

QUESTION

Should education regarding possible cognitive impairment vs. no education regarding possible cognitive impairment be used for all individuals with Friedreich ataxia?

POPULATION: all individuals with Friedreich ataxia

INTERVENTION: education regarding possible cognitive impairment

COMPARISON: no education regarding possible cognitive impairment

MAIN OUTCOMES: Improved information;

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>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of the topic.</p> <p>1/7 indicated disturbance of cognitive function was probably not serious, 3/7 indicated probably serious, 3/7 indicated serious.</p> <p>4/7 indicated disturbance of cognitive function was probably not urgent, 3/7 indicated urgent.</p> <p>2/7 indicated disturbance of cognitive function was probably not a priority, 2/7 indicated probably a priority, 2/7 indicated priority, 1/7 indicated varies/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 five databases (CENTRAL, MEDLINE, EMBASE, CINAHL, PsycINFO) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 18 September 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 can be assumed from other rare disease that education assists with cognitive problems. It appears that individuals with FRDA are unaware of cognitive impairment and the potential impacts on vocational and academic issues. It is particularly important at adolescence when critical decisions are being made about vocation prospects. Therefore, education would provide at least moderate desirable effects; however, there are no known educational packages to implement.</p>

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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 18 September 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 providing education about cognitive effects of FRDA. On the other hand, not identifying a deficit or providing education may contribute to increased psychological burden and not provide the individual with FRDA every opportunity to maximise their potential.</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%; text-align: center;"> <tr> <td style="width: 33%;">Outcomes</td> <td style="width: 33%;">Importance</td> <td style="width: 33%;">Certainty of the evidence (GRADE)</td> </tr> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	
Outcomes	Importance	Certainty of the evidence (GRADE)			

	Improved information - not measured	IMPORTANT ^a	-	
<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>				

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"> ○ 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 	<p>No published evidence.</p>	<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 Education of people with FRDA regarding possible cognitive impairment as a management strategy for all individuals with Friedreich Ataxia. Reflecting on the impact of Education of people with FRDA regarding possible cognitive impairment on Improved information, 100% (3/3) clinical experts reported a benefit (large, moderate or small).</p> <p>Clinical experts were asked to consider the harms/benefits of Education of the parents/carers/family of people with FRDA regarding possible cognitive impairment as a management strategy for all individuals with Friedreich Ataxia. Reflecting on the impact of Education of the parents/carers/family of people with FRDA regarding possible cognitive impairment on Improved information, 100% (3/3) clinical experts reported a benefit (large, moderate or small).</p> <p>Clinical experts were asked to consider the harms/benefits of Education of Teachers or employers of people with FRDA regarding possible cognitive impairment as a management strategy for all individuals with Friedreich Ataxia. Reflecting on the impact of Education of Teachers or employers of people with FRDA regarding possible cognitive impairment on Improved information, 100% (3/3) clinical experts reported a benefit (large, moderate or small).</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 type="radio"/> Probably yes <input type="radio"/> Yes <input checked="" 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 the intervention was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>1/5 indicated education regarding possible cognitive impairment for all people was not reasonable, 1/5 indicated probably not reasonable, 2/5 indicated reasonable, 1/5 indicated didn't know if reasonable. (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 <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 development and implementation of an educational program for affected individuals and their families and carers to improve information about the potential for, and management of, cognitive dysfunction in individuals with Friedreich ataxia.

Justification

There is increasing evidence of cognitive dysfunction in individuals with Friedreich ataxia that has the potential to affect academic, vocational and interpersonal pursuits. There is a need to identify and address this potential, particularly as individuals with Friedreich ataxia may be unaware of the possibility of cognitive dysfunction.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia and their parents, carers, partners and family.

Research priorities

Key research priorities include evaluating the understanding of potential cognitive impairment in both affected individuals and their families including identification of the requirements of an educational program, development of an educational program and testing of the efficacy of the program.