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Diagnostic and treatment standards in idiopathic pulmonary fibrosis in the era of antifibrotic drugs in Poland: A real-world practice survey

Abstract

Introduction: Currently, only two drugs have been shown to modify the inevitable natural history of idiopathic pulmonary fibrosis (IPF). Changes in the reimbursement policy for antifibrotic drugs in Poland have led to the availability of pirfenidone from January 2017 and nintedanib from March 2018 for the treatment of Polish patients with IPF. This study aimed to evaluate the possible changes and shortcomings in the clinical practice standards in IPF in the era of access to antifibrotic therapy in Poland.

Material and methods: A real-world data survey was performed among physicians attending the Polish Respiratory Society Congress held in May 2018. The present survey was a follow-up to the previous survey undertaken in 2016, before the availability of antifibrotics in Poland.

Results: A total of 99 physicians participated in the survey, among which 80% were pulmonologists. The majority of participants (83%) represented hospital-based clinicians and most of them (93%) were involved in interstitial lung diseases (ILD) management. As many as 63% of the respondents elaborate the final diagnosis of IPF working with the expert radiologist routinely, 47% do that in the cooperation with other pulmonologists, and if a biopsy was performed 39% discuss its results with the expert pathologist. Bronchoalveolar lavage (BAL) and surgical lung biopsy (SLB) would never be recommended in the differential diagnosis of IPF by 9% and 16% of the respondents, respectively. Corticosteroids (CS) or a combination of CS and immunosuppressants (IS) is still recommended by 22% of participants. Proton pump inhibitors (PPI) in the case of symptomatic GERD are prescribed by 44% of the respondents, and 12% prescribe PPI regardless of GERD symptoms. Pirfenidone is used by 70%, and nintedanib by 48% of the respondents. Only 39% of the respondents refer patients with IPF to professional rehabilitation centers.

Conclusions: The level of cooperation between pulmonologists and other specialists in the diagnostic workup of IPF is unsatisfactory. IPF treatment practices in the era of access to effective drugs in Poland require immediate improvement. There is an urgent need to develop the local Polish practical guidelines to improve the management of IPF.

Key words: idiopathic pulmonary fibrosis, IPF, real-world data, clinical practice, diagnosis

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Introduction

Idiopathic pulmonary fibrosis (IPF) is a specific form of chronic, fibrosing and progressive idiopathic interstitial pneumonia (IIP) that occurs primarily in older adults. IPF is characterized by histopathologic and/or radiologic pattern of usual interstitial pneumonia (UIP) [1]. The disease represents one of the most common interstitial lung diseases (ILD) [2]. The clinical course of IPF is variable and unpredictable, nevertheless, median survival is known to be between 2.5 to 3.5 years, yet some patients live much longer [3]. According to the actual international clinical practice guidelines IPF may be diagnosed based on the presence of a radiologic pattern of UIP on

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high-resolution computed tomography (HRCT) of the lungs after careful exclusion of known causes of pulmonary fibrosis. In the case of other than typical radiographic UIP pattern on HRCT of the lungs (probable UIP, indeterminate for UIP or alternative diagnosis) a surgical lung biopsy (SLB) is recommended for confirmation of the presence of histopathologic UIP pattern for definite IPF diagnosis [1].

For patients with newly detected interstitial lung disease (ILD) of unknown cause, who are suspected of having IPF, multidisciplinary discussion (MDD) is suggested for the diagnostic decision-making process. It is recommended that the multidisciplinary team (MDT) involved in the diagnostic process of IPF and serving for MDD should consist of pulmonologist, radiologist, and pathologist. Participants of MDT should have expert knowledge and experience in the differential diagnosis of ILD. Therefore, the diagnosis of IPF should be undertaken in ILD reference centers. Benefits of the direct interaction during MDD in the diagnostic work-up of IPF include a shorter delay for the correct diagnosis and therapy, limitation of additional, unnecessary diagnostic testing and incorrect therapies [1].

Historically used pharmacotherapy for IPF including corticosteroids (CS), immunosuppressants (IS), N-acetylcysteine (NAC) or combination triple therapy including CS, azathioprine (AZA) and NAC is nowadays strongly contraindicated due to the lack of benefit and increased number of adverse effects [4]. Based on the several suppositions of the potential role of gastroesophageal reflux (GER) in IPF pathogenesis and progression, actual international practice guidelines recommend that all patients should be treated with antacid therapy (AAT), which may decrease the risk for microaspiration-associated lung injury or damage, a mechanism that has been postulated to cause or worsen IPF. AAT treatment is suggested by the international IPF treatment guidelines regardless of whether GER symptoms are clinically apparent or not [4]. Non-pharmacological interventions, such as supplemental oxygen therapy, pulmonary rehabilitation programs, and lung transplantation are also recommended in the holistic management of IPF.

