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LIVING WITH CYSTIC FIBROSIS

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Equality for all!

It's not a secret to anyone: science evolves extremely fast. Since the discovery of the gene responsible for cystic fibrosis in 1989, we have seen prodigious medical advances and many medications have come to market to help people living with Cystic Fibrosis.

Of particular note, the arrival of drugs enabling the liquefaction of pulmonary secretions, such as Pulmozyme™. Although approved by Health Canada in the mid-Nineties, its high cost caused an uproar at the time, and the Quebec medical insurance program refused to reimburse it. An intervention of our organization, supported by partners, was needed for the government to review its decision. Due to our intervention, CF people in Quebec can now benefit from this treatment. About ten years ago, hypertonic saline was added to the range of mucolytic medications.

In the past years, we have seen the emergence of antibiotics that are more and more efficient to help us fight against infections that are still damageable. This new generation of antibiotics includes aztreonam (better known as Cayston™), which acts directly against Pseudomonas.

Advances following the discovery of the defective gene were expected until recently. Therapies directly targeting genetics are now conceivable. The pharmaceutical company Vertex has updated Kalydeco™, the first treatment correcting cystic fibrosis genetic defects in persons with a very specific mutation (about 2% of CFs in Quebec). Afterwards, Orkambi™, which targets about 50% of CFs in Quebec, was approved by Health Canada in January 2016.

Despite its approval, Orkambi™ is, unfortunately, not automatically reimbursable by our public insurance program for the many persons which could benefit from it. Indeed, the organization that recommended to RAMQ that the medication be covered, the INESSS (National Institute for Excellence in Health and Social Services), stated in October that the preliminary results are not concluding enough for the Quebec government to reimburse that treatment. However, some CFs have the chance of being covered by a private insurance that accepts reimbursement. It is nevertheless a minority of cases, the coverage conditions not being the same for all insurances.[1]

If we compare insurance coverage or even the Quebec hospital system with that of the United States, for example, we can consider ourselves lucky. In our southern neighbour, the survival median for people living with CF is 39 years old, against 53 here: the difference is enormous. This difference is easily explained when comparing the two types of healthcare accessibility. There, the medical coverage is uneven and some persons, once at the age of majority, do not have any coverage (including for medication). Some CFs sometimes have to reduce their medication dosage to lengthen their monthly prescription, particularly for enzymes and insulin. Even graver, in many States, it is not rare that people have to stop a medication or cannot complete a hospitalization.

We therefore have the incredible chance of benefiting from an important protection system and of being born in a country that prioritizes the social and health issues in its population. Nevertheless, this chance must not stop us from denouncing some unfortunate situations that we witness: when will we see equality for all citizens to access better treatments, independently from their medical coverage, whether it be public or private? If a doctor judges that the patient could benefit from a medication that was approved, why wouldn't it be covered by the public insurance program?

These questions of importance remain at the heart of our organization's commitment towards you.

Charlène Blais

Chairperson of the Board of Directors

1. According to our data, between 25 and 30% of our members have private insurance.

A common history of success

It is with great pleasure that I present to you the 2017 Edition of the *SVB*, the official magazine of **Vivre avec la fibrose kystique**. For Montreal's 375th anniversary, the choice of the cover page is not anecdotal. We find our president, Charlène Blais, at the top of Mont-Royal with her two young children. In the backdrop, we see the metropolis of Québec: Montréal. As a matter of fact, there are 2 dates to remember: 1642 and 1985.

In 1642, Jeanne Mance and Pierre Chomedey de Maisonneuve found the City of Montréal. This small, poorly populated, religious community had a rough start: threats from Mohawks (Iroquois) Indians and the rise of the tides of the St-Lawrence River nearly destroyed the city several times.

Above adversity, and the hesitations of the representatives of the King of France, the creativity and resilience of our ancestors allowed Ville-Marie (Montréal's first name) to survive and grow until the XXIst century. Now, the city is the second French-speaking city in the world. At the beginning, Jeanne-Mance, our founding mother, built a hospital that still exists today: the Hôtel-Dieu of Montréal. This is where, in our modern days, we find the largest cystic fibrosis clinic in Quebec.

Three and a half centuries later, in 1985, our organisation is founded by a few young and impetuous free and determined spirits. Courageously, they regrouped and faced many challenges, most notably: financing the organisation and fighting against crossed-contamination. Very few people in the cystic fibrosis community were convinced that having 2 distinct entities working for the cause were necessary. However, as time went on, we were able to build many partnerships, insuring our existence and financing. We must also be proud of our crossed-contamination policy which still is in effect today and allows us to organise meetings and events of all types while preserving the health of those living with CF.

Nearly 30 years later, we are stronger than ever! We're among the first most important information diffusers concerning CF in the world. While research has allowed us to advance considerably (the median survival age being of 53 now), we begin 2017 with confidence. We are certain that the best is to come. In fact, we can now continue our mission without any of our past fears and we'll be able to offer even more services to our community.

While we have 375 reasons to celebrate our great city of Montréal, we have thousands more reasons to be happy and enthusiast towards our organisation. Collectively, we have been able to keep track of our objectives and have ensured a healthy governorship of the organisation. I'd like to warmly thank those who participated in our story.

On a more editorial side, I'd like to remind everyone of our ongoing battle to have in-vitro fertilization covered by public Medicare again in order to allow women like Charlène to give birth to children and know the great happiness of founding a family. The two children on the cover page, magnificent twins, were conceived thanks to that medical technique. I'd also like to remind everyone that we are still waiting for newborn screening to be accepted by the Quebec government. We are one of the last few remaining places in the west to not have this service. Let's continue our battle and don't forget to speak to your MPs about this important subject.

Just like Montréal, bastion of French culture in North America, **Living with Cystic Fibrosis**, the only organisation in Quebec to be present and offer direct help to CF patients, is unavoidable for those living with CF and their loved ones.

We'll always be there for you.

Tomy-Richard Leboeuf McGregor
Editor of the *SVB*
Executive Director

Gathering for better success

Interview with Véronique Hivon, instigator of the Quebec legislation concerning end-of-life care

Seasoned lawyer, committed politician, mother and passionate about social and health justice, Véronique Hivon is a captivating figure of the Quebec political community. Graduate of McGill Law in 1994, she then studied at the London School of Economics and Political Science, in England, were she developed her knowledge on the role of social measures in communities.

Interview held by
Tomy-Richard Leboeuf-McGregor

She is often defined as an anti-politician that can claim being as popular and appreciated by supporters of her political party as well as by the population and adversaries. Entering politics with the conviction that she had to change things from within, she is the Member of the National Assembly from Joliette since December 2008. In 2012, as a member of the Cabinet of Ministers, she introduced Bill 52, titled *Loi concernant les soins de fin de vie* (Act Respecting End-of-Life Care).

This is an interview with a politician that was able to pilot this touchy file, which generated important discussions with people living with cystic fibrosis and the entire Quebec population.

Ms. Hivon, what made you decide to get into politics?

I first worked as a political staffer with two Ministers, and then worked as a lawyer for the Government of Quebec. I was also involved as an activist in my party and many told me I had the required qualities to go to the front of the stage. I love meeting people and expressing my opinions very much. With time, I wanted to develop my own professional skills and to go to commit myself in the fullest.

Three strong reasons definitely convinced me to get into it. Firstly, I felt that the newer generation should take its place in politics. As you know, I'm a sovereigntist and it is important for me that we renew our discourse. Then, the question of social justice touched me and I felt that it should be a priority. Finally, I made the jump because I wanted to change things from within. It is an important theme in my commitment, I know there is a real disconnect between citizens and politics. This disconnection, I understand it: people don't want to see politicians fit into the traditional mold. I had the will to change these things!

In the 2012 elections, you were re-elected as Member of the National Assembly for Joliette and you entered the Quebec Cabinet of Ministers. What were your responsibilities then?

The Prime Minister of Quebec, Pauline Marois, gave me many responsibilities. I was named as the Minister responsible for social services and as the minister responsible for questions regarding end-of-life care. I was also named as the Minister responsible for my region, Lanaudières. It is important to know that the Ministry of Social Services is a big responsibility, 7 billion dollars are allocated to it. Its role is wide as it touches all spheres of wellbeing, whether it be physical deficiency, autism, drug addiction or homelessness, as well as child protection services. In a nutshell, everything that touches social questions far and wide. As you can see, it was very diverse!

On June 12th 2013, in the National Assembly, you introduced Bill 52, titled Loi concernant les soins de fin de vie (Act Respecting End-of-Life Care). Where did the idea come from?

It is an idea that I had put forth at the beginning of my political commitment, and that for many reasons. During my Law studies, I understood the importance

of individual autonomy so that the person can decide her or his fate. I had read a lot of jurisprudence on the subject, including the famous Sue Rodriguez case. Suffering from an incurable disease, she started a long judicial fight in 1992, demanding the right to die with dignity with the assistance of a physician. She appealed to the Supreme Court of Canada. The justices, in a very close judgment (5 against 4) had rejected the demand. I also had that chance to take medical ethics classes.

Then, I was also face with this situation when, people close to me, at the end of their lives, had greatly suffered. I also know people that had access to exceptional end-of-life care. For all these reasons, I had, on multiple occasions and at different moments, the time and the need to reflect on this question.

Politically, I thought it was essential that elected representatives look into this issue. We could not wait if, for or against, courts would trace a path for this legislation. I am convinced that on such delicate topics, we have to roll our sleeves and work with the population. Elected representatives must tackle these types of issues rather than ignoring them because they are difficult questions that can offend part of the electors. We have to assume our responsibilities!

Many people were opposed to medical aid in dying. How did you succeed in getting such wide support from the population?

Firstly, we took all the time necessary to lead different consultations. For issues of such importance, it is imperative do have the means to succeed. When trying to effect important social and human change, you have to be in tune with the population. By working well at all stages, it is possible to build the necessary consensus.

I would also say that we were able to work in a non-partisan way. We put together a commission that went to meet people in different cities in Quebec. We have to remember that the issue does not only concern medical aid in dying, it was also a question of all end-of-life care. We had to ask ourselves how to best assist people in this situation that we will all face someday. This Bill therefore also concerned palliative care and advance directives, for example. I believe that it also reassured part of the population, we really wanted that individuals concerned by this care be at the heart of the decision-making.

According to a majority of the population, this Bill became indispensable and was a real step forward for our society. That is why we wanted to work collectively: so that it would be a success. People from all fields contributed, positively improving our approach. Thus, even those who were *a priori* against this Bill, were not refractory at its conclusion.

Allowing all the time that we needed also enabled us to better share our message with the media. All the medical, legal or ethical issues raised were communicated to the population and debated, gradually accompanying persons that still had misgivings.

What are the main differences between medical aid in dying and palliative care?

When we talk about palliative care, it is about alleviating both the physical and moral pain in a person, but also relatives. It is to treat that person in a global and holistic approach, while taking into account all the needs to feel well. It is not about provoking death, but to accompany the person in end-of-life.

Concerning medical aid in dying, it is an element in an exceptional, very precise situation where despite the best care available, we cannot alleviate all of the pain suffered. This leads to physiological and psychological distress. Hence, with the assistance of medical staff, the person can leave serenely according to her or his will, and end intolerable suffering, when it does not make sense anymore for them.

The federal government was required, by the Supreme Court, to authorize medical aid in dying everywhere in Canada. Are there differences between the Quebec Law and the Canadian Law?

Yes, there are some differences that are important. The federal law allows two forms of medical aid in dying, because it also allows assisted suicide. In Quebec, death is administered by injection by a physician: it is the medical field that acts. At the federal level, there can be cases where the person, with another protocol, administer itself death. Concretely, in Quebec, we have made the choice to include medical aid in dying as a service delivered inside the health system, in a continuum of end-of-life care. The other aspect is that at the federal level, a person with a severe handicap could request it, while in Quebec only persons suffering from a serious and incurable disease who are at the end of their life can request it.

The moment we can resort to it is also different. In our Law, we have also included that the person must also be at the end of their life, but there is no precise period defined to allow a certain margin and so that each situation can be interpreted according to different criteria. We can usually expect that it is for a life expectancy of less than a year. At the federal level, they opted for a wider definition, that of reasonably predictable death. There is still a lot of debates to define exactly what this expression signifies, but generally, we believe that it covers a greater period than that of the end of life.

