Primary Biliary Cholangitis

PBC, more than a chronic disease

12 patient stories

Dutch
Liver Patients
Association

Colofon

This is a publication of the Dutch Liver Patient Association .

These are personal stories of our members, written from their experiences. An illness can affect everyone differently. If you have any medical questions, please contact your doctor.
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Editor
José Willemse
Members of the Dutch Liver Patients Association

We wish to thank the members of the NLV for their wonderful contributions.
The NLV has received written permission for publication in the brochure.

Layout
The Happy Horseman, Rotterdam

Address
Dutch Liver Patients Association
Van Boetzelaerlaan 24J
3528 NS Hoogland
085 27 34 988

info@leverpatientenvereniging.nl
www.leverpatientenvereniging.nl
Leverpatiënten

Bank: NL44 INGB 0000361038

This brochure has been made possible by a financial contribution from AOP Orphan Pharmaceuticals GmbH, Member of the AOP Health Group Vienna, Austria

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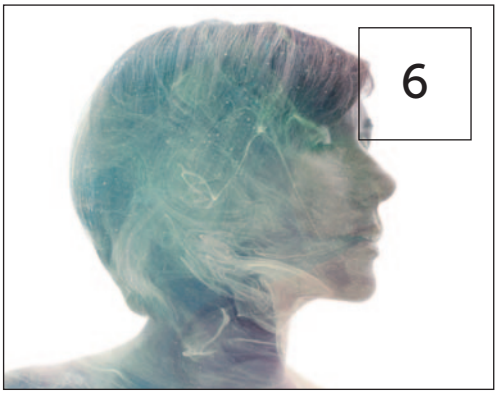
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The following stories are the personal narratives of our members, written based on their experiences with PBC. The course of any disease can differ from person to person. Contact your physician if you have any medical questions.

Foreword

Primary Biliary Cholangitis or PBC is a chronic liver disease that affects the small bile ducts of the liver. This autoimmune disease is more common in women than in men. The condition is extremely rare in children.

Before finally being diagnosed with PBC, patients often go through an extended period of experiencing vague symptoms that have an enormous impact on their daily lives. Symptoms including extreme fatigue and itching are commonly associated with this condition, which belongs to the group of cholestatic liver diseases.

A treatment is available for PBC to prevent the condition of the liver from deteriorating further. However, not all patients respond equally well to this treatment. Although various new treatments are already available or currently being developed, these treatments are not yet permitted in the Netherlands or covered by Dutch health insurance providers, or they are only available in a research context.

PBC has an enormous impact on a patient's daily life
Here, you can read the personal stories of ten people who have been diagnosed with PBC. These ten impressive stories underscore the urgent need to conduct further scientific research, improve the availability of medication and – especially –raise the general public's awareness of the liver and liver disease.

The Dutch Liver Patients Association (NLV) tirelessly strives towards these goals.

José Willemse,
executive director

@Dutch Liver Patients Association
2022

PBC has become part of my body and my soul

Marjolein (54)

At first, I was incredibly relieved to learn that my itching was not caused by an allergy, that I was not imagining the fatigue I felt and that the brain fog I so often experienced was not caused by the tumor I was convinced I had in my head; they were all symptoms of Primary Biliary Cholangitis (PBC). Gradually, I began to realize that this was a serious chronic condition that would have a major impact on my daily life. It was hard to listen to my doctor as they told me that the disease "would never go away" and that the purpose of my treatment was to stabilize the PBC to keep the condition of my liver from deteriorating further. I cried for days about this illusive disease known as PBC.

It was a liver disease, of all things. At the time, I associated the word with alcohol abuse. Was this the result of drinking too many beers and glasses of cheap wine in my youth? Was it caused by the liquor that I grew to like once we had some disposable income? I found it hard to shake that thought and began to wonder how I would ever tell the people around me. Surely, they would say they never suspected I was an alcoholic.

I read a little bit and couldn't go on...

Luckily, my husband was at my side when I received my diagnosis. He was better able to understand what the doctor was saying and knew that this was caused by a disruption of my autoimmune system. I must have been miles away in my head at the time.

After my husband had patiently repeated for the umpteenth time what the nice doctor had told us, he grabbed his laptop, typed some words into Google and told me: "Read this!"

It was the website of the NLV, which clearly explained what PBC was and how other people had found ways to live with this condition. I read a little bit and couldn't go on. Surely, this didn't apply to me. My husband said: "If you don't want to join, I will." He didn't have to tell me twice.

We joined the NLV and learned that they were organizing a members' convention to commemorate their thirtieth anniversary. I still had some doubts about attending, but my husband had already signed us up. He said: "If we don't like it, we can leave." That's how we left home that day: would I find someone else just like me, would I receive more information, would they understand my concerns? To sum up a very long, very exhausting day in a few short words: I couldn't stop talking about it in the days that followed. I began to read up on PBC to learn more about what I could do to stay as healthy as possible and how best to cope with the bad days that I was experiencing now and then.

