46 | SVB 2023

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LIVING WITH CYSTIC FIBROSIS

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LIVING WITH CYSTIC FIBROSIS

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CONTENTS SVB 2023

Testimony

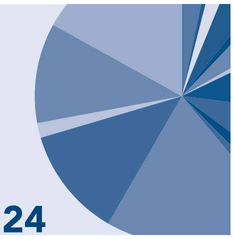
Interview

Psychology

Trikafta

Socio-economic study

Research



A new year

Last October,
Living with Cystic Fibrosis celebrated its 37th anniversary.
What a pride!

When it was created, the median age of survival for people living with cystic fibrosis was around 25 years old. The founding members of our organization were then young adults with CF who had barely emerged from their teenage crisis! In its debut, our organization aimed, among other things, to equip our members (exclusively CF adults in Quebec) to enable them to develop their financial, romantic and health management autonomy. Our organization met their need of recognition of their independence from their parents!

Since then, water has flowed under the bridge and we have become accomplished adults; we can hope to study, have a fulfilling job, travel, have friends, have a partner and a loving and helping environment. We can even become parents in our turn (I have the joy of being the mother of four children). We can recognize that overall, our realities have evolved positively.

As mentioned in my note from the summer 2022 magazine Vivre Express, our organization experienced a historic moment during our last annual general meeting:

"Since April 2022, members close to a CF person can become a member and benefit from our services! May this openness allow you to be surrounded and included!”

Today, I would like to give you the advantages of being a member with us:

- Be invited to each annual general meeting.
- Speak at these meetings.
- Vote at these meetings.
- Submit your candidacy for an administrator position, and possibly be elected.
- Be the first to receive the organization's publications.
- Be the first to receive all the important information about cystic fibrosis.
- Be the first to receive invitations to conferences, information/educational activities, workshops, symposiums, or other events organized by the organization.
- Have access to our financial assistance and support programs.

So, whether you live with cystic fibrosis or are part of the immediate family of someone with CF (parents/in-laws, children, legal guardian, designated caregivers), you can become a member with us for free!

Don't hesitate any longer, fill out the online registration:
https://vivreaveclafibrosekystique.com/devenir-membre !

What a pleasure it will be for us to better understand the needs of those around CF people in Quebec and to discuss our respective realities!

Charlène Blais
President, **Living with Cystic Fibrosis**

A new General Manager? Again!

With the particularity that I come from the private sector with a mad desire to finish my career in the universe of an NPO and to continue to work for a long time. With the pretension of not changing the essence of the mission and values of Living with Cystic Fibrosis, carried by extraordinary and dedicated people both on the Board of Directors and on the permanent staff.

My little added value as the new General Manager will be to provide grist to the mill ($$) by making sustained efforts in terms of our funding sources. And who says more funding means more assistance programs and services for our members. To use a popular expression: put our feet where our mouth is and optimize programs to serve our members.

This is my wish for the new year and take care of you and your loved ones.

Mario Bertrand
General manager, **Living with Cystic Fibrosis**

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Something new!

Dear readers, we are always pleased to offer you our latest issue of the SVB.

During this year, we have had many movements within our organization: departures, but also arrivals. First, I would like to thank Josée Côté and Valérie Lacroix for the work we were able to do together and for the good times we shared, wishing them every success in their new adventures.

Second, I want to welcome the members of our new task team. Mario has become our new General Manager and will have the difficult task of finding the necessary funding to achieve our ambitions. Amélie, whom you already know for her involvement in the magazine, has joined us on the team and is now responsible for the organization's communications. Finally, Viviane, formerly the Vice President of the Board of Directors, now supports the administration. No doubt we will do great things with this team.

In this issue, we have decided to highlight NPO Sound of CF, by introducing its two founders, Gabrielle and Jeffrey, who agreed to answer our questions for a most interesting interview. We offer, as usual, various testimonies, as well as an article on cystic fibrosis research and questions on health issues.

In addition, we decided to focus on the psychological effects of Trikafta. To do this, we were able to contact Jaqueline Noordhoek, CF Europe President, and Anna Georgiopoulos, a researcher in Boston. They agreed to participate in our magazine, and we thank them warmly. On another topic, Marie-Michèle Ricard, psychotherapist, presents an article on body image.

For our part, we have completed the compilation of the data collected as part of our questionary to determine the socio-economic profile of the CF population in Quebec. We present to you an infographic of the highlights, but you can find the entire profile here.

We hope you will enjoy reading this issue. Good reading!

Sébastien Puli

Coordinator, **Living with Cystic Fibrosis**

A complicated life

Carol Côté

Québec, Quebec
Canada

Aging with cystic fibrosis brings its share of difficulties and challenges. Our bodies age faster and expose us to more diseases and incidents of all kinds.

I was born in Saguenay, the eighth of 10 children. When I was little, I was told I had asthmatic bronchitis. When I was three months old, I had my first pneumonia. I was rickety as a child, and I often had asthma attacks. As I had no medicine, my solution was to go to bed and the next day, I would feel better.

In my teens, things got better. I grew up and gained weight; everything was better for me. However, I had a lot of allergies: animals, dust, and weeds. I did a bachelor's degree in administration specializing in marketing. I then started a career in shopping centers for a subsidiary of the Caisse de dépôt (Ivanhoé Cambridge). I was domiciled in Quebec and stayed there for 32 years.

At 30 years old, I got married to an extraordinary woman, France. She had no idea that my health would be fragile and complicated, but she accompanied me faithfully through every new problem and I could always count on her support. When the time came to have kids, it didn't work out. I was operated on to see what was wrong and was told my deferential canals were blocked. The doctor met with me to tell me that it happened in families where there was cystic fibrosis. France and I therefore decided to go for artificial insemination to have our children. It worked, we had two beautiful boys who are now 27 and 29, Benjamin and Jérémie. They are two magnificent humans of whom we are very proud.

After the children arrived, my health deteriorated due to fatigue and lack of sleep. In addition, my work led me to travel throughout Quebec. I started having repeated pneumonia, two or three a year, but over time it was between five and eight a year. The doctors therefore decided to investigate, and I was hospitalized for a full check-up. The diagnosis given: cystic fibrosis.

So, I find out, at 38, that I have this disease. From then on, things just got worse. In the last years before the transplant, I had pneumonia every month with a succession of hospital stays and IV antibiotics every two weeks. It was very difficult with my job. I had no more energy. Every time, the doctor returned me to work.

At the age of 46, I developed a bacterium that they had difficulty diagnosing, stenotrophomonas maltophilia. This bacterium causes complex infections to treat because it is multi-resistant to antibiotics. I was hospitalized, on oxygen. I went home with oxygen. I was put on long-term sick leave and since my income had gone down, we had to resign ourselves to selling our house and buying a smaller one.

During a medical meeting, Dr. Josée Villeneuve offered me a lung transplant, because my lung capacity was down to 30%. I accepted immediately. My children were 11 and 13 and I wanted to see them grow up. I was almost two years at home on standby with 24-hour oxygen therapy.

On May 28, 2008, I received the long-awaited call from the surgeon, Dr Pasquale Ferraro, who told he had perfect lungs for me. Half an hour later, my spouse and I were in the ambulance heading to Notre-Dame Hospital. Upon arriving, Dr. Ferraro was at the door with my sister Pauline, from Longueuil, who is a retired nurse. The operation went well and despite some complications, I was discharged from the hospital after five weeks.

What a beautiful gift I received! The gift of a second life where I was able to see my children grow up, finish their studies, and get to know their spouses. This true miracle was possible thanks to a donor who saved several lives and I thank him every day. I would also like to thank his family for accepting this donation.

It will soon be 15 years since I had a transplant. I had been told that I would exchange one disease for another, and they were right. Over the years, I developed different complications like lung infections, my left lung collapsed (atelectasis), pancreatitis, vascular problems in my legs that forced me to undergo bypass surgery and even the amputation of my right leg, eye surgery, and kidney failure which led me, a year and a half ago, to receive dialysis treatments three times a week in the hospital. I now live separated from my wife, in Saguenay, near my family. My wife was no longer able to accompany me through my illness, too much stress and anxiety for her. However, we are on good terms, and we see each other as often as possible with the boys.

Life has been good to me despite everything, and I have no regrets. I hope it lasts long enough to see my grandchildren looming on the horizon.

Thank you! ◂

Having a sick child

Johanie boivin
Québec, Quebect
Canada

Having a sick child is probably one of the greatest challenges one can face. It is necessarily an element that disrupts all spheres of our lives: family, couple, work, routine, etc. Living with cystic fibrosis daily means learning to juggle with a lot of additional parameters. Already being a parent is not always easy, being a parent of a girl with cystic fibrosis can push you to your limits.

There is obviously the shock of the diagnosis which triggers a trying period because there are many things to digest, to assimilate. However, looking back, I feel like it was nothing compared to the other obstacles we had to face. Of course, each person experiences hardships differently, each family does not go through all the same difficulties. In our case, our daughter has many challenges. In addition to cystic fibrosis, she has learning disabilities, attention deficit disorder and other health issues... Let's say that at 14 months, the diagnosis of CF was only just the tip of the iceberg for us.

Of course, it is difficult to see your child coughing and spitting. Having to do clapping on his little body for 20 minutes every day. Asking yourself whether to give them antibiotics, going to the emergency room, etc. Despite all this, the hardest thing in my opinion are not the physical symptoms, but the psychological difficulties. In addition to the ups and downs of this often unpredictable disease, there are fluctuations in morale. Convincing your child to do her treatments when she had a bad day or just had a crying fit. When our child accumulates difficulties, the situation becomes more complicated. We must constantly adjust, review our routine, our strategies, our priorities.

To top it all off, cystic fibrosis is an invisible disease, meaning that for others (even sometimes the immediate entourage), it is easy to forget. Most people do not understand what it really means on a daily basis and tend to trivialize its impact on our lives. Sometimes, we were told that our daughter’s health looked good when her FEV1 (maximum expiratory volume in a second) had been in free fall for several months, or even that hospitalization was planned a few days later. Under these conditions, it is sometimes difficult to find an attentive ear to listen to us, to understand us. We can easily feel inadequate, by being told that we talk about it too much, when we are only talking about our daily lives.

So, for me, managing the “physical” part of the disease, although difficult, remains less difficult than juggling all the other aspects. Our child's illness, whether we like it or not, interferes in all spheres of our lives. How many jobs did I quit because work-family-health balance was not possible, or simply because my bosses clearly lacked empathy. Managing your own emotions is one thing but supporting your child in this ordeal is another! Parenting is not the easiest job to start with, especially since there is really no diploma for it. And with this kind of additional component, it can quickly become beyond our scope.

The medical care of our daughter is all in all well provided by the clinics. When it comes to physical symptoms, FEV1, everything is going relatively well. The disease is taken care of, but not our child and not us. As soon as you go beyond the physical aspect, it becomes hazardous. Every professional does a great job, it goes without saying, but they don't have the resources to really help us. Despite all their good will, their help unfortunately stops there. Personally, I learned it the hard way. Asking for help and saying how we really feel can easily backfire.

