QUESTION

Should health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for diabetes FA) vs. non-use be used for subjects with fatigue, fall risk, poor sleep, diabetes etc who use digital and assistive technology with Friedreich ataxia?

POPULATION:	subjects with fatigue, fall risk, poor sleep, diabetes etc who use digital and assistive technology with Friedreich ataxia
INTERVENTION:	health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for diabetes FA)
COMPARISON:	non-use
MAIN OUTCOMES:	Independence in daily activities; Independence in daily activities; Independence in daily activities; Quality of life; Quality of life; Quality of life; Quality of life; Quality of life; Quality of life; Quality of life; Quality of life; Number of hospitalizations; Improved sleep control; Improved diabetes control; Improved cardiac control; Improved eating control; Improved medication control;

ASSESSMENT

Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ● Probably yes ○ Yes ○ Varies ○ Don't know		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. 4/6 indicated that the problem was serious, 2/6 indicated they didn't know if serious. 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. 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)

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ Trivial ● Small ○ Moderate ○ Large ○ Varies		

Outcomes	№ of	Certainty of	Relative	Anticipated absolute effects* (95% CI)

○ Don't know

	participants (studies) Follow-up	the evidence (GRADE)	effect (95% CI)	Risk with non-use	Risk difference with health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for diabetes FA)
Independence in daily activities assessed with: Frenchay activity index	38 (1 RCT)[1]	⨁⨁◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the FAI (p=0.50). (Nasseri et al 2020).	
Independence in daily activities assessed with: AMC Linear Disability Scale	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control group (usual care, n=51). Data was obtained at baseline, 3, and 6 months. Cohen's d effect for the ALDS was calculated as (the difference between two means) / (SD of the ALDS score in the Medido group at baseline). After 6 months of follow-up, the mean score in the Medido group improved by 2.5 points (SE 3.4). Compared to the control group, this resulted in a nonsignificant difference of 2.9 points (95%CI -11.6;5.7) in favor of the Medido group. The effect size was small at 0.13. In exploratory sub analyses, a significant improvement in the Medido group compared to the control group was observed (difference of 14.69 points (95% CI-28.5;-0.9) only in H&Y > 2.5 (Hoehn & Yahr score > 2.5, indicating more advanced disease). The effect size was 0.59. (Hannink et al 2019).	

Independence in daily activities assessed with: Godin–Leisure Time Exercise Questionnaire	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the GLTEQ (*p*=0.51). (Nasseri et al 2020).	
Quality of life assessed with: Hamburg Quality of Life Questionnaire Multiple Sclerosis	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the HAQUAMS (*p*=0.71). (Nasseri et al 2020).	
Quality of life assessed with: MS Walking Scale	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the MSWS (*p*=0.82). (Nasseri et al 2020).	
Quality of life	40	⊕◯◯◯	-	40 people with Parkinson's disease	

assessed with: Short Form 36 Health Survey	(1 RCT)[3]	Very low[b,c,f,g,h]		undertook gait training for 30 mins, 3x/week for 6 weeks, and were randomised to an intervention (CuPiD-system whereby a smartphone app offered positive and corrective feedback on gait) or active control group (personalised gait advice provided). Assessments were conducted before and after training and at 4 weeks follow up. A significant time by group interaction was found for the physical health score of the SF-36 ($F(2,108) = 1.85$, $p < 0.05$, $\eta 2 = 0.06$), indicating that the control group experienced a decrease in self-reported physical health at follow-up, while the CuPiD group did not. (Ginis et al 2016).
Quality of life assessed with: PDQ-39	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. At baseline, significant differences between groups for total PDQ-39 score ($p = 0.005$), mobility ($p = 0.018$), ADL ($p = 0.002$) and cognition ($p = 0.007$) were in favor of the control group. After 6 months, both groups deteriorated in total PDQ score, but the Medido group deteriorated 1.0 point further ($p = 0.01$ for group x time interaction). Patients in the control group did numerically, but not significantly better, or deteriorated less in emotional wellbeing, stigma, social support, communication, and physical wellbeing domains compared to the Medido group, whereas the Medido group improved numerically, but not significantly in the mobility, ADL and cognition domains. (Hannink et al 2019).
Quality of life assessed with:	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser