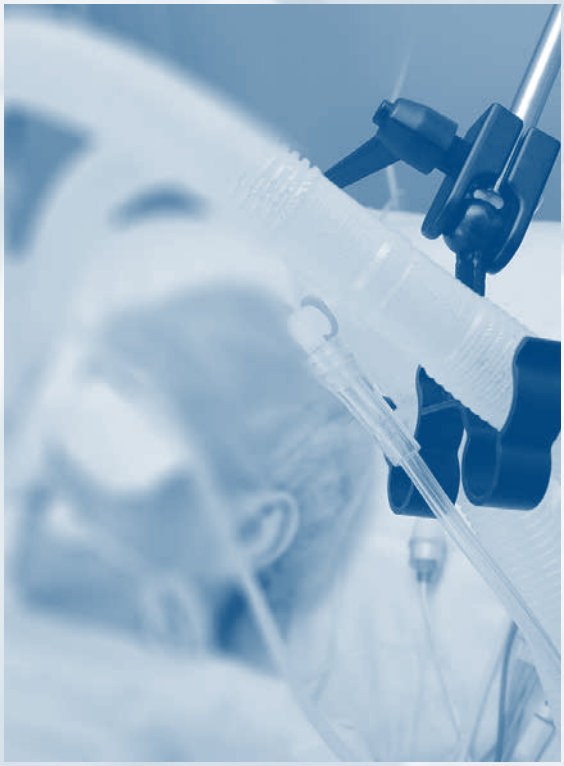
There are also some technical differences: on the federal side, a 15 day delay is required between the request and the administration of the act. When we know that a day can be like an eternity for a person suffering and in end-of-life, I believe this difference is unfortunate.

It is also important to say that our law was thought so that we remain in our exclusive jurisdiction, health. Quebec is not bound to follow those differences.

You are often cited as an example for your non-partisan approach. Do you believe that this is what allowed that support of a majority of Members of the National Assembly and of the population to this Bill? Do you believe that this way of doing could improve the climate of discussions in the National Assembly?

Yes, the non-partisan aspect was the cornerstone of this success. People felt that the elected representatives of all political parties worked for the population's superior interest, for the advancement of society. Often, at the end of consultations, people came to tell me that this approach had reconciled them with politics! They found it beautiful to watch Members work together, in collaboration. Many even highlighted that sometimes, you couldn't who was in which political party! This debate was not polluted by the will to make political gains.

Furthermore, I obviously think that we should work like that more often. Generally, elected representatives should avoid partisanship, in the negative sense of the term. Of course, I believe that we have to be in politics with strong beliefs, that we need diverging points of view, because it is the debate of ideas that can advance a society. When you go in politics, it is to fight for an ideal! However, I believe that when we discuss sensitive questions, we should work without partisanship, with everyone's baggage and ideas. It is all of society that will benefit, because the debate is constructive and it allows to go to the heart of the issues.



Apart from medical aid in dying, are there any other accomplishments that you are particularly proud of?

I am very happy to have put into place the first fight against homelessness policy and of the work I did as a Minister to help persons with deficiencies or with autism. Those were small steps and there is much left to do, but there was a will on my part to increase resources, particularly for those 21 or older that sometimes experienced a break in services.

Beyond all that, I'm very proud of all the work I accomplish daily in my riding, as Member of the National Assembly for Joliette. When I can help someone get their social security check or with an occupational health and safety problem, a distressed person or another one living a difficult situation, I find a meaning in my work as an elected representative.

In your opinion, what should be the priorities in terms of health and social services in the upcoming years?

It is a very wide question! There are so many issues…

Following my experience as Minister for Social Services, I think that we should have a more global approach when we assist a person with an illness or serious problems. I believe we need to eliminate the compartmentalization of services and care delivery. It is very difficult for someone to have complete patient care that makes sense. We take care of the medical side, then a social worker can take care of the other. Often, there is no contact between both parts.

We must now reverse the pyramid. Rather than going from the service and to impose it to someone, we should go from the person and make a game plan adapted to their needs. We also need more pivot resources that would always be in contact with the concerned person. This would ensure fluidity and more efficiency for the services that are given.

We must also better recognize the importance of social services. I often say that it is not because you don't bleed that there is no emergency! For example, we emphasize interventions to the hip, cataracts and knees, for which we have quantified targets, with an objective to meet. These targets are obviously very important, but we don't frequently find them for many illnesses and social realities, such as dependence, persons suffering from autism or from homelessness. Without these objectives, these persons are sometimes forgotten and the allocated resources aren't adequate.

Ms. Hivon, thank you very much for taking part in this interview! ◄

A roller coaster relationship

Christine
Québec, Quebec
Canada

Falling in love isn't always easy and once it happens, we fly.

We already knew each other and I knew he had cystic fibrosis (CF). Having graduated in nursing, I had studied the subject at the time and was shocked at the time by the unavoidable age of their death. How could we give someone, without choice, a deadline for life?

Time had made its way and we became a couple. Our relationship wasn't built on this contingency. I did not fall in love with a sick man, but with a guy that faced, tackled and sometimes confronted life. His attitude came without doubt from the fact that his being forced him to LIVE! Living without worrying for the worst, living in the present.

Living with a person with CF is loving adventure and being challenged continually.

My lover opened my heart and seduced me with his humor, his sarcasm, his strong taste to bite into life. Living with him, it's living a roller coaster of emotions, it's letting go of some fears and building objectives. You must sometimes close your eyes on some thoughts that can hurt and staying strong next to the proud person that doesn't seem affected by illness.

We have lived through hospitalizations, many meetings with the multidisciplinary team. I had many disagreements with the nurse who found me annoying when I worried too much because he was not concerned with his treatments and follow-ups. The nurse works in the hospital, but the one that cries and screams in silence, the one who is afraid, is the WOMAN that shares her life. It's my fault, I want to live with him no matter what and for many long years!

YES, we are at the edge of a pier and with any gust of wind we can fall.

We don't need to be married to live "together for better or for worse". My boyfriend worried in the beginning of our relationship that I would fall in love with him. He didn't want to make me live in the pain and suffering of his illness. I made him understand quickly that I wasn't there only for the best and that I wouldn't let him go, even in the worst moments. I want to share his joys and fears. We don't want to live with fatality but with optimism!

Often people tell me: "you're young, wait until you're my age". I answer "you should know that it's a privilege being your age, some people don't make it there". Their faces, their speech and their perceptions then change and they reply: "you're right". My lover looks healthy and in good shape. If you knew at what point he is super powerful in that condition. I am proud of him because he never stops biting into life and always wanting to go further. I would lie if I said that I don't worry when seeing more symptoms or that I don't wait impatiently for his FEV1 results at each meeting. I must often take a step back. I care about him and his life. He is not alone, we are TWO.

Helplessness is a feeling that the ill person or the family can feel. The illness is a boomerang and we would like to do more, but CF or not we must rely on medicine and science. My man is well and lives with full breaths, if I can make a play on words. ◄

Letting go will always surprise us!

Lyne Cuillerier and Michel Ferland
Terrebonne, Quebec
Canada

This summer, I was asking myself what would happen with my husband Michel Ferland, living with cystic fibrosis. He was breathing at barely 20% and I was seeing his health deteriorate before my very eyes. I felt helpless with this situation.

Between coming and going from the hospital to home, Michel tried to do some activities with us, despite his physical situation that prevented him from breathing normally. He never complained. He was tired quickly but he was holding on with his oxygen bottle in his back, tube in his nose! Socially, the situation wasn't always positive: it's not easy to walk around in this society filled with judgment and sometimes inquisitive looks.

I found it very sad and hard to see him that way. In everyday life, Michel never stopped and we had always gone out and done many activities together.

During the summer, I asked myself when would be the best time for a small getaway with my daughter, to change scenery. The decision for the right moment was difficult because we never knew when the call for a transplant would come!

On Thursday August 18th, my daughter and I finally left for L'Isle-aux-Coudres for our little trip. To find ourselves next to the sea, to take deep breaths and to appreciate the beauty of nature surrounding us, would lift our spirits! Arriving on the island we looked at the spectacular landscape. The sky was full of a 1001 colors. It was a full moon night, what more could we expect! I decided to send Michel some pictures so that he could also admire the beauty of nature.

That night, I slept very badly. I felt guilty to have left Michel at home.

The next morning, we had many opportunities for day trips, recommended by the information centre. As if we felt something would happen, we decided to postpone everything for later.

We chose a little trip by train, by the water, that was recommended to us. Once there, we parked the car somewhere that had phone network.

It is at that time that we received the long awaited call!

What a relief! Tears of joy were streaming on my cheeks. I took the time to call relatives, to share the good news with them. I was so emotional!

At the same time, I looked up to the sky, a cloud had the shape of a bird, as if it came to bring me this wonderful message.

We decided to still go on the trip, to relieve tension and to then calmly take the road afterwards. 1001 positive images were going through my head.

The operation was planned for 8PM. I wasn't able to get back on time to see him off to the transplant. But with a turn of fate and delays we were able to be there on time. We held him in our arms before the long awaited departure for surgery!

That night, I never thought negatively. I was confident, everything would go well. Like Michel, I only saw the brighter side of life!

When we woke up in the waiting room, the doctor was there with a large smile on his lips. At that moment, I knew everything went well.

Letting go brings great things. I simply had to believe and stop to listen to the beautiful messages that life send us.

Today, we regain our momentum on a new path of life.

A second chance for us.

A new start.

One day at a time!

Hang on, life brings us great surprises at the moment we expect it the least! ◂

Testimony by Michel Ferland

We are on August 17th. I have to get to Hôtel-Dieu Hospital because my body can't breathe anymore!

For some time, 24 hours a day, I get tired at the least effort. Entering and exiting the car, taking a little walk and brushing my teeth are very demanding tasks for me.

I even had episodes of urinary incontinence. Often, without a breath, my stubborn head went over the logic of my body's limits. I am drowning trying to breathe. The image is clear: I am at the bottom, I rise back to the surface to meet the fresh air that will free me, but a glass ceiling awaits me.

« Michel, with 20 % respiratory capacity, we must keep you in the hospital until the surgeon calls. »

I don't want that! Please, no I don't want that!

Waiting eternally for the surgeon's call. The word eternal is in my head. Yes, eternal.

Other people have lived much worse than me, but I wasn't capable of imagining living in their situation. I didn't want to believe it, but I probably would have to go through this step.

My voice changed in front of the doctor. Tears fell on my cheeks with my mother sitting beside me. Lyne, my partner, was not there as she was on a weekend trip with our eldest daughter, Annabelle. A well-deserved rest! I didn't want to give them this news, I wanted them to relax. Anyways, they know in what state I am. I had already spent all summer inside these walls. I didn't see myself waiting two, three or six months.

"A person is in charge of giving you a bed for your stay here and will call you tomorrow morning to inform you at what time you can come back…"

I wander around with my mother even more slowly than when I came in the hallways of the hospital. My shoulders were down, demolished.

I keep thinking about Lyne who is away. How to tell her that she will have to wait, alone, at home, taking care of our daughters? I decide not to tell her, I will let her finish her trip with Annabelle so that they can fully enjoy it. They will know soon enough when they come back.

The call

The next day, I was lying in bed with my mother in the room. We were waiting for the call from the person that would assign me a bed at the hospital. That wait seemed eternal! I did not want this option. I am moody and I have troubled thoughts. It isn't going the way I wanted and I accept it difficultly. My mother and my youngest daughter, Jessyca, are there.

Lying down, BiPap on my nose, talking with my duck voice, I discuss the last days' and the past years' events with them. I never believed that, 6 months ago, I would still be waiting for this call! A bed while waiting for the transplant. I was sure that I could go through this step peacefully at home.

It is almost 11 and the person in charge of admissions should soon call. I know because this isn't my first time waiting for a call for a hospital stay.

11 o' clock: Ring ring! Unknown number, it isn't the person from the hospital!

"Hello Mr. Ferland, this is Dr. Ferraro! I announce you that we have beautiful lungs for you!"

The transplant surgeon surprises me like a mirage. He tells me the surgery will be tonight. They have new lungs for me! Lungs younger than me and in full health!

I burst! I scream and cry over the phone, I jump in underwear on my bed!

Where was this energy a few minutes earlier? The BiPap comes off my face and I can't believe what happens! The pressure falls off.

"Be at the hospital at 3PM. The operation will be at 8PM, if everything matches with the donor" he tells me, calmer than me.

Jessyca, our youngest, already called Lyne in Tadoussac. In a few moments, people come home to support me. We cry, we laugh! I don't know what to think and everything goes quickly. I have to pack my luggage? No! Toothbrush, soap, shampoo, clothes. That's all. I will spend most of my time in a gown anyways.

Inevitably, I think of Lyne. How does she feel being so far? How is she managing the situation? Will she feel guilty being away or not? She shouldn't! She needed that rest. She will be back anyways, and she will be back just in time before I leave!

Her absence was only a matter of chance. Her being away maybe even got thing going, who knows? For me, the adventure begins. Each second entering the hospital, it becomes "real".

At the hospital

From the reception to the intensive care unit, strangely, I was serene and calm. The attention, the smiling faces, it is as if I was in an all-inclusive resort, no joke! Here, the care is comprehensive. I am plugged everywhere in 20 minutes: pulse oximeter, electrocardiogram, and solution.

I am sent to the waiting room and my relatives have the permission to come back at my side. I see them in chick costumes: gowns, gloves and masks! No germs can get in here! From that moment, I realized that I wouldn't see smiles for the next two weeks, they were hidden under the masks. I know what would be hard psychologically. Faces I wouldn't recognize: doctors, nurses, visitors, only eyes that speak and barely move. I was announced the surgery would be at midnight and not at 8PM.

I can't wait to leave to continue on this adventure in a more concrete way. Lyne and Annabelle have finally arrives from Tadoussac and, before going to the operating room, I can hold them in my arms!

