Clinical Management Guidelines for Friedreich Ataxia

Chapter 16. Digital and assistive technologies in Friedreich ataxia

Contents

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16. Digital and assistive technologies in Friedreich ataxia

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This chapter describes the effects of Friedreich ataxia on independent living and the role of digital and assistive technologies in improving independence, with some considerations for wheelchair and seating systems. In making recommendations for managing digital and assistive technologies for individuals with Friedreich ataxia, the authors were tasked with the following:

Overview of the use of digital and assistive technologies in Friedreich ataxia (with recommendations).

16.1 The impact of Friedreich ataxia on independent living

Like most progressive neurological conditions, the relentless nature of Friedreich ataxia (FRDA) can have a profound impact on the capacity to participate in daily activities, be they personal, social, vocational or community focused. The overwhelming impact on independence for people with FRDA is the associated motor impairment. In particular, people with FRDA have difficulty with multi-joint movements (dysynergia), coordinating the spatial and temporal components of movement, terminal accuracy of movement (dysmetria), terminal accuracy of movement amplitude (tremor), maintaining consistent and appropriate force, truncal balance, muscle tone imbalance, muscle weakness and fatigue. Successful completion of most activities of personal (bathing, grooming, dressing, feeding and toileting oneself), domestic (meal preparation, household tasks) and community (driving, using public transport, shopping) activities involve multi-joint movement, balance, muscle strength, endurance, intact sensation and adequate temporal and spatial control of movement. As the disease progresses, the limitations in everyday life become greater and with them the need for support.

Communication impairments are another common feature of FRDA that have an impact on independence. Dysarthria as a result of impaired co-ordination and weakness of muscles used to produce speech can affect a person's ability to engage with carers and family and participate socially and in the work environment. A further impact of FRDA can be in relation to other medical co-morbidities which require specialist treatment and monitoring, particularly those related to the heart (cardiomyopathy), endocrine system (diabetes), spasticity/spasm and fatigue.

The use of digital and assistive technologies can be considered for people with FRDA to compensate for motor, communication and sensory deficits, for better management of personal, domestic, community and workplace activities, and for monitoring other medical co-morbidities. Some emerging digital and assistive technologies aim to enable remediation of deficits or encourage participation in activities to maintain function and to avoid further complications. The overarching aim of digital and assistive technologies for people with FRDA is to optimize their independence and participation in all daily activities.

16.2 The role of digital and assistive technologies in improving independence

Interventions aimed at maintaining or improving independence in people with FRDA can either be compensatory, that is interventions that improve function by compensating for the underlying deficit; or restorative, that is interventions that aim to facilitate restoration of function by adaptation and recovery within the neuro-musculoskeletal system (1).

As a consequence of disease progression individuals with FRDA become increasingly dependent on adaptive equipment. Symptoms including ataxia, muscle weakness, spasticity and bony changes such

as scoliosis and foot deformity interfere with the ability to mobilize, attend to personal daily tasks and to stand and transfer independently, the latter of which is a significant contributor to ongoing maintenance of independence. Given there is limited evidence regarding the provision of adaptive equipment for people with FRDA, clinical reasoning underlying provision of adaptive equipment is largely based on evidence from similar progressive, neurological conditions. It is critical however that the clinician prescribing the equipment is familiar with the particular issues related to FRDA and experienced in the adaptive equipment prescribed. In particular, the prescription of the first wheelchair can be seen as a significant milestone in the progression of the condition; hence, ensuring active involvement of both the client and parents (if appropriate) is critical (2). Here, mobility should not be the only focus of attention, but also possible neuromuscular deformities such as poor posture due to the presence of scoliosis (e.g., adjustment of a seating system, with for example, foam or gel inserts, to improve seated posture).

16.2.1 Health monitoring and self-management

Health monitoring and alert devices have becoming increasingly available in recent years; however, their adoption into real-life use has been low in like conditions (3). Examples include phone applications that track physical activity, prompt/remind for medication, monitor clinical indicators (e.g., heart rate) or provide tools for self-management strategies (sleep, fatigue management). Digital tracking and monitoring tools are used to support self-management and personalized health care in like conditions; however, the acceptance of these apps and devices depends on the person experiencing the value of the device and being well equipped to use them (3). Further development of these tools is required to ensure they are adaptable to a person's own situation, there is support to understand the information gathered and there are links into clinical treatment plans (3). For people with FRDA there are a number of medical co-morbidities which might be better monitored or managed through the use of digital assistive technology. Smart watches that can monitor heart rate, sleep cycles or prompts for medication regimes may be useful. Technology such as blood glucose monitoring systems may also be helpful in a person managing their FRDA-related diabetes (see Chapter 10, section 10.1). Applications to record, track and prompt healthy lifestyle behaviors or therapy tools for physical exercise can be useful to provide a person with FRDA an opportunity to engage in self-management. Given there is limited evidence for the use of these systems, it is vital that any digital or assistive technology be explored with appropriate clinical reasoning and full involvement of the person with FRDA to ensure they are well equipped to use the device or technology.

