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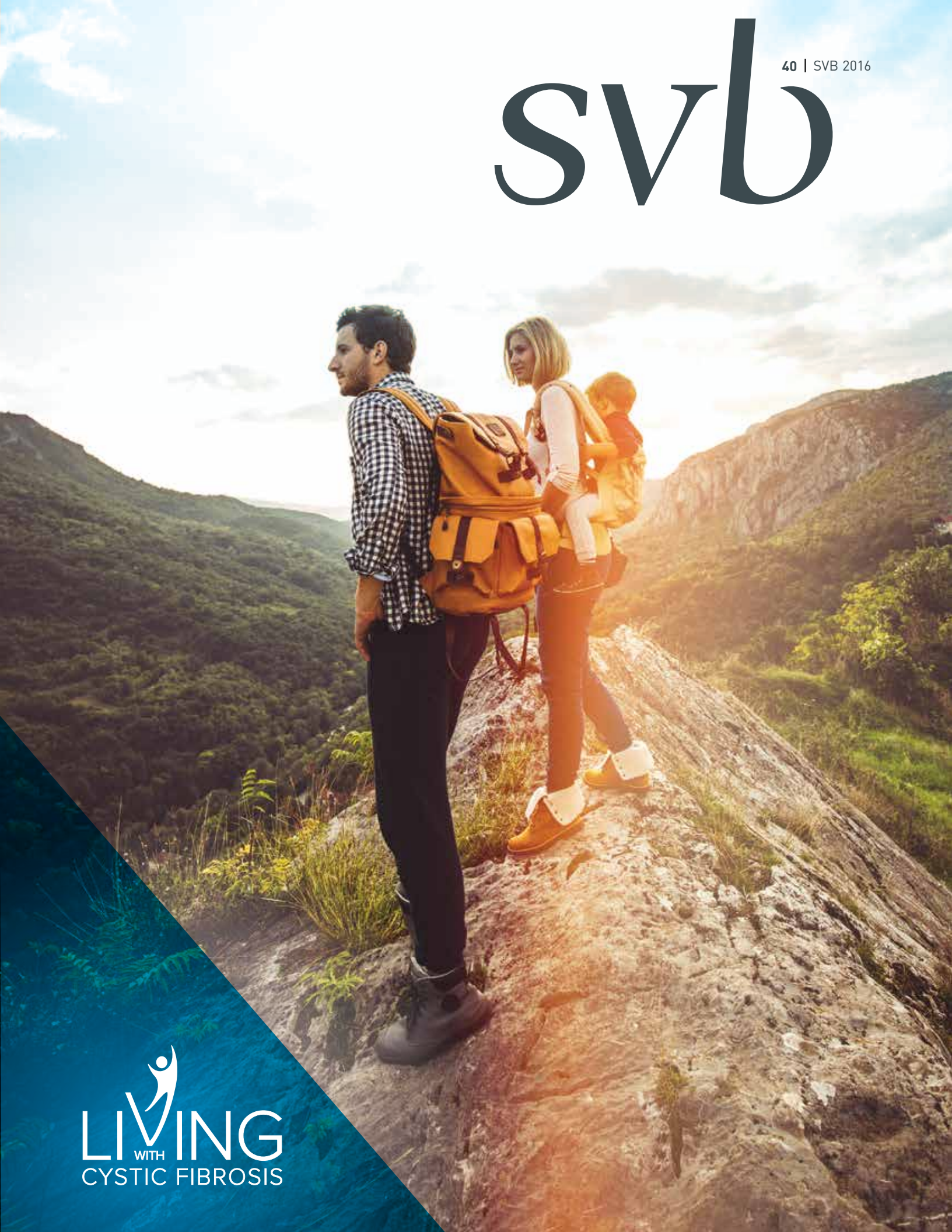

LIVING WITH CYSTIC FIBROSIS

Living with Cystic Fibrosis would like to thank Vertex for its generous contribution to the production of the 40th edition of *SVB*.

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Printing
FJDG
Montréal, Quebec, Canada

This magazine is produced by ***Living with Cystic Fibrosis*** *through participation of its sponsors.*

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February 2016

Bibliothèque nationale du Québec, D9150134
National Library of Canada, D411325D

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Cystic fibrosis is a hereditary disease that affects the lungs and digestive system as a result of poorly functioning mucus glands. Although research has made tremendous headway, it has not yet come up with a cure or a way to fully bring the illness under control.

Problems with mucus glands cause problems with digestion and lung function. Mucus is normally fluid and helps keep the lungs and airways clear by helping to eliminate germs and dust particles. But the mucus in cystic fibrosis sufferers is thick and sticky and obstructs the bronchioles, making it difficult to breath. Air ends up getting trapped in some bronchioles and the lungs become clogged up with mucus. This is turn causes bacteria to accumulate and multiply in the clogged bronchioles, leading to infections.

Treatment for cystic fibroses mainly focuses on pulmonary problems, given that the mortality associated with the illness is primarily related to lung infections and lung function. Lung treatments are aimed at loosening up and expelling the mucus blocking the airway in order to improve pulmonary operation. In many cases, frequent treatment by antibiotics is needed in order to get the upper hand on the many infections that develop in the lungs. In very severe cases, the lungs can get so badly damaged as a result that a transplant is the only solution.

Mucus also blocks the tiny ducts of the pancreas, an organ located beneath the stomach, near the intestines. The pancreas secretes enzymes that are normally delivered to the small intestine to help digest food. When the openings of the pancreas are blocked by mucus, these enzymes can no longer reach their destination and food leaves the intestines only partially digested, depriving the body of a portion of the food's nutritional value.

Cystic fibrosis symptoms appear early on in life. In some individuals, they first show up in the lungs; in others, the digestive system. However, most people with CF, whether children or adults, suffer from both types of symptoms.

"The world belongs to optimists, pessimists are only spectators"

- François Guizot

It is with renewed pleasure this year, as it has been for the past 30 years now, that we send you the *SVB*, our annual publication. Without a doubt, 2015 was a pivotal year in the world of cystic fibrosis. We began the year by learning that the median survival age was 51 years, we ended it by reaching a record of lung transplants in Quebec. This year, more than 57 people underwent lung transplant surgery at the Notre-Dame hospital, in Montreal. Both these aspects were incredible advances!

On October 17th, we celebrated our 30th anniversary. Over the past 30 years we developped a unique organisation. We now live longer and better than ever. Better because people living with CF in Quebec gave themselves an organisation that protects their interests and promotes good habits and forwards useful and trustworthee information to the community. We've built links with many national partners like the Fondation l'air d'aller and Fibrose kystique Québec in order to have accessible programs and projects specifically built for us. We have even built relations with organisation from other countries to share knowledge of how to live with CF around the world. Well served by research and the healthcare system which takes charge of us in almost all aspects of our lives, we can count on an ever present psychosocial support.

However, 2015 was not perfect in the healthcare world in Quebec. This year was rough on everyone, and important changes occurred.

Even though Quebecers with CF can count themselves as being amongst the most privileged in the health care system, 2015 was a dire year for our healthcare system and social services. Budget cuts have taken their toll over the years. In December, thanks to the *National Report on Incidents and Accidents Occurring During Healthcare Acts*, we learned that 316 people died in hospital in 2015 because of falls and medication errors. It should also be noted that fusions imposed by Bill 10 forced administrators to take care of many departments at the same time, don't allowing them to take the time to well understand their departments and correct problems.

We like to think about what we have gained during the last decades, and that everything is owed to us. However, it is imperative to remain vigilant in order to protect our gains. For example, free access to in vitro fertilization was lost to most of us without warning.

In such a context, it is indispensable for us to thank all the doctors, nurses, social workers, psychologists and other interveners from the bottom of our hearts. Every day, they have taken care of us and accomplished miracles.

The 2016 edition of the *SVB* carries hope, but remains realistic. We live longer, and thus we require more support. The new transplant recipients cannot escape this reality, as you will see in the following pages. Solutions will have to be found to give even more hope to those who have gone through this harsh period of their lives and allow them to live their dreams.

Living with Cystic Fibrosis will face many challenges in the following years. Thanks to the implication of many, we have realised our mission against impossible odds. An old African proverb says that alone we travel faster, but together we travel further. That is exactly what all of us, together, have accomplished. Sometimes slow, but concrete and efficient realisations.

Our healthcare system and social safety net are a legacy that Quebecers built for themselves because we believe that everyone deserves a good quality of life no matter their origins, skin color or social standing.

Our organisation is the gift that we gave each other was Quebecers living with CF. It is unique in the world, and thus even more of a precious gift.

Let's be proud to continue building a better life for those that live with Cystic Fibrosis, and a better life for all of us.

Tomy-Richard Leboeuf McGregor

Editor

Bill 20
the end of a dream

Charlène Blais, Chair of the board of directors
Magog, Quebec, Canada

The liberal government adopted the Quebec assisted procreation program in 2010. This program covered part of the costs linked to infertility treatment. Due to this program, between 2010 and 2015, a very high number of children were born, fulfilling the dream of couples that thought it was impossible to start a family.

On November 11th, the liberal government adopted Bill 20 greatly limiting the state's support given to infertile couples.

The Bill's language ended free treatments to replace them with tax credits for low income families. « Families with an income lower than 50,000$ per year will receive a tax credit equivalent to 80% of the cost of treatments. Progressively, this tax credit will lower to 20% of the cost of treatment for households making more than 120,000$. However, no tax credit is available for couples that already have a child »

When it is considered that 99% of men and 50% of women living with cystic fibrosis suffer from infertility, it is clear that this law directly affects our community.

Living with Cystic Fibrosis is having to live a certain amount of losses at different steps in our lives, but, with the Quebec assisted procreation program, the dream of starting a family became possible for some of us. With Bill 20, the load of the loss of being able to start a family is added as, for some of our members, the tax credit is not sufficient enough to undertake this step.

Mr. Couillard, at the next election, the CF community will remember this law adopted on Remembrance Day.

Each time I hold my two miracles born through the Quebec assisted procreation program, I think of all the infertile couples that won't have my luck, and I am frustrated. ◄

[1] http://aciq.ca/projet-de-loi-20/

Post-transplant reality... **AKA the dark side of life after a transplant**

Alex Danis
Montréal, Quebec, Canada

No, the TRUE reality of life after a transplant is not one with rainbows, a beautiful sun and an incredible future, as we all believe.

In January 2015, I received new lungs. It is a gift I appreciate above all else and I am happy to be alive, to do activities that I never thought I could accomplish before and to fully enjoy life for the first time.

But my reality, like the one of many transplant recipients (especially young ones), is sadder than you think. Behind the curtains, the situation is difficult. Even if health is finally there, it is a tough task to recover.

I am talking here about money, debt, costly medication and psychological problems that follow.

I am talking about someone who, like me, was affected by cystic fibrosis before a transplant and has spent a lot of time in hospitals, in bed, or simply at home for treatments. Before the transplant, the illness and its treatment took a large part of our day and energy. If the target age is 20 to 30 years old for successful transplants, that time and energy would be used by a normal person to study, work, save, gain pertinent experience and grow in the job market while that time and energy is foremost for a recent transplant. For us, this translate into a constant inability to work the 40 hours a week needed to subsist, having to leave jobs, university/college courses or semesters for hospital stays. The schedule is sometimes too intense for our body to cope. These situations bring us to never being perceived as worthy of a promotion in an organization - due to our unpredictable health status.

With all this, debt accumulates and accumulates again. The credit rating becomes worse and worse as debt becomes the only solution. After months or years of going from job to job, classes to classes without any concrete experience for references, the transplant arrives. Life finally arrives. Yay! Oh well no, just no.

No work for another 6 months -MINIMUM- after the transplant. Six months with no revenue, six months of getting by, finding money here and there, six months of reliving shame to ask your parents to support financially, when you are supposed to have become a 27 year old « grown ass man ». And this deteriorates your relationship. Debts, and debts, continue to grow. They become even worse! Some will declare bankruptcy after a transplant. That is the reality of many.

Psychological problems arise, even more so than before. Anti-rejection drugs often cause many new psychological symptoms or increase those already present. And the continuous stress of your financial situation brings out all your demons. In my case, it was compulsive buying disorder and made the situation 1,000 times worse. I know others for whom it was drugs, alcohol or something else...

You find yourself happy to be alive, but the reality hits you back in the face. That is why research has shown that the suicide rate for transplant recipients is 15.7 persons out of 100,000 a year... in comparison with 9,0 for 100,000 for the general population, which is extremely significant.

Being 27 years old and having the experience of an 18-year-old kid on your resume is not motivating to an unthinkable point. Finding yourself at 27 with 12,000$ in debt, without counting student loans if you want to pursue your studies, is pride that crumbles. The future you always wanted after the transplant seems further and further away.

Also, after having spent your life going through all this hardship, the last thing you want is to having to kill yourself at work in a fast food joint for minimum wage. You are finally free for your hospital jail, you don't want to lock yourself in another jail in the form of a kitchen or cubicle. So you cheat. You ask your family and friends to be fake references for fictitious employment that you held. You overstate the time you kept your jobs and the tasks you had to accomplish. You try to recreate on paper the person you would be today were you not born with the illness. However, deep down, it is humiliating. It is auto-humiliation.

You want to be professional, you are ready to work, but, because the illness always slowed you down, you must start again at the beginning of the race while those around you are at the finish line.

So you make yourself a mask. You don't talk about it. You keep it to yourself because the people around you are happy for you and you don't want to break that. You want to keep the image of the person who won against death and for whom everything rolls smoothly now. But inside, you stress. You cry. You are anxious. You are depressed. You ask yourself whether it was worth it to have had the transplant, if it is to live in this way.

So this is it, the dark side of living after a transplant.

We should talk about it! ◄

EDITOR'S NOTE

The publishing of this article filled with despair seemed pertinent and necessary given the studies done on symptoms of stress and depression in persons affected by CF. Do not hesitate with your CF clinic's specialists if you feel those symptoms. You can also send us your comments regarding this article at this address: info@vivreaveclafk.com

I also wish to personally thank the author of this gripping article and to restate my support and that of Living with Cystic Fibrosis to those who suffer at this moment. May our support and that of your relatives help you in the immense task of full recovery.

