Turk Thorac J 2020; 21(1): 44-8 DOI: 10.5152/TurkThoracJ.2019.180181

Original Article

Survey of Physician's Knowledge, Attitudes, and Practices Regarding Idiopathic Pulmonary Fibrosis in Turkey

Haluk Türktaş[1], Gülfer Okumuş[2], Oğuz Uzun[3], Özlem Özdemir Kumbasar[4], Göksel Altınışık[5], Züleyha Bingöl[2], Şermin Börekçi[6], Vincent Cottin[7], Benan Müsellim[6]

[1]Department of Pulmonary Diseases, Gazi University School of Medicine, Ankara, Turkey
[2]Department of Pulmonary Diseases, İstanbul University İstanbul School of Medicine, İstanbul, Turkey
[3]Department of Pulmonary Diseases, Ondokuz Mayıs University School of Medicine, Samsun, Turkey
[4]Department of Pulmonary Diseases, Ankara University School of Medicine, Ankara, Turkey
[5]Department of Pulmonary Diseases, Pamukkale University School of Medicine, Denizli, Turkey
[6]Department of Pulmonary Diseases, İstanbul University-Cerrahpaşa, Cerrahpaşa School of Medicine, İstanbul, Turkey
[7]Claude Bernard University, Reference Center for Rare Pulmonary Diseases, UMR754, Lyon, France

Cite this article as: Türktaş H, Okumuş G, Uzun O, et al. Survey of Physician's Knowledge, Attitudes, and Practices Regarding Idiopathic Pulmonary Fibrosis in Turkey. Turk Thorac J 2020; 21(1): 44-8.

Abstract

OBJECTIVES: The aim of this study is to evaluate the approaches of Turkish pulmonologists to the diagnosis and treatment of idiopathic pulmonary fibrosis (IPF) in daily clinical practice.

MATERIALS AND METHODS: A questionnaire containing 38 questions about the IPF diagnosis and treatment was given to pulmonologists between January 22 and 29, 2018, and the data of 158 physicians who responded to the questionnaire were evaluated.

RESULTS: This survey showed that the mean number of patients that physicians followed up and managed annually was 8.3 and 5, respectively. The mean symptom duration before the diagnosis was 9–12 months. Patients were seen on average by three physicians prior to confirmed diagnosis. Almost 80% of the physicians have an opportunity to access a pathologist and radiologist specialized in IPF. However, only 26% of them have an opportunity to access regular multidisciplinary meetings. Although antifibrotics were the most commonly prescribed drugs, approximately 10% of patients were prescribed steroids, N-acetylcysteine, and immunosuppressants. Most of the physicians (81%) were aware of international guidelines; however, the *Turkish Thoracic Society IPF Diagnosis and Treatment Consensus Report* was read by only 41% of them.

CONCLUSION: This survey may lead to the IPF awareness in Turkey, and it may help to close the gaps regarding the diagnosis and treatment.

KEYWORDS: Idiopathic pulmonary fibrosis, survey, Turkey

Received: *12.11.2018* ***Accepted:*** *26.12.2018*

INTRODUCTION

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive disease of the lungs with irreversible fibrosis. The pathogenetic mechanisms have not been fully explained [1,2]. There is a rapid loss of the respiratory function and unpredictable prognosis. The average life expectancy of the patients is 3-5 years. There are serious problems in diagnosis and treatment of IPF reported.

IPF is an orphan disease with an estimated incidence in the United States between 6.8 and 8.8/100.000 [3,4], and in the United Kingdom, 2.85/100.000 [5]. In Turkey, there are no data about the incidence and prevalence of IPF, but the estimated incidence for all interstitial lung diseases is 25.8/100.000 [6].

In 2011, an international consensus report (by ATS/ERS/JTS/ALAT) and, in 2018, a national consensus report by the Turkish Thoracic Society were published to standardize the diagnosis and treatment of IPF [7,8]. According to these guidelines, high resolution computed tomography (HRCT) plays a key role in the diagnosis of IPF. In HRCT, the diagnosis of IPF is made through the existence of the usual interstitial pneumonia (UIP) pattern and after the exclusion of known causes of interstitial lung disease. Surgical lung biopsy is recommended in patients with a possible UIP pattern in HRCT. It is emphasized that multidisciplinary council meetings where clinical, radiological, and pathological findings are discussed are very important in the diagnosis of IPF.

