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What patients with Idiopathic Pulmonary Fibrosis and caregivers want – Filling the currents gaps with patient reported outcomes and experience measures.

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Running head: Unmet needs and potential solutions in the Idiopathic Pulmonary Fibrosis patient pathway

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Abstract

Idiopathic pulmonary fibrosis is a progressive disease, with a high mortality within the first 3-5 years from diagnosis and a poor quality of life mainly because of the burden of symptoms, such as dyspnea and cough, occurring usually many months before the diagnosis. Although available antifibrotic therapies slow down disease progression, they have no impact on quality of life. Moreover, healthcare around idiopathic pulmonary fibrosis patients is very often "disease-centered", and relies on clinical surrogate outcomes that are poorly related to patients' quality of life and disease experience.

Therefore, patients with idiopathic pulmonary fibrosis have several unmet needs in all domains of health that they wish to see recognized and addressed in the context of the treatment of their disease and its complications.

In this review, we summarize the care pathway from the patients' perspective, identifying current gaps in care, education, support and communication between patients with idiopathic pulmonary fibrosis, their caregivers and care teams during the patient journey. The role of patients reported outcomes (PROs), PRO measures (PROMs) and patient reported experience measures (PREMs) in their care is discussed, as well as the need of disease specific PROs/PROMs/PREMs.

Introduction

Patient experience is an important quality index of healthcare, and the use of patient-reported outcome (PRO) and experience measures (PROMs, PREMs) is recommended for measuring the performance of healthcare systems worldwide.[1,2] In fact, there is an increasing recognition of the need to include patient perspectives among the outcomes of medical care. [1,3] Policy makers, funding and regulatory bodies recognize that the lack of patient-centered care results in unmet patient needs, high costs and ultimately ineffective care.[4,5] Historically, clinicians and researchers have favored "objective" outcomes, such as mortality, for the assessment of efficacy of medical interventions. Unfortunately, these measures do not always align with what is most important to the patients (i.e. the impact of disease on their wellbeing and their ability to live a fulfilling life[5]), creating unmet needs and care gaps.[6] The development and use of PRO and related measures is an attempt to include patient perspectives in designing systems and approaches that can truly meet their needs.[4]

Idiopathic Pulmonary Fibrosis is a progressive disease with a high mortality in 3-5 years from diagnosis.[7,8] It is characterized by poor quality of life and high symptom burden.[9,10] Current idiopathic pulmonary fibrosis care delivery models do not address patient needs. As a result, patient advocacy bodies have generated calls for action to develop a holistic approach to idiopathic pulmonary fibrosis patient care,[11–13] with the aim to include good quality care at all stages of disease.[10,14] A holistic approach should address disease management as well as individual patient concerns.[15]

The lack of appropriate and dedicated tools such as patient reported outcomes (PROs) and their respective measures (PROMs, PREMs) in idiopathic pulmonary fibrosis contributed to current gaps in the field[11,12].

In this review, we summarize the available evidence on what idiopathic pulmonary fibrosis patients and caregivers value and what they perceive as care gaps; finally, we highlight the urgent need to develop and validate disease-specific PROMS and PREMS.[16,17]

Patient Reported Outcomes, patient reported experiences and their measures.

Patient reported outcomes (PROs) are directly reported by patients and provide information about the perspective on their own health status and quality of life in the context of a disease and its treatments.[18] Patient reported experiences (PREs) describe how patients experience healthcare, and reflect patient perspectives on various facets of the care processes. They are also self-reported interpretation[19] from patients and their families, representing a valuable opinion to assess quality and to improve healthcare.[20]

Patient reported outcome measures (PROMs) are dedicated tools to assess health outcomes and quality of life of patients.[21,22] They are multidimensional as they address various domains, such as symptoms, functional status, psychological, social and spiritual wellbeing. Different types of PROMs are available: they can be classified as generic, disease or domain-specific.[18,22] Generic PROMs are useful for the whole population (healthy and diseased), and focus on function, independence and mood. Disease-specific PROMs focus on impact of a single disease or group of diseases, such as on various aspects of patient's life. Domain-specific

PROMs are focused on particular symptom (e.g. dyspnea) or organ/systems (e.g. gastro-intestinal, colorectal).

