

QUESTION

Should intensive Behavioural Speech Intervention vs. no treatment be used for all people with Friedreich ataxia?

POPULATION:	all people with Friedreich ataxia
INTERVENTION:	intensive Behavioural Speech Intervention
COMPARISON:	no treatment
MAIN OUTCOMES:	Intelligibility; Naturalness; Breath support; Vocal quality; Loudness; Speech and voice related quality of life; Communication participation;

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>Dysarthria remains a significant and important issue for people with FA. Speech disorder is prevalent across the disease course, gradually worsening as the disease progresses. Traditional behavioural speech therapy programs may be effective in improving speech in FA.</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 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														
<input type="radio"/> Trivial <input type="radio"/> Small <input checked="" type="radio"/> Moderate <input type="radio"/> Large <input type="radio"/> Varies <input type="radio"/> Don't know	<table border="1"> <thead> <tr> <th rowspan="2">Outcomes</th> <th rowspan="2">No of participants (studies) Follow-up</th> <th rowspan="2">Certainty of the evidence (GRADE)</th> <th rowspan="2">Relative effect (95% CI)</th> <th colspan="2">Anticipated absolute effects* (95% CI)</th> </tr> <tr> <th>Risk with no treatment</th> <th>Risk difference with intensive Behavioural Speech Intervention</th> </tr> </thead> <tbody> <tr> <td>Intelligibility assessed with:</td> <td>48 (3)</td> <td>⊕○○○</td> <td>-</td> <td colspan="2">21 patients with Friedreich ataxia (n=12) spinocerebellar ataxia type 6</td> </tr> </tbody> </table>	Outcomes	No of participants (studies) Follow-up	Certainty of the evidence (GRADE)	Relative effect (95% CI)	Anticipated absolute effects* (95% CI)		Risk with no treatment	Risk difference with intensive Behavioural Speech Intervention	Intelligibility assessed with:	48 (3)	⊕○○○	-	21 patients with Friedreich ataxia (n=12) spinocerebellar ataxia type 6		
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Direct Magnitude Estimation	observational studies) ^{1,2,3}	Very low ^{a,b}		<p>(n=1), idiopathic cerebellar ataxia (n=1), spastic paraplegia 7 (n=1) underwent Lee Silverman Voice Treatment (LSVT LOUD) via Skype, 2 sessions/week for 8 weeks. There were 4 assessment points - 2 sessions 2 weeks apart prior to treatment, and 2 post-therapy assessments: 1 week and 8 weeks post treatment. Friedman tests did not identify any significant changes over time in intelligibility ($p=0.813$). (Lowit et al 2020). 7 individuals with ARSACS underwent 4 week home based treatment trial. Assessments were completed at 3 timepoints: A1 (4 weeks prior to training), A2 (immediately before training), A3 (immediately after training). Friedman tests as used to determine within group differences between A1 and A3. DME score remained unchanged before training but increased significantly after training ($p=0.024$). (Vogel et al 2019). 20 people with ARSACS, FRDA or SCA completed a 4-week intensive speech rehabilitation program. Mean (SD) intelligibility was 98.0 (29.8) points at baseline and 119.5 (25.4) 4-weeks post-baseline with Pearson's correlation coefficient of 0.9. The relative (%) increase in speech intelligibility from baseline to post-treatment was statistically significant (geometric mean of the post-treatment/baseline ratio 1.25, 95% CI [1.16, 1.35], $p<0.0001$) using a two-sided paired t-test on base-2 logtransformed outcome data. (Vogel et al 2018).</p>
Naturalness assessed with: Direct Magnitude Estimation	21 (1 observational study) ¹	⊕○○○ Very low ^c	-	<p>21 patients with Friedreich ataxia (n=18), spinocerebellar ataxia type 6 (n=1), idiopathic cerebellar ataxia (n=1), spastic paraplegia 7 (n=1) underwent Lee Silverman Voice Treatment (LSVT LOUD) via Skype, 2 sessions/week for 8 weeks. There were</p>

				4 assessment points - 2 sessions 2 weeks apart prior to treatment, and 2 post-therapy assessments: 1 week and 8 weeks post treatment. Friedman tests did not identify any significant changes over time in naturalness ($p=0.989$). (Lowit et al 2020).	
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Loudness - not	-	-	-	-	-

reported				
Speech and voice related quality of life assessed with: Voice Handicap Index	21 (1 observational study) ¹	⊕○○○ Very low ^c	-	21 patients with Friedreich ataxia (n=18), spinocerebellar ataxia type 6 (n=1), idiopathic cerebellar ataxia (n=1), spastic paraplegia 7 (n=1) underwent Lee Silverman Voice Treatment (LSVT LOUD) via Skype, 2 sessions/week for 8 weeks. There were 4 assessment points - 2 sessions 2 weeks apart prior to treatment, and 2 post-therapy assessments: 1 week and 8 weeks post treatment. There were no significant differences in the VHI between pre and post-treatment sessions ($p=0.056$). On the other hand, posttreatment interviews indicated that the majority of participants felt their communication had improved after treatment. (Lowit et al 2020).
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- a. Low absolute numbers of participants and events, and only one study reported confidence intervals.

- b. One study had no follow up period for evaluation, one study was a conference abstract and there was no control group for any of the studies.
- c. No confidence intervals reported with low absolute numbers of participants and events.