Currently, only two drugs have been shown to modify the inevitable disease course. Pirfenidone and nintedanib, independently, have been shown to slow the disease progression limiting the decline of lung function in patients with IPF [5, 6]. The European Medicine Agency (EMA) registered pirfenidone in 2011 and nintedanib in 2015 for the treatment of IPF. Both medications are recognized as an actual standard of pharmacological treatment of the disease [4]. Changes in the reimbursement policy for antifibrotic drugs in Poland have led finally to the availability of pirfenidone from January 2017 and nintedanib from March 2018 for the treatment of Polish patients with IPF.

Despite the availability of effective pharmacological treatment options for IPF patients in Poland, local evidenced-based diagnostic and therapeutic standards established for Polish IPF patients with regards to the specificity of the national health service system and specific patients' needs and expectations are lacking. The previous survey on common practices of the Polish physicians in the management of IPF, undertaken before the availability of antifibrotic therapy in Poland, pointed out unsatisfactory local diagnostic and therapeutic standards in IPF [7].

The present real-world practice survey aimed to collect follow-up data regarding possible changes and shortcomings in the clinical practice standards in IPF diagnosis and treatment in the era of access to the antifibrotic therapy for Polish patients with IPF.

Material and methods

Physicians attending the Polish Respiratory Society Congress held in May 2018, representing mostly pulmonologists and other professionals involved in the management of different fields of respiratory medicine, were invited to participate in the study. The self-prepared questionnaires consisting of 20 questions divided into 3 parts: 1. Data describing the responding physician; 2. Diagnosis of IPF; and 3. Treatment of IPF were distributed at the entrance to the lecture room during the ILD sessions of the congress and collected at the end of the session. All collected categorical data are presented as absolute numbers and relative frequencies (n, %). Continuous data are expressed as mean with standard deviation (SD). For data clarity percentage values are rounded to the nearest whole number.

Results

Characteristics of participants

A total of 99 physicians participated in the survey. Summary of characteristics of study participants is shown in Table 1. The most prevalent group of respondents (38%) were in the age between 41 and 50 years old. Pulmonary medicine

Table 1. Characteristics of respondents

Question	Answer	n	%#
Age [years]	< 30	2	2
	30–40	18	18
	41–50	38	38
	51–60	33	33
	> 60	8	8
Specializations*	None	5	5
	Internal medicine	70	70
	Pulmonary medicine	79	80
	Allergology	6	6
	Family medicine	5	5
	Other	3	3
Place of work*	Hospital — academic/research center	45	45
	Hospital — non-academic/non-research center	38	38
	Pulmonary Diseases Outpatient Clinic	48	48
	Family medicine practice	18	18
	Other	7	7
Professional experience in pulmonary medicine [years]	< 5	16	16
	6–10	12	12
	11–20	33	33
	21–30	29	29
	31–40	7	7
	> 40	1	1
Involved in ILD diagnosis and treatment?	Yes	63	63
	Sometimes	30	30
	Never	6	6
If involved — number of IPF patients per year	< 5	35	35
	6–10	36	36
	11–20	17	17
	21–30	3	3
	31-40	5	5
	> 40	2	2

*The sum of particular items does not equal 100%; # rounded to a whole number

as the medical specialty was declared by 80% of the respondents (68% of pulmonologists reported having also an internal medicine specialty), and 11% were general internists only, without completed additional specialization. The other respondents' specializations included: family medicine (5%; 3 out of 5 family doctors had basic specialization in pulmonary medicine), allergology (6%; 5 out of 6 allergists had basic specialization in pulmonary medicine), 3% declared being pediatricians, 2 out of 3 having pulmonology as the basic specialization. The university hospital or research institute was the main place of work for 45% of the respondents, and 42% of them were joining it with outpatient pulmonary medicine practice. The non-academic hospital was the main place of work for 38% of the respondents, and 45% of them were joining inpatient with outpatient practice. Sole outpatient pulmonary department as the main place of work was declared by 12% of the respondents. A significant majority of respondents (70%) declared experience in the field of pulmonary medicine exceeding 10 years. The distribution of professional experience in the area of pulmonary medicine is summarized in Table 1.

