Idiopathic Pulmonary Fibrosis (IPF)
Palliative care needs

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- Other: Employee of AHS & University of Alberta
Agenda

• What is IPF and ILD?
• What are the palliative care needs in IPF?
• When should ILD/IPF palliative care start?
• How to deliver components of palliative care?
  • Describe our Multidisciplinary Collaborative Care innovation
Fibrotic Lung Disease: Sorting Out the Alphabet Soup

Interstitial lung disease

CTD-ILD

NSIP

Idiopathic interstitial pneumonias

IPF

Chronic HP

Unclassifiable

Early interstitial lung abnormalities

Where does IPF fit in the context of the ILDs?

• IPF is the most common of the idiopathic interstitial pneumonias (IIPs)

• Definition of IPF:
  • Chronic, progressive fibrosing interstitial pneumonia of unknown cause
  • Older adults
  • Limited to the lungs
  • Histopathologic and/or radiologic pattern of UIP (usual interstitial pneumonia)

Key Clinical Features of IPF

- Increasing breathlessness on exertion
- Non-productive cough
- Patient age > 50 years
- Bibasilar inspiratory crackles
- Digital clubbing in many cases
- No cure
- High morbidity & mortality


IPF Has an Unpredictable Disease Course

Variable onset of symptoms

Unpredictable patterns of progression

Disease Progression

Demise

Time
Approved Antifibrotic Therapies for Patients with IPF

**Pirfenidone**
- FDA approval 2014
- Anti-fibrotic properties; exact mechanism of action unknown
- Oral, 801 mg, three times daily
- Nausea, rash/sun sensitivity, dyspepsia/GERD

**Nintedanib**
- FDA approval 2014
- Tyrosine kinase inhibitor; targets FGFR, PDGFR, VEGFR, FLT3
- Oral, 150 mg, two times daily
- Diarrhea, nausea

Slows down progression, improves survival  
Does not stop progression or reverse disease  
No impact on symptoms  
No impact on QoL (? Possible worsening due to adverse effects)

Pirfenidone. [https://www.accessdata.fda.gov/drugsatfda_docs/label/2017/022535s005lbl.pdf](https://www.accessdata.fda.gov/drugsatfda_docs/label/2017/022535s005lbl.pdf)  
Nintedanib. [https://www.accessdata.fda.gov/drugsatfda_docs/label/2017/205832s004lbl.pdf](https://www.accessdata.fda.gov/drugsatfda_docs/label/2017/205832s004lbl.pdf)  
ATS Guidelines Recommend Palliative Care For Patients with IPF...

• Palliative care should be considered as an adjunct to disease-focused care

• Advanced directives and end-of-life care issues should be addressed in the ambulatory setting in all patients with IPF, particularly those with severe physiological impairment and comorbid conditions

• In patients who are bedbound due to IPF, hospice care should be considered

Evidence Indicates a Need to Do Better

**Rajala et al. 2018**
N = 247 patients with IPF included in study; 92 died and were included in follow-up
Marked deterioration in the QOL during the **LAST TWO YEARS OF LIFE**
Most prominent declines in HRQOL occurred in physical function, vitality, emotional role, and social functioning

**Bajwah et al. 2012**
N = 45 patients with progressive idiopathic fibrotic ILD
38% had palliative care team involvement
93% experienced dyspnea in last year of life
<50% on opiates
Only 18% had preferred place of care, and 13% had preferred place of death documented

**Lindell et al. 2015**
N = 404 decedents (patients with IPF)
57% died in the hospital
14% had formal palliative care referral
71% referred within last month of life

Palliative care is needed!

What is the appropriate time to start Palliative Care?

**START AS EARLY AS POSSIBLE**

- **Need for palliative care**
- **QOL compromised** → **further reduced** → **poor with persistent decline**

**Lung injury** → **Death**

**Asymptomatic**
- 0-10 years

**Symptomatic**
- 0-5 years

**IPF Diagnosis**

**Progression**
that breathlessness was the overwhelming symptom. Many participants stated how their breathlessness had taken on a life of its own and was consuming them.

I can’t go anywhere […] … I don’t don’t [really] have a life I’m sitting indoors to be meet friends and have coffee and in the middle of life back … (Mary, in her 70s with advanced IPF)

The patients get used to the breathlessness, their doctors and nurses get used to the breathlessness, and (...) the penny drops that maybe they need to have um (1) to use drugs for the symptomatic relief of breathlessness. (GP).