It is part of who I am

My liver is not exactly in optimal condition. After returning from her maternity leave, my sweet doctor asked me if I had been able to give the PBC a place in my life.

I told her that I could not say that I have adapted to having PBC in my life. Instead, I would say that I have learned to live with this condition as a part of me.

PBC has become part of my body and my soul. It is part of who I am. It is not in my best interest to worry about PBC or obsess about the things I am no longer able to do.

After coming along on another visit to my doctor, my husband said: "The PBC is here to stay and it will cause you to have bad days and bad moments. We will always have to keep that mind. However, if you can accept that and stop worrying about the things you cannot do, you can have a fine life."

PBC opened the door wide for more autoimmune diseases

Annelies (44)

During my three pregnancies, I began to suffer from migraines, a foggy brain, extreme fatigue and itching across my entire body. It felt like the itching was inside me and I couldn't reach it to scratch at it. I didn't have these symptoms all the time, so I simply blamed them on the stress of raising my family and living a busy life as a mom. When my youngest son turned four, my symptoms grew worse. Again, I - along with many of my doctors at the time - dismissed them as results of the fatigue caused by raising children. When my symptoms began to affect my sleep, I could no longer ignore them. My doctor did his best to help me find the answers I sought, but he was stumped.

When I was in my mid-thirties in 1998, my liver enzyme levels began to rise. During a visit to my dermatologist for something unrelated, he saw my test results. After I told him that my liver enzyme levels had been elevated for some time, he referred me to a gastroenterologist. They ordered a liver biopsy. A biopsy is not necessary in order to diagnose PBC, but the doctor was not thinking about PBC at the time, as they thought I was far too young for that.

The physician tried to assure me that my values were "not that bad". By this time, I had read up on liver diseases and I knew how severe a liver condition can be. I called the NLV and asked them whom I should see to learn more. That led me to a true hepatologist: a doctor whose hobby it is to study the liver, as the current director of the NLV put it. I had finally find that doctor.

PBC expert

The hepatologist conducted another series of tests. When the results came in, he explained to me that I had Primary Biliary Cirrhosis or PBC. In 2014, the disease's name was changed to Primary Biliary Cholangitis. He told me that PBC is a progressive and chronic autoimmune disease that affects the bile ducts of the liver. He said I had come to the right place; he turned out to be one of the foremost experts and researchers in the field of PBC!

My hepatologist told me that I would have to start taking medication and that treatment would never make my disease go away entirely or even alleviate all my symptoms, but that the goal was to slow down the disease's progression. As the years went on, I received various treatments that have helped me get most of the itching under control. I still suffered from the brain fog and my fatigue came and went, but the treatment seemed to have at least stabilized the progression of my PBC; my liver enzymes levels were checked every three to six months and the values stayed within the acceptable margins.

Unfortunately, PBC is not all I have had to deal with. It feels like PBC has opened the doors of my immune system to other conditions that are also classified as autoimmune diseases. I have since been diagnosed with Sjogren's Syndrome, Crohn's disease and rheumatoid arthritis. However, I can cope with these issues and I believe my fight against PBC has helped prepare me to take on these new challenges.

Source of inspiration

I visited my dear hepatologist until he retired. Of course, he has every right to retire, but I have to admit that I had hoped he never would. He has been both a teacher and a source of inspiration to me. From the first moment I met him, he made me feel validated and supported; validated because he confirmed that the symptoms I experienced were real and not just figments of my imagination and supported because he told me that no one better understood how I felt than I.

I have since been assigned a new doctor. Someone so young took some getting used to. By now, we have built an excellent rapport between us. This physician is highly proactive and also invests in his patients.

I am on the right path, which means the condition of my PBC has not changed. The fact that the disease has not progressed is something I see as a major victory. The main goal continues to be to slow down the progression of my disease as much as possible. My symptoms have certainly not gone away; they come and go in frequent clusters, so I have to adjust my daily activities based on how I feel. If that means doing less or withdrawing from certain obligations, so be it.

Creams

Sonja (56)

The glass shelf above my sink is full of jars and tube bottles. They were all gifts from well-meaning friends and acquaintances, given to me because I suffer from cholestatic itching.

It is the kind of itching that will drive you absolutely crazy and against which no cream is effective. I cannot seem to get that message across.
Friend #1 has eczema and says: "I know exactly what it's like to be itching. Try this cream, it did wonders for me."

Friend #2 has psoriasis and gave me a jar of ointment: "You have to keep your skin oily, that really helps against the itching."

Acquaintance #3 suffers from extremely itchy skin herself and swears by aloe vera: "It makes your skin softer, which helps with the itching."

Friend #4 takes one look at my scabby legs and gives me some calendula ointment: "Calendula is a natural remedy. There is no chemical crap in it. Once your wounds heal up, your itching will be gone."