Grandma for life

Francine Bernier
St-Jean-de-Matha
Quebec, Canada

A little girl came to the world. Anticipated, expected, desired: Finally - she's here!

I'm Grandma to this little girl called Odile and I call her "my pitchounette".

My Grandma heart is filled with immense happiness. I cannot wait to traverse all those kilometres that will lead me to her.

I think of my son and my daughter-in-law, and I am so happy for them. They are dad and mom for the first time. All hopes and dreams are open to them. The future is bright ...

June 24: The phone rings. Crying, my son tells me that Odile is very ill. After an emergency operation, a terrible diagnosis fell upon us. Odile suffers from cystic fibrosis: a fatal and incurable disease that attacks the lungs and digestive system.

Phew! At the other end of the line, what do you do? What do you say? I feel so helpless ... You cry with your son and encourage him as best you can ... Odile is at the Ste-Justine Hospital. She will spend several days in intensive care, and will be in intermediate care for several days afterwards. When I can finally see her - all hooked up - it's a little angel who is placed in my arms, and at the same time also a great fighter who has just fought her first battle for survival.

Since Odile's birth I have been a witness to the many battles that she has fought against her illness, of the great suffering that she endured, of fears, worries and anxieties. I'm also a witness to her great victories against the disease, to her hopes and her dreams. It's undeniable, Odile is a great fighter. A small happy and cheerful girl who loves life.

Odile was still a baby when my son and my daughter taught me how to do "clapping". This is a method of accurately hitting several key places to dislodge the thick mucus that accumulates in the lungs. This was a priceless gift. They trusted me and each session is a moment of grace that I share with her.

Odile will be 12 in a few days. I'm her grandma and sometimes I feel very small in front of this child of exceptional maturity.

I am so proud of my pitchounette!

I wrote this text 5 years ago. What has happened since?

Odile became increasingly ill. Her hospital stays became more and more frequent until the bi-lung transplant that she miraculously received in 2015, which radically changed her life. She was only 15 years old!

She's come so far … She's made so much effort to be who she is now: a beautiful and joyful young woman, happy, always smiling; a young woman who loves life, her studies, her friends, and most of all, her family.

Odile was never alone in her fight, even in her moments of great darkness. When a serious disease settles, it becomes the disease of the family.

There was her immediate family: dad, mom and two little sisters.

If Odile is strong in the face of her illness, and what follows from it, it is because she draws that will and energy to never give up from her parents. Always she is able to hold on to them. Trust, hope, determination and will are attributes from their daily lives. Sorrows, anxieties, and fears are experienced within the

intimacy of family, which becomes a place of healing.

Odile's little sisters have always known her to be frequently sick and suffering. Before Odile received her transplant, each painful experience their big sister went through made them sad, worried them, raised questions always answered by mom and dad.

They have suffered with her, they were afraid for her, they kept hope with her. Meanwhile there were moments of great happiness experienced daily, these never-ending bursts of laughter, complicity between sisters, and this need for the presence of the other.

There is also the family of the family:

Grandparents, uncles, aunts, cousins, who love and are worried… and who are afraid ... All have seen her in extremely painful states. That was before the transplant!!!

I am one of the two grandmas of Odile. I suffered with her, I hoped with her and prayed for her. But I am primarily a mom. Through my son, I endured the suffering in silence and impotence.

It will be 1 year this spring that Odile was grafted. She lives well, post-transplant. It is a period of calm all can enjoy to the greatest extent. Whatever future bridges Odile has to cross, all of us, her big family, will cross them again with her.

THIS is what family is. ◄

One kilometer at a time

Sophie Jacob
St-Eustache, Quebec
Canada

Hi, my name is Sophie Jacob, a 34 year old happy and young woman, truck driver and social network media facilitator. I am also the author of the book *Un kilomètre à la fois* (One kilometer at a time). I received a diagnostic of cystic fibrosis at the age of 6 months, and have succeeded to overcome the prognosis to enjoy my presence in this wonderful universe. However, the path wasn't always clear…

After incredible teenage years, I became an agitated woman. I had many dreams to fulfill, but as a victim of cystic fibrosis, I couldn't find means to achieve them or to develop myself. I could not live like other women my age. I was single, unstable and without a job. I was also angry, infuriated from many failures and from spending too much time in the hospital. I didn't accept being born under a bad star.

In 2006, my depressive state and my behaviour brought me to consult doctor for other reasons besides CF. After meeting for a few minutes with a psychiatrist, I was diagnosed as bipolar. The reason was very simple! It had been years that I had been living ups and downs, moments of exhilaration interrupted by moments of depression. Life wasn't easy, especially with CF.

From this moment, a new fight was added, to understand the meaning of my pain and to adjust my new medication so as to not live as a vegetable. Each meeting with my psychiatrist made me more depressive, bringing me back to the past and to bad memories. My soul was dying inside a sicker and sicker body.

One beautiful morning, as I was watching my life pass before me while twiddling my thumbs, a thought infiltrated my head without welcome. It constantly repeated: "You are a victim of your misfortune". It was long, however I ended up understanding its meaning… The only reason that I was dead inside a sick body was because I was convinced that my life summarized to this.

This revelation brought me something new, conscience, then ambition. I was now decided to change my way of conceiving my life, and to further fulfill my dreams. I was to stop being scared of being sick before being sick, and I was to live. It was no easy thing, but facing each obstacle I told myself "one step at a time" regardless of the step's height. When I fell, I took a breath and rose back up. When others held me back, I fled… right ahead! My decision was taken, I was going to respect the commitment I had taken from the bottom of my heart. I was to become a truck driver. However, this long path forced me to do great introspection…

I realized in my progression that, because of the many bereavements that we must regularly face with CF, I had accumulated much sadness and sorrow. Because of failures, and sometimes rejection, I had accumulated frustration and deception. Because I was sick, I saw myself as an imperfect human being. But heck! Are there any perfect humans? My perception of life was wrong and it wasn't because I was bipolar. It is because I was emotional and I didn't know how to channel my emotions.

I also realized one thing in my healing process, it is that emotions don't result from situations but from our interpretations and experiences. When a person compares to me and say: "Oh, I should stop complaining, what you live with CF is far worse", it is completely false! The emotion that results from their obstacle is the same as mine. If that person is not able to channel that emotion adequately, their pain can make them suffer far more than I suffer from CF!

What is different is not CF or the obstacle, it is our capacity to interpret adequately and to rise again! The fight is not physical… The fight is inside and is lived everyday, by everyone.

By being conscious of that, I had no choice but to cease seeing me as a victim of CF. I also changed my perception of others. If I, when I am emotional and about to erupt, consciously or unconsciously, can change my perception of people and situations by changing my behaviour to protect myself… then it is the same for others! We are all human. Our reactions to situations do not depend on whether we are good or bad! They result from our experiences and also from our interpretation.

From that moment, I started forgiving to those who didn't have a harmonious behaviour with me (without nevertheless keeping them in my life). I forgave to those who hurt me, and I forgave myself for the hurt I caused. We all want to be happy, but it a complex endeavour that can become confusing if our emotions mix into it.

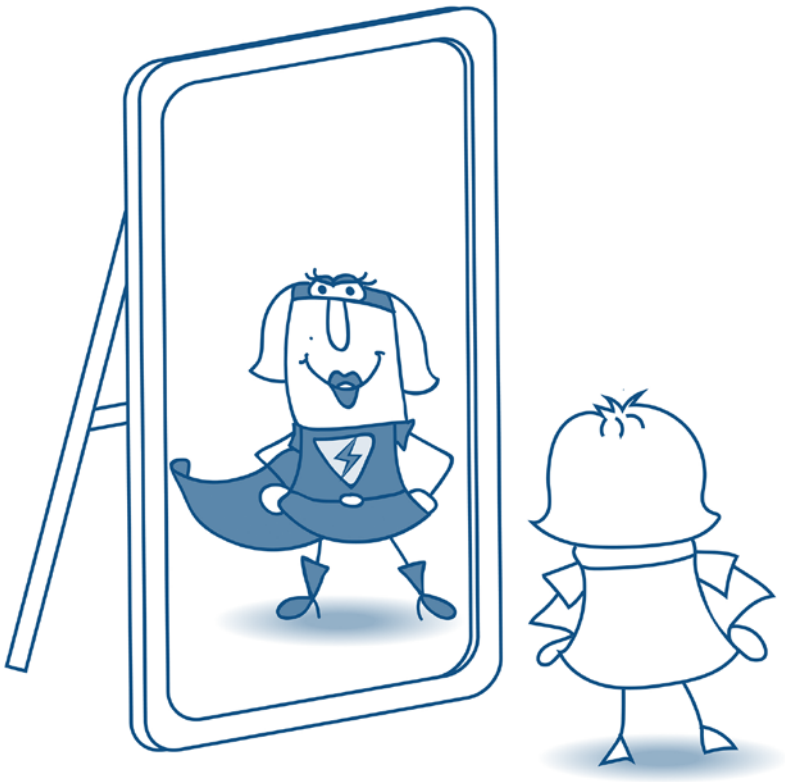
Today, I am much more calm and serene. I do not take things in a personal manner. I also give myself the right to express myself clearly, because I am not perfect, I am human. I succeeded in ridding myself of my heavy negative emotions, and my arms are now free to welcome happiness. Above all, it is now easier to take decisions, and to race to do the things I love, even with CF

I take moments one at a time, minute after minute, whatever they are. I do not anticipate tomorrow's hurdles, because life is lived today and is full of surprises, opportunities and twists. I allow myself to put a knee down when I am out of breath, but always to rise back up. My head is full of projects, and my heart is ambitious to succeed and to give. I have not achieved that with better physical health, but rather with a more positive mental flow.

No matter the obstacles or the disease to fight, victory comes from within. Love and happiness come from the heart, not from your feet or from your neighbours…

On the eve of 2016, I realize all the things I have accomplished in the last 10 years. I have fulfilled my dream of becoming a truck driver, I have crossed almost all American states and Canadians provinces, I have travelled, I have written a book, I have an understanding and outstanding employer, and extraordinary friends and family. This was unthinkable in 2006, nevertheless it was possible, as I have done it.

If I was able to do it, soc an you. We are all human, imperfect, and looking for happiness. Be happy, health will follow. It is a great gift to make for yourself in this upcoming new year. ◄



And if illness had something to teach...

Annie Dubreuil
Brossard, Quebec
Canada

I was six months old when I was diagnosed. In the beginning of the Eighties, the prognosis wasn't encouraging for affected children's future. Despite this, my parents decided to thumb their noses at reality.

Thus, they will did everything in their capacity to have a healthy Cystic Fibrosis child.

The illness is part of me without nevertheless defining me. Basically, I remain, in the eyes of my parents, a child like anyone with an array of possibilities, skills to develop and, mostly, a future to build. Without experiencing hate towards life or illness, I still feel an important inferiority complex towards people with full health. This impression pushes me further to prove to myself and to others that even with a handicap I am a respectable person with something to bring to society. Coming from a family where performance and self-realization are valued and encouraged, the apple didn't fall far from the tree. I am driven by the same fuel. I love living in the action. Living day to day is not for me. Projects, challenges and results nourish me.

Even with the illness, everything remains possible, by design a plan A, B and sometimes C. I succeeded in getting a scholarship in a great Paris university, in travelling to remote areas by passing through Russia, Uruguay, Greenland and the Galapagos Islands, to name a few, in walking the 170 kilometers with steep grades around the Mont Blanc (one of my great prides), to backpack throughout Europe alone, to have a boyfriend and a great group of friends. No doubt, life is good for me. For a long time, I took my health for granted and believed I could control the illness. Statistics were for others. For my part, I would crush them!

Around the age of 21 when I was finishing the last year of my bachelor's degree in economy in the Coop program at Concordia University, things switched for the first time. In the past, I had experienced hospitalization, but never such a quick and important decompensation

While I was succeeding in juggling my studies, my work and my couple, things took an unexpected turn. In a few weeks, my pulmonary functions went from 113% to 37%. Literally, I was seeing stars. I counted my steps when moving. Diagnosis: Overwork. Obviously, a long hospitalization followed. Fortunately, my functions rose to 94% and I could get back a normal life.

Without knowing it, the illness had woken up and I was seated in its roller coaster. With my bachelor's degree completed, I undertook my master's degree in economics which I finished by collecting all nighters and infections. While always pursuing performance, I had learned nothing. Before even submitting my final master's report, a state-owned company, where I had made many internships during my studies, offered me a job in my field. I was more than happy to be offered such an opportunity. My work was fulfilling, and gave me a status and a comfortable financial situation. With work, daily routine is strictly set. Between treatments, the clapping which I had to take over, exercise, work and traffic, I had very little time left. Most weekends were spent sleeping to regain strength and to dream about my next trips.

Even with a strict lifestyle and by carefully following the treatments, the illness continues its course and takes about 10% of my respiratory function per year. With hindsight, the most judicious choice would have been to reduce my workload. Back then, that wasn't an option for me. If healthy people were able to work full time, then so was I.

Even though I cough, spit and threw up every morning, I went to the office. I probably pushed too far the meaning of the Henri Salvador lyrics saying that work is health. Maybe, but at what price? I then had my first pneumothorax requiring pleural talcage. Before leaving the hospital, the surgeon told me something that shook me « Annie, I cannot say if it will be in two, five or ten years, but someday you will be a good candidate for a transplant. Don't forget to come and see me before it's too late ». I receive this advice as a slap in the face. I am not ready to make that an option. In my eyes, my difficulties are temporary. My lungs will recover. It cannot be otherwise. Things calm down for a while. I have the impression of taking back control.

Calm before the storm.

In the beginning of 2009, I was in a nose dive. I like to think that the beauty of Prague Christmas trees took my breath away. Flying home, I knew this was to be my last trip for a long time. My body wasn't following anymore and my lungs even less so. I went to the hospital emergency to treat a pneumonia. At the admission, my first call was to my boss saying that I had to miss two weeks for treatment. I made it a priority to return to work quickly.