16.2.2 Communication aids

There is limited research in like conditions related to assistive technology for use in communication and to facilitate workplace participation. One study in amyotrophic lateral sclerosis, a like condition with progressively deteriorating expressive communication and upper limb function, suggests that early use of tablet-based assistive technology for communication is beneficial and has positive impacts on quality of life, particularly as the condition progresses and more complex alternative strategies are required (4). For people with FRDA, customized communication devices that compensate for upper limb, vision, hearing and/or speech deficits may enhance communication and workplace opportunities (see Chapter 3.5).

16.2.3 Environmental control

Environmental control is becoming more mainstream with the development and widespread commercial availability of technical systems such as Apple Siri, Google Home and Amazon Alexa. These integrated devices allow users to control aspects of their home environment such as lighting,

heating, answering the door and controlling appliances *via* their phone or tablet, making it easier for disabled persons to live independently. Access to this technology is becoming easier thanks to particular brands of phone or tablet that already have inbuilt Apps for such function at a reasonable cost. Use of such freely available environmental control devices in addition to more customized and specially designed systems for people with FRDA may enable greater independence within a person's home.

16.2.4 Mobility aids

Customized assistive technology for mobility includes modified vehicles, wheelchairs, traction devices, transfer devices (sling or standing hoists, sit to stand aids such as Sara Stedy [https://www.arjo.com/int/products/patient-handling/standing-and-raising-aid/sara-stedy/], ETAC turners [https://www2.etac.com/en-us/products/product-archive/manual-transfer/etac-turner/]), walkers, adaptive seating and positioning aids. These devices can assist a person to mobilize, transfer, access the home or community and participate in all aspects of daily life.

Due to progressive muscle weakness, ataxia, spasticity and other neuromuscular changes the needs for assistive technology will probably change over time. There may be a progression from transfers in standing/stepping to the use of standing/sling or track hoists and lifters to enable safe transfers in a person's environment. A thorough assessment of a person's functional abilities within the context of their own environment is crucial to selecting the most appropriate device for mobility, transfers and seating. A focus on maintaining active participation in mobility and transfers is critical to maintaining engagement of muscles, which is important in maintaining muscle strength. A sling hoist removes any active participation of the person's upper body, core or lower body and is frequently used from a lying or supported sitting position. Alternatively, a standing hoist is used from a sitting position with the sling just around the middle of the body and this allows engagement of upper limb and lower limbs in the process of helping with standing. Preserving active use of upper limbs, core and lower limbs in any way is important for a person with FRDA and the use of transfers is a further opportunity for these muscles to be engaged on a daily basis. Despite the sometimes unconventional techniques for transfers, a balance between risk and independence must be considered. Moving too quickly from a standing transfer to a hoist can have unintended consequences in addition to further loss of lower limb strength, for core strength, foot and ankle muscle length and upper limb function.

Individuals with FRDA benefit from the provision of grab rails to assist in attending to activities of daily living. Given that ataxia is a core impairment, it is essential that grab rails be correctly positioned to ensure that during the transfer the person is able to maintain their center of gravity over their knees and feet. As such, grab rails positioned to facilitate pulling up, rather than pushing up may be of more benefit. Likewise, provision of wheeled commode/shower chairs requires customization to the specific requirements of a person with FRDA including stability and ease of maneuverability and transfer. This may include lowering the seat height to facilitate safe transfers, utilizing smaller seat apertures and ensuring an appropriate seat width to avoid extraneous trunk movement while in use.

16.2.5 Compensatory techniques

Compensatory techniques include interventions such as the use of weighted and adaptive equipment such as specialized cutlery, feeding and writing implements that feature both weighted, non-slip and larger grips (5, 6) and weighting of the distal component of the limb during activity (7-10). The use of simple strategies to decompose multi-joint movements to more accurate single joint movements (1, 11), minimize reaching movements and stabilize proximal joints (12) have also been proposed. Practically, this includes stabilizing the elbows on the table while feeding oneself, sliding

the hand on the table to facilitate grasp instead of grasping in space, use of orthotics to stabilize joints (in particular the wrist), using two hands to grasp objects or using one hand to stabilize the other while manipulating an object. Training in independence tasks may also consider the significance of visuomotor or proprioceptive loss on functional capacity. People with FRDA may use vision to compensate for sensory impairment; however, the inherent delay in using visual feedback to correct motor performance needs to be factored into training and provision of equipment (e.g., programming of motorized wheelchairs).