EQ5D-5L				Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. The control group had a significant (p =0.047) better initial EQ5D-5L score compared to the Medido group at baseline. After 6 months of follow-up both groups showed no change. (Hannink et al 2019).
Quality of life assessed with: EQ5D-5L visual analogue scale	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. After 6 months, the Medido group improved by 0.4 points in the VAS score, while the control group remained stable (difference 0.4, 95% CI - 0.2;1.1, p = 0.057). (Hannink et al 2019)
Quality of life assessed with: PDQ-carer questionnaire	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. At baseline, the Medido group showed a significantly higher stress score (p = 0.01). We observed no differences in change over time between both groups in any of the domains in the PDQ-carer. (Hannink et al 2019).
Quality of life assessed with: Health related quality of life	87 (1 RCT)[4]	⨁◯◯◯ Very low[b,c,h,i]	-	20 patients with multiple sclerosis were randomised to either a software application-based or paper-based home therapy program for 3 months. At month 3, all patients received the app-based program for another 3 months. Differences between patient groups were compared at baseline using Fisher's exact test or independent t-test and two-way ANOVA at month 3 for interaction of treatment and time respectively. None

				of the interventions was associated with a decrease in HRQoL. (Ehling et al 2017).	
Number of hospitalizations - not measured	-	-	-	-	-
Improved sleep control - not measured	-	-	-	-	-
Improved diabetes control - not measured	-	-	-	-	-
Improved cardiac control - not measured	-	-	-	-	-
Improved eating control - not measured	-	-	-	-	-
Improved medication control assessed with: Multiple Sclerosis Treatment Adherence Questionnaire	117 (1 RCT)[5]	⨁◯◯◯ Very low[a,g,j]	-	117 patients with multiple sclerosis were randomly allocated to either using an e-diary (n=62) or no e-diary (n=55). Patients were evaluated at baseline, 6, and 12 months. Self-reported adherence was assessed with the MS-TAQ. Agreement between medication possession rate (MPR) and e-diary-derived adherence, as well as between the MPR and self-report adherence wereevaluated by Bland–Altman plots. The median difference between e-diary-derived adherence and the MPR was –3% (95% limits of agreement: -53% to 12%). The median difference between retrospective self-reported adherence and the MPR was 0.3% (95% limits of agreement: -20% to 42%). The proportion of participants with poor adherence to DMDs was similar in the e-diary and control groups (10% vs. 13%, p= 0.6). (Golan et	

				al 2020).

1. Nasseri N.N., Ghezelbash E.,Zhai Y.,et al. Feasibility of a smartphone app to enhance physical activity in progressive MS: A pilot randomized controlled pilot trial over three months. . Peer J. ; 2020.
2. Hannink K, Ter Brake L,Oonk NGM,et al.. A randomized controlled efficacy study of the Medido medication dispenser in Parkinson's disease. . BMC geriatr.; 2019.
3. Ginis P, Nieuwboer A,Dorfman M,et al.. Feasibility and effects of home-based smartphone-delivered automated feedback training for gait in people with Parkinson's disease: A pilot randomized controlled trial. . Parkinsonism Relat Disord. ; 2016.
4. Ehling R., Edlinger M.,Hermann K.,et al. Successful long-term management of spasticity in patients with multiple sclerosis using a software application (APP): A pilot study. . Mult. Scler. Relat. Disord.; 2017.
5. Golan D., Sagiv S.,Glass-Marmor L.,Miller A.. Mobile phone-based e-diary for assessment and enhancement of medications adherence among patients with multiple sclerosis. . Mult. Scler. J. Exp. Transl. Clin.. 2020;6(3):no pagination. doi:10.1177/2055217320939309 ; 2020.

a. Participants had a diagnosis of multiple sclerosis (not FRDA).
b. Small sample size.
c. Participants had a diagnosis of Parkinson’s disease (not FRDA).
d. Loss to follow up, resulting in unequal n in groups (treatment n=36, control n=51).
e. Missing measures at third timepoint
f. Uncorrected post-hoc analyses for multiple comparisons
g. CI’s not reported
h. Lack of assessor blinding for group allocation
i. Single study, small sample size.
j. Participants and investigator aware of allocation and monitoring of medication.

