# Clinical Management Guidelines for Friedreich Ataxia

## Chapter 3.5. Dysarthria in Friedreich ataxia

Contents



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# 3.5 Dysarthria in Friedreich ataxia

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This chapter describes the effects of Friedreich ataxia on speech, the functional consequences of speech impairment (dysarthria), and strategies for managing dysarthria. In making recommendations for management, the authors were tasked with answering the question:

**For individuals with Friedreich ataxia, what management strategies could be implemented for dysarthria?**

## 3.5.1 Effects of Friedreich ataxia on speech and communication

Dysarthria is the core speech disorder in Friedreich ataxia (FRDA) (1, 2). Speech related physiological impairments in FRDA include poor breath support, incoordination of articulators, and slower movement. They lead to reduced intelligibility and naturalness of speech (deviation from healthy norm). Degraded intelligibility can make it difficult to communicate in day-to-day tasks, which is exacerbated when talking on the phone or in noisy environments. As well as motor deficits associated with speech, individuals with FRDA often present with cognitive inflexibility (3) that may lead to difficulties generating ideas, planning and organizing messages, and making inferences from spoken/written information due to the inability to process verbal information at speed (4). Auditory processing deficits are also described in FRDA related to hearing and processing speech and sound in complex noise environments (5). Two other functional domains can potentially impact on communication or access to alternative/augmentative forms of communication (AAC; e.g., unaided systems like gesture or aided systems such as speech generating devices): oculomotor and upper limb performance. Most individuals with FRDA retain adequate visual acuity (6, 7); however, this can change with disease progression. In contrast, many develop ocular motor fixation deficits (8). These deficits, coupled with the loss of upper limb dexterity (9) and dysarthria itself, complicate the use of traditional AAC technologies that rely on speech to text or mouse use on digital devices.

## 3.5.2 Functional consequences of dysarthria

Dysarthria manifests in slower speech, poor vocal control, hypernasality (10), dysphonia (11), and imprecise consonants (12) in the speaker. Dysarthria is debilitating and can render the speaker unintelligible (13). Speech is affected in almost all people with ataxia. Communication deficits can trigger altered self-identity (14) and impede or prevent both social and professional interactions (15). This leads to daily disadvantage and social marginalization through limitations in accomplishing tasks with associated emotional consequences (16, 17) and underemployment (15).

## 3.5.3 Management of dysarthria

There are three potential management strategies designed to improve speech function in Friedreich ataxia: behavioral therapy; alternative and augmentative communication (AAC); and pharmacological treatment.

Behavioral management is based on traditional models of speech therapy that focus on improving the underlying physiologic support for speech (e.g., improving trunk stability and breath support); modifying speech through compensatory speaking strategies (e.g., segmenting phrases and

controlling rate to improve articulatory accuracy); improving clarity and naturalness of speech by practicing appropriate targets; improving self-monitoring; and managing the communication environment (e.g., reducing background noise levels when speaking, conversation partner training). Advice and guidance on improving speech is typically delivered by a therapist or via technology and is delivered intensively in the clinic or home. Intervention may be delivered individually or in a group format.

AAC is designed to provide capacity for individuals to use technology, devices or alternative forms of communication (such as eye gaze, writing) to replace or supplement spoken communication. It is often beneficial to work jointly with colleagues from the multi-disciplinary team, such as physiotherapists for trunk stability and occupational therapists for fine motor control and accessing augmentative communication devices.

Pharmaceutical treatments are commonly designed to halt or reverse disease progression or alleviate the symptoms of disease. However, there are currently no known drugs that specifically target amelioration of speech disorder (18).

## Best practice statement

In the absence of strong evidence supporting widespread adoption of treatment for speech problems, interventions to improve the communication skills of listeners (i.e. communication partners of speakers with ataxia) could be incorporated into care plans. These include focused attention during conversations, communicating in quiet environments, and identification of strategies to ameliorate communication breakdowns, with practice of the strategies in a supportive environment.