Acquaintance #5 has had great success with menthol ointment: "It cools your skin and really gets rid of the itching."
Does this sound familiar? Do you ever find yourself struggling to sit still at a party or during a meeting because the itching is so bad, only to have someone tell you that they know exactly what you're going through and have just the thing to cure you? Of course, they're only trying to help.

No matter what, I cannot get them to understand that my itching comes from within and that creams and ointments may offer slight temporary relief at best, but that's it. After all, they know exactly what you're going through...

PBC: the P does not stand for the Pension I was so eagerly looking forward to

Karin (69)

After thinking about it for years, I finally decided to retire early. A recently retired friend and I were planning to go on many amazing trips and see the world together. We had been looking at possible destinations online for a long time and now it would finally happen. We figured we should go before we got too old and began to develop all kinds of physical ailments...

When the date of my retirement approached, I was not feeling very good. That's putting it mildly, to be honest. In reality, I was so tired that I could not bear to face the day when I woke up in the morning.
At first, I thought I had caught the flu. When my symptoms didn't go away, I figured it was my body's reaction to my decision to retire. I had worked hard my entire life and had had to cope with excessive stress at work for the past few years, which certainly factored into my decision. In fact, I had been feeling tired for years.

Urged on by some friends, I decided to pay a visit to my general practitioner. They said we should begin with a blood test, so we could eliminate a few things on the outset. The medical terminology behind the checked boxes didn't mean anything to me, with one exception. I knew HB had something to do with anemia and Hb1C with diabetes.
I was so beat that I didn't even have the energy to look up what they would be testing my blood for. The old me would have wanted to know every detail.

My follow-up appointment was scheduled one week later. After a few days, I got a call from the doctor's assistant, who told me that my GP wanted to see me sooner than that. "Fine by me", I thought. I listened to what she had to say and understood that I had to visit a gastroenterologist because my liver enzyme levels were far too high. I had no idea what the fuss was all about, but if that other doctor had a pill that would make me feel better, I would happy to meet with them!

I met a nice young doctor in the hospital. I remember thinking: "It's amazing how much you know at such a young age." She quickly prescribed a number of exams. I meekly showed up for every appointment. After all, that was what it would take to get better. This doctor seemed to know just what to do once I had my diagnosis. She would find the right cure for me...

Being told that I had Primary Biliary Cholangitis (PBC) and that there were no drugs that would help me get better felt like a death sentence. The medication I was given was only designed to slow down the disease's progression. It was certainly not a cure!

After four years of good and bad times, I now have my life reasonably under control. I know that there is no cure for my condition. I also learned there is something else to worry about: the stigma. I never thought I would have to deal with that. Anyone with liver disease reading this will undoubtedly know what I'm talking about. The people around you are quick to associate liver disease with excessive alcohol usage. That is not what you want. When you think about it, it is bad enough that people whose condition is caused by their alcohol addiction are treated this way by society.
Let's go back in time for a bit. Because I always like to know everything about everything, I began researching my condition online. I soon found the Dutch Liver Patients Association. I read their magazine, NLVisie, and their the newsletters and I attend the gatherings. I do that mostly to hear new information and to socialize with others like me. Anyone worried that these gatherings are just people complaining about their condition can rest assured; that is not true at all. It is just nice to talk to people who know exactly what you mean when you say you are having a bad day. In a way, the NLV saves me the trouble of constantly being on the lookout for new developments regarding PBC; they do all that work for us. Instead, I can focus on my (new) hobbies, such as studying art history and bird migration.

I never got to go on my trip around the world with my friend. That's a shame. Then again, life doesn't always go the way you planned.

Without darkness, you cannot appreciate the light

Nora (56)

I was caught in a tug-of-war between my head and my body and it felt like I was being torn apart. In my head, I could do anything I wanted, I was making plans and I had dreams. However, my autoimmune diseases gradually widened the gap between my desires and my abilities. Despite the various diagnoses I was given, I kept doing what I had always done - but I could not escape the cold hard truth. Nothing was normal anymore. Everything was painful, took effort or disproportionate amounts of time and energy. Doing something quickly had become a thing of the past. It was so incredibly frustrating!

As the years went by, I had to learn to plan and weigh my options. What should I spent my finite energy reserves on? I had to learn to say "no". Not because I didn't want to do something, but because I couldn't. I had to learn to stop trying to justify the choices I was making. Explaining why I could do some things but not others proved to be an impossible task. I often did not know myself and no two days were the same for me.

I began acting worn out and tired even on my good days, just so I wouldn't get people's hopes up. Every time I felt like I had found a good balance between my desires and my abilities, something else came up that would throw everything into disarray again.

Acceptance and Resignation

Over time, I learned just how important it is to keep trying to align your Desires and your Abilities. That requires Acceptance and Resignation, which are not exactly my strong suits. I refused to accept my disease for a long time and behaved in a euphoric manner instead. When my resistance became counterproductive, I had no choice but to face the truth.