The majority (63%) of respondents declared being involved in the diagnosis and treatment of ILD routinely, 30% sometimes, and 6% never. Of those who declared being involved routinely or sometimes, 35% have had below 5 IPF patients, 36% between 6 and 10, 17% between 11 and 20, and 10% over 20 patients with IPF under their supervision in the previous 12 months, see Table 1. Those who declared not being involved in the ILD diagnosis and treatment (6%) refer patients with suspicion of IPF directly to the nearest pulmonary medicine department (2 respondents) or refer them after having confirmed suspicion of IPF in the HRCT examination (4 respondents).

Diagnostic standards

As many as 17% of the respondents agreed, that BAL should be performed always in patients suspected of IPF, 74% would recommend BAL in selected patients, and 9% declared that BAL is useless or only of minimal diagnostic value in IPF patients, see Figure 1.

SLB would never be recommended in patients suspected of IPF by 16% of the respondents, would be recommended rarely (in less than 20% of cases) by 74%, often (in more than 20% of cases) by 8%, and would be recommended always by 2% of the respondents, see Figure 2. The majority of participants (53%) refer patients suspected of having IPF for transbronchial lung cryobiopsy (TBLC) rarely (in less than 20 % of cases), 9% do that frequently (in > 20% of cases), 1% do that always, and 37% of the respondents never refer patients with IPF suspicion for TBLC.

Only 1% of the respondents diagnose IPF on their own, 10% ask other specialists only in case of difficulties, 47% elaborate the final diagnosis in the discussion with other pulmonary medicine specialists, and 63% ask experienced radiologists for advice routinely. In addition, when the lung biopsy was performed, 39% of the respondents discuss its results with an experienced pathologist, who is a member of the MDT, see Figure 3.

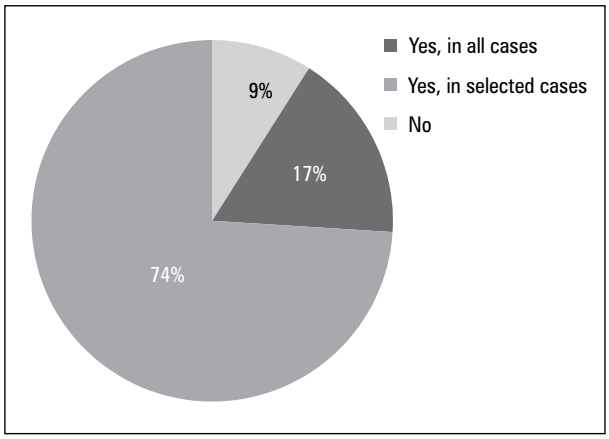

Figure 1. Distribution of answers to the question: Do you agree, that bronchoalveolar lavage (BAL) is necessary in the differential diagnosis of idiopathic pulmonary fibrosis (IPF)?

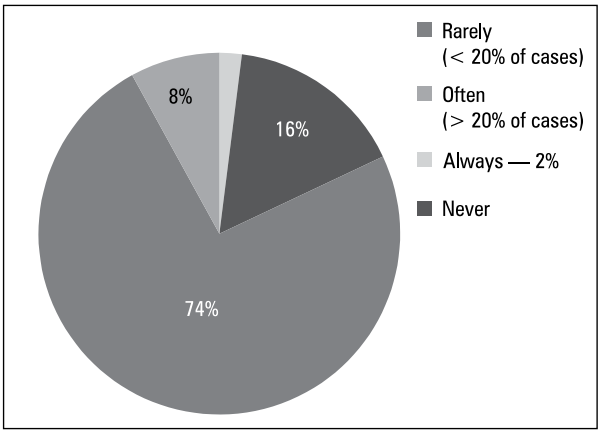

Figure 2. Distribution of answers to the question: How often do you refer patients with suspicion of IPF for surgical lung biopsy (SLB)?

