ISSUE NO.1 / SPRING 2013

!NSP!RE

the magazine of the BCNF

LONG DISTANCE LOVE: CONNECTED THROUGH NF

OUT OF THE SHADOWS: **THE FACES OF NF**

AN NF CLINIC FOR BC?

NF RESEARCH AT B.C.'S CHILDREN'S HOSPITAL

EDITOR'S NOTE

You probably notice that the BCNF newsletter has a whole new look.

Our vision for this revised and expanded newsletter was to create a publication that would inspire our readers, our families, and our communities.

I have been so inspired over the years by the incredible people I have met living with NF who are living exceptional lives despite the disorder, that I was eager to share their stories with the world!

When we are inspired our consciousness expands, our spirit lifts, and we feel more joyous, energized, and dream bigger dreams than we ever had for ourselves. If the stories in this magazine make you feel this way for even a few minutes than we have done what we set out to accomplish.

This publication has been a long time in the making and would not have been possible without the individuals whose stories are featured in this premier issue. I thank them for graciously allowing us into their lives so we could shine their light onto others.

Taking an idea and making it a reality requires the multiple talents of an incredible creative team, which I have been fortunate to work with. A heartfelt thank you to the brilliant Matt Politano at Oculus Design + Marketing for the design, Cara Grimshaw, for creative photography, and Sarah Gordon for superb interview and writing skills.

This magazine is for you. We are eager to receive your feedback and ideas for future articles. If you have an inspirational story or want to share something that has made a difference to you and your life with NF please get in touch. We are eager to hear from you.

Wishing you much joy,

Desirée Sher
Executive Director
desireee@bcnf.bc.ca

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EDITOR
Desirée Sher

CONTRIBUTORS
Sarah Gorden
Cara Grimshaw
Kimberly Jett

FOR FURTHER INFORMATION:
BC Neurofibromatosis Foundation
Box 5339, Victoria, B.C.
Canada V8R 6S4
Toll Free: 1-800-385-2263
Email: info@bcnf.bc.ca
Web: bcnf.bc.ca

Look for us on Facebook & Twitter

Together we can make a difference!

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The BCNF would appreciate your feedback on our magazine and content. Please take a few moments to complete the online survey and let us know how we are doing:
www.surveymonkey.com/s/CVRS8P3

THE FACE OF NF

SHINING A LIGHT ON MEMBERS OF THE NF COMMUNITY

FOTO SUKANEN

Until Foto Sukanen became involved with BCNF, she had never met anyone else with NF. As one of the founding members of BCNF, back in 1984, Foto has recently re-joined the BCNF Board of Directors. Foto recalls that in 1984 she heard that there was a group forming for those with NF and their family members. "I had gone to genetic counselling in regard to family planning, and called one of the contacts and got involved." Foto is getting close to retirement, so in September of 2012 she decided to re-join the BCNF board. She now has some free time and energy to devote to NF. Foto has always been a silent supporter of BCNF, but looks forward to getting actively involved once again in fund-raising and wherever else she can be of assistance.

Foto was diagnosed with NF1 in 1962 when she was six years old. Prior to her diagnosis, her mother took her to the cancer clinic in Saskatoon, Saskatchewan to keep an eye on a tumour on the left side of her nose. The doctor referred her to a plastic surgeon for removal of the tumour. When asked how NF has affected her life, Foto says that it had a major impact on her decision to not have children. Aside from two surgeries to remove the tumour from her nose and several reconstructive surgeries, her NF has not been a major concern. Foto has several signs of NF1, but none are severe. "I have high blood pressure, slight scoliosis, café-au-lait spots, liche nodules in my right eye, a slightly larger head, and short stature," tells Foto. Foto is concerned about how her NF might be affected by aging. "I have noticed that the nodules on my midriff are increasing in number," says Foto.

Aside from NF, Foto has been with her soul mate, John for 34 years. "After 34 years, we can still sit and talk for hours," tells Foto. "Our piece of paradise is Long Beach on Vancouver Island. We try to get there every couple of years." Foto enjoys cooking/baking, beading, and crafting. She hopes to get back into sewing once she is retired. Foto also has a deep love for animals. "I have always had my furry kids. The current count is four cats and no dogs," says Foto. "Our young grey tabby is called Yogi. Yes he does yoga poses," Foto humorously says. Foto also has a white and tabby male called Woody and a black elderly girl called Spike. "I look forward to adopting a rescue puppy once I have retired," shares Foto. •

ONE FOOT IN FRONT OF THE OTHER

Fifteen year old Robert McNaughton has a passion for running. Much like his love for running, he lives life putting one foot in front of the other and never gives up until he's crossed the finish line.

Robert was diagnosed with NF1 when he was only a year old. In elementary school, Robert participated in cross country running. Despite weakness in his left leg, he gave it his all and never gave up, even if it meant coming in last place. "He never quit running until he crossed the finish line," recalls his father Dave. Robert is now fifteen years old and continues to have a passion for running. "He practiced very hard on his own time and now in his grade ten year he is consistently in the top three in his class," says Dave. Robert also enjoys taekwondo, and despite being held back a year, he persevered and is gradually moving up in his belt class.

