

Paije's BHD Story: Lungs

Interview by Jazzmin Huber Charity Officer

Jazzmin: Hello Paije. Thank you so much for joining me all the way from America today. My name is Jazzmin, and I am a charity officer at the BHD Foundation which is managed by the Myrovlytis Trust and we're here today to talk to Paije about her experience living with BHD. So, we're going to start from the very beginning. When were you first diagnosed with BHD Paije?

Paije: So I was 25 I believe. The previous year when I was 24 I had been diagnosed with a rare cancer called leiomyosarcoma which really just ended up being a tumor on my leg. We just had to surgically remove everything so I didn't have to have chemo or radiation, and then shortly after that we found out that BHD had been found in our family. So my father got tested in 2007. Uh basically right when I got when I was diagnosed with cancer he decided it was a responsibility of his to get tested to see if maybe this was related. So then he tested positive so then I went through with my testing and tested positive also. So that's really what triggered the whole thing to begin with was me getting this rare unusual cancer.

Jazzmin: Did you had any symptoms before your initial diagnosis for BHD? No no no. I had no growths no none of that actually. I started showing symptoms after. The cancer was kind of like the the initiator of all of it. As a very healthy individual and also young it was unusual for me to have had that cancer at 24. So it's rare to begin with but it's also very rare to have it in young people.

Jazzmin: What were the first symptoms you did develop?

Paije: So my oncologist was the one who really was overseeing all of my preventative treatment and everything because he'd already dealt with this rare cancer of mine so he was the one that was kind of prepping me on what to expect. I had taken an overseas trip and on the plane ride home I had felt because I'd ridden on planes many times and I'd never had pain before, but on the plane ride home I had this unusual sharpness in my chest it was very small, it was not overwhelming and it went away. So I didn't do anything about it. I didn't tell anyone about it, I came home I had been traveling from Ireland to Northern Michigan at the time so there's like environment different you know just as far as the seasons go and everything and I developed a cough. I thought I had like a summer cold or something you know like just from traveling because I've been on airplanes yeah because it really just felt I'd always had kind of sinus issues anyways and it didn't feel any different. But after a week the cough started becoming overwhelming. I wasn't getting better I was getting worse type of thing, and then I would have bouts where I couldn't stop coughing and my chest would feel like something very heavy was sitting on my chest. And I'd have to lay down and just wait for it to pass type of thing. And again like I was just one of those people who were like this is I

don't know why I just downplayed everything you know and nothing had ever been wrong with me in my entire life and um you know this is warm weather season in Michigan and so

we're riding our bikes all the time like 10 miles a day you know just doing all kinds of things and I would just start coughing and so this happened for three weeks. Um and I had already a regularly scheduled scan because I had had cancer previously so we were doing preventative screening to make sure that nothing else was was growing inside of me and so I had had a scan scheduled. During the preparation for that they always ask you like are you feeling well and I told them about the cough and they said okay and they put me in just like normal. It was a circle scanner, I'm sorry I don't know what it's called.

Jazzmin: Was it it was an open one?

Paije: it was an open one. Yes an open CT that's what it was. Um and so you know I start going through and it usually takes multiple minutes you know for them to complete it but um I was not even halfway through and they immediately stopped everything came in yanked the bed out and looked at me and they said "tell us about this cough". And I said well I don't know I've just had it for like three weeks and I was going go to the doctor about it because it just isn't going away and they were like okay okay and they like don't tell me anything. Like we're gonna and go wait in the waiting room and so I sit there for you know another 10 minutes or so and this lady comes out and the very first thing she said to me was well you look perfectly fine and I was like well that can't be good like what's wrong with me you know and she's like are you experiencing pain. I was like well I mean it's just like a tight chest and she basically told me you know you're having a pneumothorax and a spontaneous pneumothorax where your lung is collapsing and you need to go to the emergency room. Do you feel well enough to drive yourself or would you like me to call you an ambulance and I was just like again I'd never had an emergency no broken bone nothing and I was like well I I think I can drive myself you know this is fine. They called ahead and everything they knew that I was coming and it just turns out that it was a pinhole bleb that was on my lung and it had just a small leak basically and so I had a slow leak um and that's what was causing the coughing. Um I will say if I'll back up a little bit because my doctor, my oncologist had told me you know spontaneous pneumothorax was an option as far as possibilities and because they were monitoring my lungs to make sure um that the cancer wasn't spreading to my lungs and so they were preparing me specifically for this moment and I still acted like I didn't know what was going on. Um but when I said to my doctor how will I know if I'm having a pneumothorax and his answer was oh you'll know it'll be so painful and it wasn't like that at all it was it was overwhelming but um it passed and I guess I just found out the doctor at the emergency room who did my surgery and everything um, he just explained to me that I probably have a very high tolerance for pain and because it was a slow leak it's not like my lung just immediately deflated collapsed suddenly. Like it happened spontaneously but not suddenly type of thing. It was a slow leak so it was a tolerable pain for me. Um so now I know what it feels like you know so and that's what I told my doctor it's like okay well now I know what it feels like so I'll go into the emergency room sooner next time.

