

## Clinical Management Guidelines for Friedreich Ataxia

### Chapter 3.2. Strength, balance, mobility and reduction of falls in ambulant individuals with Friedreich ataxia

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## 3.2 Strength, balance, mobility and reduction of falls in ambulant individuals with Friedreich ataxia

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This chapter describes the effects of Friedreich ataxia on strength, balance and mobility and risk of falls, the functional consequences of these effects, and strategies for managing disturbance of strength, balance and mobility and reducing falls risk. In making recommendations for management, the authors were tasked with answering the question:

**For individuals with Friedreich ataxia who are still ambulatory, what management strategies could be implemented for disturbance of strength, balance, mobility and reduction of falls?**

### 3.2.1 Disturbance of strength, balance and mobility in Friedreich ataxia, functional consequences and falls risk

Gait ataxia and clumsiness are usually the presenting symptoms for people with Friedreich ataxia (FRDA) (1, 2). As the disease progresses, running and jumping become challenging, limiting participation in sports and other physical activities. Individuals with FRDA often avoid poor light and uneven terrain (3) during walking or require extra concentration in these environments. As motor function decreases, ambulation is still possible but often with increased reliance on furniture, walls, mobility aids, rails for stairs or family and friends to support walking (3). As gait becomes more unsteady, falls become more frequent and descending stairs is difficult. Further progression may result in dependence on a wheelchair for mobility in some circumstances, such as community access. For individuals with typical FRDA onset, complete dependence on a wheelchair for all mobility occurs approximately 10 to 15 years after disease onset (1). In individuals with late-onset FRDA, progression to complete wheelchair dependence for mobility is slower, approximately 20 years after symptom onset (4).

#### 3.2.1.1 Neurologic basis for mobility decline

Spinal cord, peripheral nerve and dorsal column degeneration occur early in FRDA causing afferent ataxia, which is considered the primary cause of early decline in mobility (5-7). In addition, dorsal column degeneration causes impairment in proprioception (6), which plays a critical role in establishing the timing and magnitude of muscle activity during locomotion (8-10). An important function of the cerebellum is integrating sensory input with voluntary motor action. This ensures coordinated and automated timing, duration and amplitude of muscle activity in normal movement, enabling stable and accurate limb movement. The cerebellum also has a role in the dynamic regulation of balance and adaptation of posture and locomotion through practice and is critical in locomotion (11-13). Hence, cerebellar pathology also contributes to a decline in mobility in people with FRDA.

#### 3.2.1.2 Muscle strength

There is no direct evidence that demonstrates muscle weakness leads to decline in ambulation in FRDA; however, there is evidence suggesting changes to skeletal muscles in individuals who are still ambulant. This includes studies demonstrating deficits in energy production, delayed muscle oxygenation after exercise (14), increased muscle density, increased muscle specific fatigue (15), and reduced muscle strength (16), which can be up to 70% by the time of wheelchair use (17).

### 3.2.1.3 Gait pattern changes

Individuals with FRDA have deficits in gait pattern including reduced velocity, step and stride length, increased double support time, reduced swing phase as a percentage of the gait cycle and wide oscillation center of mass (COM) sway (18-22). Reduced peak ankle plantar flexion and positive values of the hip extensor impulse are also found in children and adolescents with FRDA (20). There is impairment of kinematic temporal inter-joint coordination, including an irregular proximal/distal leading joint alternating pattern within the gait cycle (20). The increase in gait variability is seen in both adults and children with FRDA (18).

### 3.2.1.4 Balance

There is a significant link between mobility decline and impaired balance in individuals with FRDA (19, 21, 23). Although the mechanisms of postural control required to maintain balance differ during static standing, dynamic tasks and gait, locomotion significantly challenges balance. Locomotion requires control of both lateral and forward stability to control a constantly moving center of mass (24). Four studies have examined balance in people with FRDA (21, 25-27). Individuals with FRDA demonstrate an increased sway path and sway area, particularly in the lateral direction (25, 27). This alteration in sway path (27), suggests individuals with FRDA are highly dependent on vision during tasks requiring postural stabilization, and balance decline may be related to spinocerebellar tract and proprioceptive impairment (25). However, significant correlations between the Scale for the Assessment and Rating of Ataxia (SARA) posture subscale and Friedreich Ataxia Rating Scale (FARS) upright stability subscale; and medio-lateral sway, antero-posterior sway and the whole course of the total travelled way of center of pressure during quiet stance (26), suggest that the cerebellum also has a role in controlling sway during quiet stance. Given the role of the cerebellum in the integration of somatosensory, visual and vestibular inputs, this is not surprising. Individuals with FRDA also experience challenges during dynamic balance tasks, including controlling their COM over their base of support (21).