Robert has had fourteen major surgeries due to his NF1. "They have caused much discomfort and time away from school due to recovery," says Dave. When Robert was younger he had trouble with physical activity which limited him in playing certain sports. "His symptoms include a nodule under the skin below his right eye, numerous café-au-lait spots all over his body, and the right side of his face is slightly bigger." Robert also has weak vision in his left eye and dental problems related to NF1.

Robert has a wonderful support system in his parents and grandparents, who are there for every surgery and are also very involved with BCNF. "Through this organization we were able to obtain much information, guidance and comfort when Robert was diagnosed with NF1." Robert and his family raised over $4,000 in just two weeks for the BCNF Scotiabank Charity Challenge where Robert ran the 5 km event. "Robert wrote a personal letter explaining a little bit about his illness with a picture of himself attached asking for donations towards research in finding a cure." Dave circulated the posters at his workplace, CN Rail. "The response and generosity of my fellow employees was overwhelming," recalls Dave. Over $2,500 was donated by employees at CN Rail. The McNaughton family also circulated the posters to friends, neighbours, and at Robert's school where generous donations continued to pour in.

In addition to running and taekwondo, Robert enjoys travelling to Pokemon tournaments across North America, playing video games, cooking, the scouting program, attending his church's young adults group, and graphic design. "Even though I have had many surgeries since I was a young child, I never let the recovery time discourage me," says Robert. "When I am fully recovered, I continue doing the things I enjoy." •

"HE NEVER QUIT RUNNING UNTIL HE CROSSED THE FINISH LINE"

WORDS BY **SARAH GORDEN**

ROBERT MCNAUGHTON

PHOTOGRAPH BY **CARA GRIMSHAW**

FREQUENT FLYER

Karen Thornton loves to fly. For her fiftieth birthday while on vacation in Lumby, B.C., Karen's husband, Ken bought her a glider flight. "What a trip that was!" explains Karen. "When we landed, I couldn't feel my feet on the ground I was so high!"

But that was only the beginning. Five years later, she was taken up in a motorized hang glider in Hope, B.C. Then, for her sixtieth birthday, her daughter gave her a flight in a hang glider. "The feeling of total freedom I get is indescribable!" exclaims Karen. Next on her list of things to try is paragliding.

Things have not always been carefree for Karen. When she was 18 years old and pregnant, doctors diagnosed Karen with Neurofibromatosis Type One (NF1). Unfortunately, Karen's daughter, Kim, was also born with NF1 and had to have a tumour removed from her mouth shortly after birth. Karen also has a son, Dan, who did not inherit NF1.

Karen enjoys spending time with Dan's two daughters, who are five and two. Karen is a spontaneous mutation, as her parents and siblings do not have NF1. Karen has lived with tumours on various parts of her body since childhood, but the pregnancy made the tumours grow and new ones to appear on her face and neck. "The biggest challenge for me was the fear of meeting people and not being accepted due to the visible tumours," says Karen. "My interest in motorcycles and motorcycle riding (as a passenger only) is what enabled me to overcome my self-image hang-up."

Karen and her husband bought their Honda Gold Wing motorcycle in 1990 and rode for a couple of years before they joined the Gold Wing Road Riders Association (GWRRA). "I was accepted for who I was," says Karen. They became provincial directors in the organization and met hundreds of people. According to Karen, "I had never had so many people wanting to hug me!" To many people, owning a Gold Wing means buying chrome, lights, and accessories for their bikes. After becoming friends with the owner of a bike shop in Bremerton, Washington, Karen decided she would like to start her own business. She now runs a small, home-based business selling motorcycle accessories called Karen's Krome. You can visit her website at *karenskrome.com*. •

WORDS BY **SARAH GORDEN** / PHOTOGRAPH BY **CARA GRIMSHAW**

HOW SWEET THE SOUND

To Sarah Gorden, being able to hear her husband say "I love you" is a miracle.

Four years ago, Sarah had to undergo a surgery which took her hearing. Living with Neurofibromatosis Type Two (NF2), she had tumours on both sides of her brain, affecting both her hearing and balance. When the larger of these two tumours had to be removed, she had to sacrifice her only remaining hearing. Stepping into a now silent world was difficult, but there was hope.

"I connected with other NF2 patients through an online support group called the NF2 Crew," Sarah explains. "It was through this group that I learned of the House Research Institute (HRI) in Los Angeles, California." The House Research Institute specializes in disorders affecting the ears and hearing. They are one of very few centers in the world that specialize in NF2. At HRI, they have developed a specialized hearing implant called an Auditory Brainstem Implant (ABI). The implant is specifically for people with damaged auditory nerves. When the NF2 tumours are removed, the auditory nerve is destroyed. Devices like hearing aids or cochlear implants are no longer an option. The ABI bypasses the auditory nerve and stimulates the brainstem to restore some sense of sound.

The ABI was placed at the same time as the tumour was removed. Two months after her surgery, Sarah returned to Los Angeles to have the device activated. "When the audiologist flipped my ABI on and began talking... *'Testing one, two, three, popcorn, baseball, hot dog'* I was amazed that I COULD HEAR HIM!" Sarah recalls. "At first, the sounds were artificial sounding and peoples' voices sounded like they had been inhaling helium. With time, this got better and everything started to sound more natural." With a lot of practice, Sarah now does very well with her ABI. "Although it did not restore my hearing, I am able to hear things that I never thought I would get back. I am especially grateful to be able to hear my husband's voice and am relearning how to hear some music."

