Common Drug Review
Patient Group Input Submissions

nintedanib (Ofev) for Idiopathic pulmonary fibrosis (IPF)

Patient group input submissions were received from the following patient groups. Those with permission to post are included in this document.
British Columbia Lung Association & Lung Groups — permission granted to post.
Canadian Pulmonary Fibrosis Foundation — permission granted to post.
Lung Association of Saskatchewan — permission granted to post.
Ontario Lung Association — permission granted to post.

CADTH received patient group input for this review on or before May 4, 2015

Disclaimer: The views expressed in each submission are those of the submitting organization or individual; not necessarily the views of CADTH or of other organizations.

While CADTH formats the patient input submissions for posting, it does not edit the content of the submissions.

CADTH does use reasonable care to prevent disclosure of personal information in posted material; however, it is ultimately the submitter's responsibility to ensure no personal information is included in the submission. The name of the submitting patient group and all conflict of interest information are included in the posted patient group submission; however, the name of the author, including the name of an individual patient or caregiver submitting the patient input, are not posted.

British Columbia Lung Association & Lung Groups

Section 1 — General Information

Name of the drug CADTH is reviewing and indication(s) of interest		OFEV (nintedanib)
Name of the patient group		British Columbia Lung Association & Lung Groups
Name of the primary contact for this submission:		[redacted]
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Permission is granted to post this submission		Yes

1.1 Submitting Organization

The Mission of the British Columbia Lung Association (BCLA) is to improve lung health and to lead lung health initiatives. Our vision is healthy lungs for everyone. Our role is to improve respiratory health and overall quality of life through programs, education, research, training, treatment, advocacy and prevention of lung disease.

The BCLA is a major Canadian charitable organization with more than a century of experience and leadership in lung disease prevention, treatment and management. Today our areas of interest and expertise include the entire scope of respiratory diseases including Idiopathic Pulmonary Fibrosis, COPD (chronic bronchitis and emphysema), asthma, lung cancer, sleep apnea and tuberculosis. We work together with the Canadian Lung Association and other partners to help the one in five Canadian who have breathing problems.

Our staff and volunteers include health professionals and interested individuals and patients with a broad range of training and experience in lung disease and lung health that enables our organization to develop and lead programs of education and health promotion at the highest standard. The British Columbia Lung Association provides approximately $1.2 million each year to internationally recognized physicians and scientist doing research in BC on lung diseases. All funding proposals go through rigorous national peer review system so that the most promising research can be explored. This world class research is discovering the causes of lung disease, finding new treatments, and giving hope for a future free of lung disease.

1.2 Conflict of Interest Declarations

The British Columbia Lung Association has several sources of funding for programs and operations and is supported by individual and corporate donations, and through service contracts with government organizations. Funding sources include direct mail campaigns such as the Christmas Seals campaign, memorial giving, bequests, Special events such as Climb the Wall: Stair Climb for the fight against lung disease!, Bicycle Trek for life and breath and our RUSH event. The Lung Association, does, from time to time receive program grants from health industry/pharmaceutical companies. Our relations and interactions with pharmaceutical companies remain transparent and positions of the Lung Association are developed without industry influence.

The BCLA has received health educator's program grants from the following pharmaceutical companies: Grifols, GlaxoSmithKline, InterMune, Astra Zeneca, Boehringer Ingelheim, Pfizer, Novartis and Merck Frosst

a) We have the following declaration(s) of conflict of interest in respect of corporate members and joint working, sponsorship, or funding arrangements:

b) We have the following declaration(s) of conflict of interest in respect of those playing a significant role in compiling this submission:

Neither the principal author, nor the BCLA, has conflicts to declare in respect to the compiling of this submission

Section 2 — Condition and Current Therapy Information

2.1 Information Gathering

The BCLA is significantly invested and involved in IPF and other respiratory disease research and provision of patient services and programs. We have Certified Respiratory Educators on staff who provide expert educational consultations to respiratory patients, their family members and caregivers dealing with IPF and other lung diseases. The vast knowledge and experience garnered through research, best practice guidelines and direct involvement with patients is the basis of the information provided.

2.2 Impact of Condition on Patients

Pulmonary Fibrosis is an interstitial lung disease that has a devastating impact on the patient. It is a debilitating and fatal disease. After diagnosis many patients with Interstitial Pulmonary Fibrosis (IPF) are in steady decline experiencing breathing difficulties (shortness of breath or dyspnea), chronic cough and tremendous fatigue. About 85%of people with IPF have a chronic cough that last longer than 8 weeks; some people may cough up sputum and phlegm. Sadly IPF has a very high mortality rate: Approximately 5,000 individuals will die from IPF each year.

Pulmonary Fibrosis affects each person differently and progresses at varying rates. Generally, the individual's respiratory symptoms become worse over time. Breathlessness is a key symptom and complaints of IPF patients with rapid decline in lung function leading to fatigue and exercise intolerance. Breathlessness can affect day-to day activities such as showering, climbing stairs, getting dressed and eating. As scarring in the lungs gets worse, breathlessness may prevent all activities. The physical deterioration of the individual with IPF is profound and commonly emotionally demanding. The goal of available therapy to date has been to prolong life, reduce disability and stabilize lung function and slow disease progression to allow physical and social functioning to the highest level possible.

