QUESTION

Should hearing aids (including cochlear implants) vs. no hearing aids or cochlear implants be used for all people with Friedreich ataxia?	
POPULATION:	all people with Friedreich ataxia
INTERVENTION:	hearing aids (including cochlear implants)
COMPARISON:	no hearing aids or cochlear implants
MAIN OUTCOMES:	Speech perception in background noise; Speech perception in background noise; Speech perception in background noise; Speech perception in background noise; Speech perception in background noise; Speech perception in background noise; Everyday listening/communication; Everyday listening/communication; Everyday listening/communication; Everyday listening/communication; Everyday listening/communication ; Everyday listening and communication; Hearing/communication-related quality of life; Hearing/communication-related stress/anxiety; Hearing/communication-related fatigue; Speech perception in background quiet; Speech perception in background quiet;

ASSESSMENT

Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ● Probably yes ○ Yes ○ Varies ○ Don't know	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). 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).	The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of the topic. 2/7 indicated disturbance of audiological function was probably serious, 5/7 indicated serious. 1/7 indicated disturbance of audiological function was probably not urgent, 2/7 indicated probably urgent, 4/7 indicated urgent. 1/7 indicated disturbance of audiological function was probably not a priority, 2/7 indicated probably a priority, 4/7 indicated priority. (Aug 2020).

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ Trivial ● Small ○ Moderate ○ Large ○ Varies ○ Don't know	**Hearing Aids** 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).	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). Auditory neuropathy is caused by a range of different

Cochlear Implants

Two case studies:

7-year-old: no useful hearing post CI

41-year-old: near normal speech perception (in quiet) post implantation.

Variability is consistent with reported CI outcomes in individuals with central auditory disorders (Rance & Starr, 2015).

Outcomes	№ of participants (studies) Follow-up	Certainty of the evidence (GRADE)	Relative effect (95% CI)	Anticipated absolute effects* (95% CI)	
				Risk with no hearing aids or cochlear implants	Risk difference with hearing aids (including cochlear implants)
Speech perception in background noise assessed with: The Glendonald Auditory Screening Procedure	10 (1 observational study)[1]	⨁◯◯◯ Very low[a,b,c]	-	10 children with auditory neuropathy were treated with cochlear implants (CI). Speech perception tests were completed on the day of the surgery and then again at least 1 year post experience with the implant. In The Glendonald Auditory Screening Procedure (GASP) analysis (p=0.002), there was a statistically significant improvement with the use of CI. (de Carvalho et al 2016).	
Speech perception in background noise assessed with: Hochmair-Schulz-Moser (HSM) sentence test	32 (1 observational study)[2]	⨁◯◯◯ Very low[a,b,c,d]	-	32 children with auditory neuropathy spectrum disorder provided with hearing aids (HA) or cochlear implants (CI) were assessed to determine the outcomes of the different therapies. The Mann-Whitney U test was used to compare	

pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015).

Negligible importance as the paper (de Carvalho et al 2016) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)

We might predict CI outcomes in FRDA based on findings from other diseases with a similar site of lesion (auditory nerve), but this would require detailed aetiological information which this paper – does not provide.

Negligible importance as the paper (Ehrmann-Muller et al 2019) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)

Negligible importance as the paper (Ehrmann-Muller et al 2019) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)

				speech discrimination of HA and CI groups. With the Hochmair-Schulz-Moser (HSM) sentence test at 65 dB SPL (sound pressure level), 75% of the children with a CI achieved a speech discrimination in noise score of at least 60% at a signal to noise ratio (SNR) of 5, and four scored 80% or higher. (Ehrmann-Muller et al 2019).	Negligible importance as the paper (Ji et al 2015) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)
Speech perception in background noise assessed with: Freiburgers' monosyllabic word test	32 (1 observational study)[2]	⨁◯◯◯ Very low[a,b,c,d]	-	32 children with auditory neuropathy spectrum disorder provided with hearing aids (HA) or cochlear implants (CI) were assessed to determine the outcomes of the different therapies. The Mann-Whitney U test was used to compare speech discrimination of HA and CI groups. A comparison of speech discrimination tested with a CI (n = 6) or a HA (n = 6) in the Freiburgers' monosyllabic test revealed that the former tended to produce better outcomes. This difference was not significant (p=0.46), probably due to the small sample size. (Ehrmann-Muller et al 2019).	Negligible importance as the paper (Ji et al 2015) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)
Speech perception in background noise assessed with: Mandarin Early Speech Perception test	8 (1 observational study)[3]	⨁◯◯◯ Very low[a,b,c,e]	-	8 children with auditory neuropathy received CIs and were assessed to evaluate its benefits. Assessments were conducted pre and post operatively. 7/8 cooperated during performance of the MESP. 2/8 passed all categories in the standard version. 3/8 made progress in the low-verbal version, but could not complete the standard version. 2/8 did not progress past the first	Negligible importance as the paper (Lima et al 2016) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)
					Negligible importance as the paper (Santerelli et al 2015) does not involve any participants with FRDA and provides only limited