Studies about the treatment of IPF have shown that steroids, N-acetylcysteine, anticoagulant, and immunosuppressive drugs have no place in the treatment. Currently, antifibrotic drugs such as pirfenidone and nintedanib are recommended for the treatment of IPF [7,9,10].

MATERIALS AND METHODS

This study was planned and performed by the Turkish Thoracic Society Clinical Problems Study Group. This is a survey study that included physicians. All the physicians voluntarily gave their informed consent and answered the question-

Address for Correspondence: Haluk Türktaş, Department of Pulmonary Diseases, Gazi University School of Medicine, Ankara, Turkey
E-mail: halukturktas@gmail.com

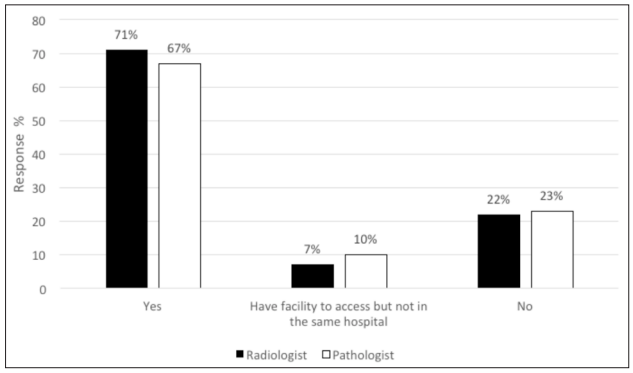

Figure 1. Proportion of physicians with access to radiologist and pathologist

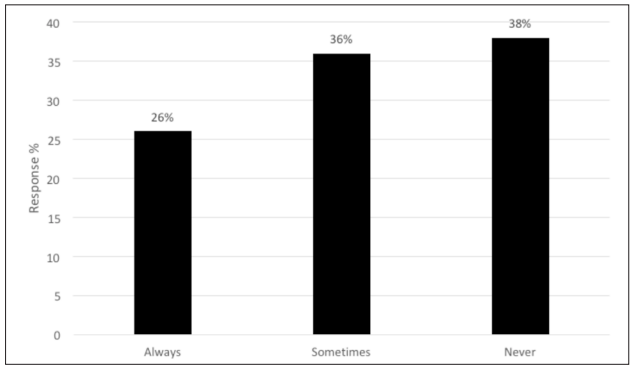

Figure 2. Proportion of physicians with access to multidisciplinary discussion

naire sent by e-mail. The study was carried out according to the principles of the Declaration of Helsinki. The Advancing IPF Research Survey questionnaire used in European countries was applied. It consisted of 28 questions [11]. We added several questions to assess the approaches and attitudes of pulmonologists about IPF. The questionnaire consisted of 38 questions relating to the diagnosis and treatment of IPF. The questions were about the approaches of physicians in daily clinical practice for the diagnosis and treatment of IPF. The questionnaire was sent by e-mail to the chest physicians listed in the Turkish Thoracic Society database. Physicians were given the week between January 22th and 29th, 2018, to complete the questionnaire. The responses of the physicians who completed the questionnaire were analyzed.

RESULTS

Characteristics of Respondents

At the end of the determined period, 247 physicians completed the questionnaire. The responses of 158 pulmonologists who followed at least 1 IPF patient were evaluated. More than half of the physicians participating in the survey were women (n=87, 55%), and 71 (45%) were men, while 112 (71%) of them were aged >40 years. Most of the physicians were working in academic centers (n=124, 80%), and 34 physicians (20%) were working in non-academic hospitals.

The survey responses showed that the total number of IPF patients followed by respondents was about 940. The number of patients followed by each physician ranged from 2 to 200 (mean 8.3). The total number of patients who received treatment in the last year was 530, ranging 1 to 50 per physician. The average number of IPF patients treated by a physician per year was 5 (Table 1).

Table 1. Characteristics of the physicians participating in the survey

Questions	Responses	n	%
Gender	Female	87	55
	Male	71	45
Age	<30	1	1
	31-40	45	28
	41-50	62	39
	51-60	44	28
	>60	6	4
Institution	Public Hospital	17	10
	MH Academic Hospital	44	29
	University Hospital	80	51
	Private Hospital	17	10
Title	Specialist	57	36
	Assistant professor	13	8
	Associate professor	35	22
	Professor	53	34
Number of IPF patients treated by a physician in one year	<5	48	41
	6-20	59	51
	21-50	9	8
	>51	0	0

MH: Ministry of Health

Diagnosis of IPF

Approximately two-thirds of the survey respondents had access to a radiologist (71%) and a pathologist (67%) at their institution, but one-third did not have this opportunity (Figure 1).