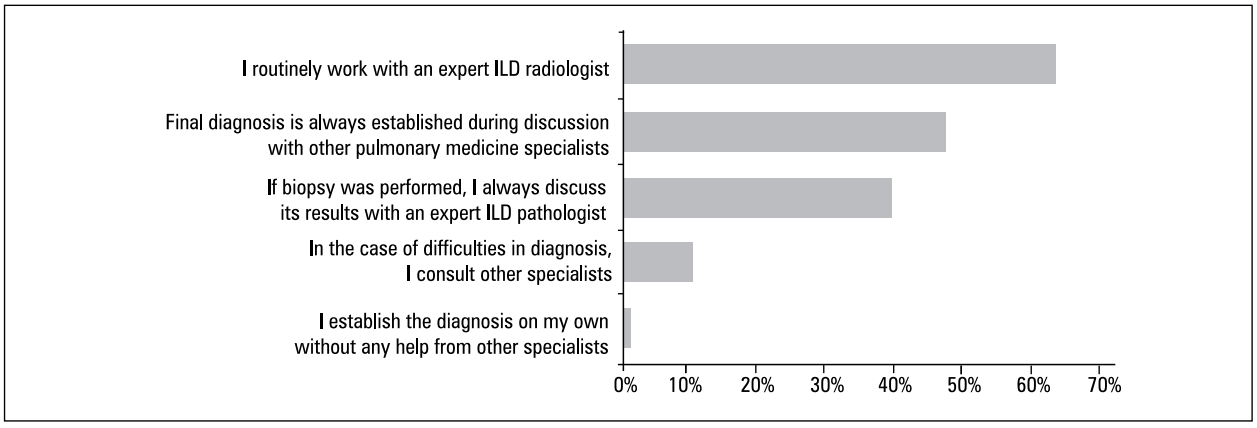

Figure 3. Distribution of answers to the question: Are you assisted by other specialists in the differential diagnosis of IPF? (multiple choice question)

Respiratory polygraphy or polysomnography examination is never recommended to IPF patients by 40% of healthcare professionals, is recommended rarely (< 20% of patients) by 51%, frequently (> 20% of patients) by 6%, and is recommended almost always (in > 80% of patients) by 3% of the respondents.

Treatment standards

Home oxygen therapy is recommended to all patients with IPF by 15% of participating physicians, only in case of exercise-induced hypoxemia by 13%, in case of resting hypoxemia by 71%, and is never recommended (because of lack of proof of its efficacy) by 1% of participants.

Ambulatory oxygen (from portable sources) is recommended to all IPF patients by 12%, to all IPF patients with exercise-induced hypoxemia by 46%, to all patients with resting hypoxemia by 31%, to all IPF patients with exercise-induced dyspnea by 5% of the respondents. Due to the lack of evidence for its efficacy, 6% of the respondents do not recommend ambulatory oxygen at all.

As far as rehabilitation is concerned, 45% of the respondents recommend different simple forms of activity to IPF patients, whereas 39% refer patients to professional pulmonary rehabilitation centers. As many as 6% of the respondents do not recommend rehabilitation due to the lack of evidence of its efficacy in this particular group of patients, and 10% do not do that due to the lack of access to the rehabilitation centers in the vicinity.

As many as 25% of the physicians refer all their IPF patients to lung transplantation centers, 59% of participants refer their patients when they notice a significant deterioration in lung function test results, 16% never refer patients to