Unfortunately, during the surgery, Sarah's right facial nerve was traumatized, leaving her unable to move half of her face and to smile. Having her ability to smile and show emotion taken away has been very difficult for her to cope with. "Despite it all, I try my best to face each day thankful for what I still have."

Since 2008, Sarah has been participating in an NF2 natural history study at the National Institutes of Health (NIH) in Bethesda, Maryland, USA. The purpose of this study is for the researchers to learn more about NF2 in hopes of finding better treatment options.

Sarah has many other tumours throughout her brain, spine, and peripheral nerves and has undergone numerous surgeries.

Sarah has a love for animals, especially her beloved dog, Bailey and her two cats, Oliver and Maddie. "They bring me a lot of joy and help me cope on the rough days." She enjoys graphic design, photography, the outdoors, exercising and spending time with her husband, Hans, family and friends. She volunteers her time as the graphic designer for AdvocureNF2, a non-profit organization advocating for the NF2 community. Sarah tries to live life to the fullest, one day at a time and her motto is "Never give up hope!" •

WORDS & PHOTOGRAPH BY **SARAH GORDEN**

NEVER GIVE UP

JESSICA & ELISSA

It's the motto that many patients with NF2 live life by. This certainly holds true for the Viitanen family. Twenty-four year old Jessica and 20 year old Elissa of Delta, B.C., both have NF2 which they inherited from their mother Kaarina.

Since they had a known family history, the doctors knew to check the girls with MRIs, and sadly they were both diagnosed as children. "Some people have assumed my sister having NF2 made it easier to deal with because we could relate to each other's experiences and we wouldn't have to deal with it alone," tells Jessica. Jessica explains that it is very difficult knowing her baby sister has tumours and will end up deaf one day. "I'd do anything to bear that burden for her."

Jessica and Elissa's mother, Kaarina, also lived with NF2. She passed away when Jessica and Elissa were small children, ten days after celebrating her thirtieth birthday. "She had dozens of tumours all over her spine and a big one in her brain that eventually killed her," tells Jessica. She didn't let that stop her from living her life though. Despite being fully deaf, Kaarina still played the organ at church. She had a friend type the sermons on a laptop so she could follow along with the church service. The same person still types for Jessica and Elissa at church. Kaarina exchanged long letters with a close friend from the United States for many years. After she passed away, the letters were published into a short book which the girls can now look back on in memory of their mother. "One thing I noticed in the book was that she never mentioned NF2 and didn't worry or complain about her situation," tells Jessica. "Instead she wrote about all the good things in her life and the things that brought her joy, like Elissa and I." She lived her life as a person who had NF2 but did not let the condition define how she lived her life.

Jessica, who recently finished schooling for applied business technology, explains the challenges of living with a hearing impairment. "It's put up a barrier between me and the rest of the world," tells Jessica. "The last job interview I had lasted less than two minutes. After the employer learned that I couldn't hear, he refused to write down what he wanted to say and wasn't willing to speak a little louder," tells Jessica. She explains how difficult it is to have self-confidence in moving ahead in life and gaining employment when so many people are not willing to put in the effort or be accepting of differences. Since NF2 patients tend to lose their hearing gradually, the change from the hearing world to the deaf world happens slowly. "For me, this means I still think like a hearing person, when in most situations I might as well be deaf because I can never understand anything," tells Jessica. Jessica, who is severely hard-of-hearing, explains that it would almost be easier to be completely deaf. "Of course I'd miss the sounds of rain falling on the roof, snow crunching under my feet, and birds tweeting in the spring."

Jessica loves to renovate and build things. "I'd love to go to Mexico with a group one day to help build small houses or classrooms for less fortunate communities," says Jessica. Elissa recently finished culinary school and is working as a baker in Vancouver. Jessica and Elissa are carrying on their mother's legacy and will never give up! •

WORDS BY **SARAH GORDEN**

SCOTIABANK VANCOUVER HALF MARATHON

RAISING FUNDS FOR RESEARCH: JUNE 23, 2013

Join us as we walk and run for NF Research. The BCNF is putting together a team of fun and energetic individuals to run or walk the half marathon or 5km event on June 23. Our team has a few members but we need you, your family, your coworkers and neighbours, to help us make this event a real success!

Last year's half marathon and 5km walk/run events raised $14,000. The event requires runners and walkers to raise pledges and then come out to complete one of the most scenic routes in Vancouver.

Set the goal to raise a minimum of $200 and your registration to the event is FREE. If you aren't eager to do the fitness part we also need volunteers to cheer on our participants and host the information booth on event day.

IT'S EASY TO JOIN!

Contact the BCNF at info@bcnf.bc.ca and let us know you want to join the team. We will send you a promotional code that will allow you to register FOR FREE!

Together, we do make a difference for those living with NF but we can't do it alone – we need YOUR support!

If you are searching for some reading material that will inspire you to excel, become a better person, or even change the world, then check out these resources.