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS

○ Large
○ Moderate
○ Small
○ Trivial
○ Varies
● Don't know

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 health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for diabetes FA)
Independence in daily activities assessed with: Frenchay activity index	38 (1 RCT)[1]	⨁⨁◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the FAI (*p*=0.50). (Nasseri et al 2020).	
Independence in daily activities assessed with: AMC Linear Disability Scale	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control group (usual care, n=51). Data was obtained at baseline, 3, and 6 months. Cohen's d effect for the ALDS was calculated as (the difference between two means) / (SD of the ALDS score in the Medido group at baseline). After 6 months of follow-up, the mean score in the Medido group improved by 2.5 points (SE 3.4). Compared to the control group, this resulted in a nonsignificant difference of 2.9 points (95%CI -11.6;5.7) in favor of the Medido group. The effect size was small at 0.13. In exploratory sub analyses, a significant improvement in the Medido group compared to the control group was observed (difference of 14.69 points	

				(95% CI-28.5;-0.9) only in H&Y > 2.5 (Hoehn & Yahr score > 2.5, indicating more advanced disease). The effect size was 0.59. (Hannink et al 2019).
Independence in daily activities assessed with: Godin–Leisure Time Exercise Questionnaire	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the GLTEQ (*p*=0.51). (Nasseri et al 2020).
Quality of life assessed with: Hamburg Quality of Life Questionnaire Multiple Sclerosis	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to evaluate group differences revealed no group differences observed in the HAQUAMS (*p*=0.71). (Nasseri et al 2020).
Quality of life assessed with: MS Walking Scale	38 (1 RCT)[1]	⊕⊕◯◯ Low[a,b]	-	38 people with chronic progressive multiple sclerosis were randomised to either a control (n=20) or intervention group (n=18). The intervention gtroup received access to an evidence based patient information app including activity feedback, texts, figures and videos. The control group received a leaflet with unspecific information about exercising in general. ANCOVA corrected for baseline values to

				evaluate group differences revealed no group differences observed in the MSWS (p=0.82). (Nasseri et al 2020).
Quality of life assessed with: Short Form 36 Health Survey	40 (1 RCT)[3]	⨁◯◯◯ Very low[b,c,f,g,h]	-	40 people with Parkinson's disease undertook gait training for 30 mins, 3x/week for 6 weeks, and were randomised to an intervention (CuPiD-system whereby a smartphone app offered positive and corrective feedback on gait) or active control group (personalised gait advice provided). Assessments were conducted before and after training and at 4 weeks follow up. A significant time by group interaction was found for the physical health score of the SF-36 ($F(2,108) = 1.85$, $p < 0.05$, $\eta 2 = 0.06$), indicating that the control group experienced a decrease in self-reported physical health at follow-up, while the CuPiD group did not. (Ginis et al 2016).
Quality of life assessed with: PDQ-39	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. At baseline, significant differences between groups for total PDQ-39 score (p = 0.005), mobility (p = 0.018), ADL (p = 0.002) and cognition (p = 0.007) were in favor of the control group. After 6 months, both groups deteriorated in total PDQ score, but the Medido group deteriorated 1.0 point further (p = 0.01 for group x time interaction). Patients in the control group did numerically, but not significantly better, or deteriorated less in emotional wellbeing, stigma, social support, communication, and physical wellbeing domains compared to the Medido group, whereas the Medido group improved numerically, but not significantly in the mobility, ADL and cognition domains. (Hannink et al

				2019).
Quality of life assessed with: EQ5D-5L	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. The control group had a significant (p =0.047) better initial EQ5D-5L score compared to the Medido group at baseline. After 6 months of follow-up both groups showed no change. (Hannink et al 2019).
Quality of life assessed with: EQ5D-5L visual analogue scale	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. After 6 months, the Medido group improved by 0.4 points in the VAS score, while the control group remained stable (difference 0.4, 95% CI - 0.2;1.1, p = 0.057). (Hannink et al 2019)
Quality of life assessed with: PDQ-carer questionnaire	87 (1 RCT)[2]	⨁⨁◯◯ Low[c,d,e]	-	87 patients with Parkinson's disease were randomised into intervention (electronic medication dispenser Medido, n=36) or control groups (usual care, n=51). Data was obtained at baseline, 3, and 6 months. At baseline, the Medido group showed a significantly higher stress score (p = 0.01). We observed no differences in change over time between both groups in any of the domains in the PDQ-carer. (Hannink et al 2019).
Quality of life assessed with: Health related quality of life	87 (1 RCT)[4]	⨁◯◯◯ Very low[b,c,h,i]	-	20 patients with multiple sclerosis were randomised to either a software application-based or paper-based home therapy program for 3 months. At month 3, all patients received the app-based program for another 3 months. Differences between patient