### 3.2.1.5 Falls

There have been no publications reporting the incidence or frequency of falls in individuals with FRDA. However, clinical observations show that frequent falling is common for many people with FRDA. This observation, together with the increased presence of osteopenia or osteoporosis in individuals with FRDA, might result in more common fall-related fractures. Data from the FRDA Clinical Outcome Measures (FA-COMS) registry found 9.7% of individuals (107/1104) had reported a fracture in the year prior. Individuals with FRDA have also identified a fear of falling as a key transitional event related to mobility decline (28). This fear typically arises from a previous fall or near falls and can limit participation in activities and can cause anxiety about social stigmatization (28). Worsening gait ataxia and impaired balance contribute significantly to falling. In addition, clinical observations suggest foot and ankle posture, ankle spasticity and joint instability cause uncontrolled ankle inversion during ambulation, resulting in ankle sprain injury and falls.

## 3.2.2 Management of strength, balance, mobility and reduction of falls

### 3.2.2.1 Monitoring

An annual multidisciplinary review can identify and address the changing needs of an individual with FRDA. Monitoring should include a functional review and an impairment-based assessment to

establish the contributing factors to functional decline. A thorough assessment will allow treatment and management to be appropriate for each individual. Individualization of therapy ensures that the rate and stage of decline; contribution of impairments to functional decline; impact on daily activities; individual goals, preferences and enjoyment; and pro-active and reactive needs are considered.

### 3.2.2.2 Management with rehabilitation, allied health therapies and exercise interventions

Rehabilitation and physiotherapy interventions may be applicable to people with FRDA at varying ages and stages of the disease and may be beneficial both proactively and reactively in delaying, maintaining and improving functional decline (30, 31). Physical activity and exercise are encouraged throughout the lifespan and can be divided into three categories: continuous/maintenance exercise and physical activity; short-term intensive rehabilitation; and targeted therapy. Intensity, type and amount of clinician support may vary between each category, as summarized below.

#### Continuous/maintenance exercise and physical activity

Daily exercise is designed to maintain physical function and prevent any complications caused by deconditioning. Several studies have reported the importance of regular and frequent ongoing training and exercise to maintain improvement in function achieved with short-term rehabilitation (32, 33). Continuous rehabilitation is particularly beneficial if the demands of usual daily activities are not sufficient to maintain or improve physical capabilities (34). Continuous rehabilitation may comprise a home exercise, gym, hydrotherapy (33), standing, videogame-based coordinative training (35, 36), and/or community-based programs. Non-FRDA-specific physical activity programs such as yoga, Pilates and swimming may also be of benefit, especially if they are suitable for the disease stage and encourage enjoyable physical activity. However, there is no published literature examining these activities.

The level of clinician support provided may vary and is dependent on finances, physical distance from suitable venues and/or clinicians, and individual preferences. Individualized and regularly reviewed exercise programs (by a qualified clinician such as physical therapist and/or exercise physiologist) are important to ensure the activities and exercise undertaken are targeted appropriately for each individual. Prescribed programs should consider individual goals, motivation, enjoyment and preferences (37), the disease stage, and presenting impairments.

#### Short-term intensive rehabilitation

A 4- to 8-week period of intensive rehabilitation (defined as 3 or more sessions per week) appears beneficial in improving function for people with FRDA and other degenerative ataxias (38). Theoretically the intensity is thought to provide stimulus to the neuromuscular system (39), but may be difficult to maintain for a longer period due to other life commitments. Rehabilitation may be provided in an inpatient, outpatient or community setting. The benefits of intensive rehabilitation appear to have some carryover into the community and may persist over time; however, there is evidence suggesting that continuous ongoing exercise is required to maintain these benefits (31, 33, 39).