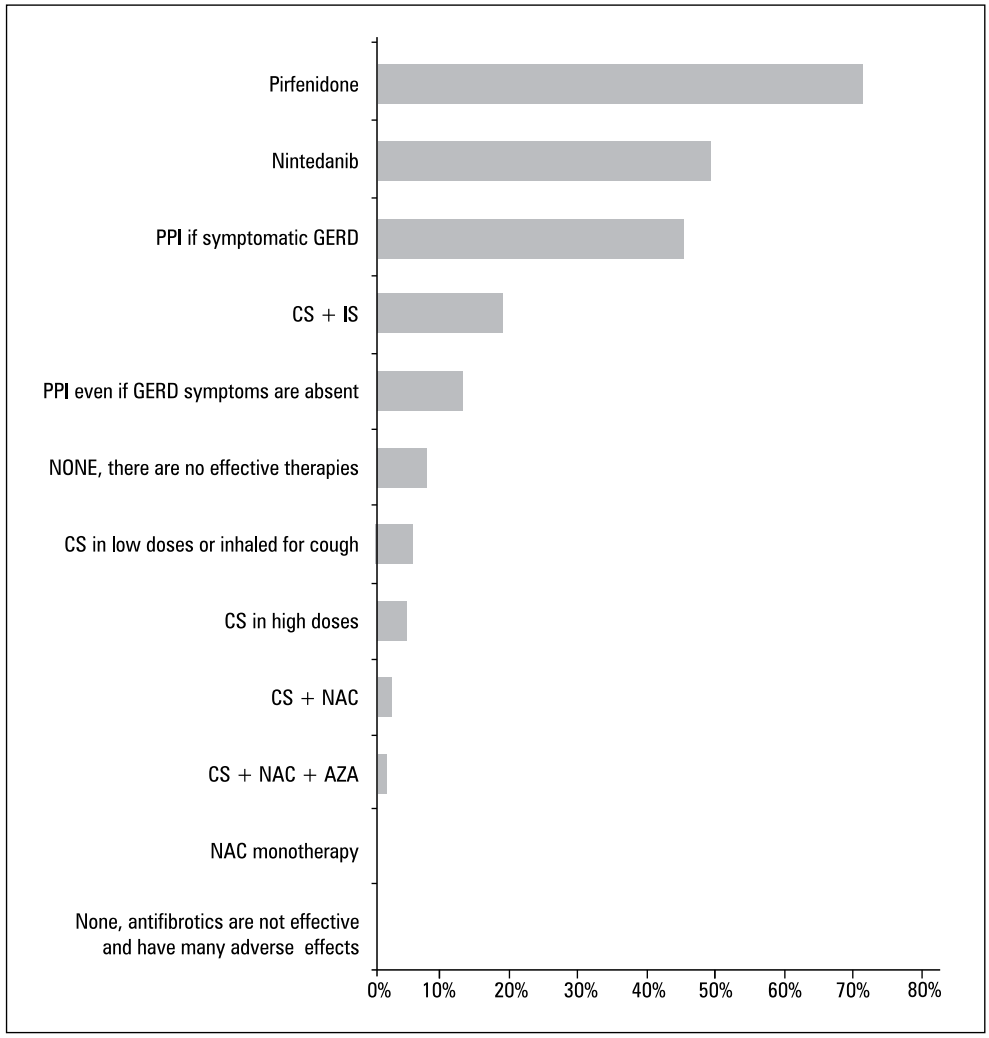

Figure 4. Distribution of answers to the question: What medications do you use in the treatment of idiopathic pulmonary fibrosis (IPF)? (multiple choice question). AZA — azathioprine; CS — corticosteroids; GERD — gastroesophageal reflux disease; IS — immunosuppressive drugs; NAC — N-acetylcysteine; PPI — proton pump inhibitors

lung transplantation centers due to lack of access to such centers.

A high dose of CS is still recommended by 4% of participants, and 5% of them prescribe low dose oral steroids or inhaled steroids for symptomatic treatment of cough in IPF. As many as 18% still recommend CS and IS. NAC in monotherapy or combination with CS or with CS and AZA is used by 3% of the respondents. Proton pump inhibitors (PPI) in the case of symptomatic GERD are prescribed by 44% of the respondents, and 12% prescribe PPI regardless of GERD symptoms. Pirfenidone is used by 70%, and nintedanib by 48% of the respondents. No treatment is recommended by 7%, and the explanation for this attitude is the lack of access to antifibrotic drugs, see Figure 4. None of the respondents declared, that antifibrotic drugs are not sufficiently effective or present unacceptable profiles of toxicity.

As many as 72% of the respondents declared having under their supervision at least 1 patient on pirfenidone therapy. In this subgroup, the mean number of IPF patients treated by a single healthcare professional with pirfenidone was 4.76 (7.96). About 38% of the respondents declared having under their supervision at least 1 patient on nintedanib therapy. In this subgroup, the mean number of IPF patients treated by a single healthcare professional with nintedanib was 1.53 (3.06).

According to 38% of the respondents < 50% of patients with IPF are eligible for the treatment based on the Polish National Health Fund (NHF) qualification criteria for antifibrotic therapy, 33% of the respondents estimate that between 50 to 80% of their IPF patients are eligible, 7% estimate that > 80% of patients are eligible. As many as 13% of the respondents are not familiar with the criteria of NHF antifibrotic therapeutic program for IPF patients, and 9% are not able to answer what percentage of their patients are eligible for antifibrotic therapy due to the lack of access to all mandatory testing required for patient's enrollment into the treatment program, see Figure 5.

The majority of respondents (64%) estimated, that < 50% of patients referred to NHF treatment program receive antifibrotic drugs without substantial delay (within 3 months since qualification), 13% presume that between 50 to 80% of patients receive drugs without delay, 7% estimate no delay in case of > 80% of patients, and 16% declare, that all their patients receive drugs without delay.

A 46% of surveyed physicians inform all their patients about the IPF Patients Association, which provides the support and education for patients with IPF and their family members, 19% of the respondents inform patients about the association only when they are asked. The minority of respondents (6%) are not convinced if patients need such association, and 29% did not realize, that such an association exists.