IPF sufferers will often require assistance and become increasingly dependent on others to the most basic human task of daily living activities. Depression and feelings of hopelessness are common among patients with IPF.

Lung attacks or flare-ups drive disease progression. As the disease progresses frequency of flare-ups increase, overall lung function and lung health typically decline and risk of hospital admission increases as well as rate of mortality.

2.3 Patients' Experiences With Current Therapy

The therapies used and recommended by the Canadian Thoracic Society

There is no known cure for IPF, The other therapies used in IPF include pulmonary rehabilitation programs that may ease some of the symptoms include Oxygen therapy which helps to reduce breathlessness, Corticosteroid pills to reduce swelling in the lungs by suppressing the immune system, pulmonary rehabilitation program to teach breathing techniques and exercises for people with IPF and Lung transplantation for patients below age 65.

What is current standard of care for IPF patient?

Esbriet (pirfenidone) is the first and only medication indicated for use in Canada for the treatment of adults with mild to moderate idiopathic pulmonary fibrosis (IPF) Health Canada approved Esbriet on October 1, 2012 following a priority review granted due to the severity of the disease and significant unmet medical need. Esbriet is an oral tablet with systematic anti-fibrotic and anti-inflammatory properties and has been shown to slow the progression of scarring (fibrosis) and has been shown to be effective in decreasing decline in lung function in some patients. It has also been shown that it does not work with all patients with IPF.

Since Esbriet is not yet funded by provincial/territorial drug program, patients can only access the treatment through private insurance or personal financial means. Today, Esbriet remains out of reach for many patients especially seniors who no longer have private coverage and rely strictly on government funding for access.

Unmet Needs: Of critical importance to the treatment of IPF are medicines that will help reduce or stop the progression of the disease and subsequent hospitalizations. Additional therapies are needed that go beyond symptomatic relief. New treatments are urgently needed that will work to improve overall lung function. New treatment options are required as the disease progresses.

The BCLA believes that access to IPF medications will serve to reduce cost on admissions to hospital and improve the overall lung health of patients with IPF. The BCLA support the quick access to respiratory medications such as that for IPF patients and recommended by the Canadian Thoracic Society

We recognize that not all patients or individuals respond the same to various types of formulations of medications and BCLA support having access to the medications to which a particular patient responds better.

We also recognize that not all patients are able to use inhalation devices effectively and we also support having access to the inhalation devices which the patient are able to use correctly. Delivery of the medication effectively is important in the treatment of IPF. The new medication is available in oral preparation.

Unmet Needs: Medications are of critical importance in the treatment and management of IPF. It improves lung function and breathing, reduce lung attacks and prevent patients with repeat admission to hospital there by improving the lives of IPF patients.

2.4 Impact on Caregivers

Our health care system places a lot of demands on both the patient and caregivers. Caregivers are often the spouse, the children and other relations. Financial challenges are the obvious ones, depending on the level of reimbursement for medicine.

Another major impact identified by patients and care givers is physical activity. The impact is most noticeable on patients' progressive inability to perform day to day tasks as they begin to notice that they had previously taken for granted (e.g. negotiating a staircase that they climb every day)

As the patient's condition deteriorates, they tend to stay at home more which means that their fitness levels further deteriorate and their body's ability to use oxygen efficiently is further compromised. As the condition progresses, further compromises are made in patient's independence with huge implications for caregivers. Patients with IPF and their caregivers experience anxiety and depression. This disease has a progressive debilitating course and sadly it increases mortality.

Caring for someone with IPF can be both physically and emotionally demanding. Caregivers may experience a great deal of stress and anxiety, resulting from their loved one's deterioration. Frequently these feelings have a negative impact on the caregiver's health and well- being. Frequent visits to medical professionals, increasing medical needs, restrictions in activities leading to the caregiver taking a larger role may impact the caregiver significantly. The BCLA sponsor a number of support groups called "Better Breather's Group" for individuals with Lung Conditions and their caregivers and helps the caregiver cope more effectively.

Section 3 — Information about the Drug Being Reviewed

3.1 Information Gathering

The BCLA is significantly invested and involved in IPF and other respiratory research and provision of patient's services and programs. We have Certified Respiratory Educators on staff who provide educational expert consultations to respiratory patients with IPF, their family members and caregivers. The vast knowledge and experience garnered through research, best practice guidelines and direct involvement with patients is the basis of the information.

3.2 What Are the Expectations for the New Drug or What Experiences Have Patients Had With the New Drug?

Nintedanib (OFEV) is an anti-scarring(anti-fibrotic) medication that slows progression of IPF. This new medication for IPF is not yet available in Canada. The British Columbia Lung Association on behalf of our lung patients with the deadly disease IPF who do not respond on Esbriet, we urge CDR at CADATH for the easy access and approval of Nintedanib (OFEV) the second medication for IPF. It is also imperative to make easy access to our IPF patients in the Provincial/Teritorial drug formularies.