#### Targeted therapy

Targeted therapy is aimed at addressing specific concerns or problems that arise or are identified as immediate risks. They may be identified by a clinician or an individual with FRDA. Issues may include but are not limited to falls, deterioration of function, losing capacity to mobilize, deconditioning post medical or surgical event, pain and musculoskeletal injury. The therapy provided is dependent on

the issue and the findings of a comprehensive assessment of function, and sensory and motor impairments. A neurologist, pediatrician, physical therapist, occupational therapist, podiatrist, orthotist, or a combination of clinicians, may be involved.

### 3.2.2.3 Evidence base for management recommendations

There is emerging and consistent evidence that rehabilitation, allied health therapies and exercise interventions are beneficial for individuals with FRDA (38). However, there is limited and minimal high-quality evidence to guide the type, duration and intensity. The recommendations in this topic aim to guide these aspects of therapy for individuals with FRDA. The following provides a summary of each of the interventions recommended.

#### **Multi-faceted approach to physical therapy treatment**

Most trials examining the effects of rehabilitation for individuals with degenerative ataxias employ rehabilitation programs that address multiple areas of impairment and function (38). This multi-faceted approach is recommended, in part, due to the extensive and diverse peripheral and central neuropathology (40), in conjunction with myopathic and mitochondrial changes in skeletal muscle (14-16), and thus multifactorial cause of locomotor decline in individuals with FRDA. Interventions are not targeted at one impairment, rather include a combination of all or many of the following: lower limb and trunk strengthening, facilitation of normal muscle activity, postural control exercises, balance exercises, functional/mobility retraining and education, cardiovascular and endurance exercises, somatosensory re-training, and stretching.

#### **High intensity (defined as 3+ days per week) exercise**

There is currently no definition of high intensity or low intensity exercise for individuals with FRDA. No studies have compared different intensity, frequency, or durations of exercise; however, anecdotally individuals who are more active and participate in greater amounts of exercise appear to maintain mobility and balance function to a greater extent than individuals who do not participate in regular physical activity. This recommendation defines high intensity as three times or greater sessions of exercise per week. Although there is no evidence to suggest any detrimental effects from exercise, care should be undertaken so as not to “over-exercise”. This is important to manage fatigue, allow muscle recovery and balance the demands of day-to-day activities.

#### **Cardiovascular fitness and endurance exercises**

For the general population, the *American Heart Association Physical Activity Guidelines for Americans* (41) recommends 150 to 300 minutes per week of moderate intensity aerobic activity spread over the week for adults and 60 minutes per day for children aged 6 years and older. There is no evidence to suggest the benefits to health and well-being would not be similar in individuals with FRDA and physical activity may also counteract secondary adaptations caused by decline in mobility and de-conditioning. As cardiomyopathy is present in around 40% of individuals with FRDA (42), a comprehensive assessment and advice from a cardiologist is recommended before commencing physical activity. When considering the type of cardiovascular exercise, choosing functional activities and postures (i.e. walking) may provide additional benefits related to improving or maintaining function. However, the exercises selected should enable the activity to be maintained at the required intensity for the recommended duration.

#### **Lower limb strengthening**

Physical therapy and aerobic training are thought to lessen fatigue and improve strength and well-being. However, there is minimal evidence for isolated strength training in FRDA. Notwithstanding,

four studies have included strength training as one component of their multi-faceted rehabilitation program designed for individuals with inherited cerebellar ataxia (31, 33, 43, 44). This suggests that clinicians believe strengthening may be beneficial for individuals with ataxia.

Several considerations may assist in guiding the strengthening program prescribed for individuals with FRDA:

- Maintaining correct movement patterns and eccentric control may concurrently address ataxia, coordination and strength impairments.
- Facilitation of postural control and/or alignment to isolate control for strengthening may concurrently address ataxia, coordination and strength impairments, as well as allow progressive resistance principles to be applied (45).
- Eccentric control and multi-joint muscle exercises should be considered as they may produce more substantial physiological adaptation of skeletal muscle (46).