One other thing I am interested in is when you reach the end stage and you’re struggling to breathe and all these NO SYMPTOM MANAGEMENT

NO ADVANCE CARE PLANNING (ACP)

she’ll panic because although she tries not to but em she would panic because it’s not nice not being able to breathe you know … (Anthony, husband to Betty)

Um I mean I have never seen quite so much phlegm (laughs nervously) and he was literally choking on it, he was deeply blue and and the hopelessness that nobody could actually do anything about it and I thought there probably were few few worse ways to die than that when I saw it in that instant, sort of haunts me a bit today. (Palliative Care Consultant)
Unmet Palliative care needs in IPF

- Progressive symptoms
- Hopelessness
- Helplessness
- Frustrations
- Poor coping
- High caregiver burden
- End of life needs & bereavement support
- Depression & anxiety
- Need for timely, titrated information & education
Referral to PC service vs integrated approach in routine care

- How to assess needs and when to start symptom therapies?
- What is the best approach to dyspnea management in IPF?
- What is the best model of ACP discussion to improve communication?
- How to coordinate care, engage and support the family better?

But how can 1 clinician meet all of these needs?!
Multidisciplinary Collaborative Care Model (MDC)

Developed and implemented in 2012
(2017 CFHI recognition)

**What?**
- Needs assessment, integrated symptom therapies, ACP, community support

**Who?**
- Clinic → MD, ILD RN and allied health team
- Community → Homecare team (allied health, NP) and primary care

**When?**
- Start early; at all clinic visits, and ongoing in the community and at home

**How?**
- Create and train multidisciplinary clinic team
- Identify and partner with community team

Kalluri. 2019 NEJM Catalyst. IPF Care Redesign
Conceptual Framework of Multidisciplinary ILD Collaborative

- **Patient-Centric Care**
  - Multidisciplinary, ILD Collaborative Clinic (Physicians, RT, PT, RD, RN)

- **Early Integrated Palliative Approach**
  - Symptom management strategies
  - Advance care planning from first encounter
  - Education

- **Collaborative Community-Based Support**
  - Engagement of community allied health care and primary care physicians
  - Close communication
  - Early detection of changes

Meet Harry and His Wife

• 72-year-old male with IPF (after MDD, FVC 79%, DLCO 49%)
  • Tolerating antifibrotic therapy well for six months

• He presents to our clinic with gradual worsening dyspnea (MRC 4/5) with his wife in attendance
  • Recent HRCT shows progression of UIP
  • 6 MWD: 467 m, baseline SpO2 92%, nadir SpO2 79% on 2 L
<table>
<thead>
<tr>
<th>Harry’s Care Needs Assessment Tool</th>
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<td>Pre-clinic ILD RN assessment</td>
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### Patient concerns
- What is wrong with me?
- How can I improve my breathing?
- Angry, frustrated with inability to do things and go out

### Caregiver concerns
- Anxious, worried and frustrated
- What is going on with Harry?
- How can I help him?
- What can we do when his breathing gets worse?*

### Symptoms and function
- MDC approach
- Assess impact on function
  - Daily activities: struggling
  - Work: retired
  - Recreation: no longer playing golf, camping or fishing

* Crisis event: 2-3x week in the mornings, when he showers and is helping with chores
Dyspnea

- Definition per ATS: A subjective experience of breathing discomfort that consists of qualitatively distinct sensations that vary in intensity
- Dyspnea is not unidimensional
  - Need to assess severity and impact
- Framework of dyspnea (MDC approach):
  - At baseline (rest)
  - Episodic (with activity)
  - Dyspnea crisis episodes

Harry’s Dyspnea Assessment

MDC assessment approach*

- MRC is not detailed enough (ADL)
- Numerical rating scale is easy to administer, track, sensitive to changes in ADL
- 0-10 (mild/mod/severe)
- Facilitates early detection of changes and personalized treatment advice
- Use of an interdisciplinary team

Dyspnea  | Harry’s Score?
---------|------------------
Baseline (at rest)  | 
Episodic  | 
Crisis episodes  | 

Breathing is Not an Option: Dyspnea is!

MDC Treatment Approach*

- Patient-centric dyspnea assessment
- Non-pharmacological strategies
  - Allied health team: PT, OT, RT, RN
- Early O2 start and frequent titrations in clinic and home
- Low dose opiate: oral for baseline, buccal for exertion/episodic and crisis dyspnea (fast onset, self administer)

Harry’s Individualized Care Plan Based on Needs

Patient-centric dyspnea assessment: measure severity and impact; at rest, various ADL, track crisis

<table>
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<tr>
<th>Activity</th>
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<td>4</td>
<td>4</td>
<td>7</td>
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<td>7</td>
<td>7</td>
<td>1</td>
<td>8/9 showering</td>
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1 Very slight  2 Slight  3 Moderate  4 5 Severe  6 7 Very severe  8 9 Extremely severe  10 Maximal

Systematic dyspnea management using MDT approach

<table>
<thead>
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<td>How to manage dyspnea with golf, camping, fishing and ADL?</td>
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<tr>
<td>• Pacing, activity and behavior modification</td>
</tr>
<tr>
<td>• Arrange for home assessment, environment modifications</td>
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<tr>
<td>• Develop an exercise plan</td>
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<tr>
<td>• Refer to pulmonary rehabilitation program</td>
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<td>• Advise on other symptoms: cough, fatigue, pain when needed</td>
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<td>• Identify crises and provide practical advice/action plan</td>
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Systematic dyspnea management using MDT approach

RT

How to manage dyspnea with daily function and recreational activities (golfing, camping, fishing)?

