

## QUESTION

### Should behavioural swallowing intervention/ swallow rehabilitation vs. no intervention be used for all people with Friedreich ataxia?

<b>POPULATION:</b>	all people with Friedreich ataxia
<b>INTERVENTION:</b>	behavioural swallowing intervention/ swallow rehabilitation
<b>COMPARISON:</b>	no intervention
<b>MAIN OUTCOMES:</b>	Oropharyngeal swallowing function; Swallowing related quality of life; Presence of aspiration/penetration; Nutrition/ Dehydration/ Weight Loss;

## ASSESSMENT

### Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<input type="radio"/> No <input type="radio"/> Probably no <input type="radio"/> Probably yes <input checked="" type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know	<p>Dysphagia presents in all people with FRDA at some stage during the disease (Keage et al, 2017). It can lead to serious health issues like aspiration pneumonia as well as impact quality of life through diet changes, extended mealtimes, and social isolation.</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of the topic.</p> <p>7/7 indicated dysphagia was serious.</p> <p>1/7 indicated dysphagia was probably urgent, 6/7 indicated urgent.</p> <p>1/7 indicated dysphagia was probably a priority, 6/7 indicated priority. (Aug 2020).</p>

### Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<input type="radio"/> Trivial <input type="radio"/> Small <input type="radio"/> Moderate <input type="radio"/> Large <input type="radio"/> Varies <input checked="" type="radio"/> Don't know	<p>A search of 4 databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 14 October 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	

### Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS

<ul style="list-style-type: none"> <li>○ Large</li> <li>○ Moderate</li> <li>○ Small</li> <li>○ Trivial</li> <li>○ Varies</li> <li>● Don't know</li> </ul>	<p>A search of 4 databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 14 October 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	
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## Certainty of evidence

What is the overall certainty of the evidence of effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li>○ Very low</li> <li>○ Low</li> <li>○ Moderate</li> <li>○ High</li> <li>● No included studies</li> </ul>	<p>No published evidence.</p>	

## Values

Is there important uncertainty about or variability in how much people value the main outcomes?

JUDGEMENT	RESEARCH EVIDENCE			ADDITIONAL CONSIDERATIONS															
<ul style="list-style-type: none"> <li>○ Important uncertainty or variability</li> <li>○ Possibly important uncertainty or variability</li> <li>● Probably no important uncertainty or variability</li> <li>○ No important uncertainty or variability</li> </ul>	<table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th style="width: 45%;">Outcomes</th> <th style="width: 20%;">Importance</th> <th style="width: 35%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Oropharyngeal swallowing function - not measured</td> <td>IMPORTANT<sup>a</sup></td> <td style="text-align: center;">-</td> </tr> <tr> <td>Swallowing related quality of life - not measured</td> <td>IMPORTANT<sup>a</sup></td> <td style="text-align: center;">-</td> </tr> <tr> <td>Presence of aspiration/penetration - not measured</td> <td>CRITICAL<sup>b</sup></td> <td style="text-align: center;">-</td> </tr> <tr> <td>Nutrition/ Dehydration/ Weight Loss - not measured</td> <td>IMPORTANT<sup>c</sup></td> <td style="text-align: center;">-</td> </tr> </tbody> </table> <p>a. Identified as critical (2/6), important (3/6) and low importance (1/6) by</p>			Outcomes	Importance	Certainty of the evidence (GRADE)	Oropharyngeal swallowing function - not measured	IMPORTANT <sup>a</sup>	-	Swallowing related quality of life - not measured	IMPORTANT <sup>a</sup>	-	Presence of aspiration/penetration - not measured	CRITICAL <sup>b</sup>	-	Nutrition/ Dehydration/ Weight Loss - not measured	IMPORTANT <sup>c</sup>	-	
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	JUDGEMENT						
DESIRABLE EFFECTS	Trivial	Small	Moderate	Large		Varies	Don't know
UNDESIRABLE EFFECTS	Large	Moderate	Small	Trivial		Varies	Don't know
CERTAINTY OF EVIDENCE	Very low	Low	Moderate	High			No included studies
VALUES	Important uncertainty or variability	Possibly important uncertainty or variability	<b>Probably no important uncertainty or variability</b>	No important uncertainty or variability			
BALANCE OF EFFECTS	Favors the comparison	Probably favors the comparison	Does not favor either the intervention or the comparison	Probably favors the intervention	Favors the intervention	Varies	Don't know
ACCEPTABILITY	No	Probably no	Probably yes	Yes		<b>Varies</b>	Don't know

## TYPE OF RECOMMENDATION

Strong recommendation against the intervention ○	<b>Conditional recommendation against the intervention</b> ●	Conditional recommendation for either the intervention or the comparison ○	Conditional recommendation for the intervention ○	Strong recommendation for the intervention ○
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## CONCLUSIONS

### Recommendation

For people with Friedreich ataxia, the guidelines panel suggests that behavioural therapies are not used as a treatment for dysphagia.

### Justification

There are no trials or published evidence supporting the use of behavioural therapies to improve swallow function in Friedreich ataxia.

## Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with swallowing problems.

## Research priorities

Treatments that improve dysphagia are a research priority. Behavioural therapies may prove useful for restoring swallowing function in some patients, however efficacy trials are needed.

### Reference

Keage MJ, Delatycki MB, Gupta I, Corben LA, Vogel AP. Dysphagia in Friedreich ataxia. *Dysphagia*. 2017;32(5):626-35.