Cystic Fibrosis Canada™

Connections

SUMMER 2015

TARA BOURQUE
LEAVING MY MARK ON THE WORLD

ONE ORGAN DONOR REGISTRATION AT A TIME

INTRODUCING OUR NEW PRESIDENT AND CEO, **NORMA BEAUCHAMP**

FOUNDER OF CANDID FACTS: **DONNA SUMMERHAYES**

2015/2016 KIN CANADA CYSTIC FIBROSIS CANADA LIAISON COMMITTEE

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On the cover: Tara Bourque, photo credit: Adele Marianne Photography

INTRODUCING OUR NEW PRESIDENT AND CEO, NORMA BEAUCHAMP

Norma Beauchamp joined Cystic Fibrosis Canada in November 2014 with extensive experience in the healthcare sector. An accomplished business and not-for-profit leader, Norma has over three decades of senior leadership positions in Canada and Germany. She held executive positions at Genzyme, Bayer and the Canadian Foundation for Women's Health. Norma is a seasoned senior fundraising volunteer, having served on the boards of the St. Joseph's Health Centre Foundation, Providence Healthcare Foundation and the Breast Cancer Society of Canada.

With a renewed focus in the fight against this fatal genetic disease, Norma is calling upon all Canadians in every corner of the country to join us in the quest to find a cure or control for cystic fibrosis (CF).

Q. What attracted you to the role of President and CEO of Cystic Fibrosis Canada?

N.B. My seven-year-old nephew has cystic fibrosis. Like most families, my family was no different in that we were devastated by the news. When I saw the job posting for the President and CEO role at Cystic Fibrosis Canada it seemed like the perfect fit and the vehicle of how I could make a difference in the lives of people living with CF. I bring my previous pharmaceutical marketing and management industry experience within Canada and globally in addition to my five years of board of director experience to this critical role.

Q. What is your vision for the future of Cystic Fibrosis Canada and what do you hope to achieve?

N.B. My vision is to inspire and enable people living with CF to lead long and healthy lives. Of course, the most important vision for the future is finding a cure or control. Having said that, we have a lot of work to do to get there. I hope that together we can educate Canadians about cystic fibrosis as many Canadians are not aware of CF and do not understand this fatal genetic disease. I also hope to **inspire more Canadians to donate to Cystic Fibrosis Canada to fund critical CF research and care.**

When I meet the many dedicated scientists, researchers and medical staff, I am truly excited about the possibilities for the future. I see the dedication of the volunteers. While I can honestly say that I have experienced a lot of great work related to healthcare and fundraising in my career, I have not seen the kind of commitment and passion that I see, among those involved with cystic fibrosis.

Q. Since taking on the role in November, what do you see as the biggest opportunity for the organization?

N.B. We need to dedicate ourselves to finding a cure or control for this disease while living the Cystic Fibrosis Canada values: Caring, Accountability, Teamwork and Excellence. There are many charitable organizations competing for the same funds for some very compelling reasons. We need to differentiate our goals and objectives effectively to our donor population so that our vision is clearly understood. I look forward to working with you in making a difference in the lives of every CF patient and family.

"My vision is to inspire and enable people living with CF to lead long and healthy lives."

HOPE THROUGH PROGRESS

NEW RESEARCH REVEALS CANADIANS WITH CYSTIC FIBROSIS LIVING LONGER THAN THEY DID 25 YEARS AGO


Megan Parker

MEDIAN AGE OF SURVIVAL SURPASSES 50 YEARS OF AGE

Based on the most recent *Canadian Cystic Fibrosis Registry* data, the predicted median age of survival for Canadians with cystic fibrosis (CF) is now 50.9 years, over 25 years higher than in the early 1980s.

"The median age of survival in Canada for CF patients has now passed 50 years of age and shows tremendous progress in the fight against cystic fibrosis," says Dr. Anne Stephenson, Director of the CF Registry and CF clinician at St. Michael's Hospital.

Data in the Registry are collected from all 42 CF clinics located across Canada. The Registry is an important resource for CF clinicians to help monitor and identify emerging patterns in health outcomes of CF patients, as well as for researchers searching for a cure or control for this devastating disease.

"Although improved survival is very positive news for the CF community, we must remember that people with cystic fibrosis still die prematurely. In fact, in 2012, half of the deaths due to cystic fibrosis happened before the patients reached their 32nd birthday. This reinforces the need for continued research in this area in order to optimize health for all individuals living with this progressive disease," says Dr. Stephenson.

Cystic Fibrosis Canada provides nearly $2 million in funding each year to CF clinics through the annual Clinic Incentive Grant program to support efforts in providing high quality CF care. The Registry continues to benchmark the tremendous strides we are making, and would not be possible without the tireless funding support from our donors.

Summary data from the Canadian CF Registry are available on our website at **www.cysticfibrosis.ca**.

UNDERSTANDING THE PREDICTED MEDIAN AGE OF SURVIVAL

The median age of survival is the estimated age beyond which 50 percent of the CF population would be expected to live, assuming the mortality rate in CF remained constant. This is not the age at which people with CF would be expected to die. For example, if a child with CF is born in Canada in 2013, they would have a 50 percent chance of living beyond 50.9 years of age based on current mortality rates, meaning half of the CF population would be expected to live to an age older than 50.9 years. Since mortality rates are not static and are constantly changing as new therapies and medicines for CF become available, this estimate is a reflection of the most accurate data that is available today.

2013 CANADIAN CYSTIC FIBROSIS REGISTRY HIGHLIGHTS:

- ✔ Over 4,000 Canadians received care at one of the 42 CF clinics across the country
- ✔ The median age of Canadians with cystic fibrosis is 21.4 years of age
- ✔ Almost 60 percent of all Canadians with cystic fibrosis are adults
- ✔ 118 new CF diagnoses were made in 2013 – 38 were through the newborn screening program and 13 were over the age of 18 years
- ✔ 44 CF patients received transplants
- ✔ 23 percent of all Canadians with cystic fibrosis have CF-related diabetes (CFRD)

INFECTION PREVENTION AND CONTROL FOR CYSTIC FIBROSIS

Q & A WITH DR. VALERIE WATERS

The Cystic Fibrosis Foundation (U.S.A.) updated the *Infection Prevention and Control Guideline* for individuals with cystic fibrosis (CF), their families and healthcare professionals in 2013 to help reduce the spread of germs in the clinic and hospital setting, as well as in everyday life. On the recommendation of its Healthcare Advisory Council, Cystic Fibrosis Canada has shared the updated guidelines with all Canadian CF clinics.

Dr. Valerie Waters is a physician and researcher at the Hospital for Sick Children in Toronto. Dr. Waters' research is specific to infectious diseases and she was an advisor on the implementation of the Infection Prevention and Control Guideline for Cystic Fibrosis in Canada.

Q. How were the new guidelines developed?

V.W. The new guidelines were published in 2013 and were the result of many years of work by a panel that was sponsored by the Cystic Fibrosis Foundation (U.S.A.); the panel included members from many countries including Canada to update the old guidelines that had not been updated since 2003.

Members of the panel included respirologists, nurses, physicians and many others – there was quite a representation from different specialities. The process involved looking back through the old guidelines and seeing what had changed in the past 10 years. Since 2003 there have been several developments in what we know about the transmission of bacteria specific to cystic fibrosis. New research and literature was also assessed and included in the updates to the guidelines.

Cystic Fibrosis Canada reviewed the new guidelines with the Healthcare Advisory Council and decided they were applicable to Canadians with cystic fibrosis, but also CF patients worldwide.

Q. How will these guidelines be implemented in cystic fibrosis clinics?

V.W. Because Canada is geographically diverse, clinics can range from a very few to 500 patients and each clinic will have different resources for implementation. For example, negative pressure in hospital rooms is not always an easy thing to institute especially in older hospitals. The guidelines will have to be adapted specifically to each clinic and incorporated into Cystic Fibrosis Canada's Accreditation Site Visit program.

Patient education is another important factor for implementing the new guidelines to inform everyone why the new policy was instituted and how it is different from the earlier version.

Q. Why would clinics be unable to implement the guidelines?

V.W. Depending on the size of the clinic and the amount of personnel or the way the clinic is laid out, there may be some challenges for implementation. The amount of patients flowing through the clinic also affects how the guidelines can be implemented. Each clinic will adapt the guidelines to its unique situation to the best of their ability, to protect patients from cross infection.

Q. What do the new guidelines mean for CF patients?

V.W. The overarching goal of the guidelines is to make cystic fibrosis patients safer. While it is easy to get caught up in the details, the main purpose of the guidelines is to prevent the transmission of harmful bacteria from one patient to another. This might result in some changes for patients, such as doctors attending to them in yellow gowns and gloves, and this can be unusual at first. At the end of the day, however, these guidelines are meant to protect CF patients from harmful pathogens and prevent infections from passing from one patient to another, and this will result in better CF care.

For more information about Infection Prevention and Control, visit www.cysticfibrosis.ca/about-us/infection-prevention-and-control.