### **Foot and ankle stretching and strengthening**

In individuals with FRDA, the presence of cerebellar and afferent ataxia, and impaired balance necessitates therapy that focuses on postural control, coordination and balance exercise. However, the presence of lower limb spasticity, peripheral neuropathy, myopathy, ankle equinovarus deformities and muscle contracture seen in individuals with FRDA, also significantly contributes to difficulties in locomotion and standing (47, 48). Due to poor ankle and foot posture and muscle activity, individuals may have difficulty maintaining adequate foot contact on the ground, controlling movement of the tibia over the foot, generating appropriately timed activity and power for push-off, and maintaining appropriate foot and ankle alignment during the swing phase of gait. This can increase the risk of falls and necessitate the use of mobility aids at an early stage of disease progression.

There is minimal evidence for stretching in individuals with FRDA; a 2010 systematic review found moderate to high quality evidence to indicate stretching had no effect on joint mobility in people with neurological disorders (no participants with FRDA) (49). The stretching intervention examined did not incorporate any active movement or immediate translation into functional activity. Yet, in clinical practice, individuals report improved comfort and better foot positioning following regular completion of a stretching program. For individuals with FRDA, the primary aim of stretching should be to allow functional re-training and strengthening of the ankle musculature in a functional position. Therefore, any stretching program should be accompanied by strengthening and mobility re-training in functional positions for ambulation.

### **Orthotic devices (i.e. braces, ankle-foot-orthotics)**

Ankle foot orthotics (AFOs) may be appropriate to provide mediolateral stability at the ankle in the stance phase of gait, facilitate toe clearance in the swing phase, promote heel strike at initial contact, prevent foot deformity, support normal joint alignment and biomechanics, improve range of motion and facilitate walking function (50, 51). There are no studies specifically looking at orthotic prescription for people with FRDA; however, there are multiple studies describing the benefits of AFO prescription in improving postural security and movement, increasing weight-bearing percentage, improving knee control, reducing toe clawing, improving gait parameters, and improving standing and balance in individuals with other neurological conditions such as stroke and spastic cerebral palsy (50-54).

### Focal pharmacological intervention for lower limb spasticity management

Please refer to Chapter 3.4 for further detail on focal pharmacological interventions for lower limb spasticity in ambulant individuals with FRDA.

#### Best practice statements

Gait aids should be considered for ambulant people with Friedreich ataxia who are at risk of, or fearful of falling. Individualized assessment is required to ensure the gait aid selected provides the support required to facilitate maximal independence but does not restrict available movement or reduce current balance capacity (due to over-dependence on the gait aid to stabilize).

Several studies have highlighted the benefits of computer-based or video-based coordination/balance training for individuals with ataxia (32, 35, 36, 39, 55). These papers highlight the importance of prescribing exercise programs that are enjoyable, motivating and sustainable.

Given the significant decline in balance evident in individuals with Friedreich ataxia, suitably challenging balance exercises should be prescribed in all therapy programs.

#### 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	↓↓		

#### Monitoring

<i>Should monitoring at least once per year versus informal monitoring be used for ambulant individuals with Friedreich ataxia?</i>	Strength	Level of evidence
For individuals with Friedreich ataxia who are ambulant (with or without an aid), we recommend regular monitoring of ambulation and contributing physical and non-physical factors for mobility decline (such	↑↑	⊕○○○



as balance, strength, lower limb spasticity and fear of falling) at least once per year over less regular or informal monitoring.		
<b>Justification:</b> Although there is no published evidence directly addressing the effectiveness of monitoring mobility for ambulant individuals with Friedreich ataxia, annual decline reported in published papers and seen in clinical practice warrants regular (at least annual) monitoring and assessment of mobility. This recommendation is consistent with recommendations made in the 2019 Ataxia UK Medical Guidelines (29).		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

### Multi-faceted rehabilitation

<i>Should a multi-faceted approach to physical therapy treatment versus single focused therapy (i.e. strengthening only) be used for ambulant people with Friedreich ataxia?</i>	<b>Strength</b>	<b>Level of evidence</b>
For ambulant people with Friedreich ataxia, we recommend a multi-faceted rehabilitation approach (targeting multiple areas of impairment) over a single focused rehabilitation approach.	↑↑	⊕⊕⊕○
<b>Justification:</b> This recommendation is based on moderate evidence in favor of a multi-faceted rehabilitation approach and positive short-term outcomes seen in clinical practice.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