Jazzmin: You said you had this cough but what about breathlessness? When you were going on these bike rides were you finding them a little bit more difficult than usually?

Paije: Oh for sure yeah and like we would always choose this one route. It was very flat for the most part except at the very end like the last two miles was this giant hill in our city and we'd go up it and I was just like why are we always going up this hill because I always

struggled with the hill but it got to the point where I like had to stop halfway up the hill and like walk my bike the rest of the way. I was like I just can't do it I don't know what's wrong with me. Almost like a shaming thing where you're just like I'm just out of shape. It was just my left lung and that's that's another thing is that they're like well I mean it's not that you can't breathe because you have your other lungs still so we understand why you thought maybe this was not a big deal. But at the same time don't wait that long next time so. So yes the breathing I did have trouble breathing but I couldn't not breathe does that make sense okay?

Jazzmin: Is that the only pneumothorax you've had or have you had any since?

Paije: I've had three. So they pretty much happened very quickly after each other once it started. So the first one was my left lung and because I waited so long to go to the doctor about it they couldn't get it to come back up. Because their procedure is to try to pull the air out of your chest cavity so that the lung will naturally come back up and seal itself off. Um and they will do that first before they do anything else. So with my left lung I had waited too long and it was so far down that I couldn't get it to come back up basically. So they went ahead and did pleurodesis on my left lung and that was you know very invasive. But then now I knew what to expect and they were still monitoring my lungs as far as like scanning and they knew I had blebs on my right lung and so they said this is probably it's not a matter of if it's a matter of when, so here's what we do next time you. So immediately when the right one started I knew right away it happened while I was sleeping. I woke up and in the morning and I was like oh this feels like an elephant sitting on my chest again you know and so I knew right away. So I prepared my hospital bag. I had time to think about it this time because the first time I spent six days in the hospital recovering from that surgery that's not normal for a pneumothorax that was just because I had the surgery. So with this one I was like prepared I had everything packed and ready to go to the hospital. I was calm when I went in and they were able to get that one to come up pretty quickly I just had to spend like an overnight observation in the hospital. So they think what might have triggered that one was the oncologist who's always doing scanning and stuff found a lump on my back. Like in my lower back and we didn't know what it was but I said I'd like to go ahead and have it removed. And so we had that done. It was a one-day outpatient type of thing they did put me under. The anaesthesiologist was the one who had first said okay well she's had a lung collapse before so we want to make sure we're really monitoring her. He made it sound like that it was a concern to put me under again and two days later I had a long collapse. So we're pretty sure that process was what kind of started. I mean you're just making

assumptions. But we're able to get that one up. It was really only like four or five months later that I had a second collapse on my right lung. That was the week of Christmas, we didn't even decorate our tree. Because that was my second one in my right lung they said we're just gonna go ahead and do pleurodesis on the right lung now and we're just gonna have all of it taken care of. So they did that right away and I was able to be discharged on Christmas day so that went much more smooth and the recovery time wasn't nearly as bad as the first pleurodesis that I had.

Jazzmin: Can we explore that first lung surgery a little bit more because it's something that a lot of patients are worried about in particular recovery time and pain. What was your

experience like after the surgery? What kind of pain did you get? How or what kind of support was available to you?

Paije: Sure, well I definitely have to give very high regards to the hospital. Um it was Genesis Hospital up in Michigan and there was a thoracic surgeon that did my surgery. The pain was worse than the pneumothorax. I'm just gonna say the recovery from the surgery the thoracic surgeon really was good about explaining to me what was gonna happen without saying too many medical terms to me. He really described it as a very abrasive procedure where they scuff everything up on the inside and the healing is what makes your lungs stick to your chest lining. So as you can imagine that would hurt way worse than just a pinhole in your lung because your whole chest cavity is just healing during that time. But again I waited too long. Like it was much more invasive and the way they had to go in through my ribs or underneath my ribs. The draining tubes were also very cumbersome you know as far as I don't know if it's common for people to stay in the hospital for six days afterwards but I just know that my body definitely took its time you know healing and the pain meds cause constipation and so then they can't release you because if you don't pass gas you know. It's all these things that I had never experienced in my whole life you know and that I didn't know and so I was just like thinking something's really not right about this. So that experience was difficult. You know you don't get to shower if you've got tubes just coming out of you all the time you know and you have to eventually get up and walk around and carry your little drainage boxes with you um to gain strength and I think probably the thing I became most frustrated with was a test that they have to make sure your lungs are inflating. For me it was a hose with a contraption and you had to suck on the hose and there were three different balls in columns and you had to make all three balls come up to the top and you had to hold them there for a couple seconds. And for the life of me I could not get the third one up. I did eventually and I literally threw a party for myself in my room you know because I was just so tired of being in the hospital. But they're like you have to do this you know multiple times every day in order to get that lung to really expand and heal all the way up. But they were good about explaining it to me. But it was painful it was painful to to inflate to breathe that deeply but it had to be done you can't heal properly if you don't achieve that and so I appreciate how much they worked with me step by step you know

