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“Suddenly, I was no longer ashamed of myself.”

Katri-Anna Lehto suffers from a rare disease causing excessive growth of keratin. When she was young she was ashamed of the way she looked and barely dreamt of a family of her own. A trip to Scotland changed it all.

Katri-Anna Lehto

Age: 37

Occupation: Stay at home mother

Family: Husband Mika, daughter Kerttu, 3, and son Reino, 1

Disease: Pachyonychia Congenita

Motto: Never give up hope



Three-year-old Kerttu is building a stone garden by the apple trees in the backyard of a red, wooden house. Earlier in the day they had been catching water lizards from a nearby pond with daddy and put little brother Reino down for his nap in the pram. Her mother Katri-Anna Lehto, 37, is resting her feet under an apple tree. Kerttu's enthusiasm is contagious, but her mother has just about used up her steps for the day. As much

as Katri-Anna would like to join Kerttu, she has to think of tomorrow. The pain in her feet increases as the steps accumulate, and the excessive steps taken today will be paid for tomorrow.

The disease in Katri-Anna's feet is called Pachyonychia (thick nails) congenita (born with). The name tells you what you see first looking at a patient: both finger and toenails are thick and hard due to excessive growth of keratin. But the worst part of the disease is the callouses and extremely painful blisters that cover the soles of Katri-Anna's feet.

Part of the nature of the disease is that you cannot predict the amount of pain you are going to be in. Today Katri-Anna may take 2000 steps, tomorrow maybe only 500, depending, for example, on what she has been doing and what the weather is like. Some PC patients use a wheelchair daily, but Katri-Anna only takes hers with her when she goes abroad or wants to spend a day in the city. At home it's easier to walk on her knees if her feet can't take the pressure.

Due to the pain caused by the disease, Kerttu, too, has had to learn that her mother can't run after her or follow her everywhere just like that. Her mother has to save her feet.

“When I'm on my feet, I desperately look for a place to sit and I cannot enjoy the moment. Fortunately the children have their father and other people close to them that can pull them in a sledge or take them for a walk in the woods,” Katri-Anna says.

Kolmivuotias Kerttu rakentaa kivipuutarhaan punaisen puutalon pihalla, omenapuiden katveessa. Vähän aiemmin on kalastettu isin kanssa vesiliskoja lähiesestä lammasesta ja nukutettu pikkuveli Reino vauvuunin rättyään alle. Äiti Katri-Anna Lehto, 37, lepauttaa jalkojaan omenapuun alla. Kertan innostus on tarttuvaa, mutta äidin askeleet alkavat olla läheltä päivältä käydyt. Niin paljon kuin Katri-Anna haluaisikin liittyä puutarhan tekoon, on ajateltava huomiosta. Kipu jaloissa yhtyy sitä mukaa kun askelia kertyy, ja tämän päivän liika-askelista maksetaan huomenna.

Sairaus, joka Katri-Annalla jalkoja pitäen, on nimeltään Pachyonychia Congenita, tuttavallisemmin PC ja suomeksi synnynnäinen paksukynnyisyys. Nimi kertoo suoraan sen, mikä ihmisestä näkyy myös päälle: sekä sormien että varpaiden kynnet ovat keratiinin liikakasvun vuoksi huomattavan paksut ja kovat. Pahinta ovat kuitenkin kovettumat ja äärettömän kivuliaat rötöt, jotka peittävät Katri-Annalla jalkoja.

Sairauden luonteeseen kuuluu, etteivät kivut ole ennalta arvioitavissa. Tähtään Katri-Anna saattaa yllättä kaiteutus-hoitanteen askeleeseen, huomenna vain viitteenastaan, riippuen vaikkapa siitä, mitä hän on tehnyt tai minkälainen ilma on. Osa sairastuneista käyttää pyörätuolia, mutta Katri-Annalla se on mukana vain matkoilla tai kaupunkikeskuksilla. Kotona on helppompi turvautua pehvikävelyyn, mikäli jalkapohjat eivät enää kestä painoa.

Sairauden aiheuttaman kivun vuoksi Kerttunkin on ollut opittava, ettei äiti pysty juoksemaan perässä ja tutkimaan pihan syrjäisimpiä kolkkia tuosta vain. Äidin jalkoja pitää säästää.

Jalkoilla ollessani katseeni etäji jo epätoivoisesti seuraavaa istänpaikkaa, enkä pysty nauttimaan hetkestä. Onneksi lapsilla on isä ja muita läheisiä, joiden kanssa he pääsevät pulkkailemaan ja metsäretkille, Katri-Anna sanoo.

NÄIDEN AVULLA SELVIYDYN

- Mies ja muut läheiset. He hyväksyvät, tuovat elämän merkitystä ja iloa, auttavat kukaan itsellään.
- PC Project. Vertaistuki ja toivo parannuskeinoista rohkaisivat hankimaan lapsia.
- Pehmeät pohjalliset ja sisäsovet. Ilman on ota askelkään.
- Auto ja irtaväpyskääntöluva. Anteeksi vaan, luonto.
- Suurimittaisuus ja priorisointi. Säästävät monta askelta.