Between 2018 and 2020, we really went through a tough time. Our daughter, who was about 10 years old, was in the middle of a period of opposition disorder. It was war morning and night for her treatments. Despite her poor performance at school, we often dodged homework. Convincing her to do her treatments was already a feat and very often, the evening was spent there. And the good mood too. And the vicious circle sets in, the hand caught in the gear. We have consulted a lot of workers since our daughter was 5 years old:

psychologists, neuropsychologist, psychoeducators, occupational therapists, social workers, etc. Everyone seemed to have the same vision: everything is fine, it will pass. At the end of 2019, during a clinic appointment, we reiterated our request for help, explained our exhaustion, our inability to convince her to do her treatments properly. We were offered a motivation calendar. I tried to list all the types of calendars that we had already created. We wanted concrete help, like a physiotherapist who comes by the house a few times a week, for a few weeks, to breathe a little. We were sold the idea of the motivational calendar again! It was too much. I said I didn't want to hear about their "damn" calendar anymore. We were taken aside to tell us that our daughter's FEV1 kept falling and that if we did not reverse the trend quickly, they would be required to report it to the DPJ. According to them, we were responsible for our daughter's poor health. I froze, unable to defend myself. Words did not come out. Unable to believe that speech. By saying these words to us, the doctor was crying, and I then understood that they had to warn us. Basically, they have no resources to offer us.

In early 2020, our daughter had sinus surgery and the hypothesis that the Pseudomonas had lodged in her sinuses and not in her lungs was then confirmed. Two months later, she finally stopped the nebulized antibiotics she had to take twice a day since 2016. At least forty minutes gained each day to live her teenage life. At the same time, we discovered that she had sleep apnea and, in the fall of 2020, her tonsils were removed. These two surgeries have changed our lives. She is sleeping better and is therefore much less irritable, more concentrated at school. She regained confidence in her abilities. She has more time for herself and is therefore more inclined to do her treatments. Ultimately, the symptoms back then which were mostly psychological had everything to do with her illness, her physical condition.

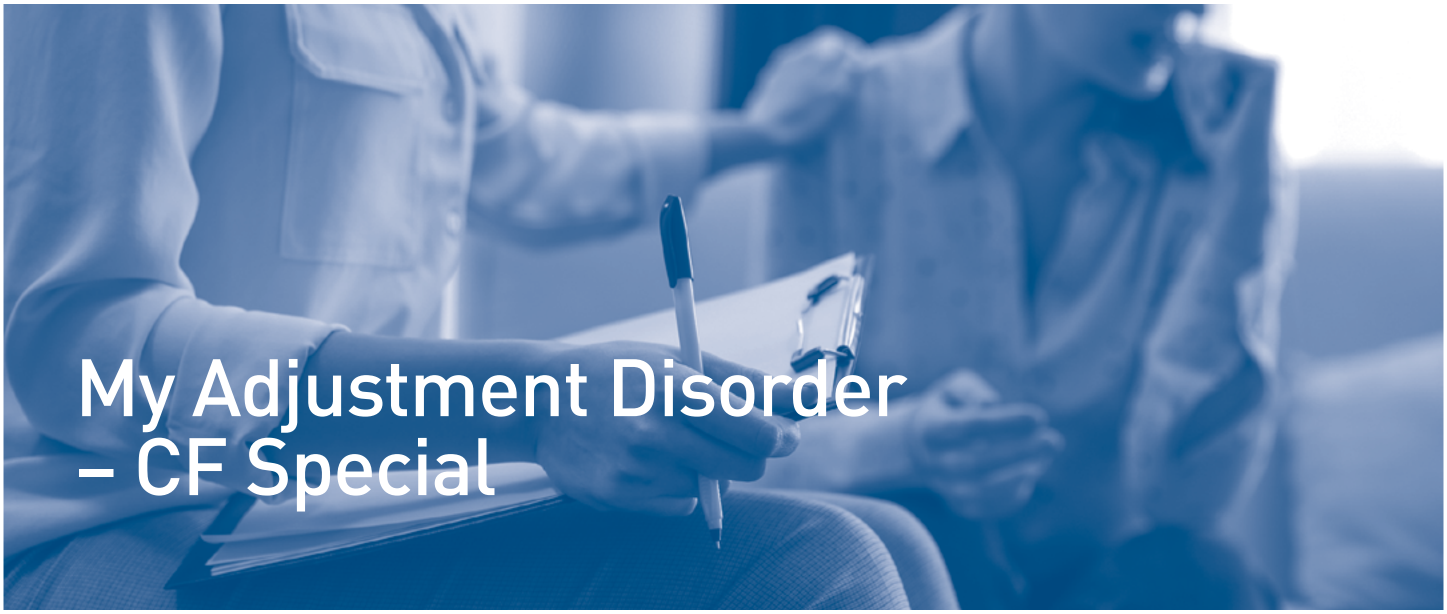
The disease is not an addition of symptoms, certainly not for a chronic degenerative disease like cystic fibrosis. Learning to live with CF, to "accept" it, for a growing child who already has to deal with a lot of emotions and hormones, is virtually impossible to do without a hitch. We often hear that learning about a diagnosis is a bit like experiencing grief, with the five "regulatory" stages. You surely know them: denial, anger, negotiation, depression, and acceptance. But that's just a theory because in practice, I doubt that this is how it works. One of my daughter's pulmonologists had already given me this ready-made formula: according to him, we were stuck in the anger phase, and for this reason, our daughter's health was not improving. If we follow this logic, as a parent, we are no longer entitled to be upset in the hospital, otherwise we are labelled as a parent who does not accept his child's illness. And you and I know that with our healthcare system, anger is often an understatement!

From experience, I believe that, although these famous five stages are rather true, it is the idea of the phases that does not make sense. As if it had to be a logical sequence that necessarily leads to the final stage: "acceptance"! In my opinion, these are emotions that we experience in disorder, without logic and above all, without finality. This is even more true when it comes to a chronic disease such as CF, which comes with its share of additional diagnoses. It is not a linear disease, but rather a physical and emotional roller coaster. Also, the whole family experiences these emotions at a different pace, where other emotions are added, such as misunderstanding and disappointment. Sometimes, like it or not, it can set us back a phase or two. Seeing your child angry at their illness, hearing them say, "It's unfair, why is it happening to me and not to someone else", even though we thought we had already "accepted" CF, it doesn't take more than that to get us back into that anger as well.

I always write "to accept" between quotation marks, because although we often hear this expression, I admit that I don't like it, I don't believe it. I'm not an expert, I don't have a degree in psychology. However, in my experience, aiming for acceptance as the ultimate goal is more damaging than anything else. For me, "acceptance" is more about understanding that we are passengers on this roller coaster, for better or for worse. Recognizing that we have already passed several "lows" and hoping to stay on the upward slope for quite some time while knowing that the trough could come back sooner than we think. Hopefully, with time and a lot of effort, we will go up the next hill more easily. The important thing is to allow ourselves to experience all these emotions, to welcome them and to try to understand them. It will be easier to discern them and cross them during the next descent. And above all, don't forget that despite bad experiences, exposing our vulnerability in order to find help is sometimes necessary. That is a real proof of courage. ◄

About Johanie Boivin

- Mother of two daughters, including Axelle, 13 years old, CF.
- Cofounder of Tousse ensemble in 2013
- Board member of **Living with Cystic Fibrosis** since 2021



My Adjustment Disorder – CF Special

Amélie Payment

Salaberry-de-Valleyfield
Quebec, Canada

Many people with cystic fibrosis also have other health problems that can be divided into two categories: physical and mental. Physical problems such as diabetes, endometriosis, high blood pressure or asthma are quite well known for people with CF. Then, for most cases, they are treatable. However, what happens to mental problems such as the famous adjustment disorder? I say it like that, but with CF, it's a much more common reality than you might think! I have the impression that the nursing staff and our loved ones may tend to trivialize things that shouldn't be. I write this today to normalize the fact that coming to terms with a physical or mental health issue is a different journey for everyone.

First, when I was diagnosed with diabetes, the waiting time to get an appointment with a psychiatrist was very long. Even to this day, I still haven't had a follow-up. However, I strongly believe that poorly managed anxiety can do as much damage as a vitamin deficiency. Let me explain myself: when we have a vitamin D deficiency, we can have symptoms such as being more tired, being more vulnerable to catching viruses, be depressed and even have dizziness. If you have ever had severe depression or anxiety like me, you may have experienced some similar symptoms. During my anxiety attacks, I experienced fatigue, demotivation, insomnia, dizziness, panic, and I even fell ill from depression. Do all these symptoms ring a bell? If so, you see, you are not alone.

In addition, I often feel that adjustment problems are overlooked. For example, a person who learns of a new diagnosis may feel depressed. This is a common and normal reaction. However, I feel that talking about it openly is perceived as a taboo subject. I remember when I found out I had diabetes. The nurse was trying to explain to me how insulin works, and that I could not leave the hospital until I had given myself at least two injections. I remember him saying to me: “It shouldn't be too difficult for you to give yourself injections, you must be used to it.” The truth is, yes, I was used to being sick and going through a lot of treatments. On the other hand, injecting myself three times a day for the rest of my life was certainly not part of my “routine”. So, here's my point: at that moment, I felt like my fear was not valid.

Then, after my depression in the face of my adjustment disorder, I had an avoidance phase. I avoided endocrinology follow-ups like the plague. I no longer wanted to do my pulmonary treatments anymore, I even left suddenly once from an appointment because I was feeling downright suffocated. The burden of care for cystic fibrosis, diabetes and anxiety had become too heavy. That's when my social worker at Sainte-Justine told me I had an adjustment disorder.

Despite everything, I got through it, because I had the support of my lover and my family. Now I'm not ashamed to talk about it anymore because for me it was the path that I had to take to feel good about myself again.

To conclude, my adjustment problem at the age of 18 was a big ordeal. So, I'm making a heartfelt cry: keep asking for help. Mental health is as important as physical health. If getting access to a psychiatrist is too difficult, **there are always resources to help you such as:**

- **Info-Sociale**: 8-1-1 extension 2
- **Aide et prévention du suicide**: 1 866 APPELLE (277-3553)
- **Tel-Jeunes**: www.teljeunes.com
- **Centre de crise du Regroupement des Services d'interventions de crise du Québec** (RESICQ): www.centredecrise.ca/listecentres

Founders of «Sound of CF»

Interview with Gabrielle and Jeffrey

Meet two founding CF members of the NPO Sound of CF (FKM), an innovative and creative project for the cystic fibrosis community!

Interviewed by **Sébastien Puli**

Can you introduce yourselves?

GABRIELLE: Gabrielle Phaneuf, 29, creative, different, ambitious, determined, very involved in my projects. I am the girl who adapts to everything, but who doesn't fit anywhere. I like yoga, dancing, cooking and electronic music! ! I met Jeffrey Carpentier in May 2020 when I called him to suggest a crazy idea.

JEFFREY: Jeffrey P. Carpentier, 26, event organizer, bartender, volunteer since 2019 with CF Quebec and organizing member of Sound of CF. I met Gabrielle Phaneuf through my volunteer involvement in 2020.

Tell us about your journey.

