

QUESTION

Should oral medications vs. none be used for pain with Friedreich ataxia?

POPULATION:	pain with Friedreich ataxia
INTERVENTION:	oral medications
COMPARISON:	none
MAIN OUTCOMES:	Reduction in pain; QOL; ADLs; Ambulation;

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>Bodily pain is a problem for individuals with FRDA (Pérez-Flores et al, 2020).</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of non-neuropathic pain.</p> <p>3/7 indicated that the problem was serious, 2/7 indicated probably serious, 1/7 indicated probably not serious, 1/7 indicated they didn't know if serious.</p> <p>2/7 indicated that the problem was urgent, 3/7 indicated probably urgent, 1/7 indicated probably not urgent, 1/7 indicated they didn't know if urgent.</p> <p>2/7 indicated that the problem was a priority, 2/7 indicated probably a priority, 2/7 indicated probably not a priority, 1/7 indicated they didn't know if 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 type="radio"/> Moderate <input checked="" 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 02 August 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 02 August 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 evidence.</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" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th style="width: 40%;">Outcomes</th> <th style="width: 20%;">Importance</th> <th style="width: 40%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td style="text-align: center;">Reduction in pain - not measured</td> <td style="text-align: center;">IMPORTANT^a</td> <td style="text-align: center;">-</td> </tr> <tr> <td style="text-align: center;">QOL - not measured</td> <td style="text-align: center;">IMPORTANT^b</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Reduction in pain - not measured	IMPORTANT ^a	-	QOL - not measured	IMPORTANT ^b	-	
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Ambulation - not measured	IMPORTANT ^c	-						
	<p>a. Identified as critical (1/6), important (3/6) and low importance (2/6) by people with FA and important by expert authors on this topic.</p> <p>b. Identified as critical (3/6) and important (3/6) by people with FA and important by expert authors on this topic.</p> <p>c. Identified as critical (2/6), important (3/6) and low importance (1/6) by people with FA and important by expert authors on this topic.</p>							

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"> <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 evidence.	

Acceptability

Is the intervention acceptable to key stakeholders?

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 	No published evidence.	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if using oral medications for pain was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>2/4 indicated the intervention was acceptable, 2/4 indicated probably acceptable. (Aug 2020).</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 ○	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 conditionally recommend the use of oral medication over no medication to manage pain in individuals with Friedreich ataxia.

Justification

Despite there being no published evidence directly related to the use of medication for pain management in FRDA, the expert authors agree that the negative effects of pain on quality of life, mobility and participation in daily activities support proactive management of pain by medication. Consideration needs to be given to side-effects of pain medication that may exacerbate existing issues, such as reduced balance, poor mobility, falls risk and increased fatigue, and people with FRDA taking medication should be monitored for possible adverse effects.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia who report pain.

Research priorities

Further research is required to ascertain the most effective management strategies for non-neuropathic pain in Friedreich ataxia.

Reference

Perez-Flores J, Hernandez-Torres A, Monton F, Nieto A. Health-related quality of life and depressive symptoms in Friedreich ataxia. *Qual Life Res.* 2020;29(2):413-20.