After three weeks of treatment, my functions didn't recover higher than a lowly 31%. Even though I had seen it coming for a few days, reality was hitting me hard. At that rate of declining functions, I had to ask myself if I could hold up for to years for that call. Pushed against the wall, I had no other choice but to reassess my priorities. Should I again privilege work over my health? I loved life too much to not give myself a chance. I had to follow the advice of my doctors.

Like that, I found myself on work stoppage. For a moment, I had the impression that the sky had fallen on me. I lost my reference points, my status, but mostly, my identity. Outside of work and travel, who was I? What defined and fuelled me? I didn't have an idea. Even with my name on the donor's list, I still had hope that the wind would turn and that I could push back the surgery by a few more years.

Meanwhile, it is out of the question for me that I waste my days watching television on my couch and envying everyone around me that was healthy. As I was forced to take a break, I would use that time efficiently by discovering what I really wanted from life. Mulling over my shortcomings was too heavy for me. I wanted to focus on what was possible. Changing drastically from action to contemplation mode wasn't always obvious or natural for me. With my oxygen tank, I started yoga and pilates with senior groups. I felt the benefits physically as well as mentally. To spend my time, my sweet tooth brought me to take pastry classes. My desserts looked great to the eye, but didn't taste as good. I love painting, Russian language and photography. The interest is there, but my inner flame wasn't burning. On a beautiful summer morning, the postman left an ad in the mailbox for evening classes at École nationale de l'humour (National Comedy School). I took it as a sign. On top of learning the basics of a good joke, evenings spend at school make me escape from daily life. Laughter becomes my therapy. Like a revelation, I found out I have a certain talent for storytelling and a particular interest for writing. It is in that way that I was tempted to write a first novel. Behind my keyboard, I feel good as I hadn't in a log while. On top of entertaining me, writing feeds my need for appreciation and can be practiced during the endless aerosol and intravenous treatments.

At the moment of writing these lines, I have been freshly transplanted for a few weeks. The wait will have lasted a little over 6 years, 3 during which I was continuous intravenous antibiotherapy. With repeated superinfections, I had a major pneumothorax which almost compromised the transplant and hemoptysis requiring embolization. Although the wait brought its share of uncertainty, difficulty and moments of great anxiety, it was also a nice period of recovery to normal. Having used this time to write and publish six novels of which I enjoyed every creation moment, I redefined my priorities, cleaned up my entourage and spent quality time with the people I love the most for the simplicity of who I am and not for what I had to bring them.

If there are two things I wish now is the stay healthy to spend the most time with my loved ones and to not fall again into the spiral of performance at all cost.

Sometimes, humans have a short memory! ◂

Helping by communicating

Interview with Harold Gagné

On October 17th last year, **Living with Cystic Fibrosis** awarded the Michel-Paquette prize to the journalist Harold Gagné. For several years now, this kind-hearted man has been supporting the cause of organ donation through his humane and respectful reports. He is also an important ally for the cystic fibrosis community.

Having been the subject of one of his stories, I now feel very privileged to be able to turn the tables and become the interviewer to learn more about this great reporter and great individual.

Interview held by
Valérie Mouton

Mr. Gagné, your love for media goes way back... How did you get interested in this field?

I was born in Gaspésie in 1959, but spent much of my childhood and adolescence on the Côte-Nord, in Baie-Comeau and Sept-îles, where I fell in love with radio early on.

The first time I walked into a radio studio, as part of a preschool activity, I was 5 years old. I was mesmerised by the environment and the fact that you could speak to thousands of people through a microphone. I came home and said to my mother: "This is what I want to do when I grow up!"

How did you get started in the media business?

A few years later, I had the chance to get involved in the students' radio during high school and college. At the same time, I found a small weekend job announcing the specials on the overhead speaker in a department store. It was there that I met a real radio host who suggested I come work at CKCN Sept-îles radio station.

I started out playing records during the night and then hosted the evening service. I played the latest hits for the young people who called in with special requests.

I then made the painful choice to leave this fascinating but uncertain and low-paying area to pursue studies in Politics at the Université Laval de Québec, where I stayed for one year. I loved the course. I had very good grades but didn't have enough money to continue. Then I came back to radio at Sept-îles where I worked for 6 years. I left radio hosting to go into public affairs and journalism. I conducted hundreds of interviews with Provincial and Federal politicians, as well as very famous artists.

When did you start your career with the TVA network?

I got noticed by the management of the television station CFER TVA Rimouski, across the River, and was hired in 1986. It was there that I learned all the basics of television reporting. Then in 1994, I decided to try a new experience by becoming a news editor at CHEM TVA Trois-Rivières.

Four years later, I was missing the field. So I returned to reporting, this time at TVA Montréal. I have been there since 1998. In August 2016, I'll have been a journalist for the TVA network for 30 years.

During my years in journalism, I added two other certificates to my certificate in Politics by taking night and weekend classes: one in administration and one in management.

Did you always know that journalism was what you wanted to do?

I've always been drawn to many other professions. After a short stint as an officer cadet in the Canadian Armed Forces in 1979, I almost studied Criminology. I

dreamed of being part of the military police and intelligence service, but destiny had other plans for me. There was no opening in this area and I wasn't keen on military life. It's a whole other world.

I also think I could have been a lawyer.

You have also written three books dealing with quite sensitive topics. Could you briefly tell us about these?

When I was younger, I told myself that one day I would write articles – perhaps a book – but it seemed almost unreal. That was until one day, in 2006, when I conducted a televised interview with Monique Lépine, the mother of Marc Lépine, the Polytechnique killer. The 30-minute program was far too short to answer all the questions. Mrs. Lépine had been hidden from the media for 17 years and this was the first time she was confiding in a reporter. She was seeking the truth, what it was that caused her son to murder several women. We decided to meet with the witnesses of the time: Marc's friends, police officers and teachers. In 2008, I wrote a book that summarised our meetings and the life of Monique Lépine. It was entitled *VIVRE*.

Two years later, I chose to write another book on the children of the DPJ (Youth Protection Directorate): *À quoi ça sert de grandir*?

I feel particularly passionate about this cause. I can't accept children being abused or abandoned, or having serious problems because they're part of the wrong crowd.

I'm probably the journalist who has visited Quebec's youth centers the most. I have interviewed dozens and dozens of children and teenagers who have been scarred for life. They also told their stories in various series of reports that were aired on TVA for 10 years before the holidays in order to raise money to buy them gifts.

Then, three years ago, I thought I would publish one last book, this time on the elderly: Laissez-nous vieillir! In it, I mention my grandmother, who died from cancer all too young, at the age of 63. She had a profound impact on my life. I also discuss palliative care issues and elder abuse. As long as families are not confronted with the diseases of elderly parents, with putting them into nursing homes, or with their death, they usually care very little about ageing. We live in a society that glorifies youth and prefers to stay well away from all this subject. But soon, there will be more elderly people in Quebec than young people. Then, we'll have to start talking about it.

How did you start to get involved with sufferers of cystic fibrosis?

Even if I haven't written a book on organ donation, I have been really interested in it since Valérie Mouton came close to death in July 2004. Her father, Denis, had brought TVA's attention to the importance of organ donation. Together with my colleague Claude Charron, I found myself in a room of the Hôtel-Dieu in Montréal, listening to Valerie, exhausted, urging for donor cards to be signed. A few days later, a miracle happened. A donor was found.

I remember hearing the good news over the phone directly from her father. With my cameraman, we jumped into our mobile for remote assignments and reached the Hôtel-Dieu a few minutes later. She left the scene by ambulance to undergo her double lung transplant at Notre-Dame hospital. On arrival, ignoring security guards, we took Valerie's picture with her thumb in the air, already triumphant even before her operation. The powerful video gave courage to many people waiting for a transplant. It meant life rather than the end.

This meeting with Valerie and her family has had a profound impact on me and later allowed me to come into contact with other patients with cystic fibrosis. I wanted to help in my own way!

You have followed the progress of several people after their transplant... A great deal of success stories! Sadly, some of them lost their fight. Among others, I think of Laura Leblanc who died shortly after her transplant. How do you react to these losses of life?

Laura, a young woman of 19 years from Saguenay, died in January 2014 after waiting far too long for a lung donor. She will always be in my thoughts. I was convinced that she was going to get the better of

cystic fibrosis. There were complications after her transplant at Notre-Dame hospital. I could only imagine the sadness of her parents and sister when she passed away.

In the past, I had already been greatly interested in kidney, liver and heart transplants. At the beginning of the 2000s, I became friends with a gentleman who was around fifty years old and had to undergo a heart transplant at the Royal-Victoria Hospital. I made him promise to call me when he found a donor, and this is what he did one morning at about 4 a.m. He was so happy! Then, in the afternoon, his daughter contacted me to thank me and tell me he had died. That greatly upset me.

This is another reason why I keep talking about organ donation. Had he found the perfect donor? Could he have been saved with better public awareness? That will always haunt me.

Organ donation is the key focus of many of your stories. What impact do you hope to have?

I hope that listeners will never forget the positive stories – and sadly sometimes also the negative ones – and that they will reflect on the importance of organ donation. The choice must not be imposed onto them, but discussions and actions should be gradually brought forward to save more people awaiting new organs.

In the last few days, I've been close to a 5-year old girl who desperately needs a heart to survive. I imagine the anxiety and determination of her parents. We need to talk about this publicly, even if it is a very sensitive issue, to obtain a greater number of donors. This way she can be saved. I am sure of it.

What do you learn from your meetings with people living with Cystic Fibrosis?

All these meetings make me appreciate life more. The patients and their families demonstrate courage that perhaps I couldn't. They are immensely important in my eyes. They all have values and desires to build a better world, a society where one day, we can save those in need of a gift of life!

What are your plans for the future?

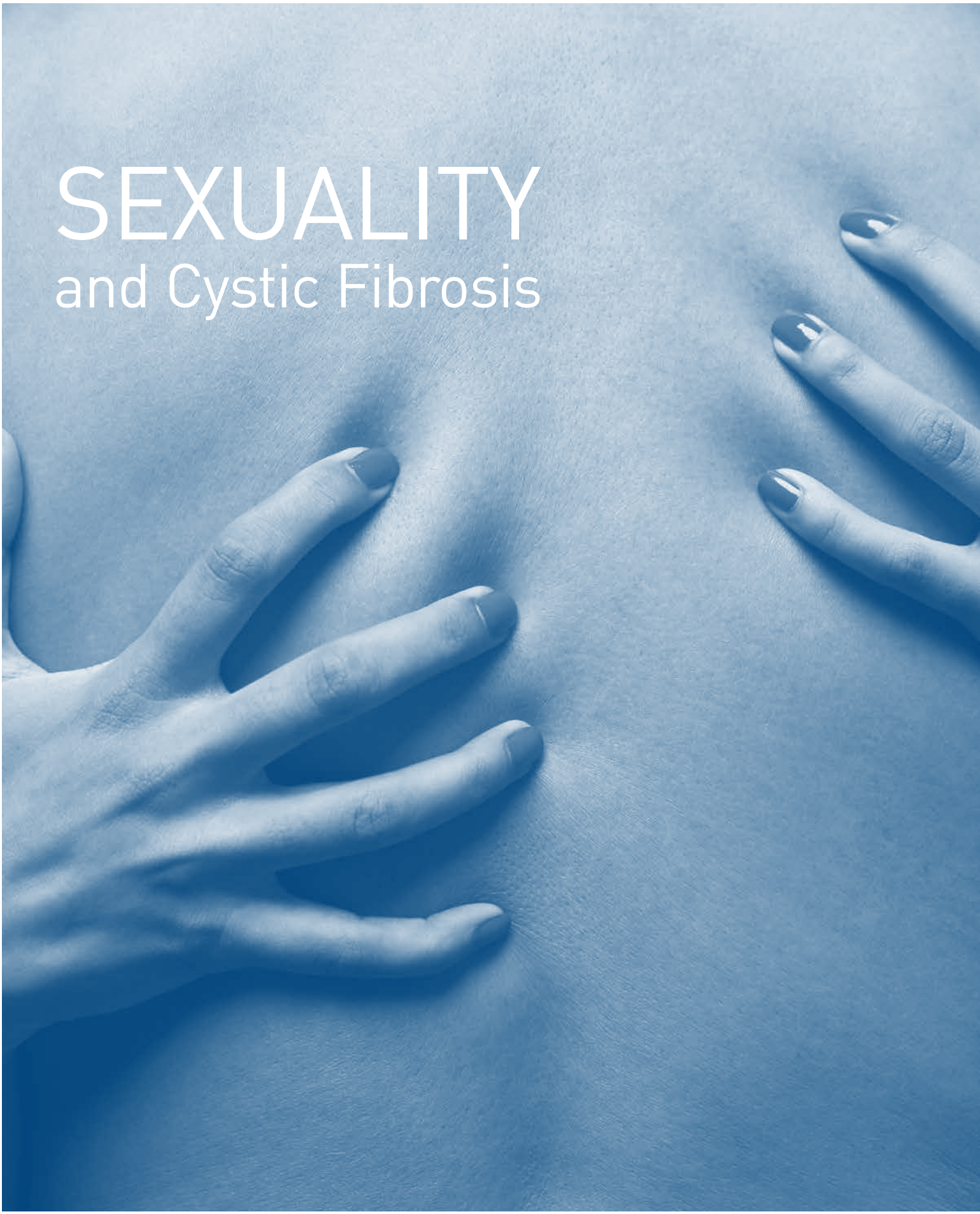
As a journalist, I now hope that hospitals and doctors will allow us to show people what a transplant is really about, with the process, decisions, hesitations, victories and failures it involves. For 10 years, we have greatly focused on emotions, on the testimonies of patients awaiting transplants, and on the importance of signing donor cards and talking about it with families. We must now explain what we're really talking about using images, while at the same time putting a system in place that encourages healthcare workers to do everything, everywhere in Quebec, to obtain the highest number of donors.

We have already gone a long way. In 2004, when Valérie Mouton received her transplant, the public relations department of Notre-Dame hospital criticized her father for providing me a video of his daughter in her hospital room thanking those who prayed for her. Ten years later, a doctor told me that these images and reports had been a catalyst for Transplant Québec and for the cause of cystic fibrosis.