Accepting my disease led to a painful process of saying goodbye. I had to bid farewell to my professional career, my hobbies, my colleagues and my friends, my health and a large part of my freedom. I had to learn to live with limitations, insecurities, a lack of understanding from and dependence on others. I had to change the way I lived my life and drastically modify my wishes and expectations. Above all, I had to learn to love my life and myself all over again.

Against all odds, I began to enjoy my life again. I have learned that continuing the fight is a waste of my energy and that it alienates me from who I am. I have also learned that pushing aside the feeling of sorrow for a loss (mourning) only exacerbates your suffering. Accepting that my disease is a part of me makes it easier to bear. Once I was able to face that truth, I could begin the process of forging a new path in life. Ultimately, I was able to find light and happiness again. In my own way, I can contribute to society and do things for other people. That, in turn, gives me new energy. It's been a long, hard road, but I have never stopped dreaming. There can be no white without black and without darkness, you cannot appreciate the light!

PBC turned out to be more than a chronic disease that would be “relatively easy to live with

Josefien (63)

I was 58 years old. I can still see myself sitting in the doctor’s office. He knew what was causing my symptoms, which I had been having for a number of years at that point. I felt so relieved to finally be able to put a label on what was wrong with me: Primary Biliary Cholangitis, or PBC. I immediately understood that this had something to do with inflammation in or of my liver, my immune system and that there was no cure. My biggest fear - that I only had a little time left to live - proved unfounded.
I was given some medication and I remember thinking: “At least this will get rid of my symptoms. I can live with this.” I had another autoimmune disease that I was coping well with and I figured this wouldn’t be too much of an extra burden.
I could no longer do the job I loved.

It did not turn out that way at all. The medication was not as effective as I had hoped. Changing the dose, switching to a different brand and – as an act of desperation – changing my entire lifestyle did not have the desired effect. My disease dragged on like that for years. I could no longer perform the job I loved as a teacher. I had already reduced my hours, but the unpredictability of my energy reserves in the morning made work impossible. I was no longer able to show up well prepared to try and convince a bunch of unmotivated teens that my subject of choice – English – would open so many doors for them. I loved to be able to treat everyone to some English shortbread cookies after all my students had passed their test. What hurt the most was gradually having to say goodbye to my students, especially from the weaker brothers and sisters among them whom I was still able to captivate and motivate. I would miss their bright eyes in an otherwise uninterested expression when things were going well and especially once they had obtained their diploma.

It has taken the joy out of my life

My silent assassin is called PBC. Although it is not going to kill me, it has taken the joy out of my life. I am usually a “glass half full” kind of person, but it is not easy to hold on to that optimism when you feel exhausted as soon as you wake up in the morning, when your muscles refuse to do your bidding and the fog in your head won’t clear. Unfortunately, the medication that is available is not doing it for me. I learned from the NLV that a new drug is available, but the Care Institute Netherlands refuses to bring it to market here because alternatives are supposedly available.

How I would love to tell those people my story, about how having this condition affects you and the people close to you, how it controls every aspect of your life... Those decision-makers have no idea what they are doing to people for whom the standard drugs are ineffective. I know that the NLV is on top of things and so are the doctors. Thus far, however, their efforts have been in vain.

I have been a member of the NLV for years. My doctor advised me to join. “At least you’ll be kept up to date on the latest developments”, he told me. I never regretted it for a day. It has brought me so much more than that. Although there aren’t that many opportunities to interact with people in the same boat as I, my membership reminds me that I am not in this alone. It reminds me that there many people out there who recognize and acknowledge my story. At times, it can be so difficult to explain to those around you what you are going through.

I would like to be more positive

Christa (57)

For a long time, I wasn't sure if I was going to write my story down. Well, I already had written it down, but I was afraid to send it in. It is not very positive, you see. Like many others, I have read countless personal narratives online from women suffering from PBC. Strong women who appear to have found ways to cope with their PBC and who are able to maintain a positive and up-beat mindset. That is not me at all.

I'm sure they experience the same conflicting feelings I do, of being angry with your own body. I cannot find a single positive aspect about my PBC. Nevertheless, I have decided to finally share my story. It is certainly not easy to cope with having PBC in your life.

I often feel miserable as soon as I wake up in the morning. Everything feels heavy and I would love nothing more than to stay in bed all day. The clock tells me that I have been lying in bed for ten hours already, but I feel far from rested. Even though I am in my bed, I cannot seem to fall asleep. I feel even more exhausted now than I did when I went to bed last night. My husband didn't say a word when I said goodnight to him at nine in the evening.

I feel guilty, because I wanted us to have an enjoyable evening together. I was absolutely knackered, though. I was so tired that everything hurt. I've been taking my prescribed dose of ursodeoxycholic acid. My doctor recently prescribed a higher dose, but that doesn't seem to help at all. I know that a different drug exists, but it is not available in the Netherlands.

