

QUESTION

Should counselling or therapy vs. none or antidepressant prescription be used for depressed patients with Friedreich ataxia?

POPULATION:	depressed patients with Friedreich ataxia
INTERVENTION:	counselling or therapy
COMPARISON:	none or antidepressant prescription
MAIN OUTCOMES:	Less depression; Better coping/ outlook; Reduced vegetative symptoms; QOL;

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 type="radio"/> Probably yes <input checked="" type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>In a cohort of 62 individuals with FA the mean BDI score was 15.32 (SD=10.33). Minimal depressive symptoms were observed in 50% of participants, 23% of participants scored in the “mild” range, 23% had moderate depressive symptoms, and 5% scored in the “severe” range (Perez-Flores et al 2020). In a cohort of 57 adults with FA the mean BDI score for FRDA patients was 13.62 (SD = 9.52) which is significantly higher than the mean score in the general population ($t(56) = 3.15, p < .01$), according to populations norms (Neito et al 2018),</p> <p>Depression varies: one study using the Beck Depression Inventory (BDI-II) showed the following frequencies for each level of depression: 0–9 (score normal) 21/33 patients, 63.6%; 10–18: mild depression 6/33 patients, 18.2%; 19–29: moderate depression 5/33 patients, 15.1%; 30–63: severe depression 1/33 patient, 3% (Alexandra Durr, personal communication, 2021).</p>	<p>The Friedreich’s ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of depression.</p> <p>5/6 indicated that the problem was serious, 1/6 indicated they didn’t know if serious.</p> <p>4/6 indicated that the problem was urgent, 1/6 indicated probably urgent, 1/6 indicated probably not urgent.</p> <p>4/6 indicated that the problem was a priority, 2/6 indicated probably 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 type="radio"/> Moderate <input checked="" type="radio"/> Large <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>A search of five databases (CENTRAL, MEDLINE, EMBASE, CINAHL, PsycINFO) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 29 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>If counseling is efficacious then the desirable effect on individuals who are depressed would be large.</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 five databases (CENTRAL, MEDLINE, EMBASE, CINAHL, PsycINFO) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 29 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>Need to consider issues of physical accessibility of services in regards to possible undesirable effects of counseling.</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"> <thead> <tr> <th>Outcomes</th> <th>Importance</th> <th>Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Less depression - not measured</td> <td>IMPORTANT^a</td> <td>-</td> </tr> <tr> <td>Better coping/ outlook - not measured</td> <td>IMPORTANT^b</td> <td>-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Less depression - not measured	IMPORTANT ^a	-	Better coping/ outlook - not measured	IMPORTANT ^b	-	<p>Clinical experience that people with FA value the outcome of counseling, particularly telephone counseling.</p>
Outcomes	Importance	Certainty of the evidence (GRADE)									
Less depression - not measured	IMPORTANT ^a	-									
Better coping/ outlook - not measured	IMPORTANT ^b	-									

	Reduced vegetative symptoms - not measured	CRITICAL ^c	-
	QOL - not measured	CRITICAL ^d	-
<p>a. 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> <p>b. 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> <p>c. Identified as critical (3/6), important (2/6) and low importance (1/6) by people with FA and important by expert authors on this topic.</p> <p>d. Identified as critical (3/6) and important (3/6) 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.	Clinical experience of benefit of counseling – often considered as first step in treatment, or in conjunction with treatment with medication.

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 type="radio"/> Probably yes <input checked="" 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 counselling or therapy for people with depression was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>4/4 indicated that the intervention was 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 recommend counseling or therapy over no counseling in individuals with Friedreich ataxia who present with symptoms of depression.

Justification

Cohort studies indicate that the presence of depressive symptoms may be greater in individuals with FRDA than those without FRDA. Whilst there is no published evidence in FRDA, if counseling is efficacious then the desirable effect on individuals who are depressed would be large. Given individuals with FRDA may be reluctant to recognize symptoms of depression, the first line of clinical management may be counseling. It is important clinicians explore what will work best for individual with FRDA, particularly in the context of the other aspects of their lives (accessibility requirements of face to face counseling, time constraints for work/study/family, etc). The clinician may need to consider treating depressive symptoms with both counseling and medication. In addition, it is important for the clinician to be aware of the role of fatigue in depression (and perhaps presenting as depression).

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia and depression. It may be difficult for a person with severe dysarthria to engage in counseling. Counseling may be very helpful at major life stages such as going to college, leaving home etc. Counseling may also be appropriate for parents/carers of individuals with Friedreich ataxia.

Research priorities

Finding ways to offer counseling in manner that is acceptable and user friendly for individuals with FRDA, such as via telephone.

Trying to understand the separation between FRDA and depression-related fatigue.

References

Nieto A, Hernández-Torres A, Pérez-Flores J, Montón F. Depressive symptoms in Friedreich ataxia. *Int J Clin Health Psychol*. 2018;18(1):18-26.

Pérez-Flores J, Hernández-Torres A, Montón F, et al. Health-related quality of life and depressive symptoms in Friedreich ataxia. *Qual Life Res* 2020;29(2):413–420.