

QUESTION

Should hospice support vs. usual care be used for people with advanced heart failure in Friedreich ataxia?	
POPULATION:	people with advanced heart failure in Friedreich ataxia
INTERVENTION:	hospice support
COMPARISON:	usual care
MAIN OUTCOMES:	Patient quality of life; Caregiver quality of life; Patient and/or caregiver satisfaction ; Health care utilization/ cost; Health care utilization/cost; Collaboration with neurologist ;
SETTING:	
PERSPECTIVE:	
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 					While heart failure is a significant illness which causes a significant mortality, it is not the biggest problem that those with FRDA have. Therefore, hospice support for heart failure alone, although important, is not the only need of this group.
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 					Heart failure may cause an admission in this group but it is more likely that the person with FRDA will be admitted as their family are no longer able to care for them – doing transfers, toileting, feeding changing – causing lack of dignity for the person with FRDA.
	Outcomes	No of	Certainty of	Relative	Anticipated absolute effects* (95% CI)

	participants (studies) Follow-up	the evidence (GRADE)	effect (95% CI)	Risk with usual care	Risk difference with hospice support
Patient quality of life - not measured	-	-	-	-	-
Caregiver quality of life - not measured	-	-	-	-	-
Patient and/or caregiver satisfaction - not measured	-	-	-	-	-
Health care utilization/ cost assessed with: Readmission rate	0 (2 observational studies)	⊕○○○ Very low ^{a,b,c,d,e,f,g,h}	-	The impact of hospice and palliative care service utilisation on 30-day all-cause hospital readmissions for patients with heart failure was studied. Patients with HF who received hospice and palliative services were compared to those who did not. Patients were matched using propensity scoring for their use of hospice services. The propensity score was calculated by regressing use of hospice and palliative care on age, 30-day survival after index admission, race, gender, and 15 comorbidities from the Deyo's modification of Charlson comorbidity index. The odds ratio for readmission was 1.29. Hospice and palliative care patients were 1.29 times more likely to be rehospitalized, 95% CI: 1.13 - 1.48, p<0.001. (Kheirbek et al 2019). Data pertaining to the treatment of pneumonia and heart failure in 2196 US hospitals was collected. Higher rates of hospice utilization were consistently associated with lower 30-day pneumonia readmission rates for hospitals in the 25th (p = 0.02), 50th (p = 0.002) and 75th percentiles (p = 0.04), after controlling for covariates, including quality of care metrics	

				Higher rates of hospice utilization were associated with lower 30-day readmission rates for heart failure in the 50th percentile (median) only (p = 0.01). (Lah et al 2018).
Health care utilization/cost assessed with: Hospital admission rates	5073 (1 observational study)	⊕○○○ Very low ^{a,b,c,d,e,f,g,h}	-	<p>5073 patients with advanced heart failure with at least 2 HF hospitalisations who subsequently enrolled in hospice were studied to compare their acute medical service utilization before and after enrollment. Panel-negative binomial models were used to account for differences in length of exposure and possible correlation between the before and after measurements for the same patients, when calculating acute medical service utilization 6 months before and after hospice enrollment. Unadjusted and adjusted marginal means of the number of hospital admissions, ICU stays, and ER visits were calculated, and the differences in means before and after enrollment were compared.</p> <p>After hospice enrollment, there was significant reduction in hospital admissions (2.56 versus 0.53; p<0.001), ICU admissions (0.87 versus 0.19; p<0.001) and ER visits (1.17 versus 0.76; p<0.001). (Yim et al 2017).</p>
Collaboration with neurologist - not measured	-	-	-	-

- a. No adjustment for multiple comparisons.
- b. Potential imbalances in unmeasured covariates.
- c. Unable to identify hospice vs palliative care treatment during admission.
- d. Participants treated under Veteran Health system (largely male).
- e. Unable to validate diagnosis as documented in medical record.
- f. Results not generalisable to other cohorts.
- g. Participants with a diagnosis of heart failure (not FRDA).
- h. Participants had a diagnosis of pneumonia (not FRDA).

Undesirable Effects

How substantial are the undesirable anticipated effects?