We support access to those medications recommended by the CTS.

There have been clinical trials on (Nintedanib) OFEV in several pulmonary research clinics in BC.

Results included Reduction in annual rate of lung function decline (FVC) Reduction in absolute change from baseline in FVC, time to first acute exacerbation(flare up) and change from baseline in health-related quality of life as assessed by St. George's Respiratory

Questionnaire (SGRQ)
OFEV is an important new advance in the treatment of IPF

It is an important new choice for the IPF patients

OFEV has a convenient dosing schedule of 150mg twice a day, and it has manageable side-effect profile. The patient's choice of delivery device must be individualized to what is best suited for the individual patient. We support having access to oral medications and inhalation devices which the patient is able to correctly use with ease and which improve the effective delivery of the medication to the lungs. Please provide access to the new medication for this rare and fatal lung disease!

Section 4 — Additional Information

Please provide any additional information that would be helpful to CADTH, CDEC, and participating drug plans. This could include suggestions for improving the patient input process, indicating whether the questions are clear, etc.

On behalf of our IPF patients, please make easy access to our IPF patients who do not respond on Esbriet!

Many …many …thanks

Canadian Pulmonary Fibrosis Foundation

Section 1 — General Information

Name of the drug CADTH is reviewing and indication(s) of interest		Nintedanib (Ofev)
Name of the patient group		Canadian Pulmonary Fibrosis Foundation
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Permission is granted to post this submission		Yes

1.1 Submitting Organization

The Canadian Pulmonary Fibrosis Foundation was established to provide hope and support for people affected by pulmonary fibrosis. Robert Davidson, president of the CPFF, created the organization in 2009 to raise money to find causes and treatments for PF, provide education and support for people affected by pulmonary fibrosis, and to help answer those non-medical questions frequently asked by those suffering with the disease. The CPFF is a not-for-profit charitable organization and is a registered Canadian Charitable Foundation. Our vision is make lung transplants the LAST resort not the ONLY resort for people with pulmonary fibrosis. Our mission is to raise funds to finance research to better understand, develop treatment and find a cure for pulmonary fibrosis. The CPFF has provided $395,400 in research grants over the past four years to leading institutions and researchers in Canada including University Health Network, University of Alberta's Lung Health Centre, St Joseph's Hospital Foundation and Toronto General Hospital Foundation.

Our key objectives in the battle against pulmonary fibrosis are to:

- Raise public awareness about this fatal disease
- Offer support to those affected by pulmonary fibrosis
- Represent Canadians affected by PF to government, healthcare professionals, the media and the public – to be "the patient voice" for PF

The CPFF has also:

- developed and printed a 20 page Patient Information Guide available in both English and French that is freely available to patients, caregivers and medical offices across the country
- a comprehensive web site in both languages,

- a Support Group Toolkit that is also freely available
- A fundraising guide that is also freely available
- A guide and training workshops for patient support group leaders
- A program to recognize and reward the important role of caregivers

The CPFF works closely with the medical community and with support from Canadians we hope to achieve our vision, mission and objectives. The CPFF has many individual and corporate sponsors, with an annual spring gala, Scotiabank half marathon and 5KM run in Halifax, Vancouver, Toronto and golf tournament raising a significant portion of our funds.

1.2 Conflict of Interest Declarations

The Canadian Pulmonary Fibrosis Foundation has a number of corporate and individual sponsors, all listed on the CPFF website. These sponsors include Boerhinger Ingelheim, the manufacturer of nintedabnib (Ofev) and Roche, the manufacturer of pirfenidone (Esbriet). Other major sponsors include a private family foundation, the Investors Group, Dynamic Funds and Emerson Canada.
Pharmaceutical donors include: Roche, Boehringer Ingelheim

No conflicts to declare.

Section 2 — Condition and Current Therapy Information

2.1 Information Gathering

An online survey directed to both Canadian and US IPF patients and caregivers was conducted between April 1 and April 24, 2015. The survey included a total of 159 responses broken down as follows: 131 respondents from Canada, 24 from the United States and 4 from countries outside of North America (one respondent skipped the question). A total of 112 were patients and 47 were caregivers. A total of 23 respondents had used nintedanib (Ofev), of the 23, 18 were from the US, 4 from Canada and 1 from the UK. Additionally, the information includes my experiences as President of CPFF and an IPF patient and my conversations with other patients across the country.

The answers to the questions in this report that are not specifically related to patients' and caregivers' experience or expectations with nintedanib (Ofev) are derived from our previous survey conducted for the purpose of providing our input for pirfenidone (Esbriet). Information gathering consisted of a national online survey conducted between August 17 and September 10, 2014. The survey included responses from 217 individuals, primarily patients living with IPF, but also caregivers. Additionally, the information includes my experiences as President of CPFF and an IPF patient, my conversations with other patients across the country and seven individual telephone interviews with patients (and caregivers) the week of September 8, 2014.