GABRIELLE: I have long wanted to rebel against my chronic conditions. I was diagnosed with cystic fibrosis when I was 12 and with diabetes when I was 18. I spent many years living with symptoms that I did not understand. I spent many years living with chronic pain and a deep pain of living. After living in the dark for many years and hitting rock bottom, I began to see my life differently. By volunteering for cystic fibrosis from 2013 to 2019, I developed a sense of belonging and empowerment towards my disease and discovered a tight-knit community. Stronger from these life experiences, I chose to see my illness in a different light and to use it wisely as a driving force for action. I had no idea that, two years later, I would take over the management of a non-profit organization with a golden volunteer team (Jeffrey, Julie, Charley and Anaïs).

Since Jeffrey also has cystic fibrosis, we bonded very quickly. Being from the same generation, loving the same music and sharing the same diagnosis brought us closer from the first call.

JEFFREY: It all started five years ago when I joined the entrepreneur club at Cégep Montmorency. I was an event promoter at the La P'tite Grenouille bar in Montreal and I was itching for an idea: to organize a party with all my friends. I had the opportunity to organize the official party of "End of session Four Loko Montmorency". The love was immediate and passionate. After realizing this first event, I knew it was the way to go. I then changed my program to go into Event Management and Business Launch.

I graduated in 2019, and that same year, I oversaw the creation of an electronic music festival that I named "Cap sur l'île" because of the place where it took place. As part of this festival, it was important for me to include a donation for cystic fibrosis. I donated some of the money from ticket sales to the cause. The festival was a great success. It gave me the drive to get more involved in the cystic fibrosis community.

What is Sound of CF?

GABRIELLE: Sound of CF (FKM) is a unique concept in philanthropy that unites music and cystic fibrosis. On May 10, 2022, we registered FKM as an NPO with the Registraire des entreprises du Québec. FKM refers to music played for the CF cause in our fundraising and awareness campaigns. We are a none-profit organization whose goal is to raise awareness of cystic fibrosis while raising funds. We aim for an average of two events per year.

We organize virtual and face-to-face events that bring together passionate people, artists, DJs, musicians, and people who offer musical performances on a voluntary basis. Our events are festive, dynamic, educational, unifying, and young.

JEFFREY: Our service offer in events is rich and diversified. We organize several types of events, and all are for the cause: family day, underground DJ evening club version or public version, variety show, yoga event, music, and more. We explore new ideas with each edition. We like to do business with Montreal merchants and local partners who encourage and support us in exchange for a little visibility or simply to do the right thing.

What is your relationship to music?

JEFFREY: My mother instilled in me the benefits of music at a very young age as well as an openness to the diversity of music that surrounds us. I am a music lover and I believe that each style of music has its time to be listened to. Rarely can I move without music because I feel like something is missing. It is also the primary tool that feeds all my events. Music is unifying and universal.

GABRIELLE: My relationship to music is natural and visceral. It doesn't matter where I am in the world, it's part of me and my personality. My musical preferences have evolved over the years. When I was a kid, my father played music on Saturday mornings during breakfast. My father was himself a DJ in his time. He transmitted his passion to me during our karaoke evenings where the whole family sang. I knew all the lyrics by heart. My passion for music developed further in high school, during which I spent two years in the school symphony orchestra where I played the French horn and the clarinet. I took dance lessons for 15 years and was part of the dance troupe. Whether it's going well or whether it's going badly, music consoles me, excites me, makes me cry or makes me smile. It's one of the most beautiful things in the world.

How did you come up with the idea?

GABRIELLE: When the pandemic hit, we were about to start a big event for cystic fibrosis. I was very involved, but when the date of March 13, 2020, came, it was no longer possible to do so, and the company decided to cancel all its fundraising events. I did not accept the idea of giving up and looked for another way to do something. The idea came to me while thinking of my DJ friends and imagining myself dancing for the cause. I called Jeffrey to offer him a virtual fundraising concept for CF with live electronic music performances. We were going to call it: Sound of CF.

JEFFREY: When 2020 arrived, Gabrielle and I were well involved in a CF Walk committee. We had to be ambassadors for the cause and raise large sums of money for the organization. It immediately clicked between us when she called me. I found her idea original and fun. I got on board right away, because I already knew several DJ artists thanks to the festival I had organized. We set out to work together to create this new concept of virtual fundraising.

GABRIELLE: By mobilizing electronic music producers in Montreal, we put together over 25 DJ sets and managed to raise over $2,000 in just four days. We continued while refining our concept. We are now at our third annual fundraiser to benefit cystic fibrosis and wellness and our goal is $7,000.

What are your fondest memories?

GABRIELLE: My fondest memory is the December 2020 edition, when we filmed music performances at Salon Daomé, a very well-known and popular club in Montreal. In less than six weeks, we managed to put together a full four-day program of musical entertainment, with big names from the local scene, such as the artist duo Hicky & Kalo, Horla, Jimmy Strip and Guillaume Michaud. My DJ friend Julie/Anne helped me with the visual production of the performances. We managed to find cameras thanks to a contact at Solotech and to make a montage, all on a voluntary basis. This edition will forever be engraved in my heart and in my head because it was the real kick-off of FKM. Following this edition, we won the Prix Jeunesse par excellence in philanthropy – category 18 to 35 years old to highlight our efforts.

What are your next projects ?

JEFFREY: We have a long-term vision. We want FKM to become an annual fundraising tradition for the CF community and the music community in Montreal. We imagine that within five years we can be talking about a fundraising goal of $25,000.

GABRIELLE: We are developing the market in Quebec for now. Eventually, I would like to extend our concept to other Canadian provinces in French and English.

In five years, we will have developed a turnkey organization and musical entertainment components for events related to cystic fibrosis or awareness. People will hire the company and its brand to host their events.

In 10 years, our company will have created an annual philanthropic tradition, where people give to the cause every year. The business will have grown. Our evenings will have gained notoriety and credibility with major donors and actors affected by CF in Quebec.

We will have a committee in several municipalities and cities to be present everywhere in Quebec. We will have a branch that will take care of the rest of Canada as a whole.

We will be the leading and preferred organization in the field of musical events for cystic fibrosis. We stand out in several innovative ways to get our message across. We will do what no one else has done before. The feats will be achieved over the next few years.

Would you like to add something?

JEFFREY: Absolutely, but a face-to-face interview would probably be better to elaborate further.

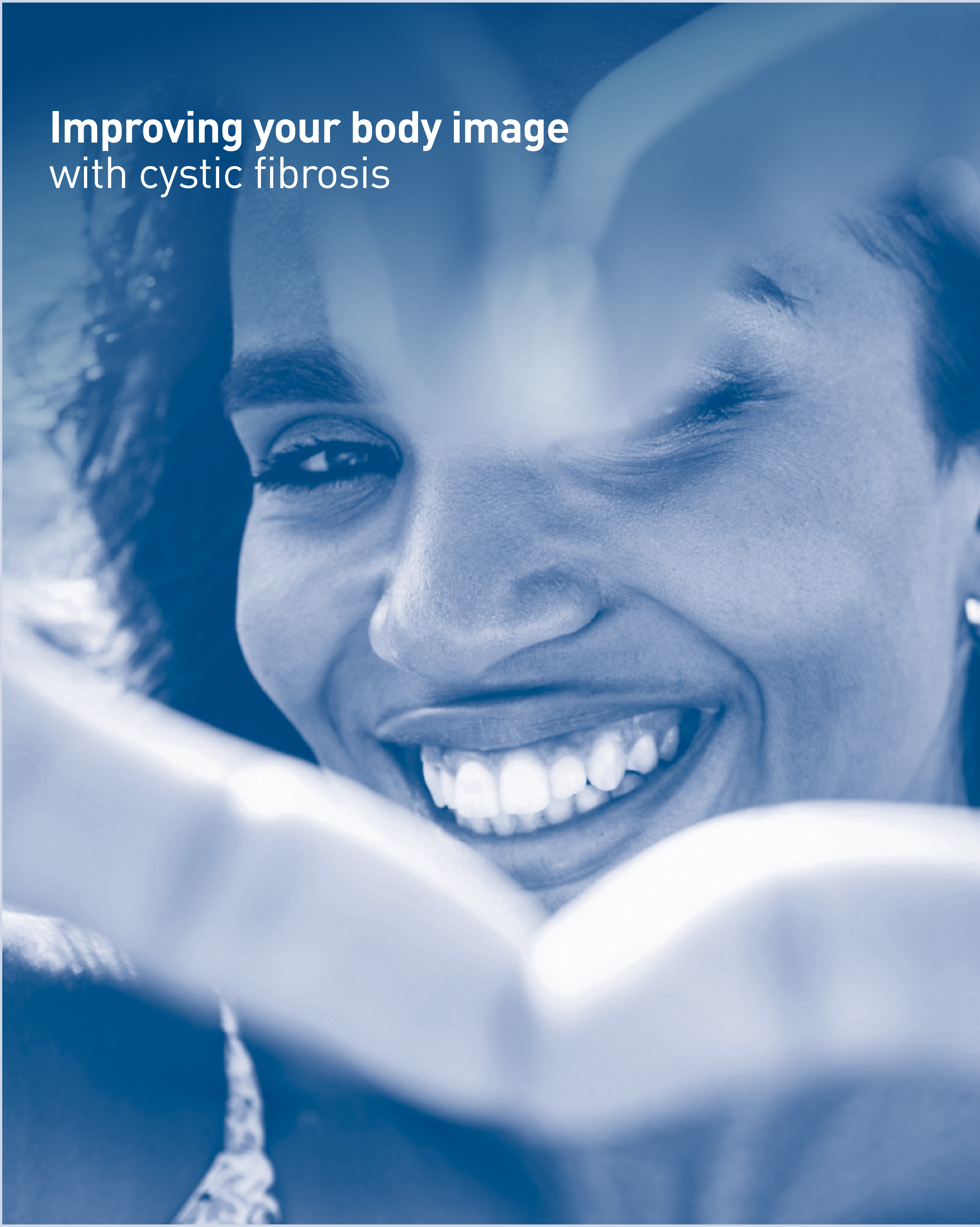
GABRIELLE: I never thought I would get this far with just one idea! This is a sign that when you believe in something, go for it. If I had listened to those who did not believe in it, I would never have succeeded. Believe in yourself ! ◀

FKM

FIBROSE KYSTIQUE EN MUSIQUE


SOUND OF CF FK EN MUSIQUE
yoga
MARC & Sebastien Kostello
Lundi 31 mai 18h30-19h45
Yoga Yin et Méditation
Musique envoutante de Sebastien Kostello
Plateforme ZOOM

Improving your body image with cystic fibrosis



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Do you love your body?
Are you satisfied with it?
Do you feel dissatisfaction?

The answers to these questions tell you about your body image, the perception that brings together the thoughts, emotions and physical sensations that are triggered when you think about or look at your body. Several body characteristics are related to body satisfaction or dissatisfaction. These include hair, face, body shape, height, weight, teeth, physical disabilities, chest, muscles, and scars.

How you feel about your body can change over time and vary in different situations and contexts. Even if you're not always 100% satisfied with your body, your body image can be quite positive. However, it can also be negative if the way you feel about your body causes you pain, affects your mood, health, work, activities, or relationships.

Cystic fibrosis can affect how your body develops, looks, and functions and presents several challenges to your body image, especially regarding your body shape, the appearance of your skin, your weight, your posture, and your muscle mass. CF, its development, and its different treatments are important factors that can negatively influence your body image. With CF, it can become difficult to see your body as an ally. Does CF influence your body image? Do you think you have a positive or negative body image? How to accept the changes imposed by CF and certain treatments? Do you sometimes feel like your body is betraying you, or that it is not what you would like it to be?