				groups were compared at baseline using Fisher's exact test or independent t-test and two-way ANOVA at month 3 for interaction of treatment and time respectively. None of the interventions was associated with a decrease in HRQoL. (Ehling et al 2017).	
Number of hospitalizations - not measured	-	-	-	-	-
Improved sleep control - not measured	-	-	-	-	-
Improved diabetes control - not measured	-	-	-	-	-
Improved cardiac control - not measured	-	-	-	-	-
Improved eating control - not measured	-	-	-	-	-
Improved medication control assessed with: Multiple Sclerosis Treatment Adherence Questionnaire	117 (1 RCT)[5]	⨁◯◯◯ Very low[a,g,j]	-	117 patients with multiple sclerosis were randomly allocated to either using an e-diary (n=62) or no e-diary (n=55). Patients were evaluated at baseline, 6, and 12 months. Self-reported adherence was assessed with the MS-TAQ. Agreement between medication possession rate (MPR) and e-diary-derived adherence, as well as between the MPR and self-report adherence wereevaluated by Bland–Altman plots. The median difference between e-diary-derived adherence and the MPR was –3% (95% limits of agreement: -53% to 12%). The median difference between retrospective self-reported adherence and the MPR was	

					0.3% (95% limits of agreement: -20% to 42%). The proportion of participants with poor adherence to DMDs was similar in the e-diary and control groups (10% vs. 13%, *p*= 0.6). (Golan et al 2020).

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4. Ehling R., Edlinger M.,Hermann K.,et al. Successful long-term management of spasticity in patients with multiple sclerosis using a software application (APP): A pilot study. . Mult. Scler. Relat. Disord.; 2017.
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a. Participants had a diagnosis of multiple sclerosis (not FRDA).
b. Small sample size.
c. Participants had a diagnosis of Parkinson’s disease (not FRDA).
d. Loss to follow up, resulting in unequal n in groups (treatment n=36, control n=51).
e. Missing measures at third timepoint
f. Uncorrected post-hoc analyses for multiple comparisons
g. CI’s not reported
h. Lack of assessor blinding for group allocation
i. Single study, small sample size.
j. Participants and investigator aware of allocation and monitoring of medication.

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	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

○ Important uncertainty or variability
○ Possibly important uncertainty or variability
● Probably no important uncertainty or variability
○ No important uncertainty or variability

RESEARCH EVIDENCE

Outcomes	Importance	Certainty of the evidence (GRADE)
Independence in daily activities assessed with: Frenchay activity index	CRITICAL[a]	⨁⨁◯◯ LOW[b,c]
Independence in daily activities assessed with: AMC Linear Disability Scale	CRITICAL[a]	⨁⨁◯◯ LOW[d,e,f]
Independence in daily activities assessed with: Godin–Leisure Time Exercise Questionnaire	CRITICAL[a]	⨁⨁◯◯ LOW[b,c]
Quality of life assessed with: Hamburg Quality of Life Questionnaire Multiple Sclerosis	CRITICAL[a]	⨁⨁◯◯ LOW[b,c]
Quality of life assessed with: MS Walking Scale	CRITICAL[a]	⨁⨁◯◯ LOW[b,c]
Quality of life assessed with: Short Form 36 Health Survey	CRITICAL[a]	⨁◯◯◯ VERY LOW[c,d,g,h,i]
Quality of life assessed with: PDQ-39	CRITICAL[a]	⨁⨁◯◯ LOW[d,e,f]