2.2 Impact of Condition on Patients

Note the response to this question is based on the results of our survey conducted in August/September 2014. See description above in section 2.1 Information Gathering.

All patients diagnosed with IPF experience many problems and symptoms as a result of the illness. Depending on the stage of the disease, patients will have different symptoms and increasing severity of symptoms. Additionally, patients not only deal with physical issues, they also have to manage psychosocial issues associated with their prognosis.

Based on the national survey and supported by the telephone interviews, patients describe the following most common symptoms:

- 96% experience breathlessness, or shortness of breath during exercise
- Almost 84% deal with tiredness and loss of energy, as well as reduced physical activity. Several patients commented that reducing physical activity also negatively affected their mental well-being in that they knew their illness was progressing and further limiting their quality of life.
- Over 62% have a dry, hacking cough, usually lasting longer than eight weeks. Many other patients commented about a productive cough, and several noted that clearing mucous became more difficult with the progression of their disease, making them more susceptible to infection.
- Almost all patients noted that slowing the progression of the disease would be the most desired current goal. All of the patients interviewed were under no illusion of finding a cure, however they all hoped to slow the progression of the disease to allow them greater quality of life. Several also hoped that slowing the progression would allow them to qualify for a lung transplant.
- All patients noted some symptom that affected their quality of life, notably reduced physical activities, including even routine activities such as climbing the stairs in their homes, walking their dogs, gardening or taking out the garbage
- 73% of patients noted a decrease in their ability to work or to be productive at work. Several patients commented that they lost their jobs as a result of the symptoms of their IPF.
- Over 60% of patients said they had to attend frequent medical appointments, and that it took a lot of effort and energy to get out of the house
- Patients with more advanced IPF (moderate to severe), had great difficulty getting around their home
- Patients with mild to moderate IPF said that while they haven't experienced many symptoms, the stress of the diagnosis and prognosis greatly affected their quality of life. One newly-diagnosed patient commented that while he was still able to get out golfing, he now needed to take a cart and knew it would be just a matter of time before he wouldn't be able to golf at all.
- Trips outside the home require planning and are often avoided, and for many the social isolation is further limited by their inability to travel by airplane to visit family and friends, and sort out where and how they can have ready access to oxygen.

2.3 Patients' Experiences With Current Therapy

Note the following is based on the results of August/September 2014 survey. See description above in section 2.1 Information Gathering.

Over 55% of patients surveyed were using oxygen. Of those patients using oxygen, 71% describe it as very or somewhat effective. Many were using oxygen at night only, and those with moderate to advanced disease also required daily use of oxygen if they were doing anything physical, such as walking around the house or going out for a walk.

Other treatments noted in the survey include:

- Over 47% currently or previously used N-Acetylcysteine. 26% of patients said it was somewhat effective in reducing symptoms.
- Over 35% have been or are in pulmonary rehabilitation and 32% found it effective 12% found it not very effective.
- Over 35% have used or continue to use prednisone (long-term use, not for exacerbations). Several patients commented that prednisone was somewhat effective, however all patients noted the negative side effects (such as weight gain) as adding to their stress.
- 7% have tried Imuran (azathioprine)
- Just over 4% have had a lung transplant

Most patients commented that their physicians told them there was very little they could do for them with current therapy, and were optimistic that pirfenidone (Esbriet), as the only treatment specifically for IPF, would help to slow the progression of disease. While a lung transplant is also an option, most patients were not holding out hope that they would be good candidates or able to get the transplant in a timely way. (Please note this comment is from our previous survey when pirfenidone was the only treatment specifically for IPF patients. I thought the comment about lung transplant to be relevant.)

The majority of IPF patients indicated that choice of drug to treat their IPF was very important as demonstrated by their response in our April 2015 survey. Patients were asked to rate on a scale of 1 – 5 the importance of choice in deciding which drug to take to treat their IPF. 1 was "not important as long as there is a drug for me" and 5 was "very important"; 87% selected 5, and 2% selected 1. This clearly demonstrates that choice is a number 1 need for patients with IPF

Similarly, results from the April 2015 survey, demonstrated that the majority of IPF patients considered improvement to quality of life to be extremely important. Patients were asked to rate on a scale of 1-5 the importance of improvement to overall quality of life from drug treatment. 1 was "not at all important" and 5 was "extremely important"; 92% selected 5.

2.4 Impact on Caregivers

Note the response to this question is based on the results of our August/September 2014 survey. See description above in section 2.1 Information Gathering.

Over 90% of caregivers worry about their loved one's prognosis and experience increased stress. Close to 2/3 of caregivers limit social activities and don't leave the home as often as they would like. Over 62% of caregivers attend frequent medical appointments with their family member. More than half of caregivers have experienced a decrease in their ability to work or to be productive at work. Many have to make accommodations around the house such as doing all household chores and all the yard work themselves, coping with depression and anxiety, feeling helpless to do more for their family member and worrying about coverage if their health plan stops paying for the treatment.