				category (“cannot test”) in the low-verbal version. (Ji et al 2015).	aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology. (Rance & Starr, 2015)
	Speech perception in background noise assessed with: Mandarin Pediatric Speech Intelligibility	8 (1 observational study)[3]	⨁◯◯◯ Very low[a,b,c,e]	-	8 children with auditory neuropathy received CIs and were assessed to evaluate its benefits. Assessments were conducted pre and post operatively. The MPSI was administered if the speech perception abilities of the children were sufficient. 1/8 reached the level of effective speech, and was the only subject with marked progression in sentence recognition under the noisy condition as determined by the MPSI test. (Ji et al 2015).	Negligible importance as the paper (Ji et al 2015) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015).
	Speech perception in background noise assessed with: Recognition threshold of sentences in noise	1 (1 observational study)[4]	⨁◯◯◯ Very low[a,b,f]	-	A 47-year-old woman reported moderate sensorineural hearing loss in the right ear and high-frequency loss of 4 kHz in the left ear, with bilateral otoacoustic emissions. A hearing aid (HA) was indicated on the right. The patient was tested within a 3-month period before the HA fitting. For RTSN, there was a 5.6-dB improvement in the first and second months after HA use and a 6.0-dB improvement 3 months later. (Lima et al 2016).	
	Speech perception in background noise assessed with: Speech perception tests	21 (1 observational study)[5]	⨁◯◯◯ Very low[b,c,g]	-	21 subjects with dominant optic atrophy (n=11 with *OPA1* mutations inducing haploinsufficiency "*OPA1*-H", n=10 with *OPA1* missense mutations "*OPA1-M*") underwent cochlear implantation. Open-set	Negligible importance as the paper (de Carvalho et al) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015). Negligible importance as the paper (Ji et al 2015) does not

				disyllable recognition scores were evaluated before cochlear implantation and after 1 year of cochlear implant use. Although there was considerable variation between subjects, scores significantly improved for all cochlear implant recipients in a quiet environment and in the presence of background noise, except for 1 subject (paired t-test, p<0.01). (Santerelli et al 2015).	involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015). Negligible importance as the paper (Kontorinis et al 2014) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015).
	Everyday listening/communication assessed with: Meaningful Auditory Integration Scale for Young Children	18 (2 observational studies)[1,3]	⨁◯◯◯ Very low[a,b,c]	-	10 children with auditory neuropathy were treated with cochlear implants (CI). Assessments were completed on the day of the surgery and then again at least 1 year post experience with the implant. There was better performance trend with the CI on the assesment by the IT MAIS (p=0.054) however this did not reach significance. (de Carvalho et al 2016). 8 children with auditory neuropathy received CIs and were assessed to evaluate its benefits. Assessments were conducted pre and post operatively. The average preoperative MAIS/IT-MAIS total, detection, and perception scores were 12.8 ± 14.7%, 11.8 ± 12.0%, and 7.9 ± 16.0%, respectively. Overall postoperative total, detection, and perception scores of MAIS/IT-MAIS for all eight children on the final test session were 66.5 ± 18.4%, 75.1 ± 22.2%, and 61.9 ± 14.7%, respectively, which were significantly higher than the preoperative scores (all	Negligible importance as the paper (Kontorinis et al 2014) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015). Negligible importance as the paper (Lima et al 2016) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015). Negligible importance as the paper (de Carvalho et al 2015) does not involve any participants with FRDA and provides only limited aetiological data. Auditory neuropathy is caused by a range of different pathophysiological mechanisms and CI outcomes are highly dependent on site of lesion/aetiology (Rance & Starr, 2015).