I have heard about a different drug, but that is not available in the Netherlands.
I recently contacted the NLV to ask if they knew anything more about that. They certainly did. Together with physicians and the pharmaceutical company, they have been trying to convince the Care Institute Netherlands that the new drug is the only hope of a remedy for some people. The Care Institute appears resolute in its position, however. I believe they think there is not enough scientific evidence available.
Apparently, other countries are not so scrupulous, because the drug is available there.
As I understand it, the EMA has already approved the drug and the Medicines Evaluation Board (CBG) was also positive about it. How I would like to trade lives with the people making these decisions - if only for a day. Their partners can join in as well, so they can experience for themselves what it is like to watch your partner go to bed extremely early, knowing that they will probably feel just as exhausted – if not more so – in the morning.

Focusing on the positive

Don't get me wrong; I do try to focus on the positive things in my life. It is just so hard most of the time. I was recently referred to a medical psychologist. I like that. They are helping me find ways to better cope with the impact that PBC has had on my life. I know that the PBC is never going to go away. I have to find a way to live with it, because I really do want to have a more positive mindset. It is just hard for me to keep that up all the time. Together, we discussed other issues in my life that are a direct result of the PBC. The fact that I had to quit my job certainly doesn't help. What frustrates me the most, however, is knowing that there is a drug out there that may improve my quality of life – and that of my family – and that I simply cannot get my hands on it. I am truly grateful to everyone who is fighting to make that drug more widely available. I keep hoping for good news that doesn't come. I call the NLV sometimes. I know that they don't mind and that there is someone there who will lend me their ear. They understand how bad I feel. I feel like such a weakling at times.

When I spoke to José* and told her how I felt, she said I was not giving myself enough credit. She told me I was strong and that I was simply going through a rough patch, which is perfectly understandable! It's funny how hearing that perked me up a bit. See, I managed to end my story on a positive note after all.

(*director of the NLV, ed.)

Doing what you can and want

Franciska (44)

I was diagnosed with PBC about four years ago. I had never heard of this disease before then. Pieces of a puzzle began to fall into place. Why did I struggle so much with things that seemed so easy for others? Why did I feel so tired all the time? I was working eighteen hours a week at the time, divided across three days. When I reached the point where even that became too much for me, I called in sick.

After a period of rest, I began a reintegration process. I was assigned an OHS physician who was unfamiliar with my condition and could not be bothered to learn more about it. At the same time, he thought he knew what I needed to do to get better (from a chronic disease!): meet with the GP's nurse practitioner and improve my condition with a sports medicine expert. Unfortunately, none of that did me much good. He wanted me to undergo an intensive training with a team of physical therapists, psychiatrists, etcetera. That plan quickly fell through, because I absolutely did not have the energy to do all that. On top of that, I was „lucky" that one member of the team had heard of PBC before. They said the program they offered would be far too taxing for me. The GP's nurse practitioner had told me to take frequent breaks in between my various activities. I did that, but it hardly seemed to help.

Eventually, I ended up at the hospital's medical psychology department for cognitive behavioral therapy. The physician immediately told me that plan would be scrapped, because the problem was in my body, not my head. She asked me to draw up a list of my daily activities. Looking at that list, she quickly discovered that my balance was off. I was doing too much and not getting enough rest. She emphatically told me that I had to remember I was sick. She really did a lot to help me find the right balance in life. In the meantime, my return to work was not going smoothly. After a reorganization, I was assigned a different OHS physician. They were not familiar with my condition either, but they made an effort to read up on it. He saw that although I wanted to work, I simply was not able to. I would be coming into contact with the UWV in a little while. He suggested that I began creating a file immediately.

The struggle with the UWV

After receiving sickness benefits for almost two years, I found myself at the UWV. That was a struggle in its own right. Once again, I ran into the problem of people simply not knowing about this condition. I had to fight to prove that I am willing to work, but unable to. The UWV drew up a report that did not reflect my situation at all. After filing an objection with the help of a lawyer, I was diagnosed as being fully incapacitated for work. I have to deal with various symptoms on a daily basis. These include extreme fatigue, struggling to find the right words for things, concentration and memory issues, dry eyes and a dry mouth. I never know when the symptoms will flare up again. I have learned to listen to my body when it tells me to lie down for an hour, because I won't make it through the day otherwise.

I am doing reasonably well because I pay attention to what I do, when I do it and how. My dear husband and son are both very understanding of my needs. Several other people around me have also made the effort to learn more about my condition and they are there to support me. The Dutch Liver Patients Association and a fellow patient have been a major help to me. They were and are there to answer any questions I have. By now, I have found a way to cope with my disease. Whereas it used to feel like I was pushed into the deep end and left to fend for myself, I now feel more at peace. I feel optimistic about the future. I get to do what I can and what I want – at my own pace.