You Can Heal Your Life
Louise Hay

A Long Walk to Freedom
Nelson Mandela

Man's Search for Meaning
Viktor E. Frankl, Harold S. Kushner

Always Looking Up: The Adventures of an Incurable Optimist
Michael J. Fox

The Long Run
Matt Long

A Beginner's Guide to Changing the World
Isabel Losada

Learning to Breathe
Alison Wright

Stroke of Insight
Jill Bolton Taylor

Seven Thousand Ways to Listen
Mark Nepo

The Alchemist
Paulo Coelho

Long Distance Love

GAIL AND TIM HAVE SO MUCH IN COMMON, but it was fate that brought them together. "NF1 helped me find my soul mate and one true love," explains Tim. On August 11, 2010, Tim Golumbia, who is a Social Worker, was asked to assist with a task at work that he doesn't normally do. This task involved a phone call to an Edmonton office. In the conversation with the worker in Edmonton, the topic of NF1 came up and the worker shared with Tim that she had a co-worker with the same condition. "She asked me if I was single and told me she was cupid," Tim recalls. Tim explains that this worker has a history of trying to find a man for Gail. Gail called Tim and left a message, and when they finally managed to connect, it was at the end of the day on Friday, August 13th. "We only talked for a few minutes, as she was meeting friends for dinner," says Tim. "She called me again on Sunday and we talked for around an hour and a half. It was like WOW, she is amazing and we have so much in common," Tim explains.

Tim and Gail both have Neurofibromatosis Type One (NF1). Tim was diagnosed when he was around ten years old. "I did not have a lot of bumps until I was older, but I did have other signs," says Tim. Tim went through puberty earlier than most and therefore was much taller than his classmates. He also had two growths removed from his left arm and one from his abdomen which led to a confirmation of a diagnosis of NF1. Tim explains that he was teased a lot in school, especially when changing for swimming or gym class. "Going shirtless was difficult for me because I was very self-conscious about my body." Tim also has a learning disability due to NF1 and it made school more difficult for him. "I am actually quite bright, and looking back I feel it was just a matter of learning how I learn," explains Tim. "As a kid it was so difficult to get my thoughts onto paper, as my mind runs so much faster than I can produce hand written documents." Computers helped change this for Tim as he could properly organize his thoughts and use spell check. Dealing with peers was difficult for Tim and he was treated quite badly. "I never had a lot of friends, but the friends I had were close," Tim explains. "Despite being treated badly by my peers, I was a very giving person. I did volunteer work in nursing homes and had a number of friends with physical disabilities. I learned so much from these experiences," Tim recalls. After high school, Tim had great difficulty finding his spot in life. He tried many different careers before returning to University for a degree in Social Work. "My decision to return to University was prompted by a friend who had been through a very abusive childhood but was moving ahead with her life," explains Tim. "I decided that it made sense for me to try again." Today, Tim uses his NF1 and bumps as a tool to "break the ice" when talking to the kids he works with. "They always look and ask questions, so I use that as an opportunity," explains Tim. It was because of this that he ultimately met Gail.

Gail Appelgren has a similar story to tell of the difficulties growing up with NF1. "I was born with an NF tumour on the left side of my chest, where my breast should have been," explains Gail. "This tumour grew as I aged. I was teased as

a child and called names such as lopsided." Gail was under the care of doctors in Moose Jaw, Saskatchewan. "The doctors in Moose Jaw in the 1960's didn't know what it was and since it didn't seem to affect my health, my aunt, who was raising me, didn't feel the need to seek out other professional opinions," says Gail. "When I was about twelve years old, I saw a new doctor who knew immediately what it was. He told me what I could expect from having Neurofibromatosis and talked to me about seeing a plastic surgeon when I was an adult." Gail explains how difficult it was growing up with NF1. "As a teenager it was difficult to have a relationship with a boy as they just wanted to try and see what was under my shirt. When I hit puberty, my body started to develop more. I developed many smaller tumours all over my body," Gail shares. "People who didn't know me were afraid of me. They didn't want to sit beside me on a bus. I was asked to leave a swimming pool and not to touch the tomatoes at the grocery store," Gail recalls. Although she was often saddened by people's reactions to her, Gail just wanted people to know she was just like them. "I just happened to look a little different or special," explains Gail. "I would just try and talk to them and give them one of my big smiles," says Gail. Gail feels that living with NF has made her more accepting of others who are different. "It has made me more caring, forgiving and tolerant. I am always looking at the positives in every situation, always having hope and always making the best out of whatever I am faced with," Gail explains. Gail deals with a lot of ignorant people who assume she is unable to care for herself. "They are often surprised to hear that I have two University degrees and I live on my own," says Gail. "I always see this as an opportunity to educate individuals on Neurofibromatosis."

Tim and Gail met in person for the first time in November of 2010 and it was confirmed, "We had a relationship starting," Tim says. When asked to describe their relationship, Tim says, "It is amazing. I have been in relationships before but have never felt such a connection with the other person." Tim explains that being with another person with NF helps in that they both understand how the disorder affects each other. "Gail is a warm, caring, thoughtful, loving, and polite person," Tim says. He loves her eyes, smile, and hearing her laugh. "Being in a relationship with Gail means having a relationship with three dogs as they are like her children," Tim says. "I love spending time with her 'kids' when I am in Edmonton."

