia. These patients will likely never need a gastrostomy unless they opt for trach, in which case it can be done at the same time. The gastrostomy would increase burden on caregivers for no good reason in my opinion. I think we should remove that 1st sentence."

- I wonder whether we should remove "prompt" or change to "approaching less than 50%"

Re – comment that RIG may be safer than PEG with respiratory insufficiency: "I agree that RIG may be safer if resp. insufficiency. But should we not also say that there is some evidence that PEG has less complications."

Re – Symptom management section: "It may sound very basic but there is no recommendation on helping patient obtain assist devices (cane, walker, wheelchair, lifts, electric beds, etc... ) for gait and other ADL management."

Re – potential treatments for patients bothered by fasciculations: "Why Gabapentin over Pregabalin or Baclofen? I would be more vague: ''medications can be considered, such as gabapentin, pregabalin, baclofen or others..''

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Is there are reason you couldn't include a statement on engaging in clinical trials/clinical research? Was it lack of articles?

XXXXX

I was wondering if you would consider adding recommendation regarding creating regional access to care and services, as such recommendation would support our advocacy efforts for obtaining resources for the patients. As it stands, the BPR states that patients with ALS should be referred to specialized ALS multidisciplinary clinics for optimized health care delivery. However, there are still regions in Canada without an ALS clinic. It is quite onerous for ALS patients in these regions who have significant disabilities and difficulty with travelling to receive all of their services at a distant ALS multidisciplinary clinics. On the other hand, the recommendation that patients with ALS should be referred to specialized ALS multidisciplinary clinics effectively relieves hospitals in such regions from their duty to create access to services that ALS patients need. A request made to the hospital administration or the ministry of health for any service for ALS patients can be easily dismissed by the assertion that ALS patients have to be referred to a specialized ALS multidisciplinary clinic where they can receive the needed resources. Therefore, I think the recommendation for patients to be referred to specialized ALS multidisciplinary clinics would be unfair to the ALS patients unless there is a parallel recommendation on creation of accessible services.

XXXXX (respirologist)

Two comments:

#1. Under the heading of 'Respiratory Management'

'There should be ongoing assessments by a specialized respiratory therapist who can optimize modes, pressures, and interfaces of NIV. Monitoring can include device download and nocturnal oximetry (level C).'

I would be more detailed here. See the attached article. I have highlighted the important information.

O'Brien D, Stavroulakis T, Baxter S, et al. The optimisation of non-invasive ventilation in amyotrophic lateral sclerosis: A systematic review. Eur Respir J 2019; in press (https://doi.org/10.1183/13993003.00261-2019).

For NIV to be effective, regular respiratory monitoring MUST be performed, which MUST include download of the device to ensure that specific targets are met. I have listed some of these below with references. One cannot assume that the implementation of NIV alone is going to modify outcome; there MUST be regular expert interaction with both the patient and data to ensure maximal benefit.

RESPIRATORY MONITORING and Targets

- Face-to face clinic evaluation
- Analyzable data from the bilevel device
- Downloaded to memory card at time of clinic visit
- Remote access via tele-monitoring (Pinto 2010; Vitacca 2016; Mansell 2018 )
- Basic guidelines

- Acceptable mask leak (Janssens 2011)
- Adequate device usage for benefit [>4 hr/24 hr] (Aboussouan 2001; Lo Coco 2006)
- Adequate delivered VT (8 mL/kg/IBW) and minimized f/VT ratio <40 (Nicholson 2017)
- Slower rise time and longer inspiratory time (Berry 2010)
- Normalize daytime PaCO2 (Tsuboi 2014) /nocturnal PtcCO2 monitoring (Aarrestad 2016; Chhajed 2016)
- Residual AHI <10 (Georges 2015)

I realize that you want to limit words here but I would, at a minimum, suggest the following:

'There MUST be ongoing assessments by a specialized respiratory therapist who can optimize modes, pressures, and interfaces of NIV. Monitoring MUST include device download and may include nocturnal oximetry (level C).'

#2. Under the heading ' Airway Clearance Management'

'Pharmacotherapy with mucolytics (guaifenesin or N-acetylcysteine), a β-receptor antagonist (metoprolol or propranolol), nebulized saline, or nebulized ipratropium can be considered (EC).'

I cannot find research with the use of a beta-blocker that benefits secretion clearance. The recent NICE guidelines do not mention this. See link below. It may have negative consequences; on the contrary beta2 agonists are frequently used. Could you direct me the evidence for this recommendation?

SLP

In the Nutritional Management section when monitoring for when to consider enteral feeding there is no criteria for dysphagia listed. While on the flow sheet it does list that when a swallow is unsafe enteral feeding could be considered. So I wonder about adding something to make the flow sheet and recommendation sheet match. Also I wondered about adding something regarding if dysphagia seems to be progressing rapidly enteral feeding could be considered?

Also in the same section there is a mention of instrumental swallowing assessments. I have come across some articles indicating that people with ALS can be at increased risk for silent aspiration (consistent with reduced pharyngeal sensation and/or reduced cough reflex) which would render a clinical assessment less effective and warrant instrumental assessment. I wonder if the team would consider adding something like 'if pharyngeal sensation impairment is suspected an instrumental assessment should be considered'. I don't have the papers handy but if it's of interest I can definitely dig them up.