JUDGEMENT

- Large
- Moderate
- Small
- Trivial
- Varies
- Don't know

RESEARCH EVIDENCE

Outcomes	No of participants (studies) Follow-up	Certainty of the evidence (GRADE)	Relative effect (95% CI)	Anticipated absolute effects* (95% CI)	
				Risk with usual care	Risk difference with hospice support
Patient quality of life - not measured	-	-	-	-	-
Caregiver quality of life - not measured	-	-	-	-	-
Patient and/or caregiver satisfaction - not measured	-	-	-	-	-
Health care utilization/ cost assessed with: Readmission rate	0 (2 observational studies)	⊕○○○ Very low ^{a,b,c,d,e,f,g,h}	-	The impact of hospice and palliative care service utilisation on 30-day all-cause hospital readmissions for patients with heart failure was studied. Patients with HF who received hospice and palliative services were compared to those who did not. Patients were matched using propensity scoring for their use of hospice services. The propensity score was calculated by regressing use of hospice and palliative care on age, 30-day survival after index admission, race, gender, and 15 comorbidities from the Deyo's modification of Charlson comorbidity index. The odds ratio for readmission was 1.29. Hospice and palliative care	

ADDITIONAL CONSIDERATIONS

				<p>patients were 1.29 times more likely to be rehospitalized, 95% CI: 1.13 - 1.48, $p < 0.001$. (Kheirbek et al 2019). Data pertaining to the treatment of pneumonia and heart failure in 2196 US hospitals was collected. Higher rates of hospice utilization were consistently associated with lower 30-day pneumonia readmission rates for hospitals in the 25th ($p = 0.02$), 50th ($p = 0.002$) and 75th percentiles ($p = 0.04$), after controlling for covariates, including quality of care metrics. Higher rates of hospice utilization were associated with lower 30-day readmission rates for heart failure in the 50th percentile (median) only ($p = 0.01$). (Lah et al 2018).</p>
Health care utilization/cost assessed with: Hospital admission rates	5073 (1 observational study)	 <p>Very low^{a,b,c,d,e,f,g,h}</p>	-	<p>5073 patients with advanced heart failure with at least 2 HF hospitalisations who subsequently enrolled in hospice were studied to compare their acute medical service utilization before and after enrollment. Panel-negative binomial models were used to account for differences in length of exposure and possible correlation between the before and after measurements for the same patients, when calculating acute medical service utilization 6 months before and after hospice enrollment. Unadjusted and adjusted marginal means of the number of hospital admissions, ICU stays, and ER visits were calculated, and the differences in means before and after enrollment were compared.</p> <p>After hospice enrollment, there was significant reduction in hospital admissions (2.56 versus 0.53; $p < 0.001$), ICU admissions (0.87 versus 0.19; $p < 0.001$) and ER visits (1.17 versus 0.76; $p < 0.001$). (Yim et al 2017).</p>

	<table border="1" data-bbox="520 107 1417 235"> <tr> <td>Collaboration with neurologist - not measured</td> <td>-</td> <td>-</td> <td>-</td> <td>-</td> <td>-</td> </tr> </table> <p data-bbox="562 277 1388 480"> a. No adjustment for multiple comparisons. b. Potential imbalances in unmeasured covariates. c. Unable to identify hospice vs palliative care treatment during admission. d. Participants treated under Veteran Health system (largely male). e. Unable to validate diagnosis as documented in medical record. f. Results not generalisable to other cohorts. g. Participants with a diagnosis of heart failure (not FRDA). h. Participants had a diagnosis of pneumonia (not FRDA). </p>	Collaboration with neurologist - not measured	-	-	-	-	-	
Collaboration with neurologist - not measured	-	-	-	-	-			

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>There is very low certainty of evidence as per the evidence profile table.</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" data-bbox="520 1308 1417 1463"> <thead> <tr> <th>Outcomes</th> <th>Importance</th> <th>Certainty of the evidence (GRADE)</th> </tr> </thead> <tbody> <tr> <td>Patient quality of life - not measured</td> <td>CRITICAL^a</td> <td>-</td> </tr> </tbody> </table>	Outcomes	Importance	Certainty of the evidence (GRADE)	Patient quality of life - not measured	CRITICAL ^a	-	
Outcomes	Importance	Certainty of the evidence (GRADE)						
Patient quality of life - not measured	CRITICAL ^a	-						

