ONTARIANS WITH RARE LUNG DISEASE FINALLY GIVEN THE CHANCE TO BREATHE

IPF patients in other provinces hope publicly-funded access to first and only treatment isn't far behind

TORONTO, ON – (September 3, 2014) – Ontarians affected by idiopathic pulmonary fibrosis (IPF) – a rare, progressive and fatal lung disease – have finally been given the chance to breathe after an emotional rollercoaster ride lasting almost two years, as they fought for access to the first available treatment for their deadly disease.

In Ontario, the Executive Officer has announced that Esbriet (pirfenidone), the only medicine available in Canada to treat mild to moderate IPF in adults, will be funded through the Exceptional Access Program (EAP) effective August 19, 2014. This funding decision will allow patients who meet certain criteria (which has yet to be published) and who rely on the Ontario Public Drug Programs to finally have access to the only medication currently available for the treatment of the disease.

The initial elation felt by Canadian patients at hearing that Esbriet had been approved by Health Canada in October 2012, quickly turned to acute dismay once they learned that the Common Drug Review (CDR) recommended provinces not publicly fund the drug, leaving many patients without access to the only treatment available. Hope had been cruelly taken away and, as a result, the issue of access to Esbriet has caused patients, caregivers, physicians and other healthcare professionals to relentlessly advocate to provincial governments for public funding. The entire IPF community desperately hopes the Ontario government will act on their commitment and begin providing immediate access to Esbriet, and that all other provinces will quickly follow suit.

"This announcement is the best news the Canadian IPF patient community has heard since this journey began two years ago, and we applaud the government of Ontario on its leadership decision to provide access to funding for patients who can benefit from Esbriet," said Robert Davidson, president and founder of the Canadian Pulmonary Fibrosis Foundation (CPFF). "Unfortunately, we've seen too many people lose the battle to this devastating disease while fighting for access to the only treatment that could have extended and improved their lives. We won't give up our efforts until all Canadians with IPF are given the same chance to breathe."

Newly published data supports need for access to treatment

Earlier in May, data presented at the 2014 International Conference of the American Thoracic Society (ATS) and concurrently published in the *New England Journal of Medicine* (NEJM) gave the IPF patient community renewed hope that publicly funded-access may soon be reality. The Phase 3 ASCEND Study: A Randomized, Double-Blind, Placebo-Controlled Trial of Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis (IPF) confirmed the proven efficacy and safety of Esbriet through a number of key primary endpoints.

According to the published data, the study demonstrated that Esbriet significantly reduced the decline in lung function by 47.9 per cent, as measured by a decline in forced vital capacity of 10 per cent or more. Additionally, the study showed that 27.5 per cent of patients experienced a reduced decline in the 6-minute walk distance (6MWD) test, and almost half (43 per cent) saw an improvement in progression-free survival with IPF. Most importantly, a pooled analysis of data from ASCEND and two previous phase 3 trials

(CAPACITY) with Esbriet showed that the risk of all-cause mortality in IPF patients was reduced by 48 per cent compared with placebo.

“As a physician, I am gratified that all IPF patients in Ontario who meet the criteria will have the opportunity to benefit from Esbriet and have access to this much-needed treatment,” says Dr. Shane Shapera, respirologist and IPF specialist at Toronto General Hospital, University Health Network. “The ASCEND data presented at ATS further reinforces the extensive body of clinical evidence and international expert consensus supporting the use of Esbriet for adults with mild to moderate IPF, and should be reconsidered by the Common Drug Review to decide if it will be listed on all provincial formularies.”

Permanent funding resolution in all provinces still needed

While the public funding of Esbriet in Ontario is good news and will serve the needs of some IPF patients in the short-term, a permanent solution is needed for all patients across Canada who could benefit from this treatment before their time runs out. Based on the strength of recently published data, compelling patient evidence submissions and expert clinical opinion, the CPFF urges the CDR to provide a positive recommendation for listing Esbriet on provincial formularies so that all Canadians with IPF can have long-term access to this proven medication. This recommendation is expected by spring of 2015.

“We are beyond thrilled to hear that the Ontario government has taken the lead to finally make steps towards granting IPF patients access to the treatment they deserve, “ says Larkell Bradley, North York resident and IPF patient. “It is our hope, that since we have fought so hard to come this far, that a full listing will soon be imminent.”

In order for Ontarians to receive funding for Esbriet, eligible patients must first receive a prescription from their respirologist, and be enrolled in the Inspiration Program, a patient support program offered by InterMune Canada. Esbriet will then be funded through the Exceptional Access Program (EAP), based on clinical criteria which are expected to be available in the coming weeks.

About IPF in Canada

Idiopathic pulmonary fibrosis (IPF) is an interstitial lung disease with no known cause. Studies suggest that up to 30,000 Canadians are believed to be affected by all forms of pulmonary fibrosis, with an estimated 3,000 to 5,000 suffering from mild to moderate forms of IPF. The disease is more common in men than women and is usually diagnosed between the ages of 40 and 80 years, with a life expectancy of just two to five years. In patients with IPF, the lung tissue becomes scarred and over time, as the scarring becomes thicker and more widespread, the lungs lose their ability to transfer oxygen into the bloodstream. As a result, patients become short of breath and the brain and vital organs are deprived of the oxygen necessary for survival. IPF has a higher mortality rate than many other malignancies, including pancreatic, lung and liver cancers.

About the CPFF

The Canadian Pulmonary Fibrosis Foundation (CPFF) is a registered not-for-profit charitable organization established to provide support, hope and resources for those people affected by pulmonary fibrosis. Robert Davidson, president of the CPFF, who had IPF and received a double lung transplant in January 2010, founded

the organization in 2009 to help support and educate others, and to answer non-medical questions frequently asked by those suffering with the disease. For more information, please visit www.cpff.ca.

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