attainable goals and encouraging and I kept that little thing with me and I just kept doing it, just to make sure like how my lungs doing okay.

Jazzmin: How long did it take for you to kind of feel normal like yourself again?

Paije: I want to say probably a month before I didn't have any aching anymore. Those first two weeks are still pretty rough at home. You're definitely much more comfortable because you don't have the drainage tubes and everything but you have a lot of holes in you that need to be healing. For me the hardest part was my arms. My sides were very sensitive and so I always had to prop my arms up on pillows on either side and I would like prop myself. I couldn't sleep laying all the way down because it would hurt my chest and so I had to create this little pillow throne for myself in the bed and sleep like that for two weeks before I could you know roll over. It's an achy pain um but I would say it was a month before I actually resumed normal things where I didn't have to have help. I had to have help cleaning myself

because I couldn't lift my arms very well. So things to think about is it's good to have someone around that you're okay with being intimate with.

Jazzmin: I think knowing these type of things is really important for people because doctors will tell you all the medical stuff but sometimes it's the little things, even a pile of pillows just so you can rest your arm on, which are really important and not necessarily what people think about.

Paije: I was constantly thirsty, just always make sure you're drinking tons of water and just comfort is a major thing and you're just not going to be able to wear the clothing like a bra or a tank top you know depending on where your your surgery happened if you have a pleurodesis. I mean this was over 10 years ago right uh yeah so um maybe they've found better ways to do you know as far as the surgery goes where the healing is faster. My experience is that it's a slow process.

Jazzmin: Have you had any lung symptoms since then?

Paije: I'm kind of a preparer, I like to plan for things. I really grilled that thoracic surgeon you know at my follow-up appointment when he took the stitches out, and I said I really need to know what is my life going to be like now like what is it going to be because my body isn't the same so what is it gonna feel like. I said if I experience pain is that bad you know uh and I don't know if this if this is true for all doctors but he did say that I could still experience kind of what I would call zingers, kind of a quick pain when breathing. And he said it's possible if the lung doesn't fully heal all the way around the chest lining and if there's pockets that are loose still and you develop blebs it's possible that you could still get a pop you know every once in a while but your lung won't collapse. I don't get a scan every time that happens so I can't tell you definitively but I do get quick chest pains that could last for 10 minutes or a couple hours they always go away and my breathing never becomes hindered or laboured or anything like that. As fitness goes I can't just max out anymore as far as like pushing myself to the limits. My chest just starts hurting you know when I'm really putting my body through

something fitness-wise. I went for a 10-mile hike two weeks ago and it was at mile eight when it starts to ache you know and I do the yoga breathing. I breathe in real deep, real slow and it definitely like calms the pain but it just stays until your body is recovered and I don't really know why that happens but that's how it feels. Um does that make sense but yeah I don't really have anything other than that just some aches and sometimes some zingers.

Jazzmin: So other than full-on exercise does it stop you doing anything else?

Paije: Well my doctor told me that I couldn't go in unpressurised planes or scuba dive anymore, which is totally fine because I wasn't planning on doing that anyways but for those adventurers out there who are BHD you might be disappointed in that. I did recently try to donate plasma and they told me I couldn't. I got through a lot of the screening process until they found out about the lung collapse and it's not because of BHD, if you don't have any lung collapse you could probably still get through the physical part but I was permanently rejected from ever giving plasma.

Jazzmin: You were going to and it's a lovely thing to do if you can.

Paije: Yeah I mean this is just the time you know there's a great need all the time but there's a great need right now with covid. So I still find out things that I'm not allowed to do. That was never something that we talked about with the doctors so they can't possibly think of everything you know.

Jazzmin: How did you find a did you find a pulmonologist who knew about BHD or did he have to of educate himself?