ADVANCES IN INFECTION CONTROL LEAD TO SAFER CLINIC SPACES

Q & A WITH DR. LARRY LANDS

Despite significant progress in treating cystic fibrosis (CF), lung infections caused by germs, such as bacteria, viruses, yeasts and molds, remain a serious problem for those with CF and can lead to worsening of the disease. The *Infection Prevention and Control Guidelines* contain recommendations to reduce the risk of CF patients receiving or spreading potential pathogens in the CF clinic or hospital setting. Negative pressure rooms in CF clinics further reduce the patient's risk of cross-contamination of harmful bacteria.

Dr. Larry Lands is the CF Clinic Director at Montreal Children's Hospital. A long-serving volunteer and clinical advisor to the Canadian CF community, Dr. Lands has assumed several leadership roles for Cystic Fibrosis Canada including a Chair position on a past Clinical Advisory Committee. Dr. Lands has also served as a medical leader on numerous Accreditation Site Visits as part of Cystic Fibrosis Canada's program to ensure consistent high quality CF care across the country. Dr. Lands is recognized nationally and internationally for his renowned research in cystic fibrosis.

Q. What is the purpose of negative pressure rooms, and how do they work?

L.L. We now have a better understanding of how germs, like *Pseudomonas aeruginosa bacteria*, or viruses, are spread. Most of these are spread by droplets which are produced when someone sneezes or coughs, or performs pulmonary function tests. It used to be thought that these could not travel beyond a range of three feet, but we now know that they can be projected up to six feet.

A standard room has the same pressure as the corridors outside. A negative pressure room causes air to flow from outside and into the room, like taking a breath in. This limits how far a droplet produced in the room can travel. The air that is brought into the room is then either sent outside the building, or cleaned through a high-efficiency particulate arrestance (HEPA) filter before being re-circulated.

Q. How will negative pressure rooms help with infection control?

L.L. By limiting the movement and spread of droplets, negative pressure rooms, particularly in the pulmonary function laboratory where people often cough, can keep the droplets from moving around.

Q. Will care for CF patients be affected by negative pressure rooms?

L.L. The patients will not notice any difference. In fact, having negative pressure in the pulmonary function laboratory and clinic rooms allows patients to move through more quickly. In pulmonary function laboratories and clinic rooms without negative pressure or HEPA filters, a wait time between patients is recommended.

Q. Is cross-infection reduced for CF patients with these new rooms?

L.L. This is the desired result. Of course, negative pressure rooms are just part of an overall strategy to reduce the risk of cross-infection. Patients should wear masks when in common areas of the hospital and clinic (but not in the clinic visit room), and staff are encouraged to wear gowns and gloves. Generally the patient will remain in the clinic room while the members of the CF team come by for the visit. In addition, surfaces in clinic rooms and stethoscopes are wiped between patients. If patients must wait in the waiting room, then they should sit at least six feet apart.

For more information about *Infection Prevention and Control* for CF patients, families and health care providers in different settings visit **www.cysticfibrosis.ca.**

Cystic Fibrosis Canada President & CEO Norma Beauchamp with CF Clinic team at Hotel Dieu Hospital in Kingston, ON

Norma Beauchamp with CF patient Daniel Williamson

CLINIC SPOTLIGHT: HOTEL DIEU CF CLINIC, KINGSTON, ONTARIO

A sense of caring and community is what you immediately feel upon walking through the doors of the Cystic Fibrosis (CF) Clinic at Hotel Dieu Hospital in Kingston, Ontario. Home to both an Adult and Children's CF Clinic, Hotel Dieu Hospital provides outstanding patient-centred care to individuals with cystic fibrosis who reside in Kingston and its surrounding areas. Hotel Dieu's CF Clinics are among Cystic Fibrosis Canada's 42 accredited programs in the country.

At each CF clinic visit at Hotel Dieu, patients are cared for by various members of an interdisciplinary healthcare team. Dr. Richard Van Wylick is the Director of the Paediatric Cystic Fibrosis Clinic and cares for CF patients from birth through 17 years of age. Dr. Diane Lougheed is the Director of the Adult Cystic Fibrosis Clinic and cares for individuals with CF who are 18 years and older. Other members of the CF team include: Lisa Smith, Nurse Coordinator; Kristy Brundage, Physiotherapist; Julie Nedvidek, Dietician and Tania Nicholls, Social Worker.

"For many years, we have served the CF patients and families of this region and have become known for high-quality CF care. We have a very high-calibre team and we are all working together to provide the best possible care for each CF patient that comes to our clinic," says Dr. Van Wylick.

Daniel Williamson, a CF patient in the Adult Clinic at Hotel Dieu was concerned about the care he would receive after turning 18 and moving to Kingston to start University.

"Starting my University career at Queen's, I was worried about the treatment I would receive after turning 18 and transitioning to an Adult CF Clinic especially away from my hometown. After my first appointment at Hotel Dieu Hospital, I've never felt more accepted. Each individual on the cystic fibrosis team was welcoming, caring and passionate, which made me feel comfortable. They provided me with a new confidence – I was no longer nervous about telling people that I had cystic fibrosis, and for this I will always be thankful," says Daniel.

The key to the success of the CF Clinics at Hotel Dieu Hospital is through teamwork and compassion for each patient.

To learn more about how CF care is delivered in Canada or to connect with a CF clinic in your community, visit **www.cysticfibrosis.ca/cf-care**.

FUNDING RESEARCH TO IMPACT THOSE LIVING WITH CYSTIC FIBROSIS

Q & A WITH SCIENTIFIC REVIEW PANEL STAKEHOLDER MEMBERS: JAMES CUMMINE AND ROB LAKE

When making decisions on research funding, Cystic Fibrosis Canada relies on scientists and clinicians on the Scientific Review Panel to expertly review each grant application. In addition to critical review of the science, the Panel looks to its stakeholder members for their unique and significant perspectives, based on their experiences, on the relevance of the proposed research projects to those living with cystic fibrosis. James Cummine and Rob Lake have been engaged stakeholder members of the review panel for the past three years and two years, respectively, and we thank them for volunteering their time and invaluable insight throughout their tenure in the roles.

Q. How did you first become involved with CF Canada and later with the grant competition?

James: It began shortly after my daughter was born. She was diagnosed with cystic fibrosis just days after her birth. About four months later, we found ourselves attending the Chapter Open House in search of information, and I have been involved in CF Canada's work for over 20 years now. Initially, I got involved with fundraising events at the Chapter level, then became a Chapter President, and then Regional Director for Manitoba. That got me involved at the National level. However after more than 15 years on the fundraising side, I needed a change and the lay stakeholder position on the research grants review committee was available. I welcomed the chance to learn more about the disease while helping to make a difference.

Rob: I moved to Canada in November 2011, and was almost immediately put in touch with the various outreach arms of CF Canada in Southwestern Ontario, where I was living. I was nominated by my local Chapter to serve on CF Canada's Adult CF Advisory Committee (ACFAC), a role I have held since 2013. Based on my background, experience on the ACFAC, as well as my experience as a kinesiology professor, my name was put forward for the position of stakeholder member on the panel. That was two years ago now, so I just finished my second year as a member on the panel.

Rob, Siobhan, Aoife, and Carys Lake

Glenna, James, and Amy Cummine

Q. Describe the process of reviewing a grant from a stakeholder's perspective.

Rob: Grant reviewing in basic terms involves reading the lay summary provided by the author within the proposal, and judging whether, and to what extent, the research proposals and potential outcomes are relevant and impactful to CF patients. If the lay summary is well written, this process does not take too long but if there is a great deal of jargon, a good deal of research goes into the process. It is important to ensure that I understand as best I can what the research aims to do, what problems it aims to tackle, or what questions it aims to answer.

James: Initially, it is a learning process, understanding the grant application and the research world. The rest was easier. Once I understood the basic premise of the grant application, which was possible because of the lay summary, it was pretty straight forward determining which ones would lead to a cure and which ones a control, which ones were about basic information and which ones were focused on the symptoms of the disease. Then it is about trying to assess how this will impact the CF population.

Q. How do stakeholders contribute to the scientific review process?

James: At first, I was very concerned about that myself. What did I really have to contribute? However, after living with my daughter and seeing her daily battle, I quickly realized that there were certain aspects of her quality of life that I have unique insight into, and that insight guides me to comment on how each research project would impact the lives of those living with CF. That was the perspective that the rest of the panel members were looking for from us. It wasn't about the research itself but how it would help those living every day with this horrible disease.

Rob: After the scientific advisors read their summaries of the proposal and rate them with scores (out of 10) based on how good the research is or proposes to be, stakeholders provide a score and give verbal input into the discussion. In some cases, our input is decisive, particularly where the scientists rank the proposal very high because of how good the 'science' is, but we score it lower because it aims to tackle a marginal issue for CF patients, or the results may only be relevant to a small proportion of those who have CF. In these cases, our input can mean some proposals that are good scientifically, but less relevant to patients, come out with lower scores and therefore have less chance of getting funded, but also that really relevant research projects, which might have a couple of flaws but potentially could be a 'game changer' for how CF is treated or understood, may have a better chance of getting funded.

Q. What factors do you consider when assigning a relevance score to a grant proposal?

James: First, will it lead to a cure or control? Second, is it basic understanding or quality of life research? And third, will it impact a large portion of the population or only a small percentage with certain mutations?