Sometimes we have to push the limits of the system to achieve ambitious goals. This is what I will continue to do, while respecting the choice of individuals. I am convinced that donating one's organs at the end of one's life is a personal decision that must not be imposed but simply considered.

Thank you very much, Mr. Gagné. ◂

SEXUALITY
and Cystic Fibrosis



Geneviève Nadeau
sexologist

Gatineau, Quebec
Canada

Sexuality: a subject causing much ink to flow! Whether you are a teenager, adult, male, female, transgender, heterosexual, homosexual, single, in a relationship, a parent looking for questions to answer those of his children, etc.; everyone wants to know more. In a society like ours, where social norms and stereotypes prevail, who can honestly say that they have never questioned themselves about their love and sex lives? We are constantly bombarded with images that dictate what a woman should look like, what a man can or cannot do; texts that explain in 10 points what to do to excite a man, or how to demonstrate "true" masculinity. It is difficult to navigate all of this whilst fully living our sexuality by ignoring the social pressures, being oneself, and not becoming a copy of what the media conveys.

When one adds to these questions, already ubiquitous, concerns and challenges related to physical condition, it is normal to feel overwhelmed. Finding a partner who shares our values, who respects us for what we are with our flaws, and with whom the sexual chemistry is present, is a challenge we all face. We all want to show ourselves at our best at the beginning of the relationship, but as intimacy develops, so comes the less good days when fatigue, stress, or simply bad news, sometimes takes over. Opening up to our intimate ones without fear of judgment: expressing what we live, what we fear, what bothers us, is not an easy task. Communication is the key to any relationship. That being said, it is not always so easy to implement. It is normal to wonder about the best time to tell our partner that we have cystic fibrosis. The first rendezvous is perhaps too fast, but after two months of dating, it may be that the other would feel hurt not to have known earlier. There are obviously no good answers to these questions: we must follow our instincts and discuss this when we feel that the time is right. If the loved one is the right one, they will receive your testimony without judgment, and with all the listening you need.

Once in a relationship, problematic or embarrassing situations can still occur. The ideal woman is often represented as always ready and available for sex, and the man as always the instigator with his beloved. But when vaginal infections, urinary incontinence, the irritation of scars, or simply a sense of discomfort occurs, it is even more difficult to indulge in sexual pleasure. Listening to and trying to understand our loved one is even more important with cystic fibrosis. No need to share all the discomforts, such as urinary incontinence that is sometimes caused by coughing, but it's ok to leave ourselves the space for mentioning to the other that we need a moment to get more comfortable and feel good. It goes without saying that doing any exercise, such as sex, often leads to coughing. But once complicity has been developed within an intimacy, these moments are no longer an issue.

What about fertility? The majority of men with cystic fibrosis are infertile, and while some procedures may be undertaken for successful in vitro fertilization, these procedures are expensive and not fully effective. Mourning the idea of parenthood, at least from a biological sense, may already be an ordeal in itself. And in all relationships, that question arises: do we want to be parents? Of course, this issue becomes a couple

question when the relationship turns out to be more serious. Again, listening and communication will allow us to express our desires and see if they can be compatible with those of the other.

Although women are not infertile due to cystic fibrosis[1], pregnancy can be quite a trial. In addition to discussing with her partner the best time to start a family, her doctor must be included in the discussion. He may give good advices on the health status of the mother and the risk of pregnancy. To all this is added the difficult but important discussion of who will take care of children should the health of the parent with CF deteriorate.

Unquestionably, being in a couple while having cystic fibrosis brings its share of discussions and issues! But that does not stop the possibility of having more than satisfactory romantic and sexual experiences. Pamper yourself, and let yourself be carried away by the beauty and happiness that love and sexuality can bring. ◂

[1] A couple is said to be infertile after two years of unprotected intercourse without result, while infertility is a permanent inability to procreate.

No Stress!

" When happiness is there, we see it, but when we don't see it, it is still there. And you, during that time?… where were you? "

Manu Lemire
professional personal development coach, author and speaker

Saint-Augustin, Quebec
Canada

Today my friends, I will simply try to help you understand a very simple thing: « stop worrying about life», it is exactly what it should be!

Beauty residing in each achievement is not represented only by the finished product, it is also in all the work that was accomplished during its creation. Your life is no exception, it is the fruit of daily work, some success, painful failures, great accomplishments and bitter losses. Personally, why try understanding it, this is obviously beyond us...

Visualizefor just two seconds where you are at in your life. Think of your financial situation, your friends, your family, your daily occupations, your way of thinking or anything else that can cross your mind. Now, imagine the incalculable details, such small details that could all, one as well as the other, could have brought you completely elsewhere, living a completely different life or even being a completely different person...

The possibilities are infinite, you have to admit it.

Why not try to understand something so complicated as our existence. I believe that one of the keys to happiness resides in the fact that it should be taken for granted that everything will go well, at the right time and that whatever the obstacles in our path, it will represent a necessary pain in your training. Worrying will change nothing in what must happen, the only thing it will bring is passing by your life because you were too busy looking forward instead of taking advantage of the current moment, now and here. Furthermore, is negative that harmful? For my part, the greatest lessons I learned were taught in the hardest moments of my existence. If everything went well, could we really prove our value or correct our shortcomings and weaknesses? If bad guys didn't exist, Superman would be a janitor, bus driver or hot dog vendor on a street corner. Do you follow me?

Falling is not a failure in itself. Staying on the ground is! And it is not life who will decide, it's you!!

Who among you has never lived difficult moments? No one, of course! And still, today, you are here, reading this article, you are standing, ready to fight for what you believe in... That is something my friends... That is something...

If you are still standing after all this hardship it is that you have made the choice of believing in life. You told yourself that by hanging on, one day or another, everything will go well... You are so right... When we go through a painful moment, we always have the impression that it is unfair, that it shouldn't be so, that we don't deserve it... How is it that today we see things differently? Time! Time is sometimes the only thing that can bring us the answers we look for... You have to hold on, keep the faith and look forward. I am teaching you knowing by telling you tat life is made of ups and downs, positive and negative, and that these two extremes are what gives value to happiness. The beauty of the current moment is an invaluable gift that we often forget to unwrap everyday.

I repeat it to you, nothing comes for nothing. Sides of the person I am most proud of were forged in adversity. The importance that my hardships had on the man I became are undeniable and I am willing to bet that it is the same for each of you. You are stronger, bigger and better prepared to weather the next storm...

Why worry? Maybe you will soon live an event that you will consider horrifying, but that will, in the end, have been necessary to make you the person you are meant to be. Can someone predict the future? Can someone know exactly the surprises that will come tomorrow? How many believed they had met the love of their life before, months or years later, finally meeting THE right person? Life is unpredictable even when you try to control everything, it is simply impossible! So I repeat my question: why worry?

Whether you want something, whether you don't want it, whether you think this, whether you do that, life is full of twists that I stopped bothering about controlling everything... I known spend the most time enjoying each moment, positive or negative, learning about each experience, listening more closely to each person, laughing for nothing... In the end, what I am trying to do is to love my life, for what it is as a whole, and to thank heaven for still being here, healthy, fully enjoying it with the ones that are dear to me.

I hope that after reading this you will go through life with a light heart, knowing that whatever tomorrow brings, one day or another, all will be well. ◄

Love... and health!

Diane Gagnon
coach and autor

Québec, Quebec
Canada

These are often the wishes we extend to our loved ones almost automatically at the start of each new year: love and health!

Many of us mistakenly believe that only a few privileged individuals receive these precious gifts to be happy in life. Sometimes we have the feeling that we don't have either of them, and our lives don't seem to live up to our expectations.

And yet, we ALWAYS have much more than what we think!

In fact, we often hope to regain our health or receive love, when it is actually us who need to BE the love and APPRECIATE the health we have – a sort of special training for the areas of ourselves that require even more love.

Let me tell you what I mean.

The reason for our existence on Earth is to learn to love. More. Better. Without judging. Everything that happens to us in life – absolutely everything, without exception – happens for this valuable lesson: to learn to love.

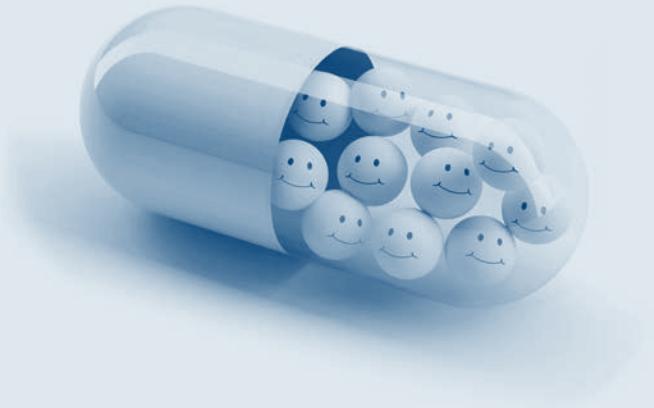
First of all, this means learning to love ourselves, because it is difficult to love others if we do not love ourselves. Learning to love yourself can be the greatest challenge you face in your lifetime. Regardless of our physical, financial, social, personal or family status, we must learn to love ourselves for what we are, with whatever life lessons are placed on our path in their various forms – they're there to teach us.

Don't expect to be loved for loving yourself or loving others. A fireplace gives off no heat unless we have wood, paper and fire! Let's start first by loving ourselves, us, the way we are, loving the life we have, even if we don't like it, it's no everyday challenge. Because in the love we have for ourselves lies the key to our happiness.

This lesson can't be avoided or postponed for our whole life. It is the basis for everything, for all our happiness, our openness to life, our approach to what happens in our life, our openness to others and our ability to receive the love of others. Have you noticed how we doubt the love of others when we don't love ourselves? To accept being loved, you must first love yourself.

It is often a long and difficult lesson, as it is not widely taught, but it is a daily journey that gives us great joy... and love!

Let's learn to love ourselves by doing an act of kindness for ourselves every day. By taking care of ourselves, for example, by doing something that pleases us, by recognizing our qualities, or by finding the child within us to look after. Also, by listening to our heart, by putting ourselves at the service of our soul, we can be seamlessly guided through our own life mission.



Because we all have a mission in life, regardless of our physical capabilities.

We are always given us the best possible conditions for our development. Oh! Of course, we often strongly disagree with what is "imposed" on us. But if we open ourselves up to life, we will understand that everything serves our greater good, whether it's to achieve our mission in our own way, to finally learn to love ourselves as we are, or to learn how to love others and life more, nurturing the life we have.

Our health, or rather our health conditions, often seem to be a limitation on what we want to do. But what if these were the very conditions we need to accomplish our life mission? And even if our health problems worry us, don't we have, elsewhere in our life, a healthy mind, a healthy heart and healthy relationships?

Byron Katie, an internationally-renowned American author, reminds us of this: "How do we know if we really need this thing (illness, problem or event) in our lives?" If we have it, if we are experiencing it, it is what we need! Likewise, what we don't have, we don't need at the present time.

I know, for many of us, this is probably a drastic change in perceptive towards life. And any change, especially of this magnitude, needs time and openness. Indeed, the most important thing is to remain open to these thoughts and not to discard them right away. Let them run their course within us and let's see if they can bring us something of value.

I know, I don't suffer from cystic fibrosis, but I've had my share of challenges and illnesses in my life. When I finally opened myself up, just a little bit, to a new perspective on life, my life began to change. For the better. In all aspects.

How open we are to life determines how much happiness we let in. The more open we are, the more we can welcome what life sends us and understand that everything has its place in our lives. And we too have a place in our lives.

So every hard moment in life is necessary to make room for something bigger. Every abandonment teaches us to love ourselves more and to never again abandon ourselves. Every failure is a springboard to go further, so it is not a real failure. Every betrayal teaches us to love more and to forgive. Every disease teaches us to love more, to choose what is essential, not to judge, and to develop qualities and skills we may not have developed otherwise. Every challenge is part of our development. Every heartache teaches us to keep our heart open. There's a wonderful Sufi proverb which says: "Life breaks the heart again and again and again until it stays open". And doesn't Leonard Cohen sing that the cracks are what lets the light get into our soul?

So, a day will come when we "welcome" what we have been given, day by day, with more and more openness, as we gain an intrinsic awareness that there is a gift for us behind every situation. Sometimes the gift is poorly packaged, I agree. But there is ALWAYS a gift. The heavier burden, the larger the gift!

I wish, with all my heart, that you find your unique gift in life! ◂

Study of the Psychological Needs
of Adults Living with Cystic Fibrosis

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The aim of the current study was to assess the psychological needs of adults living with CF. As per the World Health Organization recommendations, we included a clinical assessment of depressive symptoms and anxiety, and examined the relationship between scores on these clinical assessments and likelihood to use psychological services if available at the clinic.

The participation rate in the current study was 94%. This suggests that the study sample, although small, may be representative of the adult population living with CF in Ottawa. First, several observations can be made about the prevalence of symptoms of depression and anxiety in the study sample. In our study, 40% of the sample reported elevated symptoms of depression. This prevalence is higher than in the general population, and slightly greater compared to a large study in which 29% of adults with CF reported elevated depressive symptoms. Prevalence rates in women tend to be higher compared to men, but the current study sample was fairly balanced with regards to sex (42.2% females). Importantly, 20% of study participants reported low levels of depression and regular ('occasionally' or 'often') past access to psychological services. A positive interpretation of this is that accessing psychological services had a protective effect on participants' symptoms of depression.

In terms of anxiety scores, a total of 13% of our study sample reported elevated symptoms of anxiety on the GAD-7 (seven questions on generalized anxiety disorder). This prevalence rate is lower than the rate reported in a large sample of adults with CF (32%). On the other hand, it is above the prevalence of elevated anxiety symptoms (5%) in another study on 153 adults with CF, and above the prevalence of Generalized Anxiety Disorder in the general Canadian population.

