QUESTION

Should hospice	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
 No Probably no Probably yes Yes Varies Don't know Desirable Effects How substantial are the desirable anticipated eriticipated eriticipate	ffects?	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.
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
 o Trivial o Small Moderate o Large o Varies o Don't know 	Outcomes Nº of Certainty of Relative Anticipated absolute effects* (95% CI)	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.

	participants (studies) Follow-up	the evidence (GRADE)	effect (95% Cl)	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 Jow ^{a,b,c,d,e,f,g,h}		care service ut cause hospital patients with I Patients with I and palliative to those who of matched using their use of hot propensity sco regressing use care on age, 3 admission, rac comorbidities modification of index. The odo was 1.29. Hosp patients were be rehospitaliz p<0.001. (Khei pertaining to t pneumonia an US hospitals w rates of hospit consistently as day pneumoni hospitals in th = 0.002) and 7 0.04), after co	hospice and palliative tilisation on 30-day all- readmissions for heart failure was studied. HF who received hospice services were compared did not. Patients were g propensity scoring for ospice services. The ore was calculated by of hospice and palliative 0-day survival after index se, gender, and 15 from the Deyo's of Charlson comorbidity ds ratio for readmission pice and palliative care 1.29 times more likely to zed, 95% CI: 1.13 - 1.48, irbek et al 2019). Data the treatment of ad heart failure in 2196 vas collected. Higher ce utilization were ssociated with lower 30- ia readmission rates for e 25th (p = 0.02), 50th (p '5th percentiles (p = ntrolling for covariates, ity of care metrics

				associated wit readmission ra	f hospice utilization were th lower 30-day ates for heart failure in entile (median) only (p = al 2018).
Health care utilization/cost assessed with: Hospital admission rates	5073 (1 observational study)	⊕⊖⊖⊖ Very Iow ^{a,b,c,d,e,f,g,h}	-	failure with at hospitalisation enrolled in ho- compare their utilization befa Panel-negativa used to accoun length of expor- correlation be after measure patients, when medical servic before and aft Unadjusted an means of the r admissions, IC were calculate means before were compare After hospice significant red admissions (2. ICU admission p<0.001) and	hs who subsequently spice were studied to a cute medical service ore and after enrollment. e binomial models were nt for differences in osure and possible tween the before and ments for the same in calculating acute the utilization 6 months the hospice enrollment. ad adjusted marginal number of hospital CU stays, and ER visits ed, and the differences in and after enrollment
Collaboration with neurologist - not measured	-	-	-	-	-
b. Potent c. Unable d. Partici e. Unable f. Result g. Partici	ustment for r ial imbalance to identify h pants treated to validate o s not general pants with a o pants had a o	s in unmeasu ospice vs pal under Vetera liagnosis as c isable to othe diagnosis of h	red cova liative ca an Health locument er cohorts neart failu	re treatment system (larg ed in medica s. ure (not FRD/	al record. (A).

Undesirable Effects How substantial are the undesirable a	nticipated effects?						
JUDGEMENT	RESEARCH EVIDENC	E					ADDITIONAL CONSIDERATIONS
o Large o Moderate							
• Small o Trivial o Varies o Don't know		№ of participants	Certainty of the evidence	Relative effect	Anticipated a	bsolute effects [*] (95% CI)	
		(studies) Follow-up	(GRADE)	(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	-	-	-	-	-	
	utilization/ cost assessed with:	0 (2 observational studies)	⊕⊖⊖⊖ Very low ^{a,b,c,d,e,f,g,h}	-	care service u cause hospita patients with Patients with and palliative to those who matched usin their use of h propensity sc regressing use care on age, 3 admission, ra comorbidities modification index. The od	hospice and palliative tilisation on 30-day all- l readmissions for heart failure was studied. HF who received hospice services were compared did not. Patients were g propensity scoring for ospice services. The ore was calculated by e of hospice and palliative 80-day survival after index ce, gender, and 15 from the Deyo's of Charlson comorbidity ds ratio for readmission price and palliative care	

		1	1	
				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}	-	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. 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).

	Collaboration with neurologist - not measured - - a. No adjustment for mu b. - b. Potential imbalances in c. Unable to identify hose d. c. Unable to identify hose d. Participants treated u e. g. Participants treated u participants with a dia h. Participants with a dia	in unmeasured cova spice vs palliative ca nder Veteran Healtl ignosis as documen able to other cohort agnosis of heart fail			
Certainty of evidence What is the overall certainty of the evidence of	effects?				
JUDGEMENT	RESEARCH EVIDENCE				ADDITIONAL CONSIDERATIONS
 Very low Low Moderate High No included studies 	There is very low certainty of evidence	e as per the evidence pro			
Values Is there important uncertainty about or variabili	ty in how much people value the main o	outcomes?			
JUDGEMENT	RESEARCH EVIDENCE			ADDITIONAL CONSIDERATIONS	
 Important uncertainty or variability Possibly important uncertainty or variability Probably no important uncertainty or variability No important uncertainty or variability 	Outcomes				
	Patient quality of life - not	measured	CRITICAL ^a	-	

Caregiver quality of life - not measured	CRITICALª	-
Patient and/or caregiver satisfaction - not measured	CRITICALª	-
Health care utilization/ cost assessed with: Readmission rate	IMPORTANT ^b	$ \bigoplus \bigcirc \bigcirc \bigcirc \\ 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 b. Identified as important by expert authors c. No adjustment for multiple comparison d. Potential imbalances in unmeasured of e. Unable to identify hospice vs palliative f. Participants treated under Veteran Heig g. Unable to validate diagnosis as docum h. Results not generalisable to other consist i. Participants with a diagnosis of heart j. Participants had a diagnosis of pneum 	hors on this topi ons. covariates. e care treatmen ealth system (lar nented in medica norts. failure (not FRD	t during admission. gely male). al record. A).

Balance of effects

ioes the balance between desirable and undesirable effects favor the intervention or the comparison?						
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS				
 o Favors the comparison o Probably favors the comparison o Does not favor either the intervention or the comparison o Probably favors the intervention o Favors the intervention o Varies o Don't know 						
Resources required						

How large are the resource requirements (costs)?

JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
 O Large costs O Moderate costs O Negligible costs and savings O Moderate savings O Large savings O Varies O Don't know 		
Certainty of evidence of requ What is the certainty of the evidence of resource	ired resources e requirements (costs)?	
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
o Very low o Low o Moderate o High o No included studies		
Cost effectiveness Does the cost-effectiveness of the intervention f	avor the intervention or the comparison?	
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
 o Favors the comparison o Probably favors the comparison o Does not favor either the intervention or the comparison o Probably favors the intervention o Favors the intervention o Varies o No included studies 		

Equity What would be the impact on health equity?		
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
 Reduced Probably reduced Probably no impact Probably increased Increased Varies Don't know 		
Acceptability Is the intervention acceptable to key stakeholde	rs?	
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
o No o Probably no • Probably yes o Yes o Varies o Don't know	No published evidence.	
Feasibility Is the intervention feasible to implement?		
JUDGEMENT	RESEARCH EVIDENCE	ADDITIONAL CONSIDERATIONS
o No o Probably no o Probably yes o Yes o Varies o 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	Conditional recommendation against the intervention	Conditional recommendation for either the intervention or the comparison	Conditional recommendation for the intervention	Strong recommendation for the intervention
0	0	0	•	0

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.