## 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 | ↓↓ | | |

### Intensive behavioral interventions

| *Should intensive behavioral speech intervention versus no treatment be used for all people with Friedreich ataxia?* | **Strength** | **Level of evidence*** |
|---|---|---|
| For people with Friedreich ataxia, we suggest the use of targeted intensive behavioral therapy for improving speech in individuals with dysarthria. | ↑ | ⊕⊕◯◯ |
| **Justification:** There is some limited evidence supporting the use of behavioral therapies for improving speech in Friedreich ataxia. Three small non-randomized trials show some preliminary evidence that some aspects of speech can improve in hereditary ataxias following intensive therapy (19-21). These therapies include methods for improving self-monitoring, biofeedback and diverse tasks designed to improve specific aspects of speech like breath support, vocal control and intelligibility. The studies suggest that some gains in intelligibility or voice quality can be achieved with intensive treatment. However, the data are derived from non-controlled, underpowered pilot studies. | | |
| **Subgroup considerations:** This recommendation is for individuals with Friedreich ataxia with dysarthria. | | |

### Augmentative and alternative communication

| *Should augmentative and alternative communication (AAC) versus no AAC be used for all people with Friedreich ataxia?* | **Strength** | **Level of evidence*** |
|---|---|---|
| We cannot recommend either the use or non-use of AAC to treat dysarthria in individuals with Friedreich ataxia. | — | ⊕◯◯◯ |
| **Justification:** There are no trials or published evidence supporting the use of AAC in Friedreich ataxia. | | |
| **Subgroup considerations:** This recommendation is for individuals with Friedreich ataxia with dysarthria. AAC may be best suited to individuals who have severe dysarthria and are not receptive to behavioral or pharmacological interventions. | | |

### Pharmacological interventions

| *Should pharmaceutical intervention versus no pharmaceutical intervention be used for all people with Friedreich ataxia?* | **Strength** | **Level of evidence*** |
|---|---|---|
| For people with Friedreich ataxia, we suggest that pharmaceutical therapies are *not* used to treat dysarthria. | ↓ | ⊕◯◯◯ |
| **Justification:** Very few published studies have used speech as an outcome measure in pharmaceutical trials for Friedreich ataxia. There is very low evidence supporting the use of any pharmaceutical therapies to improve dysarthria in Friedreich ataxia. One open label trial showed minor changes in acoustic outcomes related to timing in the high dose group (versus low dose group) (22). These findings were not verified against listener-based judgements. | | |
| **Subgroup considerations:** This recommendation is for individuals with Friedreich ataxia with dysarthria. | | |

## Lay summary

### Lay summary of clinical recommendations for dysarthria in Friedreich ataxia

Individuals with Friedreich ataxia often develop problems with their speech (dysarthria). There are some potential treatments for speech problems, but as yet there is not enough research to show the benefits of some of these possible treatments.

Behavioural treatment is currently the primary management option available to individuals with Friedreich ataxia who have problems with their speech. There is some evidence suggesting that intensive drill-based therapy (regular and frequent therapy) delivered for at least three weeks may be effective in improving some aspects of speech, including intelligibility (how well a person can be understood by a listener) and voice quality.

**Why these recommendations?**

Augmentative and alternative communication (AAC) is the use of technology (such as speech to text), devices (often needing the use of a mouse) or alternative forms of communication (such as eye gaze, writing) to replace or help spoken communication. There are currently studies investigating the use of AAC for improving communication in Friedreich Ataxia, but there is not yet enough evidence showing benefit to people with Friedreich ataxia who have problems with their speech to recommend its use.

There are no studies showing that any medication can help to improve speech in Friedreich ataxia.

Behavioural treatments for dysarthria are supported by some preliminary studies suggesting they can help to improve speech in individuals with Friedreich ataxia. Behavioural therapy is typically safe and is very unlikely to lead to any harm to the individual.

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

For individuals with dysarthria it is important to have the type and severity of your speech problems assessed, and to discuss options for intensive behavioural therapies with your doctor or other health professional. These therapies are focussed on improving intelligibility and naturalness of speech.

**Who are these recommendations specifically for?**

These recommendations are for Individuals with Friedreich ataxia with changes to their speech.

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