Keep going as long as you can!

Jacqueline (57)

I was diagnosed with Primary Biliary Cholangitis (PBC) in 2010. Since then, I have constantly been running up against the restrictions that this condition imposes on me. I am single, which means I do not have a partner to rely on financially. I work a full-time job with a thirty-six-hour work week. If I were to start working less, that would have a serious impact on my income. I want to avoid that if I can. It does come with a price, however.

I work at a hospital where I am responsible for the catering. I deliver coffee, tea and lunch. It is a physically demanding job, especially during peak times. That means I am in more pain and suffer from heavy, restless legs. No one knows what causes my restless legs. I don't know if it is a side effect of my condition. To find out, I was told to undergo a sleep study at one time. I did not appreciate being told that I had to lose some weight first. They said they couldn't conduct the sleep study before then. What are you supposed to say to that? I want to know what causes my heavy, restless legs, so I can do something about that.

The pain is not only caused by my condition

I do think my colleagues and my direct supervisor at work understand why I have to take a break from time to time when things are particularly hectic. I believe that because I never hear them comment on it to my face. Sometimes, though, I wonder what they really think. Do they talk about me behind my back? I hate feeling this way, so I try to convince myself that they truly are as understanding as they appear.

The pain I feel is not only caused by my condition. In 2019, I was involved in two accidents - one shortly after the other. That not only resulted in two stays in the hospital, but also caused lasting damage, including hairline fractures in my hip and diaphragm and painful joints. My recovery was further complicated by my osteoporosis, which has resulted in even more restrictions. That same year, I had a liver biopsy and was diagnosed with autoimmune hepatitis (AIH). I feel my body growing weaker as time goes on.

My work takes up most of my energy

I try my best to recover and return to work full time. I have no choice, because I cannot afford the financial consequences if I don't go back to work. That does worry me and causes a lot of stress. Can I keep going and if so, how? It requires a great deal of willpower and perseverance. Luckily, I am able to muster those. I have to.

I also keep telling myself that it is hard to keep working, but sitting at home all the time would not be ideal either. My world would become even smaller than it already feels at times. I hardly have time for private activities or social interaction. That is mainly because I cannot stand the hustle and bustle. I am quickly overwhelmed by loud noises of large crowds. My work takes up most of my energy, because it can be quite hectic there as well. Because I am trying so hard to keep working, it does bother me when my employer asks what is wrong with me this time when I call in sick. He knows about my disease and my desire to keep working. He also knows that my situation has a major impact on my personal life.

I have had to give up some important things

I am fortunate to have two very good friends, one of whom I also work with. They are there for me when I need their support. I have had to give up some important things, though. For example, I can no longer watch my grand-children. Although my son and daughter-in-law understand my situation, it is very painful for me.

I sometimes ask myself what is more important: devoting all my energy to my job or working a bit less and having more energy left for the things I love to do? The UWV, an organization that is helping me look for and find work that suits me better, has asked me that same

question. I only have one answer: save as much energy as I can for work. I have to earn a living, after all. I am part of a generation of go-getters, even when times are tough. I want to keep going as long as I can.

PBC ruined New Year's Eve for me and has controlled my life ever since

Marjoleine (55)

It was New Year's Eve, 2003. Like everyone else, we were celebrating the coming of the new year with our best friends and we went outside at midnight to admire the fireworks with our neighbors. I was 43 at the time and the festive evening I had been looking forward to did not play out as expected. I felt miserable all evening. I left my husband and my kids to entertain our guests and went upstairs to lie down "for a bit". At eleven thirty, I forced myself to get up and put on a happy face. I saw everyone staring at me: "Are you alright?"

I spent most of the early days of the new year in bed. It felt like my arms and legs had been cast in concrete. I had a dry mouth and itching skin. I was not digesting my food properly, either. It was clear that something was wrong with me, but what was it?

My general practitioner had my blood tested and she called me up even before the appointment we had scheduled to discuss the results: "I am looking at your test results and I want you to see a medical specialist. Your liver values are off." Liver? Do I have liver cancer? What else could it be but that? I had no idea at the time that there are around twenty-five other liver diseases besides cancer, nor that I had one of those twenty-five. An appointment was made for me to see a gastroenterologist. In week eight of the new year of 2014, I got my diagnosis: PBC, or Primary Biliary Cholangitis. At the time, the C still stood for "cirrhosis", but that was changed around 2015 or so.

"How did that happen?"

"What is wrong with you", my family and friends would ask with concern on their faces. "How did that happen?" By now, I had learned from my liver specialist that I got this disease through no fault of my own. It was just dumb luck. I had memorized an explanation to offer people: "PBC is a chronic autoimmune liver disease that makes the body's immune system turn on the healthy cells in the bile ducts of the liver, which can lead to cholestasis or impaired bile flow. This can result in an accumulation of bile acids in the liver, which leads to inflammation and scarring, known as fibrosis. There is no known cause of PBC and there is no definitive cure, although medication is available to slow down the disease's progression." As I told people this, I could see the confusion dawning on their faces.