- Oxygen titration to keep nadir exertional SpO2>90%
- Review flows (Cont vs pulse) 24/7 use
- Type of equipment
- Nasal care
- Oxygen and travel (air travel)
- Identify crises and provide practical advice/action plan

6 MWD: 467 m
Baseline SpO2 92%, Nadir SpO2 79% on 2 L
Harry’s Individualized Care Plan Based on Needs

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Systematic dyspnea management using MDT approach

RD (Dietitian)
- Reflux education
- Weight management
- Safe swallow practices
- Diet modifications for dyspnea as needed
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**Systematic dyspnea management using MDT approach**

**RN**
- Disease & symptom education
- Antifibrotic therapy management
- Provide clinical trials info
- Written action plans and instructions
- Caregiver education
- Facilitate connection to support group
- Care coordination

8/9 showering
# Harry’s Individualized Care Plan Based on Needs

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- Baseline
- Episodic
- Crisis

Systematic dyspnea management using MDT approach

**MD (ILD specialist/palliative)**

- Diagnosis, disease specific therapies, education
- Review and address identified needs
- Advance care planning
- Review symptom management, action plans in discussion with team
- Refer to community supports
Harry’s Individualized Care Plan

Dyspnea Prescription

• Pacing, behavior and activity modification
• Oxygen flows (rest 3L/exertion 9L/exercise 10L)
• Low dose opiates:
  • Baseline (rest): Oral 0.1 mg hydromorphone QID
  • Exertional (episodic): 0.2 mg buccal hydromorphone 10 min preactivity
  • Crisis: 0.2 mg buccal hydromorphone every ten minutes until relieved, lorazepam 0.25-0.5 mg SL; call home care RT
• Provide action plan†

Action Plan for Episodes of Crisis Dyspnea

CALL for help:
- Calmly approach
- Patient and caregivers

OBSERVE closely and assess dyspnea for ways to respond

MEDICATIONS to be tried:
- Recommendations from providers for opioid/other use

FAN face may decrease shortness of breath

OXYGEN therapy as previously found useful

REASSURE and use relaxation techniques

TIMING interventions to reduce dyspnea. Work together.
- Reassess.
- Repeat.

Your Doctor’s customized treatment plan

Advance Care Planning

• An organized ongoing process of communication to help an individual identify, reflect upon, discuss, and articulate her or his values, beliefs, goals, and priorities to guide personal care decision making including end-of-life care

• For patients with IPF, either this isn’t happening or when it does it is implemented too late

Advance Care Planning in IPF

PATIENTS WANT:

• Open and honest discussion on progression, what to expect, how to prepare and what death looks like (“elephant in the room”)
  – Well I haven’t got a very long future so ah, I suppose I’d like a bit of honesty from them... I’ve been sort of kept in the dark a little

• Provide hope by discussing how symptom treatment can improve QOL
  – One other thing I am interested in is when you reach the end stage and you’re struggling to breathe and all these things, what can be done about it to reduce my anxiety level? No one has talked to me about that
  – What I still need to find out is how to manage that cough, so that it’s not something that embarrasses me and other people when I’m in public... there might be strategies that you can use to control it

Advance Care Planning in IPF - MDC approach

COMMUNICATION & PLANNING: MDC MODEL

Patient & Caregivers
Primary Care Community Team
Specialty Clinic Team

From diagnosis
Through death and bereavement

- Disease treatment; symptom control; action plans
- Understanding disease status & functional status
- EOL planning: affairs, estate, location, personal, spiritual, etc.
- Goals, hopes, fears, preferences
- Supports: family, community, spiritual, medical

Kalluri M. Manuscript in submission
Advance Care Planning at First Visit
Harry, his wife and the team

**Goals and Wishes**
- Remain active: HUNTING, FISHING, TRAVELING, CAMPING
- Avoid hospitalization
- HOME DEATH

**Fear**
- Suffocation
- Death and dying, being a burden to family

**Self-Management**
- DISCUSSED STRATEGIES for symptoms (QOL, achieve goals)
- Provided information, options for care/location, implications

**Engage Caregiver**
- Wife willing to support home death
- Wanted to learn how to help with breathing

**Documentation**
- Goals of care (AHS), preferred place of care and death
- Encourage EOL planning: POA, advanced directives, personal affairs, bucket list
Patient and family-centered care