Paije: Well my experience in Michigan was excellent. I was so lucky to have found both an oncologist and a pulmonologist in my county that I lived in. Not only in my state but in my county. I didn't have to drive far to find them. I didn't actually have to find them my general practitioner my doctor my normal family doctor is the one who found them for me so I was so grateful for that because I really have no idea how to find a specialist.

Jazzmin: For anyone watching we can help find a BHD specialist.

Paije: I want to just testify to that because four years ago I moved back down to Missouri which is where I grew up and I've had a very difficult time. I'm in southern Missouri and I have found that it's very different down here. My general practitioner just is not proactive about it and she doesn't seem concerned. When up in Michigan they were just like okay we're gonna make sure we get whatever scanning you get done we're gonna you know they were always on top of it and willing to help. And down here they're just like well we have a shortage of specialists in the city that I live in and so yes the doctor looked around and she's like well we couldn't find anyone down here so it's up to you now to find somebody like try St Louis or Kansas City which is the two major cities north of us and that is when I saw you guys were very active on Facebook recently about reaching out if we need help and so I emailed the foundation and they gave me I think three different names.

Jazzmin: What do you know now that you wish you had known when you were first diagnosed with BHD?

Paije: Tough question. Well I do wish even though it turned out okay for me I wish I had been not waited so long. I just was the type of person who didn't want to make a scene, cause a big deal about nothing you know make mountains out of mole hills I didn't want especially with health I didn't want to act like someone who was you know something's really wrong with me. Which was not right thinking since I knew that I had BHD. I think it was really because I was so young I just had never experienced any health problems and so I feel I mean I really couldn't have known any better I don't think because I had to go through that experience to really learn what it felt like but I would say to to not doubt as much. You know to listen to my body more. I really was disengaged with my body before this and now I'm really engaged with my body and how I feel, how I breathe you know. They're just very small tweaks that I do now and I am grateful for the experience because I think I have a better chance of having a healthier life because I pay attention now to my body. I really didn't before you know. I was kind of like push through the pain work through it you know don't be a wussy you know like um and that was that really wasn't healthy thinking you know for someone, or anybody, but for especially for someone who has BHD.

Jazzmin: What advice would you give someone now knowing all that? What would you say to someone who's just been diagnosed?

Paije: Well I would still not worry until there's something to worry about because there's so many symptoms so many things that could happen with BHD. I really appreciated my oncologist. After my first pleurodesis he said keep living your life you know, be strong, be healthy, test things out. If they cause you pain then don't do them but don't be afraid to do them you know and I think that I was a little bit shell-shocked you know for a good five years after having a rare cancer and a rare genetic disorder diagnosed. I was just like I'm not gonna live very long. It was fear you know and I just came to the point where I was just like you know what I might not but I'm gonna I'm gonna take that advice from my doctor and live, be healthy and be intentional about about what I'm doing both with my health and the rest of my life too you know so I think that's not really a perspective that a lot of 24 year olds, 25 year olds get that early in life. My main advice, I mean it's definitely better situations in other countries but here in America is it is so so important to prioritise healthcare making sure you have insurance. I will never be without health insurance and that is unusual I mean it might be hard to hear but there's many many young people in America who do not have health insurance and they they could pay for it but they don't but it's just not accessible the way it is in other countries. A lot of people go without it because they're healthy. Honestly that first surgery here in America if you look at the insurance bills and how much they charged for the surgeries and stuff I would have been financially buried for the rest of my life just after the first pleurodesis if I didn't have excellent healthcare. I don't want to step on anybody's toes or anything but I would say from from now on after your diagnosis it's not a

matter of if it's a matter of when. That's another thing that my doctor said and just to be prepared in that way. It's an investment in your health to make sure you have that coverage and there was another thing to um...Oh life insurance. These are things people don't think about. I was denied life insurance for 10 years because of the cancer so it's very possible that you won't have that problem with other BHD patients but I would say get life insurance as soon as possible before something happens not because you're going to die but because you might have to wait 10 years to get it again so I had to have a 10 years of cancer free before I could qualify for any life insurance again. So again for someone who has health problems you know in America life insurance is really important. You just don't think about stuff like that.

Jazzmin: We're based in the UK and we try to understand the American perspective as well and all the other countries but it's great to hear from someone who's actually lived it and the importance of getting it and the challenges of actually accessing something like life insurance if you've got something already. I think whoever watches this will learn a lot. What you've said is incredibly inspirational and it's amazing what you've gone through and look at where you are now. It's amazing to see you come here with a smile on your face and be able to talk about something that sounded really quite difficult with your first lung procedure.

Paije: Yes I was very fortunate and it was definitely one of the hardest things I've ever dealt with but I do have a very good life and I do consider myself to be a healthy individual and I'm grateful for that so I think it is possible to have you know a healthy life after even after symptoms start showing.