A multidisciplinary team (MDT) discussion including a pulmonologist, radiologist, and pathologist is crucial for the diagnosis of IPF. However, 26% of the respondents had regular, 36% had irregular, and 38% have no MDT meeting (Figure 2).

The average symptom duration was 9-12 months before the diagnosis, and patients were seen on average by three physi-

MAIN POINTS

- This study shows the approaches of Turkish pulmonologists regarding the diagnosis and treatment of IPF in daily clinical practice.
- Although approximately 71% of the Turkish pulmonologist have an opportunity to access a radiologist, and 67% to access a pathologist only 26% of them have a regular multidisciplinary meeting discussion.
- The most common initiated drugs for the treatment of IPF are antifibrotics.

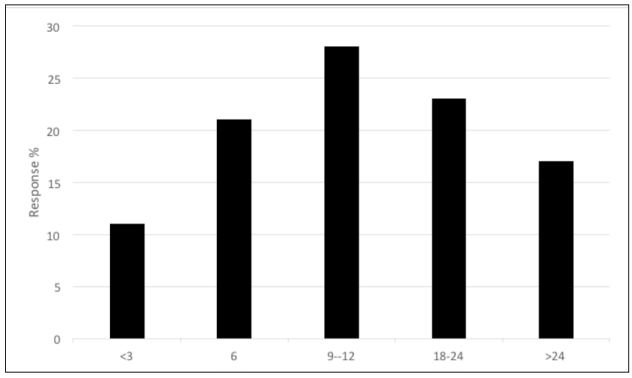

Figure 3. Average symptom duration (months) before diagnosis of IPF

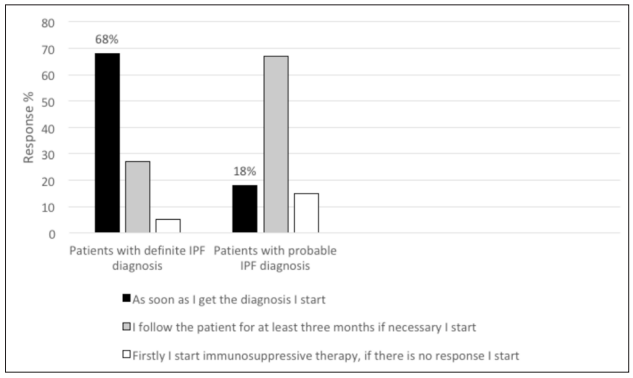

Figure 5. Percentage of physicians who prescribe antifibrotic drugs for patients with definite and probable IPF

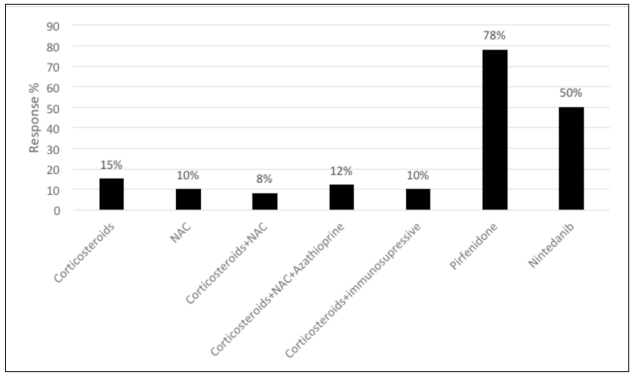

Figure 4. Percentage of physicians who prescribe treatment for IPF

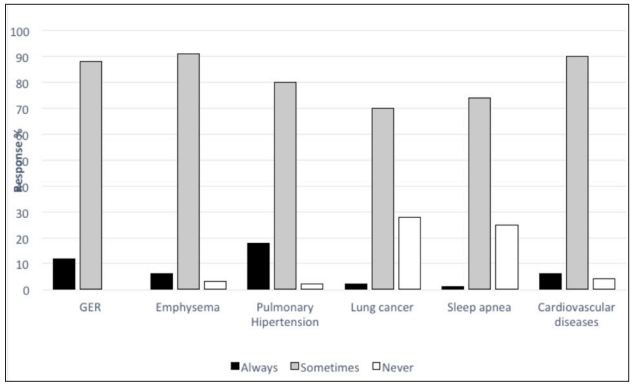

Figure 6. The most common comorbidities in IPF patients

cians before the confirmed diagnosis of IPF. One-third of the patients was seen by four or more physicians (Figure 3).

