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## **MEET JOHAN**

### **How old are you now?**

I'm fifty-four.

### **Do you have children?**

Yes, I have two daughters. One is twenty-three and our youngest daughter is eighteen. My youngest daughter is still living at home while she is studying Technical Medicine in University. My oldest daughter lives in a student home. She is in the last year of finishing her master's degree. She plans on being a doctor. I am very proud of them. They are very smart girls, just like their mother.



### **Do you work?**

No, I am home now on disability. My heart function worsened a few years ago and it wasn't possible for me to continue working. I am thankful because I had been stable for a very long time.

### **What was your profession before you stopped working?**

I have always done administrative work. I started out as a bookkeeper in an accounting firm. Over the past twenty years I was a salary administrator. I like numbers.

### **Tell me about your typical day.**

I enjoy walking and working in the garden. I have a schedule for each day. I get up at the same time every day, I eat and shower. The first activity of the day is to walk the dogs. I do my daily chores around the house. I do physical therapy maneuvers every day.

### **What kind of dogs do you have?**

One is a Pekingese and the other is Tibetan Spaniel. They are good dogs for me because they don't walk too far. They don't have to run like a shepherd dog. They sleep a lot just like me, that's perfect.

### **Do you have any hobbies?**

Oh yes, I enjoy reading detective and mystery novels. I love reading books about space and everything around it. That is very difficult to understand but I find it fascinating. I enjoy learning about space travel. I watched the launch of SpaceX a few days ago. Wow, that was fascinating to see.

I play keyboards. I really enjoy gardening. I build miniatures of old buildings. I have all kinds of things to do to keep me occupied. I always try to do things that I enjoy doing later in the day after I have done what must be done. This way, if I can't get to it, so be it, no obligations.

I volunteer one day a week in an elderly home, coloring with the seniors which requires very limited energy. I generate a newsletter for a volunteer organization. So, that is how I fill my days. They are rather full.

### **Whew, so I was going to ask if we could get you to volunteer for BSF but I'm not sure you would have time.**

If ever there is anything, I can do for BSF let me know, I will make time.

### **Could you tell us more about these miniature buildings?**

They come in kits. You have to glue each little piece brick-by-brick. It requires a lot of patience because there are around 5,000 pieces. My hands are not steady. I really enjoy doing this. So, I have adapted by using tools that help steady my hands when I put the buildings together.

### **What was your personal history with Barth syndrome?**

As we have looked back in our family history there have been others suspected to have Barth syndrome. My sister had two sons with Barth syndrome. When her second son became ill the doctors began thinking that I may have the same thing as my nephews. My sister's first son passed away when he was 2 and her second son passed away when he was a teenager.

During my first year of life, I was in the hospital for nine-months. When I was young, I couldn't walk very well, cycling was a problem. Looking back, I realize there were all those kinds of little things that now make sense. I had no major troubles to speak

of until I was in my thirties. That was around the time when I became a parent for the first time. It was a very intense period. After a short period, my health stabilized and remained stable for quite a long time. Over the past five years there has been a gradual change.

### **What other changes have you appreciated as you have grown older?**

I have a problem keeping my hands steady. That started about fifteen or twenty years ago. It started with a trembling of the hands. It has become more of a problem. For example, I have had problems writing for some time now. I can feel the muscles in my arm even when I try to write a short note on a post card. Thankfully, we now live in a digital age where I can type.

### **What was it like growing up without having a diagnosis?**

On one hand, it was very difficult not knowing the reason why I couldn't do things but on the other hand, I never knew any better. You cannot miss something you have never had. Everyone, myself included, just thought it was residual from the the illness I experienced when I was a baby. In school, I was excused from strenuous activities to do other things. I never participated in sports. So, I never missed it. As strange as it may seem, perhaps, it made things for me.

Yes, there were times when I experienced frustration because I couldn't do what others could. I was disappointing not being able to keep up with the rest of my peers as I was growing up. But overall, it wasn't too bad.

### **Were there times when you got really sick when you were growing up?**

Yes, there were times when I would get sick and it would take longer or be more difficult for to recover than my peers. Then I was introduced to penicillin and that was almost like a miracle.

### **Have you noticed any change over time with your stamina or fatigue?**

That is something that is steadily getting more pronounced over time. The fatigue is definitely a problem. I can walk maybe 800 or 900 meters now but if I try to walk any further it is a problem.

### **How does the fatigue affect your daily routine?**

I have short periods of energy. If I go in the garden or walk the dogs, I am incredibly tired. About two or three years ago I realized I was expecting too much of myself to go for a full day. It is too much.

Now I split my days. In the morning I start my day with doing what I must do and then I rest. On a good day, I will sleep about one hour. On a bad day, I will sleep about two hours. After I rest, I have a routine for the afternoon part of the day. On holidays or special occasions, I have to be mindful not to do too much all in one day. I am not able to participate in the special events if I expend my energy. I have learned I must dose my energy very well to the activities I have planned.

The normal for someone else is not my normal. My normal has changed over time. I have found it easier for me to enjoy life when I adhere to this approach of splitting my days. It took me a long time to get to this point. For many years I pushed myself to keep going or to try to work through the fatigue. I thought I'll just get through the day and rest at night. When I was doing that I had more pronounced periods of illness, flu-like illness, my heart problems got worse and so on. At one point, I spoke with a doctor and he told me *"You're doing too much. Your body can't cope. You can't keep going on like this. If you do, something bad is bound to happen."* It opened my eyes.

After speaking with him I decided I needed to listen to my body and not push myself beyond my limits. My mind could not control my body's fatigue. So, I made adjustments in my life to spread my energy better over the day. That approach has gone rather well for me.

### **Would you share some other examples of adjustments you have made in your life?**

When I go on an outing that requires me to walk long distances, I use a mobility scooter. I have made all kinds of adjustments in the things that I do. I like to work in the garden, but I do a maximum of one hour every day. I use tools that will allow me to conserve my energy. For example, to remove weeds I use a tool with a long handle. That way, I don't have to get down on my knees. I think about everything I can do to conserve energy in order to continue doing the things that I enjoy in life.

I make choices about what is important. I can't do everything. I can't do too much in one day. For example, I can't plan to go on an outing to visit people then plan to work in the garden later in the day. That won't happen. These adjustments evolve over time. As I have made adjustments to conserve my energy, I find I can do more. I have learned by reading about what others do through the Barth Syndrome Foundation and the Heart Council here where we live. I pick tips up that I can apply in my life to make modifications to optimize my energy.

**Is there anything you would like to do or a place you would like to see?**

I would like to see the Great Wall of China and the Grand Canyon in the United States. I would like to go to a place where I could see the northern lights. My doctor has advised me that I can't fly anymore. So, I read about these places or see them on the television.

**I am sorry to learn you cannot fly anymore which prevents you from coming to the conference.**

Well, you will have to host a conference in Amsterdam.

**What advice would you offer to a young person with Barth syndrome?**

The most important advice I could offer is be happy with what you can do. Learn to accept that you cannot do more. Focus on the things you have and focus less on what you don't have. Try to focus on the positive. There will still be difficult periods, but it makes it less difficult when you focus on the positive.

When I was in school, I was always the smallest and the weakest. Yet, I always had classmates who were there to help me out. They took up for me when others bullied me. In this situation, there is a choice. You can focus on being the weakest or you can think about someone being there to help you when you were in a difficult spot. You may not be able to make a choice about having Barth syndrome, but you do have a choice about how you see things. It is a mindset. That is where you will find your strength.