

QUESTION

Should customized assistive technology for mobility (e.g. modified vehicles, wheelchair, transfer devices, walkers, adaptive seating and positioning) vs. non-use be used for subjects with impaired independent gait/mobility who use digital and assistive technology with Friedreich ataxia?

POPULATION:	subjects with impaired independent gait/mobility who use digital and assistive technology with Friedreich ataxia
INTERVENTION:	customized assistive technology for mobility (e.g. modified vehicles, wheelchair, transfer devices, walkers, adaptive seating and positioning)
COMPARISON:	non-use
MAIN OUTCOMES:	Independence in mobility; Independence in mobility; Quality of life; Quality of life; Physical devices; Falls; Posture and avoiding pressure; Social and occupational participation ;

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 digital and assistive technologies.</p> <p>4/6 indicated that the problem was serious, 2/6 indicated they didn't know if serious.</p> <p>2/6 indicated that the problem was urgent, 1/6 indicated probably urgent, 1/6 indicated probably not urgent, 2/6 indicated they didn't know if urgent.</p> <p>3/6 indicated that the problem was a priority, 1/6 indicated probably a priority, 2/6 indicated they didn't know if a 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>The two papers deal with two different interventions. The standing frame study in MS improved motor function but to a degree not clinically meaningful. Had control group.</p> <p>The power operated vehicle study in ALS was open label but indicated non-significant changes in QOL but improved social participation.</p>

Outcomes	№ of participants (studies) Follow-up	Certainty of the evidence (GRADE)	Relative effect (95% CI)	Anticipated absolute effects* (95% CI)	
				Risk with non-use	Risk difference with customized assistive technology for mobility (e.g. modified vehicles, wheelchair, transfer devices, walkers, adaptive seating and positioning)
Independence in mobility assessed with: Amended Motor Club Assessment	140 (1 RCT) ¹	⊕⊕⊕○ Moderate ^a	-		140 people with multiple sclerosis were randomised to standing frame (n=71) or usual care (n=69) groups. 122 (n=61 both groups) completed primary outcome assessment for the modified intention to treat analysis. The standing frame group had a increase in AMCA score compared to usual care: fully adjusted between-group difference in AMCA score at 36 weeks of 4.7 points (95% CI 1.9–7.5; <i>p</i> =0.0014). (Freeman et al 2019).
Independence in mobility assessed with: Psychosocial Impact of Assistive Devices Scale	50 (1 observational study) ²	⊕○○○ Very low ^{b,c,d,e,f,g,h}	-		50 patients with amyotrophic lateral sclerosis (ALS) were sent surveys 1 month and 6 months after receiving a new power wheelchair. Mean PIADS scores were shown to increase in the mean value in every positive category from 1 to 6 months, indicating that the power wheelchair affected the patients more in these areas. In the negative areas such as embarrassment, frustration, and confusion, the impact of the power wheelchair decreased over time. In the total absolute value of the three categories of competence, adaptability, and self-esteem, all means increased slightly over time with the greatest increase in competence and self-esteem. A Wilcoxon signed ranks test was performed on the mean scores of the three categories, and in the area of competence with the power wheelchair, Wilcoxon <i>Z</i> = 2.5 and <i>p</i> = 0.001 was a statically significant change between the 1- and 6-month scores. In

				adaptability, Wilcoxon Z = 7.0 and p = 0.2, which was no significant change between 1 and 6 months, and in the self-esteem area, Z = 1 and P = 0.05 showed a significant change between 1 and 6 months. (Ward et al 2015).
Quality of life assessed with: 29-item Multiple Sclerosis Impact Scale	140 (1 RCT) ¹	⊕⊕⊕○ Moderate ^a	-	140 people with multiple sclerosis were randomised to standing frame (n=71) or usual care (n=69) groups. 122 (n=61 both groups) completed primary outcome assessment for the modified intention to treat analysis. Short-term, statistically significant differences in favour of the standing frame group at 20 weeks in both the physical and psychological components of the MSIS-29 scale. These differences in the MSIS29 scale were not sustained at 36 weeks. (Freeman et al 2019).
Quality of life assessed with: Self report	50 (1 observational study) ²	⊕○○○ Very low ^{b,c,e,f,g,h}	-	50 patients with amyotrophic lateral sclerosis (ALS) were sent surveys 1 month and 6 months after receiving a new power wheelchair. At the 1- and 6-month follow-ups, when asked about how the power wheelchair affected their quality of life, 89% and 90% respectively noted an improvement in their quality of life because of the power wheelchair. Although the power wheelchair did improve quality of life as a whole, a Chi-square was calculated, and the statistic was 0.0532 with a p—value of 0.817576 at p < 0.05, which does not demonstrate a significant change in percentage of quality of life between 1 and 6 months. (Ward et al 2015)
Physical devices - not measured	-	-	-	-
Falls assessed with: Self report	140 (1 RCT) ¹	⊕⊕⊕○ Moderate ^a	-	140 people with multiple sclerosis were randomised to standing frame (n=71) or usual care (n=69) groups. 122 (n=61 both groups) completed primary