"Are you ready Mr. Ferland?
Here we go!"

I am excited and calm at the same time. I leave the room and pass in front of my friends while coughing: people laugh and say this is probably the last time they will hear me couch like that!

I can hear applause. I move in the hallway, I show a thumbs up. I back into the elevator, showing a heart with my hands and the door closes with a goodbye. Going to the operating room isn't very long. Everyone starts working, very concentrated, without a smile. I am surprised because their attitude is very different than the one they had since my arrival. But I understand they have a very serious job to accomplish and that they must be ready, as well me, for this challenge. I talk with the nurse while the anesthetist installs the epidural. I confess to her that this moment is incredible, that I am the happiest man. She looks at me with laughing eyes where, under her mask, I can imagine a smile. I am well installed, I am injected the magical liquid that will put me to sleep.

"Goodnight, Mr. Ferland, see you later."

I say goodnight with a smile that disappears in three seconds.

3, 2, 1. ◄

Life goes on!

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Seventeen years ago, a little girl was born. It could've been any common story, only this one wasn't. When two days old only, the little girl had to be operated because her digestive system didn't work properly. First sign to the doctors, they made some tests to confirm their first hypothesis and brought a name to her parents that they did not know before: cystic fibrosis.

Today, many years later, the small scar is still there on my stomach, among others. That of my feeding plug, which artificially fed me for years. Those of my many antibiotic perfusions, which dotted my arms in my childhood. That of my old catheter, higher up, when my veins weren't enough for perfusions. That of my lung transplant, when everything else wasn't enough.

I was thirteen years old when my state started deteriorating. Even with daily treatment, countless vitamins, antibiotics and others, despite regular visits to the cystic fibrosis clinic, I had to spend some time hospitalized at Sainte-Justine Hospital. I left less than a month later, reassured about my health.

Only a year went by between that moment and when I started sleeping every night with oxygen. For the first time, we were talking about greater measures, more important than simple medication. The word transplant was added to my vocabulary, and while I was conscious of the necessity to resort to an operation someday, I didn't expect it to happen so soon.

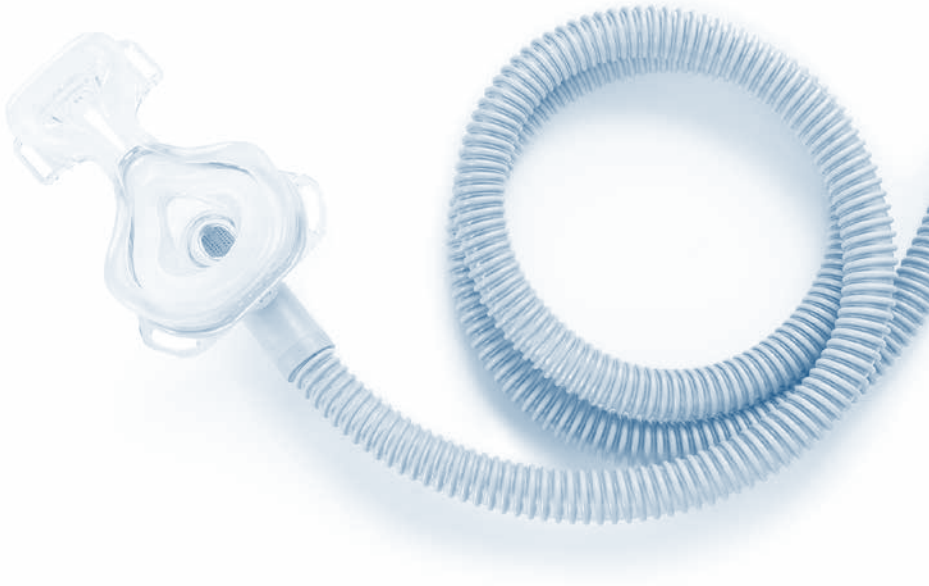
That same year, I was in class for only two of the ten months in 9th grade. Then, I had the precious help of an amazing university student even when I didn't see the importance anymore. I went at least half a day to the hospital every week, to make sure my state didn't worsen too fast. I saw a friend, amazing also, and the rest of the time I could only sleep, exhausted.

It is difficult to realize the energy that breathing requires. At the end of my school year, I kept my oxygen all day, totally dependent on these little O2 molecules. I never left it and, despite this help, I still had difficulty breathing. Between long coughing fits and rest periods, in little time I was drained at the least effort.

Parallel to that, my appetite was ruined by the enormous quantities of medication I was administered each day while I needed more calories than ever. With a feeding device that I had since I was eleven years old, I could now have access to a greater nutritional value. In total, I received 4,000 calories daily, less those that I vomited. I still had to gain two kilos if I wanted to be accepted on the waiting list for a transplant.

My 10th grade just didn't happen. I followed some classes, tried to stay up to date, but I didn't have the energy anymore. I watched many series, of which there are many seasons. I read a lot also, many novels, but I was quickly tired. During the day, I did everything to stand up the least, and I took many naps, not even going back to my bed to sleep.

In the beginning of the month of December, I started using a new machine to help me breathe at night. That way, I could have many hours, each night, where I would have real rest. In the beginning, I wasn't a fan of this big mask, but I quickly accepted its presence. Sometime after, I was in a hurry to wear it, putting it as soon as possible, and always waiting as much as possible to take it off.



I had become evident: breathing by myself was now a difficult task to accomplish. At that time, my lungs were working at less than 20%. It was impossible to know how low I had come down to because, under that limit, measures aren't accurate enough to calculate pulmonary capacity. Nevertheless, I knew it had worsened to far less.

After the Holidays, I was admitted for another stay at the hospital. That time, my status had deteriorated too quickly, to a point where it was worrying. Climbing stairs, or simply walking down the hall, everything exhausted me alarmingly. I liked reading so much, but I couldn't concentrate on a page for more than a few seconds. The words, the ideas, everything was mixed up in my head.

My body had, more and more, become resistant to normal antibiotics and I had to test some new, less traditional ones… Some hadn't been used for many years, due to their sides effects that were too numerous and important. Regardless, they had very good results on me and, by monitoring them, we avoided any complication.

After two months in a normal unit, I was transferred to intensive care at the hospital. We couldn't wait anymore, my doctors completed all the steps necessary and I was admitted to the emergency transplant list, passing all others. From then, I only waited a short time, even though it felt like eternity.

I have very little memories of that period, drugged by medications I received. Tubes in my small hand veins would burst after only a few hours, with pain coming with it. The book purchased by my parents, placed against the sink, stayed new and abandoned, words being confused in my eyes. These short instants, awoken in the middle of the night in the hospital's relative silence, which I hadn't left in two months. My two younger sisters' eyes were full of incomprehension and sometimes breaking tears.

I was transferred again 15 days later to an adult hospital. I had imagined that it would be a great change, but I was barely conscious enough to notice the difference. At one place or another, my main activities were to sleep, stare at the ceiling and talk out loud without making sense. The day after I arrived, my surgeon came to tell me he had lungs for me, and it must be the only thing I remember that well.

According to my attending physicians, two weeks later, I would have been dead.

I spent a little more than a month hospitalized before finally going home. I had to relearn everything; talking and eating despite my irritated throat due to intubation, to walk despite my leg muscles having suffered from all this time without exercise, to socialize even though my only company for moths was my family and some rare friends. But I succeeded, and fully enjoyed my summer before going back to a normal life.

Two years after having left them, I was back on school benches. Like any normal teenager, I worried about my grades in math, complained about some teachers and argued for anything for reasons I forget. I am only a year late compared to my friends, but I also made some new connections. Next year, after getting my high school diploma, I will join them for college studies, in a field that I am passionate about.

Of course, I still take many medications to avoid my body rejecting these new lungs, and I have to take care of my health as I have no or almost no immune system. Having come all this way, I think it is very little for me to be here. I also started reading again, losing a lot of time watching television and procrastinating with homework until having no choice left but to do them.

As we say, life goes on! ◂

Who controls my life?

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Cystic Fibrosis is without doubt a chronic illness with significant impact on the psychosocial functioning of individuals living with it. Indeed, it affects the personal, family and social spheres as much. It also has consequences on professional and academic life, as well as daily and domestic activities.

According to the American psychologist Julian Rotter (1916-2014), each human person tends to attribute elements affecting them to causes that are mainly *interior*, or *rather exterior*. Obviously, individuals with cystic fibrosis do not escape that reality. We can, as a matter of fact, identify in their speech elements attached to one or another of these opposing beliefs.

Also according to Rotter, we will say that persons who believe having full control of their life have an *internal locus of control*, and people attributing what happens to them to external causes, an *external locus of control*. In the literature, we also talk about *internal and external control position*. For the psychologist, these two notions must be understood as a spectrum of possibilities, and not as absolute values. Thus, the locus of control in most individuals is somewhere between both.

The primary objective in the current text is to help you better situate yourself on the scale of different loci of control by examining you beliefs and questioning your certainties. Secondly, it will allow you to take conscience of the forces of your belonging to one or the other of the two groups that allowed you to develop, through your process of accepting the illness. Finally, it will bring you to identify the limits inherent to each of these schools of thought, so as to better recognize the different challenges that you will possibly have to face in the future.

The internal locus of control

Persons possessing a locus of control that is internal in majority generally believe that nothing happens for nothing. They establish a very clear causality link between their actions and the consequences they have on their life. If they failed an important exam at school, it is that they did not study enough. If they did not get the promotion they expected at work, it is that their performance at the interview was sub-par. If they arrive late at their medical appointment, it is because they did not wake up earlier than usual to forecast the traffic generated by the snowfall. In regards to illness, these persons will also expect that their clinical status reflect their diligence and determination in accomplishing their treatments.

When their medical state is stable or, even better, improved, there is no word to describe the pride that shows through their smile. The spirometry result matches with the efforts made and all is right in the world. However, when there is a deterioration of the clinical state, these persons have the reflex to blame and guilt themselves. They will remember all the occasions where they did not give their maximum, and set a direct link between these occasional digressions and the state they are now in.

Thus, when it goes well, having an internal locus of control allows the individual to keep a positive and confident attitude towards the future. This also fosters their commitment at school and at work, as well as therapy adhesion, because they are convinced that their state is the product of perseverance. Furthermore, by profoundly analyzing each of success and failure, the individual presenting an internal locus of control develop a capacity for introspection as well as extraordinary adaptation strategies.

There are however some disadvantages to having an internal locus of control. An individual belonging to that group can sometimes be extremely perfectionist and feel a string sense of culpability or of shame when confronted with failure. Furthermore, because their personal esteem is intimately linked to success as well as accomplishments, they can easily become anxious or depressed when efforts are not rewarded by the expected results. Finally, this individual is usually more vulnerable to critics and to emotional blackmail, because on top of considering themselves entirely responsible for their state, they have a tendency for feeling responsible for the happiness or unhappiness of others.

To counterbalance the disadvantages of an internal locus of control, the individual can learn to make the distinction between feelings of culpability and the feeling of responsibility. For example, instead of feeling guilty uselessly by saying to themselves: *"If my pulmonary function is reduced this month, it is because I did not do my physiotherapy exercises everyday. I showed cowardice and now I get what I deserve"*, a person can rather be empowered by saying: *"It is possible that the fact of not doing my respiratory physiotherapy might have contributed to the reduction of my pulmonary capacity this month. My health is important for me and my actions should better reflect that reality. I give myself an objective to do better next month."*

In this example, the individual presenting a mostly internal locus of control exploits the forces stemming from his adhesion to this group while refusing to make violence by being overtaken by feelings of guilt or shame. It therefore increases considerably chances of bringing positive change in life.

The external locus of control

Persons that present a mostly external locus of control will, on their part, explain things affecting them by environmental factors over which they have no control, like for example, chance, randomness, others or institutions. If they failed an important exam at school, it is that the subject was too difficult. If they do not get the expected promotion at work, it is that managers doing the interviews made a bad choice. If they arrived late at their medical meeting, it is because of traffic after a snowfall. With regards to illness, these persons will entertain the belief that it is useless to have flawless therapy adhesion, given that a medical emergency can bring irreversible deterioration to their state at any time, regardless of efforts made to take care of their health.

To present a mostly external locus of control brings few advantages. We can think that these persons are less prone to anxiety, because they seem to be carried by life, without hassles or worries, but that is not the case. In fact, while many of these persons tend to feel less guilty for what happens to them, they are nevertheless anxious about the perspective of not having power over their future, no influence on their fate. The challenge to lift these persons will be to develop their initiative as well as their power to act so as to take back a certain control of their lives.

The good news is that no one has a 100% external or internal locus of control. Most of the time, the effects of an external locus of control are counterbalanced by elements of an internal locus of control, and vice versa. I tis also important to keep in mind that there is no good or bad way to interpret events that affect your life. In fact, each human experience is unique, complex and valid.

Conclusion

Concerning all these elements, we can say that knowing where our locus of control is situated regarding what we are experiencing is a good way to identify our strengths, as well as the challenges to overcome in accepting our reality. This quest is certainly not an easy task. It requires a great dose of courage. In fact, it appeals to our capacity to question ourselves, a skill that is not available to all and can sometimes make us fell discomfort.