### High intensity rehabilitation

<i>Should high intensity (defined as 3+ days per week) exercise versus less intensive (defined as &lt;3 days per week) exercise be used for ambulant people with Friedreich ataxia?</i>	<b>Strength</b>	<b>Level of evidence</b>
For individuals with Friedreich ataxia who are ambulant, we recommend completing rehabilitation or exercises 3 days per week or more frequently over completing these exercises less than 3 days per week.	↑↑	⊕⊕⊕○
<b>Justification:</b> Although there are no studies that have compared three days or greater frequency of exercise versus less than 3 days per week, a systematic review found the average frequency of rehabilitation interventions for individuals with hereditary cerebellar ataxia was 4.4 days per week (38) and three studies have found greater frequency of independent training was associated with better outcomes (32, 33, 35, 39). This is supported by observations in clinical practice.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

### Cardiovascular fitness

<i>Should cardiovascular fitness and endurance exercises versus no treatment be used for ambulant people with Friedreich ataxia?</i>	<b>Strength</b>	<b>Level of evidence</b>
We suggest cardiovascular and endurance exercise training be used over no cardiovascular exercise in ambulant individuals with Friedreich ataxia. Gradual onset and increase in the level of activity, with	↑	⊕⊕⊕○

monitoring for any adverse symptoms, is likely to be a safe approach in those with and without cardiac abnormalities.		
<b>Justification:</b> Many of the studies included cardiovascular/endurance training as a component of a multi-faceted exercise/rehabilitation program, making it hard to determine the effects of individual components of the programs. However, clinical practice and the few studies examining cardiovascular training alone suggest cardiovascular training provides beneficial effects. Although we are providing a conditional recommendation, there is no evidence to suggest cardiovascular training is superior to other targeted exercises and there is no evidence to suggest a particular dosage or intensity of exercise.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

### Lower limb strengthening

<i>Should lower limb strengthening versus no treatment be used for ambulant people with Friedreich ataxia?</i>	Strength	Level of evidence
We suggest lower limb strengthening over no lower limb strengthening in individuals with Friedreich ataxia who are ambulant. In most instances, lower limb strengthening should be one component of a whole exercise or rehabilitation program.	↑	⊕○○○
<b>Justification:</b> There is no published evidence examining the effects of lower limb strength training alone for individuals with Friedreich ataxia. In clinical practice (and some in studies) strength training is included in exercise or rehabilitation programs with an emphasis on correct movement control and pattern with beneficial effects. However, it is difficult to ascertain the direct impact of this management strategy.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

### Foot and ankle stretching

<i>Should foot and ankle stretching and strengthening versus no intervention be used for ambulant people with Friedreich ataxia?</i>	Strength	Level of evidence
We recommend ankle and foot strengthening and stretching in combination over no strengthening or stretching program in individuals with Friedreich ataxia who are ambulant.	↑↑	⊕○○○
<b>Justification:</b> There is no published evidence examining the effects of ankle strengthening and stretching exercises for individuals with Friedreich ataxia. However, in clinical practice ankle strengthening and stretching exercises are included in exercise or rehabilitation programs with an emphasis on full active range of ankle dorsiflexion and eccentric calf activity to allow the appropriate ankle range of movement during stance phase of gait, adequate push-off and ankle dorsiflexion for foot clearance. This needs to occur alongside postural control exercises and a focus on improving balance during single leg stance.		
<b>Subgroup considerations:</b> For ambulant individuals with Friedreich ataxia who present with more severe levels of calf spasticity, stretching and strengthening may provide greater benefit. Orthotics or pharmacological management of spasticity should be considered if strengthening and stretching are not able to address the impact of ankle dysfunction on gait pattern, falls frequency or independence.		