Patient reported experience measures (PREMs) are tools to gather data on patient views of their healthcare experience, rather than on outcomes. They indirectly assess the quality of care and they can be classified as relational or functional.[19] Relational PREMs focus on patients' experience of their relationship with the care team. Functional PREMs focus on objective experience of care facilities (e.g. type of care model, support, waiting times, access, cleanliness, comfort). PREMs can be generic or disease specific.[19]

The Food and Drug Administration (FDA) and the Patient-Centered Outcome Research Institute (PCORI) have released guidelines to help researchers develop and use PROs and PROMs in research.[18] They recommend that studies of any intervention should include the impact on PROs, PROMs and PREMs.[16,18] There is increasing recognition that clinical care also benefits from their use.[23–25] Incorporation of PROs in clinics can facilitate a multidimensional, patient-centered evaluation of the disease, identify unmet needs, direct clinician attention to these needs and better inform policy makers in the light of identified patient goals.[11,16,17] The use of PREMs can impact the quality of patient-provider communication by facilitating inclusion of patient perspectives and their narratives.[26–29]

Patient Reported Outcomes and Experience Measures for idiopathic pulmonary fibrosis.

Targeted strategies to improve patient-centered care, quality of life and patient experience are paramount in a progressive disease such as idiopathic pulmonary fibrosis.[9] Unfortunately, all major idiopathic pulmonary fibrosis clinical trials focused on objective

measures such as lung function, and PROMs have only been used as secondary endpoints.[30,31] The past decade has seen an increase in PROMs use in idiopathic pulmonary fibrosis, generally including dyspnea and cough questionnaires, quality of life measures and depression/anxiety questionnaires.[9] Generic PROMs such as Euroqol-5D and short form-36 have been used in this condition.[32,33] Disease specific PROMs, such as Saint George Respiratory Questionnaire (SGRQ)[34] and COPD assessment Test (CAT),[35] although not specific for idiopathic pulmonary fibrosis , have also been used. In addition, idiopathic pulmonary fibrosis specific tools such as SGRQ-IPF[36], the cross Atlantic tool to assess quality of life in IPF (ATAQ-IPF-ca)[37] and King's Brief Interstitial Lung Disease (K-BILD) questionnaire[38] were also developed. There are several other specific questionnaires in various stages of development: "Living with IPF" (L-IPF)[39] was reviewed by the FDA and is undergoing further validation testing; "IPF-PROM"[40] is currently under development. The other domain specific PROMs used in idiopathic pulmonary fibrosis are the University of California San Diego Shortness of Breath Questionnaire (UCSD SOB),[41] the modified Medical Research Council Dyspnea Questionnaire (mMRC)[42] and again the K-BILD,[38] the SGRQ-IPF[36] and the ATAQ-ca[37] for dyspnea, the Leicester Cough Questionnaire (LCQ)[43] and the Cough Quality of Life Questionnaire (CQLQ)[44] for cough, as well as the Hospital Anxiety and Depression Scale (HADS) for anxiety and depression.[45]

Currently, there are no PREMs available for clinical use. The Patient Experiences and Satisfaction with Medications (PESaM)[46] questionnaire is an example for idiopathic pulmonary fibrosis PREM that was developed to assess patient experiences with pirfenidone for the treatment of the disease and it is awaiting a validation study.

The patient pathway in idiopathic pulmonary fibrosis and the patient-centered approach

The diagnosis and treatment of this disease is a complex clinical process, involving a multidisciplinary team.[47,48] Most patients describe the diagnostic process as a major struggle due to lack of awareness, delayed access to tertiary centers, lack of patient-centered care and lack of reliable information about the disease (Table 1).

At least three important overlapping phases are included in the patient pathway: 1) early and confident diagnosis 2) progression of the disease 3) EoL. Needs assessment, individualized care delivery, timely communication and planning should support the patients in all the phases of their journey with the disease.[9,12] Appropriate and precise tools, such as PROMs and PREMs, should be used by the multidisciplinary team to understand the complexity of the patient´s condition and to tailor supportive care.[9,13]

Table 1 and 2 report the main pitfalls of the current care of patients with idiopathic pulmonary fibrosis. They identify gaps in the current clinic models, and provide suggestions for the use of PROMs.

Early and accurate diagnosis with patient-centered education and support

Recent studies showed that delayed recognition of symptoms in primary care leads to delayed referral and diagnostic delays.[49,50] Patients with idiopathic pulmonary fibrosis have a poor quality of life already at diagnosis, therefore early diagnosis and disease specific treatment/supportive care strategies are crucial to preserve it.[9,10] Cough, dyspnea and fatigue are frequently underrecognized in primary care, leading to delays in referral[51,52]. The use of

generic PROMs and PREMs in primary care may bring dyspnea and cough to light, raising the suspicion and early detection of the disease.