Discussion

The present real-world data survey on the clinical practice standards in IPF diagnosis and treatment in the era of access to the antifibrotic therapy for Polish patients with IPF is a follow-up to the previous survey on the same topic undertaken in 2016, before the availability of antifibrotics in Poland [7]. During two years in between both surveys, the situation has changed for Polish patients with IPF in regards to the access to effective pharmacological therapy. NHF reimbursement for antifibrotic drugs in Poland has led to the availability of pirfenidone from January

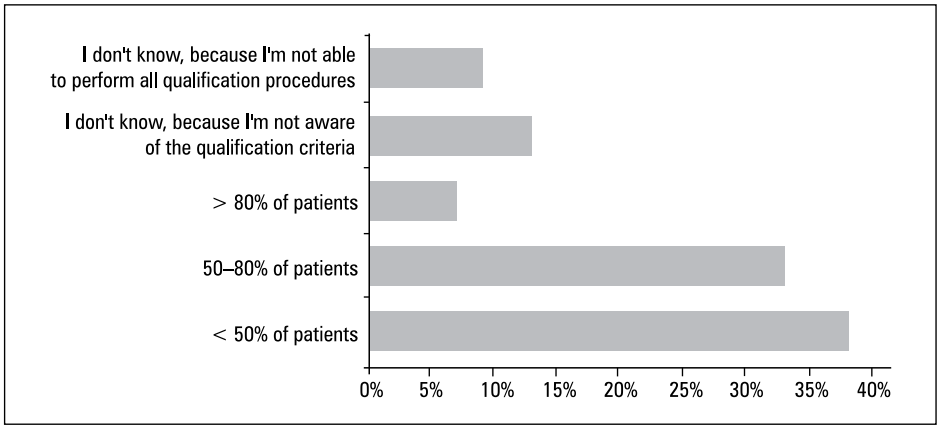

Figure 5. Distribution of answers to the question: What percentage of patients with IPF under your supervision qualify for the treatment with antifibrotics based on the Polish National Health Fund (NHF) qualification criteria?

2017 and nintedanib from March 2018 for the treatment of Polish patients with IPF. The present survey aimed to determine whether access to antifibrotic therapy together with growing awareness of the disease has influenced the diagnostic and treatment practices of the Polish healthcare professionals in the management of IPF.

A total of 93% of the respondents declared to be involved in the diagnosis and treatment of ILD routinely or sometimes which confirms that the survey's targeted population has been adequately chosen. The first part of our survey evaluating diagnostic standards confirmed that the clinical workup of Polish professionals for IPF diagnosis has only slightly changed over that time. Compared to the previous survey in which almost 30% of the respondents agreed that BAL is necessary in all cases in the differential diagnosis of IPF [7], in the actual one, only 17% of physicians agreed to the same opinion. The majority of respondents (74%) use BAL only in selected cases of patients. It is worth to mention, that although the 2011 international evidence-based guidelines for diagnosis and management of IPF do not recommend the use of BAL in the differential diagnosis [8], the update of this document from 2018 gives conditional recommendation for BAL in patients with other than typical UIP pattern in HRCT [1]. The majority of respondents in both surveys would recommend SLB in selected patients with a suspicion of IPF, which is in line with current conditional recommendation for SLB in patients with other than typical UIP radiologic patterns [1]. Our results point out that TBLC is not a common alternative for SLB in the diagnosis of IPF in Poland. More than half of the respondents refer patients with a suspicion of IPF for TBLC rarely and almost 40% never refer them for TBLC. TBLC represents a novel and exciting tool in the diagnostic pathway in IPF and offers a potentially safer, less-invasive and cheaper alternative to SLB, although less accurate [9]. The low use of TBLC in the diagnosis of IPF in Poland may be caused by less availability and experience with TBLC in our country. While the body of evidence regarding the use of TBLC in the differential diagnosis of IPF grows, the quality remains low. The majority of publications are retrospective single-center studies, which suffer from a significant risk of bias. This is a reason, that no recommendation either for or against TBLC in the diagnosis of IPF has been made in the updated international recommendations from 2018 [1]. More rigorous studies with adequate patient sample size are warranted to settle the issue of the diagnostic utility of TBLC in IPF.