Our study data suggests that approximately 30% of those experiencing elevated depression symptoms in the sample had 'never' or 'rarely' accessed psychological services. There were fewer participants with elevated symptoms of anxiety in the study sample, and half of them had 'never' or 'rarely' accessed psychological services. It is it not uncommon for people who experience symptoms of depression or anxiety to not receive the psychological support they need. In an Australian study, 75% of clinically depressed cancer patients did not receive psychological treatment, despite the availability of counseling and support groups. Two US based studies showed that the mental health care costs and visits for patients with and without depression were similar, despite differences being significant for other health care services. In another study, a large proportion (64%) of University students with

Following a transplant, patients should avoid consuming unpasteurized foods such as cider vinegar.

major depression were not receiving therapy, despite having health insurance and free access to short-term therapy from campus providers. In addition, people suffering from anxiety may be less likely to use mental health services than people with mood disorders . These data suggest that problems in accessing psychological treatment are not restricted to CF and Canadian populations. If psychological services are available at a CF clinic, other interventions promoting their use should be delivered in parallel.

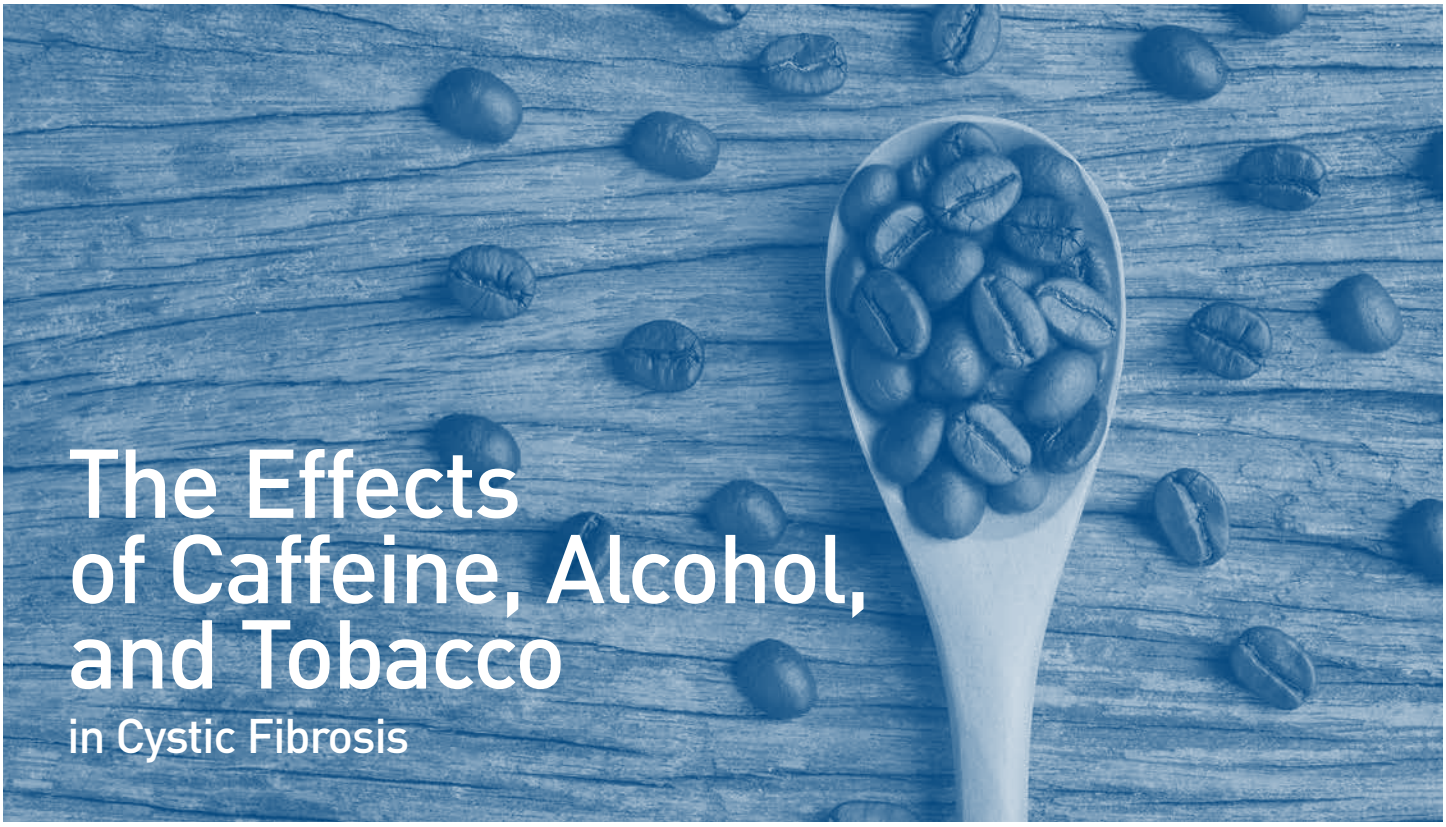
The majority of participants reporting elevated symptoms of depression and anxiety reported that they would be likely to use psychological services if they were made available at their CF clinic. A large proportion of those who did not report symptoms of depression or anxiety showed interest in using psychological services. As Oxley and colleagues state, the range of psychological stresses and difficulties people with CF experience may not be identified by measures of depression and anxiety. Participants in our study were not only interested in discussing worries and mood with a psychologist, they were also interested in discussing life transitions, quality of life, life stress, and adjustment to CF. As such, a first recommendation supported by existing guidelines would be to implement annual routine psychological screening of patients with CF. This would help identify patients at risk of depression and anxiety, and allow them to be given priority access to a psychologist. To be effective, screening strategies need to account for barriers to implementation identified in recent work, the most commonly cited by CF health care professionals being limited staff time, limited personnel, and lack of qualified personnel to provide referrals or interventions. The high proportion of participants in our study with low symptoms of depression or anxiety that showed interest in seeing a psychologist underlines the need to deliver the preventative and supportive interventions recommended by the Cystic Fibrosis Foundation. According to these guidelines on mental health, these interventions could include training in stress management, development of coping skills aligned with appropriate developmental stages and life disease events, and behavioural approaches to reduce the risk of distress, particularly for those undergoing medical procedures.

There is little research on the effectiveness of psychological interventions in patients with CF. As was underlined by Oxley and colleagues, there is no reason people with CF would not benefit from such interventions when they have been shown to be effective in other populations. Several interventions have been found to improve depression and anxiety in people with other medical conditions. Offering psychological interventions to patients with CF may also improve the effectiveness of medical co-interventions.

This study is the first description of the psychological needs of adults with CF attending a Canadian adult multidisciplinary CF clinic where a social worker is available, but access to a clinical psychologist is only available through a private practice referral system. This situation is common in CF clinics across Canada. In 2011, Cystic Fibrosis Canada released a summary of the services available at the 42 CF accredited clinics in Canada. In this document, the services offered by psychologists and psychiatrists are presented together despite the important differences in roles between these professions. The data do however provide some overall indication of the level of access to mental health services in Canadian CF clinics. Only 16 of 42 CF clinics had a psychologist/psychiatrist integrated to the CF multidisciplinary care team, and community private practice referrals were available in 20 CF clinics. In 3 of the Canadian CF clinics, there was no psychologist/psychiatrist integrated to the CF team, and no referral system in place.

European countries have agreed that there is a need for CF teams to include a clinical psychologist, and this is now part of their standards of care. As such, many Canadian CF clinics do not meet international standards for CF care. Referral systems are sometimes in place but they have several disadvantages. First, they are often associated with excessively long waiting times, sometimes up to 12 months. A second disadvantage of referral systems is that it is more difficult to create and maintain a working partnership between the CF care team and the psychologist/psychiatrist, especially when these services are offered in different locations. This is important, as an integrated team approach is key in the care of people with chronic health conditions. A third disadvantage of referral systems relates to the reimbursement scheme associated with these referrals in Canada. Psychological services offered in hospital-based CF clinics are covered by universal provincial health insurance plans; referrals to private practice psychologists outside the CF clinic team are not. The majority of people with CF are unlikely to be able to afford expensive private practice psychological services, or to have access to extended employee health benefits that could help cover the costs (40% of participants in our study were unemployed or on disability). The limited availability of publicly funded psychological services in health settings, and the lack of affordability of private services, was underlined by the Canadian Government in 2004, yet this has not translated to improvements in Canadian CF care.

Our study highlights the psychological needs of adults with CF, with regards to both prevention and treatment. Despite the availability of mental health services in some of the CF care teams across the country, psychological services are not available to patients with CF in the majority of Canadian CF clinics. This means patients with CF are receiving sub-standard care. There is an important need to continue to promote an integrated team approach to CF care, and to underline the need for multidisciplinary CF care teams to include a clinical psychologist. ◂



The Effects of Caffeine, Alcohol, and Tobacco

in Cystic Fibrosis

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Introduction

Caffeine comes from many sources, but tobacco and alcohol are from a single source (the tobacco plant and ethanol, respectively) and then made available in various forms. Logically, most chronically ill patients should avoid substances that have the potential to worsen their disease or cause significant adverse effects when combined with medications they are currently being treated with. Caffeine is a bitter substance found typically in coffee, tea, soft drinks, chocolate, kola nuts, and some medications. It has several effects on the body's metabolism but is known to stimulate the central nervous system, giving individuals a boost of energy and alertness[1]. Two to four cups of regular coffee a day may not be harmful for some, but for others it may foster the effects of caffeine: jittering of the limbs, difficulty sleeping, headaches, dizziness, tachycardia, and dehydration[1]. With an underlying disease such as cystic fibrosis (CF), it can be very hard on a patient's body, which is already being strained by chronic illness. Tobacco and alcohol are no different in that they cause destruction to the healthy human body let alone one plagued with CF. Smoking tobacco harms nearly every organ in the body, and 87% of lung cancer deaths are attributed to this substance. Other damaging effects of tobacco smoke include lung disease, heart disease, stroke, and even cataracts[2]. Alcohol at high concentrations can take a toll on a healthy individual, as can chronic use. Small amounts of alcohol are generally not beneficial to health, but these amounts typically have no adverse effects on a healthy individual[3]. Individuals who consume alcohol from age 18 and into their early adult years are at an increased risk of liver disease, heart disease, pancreatitis, and cancer. These risks are multiplied for individuals who are chronic consumers of alcohol, those who take prescription medications that interact with alcohol, and those with certain medical conditions[3]. By examining the relationship between the consumption of these staple drugs of American culture by healthy individuals and patients with CF, information can be obtained for future use in the treatment for patients with CF that may help them better understand how to live with CF and what lifestyle choices can determine their survival.

Cystic Fibrosis Overview

CF is an inherited chronic disease that affects an estimated 30,000 children and adults in the United States and another 70,000 worldwide[4]. Approximately 1 in every 3,500 babies in the United States are born with CF, and CF primarily affects Caucasians but has the ability to affect all racial and ethnic groups[4]. The disease causes severe damage to the lungs and digestive system that can be life threatening. It is caused by a defective gene that affects the cells that produce mucus, sweat, and digestive juices, causing an abnormal sticky, thick mucus that clogs the lungs, obstructs the normal function of the pancreas, and stops natural enzymes from helping the body break down and absorb nutrients. In a healthy individual, secreted fluids are normally thin, slippery, and act as lubricants; however, in patients with CF these fluids plug up tubes, ducts, and passageways, primarily in the lungs and pancreas. The median survival age for patients with CF is now 36.8 years due to advancements in medical research and technology[4].

On a cellular level, CF is an autosomal-recessive disorder affecting chloride transport in the pancreas, lung, and other tissues. In the majority of cases, a ΔF508 mutation occurs in the first nucleotide-binding fold (NBF-1) of the cystic fibrosis transmembrane regulator (CFTR)[5]. The CFTR is expressed in airway epithelia on the luminal side of the plasma membrane,

where it serves as a phosphorylation-regulated Cl– channel and a regulator of channels and transporters. Activation of CFTR leads to parallel inhibition of the epithelial Na+ channel, which is lost when CFTR is absent or dysfunctional[6]. It is postulated that the loss of Cl– secretion and Na+ absorption reduces the thickness of the airway surface liquid overlying airway epithelia, resulting in impaired mucociliary clearance. Reduced CFTR-dependent bicarbonate secretion might affect the hydration of the secreted mucus, affecting its physical properties. Since CFTR is also expressed in submucosal glands in the airways, it plays an important role in defending the body from foreign substances that are inhaled[6]. Loss in the function of ductlining serous cells that contain CFTRs prevents the secretion of mucus and antimicrobial factors by the submucosal glands. These pathological features contribute to the formation of thick, dehydrated mucus that provides a pristine environment for continued growth of bacteria, which ultimately triggers chronic inflammation and organ failure in patients with CF[6].

Patients suffering from CF have various symptoms, depending upon their age. For newborns, symptoms include delayed growth, failure to gain weight normally during childhood, no bowel movements during the first 24–48 hours of life, and salty tasting skin[7]. For individuals who are diagnosed at a later age, any of the following may be symptoms, based on the particular system inundated by the disease: nausea or loss of appetite, weight loss, belly pain from severe constipation, coughing or increased mucus in the sinuses or lungs, fatigue, recurrent episodes of pneumonia, and sinus pressure[7]. Individuals with CF may develop infertility, repeated inflammation of the pancreas (pancreatitis), respiratory symptoms, and clubbed fingers later in life[7]. Since there is currently no cure for CF, treatment for these patients includes (1) antibiotics, which prevent and treat lung and sinus infections, and inhaled medications to open airways; (2) DNAse enzyme therapy to thin mucus; (3) hypertonic saline solutions; (4) oxygen therapy; and (5) alterations in dietary intake, especially with regard to protein and calories, and other therapies relating to increased consumption of antioxidants[7]. Over the years, as more has been learned about CF and treatment options, researchers have been looking into the consumption of other foods and substances to determine how patients with CF are affected compared to individuals who do not have CF.

As caffeine, alcohol, and tobacco are further researched in patients with CF, hypotheses can be created to determine whether the typical side effects of these substances affect patients with CF and people without CF in the same or worse ways.