As mentioned, a detailed assessment of functional capacity in the context of the individual's everyday environment such as home, work or community is critical to developing appropriate strategies to improve independence. The goals of intervention aimed at maximizing function in each environmental context can be incorporated into subsequent strategies and modifications to facilitate independence. Clinical reasoning is essential to avoid a prescriptive approach to the provision of adaptive equipment. For example, it is of greater benefit if people with ataxia are able to pull up, rather than push up on grab rails in order to maintain the center of gravity over their feet and thus ensure safety while transferring. Such clinical reasoning is necessary in most situations when providing adaptive equipment and modification of environment. Table 16.1 provides a summary of compensatory techniques for specific activities of daily living.

Table 16.1: Suggested interventions/aids to facilitate independence in daily activities

Activity	Aid/strategy to facilitate independence
Grooming	Large, weighted grips on implements; use of two hands; stabilizing elbows while performing task
Feeding	Large, weighted grips on implements; two handled mugs; modifying food (stir-fries, pasta that only require one utensil); stabilizing elbows to perform task
Bathing	Grab rails/hand-held shower hose; if standing, maintain safety by leaning against wall when shutting eyes to wash hair; equipment to sit on while bathing; liquid soap; toweling robe to assist with drying; use of a hand-held shower hose; transitioning from a bathtub to a shower.
Toileting	Grab-rails (including behind toilet and drop down to side); slide bench next to toilet; double-hinged toilet seat to accommodate side transfers
Dressing	Position on bed to dress lower half of body; consideration of clothes that are easy to put on and remove; consideration of wheelchair scripting if dressing self in wheelchair
Bed	Bed sticks; adjustable high-low bed with head raiser; satin sheets for ease in rolling over; positioning of grab rails on adjacent walls for safe and effective transfers; provision of king single bed as minimum size
Home access	No steps or stairs at the access points or internally; ramps at a gradient of 1:14; door openings at least 850 mm.
Bathroom design	Stepless shower-area; non-slip flooring; basin that allows wheelchair access if indicated; lever or sensor taps; angled mirror for seated user; accessible storage areas

Activity	Aid/strategy to facilitate independence
Kitchen	Meal preparation, sink and storage area wheelchair accessible; induction cooktops; adequate bench space between refrigerator, hot plates, sink to avoid carrying items; wall oven with side opening doors; rocker action, push-pad or toggle electrical switches; non-slip mats; kitchen trolleys
Writing	Larger grips pens; sloped writing boards; computer assisted technology such as modified keyboards and mouse.
General	Environment controls; keyless entries
Driving	Hand controls; wheelchair hoist

16.2.6 Restorative techniques

Restorative techniques include the use of physical rehabilitation, relaxation and biofeedback (13-15). However, these are not widely used in clinical practice for people with FRDA.

16.3 Considerations for prescribing wheelchair and seating systems

16.3.1 Review of literature related to Friedreich ataxia

There is only one published study that specifically examines the prescription of wheelchairs for people with FRDA (2). This prospective two-year randomized crossover study included 19 male participants, four of whom had been diagnosed with FRDA, although the method of diagnosis was not reported (the study predated genetic diagnosis). This study examined the efficacy of provision of a customized wheelchair seating system on sitting posture, respiration and upper limb function. While having some methodological shortcomings, this study is important in highlighting how critical adaptive seating is to an improved seated posture. There was, however, little immediate effect on respiratory function and only small differences in upper limb function. An important conclusion from this study was the potential to over-correct musculoskeletal changes, and moreover, in the process of correction for deformity, to further compromise functional capacity. The study findings reinforce that in prescribing a manual wheelchair and seating system, it is essential that functional capacity is not impeded for the sake of an anatomically correct seated posture (2, 16). No new studies specifically relating to prescription of wheelchairs for people with FRDA have been identified for inclusion in the updated clinical management guidelines. However, most clinicians treating individuals with FRDA support this notion.

16.3.2 Review of literature from like conditions

As with FRDA there are few documented studies evaluating the efficacy of wheelchair and customized seating in people with like chronic neurological conditions. There are however, retrospective audits (17), review articles (18, 19) and clinical guidelines (20).