Rob: There are three main things, ranked in order of importance: 1) To what extent will this research project work towards finding a cure? 2) To what extent will this research project work towards better control for the disease? 3) To what extent will this research project work towards improving the quality of life for CF patients? We also look at the potential proportion of CF patients that the research applies to, and to what extent positive change can be had with the proposed outcomes. So, while we rate all science as at least somewhat 'relevant', those that aim to treat/control the main (and most troublesome) illnesses that CF patients get (e.g. *Pseudomonas* or *B. cepacia*) will score higher than more marginal or less worrisome or problematic bugs. Also, projects that target the most common mutation of CF (i.e. F508del) are considered more relevant than some of the more obscure mutations, simply because the outcome can positively impact a greater proportion of CF patients.

Q. What did you enjoy most about participating on the scientific review panel?

James: Meeting and talking with researchers and other CF clinicians from across the country. I think the priceless part is the contacts and discussions I have had with these new friends. It has allowed me to learn so much that will hopefully help improve my daughter's care and quality of life.

Rob: I most enjoy having an impact on the discussions, and the feeling of my presence in this panel being of relevance to the scientific community. At the end of the day, the fate of most people with CF lies in the hands of the scientists working towards a cure, so it is awesome to feel as though I am able to help direct their attentions (and CF Canada's money!) to the most relevant and worthwhile causes. I also enjoyed the opportunities to develop my knowledge and understanding of the disease, from a scientific (molecular, biochemical, etc.) perspective.

Q. What have you learned by taking part in this year's grant review?

Rob: I have learned a great deal more about CF, and also a bit more about those scientists working on behalf of the CF patient community to find a cure. I have also gained an appreciation for how important this work is to the CF community, and how much responsibility we, collectively, have in this process in terms of how decisions are arrived at and how money is best spent.

James: Given this is my third and final year, it is the time it takes to go from research to clinical trials. I have heard so many potential advances in both the treatment of the basic defect and the symptoms of the disease. However, they still seem too far away as I see my daughter still battling hard every day. There is so much that has been learned, but still so much to learn before there will be a cure. That day can't come soon enough but from what I have seen, I feel confident that it will come.

Q. James, this was your last year serving as a member of the scientific review panel. How do you plan to stay involved with Cystic Fibrosis Canada in the future?

James: Well, I have done a lot of fundraising and now I have reviewed research grants. The other area I would like to move into next is an advocacy role, both for the CF population and for my daughter. I have realized that CF patients need advocates to fight for them because they are so busy fighting the disease. I am really intrigued by the quality improvement initiative launched by CF Canada and I would like to see it expanded to every clinic across Canada. As well, I would like to make sure every CF patient has access to the latest drugs and treatments as quickly as possible as every day really does matter! I am working to achieve these last two goals in my new role as a member of CF Canada's Healthcare Advisory Council.

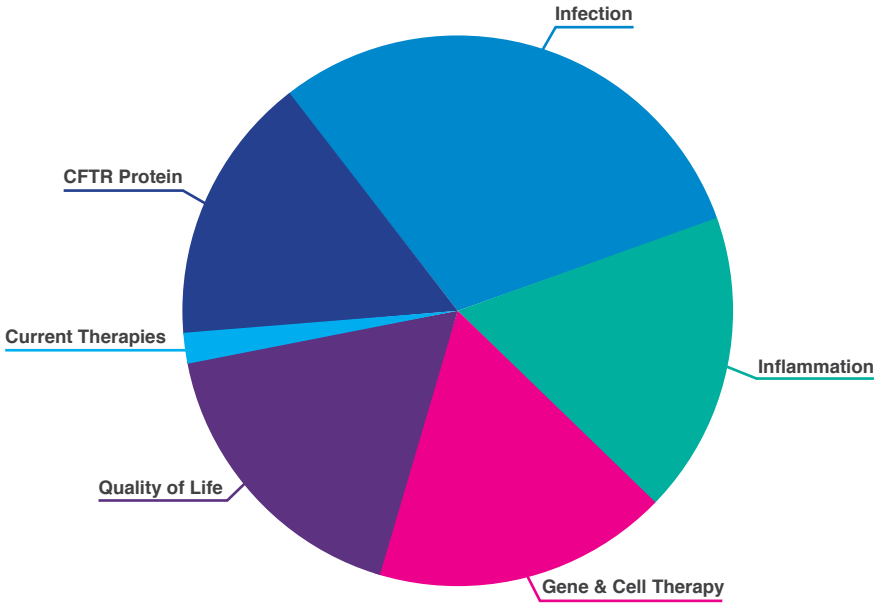
2014 CYSTIC FIBROSIS CANADA RESEARCH COMPETITION RESULTS

In January 2015, Cystic Fibrosis Canada's Scientific Review Panel convened to review grant applications submitted to the 2014 Fall Research Competition, the largest competition of the year held by the organization. The top applicants were awarded funding for their projects following a rigorous, independent review process where all grants were evaluated for scientific merit and relevance to CF by a panel of scientists, clinicians, and stakeholder lay members.

The 2014 competition grant recipients represent 12 institutions from across Canada and have interests in a variety of research areas that are central to finding a cure or control for CF; 13 discovery research grants, two clinical research grants, and one early career investigator award were funded. Additionally, in our continued efforts to train future CF scientists, CF Canada awarded three fellowships and five studentships to trainees working on promising CF research projects.

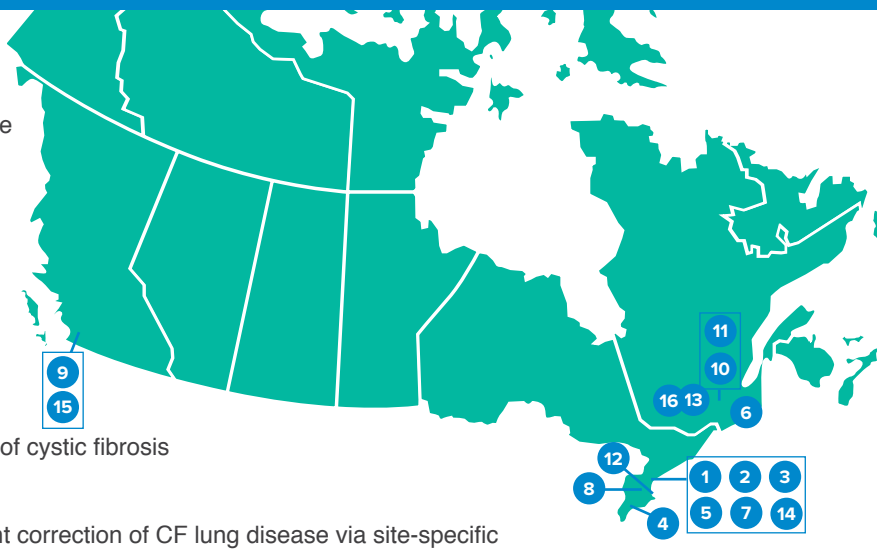
INVESTMENTS BY AREA OF RESEARCH* 2014 COMPETITION GRANT RECIPIENTS

TOTAL INVESTMENT†: $4,197,945



**Graph depicts new investments in the Discovery Research, Clinical Research, and Early Career Investigator awards, but does not include CIHR partnerships, training awards, previous research commitments, and other research programs; †Multi-year commitment to Discovery Research, Clinical Research, and Early Career Investigator grants awarded in the 2014 competition*

CONGRATULATIONS TO THE 2014 RESEARCH COMPETITION GRANT RECIPIENTS:



DISCOVERY RESEARCH GRANTS

1. **Dr. Shaf Keshavjee**, *University Health Network, Toronto*, Tissue remodeling and the immune response in obliterative bronchiolitis after lung transplantation
2. **Dr. David Clarke**, *University of Toronto*, Repair of CFTR defects caused by cystic fibrosis processing mutations
3. **Dr. Daniela Rotin**, *Hospital for Sick Children, Toronto*, Kinase inhibitors as correctors of ΔF508-CFTR
4. **Dr. Cory Yamashita**, *Western University, London*, Host-defense peptide fortified surfactant for the treatment of cystic fibrosis associated lung infections
5. **Dr. Jim Hu**, *Hospital for Sick Children, Toronto*, Permanent correction of CF lung disease via site-specific insertion of a human CFTR gene expression cassette into the genome of the airway epithelial progenitor cells
6. **Dr. François Malouin**, *Université de Sherbrooke*, Development of a combination therapy for MRSA and Pseudomonas aeruginosa in cystic fibrosis
7. **Dr. Neil Sweezey**, *Hospital for Sick Children, Toronto*, Narrowing the gender gap: Modulating estrogen/IL-17 signaling to control Th17 cells and neutrophil extracellular traps in the cystic fibrosis lung
8. **Dr. Sarah Wootton**, *University of Guelph*, Nuclease-based gene therapy for permanent correction of CF lung disease
9. **Dr. Stuart Turvey**, *Child & Family Research Institute, Vancouver*, Discovery of new anti-inflammatory therapies for cystic fibrosis empowered by enhanced understanding of the mechanisms of lung-damaging inflammation
10. **Dr. John Hanrahan**, *McGill University, Montreal*, Role of bicarbonate and mucus in airway innate immunity
11. **Dr. Don Sheppard**, *McGill University, Montreal*, Development of therapeutic agents for Pseudomonas and Aspergillus lung disease in cystic fibrosis
12. **Dr. Philip Britz-McKibbin**, *McMaster University, Hamilton*, Metabolomics for improved screening of cystic fibrosis: Better diagnostics without carrier identification
13. **Dr. Emile Levy**, *CHU Ste-Justine Research Centre, Montreal*, Unraveling the role of microRNAs in intestinal cystic fibrosis disorders

