

Pompe Disease: Diagnosis and Management. Evidence-Based Guidelines from a Canadian Expert Panel. Tarnopolsky et al. Can J Neurol Sci. 2016; 43: 472-485

Summary of conclusions using the Appraisal of Guidelines for REsearch & Evaluation (AGREE) Instrument (AGREE II) tool: the AGREE II tool is intended for guideline developers to conduct an internal assessment to ensure that their guidelines are sound or to evaluate guidelines from other groups for potential adaptation to their own context.

Number of Appraisers: 2

Endorsed by the Garrod Guideline committee on December 16, 2022

Summary:

The guidelines summarize the evidence (literature) but do not critically appraise it or talk about how the authors evaluated the validity of the evidence. While the authors use the Oxford center for evidence-based medicine and the international GRADE group approach to clinical guidelines to grade the quality of evidence, they do not outline which body of literature was used to provide that evidence. Patients' views were not thought of in this manuscript. It is clear this manuscript is intended for practitioners and is meant to inform them about evidence from the medical literature. It provides specific and clear recommendations, including indemnifying key recommendations for both IOPD and LOPD, and provides clear recommendations for management options. It also is clearly outlined that these are guidelines and not standards of management. External review was not done and the procedure for updating the guidelines is not discussed. Editorial independence is strong and competing interests are declared.

Summary of the AGREE II domain scores given by the appraisers:

Domain	score
Scope and purpose	20/21
Stakeholder involvement	12/21
Rigour of development	21/56
Clarity of presentation	21/21
Applicability	7/21
Editorial independence	14/14