

QUESTION

Should Augmentative and Alternative Communication (AAC) vs. no AAC be used for all people with Friedreich ataxia?

POPULATION:	all people with Friedreich ataxia
INTERVENTION:	Augmentative and Alternative Communication (AAC)
COMPARISON:	no AAC
MAIN OUTCOMES:	Quality of life; Safety ; Communication Effectiveness; Barriers and Facilitators of AAC; Functional Communication Ability;

ASSESSMENT

Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> No <input type="radio"/> Probably no <input checked="" type="radio"/> Probably yes <input type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 		<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 dysarthria was serious.</p> <p>1/7 indicated dysarthria was probably not urgent, 1/7 indicated probably urgent, 4/7 indicated urgent, 1/7 indicated varies/sometimes urgent.</p> <p>2/7 indicated dysarthria was probably a priority, 5/7 indicated priority. (Aug 2020).</p>

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> Trivial <input type="radio"/> Small <input type="radio"/> Moderate <input type="radio"/> Large <input checked="" type="radio"/> Varies <input type="radio"/> Don't know 	<p>A search of four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 12 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 four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 12 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> <th style="text-align: center;">Outcomes</th> <th style="text-align: center;">Importance</th> <th style="text-align: center;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td style="text-align: center;">Quality of life - not measured</td> <td style="text-align: center;">IMPORTANT^a</td> <td style="text-align: center;">-</td> </tr> <tr> <td style="text-align: center;">Safety - not measured</td> <td style="text-align: center;">CRITICAL^b</td> <td style="text-align: center;">-</td> </tr> <tr> <td style="text-align: center;">Communication Effectiveness - not measured</td> <td style="text-align: center;">CRITICAL^c</td> <td style="text-align: center;">-</td> </tr> <tr> <td style="text-align: center;">Barriers and Facilitators of AAC - not measured</td> <td style="text-align: center;">IMPORTANT^d</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Quality of life - not measured	IMPORTANT ^a	-	Safety - not measured	CRITICAL ^b	-	Communication Effectiveness - not measured	CRITICAL ^c	-	Barriers and Facilitators of AAC - not measured	IMPORTANT ^d	-	
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	<table border="1" data-bbox="520 110 1417 180"> <tr> <td data-bbox="520 110 1018 180">Functional Communication Ability - not measured</td> <td data-bbox="1018 110 1167 180">IMPORTANT^e</td> <td data-bbox="1167 110 1417 180">-</td> </tr> </table> <p data-bbox="562 220 1369 477"> a. Identified as critical (3/6) and important (3/6) by people with FA and important by expert authors on this topic. b. Identified as critical (3/6) and important (3/6) by people with FA and critical by expert authors on this topic. c. Identified as critical (4/6) and important (2/6) by people with FA and important by expert authors on this topic. d. Identified as critical (2/6), important (3/6) and not important (1/6) by people with FA and important by expert authors on this topic. e. Identified as critical (2/6) and important (4/6) by people with FA and important by expert authors on this topic. </p>	Functional Communication Ability - not measured	IMPORTANT ^e	-	
Functional Communication Ability - not measured	IMPORTANT ^e	-			

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 data-bbox="1436 1230 1976 1338">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 data-bbox="1436 1365 2003 1414">3/4 indicated Augmentative and Alternative Communication was probably reasonable, 1/4 indicated reasonable. (Aug 2020).</p>

SUMMARY OF JUDGEMENTS

	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 ○	Conditional recommendation against the intervention ○	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

We cannot recommend either the use or non-use of AAC to treat dysarthria in individuals with Friedreich ataxia.

Justification

There are no trials or published evidence supporting the use of AAC in Friedreich ataxia.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with dysarthria. AAC may be best suited to individuals who have severe dysarthria and are not receptive to behavioural or pharmacological interventions.

Research priorities

Treatments that improve dysarthria are a research priority. Alternative and augmentative communication devices or approaches may prove useful for restoring communicative capacity in some patients, however efficacy trials are needed.