

## QUESTION

### Should bladder scan vs. standard assessment be used for people presenting with UTI with Friedreich ataxia?

POPULATION:	people presenting with UTI with Friedreich ataxia
INTERVENTION:	bladder scan
COMPARISON:	standard assessment
MAIN OUTCOMES:	UTI symptoms; Hospitalisations; Accurate diagnosis;

## ASSESSMENT

### Problem

Is the problem a priority?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> No</li> <li><input type="radio"/> Probably no</li> <li><input checked="" type="radio"/> Probably yes</li> <li><input type="radio"/> Yes</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	<p>Lad and colleagues (2017) reported 80% of individuals with Friedreich ataxia over the age of 16 years report lower urinary tract Symptoms. Although the incidence or frequency of emergency department presentation due to UTIs has not been reported, risk factors for UTI, including neurogenic bladder (Lad et al, 2017) and diabetes mellitus (Cnop et al, 2013), have an increased prevalence in individuals with FRDA.</p>	<p>The Friedreich's ataxia Clinical Management Guideline Patient and Parent Advisory Panel were interviewed on the consequences, urgency and priority of fever, chest infections and UTIs.</p> <p>4/6 indicated that the problem was serious, 1/6 indicated probably not serious, 1/6 indicated they didn't know if serious.</p> <p>4/6 indicated that the problem was urgent, 1/6 indicated probably not urgent, 1/6 indicated they didn't know if urgent.</p> <p>4/6 indicated that the problem was a priority, 1/6 indicated probably a priority, 1/6 indicated they didn't know if 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"> <li><input type="radio"/> Trivial</li> <li><input type="radio"/> Small</li> <li><input checked="" type="radio"/> Moderate</li> <li><input type="radio"/> Large</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	<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 14 December 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 experience indicates individuals with FA may experience urinary retention that requires assessment and intervention.</p>

### Undesirable Effects

How substantial are the undesirable anticipated effects?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li>○ Large</li> <li>○ Moderate</li> <li>● Small</li> <li>○ Trivial</li> <li>○ Varies</li> <li>○ Don't know</li> </ul>	<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 14 December 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"> <li>○ Very low</li> <li>○ Low</li> <li>○ Moderate</li> <li>○ High</li> <li>● No included studies</li> </ul>	<p>No published research.</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"> <li>○ Important uncertainty or variability</li> <li>○ Possibly important uncertainty or variability</li> <li>○ Probably no important uncertainty or variability</li> <li>● No important uncertainty or variability</li> </ul>	<table border="1" data-bbox="520 1187 1421 1479"> <thead> <tr> <th data-bbox="527 1192 940 1271">Outcomes</th> <th data-bbox="947 1192 1119 1271">Importance</th> <th data-bbox="1125 1192 1415 1271">Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td data-bbox="527 1276 940 1344">UTI symptoms - not measured</td> <td data-bbox="947 1276 1119 1344">CRITICAL<sup>a</sup></td> <td data-bbox="1125 1276 1415 1344">-</td> </tr> <tr> <td data-bbox="527 1349 940 1417">Hospitalisations - not measured</td> <td data-bbox="947 1349 1119 1417">IMPORTANT<sup>b</sup></td> <td data-bbox="1125 1349 1415 1417">-</td> </tr> <tr> <td data-bbox="527 1422 940 1479">Accurate diagnosis - not measured</td> <td data-bbox="947 1422 1119 1479">CRITICAL<sup>a</sup></td> <td data-bbox="1125 1422 1415 1479">-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	UTI symptoms - not measured	CRITICAL <sup>a</sup>	-	Hospitalisations - not measured	IMPORTANT <sup>b</sup>	-	Accurate diagnosis - not measured	CRITICAL <sup>a</sup>	-	
Outcomes	Importance	Certainty of the evidence (GRADE)												
UTI symptoms - not measured	CRITICAL <sup>a</sup>	-												
Hospitalisations - not measured	IMPORTANT <sup>b</sup>	-												
Accurate diagnosis - not measured	CRITICAL <sup>a</sup>	-												

	<ul style="list-style-type: none"> <li>a. Identified as critical by expert authors on this topic.</li> <li>b. Identified as important by expert authors on this topic.</li> </ul>	
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### 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"> <li><input type="radio"/> Favors the comparison</li> <li><input type="radio"/> Probably favors the comparison</li> <li><input type="radio"/> Does not favor either the intervention or the comparison</li> <li><input checked="" type="radio"/> Probably favors the intervention</li> <li><input type="radio"/> Favors the intervention</li> <li><input type="radio"/> Varies</li> <li><input type="radio"/> Don't know</li> </ul>	No published research.	Given the increased prevalence of lower urinary tract symptoms in individuals with FRDA, for people presenting with UTI symptoms consideration should be given to reviewing catheterisation technique if indicated and performing diagnostic evaluation to assess for elevated residual urine (including a bladder scan) and morphological causes (including an ultrasound of the kidneys, ureters and bladder (KUB) or outpatient cystoscopy).

### Acceptability

Is the intervention acceptable to key stakeholders?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <li><input type="radio"/> No</li> <li><input type="radio"/> Probably no</li> <li><input type="radio"/> Probably yes</li> <li><input type="radio"/> Yes</li> <li><input type="radio"/> Varies</li> <li><input checked="" type="radio"/> Don't know</li> </ul>	No published research.	

## SUMMARY OF JUDGEMENTS

PROBLEM	JUDGEMENT						
	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

	JUDGEMENT						
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	<b>No important uncertainty or variability</b>			
BALANCE OF EFFECTS	Favors the comparison	Probably favors the comparison	Does not favor either the intervention or the comparison	<b>Probably favors the intervention</b>	Favors the intervention	Varies	Don't know
ACCEPTABILITY	No	Probably no	Probably yes	Yes		Varies	<b>Don't know</b>

## 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 ○	<b>Strong recommendation for the intervention ●</b>
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## CONCLUSIONS

### Recommendation

The assessment of UTIs in the emergency department for individuals with Friedreich ataxia should not be fundamentally different from standard care in individuals without Friedreich ataxia. However, we recommend reviewing self-catheterization technique if appropriate and performing diagnostic evaluation to assess for elevated residual urine (including a bladder scan) and morphological causes (including an ultrasound of the kidneys, ureters and bladder (KUB) or outpatient cystoscopy).

### Justification

Individuals with FRDA have risk factors for increased incidence of UTIs including neurogenic bladder and diabetes mellitus. The presence of urinary retention could indicate that FRDA symptoms are getting worse rather than a symptom of UTI. Individuals with FRDA and lower urinary tract symptoms should therefore be screened for progression of the disease, including having a bladder scan in addition to assessment for the presence of UTI.

### Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with urinary tract infection. Bladder scans may be contraindicated if there is open skin or a wound in the suprapubic region. In addition, care needs to be taken with bladder scans in pregnant women to ensure the bladder, not the uterus, is visualized.

## Research priorities

Future research examining the frequency of presentations of individuals with FRDA to the emergency department with UTIs, alongside its treatment and prevention strategies and outcomes, would strengthen any recommendations.

### References

Cnop M, Mulder H, Igoillo-Esteve M. Diabetes in Friedreich ataxia. *J Neurochem*. 2013;126 Suppl 1:94-102.

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.