At the diagnosis of IPF, almost all the physicians explored the occupational risk factors, and 82% of them asked whether there was a family history of fibrotic interstitial lung disease. Genetic tests were performed by 20% of the physicians if there was a family history.

Only 15% of the physicians always performed the bronchoalveolar lavage (BAL) examination in the diagnosis of IPF, and 85% of them performed BAL in only selected patients such as younger non-smokers, patients with a pre-diagnosis of hypersensitivity pneumonia, and patients with a possible UIP pattern in HRCT. The ratio of physicians who always recommend a surgical lung biopsy for the diagnosis of IPF was 22%. Surgical biopsy was usually recommended in younger patients.

The ATS/ERS/JRS/ALAT IPF guidelines were considered useful by 81% of physicians, while 16% stated that they did not read the guidelines in detail. Only 41% of the physicians read the *Turkish Thoracic Society IPF Consensus Report*, while 46% of them were aware of the report but have not read it yet, and 13% have not heard about it.

IPF Treatment

The IPF treatment decision was made on their own by 27% respondents, through a discussion with other pulmonologist by 31%, and multidisciplinary team discussion by 27%. The most commonly initiated medications were pirfenidone and nintedanib.

Corticosteroids, NAC, and immunosuppressive drugs are used alone or in combination in 10%-15% of patients (Figure 4).

For patients with the diagnosis of definite IPF; 68% of the physicians started the antifibrotic drugs early, and 27% of them followed the progression of the disease for at least 3 months and decided to treat according to the progression of the disease. For the patients diagnosed with possible IPF, 18% of the physicians started the treatment early, and 67% followed the patients for at least 3 months and started the treatment according to the progression of the disease (Figure 5).

Approximately 80% of physicians assess the progression of the disease by the FVC, DLCO, and 6-minute walking test. Lung transplantation was performed in 17 of the patients followed up by the physicians in this survey.

When the IPF treatment is considered, 75% of physicians give priority to early diagnosis, 70% to effectiveness of the treatment and monitoring of side effects, 61% to the follow-up plan, 59% to reaching a definitive diagnosis, and 57% to treatment of comorbidities.

Comorbidities

Comorbidities are very common in IPF patients. The most common comorbidities were pulmonary hypertension, gastroesophageal reflux (GER), emphysema, cardiovascular diseases, and lung cancer (Figure 6).

In patients diagnosed with IPF, 83% of the physicians always investigate the symptoms of GER. Twenty-six percent of the respondents routinely treat GER, regardless whether the

patients have GER symptoms or not, but 73% only treat patients with symptomatic or documented GER.

In patients with pulmonary hypertension, 11% of the physicians regularly treat pulmonary hypertension in patients with IPF, 41% treat it occasionally, and 48% never prescribe any specific treatment for pulmonary hypertension.

DISCUSSION

To the best of our knowledge, this is the first survey to assess the attitudes and current approaches of Turkish pulmonologists about the IPF diagnosis and management. With this survey, we had the opportunity to compare our results with other international surveys.

Seventy-nine percent of respondents were working in university hospitals or in education hospitals of the Ministry of Health. Because of the difficulties in the diagnosis and management of IPF, the majority of the patients was followed in tertiary care settings, and this may explain the higher rate of respondents from academic centers.

The mean number of patients treated annually by each physician was 5. Forty-one percent of physicians reported that they treated <5 patients per year, and 51% of them treated between 5 and 20 patients. Similarly, in a Polish survey with 150 participants, 52% of the physicians managed <5 IPF patients per year, and 9% of them managed >20 patients [12]. In a Latin American survey, the average number of IPF patients managed previous year was 13.8 [13]. In a French survey, the mean number of patients managed annually was 56 [14], and in a European survey, it was 39 [11]. The reason for this higher number of patients in France and Europe may be explained by the fact that the majority of the respondents in the French and European survey was from IPF centers.

To examine the awareness of participants about the national and international guidelines, our survey had several questions. Most of the respondents (81%) declared that they have read the ATS/ERS/JRS/ALAT IPF guidelines and considered it useful for the management of IPF. In contrast, only 41% have read and another 46% were aware of have not read the Turkish Thoracic Society IPF Consensus Report, and 13% have not heard about the report. In our opinion, the reason for this was that the survey was conducted just a few months after the publication of the national consensus report, so there was a very short period between the publication of the national consensus report and the survey. We believe that future studies will reflect the actual usage of the national report more accurately.