Section 3 — Information about the Drug Being Reviewed

3.1 Information Gathering

An online survey directed to both Canadian and US IPF patients and caregivers was conducted between April 1 and April 24, 2015. The survey included a total of 159 responses broken down as follows: 131 respondents from Canada, 24 from the United States and 4 from countries outside of North America (one respondent skipped the question). A total of 112 were patients and 47 were caregivers. A total of 23 respondents had used nintedanib (Ofev), of the 23, 18 were from the US, 4 from Canada and 1 from the UK. Additionally, the information includes my experiences as President of CPFF and an IPF patient and my conversations with other patients across the country.

3.2 What Are the Expectations for the New Drug or What Experiences Have Patients Had With the New Drug?

In Summary, based on the following survey results, it is very clear that patients want a choice of treatment so that if one is not tolerated they can choose an alternative effective treatment for IPF. Patients want to have nintedanib available as a second choice and they are satisfied that it is of benefit to them. This is important due to the evidence that some patients could not tolerate pirfenidone.

IPF patients are also very engaged and interested in treatments as shown by the excellent response to our surveys, two very close together. Patients want and need treatments for this 100% fatal disease

a. Based on the answer to an open-ended question, patients described the most important symptoms or issues that they would like addressed by drug treatment to be:
   - shortness of breath or difficulty breathing (44%)
   - delay disease progression (35%)
   - coughing (15%)
   - fatigue/energy 13%)

Respondents were asked to select the adverse/side effects that they would be willing to tolerate if a new drug gave them other benefits to help delay the progression of IPF. They provided the following responses:

Decreased appetite	53%	Nausea	17%
Weight loss	57%	Stomach pain	14%
Diarrhea	27%	Vomiting	7%
Headache	19%	I don't know	35%

Additional responses under other include, 3 respondents who indicated that they would not accept any side effects, 6 who would accept side effects with certain conditions, and 4 who responded that they would accept all side effects.

b. Overall patients found ninetanib effective at managing their common symptoms of IPF when asked to rate its effectiveness on a scale of 1-5. 1 was “not effective at all” and 5 was “very effective”. The weighted average for the response was 3.30, which indicated a better than average response. 55% said very effective.

1 - Not effective at all	**2**	**3**	**4**	**5 - Very effective**	**I don't know**	**Total**	**Weighted Average**
5% (1)	9% (2)	9% (2)	14% (3)	9% (2)	55% (12)	22	3.30

Patients were asked what adverse/side effects would they be willing to tolerate with nintedanib (Ofev) if it gave them other benefits to help delay the progression of IPF.

Diarrhea	83%	Headache	35%
Decreased appetite	74%	Stomach pain	22%
Weight loss	61%	Vomiting	22%
Nausea	52%	I don't know	9%

One respondent provided an additional written response stating “I had most of those effects with Esbriet & couldn't handle them so I stopped taking it. Ofev is tolerable.”

Patients found that nintedanib (Ofev) did improve their quality of life. When patients were asked to rate on a scale of 1-5 how nintedanib (Ofev) has affected their quality of life, 1 was “no improvement” and 5 was “highly improved”. The weighted average was 2.82, which was a slightly better than average response.

1 - No improvement	**2**	**3**	**4**	**5 - Highly improved**	**I don't know**	**Total**	**Weighted Average**

1 - No improvement	2	3	4	5 - Highly improved	I don't know	Total	Weighted Average
14% (3)	14% (3)	27% (6)	18% (4)	5% (1)	23% (5)	22	2.82

When asked about the most positive effects of nintedanib (Ofev) on their IPF, 6 patients responded that it helped to improve symptoms, particularly cough, 5 indicated that it provided hope, 3 felt that it would help extend their life or slow the disease, 2 thought it was positive to have fewer pills to take, 1 respondent didn't know, 1 wasn't sure and thought he/she was getting worse and 8 patients indicated that it was too soon to tell.

When asked about other benefits they are hoping for from nintedanib (Ofev), patients responded that they would like to slow or stop disease progression (17), prolong their lives (5), decrease exacerbations and reduce symptoms (4), be a cure (1).

When asked in an open-ended question about the most negative side effects that nintedanib has had on their IPF, 12 patients responded that they had none. A total of 9 experienced diarrhea and of those 2 reduced their dosage to resolve the problem, 1 experienced minimal side effects that were tolerable and 1 had a need for more oxygen.

Overall patients rated nintedanib (Ofev) as being much better than other drugs they have taken for their IPF. The weighted average for the response was 3.67, which indicated a much better than average response.