Tim and Gail are now engaged to be married. When asked to share their proposal story, Tim shares "In April of 2012, Gail was arriving at the airport in Comox and I went to pick her up. I had told her to make sure she wore good shoes as we would be going for a walk at Cathedral Grove on our way back to Nanaimo. The massive old growth forest is amazing and we had been there before. It was a cool but sunny day.... quite nice. Gail went off the beaten path at one point and I followed. I tried to get her to turn around at one point and she walked further into the clearing. I walked up to her and she turned, I said "Gail..." and she looked at me and said, "Are you sure you want to do this?" I started to say the first verse of John Denver's "Annie's Song." She loves John Denver's music. Between the first and second verse I asked the question and gave her the ring. While she was on the phone with our matchmaker in Edmonton, I noticed another couple had walked into the area where we were standing... so I asked if they could take our picture. We continued our walk through the trees, hand in hand."

Tim, who currently lives in BC, plans to move to Edmonton to be with Gail. He is currently in the process of finalizing employment in Edmonton. They will make wedding plans once Tim is in Edmonton. Tim and Gail's story proves that everything happens for a reason. Although they have both had their fair share of struggles with NF1, it is ultimately what brought them together. Tim is currently the president of BCNF. "I will miss BCNF when I leave the province but I will maintain contact and continue to support them financially," Tim says. •

WORDS BY **SARAH GORDEN**

DARING TO DREAM

WHERE IS THE DEDICATED CLINIC FOR NF?

ARMED WITH A LIST OF QUESTIONS, THE PHILLIPS FAMILY WALKED INTO THE NF CLINIC. FOR THE FIRST TIME SINCE THEIR SON'S DIAGNOSIS, AN EXPERT – A PHYSICIAN FAMILIAR WITH THIS COMPLICATED MULTI-SYSTEMIC DISORDER – WOULD EXAMINE HIM.

They had completed the pre-screening questionnaire, forwarded all reports to the clinic coordinator, and were ready and excited for an appointment they had waited ten years for.

While many patients require no more than an annual neurological evaluation and imaging, or a hearing evaluation, others who visit the clinic have ongoing complex medical issues that require a visit to one of the physicians in the NF network. This network includes specialists in child life, nutrition, neuropsychology, plastic surgery, otolaryngology (ENT), orthopedics, audiology, dermatology, ophthalmologists, oncology, radiology, and occupational therapists. The clinic is situated within a major health center In Vancouver and makes regular tips to Victoria, Prince George and Kelowna.

THIS IS THE DREAM.

Currently there is no coordinated care clinic serving the children and adults struggling daily with the effects of neurofibromatosis, but the BCNF is on a path to change that.

The NF clinic is a subject the BCNF office receives countless inquiries about. "Who is the NF expert?" "Where is the NF clinic?" are the calls from families desperately seeking a health care professional familiar with the disorder. And it's not just families seeking the experts. Questions also come from hospital-based professionals who are transitioning patients back to the community. They too are seeking the NF expert to offer a continuation of specialized care. It pains me to tell them that what they are looking for just doesn't exist, and we do it every week.

At the BCNF we believe its time to create a clinic for the children and adults living with NF1 and NF2. While less common genetic disorders such as cystic fibrosis and Huntington's disease have specialized clinics, the NF community is left unsupported. Many families have shared that their family doctors look to them for information. This adds stress to sick patients who find themselves educating the one caring for them.

BCNF will need to consider how it can develop NF expertise within the medical community. One way this could be addressed is by identifying professionals who have an interest in the disorder and then fostering their expertise and skills by sending them all the patients with NF. We'll make them the "go to" expert. The more patients the specialist sees with NF the greater their knowledge and skillset will become. In turn this will provide improved health outcomes for the patient.

Family doctors also need education and increased confidence to care for the NF community. Working to develop a network of GPs familiar with NF would also improve care options for families closer to home. One idea that was generated at a meeting with Drs. Jan Friedman, Linlea Armstrong and Patricia Birch, from B.C.'s Children's Hospital, was to host a lavish half-day conference on NF focused specifically at these general practitioners. A costly endeavor but an idea we will pursue.

I got home from my holiday to find a letter from my GP releasing me from his care. He no longer wanted me as a patient as he knew too little about NF to provide adequate care, and he didn't have time to learn. ***Who will take care of me now?***

At the BCNF we are not strangers to leaps of faith. We believe with enough vision, passion and perseverance we can make all our dreams come true, including the clinic, won't you join us?

if you have a comment about the clinic project please contact us at *info@bcnf.bc.ca* •

Nicole

DREAMS
FOR AN NF CLINIC

IN THE FALL OF 2012, NUMEROUS INDIVIDUALS WITH NF1 AND NF2 AND THEIR FAMILIES CAME TOGETHER TO SHARE THEIR DREAMS AND HOPES FOR AN NF CLINIC. HERE ARE THE HIGHLIGHTS OF THIS SESSION.

I WISH FOR...

