Author/s:
Guo, H;Mann, J;Goh, N;Smallwood, N

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Investigation burden for patients with fibrotic interstitial lung disease at the end of life

Authors & Affiliations

Dr Hui Guo MBBS BMedSci

Department of Respiratory and Sleep Medicine, The Royal Melbourne Hospital, Parkville, Victoria 3050, Australia.

Email: Hguo@phcn.vic.gov.au

Dr Jennifer Mann BPharm MBBS FRACP

1. Department of Respiratory and Sleep Medicine, Austin Health, Heidelberg, Victoria 3084, Australia.

2. Institute for Breathing and Sleep, Heidelberg, Victoria 3084, Australia.

Email: J.mann@rbht.nhs.uk

Assoc Prof Nicole Goh MBBS FRACP PhD

1. Department of Respiratory and Sleep Medicine, Austin Health, Heidelberg, Victoria 3084, Australia.

2. Institute for Breathing and Sleep, Heidelberg, Victoria 3084, Australia.

Email: Nicole.goh@austin.org.au

Assoc Prof Natasha Smallwood BMedSci MBBS SpecCertPallCare MSc(Epid) MRCP FRACP PhD

1. Department of Respiratory and Sleep Medicine, The Royal Melbourne Hospital, Parkville, Victoria 3050, Australia.

2. Department of Medicine (Royal Melbourne Hospital), University of Melbourne, Victoria 3050, Australia.

Email: Natasha.smallwood@mh.org.au

Corresponding Author: A/Prof Natasha Smallwood

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Abbreviations

f-ILD=Fibrotic Interstitial Lung Disease (f-ILD)

AH = Austin Health

ILD = interstitial lung disease

IPF = Idiopathic pulmonary fibrosis

RMH = Royal Melbourne Hospital.

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Email: Natasha.smallwood@mh.org.au

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Interstitial Lung Disease (ILD) are characterised by chronic inflammation and/or fibrosis and are often progressive. Whilst varied aetiologies and clinical phenotypes exist, most ILD is idiopathic and incurable[1]. Notably, fibrotic subtypes carry a worse prognosis, with idiopathic pulmonary fibrosis

(IPF) being progressive and commonly fatal[2]. People with IPF also experience a significant symptom burden with dyspnoea and cough[3].

Despite the terminal nature of the disease, people with fibrotic ILD (f-ILD) are often unaware of their prognosis and unprepared for end of life care decisions[4]. While the American Thoracic Society and British Thoracic Society ILD guidelines recommend early palliative care involvement[1, 4, 5], some studies from overseas demonstrate that few people with f-ILD access such care and those who do, are referred only late at the very end of life.[4, 6, 7]. Furthermore, the majority of people with f-ILD die in acute hospitals, not at home or in hospices[7].

As little is known regarding end-of-life care for people with f-ILD in Australia or whether they continue to undergo diagnostic tests in the last days of life, this study aimed to examine the management approach and diagnostic test burden for f-ILD patients during their terminal admission at two Australian hospitals. In this manuscript we report the findings regarding investigation burden.

A retrospective cohort study was undertaken at the Royal Melbourne and Austin Hospitals in Victoria, both being tertiary referral centres with specialist ILD services. Data were retrospectively extracted from electronic and paper records for all patients who died from f-ILD (including disease progression or exacerbation, respiratory infection or cor pulmonale) between 1st January 2012 and 31st December 2016. Patients who died from a co-existing malignancy or an unrelated condition were excluded. ILD types were classified using the combined American Thoracic Society and European Respiratory Society criteria[8].

Data collected included: patient demographics, disease severity as determined by lung function tests and number of admissions, specialist palliative care referral and advanced care planning. During the terminal admission data were collected regarding: admitting team, duration of admission, frequency and timing of investigations and timing of palliation.

Data are expressed as frequencies and percentages with summary descriptive statistics (means with standard deviation for normally distributed data or medians with interquartile ranges for non-parametric data). Continuous non-parametric data were compared using the Mann-Whitney test and categorical data were compared using the Chi-Square test, with $p<0.05$ taken to indicate statistical significance. Ethics approval was granted by the Melbourne Health Research Office (Approval number: QA2017091) and the Austin Health Human Research Ethics Committee (Approval number: LNR/17/Austin/300).

Of one hundred and sixteen patient deaths identified from each hospital's mortality database, 67 met eligibility criteria for inclusion, with 33 deaths at the Royal Melbourne Hospital (RMH) and 34 at Austin Hospital (AH). Patients' demographic and disease severity characteristics were similar at both sites, with a median age of seventy-eight years and slight male predominance (Table 1). Most patients had a diagnosis of IPF, with severe lung function impairment identified on tests undertaken within two years of the terminal admission. The minority of patients accessed outpatient palliative care services (36%) or made an advanced care plan prior to the terminal admission (16%).

Most patients (60, 89.6%) underwent at least one investigation during their terminal admission, with a median of 4 (IQR=1-12) investigations per patient (Table 2). The median time to death was 4 days

(IQR=2-10). Final arterial blood gas tests were performed a median of 2 days (IQR=0.8-6) before death and venepuncture blood tests continued until a median of 1 day before death (IQR=0-2). Significantly more investigations were performed at the end of life at RMH than at AH.

