

**Proposed recommendations for diagnosing and managing individuals with glutaric aciduria type I: second revision**

**Boy et al, JIMD 2017 Jan; 40(1): 75-101**

**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: 3**

**Endorsed by the Garrod Guideline committee on April 13, 2023**

**Summary:**

This guideline is well constructed and provides specific and clear recommendations. The recommendations are supported by evidence and the methodology for constructing the evidence is clearly defined. In-depth management tools to follow recommendations made in the guideline are provided.

Please find our evaluation based on the following domains:

1. **Scope and purpose** clearly state the objective, and the intended population and report the health questions covered by the guidelines.
2. **Stakeholders' involvement**- A representative from a support group was included but specific information or type of strategy used to identify patients' views were not included.
3. **Rigor of development**- The guideline paper is very strong in outlining the search methods, clearly indicating criteria for selection of the evidence, highlighting strength and limitation of evidence, formulation of recommendations and linking this to existing evidence, and considering the benefit and harm. The guideline is weak in addressing if an external review was implemented and the procedure for updating the guidelines is not discussed.
4. **The presentation was clear.** This guideline provides specific and clear recommendations, including indemnifying key recommendations and clear recommendations of management options.
5. **Applicability**- The appraisers found certain sections in this domain to be non-applicable in the guideline, for example, discussions around resource implications or implementations of devices or tools. The guideline does an acceptable job at discussing the facilitators, and barriers to the application of the recommendations.
6. **Editorial independence**- is strong and competing interests are declared.

Summary of the AGREE II domain scores given by the appraisers (averaged between the three appraisers):

<b>Domain</b>	<b>score</b>
Scope and purpose	21/21
Stakeholder involvement	17.3/21
Rigour of development	35.7/56
Clarity of presentation	20.6/21
Applicability	Score not determined*
Editorial independence	14/14

\*Insufficient information in the guideline to assess properly