*A place where great thinkers would come together and there would be research discoveries, which offer **hope***

A clinic which housed a resource library for information so parents don't have to turn to the web after diagnosis

A clinic where the NF experts would be identified and would be a resource for community physicians who want to learn more

*A **go-to** place for families*

A place where a life plan for the patient would be developed reflecting the ages and stages of NF; provide information for the future including tests and procedures

A place where families can find help to navigate the medical system, coordinate care, and get help to formulate the questions that need to be asked of doctors and surgeons

A place where testing and psycho educational assessments are understood and could be found

A clinic would have a nurse or genetic counselor who works with families to coordinate care and advocate during times of transition, i.e. initial diagnosis, going to school, transition out of BCCH and into adult care

A DIFFERENT KIND OF DREAM HOUSE

WORDS BY **SARAH GORDEN** / PHOTOGRAPH BY **CARA GRIMSHAW**

JENEECE EDROFF IS A NINETEEN-YEAR-OLD philanthropist from Victoria, B.C. who is known to many on Vancouver Island as "the Penny Girl" for her fundraising efforts – she has been credited with raising more than $1.5 million through penny drives, beginning at age 7.

Jeneece has Neurofibromatosis Type One (NF1) and has undergone multiple surgeries due to the condition. Despite her struggles, she has always been determined to help others facing similar hardships.

Jeneece's latest fundraising accomplishment was to create a home-like facility called Jeneece Place for families enduring medical treatments in Victoria. Dealing with a childhood illness is stressful for the entire family. Having a home away from home while undergoing medical treatment helps ease that stress for families.

Jeneece's mother, Angie Edroff says, "Over the years when traveling to Vancouver for Jeneece's medical care we spent a lot of time at the Ronald McDonald House. There were times we could not get a reservation there because the house was full. It was at these times that Jeneece mentioned that maybe we should help them build a new Ronald McDonald House in Vancouver."

The Edroff family tried several times to make that happen, but they kept running into obstacles along the way. "When talking with some folks in Victoria, they asked Jeneece what she wanted to do next. She spoke about her wish to build a new Ronald McDonald House in Vancouver and the roadblocks. It was then that we learned that there was a need for such a house right here in Victoria."

To Jeneece, building a home in her own community that would serve the people on Vancouver Island and the Gulf Islands was far more than she could have dreamed of.

Jeneece Place opened its doors in January of 2012. It is a comfortable "home away from home" for families facing medical treatments at Victoria General Hospital. Jeneece created the floor plan of the house, for all three levels. The architect took those plans and did his best to put what she wanted into the house. This included a movie room, games room, and an arts and crafts room. The house is a 10,000 square foot, ten-bedroom facility. It has a very welcome and peaceful environment along with the many comforts of home. The house is decorated with artwork donated by local schools and artists, including a giant penny that Jeneece helped sculpt.

//BUILDING A HOME IN HER OWN COMMUNITY THAT WOULD SERVE THE PEOPLE ON VANCOUVER ISLAND AND THE GULF ISLANDS WAS FAR MORE THAN JENEECE COULD HAVE DREAMED OF//

Jeneece is an ambassador in the house while a housemother is in charge of the house and all the volunteers. "On holidays we go and cook for all the families that are in the house at that time," says Edroff.

The house took nine months to build and was built with the help of more than 2200 gifts, from a few dollars to $1 million. The Edroff family is looking to build another house on the island by the hospital. "Jeneece Place is now full more than fifty percent of the time so there is a need to build another one". Jeneece also has dreams to build a house in Nanaimo. In addition to her fundraising for Jeneece Place, Jeneece has been involved in fundraising for the Easter Seal's 24-hour Relay, the BCNF, BC Children's Radiothon for the Kids, Cops for Cancer and is a Junior rider in the Tour de Rock.

Jeneece finished high school in June, 2012. She would like to become a Child Life Specialist and plans to help out at Camp Goodtimes, a camp for kids living with cancer, this summer. She also has dreams to volunteer at the local hospital. This fall Jeneece wants to start at Camosun College to further her learning. "She likes to play guitar, and is learning the ukulele," says Edroff.

Jeneece continues to face medical challenges due to her NF and will soon be undergoing yet another surgery, this time at the Mayo Clinic in Rochester, Minnesota to remove a plexiform neurofibroma from her right leg.

For more information on Jeneece Place or to make a donation, please visit **jeneeceplace.org** •

SUMMER CAMP

ADAM NIXON IS A SEASONED camper. For four years, he has attended Camp Kostopulos, the annual NF summer camp, located just outside Salt Lake City, Utah. He loves every minute of it. "My favourite part is seeing people with NF and how they cope with it. I saw many people worse off than myself. It was both sad and interesting," Adam says. Last summer, Adam went to the camp as a junior counsellor. "My duties were mainly to help out and take on a leadership role in activities," explains Adam. Adam really enjoyed being a junior counsellor and says that if he goes back to camp,

WORDS BY **SARAH GORDEN**

he'd like to take on that role again. Adam says that it was BCNF that initially suggested that he step into the leadership role as a camp counsellor. "I enjoy helping people and have a big interest in psychology," Adam says. Adam's favourite memory from camp is helping a child named Chris. "Chris is a very good person and it was his last year at camp. I got all the counsellors to sign an award for him and all the campers gathered around to present it to him," Adam recalls. "He was so happy he cried. When we went to the airport, it was the first thing that he showed his mother." By participating in the NF Camp, Adam gained perspective on how NF affects everyone regardless of their race, gender, culture, or religion. "I also gained patience," says Adam. "I needed a lot of patience as a camp counsellor." Adam says that the most fun part about camp is the trip to a large waterpark that they go to each year. When asked what Adam would tell kids thinking about attending camp, he says "COME!! JUST GO!!! You will meet others just like you and it will help you grow. People are nervous the first time they go, but believe me, it is worth it. After attending once, you'll be counting down the days until next year!"

