

QUESTION

Should remote-microphone (FM) listening devices vs. no devices be used for all people with Friedreich ataxia?

POPULATION:	all people with Friedreich ataxia
INTERVENTION:	remote-microphone (FM) listening devices
COMPARISON:	no devices
MAIN OUTCOMES:	Speech perception in background noise; Everyday listening/communication; Hearing/ communication-related quality of life; Hearing/ communication-related stress/anxiety; Hearing/ communication-related fatigue;

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 type="radio"/> Probably yes <input checked="" type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>A review of clinical features in FRDA (beyond afferent ataxia) revealed that 11% of 650 affected individuals reported significant 'hearing loss' (Reetz et al., 2018).</p> <p>A group of 10 individuals with FRDA reported high levels of listening/communication difficulty in everyday listening circumstances on a Hearing Disability questionnaire (Abbreviated Profile of Hearing Aid Benefit), with significantly higher (worse) scores than matched controls (Rance et al., 2010).</p> <p>References</p> <p>Rance G, Corben LA, Du Bourg E, et al. Successful treatment of auditory perceptual disorder in individuals with Friedreich ataxia. <i>Neuroscience</i> 2010;171(2):552-5.</p> <p>Reetz K, Dogan I, Hohenfeld C, et al. Nonataxia symptoms in Friedreich Ataxia: Report from the Registry of the European Friedreich's Ataxia Consortium for Translational Studies (EFACTS). <i>Neurology</i> 2018;91(10):e917-e30.</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>2/7 indicated disturbance of audiological function was probably serious, 5/7 indicated serious.</p> <p>1/7 indicated disturbance of audiological function was probably not urgent, 2/7 indicated probably urgent, 4/7 indicated urgent.</p> <p>1/7 indicated disturbance of audiological function was probably not a priority, 2/7 indicated probably a priority, 4/7 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 checked="" type="radio"/> Moderate <input type="radio"/> Large <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>A group of 10 individuals showed significantly improved speech perception in background noise with the provision of a remote microphone (FM) listening system. They also reported improved everyday listening and communication when wearing the device over the course of a six-week trial (Rance et al., 2010).</p>	<p>Mean speech perception in noise and self-reported Hearing Disability ratings did not improve to normal levels, but did show significant improvements compared to the unaided condition.</p>

Outcomes	No of participants (studies) Follow-up	Certainty of the evidence (GRADE)	Relative effect (95% CI)	Anticipated absolute effects* (95% CI)	
				Risk with no devices	Risk difference with remote-microphone (FM) listening devices
Speech perception in background noise assessed with: Consonant-Nucleus-Consonant word test	10 (1 observational study) ¹	⊕○○○ Very low ^{a,b}	-	10 people with Friedreich ataxia completed a battery of auditory function tests and the results were compared with a cohort of matched controls. The FA subjects were then fitted with personal FM-listening devices and evaluated over a 6-week period. Speech perception in noise was poorer in the FRDA listeners than controls. Mean phoneme score (the percentage of speech-sounds correctly imitated) for the FRDA group was 42.5 ± 26.1% and for the control group was 80.1 ± 1.8% (T=-4.53, p<0.005). Importantly, perceptual ability was significantly improved in the FRDA listeners when wearing the FM device (T=-4.87, p<0.005). (Rance et al 2010).	
Everyday listening/communication assessed with: Abbreviated profile of hearing aid benefit	10 (1 observational study) ¹	⊕○○○ Very low ^{a,b}	-	10 people with Friedreich ataxia completed a battery of auditory function tests and the results were compared with a cohort of matched controls. The FA subjects were then fitted with personal FM-listening devices and evaluated over a 6-week period. The mean overall APHAB score (representing the proportion of situations in which the individual perceived a difficulty) was 40.7 ± 11.5%.	

In contrast, the mean APHAB score for the control group was significantly lower: $8.3 \pm 4.5\%$ ($T=8.16, p<0.001$). FM device use produced significant listening and communication improvements for the FRDA cohort. APHAB scores across the four data collection points were; Unaided1: $40.2 \pm 11.5\%$; Aided1: $20.6 \pm 7.5\%$; Aided2: $17.1 \pm 5.2\%$; Unaided2: $38.3 \pm 12.1\%$. Analysis of variance showed a significant group difference ($F(3,36)=15.61, p<0.001$). Post hoc analysis found that both aided data points were significantly lower than the unaided ($p<0.05$). (Rance et al 2010).