Caffeine and patients with CF

Caffeine is a white, crystalline xanthine alkaloid and is considered to be a psychoactive drug. Caffeine is believed to work by blocking the adenosine receptors in the brain and other organs, which in turn reduces cellular activity. This action then stimulates nerve cells to release epinephrine, which causes the body to exhibit increased heart rate and blood pressure as well as the other common effects of caffeine. It also increases the levels of dopamine and causes the liver to release glucose[1]. For individuals who become tolerant of the effects of caffeine, headaches may be a withdrawal symptom if they lack it in their body. This is one reason why caffeine is sometimes added to medications that treat headaches. On the other end of the spectrum, individuals who consume too much caffeine and develop intoxication can experience nervousness, increased urination, flushed face, and even hallucinations[1]. According to the US Food and Drug Administration (FDA), the consumption of caffeine in the United States between 2003 and 2008 was approximately 300 mg per person per day[8]. The FDA also reported that solid foods contribute a very small amount of caffeine, whereas the major source of caffeine is coffee, with roughly 64–145 mg in an 8-oz cup of it. Other sources of caffeine include cocoa (11–115 mg); guarana (5%, whereas coffee beans are between 2% and 4.5%); kola nuts (2–3.5%); tea (4%, with a typical cup containing 20–80 mg); taurine, which is an additive in energy drinks; and yerba mate[8].

Patients with CF are at an increased risk for loss of sodium and chloride, two critical components of sweat. These individuals are at a higher risk of losing these vital elements during exercise, fevers, and infections or when exposed to high temperatures[9]. Sodium in the body acts as a sponge and helps maintain bodily fluids wherever it is present, primarily the blood and tissues. As patients with CF sweat out water and salt (sodium and chloride), their bodies are unable to reabsorb the salt. Their bodies do not notice the deficit in sodium and water being lost, which ultimately results in dehydration[9]. This characteristic of patients with CF makes consuming large quantities of caffeine an unwise choice. The diuretic properties of caffeine, which causes frequent urination, can endanger a patient who is already at risk of dehydration based on the underlying pathogenesis of their inherited disease. Some of the characteristics of salt and fluid deficiency mirror those of CF, which help to explain further the complications of the disease and the risk involved with dehydration and caffeine consumption. Hyponatremic dehydration also contributes to sputum that is thicker and more difficult to expel. In addition, thicker secretions from the bowel lead to blockages in the intestines[9].

A study was conducted in 1997 involving children diagnosed with CF and the possibility of enhanced drug metabolism. The study used caffeine as the mechanism to determine the increased or decreased usage of the CYP1A2 metabolic pathway. The results showed that patients with minimal pathophysiological changes in liver and lung function have enhanced metabolism of the pathway involving the CYP1A2 enzyme. The changes in liver function or pulmonary diseases can affect drug metabolism; however, patients with CF often have enhanced clearance of drugs[10a]. Based on this study, it seems that individuals with cystic fibrosis may have an increased amount of medication their body takes in because of the activity of the CYP1A2 enzyme and not specifically related to caf- feine. Another study hypothesized that coffee was a food replacement in individuals with chronic pancreatitis[10b]. It was noted that in the later stages of the disease, patients with CF may experience repeated inflammation of the pancreas, which can develop into chronic pancreatitis[7]. Olesen et al.[10b] stated that, due to the pain associated with chronic pancreatitis, individuals often avoid consuming food and replace it with alcohol, tobacco, and coffee. In the study, drug absorption was being researched in patients with chronic pancreatitis, and it showed that these patients may have increased drug absorption due to the pathophysiology of the diseas[10b]. The significance of this is again not directly correlated to caffeine and its effect on patients with CF, but on how patients with CF who develop chronic pancreatitis can worsen their disease by consuming coffee to replace meals and suppress their appetite[7]. This can indirectly affect the nutritional efforts of individuals with CF who are lacking nutrient absorption and must consume nutrient-rich foods to nourish their bodies.

Alcohol and patients with CF

Ethyl alcohol, or ethanol, is the world's most important psychoactive depressant drug and is the intoxicating component of beer, wine, and distilled liquors. It is obtained through the fermentation of various products including yeast, sugars, and starches. As ethanol products are distilled their concentration increases, producing a concoction containing more than 15% alcohol[3]. Depending on the amount of alcohol consumed, the weight of the person, and the strength of the alcohol, the effects will differ. Alcohol is initially consumed for its stimulant effect, the notion that one is "loosened up or relaxed"[11]. But as an individual consumes more alcohol, the depressant effects of this drug present themselves throughout the central nervous system and manifest as slurred language, impaired judgment, and other disturbed perceptions[11].

The absorption of alcohol begins in the stomach but primarily occurs in the small intestine. In the presence of food, the absorption is reduced; however, in the presence of carbonated liquids, the absorption increases[3]. As alcohol is absorbed into the body, it is distributed throughout the bodily fluids and

metabolized by the liver at a rate of 0.25 ounces per hour, with 90% metabolized. One sex difference with respect to alcohol is the stomach enzyme that metabolizes alcohol. This enzyme is less active in women than in men, which results in women being more susceptible to the effects of alcohol consumption[3]. As it depresses the central nervous system, alcohol has many effects on the brain. Its mechanism of action is similar to that of barbiturates and benzodiazepines, which enhance the inhibitory effects of gamma-aminobutyric acid (GABA) and the GABA- A receptor. At high doses, it blocks the effects of the excitatory transmitter glutamate and affects dopamine, serotonin, and acetylcholine neurons. Also, alcohol influences peripheral circulation and fluid balance and has hormonal effects. As an individual becomes more and more intoxicated, peripheral blood vessels dilate, causing the user to lose body heat but still feel warm[3]. The diuretic effects of alcohol cause the user to have decreased blood pressure and excessive urination. Excessive urination is caused by the body's inability to release antidiuretic hormone, which prevents the body from retaining fluid. The long-term effects of alcohol are brain tissue loss and intellectual impairment, liver disease, heart disease, cancer, and impaired immunity[3].

According to the Centers for Disease Control and Prevention, two surveys indicated that more than half of the US population drank alcohol in the past 30 days, with approximately 5% of the total population being heavy drinkers and 17% of the population being binge drinkers[12a]. Another study indicated that from 2001 to 2005, there were approximately 80,000 deaths annually that were attributable to excessive alcohol consumption, making it the third leading lifestyle-related cause of death for people in the United States each year[12a]. In the United States overall, chronic alcohol abuse is a major cause of alcoholic hepatitis and chronic pancreatitis, with rates at about 50% and 5%, respectively, which further indicates that only some individuals are susceptible to the development of these disease[12b]. Interestingly enough, the third leading cause of death in patients with CF is liver disease, which is not necessarily caused by alcohol but is one of the adverse effects of excessive consumption. The prevalence of liver disease among patients with CF is 2–37%, varying largely on liver testing[13]. Since the adverse effects of excessive alcohol consumption include direct toxicity to organs, especially the liver, patients with CF are already at an increased risk because of their condition. Of the percentage of patients with CF with liver disease, up to 3% will see a progression to liver decompensa- tion as a result of abnormal CFTR[14]. Consuming excess amounts of alcohol can be considered a "risky behavior" among the general population; however, individuals who are already suffering from bodily distress are at a greater risk when participating in this behavior. Mc Ewan et al.[14] conducted a study looking into the behaviors of patients with CF and found that of the 599 participants in the study, 77 males (94%) and 98 females (98%) had tried alcohol and 151 participants (83%) continued to drink on a daily basis. In addition, 144 participants (79%) did not drink or drank within the recommended limit, which is significantly higher than that of the general population[14]. The results of this study suggested that individuals with CF who participate in these behaviors are imposing additional health risks with respect to the disease. Risk-taking behavior tended to start at a later age when compared to people without CF and was more prevalent in women[14]. In another study, researchers showed that individual alcohol intake was low in all groups tested but decreased as patients switched from dieting to enteral feeding[15a]. As a result of the complications associated with CF, enteral feeding allows for more nutrients to be absorbed by the body[15a]. Alcohol posses a similar threat to both patients with CF and the general population, but the primary difference here is the current underlying characteristics of the disease that patients with CF already have and that alcohol can greatly increase. The meal replacement hypothesis studied by Olesen et al.[10b] mentions alcohol as another food replacement option for patients with chronic pancreatitis.

Chronic pancreatitis causes irreversible anatomical changes and damage to the pancreas that includes infiltration of inflammatory cells, fibrosis, and calcification with destruction of the glandular structure[15b]. This damage then causes normal digestion and absorption of nutrients to cease altogether. This has become evident in alcoholics who have been consuming alcohol for 8–9 years[15b]. The malabsorption and malnutrition

associated with patients with chronic pancreatitis indicates the increased risk for patients with CF who are currently experiencing chronic pancreatitis or who are headed in that direction. Using alcohol as either a meal replacement to prevent the pain associated with chronic pancreatitis or for its other uses poses a significant threat to the long-term survival of patients with CF[10b, 15b]. Lifestyle changes are crucial in an attempt to extend the life of patients with CF who are already experiencing the later complications of the disease.

Tobacco and patients with CF

Two main species of tobacco are grown today: *Nicotiana tabacum*, indigenous to South America, and *Nicotiana rustica*, found in the West Indies and eastern North America[2]. Tobacco is found not only in cigarettes but also snuff, chewing tobacco, smokeless tobacco, hookah, and cigars. Similar to caffeine and alcohol, tobacco contains a substance that has significant effects on the body. Nicotine, a naturally occurring liquid alkaloid, is present in tobacco and is the addictive ingredient that quickly builds tolerance and dependence. It is highly toxic, with a lethal dose of 60 mg; cigars contain about twice that amount. Inhalation of nicotine through the use of tobacco products results in the absorption of 90% of what is inhaled. Most of the nicotine absorbed (80–90%) is deactivated by the liver and then excreted by the kidneys. Tolerance is created by an increased use of liver enzyme activity responsible for nicotine deactivation[2]. Nicotine, like caffeine and alcohol, affects the central nervous and circulatory systems. It increases the heart rate and blood pressure; increases the oxygen needs of the heart, decreasing the oxygen-carrying ability of blood and resulting in shortness of breath; reduces hunger; increases blood glucose; and deadens taste buds[2]. Smokers report that nicotine has both a calming and stimulating effect, which may contribute to why users continue to use tobacco and other products containing nicotine despite the possible side effects. The side effects of continued tobacco use are similar but differ slightly based on the forms used. Lung cancer, cardiovascular disease, and chronic obstructive pulmonary disease are the primary adverse effects of continued tobacco use, but others include bad breath, spitting, increased risk of dental disease, and oral cancer. Additional costs of smoking include financial costs, social isolation, physical isolation, increased risk of fires or fire-related injuries, and pollution from the toxins[2].

Tobacco use is another risky behavior many Americans partake in on a daily basis. According to statistics tabulated by the Centers for Disease Control and Prevention, an estimated 45.3 million people, or 19.3% of all individuals 18 years of age and older, smoke cigarettes (21.5% of men and 17.3% of women). Cigarette smoking is the leading cause of preventable death in the United States and accounts for approximately 443,000 deaths each yea[16]. In individuals with CF, exposure to smoke irritates the linings of mucosal membranes, which increases coughing and sputum production and increases the risk of bacterial infections and worsening symptoms[14]. Tobacco smoke may also affect the function of CFTR, accelerating the decline in pulmonary function[14]. The study of risky behavior conducted by Mc Ewan et al.[14] reported that 45% and 47% of males and females in her study, respectively, tried cigarette smoking, but only 6% continued smoking throughout the study. These numbers were significantly lower than the general population, which was roughly at 21%, based on data at the time of the above-mentioned study. Feldman and Anderson[17] analyzed affects caused by cigarette smoke on various systems of the body. One system they looked at was the mucociliary escalator function. In patients with CF, mucociliary function is impaired because of the loss of Cl− secretion and Na+ absorption that reduces the thickness of the airway surface liquid overlying airway epithelia[17]. The researchers explained that the primary function of the mucociliary escalator system, which lines the luminal surface of the airways, is to entrap and expel pathogens from the lower airways. The mechanism involves the interaction between proadhesive mucus secreted from goblet cells and submucosal glands operating simultaneously with ciliated respiratory epithelium[17]. As pathogens are inhaled and reach this mucosal layer, they adhere to the viscous luminal gel phase

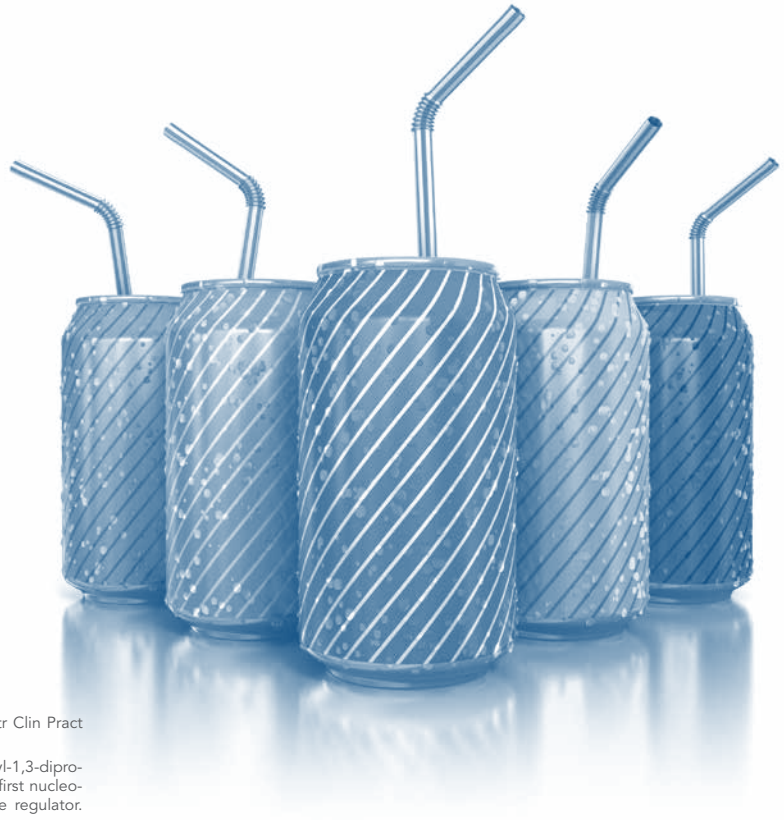
of mucus and are propelled upward toward the larynx, resulting in sputum [17]. When an individual inhales cigarette smoke, the damage it causes to this system is presumed to be caused by the cytotoxic, irritant, and intracellular redox signaling actions that decrease ciliary beating/denudation of ciliated epithelium, mucus hypersecretion/submucosal gland hypertrophy, and squamous cell metaplasia [17]. Furthermore, cigarette smoke has been reported to increase epithelial permeability by compromising the structural integrity of tight junctions of the respiratory epithelium [17].