If you wish to start a process of personal growth to better understand the reasons that motivate your thoughts, your decisions and your behaviours, surround yourself with persons with which it is possible to exchange and share your reflections without fear of being judged. Do not hesitate to ask for professional assistance by a psychotherapist that will listen and guide you, especially if your family and social network is unable to do so. Furthermore, some persons need less contact when they are introspecting and prefer reading or writing their thoughts in a journal. Finally, practicing an artistic or sports activity can also help center you and discover yourself.

Better knowing yourself is better understanding yourself. Better understand yourself is better accepting yourself. It is by working on self accepting yourself that you can develop more love and respect for the human being that you are. ◄

Source

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An alternative approach to illness

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Cystic fibrosis, despite its orphan disease status, is a disease that touches an important part of the population. This illness attacks intestinal mucus and bronchial mucus is due to the mutation of a gene, the CFTR, that provokes severe respiratory and gastric troubles in the patient while sometimes attacking the liver, sinuses and the kidneys.

Thanks to progress in Western medicine, the median living age was, in Québec, of more than 53 years in 2016. Regardless of all this progress, cystic fibrosis remains an incurable disease with which we have to learn to deal with, in a sometimes failing body.

These days, many look at treatments of illness and the body in a holistic manner. We must have an overview, with different approaches, of alternative medicines that seek to improve quality of life for patients and their relatives.

We will present here a set of techniques and advice that each is free to use without forgetting that these approaches represent precious help for daily life in complement with classic treatments: in no case should they replace medical treatment prescribed by physicians.

These approaches seek to help the ill person's body in suffering and in supporting it in the psychological process of self-development. In fact, most alternative medicines rest on spiritual and psychological development that feeds the body, and not the opposite. Therein lies the interest in alternative medicines presented here: feeding the soul to rebuild the body.

Yoga and pranayamas: revitalizing the body

Yoga is an ancestral art that can be practiced by all no matter what age, gender or state of health. This practice proposes postures that stretch and tone the body while offering the possibility, for some, to lead to a state of awakening and serenity.

How does yoga lead to a meditative state? The different postures allow for an increase in abdominal and cerebral blood circulation while soothing the nervous system. The flow of blood in these vital parts associated with greater attention to breathing (pranayamas), enable a better oxygenation of the brain and develop better concentration. Furthermore, focusing on the position of each asana (postures) develop a better listening of the body: through each posture, all parts of our limbs are felt differently. Thus, the awareness of different body muscles and the increase in oxygenation of the brain allows the access to a state of meditation and awakening that brings mental clarity and generated a feeling of inner peace.

The term asanas is a sanskrit word that means "the act of sitting" or the manner of sitting". For yoga practitioners, this word has a more particular signification as a "ritual of posture". In the common language, it could be translated as "posture" or "way to sit"[1].

Three breathing exercises and asanas are particularly beneficial for intestinal and respiratory troubles. These exercises associate the benefits of conscious breathing to the circulatory benefits of yoga, both being intimately linked and allow access to meditation.

1

FIRST EXERCISE:
Malasana
and Bhastrika

Malasana
(posture of Mala, the Indian rosary)[2]

Hold your feet spread at the width of your hips, turn your feet outside, and lower your buttocks while completely crouching. You can join hands in front of your chest by pushing them against each other or by putting them on the ground in front of you. This position allows for better digestion because it aligns your digestive system perfectly. It is particularly advised to do it in the morning to start the digestive system.

The bhastrika pranayama[3]
(scream breathing)

This breathing technique oxygenates the brain and warms the body. Breathe in deeply with both nostrils, then breathe out quickly. Breathing out should be quicker than breathing in. Repeat between 5 and 10 times during the asana.

2

SECOND EXERCISE:
Tuck pose
and Viloma Pranayama

Tuck pose[4]

Put yourself on your back, bend your legs and bring your keens together on your chest, pass your arms around your keens. Lift your head and put your nose between your knees. This pose targets the digestive organs because it stimulates the apana (exhaling breath) which activates the digestive system.

Viloma Pranayama
(breathing by steps)[5]

Instead of breathing in only once you will inhale in steps: breathe 2 seconds, hold the air in 2 seconds and continue breathing until your lungs are full. For exhaling, it is the same: breathe out for 2 seconds then hold in for 2 seconds. Repeat until your lungs are empty. This breathing technique helps learn how to control your breath and prolong exhaling.

3

THIRD EXERCISE:
Suptamatsyendrasana
and AnulomaViloma

Supta matsyendrasana
(lengthened torsion) [6]

Once on your back, while keeping it on the ground (both shoulders must touch the ground), send a leg to the other side. The arm on the side where you rest your leg will put pressure on it. This position improves digestion, flexibility of the spine and relieves back pain. Torsions are excellent to fluidize the intestinal tract.

Anuloma Viloma
(alternate breathing)

Breathe in with the left nostril while closing your right with you thumb, then breathe out with the right nostril while closing your left nostril with your middle finger. Then, repeat the exercise by starting to breathe with the right nostril. Repeat 10 times. This technique establishes a regular rhythm for breathing and helps overcoming stress.

Full conscience and sophrology technique: feeding the soul

The full conscience meditation technique, whose origins are millennial, has often been used to improve the life of patients. This meditation technique adapted by Jon Kabatt Zinn, gives tools to learn managing pain induced by the body. As defined by Catherine Verhaeghe, specialist in human genetics whose doctoral thesis was on molecular inflammatory mechanisms associated to cystic fibrosis, this technique is the "action of giving attention to the present moment, with intention and without judgement." This approach enables improvements both psychological and biological: improvement in quality of life, of sleep, stimulation of the immune system and decrease in blood pressure, reduction of stress, of ruminations and of anxiety level.

It is therefore a daily meditative practice to leave what could be qualified as an auto-pilot mode to act in full conscience. More concretely, it takes the form of repeated and deliberate exercises that develop our capacity to focus on ourselves. Specifically, it changes our "mental speed", which allows us to enter an advanced meditative state. It is presented in the form of daily 30 to 60 minute sessions during which we participate actively in guided meditation.

Here is a simple exercise[8].

Breathe calmly, with forced inhaling or exhaling. Observe carefully the air going inside you, feel it literally pass through you. You will be surprised to see that you can.

Once the air fills you, feel the lifting of your chest, the brushing of your clothes against it, the way your torso is slightly shifted. Don't hold the air in, let it escape and do the same visualization work.

By concentrating, you can sometimes feel the beat of your heart. Observe them as well, see how the rhythm of your breathing affects them. Observe each sensation with your spirit and body. It is never a question of judging what you observe. Look at each thing as it is, that's all. You can practice this with anything you want. Each time, be fully awakened and open to the sensations that occur. See them one by one and observe how they are intricately linked and how it fits in a grander scheme. Don't lose the primary object of your attention, to go back to it when you get lost and to discover new things every time.

By doing an effort not to judge, you can see what comes from your own judgement and what comes from the nature of things. You will be better conscious of this judging constantly going on in your spirit.

Sophroplogy

Sophrology is another form of meditation. This technique brings patients to a state of "unencumbered" conscience in which they can focus on a specific need. In the case of cystic fibrosis and other chronic illnesses, it accompanies body visualization and representation exercises during which the patient learns to control pain intensity.

Here is a sophrology exercise, the "inner smile".

Install yourself in your favourite couch or lie down if you want. Take a few deep breaths: inhale through your nose, exhale with your mouth. Calm your breaths, this will calm your spirit. Breathe in and out again.

Let your memory go back to a happy moment. This can be a recent memory, from yesterday or from your childhood. This is a moment full of joy and good feelings. Relive it quietly, combining your five senses: give it colors, perfumes or smells, sounds or melodies, textures or tactile sensations, even flavours or tastes.

Observe the people present in your memory. Observe their faces, their joy, their good mood. Feel the link uniting them with you and let the joy of knowing them fill you. Breathe in and out deeply: enjoy this memory.

Bathe in all the feelings of this memory, observing all positive reactions in your body and spirit. Fully live the enjoyable feelings, the wellbeing, the joy, the happiness. Breathe deeply.

Bring your attention to the heart region. Inhale and exhale for a long time. Feel your heart bathing in positive feelings. You can make it smile, an inner smile of the heart. Your heart opens with this smile that fills your being... And maybe you will feel like smiling too!

These techniques and advice unfortunately don't create miracles. However, they are useful in daily life not to feel helpless with illness. They enable another treatment of the sick body and bring great support to the patient. Combining them allows not only to alleviate some harder to treat symptoms, but also to structure each day by offering an alternative to persons living with cystic fibrosis. Wellbeing has a thousand paths! ◂

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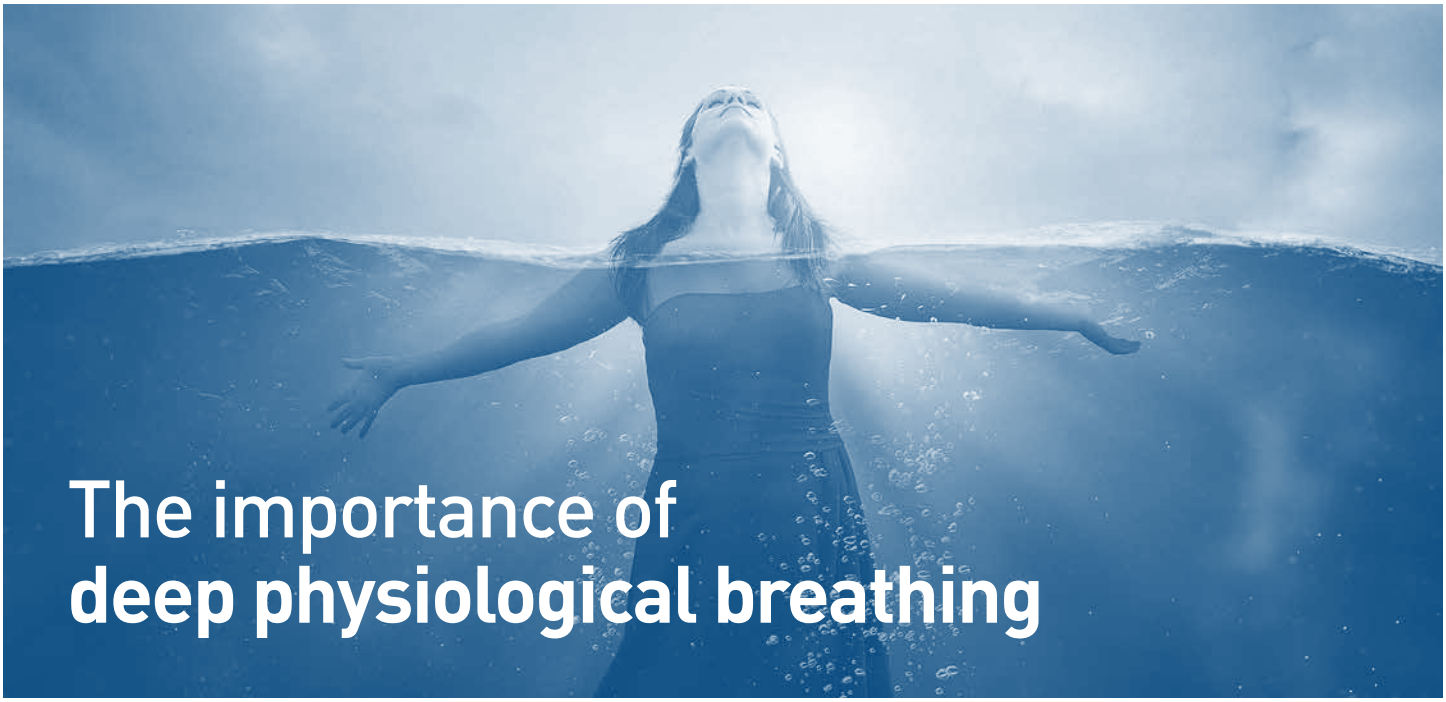

The importance of deep physiological breathing

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In all the great functions that sustain life, breathing is, with circulation, the most important. It is an evidence that the respiratory function plays an important role in our life. Especially when we have a respiratory deficiency, we understand how a difficulty in breathing handicaps our health. Mrs. Metge-Sandra spent all her life to developing her method. I was offered the chance to meet her in Paris and to be trained to teach her method.

Born in the South of France in 1909, she passed away in Paris in 2002. Grad Prix winner at the Paris Music Conservatory in singing and piano, singing teacher, she observed the impact of the respiratory function on different persons to help her students develop control of their body as an instrument. She also created breathing gymnastics enabling a deep physiological breathing mechanism rather than partial thoracic breathing.

She realized very quickly that her work went well beyond singing. Her method is based on deep physiological breathing, that which we have at birth. In this respiratory reeducation, all the importance is given to spontaneous exhaling. This respiratory work ensures the development of deep muscles: dorsal, abdominal, transversal and oblique muscles, enabling the diaphragm to be the engine of the breathing motion.

This method has preventive and curative therapeutic properties, providing physiological breathing and voice permanently to each human being.