				p<0.0001) (Ji et al 2015).
Everyday listening/communication assessed with: Meaningful Use of Speech Scale	10 (1 observational study)[1]	⨁◯◯◯ Very low[a,b,c]	-	10 children with auditory neuropathy were treated with cochlear implants (CI). Assessments were completed on the day of the surgery and then again at least 1 year post experience with the implant. There were no statistical significance differences in the MUSS (*p*=0.35*).* (de Carvalho et al 2016).
Everyday listening/communication assessed with: Speech Intelligibility Rating	8 (1 observational study)[3]	⨁◯◯◯ Very low[a,b,c,e]	-	8 children with auditory neuropathy received CIs and were assessed to evaluate its benefits. Assessments were conducted pre and post operatively. None of the children performed at the level of effective communication; all of their SIR scores stayed at the preoperative lowest level 1, indicating that none of these children developed effective speech-producing ability or further social communication skills. (Ji et al 2015).
Everyday listening/communication assessed with: Manchester spoken language development scale	27 (1 observational study)[6]	⨁◯◯◯ Very low[a,b,c,d]	-	27 children with ANSD were treated with CI. Pre- and post-CI hearing outcomes were assessed with the MSLDS and compared using the Mann-Whitney U test. The difference between pre-CI and post-CI MSLDS scores were significant (pre scores 2.5 (range 0-6) and post scores 7.7 (range 3-10), *p*<0.001). (Kontorinis et al 2014).
Everyday listening/communication assessed with: Categories	27 (1 observational	⨁◯◯◯ Very low[a,b,c,d]	-	27 children with ANSD were treated with CI. Pre- and post-CI hearing outcomes were

of auditory performance	study)[6]			assessed with the CAP and were compared using the Mann-Whitney U test. The difference between pre-CI and post-CI CAP scores were significant (pre scores 2.5 (range 0-5) and post scores 5.8 (range 2-9), p<0.001). (Kontorinis et al 2014).
Everyday listening and communication assessed with: Hearing Handicap Inventory for Adults	1 (1 observational study)[4]	⨁◯◯◯ Very low[a,b,f]	-	A 47-year-old woman reported moderate sensorineural hearing loss in the right ear and high-frequency loss of 4 kHz in the left ear, with bilateral otoacoustic emissions. A hearing aid (HA) was indicated on the right. The patient was tested within a 3-month period before the HA fitting. The HHIA was administered before HA adaptation and after 3 months of HA use. The evaluation of perception of hearing handicap before HA adaptation demonstrated that all responses were attributed to the "always" option regarding both the emotional and social subscales, corresponding to a severe degree of perception (100 points). Three months after HA adaptation, there was a 40-point reduction of hearing handicap, although perception continued to be of a severe degree. (Lima et al 2016).
Hearing/communication-related quality of life assessed with: Satisfaction with Amplification in Daily Life	10 (1 observational study)[7]	⨁◯◯◯ Very low[a,b,c,d]	-	10 children with auditory neuropathy who had previously been treated with cochlear implants (CI) completed the SADL to determine their overall satisfaction level with the CI. The positive effects were

				shown to be very high, with low negative factors, costs or negative effects on personal image of the patients. (de Carvalho et al 2015).	
Hearing/communication-related stress/anxiety - not measured	-	-	-	-	-
Hearing/communication-related fatigue - not measured	-	-	-	-	-
Speech perception in background quiet assessed with: Bamford–Kowal–Bench (BKB) sentences	1 (1 observational study)[8]	⨁◯◯◯ Very low[f]	-	A 41-year-old female with FA presenting with moderate low-frequency then mild high-frequency sensorineural hearing loss. Speech discrimination was assessed by presenting recorded Bamford–Kowal–Bench (BKB) sentences at 70 dBA in an auditory alone condition. She achieved a score of 11% of keywords correct. After a 4-week trial with hearing aids, speech perception was re-assessed with hearing aids. Functional hearing was impaired by hearing aids, confirmed by a BKB score of 0% keywords correct. Sequential bilateral cochlear implantation (CI) was done over 8 months. After 2 months of bilateral CI use, a score of 88% keywords correct on the BKB was achieved.	
Speech perception in background quiet assessed with: Phonetically Balanced Kindergarten (PBK) words	1 (1 observational study)[9]	⨁◯◯◯ Very low[f]	-	At 6 years of age, a boy with auditory neuropathy and moderate hearing loss had poor outcomes with conventional hearing aids for 2 years, despite aidable hearing (L: PBK = 4% word	