**Orthotic devices**

<i>Should orthotic devices (i.e. braces, ankle-foot-orthotics) versus no orthotic support be used for ambulant people with Friedreich ataxia?</i>	<b>Strength</b>	<b>Level of evidence</b>
In individuals with Friedreich ataxia who are ambulant, we suggest customized orthotics (including lateral ankle support braces, ankle-foot-orthotics) over no orthotic devices when ankle joint kinematics are altered during gait and this has a significant impact on gait. It is important that orthotics are customized and fitted professionally to avoid complications, such as pressure ulcers and incorrectly altered ankle range of motion.	↑	⊕⊕○○
<b>Justification:</b> Although there is no published evidence examining the effect of orthotics on gait and independence of ambulation, expert-based clinical observations indicate benefits for individuals with Friedreich ataxia.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

**Botulinum toxin injection**

<i>Should botulinum toxin injections of the ankle/foot musculature versus no pharmacological intervention be used for ambulant people with ankle/foot spasticity with Friedreich ataxia?</i>	<b>Strength</b>	<b>Level of evidence</b>
We cannot recommend either botulinum toxin injection or no pharmacological therapy for ambulant individuals with Friedreich ataxia with ankle or foot spasticity. In selected cases, botulinum toxin injection could be considered after weighing up the potential benefits and harms related to ambulation and dynamic standing balance and therapy-based treatments (such as physiotherapy) have been tried but have not been completely effective. Clinicians should discuss potential negative effects and ensure the individual is aware of the risks prior to this treatment.	—	⊕○○○
<b>Justification:</b> There are no publications examining the effectiveness of botulinum toxin injections in the ankle or foot muscles in individuals with Friedreich ataxia who are ambulant. Clinical experience indicates some beneficial effects from this treatment, although there are not consistent observations of benefit.		
<b>Subgroup considerations:</b> This recommendation is for individuals with Friedreich ataxia who are ambulant.		

## Lay summary

### **Lay summary of clinical recommendations for strength, balance, mobility and reduction of falls in ambulant individuals with Friedreich ataxia**

#### **Why these recommendations?**

The following recommendations are aimed at maximizing walking and balance ability in people with Friedreich ataxia, as well as preventing any secondary consequences from reduced walking and balance ability.

*Monitoring:* We recommend at least annual monitoring of physical function, balance and ability and quality of walking, for all people with Friedreich ataxia who are walking.

*Management:* There are a number of recommendations related to managing balance, strength and physical function in individuals who are walking. Two are general recommendations and the others are targeted at specific impairments that commonly (but not always) affect walking ability in individuals with Friedreich ataxia.

- We recommend physical therapy/exercise that focuses on multiple areas of the body and multiple impairments (such as balance exercises, strengthening and stretching) over focusing on one area.
- We recommend exercising three times or more per week because the overall benefits may be greater than exercising less often. However, we encourage exercise and physical activity (even if it is less frequent) over doing no exercise.
- We suggest strengthening exercises for the leg muscles as part of a regular exercise program. Strengthening exercises should concentrate on increasing resistance as well as ensuring correct movement patterns.
- Orthotic devices, such as ankle and foot braces, might be used to help support independent walking. This can be assessed by a physical therapist in conjunction with an orthotist.
- If spasticity (abnormal muscle tightness due to prolonged contraction) is present in the leg muscles and is affecting walking ability, the use of medications given by injection may be beneficial. Suitable medications may include botulinum neurotoxin or phenol injections. However, the most appropriate treatment of spasticity should be carefully assessed by a neurologist and physiotherapist. The benefits and side-effects of medication should be discussed.
- We suggest cardiovascular exercise as part of a regular exercise program. Gradual onset and increase in the level of exercise, with monitoring for any adverse symptoms, is likely to be a safe approach.

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

People with Friedreich ataxia who are walking should have regular (at least once per year) monitoring and assessment by a clinician experienced in managing ataxia. There are subtle differences in how walking ability is affected in each person with Friedreich ataxia, therefore, there is no one approach that will work for everyone. Regular contact with a healthcare provider will give you an opportunity to discuss the management options available to improve or maintain your walking ability and to work out which recommendation (or a combination of recommendations) may be best for you. Your healthcare provider/s will ensure that management is tailored specifically for you.

#### **Who are these recommendations specifically for?**

All people with Friedreich ataxia who are walking should have annual monitoring for strength, balance and mobility and have a regular exercise program. Specific management recommendations are for those who are walking but have particular problems of mobility, balance or strength.

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