

QUESTION

Should counselling for sexual dysfunction vs. no counselling be used for all sexually active people with report sexual dysfunction with Friedreich ataxia?

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|----------------|---|
| POPULATION: | all sexually active people with report sexual dysfunction with Friedreich ataxia |
| INTERVENTION: | counselling for sexual dysfunction |
| COMPARISON: | no counselling |
| MAIN OUTCOMES: | Quality of life; Improved self-esteem; Improved management of physical or sensory impairment compromising sexual function; 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 FRDA (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 findings of two studies exploring sexual function in FA the desirable anticipated effects of counselling for sexual dysfunction is moderate.</p> |

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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 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"> ● 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%; text-align: center;"> <tr> <td style="width: 50%;">Outcomes</td> <td style="width: 15%;">Importance</td> <td style="width: 35%;">Certainty of the evidence (GRADE)</td> </tr> </table> | Outcomes | Importance | Certainty of the evidence (GRADE) | |
| Outcomes | Importance | Certainty of the evidence (GRADE) | | | |

| | | | | | | | | | | | | | | |
|---|---|--------------------------------|------------------------|---|-------------------------------------|------------------------|---|---|------------------------|---|--|------------------------|---|--|
| | <table border="1"> <tr> <td>Quality of life - not measured</td> <td>IMPORTANT^a</td> <td>-</td> </tr> <tr> <td>Improved self-esteem - not measured</td> <td>IMPORTANT^b</td> <td>-</td> </tr> <tr> <td>Improved management of physical or sensory impairment compromising sexual function - 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> <p>a. Identified as critical (2/6) and important (4/6) by people with FA and important by expert authors on the topic.</p> <p>b. Identified as critical (2/6), important (3/6) 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 (3/6) and requiring more information (1/6) 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> | Quality of life - not measured | IMPORTANT ^a | - | Improved self-esteem - not measured | IMPORTANT ^b | - | Improved management of physical or sensory impairment compromising sexual function - not measured | IMPORTANT ^c | - | Improved intimate relationships - not measured | IMPORTANT ^d | - | |
| Quality of life - not measured | IMPORTANT ^a | - | | | | | | | | | | | | |
| Improved self-esteem - not measured | IMPORTANT ^b | - | | | | | | | | | | | | |
| Improved management of physical or sensory impairment compromising sexual function - not measured | IMPORTANT ^c | - | | | | | | | | | | | | |
| Improved intimate relationships - not measured | IMPORTANT ^d | - | | | | | | | | | | | | |

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 | No published evidence. | The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were asked if the intervention was |

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|--|--|---|
| <ul style="list-style-type: none"> <input type="radio"/> Probably yes <input checked="" type="radio"/> Yes <input type="radio"/> Varies <input type="radio"/> Don't know | | <p>acceptable (weighing up the balance between benefits, harms and costs).</p> <p>4/5 indicated counselling for sexual dysfunction for all sexually active people who report sexual dysfunction was probably reasonable, 1/5 indicated didn't know if reasonable. (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

| | | | | |
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| <p>Strong recommendation against the intervention</p> <p><input type="radio"/></p> | <p>Conditional recommendation against the intervention</p> <p><input type="radio"/></p> | <p>Conditional recommendation for either the intervention or the comparison</p> <p><input checked="" type="radio"/></p> | <p>Conditional recommendation for the intervention</p> <p><input type="radio"/></p> | <p>Strong recommendation for the intervention</p> <p><input type="radio"/></p> |
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CONCLUSIONS

Recommendation

We cannot recommend either counselling or no counselling to improve quality of life, self-esteem, management of physical or sensory impairment compromising sexual function, or intimate relationships in sexually active people with Friedreich ataxia reporting sexual dysfunction.

Justification

There is little evidence supporting the role for counselling in the management of FRDA-related sexual dysfunction, although there are studies in other cohorts that suggest a possible role.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia who report sexual dysfunction.

Research priorities

Future research should focus on not only the benefits of counselling for sexual dysfunction in Friedreich ataxia but also which patients would benefit from counselling, at what time point (i.e., prior to a specific intervention), and by whom. In addition, further research is required to identify alternative strategies in case counselling is not appropriate.

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>