Roughly 21% of patients with CF have been actively or passively exposed to cigarette smoke. The relationship between smoking and the severity of CF may be apparent in impaired growth, higher rates of lung infections, and a higher frequency of intravenous antibiotic usage [17]. The use of this substance, as well as alcohol and tobacco, can be seen as a risk behavior because of the increases in symptoms among patients with CF [14].

Summary

The consumption of caffeine may not be as severe as the other two substances, but continued use, or use associated with the general population, can pose threats to patients with CF and should be considered when creating various forms of therapy. The commonality shared by these substances is their exacerbation of pre-existing conditions experienced among patients with CF. Caffeine and alcohol's diuretic effect poses a great risk to dehydrated individuals with CF who are typically unaware of their fluid balance. Those who have used tobacco in the past or still do have increased risks of disease from the tobacco itself and the exacerbation of preexisting respiratory symptoms. Similar to what Mc Ewan et al. [14] stated at the conclusion of their study, risk behaviors should be carefully evaluated on an individual basis to determine the possible adverse outcomes of a patient.

The life expectancy of patients with CF has been increasing despite the underlying cellular defects caused by the disease. Patients born today are expected to have a median survival age well into their 60s [18]. The improvements are due to the introduction of pancreas enzymes, better nutrition, and specialized care, and some hypothesize that there are other factors that influence the survival of patients with CF [18]. Other than variances among patients and their genetic makeup related to CFTR genetics, some nongenetic determinants of survival include environmental influences [9,14,18] such as biological, social, cultural, and health care-related factors and include microorganisms, nutrition, pollutants, and others. The long-term survival of patients with CF can be strongly correlated to maintaining good health during childhood if the patient so happens to be diagnosed early [18]. By instilling good health behaviors in children who are diagnosed at an early age, long-term survival may be achieved. Further research may eventually demonstrate a stronger correlation between the consumption of caffeine, alcohol, and tobacco in patients with CF and reasons why they should be avoided to allow for long- term survival. But based on the current data, it is clear that lifestyle behaviors affect CF symptoms in various ways, often putting the individual at an increased risk above the general population. Individuals using these substances as meal replacements are at the greatest risk of further complications of both CF and chronic pancreatitis. Olesen et al. [10b] also noted that the further along the complications of chronic pancreatitis are, the higher the potential bioavailability of drugs in the body. As patients are being treated with various medications to treat the symptoms of CF, there are a variety of factors that must be considered, especially as they relate to the three substances reviewed herein. Future studies will dictate the course of treatment for patients with CF and what substances they should avoid to prevent further deterioration. ◀



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Natural products and complimentary therapies

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You are far from being the only one interested in complementary therapeutical approaches to relieve physical and affective problems related to cystic fibrosis (CF). More and more people follow non traditional approaches to treat some of their health worries. That said, remember that herbal or holistic medicine aren't always without risk. Most complimentary therapies are not subject to the same clinical tests as classic medication. Some supplements or herbs can even interact with medication you take for CF. Furthermore, don't forget that complementary therapies serve, as their name suggests, to complement your current treatments, and not to replace it.

When trying a new product or complimentary therapy, it is essential to pursue your medical treatment, to consult with competent professionals and to obtain information from existing associations, federations or professional corporation, to discuss treatment with your physician and with your CF clinic's medical team. These people will without a doubt have suggestions, answers and advice to give you.

Phytotherapy

Phytotherapy can be qualified as medicine with plants, or botanical medicine. It is the most common medicine in the world. It has been used in Asia for thousands of years. But beware: natural therapies are not necessarily without risk. Few studies have been done on natural products: it is therefore impossible to guarantee their effectiveness. As they are sold as nutritional supplements, they are not subject to regulation on prescription medication. Manufacturers are not obligated to mention the different ingredients on the labels. Beware of potential allergic reaction. Furthermore, what is indicated on the bottle label does not necessarily correspond to the content.

One of the best-known plants is obviously garlic. Without proven clinical arguments on garlic's health benefits, it remains one/ of the most popular supplements for the stimulation of the immune system and a contribution to recovery following respiratory tract infection. Garlic contains a component names allicine which helps to fight against cold symptoms (and even prevent its emergence).

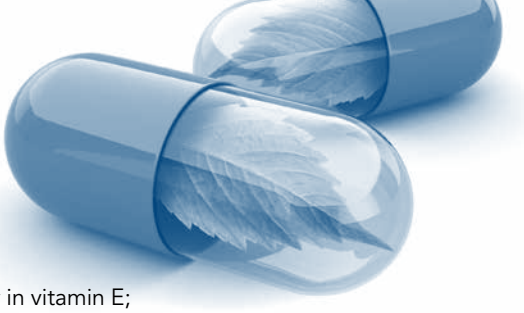
Some experts estimate that, to be efficient, garlic must be taken once a day, even without any cold symptoms present. Also, there is echinacea, a root that, it is believed, could reinforce the immune system and protect from the cold. It can also help to resist other types of infection. Opinions differ on its effectiveness. Some experts estimate that in association with ginseng, it indeed possesses immune system reinforcement properties. Others state that it has no virtues whatsoever and that it can provoke allergic reactions. Echinacea has no known side effects and is, generally, considered as a safe choice (although a little costly).

Pine bark extract and grape seeds are regarded as powerful antioxidants. Studies have shown that pine bark is beneficial for asthmatic patients, besides boosting resistance, increasing athletes' performance and contributing to the treatment of diabetes. According to experts, grape seeds contain an active component, which is a powerful antioxidant. Grape seeds have been used as an anti inflamatory substance to prevent infections; in Europe, they are frequently recommended for the treatment of allergic rhinitis and asthma. They have no known side effects.

Cider vinegar is also a very popular home remedy. It can stimulate digestion and the absorbtion of nutrients, and even relieve symptoms of asthma, flu and certain sinus infections. The usual treatment consists of taking two teaspoons of cider and one teaspoon of honey, in a glass of water, three times a day, half an hour before each meal. The vinegar must be unpasteurized and, ideally, organic and non-filtered. Other than the slightly acid taste of vinegar, there are no known side effects. Finally, some studies state that primrose oil reinforces the immune system, increasing circulation and reducing pain and inflammation caused by arthritis.

There also exist plant mixtures that improve some symptoms. Used for decades already in Europe, Gélo-Myrtol™ arrived in Quebec a few years ago. Available in capsule form, it contains eucalyptus leaves essential oils, myrtle, sweet orange and lemon peel, as well as other active ingredients. It is a mucolytic, meaning that it liquefies the mucus in bronchial tubes and sinuses, improving evacuation and facilitating expectoration.

Following a transplant, patients should avoid consuming unpasteurized foods such as cider vinegar.



Vitamins

To improve your nutritional levels and to prevent vitamin deficiency, you can take vitamin supplements and multiminerals, as well as other supplements, according to your needs. If you consider adding supplements to your diet, discuss it with your CF team first. Also, consult with your CF team before starting any alternative therapy based on supplements.

Vitamin A is an antioxidant that can protect against bacterial infections. You have probably already taken a vitamin A supplement in a multivitamin. Vitamin A is fat soluble, meaning that it is soluble in fats. Therefore, it is absorbed in smaller quantities by someone with CF. Nevertheless, it is important not to consume an excessive quantity of this vitamin to prevent potential poisoning. It plays an important role in the human body: it helps to see at night and allows the eyes to adjust when moving from a dark to a lit space. I you suffer from vitamin A deficiency, it is possible that your eyes cannot adapt from light intensity changes. Some people affected by CF have signaled this problem. If you think you suffer from these symptoms, speak to your physician. Vitamin A also helps to make the skin healthy and to fight infections. During childhood, it spurs growth. It can be found in eggs, liver, tomatoes, milk and some fruits and vegetables..

Vitamin C is also an antioxidant, which favours absorption of iron and contributes to reinforce the immune system. Some experts believe that, in doses sufficiently important (more than a gram per day), it contributes to mitigate cold symptoms. However, taking higher doses also comprises risks in certain cases. Because it improves iron absorption, vitamin C can lead to iron overdose and provoke a blood imbalance. Consult your physician before taking important doses of vitamin C.

Vitamin D comes from two sources. It is produced by the skin when exposed to the rays of the sun, and is also found in foods we consume. It helps the body to absorb calcium and to transport calcium from the blood to bones. If the body is deficient in vitamin D, bones can weaken and fracture risks increase. Although some food such as eggs and cereal products contain vitamin D, the most important source of this nutrient is milk. It is usually recommended for persons with CF to take vitamin D supplements to ensure a sufficient and continuous supply.

Vitamin E is more and more popular due to its role as an antioxidant. During growth, many changes occur in the body. As these changes occur, the body makes damageable products that can destroy healthy cells. Vitamin E helps the body to get rid of those products, and that is why it is called an antioxidant. A deficiency in vitamin E can affect the body's capacity to protect itself during an illness. For persons affected by cystic fibrosis, such a deficiency can have effects on their lungs and their respiratory capacity. Food that contains vitamin E includes vegetable oils, margarine and some fruits and vegetables. For persons affected by CF, food only cannot prevent a deficiency in vitamin E; that is why a supplement is usually recommended.

Most of the vitamin K we need can be found in food. It is found in green vegetables, in vegetable oils and in margarine. «Good» bacteria in our intestines also produce vitamin K. These bacteria can be destroyed by antibiotics taken to fight pulmonary infection. To optimally absorb supplements, it is preferable to take them with food and enzymes.

Vitamin K plays a very important role, because it helps blood coagulation. When you cut yourself, the time necessary for the cut to stop bleeding depends on the quantity of vitamin K present in your body. The longer the coagulation time, the more you risk having a vitamin K deficiency. Furthermore, vitamin K is essential in children's normal bone growth. To prevent a deficiency, many persons with CF take a supplement containing vitamin K .

Holistic therapies: a healthy mind in a healthy body

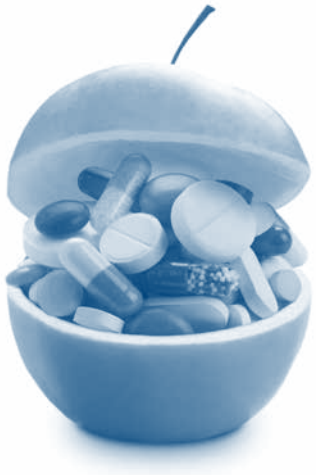
The reduction of tensions suffered by the body can contribute to relax the mind and to bring a sense of wellbeing. The following methods are part of the preferred holistic treatments, a doctrine that encompasses the entire human being, in its physical, emotional and mental dimensions.

The most known is certainly acupuncture, a 2,000 year old Chinese medical technique characterized by the insertion of needles in the body, at specific points. Some believe that a simple touch or stimulation of these points, called meridians, contribute to liberating the natural energy flow, for the system to itself trigger the healing process. It is also believed that acupuncture can successfully treat some respiratory problems (particularly mild cases of bronchial asthma) and gastro-intestinal problems (such as hiccups, duodenal ulcers and constipation). It is not painful but some people find it uncomfortable.

Although everyone is different, in most cases treatment requires five to fifteen sessions. When practiced by a certified acupuncturist there are few side effects. However, the following events might happen:

- The acupuncturist may forget a needle.
- Your symptoms may get worse following a session. This generally doesn't last more than two days.
- You may feel euphoric after a session. This is related to the release of endorphins, or the "feel-good" chemical in your brain. Patients are encouraged to relax for 20 minutes following a session.
- Pneumothorax has been reported.

Certain medications may interfere; check with your physician or your acupuncturist and perhaps your pharmacist.



For centuries, people have used the healing power in their hands and fingers to relieve all kinds of health problems. It makes sense when you think of how we naturally want to rub an area of our body that hurts, for example, your stomach when you have a stomachache. Some experts believe that massage can block the painful stimuli carried by certain nerve fibres. As well as relieving aches and pains, it can improve your circulation, digestion, reduce fatigue, and relieve tension. Massage techniques are numerous. For example: the Japanese shiatsu technique by which the masseur applies strong pressure using thumbs, palms, elbows or even feet. This pressure is applied on acupressure points on the body to restore the balance of the energy flow.

There is also the reflexology theory, where each organ corresponds to precise points on the hand and feet. The application of pressure on these points contributes to appease tension in other body parts. Californian and Swedish massages can also give good results, as well as reiki, which is different from massage sessions as muscles are not directly manipulated. Instead, the Reiki master gently applies his hands on different body parts for five minutes.

Aromatherapy

Aromatherapy is a system of caring for your mind and body with the scents of essential oils derived from plants. According to some experts, smelling certain aromas have a direct effect on brain activity. Whether they are inhaled directly, massaged into your skin or added to your bath, these oils can help relieve pain, ease tension, and restore your energy and sense of well-being.

There are literally as many types of oils as there are plants. Here are some choices to consider. ◂

Pine

May have antiseptic, antibacterial and inflammatory properties. May also be beneficial when inhaled for treating colds and flu.

Lavender

Used for centuries to alleviate stress.

Eucalyptus radiata

It is the kind of eucalyptus found in dry saunas. It is known for its bronchodilator properties. It is generally well tolerated and rarely provokes spasms.

Eucalypsus globulus

It is known for its antiviral, antibacterial and antifungal properties. Beware, it is less well tolerated and must be avoided with young children!

Hyssop

It possesses expectorant properties and is used to relieve asthma, cough, flu and different allergy symptoms.