It is quite possible that CF raises questions about your body image. Indeed, many bodily dissatisfactions are reported by people living with CF. The latter refer to bodily characteristics that do not correspond to the unique model of beauty conveyed by society: excessive weight, insufficiently developed musculature, a smaller size, or the presence of scars.

Did you know that your body image develops as you age and that it is the interaction of different developmental factors that determines your assessment of your body? Your physical characteristics (weight, puberty, gender, age), individual psychological factors (personality traits, self-esteem), interpersonal experiences and socio-cultural factors are the factors that determine the body image you have developed.

It is also important to realize that the bodily dissatisfaction you may feel is triggered by a gap between your perception of your body and your perception of “your ideal body.” The latter is mainly decided by social norms. The greater the gap, the greater the bodily dissatisfaction felt.

Improving your body image takes time. No magic wand can change the relationship you develop with your body in one movement. However, several activities and exercises (behavioural, emotional, or cognitive) can improve it. The important thing is to take one small step at a time. Slowly, but surely.

Developing a positive body image makes it possible to feel good about one's body, and this, with certain dissatisfactions, and especially, with an illness. Positive body image allows you to feel pleasant emotions about your body, to appreciate its unique beauty, and to use your body wisely. This positive body image is closely linked to good mental health. In fact, people who develop it feel less distress about their bodies, adopt more balanced eating habits and express greater well-being. They also report better physical health.

To have a healthier and more diversified body image, certain skills must be developed. Reflections can be prompted, and certain behaviours are important to put in place, or even to stop.

Far from being a rigid protocol or a magic recipe, here are some suggestions to improve your relationship with your body.

TIP #1: Observe yourself

- Just close your eyes.
- Imagine yourself in front of your mirror.
- Let emotions and thoughts be activated.
- Be aware of the discomfort.
- What behaviours do you want to implement now?

Think about a typical day. What are your body image behaviours? What place do your physical concerns take in your life? How important are your physical dissatisfactions now?

This exercise allows you to connect to your body image. The fact that the exercise is pleasant or unpleasant gives you a clue whether you have a good or bad relationship with your body. This observation will then allow you to make some changes.

TIP #2: Understand the development of your body dissatisfaction

Give yourself the opportunity to understand where your body dissatisfaction comes from. What factors contributed the most? Several are decisive in the development of body dissatisfaction. Allowing yourself to trace the developmental trajectory of your body image is a good step towards a full understanding that leads to improvement. Think about your physical and psychological characteristics, your past interpersonal experiences, and the impact society may have had on you. Maybe you can identify an important trigger, or maybe not. Remember, it is the interaction of several factors, and not just one, that explains your body dissatisfactions.

TIP #3: Connect to your bodily sensations

Every day, take a few minutes: sit down and breathe slowly. Connect to your bodily sensations. When you are hungry, tired, in pain, excited, take a few seconds to identify what sensations are present. Do you know what messages your body sends you with these sensations? Are you able to meet its real needs? Try to identify your sensations without judging them. Reconnect to your body by simply listening.

TIP #4: Take care of your body

Taking care of your body means being able to give it what it asks for. Food, liquid, rest, warmth, sweetness. It can be difficult and even complex to meet the needs of your body, especially if you have lost the habit of listening to them. Go slowly. And allow yourself, sometimes, to be wrong. Adjust your actions. Take the time to measure what you offer to your body and see if it reacts well. Listen to him.

Your body may be sick, but it is still very much alive. You have every right to be mad at CF. You may need to mourn certain bodily features, mourn a disease-free body, mourn another body. Allow yourself this moment, whenever it is needed. At the same time, allow yourself to ally yourself with your body. Even with CF, it talks to you and expects you to listen.

TIP #5:
Identify your maintenance behaviours

If you're worried about your body image, you probably have behaviours that keep you in that worried state. Identify these behaviours more clearly. Think about your eating or physical activity behaviours, or what you might be doing to watch your weight. Think about behaviours you avoid doing, such as clothes you avoid wearing (although you would like to) or places you avoid going (although you would like to).

These behaviours are called maintenance behaviours. By their action, they maintain your bodily concerns, as a constant reminder to think about your body, your weight, your shape. Correcting or stopping these behaviours also alleviates your concerns and gives you more mental space for something else, for other interests.

TIP #6:
Tame your body

Improving the relationship you have with your body takes time. It's a relationship. It therefore requires you to tame your body, or to re-tame it slowly. Give yourself this time and identify the basic need you are trying to meet through your bodily control. Remember that to improve the relationship with your body, you will most likely have to tolerate discomfort and connect to a need, such as the need to be loved, to be accompanied, to be validated, to feel important. Remember that these needs are fundamental and that it is up to you to express them and ensure that they are met.

Improving your body image is possible. And when you take that path, no one can take away your new image. This is precisely one of the objectives of this work: to regain power over your own body without regard to external control, based on idealized images, social norms, rules decided by others.

Your body belongs to you. It is up to you to form a team and learn to live in harmony with it.

To learn more about body image and the relationship you have with your body, here is a very relevant reading suggestion:

De l'insatisfaction à l'acceptation corporelle, développer une relation plus positive avec son corps, from Éditions JFD, by Marie-Michèle Ricard.

Marie-Michèle Ricard, psychoeducator and psychotherapist, has more than twenty years of experience in the field. She specializes in the treatment of body and food-related problems, is the co-founder of the Imavi Clinic (www.imavi.ca), is behind the www.acceptersoncorps.com platform and is the author of several books ◀

Social networks @mmricardpsed

TRIKAFTA and the **identity crisis**

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Having been part of the CF community for nearly three decades, I think I can safely say that these are truly exciting times. Before the era of modulating therapies, we welcomed news about the most effective droplet size of inhaled antibiotics presented at international CF conferences. Many advances have been made and, thanks to improved treatments, the survival rate has increased. Treatment with an effective and evidence-based multidisciplinary approach contributed to this. It's amazing what has been achieved through the perseverance of so many researchers, clinicians, and patient organizations around the world.

We then entered the era of modulating therapies, and the news was, and still is, overwhelming, exciting, and encouraging. Access to these therapies is improving, but not fast enough. Right now, it really depends on where you were born. A tragic fact of life, in my opinion. Unfortunately, we tend to forget this in the media hype we know, but we must face the facts: in many countries, the Trikafta (called Kaftrio in Europe) will probably never be available to people with CF. Imagine the frustration of parents, patients, and doctors when they surf the Internet and read what is happening in the western part of the world.

Nevertheless, we have reached the era of the Trikafta. In many homes, champagne bottles were uncorked when the family member with CF was finally able to swallow the first dose of this miraculous drug. Photos of patients receiving their first dose became viral. My organization received beautifully decorated cakes for successfully lobbying for the reimbursement of this drug in the Netherlands. Doctors, parents, and people living with the disease, we all have high expectations for this new drug. The results of study after study are impressive. The personal stories on social media are amazing.

As I pondered the immense change the drug can have on a life, a family, a future, I first found myself a little surprised when a number of patients reported specific side effects to me. I'm not talking about the kind of side effects that are a result of the drug itself, such as the dose, interaction with other drugs, or effects on mental well-being due to certain compounds in the drug. This type of effect is the subject of in-depth studies. I want to focus here on the mental thoughts that people with CF and those close to them can have when Trikafta is at hand, specifically the thoughts and feelings that are likely to exist but are not always planned or consciously experienced. Yet they have a huge impact.

When preparing a presentation for the European CF conference last June, I asked a group of Dutch patients and parents of children with CF what they thought when Trikafta was at hand. As I said, I was a little surprised by some of the answers, but they make perfect sense when you look at them. In fact, some of their answers opened my eyes.

“I would like to know more about the effects of Trikafta on your life,” said a young woman with CF. Then she rushes to convince us how lucky she is to have access to this drug, "It’s such a huge gift that I feel I should not complain or ask for understanding." She has doubts and questions, but it seems that she does not want to bother us with this, because the gift is such a beautiful thing. Another patient replied: “The people around me are so ecstatic about this; I’m afraid I can’t meet their expectations. What if my body let me down?"

I totally understand that. I understand that very well. People with CF have grown up with the idea of not living very old. They had to get used to their bodies failing them — or they had to adapt to it, physically and mentally. Parents have brought up their children with the fear of losing them, a thought always present in their minds. These parents might now want to change their mindset, which seems easy to do because Trikafta is so promising, and the results are truly amazing. But see how this parent describes this change: “We lived for twenty years with the idea that she would not grow old. Now that it could happen, we began to realize how worried we had been. For years and years. Only now do we feel the weight that has been on our shoulders for so many years. Suddenly, we realized it. It was so sad and upsetting. This caused depression for one of us and some slight relationship issues as we seemed to be coping with it differently and had to figure it out ourselves.” People with CF tend to dwell on old worries and feel sorry for the painful episodes they had to endure in the past — at least that's what some people in the Dutch panel said when asked the question.

Let's take a closer look at this question. Talking to people with CF, they made it clear to me that they also had to get used to their “new” bodies. This may be the same feeling people have after a lung transplant. They were used to a body with limits, pain, and very uncomfortable moments. Frustration too. Now that same body has to be explored again and expectations are high: a whole new future seems to be upon them. The same is true for mental processes when people with CF have access to Trikafta. It's not a side effect of the drug itself, but the brain has to adapt to a new and unpredictable situation. As glorious as the effects of the drug are on the body, the mental processes will also be affected. New questions arise. Some panelists tell us that, for the first time in their lives, they feel like they have a future. Yet they are not comfortable, they are not used to considering it. "Are you starting to make plans? we asked. "Or are you changing the plans you had? Are you rethinking your future? ". "No, it's too early for that," some people with CF replied, "but it seems those around me expect me to make plans and create my own future with enthusiasm and eagerness, and their expectations are high and I'm afraid of failing. »

“My life has a new chance. And what do I do now?” a woman with CF posted on Facebook. "Now that there is a possibility that I will get old […] what kind of work should I like? I mean, think of something that I would like to do for many years. I just can't imagine it.”

The (possible) use of Trikafta makes people realize that too many people with CF have not survived. There are dear friends for whom the medicine came too late. “As I think of these people, I feel even more thankful, and I also feel sorrow again.… I wish they had the chance to enjoy the benefits of Trikafta.” There are also people with CF who don't have the "right" mutations and can't yet benefit from the drug. I quote again an excerpt from a message on Facebook: “I know they are happy for us, but it's sad that there is nothing for them.”

Most of the people in our panel responded that they had started to think differently about their future, and in a more positive way. They begin to dream of working, of having children, of participating in society, and feel more confident because their illness now seems to be more of a chronic illness. At the same time, some people are concerned about the possible long-term side effects of the drug itself; some also experience feelings of anxiety when they realize that they are dependent on the drug. What happens if the drug cannot be prescribed or dispensed for some reason?