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE					ADDITIONAL CONSIDERATIONS												
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Communication participation	21 (1	⊕○○○	-	21 patients with Friedreich ataxia (n=18) spinocerebellar ataxia type 6

	assessed with: Communication Participation Item Bank	observational study) ¹	Very low ^c	(n=1), idiopathic cerebellar ataxia (n=1), spastic paraplegia 7 (n=1) underwent Lee Silverman Voice Treatment (LSVT LOUD) via Skype, 2 sessions/week for 8 weeks. There were 4 assessment points - 2 sessions 2 weeks apart prior to treatment, and 2 post-therapy assessments: 1 week and 8 weeks post treatment. There were no significant differences in the CBIP between pre and post-treatment sessions ($p=0.154$). (Lowit et al 2019).	
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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"> <input type="radio"/> Very low <input checked="" type="radio"/> Low <input type="radio"/> Moderate <input type="radio"/> High <input type="radio"/> No included studies 	Low certainty of the evidence of effects as per the evidence profile table.	

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="width: 50%;">Outcomes</th> <th style="width: 20%;">Importance</th> <th style="width: 30%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Intelligibility assessed with: Direct Magnitude Estimation</td> <td>IMPORTANT^a</td> <td>⊕○○○ VERY LOW^{b,c}</td> </tr> <tr> <td>Naturalness assessed with: Direct Magnitude Estimation</td> <td>IMPORTANT^d</td> <td>⊕○○○ VERY LOW^e</td> </tr> <tr> <td>Breath support assessed with: Vowel prolongation</td> <td>IMPORTANT^a</td> <td>⊕○○○ VERY LOW^e</td> </tr> <tr> <td>Vocal quality assessed with: Grade Roughness Breathiness Asthenia Strain</td> <td>IMPORTANT^a</td> <td>⊕○○○ VERY LOW^e</td> </tr> <tr> <td>Loudness - not reported</td> <td>IMPORTANT^f</td> <td>-</td> </tr> <tr> <td>Speech and voice related quality of life assessed with: Voice Handicap Index</td> <td>IMPORTANT^g</td> <td>⊕○○○ VERY LOW^e</td> </tr> <tr> <td>Communication participation assessed with: Communication Participation Item Bank</td> <td>IMPORTANT^d</td> <td>⊕○○○ VERY LOW^e</td> </tr> </tbody> </table> <p style="margin-top: 20px;"> a. Identified as critical (2/6) and important (4/6) by people with FA and important by expert authors on this topic b. Low absolute numbers of participants and events, and only one study reported confidence intervals. c. One study had no follow up period for evaluation, one study was a conference abstract and there was no control group for any of the studies. d. Identified as critical (1/6), important (4/6) and low importance (1/6) by people with FA and important by expert authors on this topic e. No confidence intervals reported with low absolute numbers of participants and events. f. Identified as critical (1/6) and important (5/6) by people with FA and important by expert authors on this topic </p>	Outcomes	Importance	Certainty of the evidence (GRADE)	Intelligibility assessed with: Direct Magnitude Estimation	IMPORTANT ^a	⊕○○○ VERY LOW ^{b,c}	Naturalness assessed with: Direct Magnitude Estimation	IMPORTANT ^d	⊕○○○ VERY LOW ^e	Breath support assessed with: Vowel prolongation	IMPORTANT ^a	⊕○○○ VERY LOW ^e	Vocal quality assessed with: Grade Roughness Breathiness Asthenia Strain	IMPORTANT ^a	⊕○○○ VERY LOW ^e	Loudness - not reported	IMPORTANT ^f	-	Speech and voice related quality of life assessed with: Voice Handicap Index	IMPORTANT ^g	⊕○○○ VERY LOW ^e	Communication participation assessed with: Communication Participation Item Bank	IMPORTANT ^d	⊕○○○ VERY LOW ^e	
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	g. Identified as critical (3/6) and important (3/6) by people with FA and important 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 checked="" type="radio"/> Probably favors the intervention <input type="radio"/> Favors the intervention <input type="radio"/> Varies <input type="radio"/> Don't know 	On balance is it unclear whether evidence favours either the intervention or the comparison. There is some preliminary evidence that some changes to some aspects of speech can be improved with behavioural therapy; however, the quality of that evidence is weak.	

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>4/4 indicated Intensive Behavioural Speech intervention for all people with FA was reasonable. (Aug 2020).</p>

SUMMARY OF JUDGEMENTS

PROBLEM	JUDGEMENT						
	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

JUDGEMENT							
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

For people with Friedreich ataxia, we suggest the use of targeted intensive behavioural therapy for improving speech in individuals with dysarthria.

Justification

There is some limited evidence supporting the use of behavioural therapies for improving speech in FRDA. Three small non-randomised trials show some preliminary evidence that some aspects of speech can improve in hereditary ataxias following intensive therapy (Vogel et al, 2018; Vogel et al, 2019; Lowit et al, 2020). These therapies include methods for improving self-monitoring, biofeedback and diverse tasks designed to improve specific aspects of speech like breath support, vocal control and intelligibility. Data suggest that some gains in intelligibility or voice quality can be achieved with intensive treatment. However, data are derived from non-controlled, underpowered pilot studies.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with dysarthria.

Research priorities

Large adequately powered randomised controlled trials comparing intensively delivered therapies compared to alternative treatments are required. The primary outcome measure needs careful consideration, focussing on patient identified priorities.

References

Lowit A, Egan A, Hadjivassiliou M. Feasibility and acceptability of Lee Silverman voice treatment in progressive ataxias. *Cerebellum*. 2020;19(5):701-14.

Vogel A, Rommel N, Synofzik M. Intensive speech rehabilitation in degenerative ataxias improves intelligibility (Abstract). *Mov Disord*. 2018;33(Suppl 2):S281.

Vogel AP, Stoll LH, Oettinger A, et al. Speech treatment improves dysarthria in multisystemic ataxia: a rater-blinded, controlled pilot-study in ARSACS. *J Neurol*. 2019;266(5):1260-6.