1 - Much worse –	2 –	3 –	4 –	5 - Much better –	I don't know –	Total –	Weighted Average –
4% (1)	4% (1)	0%	5% (1)	14% (3)	73% (17)	23	3.67

When patients who have used or are using nintedanib were asked to provide additional information about living with IPF, existing treatments or nintedanib (Ofev), 17 provided a response. Here is a sample of a few verbatim answers that reflect the sentiments of a number of respondents:

"I have only been on OFEV for two months, so it is too early to say whether or not it is effective at controlling my IPF. However, the data to date has shown some evidence that it can slow the progression of IPF in some cases. Because of this I would be willing to put up with a certain amount of side effects . The reality is I have experienced no side effects at all, and only time will tell if it is going to be helpful. In the mean time taking this drug gives me hope, and is the best thing that I believe that I can do for my health at this time. "

"Ofev makes sure it's patients know as much as possible re. it's use.A nurse visited me at home, a Co. rep had a one-on one in the Dr's office & there's been a support meeting downtown. They care! "

"Keep investigating to find if not a cure, relief from this stupid disease. "

"I have had no adverse reaction to Ofev, which I began using on 23 Dec 2014. Too soon to know what its efficacy is."

"My Pulmonary doctor chose OFEV for me because of the results of trials in Europe she felt were better than Esbiret, and that 2 pills at 150 mg ea was simpler to take than 3 pills at 100 mg ea of Pirfenidone."

Lung Association of Saskatchewan

Section 1 — General Information

Name of the drug CADTH is reviewing and indication(s) of interest		Ofev (nintedanib)
Name of the patient group		Lung Association of Saskatchewan
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Permission is granted to post this submission		Yes

1.1 Submitting Organization

The Mission of the Lung Association of Saskatchewan (LAS) is to improve lung health one breath at a time. Our Vision is Healthy lungs for everyone**.** Our Role is to improve respiratory health and the overall quality of life through programs, education, research, training, treatment, advocacy and prevention of lung disease. The LAS is a not for profit organization with a voluntary Board of Directors, approximately twenty full time equivalent staff and many volunteers.

The Lung Association of Saskatchewan provides the most recent information on lung diseases [including Idiopathic Pulmonary Fibrosis (IPF)] to person's living with lung disease, caregivers, families, the public and the healthcare community and professionals. The Lung Association provides support to people living with lung disease and their caregivers through a number of initiatives: A Helpline is staffed by licensed health care professionals who are Certified Respiratory Educators (CREs). CREs provide educational consultations to IPF patients and their family members on various aspects of IPF diagnosis and management. A recent Webinar was conducted to increase the awareness about IPF for both the public and health care professionals. Information on what IPF is, signs and symptoms, incidence, potential risk factors, diagnostic tools and criteria, natural history and treatment options and management suggestions was provided. Written and on-line information is available. The Lung Association is contracted by the Saskatchewan Ministry of Health to provide training and services for the SAIL home oxygen program for patients requiring supplemental oxygen. The Lung Association also provides some funding/support (i.e. equipment) for patients with IPF who have had a lung transplant. The Lung Association's work in health promotion and prevention includes advocating for public policies which reduce the exposure to potential risk factors such as tobacco smoke and environmental agents.

The Lung Association of Saskatchewan has developed and administers RESPTREC™ (the Respiratory Training and Educator Courses) for health care professionals across the country including courses on specific chronic diseases and Foundations of Chronic Disease Management Education. Resources are available such as the IPF webinar to both patients and others through the RESPTREC website.
The Lung Association has provided funding support for research in IPF.

The Lung Association of Saskatchewan is a member of the Canadian Lung Association partnership. All educational initiatives of the Lung Association of Saskatchewan are based upon evidence-based guidelines published by Lung Association's medical society, the Canadian Thoracic Society (CTS). In the absence of IPF Canadian guidelines, educational initiatives are guided by best practice as determined by physician specialists and researchers working in the area of IPF along with the most recent peer reviewed publications related to IPF.

1.2 Conflict of Interest Declarations

c) We have the following declaration(s) of conflict of interest in respect of corporate members and joint working, sponsorship, or funding arrangements:

The Lung Association of Saskatchewan has multiple sources of funding for programs and operations and is supported by individual and corporate donations, and through service contracts with government organizations. Funding sources include: direct mail campaigns such as the Christmas Seals campaign, raffles, such as Share the Air, door-to-door fundraising, memorial giving, bequests, and service contracts with government organizations. The Lung Association, does, from time-to-time receive program grants from health industry/pharmaceutical companies. Our relations and interactions with pharmaceutical companies remain transparent, and policy positions of the Lung Association are developed without industry influence.

The Lung Association of Saskatchewan has received unrestricted grants from the following pharmaceutical companies: AstraZeneca, Boehringer Ingelheim, GlaxoSmithKline, Grifols, InterMune, Merck Frosst, Novartis, Nycomed, Pfizer, Roche and Takeda.

d) We have the following declaration(s) of conflict of interest in respect of those playing a significant role in compiling this submission:

Neither the principal author, nor the Lung Association of Saskatchewan, has conflicts to declare in respect to the compiling of this submission.

Section 2 — Condition and Current Therapy Information

2.1 Information Gathering

The Lung Association of Saskatchewan is invested and involved in both IPF research and educational initiatives for those affected by IPF. We have Certified Respiratory Educators on staff who provide educational consultations to IPF patients, their family members and caregivers. The knowledge and experience garnered through research, best practice and direct involvement with patients is the basis of the information provided.