Another issue that has been raised is the meaning of the term “severity of illness” when the diagnosis is to be discussed with parents of newborns. Life expectancy is much better today, and more newly diagnosed children will be able to live more or less healthy lives. The message from health professionals about CF has a huge impact on parents' perception of CF; it affects their mental well-being and probably the way they raise their child. What is the best approach to describe the impact of CF on the lives of parents of newborns with CF? I invite health professionals to think about the possible effects of their message, when describing the impact of the disease and explaining the difference it can make if someone qualifies for the drug and has access to it.

I would like to thank the interviewees for giving us a better understanding of the effects that the use of this drug may have on their mental well-being. As involved spectators, we should know that possible side effects are not always obvious. And that's completely understandable.

“But when all those thoughts go away, I feel grateful. So deeply, I can't explain it. I think it's because there have been many occasions in my life where we thought 'this is it' and then that slippery ladder down. Or even the times when we had to face death. And I'm still here. Life was and is not over with me, of course.*” ◀

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* Courtesy of Yvonne Verschoor-Broeksma

CFTR modulators and mental health

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IMPACT OF CFTR MODULATORS ON QUALITY OF LIFE AND MENTAL HEALTH

Cystic fibrosis transmembrane conductance regulator (CFTR) modulators, in particular the triple therapy Trikafta® (elexacaftor/ tezacaftor/ ivacaftor), have been transformative for the many people living with CF who have experienced improvement in physical symptoms and quality of life (Regard, Martin, Burnet, Da Silva, Burgel, 2022) while taking them. For others who are not eligible because of their CFTR variants or who lack access (Guo, Garratt, Hill, 2022), however, the approval of Trikafta® may be accompanied by mixed emotions including disappointment, frustration, and worry about the future (Keyte R, Kauser S, Mantzios M, Egan H, 2022). Those who are eligible but have side effects that make Trikafta® unsafe to use or difficult to tolerate may also experience distress.

In addition to physical benefits and side effects, questions have emerged regarding the mental health impact of taking CFTR modulators. The evidence on this topic remains limited to describing the experiences of individuals or small groups of people with CF who have started taking a modulator. Such reports have described improved or worsening mood or anxiety; increased or decreased energy levels; changes in thinking, memory or concentration ("brain fog"); and trouble sleeping (Bathgate CJ, Hjelm M, Filigno SS, Smith BA, Georgiopoulos AM, 2022).

HOW COULD MENTAL HEALTH SYMPTOMS BE RELATED TO MODULATORS?

Larger studies will be needed to more clearly demonstrate and understand the relationships between CFTR modulators and mental health. However, there are several ways to think about whether and how CFTR modulators could be linked to mental health symptoms (Talwalkar, Koff, Lee, Britto, Mulenos, Georgiopoulos, 2017).

Hypothesis 1: Modulator is unrelated to changes in mental health

The first possibility is that an individual's mental health symptoms are unrelated to starting a CFTR modulator. People with CF are at higher risk than people in the general population for several mental health conditions, including depression, anxiety, attention problems, and sleep disturbance (Bathgate CJ, Hjelm M, Filigno SS, Smith BA, Georgiopoulos AM, 2022). These challenges could be brought on or made worse by many factors, including the stress of living with CF, pain, inflammation, or periods of poor nutrition or low oxygen. The timing of approval of Trikafta® also coincided with the COVID-19 pandemic, which could complicate distinguishing the triggers for mental health changes during this time period. The pandemic created uncertainty, fears and losses affecting emotional wellness for many people, and also had profound effects on the delivery of CF care and mental health care (Smith BA, Georgiopoulos AM, Mueller A, Abbott J, Lomas P, Aliaj E, Quittner AL, 2021). Mental health symptoms are common and often get better or worse over time or in response to starting or stopping mental health treatment. For these reasons, it would be expected for some people to notice changes in mental health around the time they started taking Trikafta® due to coincidence alone.

Hypothesis 2: Psychological impact of starting a modulator

Even when changes are welcome and positive, they can be stressful, resulting in anxiety, sadness, or a sense of distraction and preoccupation. This can be true for people with CF who start modulator therapy. Improvements in health can result in reevaluating one's past and future, worries that the benefits of modulators might not last, and thinking about how your identity and life might change if your health is more stable. Some may experience an overwhelming sense of pressure to "do more" at work, school, or home, or have concerns about losing social supports they have come to rely on or benefits such as disability payments. Your sense of self may change; for example, some who gain even a healthy amount of weight after starting Trikafta® may struggle with body image. Despite gratitude for the opportunities better health may present, knowing that others with CF have not had the chance to benefit from modulators may also lead to survivor's guilt.

Hypothesis 3: Direct effects of modulators on the brain

The CFTR protein is found not only in the lungs, but other in other parts of the body including nerve cells within and outside the brain. Although the role that CFTR serves in the brain is uncertain, it is possible that changing the availability of CFTR by having cystic fibrosis and/or by taking a CFTR modulator might change how the brain functions. CFTR modulators could also affect how nerve cells communicate with each other. For example, ivacaftor, an ingredient in all currently available CFTR modulators, has been shown to connect to receptors and transporters of neurotransmitters such as serotonin, norepinephrine, and dopamine (Schneider EK, McQuade RM, Carbone VC, Reyes-Ortega F, Wilson JW, Button B, Saito A, Poole DP, Hoyer D, Li J, Velkov T, 2018). Many medications used to treat depression, anxiety and Attention Deficit/Hyperactivity Disorder (ADHD) also have effects on these neurotransmitters. It is therefore plausible that CFTR modulators could have a direct impact on mental health symptoms.

Hypothesis 4: Drug-drug interactions between modulators and psychiatric medications

Finally, drug interactions can change the effectiveness of modulators or psychiatric medications or increase the risk of side effects (Bathgate CJ, Hjelm M, Filigno SS, Smith BA, Georgiopoulos AM, 2022). For example, lumacaftor, found in Orkambi®, is known to decrease the levels of the antidepressant medications citalopram, escitalopram, and sertraline. When a person with CF taking one of these antidepressants starts Orkambi®, they could notice a relapse of depression or anxiety symptoms and need to increase the antidepressant dose. On the other hand, both the anticonvulsant/mood stabilizer carbamazepine and St. John's wort, an over-the-counter health supplement sometimes used for depression, could reduce the levels of Trikafta® and make it less effective. Medications are processed through the body using many pathways, so when side effects occur after starting a new medication, the potential role of drug-drug interactions should be considered.

TALK TO YOUR HEALTHCARE PROVIDERS ABOUT MENTAL HEALTH

If you are taking a CFTR modulator and are concerned about your mental health, ask for an evaluation by a mental health professional who is able to collaborate with your CF care team. At many CF centers, a CF social worker or Mental Health Coordinator can help get this process started. Some CF centers work closely with psychologists or psychiatrists who specialize in treating people who have both physical and mental health conditions. (Figure 1)

What is mental health provider, and what services do they provide?

Figure 1: Mental health professionals in Quebec.

- Therapists are professionals with graduate or doctoral level education focused on human behavior who provide care through individual, family, or group therapy.
- Psychiatrists are licensed medical doctors who prescribe and manage medication.
- Psychotherapists are professionals with a master's or doctoral degree (social workers, marriage and family therapists, nurses, occupational therapists, psychoeducators, guidance counsellors, sexologists and criminologists) and hold a permit issued by the Ordre des psychologues du Québec. They provide care through individual, family or group therapy.

Figure 2: Mental health professionals elsewhere in Canada

Professional Credentials	Provides Therapy	Prescribes Medication
Licensed Clinical Social Worker	Yes	No
Licensed Marriage and Family Therapist	Yes	No
Licensed Professional Counselor	Yes	No
Clinical Psychologist	Yes	Rarely
Psychiatrist	Less Common	Yes
Nurse Practitioner Physician Assistant	Less Common	Yea

Reproduced with permission from: Finding a Mental Health Professional: A Guide for the CF Community. Cystic Fibrosis Foundation, 2022.

It's important to be open about what you've been experiencing. Tell your treatment team when your symptoms started, how frequent and severe they are, how they are affecting your day-to-day life, and what you think may have brought them on. What is the timing of your mental health symptoms in relation to taking Trikafta® or to other life stressors? Have there been recent changes in your other CF medications, psychiatric medications, or any over-the-counter or non-prescribed substances you may be using? Your team will also want to know what you have tried so far to address these problems and whether symptoms are getting better, worse or staying the same. If you are concerned that your symptoms may be related to taking Trikafta®, explain what led you to wonder about that. Have you had any physical side effects, such as liver problems?

Your healthcare providers will ask if you have had mental health symptoms or treatment in the past, and if so, what helped. Think about whether what you are experiencing now is similar to or different than any mental health symptoms you may have had before. Knowing about any family members who have had mental health conditions—for example, if a person with symptoms of depression or anxiety has a close biological relative with bipolar disorder—can also provide useful clues about diagnosis and what kind of treatment might be most effective.

Making a decision about next steps

A variety of treatment approaches are available to address mental health conditions that begin or worsen while someone is taking a CFTR modulator. If symptoms are mild or improving with time, no intervention might be necessary. Support from friends, family, or healthcare professionals can help with adjusting to living with CF in the age of CFTR modulators. Seeking out resources with information about CF and mental health such as the CF Foundation website (https://www.cff.org/managing-cf/mental-health) or Facing CF (Sher Y, Georgiopoulos AM, Stern TA, 2020) may also be useful. Behavioral strategies and psychotherapy have a strong evidence base to effectively manage depression, anxiety, insomnia, and other mental health conditions. Similarly, whether or not symptoms are felt to be related to taking a CFTR modulator, psychiatric medications can be started or adjusted to target bothersome symptoms.

In some cases, stopping the CFTR modulator or changing the dose may be tried to help assess whether mental health symptoms are related to the modulator and if this strategy will relieve them. However, stopping the modulator risks losing its physical health benefits, and there is limited data available to guide safe and effective use of a dose lower than recommended by the instructions on the product label (Spoletini G, Gillgrass L, Pollard K, Shaw N, Williams E, Etherington C, Clifton IJ, Peckham DG, 2022). This strategy should therefore should be approached with caution.

If you think you may be having mental health side effects and are considering taking Trikafta® differently than it is prescribed for you or stopping it, it's important to talk to your CF care team first. Ask them to think together with you about the risks and benefits of making this change, which may depend on your individual circumstances.

- How risky would it be to try stopping Trikafta®? Consider any ways that your health or quality of life has improved since you started taking it. The consequences of a short trial period without a modulator might be quite different for a person with CF who, before starting Trikafta®, had normal lung function and rare pulmonary exacerbations as compared to someone with advanced disease who had frequent hospitalizations and was waiting for lung transplantation.

- How severe and bothersome are the mental health symptoms? For example, occasional trouble concentrating might be a minor nuisance, but if attentional problems are persistent and severe, they could lead to trouble doing well in school or keeping a job.

- How risky would it be if the mental health symptoms stayed the same or got worse? Are there any concerns about safety? Regardless of the cause, some symptoms, such as feeling suicidal, agitated, or out of touch with reality, require immediate attention.

- What are the alternatives to stopping Trikafta®? Are there coping skills you could use to manage your mental health symptoms, or other psychological or medication treatment strategies that could make sense to try?

Because people's preferences and tolerance for different kinds of risk may vary, two people in a similar set of circumstances might make different choices. It's important to discuss your goals and values with your team as you make decisions about your next steps.

