

QUESTION

Should assessment should consider (cardiac cause, low threshold for X-ray due to spasticity) vs. standard assessment be used for people presenting with trauma due to a fall with Friedreich ataxia?

POPULATION:	people presenting with trauma due to a fall with Friedreich ataxia
INTERVENTION:	assessment should consider (cardiac cause, low threshold for X-ray due to spasticity)
COMPARISON:	standard assessment
MAIN OUTCOMES:	Accurate diagnosis; Unnecessary testing;

ASSESSMENT

Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> No <input type="radio"/> Probably no <input checked="" type="radio"/> Probably yes <input type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>People with FRDA are more likely to suffer fractures from falls due to osteopenia/osteoporosis (Eigentler et al, 2014)</p> <p>Musculoskeletal injuries resulting from falls may also exacerbate underlying spasticity or present with painful muscle spasms.</p> <p>Reference</p> <p>Eigentler A, et al. Low bone mineral density in Friedreich ataxia. <i>Cerebellum</i> 2014;13(5):549-57.</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of fever, chest infections and UTIs.</p> <p>4/6 indicated that the problem was serious, 1/6 indicated probably not serious, 1/6 indicated they didn't know if serious.</p> <p>4/6 indicated that the problem was urgent, 1/6 indicated probably not urgent, 1/6 indicated they didn't know if urgent.</p> <p>4/6 indicated that the problem was a priority, 1/6 indicated probably a priority, 1/6 indicated they didn't know if a priority. (Aug 2020)</p>

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> Trivial <input type="radio"/> Small <input checked="" type="radio"/> Moderate <input type="radio"/> Large <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>A search of four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 14 December 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ Large ○ Moderate ○ Small ● Trivial ○ Varies ○ Don't know 	<p>A search of four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 14 December 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	

Certainty of evidence

What is the overall certainty of the evidence of effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ Very low ○ Low ○ Moderate ○ High ● No included studies 	<p>No published research.</p>	

Values

Is there important uncertainty about or variability in how much people value the main outcomes?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS									
<ul style="list-style-type: none"> ○ Important uncertainty or variability ○ Possibly important uncertainty or variability ● Probably no important uncertainty or variability ○ No important uncertainty or variability 	<table border="1"> <thead> <tr> <th>Outcomes</th> <th>Importance</th> <th>Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Accurate diagnosis - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> <tr> <td>Unnecessary testing - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Accurate diagnosis - not measured	CRITICAL ^a	-	Unnecessary testing - not measured	CRITICAL ^a	-	
Outcomes	Importance	Certainty of the evidence (GRADE)									
Accurate diagnosis - not measured	CRITICAL ^a	-									
Unnecessary testing - not measured	CRITICAL ^a	-									

	a. Identified as critical by expert authors on this topic.	
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Balance of effects

Does the balance between desirable and undesirable effects favor the intervention or the comparison?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<input type="radio"/> Favors the comparison <input type="radio"/> Probably favors the comparison <input type="radio"/> Does not favor either the intervention or the comparison <input checked="" type="radio"/> Probably favors the intervention <input type="radio"/> Favors the intervention <input type="radio"/> Varies <input type="radio"/> Don't know	No published research.	

Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<input type="radio"/> No <input type="radio"/> Probably no <input checked="" type="radio"/> Probably yes <input type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know	No published research.	Clinical experience suggests it would be acceptable to patients with FRDA.

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

JUDGEMENT							
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 ○
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CONCLUSIONS

Recommendation

We suggest thorough and careful multi-disciplinary assessment of the causes and effects of falls should be performed for individuals with Friedreich ataxia presenting to the emergency department with a fall over standard assessment, taking into consideration factors such as neurological progression, cardiac arrhythmia, hypotension, uncontrolled or newly presenting diabetes. More advanced imaging such as CT or MRI scan may be necessary to assess for any spinal cord or nerve root compression, or for complications related to any prior scoliosis surgery, such as rod infection or migration.

Justification

There is no published evidence examining the best assessment or management of falls in individuals with FRDA. However, a thorough assessment is clinically appropriate.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia presenting to an emergency department with trauma.

Research priorities

Further examination of falls frequency, alongside the causes and resulting injuries and other sequelae for individuals with FRDA, is warranted to help understand options for prevention and management.

Reference

Eigentler A, Nachbauer W, Donnemiller E, Poewe W, Gasser RW, Boesch S. Low bone mineral density in Friedreich ataxia. *Cerebellum*. 2014;13(5):549-57.