

QUESTION

Should enquiring about bowel symptoms vs. none be used for all patients with Friedreich ataxia?

POPULATION:	all patients with Friedreich ataxia
INTERVENTION:	enquiring about bowel symptoms
COMPARISON:	none
MAIN OUTCOMES:	Constipation; Bowel (faecal) incontinence;

ASSESSMENT

Problem

Is the problem a priority?

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	In a study of 59 individuals with FRDA, 64% (n = 38) reported bowel symptoms (Lad et al, 2017).	Clinical experience indicates individuals with FRDA may experience constipation.

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<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	A search of four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 13 July 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.	Enquiring about bowel symptoms could help to: <ol style="list-style-type: none"> 1. Establish the presence of these, often embarrassing symptoms, which may not be spontaneously voiced by the patients 2. Establish whether there is a bowel contribution to urinary or other symptoms 3. Identify and treat symptoms at an early stage before they become more problematic (potentially impacting bowel continence, appetite) Aside from QOL factors, identifying constipation, through asking for bowel symptoms, may result in treatment interventions to avoid potential complications including hemorrhoids, anal fissures, fecal impaction, bowel incontinence, rectal prolapse,

bowel obstruction and urinary incontinence.

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 13 July 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 unlikely to be any undesirable effects from enquiring about bowel symptoms.</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: 50%;">Outcomes</th> <th style="width: 20%;">Importance</th> <th style="width: 30%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Constipation - not measured</td> <td>IMPORTANT^a</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Constipation - not measured	IMPORTANT ^a	-	
Outcomes	Importance	Certainty of the evidence (GRADE)						
Constipation - not measured	IMPORTANT ^a	-						

	<table border="1"> <tr> <td>Bowel (faecal) incontinence - not measured</td> <td>IMPORTANT^a</td> <td>-</td> </tr> </table>	Bowel (faecal) incontinence - not measured	IMPORTANT ^a	-	
Bowel (faecal) incontinence - not measured	IMPORTANT ^a	-			
	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"> <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.	

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

	JUDGEMENT						
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 conditionally recommend that clinicians enquire about the presence of bowel symptoms when consulting individuals with Friedreich ataxia.

Justification

Clinical experience indicates that bowel symptoms may adversely affect quality of life in individuals with Friedreich ataxia and herald an increased risk of complications, such as hemorrhoids, anal fissures, fecal impaction, bowel incontinence, rectal prolapse, bowel obstruction and urinary incontinence. Enquiring about bowel symptoms may lead to timely intervention to alleviate symptoms and avoid complications.

Subgroup considerations

None

Research priorities

Examining the acceptability and utility of enquiry regarding bowel symptoms in individuals with Friedreich ataxia.

References

Lad M, Parkinson MH, Rai M, Pandolfo M, Bogdanova-Mihaylova P, Walsh RA, et al. Urinary, bowel and sexual symptoms in a cohort of patients with Friedreich's ataxia. *Orphanet J Rare Dis.* 2017;12(1):158.