

QUESTION

Should early/aggressive (out of bed) physical therapy vs. standard post-operative management be used for patients undergoing surgery with Friedreich ataxia?

POPULATION:	patients undergoing surgery with Friedreich ataxia
INTERVENTION:	early/aggressive (out of bed) physical therapy
COMPARISON:	standard post-operative management
MAIN OUTCOMES:	Function; Safety of care - pressure injuries/falls; Quality of life; Length of hospital stay;
BACKGROUND:	

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>Low mobility levels are prevalent in the hospitalized adult population (Jones et al, 2020).</p> <p>The negative consequence of bed rest affecting the cardiovascular, respiratory, gastrointestinal, musculoskeletal, renal, endocrine and nervous systems are well documented (Graf, 2006).</p> <p>Prolonged immobility is associated with adverse outcomes such as functional decline, prolonged length of stay, falls, and an increased risk of mortality (Jones et al, 2020).</p> <p>'Standard post-operative management' in regards to mobilization can vary considerably across settings and institutions. Early mobilization would involve a patient participating in out of bed activity as early as possible once deemed medically appropriate.</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of the topic. 6 out of 7 indicated the consequences of fluid and operative management was serious; 1 out of 7 was probably not serious. 6 out of 7 indicated fluid and operative management was urgent; 1 out of 7 was probably not urgent. 5 out of 7 indicated fluid and operative management was a priority; 1 out of 7 was probably a priority; 1 out of 7 was probably not a priority. (July 2020)</p> <p>Concern regarding delay in getting someone out of bed due to the complexity of presentation.</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, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 30 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>Depends on medical context.</p> <p>FA-phobia/fear of moving ataxia patient. The effects of early mobilization including maintenance of mobility level and physical ability are important in maintaining a patient's level of function and independence on discharge from the hospital setting. These desirable outcomes are somewhat dependent on the medical</p>

		context.
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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 four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 30 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>After surgery need to consider ataxia/difficulties in movement that may cause instability in bone healing/stability – staples, rod placement.</p> <p>Following surgery, it is important to gain medical clearance. Consideration needs to be given to the impact that significant ataxia may have on musculoskeletal stability (ie after rod placement or in fracture management). Ensuring appropriate physical support and assistance, environmental set-up and equipment should be considered to minimise the undesirable effects.</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: 50%;">Outcomes</th> <th style="width: 20%;">Importance</th> <th style="width: 30%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td style="text-align: center;">Function - not measured</td> <td style="text-align: center;">CRITICAL^a</td> <td style="text-align: center;">-</td> </tr> <tr> <td style="text-align: center;">Safety of care - pressure injuries/falls - not measured</td> <td style="text-align: center;">IMPORTANT^b</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Function - not measured	CRITICAL ^a	-	Safety of care - pressure injuries/falls - not measured	IMPORTANT ^b	-	<p>Pérez-Flores, et al. (2020) found that health related quality of life is significantly reduced in FRDA patients. Scores in all dimensions of health related quality of life measured were significantly lower than scores in the general population. This suggests certainty that people value quality of life as an important outcome.</p> <p>Additionally, Xiong et al. (2020) found that the physical domains of the SF-36 HRQOL were the most affected from the patient</p>
Outcomes	Importance	Certainty of the evidence (GRADE)									
Function - not measured	CRITICAL ^a	-									
Safety of care - pressure injuries/falls - not measured	IMPORTANT ^b	-									

	Quality of life - not measured	IMPORTANT ^c	-	perspective. This suggest certainty that people value physical functioning as an important outcome.
	Length of hospital stay - not measured	IMPORTANT ^d	-	
<ul style="list-style-type: none"> a. Completing everyday activities independently identified as critical (1/3) and as important (2/3) for decision-making by people with FA and function identified as critical by expert authors on this topic. b. Falls identified as critical (3/3) for decision-making by people with FA and safety of care identified as important by expert authors on this topic. c. Identified as critical (1/3) and as important (2/3) for decision-making by people with FA and identified as important by expert authors on this topic. d. Identified as critical (2/3) and as important (1/3) for decision-making by people with FA and identified as 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
<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.	In most cases the importance of preventing factors such as reduced physical functioning, falls and increased length of hospital stay associated with prolonged bed rest outweighs the possible undesirable effects.

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.	The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if the intervention was reasonable (weighing up the balance between benefits, harms and costs). 1 out of 3 individuals indicated early/agreesive physical therapy after surgery was probably reasonable; 2 out of 3 was sometimes reasonable. (August 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 ○
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CONCLUSIONS

Recommendation

We suggest early mobilization following surgery over standard post-operative management for individuals with Friedreich ataxia undergoing surgery.

We cannot recommend a particular approach to the timing of getting someone out of bed post-surgery; mobilize as soon as possible with consideration for the following: cardiac/medical status, mobility needs, and decreased reserve of strength.

Justification

The complex clinical features of FRDA and the decreased strength reserve predispose this population to deconditioning and deterioration in physical functioning with prolonged immobility. There is no clinical reason to limit early mobilization in the FRDA population, other than individual circumstances. Early mobilization is recommended to promote maintenance of physical functioning and independence during and after hospitalisation. However, this does not mean there should be early discharge from hospital.

The resources, hospital staff assistance and care-giver support that may be required to allow early mobilization to take place need to be considered.

Subgroup considerations

This recommendation is for individuals with FRDA undergoing surgery. For non-ambulant (hoist transfer dependent) individuals with FRDA, a hoist transfer to sit out of bed is a form of mobilization.

Research priorities

There is currently no available evidence to support the timing or the type of post-surgical mobilization in the FRDA population. Research in this area is warranted to support clinical practice.

References

Graf C. Functional decline in hospitalized older adults. *Am J Nurs.* 2006;106(1):58-67

Jones RA, Merkle S, Ruvalcaba L, Ashton P, Bailey C, Lopez M. Nurse-led mobility program: Driving a culture of early mobilization in medical-surgical nursing. *J Nurs Care Qual.* 2020;35(1):20-6.

Perez-Flores J, Hernandez-Torres A, Monton F, Nieto A. Health-related quality of life and depressive symptoms in Friedreich ataxia. *Qual Life Res.* 2020;29(2):413-20.

Xiong E, Lynch AE, Corben LA, Delatycki MB, Subramony SH, Bushara K, et al. Health related quality of life in Friedreich Ataxia in a large heterogeneous cohort. *J Neurol Sci.* 2020;410:116642.