People with FRDA usually present with some degree of scoliosis with associated hypertrophy of back musculature, and pelvic asymmetry. Uncorrected, this can lead to further loss of function, pain, pressure areas and compromised respiratory function (21, 22). Provision of an adjustable contoured seat that has the flexibility to correct pelvic asymmetry by the use of gel or foam inserts can be effective in preventing further deformity (20, 21, 23, 24). Moreover, the use of a contoured padded back support with either or both pelvic and thoracic lateral supports can assist in providing support in a coronal plane (22, 25). Holmes and colleagues (22) identified the efficacy of using four

adjustable lateral pads in managing scoliosis related to cerebral palsy (CP). Whilst the mechanisms of scoliosis in people with FRDA have some differences to those with CP, the intervention may prove just as effective. A correctly positioned four-point lap belt can maintain the person in an optimum position despite a tendency to change position as a response to coughing, laughing or sneezing. De Sousa & Frank (24) describe the prescription of tilt-in-space wheelchairs for people with rare diseases to help manage problematic pain and scoliosis. Consideration should be given to the most appropriate type of wheelchair frame (18). Whilst people with FRDA benefit from a lightweight, responsive model (often with a rigid frame), it is essential it has the strength to manage at times poorly controlled, forceful movement (18, 26, 27). Removable footplates are critical to ensure an optimum sit to stand stance during a standing transfer. Likewise, the use of one-piece lift-off footplates may prove easier to remove and replace as well as achieving optimum positioning for hips, knees and feet.

16.3.3 Provision of powered wheelchairs and scooters

Individuals with FRDA may benefit from provision of either a powered wheelchair or scooter. This equipment is particularly important in facilitating community access for those who find it difficult to wheel distances in a manual wheelchair (28). As discussed previously, careful attention needs to be given to appropriate seating, in addition to the programming, control, stability and storage of the chair and the mode of transfer on and off the chair or scooter (17). In one study (24), there was recognition that whilst some people with FRDA may lose the ability to control their powered wheelchair, many do not and are able to continue to use the wheelchair with standard controls. Modifications to the controls may allow appropriate responsiveness despite a reduction in upper limb co-ordination or strength. With either manual or powered wheelchairs, it is critical that structured training programs are conducted to ensure the user is safe, competent and confident to use the wheelchair (29). It is also important to consider the transportation of a powered wheelchair, particularly if needing to access more distant destinations where modifications to a car would be necessary.

16.3.4 Pediatric versus adult individuals with Friedreich ataxia

It should be noted that the prescription of wheelchairs for children with FRDA raises different issues than those for adults. An earlier age of onset is associated with faster rate of progression to wheelchair use (30, 31). In addition, the repeat size on GAA1 has been associated with the presence of scoliosis, wheelchair dependence, impaired vibration sense and presence of foot deformity (32). The earlier scoliosis is evident, the greater is the likelihood of progression and requirement for surgical intervention (21). Prophylactic management of scoliosis in children is critical even before symptoms are apparent. The goal of seating in both pediatric and adult populations, regardless of evidence of scoliosis, is to maintain the spine in a balanced position in the coronal and sagittal planes over an even pelvis (21). Prescribing a wheelchair for a child has to incorporate requirements for training, growth, postural stability, completion of activities of daily living and school/leisure/home access issues (19).

Best practice statements

Comprehensive assessment to identify barriers to independence should be conducted by a multidisciplinary team to identify appropriate customizable digital and assistive technology to optimize independence and occupational participation and performance, and to enhance quality of life.
Compensatory or remedial interventions with digital and assistive technology may improve independence for individuals with Friedreich ataxia.
Prescription of a manual or powered wheelchair or scooter should be preceded by an assessment of the home/school/work and community environment the equipment will be used in.
A comprehensive prescription of a manual or powered wheelchair or scooter should be completed by a qualified clinician familiar with the specific issues related to Friedreich ataxia.
A validated assessment and evaluation tool for wheelchair and seating prescription may be used to guide the process of prescription and evaluation.
In prescribing a manual wheelchair and seating system, functional capacity should not be impeded for the sake of an anatomically correct seated posture.
Appropriate training should be provided for the safe use of a wheelchair or scooter in the home or community environment.
Suitability of the seating and wheelchair system should be evaluated on an annual basis in adults and biannually in children.

Recommendations

Grading for strength of recommendation and level of evidence

For the rating of the **strength** of the recommendation, in addition to evidence from studies in FRDA, evidence from like conditions, clinical experience and expert consensus are taken into account when published evidence is not available.