CLINICAL RESEARCH GRANTS

14. **Dr. Laurent Brochard**, *St. Michael's Hospital, Toronto*, Randomized cross-over physiologic study of HIgh Flow nasal oxygEN cannula versus non-invasive ventilation in cystic fibrosis: The HIFEN study
15. **Dr. Bradley Quon**, *Centre for Heart Lung Innovation, Vancouver*, External replication of a plasma protein biosignature to predict cystic fibrosis pulmonary exacerbations

EARLY CAREER INVESTIGATOR AWARD

16. **Dr. Genevieve Mailhot**, *CHU Ste-Justine Research Centre, Montreal*, Toward a better understanding of cystic fibrosis bone disease: Role of CFTR

CYSTIC FIBROSIS CANADA'S RESEARCH AWARDS RECOGNIZE OUTSTANDING CONTRIBUTIONS BY CANADIAN RESEARCHERS.

Senior Scientist Research Award – Dr. David Clarke
The Robbie Award for Most Promising New Research Project – Dr. Cory Yamashita
Marsha Morton Early Career Investigator Award – Dr. Genevieve Mailhot
Kin Canada Fellowship Award – Dr. Amber Park

CIRCLE OF FRIENDS

LEAVING MY MARK ON THE WORLD – ONE ORGAN DONOR REGISTRATION AT A TIME

Tara Bourque speaks at Lawn Summer Nights in London, ON

BY TARA BOURQUE

My name is Tara Bourque and I am a 21-year-old living with cystic fibrosis (CF). I was diagnosed at 14 months old, so CF has been my norm my entire life. I was very fortunate to grow up with very few complications as a result of my CF. I underwent one hospital stay for pneumonia when I was in grade 2 and another stay as a "tune up" of my lungs and an attempt to gain some weight in the summer of 2008. I was always considered a healthy CF'er until I reached my grade 12 year.

My lungs quickly deteriorated and I was placed in the hospital for 2.5 weeks. After being released and sent home on IV antibiotics it was clear that this hospital stay did not accomplish much in the way of improving my health and three months later it was back in the hospital – this time in Toronto – for a month long stay and many tests. I was told that a double-lung transplant might be a treatment I would require in the near future. Before this point I never knew that a transplant was a treatment option for CF patients. I didn't know that a healthy patient could suddenly be told that their disease had progressed to end stage function. I never thought I would be told I was dying before I turned 18.

After being released from this hospital stay, I graduated from high school and made the decision to go away to University while I had the chance to do so. I lasted 1 month and 10 days at the University of Guelph before attending my first clinic in Toronto as an adult patient. I was told I was the sickest patient in their clinic and that I needed to be hospitalized immediately.

I became dependent on oxygen to keep me breathing easier, IV medications to keep my health stable and a Bi-PAP machine to do the breathing for me overnight to allow my body to rest. It was at this time that I realized my life was no longer under my control. I was told I would need a transplant to survive and I would need one soon. I spent 2 months in hospital, decided to spend Christmas at home with family and friends and come January, I returned to Toronto to be placed on the transplant waiting list. The doctors informed me that it could be a 6-9 month wait, even longer sometimes, but that I needed lungs within 6 months or there was a fear I would not be strong enough to handle the surgery.

Tara (middle) with her sisters Erin (left) and Leah (right) Photo by Adele Marianne Photography

Luckily enough, I received my call for the perfect set of lungs a mere 11 days after being placed on the transplant waiting list. I had a speedy recovery of 18 days in hospital and three months in Toronto for physical rehabilitation while being blessed with only a few, very minor complications. The doctors were all amazed with my progress and at how well the new lungs were adapting to my body. I returned home in May to live a life I never could have dreamed of. No treatments for six hours a day. No inhaled medications. No IV poles to drag around. No wires attached to me. I was a normal, healthy individual that had to take a handful of pills twice a day to maintain my health and attend occasional clinic follow-ups.

I had two years of bliss. I returned to University, lived in residence, and experienced life and truly living as a young adult without a care in the world. Then two months shy of my second year "Lung-aversary" my lung function took a sudden, unexplainable drop. I underwent a steroid treatment and then a blood plasma filtering process in an attempt to figure out why my lung function had dropped and to reverse the damage. With no progress after two months of intense treatment I was diagnosed with chronic rejection of my lungs with no explanation as to why or how this happened.

Double-lung transplant recipient Tara Bourque has helped find more than 600 potential organ donors, making her one of the most successful recruiters in Ontario.

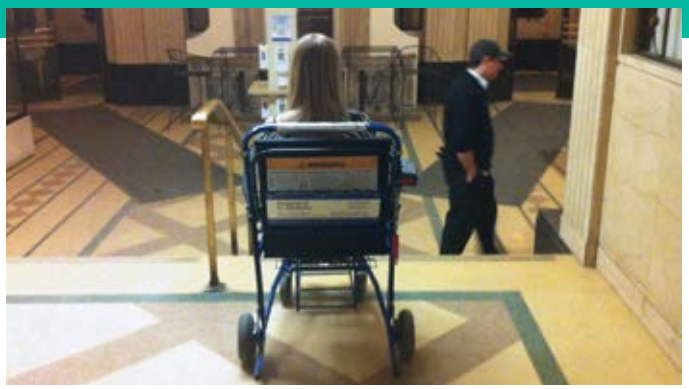
I am now three years post-transplant and in the process of being listed for my second double lung transplant. I am 21 years old and fighting for my life for the second time. For my third "Lung-aversary," I created a challenge for anyone who follows my journey – to raise awareness about both cystic fibrosis and the need for organ donors. On my Facebook page that I blog on and keep everyone updated on my medical life happenings, I put out the challenge for each follower to talk to three people – one for each year post-transplant – about organ donation.

Start the conversation that has the potential to change up to eight lives through registering as an organ donor. I wouldn't be alive today if it hadn't been for the selfless act of my donor and their family deciding to give a stranger a fighting chance at living. I would have never been able to experience living life feeling free of any medical worries. I would not have had my perspective and outlook on life altered for the better, my appreciation of the simple, small things in life discovered. I wouldn't have a story to share and for that I am forever grateful.

To visit Tara's Facebook Challenge page: search for *Helping Tara Breathe Easy*

To visit Tara's personal donor registration page: **www.beadonor.ca/tara-bourque**

MY CONSTANT BATTLE WITH CYSTIC FIBROSIS
BY MEAGHAN MACRURY



For me, cystic fibrosis (CF) is a constant struggle. It causes me to constantly worry about my future. I had my life all planned out – I wanted to become a nurse. But that all changed when I turned 17 and was told that it wouldn't be in my best interests for my health to pursue that dream. Then the constant hospital admissions began…four or five times a year.

My big plans turned into me just hoping I'd be out of the hospital to go on holidays or when I had made plans with my friends or family. CF has caused me to have to leave university four different times because I was unable to keep up due to being hospitalized so often. I often question my decisions for the future – like buying a home or any sort of decision that needs a concrete answer, because I cannot depend on my body physically to plan beyond the present. CF keeps me from living like a regular young adult my age, although it has caused me to become the woman I am, and I am proud of that person.

In December 2013, I began the downhill spiral that landed me in the Intensive Care Unit (ICU) for 40 days, on a ventilator, unable to breathe on my own. I spent 74 days in hospital learning how to relive and it's because of those three months that I know I can get through anything. CF causes a lot of unnecessary stress and heartache, but it does not define the person you are. I've realized that although it is a big part of me, and although I have many scars on my body because of it, I'm still Meaghan. I'm still a determined, loving, sarcastic 22-year-old that loves her friends and family. Maybe CF has caused my life to go a different direction than I had originally anticipated, but it has caused me to be right where I am today and I'm okay with that.

My favourite quote about CF is "It's not a glamorous fight, but it's my fight and I'm going to win."

In five years I would love to own a home, to have completed my education finally after working at it for five years and to have settled down with someone I love. I want to enjoy life to the best of my ability. I pray in five years there will be progress in the curing of CF.

Cystic Fibrosis Canada has given not only me, but my family a voice. It has allowed us to have constant information available at our fingertips as far as progress in research, events in our area and fundraising opportunities. Cystic Fibrosis Canada has allowed us to become a part of a network of other CF families that we otherwise would not have had the opportunity to meet and get to know. CF is a disease that doesn't affect you on the outside, but on the inside and it is because of the support of our own family, other CF families and Cystic Fibrosis Canada that those battling CF have the strength to continue to do so.

"It's not a glamorous fight, but it's my fight and I'm going to win."

