

# HANDS OF HOPE



THE BETHANYS HOPE FOUNDATION NEWSLETTER | HOLIDAY 2007

## St. Thomas Aquinas Leadership Class Becomes Bethany's Butterflies

It started with a simple question. And, when the question was asked, no one expected the results. "Who is your greatest role model?"

Teacher Antonietta De Bellis posed the question in a journal entry exercise for the students in her grade 11 Peer Leadership and Development course at St. Thomas Aquinas Catholic Secondary School - an elective class designed to help students nurture leadership abilities. One of her students' responses stood out, and she had him explain his answer to the class. The response was: "My sister." The student was Ben McIntyre, son of Dave and Lindey.

"When I was telling my story I was getting nervous and I started shaking," said Ben about his response. "But, I wasn't expecting anything big to come out of it."

Ben's telling of his sister Bethany's heroic story and how it affected him struck a chord with his 11 classmates, though.

"I'd known Ben since grade nine but I never knew his story," said classmate Shannon Marchant. "To hear his story about how strong he was, and his sister...it was just amazing. I appreciate that he shared it with us, and I knew then and there that our class could make a difference."



St. Thomas Aquinas' Peer Leadership and Development Class became the Bethany's Butterflies Team. Front Row: Ben (left holding portrait of Bethany) and teacher Antonietta De Bellis (on right behind poster).

The group of leaders decided to create the "**Bethany's Butterflies**" team with a mission of raising awareness within their school of Metachromatic Leukodystrophy (MLD) and funds for Bethany's Hope Foundation.

To educate themselves in genetics issues and the research being done to find a cure, they visited Bethany's Hope Leukodystrophy Research Laboratory.

"Their visit helped the students understand the genetic background behind disease," said Dr. Tony Rupar. "Ultimately, some of them might have been inspired to consider pursuing science."

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*It is better to light a candle than curse the darkness*  
~ Eleanor Roosevelt



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### Celebrating a Year of New Faces

In reflecting on the past year, a theme that I think best captures it all for Bethanys Hope Foundation is a "Celebration of New Faces."

Because of the many successful fundraising events and the generosity of Foundation supporters Bethanys Hope Leukodystrophy Research Laboratory will see a new face added to Dr. Tony Rupar's research team. On April 1, we will look forward to announcing the addition of a **new investigator who will join the team** in their quest to cure MLD. This investigator position will be funded entirely by the Foundation.

New faces this year also came in the form of new neighbors in Old South London, as we opened the doors to **our new office at 141 Wortley Road**.

You can find the Foundation logo on the window awnings; it is wonderful to have a visual presence in the community. Please stop by for a visit when you are in the neighbourhood. We also thank everyone at Bishop Cronyn Memorial Church for serving as outstanding hosts for our original office all these years.

This has been a year of some **amazing events**, and we were delighted to see faces new and familiar. Each event brought increased funding for the research efforts in the lab, and greater awareness in the community of MLD. Please read through the "Event Roundup"...you'll truly get a sense of how much heart and soul goes into each event, and for that I extend a huge thank you to our sponsors and event partners.

With a Foundation our size we rely on the generosity of time and talent of many volunteers. We added new faces to the fold this year, including the Acadia Fraternity from UWO, increasing the **Bethanys Hope volunteer community** to more than 230 people! The successes this year could not have happened without their dedication and support, and we celebrated their efforts at our Volunteer Appreciation Night.

Our 2008/09 returning Foundation Directors would like to take this opportunity to welcome a new face. We are pleased to have Mr. Mark Fagan of @ Print join our Team as a Director. Mark has been a long time volunteer and a very generous supporter of Bethanys Hope Foundation. Welcome Mark!

New faces came in another way this year - as we were introduced to more **families of children with MLD**. We heard them share stories of their children's heroic efforts, and as always they inspire us to continue. These children's stories, like Bethany's, give us and Dr. Rupar's research team a continued - and urgent - sense of purpose.