The **level of evidence** is based on published evidence from studies in FRDA. If there is no published evidence in FRDA, evidence from other like conditions or clinical expertise may have been used to make the recommendation – this is graded as 'very low' or in some cases 'low' level evidence. See the table below for an explanation of the symbols used to grade recommendations.

Strength of recommendation	Symbol	Level of evidence	Symbol
Strong for intervention	↑↑	High	⊕⊕⊕⊕
Conditional for intervention	↑	Moderate	⊕⊕⊕◯
Neither intervention nor comparison	—	Low	⊕⊕◯◯
Conditional against intervention	↓	Very low	⊕◯◯◯
Strong against intervention	↓↓		

Assistive technology for mobility

Should customized assistive technology for mobility (e.g., modified vehicles, wheelchair, transfer devices, walkers, adaptive seating and positioning) versus non-use be used for individuals with impaired independent gait/mobility with Friedreich ataxia?	Strength	Level of evidence*
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.		

Personal care and environmental control

Should customized assistive technology (personal care technology, environmental control [iPAD, home APPs, smart watches], ALEXA/ SIRI) versus non-use be used for individuals with impaired upper limb functionality with Friedreich ataxia?	Strength	Level of evidence*
For individuals with Friedreich ataxia with impaired upper limb functionality, we suggest the use of customized assistive technology for personal care and environmental control (e.g., iPad, home apps, smart watches, Alexa/Siri) to enhance independence in daily activities, quality of life, and social and occupational participation.	↑	⊕◯◯◯
Justification: Despite the low level of evidence in only one like condition, expert clinicians who provide clinical care for individuals with Friedreich ataxia agree that customized assistive technology for personal care can benefit independence in daily activities, quality of life, and social and occupational participation in individuals with Friedreich ataxia.		
Subgroup considerations: This recommendation is specifically for individuals with Friedreich ataxia with impaired upper limb functionality. The guideline expert panel considers that all individuals with Friedreich ataxia might benefit from the use of digital and assistive technology.		

Health monitoring

Should health monitoring/alert devices (e.g., monitoring heart rate, steps, sleep, ECG, activity, healthy eating, medication, glucose for diabetes) versus non-use be used for individuals with fatigue, fall risk, poor sleep, diabetes, etc, with Friedreich ataxia?	Strength	Level of evidence*
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 Friedreich ataxia-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.		

Communication and workplace assistance

Should customized assistive technology for communication and to expand workplace opportunities (e.g., writing, speech, computer use, tablets, workplace design [adaptive seating and positioning], vision and hearing) versus non-use be used for individuals with impaired communicating ability and workplace difficulties with Friedreich ataxia?	Strength	Level of evidence*
For individuals with Friedreich ataxia with impaired communication and workplace difficulties, we suggest the use of customized assistive technology for communication and the workplace (e.g., writing, speech, computer use, tablets, workplace design [adaptive seating and positioning], vision and hearing) to enhance independence in communication, improve quality of life and increase occupational participation.	↑	⊕◯◯◯
Justification: Despite the low level of evidence in like conditions, expert clinicians who provide clinical care for individuals with Friedreich ataxia agree that customized assistive technology for communication can benefit independence in communication, quality of life and occupational participation in individuals with Friedreich ataxia, particularly in the later stage of the disease.		
Subgroup considerations: The provision of customized assistive technology is particularly relevant to individuals with advanced Friedreich ataxia who may have concurrent issues related to upper limb ataxia, visual difficulties and eye movement problems.		

Lay summary

Lay summary of clinical recommendation for digital and assistive technologies in Friedreich ataxia

Why these recommendations?

These recommendations are about the use of digital and assistive technologies such as modified wheelchairs, adaptive seating, environmental controls, apps and smart watches. These technologies may help individuals with Friedreich ataxia with work, leisure, social and daily activities that involve communication, mobility and upper limb function. They may also be useful for individuals for whom it is important to maintain health and well-being, such as for management of diabetes or maintaining healthy eating.

Due to a lack of studies, there is little direct evidence showing the benefits of using digital and assistive technologies in individuals with Friedreich ataxia. However, the clinical experience of the authors and published research in other neurological conditions supports the consideration of using such technologies.

What does this mean for you as a person living with Friedreich ataxia or caring for someone living with Friedreich ataxia?

It might be important for you to talk to your allied health and medical clinicians to see if the use of digital and assistive technologies may be appropriate for you.

Who are these recommendations specifically for?

These recommendations are relevant to all individuals with Friedreich ataxia, particularly those who have lived with the condition for a long time and have many inter-related issues that may interfere with independence, such as speech, mobility and upper limb problems.

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