For sixty-three (94%) patients the goal of care was changed (and documented) to aim for comfort care (palliation) only at a median of 1 day prior to death. Twenty-four (36%) patients underwent their last diagnostic investigation on the day the goal of care was changed, and 12 (18%) underwent further investigations (including ongoing venepuncture and for one patient arterial blood gas sampling) after the goal of care was changed to comfort.

Patients who had received outpatient specialist palliative care ($p<0.001$), used domiciliary oxygen $p<0.001$), and those with a documented advance care plan ($p=0.004$) prior to their terminal admission underwent significantly less investigations than those who had not. Admission to either intensive care ($p=0.001$) or a high-level care/respiratory support unit ($p<0.001$) as compared with a medical ward was associated with significantly increased investigations. Additionally, admission under a general medical team compared with a palliative care team ($p<0.001$) was associated with increased investigations at the end of life. Other patient factors such as age, gender and lung function impairment were not significantly associated with investigation burden at the end of life.

Discussion

This study suggests that few patients with f-ILD access outpatient palliative care services prior to the terminal admission. Recognition that patients were actively dying and adjustment of goals of care to aim for comfort only occurred late, only one day prior to death. Additionally, diagnostic

investigations were performed until the very end-of-life, even sometimes after the decision to aim for comfort care only. Yet there is no clear rationale for continuing diagnostic investigations once this decision has been made. Notably, similar findings have also been demonstrated in patients dying from chronic obstructive pulmonary disease and cardiac failure[9-11]. Factors that may contribute to excessive investigations at the end of life include physicians' uncertainty recognising when patients are actively dying, lack of consistent communication and poor advance care planning.

The patients in this study had survived, on average, one ILD-related hospitalisation in the two years preceding their terminal admission, highlighting that it can be challenging to determine which exacerbation will be fatal. Thus when investigations are being requested, the clinical team does not know if the patient will survive or not, and investigations are clearly important in identifying reversible factors contributing to an exacerbation. Diagnostic tests can also help clinicians determine that the chance of survival is low and that a palliative approach is required. Therefore, investigations have a role, however, once a person is recognised to be actively dying, investigations are burdensome and unnecessary. Postulated explanations for requesting ongoing investigations at the end of life include clinicians' lack experience, confidence and training in recognising active dying[12]. Furthermore chance of survival is often overestimated[13], especially within the hospital culture where there is a focus on life-sustaining treatments, and this equally may contribute to excessive investigations at the end of life. Additionally, some patients and/or their families may wish for "everything to be done" right until the end, even if that includes burdensome tests and treatments.

Notably, even after recognising that a person was dying and thus the goal of care was changed to aim for comfort, some patients (18%) in this study continued to undergo tests. A similar study of IPF

patients dying in hospital in Finland found that 42% of patients underwent ongoing investigations until death[7]. This practice should be reconsidered as it is burdensome for patients (who often experience physical discomfort during tests) and family (who may be separated from their loved one for tests to occur). One explanation for indiscriminate ordering of investigations at the end of life is poor communication within the treating team. Thus junior staff and sometimes also patients' caregivers may not fully appreciate that death is expected and that investigations are no longer required. Notably reduced investigation burden was significantly associated with previous outpatient specialist palliative care referral and documented advance care planning, thus highlighting the importance of discussing and planning for end of life care in all illnesses, but particularly in f-ILD which can be rapidly fatal.

To address these challenges in f-ILD, it is essential that respiratory and general physicians have greater knowledge, experience and competence in providing a palliative approach themselves to these patients. Notably, international guidelines recommend that discussion and referral to palliative care services should occur early, at time of diagnosis, to ensure patients can access these services in a timely manner and have opportunities to discuss palliative care and advanced care planning[5]. As demonstrated in this study, these early interventions have clear implications for subsequent inpatient care. Within the hospital setting, clear communication is vital especially once recognising that a person is actively dying. The cessation of investigations does not mean the cessation of care, and this should be communicated consistently within the treating team and with the family.

The main limitation of this study is its retrospective design and reliance on clinical documentation. There was no documentation of the exact time of day a decision was made to change the goal of care to comfort. However, documentation of such discussions was high, suggesting overall good quality medical record keeping. This study included two tertiary Australian health services with large specialist ILD services, where expert inpatient end-of-life care is expected for f-ILD. Therefore these results may overestimate the quality of end-of-life care at other Australian hospitals, thus further highlighting the urgent need to respond to f-ILD patients' unmet palliative care needs.

Ultimately, the model of care for f-ILD patients should integrate disease-directed and supportive management strategies concurrently, with palliative care introduced early, at time of symptom onset[5]. The challenges lie in planning for and recognising deterioration and ultimately overestimating survival, leading to increased investigation burden and potential suffering for both patients and their families. This study highlights several potential areas for improvement, including early anticipation of patients' palliative care needs, actively discussing future care wishes at the time of diagnosis, and clear communication within health professional teams as well as with patients' relatives and carers.