- Lääkärit olivat neuvottomia. Kivulain muisteni lapsuudesta on, kun lääkäri poisti kaikki kynnet. Olin kahdeksanvuotias, mutta muistan sen tuskan yhä. Ei hoito tietenkään autanut, sillä kynnet kasvoivat piian takaisin, aivan yhä paksuina.

Koulussa Katri-Anna peitti kynnensä nykyin sisällä ja osallistui sinikkästä leikkien, vaikka kipu sai välillä kynnet silmiin. Piinaavimmat muistot liittyvät uimahallikylpyteihin, jossa jalkoja oli mahdotonta peitellä.

- Tunsin olevani todellinen friikki. Pelastukseni oli yksi hyvä ystävä, joka kulki rinnallani koko peruskoulun. Hän oli puoli-ikäinen, ja sovimme hyvin yhteen. Hän ajoi minulle tuolin, ja minä yritin pataus siihen kaukalevan korvaan.

Lukiokärsäni Katri-Anna alkoi käydä bareissa. Hän naamioi paksut kynnet kytensä kynsilakalla ja vaimensi kivun alkoholiin. Vanhukontun usko siihen, ettei kukaan voisi rakastaa häntä, kuitenkin pysyi.

- Päätin poika-avereidokkaiden puolesta olevani heidän miesten ystävä. En antanut kellekään tilaisuutta edes tutustua minun.

KAKSI UNELMAA Katri-Annalla kuitenkin oli: asua ulkomailla ja saada oma perhe. Ulkomaille hänet viivitti parikymppisenä ylempösten kielipöytä, joiden kautta Katri-Anna pääsi vaihtoon Portugaliin. Mukulajäädä, mäkainen maasto ja kun-

“Yli kuusi vuotta sitten en itsekkään tiennyt sairauteni nimeä.”

mus vaikeuttivat elämää, mutta kokemuksesta jäi enemmän hyviä kuin huonoja muistoja. Katri-Anna sai suosituksen vauvouden tunteesta, jonka sairaus oli hänet viennyt.

Urelna perheestä puolestaan tuntui pitkään kaukaiselta. Heinäkussa vuonna 2004 Katri-Annalla äänensä tapahtui kuitenkin jättyvä käänte, joka mahdollisti ensimmäisen hyväksymisen: löytyi nimi sairaukselle ja sitä myötä muut sairastuneita.

- Olin uskonut 30 vuotta olevani äitini kanssa ainoa omaltai- nen koko maailmassa. Sitten näin tietokoneen ruudulla kuvia muista katoistamme. Soitin äidilleni ja ihkinin, että “meidän sairauksella on nimi! Me emme ole ainoita.”

Selvisi, että kyseessä oli harvinaisen, mutta myös onneksen sairaus. Eridän sairauten lähisukulainen oli rikas lääkäri- nainen, joka oli lahjottanut omaisuuden lääketieteen tutkijoille, jotta nämä eivätsivät sairautelle hoitokirjoita. Nainen oli perustanut vain muutamaa vuotta aiemmin yhdistyksen PC Project, jonka nettisivu Katri-Annankin elämäni muutti.

KATRI-ANNA OTTI YHTEYTTÄ yhdistyksen yhteyshenkilöön ja kertoi mahdollisesti sairastavansa samaa tautia. Hän sai äitinsä kanssa kutsun vertaistapaamiseen Skotlanttiin, jossa y-

her and her mother's symptoms were caused by a mutation in the gene K16. The diagnosis of Pachyonychia Congenita was now certain.

"It was just unbelievable. Suddenly I was no longer ashamed of myself. Instead, I wanted to tell everybody about my disease. And for the first time ever, there was also hope for a cure to be found some day."

Finding a support group was the final push Katri-Anna needed to reach the other end of her dreams. Already in the previous spring, Katri-Anna had started seriously working on her self-esteem. She read numerous self-help books and tried everything from Feng Shui to visualization. Three weeks after the trip to Scotland, on Mika's day, she finally got enough courage to place a profile on an internet dating site. That same day, she received a reply from an interesting man from the Turku region, named Mika. Katri-Anna decided to reply.

"In my third message to him I added a link to PC Project's website and told him I understood if he no longer wanted to keep in touch with me. To my surprise, he did not comment on the way the disease made the patients look at all, but instead told me how sorry he was that I was in pain. I felt this was a guy I could meet some day."

On their second date Katri-Anna asked Mika if he would like to see her feet. He stroked her feet gently and said, "Oh, poor foot." Katri-Anna is still moved by the memory.

"From then on, it was clear to me that he accepted me the way I was. He proved my belief that nobody would ever love me wrong."

