

QUESTION

Should enquiring about sexual function including erectile or vaginal lubrication dysfunction vs. no enquiry be used for all sexually active people with Friedreich ataxia?

POPULATION:	all sexually active people with Friedreich ataxia
INTERVENTION:	enquiring about sexual function including erectile or vaginal lubrication dysfunction
COMPARISON:	no enquiry
MAIN OUTCOMES:	Presence of erectile dysfunction; Presence of inadequate vaginal lubrication; Quality of life; Improved intimate relationships;

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>Data from the FA Clinical Outcome Measures (FA-COMS) registry found 3.3% (15/456) females and 6.8% (30/439) males reported sexual dysfunction (Lynch, 2017).</p> <p>Two studies exploring sexual dysfunction in FA:</p> <p>Sexual dysfunction reported in 30/36 (83%) of individuals with FA (Lad et al, 2017)</p> <p>Sexual functioning, sexual satisfaction and the capacity to form intimate relationships is impacted by FA as evident by: erectile dysfunction reported in 57% (20/35) of males, inadequate vaginal lubrication interfering with sexual responsiveness in 57.7% (26/45) of females, and reduced genital sensation in 47% (51/107) of people with Friedreich ataxia. In addition, 88% (94/107) reported problems moving their body during sexual activity and 73%, (78/107) reported reduced confidence about their sexuality due to FRDA. (Corben et al, 2021)</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.</p> <p>3/7 indicated disturbance of sexual function was probably not serious, 3/7 indicated probably serious, 1/7 indicated didn't know if serious.</p> <p>4/7 indicated disturbance of sexual function was probably not urgent, 1/7 indicated probably urgent, 2/7 indicated didn't know if urgent.</p> <p>3/7 indicated disturbance of sexual function was probably not a priority, 1/7 indicated probably a priority, 2/7 indicated didn't know if a 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 four databases (CENTRAL, MEDLINE, EMBASE, CINAHL) identified no randomized, non-randomized controlled, cohort and case studies published from 2014 through to 15 October 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</p>	<p>Given the findings of two studies exploring sexual function in FRDA the desirable anticipated effects of enquiry about sexual dysfunction is moderate.</p>

--	--	--

Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> Large <input type="radio"/> Moderate <input type="radio"/> Small <input type="radio"/> Trivial <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 15 October 2020. No further published evidence meeting the search criteria was identified in the Consensus Clinical Management Guidelines for Friedreich's ataxia, 2014.</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"> <input checked="" type="radio"/> Very low <input type="radio"/> Low <input type="radio"/> Moderate <input type="radio"/> High <input type="radio"/> 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"> <input type="radio"/> Important uncertainty or variability <input type="radio"/> Possibly important uncertainty or variability <input type="radio"/> Probably no important uncertainty or variability <input checked="" type="radio"/> No important uncertainty or variability 	<table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr style="background-color: #f2f2f2;"> <th style="width: 50%;">Outcomes</th> <th style="width: 15%;">Importance</th> <th style="width: 35%;">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Presence of erectile dysfunction - not measured</td> <td>IMPORTANT^a</td> <td style="text-align: center;">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Presence of erectile dysfunction - not measured	IMPORTANT ^a	-	
Outcomes	Importance	Certainty of the evidence (GRADE)						
Presence of erectile dysfunction - not measured	IMPORTANT ^a	-						

	<table border="1"> <tr> <td>Presence of inadequate vaginal lubrication - not measured</td> <td>IMPORTANT^b</td> <td>-</td> </tr> <tr> <td>Quality of life - not measured</td> <td>IMPORTANT^c</td> <td>-</td> </tr> <tr> <td>Improved intimate relationships - not measured</td> <td>IMPORTANT^d</td> <td>-</td> </tr> </table>	Presence of inadequate vaginal lubrication - not measured	IMPORTANT ^b	-	Quality of life - not measured	IMPORTANT ^c	-	Improved intimate relationships - not measured	IMPORTANT ^d	-	
Presence of inadequate vaginal lubrication - not measured	IMPORTANT ^b	-									
Quality of life - not measured	IMPORTANT ^c	-									
Improved intimate relationships - not measured	IMPORTANT ^d	-									
	<p>a. Identified as important (4/6), low importance (1/6) and requiring more information (1/6) by people with FA and important by expert authors on the topic.</p> <p>b. Identified as important (3/6), low importance (2/6) and requiring more information (1/6) by people with FA and important by expert authors on the topic.</p> <p>c. Identified as critical (2/6), important (4/6) by people with FA and important by expert authors on the topic.</p> <p>d. Identified as critical (2/6), important (2/6), low importance (1/6) and requiring more information (1/6) by people with FA and important by expert authors on the 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"> <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 	Expert opinion based on five clinicians favours the intervention despite no published evidence.	

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 checked="" type="radio"/> Probably yes <input type="radio"/> Yes 	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

<ul style="list-style-type: none"> ○ Varies ○ Don't know 		<p>and costs).</p> <p>2/5 indicated enquiring about sexual function for all sexually active people with FA was probably reasonable, 1/5 indicated varies or sometimes reasonable, 2/5 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 type="radio"/>	Strong recommendation for the intervention <input checked="" type="radio"/>
---	--	---	--	---

CONCLUSIONS

Recommendation

We recommend clinicians enquire about sexual function, including but not limited to erectile or vaginal lubrication dysfunction, the physical capacity to engage in sexual activity and the psychological aspect of the sexual response in sexually active individuals with Friedreich ataxia.

Justification

There are two recent published studies that indicate sexual dysfunction is a greater issue in individuals with Friedreich ataxia than those without (Corben et al, 2021; Lad et al, 2017). It is important that the clinician enquires about the possibility to ensure appropriate intervention can be implemented aimed at improving intimate relationships.

Subgroup considerations

This recommendation is for sexually active individuals with Friedreich ataxia.

Research priorities

Further studies are required to confirm the findings of the first two studies regarding the incidence and extent of reported sexual dysfunction in individuals with Friedreich ataxia.

References

Corben LA, Hermans MM, Marks A, Crowe LM, Delatycki MB. Sexual function, intimate relationships and Friedreich ataxia. *J Neurol*. 2021;268(3):1088-95.

Lad M, Parkinson MH, Rai M, Pandolfo M, Bogdanova-Mihaylova P, Walsh RA, et al. Urinary, bowel and sexual symptoms in a cohort of patients with Friedreich's ataxia. *Orphanet J Rare Dis*. 2017;12(1):158.

Lynch D. FA Clinical Outcome Measures (FA-COMS) Registry (unpublished data): clinicaltrials.gov; 2017 [Available from: <https://clinicaltrials.gov/ct2/show/NCT03090789>