I have since learned a better way to explain myself, while also avoiding the stigma that is commonly associated with liver disease. I always start with: "I have a chronic illness whose cause is unknown and for which there is no cure. It is a liver disease called PBC. This has nothing to do with my lifestyle choices; it is simply the result of how my body functions." That has allowed me to avoid the accusatory question of "Are you an alcoholic?" I have gradually begun to understand the awful stigma that people suffering from alcohol addiction have to cope with on a daily basis.

Things really took a turn for the worse in 2018

I was doing reasonably well for the first five years or so. I took my medication on time and had no real symptoms to speak of. I did change lifestyle a bit to go easy on my liver. I began to watch my diet and tried to get more exercise. That does nothing to alleviate the PBC, but it certainly doesn't hurt my body or my liver either. After five years, I began to experience "flare-ups". I felt absolutely miserable, but my physician and I were able to find a new way to cope. Things really took a turn for the worse in 2018. A test showed that my liver had become "cirrhotic" and it looked like the "urso" had stopped doing what it used to do so faithfully. My condition was not bad enough to make me eligible for a liver transplant, but it did mean I had to change my medication.

The doctor told me that a suitable drug existed on the market, although it was not (yet) available in the Netherlands. I now know that this drug has still not been approved for the Dutch market. It is really hard for me to know that there is something out there that could help me, but it is unobtainable for me. I am so eager to try that drug and I was hoping that it would at least help to improve my quality of life. Alas, I cannot get it because the government does not approve... Do those people even realize how their decisions impact people like myself? At the moment, another drug is going through the study phase. I have been told that I will get to try that.

Talking to others about PBC

I have learned to adjust in order to keep doing the things that bring me joy and I will not let anything stop me from living my life. Talking to others about PBC can help with that. In hindsight, I regret the fact that it took me almost ten years before I came into contact with other people with PBC. I knew about the Dutch Liver Patients Association, but "there was no way I would be joining that." I have certainly learned to eat those words. They are really good at explaining a world that is – at first – entirely new to you and they fight tirelessly to raise people's awareness of liver disease. They are always willing to find the answers to your questions or lend you their ear if you feel the need to talk. I attend every gathering they organize, which are known as NLV conventions these days. I always look forward to the next one. They are an opportunity for people to get together and share their laughter and their tears. I love going there and talking to other people with PBC or a different form of liver disease. It is great to be among people who know what you're going through.

I know that the NLV is doing everything it can to make drugs more widely available. It is heartening to know that they represent "me" and that they are telling "my" story to the wide world. I hope the people making decisions about my life sit up and take note!

The long road to being diagnosed with PBC

Iris (61)

I had been feeling under the weather for a while. I was living a busy life, so I didn't think too much about it. Still, I figured I had best go to my GP anyway. They found all kinds of abnormal blood values and sent me to the internist at our local hospital.

Another round of blood tests. The internist told me that although many of my values were a bit high, they did not fall outside the normal threshold values. They advised me to have my GP keep an eye on things.

Relieved knowing that nothing was seriously wrong with me, I went home again. Before long, though, I got a nagging feeling that something was not right. My general practitioner told me not to worry...

For at least five years, I kept going back to my GP every now and then. In all that time, I had no new blood tests. After all, the internist had assured me that nothing was wrong in that area...

You only have one heart and one liver

Then I became seriously ill with an inflamed kidney. It was bad enough that I ended up in hospital. See, I thought to myself, this was bound to happen some day. At my local hospital, they submitted me to a whole battery of tests. The physician told me that I had type 2 diabetes, that they had found elevated liver enzyme levels and that the gastroenterologist would be paying me a visit. I didn't really pay attention to what they were saying. My entire family has type 2 diabetes and I didn't think much about the elevated liver enzyme values either.

While I was still recovering in the hospital, I developed heart problems. The elevated liver enzyme levels suddenly seemed far less important. I was doing my best to survive and thought to myself: I only have one heart. It never occurred to me that I only have one liver as well. In fact, I didn't even consider that something might be wrong with my liver. Having elevated liver enzyme levels didn't mean anything to me.

The gastroenterologist came by and told me that he suspected I had PBC. He failed to mention that he didn't really know much about that condition. I underwent various exams and tests, but my heart remained my main concern throughout it all. I was given a pacemaker and began to feel better afterwards. After I was discharged, the gastroenterologist wanted me to come see him again. More than anything else, I showed up simply because he had asked me to. He told me that I did indeed have PBC. I couldn't remember what those letters stood for. I was prescribed medication and gradually began to realize that PBC has to be taken absolutely seriously. Six years had passed since I first developed symptoms.