Two years after their first date, the couple bought Mika's childhood home in the country and got married. The following year, Kerttu was born. Now, in the backyard of the house built by Mika's father, there is Kerttu's stone garden, but also Katri-Anna's little herb patch – right next to the door to save her some valuable steps.

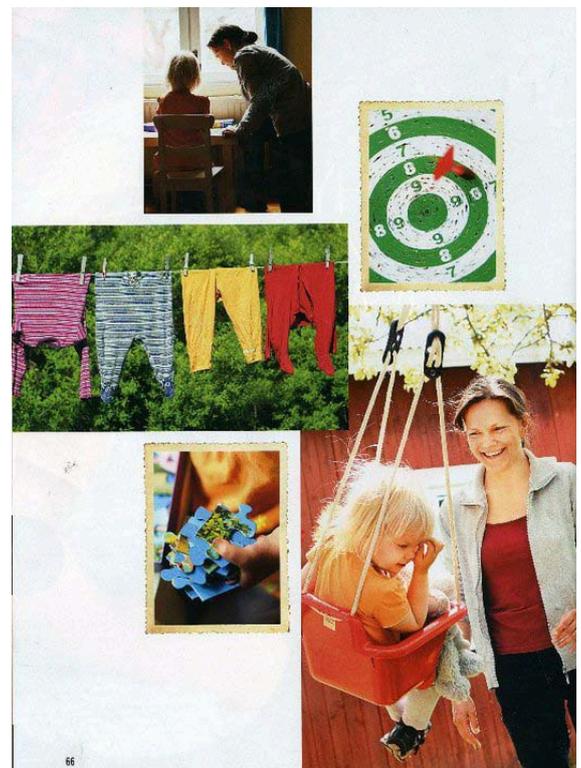
"From the beginning, Mika tried to make my life easier in my new home. He changed the gravel in the yard to fine sand and removed the knotted rugs that made walking difficult for me. He also acts as my feet – he'll carry me over the roots in the forest if I can't make it myself and all. He is a kind man, a true gem."

As a biologist, Mika has special interest in his wife's rare condition. He is the one in the family that knows how to explain about the mechanism behind the pain in the disease, and also understands how difficult it is to find a treatment that actually works. Family life, on the other hand, has not been influenced by his wife's condition in any significant way.

"We lead a normal life. You just have to take into consideration the other person as an individual, as you do in life anyway," says Mika.

"We all have our problems," adds Katri-Anna. "When I was younger I could spend ages contemplating my own misery and I even managed to create myself a coffee cup neurosis. As I've grown older, it's been easier to remember that things could be worse."

Instead of complaining, Katri-Anna concentrates on making her life easier and saving her valuable steps. A disability parking badge, lists written on a daily basis and an easygoing attitude towards life take her a long way. Persistence has been helpful, as well.



“When I want something bad enough, I get it done. Fortunately, I am also somewhat lazy. It doesn’t bother me so much if things don’t get done because of my feet.”

Life in the countryside seems nice and peaceful. Katri-Anna is at home with the children, and Mika, too, works at home as an entrepreneur. Walking after the children makes Katri-Anna’s feet hurt and shrinks her circle of life even more, but she would not change the life of a mother for anything in the world.

“The fact that my dream of a family was fulfilled is, to a great extent, thanks to the patient support group. From early on, I will try to teach Kerttu the attitude that this is just a part of us and doesn’t make us any worse than others.”

If, and when, a treatment for the disease one day becomes reality, Katri-Anna will enroll in dancing lessons. She can catch some of the joy of movement now in the summer in the surroundings of her house: sitting on a bike with the saddle low, she can take pain free steps and let the wind blow in her hair. Just take a look at what’s happening around the corner.



An ultra rare gene mutation:

- Pachyonychia Congenita is a very rare skin disease caused by a mutation in one of four Keratin genes. The mutation can be spontaneous or inherited from a parent with PC. The chance of passing the disease on is 50 %.
- The symptoms vary, and depend, among other things, on which one of the genes is affected. The most common symptoms are thick nails, callouses and blisters on the soles of the feet and palms, and plantar pain. There may be excessive keratin in the oral mucosa, and different types of cysts and bumps may appear around the body.
- There is not yet a treatment for the disease, but keeping feet cool, removing the excess keratin and rest may relieve the pain. Pain killers only offer temporary relief.

PC Project is a charitable organization that collects funds and coordinates the research of Pachyonychia Congenita for finding a cure for the disease. More information can be found at www.pachyonychia.org.



These help me survive:

- My husband and other loved ones. They accept me the way I am, bring meaning and joy into my life and all help in their own ways.
- PC Project. The existence of a support group and the hope for a cure encouraged me to have children, too.
- Soft insoles and slippers. Without them I don’t take a single step.
- Car and disability parking badge. Sorry about that, nature.
- Planning and prioritizing save me many steps.