When patients are informed of this diagnosis, communication is frequently insensitive and incomplete, lacking empathy or expression of support, and this has detrimental effects on how patients perceive their disease.[53,54] The use of relational PREMs can explore patient experience, measuring if they were treated with empathy and the quality of communication and care provided.

The majority of patients do not receive holistic care, and their symptoms, psychosocial-emotional and caregiver needs are frequently overlooked.[12,53,54] The CaNoPy study[55] revealed that the standard clinical assessments do not truly reflect patient experience of the disease. Bajwah et al[27] found that while all patients and their caregivers identified dyspnea as their main concern, their care providers had limited appreciation of its impact on patient's quality of life, showing that attention must be paid to detecting such needs and align provided care to idiopathic pulmonary fibrosis patient's individualized goals from the time of the diagnosis. In this context, PROMs and PREMs offer the possibility to make clinicians aware of what patients really feel, need and want from the beginning of their journey.

On the other hand, the patients frequently experience fear, anxiety, worry, hopelessness and helplessness and would like to discuss strategies to address their daily needs.[12,53,54] To fill this gap, the multidisciplinary teams, including specialized nurse, physiotherapist, respiratory therapist, social worker, need training in best supportive and palliative care including meticulous needs assessment, symptom relief strategies and patient education. Patients' group educational meetings should be used to integrate the patient-

centered approach. The use of PROs, PROMs and PREMs in these situations can help identify common priorities and establish a shared understanding of anticipated outcomes and care experiences, thereby improving communication.

Tools such as the palliative care needs assessment tool in interstitial lung disease (NAT: PD –ILD),[56] symptom assessment scales and quality of life measures can provide useful measures to assess patients health and their lives at the time of the first assessment (Table 1). Other needs may not be captured by the above tools; in that case, the measure yourself medical outcome profile (MYMOP) questionnaire[57] can be used. The advantage with MYMOP is that it asks the patient to identify the most important symptom instead of forcing fixed choices. In idiopathic pulmonary fibrosis, where education and information needs are largely unmet, use of MYMOP may allow patients to underscore this needs.

PROMs alone will not lead to improved outcomes without a change in clinicians approach to care. Ramadurai et al[58] explored the education and information gaps and described both disease specific as well as individualized content for both patients and caregivers: Education is key as it increases understanding, sense of control and empowerment. Such information needs to be delivered at the first visit and revisited throughout the course of the disease.

Advanced care planning is another very important part of the patient-centered approach, and needs to be addressed early at the beginning of the patient journey with idiopathic pulmonary fibrosis. It implies an interactive patient-centered communication process between patients, their families and the healthcare[14,53,59] All major guidelines[48] endorse early advanced care planning discussions to (1) alleviate patient and caregiver burden, (2) provide

relevant information, (3) help prepare families for end of life and (4) eventually deliver care in alignment with patient preferences. Unfortunately, over 80% of the patients with pulmonary fibrosis do not have advanced care planning discussions, and end up dying in hospital without any palliative care involvement.[60] At the core of these issues is an underlying misperception that palliative care is relevant only in EoL care. Therefore, most patients are referred too late and consequently also believe that palliative care is an EoL care only[10].

Given the existing shortages of palliative care experts, advanced care planning should be regarded as the responsibility of the specialist and primary care teams.[61] Preliminary evidence suggests that implementing these conversations tertiary clinics has direct impact on care.[14] Further efforts are needed to develop advanced care planning models and PROs to assess its quality in idiopathic pulmonary fibrosis care.[10]

Pulmonary rehabilitation is an excellent opportunity to improve functional capacity and quality of life. Patients should be referred to such programs early and whenever possible as needs escalate. These programs can be tailored to meet the education and information needs including advance care planning needs in this patient population.

Caregivers should be involved and integrated early on in the patient journey. The devastating effects of idiopathic pulmonary fibrosis extend beyond patients, to their families and friends. Use of caregiver burden surveys may highlight their needs and lead to provide support or more resources to improve their living. Caregiver presence and ability to help is vital to patient well-being in this condition, therefore, care providers must actively seek out ways to identify and address caregiver concerns and to educate them on how to face the challenges of the disease.