Keeping track of changes

Regardless of which intervention you decide to try next—or even if you decide it seems best not to make any change in your CF or mental health treatment plan at the moment—discuss how you and your healthcare team will monitor your physical and emotional health. Make a list of the changes that would be most important to look for as early signs that your health could be starting to worsen (such as increased cough or fatigue) or improve (such as sleeping better or feeling less anxious). It can be easy to forget details, so if you can, keep a daily log to keep track of how things go over time. Include the physical and emotional symptoms most relevant to you—for example depression, irritability, anxiety, sleep or pain. Also make a note of any medication changes and key life events or stressful situations that could affect how you are feeling, such as a new relationship, financial problems, or final exams. It may be wise to meet more often than usual to check in with your treatment team.

LEARNING MORE ABOUT CFTR MODULATORS AND MENTAL HEALTH

Healthcare providers and people with CF are still learning together about the impacts of CFTR modulators. Research is underway and new studies are being designed to help the CF community better understand the complex effects of modulators—both positive and negative. Partnering with your treatment team is essential to help optimize your physical and mental health. ◀

NEED HELP?

SIf you or someone you know is in crisis, support is available 24 hours a day, 7 days a week.

Besides contacting your treatment team, calling 911 or visiting an emergency room, you can call

Talk Suicide Canada (talksuicide.ca) at 1-833-456-4566.

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For residents of Quebec, call 1-866-277-3553 or visit suicide.ca/en.

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Across the US, you can reach the Suicide & Crisis Lifeline by calling or texting 988.

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Profile of the CF population in Quebec

Up to this day, no recent study provides information on the sociodemographic profile of the population with cystic fibrosis in Quebec. The last such study dates to 2006, more than 15 years ago.

Many things have changed since then, societal, and economic issues, knowledge of the disease and its management. Therefore, **Living with Cystic Fibrosis**, which has been working for and by people with CF since 1985, decided to investigate the matter.

To identify and understand the issues experienced by people with CF and their loved ones, to defend their rights and interests and to better direct our services, we decided to conduct a vast survey of adults and parents of children living with cystic fibrosis in Quebec.

GOALS

1. Get to know the profile of the CF population in Quebec.
2. Adapt our service offer according to the realities and needs of people living with cystic fibrosis in Quebec.
3. Collect data that will allow us to carry out further studies later.

METHODOLOGY

Step 1: Creation of the survey
Several meetings were held with various representatives of the CF community to design a survey that covers the essential data necessary for our objectives.

Step 2: Administration of the survey
The survey was published online. We reached our participants via our social networks, emails and posters posted in the various CF clinics in Quebec.

Step 3: Compilation and exploitation of data
After collecting the data, they were recovered in their raw format, then dissected and analyzed to produce a file that can be used by our organization.

RESULTS

The compilation of the data reveals several problems.

Economic fragility

Adults living with cystic fibrosis constitute a population that can be considered economically fragile. In fact, two-thirds of them have incomes below the average income in Quebec, and a quarter below the MPC (market basket measure). Moreover, only about half work full time. Difficulties in accessing employment for many of them are a major issue.

Coverage for drugs and devices

For the vast majority of households with a CF member (adult or child), drugs and devices prescribed by a doctor are not reimbursed, which represents a considerable cost each year. This situation adds to the economic problems already mentioned. That some simply cannot afford the necessary drugs and devices because of the cost is very worrying.

Daily impact of the disease

Cystic fibrosis has considerable daily consequences for adults, but also for parents. The number of days per year when daily activities must be limited for reasons related to illness represents half the year for parents and three-quarters of the year for adults. The consequences of this situation on mental health should not be overlooked.

Parents' concerns

When comparing levels of concern between adults with CF and parents of children with CF, the latter have a much higher level of concern. Although some sources of worry are the same in both categories, it should be noted that parents worry much more than adults about matters of daily life. It would be interesting to push the analyzes and reflections further, but also to consider a possible need for information and support at home.

Mental health

The psychological health of people living with cystic fibrosis is an increasingly perceptible issue. According to the data, just over 20% of them have been diagnosed with a mental disorder, mainly depression and anxiety. Added to this are those who have no diagnosis, but for whom these issues are also present.

Geographic disparity

The geographic distribution of people living with cystic fibrosis shows that nearly half of them live in three administrative regions: Montérégie, Montréal and Saguenay–Lac-Saint-Jean. If the first two regions are traditionally more populated, the third constitutes a specificity. It would be interesting to carry out a study on the management of the disease in the different regions and see if there are any disparities. It is also important not to forget those from other regions, because even if they are less numerous there, they are present in all regions of Quebec.

Retirement

We could note in the answers that CF retirees are more and more numerous, which was not the case in the past. Many reported a lack of support and information regarding their reality.

This data will allow our organization to adapt its services and to better understand the community and thus better represent people living with cystic fibrosis with institutions.

It’s also a good starting point for more in-depth studies on specific issues, as our raw data can be cross-referenced to bring out precise figures on a given situation

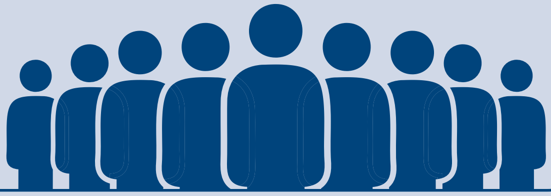
Profile of the CF population in Quebec

HIGHLIGHTS INFOGRAPHIC

Sources: Data collected by questionnaire from 340 respondents from March to August 2022. Level of confidence 95% and margin of error 5%.

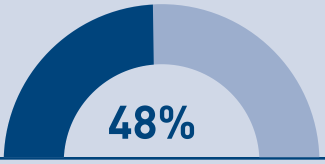
Income

A quarter (25%) live on annual income **less than $20,700** (Market Basket Measure), and **more than two-thirds (67%)** earn **less $55,000** gross per year (the average salary in Quebec)



Geographical distribution

48% of people living with cystic fibrosis live in areas **Montérégie, Saguenay-Lac-Saint-Jean and Montreal**



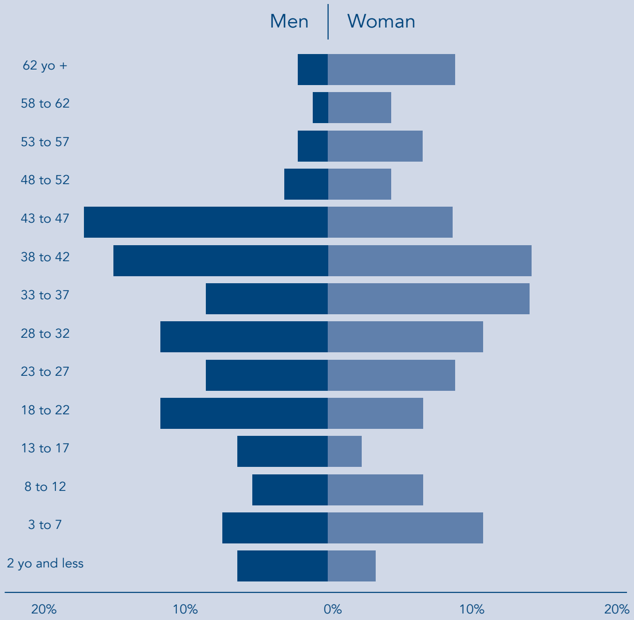
Concerns

These are the **4 major concerns** that are found in **adults with CF and parents of children**.

- Evolution of the illness
- Body's degeneration
- Efficiency of treatment
- Future

Age structure

Very close to Canadian data. We take note that women tend to age more.



Mental health

20% report having a mental health diagnosis in addition to cystic fibrosis.

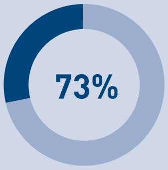
Depression and **anxiety** are the large majority

Annual expenses

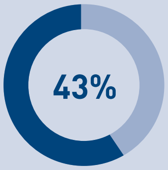
Per CF adult and child, an average of **$1,350** is spent **each year** to obtain the drugs and devices **prescribed but not reimbursed by insurance**.

Limitation of activities daily, in days per year

Each year on average, an adult with CF will limit its activities for 265 days (73% the year), and a parent of CF child for 158 days (43% per year), for illness-related reasons.