Mrs. Metge-Sandra is one of the rare persons, in the respiratory field, to signify the importance of respiratory reeducation insisting on the "spontaneous exhaling" in the oral position of the smile.

Here is what Mrs. Metge-Sandra observed:

The majority of human beings do not know how to breathe and practice a respiratory mode sufficient to sustain life, but insufficient to ensure profound exchanges that guarantee a physiological and psychological balance that the modern man is far from knowing.

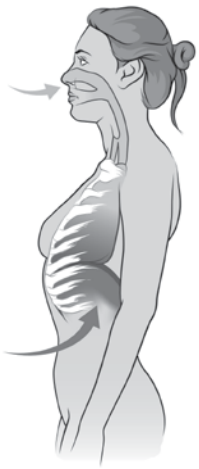
Less muscled due to a sedentary lifestyle, in a polluted atmosphere, with a relentless rhythm of life that uses its nervous system, the 21st century man does not know how to breathe, cannot breathe fully and practices a lazy and partial breathing, the thoracic breathing.

The truth is, any subjects practicing this incomplete breathing will become tired, whatever their activities, a famous fatigue, so common in the modern man, a general fatigue resulting in daily life in insufficient exchanges and a lack of oxygen in the body, a nervous fatigue that worries the medical world without, nonetheless, finding an efficient remedy.

It is with the practice of deep breathing, physiological breathing, that any subject can access, by "remuscling", to deep exchanges in the respiratory system in daily life, to calm and to the possibility of physical activity without fatigue.

Contemporary literature considers breathing as being the work of lungs with the help of the nose and mouth. The lungs collect and exchange gas (using the blood globules as a vehicle), but in reality, each living cell in the body plays a role in breathing. All body cells need oxygen to live, develop and produce the required energy for body functions. The role of lungs is to extract oxygen in air, to transfer it to blood that will carry it to body cells.

Breathing is life



Breathing has, among other functions, the regulation of water quantities in the blood system and a contribution to the body's thermal balance.

It is also noted that the fundamental process of breathing is the chemical use of oxygen by the cell to oxidize energy substances and to liberate they energy they contain. This results in the liberation of H_2O and CO_2. This is produced in mitochondria [1]. This is therefore a function of primary importance for each of our cells.

By its respiratory system, our body communicates mostly with the exterior world. It is with a surface of 130 square meters that our tissues are intimate, the blood, in contact with ambient air through the pulmonary air cells.

It is considerable physiological evidence: this respiratory function mechanism, our will can direct it. In fact, our will can modify its frequency: the number of respirations, exhalations and inhalations that are numbered at 12 to 16 each minute, can be increased or decreased, by accelerating or slowing them. Our will can modify the breathing rhythm by amplifying or shortening exhalation and inhalation. Finally, our will can suspend during a few moments the respiratory function by blocking its mechanism with the dorsal, abdominal or diaphragm muscles.

While our will cannot influence our digestive system, whether it be the peristaltic movements of the intestines or the secretions of the liver, our will cannot intervene in our gland functions, that it remains powerless in changing our heart and arteries beats, our will can, however, have a precise action on our breathing.

A regrettable observation: this function being life itself. The only function over which our will can act is the function that is the least known and the most neglected.

But how can we neglect a function that, we have determined, has a primary role in life? The 21st century man does not know how to breathe fully, cannot breathe fully and practices a lazy and partial breathing: the thoracic breathing.

Respiratory illnesses keep increasing and the weakness in respiratory muscles increases this degeneration.

Deep muscles, if not developed with precise respiratory work, will not allow deep breathing, the physiological breathing that gives exhalation the most important role in the act of breathing.

By practicing this breathing mode, persons focus all their attention on emptying pulmonary air during exhaling without worrying about inhaling which occurs automatically and in a direct link with the scale of emptying provoked by exhaling.

PARTIAL THORACIC BREATHING

In thoracic breathing, the attention is on inhaling in the goal of storing the most air possible in the respiratory system. Inhalation is active and systematically provokes the displacement of the thorax upward and the lifting of the shoulders.

However, exhaling is always neglected and only partially empties, passively, with exchanges reduced to a maximum: there is therefore sub-oxygenation. This lack of oxygen is at the basis of physiological and psychological perturbations. Any person with this mechanism experiences shortness of breath.

This thoracic breathing mechanism solicits only a small part of the muscles, neglecting the diaphragm, abdominal muscles and dorsal muscles.

This respiratory mechanism is practiced in different activities is the mechanism we consider "normal". The deep muscles are less solicited due to a bad use of the respiratory function and an intense cerebration.

While we insist on air intake before each exhalation, it is difficult, even impossible, for the subjects with fewer muscles, to find and maintain tone in all their dorsal and abdominal muscular system. All the respiratory exercises should have the objective of toning, of setting muscles in their real anatomical place.

We can consider thoracic breathing as a sort of organic deficiency, of respiratory handicap. Disciplined respiratory reeducation can correct and bring a person to practice the only total breathing, the one we had at birth, the deep physiological breathing.

DEEP PHYSIOLOGICAL BREATHING

Deep physiological breathing gives exhaling a more important role in breathing. The subjects that practice this breathing focus all their attention on emptying the pulmonary air during exhaling without worrying about inhaling which occurs automatically and in a direct link with the scale of emptying provoked by exhaling.

The exhalation is active to empty at the maximum pulmonary air during the effort; the inhaling is done with no mobility in the auxiliary respiratory muscles and with the diaphragm being sort of passive: the exchanges are increased to the maximum.

Deep respiration muscles are a real brace for the bowels. During physiological breathing, the body's most important muscles participate in the action of breathing.

There is no life without breathing

This breathing mechanism is not inaccessible. In some daily moments, the body finds this respiration: laughs, cries, screams, coughs. Nature sets muscles to their real anatomical place. Some sports, some occupations keep this mechanism by the precision and tone that requires deep breathing.

Exhalation is the vital phase for the resistance of the respiratory function: *"he who breathes will live long, intensely and peacefully"*.[2]

> "The majority of human beings only half breathe: we inhale because that part of the action of breathing is the condition for living. But we do not take care of exhaling because of muscle laziness notwithstanding that we better repair our strengths by exhaling with measure than by filling our lungs. […] With training, we are fortunately capable of making our breathing as good as it was unconsciously bad before."[3]

The voice and breath are extremely linked physiologically. Voice is the sound reflect of an exhalation. Without breath, voice does not exist. The human voice remains the most precious way of communicating between people. Human voice and breath have been, throughout the centuries, an important revelation of human condition. Man is shaped by his environment. In the Aesop story, Xanthos orders Aesop to buy what is best at the market. Aesop buys tongues saying there is nothing better than tongues, link of civil life, key to science, organ of truth, reason and prayer.

How can we improve the respiratory function of persons with cystic fibrosis?

We saw that, when the respiratory work is foremost inhalation while neglecting exhalation, the diaphragm and all other auxiliary, dorsal, transverse, oblique abdominal muscles cannot deeply accomplish their work. The tone and exchanges in pulmonary air cells remains weakened. All the importance given to sound exhalation with the oral position of laughter with the help of different movements, leads to a respiratory gymnastic where the entire body breathes and reinforces itself. This exhalation is not only a simple breath, but is an expiratory noise where deep muscles are more solicited and moreover trained for better tone. All persons with cystic fibrosis, with noisy exhalation, benefit from large ventilation, freeing up mucositis. It is a real autoclapping: expectorations are without violence and with relief. We give an important role to nasal passages to enable better respiratory health.

Some *important* advice:

- All respiratory work must begin with exhalation: do not force your breath. Keep your mouth open in the laughter position and breathe before doing an activity. When you are out of breath, exhale quietly. Your rhythm will bo back to normal.
- Take care of keeping good dorsal and abdominal tone. Deep breathing progressively adjusts muscles avec interventions to regain endurance and strength. It is good to exercise, but to avoid fatigue during training. We often do respiratory exercises on the ground. It is important to fell the spine on the ground, which enables reinforcing the pulmonary system.
- It is paramount to train your breath. The control of the respiratory function based on the deep physiological breathing mechanism with spontaneous exhalation guarantees a good control of respiration.
- Respiratory reeducation is a great necessity for pulmonary transplant recipients. After the surgery, when they have to breathe without a machine, there is often a panic. All the importance given to controlling exhalation, without rushing the respiratory muscles, allows them to avoid panic and make contact with their respiratory function at the same time. Calm and relaxation are an extreme necessity for them.

All persons, one day or another, are confronted to challenges. Illness, unfortunately, is part of that. A good muscle tone constitutes a safety engine for deep physiological breathing. Reeducating your breath, no matter what your physical condition, is a step towards regaining better health. In this respiratory reeducation is an important aspect, ethics. The Jules-Vernes de Nantes clinic team, made up of Alan Boone, teaching the Metge-Sandra method, and Drs. Philippe David and Gérard Dabouis, have brought together different specializations, a work of great importance for the health of patients and caregivers. ◄

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1. Element in cell cytoplasm with the essential role of ensuring oxidation, cell respiration, the start of energy stocking by the cell and of other substances (Le Larousse)
2. It refers to contractions that occur in the tubular organs and cause movement of the content in the organ)
3. (TaisenDeshimaru, La pratique du zen).
4. (William Knowles, The Knowles Method of Breath Training)

Six myths
related to urinary incontinence
in persons with cystic fibrosis

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Urinary incontinence affects only women.

FALSE. Despite the fact that we report more incidences in women, men can also suffer from urinary incontinence. On cystic fibrosis, many studies have shown that between 30 and 68% of adult women suffer from it versus 2 to 16% of men. In the population at large, this ratio is between 25 to 45% of women and 4 to 32% of men. Moreover, we observe that it can happen frequently to children. In fact, 8 to 47% of girls who participated in the study had symptoms of urinary incontinence as opposed to 2 to 14% of boys. In the population at large, this ratio is 3 to 9% of children, regardless of gender.

Constipation leads to urine losses.

TRUE. There exist other urinary incontinence risk factors besides cough. A constipated person must increase the force used to stool. This force leads to a stretch in the pelvic floor and, with time, the muscles loosen, which prevent it from working normally.

Also, the chronic inflammation experienced by patients with cystic fibrosis can have consequences on muscle contraction, and therefore on the pelvic floor.

Some other risk factors have been studied and explained, such as quantity and quality of muscles as well as autonomous nervous system control.

Persons with CF have a smaller bladder, pressures becomes greater more quickly, which leads to urinary incontinence.

FALSE. Bladder size is different from one individual to another and there is no link with the illness.

There are three types of urinary incontinence: stress incontinence, emergency incontinence and a combination of both. Stress incontinence translates into an involuntary loss of urine during sneezing, coughing or a physical effort. Emergency incontinence is explained by the sudden need to urinate followed by loss of urine. With cystic fibrosis patients such as you may have guessed, it is mostly stress incontinence.

The normal urination cycle fills the bladder, which generates the first urination sensations followed by the evacuation of urine. One part of the important physiological components to consider is the bladder and pelvic cavity. The bladder has a muscle with is named the detrusor muscle. This muscle enables the bladder to contract when it is full allowing the evacuation of urine. The urethra is the passage that allows the exit of urine. It leaves the bladder and becomes the urinary meatus. The urethra is support by many muscles that, together, are called the pelvic floor. When it is contracted, it prevents the evacuation of urine because it envelops and squeezes the urethra. The pelvic floor also protects the rectum as well as the uterus, in women. Urine losses can also be led by a problem with pelvic floor muscles.

The intervention of a physiotherapist is pertinent to prevent and treat urinary incontinence.

TRUE. There are physiotherapists specialized in urinary and fecal incontinence. They are trained to evaluate and treat problems with urinary and stool losses. There are many ways to treat incontinence, such as exercise and specialized interventions. The pelvic floor is a muscle that requires training and rehabilitation.

Urinary incontinence is still an overlooked subject that we do not discuss enough in cystic fibrosis. It is important to exchange with your treatment team if this problem occurs. There are links between urinary incontinence and illness. There are many alternatives to treat this problem and help the patients improve their quality of life.

Hoping that this article will enlighten you and help you. Do not hesitate to communicate with your contact person.