Progression of the disease

idiopathic pulmonary fibrosis remains an unpredictable disease with varying rates of progression and inability to predict decline at individual level,[7,48] also for the occurrence of acute exacerbations.[62,63] Most patients are not well informed about this, and uncertainty creates anxiety that affects their living. There is a need to educate patients on what to expect and how to prepare for future decline in an easy-to-understand fashion. This type of education must be individualized as the rates of progression will vary among individuals.[64]

Impairment of quality of life in idiopathic pulmonary fibrosis is largely driven by worsening of symptoms, such as dyspnea, cough and fatigue over time.[10,27,65] Daily activities, recreation, pleasure and employment are often directly affected by the burden of symptoms; anxiety, depression and social isolation result from the limitations caused by the disease (Table 2). Currently, there are no dyspnea questionnaires that capture the newly defined episodic breathlessness that may require different treatment strategies.[66,67] Many patients report that the practical information needed to live with their disease is generally missing[68], perceiving that care in the specialist centers tends to be "disease-centric" without enough emphasis on symptom management, living life well, maintaining hope and preparing for death with dignity.[69] Mularksi et al. recommend that patients and caregivers should be provided dedicated dyspnea education and action plans as part of home dyspnea management.[70] Dyspnea self-management in idiopathic pulmonary fibrosis is not described outside of a few case reports.[59,71] As disease

and symptom needs progress, the efforts to address education and information need to follow in parallel. The use of PREMs and PROMs (Table 2) in this context can lead to recognition of patient needs and prompt required action by the multidisciplinary team.

In addition, the patients want to be supported outside the clinic: the CaNoPy study[69] highlighted the patients' need to identify early deterioration and to start simple interventions - such as timely supplemental oxygen - to preserve quality of life without waiting for follow-up clinic appointments. These findings suggest that self-monitoring and management at home are important patient's needs to regain a sense of control and to feel empowered. In this context, PROMs can be easily monitored, detect clinical changes and lead to meaningful early interventions. Scales to measure self-efficacy, confidence and patient activation measure[11] along with symptom scales and needs assessment tools can also be useful to assess impact of interventions.

Furthermore, most patients in this phase want information about decline and EoL. Advanced care planning discussions can address these needs by identifying wishes, providing context discussing clinical and functional status and engaging patients in shared decision making.[59] It is paramount to ask patients directly; therefore, PROMs are invaluable in this situation.

Education of caregivers on how to recognize and handle worsening is essential in idiopathic pulmonary fibrosis care. Caregivers are often not prepared to help their loved ones, and both describe being overcome with frustration, hopelessness and helplessness.[72] Continued caregiver engagement in care process and decision making is, therefore, desirable in idiopathic pulmonary fibrosis.[59] Preliminary data show that collaboration between support

groups and community services improves care and reduces hospitalizations at EoL.[73,14] Additionally, patients with pulmonary fibrosis find support groups to be instrumental in developing positive thinking, learning how to cope well, using emotion focused strategies to overcome depression and to reduce social isolation.[74]

End-of-Life

EoL care in idiopathic pulmonary fibrosis is poor and it is marked by lack of timely and appropriate palliative care access, symptom management and advanced care planning.[75,76] Several studies pointed to a very high caregiver burden due to the lack of a patient and family-centered approach to care and easy access to community resources for support.[54,69,72] Ahmadi et al[75] and Rajala et al[76] have shown that most patients die in hospitals with aggressive, costly and ineffective therapies, poor symptom assessment and control, and low family presence at death. Death is "unexpected" and there is no anticipatory guidance for families and inadequate bereavement support. When compared to lung cancer, EoL care in idiopathic pulmonary fibrosis is poorer.[75] Qualitative data from patient, caregiver and care provider interviews indicate the lack of expertise in dealing with idiopathic pulmonary fibrosis-specific EoL issues and the urgent need to develop symptom management protocols.[59,77] An explorative analysis suggested that early implementation of palliative care can result in greater concordance between patient preferences and care provided, increase home deaths and reduce EoL acute care use.[14,76] Bereaved caregivers who experienced early integrated palliative approach in idiopathic pulmonary fibrosis care reported being better prepared to EoL, improved quality of life, and better quality of dying, in collaboration with the multidisciplinary team.[14,77]

In conclusion, palliative care processes must begin early with meaningful advanced care planning conversations, systematic and ongoing needs assessment, integrated symptom management, anticipatory guidance, education and support that can effectively address issues and questions when the disease worsens. The use of PROMs and PREMs can play a major role in this process (Table 2), helping in identifying patient and caregivers needs.