As always, with hope,

*Lindsey*

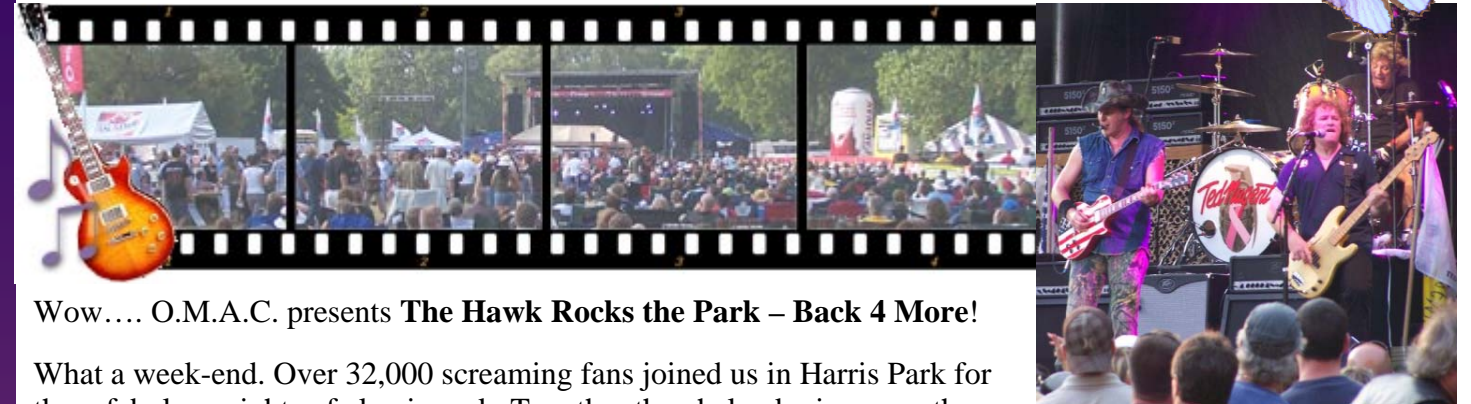


Dan, Shelley & Paul from the R.C.M.P.



(L to R): Sherard, Paul, Haley, and Acadia Fraternity President Ryan Gauss at Volunteer Appreciation Night.

Volunteer Appreciation Night photos courtesy of Snap London North newspaper



Wow.... O.M.A.C. presents **The Hawk Rocks the Park – Back 4 More!**

What a week-end. Over 32,000 screaming fans joined us in Harris Park for three fabulous nights of classic rock. Together they helped raise more than \$252,000 for MLD Research.

From July 26 thru 28, 2007 these dedicated rockers stood in the sun and the rain to enjoy the best Rock the Park ever. They came from Toronto, Detroit, Buffalo, Chicago, Montreal, Thunder Bay, Quebec City and all around the great city of London to rock with the likes of *Deep Purple, Cheap Trick, Nazareth, Pat Benator & Neil Giraldo, George Thorogood, Ted Nugent, CCR* and more.

In just three years, this festival - designed by Brad Jones of Don Jones Productions - has raised over \$680,000 for Bethanys Hope Leukodystrophy Research Laboratory. This would not be possible without our volunteer team and our wonderful partners, including: Michael Scott from Ford Keast, the terrific staff of CKDK-FM 103.9 – The Hawk (including Michele Dyer, Kent Guy, and Jeff McArthur) and of course Brad Jones.

Events like The Hawk Rocks the Park would not be possible without corporate sponsorship, and we were very fortunate to have the best. It was indeed a privilege to once again work ISM Security Group and their president Gord Fisher, O.M.A.C. Mortgages and their president David Providenti, Molson Canada, Mike's Hard Lemonade, Domino's Pizza, Rogers, Coca Cola, Delta Hotels, Leon's, London Major Appliance, Raymond Brothers, Aboutown Taxi, The London Free Press, The City of London and St. John Ambulance - and others as well. Special thanks to our long time friends Andy and Kirk Thompson of Can-Am RV Centre for donating all the RV units for the week-end.

Our foundation was blessed once again with 210 dedicated, hard working volunteers - more than half of whom came from the Royal Canadian Mounted Police. A large contingent also joined us from area McDonald's Restaurants, Scotiabank and Ford Keast Chartered Accountants. Without their energy and enthusiasm, there would be no show! Our foundation is truly grateful to them for their tireless efforts on behalf of the children of Leukodystrophy.

Thanks to all who came to party or work at O.M.A.C. presents The Hawk Rocks the Park 4... you are all making a difference in the lives of the children and families who suffer each day with the devastation of Leukodystrophy. We could not do this without you!!



Hope to see you all at The Hawk Rocks the Park 5 ... July 24 to 26, 2008!