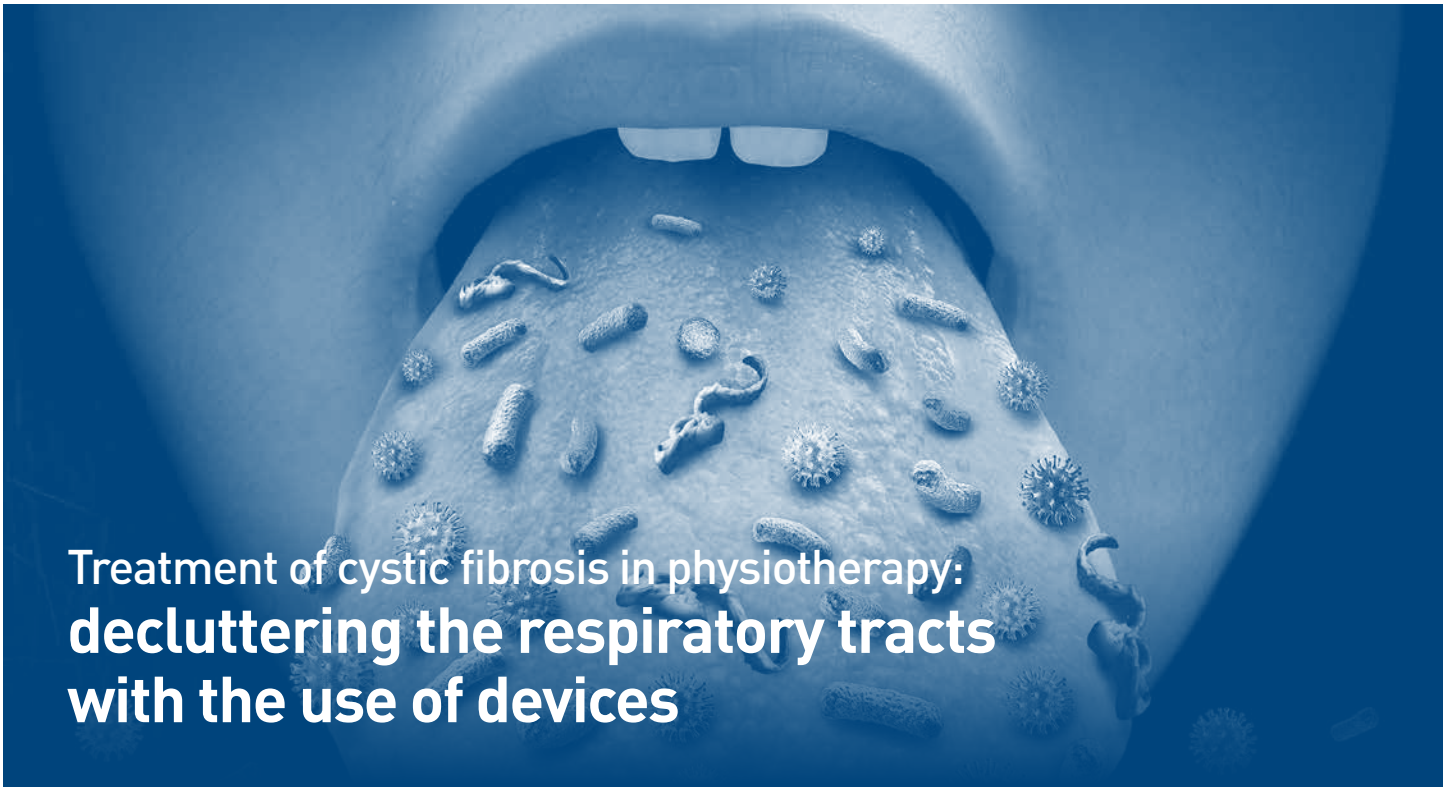
There are important consequences on the quality of life of patients suffering from urinary incontinence.

TRUE. To avoid urinary losses, a person can restrain from coughing, do respiratory physiotherapy and limit physical exercise. It is evident that these behaviors can have consequences on pulmonary health such as increasing secretions and elevate the risk of pulmonary superinfection. Also, patients can have a tendency to isolate themselves, go out less, especially where there are no bathrooms nearby, which can harm their social life. Some patients shared that they had to wear pantiliners so as not to get wet. Furthermore, some patients drink less, which is not recommended with cystic fibrosis. It is important to hydrate for many reasons, such as to compensate for the loss of salt through skin pores, which leads to dehydration, and to prevent constipation.

Cough causes urinary incontinence.

TRUE. Cough is a primary factor of risk of urinary incontinence with cystic fibrosis. The pelvic floor, mentioned previously, also supports abdominal (liver, pancreas, stomach, intestines) and pelvic (uterus, bladder, rectum) cavities organs. When a person coughs, an array of muscles contract, including some abdominal and back muscles. Contraction of these muscles during coughing leads to an increase in pressure on the pelvic floor, which consequently stretches it and, over time, provokes a loss in muscle strength. Thus, the pelvic floor did not fill one of its functions, which is to retain urine by squeezing the urethra.

We emitted a hypothesis that could explain urinary incontinence related to cough otherwise in patients with cystic fibrosis. Abdominal muscles become so strong with coughing that it leads to an imbalance in abdominal and pelvic cavities pressure. Because of the imbalance, the pelvic floor would be unable to normally function and have difficulty contracting. Therefore, it can have urine loss as a consequence.



Treatment of cystic fibrosis in physiotherapy: decluttering the respiratory tracts with the use of devices

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For many years, new methods of decluttering secretions have arrived to facilitate the daily lives of individuals suffering from cystic fibrosis (CF). These new methods include many devices, which are easy and fast to use as well as provide user autonomy. Among the principles at the basis of these devices, we find Positive Expiratory Pressure (PEP), Oscillating Positive Expiratory Pressure (OPEP) as well as High Frequency Chest Wall Oscillation (HFCWO).

Positive Expiratory Pressure

PEP is a therapy used by individuals with CF to free secretions from the respiratory tract. As the majority of PEP devices are easy to use, they can be used from the age of three or four years old [1, 2]. Normally, this method does not require the presence of another person, but children at a lower age often require supervision. PEP is a relatively low cost apparatus. In fact, a PEP device costs about 25$ to 60$ [3, 4]. Many models are available, for example: the PEP mask, the PariPEP®, the TheraPEP® and the Threshold PEP®. All these models rest on the same basic principle, but each have their own components and characteristics. Among these devices, we generally distinguish two categories : low pressure and high pressure devices.

The low pressure PEP can be executed in the acute phase of the illness [5]. However, since the PEP creates a resistance when exhaling, its use is not recommended for subjects with severe dyspnea [8] and is not recommended in presence of an undrained pneumothorax [7]. As the high pressure PEP therapy creates greater resistance when exhaling, it could be difficult to execute or badly tolerated in acute phases of the illness [8]. Furthermore, a high pressure PEP is not recommended in cases of cardiac trouble, hemoptysis, asthma, pneumothorax, or following pulmonary surgery [7].

Exhaling through a PEP device creates a resistance when exhaling that generates positive pressure in the respiratory tracts. This allows increasing the intrapulmonary and avoiding dynamic compression of the airways [3, 13]. PEP reduces hyperinflation and improves the airflow at the end of exhalation [3]. Furthermore, the increase in intrapulmonary pressure allows the air to infiltrate between the respiratory tract walls and the mucus to enable its displacement and elimination.

Oscillating Positive Expiratory Pressure

OPEP is one of the therapies that allows the decluttering of respiratory tracts in the pulmonary system. Like all PEP devices, OPEP devices are easy to use and can be used by individuals older than three years old [9]. Normally, this method is used autonomously, but children often require supervision. Furthermore, OPEP devices are portable and low cost with prices between 35 $ and 100 $. Many models are available. We can find, among others, the Flutter®, the Acapella®, the Cornet® as well as the Quake®. Although all these devices use the principle of OPEP, they each have their own particularity. OPEP can be badly tolerated during the acute phases of the illness [10]. Moreover, if the patient does not master the technique well, he can feel dizziness caused by hyperventilation [11]. As all OPEP devices increase intrapulmonary pressure, they can contribute to the development or worsening of a pneumothorax [12]. This is why it is not recommended for patients that have one, but also with patients with hemoptysis or cardiac insufficiency. [13, 14]

The Flutter®, the most known OPEP device, is a portable pipe-shaped made of an mouthpiece and a perforated. Inside the device, we find a stainless steel ball on a cone-shaped plastic receptacle [15]. Vibrations are

produced by the ball, which, because of the exhaled air and gravity, oscillates vertically[13]. When the ball comes down, it is deposited on the cone and blocks the passage of air and when it goes back up, it creates an opening for air to pass. These vibrations provoke fluctuations in resistance to the expiratory flow, which induces PEP and vibrations in the respiratory tracts. To optimize the latter, the inclination of the device must be adjusted to get a frequency similar to pulmonary resonance. However, as the mechanism for Flutter® depends on gravity, it can only be very slightly inclined. By inclining it lightly to the bottom, we get a lower frequency of oscillation or a greater frequency if inclined to the top.

The Acapella® device is a portable device that uses a magnet and a counterweighted valve to produce OPEP. When the patient breathes through the device, the valve induces OPEP by blocking the passage of air intermittently[16]. At the end of the device, a button adjusts the distance between the magnet and the valve to regulate frequency, amplitude and pressure[16]. As opposed to Flutter®, the Acapella® device is not influenced by gravity. It can therefore be used in many positions.[17, 18]

The Cornet® is a device made of a semi-circular plastic tube with a flexible hose. One of the ends of the tube is made of a mouthpiece while at the other end, a regulator wheel adjusts flow, pressure and frequency of the oscillations. When using Cornet®, the OPEP is produced by movements of the flexible hose. During exhalation, fills with air and bends in certain places because of the constraint of the semi-circular plastic tube.

The Quake® is a device made of a mouthpiece and a crank. During exhalation, the user can manually generate the frequency of oscillation by turning the crank at the desired speed. A slow rotation of the crank induces low frequency oscillations while a fast rotation induces high frequency oscillations. Therefore, the Quake® is recommended for patients who are importantly affected and are incapable of generating an expiratory flow sufficient enough for the previous devices[19].

High Frequency Chest Wall Oscillation

The High Frequency Chest Wall Oscillation (HFCWO), which is also called High Frequency Chest Wall Vibration or, more commonly, the vest. As with the two previous methods, the HFCWO does not require active participation. Therefore, it can be used from two to three years old. The HFCWO is an equipment costing thousands of dollars and can be used autonomously. Created by Hansen and Warwick in 1990, many models are now available on the market: The Vest®, the Smartvest® and InCourage®.

HFCWO therapy uses an inflatable vest and a compressed air generator. The latter sends air, intermittently, in the vest to inflate and deflate it. Therefore, during inhalation and exhalation, high frequency mechanical oscillations are transmitted to the chest walls. These oscillations provoke, as with OPEP devices, the vibration of respiratory tracts that remove the mucus and displace it to the higher airways. Furthermore, the chest compression produced by the vest filled with air helps improve the small pulmonary volume respiratory flow and increase the efficiency of mucociliary transport by inducing force similar to cough. A nebulizer can be annexed to the device to administer the patient a hypertonic solution during treatment.

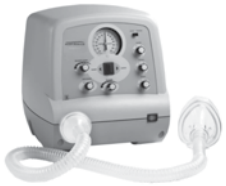
Other devices

There are multiple devices that can help the decluttering of respiratory tracts and improve the pulmonary function. Unfortunately, some of them are less known, less used or less studied. Novelty, specificity for a precise clientele, proven efficiency with other clienteles or choice of methods that are already impressive or difficult are some factors explaining the lack of conclusive data on the use of some devices with CF patients.

The Frequencer® is a device possessing a transducer that emits, with frequencies varying from 25 to 40 Hz, mechanical sinusoidal waves as well as acoustic vibrations. These mechanical and acoustic stimulations lead to vibrations of the respiratory tracts[20], which removes mucus from the walls and displace it to the higher airways to be eliminated.

The CoughAssist® is a device facilitating the removal of secretions from respiratory tracts in patients with inefficient cough. The device clears secretions from the lungs by gradually applying positive air pressure to the airway and then rapidly shifting to negative air pressure. The resulting high expiratory flow simulates a deep, natural cough.[21]

PEP Mask



CoughAssist®

Flutter®

PariPEP®

TheraPEP®

Frequencer®

Efficiency of respiratory tract decluttering devices

Many studies have been made with the goal of determining if PEP, OPEP and HFCWO are decluttering methods more efficient and/or superior to other methods. The variables analyzed to evaluate efficiency are numerous, but the one most often used is the pulmonary function. The latter can be evaluated by forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1) and the average expiratory flow (AEF). Other variables frequently used are the quantity of secretions, the duration or number of hospitalizations, the hemoglobin oxygen saturation (SpO_2) and the undesirable effects.

In the majority of studies, the results for FEV1, AEF and FVC haven't allowed to observe a significant difference in efficiency between the methods of decluttering respiratory tracts.

The quantity of secretions expectorated following the use of decluttering methods is a variable often used to evaluate the efficiency of treatment. However, it is important to keep in mind that the reliability of this measure is debatable. In fact, the quantity of secretions can greatly vary because they can contain a variable quantity of saliva and can be swallowed. By examining the secretions expectorated, only one study was able to put into evidence a variation between two methods of decluttering respiratory tracts. In fact, App et al. noted a significant reduction in the viscoelasticity of secretions following treatment with Flutter® compared to autogenic drainage. [22]

The number of hospital admissions following pulmonary deterioration can also vary according to the chosen method of secretion removal. In the study by McIllwaine et al., there is no significant difference between the group using postural drainage and percussion and the one using a PEP device [23]. However, another study by McIllwaine et al. demonstrated, by comparing PEP with a Flutter® over a period of a year, a significant decrease in the number of hospital admissions caused by pulmonary deterioration. In fact, in the PEP group, five participants had to be hospitalized compared to 18 in the Flutter® group [24].