Hearing/ communication-related quality of life - not measured	-	-	-	-	-
Hearing/ communication-related stress/anxiety - not measured	-	-	-	-	-
Hearing/ communication-related fatigue - not measured	-	-	-	-	-

1. Rance G, Corben LA, Du Bourg E, King A, Delatycki MB. Successful treatment of auditory perceptual disorder in individuals with Friedreich ataxia. *Neuroscience*; 2010.
 - a. Small sample size.
 - b. Confidence intervals not reported.

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE					ADDITIONAL CONSIDERATIONS																				
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 - a. Small sample size.

	b. Confidence intervals not reported.	
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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 	Very 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"> <input type="radio"/> Important uncertainty or variability <input type="radio"/> Possibly important uncertainty or variability <input checked="" type="radio"/> Probably no important uncertainty or variability <input type="radio"/> 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;">Speech perception in background noise assessed with: Consonant-Nucleus-Consonant word test</td> <td style="text-align: center;">CRITICAL^a</td> <td style="text-align: center;">⊕○○○ VERY LOW^{b,c}</td> </tr> <tr> <td style="text-align: center;">Everyday listening/communication assessed with: Abbreviated profile of hearing aid benefit</td> <td style="text-align: center;">IMPORTANT^d</td> <td style="text-align: center;">⊕○○○ VERY LOW^{b,c}</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Speech perception in background noise assessed with: Consonant-Nucleus-Consonant word test	CRITICAL ^a	⊕○○○ VERY LOW ^{b,c}	Everyday listening/communication assessed with: Abbreviated profile of hearing aid benefit	IMPORTANT ^d	⊕○○○ VERY LOW ^{b,c}	
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	<p>a. Identified as low importance (1/6) and critical (5/6) by people with FA and critical by expert authors on this topic</p> <p>b. Small sample size.</p> <p>c. Confidence intervals not reported.</p> <p>d. Identified as low importance (2/6) and important (4/6) by people with FA and critical by expert authors on this topic</p> <p>e. Identified as low importance (1/6), important (3/6), critical (2/6) by people with FA and critical by expert authors on this topic</p> <p>f. Identified as low importance (1/6), important (4/6), critical (1/6) by people with FA and critical by expert authors on this topic</p> <p>g. Identified as low importance (2/6), important (3/6), critical (1/6) by people with FA and important by expert authors on this topic</p>										

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 		

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 checked="" type="radio"/> Probably yes <input type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>Limited published evidence.</p> <p>Rance et al. (2010) demonstrated that remote-microphone devices were acceptable to each of their study participants in a 6-week take-home trial. All wore the systems for at least 1 to 5 hours per day across the trial.</p>	<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>1/5 indicated remote-microphone listening devices for all people with hearing impairment were probably reasonable, 4/5 indicated didn't know if reasonable. (Aug 2020).</p>
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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

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

Recommendation

We suggest using remote-microphone listening devices over not using any devices for individuals with Friedreich ataxia with auditory deficits.

Justification

The management of auditory deficits was identified by the Patient and Parent Advisory Panel as a priority. Published evidence indicates that the prevalence of hearing difficulties is higher in people with FRDA compared to the general population.

Available evidence suggests that remote-microphone listening systems can improve speech perception in everyday (noisy) listening circumstances and aid communication.

Subgroup considerations

Developmental issues in pediatric populations should be considered: hearing impairment in children can affect speech, language, social and academic development.

Non-ambulant individuals are potentially less able to control listening/communication situations; for example, physical distance from the speaker and from noise sources in the environment. As such, they are likely to derive greater device benefit as the signal provided by the system is consistent regardless of the distance between speaker and listener.

Research priorities

Randomised controlled trials to assess the efficacy of hearing devices in individuals with FRDA.

Studies of the acceptability of auditory devices in individuals with FRDA.