ADDITIONAL CONSIDERATIONS

Quality of life assessed with: EQ5D-5L	CRITICAL[a]	⊕⊕◯◯ LOW[d,e,f]
Quality of life assessed with: EQ5D-5L visual analogue scale	CRITICAL[a]	⊕⊕◯◯ LOW[d,e,f]
Quality of life assessed with: PDQ-carer questionnaire	CRITICAL[a]	⊕⊕◯◯ LOW[d,e,f]
Quality of life assessed with: Health related quality of life	CRITICAL[a]	⊕◯◯◯ VERY LOW[c,d,i,j]
Number of hospitalizations - not measured	CRITICAL[k]	-
Improved sleep control - not measured	IMPORTANT[l]	-
Improved diabetes control - not measured	IMPORTANT[m]	-
Improved cardiac control - not measured	CRITICAL[n]	-
Improved eating control - not measured	IMPORTANT[o]	-
Improved medication control assessed with: Multiple Sclerosis Treatment Adherence Questionnaire	IMPORTANT[p]	⊕◯◯◯ VERY LOW[b,h,q]

a. Identified as critical (2/5) and important (3/5) by people with FA and critical by expert authors on this topic
b. Participants had a diagnosis of multiple sclerosis (not FRDA).
c. Small sample size.
d. Participants had a diagnosis of Parkinson's disease (not FRDA).
e. Loss to follow up, resulting in unequal n in groups (treatment n=36, control n=51).
f. Missing measures at third timepoint
g. Uncorrected post-hoc analyses for multiple comparisons
h. CI's not reported
i. Lack of assessor blinding for group allocation
j. Single study, small sample size.
k. Identified as critical (3/5), important (1/5) and low importance (1/5) by people with FA and critical by expert authors on this topic
l. Identified as critical (1/5) and important (4/5) by people with FA and important 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 (3/5) and important (2/5) by people with FA and critical by expert authors on this topic o. Identified as important (5/5) by people with FA and important by expert authors on this topic p. Identified as important (4/5) and low importance (1/5) by people with FA and critical by expert authors on this topic q. Participants and investigator aware of allocation and monitoring of medication.	

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		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 **Health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ecg, activity, healthy eating, medication, glucose for diabetes FA) as a management strategy for individuals with fatigue, fall risk, poor sleep, diabetes etc**. Reflecting on the impact of **Health monitoring / alert devices (e.g. monitoring heart rate, steps, sleep, ecg, activity, healthy eating, medication, glucose for diabetes FA)** on Independence in daily activities, 75% (3/4) clinical experts reported a benefit (large, moderate or small), and 0% (0/4) reported observing a harm (large, moderate or small). 1 clinician could not provide any information on this outcome. Reflecting on the impact on Quality of life, 75% (3/4) clinical experts reported a benefit. 1 expert clinician could not provide any information on this outcome. Reflecting on the impact on Number of hospitalizations, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could not provide any information on this outcome. Reflecting on the impact on Improved sleep control, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could not provide any information on this outcome. Reflecting on the impact on Improved diabetes control, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could

		not provide any information on this outcome. Reflecting on the impact on Improved cardiac control, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could not provide any information on this outcome. Reflecting on the impact on Improved eating control, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could not provide any information on this outcome. Reflecting on the impact on Improved medication control, 50% (2/4) clinical experts reported a benefit. 2 expert clinicians could not provide any information on this outcome.

Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ● Probably yes ○ Yes ○ Varies ○ Don't know	No published research.	The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if health monitoring or alert devices for people with fatigue, fall risk, poor sleep and diabetes were acceptable (weighing up the balance between benefits, harms and costs). 3/5 indicated the intervention was acceptable, 2/5 indicated probably acceptable. (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

For individuals with Friedreich ataxia and fatigue, a risk of falls, poor sleep, diabetes and/or cardiomyopathy, we suggest the use of health monitoring/alert devices (e.g., monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for FRDA-related diabetes) to enhance independence in daily activities and quality of life, and improve medication control.

Justification

Despite the low level of evidence, expert clinicians who provide clinical care for individuals with Friedreich ataxia agree that customized assistive technology for health monitoring can benefit independence in daily activities, quality of life, number of hospitalizations, sleep, diabetes and medication control in individuals with Friedreich ataxia.

Subgroup considerations

This recommendation is particularly relevant to individuals with Friedreich ataxia who report fatigue or poor sleep, are at risk of falls, and/or have diabetes or cardiomyopathy.

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 daily activities; quality of life and medication control.