Conclusion

The methods of secretion removal with devices are easy to use, fast, portable and allow more autonomy than the conventional methods. The three main types of devices presented - the PEP, the OPEP and the HFCWO - seem to be safe. However, no clear conclusions can be emitted as to the efficiency and superiority of a device compared to another treatment. By considering that the majority of studies presented are crossover studies evaluating short-term effects, it would be pertinent to dispose of randomized long-term clinical studies to give more credibility to the results. Furthermore, Flutter® is the best known device, it is the most used in studies, but very little data is available on the other devices in the same class such as the Acapella®, the Cornet® and the Quake®. The novelty of these devices could explain the lack or absence of literature, [25] but it would be interesting that these devices be studied as well. ◄

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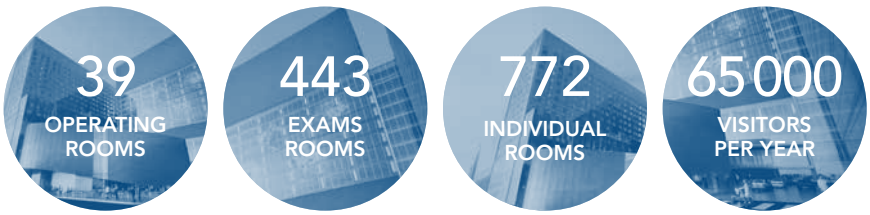
The new CHUM
innovations serving the patients

The original texts were published in volumes 1 and 2 (6th edition) of CHUMagazine.

Soon, the new CHUM will welcome its first patients. At the corner of Viger and Saint-Denis streets, a few steps from the CHUM Research Centre inaugurated in 2013, the hospitalization towers are at their full height since the end of April 2015. From three hospitals many centuries old, we will move onto a 21st century hospital: it goes well beyond construction and a mega move.

With the arrival of the first patients at the new CHUM, Notre-Dame Hospital will become a community hospital where proximity care, first and second line care, will be offered to the population. Saint-Luc Hospital will be demolished to make space for phase 3 of the new hospital, which will include the amphitheatre and the clinical-administrative wing. Hôtel-Dieu will close its doors and will be given to the Ministry of Health and Social Services, which will decide on its use.



CENTRE HOSPITALIER DE L'UNIVERSITÉ DE MONTRÉAL (CHUM) HOSPITAL

The CHUM will offer specialized and superspecialized (tertiary and quaternary) care to patients from the greater Montreal region and all of Quebec. If you are victim of a severe burn requiring intensive care, it is at CHUM that you will be treated, even if you live in Outaouais, for example. Furthermore, the pulmonary transplant and cystic fibrosis clinics at Hôtel-Dieu Hospital will be transferred here. Already, CHUM has a recognized expertise in cutting edge fields, including in chronic pain management and in transplants (lungs, heart, kidneys, pancreas, liver). There are also research, educational, technological evaluation and healthcare intervention evaluation functions.

In the new hospital, there will be 772 individual rooms including a family space, 39 operating rooms and 443 exam rooms. The emergency can welcome 65 000 visitors per year.

For patients, it will be a location enabling recovery and wellbeing. Individual rooms, to prevent the transmission of infections and offer more privacy for severely ill patients, are an important innovation. There will be a separation between the hospital and ambulatory (day clinic) zones to improve quietness for hospitalized patients. And good news, direct access, from inside, to the Champ-de-Mars metro station, will allow users to avoid outside temperatures!

100% new air at all times

With unique air treatment plants, CHUM is the only hospital in North America to offer patients, visitors, staff and caregivers a 100% new air environment. What does this mean? Once treated and distributed, he air is automatically rejected outside without being recycled. With time, 41 air treatment plants will provide outside air supply of about 80,000 square meters per minute. These air treatment plants will ensure continuous air renewal inside the hospital. An innovation that is an efficient tool against the spread of infections.

Hybrid operating room

The new CHUM will have 39 operating rooms, including three hybrid rooms, combining conventional operating rooms with an angiography imaging room (for imaging blood vessels). With an adjacent control room, these lead-lined rooms will be used for operations guided by angiography as well as for vascular, cardiac or neurological interventions. Teams will have access to these three hybrid rooms of more than 100 square meters designed specifically for these types of interventions.

Small chip, big revolution!

The new double case computer supply system will ensure the constant availability of sterile material and other medical furniture. When the first compartment becomes empty, the product tag will be put on the control panel. The radio-identification chip in the tag will trigger the supply process. For caregiver staff, time allocated to manual labor such as date entry or inventory will be finished. The supply computer system, already present in some units of the three hospitals, will enable the rapid resupply of furniture.

Double security for patients

Everything was implemented in the new hospital to ensure constant services to patients, including by doubling the main supply, security and power systems. In other words, a second system will take over, without system interruption, in case of incidents or a shortage. A safety system has been planned for medical gas supply, fire pumps, water supply, electricity and natural gas as well as air treatment and telecommunications infrastructures. In Québec, only the new CHUM will be equipped with such complete building services.

Cutting edge on call system

An all new on call system will be installed in each of the 772 individual rooms as well as in all clinical environments. Through a mural interface, the staff will be able to communicate directly with different extensions for professionals, to signify presence, to request help or even to send a code blue (cardiac arrest). An LED light installed on the exterior of the room will be linked to these mural interfaces and show a different color according to the command. From their bed or their stretcher, the patients will be able to signal their needs to the staff with a remote or an intercom. Different icons correspond to different commands: medication, water, bathroom or general request.

Guided vehicles and other logistic tricks

Two major innovations are at the center or the logistic reorganization of the new hospital: the auto guided vehicles (AGV) and the pneumatic transport system. Close to 70 AGVs will make more than 3,500 supply transports daily through precise paths. Moreover, delivery of more than 6,000 medications, samples, blood products and others will be made inside the hospital with a circuit of ducts totaling more than 9 kilometers. Auto guided vehicles will circulate in distinct hallways and elevators to transport supply carts, without ever blocking hospital hallways. The staff will deliver and receive these products from one of the 129 stations in the pneumatic transport system. An independent and clean circuit for isotope transport will have a direct connection to the cyclotron room at the CHUM Research Centre (CRCHUM) in an alcove situated in proximity to the nuclear medicine sector. The cyclotron is a device that is used for cancer treatment and imaging research. ◄

In remembrance of **our heroes**

Tomy-Richard Leboeuf McGregor
Executive Director
Living with Cystic Fibrosis

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Canada

For many people, the names of Marc Fortin, Michel Paquette, Marie-Hélène Roger and Marika Archambault are related to the world of cystic fibrosis through prizes and scholarships given to people in our community. Behind these names are hidden persons that are no longer with us, but that contributed significantly to advance, in their way, the cystic fibrosis field.

MARC FORTIN

Pay honour to whom honour is due. It is in 1985 that Marc Fortin became the first Chairperson of the Board of Directors of the **Adult Cystic Fibrosis committee of Quebec** (ACFCQ) that later became **Living with Cystic Fibrosis**. Then aged 24, it is with other adults and Laval de Launière, their social worker, who directed our organization. For the time, it was quite a feat: the median age of survival was less than 25 years and Quebec had only 150 persons with cystic fibrosis that were older than 18 years old. As founding president, his actions marked a turning point in the Quebec world of CF, which, until then, had stayed a closed club for the medical community and researchers. It is under his presidency that on September 19th, 1987, a certain Céline Dion visited the directors to show her support. Unfortunately, on December 24th that same year, Marc Fortin passed away at the age of 26.

It is the Quebec Association of Cystic Fibrosis (which later became a division of Cystic Fibrosis Canada) that put together the first Marc-Fortin prize. Awarded every two years, it was received among others by Laval de Launière, who became our director general, and Christian Auclair. In 2000, our organization took back the prize, which would now only go to persons living with cystic fibrosis for their exceptional contribution to the cause. The prize is awarded every three years. Since then, it was given to many people involved in our community. In order, Chantal Germain, Dave St-Pierre (dancer, choreographer and director), Pierre-Alexandre Tremblay, Claire Boulerice, Charlène Blais and Sophie Jacob have seen their names associated with our first president.

Also, the *Fondation l'air d'aller* gives out a scholarship with his name to persons involved socially in a community organization or in a private or public institution.

MICHEL PAQUETTE

It is in 1993 that Michel Paquette became the fifth president of ACFCQ. Being a long-time member of our organization, he was also for many years one of the first and rare members from outside of Montreal! A proud representative of Outaouais, he was a partisan of regions, wanting to avoid a centralization of powers in Montreal. Under his presidency, the organization started pressuring Quebec governments so that Pulmozyme be on the list of covered medications covered by the public medication insurance in Quebec, RAMQ.

The Michel-Paquette prize was created in 2000. It was decided that it would be awarded each year to the person or organization that works in the cystic fibrosis community and that contributed importantly to the wellbeing of the affected population. Many known personalities and groups had the honour of receiving it in the following years: Marcel Barette, France Paquet, Francine Brosseau, Diane Lalonde, Denis Mouton, Dr. Michel Ruel, the *Fondation l'air d'aller*, the Chambre des notaires du Québec (Quebec Chamber of Notaries), Dr. Alphonse Jeanneret, Dr. André Cantin, Mylène Cloutier, Laval de Launière, the journalist Harold Gagné and the *Ordre des infirmières du Québec* (Quebec Order of Nurses).

MARIE-HÉLÈNE ROGER

Nature lover, Marie-Hélène Roger completed her studies in biology, in ecology and in environment. That is why a scholarship in these fields bears her name. Not only was she involved in the PCACF as secretary for many years, but also her commitment extended to many committees and environmental organizations. She also proudly represented us as part of the former Canadian Committee on Cystic Fibrosis. Some even called her the ''little lady of great causes"!

Passionate about environmental causes, there was only her to speak with such enthusiasm about recycling, compost and birds: so much so that to save paper, our secretary wrote on both sides of pages from top to bottom and from left to right!

It is with determination that he defended the cause of those living with cystic fibrosis, despite her illness.

She touched the heart of many with her social abilities, her joy for life, her sincerity and her inspiring strength.

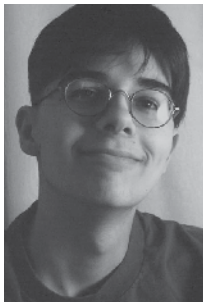
MARIKA ARCHAMBAULT-WALLENBURG

It is in memory of Marika Archambault-Wallenburg, exceptional young woman loved by all, that the Fondation l'air d'aller offers a scholarship to science students so that other persons can be inspired by her background and be encouraged by her many qualities.

Marika's great curiosity brought her to Europe, Guatemala, Nepal, Japan and the United States, alone on her motorcycle, but also brought her to dive head first in her many science books. Her academic path was flawless. The excellence of her academic results gave her many scholarships: admission scholarship to McGill, for her bachelor's in physics, Canadian scholarship for her master's in medical biophysics at the University of Toronto and one of the most prestigious Canadian scholarships for her doctoral studies in biomedical engineering at McGill. Cystic fibrosis never stopped her. Marika always followed her objectives, without complaining, for the simplest and most noble reason: the passion of learning!

The quality that best defines her is without doubt perseverance: for accomplishing what she had chosen to do, for pursuing studies as if there was nothing and, mostly, for facing each day with a smile, regardless of the challenges imposed by the deterioration of her health.

Marika's joy for life shined so much that she contributed to making others happier and better.



GAÉTAN VERREAULT

Passionate about computer and communications, it is in 2003 that Gaétan Verreault created one of the first Internet forums for persons living with Cystic fibrosis and those interested in their reality. It is in his honour that in the Fall of 2006, the forum he helped put together now bears his nickname, "Kifar".

For ten years, before the arrival of social media such as Facebook or Twitter, this forum would be the rallying point for many CF persons that found a way to escape isolation, to benefit from the support of others and, most certainly, to discover exceptional people. Throughout these years, more than 350 persons participated, with more than 6,300 messages to many discussions. His determination and his tenacity contributed to bringing the Quebec CF community together. ◂

CYSTIC FIBROSIS RESEARCH IN 2016

Dr. André Cantin
Pneumologist
Full Professor

Pneumology
Sherbrooke University
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Sherbrooke, Quebec
Canada

More than 4,000 physicians, nurses, researchers and other healthcare professionals met from October 27th to the 29th, 2016, in Orlando for the 30th annual *North-American Cystic Fibrosis Conference* (NACFC). It is the largest conference on research and care for persons with cystic fibrosis. This conference allowed participants to mark the thirtieth anniversary of the modern era in cystic fibrosis research. Thirty years ago, in 1986, the gene responsible for cystic fibrosis was not known and the number of adult patients followed at the Sherbrooke cystic fibrosis clinic was only 6 while there were close to 40 children. Thirty years later, not only is the gene responsible for cystic fibrosis identified, we have started prescribing medication with the therapeutic goal is to correct the fundamental default provoked by mutations of this default, the CFTR (Cystic Fibrosis transmembrane conductance regulator) protein. Furthermore, we now have more than ten times more adults at out cystic fibrosis clinic while the number of children has remained the same. The identification of the gene responsible for cystic fibrosis, the development of medication correcting the illness' fundamental default and the astonishing increase in adults followed by our clinics represent spectacular advances in research as well as in treatment for cystic fibrosis patients.