Caregiver quality of life - not measured	CRITICAL ^a	-
Patient and/or caregiver satisfaction - not measured	CRITICAL ^a	-
Health care utilization/ cost assessed with: Readmission rate	IMPORTANT ^b	⊕○○○ Very low ^{c,d,e,f,g,h,i,j}
Health care utilization/cost assessed with: Hospital admission rates	IMPORTANT ^b	⊕○○○ Very low ^{c,d,e,f,g,h,i,j}
Collaboration with neurologist - not measured	IMPORTANT ^b	-

- a. Identified as critical by expert authors on this topic.
- b. Identified as important by expert authors on this topic.
- c. No adjustment for multiple comparisons.
- d. Potential imbalances in unmeasured covariates.
- e. Unable to identify hospice vs palliative care treatment during admission.
- f. Participants treated under Veteran Health system (largely male).
- g. Unable to validate diagnosis as documented in medical record.
- h. Results not generalisable to other cohorts.
- i. Participants with a diagnosis of heart failure (not FRDA).
- j. Participants had a diagnosis of pneumonia (not FRDA).

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"> ○ 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 		

Resources required

How large are the resource requirements (costs)?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ Large costs ○ Moderate costs ○ Negligible costs and savings ○ Moderate savings ○ Large savings ○ Varies ○ Don't know 		

Certainty of evidence of required resources

What is the certainty of the evidence of resource requirements (costs)?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ Very low ○ Low ○ Moderate ○ High ○ No included studies 		

Cost effectiveness

Does the cost-effectiveness of the intervention favor the intervention or the comparison?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> ○ Favors the comparison ○ Probably favors the comparison ○ Does not favor either the intervention or the comparison ○ Probably favors the intervention ○ Favors the intervention ○ Varies ○ No included studies 		

Equity

What would be the impact on health equity?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
<ul style="list-style-type: none"> <input type="radio"/> Reduced <input type="radio"/> Probably reduced <input type="radio"/> Probably no impact <input type="radio"/> Probably increased <input type="radio"/> Increased <input type="radio"/> Varies <input type="radio"/> Don't know 		

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 <input type="radio"/> Varies <input type="radio"/> Don't know 	No published evidence.	

Feasibility

Is the intervention feasible to implement?

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 type="radio"/> Varies <input type="radio"/> Don't know 		

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

	JUDGEMENT						
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
RESOURCES REQUIRED	Large costs	Moderate costs	Negligible costs and savings	Moderate savings	Large savings	Varies	Don't know
CERTAINTY OF EVIDENCE OF REQUIRED RESOURCES	Very low	Low	Moderate	High			No included studies
COST EFFECTIVENESS	Favors the comparison	Probably favors the comparison	Does not favor either the intervention or the comparison	Probably favors the intervention	Favors the intervention	Varies	No included studies
EQUITY	Reduced	Probably reduced	Probably no impact	Probably increased	Increased	Varies	Don't know
ACCEPTABILITY	No	Probably no	Probably yes	Yes		Varies	Don't know
FEASIBILITY	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 checked="" type="radio"/>	Strong recommendation for the intervention <input type="radio"/>
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CONCLUSIONS

Recommendation

We suggest that people with Friedreich ataxia with advanced heart failure would benefit from hospice support when their goals align with a comfort-focused approach and the individual's prognosis meets eligibility criteria – usually a life expectancy of 6 months or less if the disease runs its natural course.

Justification

While there is little evidence directly pertaining to this question, studies indicate that the readmission rate to the hospital is lower in patients with heart failure enrolled in hospice, and for those valuing low burden treatments in a familiar environment this is an important outcome. In addition, our clinical experience is that hospice care can greatly improve quality of life by providing expert symptom management and providing care in the patient's home environment.

Subgroup considerations

This recommendation is for individuals with Friedreich ataxia with advanced heart failure. Hospice enrolment criteria vary by country; however, it is worth noting that certain populations may be eligible to continue life prolonging treatments and re-hospitalization while still receiving hospice benefits. In the United States these populations include military veterans and children.

Implementation considerations

Monitoring and evaluation

Research priorities

Studies on quality of life for people with FRDA who have heart failure, including impact on feelings of dignity and preferences for place of death.

Studies of degree of symptom management in FRDA.