				recognition; R: PBK=12%). Further progression of hearing loss occurred and bilateral cochlear implants (CI) were inserted at age 7 years. CI was associated with minimal benefit (4% words and 20% phoneme score on PBK at one year post-implantation). Children in a control group identified 39% of the words and 61% of the phonemes on the PBK one years post-implantation.

1. de Carvalho G.M., Ramos P. ,Arthur C.,Guimaraes A.,Sartorato E. Performance of Cochlear Implants in Pediatric Patients with Auditory Neuropathy Spectrum Disorder. The Journal of International Advanced Otology; 2016.
2. Ehrmann-Muller D., Cebulla M.,Rak K. et al. Evaluation and therapy outcome in children with auditory neuropathy spectrum disorder (ANSD). International Journal of Pediatric Otorhinolaryngology; 2019.
3. Ji F., Li J.,Hong M. et al. Determination of benefits of cochlear implantation in children with auditory neuropathy. PLoS ONE; 2015.
4. Lima A.P., Mantello E.B.,Anastasio A.R.T. Monitoring the hearing handicap and the recognition threshold of sentences of a patient with unilateral auditory neuropathy spectrum disorder with use of a hearing aid. International Archives of Otorhinolaryngology; 2016.
5. Santarelli R., Rossi R.,Scimemi P. et al. OPA1-related auditory neuropathy: Site of lesion and outcome of cochlear implantation. Brain; 2015.
6. Kontorinis G., Lloyd S.K.,Henderson L. et al. Cochlear implantation in children with auditory neuropathy spectrum disorders. Cochlear Implants Int; 2014.
7. de Carvalho G.M., Zago T.M.,Ramos P.Z.,Castilho A.M.,Guimaraes A.C.,Sartorato E.L. Satisfaction of Children with Auditory Neuropathy and Cochlear Implant. The Journal of International Advanced Otology; 2015.
8. Frewin B., Chung M. Donnelly N.. Bilateral cochlear implantation in Friedreich's ataxia: a case study. Cochlear Implants Int; 2013.
9. Miyamoto R.T., Kirk K.I. Renshaw J. Hussain D.. Cochlear implantation in auditory neuropathy. Laryngoscope; 1999.

a. No participants with a diagnosis of FRDA (all participants with auditory neuropathy).
b. Small sample size.
c. Confidence intervals not reported.
d. Retrospective observational study.
e. Two participants not included in the analysis as didn't meet the exclusion criteria after receiving the cochlear implants.
f. Case study only.
g. No participants with a diagnosis of FRDA (all participants with dominant

	optic atrophy).	

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ Large ○ Moderate ● Small ○ Trivial ○ Varies ○ Don't know	**Hearing Aids** 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). **Cochlear Implants** Two case studies. 1. 7-year-old: no useful hearing post CI 2. 41-year-old: near normal speech perception (in quiet) post implantation. Additional references provided by authors Frewin, B., Chung, M., & Donnelly, N. (2013). Bilateral cochlear implantation in Friedreich's ataxia: A case study. Cochlear implants international, 14(5), 287-290. Miyamoto, R. T., Kirk, K. H., Renshaw, J., & Hussain, D. (1999). Cochlear implantation in auditory neuropathy. The Laryngoscope, 109(2), 181-185. a. No participants with a diagnosis of FRDA (all participants with auditory neuropathy). b. Small sample size. c. Confidence intervals not reported. d. Retrospective observational study. e. Two participants not included in the analysis as didn't meet the exclusion criteria after receiving the cochlear implants. f. Case study only. g. No participants with a diagnosis of FRDA (all participants with dominant optic atrophy).	Potential for harm is low with hearing aids, but the likelihood of a positive outcome with amplification is also low for individuals with auditory neuropathy due to neurodegenerative disease. These individuals typically suffer severe sound distortion (in addition to impaired audibility) which is not alleviated by amplification (Rance and Starr, 2015) Potential for harm with surgical intervention. The variability in the outcomes for the cases studies is consistent with reported CI outcomes in individuals with central auditory disorders (Rance & Starr, 2015). Currently there are no pre-operative predictors of outcome in affected individuals.

