

QUESTION

Should amplification vs. cochlear implants be used for all people with Friedreich ataxia?

POPULATION:	all people with Friedreich ataxia
INTERVENTION:	amplification
COMPARISON:	cochlear implants
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 checked="" type="radio"/> Probably yes <input 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>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 checked="" type="radio"/> Small <input type="radio"/> Moderate <input type="radio"/> Large <input 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 11 September 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p> <p>Two case studies: 1 child (Miyamoto et al 1999) and 1 adult (Frewin et al 2013) with mild/moderate hearing loss. Both showed no perceptual benefit with amplification despite aidable levels of residual hearing. This is consistent with reported outcomes for individuals with auditory neuropathy due to other aetiologies (Rance & Starr, 2015).</p> <p>With respect to the comparison for the intervention, cochlear Implant outcomes in these cases were</p>	<p>Consistent with hearing aid outcomes reported for auditory neuropathy where sound distortion is typically the limiting factor. That is, users may be able to detect speech at normal levels with amplification, but obtain little functional benefit (Rance and Starr, 2015).</p>

	<p>variable:</p> <p>Miyamoto et al 1999 (child): no improvement on pre-operative (hearing aid) speech perception performance levels.</p> <p>Frewin et al, 2013 (adult): significant perceptual improvement with cochlear implant.</p>	
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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 11 September 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	<p>Amplification</p> <p>The risk of undesirable effects with correctly fitted amplification is low. Patients may experience some discomfort if sounds are over-amplified</p> <p>Cochlear Implantation</p> <p>Miyamoto et al (1999) case highlights the possibility that some individuals with FRDA will show no post-operative benefit.</p> <p>CI outcomes for other aetiologies involving neurodegenerative disease and central auditory neural deficits have been variable. (Rance & Starr, 2015). Currently no pre-operative predictors of outcome in affected individuals.</p>

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>Limited 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" data-bbox="520 321 1415 846"> <thead> <tr> <th data-bbox="520 321 1066 435">Outcomes</th> <th data-bbox="1066 321 1199 435">Importance</th> <th data-bbox="1199 321 1415 435">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td data-bbox="520 435 1066 505">Speech perception in background noise - not measured</td> <td data-bbox="1066 435 1199 505">CRITICAL^a</td> <td data-bbox="1199 435 1415 505">-</td> </tr> <tr> <td data-bbox="520 505 1066 574">Everyday listening/communication - not measured</td> <td data-bbox="1066 505 1199 574">IMPORTANT^b</td> <td data-bbox="1199 505 1415 574">-</td> </tr> <tr> <td data-bbox="520 574 1066 675">Hearing/communication-related quality of life - not measured</td> <td data-bbox="1066 574 1199 675">IMPORTANT^c</td> <td data-bbox="1199 574 1415 675">-</td> </tr> <tr> <td data-bbox="520 675 1066 776">Hearing/communication-related stress/anxiety - not measured</td> <td data-bbox="1066 675 1199 776">IMPORTANT^d</td> <td data-bbox="1199 675 1415 776">-</td> </tr> <tr> <td data-bbox="520 776 1066 846">Hearing/communication-related fatigue - not measured</td> <td data-bbox="1066 776 1199 846">IMPORTANT^e</td> <td data-bbox="1199 776 1415 846">-</td> </tr> </tbody> </table> <p data-bbox="562 883 1415 1143"> a. Identified as critical (5/6) and low importance (1/6) by people with FA and critical by expert authors on this topic. b. Identified as important (4/6) and low importance (2/6) by people with FA and critical by expert authors on this topic. c. Identified as critical (2/6), important (3/6) and low importance (1/6) by people with FA and critical by expert authors on this topic d. Identified as critical (1/6), important (4/6) and low importance (1/6) by people with FA and critical by expert authors on this topic. e. Identified as critical (1/6), important (3/6) and low importance (2/6) by people with FA and important by expert authors on this topic. </p>	Outcomes	Importance	Certainty of the evidence (GRADE)	Speech perception in background noise - not measured	CRITICAL ^a	-	Everyday listening/communication - not measured	IMPORTANT ^b	-	Hearing/communication-related quality of life - not measured	IMPORTANT ^c	-	Hearing/communication-related stress/anxiety - not measured	IMPORTANT ^d	-	Hearing/communication-related fatigue - not measured	IMPORTANT ^e	-	
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Hearing/communication-related fatigue - 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
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<ul style="list-style-type: none"> ○ 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 	No published evidence.	
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Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ No ○ Probably no ○ Probably yes ○ Yes ○ Varies ● Don't know 	Limited 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>2/5 indicated devices to amplify sound for all people with a hearing impairment were probably reasonable, 1/5 indicated reasonable, 2/5 indicated didn't know if 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 <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

We cannot recommend the use of amplification devices over cochlear implants 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.

There is very limited evidence related to the efficacy of either amplification devices or cochlear implants in FRDA populations, but findings in other groups with auditory neuropathy due to neurodegenerative disease suggest that outcomes with the use of amplification devices are poor and outcomes with cochlear implants are variable.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with auditory deficits. Developmental issues in pediatric populations with auditory deficits should be considered: hearing impairment in children can affect speech, language, social and academic development.

Research priorities

Randomised controlled trials to assess the efficacy of amplification devices or cochlear implants in individuals with FRDA.

Studies of the acceptability of amplification devices and cochlear implants in individuals with FRDA.

References

Frewin B, Chung M, Donnelly N. Bilateral cochlear implantation in Friedreich's ataxia: a case study. *Cochlear Implants Int.* 2013;14(5):287-90.

Miyamoto RT, Kirk KI, Renshaw J, Hussain D. Cochlear implantation in auditory neuropathy. *Laryngoscope*. 1999;109(2 Pt 1):181-5.

Rance G, Corben LA, Du Bourg E, King A, Delatycki MB. Successful treatment of auditory perceptual disorder in individuals with Friedreich ataxia. *Neuroscience*. 2010;171(2):552-5.

Rance G, Starr A. Pathophysiological mechanisms and functional hearing consequences of auditory neuropathy. *Brain*. 2015;138(Pt 11):3141-58.

Reetz K, Dogan I, Hohenfeld C, Didszun C, Giunti P, Mariotti C, et al. Nonataxia symptoms in Friedreich Ataxia: Report from the Registry of the European Friedreich's Ataxia Consortium for Translational Studies (EFACTS). *Neurology*. 2018;91(10):e917-e30.