2.2 Impact of Condition on Patients

Idiopathic pulmonary disease (IPF) is a progressive, fatal condition that causes a buildup of scar tissue in the lungs. This scarring affects the small air sacs (alveoli) in the lungs where oxygen enters the blood and results in the lungs becoming stiff and making it difficult to breathe.
The most common patient complaints include shortness of breath (especially with activity) and cough. Other symptoms include fatigue, chest pain or tightness in the chest, loss of appetite, rapid weight loss and leg swelling.

IPF is a debilitating and degenerative lung disease where lung function progressively deteriorates. The majority of patients experience a slow, gradual progression over many years. Some patient's condition remains stable while others decline quickly. Some patient may experience episodes of acute worsening of symptoms and lung function decline which may leave patients with significant worsened disease[1].

Shortness of breath and limitation of activity have an impact on all parts of life: daily activities, hobbies, travel, and relationships. As the disease progresses, it has an intense impact on the quality of patients' lives.

2.3 Patients' Experiences With Current Therapy

There is no cure for IPF and limited therapies are available.. The therapies used for IPF include:

- Quitting smoking
- Flu pneumococcal pneumonia vaccines
- Education regarding patient self-management
- Pulmonary rehabilitation to maintain muscle strength
- Supplemental oxygen for those with hypoxemia
- Good nutrition for those who lose weight because of their disease
- Lung transplant for selected patients
- Limited medications options

Adverse Effects/Side Effects of IPF Medications

- The frequency and severity of the side effects are dependent on a number of factors including individual response. As with any medication benefits of the medication must be weighed with the side effects.

Accessing Therapy

- The Lung Association of Saskatchewan believes that access to IPF medications which will serve to improve IPF management should be available.
- We recognize that not all patients respond the same to various types of formulations of medications and we support having access to medications to which a particular patient responds better.

Unmet Needs

- Of critical importance to the treatment of IPF are medicines that will reduce the progression of loss of lung function and breathing difficulty.

[1] Raghu G, Collard HR, Egan JJ, Martinez FJ, Behr J, Brown KK, 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 Mar 15;183(6):788-824.

2.4 Impact on Caregivers

Our health care system places a lot of demands on both patients and caregivers to manage the patient's illness. Recognizing the severe limitations the disease imposes on patients, and the level of care required by advanced IPF patients, there is a dramatic impact on the caregiver's lifestyle and routine. Caregivers are often the spouse of the IPF patient. Recognizing that IPF is more prevalent in older Canadians, caregivers themselves are often older Canadians.

The following challenges are often faced by caregivers of patients with IPF:

Financial Challenges for Family Caregivers

- depending on the level of reimbursement for medicines in any province, and recognizing that many older Canadians are often on fixed incomes, IPF can put financial strains on both the patient and the caregiver
- caregivers often become very reliant on social supports

Physical and Mental Wellbeing

- because of the level of care required by advanced IPF patients, and the limitations on mobility, frequently caregivers will experience feelings of isolation
- recognizing that caring for a patient can be significant, many caregivers find their ability to manage their own physical health and mental wellbeing is very limited
- exhaustion is a common experience with caregivers of patients with chronic disease

Section 3 — Information about the Drug Being Reviewed

3.1 Information Gathering

The Lung Association of Saskatchewan is invested and involved in both IPF research and patient and health care professional education initiatives. We have Certified Respiratory Educators on staff who provide educational consultations to IPF patients, their family members and caregivers. The knowledge and experience garnered through research, best practice and direct involvement with patients is the basis of the information provided in Section 3.

3.2 What Are the Expectations for the New Drug or What Experiences Have Patients Had With the New Drug?

Slowing the progression of the disease, management of shortness of breath and cough and reduction in acute worsening (exacerbations) would be common treatment outcomes which patients and their caregivers would seek. Tolerance of side effects would be based on individual response to therapy and seriousness of side effects.

Section 4 — Additional Information

The LAS supports having access to a choice of medications for IPF.

Ontario Lung Association

Section 1 — General Information

Name of the drug CADTH is reviewing and indication(s) of interest		Name = Ofev / Nintedanib Indication = Idiopathic Pulmonary Fibrosis (IPF)
Name of the patient group		Ontario Lung Association
Name of the primary contact for this submission:		[redacted]
Position or title with patient group		[redacted]
Email		[redacted]
Telephone number(s)		[redacted]
Name of author (if different)		
Patient group's contact information:	Email	info@on.lung.ca
	Telephone	416-864-9911 or 1-800-344-5864
	Address	18 Wynford Drive, Suite #401, Toronto, ON M3C 0K8
	Website	www.on.lung.ca
Permission is granted to post this submission		Yes

1.1 Submitting Organization

The Ontario Lung Association is a registered charity that assists and empowers people living with or caring for others with lung disease. It is a recognized leader, voice and primary resource in the prevention and control of respiratory illness, tobacco cessation and prevention, and air quality and its effects on lung health. The Association provides programs and services to patients and health-care providers, invests in lung research and advocates for improved policies on lung health. It is run by a board of directors and has approximately 70 employees, supported by thousands of dedicated volunteers. The Ontario Lung Association has a provincial office in Toronto, as well as several community offices throughout Ontario.