				outcome assessment for the modified intention to treat analysis. The proportion of participants with two or more falls during weeks 21–36 was significantly lower in the standing frame group, with odds ratio of 0.43 (95% CI 0.20–0.94, $p=0.035$). There was no significant between-group difference over weeks 1–20 or the full 36 week study period. Falling days per person year, pooled across both groups, was 9.9 during 36 weeks. (Freeman et al 2019).
Posture and avoiding pressure - not measured	-	-	-	-
Social and occupational participation assessed with: Psychosocial Impact of Assistive Devices Scale	50 (1 observational study) ²	⊕○○○ Very low ^{b,c,d,e,f,g,h}	-	50 patients with amyotrophic lateral sclerosis (ALS) were sent surveys 1 month and 6 months after receiving a new power wheelchair. Mean PIADS scores were shown to increase in the mean value in every positive category from 1 to 6 months, indicating that the power wheelchair affected the patients more in these areas. In the negative areas such as embarrassment, frustration, and confusion, the impact of the power wheelchair decreased over time. In the total absolute value of the three categories of competence, adaptability, and self-esteem, all means increased slightly over time with the greatest increase in competence and self-esteem. A Wilcoxon signed ranks test was performed on the mean scores of the three categories, and in the area of competence with the power wheelchair, Wilcoxon $Z = 2.5$ and $p = 0.001$ was a statically significant change between the 1- and 6-month scores. In adaptability, Wilcoxon $Z = 7.0$ and $p = 0.2$, which was no significant change between 1 and 6 months, and in the self-esteem area, $Z = 1$ and $P = 0.05$ showed a significant change between 1

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Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS														
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Posture and avoiding pressure - not	-	-	-	-

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 - c. Single study published.
 - d. Unable to decipher which participants contributed data at each time point.
 - e. No CIs reported. Some qualitative and quantitative data reported.
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 - g. One of the outcome measures (surveys) not validated

	h. Participants lost to follow up.	
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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 	There is moderate to very low certainty of evidence 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"> <thead> <tr> <th>Outcomes</th> <th>Importance</th> <th>Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Independence in mobility assessed with: Amended Motor Club Assessment</td> <td>IMPORTANT^a</td> <td>⊕⊕⊕○ MODERATE^b</td> </tr> <tr> <td>Independence in mobility assessed with: Psychosocial Impact of Assistive Devices Scale</td> <td>IMPORTANT^a</td> <td>⊕○○○ VERY LOW^{c,d,e,f,g,h,i}</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Independence in mobility assessed with: Amended Motor Club Assessment	IMPORTANT ^a	⊕⊕⊕○ MODERATE ^b	Independence in mobility assessed with: Psychosocial Impact of Assistive Devices Scale	IMPORTANT ^a	⊕○○○ VERY LOW ^{c,d,e,f,g,h,i}	
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Quality of life assessed with: 29-item Multiple Sclerosis Impact Scale	CRITICAL ^j	⊕⊕⊕○ MODERATE ^b
Quality of life assessed with: Self report	CRITICAL ^j	⊕○○○ VERY LOW ^{c,d,f,g,h,i}
Physical devices - not measured	IMPORTANT ^k	-
Falls assessed with: Self report	CRITICAL ^l	⊕⊕⊕○ MODERATE ^b
Posture and avoiding pressure - not measured	IMPORTANT ^m	-
Social and occupational participation assessed with: Psychosocial Impact of Assistive Devices Scale	IMPORTANT ⁿ	⊕○○○ VERY LOW ^{c,d,e,f,g,h,i}

- a. Identified as critical (1/5) and important (4/5) by people with FA and critical by expert authors on this topic
- b. All participants with a diagnosis of Multiple Sclerosis (not FRDA).
- c. Participants with Amyotrophic Lateral Sclerosis (not FRDA).
- d. Single study published.
- e. Unable to decipher which participants contributed data at each time point.
- f. No CIs reported. Some qualitative and quantitative data reported.
- g. Inadequate eligibility criteria
- h. One of the outcome measures (surveys) not validated
- i. Participants lost to follow up.
- j. Identified as critical (2/5) and important (3/5) by people with FA and critical by expert authors on this topic.
- k. Identified as critical (1/5) and important (4/5) by people with FA and important by expert authors on this topic.
- l. Identified as critical (3/5) and important (2/5) by people with FA and critical by expert authors on this topic.
- m. Identified as critical (1/5) and important (4/5) by people with FA and critical by expert authors on this topic.
- n. Identified as critical (2/5), important (2/5) and low importance (1/5) by people with FA and critical 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