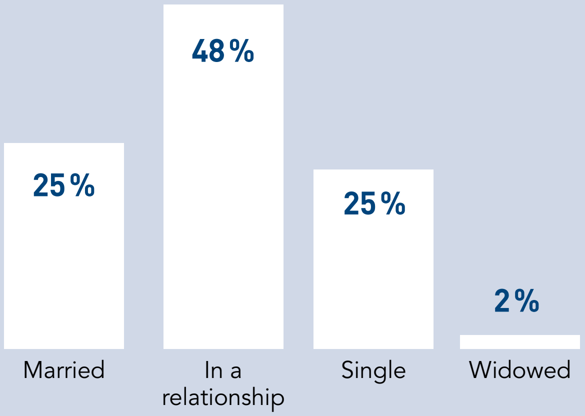


of days in a year

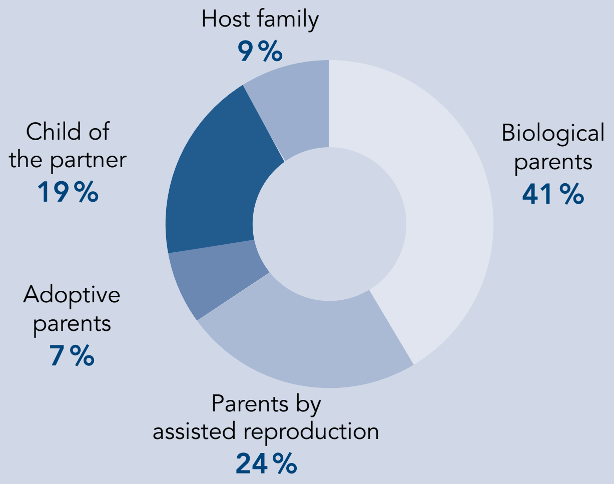


of days in a year

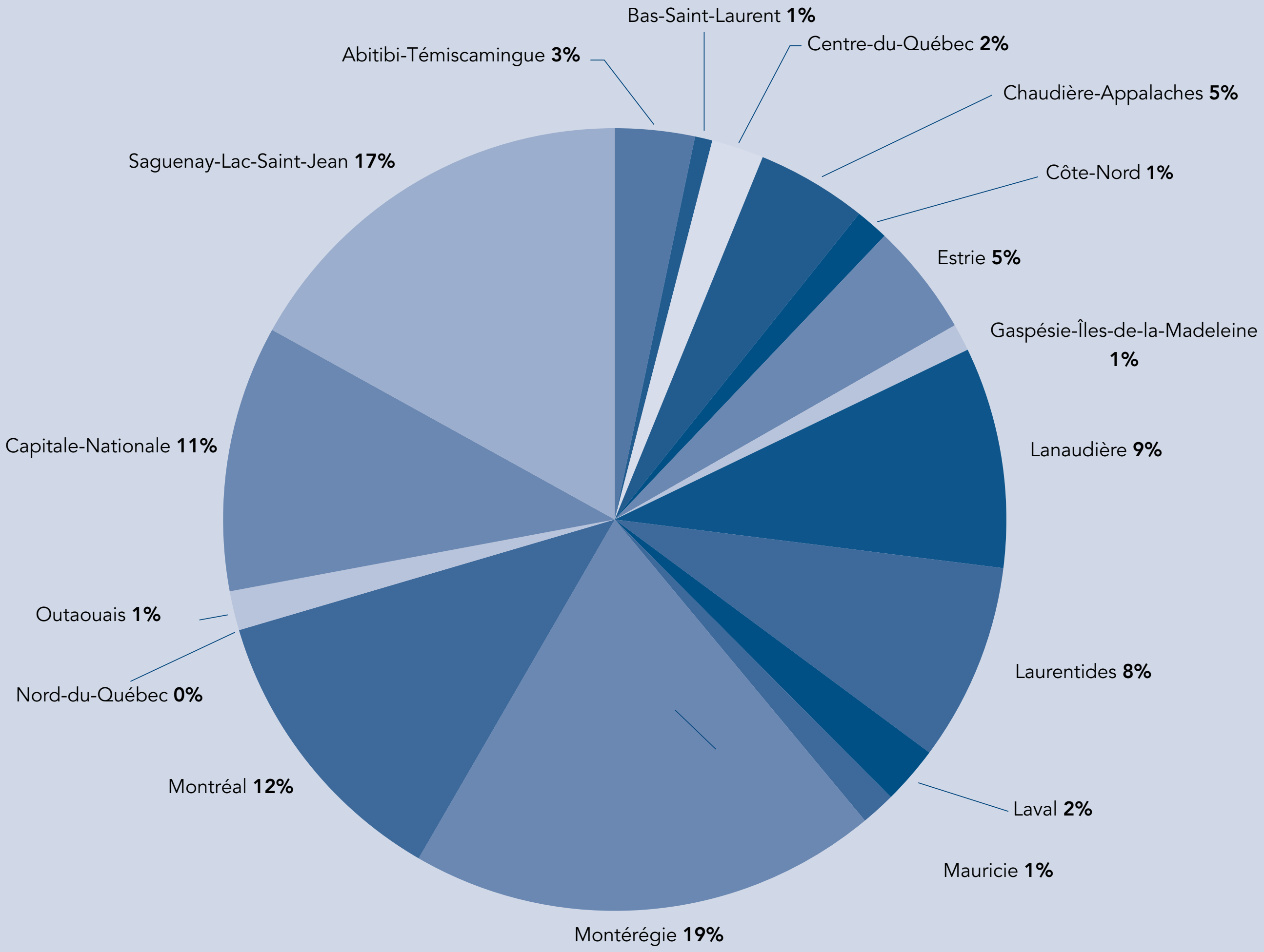
Personal situation of adults with CF



27 % of CF adults are parents

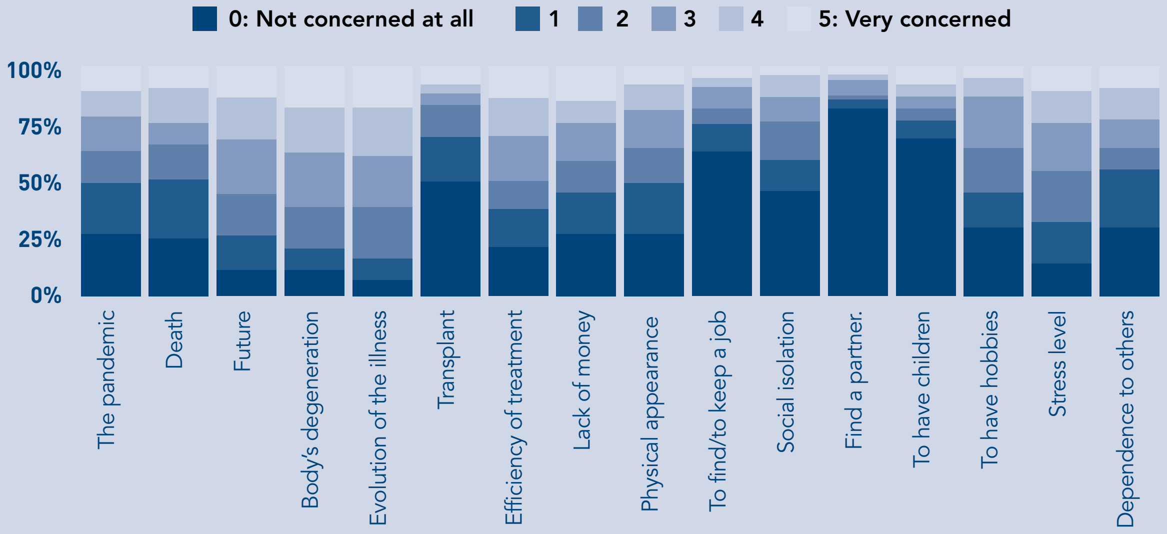


GEOGRAPHICAL DISTRIBUTION



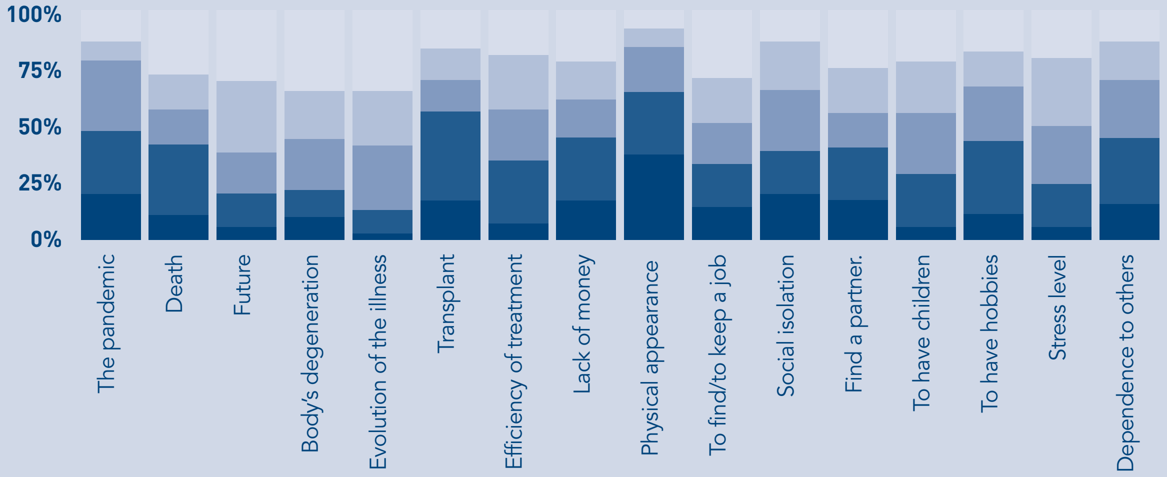
CF adults’ concerns

On a scale of 0 to 5



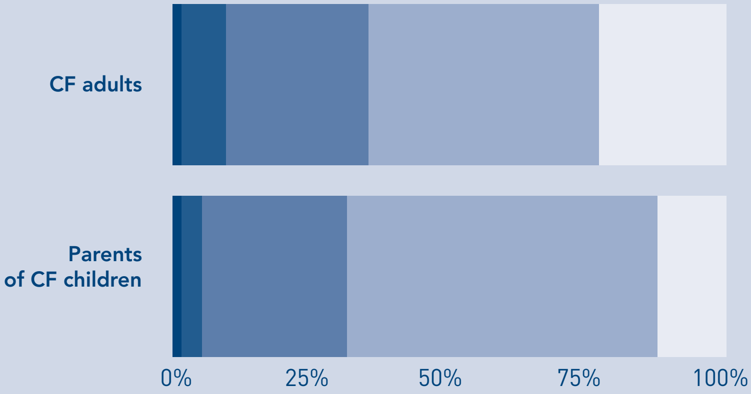
Parents of CF children concerns

On a scale of 0 to 5



LIFE’S SATISFACTION

Not satisfied at all
Not very satisfied
Moderately satisfied
Satisfied
Very satisfied



The complete data will be presented during our annual general assembly of members, on **Saturday April 15, 2023**

The Research
in Cystic Fibrosis: 2022 Update

Dr. Christelle Bergeron
and Dr. André Cantin
Pulmonologists

Pneumology department
Cystic Fibrosis Clinic (Adult)
Sherbrooke University
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Sherbrooke, Quebec
Canada

The major advances in cystic fibrosis research in the past year are undeniably related to new modulators of CFTR function. Over the past two years, the arrival of triple therapy (elexacaftor, tezacaftor and ivacaftor, a trio named ETI) has transformed the lives of most people with CF. We are experiencing a real revolution in the treatment of CF. These long-awaited changes encourage us enormously. However, solutions of similar effectiveness remain to be found for all people with CF, including the 10% whose defective CFTR does not respond to ETI. In addition, as it often happens, the new ETI treatment also brings new challenges which we will discuss.

About 90% of people with CF have at least one F508del-like mutation and therefore qualify for treatment with ETI. For these people, the beneficial effects are remarkable. The chloride concentration in sweat is a marker of the fundamental defect associated with defective CFTR. The results of the sweat test of people on ETI approach the values observed in people with a mild form of the disease. Lung function improves by around 14% in most people treated with ETI, and globally the number of people with CF waiting for a lung transplant has dropped significantly. The number of respiratory exacerbations, the main cause of hospitalization and deterioration of respiratory functions, fell by at least 50% following the new treatment. Bronchial images obtained by computed tomography of the chest after one year of ETI treatment demonstrate a dramatic disappearance of the excess mucus normally seen in the airways of people with CF. Similar observations are also reported for sinus imaging. The beneficial effects of ETI also trickle down to the digestive system with significant weight gain and improved serum vitamin profile. Diabetes also appears to improve in several individuals on ETI.

ETI medication is generally well tolerated. A few side effects, including occasional headaches, skin rash, and sometimes a feeling of respiratory congestion are observed, especially in the first weeks of treatment, and improve thereafter. Taking ETI also requires regular medical monitoring with dosage of serum markers of liver function and with dose modification according to the interactions of ETI with other drugs. Despite taking ETI, people with CF should continue with many other CF treatments unless advised otherwise by their medical team.