Conclusions

Advocacy bodies, policy makers, regulatory authorities, pharmaceutical industry and healthcare systems, including patients and their caregivers, recognize the need for the use of PROs, PROMs, PREMs in idiopathic pulmonary fibrosis. Their use can improve care and research, facilitate patient-centered care by identifying needs and informing care decisions. It must be recognized that patient needs and experiences are as important as objective measures in the health outcomes assessment and shared care decisions with the patients. Knowledge translation cannot occur without changing attitudes, culture of practice and removing practical barriers to provision of patient-centered care.

An ideal setting to implement PRO/PROM/PREM is the idiopathic pulmonary fibrosis multidisciplinary team. Many questions regarding its composition and governance remain unanswered, requiring further studies to better define the role of these tool in earlier diagnosis, better management and follow-up. The ability to assess and control symptoms, develop self-efficacy, access to psychosocial, emotional and spiritual support and information on the disease and its course are important unmet needs for both idiopathic pulmonary fibrosis patients and caregivers.[11,12,49,69] Furthermore, PROs/PROMs/PREMs need to be validated to accurately identify patient needs, to measure efficacy of interventions, to improve various domains of

quality of life and care, and to address educational needs at various time points in the disease course.

There is a huge need to support the multidisciplinary work focused on identifying gaps across the patient journey, and PREMs/PROMs can be instrumental to this goal.[9,10] Digital patient interfaces are already available, allowing their measurement at chosen time-points,[78] with scores made readily available for decision making at the time of patient clinical encounters.

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Table 1. Patient Reported Outcome Measures (PROMs) and Patient Reported Experience Measures (PREMs) in diagnosis and treatment

Patient perspective (PRO)	Pitfalls	How to improve patient pathway	Potential PROMs and PREMs
Frequent delays in primary care[54]	Lack of: 1) attention to patient illness narratives; 2) IPF awareness	Improve access to secondary and tertiary care. Include patient perspective and increase IPF awareness	Assessment of well-being, health status (SF-36)[33] and symptoms (MYMOP)[57].
Healthcare experience disconnected from patient reality and daily needs[54,79]	Lack of: 1) MDT and PC approach; 2) Prioritization of symptom/need assessment; 3) Integrated advanced care planning; 4) Communication and information skills	Integrate PC approach within MDT, importance of symptom management, initiate early SCP discussions. Importance of discussion strategies about how to improve QoL, self-management and empowerment	Symptom scale to detect needs (NAT, PD- interstitial lung disease [56], mMRC[42], UCSD SOB[41], LCQ, CQLQ) QoL measures: SGRQ-IPF[36], K-BILD[38], ATAQ-IPFca[37], L-IPF[39] Relational PREMs to identify communications gaps, perceptions of poor care. PEI[80]: to determine if patient received sufficient education
Adverse effects from antifibrotic therapies and resulting poor perceived QoL[12,81]	Lack of: 1) Empathy and training in communication; 2) Attention to patient narratives; 3) more therapeutic options; 4) Integrated advanced care planning conversations; 5) MDT support & education	Perform routine needs assessment with good communication skills. Provide patient education & support. Facilitate inclusion of patient narratives. Integrate advanced care planning in specialist care. Provide well-trained care teams.	IPF-PREM: to explore patient and caregiver perceptions of care. HADS[45]: to detect depression, anxiety, hopelessness. PESaM questionnaire (awaiting full validation)[46]
Need for support groups[53]	Lack of: 1) Availability of	Promote access to patient support	PEI, patient activation measure

	support groups; 2) Information about available support groups	groups. Provide information about available support groups	[11,46]: to measure enablement and patient activation of coping

PRO: patient reported outcome; PROMs: patient reported outcome measures; PREM: patient reported experiences measures; IPF: idiopathic pulmonary fibrosis; MYMOP: measure yourself medical outcome profile; SF-36: short form health survey 36; MDT: multidisciplinary team; PC: palliative care; advanced care planning*: advanced care planning; NAT:PD-* interstitial lung disease *: palliative care needs assessment tool in interstitial lung disease; mMRC: modified Medical Research Council Dyspnoea Questionnaire; UCSD SOB: University of California San Diego Shortness of Breath Questionnaire; LCQ: Leicester Cough Questionnaire; CQLQ: cough quality of life questionnaire; QoL: quality of life; SGRQ-IPF: Saint George Respiratory Questionnaire – IPF; K-BILD: King´s College Brief Interstitial Lung Disease Questionnaire; ATAQ-IPF: the cross Atlantic tool to assess quality of life in IPF; L-IPF: Living with IPF, Tool to assess Quality of Life in IPF; PEI: patient enablement instrument; PESaM: patient experiences and satisfaction with medications questionnaire; HADS: Hospital Anxiety and Depression Scale;* patient activation measure*: Patient satisfaction, empowerment and activation.*