At diagnosis, 96% of physicians explored lung fibrosis in families of IPF patients and performed genetic tests in 20% of those patients who had a family history of fibrotic interstitial lung disease. In the European survey, the rate of exploring the existence of fibrotic disease in the family was 94%, and 29% of physicians performed genetic tests [11]. This rate was 77% in the French study [14]. Although there were no recommendations for genetic testing in the ATS/ERS/JRS/ALAT IPF guidelines, such high rates are surprising [15].

A patient who presented with symptoms compatible with IPF had a period of approximately 1 year before the confirmed diagnosis of IPF, and every patient was seen by three physicians before the diagnosis. Similarly, in the IPF survey conducted in Europe, it is stated that there is an average delay of 1 year for the diagnosis, and the patients were seen by two or more physicians prior to the diagnosis of IPF.

In Turkey, approximately 71% of the respondents have an opportunity to access a radiologist, and 67% to access a pathologist in their institution. The access rates to the radiologist and pathologist in Latin America are 39% and 28%, in France 63% and 66%, and in Europe 85% and 74%, respectively [11,13,14]. Although there is a higher availability and easy accessibility to radiologist and pathologist in our country, we have two main problems in the diagnosis of IPF. First, we have fewer numbers of experienced radiologist and pathologist in IPF, and second, there is the inability to gather an MDT. This survey results show us that only 26% of participating institutions have a regular multidisciplinary meeting discussion. Nearly one-third (36%) have irregular meetings, and 38% have no MDT meeting in their hospitals. In French and European surveys, the vast majority of IPF cases were diagnosed on a multidisciplinary discussion. Only 7% of European responders and 3% of French pulmonologists reported that the diagnosis of IPF was made without multidisciplinary discussion [11,14]. In a Latin American survey, the access to multidisciplinary team was 41.1% among pulmonologist [13].

In Turkey, the IPF treatment decision was made alone by 27% of the physicians, 31% by consulting with other chest diseases specialists, and 27% by the multidisciplinary discussion. The rate of physicians who stated that they gave treatment decisions alone in Europe was 7%, and in Latin America, it was 20%. The treatment decision should be made through multidisciplinary council discussions, but the rate of decision making alone by a physician is quite high.

The most common initiated drugs for the treatment of IPF are antifibrotics. Most of the Turkish physicians (78%) reported that they prescribe pirfenidone as an antifibrotic, and 50% reported nintedanib. The prescription rate of antifibrotic drugs was 81% in Europe, 60% in Latin America, and 11% in Poland [11-13]. The low rate of prescription in Poland can be explained by the late reimbursement of these drugs for IPF. Corticosteroid therapy in monotherapy or in different combinations was recommended by 37% of respondents. Similarly, 43%, 57%, and 83% of the physicians from Poland, Europe, and Latin America, respectively, prescribe corticosteroids alone or in a combination for the treatment of IPF [11-13]. As mentioned in the European survey report, the possible reasons for prescription of non-antifibrotic drugs may be that in clinical practice, such therapies demonstrate a benefit in some patients with conditions similar to IPF, or that the patients themselves may be reluctant to stop certain medications. Fifteen percent of responders tried immunosuppressive therapies before starting antifibrotic drugs in patients with possible IPF.

An early initiation of antifibrotic drugs for the treatment of IPF is recommended [16]. From the point of view of initiation time of antifibrotic drugs, 68% of physicians start the treat-

ment upon diagnosis irrespective of symptoms in patients with the diagnosis of definite IPF, and 27% of the responders follow the patient for at least 3 months to evaluate the progression of the disease and start treatment if progression occurs. In patients with the diagnosis of possible IPF, 18% of physicians initiate treatment early, and 67% start after the 3-month follow-up.

The most commonly reported comorbidities in patients with IPF are emphysema, GER, cardiovascular diseases, and pulmonary hypertension. Most of physicians (83%) always ask patients if they have GER symptoms. Twenty-six percent of the physicians routinely treat GER irrespective of symptoms, but 73% of them treat only patients with symptoms. Regarding pulmonary hypertension, 11% of the physicians regularly treat pulmonary hypertension. In the European survey, this rate is 4%, and in the Latin America survey, it is 50% [11,13]. Although there is no recommendation for the treatment of pulmonary hypertension in the guidelines, it is also interesting that the rates of treatment are as high as 50%, and there are significant differences between countries.

In summary, the results of this survey provide a snapshot showing the approaches of Turkish pulmonologists regarding the diagnosis and treatment of IPF in daily clinical practice. The majority of Turkish pulmonologists is aware of the international guidelines and recommendations. Despite this awareness and despite having a new national consensus report, there are gaps in the diagnosis and treatment of IPF.