- 3L rest
- 9L exertion
- 10L exercise
- Education, nasal care
- Addressed O2 needs for golf, travel, camping

- Pacing, activity modification, exercise, discussed energy conservation measures for golf, camping

- Opiates and BDZ, ACP, written actions plans, network with community teams

Diagnosis
- Antifibrotic
- PPI
- Rehab
- Education
- Immunization

Assess family needs, connect to home care and patient support group
Care should not stop between clinic visits

1 Month Later- a Home Visit by RT

- Harry played golf twice in a month- using high flow O2, pacing
- Home walking oximetry on 9 lpm shows nadir Spo2= 84%, exertional oxygen flow increased to 10L

- Early detection and rapid intervention
- Support at home
- Care aligned to patient wishes
Collaboration with Community Teams

• Addressing symptom crisis
  Outside of Clinic Visits

- Maintain QOL
- Increase days spent at home: patient-centered goal
- Avoids needless acute care use

Worsening dyspnea

Communication between patient/caregiver and team

Multidisciplinary team assessments at home

Disease progression

Dyspnea improved, resumes prior activities outside home (camping, fishing, running errands)

Identify and treat reversible causes. Modification of dyspnea management action plans

Maintain QOL

Increase days spent at home: patient-centered goal

Avoids needless acute care use
Clinic 3 Month F/U

- Worsening dyspnea, PFTs
- Investigations: HRCT, no reversible cause
- Oxygen titration 10LPM at rest
- Opioid titration (0.5 mg hydromorphone oral tid, 0.5-1 mg buccal before activity)
- ACP: Transition to home care based on patient wishes

HM: hydromorphone
Harry’s Journey Over the Next 2 Months

Care at home
- NP/RT/MD
- Dyspnea assessment
- Oxygen titration (15 L)
- Opioid dose titration to meet patient needs
- Education and support
- Reassess patient goals: Last wish to travel to BC

Team work to facilitate patient goal
- NP/RT/MD
- Anticipate, educate, prepare
- Arrange oxygen for trip
- NP and MD: developed crisis action plans, meds on hand, who to call
Harry’s Journey Over the Next 2 Months

End-of-life at Home

- 1 week after trip to BC, family calls home care
- Dyspnea assessment
- Team: Anticipates death and prepares family
- Oxygen: 18-19 lpm via O2 concentrators + 5 lpm via E cylinder for rest and increase to 10-15 lpm for exertion
- Oral hydromorphone 2 mg qid for baseline; 2 mg buccal preactivity; 4 mg buccal q10 min prn
- Patient passes away peacefully at home, according to his preference

HM: hydromorphone
Impact of Multidisciplinary Collaborative Care

**Living with IPF with Dignity**
- Patient needs assessed and goals prioritized
- Early integrated palliative approach & Support system engaged early
- Regain hope, meaning and purpose
- Maximize function with QOL maintained
- Maintain sense of control and empowerment

**Dignity in Dying and Death**
- Patient EOL wishes respected and goals prioritized
- Symptoms well managed
- Avoid hospitalization
- Remain independent and functional
- Die peacefully at home with family prepared and supported
Impact of Multidisciplinary Collaborative Care

- Analysis of outcomes pre- and post-MDC care model (32 IPF decedents 2009-2016)
- 55% increase in home deaths and 62% decrease in hospital deaths

 Needs assessment led to early opiate start in 95%

 Prioritization of ACP led to documentation in 100%

 Early interventions lead to improved EOL Care
 Greater adherence to patient wishes; 71% died in preferred location
 Reduced acute care use and decrease in hospital deaths by 62%

Early Integrated Palliative Approach for Patients with IPF: Experiences and Perceptions of Bereaved Caregivers

Key Findings:

- Narratives support early integrated palliative approach in care
- Reduced symptom burden and related anxiety and distress
- Good quality of life, death and dying, and bereavement due to collaboration and open communication among patients and care team
- ACP enabled caregivers to feel informed, prepared and supported when death was near

“ILD is absolutely a death sentence, but how you get from the diagnosis to the end can be managed in a whole bunch of different ways. When you create care teams who are really good at what they do, and really believe in being patient centered, then this is the thing that you end up with, people who are absolutely devastated by the passing of their mom but not traumatized by it.” (SR, bereaved caregiver)
Summary

• IPF is a fatal disease with high symptom burden and unmet care needs

• Use of needs assessment tool can trigger early and individualized symptom based interventions

• Early interventions for symptoms, ACP, better communication and coordination of care facilitate better QOL & QODD

• Interdisciplinary approach with appropriate oxygen titrations and low-dose opiates are effective in dyspnea management

• Prioritization of ACP by inclusion in care can increase rates of success and improve communication

• Networking with community team can support patients at home and improve coordination of care
Questions?

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