A Canadian-American team whose director was a young professor at the University of Toronto supported by grants from Cystic Fibrosis Canada, Dr. Lap-Chee Tsui, identified the gene responsible for cystic fibrosis. Dr. Tsui was one of the first researchers to use this innovative and modern technology, which has become the standard in medical genetics, the identification of a gene by a reverse genetics approach. Before, discoveries in genes associated to hereditary diseases required the knowledge of the defective protein prior to identifying the genetic code responsible for the defect. Dr. Tsui innovated by using an approach in which he did not need to know the defective protein to identify the gene. This approach was based on epidemiologic studies with populations of families and individuals with cystic fibrosis. Many families in Québec participated in this research work in the Eighties by giving personal medical information and by providing blood samples for DNA analysis. This extraordinary implication by patients and families living with cystic fibrosis is an amazing example of the benefits that a participation in clinical research can bring to an entire population. Dr. Tsui used the information obtained to identify genetics traits that were simultaneously transmitted to children with the illness. Some of these genetic traits had already been mapped and the location of each genetic characteristic in the human genome allowed Dr. Tsui's team to get closer to where the defective gene was. In 1989, these researchers realized they had identified the exact site of the expression of the defective gene and were able to clone that gene, produce the protein and start a new sequence of fundamental research targeted on knowledge offered by this discovery. Among the discoveries, we remember the development of many models of cystic fibrosis in animals, which did not exist before. There are also many new tools such as antibodies and specific cells to the CFTR default that were developed and made available to other researchers. Following a set of discoveries coming from the identification of the cystic fibrosis gene, new molecules enabling the correction of defaults were identified and clinical research helped market two new medications for part of the cystic fibrosis population. These new medications, produced by Vertex, are known as ivacaftor and a combination of ivacaftor (Kalydeco) and lumacaftor (Orkambi).

During the NACFC conference, much work including clinical studies were presented and it is noted that patients carrying class III mutations treated by ivacaftor not only have an important correction of their sweat tests, which is a correction of the fundamental defect, but also a significant and sustained improvement of their respiratory functions, their weight gain and their respiratory symptoms. Furthermore, it was reported that the inflammatory charge in the bronchial tubes of patients treated with ivacaftor decreased and that clinicians even observed a disappearance of bacterial micro-organisms, that were chronically observed previously in their respiratory secretions, in some patients. There is thus evidence that it is possible not only to improve symptoms and signs of cystic fibrosis, but also to reverse some that we thought were irreversible before. Unfortunately, ivacaftor is only useful in a minority of persons with cystic fibrosis, because this medication does not work for other classes of mutations.

The most common mutation in Canada for persons with cystic fibrosis is the F508d, a class II mutation. Mutations in class II make it that the CFTR protein is of abnormal structure and the misshaped protein deteriorates inside the cell before it can reach the membrane to execute its work of producing salt and bicarbonate in mucus. The absence of salt and bicarbonate in mucus creates thicker mucus and prevents antibacterial defenses on the surface of airways. During the

conference, many researchers demonstrated that the excessive acidification of mucus in cystic fibrosis could contribute to bacterial growth. There is much ongoing work to discover ways to counter excessive mucus acidification.

The most effective therapy to correct the sodium chloride and bicarbonate secretion defect in mucus is a therapy, which enables the type F508d defective CFTR molecule to reach the membrane and function. Lumacraftor is a new molecule that allows the class II CFTR protein to not deteriorate and reach the membrane. Once at the membrane, the F508d CFTR still has two defects to correct, the slowness to open and the tendency to disappear quickly from the membrane. The addition of ivacaftor to lumacraftor (Orkambi) lets the F508d CFTR not only reach the membrane, but also to stay open and function for a prolonged period. The clinical studies have shown that there is a decrease between 20 and 25% of the quantity of salt in sweat when homozygous (F508d/F508d) persons for this class II mutation are treated with ivacaftor/lumacaftor. New clinical data has confirmed the partial efficiency of this medication for this part of the population with cystic fibrosis. It is a partial correction but it is associated with a statistically significant improvement in respiratory function and also a decrease of about 40% in the number of infectious exacerbations. Unfortunately, this medication is not for all individuals carrying cystic fibrosis. This new medication does not function when there is only one copy of the F508d genetic defect. Thus, there are other current clinical studies with a new generation of similar molecules with a greater capacity to correct the fundamental defect observed in individuals carrying the F508d class II mutation. Finally, many other researchers have presented preliminary data on different molecular approaches also correcting the CFTR function in cell models and in animal models. This research work is still at the preclinical level, but should quickly be the subject of clinical studies in which individuals with cystic fibrosis are called to participate.

There is other research work as well with molecules having an impact on pulmonary and systemic inflammation and also on diabetes and liver diseases (cirrhosis). This different work will also require the participation of persons with cystic fibrosis.

In this context, it is very important to acknowledge possibilities in different clinical research studies. Information concerning clinical research programs is available at Cystic Fibrosis Quebec and Cystic Fibrosis Canada. Furthermore, cystic fibrosis clinic directors across Canada are well aware of ongoing studies and can share this information when requested.

We are living in very promising times for cystic fibrosis and we already see very important advancement in the survival and quality of life of some treated persons with or without medication to correct the CFTR defect. It is essential that we do more and we will arrive at our goal only by working together. It is paramount that researchers and clinicians continue their medical research and that charities (Cystic Fibrosis Canada for example) supports these researchers. It is more important than ever that persons with cystic fibrosis inform themselves and also participate in this common effort to fight cystic fibrosis. Together we will prevail! ◂

HEALTH COLUMN

VAPING AND E-CIGARETTE

I am the mother of a teenager living with Cystic Fibrosis. For a few months now, he began to vaporize the e-cigarette. What are the risks? If there are, are they less important than those known about conventional cigarettes? As a mother, I am worried. Could you tell me more about that?

Unlike regular cigarettes, e-cigarettes do not have tobacco. E-cigarettes are battery powered devices that have a heating element and a cartridge that contains liquid. Puffing on the device heats the liquid, which produces vapour. Compare this to regular cigarettes where puffing burns the tobacco and produces smoke—the big danger for the cigarette smoker and everyone around them—not to mention the tar and carbon monoxide that the smoker inhales.

The e-cigarette might seem harmless by comparison but taking a closer look at what's in the liquid raises other concerns. Like regular cigarettes, many e-liquids contain nicotine, even though nicotine for e-cigarettes is not officially approved in Canada. The liquids often contain other ingredients too, such as propylene glycol (PG), a popular food additive. They also come in hundreds of flavours such as strawberry, root beer and chai tea, which make them very tempting to children and teenagers.

The production and sale of e-liquids is not yet closely monitored in Canada, which means they may not always contain the ingredients and proportions listed on the label. What's more, the e-cigarette industry is still so young that there's no data on the long-term effects of inhaling e-liquids.

A key similarity between the two types of smoking that can't be ignored is that e-cigarettes mimic regular smoking. Public health officials are now speaking out about the dangers of making smoking acceptable again, a trend that could potentially roll back decades of work achieved by anti-smoking campaigns. E-cigarettes should never be viewed as a better way to start smoking. Pediatric specialists all agree that whether it's e-cigarettes or regular cigarettes, children, teens and adults should never take up smoking under any circumstance.

— Les spécialistes Info-Santé.
Info-Santé Specialists – Montréal Children's Hospital (MUHC)

HEMOPTYSIS

Lately, my sister had a hemoptysis that her physician qualified as minor. Nevertheless, this situation has enormously anguished our family. Can you explain to us what are hemoptysis and how should we react in such a situation?

We talk about hemoptysis when there is expectoration by the mouth of blood from the lungs. In the lungs of a person with cystic fibrosis, chronic infection and inflammation change pulmonary blood vessels by eroding their walls and dilating bronchial arteries; leaving them more brittle and vulnerable to bleeding. It is a relatively common complication and this situation can, of course, cause a lot of stress in the individuals and their relatives.

If you experience hemoptysis, it is important to note its quantity to inform the medical team who can determine the emergency of the situation. The treatment will vary according to the severity of the situation. It can be simple monitoring (with or without antibiotics), an embolization (obstruction of blood vessels causing the bleeding) or in rare cases, a surgery.

There are three types of hemoptysis: minor, major and massive.

- **Minor hemoptysis** (<5ml): The quantity of blood can vary during the day. It can be small traces of blood in secretions or about a teaspoon of liquid blood. It is the most frequent one. Recommendation: Communicating with your cystic fibrosis clinic. If the expectorated blood is bright red and/or that it persists and/or that it is accompanied by an increase in your respiratory symptoms, it is preferable to consult your physician quickly. It is possible that, especially with the first occurrence, that your physician decides to examine you. Even if most cases of minor hemoptysis do not require treatment, it is recommended that you inform your CF team. An antibiotherapy can be required if there is an underlying pulmonary infection.

- **Major hemoptysis** (<240ml): Can be defined as an expectoration of liquid blood of a few teaspoons of blood, over a 24-hour period.

- **Massive hemoptysis** (>240ml): This severe situation requires immediate care. The quantity of blood expectorated can easily reach 1-2 cups or more over a 24-hour period. It can happen quickly and without prior symptoms.

Recommendation: If a major or massive hemoptysis occurs, you have to quickly get to a hospital and notice your CF team. In all cases, an antibiotherapy will start and you will remain under medical observation. In cases of major hemoptysis, it is recommended to do an embolization or the bronchial arteries that are responsible for the bleeding. Your physician will discuss with you the best approach to follow according to your condition.

Persons that experience hemoptysis risk having a new episode. Although no measures exist to prevent massive hemoptysis, following your medical treatments to prevent infections could reduce the risks.

As a relative, it is normal that major or massive hemoptysis makes you worried. In such an emergency situation, it is essential that you act quickly and keep your calm. It is very likely that a person in this situation will need your help. Contact emergency services by dialing 911 instead of transporting the person to the hospital yourself. The medical services have the means to adequately take in charge while ensuring safe and quick transport to the nearest hospital.

As stated by psychologist Helen Oxley in *SVB 2010*, "whatever the person's situation, if hemoptysis is a permanent stress or was a traumatic event, it is completely normal that it come with emotional reactions: our body and spirit are designed to react that way." If your relative has difficulty managing stress following an important episode of hemoptysis, do not hesitate to contact a health professional.

— Joanie Bernier, Inf., B.Sc
Cystic Fibrosis Clinic at the McGill University Hospital Center (MUHC), Montréal, Quebec

CROSS-CONTAMINATION

I hear people talking about cross-contamination between CF patients a lot. Is it true that CF patients are not allowed to meet in the same place?

Crossed-contamination is the transmission of microbes or infections from one person to another. It's been known in the CF world for a long time. In March 2000, researchers in Denmark published a study on passed observations: in 1990, 22 children CF patients participated in a winter camp lasting 1 week. 17 of them had Pseudomonas geruginosa at the beginning of the camp. At the end, everyone was infected with the same strain.

To prevent crossed-contamination between CF patients, many organisations have decided to promote forbidding any and all contact between them. This isolation allowed limiting the spreading of certain infections. However, according to us, forced isolation is not the most effective method to fight crossed-contamination. We prefer education. As a grass-roots organisation, we know that, even with rules forbidding contact, CF patients meet each other regularly, especially outside the hospital. Strong ties are built by people who have similar social and medical backgrounds. We also know it's important for CF patients to live an active and productive life in an inclusive society that respects their rights. This is why we developed a hygiene policy on crossed-contamination and prevention of infections.

This policy clearly states the behaviour one should have while with other CF patients, whether it be during friendly gatherings or public events like fundraisers. Being particularly vulnerable to bacterial infections or to any viral infection or other source of contamination, it's essential that you respect the protocol to prevent infections.

While CF patients have to maintain at least a 1-meter distance between one another, avoid handshakes or close contacts. Wash your hands often, especially when you touch an object that could have been manipulated by others. You should also wear a mask when you feel symptoms of an infection to protect yourself and others or simply to feel more protected. This behaviour should be the same with anyone who has influenza or even the common cold.

There are certain advantages and disadvantages to both approaches. The different studies preach prudence, but nothing is totally black or white. Crossed-contamination is a complex issue. It's a medical issue, but also a personal philosophy issue. Luckily, in our day and age, it's possible to break isolation thanks to social media. However, we remain convinced that education is far better than forbidding. Well-informed people who meet each other will be more prudent than those who are not well informed on the best protection methods.

We invite you to visit our website in order to read the whole policy and take the best measures to protect yourself.

Thanks to you, we can make the lives of those living with Cystic Fibrosis in Québec better

Thank you for your generosity!

Nathalie Larose
Karine St-Jean
Jean-Pierre Masson
Line Boutin
Louise Portal
Alain Ferland
Entreprise Magotteaux
Denis Dubé
Jennifer Barns Francoeur
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Mathieu Cordeau
Rachel Clermont Tourangeau

LIVING WITH CYSTIC FIBROSIS

vivreaveclafibrosekystique.com

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.