CYSTIC FIBROSIS **RESEARCH** IN 2015

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The 29th North American Cystic Fibrosis Conference took place in October 2015 in Phoenix, Arizona. It was the occasion for scientists and clinicians throughout the world to share their discoveries and to establish new collaborations for research in fundamental science and clinical applications. One the major themes that emerged from this conference is the increase in clinical research collaboration on persons affected by cystic fibrosis. It arose that cystic fibrosis is a particularly variable illness, both in its clinical expression as well as the composition of genetic anomalies. However, new medication developed for cystic fibrosis are specific to particular mutations which forces the international community to establish collaborations to accelerate the discovery of new efficient products adapted to the particular genetics of each individual (personalized medicine).

The discovery that had the most clinical impact in 2015 was that of clinical data linked to the partial correction of the CFTR defect in the population with two F508d mutations, the most common defect affecting 50% of persons with cystic fibrosis in North America (Figure 1). A combination of two medications, Lumacaftor and Ivacaftor, known commercially as Orkambi, is available on the American market. This new medication allows two chemical molecules to work in synergy where Lumacaftor favours the progression of the abnormal protein to the cellular membrane and Ivacaftor (Kalydeco) activates the opening of CFTR once this protein is situated in the membrane. Clinical research implicating more than a thousand patients over a period of 24 weeks demonstrated a modest, but significant, improvement of the sweat test as well as other clinically pertinent parameters such as respiratory function,

weight increase and a reduction of around 50% in the number of respiratory exacerbations. This last data is particularly pertinent in the clinic as we know the number of respiratory exacerbations is strongly associated with a decline of respiratory function.

The undesirable effects of Orkambi are relatively less pronounced and, interestingly, one of the undesired pharmaceutical effects include changes in intestinal habits where patients who were previously constipated report more frequent stools and sometimes complain about diarrhea. This undesirable pharmaceutical effect could be directly related to a partial correction of the defect in chloride transport in the intestine. Daily clinical experience is very limited as it is not approved in other countries besides the United States and that the medication has been on the American market for only a few months. Preliminary data suggests that its clinical efficiency is perceptible by users.

Three great challenges persist about the new medication to treats patients with two F508d mutations. First, the molecular efficiency of the medication is still very limited if relied on the sweat test correction which is only 11%. A correction of 50% or more would be desirable to reverse the majority of symptoms and clinical expression of cystic fibrosis. Second, before the medication can be prescribed, it must be approved by government medication regulation agencies such as Health Canada[1]. The evaluation made before the approval of new medication includes an analysis of benefits versus the undesirable pharmaceutical effects. Health Canada[1] currently studies this file in order to state on the potential approval of Orkambi in Canada. The third challenge, but not least, is to reimburse the high cost of the medication to persons or families affected by cystic fibrosis. At $710 USD daily in the United States (according to Associated Press in November 2015), Orkambi isn't cheap. Any new medication developed for a rare illness induces very high costs, because the amount of necessary investment to bring new molecules to market remains the same regardless of the frequency of the illness for which it is developed. However, the pool of persons affected by a rare illness being very restricted by definition, companies having invested in the development of specific molecules for this illness must recover the costs of this investment by predicting that the sales volume won't be comparable to that of frequent conditions such as arterial hypertension or hypercholesterolemia. It is challenge not only for families affected by cystic fibrosis, but also for all individuals in society. We must show social solidarity in a sustainable financial framework, as we are susceptible to be treated for a rare illness someday.

The efficiency of new medication seeking to treat the base defect in cystic fibrosis remains suboptimal and many clinical research studies are ongoing to evaluate new molecules which is particularly true for homozygous persons (two identical mutations) for the F508d mutation. Two great international studies are underway to study a new generation of improved molecules which could turn out to be better than Orkambi. Furthermore, there is currently international research to study Ataluren in persons with cystic fibrosis with a class I mutation, a mutation which prematurely blocks the transcription of the gene encoding the CFTR protein.

Many nutritional intervention projects are underway. The link between the nutritional defect and the excessive inflammation in patients carrying cystic fibrosis is better and better known. A deficiency of certain essential fatty acids exists in the membranes of those affected by cystic fibrosis and a deficiency of those fatty acids of which omega 3 oils are part could be a factor favouring inflammation. Clinical research is in develop-

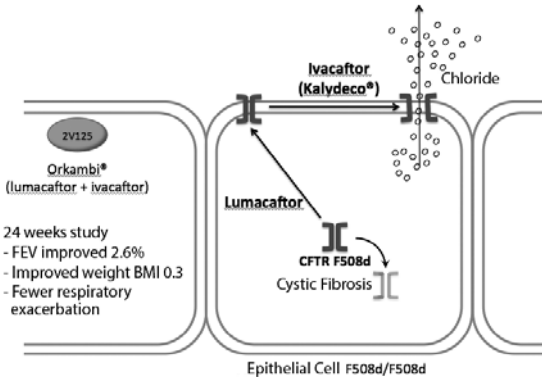
ment in Montreal, Sherbrooke and elsewhere in the world to restore a favorable lipidic membrane profile and, therefore, counter inflammation caused by cystic fibrosis. If participation in a research project interests you, you are encouraged to discuss it with your clinical director or with the staff at your cystic fibrosis clinic. These professionals are aware of the different research programs.

Finally, all participants at the North American Cystic Fibrosis Conference have benefited from the plenary session on the topic of psychological well-being, depression and anxiety associated with cystic fibrosis. These elements which previously were only rarely discussed have become priority themes for scientists and clinicians associated with the different cystic fibrosis clinics throughout the world. It is recognized that this often neglected aspect is particularly important in the care offered in cystic fibrosis clinics.

To resume, the international North American cystic fibrosis conference was one that disclosed the greatest quantity of new information allowing health professionals to hopefully change the course of the illness affecting cystic fibrosis patients. We left Phoenix optimistic, but realist, while recognizing the quantity of work left to do to fulfill these promises translating into an improved life quality for all individuals in all our cystic fibrosis clinics. ◄

Figure

Illustration of the consequences of mutation F508d of the CFTR and its pharmacological correction. The CFTR derived from the F508d mutation has two important defects. First, it is deformed and is degraded before being able to migrate to the membrane. Second, it doesn't open normally. Lumacaftor facilitates migration to the membrane and Ivacaftor (Kalydeco®) stimulates its opening. Orkambi® is a new medication cmposed of two molecules, Lumacaftor and Ivacaftor, which allows a partial correction of the fundamental defect associated to the CFTR derived from the F508d mutation. This is followed by an improvement of the permeability of the F508d/chloride F508d epithelial cells.



[1] Orkambi has been approved by Health Canada on January 26, 2016.

HEALTH COLUMN

PANCREATIC ENZYMES

My 6-year-old son often refuses to take his pancreatic enzymes with meals. Do you have some tips and advice for him to accept taking them? What exactly do these enzymes do?

When food is ingested, it goes into the stomach where it is reduced into small pieces. These pieces then pass into the small intestine. In a healthy organism, digestion is done with the aid of bile (liver) and enzymes (pancreatic). Essential nutrients are then absorbed by the body and converted into energy used for growth. Undecomposed food (undigested) are excreted from the body in the faeces. In most people with cystic fibrosis (CF), the pancreatic ducts are clogged with the same type of sticky mucus that is found in the lungs. The enzymes secreted by the pancreas are then unable to travel to the small intestine, and the food is not digested properly, resulting in problems of nutrition and growth. This is why people with CF generally must take pancreatic enzyme supplements to promote healthy digestion. The required amount varies depending on age, body weight, and the quantity and quality of the food ingested. It may vary over time, especially with children.

Most children can learn to swallow enzyme capsules between the ages of three and seven. The capsules must not be chewed or crushed and their contents should not be sprinkled on food. When the child starts the enzyme intake, weight gain is a sign that the body digests and absorbs food well. Never increase the amount of enzymes without talking to your healthcare team. Indeed, it can be dangerous to take too much.

It is often difficult to make children take enzymes. The younger, for example, may simply reject it if they feel that it angers the person who shows them how to take them, while school children are sometimes embarrassed to take their enzymes at school. Your child will probably be willing to take their enzymes if, from the start, you show yourself as constant and are frank about the issue. During early childhood, explain why enzymes have to be taken while eating. Explain that they will help them grow and gain weight, that they will prevent flatulence, stomach pain and loose stools, and that by taking them, they will have more energy to play with other children.

School-age children are sometimes embarrassed to take their enzymes in the presence of their friends, which may deter them from taking them at school. To avoid this situation, it is better to adopt an open attitude and to integrate the taking of enzymes in the daily routine at home and at school.

Some foods can be digested without enzyme supplements. Fruits (including dried fruit), juices, soft drinks, hard candy, popsicles and jellies (Jell-O©) are good examples.

Finally, for mothers who are breastfeeding, it is important to give babies some enzymes with each drink. If your baby is underweight at the diagnosis of CF, you may need, in addition to breastfeeding, to give them enriched breast milk bottles for at least a few weeks. After the weight is returned to normal, you can stick to breastfeeding.

— By specialist dietitians for Cystic Fibrosis in Canada.

PETS

I would love to get a pet. However, people around me said that an animal could be detrimental to my health. Are there any animals that pose risks to the health of people with Cystic Fibrosis?

First, be aware that it is important to choose a pet well. Some animals do not really have an impact on health if the necessary precautions are taken. However, other types of animals are simply not recommended. People with Cystic Fibrosis (CF) may be concerned about two types of risk: the risk of infection, and allergic risk. Many studies have proven the therapeutic properties of living with a pet for people with a disease (zootherapy). It is therefore important to consider the psychosocial benefits of having a pet and the potential risks to health.

Exposure to environmental allergens (like animal dander) leads to the worsening of lung functionality in other obstructive lung diseases such as asthma. The effect of animal exposure on lung functionality in people with CF remains uncertain. Therefore, reducing environmental risks remains essential to maximize lung health in CF.

A multicentre study of 703 patients with CF in the United States has shown that owning a cat is associated with a greater prevalence of nasal polyps, and owning a dog or cat is associated with greater prevalence of wheezing, which could lead to reduced lung functionality, especially between the ages of 6-8 years old. In this study, the authors have not demonstrated a significant correlation between owning a cat or dog and the prevalence of MRSA or Pseudomonas. Nevertheless we must set out the possibility of animals as a source of respiratory pathogens, and also as being a potential problem. A limited sample of CF patients and their companion animals (n = 20) identified them as carriers of the same bacteria / fungi.

However, it turns out that when the usual hygiene measures are met (ex.: frequent hand washing), owning a cat or dog has no major risk. By taking precautions, you do not run a higher risk of infections or allergies than a person without CF. The presence of an animal can be problematic if you are already allergic. Before adopting a pet, first make sure you do not suffer from such allergies! It is best to discuss with your healthcare team about the possibility of allergy tests before obtaining a pet.

On the other side, some animals, like birds or aquarium animals, are not recommended. These two types of animals can be problematic for people with CF because of their risk of transmitting bacteria. It is best to avoid contact with birds as they are often carriers of problematic germs such as Chlamydia, atypical mycobacteria, and aspergillus. Animals living in aquariums increase the risk of transmission of Pseudomonas aeruginosa and atypical mycobacteria by their humid environments.

So a new pet for you or your child? No problem, but you have to choose wisely, and discuss it beforehand with your CF team, who know your health condition.

— Joanie Bernier, Nurse, B.SC
Cystic Fibrosis Clinic at the McGill University Health Centre, Montréal

PREVENTION OF FOOD POISONING AFTER TRANSPLANT

I am a big fan of meat and fish. Soon I'll be waiting for a transplant, and while doing some research I learned that the transplantee must avoid certain foods, including sushi, deli meats, and even certain cheeses. Why is this?

When the immune system is weakened, as will be the case following the transplant, with the taking of immunosuppressive drugs, the body isn't as efficient at defending against infections. The risk of food poisoning is thus higher. Also known as foodborne illness, food poisoning is an infection of the digestive system. It occurs when one consumes food contaminated with bacteria, viruses, parasites, or the toxic products they secrete. This is an infection that can be dangerous and that may cause long-term medical complications, and sometimes even death. Symptoms may include nausea, vomiting, abdominal pain and cramps, diarrhoea, fever, or chills.

The best way to avoid this type of infection is prevention.

In addition to the health and safety measures in handling and storing food, some foods may have a higher risk for immunosuppressed people by the way they are produced and stored. Food poisoning risk can be reduced by avoiding certain types of food.

Thus, when raw or undercooked, some foods - including meat, poultry, fish, seafood, and eggs - can be risky. Steps to take are to ensure foods are cooked up to a safe temperature. The heat deployed by cooking will ensure the destruction of microorganisms. Sushi lovers may opt for sushi made with cooked fish or seafood, or vegetarian sushi, while ensuring that they have been prepared with care to prevent any risk of cross-contamination with raw fish.

As for processed meats, including deli meats, liverwurst (pâtés), and refrigerated meat spreads (including cretons), though cooked in the process of preparing, their handling and their nature (moisture, acidity) increase the risk of contamination and growth of micro-organisms. If you still desire to consume deli meats, it will be advisable to warm them at a high temperature until they are steaming. Liverwurst (pâtés) and meat spreads sold in cans or for refrigeration after opening only, are considered safer.

Likewise, raw milk cheese (made with unpasteurized milk), soft, semi-soft, and veined (blue) cheese, are also considered foods at risk for immunocompromised individuals. The risk will be lower if these cheeses are integrated in recipes where sufficient heat is deployed (gratin, cheese fondue, raclette, baked brie). Otherwise, it is recommended to opt for cheeses made with pasteurized milk and hard cheese.

— Valérie Jomphe, DtP, MSc, CNSC
Nutritionist of the lung transplant program at the CHUM, Montréal.

Made possible through the financial support of Merck Canada Inc.

LIVING WITH CYSTIC FIBROSIS

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Living with Cystic Fibrosis
mission is to promote quality of life
for people living with cystic fibrosis.

VISION

- Reach all people living with CF and their families.
- Be a leader in transmitting information on CF.
- Be a first hand support for people living with CF.

OBJECTIVES

- Promote and protect the rights and interests of those living with CF.
- Represent and support people living in Quebec with CF in their relations with government organisations.
- Transmit accurate information.
- Offer support to those living with CF.
- Promote a healthy lifestyle for those living with CF.