Adam was diagnosed with NF1 when he was just six months old. He is now seventeen years old and lives in the small town of Sherwood Park, Alberta just outside of Edmonton. He has had one surgery to remove a tumour from his lower back. "The surgery went well, but the tumour was bigger than the surgeons expected." The tumour that was removed was the size of a football and weighed seven to ten pounds. Adam says his recovery went great and the only sign left from the surgery is the scar. NF1 has impacted Adam's life in many other ways. When he was a child, he had difficulties with his balance and could not ride a bike. Adam also suffers from dyslexia and struggles with math. He also has scoliosis and endures painful migraines on a regular basis.

Adam enjoys reading, watching TV, playing the card game Magic the Gathering, and watching anime. "I have a few life goals for myself. I'd like to get my doctorate in scientific psychology specializing in dreams," explains Adam. He'd also like to write a beloved teen novel series and become a famous actor. Adam is a grade eleven student at Salisbury Composite High School where he is in the advanced placement program for Social Studies. "In addition to my school's acting classes, I am the president of Salisbury Human Rights and Gay Straight Alliance Club." To sum up his life, Adam simply states "NF is a part of my life, but not a defining factor." •

2013 CAMP JULY 13-19

Last summer almost 80 campers from all over the U.S., Canada and the U.K. experienced fun and friendship at the annual NF Camp in Emigration Canyon, Utah. This unique camp offers kids the opportunity to try new activities including rock climbing, horseback rising, zip-lining and fishing and to spend time with other youth also affected with NF. Last year campers were also treated to a surprise visit by Utah Jazz star and 2012 NBA Slam Dunk Champion, Jeremy Evans (a.k.a the Human Pogo Stick), who signed autographs and exchanged stories with the campers. Campers were also honoured by a visit from noted NF researcher and physician Dr. Eric Legius of Belgium.

This year the BCNF will once again offer scholarships for kids who otherwise may not have the opportunity to attend this special camp.

Visit the website for more information or contact the BCNF office at ***info@bcnf.bc.ca***

A limited number of scholarships are available. **Apply today!**

BCNF.BC.CA/GO/CAMP

UNDER THE MICROSCOPE

THE NF LAB AT VANCOUVER WOMEN AND CHILDREN'S HOSPITAL

As I entered the beautifully built building, the Women and Children's and Health Research Institute, that is home to the research lab Kimberly Jett, NF researcher, works out of, I was a little taken aback. The only labs I had ever been to were in high school, where I was forced to learn how not to use a Bunsen burner. The lab Kim spends many hours working in, is modern, bright and full of working science type experiments.

I was visiting Kim to have a tour of the lab where NF research is being carried out. My tour went 'backwards', seeing the cells under a microscope first and working my way back to where the samples come from.

Kimberly welcomed me with open arms and I immediately felt comfortable in such daunting surroundings of white coats, warning signs and expensive microscopes. She guided me around the new facility in Vancouver showing me the equipment used to view the cells she collects from vascular (heart) samples known as sections prepared in paraffin wax. It's time consuming, detailed and intricate work.

We moved on to where the samples are prepared for viewing under the microscope. I had never seen a microtome before. It is a machine that slices frozen tissue into incredibly thin slices. Microtome sections can be made thin enough to section a human hair across its breadth. It uses diamond blades and can even cut through bone. Once cut, the samples are prepared in paraffin wax and dyed to help preserve them on slides. I was starting to have a much better understanding of why medical research takes time, patience and it is never a case of simply finding the answer. It was way past regular working hours and yet there were still other scientists hunched over their lab benches deep in concentration.

I WAS STARTING TO HAVE A MUCH BETTER UNDERSTANDING OF WHY MEDICAL RESEARCH TAKES TIME, PATIENCE AND IT IS NEVER A CASE OF SIMPLY FINDING THE ANSWER.

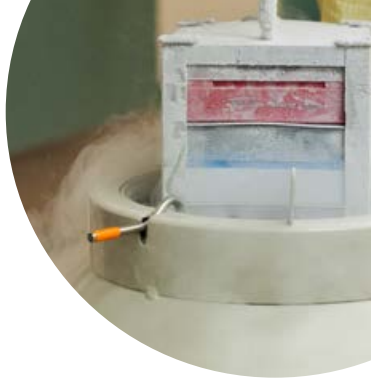
The next stop on the tour was to see where the samples came from and how they were collected. Returning back to her lab, Kimberley donned a protective outfit, which included a rather elaborate pair of 'oven gloves'. I have seen a lot of cool things in my time – seeing liquid Nitrogen at work is one to add to that list. Kimberly gently lifted out of the industrial sized thermos flask a case of samples that had been kept frozen for dissection, slicing and to help retain cell structure integrity. These samples were yet to be put through the microtome and paraffin.