Certainty of evidence

What is the overall certainty of the evidence of effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ Very low ● Low ○ Moderate ○ High ○ 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
○ Important uncertainty or variability ● Possibly important uncertainty or variability ○ Probably no important uncertainty or variability ○ No important uncertainty or variability		

Outcomes	Importance	Certainty of the evidence (GRADE)
Speech perception in background noise assessed with: The Glendonald Auditory Screening Procedure	CRITICAL[a]	⨁◯◯◯ VERY LOW[b,c,d]
Speech perception in background noise assessed with: Hochmair-Schulz-Moser (HSM) sentence test	CRITICAL[a]	⨁◯◯◯ VERY LOW[b,c,d,e]

Speech perception in background noise assessed with: Freiburgers' monosyllabic word test	CRITICAL[a]	⊕◯◯◯ VERY LOW[b,c,d,e]
Speech perception in background noise assessed with: Mandarin Early Speech Perception test	CRITICAL[a]	⊕◯◯◯ VERY LOW[b,c,d,f]
Speech perception in background noise assessed with: Mandarin Pediatric Speech Intelligibility	CRITICAL[a]	⊕◯◯◯ VERY LOW[b,c,d,f]
Speech perception in background noise assessed with: Recognition threshold of sentences in noise	CRITICAL[a]	⊕◯◯◯ VERY LOW[b,c,g]
Speech perception in background noise assessed with: Speech perception tests	CRITICAL[a]	⊕◯◯◯ VERY LOW[c,d,h]
Everyday listening/communication assessed with: Meaningful Auditory Integration Scale for Young Children	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,d]
Everyday listening/communication assessed with: Meaningful Use of Speech Scale	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,d]
Everyday listening/communication assessed with: Speech Intelligibility Rating	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,d,f]
Everyday listening/communication assessed with: Manchester spoken language development scale	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,d,e]
Everyday listening/communication assessed with: Categories of auditory performance	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,d,e]
Everyday listening and communication assessed with: Hearing Handicap Inventory for Adults	IMPORTANT[i]	⊕◯◯◯ VERY LOW[b,c,g]
Hearing/communication-related quality of life assessed with: Satisfaction with Amplification in Daily Life	CRITICAL[j]	⊕◯◯◯ VERY LOW[b,c,d,e]
Hearing/communication-related stress/anxiety - not	IMPORTANT[k]	-

measured		
Hearing/communication-related fatigue - not measured	IMPORTANT[l]	-

a. Identified as critical (5/6) and low importance (1/6) by people with FA and critical by expert authors on this topic.
b. No participants with a diagnosis of FRDA (all participants with auditory neuropathy).
c. Small sample size.
d. Confidence intervals not reported.
e. Retrospective observational study.
f. Two participants not included in the analysis as didn't meet the exclusion criteria after receiving the cochlear implants.
g. Case study only.
h. No participants with a diagnosis of FRDA (all participants with dominant optic atrophy).
i. Identified as important (4/6) and low importance (2/6) by people with FA and critical by expert authors on this topic.
j. 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.
k. 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
l. 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.

Balance of effects

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

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ 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

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ○ Probably yes ○ Yes ○ Varies ● Don't know	Limited published evidence.	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). 1/5 indicated hearing aids for all people with a hearing impairment was reasonable, 4/5 indicated didn't know if reasonable. (Aug 2020).

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
○	○	●	○	○

CONCLUSIONS

Recommendation

We cannot recommend either the use or non-use of hearing aids (or 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 hearing aids 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 hearing aids 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 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 hearing aids or cochlear implants in individuals with FRDA.

Studies of the acceptability of hearing aids 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.