Not a coincidence

I felt like a whole lot had landed on my plate all at once. I read stories from other patients online and noticed that many of the things that were wrong with me are classified as autoimmune diseases. Could all this be more than coincidence? That's right: it was not a coincidence at all. The physician had failed to tell me that, but the patients' association I had since joined finally clued me in.

Although I really liked my physician and did not question their abilities or expertise, I felt a need to see one who knew more about PBC. So I did. It turned out my diagnosis was correct, but I had not been receiving the treatment I needed. I had no idea my weight was so important to determine the correct dose of the medication I was taking. I had gained quite a bit of weight since all this began and my dose of ursodeoxycholic acid had never been upped accordingly.

The expert also believed that I was not doing as well as I thought I was.

I was told to take more medication, but to no avail. Nine years have passed since then. For the past three years, I have been taking medication to prevent the condition of my liver from deteriorating further. I know there is no cure for this disease.

Everyone around me keeps telling me that I have to rediscover the right balance in life. Well, I still haven't found it. The PBC hangs over my life like a dark cloud. You have to give in to it, someone else told me. I would like for someone to give me the pamphlet with step-by-step instructions on how to find your balance in life and how to give in to a disease that turns your life upside down and takes over completely.

I am glad that I am not reduced to sitting on the sofa in a state of depression, but it has certainly not been easy. I can imagine how other people might have more trouble coping with their condition.
It is not easy to live with PBC!

I am greater than the sum of my parts

Wilma (57)

The hematologist (blood specialist) at the Erasmus MC suggests an MRI scan. "Well," I tell them, "I had one done at the UMC Utrecht just last month." The hematologist tells me that they were looking for different things at the time and that they do not have access to my test results. She wants to have a scan "of her own." Alright then. I'm not happy about this, or about the fact that I now have to see a second specialist at the Erasmus MC, even if only for a little while.

Last year, I lost track of the many conflicting messages I got from the countless specialists I had to meet with*. I was finally able to arrange that I would be seeing the specialists at the UMC Utrecht, except for the Erasmus MC's gastroenterologist. One of the specialists in Utrecht would oversee my treatment. The rheumatologist was selected for this role, because she was the one I saw most frequently. She would be in charge of my general wellbeing and my medical file. After a promising start, it became clear after just a few months that my vision - of having a single coordinating specialist to whom all others would report - was a pipe dream. Every specialist turned out to have their own unique method of (not) communicating with their colleagues within and outside their own hospital and of reporting in my (digital) file. The latter made it very hard for me to take matters into my own hands. Although there is such a thing as a multidisciplinary approach, it only works with a small number of specialists, or only within their own department and only when there are new findings.

GP cannot do their thing

At one point, I had hoped that my GP would be able to oversee things. However, they lack the necessary knowledge in some areas. More importantly, many of the specialists only inform him sporadically and highly irregularly at that, which means he has no idea what is going on. That has turned out to be a big problem in case of an emergency!

The reality is that I have to visit many specialists and undergo countless exams as a result of the many autoimmune diseases* I have. The number of exams could be reduced if people would coordinate better and perform a single scan or take a single blood sample to serve multiple purposes.
Another thing is that I have to listen to different visions and conclusions based on my test results, which sometimes completely contradict each other.
Many of my symptoms can be caused by any number of conditions. That makes proper coordination all the more important to prevent people from endlessly pointing a finger at each other. I frequently find myself going from my gastro-enterologist to my neurologist, my endocrinologist and my rheumatologist, only to have each of them conclude that my symptoms could be caused by something they specialize in – or by something else that would make it their colleague's responsibility. As a patient, that is useless information. Who is finally going to do something to help me?

Feeling like a jigsaw puzzle

All this makes me feel insecure. I feel like a jigsaw puzzle at times. I am made up of many pieces and every specialist has a few in hand, but I often have no idea who has what. What I do know is that no one is making a concerted effort to put all the pieces in one place and work together to solve it. I also have a nagging feeling that some pieces are missing. There is no way to know for sure, however, until the pieces we do have are in their proper place. Having to look at myself from a different perspective every time makes it hard to keep seeing myself as a whole person. Of course, I understand that the specialists do not have it easy either, but a holistic approach with proper direction and coordination would make all the difference to me as a patient. After all, I am greater than the sum of all those tattered puzzle pieces. Besides my ailing physical component, I also have a mental and an emotional component. These are not separate spheres, but inextricably linked parts of a single human being: ME!

* Gastroenterologist for PBC, liver cirrhosis, portal hypertension and esophageal varices, lymphocytic colitis / Hematologist for leukopenia and thrombocytopenia / Endocrinologist for Hashimoto's disease and steroid-induced diabetes / Rheumatologist for Sjogren's syndrome and Raynaud's disease / ENT specialist for chronic sinusitis / Dermatologist for vasculitis / Neurologist for multiple sclerosis / Ophthalmologist for nystagmus / Pneumonologist for asthma