IAN PETTIGREW: THE VISIONARY BEHIND THE JUST BREATHE PROJECT

Award-winning photographer and graphic designer Ian Pettigrew knew something wasn't quite right when he was unable to have children. He was 38 years old when he discovered the real culprit behind his health issues – cystic fibrosis (CF).

"Luckily my diagnosis is considered a milder variant of the disease, but it is cystic fibrosis nonetheless," says Ian. "Before that I had no idea, but looking back on my life I always had the symptoms like persistent coughing, frequent lung infections and digestive issues, just no one ever knew it was cystic fibrosis."

Ian is not alone. Today, nearly 60 percent of all Canadians with cystic fibrosis are adults. To help raise awareness about his disease, Ian drew upon his artistic talents and started the project: *Just Breathe: Portraits of Adults Living with Cystic Fibrosis*, to shine the spotlight on adults living with cystic fibrosis.

"Because of medical advances, overall life expectancy has greatly increased for CF patients," says Ian. "But it is still very much an uphill battle; this is not just a child's disease. What I want my portrait project to touch upon are the profound psychological aspects of cystic fibrosis in adults — especially those who are diagnosed later in life. It's sometimes, mentally, very challenging."

By putting a new face on cystic fibrosis through his portraits, Ian's goal is to bring attention to some of the additional issues adults with cystic fibrosis deal with apart from the main physiological symptoms – depression, anxiety, low self-esteem, and more.

Ian believes that research through donations is making a positive difference for Canadians with this life-shortening disease and that hope lies in knowing there are so many people willing to help.

Ian's book is now complete, featuring the portraits of 92 CF patients - and their stories - from across North America. For more information about Just Breathe: Portraits of Adults Living with Cystic Fibrosis and to order a copy, visit **www.thecfproject.bigcartel.com/products**.

"What I want my portrait project to touch upon are the profound psychological aspects of CF in adults."

LOUISE LANDRY **WASN'T SUPPOSED TO LIVE PAST 18, SHE'S NOW 43**

Louise Landry, Ottawa, ON

The good thing about being a 43-year-old with cystic fibrosis (CF) is seeing how far CF treatments have come. Or so says Louise Landry, a CF fighter from Ottawa, Ontario.

Louise was born in 1971, at a time when little was known about the disease. It took two years for doctors to finally put a name to the symptoms that had plagued her since her birth: malnutrition, weight loss, recurring colds and lung infections. When doctors at the Hospital for Sick Children finally uttered the words "cystic fibrosis", her parents were stunned... and scared to death. They had never heard of this illness that could potentially kill their child before she reached high school.

At the time, the median age for children with CF was 12 years old. Doctors warned the Landry's that their daughter probably wouldn't make it to 18. She would need to take Ventolin, a bronchodilator, by mask three times a day as well as daily antibiotics; she would also need percussive treatment using a machine that looked like something out of a science fiction movie, also three times a day. And she would need to take enzymes every time she ate, to help digest the food that her body couldn't break down on its own.

By the time Louise turned eight, she was spending three hours per day on her treatments. She was also taking 60 enzymes per meal. At that time, enzyme capsules weren't as powerful as those we see today. Rather than coated microspheres, they contained white powder that was 10-20 times less effective yet still acid enough to burn the corners her mouth.

Of course, it was impossible for Louise to swallow all 60 capsules every time she sat down to eat. To overcome this, her mother would empty the powder from the capsules into a bowl and mix it with baby food, creating a lumpy (and not-too-appetizing) stew-like mixture that she could swallow by spoonfuls as she ate.

Today, Louise takes 4-6 enzyme capsules per meal. The mask has been replaced by a pump, and she continues her percussive treatment with the old machine that her parents bought for her over 30 years ago.

Louise is also a manager, writer and single mom. She watches over her 21-year-old daughter who, like her, was diagnosed with CF when she was born. Fortunately, mother and daughter are lucky to benefit from the many CF-related developments that have come to pass over the past 40 years and those that are still to come.

Louise works full time as an aviation security professional in Ottawa. She also produces a quarterly flash fiction magazine called Saturday Night Reader (SNR) under her pen name, Suzanne Lucas. SNR is a fun and engaging journal for readers who enjoy genre fiction as well as character-driven stories; it is currently available at major Chapter/Indigo stores and independent retailers across Canada. A portion of the proceeds from sales will go to Cystic Fibrosis Canada to support their efforts to find a cure or control for cystic fibrosis. To support SNR, visit **www.saturdaynightreader.com**.

MY PASSION FOR GOLF

BY LISA LEGROS

Sometimes life has a way of giving you small gifts along the way. I have been blessed with many in my life; one of these surprises, although considerably later on in my life, was a passion for golf. I am a 48-year-old cystic fibrosis (CF) adult and a proud Adult Cystic Fibrosis Advisory Committee (ACFAC) member. I live in New Brunswick along the beautiful coast line of the Northumberland Strait which is the waterway between us and Prince Edward Island.

I have always been physically active throughout my life, which in my view has been a contributing factor in maintaining my health while living with CF. As a child, my parents always encouraged me to try a variety of sports. As most kids of my generation, we were outdoors all the time. I remember ballet lessons, figure skating lessons and downhill skiing for most of my teenage years. We lived near a ski hill in the Outaouais region, the perfect winter outdoor activity.

Later in life, I was introduced to golf. In my view, golf is the perfect activity for CF patients. Here's why: golf includes a great deal of walking - 9 and 18 holes of golf makes for a great walk. A golf course is a beautiful place to spend a sunny summer afternoon. And best of all, you don't need to mow the lawn!

One of the myths of golf is that you need considerable strength to achieve distance, but this is not so. What is required is the right technique and some practice.

My spouse, who introduced me to golf, would take me to the driving range to hit a few golf balls. From the start, I was totally hooked. It may not look like it but hitting 100 golf balls takes a lot of energy and is a great stress reliever. The driving range is a great place to start when you are learning this sport.

After a few years of golfing, it was suggested by a local golf professional that my new found passion and my personality would be a good fit as a golf instructor. Shortly thereafter I obtained my CGTF/USGTF teaching certification and have been enjoying teaching ever since.

Teaching golf continues to be a gift for me and a great place to spend the summer months. I hope that this article will convince you to try golf this season!

Lisa is the PrivateGolfPro who teaches in the greater Moncton area and south-east New Brunswick. Her 'Business of Golf' articles have been published in print in the Times & Transcript newspaper and available on her website.

To contact Lisa directly email her at lisalgolfpro@gmail.com, visit her website at **www.PrivateGolfPro.ca** or join her on Facebook at lisa.golfpro.

JIM BEST RIDES COAST-TO-COAST TO RAISE FUNDS FOR CYSTIC FIBROSIS CANADA

Q & A WITH JIM BEST

Jim Best, from Centreville, Nova Scotia, is not your average community pharmacist; he is an adult with cystic fibrosis (CF) on a mission. In July, 2014, he embarked on an incredible journey, with a lofty goal; to ride a motorcycle for 8,700 km from Boston to San Francisco and raise no less than $10,000 for Cystic Fibrosis Canada.

Jim accomplished both goals (raising over $10,300) in this adventure of a lifetime and we are very grateful for his incredible support! Jim shared some of his inspiring story with us.

Q. Why do you support Cystic Fibrosis Canada?

J.B. I support Cystic Fibrosis Canada because I have always been a strong active advocate for my condition and I wanted to do something to support the organization and give back.

Q. What inspired you to take on this sort of adventure/fundraising event?

J.B. I love to be on two wheels and the freedom my motorcycle offers. The inspiration came from having friends with cystic fibrosis that were not able to do some of the things they had hoped to do in their lifetime. Things like finishing university or having a regular job are feats that I was fortunate enough to accomplish. In realizing how lucky I have been, I wanted to do something to give back and honour and remember those that hadn't had the opportunity. Cystic Fibrosis Canada does a great job of sharing both the successes and hardships of people with cystic fibrosis. We have come a long way, but we still have so much further to go with a lot of promising potential.

Q. What did you do to prepare yourself?

J.B. I spent about five solid months preparing: booking, mapping, budgeting and getting in shape for the trip. Being on a motorcycle is quite fun but over 28 days is hard on the shoulders, arms and back (not to mention the incoming fumes from traffic over a period that long can have an effect). I spoke with others who had done the trip and picked up tips along the way for what to expect.

Q. What was your biggest Aha! Moment?

J.B. I had a lot of those moments. I blogged and shared a lot of them on Facebook and Twitter. Comments from complete strangers and kind words from family, friends and colleagues in support of the cause were moments that got me out of bed every day. The trip was not a mentally challenging trip for this reason. It was easy when I saw the effect it had on people and made it quite simple to push on.

Q. Besides the obvious, what could you have not lived without during your trip?

J.B. My aerosol mask and the mask under my helmet. I packed light, but without those two things, I don't think I could have done the trip. The air on the interstates is not clean and the barren roads of Utah and Nevada were dusty and hot. The mask I wore, along with my aerosol and physiotherapy were crucial to keeping fit enough physically to complete the trip.

Q. We hear you are a baseball fan, how many games were you able to take in?

J.B. I went to two games in Boston before leaving on the bike then followed it up with Cincinnati, St. Louis, Anaheim, and San Francisco.

