

QUESTION

Should chest physiotherapy vs. no intervention be used for people with respiratory weakness and restrictive lung disease with Friedreich ataxia?

POPULATION:	people with respiratory weakness and restrictive lung disease with Friedreich ataxia
INTERVENTION:	chest physiotherapy
COMPARISON:	no intervention
MAIN OUTCOMES:	Respiratory function; Prevalence of chest infection; Dyspnea; Airway clearance function;

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>Preliminary data collected by one of the authors indicates decline in respiratory volumes in severe FA and also impaired cough mechanisms.</p> <p>Patient survey indicates that this is considered to be a significant problem by many patients.</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of pulmonary function.</p> <p>6/7 indicated that the problem was serious, 1/7 indicated they didn't know if serious.</p> <p>6/7 indicated that the problem was urgent, 1/7 indicated they didn't know if urgent.</p> <p>6/7 indicated that the problem was a priority, 1/7 indicated they didn't know if 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 type="radio"/> Large <input type="radio"/> Varies <input checked="" 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 28 October 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 	A search of four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 28 October 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.	

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 	No published evidence.	

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>Respiratory function - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> <tr> <td>Prevalence of chest infection - not measured</td> <td>CRITICAL^b</td> <td>-</td> </tr> <tr> <td>Dyspnea - not measured</td> <td>CRITICAL^b</td> <td>-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Respiratory function - not measured	CRITICAL ^a	-	Prevalence of chest infection - not measured	CRITICAL ^b	-	Dyspnea - not measured	CRITICAL ^b	-	
Outcomes	Importance	Certainty of the evidence (GRADE)												
Respiratory function - not measured	CRITICAL ^a	-												
Prevalence of chest infection - not measured	CRITICAL ^b	-												
Dyspnea - not measured	CRITICAL ^b	-												

	<table border="1" data-bbox="520 107 1417 180"> <tr> <td data-bbox="520 107 1012 180">Airway clearance function - not measured</td> <td data-bbox="1012 107 1142 180">CRITICAL^b</td> <td data-bbox="1142 107 1417 180">-</td> </tr> </table> <p data-bbox="562 220 1388 321"> a. Identified as critical (4/6), important (1/6) and low importance (1/6) by people with FA and critical by expert authors on this topic. b. Identified as critical (3/6), important (1/6) and low importance (2/6) by people with FA and critical by expert authors on this topic </p>	Airway clearance function - not measured	CRITICAL ^b	-	
Airway clearance function - not measured	CRITICAL ^b	-			

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 data-bbox="520 597 716 618">No published evidence.</p> <p data-bbox="520 646 1388 699">Based on FA expert opinion, only 10/26 had an opinion. Majority believe that chest PT has beneficial impact on respiratory function, on prevalence of chest infections and dyspnea.</p>	<p data-bbox="1442 597 2003 789">A survey designed to systematically collect expert-based opinions from clinicians involved in the development of these guidelines 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 referral for chest physiotherapy as a management strategy for people with respiratory weakness.</p> <p data-bbox="1442 816 2003 951">Reflecting on the impact of referral for chest physiotherapy on <u>respiratory function</u>, 38.46% (10/26) clinical experts reported a benefit (large, moderate or small), and 0% (0/26) reported observing a harm (large, moderate or small). 16 clinicians could not provide any information on this outcome.</p> <p data-bbox="1442 979 2003 1053">Reflecting on the impact on <u>prevalence of chest infections</u>, 38.46% (10/26) clinical experts reported a benefit. 16 expert clinicians could not provide any information on this outcome.</p> <p data-bbox="1442 1081 2003 1187">Reflecting on the impact on <u>dyspnea</u>, 34.62% (9/26) clinical experts reported a benefit, 3.85% (1/26) reported no effect and, 0% (0/26) reported observing a harm. 16 expert clinicians could not provide any information on this outcome.</p> <p data-bbox="1442 1214 2003 1289">Reflecting on the impact on <u>airway clearance</u> function, 38.46% (10/26) clinical experts reported a benefit. 16 expert clinicians could not provide any information on this outcome.</p>

Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
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<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 chest physiotherapy in people with weak breathing and restrictive lung disease was acceptable (weighing up the balance between benefits, harms and costs).</p> <p>2/3 indicated the intervention was acceptable, 1/3 indicated probably acceptable. (Aug 2020).</p>
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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

In people with respiratory weakness and restrictive lung disease with Friedreich ataxia, we conditionally recommend chest physiotherapy to improve respiratory function, reduce prevalence of chest infection, reduce dyspnea and improve airway clearance function.

Justification

There are no published data on restrictive lung disease and interventions in FRDA. A patient survey and unpublished data from one of the authors indicate that this occurs in some patients with advanced FRDA. A survey of experts suggests that chest physiotherapy provides benefit for respiratory function, chest infections, and airway clearance.

Subgroup considerations

This recommendation is for people with Friedreich ataxia with respiratory weakness and restrictive lung disease. Chest physiotherapy may be particularly useful in the event of an acute superimposed medical issue such as surgery or chest infection.

Research priorities

Further research is required to establish the efficacy of chest physiotherapy to improve respiratory function, reduce prevalence of chest infection, reduce dyspnea and improve airway clearance function in people with Friedreich ataxia and with respiratory weakness and restrictive lung disease.