From the frozen thermos the samples were prepped for further dissection under one of the many microscopes. Again, delicate hands and patience are required. Kimberly was at home in the lab – focused on the task at hand yet energetic, despite the long days in the lab. I was able to watch the dissection of vascular tissues being removed from a small sample. What continues to amaze me was the amount of time it takes to prepare the tissues before any research is done on its cell structure.

Kimberly is genuinely excited about the work being done researching the vascular effects caused by NF1 and I couldn't help but be excited with her. I was starting to have a deeper appreciation for where and how the donations to the BCNF get utilized. I never fully understood until seeing it first hand what is needed to help find a cure. It takes time and many, many little steps – all of which costs money. Raising awareness and raising funds for NF is vital and allowing others a sneak peek into the world of NF research with photographs is my way of raising awareness. Now we need to join together to raise the money. •

WORDS & PHOTOS BY **CARA GRIMSHAW**


KIMBERLY JETT



AN INSIDE PERSPECTIVE

BY KIMBERLY JETT

Our lab is involved in a variety of research projects studying NF1. Some projects are clinical and examine how to better diagnosis or treat different characteristics which may occur in individuals with NF1. Other projects are pre-clinical and use models of NF1 to reproduce observation in people to study how or why it occurs further. By doing these types of studies we hope to be better equipped to treat the different conditions which occur in NF1. One such project examines NF1 vasculopathy. Funding provided from BCNF and NF Canada has allowed our lab to determine why and how vasculopathy occurs in NF1 and test potential therapeutic agents.

NF1 vasculopathy, a condition in which the structure and function of blood vessels may be disrupted, presents in a number of different ways. In extreme cases this may include stroke (blocked vessels in the brain), rupture of major arteries or aneurysm (ballooning out of vessels). The most common presentation, however, is high blood pressure. Our results in younger adults with NF1 suggest many people have signs of vascular disease even when there are no symptoms of it. We do not know yet if the signs of vascular disease we observed will ever lead to symptoms and are going to examine this further.

The cause of vascular disease in NF1 is not clear but is thought to be related to reduced neurofibromin (the protein produced by the NF1 gene) in cells. Neurofibromin controls cellular functions, such as, the number of cells made, cell movement, and cell maturity. As a result of reduced neurofibromin in cells that make up blood vessels (that is, the endothelial and smooth muscles cells) and in the inflammatory cells (the cells that fight off infection) in people with NF1, there is an increased response to stress and repair in the cells. As our results found people with NF1 have elevated amounts of several inflammatory factors in their blood, this may be important.

To study how these factors affect vessel function we examined how the aorta (the biggest vessel in the body) functions in a model of NF1. The results of this study have shown that the function of the vessel (how it opens and closes) is altered in NF1 mice and that the vascular cells respond to stimuli differently than controls. The results of this study will be submitted for publication soon. We have also examined vascular cells from patients with NF1 and have found alterations in how these cells respond to many stimuli. We found it is possible to block the activity of the inflammatory factors and alter the effect of the factor on the abnormal behavior of the cultured cells.

More experiments on these cells are needed to draw definitive conclusions, but using these results we will be able to determine if blocking the activity of these inflammatory factors can reduce the abnormalities that occur in cultured blood vessel cells from people with NF1. NF1 research is moving forwards with exciting results. •

WORDS BY **KIM JETT** / PHOTOS BY **CARA GRIMSHAW**

THE NF REGISTRY

The Children's Tumor Foundation has created an online NF Registry. The purpose of this registry is to find people who may be eligible for clinical trials or other research studies being conducted in the field of neurofibromatosis (NF), and to determine the commonality of specific NF characteristics.

If you or your child has been diagnosed with neurofibromatosis (NF) you can join the NF Registry today.

The key to treatments lies within you.

This registry has been created to collect data on large numbers of people with NF. This has never existed before and will help:

1. Identify people who may be eligible for clinical trials or other research studies being conducted in the field of NF. New clinical trials are launched each year yet challenges identifying participants too often mean that these studies finish late – or never really get started. Today, 80 percent of trials fail to recruit enough volunteers within planned timelines. Under enrollment is one of the most significant problems facing NF drug development. Together, we can solve it.

2. Determine the commonality of specific NF characteristics. This will help researchers and doctors devise better ways to care for people with NF. The NF Registry's mission is to identify people with NF who are interested in participating in clinical trials, as well as determining the commonality of specific characteristics of neurofibromatosis. The NF Registry is committed to secure methodologies and pioneering research that will lead to improving the health and well-being of individuals and families affected by NF.

This is your chance to get involved and make a difference to finding improved treatment options for NF.

Please go to ***www.nfregistry.org*** for more information and to get involved. •

YOU ARE INVITED

The NF Tea Party is a fundraiser to aid the BCNF in supporting those living with the genetic disorder Neurofibromatosis.

For more information, please visit us online:

bcnf.bc.ca/go/teaparty