This summer Jim will be travelling across Ireland on his motorcycle for eight days raising funds and awareness for Cystic Fibrosis Canada.

To read more about Jim's motorcycle fundraiser, visit his blog: **www.cfmotorcycle.blogspot.ca**. Follow Jim's journey on **Twitter @cfmotorcycle**, on **Instagram @jhbest** or **Facebook by searching CFMotorcycle: Ireland Cross Country**. To make a donation in support of Jim's journey, visit: **http://bit.ly/1BDSxHz**.

A LIFE OF BEAUTY AND LOSS IN FACING CYSTIC FIBROSIS

BY JOHN ROMANO

John Romano, Hamilton, ON

I was diagnosed in 1968 at the age of nine years. I had a bowel blockage, which passed after spending four days in hospital. My family physician made a note on my chart, “possible cystic fibrosis”, and I then spent six weeks at the Hospital for Sick Children in Toronto. There were the usual tests and x-rays, as well as the gold standard test of the day - the sweat test. The sweat test confirmed that I did in fact have cystic fibrosis. I was placed on a regiment of sleeping in a mist tent (later discovered to do more harm than good), nebulizers three times a day, and physiotherapy. My medications included enzymes, vitamins, Ventolin and an antibiotic. My lung function when I was diagnosed was absolutely normal. If I were born 15 to 20 years later, it’s likely I would have only been given enzymes. When I came home from the hospital I remember praying at night to the Lord, because I did not want to die.

I lived a regular life for many years, without nebulizers or puffers, staying healthy with just enzymes and vitamins. Unfortunately, everything started to change in 2010 when I was diagnosed with cystic fibrosis-related diabetes (CFRD). I began taking insulin to keep my blood sugar under control. In late 2012, I had a flu that would not go away and eventually turned into pneumonia. The standard procedure of treatment included three weeks of antibiotics. The pneumonia and infection cleared for a month, but then came back three more times. Finally, I had a bronchoscopy followed by another three weeks of antibiotics. Luckily, I did not have to be hospitalized during these episodes.

I felt well again, but had a little weight to gain back. However, in October 2013, I caught a cold that lasted and lasted, and pneumonia set in again. I was prescribed antibiotics, along with two nebulizers a day of hypertonic saline and antibiotic. The oral antibiotics did not help, and then came a slap of reality; I would have to be admitted to hospital at the age of 54 for the first time since being diagnosed in 1968. I was hospitalized for a week, with an IV antibiotic drip and underwent another bronchoscopy.

My spouse, Jacquie, died of cystic fibrosis, as well as many other friends - they all remain in my heart forever.

Since 1990, I have been a member of The Kin Club of Flamborough & District. I currently serve as the Risk Manager and Membership Director. I am blessed that I have been healthy for so many years and thankful for every day of life.

CANDID FACTS

FOUNDER OF CANDID FACTS: DONNA SUMMERHAYES

Donna and Doug Summerhayes, co-founders of Cystic Fibrosis Canada

Donna Summerhayes, co-founder of Cystic Fibrosis Canada along with her husband Doug Summerhayes, created *Candid Facts* in 1959. At a time before computers, the internet and email, Donna would sit at her kitchen table and hand-write each issue between raising a young family and caring for two children with cystic fibrosis (CF).

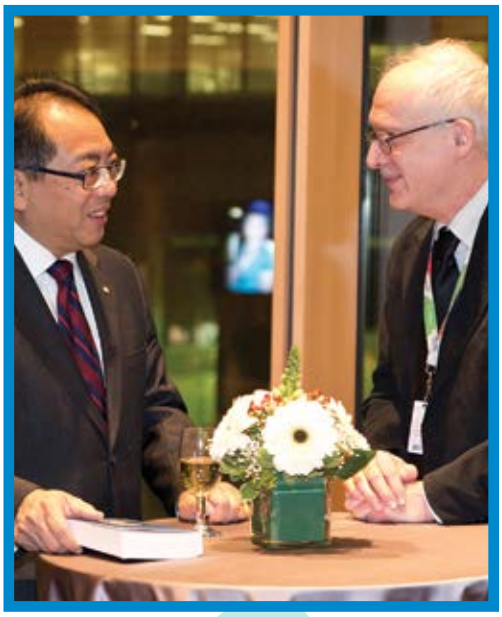
"Candid Facts bore the initials of the disease we are fighting and was about building hope, because I know that's what most of us were looking for as parents. It was also about building trust for those that were going to be volunteers and start Chapters. If we could build that trust, we could inspire and engage the community across Canada," says Donna.

In the 1960s, there was little information about the disease for parents and the first issue of Candid Facts went to almost 300 families living with cystic fibrosis to let them know they were not alone. Donna was instrumental in planting the seed for many of the communications materials about cystic fibrosis that we continue to share across the country today.

Candid Facts will now be featured as a special section of Cystic Fibrosis Canada's *Connections* magazine. We thank Donna for the legacy she created with Candid Facts and for crafting an impactful publication for many years that informed the Canadian CF community, and so many others, about cystic fibrosis.

LR: The first issue of Candid Facts; Pamela Summerhayes; the Summerhayes family in 1966; Donna Summerhayes

SICKKIDS 25TH ANNIVERSARY OF THE CF GENE EVENT



One of the key achievements in cystic fibrosis (CF) research that has played a major role in the health of people living with the disease in Canada, and around the globe, happened in 1989.

Cystic Fibrosis Canada and SickKids celebrated the 25th anniversary of the CF gene discovery. Dr. Lap-Chee Tsui, a Cystic Fibrosis Canada-funded researcher led the team at SickKids and helped discover the gene responsible for cystic fibrosis. More than 25 years later, this remarkable breakthrough is still considered one of the most significant advancements in human genetics in the last 50 years.

CYSTIC FIBROSIS CANADA LIAISON COMMITTEE

The Kin Canada – Cystic Fibrosis Canada Liaison Committee works with Cystic Fibrosis Canada to encourage involvement in fundraising and awareness activities from Kin Canada clubs across the country. Cystic Fibrosis Canada is Kin Canada's National Service Project, and finding a cure or control for cystic fibrosis is near and dear to the hearts of every Kin Canada member.

The committee is made up of appointed Kin Canada Service Directors representing eight regions across Canada, an elected Chair, the Kin National Service Director with guidance from staff members from Cystic Fibrosis Canada and Kin Canada.

Thank you to the 2014/2015 Kin Canada – Cystic Fibrosis Canada Liaison Committee on your hard work during your term as Kin Service Directors and for continuing to educate Kin members about cystic fibrosis, the work of Cystic Fibrosis Canada and motivating your fellow Kin to raise the crucial funds needed to find a cure or control for this devastating disease.

Davina Thuroo is completing her second-year term as the Chair of the Liaison Committee. Davina's dedication and support for the CF cause has been unparalleled. She has been a terrific resource to the Committee and we thank her for her energy, perseverance and passion during her two-year term as Chair. Davina will also take on a new role as the Kin National Service Director.

Helen Meinzinger will be the incoming Chair for the 2015-2017 term. We are excited to welcome the 2015/2016 liaison committee and wish them luck for the exciting year ahead.

THE KIN CANADA
CYSTIC FIBROSIS CANADA LIAISON COMMITTEE

	OUTGOING (JULY 1, 2014 TO JUNE 30, 2015)
Chair	Davina Thuroo
National Service Director	Sean Thompson
District 1	Tiffany Allan Dan Strugar
District 2	André Theriault
District 3	Charlene Duckworth
District 4	Cameron Kemp
District 5	Kelly Woywitka
District 6	Claude Legault
District 7	Jennifer Burry
District 8	Rhonda Croghan Erin Thomson

LR: Davina Thuroo (Outgoing Chair), Ron Hanishewsky (National President), Helen Meinzinger (Incoming Chair)

KIN CANADA TURNS 95

CF Canada's longest-standing national partner in the fight against cystic fibrosis, Kin Canada, celebrated its 95th anniversary in 2015. We would like to extend our congratulations to this incredible service organization for an outstanding 95 years of "serving the community's greatest needs."

Cystic Fibrosis Canada is proud to continue our 51 year partnership with Kin Canada. Last year, Kin Canada raised over $1.2 million for life-saving cystic fibrosis research and care through numerous 2014/15 fundraisers and the Great Strides™ walk.

Since Kin Canada's inception in 1920, they have contributed over $1 billion dollars to Canadian communities and disaster relief across the globe; over $42 million of these funds have been donated to Cystic Fibrosis Canada to advance the battle against cystic fibrosis.

Dedicated Kinsmen and Kinettes' commitment to Cystic Fibrosis Canada has been a tremendous driving force behind cystic fibrosis research, clinical care and advocacy advancements. We are honoured to have Kin Canada as our national partner and are proud to join them in celebrating a remarkable 95 years of positively impacting communities across the globe. Congratulations Kin Canada!

OUR LITTLE SUPERHERO **CHANCE**

BY ASHLEY AND GORDON WOOD

Chance Orion Wood was born on September 5, 2014. After many years of wishing for a beautiful baby, we were blessed with an amazing son. Chance was born a little early and weighed only five pounds 13 ounces. Within two weeks Chance had dropped to almost four pounds and we had no reason why.

