

## QUESTION

Should management should consider (intra-operative management of orthopaedic trauma, fast return to weight-bearing and minimising immobilisation where possible) vs. standard management be used for people presenting with trauma with Friedreich ataxia?

POPULATION:	people presenting with trauma with Friedreich ataxia
INTERVENTION:	management should consider (intra-operative management of orthopaedic trauma, fast return to weight-bearing and minimising immobilisation where possible)
COMPARISON:	standard management
MAIN OUTCOMES:	Disease progression; Independence of function; Independence of mobility;

## ASSESSMENT

<b>Problem</b> Is the problem a priority?		
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> No</li> <li><input type="radio"/> Probably no</li> <li><input checked="" type="radio"/> Probably yes</li> <li><input type="radio"/> Yes</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	There is no published evidence.	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.  4/6 indicated that the problem was serious, 1/6 indicated probably not serious, 1/6 indicated they didn't know if serious.  4/6 indicated that the problem was urgent, 1/6 indicated probably not urgent, 1/6 indicated they didn't know if urgent.  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)
<b>Desirable Effects</b> How substantial are the desirable anticipated effects?		
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> Trivial</li> <li><input type="radio"/> Small</li> <li><input checked="" type="radio"/> Moderate</li> <li><input type="radio"/> Large</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	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.	Although there is no published evidence, the effects of prolonged immobilisation and hospitalisation can have an impact on functional independence and can increase the rate of decline towards wheelchair dependency. Therefore, minimising the impact of immobilisation can be significant for individuals with FRDA.

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**Undesirable Effects**  
How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li>○ Large</li> <li>● Moderate</li> <li>○ Small</li> <li>○ Trivial</li> <li>○ Varies</li> <li>○ Don't know</li> </ul>	<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>	<p>There are risks associated with surgery and unprotected or early weight-bearing, including wound infection, pain and increased falls.</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"> <li>○ Very low</li> <li>○ Low</li> <li>○ Moderate</li> <li>○ High</li> <li>● No included studies</li> </ul>	<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"> <li>○ Important uncertainty or variability</li> <li>○ Possibly important uncertainty or variability</li> <li>● Probably no important uncertainty or variability</li> <li>○ No important uncertainty or variability</li> </ul>	<table border="1" style="width: 100%; text-align: center;"> <tr> <td style="width: 33%;"><b>Outcomes</b></td> <td style="width: 33%;"><b>Importance</b></td> <td style="width: 33%;"><b>Certainty of the evidence (GRADE)</b></td> </tr> </table>	<b>Outcomes</b>	<b>Importance</b>	<b>Certainty of the evidence (GRADE)</b>	
<b>Outcomes</b>	<b>Importance</b>	<b>Certainty of the evidence (GRADE)</b>			

	Disease progression - not measured	IMPORTANT <sup>a</sup>	-
	Independence of function - not measured	IMPORTANT <sup>a</sup>	-
	Independence of mobility - not measured	IMPORTANT <sup>a</sup>	-
a. Identified as important by expert authors on this topic.			

### Balance of effects

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

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> Favors the comparison</li> <li><input type="radio"/> Probably favors the comparison</li> <li><input type="radio"/> Does not favor either the intervention or the comparison</li> <li><input type="radio"/> Probably favors the intervention</li> <li><input type="radio"/> Favors the intervention</li> <li><input checked="" type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	<p>There is no published evidence related to management of orthopaedic injuries or trauma in individuals with FRDA.</p>	<p>Individual risk factors and potential benefits of reducing the time immobilised after trauma need to be discussed with the individual and carefully weighed up. The potential impact on functional capacity and preventing mobility decline should be considered.</p>

### Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> No</li> <li><input type="radio"/> Probably no</li> <li><input checked="" type="radio"/> Probably yes</li> <li><input type="radio"/> Yes</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	<p>No published research.</p>	<p>Anecdotally individuals with FRDA report significant concern over loss of independence due to immobilisation or weight-bearing restrictions.</p>

## 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 <input type="radio"/>	Conditional recommendation against the intervention <input type="radio"/>	Conditional recommendation for either the intervention or the comparison <input type="radio"/>	Conditional recommendation for the intervention <input checked="" type="radio"/>	Strong recommendation for the intervention <input type="radio"/>
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## CONCLUSIONS

### Recommendation

We suggest that trauma management approaches that minimize the time spent immobilized might be considered, with careful consideration of the risks and benefits related to each individual with Friedreich ataxia.

### Justification

There is no published evidence in FRDA. However, secondary consequences associated with reduced mobility and immobilization for individuals with FRDA are clear.

## Subgroup considerations

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

## Research priorities

Understanding the incidence and cause of emergency department admissions. Outcomes following trauma-related orthopedic surgery and non-surgical management would help to understand the risks for people with FRDA.