Table 2. Patient Reported Outcome Measures (PROMs) and Patient Reported Experience Measures (PREMs) in disease progression and End-of-Life

Patient perspective (PRO)	Pitfalls	How to improve patient pathway	Potential PROMs and PREMs
Suboptimal or missing symptom management and resulting depression[27]	Lack of: 1) recognition of patient experience of disease; 2) training and education in PC; 3) evidence-based guidelines on PC treatment; 4) patient and caregiver reluctance to accept PC	Change philosophy and care delivery models with systematic symptom assessments and early integration of PC. Provide training in PC. Correct misperceptions of PC	NAT: PD- interstitial lung disease [56], MYMOP[57], MSAS[82], UCSD SOB[41] and cough specific tools: LCQ[43], CQLQ[83] can detect burden and track changes with therapies
Poor perceived and worsened QoL (physical, psychosocial, emotional, spiritual and existential needs)[27,58]	Lack of: 1) whole person approach; 2) standardized approach to needs assessment; 3) time to properly assess these needs	Provide strategies to preserve QoL Provide anticipatory guidance, use of symptom action plans. Educational resources for patients and caregiver	K-BILD[38], ATAQ-IPFca[37], SGRQ-IPF[36], L-IPF[39], NAT: PD-interstitial lung disease [56] to assess and monitor QoL. Use of physical activity tracking
Significant burden from use of cumbersome oxygen delivery equipment[12]	Lack of: 1) portable equipment for high flow oxygen delivery; 2) education and support	Optimize oxygen delivery. Include dedicate staff in MDT for patient education & support	MYMOP[57] to identify gaps and goals; IPF PREMs for communication and education gaps
Lack of psychological support[12]	Lack of: 1) patient-centered care; 2) dedicated professionals in the MDT	Adopt patient-centered care with adequate MDT staffing	NAT: PD- interstitial lung disease [56] (to detect unmet needs) HADS[45] and MYMOP[57] (depression)
Poor EoL care. Poor quality of Dying and death[27,75,76]	Lack of: 1) PC approach and delayed referral; 2) clinical guidelines; 3) training in sensitive, patient-centered	Implement end-to-end patient-centered care models Adopt early advanced care planning	NAT: PD- interstitial lung disease [56] (for palliative care needs), MSAS[82],MYMOP[57], L-IPF[39], K-B interstitial lung

	communication	Provide education and support to the MDT	disease (to assess QoL and quality of dying)[38], IPF-PREM (to assess care delivery processes)
Caregiver distress and increased burden[27]	Lack of: 1) understanding of the caregiver needs and engagement; 2) anticipatory guidance; 3) action plans and support for caregivers	Recognize caregiver in care Assess needs and provide relevant support Engage advanced care planning early Provide anticipatory guidance and services at home	Caregiver burden survey, to detect needs.[49,69] Need to develop tools to track panic episodes at home

PRO: patient reported outcome; PROMs: patient reported outcome measures; PREM: patient reported experiences measures; NAT:PD- interstitial lung disease *: palliative care needs assessment tool in interstitial lung disease; MSAS: memorial symptoms assessment scale; PC: palliative care; MD: multidisciplinary; MYMOP: measure yourself medical outcome profile; IPF: idiopathic pulmonary fibrosis; K-BILD: King´s College Brief Interstitial Lung Disease Questionnaire; ATAQ-IPF: the cross Atlantic tool to assess quality of life in IPF ; SGRQ-IPF: Saint George Respiratory Questionnaire – IPF; L-IPF: Living with IPF- QoL tool; QoL: quality of life; LCQ: Leicester Cough Questionnaire; CQLQ: cough quality of life questionnaire; HADS: hospital anxiety and depression scale;* advanced care planning*: advanced care planning.*

Clinical Significance

- Several unmet needs of patients, caregivers and healthcare providers were identified in the idiopathic pulmonary fibrosis patient pathway, from early diagnosis to end-of-life.
- Disease-specific patient reported experience measures (PREMs) and patient reported outcomes measures (PROMs) offer the opportunity to fill the current gaps in the healthcare of these patients.
- A clear understanding of the patient pathway is paramount to implement patient-centered healthcare, research and education in this particular patient group.