According to the guidelines, the decision-making process of the IPF diagnosis should involve MDT, including a clinician, radiologist, and pathologist. The previous survey findings showed significant shortcomings regarding the involvement of MDT in the elaboration of IPF diagnosis [7]. This situation has not improved much in Poland over two years since the previous survey was undertaken. Only 8% more of the respondents dealing with IPF differential diagnosis (55% in 2016 and 63% in 2018) routinely work with and ILD expert radiologist and a similar percentage of clinicians (40% in 2016 and 39% in 2018) discuss the results of biopsy with an expert pathologist. MDT is critical for diagnosing cases of IPF and other ILD when the HRCT scans and clinical workup do not provide a clear diagnosis. Previously published data showed that a large percentage of patients experience long delays and are evaluated by three or more physicians before receiving the correct ILD diagnosis [10]. Early and accurate diagnosis of IPF through MDT discussion is essential to enable the initiation of therapies that have the potential to affect inevitable disease natural history and the avoidance of inappropriate and potentially harmful drugs [11]. Thus, the diagnostic process of IPF should be elaborated in the expert ILD centers, where the involvement of experienced MDT participants increases the diagnostic confidence [12]. It is of note, that more than 80% of the participants of the survey were hospital-based clinicians representing both academic and non-academic centers responsible for diagnosis and treatment of the broad spectrum of the respiratory conditions. Up to date, there is no reference center in Poland solely dedicated to ILD diagnosis and therapy. The present survey results indicate that more efforts are needed to improve the MDT approach to ILD diagnosis in Poland. In the authors personal opinion, formation of experienced MDT in Polish respiratory centers involved in ILD diagnosis should become an actual priority as long as IPF diagnostic standards are considered. This is especially important in the era of access to effective disease-modifying drugs.

The second part of our survey focused on treatment standards. A radical change has been noted in the pharmacological treatment of IPF patients in Poland in the meantime between surveys. Pirfenidone and nintedanib have become available in the therapeutic program refunded by NHF for patients with mild-to-moderate IPF. About one-third of the respondents of the first survey were not offering pharmacological therapy to patients with IPF because effective drugs

were not available [7]. In the present survey, the majority of the clinicians use antifibrotic drugs in the pharmacological treatment of IPF. Only 7% of participants believed that effective therapies are not available. More of the respondents declared experience with pirfenidone (70%) than nintedanib (48%) in the clinical practice, which is probably a consequence of the earlier reimbursement of the first one in Poland. It is of note, that 13% of the respondents were not familiar with the criteria of NHF antifibrotic therapeutic program for IPF patients in Poland and according to the majority of the respondents (64%), less than 50% of patients referred to NHF therapeutic program receive antifibrotics without substantial delay. These findings indicate that more attention and efforts are needed to minimize delays in accessing care by patients with IPF in Poland. In the previous survey undertaken in 2016 among Polish physicians taking care of patients with IPF, before the availability of antifibrotics, 24% of the respondents declared the use of CS or combination of CS and IS for the treatment of IPF [7]. Surprisingly, in the era of access to antifibrotic therapy since early 2017, still a total of 22% of the respondents, in the actual survey, declare use of high-dose CS in monotherapy or a combination of CS and IS for the treatment of IPF in 2018. Such pharmacological therapy is not recommended by international treatment guidelines in IPF released in 2015 due to the lack of benefit, significant toxicity, and adverse effects [4]. More educational initiatives are needed to change inappropriate practices in IPF therapy in Poland. It is likely, that Polish practical IPF guidelines, which are lacking, could improve diagnostic and treatment standards of IPF, providing Polish-language physicians a document in their language oriented towards clinical practice and decision-making process in IPF. Similar national initiative in France has led to important changes in the diagnosis and practical management of IPF by French pulmonologists [13]. Widespread awareness of national guidelines among French pulmonologists reduced the use of CS in monotherapy or combination therapy in IPF from 49% in 2011-2012 to 7% in 2014 [13, 14]. No change has been noted in the prescription habits in regards to AAT in IPF in the actual survey compared to the previous one [7]. PPI are used in the case of symptomatic GERD by 44% of the respondents and 12% use PPI regardless of GERD symptoms (43% and 11% in the previous survey, respectively). Although the potential role of gastroesophageal reflux (GER) in IPF pathogenesis and progression is not fully explained, it is suggested that AAT may decrease the risk for microaspiration-associated lung injury or damage, a mechanism that has been postulated to cause or worsen IPF [15]. Based on the encouraging retrospective clinical data, that PPI can stabilize lung function and reduce disease flares and hospitalizations, the international treatment guidelines recommend the use of AAT in IPF with the treatment indication being IPF and not GERD [4]. However, this recommendation carries very low confidence in effect estimates. On the contrary, recent studies not only question the relevance of the above mention retrospective findings but also associate the use of PPI with an increased risk of lung infections and a negative prognostic outcome [15]. Therefore, AAT prospective randomized trials in IPF are urgently needed.