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Table 1. Patients' characteristics before the terminal admission

Characteristic	All patients (n=67)	AH (n=34)	RMH (n=33)	Comparison of sites
Male gender	37 (55%)	19 (56%)	18 (55%)	p=0.912
Age*	78 (74-78)	79 (75-84)	78 (73-84)	p=0.353
English as first language	50 (75%)	25 (75%)	25 (76%)	p=0.834
Type of ILD:				
IPF	44 (66%)	25 (74%)	19 (58%)	p=0.169
NSIP	6 (9%)	4 (12%)	2 (6%)	p=0.414
COP	1 (1%)	0	1 (3%)	p=0.306
Sarcoid	2 (3%)	1 (3%)	1 (3%)	p=0.983
LIP	1 (1%)	0	1 (3%)	p=0.306
Hypersensitivity pneumonitis	1 (1%)	1 (3%)	0	p=0.321
Not specified	12 (18%)	3 (9%)	9 (27%)	p=0.049
Number of co-morbidities*	3 (3-7)	4 (2-5)	5 (3-7)	p=0.021
COPD	6 (9%)	3 (9%)	3 (9%)	p=0.969
Congestive heart failure	10 (15%)	4 (12%)	6 (18%)	p=0.461
Pulmonary hypertension	1 (1%)	1 (3%)	0	p=0.321
Ex-smoker	33 (49%)	18 (53%)	15 (45%)	p=0.540
Never smoked	26 (39%)	15 (44%)	11 (33%)	p=0.365
Smoking status not documented	8 (12%)	1 (3%)	7 (21%)	p=0.021
BMI*	27 (23-31)	27 (25-31)	27 (21-30)	p=0.433
Domiciliary oxygen use	44 (66%)	23 (68%)	21 (64%)	p=0.730
FEV_1*	69 (58-77)	72 (60-79)	67 (57-75)	p=0.488
FVC*	64 (47-74)	60 (46-68)	66 (48-80)	p=0.227
DLco*	36 (31-44)	35 (31-40)	38 (31-46)	p=0.321
Number of hospital admissions for ILD within past 2 years*	1 (0-2)	0.5 (0-2)	1 (0-2)	p=0.330
Outpatient specialist palliative care review	24 (36%)	13 (38%)	11 (33%)	p=0.676
Advanced Care Plan written	11 (16%)	7 (21%)	4 (12%)	p=0.350
Admission team:				
General medicine	31 (46%)	16 (47%)	15 (45%)	p=0.895
Respiratory medicine	22 (33%)	10 (29%)	12 (36%)	p=0.545
Palliative care	10 (15%)	5 (15%)	5 (15%)	p=0.959
Other	4 (6%)	3 (9%)	1 (3%)	p=0.317
Location at time of death:				
Emergency department	0	0	0	
Medical ward	38 (57%)	19 (56%)	19 (58%)	p=0.889
Palliative care ward	15 (22%)	8 (24%)	7 (21%)	p=0.820
RCU	8 (12%)	1 (3%)	7 (21%)	p=0.021
ICU	5 (7%)	5 (15%)	0	p=0.022
Other	1 (1%)	1 (3%)	0	p=0.321

Data are presented as counts with percentages in parenthesis unless otherwise stated. * = median with IQR in parenthesis. AH = Austin Health, BMI = Body mass index, COP = Cryptogenic organizing pneumonia, COPD = chronic obstructive pulmonary disease, DLco = diffusing capacity for carbon monoxide, FEV1 = forced expiratory volume in the first second, FVC = forced vital capacity, ICU = intensive care unit, ILD = interstitial lung disease, IPF = Idiopathic pulmonary fibrosis LIP = Lymphocytic interstitial pneumonitis, IQR = interquartile range, NSIP = Non-specific interstitial pneumonia, RCU = respiratory care unit, RMH = Royal Melbourne Hospital.

Table 2. Investigation burden in the terminal admission

Investigation type	Number of investigations per person			
	All patients (n=67)	AH (n=34)	RMH (n=33)	Comparison of sites
All investigations	4 (1-12)	2 (1-7)	7 (2-14)	p=0.049
Venepuncture	2 (1-7)	2 (1-4)	5 (1-11)	p=0.063
Arterial blood gas	1 (0-2)	0 (0-1)	1 (0-2)	p=0.041
CT	0 (0-0)	0 (0-1)	0 (0-0)	p=0.722

All data are presented as median with IQR in parenthesis. AH = Austin Health, CT = computerised tomography, IQR = interquartile range, RMH = Royal Melbourne Hospital.

Abstract

Fibrotic Interstitial Lung Disease (f-ILD) has a guarded prognosis and the goal of therapy in advanced disease should be symptom based. Despite this, patients may still undergo burdensome investigation at the end of life. A retrospective audit was performed for sixty-seven patients who died from f-ILD at the Royal Melbourne and Austin Hospitals from 2012 to 2016. Increased investigation burden was associated with lack of both outpatient palliative care referral and documented advance care plan, and admission to a high dependency unit. Eighteen percent of patients underwent ongoing investigations after the institution of comfort care. These findings highlight the unmet end-of-life care needs of people with f-ILD.