

QUESTION

Should additional support services as psychology, psychologist, ethicist vs. no additional support services be used for minors at risk of Friedreich ataxia?

POPULATION:	minors at risk of Friedreich ataxia
INTERVENTION:	additional support services as psychology, psychologist, ethicist
COMPARISON:	no additional support services
MAIN OUTCOMES:	Anxiety;

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>Nil</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of genetic testing of minor siblings.</p> <p>2/7 indicated that the problem was serious, 2/7 indicated probably serious, 1/7 indicated they didn't know if serious, 2/7 indicated varied or sometimes serious.</p> <p>2/7 indicated that the problem was urgent, 2/7 indicated probably not urgent, 2/7 indicated they didn't know if urgent, 1/7 indicated varied or sometimes serious.</p> <p>2/7 indicated that the problem was a priority, 2/7 indicated probably not a priority, 2/7 indicated they didn't know if a priority, 1/7 indicated varied or sometimes 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, PsycINFO) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 20 August 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	<p>Clinical experience indicates that minors at risk of FRDA may benefit from psychological support in relation to being at risk.</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, PsycINFO) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 20 August 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	<p>It is expected that psychological support is unlikely to have undesirable effects but it is possible that discussion of at-risk status could be a source of anxiety for the individual.</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: 33%;">Outcomes</th> <th style="width: 33%;">Importance</th> <th style="width: 33%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Anxiety - not measured</td> <td>IMPORTANT^a</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Anxiety - not measured	IMPORTANT ^a	-	<p>It is likely that different individuals will have different need or desire for such intervention.</p>
Outcomes	Importance	Certainty of the evidence (GRADE)						
Anxiety - not measured	IMPORTANT ^a	-						

	a. Identified as important 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
<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.	<p>A survey designed to systematically collect expert-based opinions from clinicians involved in developing the recommendations for this topic and providing clinical care for individuals with Friedreich ataxia, was conducted. Clinical experts from Australia, Europe, UK, South America, Canada and the USA were asked to consider the harms/benefits of Additional support services such as psychology/psychologist/ethicist as a management strategy for minors at risk of Friedreich ataxia.</p> <p>Reflecting on the impact of Additional support services such as psychology/psychologist/ethicist on Psychological harm/benefit, 75% (3/4) clinical experts reported a benefit (large, moderate or small), 25% (1/4) reported no effect and, 0% (0/4) reported observing a harm (large, moderate or small).</p> <p>Reflecting on the impact on Anxiety, 75% (3/4) clinical experts reported a benefit, 25% (1/4) reported no effect.</p> <p>Clinical experience indicates additional support services can be beneficial for those at risk of FRDA.</p>

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
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 suggest minors at risk of Friedreich ataxia (siblings of people with Friedreich ataxia) should be offered psychological support to assist with dealing with anxiety that may arise from being at risk of developing the condition.

Justification

Being at risk of FRDA may result in considerable psychological distress for the individual.

Subgroup considerations

This recommendation is for siblings (minors) of people with Friedreich ataxia. The need for psychological support may vary with the age of the sibling and their testing status, particularly as the sibling moves from being an immature to a mature minor. This should be assessed on an individual basis.

Research priorities

A study of siblings about this topic would be of interest.