

QUESTION

Should modified diet and thickened fluids vs. no diet/fluid modification be used for all people with Friedreich ataxia?

POPULATION:	all people with Friedreich ataxia
INTERVENTION:	modified diet and thickened fluids
COMPARISON:	no diet/fluid modification
MAIN OUTCOMES:	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 FA 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"> ○ Large ○ Moderate ○ Small ● Trivial ○ Varies ○ 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>	
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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"> ○ Very low ○ Low ○ Moderate ○ High ● No included studies 	<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"> ○ Important uncertainty or variability ○ Possibly important uncertainty or variability ● Probably no important uncertainty or variability ○ No important uncertainty or variability 	<table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr style="background-color: #e0e0e0;"> <th data-bbox="518 1138 1031 1222">Outcomes</th> <th data-bbox="1037 1138 1171 1222">Importance</th> <th data-bbox="1178 1138 1423 1222">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td data-bbox="518 1227 1031 1295">Oropharyngeal swallowing function - not measured</td> <td data-bbox="1037 1227 1171 1295">IMPORTANT^a</td> <td data-bbox="1178 1227 1423 1295" style="text-align: center;">-</td> </tr> <tr> <td data-bbox="518 1300 1031 1369">Swallowing related quality of life - not measured</td> <td data-bbox="1037 1300 1171 1369">IMPORTANT^a</td> <td data-bbox="1178 1300 1423 1369" style="text-align: center;">-</td> </tr> <tr> <td data-bbox="518 1373 1031 1442">Presence of aspiration/ penetration - not measured</td> <td data-bbox="1037 1373 1171 1442">CRITICAL^b</td> <td data-bbox="1178 1373 1423 1442" style="text-align: center;">-</td> </tr> <tr> <td data-bbox="518 1446 1031 1507">Nutrition/ Dehydration/ Weight loss - not measured</td> <td data-bbox="1037 1446 1171 1507">IMPORTANT^c</td> <td data-bbox="1178 1446 1423 1507" style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Oropharyngeal swallowing function - not measured	IMPORTANT ^a	-	Swallowing related quality of life - not measured	IMPORTANT ^a	-	Presence of aspiration/ penetration - not measured	CRITICAL ^b	-	Nutrition/ Dehydration/ Weight loss - not measured	IMPORTANT ^c	-	
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Presence of aspiration/ penetration - not measured	CRITICAL ^b	-															
Nutrition/ Dehydration/ Weight loss - not measured	IMPORTANT ^c	-															

	<ul style="list-style-type: none"> a. Identified as critical (2/6), important (3/6) and low importance (1/6) by people with FA and important by expert authors on this topic b. Identified as critical (3/6), important (1/6), low importance (2/6) by people with FA and critical by expert authors on this topic c. Identified as critical (2/6), important (2/6), low importance (1/6), and requiring more information (1/6) by people with FA and critical by expert authors on this topic 	
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Balance of effects

Does the balance between desirable and undesirable effects favor the intervention or the comparison?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> Favors the comparison <input type="radio"/> Probably favors the comparison <input type="radio"/> Does not favor either the intervention or the comparison <input type="radio"/> Probably favors the intervention <input type="radio"/> Favors the intervention <input type="radio"/> Varies <input checked="" type="radio"/> Don't know 	No published evidence.	

Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> No <input type="radio"/> Probably no <input type="radio"/> Probably yes <input type="radio"/> Yes <input checked="" type="radio"/> Varies <input type="radio"/> Don't know 	No published evidence.	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if the intervention was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>3/5 indicated modified diet and thickened fluids for people with swallowing difficulties was probably reasonable, 2/5 indicated reasonable. (Aug 2020).</p>

SUMMARY OF JUDGEMENTS

JUDGEMENT

	JUDGEMENT						
PROBLEM	No	Probably no	Probably yes	Yes		Varies	Don't know
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	Probably no important uncertainty or variability	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		Varies	Don't know

TYPE OF RECOMMENDATION

Strong recommendation against the intervention <input type="radio"/>	Conditional recommendation against the intervention <input type="radio"/>	Conditional recommendation for either the intervention or the comparison <input checked="" type="radio"/>	Conditional recommendation for the intervention <input type="radio"/>	Strong recommendation for the intervention <input type="radio"/>
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CONCLUSIONS

Recommendation

For people with Friedreich ataxia, we cannot recommend either modified diets/thickened fluids or no diet modification to improve swallow safety.

Justification

There are no data supporting the use of modified diet in Friedreich Ataxia.

Subgroup considerations

This recommendation is considered appropriate for people with Friedreich ataxia with moderate to severe dysphagia.

Research priorities

Large adequately powered randomised controlled trials comparing modified diet and thickened fluids to alternative approaches are required. The primary outcome measure needs careful consideration, focusing on patient identified priorities.

Reference

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