

QUESTION

Should rhythm control vs. rate control be used for atrial fibrillation/flutter with Friedreich ataxia?	
POPULATION:	atrial fibrillation/flutter with Friedreich ataxia
INTERVENTION:	rhythm control
COMPARISON:	rate control
MAIN OUTCOMES:	Morbidity - stroke; Mortality; Quality of life;
BACKGROUND:	
CONFLICT OF INTERESTS:	

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>Atrial fibrillation is common in individuals with FRDA (Harding et al, 1983; Ribaï et al, 2007).</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 cardiac arrhythmias was serious; 1 out of 7 was probably serious. 6 out of 7 indicated cardiac arrhythmias was urgent; 1 out of 7 was probably urgent. 6 out of 7 indicated cardiac arrhythmias was a priority; 1 out of 7 was probably a priority. (July 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 checked="" type="radio"/> Large <input type="radio"/> Varies <input type="radio"/> Don't know 	<p>A search of three databases (CENTRAL, MEDLINE, EMBASE) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 16 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>Clinical practice – see symptomatic benefit</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 three databases (CENTRAL, MEDLINE, EMBASE) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 16 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>Ablation – higher risks: stroke.</p> <p>Medication – moderate risks.</p> <p>Prolonged anesthesia</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"> <thead> <tr> <th>Outcomes</th> <th>Importance</th> <th>Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Morbidity - stroke - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> <tr> <td>Mortality - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Morbidity - stroke - not measured	CRITICAL ^a	-	Mortality - not measured	CRITICAL ^a	-	
Outcomes	Importance	Certainty of the evidence (GRADE)									
Morbidity - stroke - not measured	CRITICAL ^a	-									
Mortality - not measured	CRITICAL ^a	-									

	JUDGEMENT						
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 conditionally recommend attempts to maintain a normal cardiac rhythm over rate control in individuals with Friedreich ataxia and atrial tachyarrhythmias, and also recommend consideration of ablation for those who remain severely symptomatic due to a persistent atrial tachyarrhythmia or frequent paroxysms of an atrial tachyarrhythmia.

Justification

Highly symptomatic patients who are younger in particular require careful consideration regarding intervention. Some consideration should be given for moderate risks of pharmacological intervention versus higher risks with ablation, including prolonged anaesthesia.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with atrial fibrillation/flutter, with particular considerations as described in the justification above.

Research priorities

References

Harding AE, Hewer RL. The heart disease of Friedreich's ataxia: a clinical and electrocardiographic study of 115 patients, with an analysis of serial electrocardiographic changes in 30 cases. *Q J Med.* 1983;52(208):489-502.

Ribaï P, Pousset F, Tanguy M, Rivaud-Pechoux S, Le Ber I, Gasparini F, et al. Neurological, cardiological, and oculomotor progression in 104 patients with Friedreich ataxia during long-term follow-up. *Arch Neurol.* 2007;64:558-64.