Ethics Committee Approval: Authors declared that the research was conducted according to the principles of the World Medical Association Declaration of Helsinki "Ethical Principles for Medical Research Involving Human Subjects", (amended in October 2013).

Informed Consent: This study is a questionnaire study. For this reason, informed consent was not obtained.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - H.T.; Design - H.T.; Supervision - H.T., G.O., O.U., Ö.Ö.K., G.A., V.C., B.M.; Materials - H.T., G.O., O.U., Ö.Ö.K., G.A., Z.B., Ş.B., B.M.; Data Collection and/or Processing - H.T., G.O., O.U., Ö.Ö.K., G.A., Z.B., Ş.B., B.M.; Analysis and/or Interpretation - H.T., V.C.; Literature Search - H.T., V.C.; Writing Manuscript - H.T., Ş.B., Z.B., V.C.; Critical Review - H.T., G.O., O.U., Ö.Ö.K., G.A., Z.B., Ş.B., B.M., V.C.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Sgalla G, Lovene B, Calvello M, et al. Idiopathic Pulmonary Fibrosis: pathogenesis and management. Respir Res 2018;19:32. [CrossRef]
2. Walters PJ, Blackwell TS, Eickelberg O, et al. Time for change: is idiopathic pulmonary fibrosis still idiopathic and only fibrotic? Lancet Respir Med 2018;6:154-60. [CrossRef]
3. Nalysnyk L, Cid-Ruzafa J, Rotella P, et al. Incidence and prevalence of idiopathic pulmonary fibrosis: review of the literature. Eur Respir Rev 2012;21:355-61. [CrossRef]
4. Raghu G, Chen SY, Hou Q, et al. Incidence and prevalence of idiopathic pulmonary fibrosis in US adults 18-64 years old. Eur Respir J 2016;48:179-86. [CrossRef]
5. Strongmen H, Kausar I, Maher TB. Incidence, prevalence and survival of patients with idiopathic pulmonary fibrosis in the UK. Adv Ther 2018;35:724-36. [CrossRef]
6. Musellim B, Okumus G, Uzaslan E, et al. Epidemiology and distribution of interstitial lung diseases in Turkey. Clin Respir J 2014;8:55-62. [CrossRef]
7. Raghu G, Collard HR, Egan JJ, et al. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. Am J Respir Crit Care Med 2011;183:788-824. [CrossRef]
8. Ergur GA, Bingöl Z, Kumbasar ÖÖ, et al. Türk Toraks Derneği idiyopatik pulmoner fibrosis tanı ve tedavi uzlaşı raporu 2018. Türk Toraks Derneği yayınları 2018.
9. The idiopathic pulmonary fibrosis clinical research network, Raghu G, Anstrom KJ, et al. Prednisone, Azathioprine, and N-Acetylcsyteine for pulmonary fibrosis. N Engl J Med 2012;366;1968-77. [CrossRef]
10. Noth I, Anstrım KJ, Calvert SB, et al. A placebo controlled randomized trial of warfarin in idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2012;186:88-95. [CrossRef]
11. Cottin V. Current approaches to the diagnosis and treatment of idiopathic pulmonary fibrosis in Europe: AIR survey. Eur Respir Rev 2014;23:225-30. [CrossRef]
12. Piotrowski WJ, Martusewicz-Boros MM, Bialas AJ, et al. Idiopathic pulmonary fibrosis-common practice in Poland before the "antifibrotic era". Adv Respir Med 2017;85:136-42. [CrossRef]
13. Cherrez-Ojeda I, Cottin V, Calderon JC. Management and attidudes about IPF (Idiopathic Pulmonary Fibrosis) among physicians from Latin America. BMC Pulm Med 2018;18:5. [CrossRef]
14. Cottin V, Cadranel J, Crestani B, et al. Management of idiopathic pulmonary fibrosis in France: A survey of 1244 pulmonologists Respir Med 2014:108;195-202. [CrossRef]
15. Raghu G, Remy-Jardin M, Myers JL, et al. Diagnosis of idiopathic pulmonary fibrosis. An official ATS/ERS/JRS/ALAT clinical practice guideline. Am J Respir Crit Care Med 2018;198:e44-e68.
16. Torrisi SE, Pavone M, Vancheri A, et al. When to start and when to stop antifibrotic therapies. Eur Respir Rev 2017:26;170053. [CrossRef]