We were soon informed that Chance was one of the rare babies with cystic fibrosis (CF). We weren't sure how this was possible as we were not quite sure what CF even was. We met with the CF team at McMaster Children's Hospital in Hamilton and after more blood tests, it was confirmed that Chance carried a double copy of the most common CF-causing mutation deltaF508. Thankfully this had been confirmed in his newborn screening test and Chance could start on medications right away. He was quickly prescribed enzymes and soon after started gaining weight. At two months old Chance started on his nebulizer and began physiotherapy.

At first we were both in denial. How were we as a family going to cope? Will Chance still grow up to be everything we ever hoped he would be? We felt an overwhelming amount of guilt and like many parents we wished that we could take it away and have it ourselves. We are incredibly lucky to have the most loving and supportive family and friends. They were willing to learn about Chance's medications and needs, and are there to help out whenever we need them. We also could not do it without our amazing team at McMaster. They have helped us every step of the way and we could not be more thankful for everything they have done.

Chance is our little superhero! All superheroes are unique – some need gadgets, and some need to charge their powers. Chance has enzymes, his mask and "super pats"! With these Chance can spread his love and cuteness to the world. He is now a strong and happy six month old. He knows that he needs to take his enzymes before he eats and he even holds his mask himself.

One of Chance's favourite places is the martial arts school that Gord and I own – *Evolution Martial Arts* in Ancaster. Everyone knows Chance at the school and he loves to watch the kids train. He has even won a gold medal at a karate tournament for cuteness! We cannot wait for Chance to get in there and start training when he gets older.

We strive hard at our martial arts school to teach our students about respect and accepting others for who they are, and our students have outdone themselves. Some of our students have even gone above and beyond. Jeremy, a black belt, recorded himself playing Christmas songs on a piano and sold the CDs, donating all the money to cystic fibrosis. Mackenzie and her brother Austin put on a play for family and friends for donations to Cystic Fibrosis Canada. Isabel, a black belt, put on an assembly at her school to talk to the students about what CF is and what they could do to help. Isabel's school also had a dance to raise money for CF research. We are amazed what children are capable of doing and are lucky to have them supporting our son.

As black belts ourselves, we try to see the positive in every situation. Every day is tough – tough to make time for physiotherapy twice a day, feeding more often and tough to know that everyday our son struggles to breathe. We treat every day as a special one and enjoy every moment no matter what the day brings. Chance is our son, he is our *everything*, and we will do whatever it takes to keep him healthy.

Although CF is a fatal disease, we hope that one day CF will stand for "cure found". Thanks to research and donations we will be able to see our son grow into an amazing man and reach all of his dreams and desires.

A SECOND CHANCE AT LIFE: MICHAEL'S STORY

Kathryn and Glen Davis remember their heartache and confusion when they adopted their son, Michael, a beautiful baby boy, was diagnosed with cystic fibrosis (CF) in 1990 at eight months old. Due to the late diagnosis and having some health complications, Michael had to spend several weeks in hospital. Kathryn remembers that it wasn't until their first clinic visit at SickKids a few weeks later that they learned about Cystic Fibrosis Canada and the advancements that were happening in CF research and care.

"We left the CF clinic feeling quite scared, but also very hopeful. Dr. Lap-Chee Tsui had just led the discovery of the gene responsible for cystic fibrosis and there was a lot of hope that a cure or control for CF was within reach. "

Glen and Kathryn brought Michael home from the hospital along with the task of caring for a new baby with CF; daily physiotherapy, enzymes with each meal, and managing all of the medications. Soon after, Kathryn gave birth to their biological son, Scott, who arrived six weeks later. Glen and Kathryn now had the added challenge of caring for their healthy newborn while also caring for Michael.

Michael was fairly healthy until he reached grade 5 when he started having more serious digestive issues and required a feeding tube to help him gain weight. For the next few years, Michael had to be hospitalized for weeks at a time. Michael's health started to decline as he developed more serious lung infections.

In 2006, when Michael was 16 years old, his health deteriorated even more and he required a double- lung transplant. The operation was successful, but Michael experienced some setbacks with organ rejection.

"We felt very blessed. Michael was given a second chance at life and a lot of people with CF don't get that chance". Michael has since graduated from college and is living and working in Kitchener, Ontario.

Since the Davis family learned about Cystic Fibrosis Canada at their first clinic visit, they have been tremendous donors, volunteers and fundraisers for the organization. They have encouraged their friends and family to volunteer with the Toronto & District Chapter, and the Davis' also fundraise and participate in CARSTAR's Great Strides™ Walk for Cystic Fibrosis Canada. Glen has organized an industry golf tournament for the past 10 years to raise awareness and funds for Cystic Fibrosis Canada. "Over the years, my colleagues and associates have gotten to know our family's connection with CF and have seen the great advancements in CF research and care. They have remained committed to supporting the cause and so we continue to donate the proceeds from our industry golf tournament to Cystic Fibrosis Canada."

Since Michael was born, the Davis family has seen tremendous advancements in research and care that are helping people living with CF, like their son, live longer and healthier lives. But more work still needs to be done, as too many lives are still cut too short because of cystic fibrosis.

"We continue to fundraise and volunteer for Cystic Fibrosis Canada because it helps bring us closer to finding a cure or control for CF. We want to help make sure that all children born with CF are able to live a long, happy and healthy life; free from the struggles of living with cystic fibrosis."

KALYDECO®: WHERE WE ARE NOW

When KALYDECO® first entered the Canadian market via a Notice of Compliance from Health Canada on November 26, 2012, Cystic Fibrosis Canada and the cystic fibrosis (CF) community - individuals with CF, their loved ones and CF clinicians, began a 19 month advocacy campaign for access to this life-changing therapy. At this time, KALYDECO® was only approved for use in individuals with the G551D gene mutation ages 6 and older. This mutation on the CFTR gene affects four percent of the CF population, but this did not stop the CF community from supporting the 115 individuals who could benefit from this therapy.

After several months of negotiations between the drug manufacturer and the Pan-Canadian Pharmaceutical Alliance (PCPA), an agreement for KALYDECO® was reached. Ontario became the first PCPA province to list KALYDECO® on the public drug formulary for individuals with the G551D mutation. Today, seven provinces and one territory have made KALYDECO® available for individuals with the G551D gene mutation, including: Ontario, Alberta, Saskatchewan, Nova Scotia, Manitoba, New Brunswick, Yukon and British Columbia.

In June 2014, Health Canada approved the extended use of KALYDECO® for nine additional gene mutations in individuals with CF ages 6 and older, including: **G178R, S549N, S549R, G551S, G1244E, S1251N, S1255P, G1349D and G970R**. Similar to KALYDECO® for the G551D gene mutation, Cystic Fibrosis Canada submitted impactful patient input to the Common Drug Review. In December 2014, the extended indications were given a positive recommendation by the Common Drug Review. The provinces are once again negotiating an affordable price for KALYDECO® through the PCPA, with Alberta leading negotiations. Cystic Fibrosis Canada is in regular contact with officials involved in these discussions and encouraging them to reach an agreement as soon as possible. Both parties have voiced their commitment to accelerated discussions.

In the United States, the Food and Drug Administration (FDA) has recently approved KALYDECO® for use in children ages 2-5 for the mutations mentioned above. The FDA advisory committee, made up of independent medical experts and community representatives, recommended that the FDA approve the ivacaftor/lumacaftor combination therapy - trade name ORKAMBI™, that targets the underlying cause of the disease in people with two copies of the delta F508 mutation in the *CFTR* gene. The FDA decision is expected on July 15, 2015.

Health Canada approved KALYDECO® for use in adults with the **R117H** gene mutation in March 2015. Cystic Fibrosis Canada submitted patient input to the Common Drug Review on May 22, 2015.

For more updates on KALYDECO®, Health Canada, the Common Drug Review, the pan-Canadian Pharmaceutical Alliance and how medications are reimbursed, please visit Cystic Fibrosis Canada's Advocacy Blog at www.cysticfibrosis.ca.

To submit a testimonial for ORKAMBI™ or homozygous delta F508, please email advocacy@cysticfibrosis.ca.

CHAPTER OF THE YEAR GOES TO....
CALGARY AND SOUTHERN ALBERTA CHAPTER

The Calgary and Southern Alberta Chapter was the recipient of the Fred Blizzard Chapter of the Year Award at Cystic Fibrosis Canada's Annual Meeting and Conference in April.

The Calgary and Southern Alberta Chapter is the recipient of the Fred Blizzard Chapter of the Year Award

Demonstrating high standards of growth in fundraising, volunteer development, partner recognition, public awareness, and social media campaigns, the Calgary and Southern Alberta Chapter ranks as one of the top five Chapters across Canada with gross revenues of over half a million dollars annually.

A selection of the Calgary and Southern Alberta Chapter's successful fundraising events include: Wrapping for a Cure, 65 Roses Ladies Golf Tournament, the 65 Roses Princess Ball, Ride for Breath of Life, North Hill Calgary Cake Off, and partnerships with CARSTAR's Great Strides™ Walk for Cystic Fibrosis Canada, GearUp4CF, TJ Brodie's Skate in Strides program and Lawn Summer Nights. The Chapter has also cultured a startup with Shinerama at the University of Calgary, Medical Faculty while supporting their first campaign this year.