1.2 Conflict of Interest Declarations

a) We have the following declaration(s) of conflict of interest in respect of corporate members and joint working, sponsorship, or funding arrangements:

The Ontario Lung Association receives sponsorship and grants from a number of pharmaceutical companies which support educational and research initiatives. Companies who provide funding to the Ontario Lung Association include: Pfizer, GlaxoSmithKline, Boehringer Ingelheim, AstraZeneca, Merck, Novartis, Takeda, InterMune, Grifols, Actelion, Astellas, Bayer, J&J, Roche, RX&D, Valent Pharmaceuticals, and Eli Lilly. This year we also received program funding from the Ontario Home Respiratory Services Association (OHRSA). None of these organizations participated in any way in this submission.

b) We have the following declaration(s) of conflict of interest in respect of those playing a significant role in compiling this submission:

(Nothing to declare)

Section 2 — Condition and Current Therapy Information

2.1 Information Gathering

The information provided in section two was obtained from five completed on-line surveys that have been sent to patients living with IPF, input from a certified respiratory educator as well as previous patient surveys completed by OLA.

2.2 Impact of Condition on Patients

The symptoms and challenges that patients experience as a result of IPF are many, but at the top of the list were fatigue and shortness of breath. These were followed closely by excessive mucus, wheezing, difficulty fighting infections and coughing. Many tasks like walking up the stairs, opening doors or getting the mail cause shortness of breath and fatigue. All day to day tasks take much longer. Increased incidence of infections was noted as an ongoing issue as was mood /emotional well-being. The need for medications is constant and the inability to do daily activities like housework, cooking or shopping leave some people feeling depressed, frustrated and without hope. IPF impacts almost all aspects of day-to-day life for people living with it. It affects: the ability to participate in physical and leisure activities (as noted by every respondent in the survey responses), the ability to work, travel and socialize. It also affects relationships with families and friends, and slowly robs people of their independence.
All respondents indicated that shortness of breath is the aspect of this condition that is the most important to control, followed by the coughing and the fatigue.

All patients indicated that their level of activity was reduced dramatically as a result of idiopathic pulmonary fibrosis. In several cases, individuals were quite active with golf, dancing, walking, and baseball. They are no longer able to do these activities, or if they do, it is on a much smaller scale and with the use of oxygen.

2.3 Patients' Experiences With Current Therapy

Current treatments do provide some relief for: fatigue, shortness of breath, cough, low energy, and the inability to fight infection, but the side effects such as: mood swings, confusion, difficulty sleeping and bowel issues need to be better managed.

The desire for fewer medical appointments was mentioned, as was a wish for less cost burden. Overall, patients would like their treatments to provide enough help that they can experience improved independence and require less assistance from others. The desire for more / increased energy was noted many times throughout the survey.

2.4 Impact on Caregivers

Caregivers of those living with IPF experience many of the same negative impacts on their lives. They too indicate that caring for these people has affected their work, relationships with family and friends, and their physical and leisure activities. As well, their independence and the ability to travel and socialize were impacted.

Section 3 — Information about the Drug Being Reviewed

3.1 Information Gathering

The information provided in section two was obtained from five completed on-line surveys that have been sent to patients living with IPF, input from a certified respiratory educator as well as previous patient surveys completed by OLA.

The patients indicated that they would be able to live with some side-effects, but nothing worse than they are already experiencing and nothing that was irreversible.

3.2 What Are the Expectations for the New Drug or What Experiences Have Patients Had With the New Drug?

Key treatment outcomes of IPF that patients and their caregivers would most like addressed are: to stop or slow the progression of the disease, to reduce fatigue, cough and shortness of breath, and to have increased energy levels. Shortness of breath is the number one improvement patients would like to experience from this new drug, followed by reduced need for oxygen.

The patients indicated that they would be able to live with some side-effects, but nothing worse than they are already experiencing and nothing that was irreversible.

Patients who are taking Ofev indicated they have experienced various benefits, including: reduced fatigue, cough, and shortness of breath, as well as an improved appetite and energy level. They also indicated there were no side effects as a result of taking this drug. Finally two of the respondents indicated that when comparing Ofev to other available drug treatments, it is superior in the following areas: administration of the drug, time to accommodate the treatment, cost burden, side effect and treatment of the condition.

Section 4 — Additional Information

All patients indicated that shortness of breath and fatigue were the symptoms they would most like to improve. Patients would like to be less dependent on oxygen. Patients also indicated that they would like to know more about new treatments for their condition – when they become available / how to be a part of clinical trials or experimental groups. Many patients also expressed interest in accessing information on new therapies.