<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 	<p>Expert opinions suggest many assistive technologies are high impact including power operated vehicle , customized gait aids, customized showers/grab rails, commodes, modified kitchens and modified vehicles</p>	<p>A survey designed to systematically collect expert-based opinions from clinicians involved in developing the recommendations for this topic and providing clinical care for individuals with Friedreich ataxia, was conducted. Clinical experts from Australia, Europe, UK, South America, Canada and the USA were asked to consider the harms/benefits of customised gait aids (wheeled frame or walking stick etc.) as a management strategy for individuals with impaired independent gait/mobility.</p> <p>Reflecting on the impact of customised gait aids (wheeled frame or walking stick etc.) on <u>Independence in mobility</u>, 100% (4/4) clinical experts reported a benefit (large, moderate or small), and 0% (0/4) reported observing a harm (large, moderate or small). Reflecting on the impact on <u>Quality of life</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Physical devices</u>, 25% (1/4) clinical experts reported a benefit, 25% (1/4) reported no effect. 2 expert clinicians could not provide any information on this outcome. Reflecting on the impact on <u>Falls</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Posture and avoiding pressure</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Social and occupational participation</u>, 100% (4/4) clinical experts reported a benefit.</p> <p>Clinical experts were asked to consider the harms/benefits of customised wheelchairs as a management strategy for individuals with impaired independent gait/mobility.</p> <p>Reflecting on the impact of customised wheelchairs on <u>Independence in mobility</u>, 100% (4/4) clinical experts reported a benefit (large, moderate or small), and 0% (0/4) reported observing a harm (large, moderate or small). Reflecting on the impact on <u>Quality of life</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Physical devices</u>, 75% (3/4) clinical experts reported a benefit. 1 expert clinician could not provide any information on this outcome. Reflecting on the impact on <u>Falls</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Posture and avoiding pressure</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Social and occupational participation</u>, 100% (4/4) clinical experts reported a benefit.</p> <p>Clinical experts were asked to consider the harms/benefits of customised shower chairs, commodes, grab-rails as a management strategy for individuals with impaired independent gait/mobility.</p> <p>Reflecting on the impact of customised shower chairs, commodes, grab-rails on <u>Independence in mobility</u>, 100% (4/4) clinical experts reported a benefit (large, moderate or small), and</p>
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		<p><u>occupational participation</u>, 100% (4/4) clinical experts reported a benefit.</p> <p>Clinical experts were asked to consider the harms/benefits of adaptive seating as a management strategy for individuals with impaired independent gait/mobility.</p> <p>Reflecting on the impact of adaptive seating on <u>Independence in mobility</u>, 100% (4/4) clinical experts reported a benefit (large, moderate or small), and 0% (0/4) reported observing a harm (large, moderate or small). Reflecting on the impact on <u>Quality of life</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Physical devices</u>, 75% (3/4) clinical experts reported a benefit. 1 expert clinician could not provide any information on this outcome. Reflecting on the impact on <u>Falls</u>, 75% (3/4) clinical experts reported a benefit, 25% (1/4) reported no effect. Reflecting on the impact on <u>Posture and avoiding pressure</u>, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on <u>Social and occupational participation</u>, 100% (4/4) clinical experts reported a benefit.</p>
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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 checked="" type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 	No published research.	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if customised assistive technology for mobility in people with impaired mobility was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>4/5 indicated the intervention was acceptable, 1/5 indicated probably acceptable. (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	Possibly important	Probably no important	No important			

JUDGEMENT							
	or variability	uncertainty or variability	uncertainty or variability	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 individuals with Friedreich ataxia with impaired independent gait/mobility, we suggest the use of customized assistive technology for mobility (e.g., modified vehicle, wheelchair, transfer devices, walkers, and adaptive seating and positioning) to enhance independence in mobility, quality of life, and social and occupational participation, and to reduce falls.

Justification

Despite the low level of evidence in like conditions, expert clinicians who provide clinical care for individuals with Friedreich ataxia agree that the use of customized assistive technology for mobility can benefit independence in mobility, quality of life and social and occupational participation, and reduce falls.

Subgroup considerations

This recommendation is specifically for individuals with Friedreich ataxia with impaired independent gait/mobility. The guideline expert panel considers that all individuals with Friedreich ataxia might benefit from the use of digital and assistive technology.

Research priorities

Key research priorities in this area include identifying and evaluating the most appropriate customized assistive technology for individuals with Friedreich ataxia and measuring efficacy against effects on independence in mobility, quality of life, social and occupational participation and reduction in falls.