QUESTION

Should upper limb splinting/orthoses vs. no splinting/orthoses be used for individuals with spasticity, spasm or contracture with Friedreich ataxia?	
POPULATION:	individuals with spasticity, spasm or contracture with Friedreich ataxia
INTERVENTION:	upper limb splinting/orthoses
COMPARISON:	no splinting/orthoses
MAIN OUTCOMES:	Presence/severity of contracture, range of motion; Pain; Independence in daily activities;
SETTING:	
PERSPECTIVE:	
BACKGROUND:	
CONFLICT OF INTERESTS:	

ASSESSMENT

Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ○ Probably yes ● Yes ○ Varies ○ Don't know		The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of the topic. 8/8 indicated upper limb dysfunction was serious. 1/7 indicated upper limb dysfuction was not urgent; 1/7 indicated probably not urgent; 1/7 indicated probably urgent; 4/7 indicated urgent. 1/7 indicated upper limb dysfunction was probably not a priority, 3/7 indicated probably a priority, 3/7 indicated priority. (Aug 2020)

Desirable Effects

How substantial are the desirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS

○ Trivial ○ Small ● Moderate ○ Large ○ Varies ○ Don't know	A search of 4 databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 16 September 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.	Clinical experience indicates judicious use of customised orthoses may assist in the management of spasticity and prevention of contracture.

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ Large ○ Moderate ○ Small ● Trivial ○ Varies ○ Don't know	A search of 4 databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 16 September 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.	Clinical experience indicates trivial if any undesirable effects provided general principles of prescribing orthoses are adhered to.

Certainty of evidence

What is the overall certainty of the evidence of effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ 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
○ Important uncertainty or variability ○ Possibly important uncertainty or variability		

○ Probably no important uncertainty or variability ● No important uncertainty or variability		

Outcomes	Importance	Certainty of the evidence (GRADE)
Presence/severity of contracture, range of motion - not measured	IMPORTANT[a]	-
Pain - not measured	CRITICAL[b]	-
Independence in daily activities - not measured	IMPORTANT[c]	-

a. Identified as critical (2/6), important (2/6) and low importance (2/6) by people with FA and critical by expert authors on this topic
b. Identified as critical (3/6), important (1/6), low importance (2/6) by people with FA and critical by expert authors on this topic.
c. Identified as critical (3/6) and important (3/6) by people with FA and 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
○ 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	No published evidence.	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 **upper limb splinting/orthoses as a management strategy for individuals with spasticity, spasm, contracture**. Reflecting on the impact of **upper limb splinting/orthoses** on Presence/severity of contracture, range of motion, 100% (4/4) clinical experts reported a benefit (large, moderate or small), and 0% (0/4) reported observing a harm (large, moderate or small). Reflecting on the impact on Pain, 100% (4/4) clinical experts reported a benefit. Reflecting on the impact on Improvement in daily activities, 75% (3/4) clinical experts reported a benefit, 25% (1/4) reported no

		effect.

Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
○ No ○ Probably no ● Probably yes ○ Yes ○ Varies ○ Don't know	No published evidence.	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). 1/4 indicated that upper limb splinting was probably reasonable, 1/4 indicated reasonable, 2/4 indicated that they didn't know if reasonable. (Aug 2020).

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
○	○	○	●	○

CONCLUSIONS

Recommendation

We conditionally recommend considering upper limb splinting/orthoses for individuals with Friedreich ataxia who experience spasticity, spasm or contracture.

Justification

Clinical experience indicates judicious use of customised orthoses may assist in the management of spasticity and prevention of contracture. Upper limb splinting/orthoses should be an adjunct to other therapies such as a hand exercise program incorporating stretch and strengthening (as indicated). Consideration should also be given to ensuring the device does not interfere with active movement opportunities. An assessment of sensation, skin integrity and the ability to monitor the correct device positioning (either self-monitoring or a carer) should also inform decision making

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with upper limb spasticity, spasm or contracture.

Research priorities

Further research into the effect of customised splinting and orthoses for management of upper limb spasticity, prevention of contracture, range of motion, pain and activities of daily living in individuals with FRDA to guide clinical use.