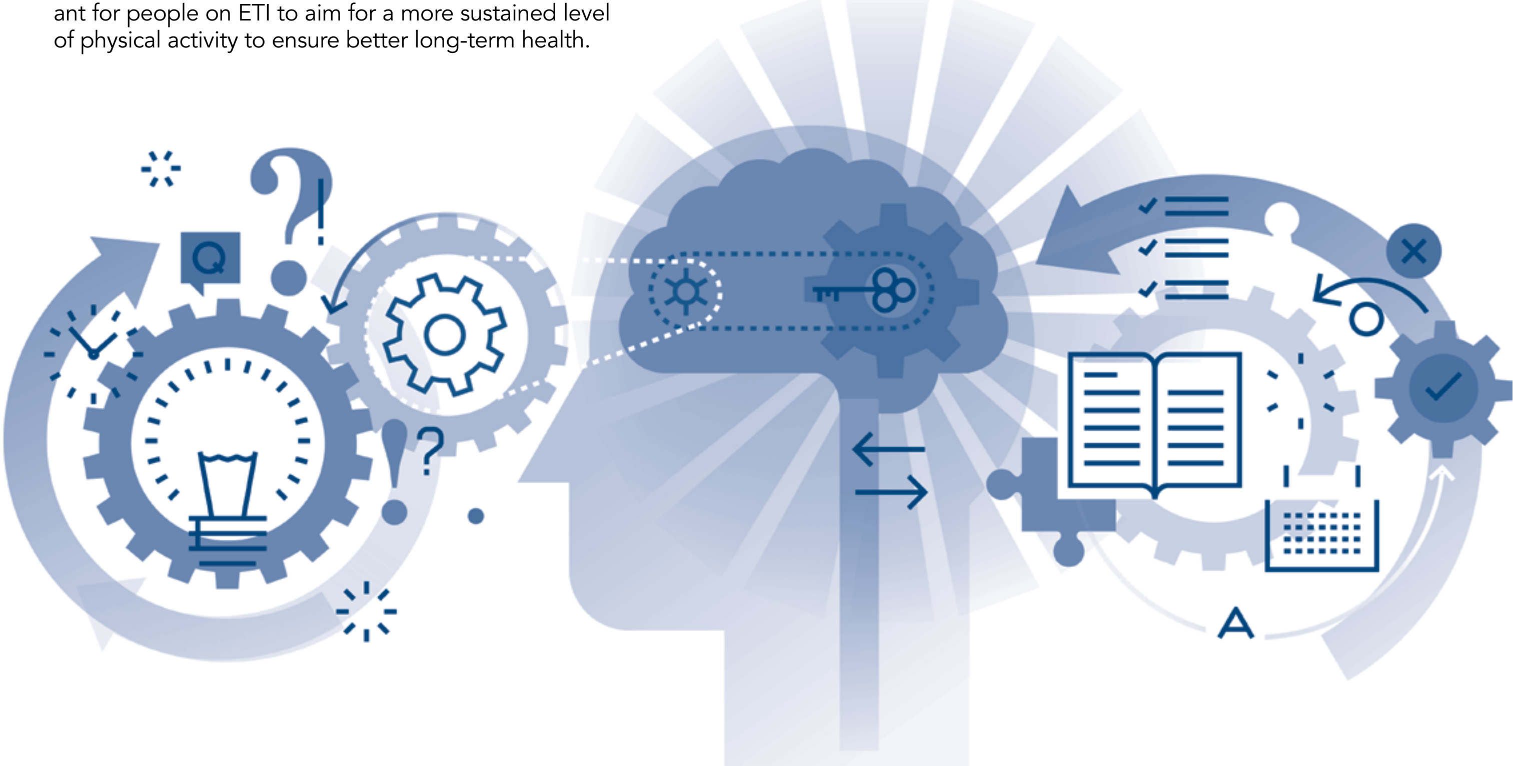
Several challenges persist in the treatment of CF, the most pressing of which is finding effective therapies for all people, regardless of their type of CFTR gene mutation. The majority of people with CF who do not meet the ETI treatment eligibility criteria have one or two class 1 mutations. These mutations cause the genetic code controlling the production of the CFTR protein to be misread. This results in an absence of CFTR protein, which explains the ineffectiveness of ETI for these people. Fortunately, there are molecules that can erase this misreading in cells examined in the laboratory. Following these scientific observations, the researchers collaborated with the pharmaceutical industry and clinicians to develop clinical studies that will determine the safety and clinical efficacy of ELX-02 combined with ivacaftor. Preliminary results from this work have not achieved the desired efficacy, and the researchers are reviewing the data to determine if the lack of efficacy is related to an insufficient amount of the drug reaching the targeted cells. Several groups of researchers continue to work on other approaches, including CRISPR-like molecular scissor gene editing and transfection of the healthy CFTR gene to bronchial cells. This work is a priority for many foundations working in CF.

The biggest challenge associated with the arrival of ETI is still access to this expensive medication. Fortunately, in Quebec, ETI treatment is now reimbursed for most people with CF who carry at least one F508del mutation. Recently, reimbursement has now been extended to people aged six and over, regardless of their respiratory function (FEV). It is expected that the earlier in life the defective CFTR is corrected, the less people with CF will have sequels from this disease.

The arrival of ETI treatment is also associated with emerging medical situations that we will have to learn to recognize and manage. In some people, particularly those with residual pancreatic function, the introduction of ETI therapy can relieve the obstruction of the pancreatic and bile ducts. A possible consequence of this effect is that the pancreatic tissue risks being exposed to an abundant quantity of pancreatic enzymes, which could generate pancreatitis. Pancreatitis causing severe abdominal pain has been reported for some people on ETI whose pancreatic function was preserved. We also observed that ETI treatment is often associated with an increase in the absorption capacity of vitamins A and E as well as a decrease in serum glucose for people with diabetes. It is therefore important to ensure medical and nutritional monitoring after starting to take this medication to adjust vitamin supplements and hypoglycemic treatments. Finally, taking ETI can lead to drug interactions, for which it is recommended to always check with your medical team before starting a new medication.

With the benefits of ETI in CF, it is expected that we will see a marked improvement in life expectancy, which is great. This change will also bring new medical considerations, including the control of risk factors for cardiovascular disease in CF. The improved absorption associated with taking ETI leads to weight gain, and we are already starting to see people with CF who are overweight and sometimes obese. In addition, we have always advised our patients to eat a diet high in fat, calories, and salt. These recommendations for people on ETI could lead to an increased risk of obesity, dyslipidemia, diabetes and high blood pressure, all factors involved in the onset of arteriosclerosis. Nutritional recommendations should be modulated according to the situation of each person. Finally, in the past, numerous bronchitis, shortness of breath, abdominal pain, difficulties in maintaining adequate muscle mass and the cumbersome treatment associated with CF are all factors that favored a sedentary lifestyle. It will be important for people on ETI to aim for a more sustained level of physical activity to ensure better long-term health.

The adjustments made by ETI treatments represent great challenges, because they most often come with a marked improvement in quality of life. However, much remains to be done to ensure that everyone benefits from the significant advances in CF research made in recent years. The best way to do this is to continue our teamwork with researchers, healthcare professionals and people living with CF. Together, we can defeat CF. ◂



HEALTH
MATTERS

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Respirologist
Assistant Professor

Director of the Adult Cystic
Fibrosis Clinic Co-director
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With the collaboration of
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Fibrosis Clinic of the
CIUSSS de l'Estrie — CHUS

What is thrush? How to recognize it? Which drugs are at risk? How do we treat it?

Oropharyngeal candidiasis, more commonly known as thrush, is an infection caused by Candida, a fungus present in the normal gastrointestinal and genitourinary flora. The infection is recognized by the presence of whitish plaques in the mouth, on the palate, on the tongue or in the oropharynx. These plaques do not disappear if they are brushed with a toothbrush or scraped with a tongue depressor. The diagnosis is usually clinical, but it can be confirmed by scraping the lesions and then making a KOH (potassium hydroxide) preparation to visualize the yeast. Other symptoms are sometimes present such as the sensation of having a pasty mouth, loss of taste and pain when swallowing.

The individuals most at risk are the elderly wearing dentures, people on antibiotic therapy or chemotherapy and those with an immune deficiency (e.g., AIDS). The medications most at risk are inhaled glucocorticoids (e.g., Flovent®, Alvesco®, Pulmicort®, etc.). Therefore, it is important to use the spacer (aerochamber) with metered-dose inhalers and to rinse your mouth after taking these inhalers.

Thrush is usually treated quite easily with an antifungal (Nystatin) to gargle and swallow 4 times a day for ten days. For more refractory cases, it is possible to prescribe an antifungal from the class of azoles in tablets, most often fluconazole. However, care must be taken since there are drug interactions to be considered with fluconazole, in particular with CFTR modulators (Kalydeco®, Orkambi®, Symdeko®, Trikafta®).

THE CENTRAL VENOUS CATHETER (PICC LINE), SOURCE OF QUESTIONS:

My skin is red and/or infected around the entry point, what can I do?

The installation of the central venous catheter (PICC line for central peripherally inserted central catheter) and the dressing change being done by sterile technique, it is essential to refer to the nurse of the cystic fibrosis clinic or to the specialized nurse in venous access for any problem related to the catheter.

If redness appears on the skin and/or near the insertion site, it may be an infection. If it is superficial, it is sometimes enough to change the dressing more frequently or by disinfecting it with chlorhexidine, but it is important to notify the team to do a close follow-up and eliminate an infection. deeper or another complication (e.g., allergy, thrombosis) on a case-by-case basis.

I have pain at the entry site, what can I do?

The causes of pain at the entry site are multiple: intolerance or allergy to dressings, blockage or thrombosis of the catheter, poor positioning, infection, etc. As mentioned, it is important to contact your nurse to reassess the patency of the catheter and to ensure that there are no signs and symptoms of infection.

What can I do when there are bubbles in the tubing?

Baths are best avoided. However, if you insist on taking a bath, the arm where the catheter is located should not be immersed in water. It should always be kept out of water.

For the shower, protect the arm where the catheter is located with a transparent film such as Saran Wrap and an empty plastic bag.

If your dressing accidentally gets wet or shifts, it will need to be changed. You must once again contact your nurse to change the dressing.

What is the impact of alcohol consumption when living with cystic fibrosis?

There are several repercussions of alcohol consumption in individuals with cystic fibrosis.

First, alcohol is a depressant that can affect breathing when consumed in large quantities. Respiratory rate and range of respiratory motion may be affected, making breathing, and managing secretions more difficult.

Additionally, since alcohol is high in sugars, diabetes control in patients with CF-associated diabetes may become more challenging, including a high risk of hypoglycemia.

In patients with known CF-associated liver disease, it is recommended that alcohol consumption be avoided or minimized, as it may exacerbate existing liver damage or hasten its progression.

Many antibiotics and drugs are metabolized by the liver and their effect can be affected by alcohol consumption. Some may amplify the effects of alcohol, while others will not work as effectively in the presence of alcohol. Caution is advised with CFTR modulators (e.g., Trikafta®), which on their own can impair liver function, which could be aggravated by heavy alcohol consumption.

Finally, several other medium and long-term complications are possible: anxiety, sleep disorders, vitamin deficiencies, high blood pressure, gastric ulcers, cancers, etc.

Some people have started taking the appetite-stimulating drug cyproheptadine. What is this drug? Is it useful and suitable for CF?

Cyproheptadine is a drug belonging to the class of first-generation antihistamines.

It is mainly used in the pediatric population to stimulate appetite and improve nutritional status but is not only used in individuals with cystic fibrosis. However, some studies have been conducted specifically in the cystic fibrosis population and have demonstrated good efficacy.

Before using this medication, you must have tried other options (assessment by a nutritionist, adjustment of intake, supplements, etc.) and discuss them with your attending physician, who will assess the situation and can determine if such treatment is appropriate.

With the advent of high efficacy CFTR modulators (Trikafta®) and their impacts on nutritional status (weight gain, improved BMI), there should be fewer and fewer reasons to start taking cyproheptadine in the future.

Exercise doesn't take time away from your life, it adds life to your time!

Health has been at the heart of our organization since the beginning!

SVB, June 1991, Issue 13

SVB, Winter 1995, Issue 19

SVB, Winter 1996, Issue 20

LIVING WITH CYSTIC FIBROSIS

LIVING WITH CYSTIC FIBROSIS

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to promote and support the quality of life
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