The Calgary and Southern Alberta Chapter has fostered partnerships that have continued to grow with Kin Canada, CARSTAR and TJ Brodie's Skate in Strides campaigns. Strong sustained growth in these areas has broadened and strengthened the Chapter's volunteer base and created new local events. A focus on social media to promote the CF cause has informed the public on varying issues of life, and living with cystic fibrosis at any age.

Congratulations to the Calgary and Southern Alberta Chapter!

CYSTIC FIBROSIS CANADA'S QUEBEC CHAPTER HOSTS PRESTIGIOUS FIVE STAR DINNER

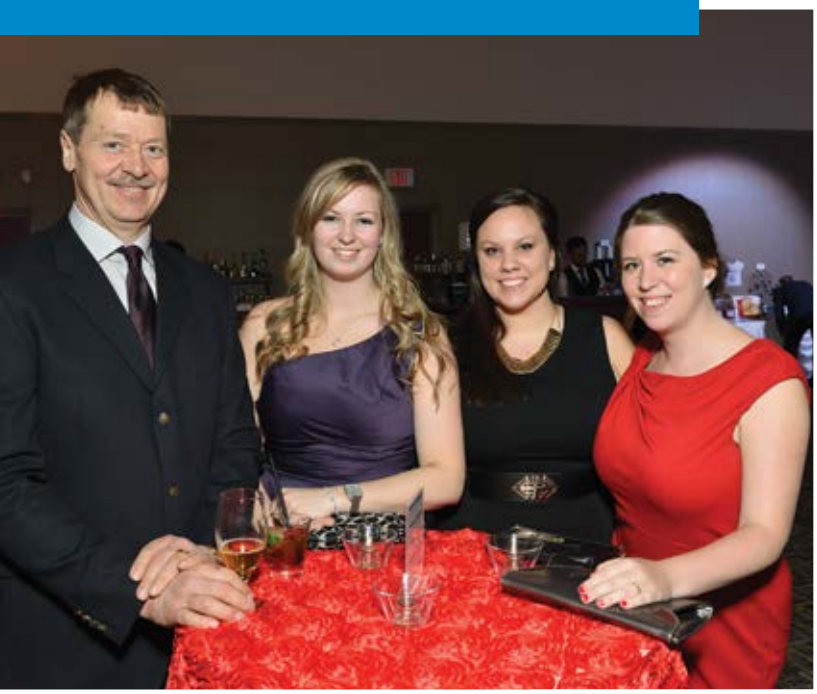
Last year, Fibrose kystique Quebec started a major gifts program launched with a prestigious fundraising dinner event sold at $1,000 per person. In attendance were members of the *Cercle des Gouverneurs*, an exclusive club for business executives in Quebec. The guest list boasted of notable people including Louis Vachon, President of the *National Bank*.

Marc Giroux Vice-President Marketing at Metro, Daniel Lalonde Senior Vice-President at National Bank Financial, Louis Vachon President of National Bank, François Castonguay CEO of Uniprix

This year's five star event will be held on June 17, with guest speaker, Gaétan Frigon, former *Dans L'œil du Dragon,* show host (Quebec's version of Dragon's Den). As an avid supporter of the cause over the years, Mr. Frigon will be speaking about his mentorship of a young adult with cystic fibrosis.

The event will be taking place at the Mount Bruno Country Club in Saint-Bruno-de-Montarville. Thanks to the event sponsor, Vice President of the National Bank, Daniel Lalonde, all ticket purchasers will be eligible for a tax receipt.

65 Roses CYSTIC FIBROSIS GALA



The 16th Annual 65 Roses Gala took place on February 28 in Ottawa, ON. The theme of this year's Gala was "Red is the New Black" which raised $105,000 for CF research and care. With over 300 guests in attendance, including five celebrities, guests enjoyed an entertaining evening that included a silent auction, a Key to a Cure, a Bail or Jail auction, fantastic food and inspiring speakers.

Since its inception, the Gala has raised upwards of $1,000,000 dollars to help fund vital research and patient care for the over 200 families living with this disease in the Ottawa area.

DREAMING OF THE ORDINARY: FIFTY YEARS OF CYSTIC FIBROSIS

BY ALLYSSA GRANT

Life expectancy has chased my uncle Bob Costerton. With it constantly at his heels for the past 50 years, Bob has watched the predicted median age of survival of a person living with cystic fibrosis (CF) grow from less than five years to its present all-time high at 50.9 years of age, among one of the highest in the world.

For Bob, there will be 51 candles on his birthday cake this year, and he has had to fight for every single one.

Sitting across from Bob, you would never guess he was diagnosed with CF. A tanned picture of health, to a passerby Bob's lean physique and smile lines simply look like a life well-lived, not the diagnosis his parent's received when they were told he *might* see his 4th birthday.

"I have always wondered what is possible for my life," says Bob.

"Could I have a happy life, live to adulthood, go to university, fall in love? Could I be a father?"

Few of us dream of living "just" an ordinary life, but for Bob dreaming of the ordinary life has empowered him to fight. And fight he has, for every single breath.

As an adolescent, Bob recalls bringing home a school project and telling his mom that he was researching cystic fibrosis. Bob's mother later confessed a feeling of dread because he might find out he was already on borrowed time.

In 50 years, Bob has watched that fateful number steadily grow. Because of generous donations, CF researcher's steadfast efforts have evolved our understanding of the fatal disease.

"Sometimes I think my life is owed to good timing," says Bob of the arrival of new CF treatment and therapies just as his condition demanded it.

While the timely arrival of this research undoubtedly added candles to his cake, Bob is known for his unwavering dedication to life that has truly influenced how his story is written.

Bob must push his lungs to their limit every day. For the more than 4,000 Canadians living with cystic fibrosis, daily exercise and treatment are necessary and life-saving measures. Regular airway clearance helps prevent a build-up of mucus in the lungs.

And so, Bob decidedly battles. A war waged mostly upon the pedals of his bike. Ascending the hills of Kamloops day after day he challenges his lungs – and his diagnosis.

But sometimes his rigorous coughing attracts unwanted attention. An opponent once commented during soccer that Bob really should see a doctor for that cough. When the player had been reassured that a doctor had been consulted, he said, "You should see a different doctor!"

"If he only knew…" laughs Bob.

Aptly named the invisible disease, there is little outward evidence of CF in Bob's life (that is until you open his car's glove box to find his plethora of medical treatments).

Many of the items on Bob's bucket list have been realized. A successful engineer and entrepreneur; Bob lives in Kamloops, BC with his wife Kathy, his son Liam, and daughter Nicola. Their home is scattered with the evidence of countless family trips, but the richness of Bob's life manifests itself in his garage. Here you will find this talented family of four's hiking boots, bikes, skis, and 19 canoes and kayaks – Bob's wife Kathy makes note that the boats are all Bob's.

Yet, on Bob's bucket list there remains a few items unchecked.

"I didn't want to share my story until it had a happy ending," says Bob, and while his story isn't finished yet, he has finally chosen to tell it for the first time.

The mother of a son and daughter, both diagnosed with CF, recently reached out to Bob because her daughter had succumbed to the disease, and her 17 year-old son was struggling.

"I don't want CF patients to have to live with a number looming over their head counting years,"

Bob calls the young man with CF. He tries to tell him, it's worth the fight.

"Did I get through to him?" questions Bob. "I don't know."

And, Bob has decided that now is the perfect time to cross one of the last items of his list.

"I don't want CF patients to have to live with a number looming over their head counting years," says Bob. "I want them to have every opportunity to live."

"I've tried to never let the disease define my life," says Bob.

Staying alive for Bob has always been mission impossible – but with countless donations to research, Bob's will, and the help of others it became mission possible.

This time Bob faces a new mission impossible – by choice.

Bob has decided to take on the 1,200 km, 9-day cycling marathon from Vancouver to Banff, called GearUp4CF in support of CF research. At 50 years old, he is the oldest person with CF, with or without a lung transplant, to attempt this feat.

This challenge makes one thing clear: Bob is living one extraordinary, ordinary life.

For more information about the GearUp4CF event raising funds for CF research and care and to sponsor a rider, please visit: www.cfchapters.ca/vancouver/gearup4cf.

Decant

GALA & FINE WINE AUCTION

The Decant Gala & Fine Wine Auction – An Evening in France, took place on May 2, 2015 at the Delta Toronto Downtown. The event was hosted by Ben Mulroney, anchor of CTV's ETALK and National Ambassador to Cystic Fibrosis Canada. The Gala raised an incredible $365,000 for cystic fibrosis research and care.

Guests enjoyed an elegant evening, which was akin to a walk down the Champs-Elysées, complete with sampling vintages from the most famous wine regions in France. The gala was highlighted by performances from French Can-Can dancers and a Parisian opera singer. Keynote speaker included Chris MacLeod, an inspiring adult living with CF. A live silent auction that featured "a Key to a Cure" helped leverage important financial support for Canadians fighting this devastating disease.

Cystic Fibrosis Canada™

www.cysticfibrosis.ca

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