As long as non-pharmacological therapy in IPF is concerned, the international guidelines recommend supplemental oxygen, pulmonary rehabilitation programs, and lung transplantation [4]. According to our results, non-pharmacological treatment practices have not changed substantially over two years between the surveys. The majority of respondents would recommend home oxygen therapy for IPF patients with resting hypoxemia (83% in 2016 and 71% in 2018) [7]. Although, guidelines recommend the use of supplemental oxygen in IPF patients with significant resting hypoxemia, specific criteria for oxygen indication in IPF are not established [4]. For the clinical practice in this matter criteria for the long-term oxygen therapy (LTOT) established for chronic obstructive pulmonary disease (COPD) patients based on two landmark studies conducted in the late 1970s, which demonstrated a survival benefit of LTOT in severe COPD associated with resting hypoxemia, are widely adopted [16, 17]. Although, no convincing data exist if similar to observed in COPD cohorts LTOT survival benefit translates to patients with IPF. Nevertheless, supplemental oxygen therapy may improve symptoms and exercise tolerance in IPF patients without resting hypoxemia [18]. The majority of respondents seem to know this relation and would recommend ambulatory oxygen (from portable sources) for the symptomatic therapy of patients with IPF.

Pulmonary rehabilitation programs are an evidence-based recommendation for the non-pharmacological treatment of patients with different chronic respiratory diseases, especially COPD, but also IPF [1]. The benefits of pulmonary rehabilitation in IPF include improved symptoms, especially dyspnea, increased exercise tolerance

and level of physical activity [19, 20]. Moreover, these benefits translate then to a lower level of anxiety and depression and therefore increased quality of life of IPF patients. Nevertheless, the majority of respondents of both the surveys would recommend only different simple forms of activity to IPF patients (74% in 2016 and 45% in 2018), and the minority of them would refer patients to professional pulmonary rehabilitation centers (21% in 2016 and 39% in 2018) [7]. Poorly developed network of pulmonary rehabilitation centers in Poland (only a few centers in the whole country) resulting in significant shortcomings to access to this type of non-pharmacological management of patients with IPF are probably responsible for the above clinical practices of Polish physicians. Pulmonary rehabilitation must be included in the integrative treatment of IPF, taking into consideration an inevitable disease course and only a modest response to new drugs. Better access to the rehabilitation programs is an unmet need in non-pharmacological therapy of patients with IPF in Poland.

Despite the advances in recent years in the treatment of IPF it continues to be a progressive disease with poor prognosis. In selected patients, lung transplantation may be a treatment option, with optimal results in survival and quality of life. Currently, pulmonary fibrosis is the main indication for lung transplantation [21]. However, mortality on the waiting list among these patients is high, since many patients are referred to the transplant centers with advanced disease. Therefore, all patients with IPF without contraindications should be referred to a transplant center early for evaluation. Based on our results, we noted a significant change in the approach to lung transplantation in IPF over two years in Poland. In 2016 almost one-third (31%) of the respondents would not refer patients for lung transplantation due to poor access to transplantation centers, whereas in 2018 only 16% of the respondents admitted to such practice. Better cooperation between pulmonary departments and transplantation units in Poland is crucial for optimal patients outcomes.

The results of the comparison of the present follow-up survey assessing possible changes and shortcomings in clinical practice standards in the management of IPF patients with a similar survey undertaken in 2016 must be considered in the context of obvious limitations. Both surveys were not exactly the same, and fewer physicians were surveyed in 2018 (99 *vs* 150). We did not control if participants of the second survey were participating in the first one, as both surveys were voluntary and anonymous. Nevertheless, we consider both surveys participants as a representative sample of motivated Polish physicians taking care of patients with IPF, and the findings commented above as a broad picture of changes that occurred between 2016 and 2018, not limited to change in the access to antifibrotic therapy.

Conclusions

Taken together, Polish physicians responsible for the management of patients with IPF are aware of the most important current international guidelines diagnostic recommendations. The level of cooperation between pulmonologists and other specialists to confirm the diagnosis of IPF, in the context of MDT work-up, is unsatisfactory. The treatment practices in the era of access to effective drugs in Poland require immediate improvement. There is an urgent need to develop national Polish practical guidelines for IPF which will offer Polish-speaking physicians and authorities an evidenced-based source document in their language oriented towards clinical practice and decision making for the best outcomes in the management of IPF.

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Conflicts of interest

Authors declare no conflicts of interest related to this research.

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