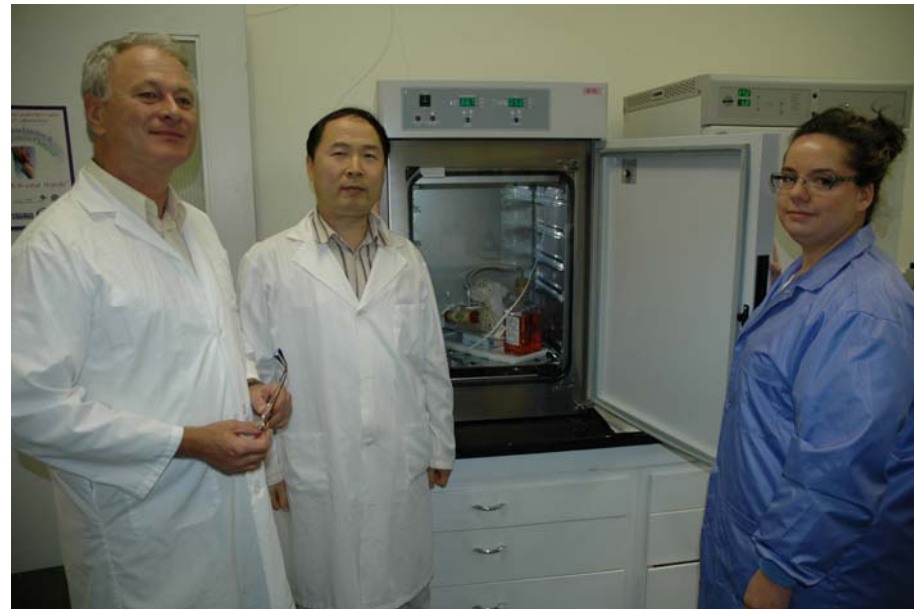




### Intra-Cranial Gene and Enzyme Delivery Could Sidestep the Blood/Brain Barrier

The very thing that protects your delicate brain from potentially harmful substances in your blood is also a tremendous challenge in developing the techniques that may cure diseases affecting the brain, such as Metachromatic Leukodystrophy (MLD).

The blood/brain barrier is a natural, biological system designed to prevent blood-borne pathogens that circulate throughout the body from affecting the brain - where they could do far greater harm than they could in the rest of the body. The problem with this barrier; however, is that it prevents a certain level of "access" to the brain that researchers need to deliver therapies to treat MLD.



*Dr. Rupar and his lab team - Dr. Jiahui Liu and Cathy Regan - stand beside an incubator that holds cells secreting the enzyme arylsulfatase A (ASA).*

"We've accomplished several important steps in unraveling the mystery of MLD," says Dr. Tony Rupar of Bethanys Hope Leukodystrophy Research Laboratory here in London. "Now we are focusing on overcoming the remaining obstacles."

It is known that genetic errors (mutations) within the arylsulfatase A (ARSA) gene cause MLD, and that these errors prevent functional arylsulfatase A (ASA) enzyme to be produced. Dr. Rupar and his team have shown in a mouse model of MLD that this enzyme deficiency causes the molecule sulfatide to build up in the brain, where it causes apoptosis (cell death).

With this understanding under their belts, the drive is to fully develop strategies for treating, and ultimately curing, MLD. "One of these strategies is genetic therapy by transferring in a copy of the proper ARSA gene through a process that uses gene transfer vectors," says Dr. Rupar. "Gene transfer vectors use harmless viruses that have been proven safe to carry a correct copy of the gene to the host site. Viruses work by imputing a part of their DNA into our body, so we are using the same concept to have a safe virus (with a correct copy of ARSA) input that DNA strand to a person with MLD."

A second strategy is direct enzyme delivery. "We know that the deficiency of ASA enzyme is what causes devastating apoptosis in the brain," says lab team member Cathy Regan. "So, by reversing the enzyme loss the cell death could be lessened or stopped. This is why we are trying to directly deliver the ASA enzyme to replace the loss that MLD causes."

These two promising therapies for MLD illustrate why the blood/brain barrier is a problem: how do you get the treatment to the brain, where the cell death is actually taking place, if there's basically a wall in your way?

The answer may sound somewhat intimidating, but it is being perfected to be as safe and effective as it is  
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promising: direct access to the brain through a process called "intra-cranial delivery."

"The blood/brain barrier essentially blocks therapies (gene vector or enzyme replacement) delivered in 'traditional' ways, to different parts of the body, from accessing and having an effect on the brain," says Cathy. "Obviously, that's a big problem when you're dealing with a disease of the brain. That's why we're striving towards intra-cranial delivery."

In the case of enzyme replacement, the team has developed a method for cells in the lab to produce and secrete the ASA enzyme. The enzyme is then purified to be suitable for direct delivery to the ventricles of the brain. Experiments are now determining if delivery to the ventricles allows the enzyme to be dispersed to the parts of the brain where apoptosis is taking place, allowing the sulfatide buildup to be broken down. Assessment is also being done to determine how long the delivered enzyme remains active, how much needs to be delivered for effectiveness and where the most ideal delivery points are.

In sum, a rather literal example of research overcoming a barrier.

### The Promise of Treating MLD Before it Harms the Brain and Nervous System

Dr. Rupar and his team see the exciting promise of therapies to treat, and eventually cure, MLD - therapies such as direct ASA enzyme replacement and genetic vectors to introduce a correct ARSA gene.

There is a major clinical paradigm; however, that faces geneticists and pediatricians when treating a child with MLD. The dilemma is this: although it may become possible to cure MLD, it's incredibly difficult - if not impossible - to reverse the devastating brain damage that the disease causes.

"MLD presents a very difficult situation," says Dr. Rupar. "First of all, it's not an easy diagnosis for a pediatrician. Parents might be concerned that their child is not reaching the developmental milestones for their age. That could be caused by any number of things, including simply being a little behind in development at a particular stage. Their pediatrician will go through a multitude of diagnosis techniques to see if something is wrong. Meanwhile, if the cause is MLD...throughout all the time of necessary diagnosis, brain damage is taking place! We'll eventually be able to treat the MLD and stop further damage, and we don't know whether treatments will be able to one day reverse cell death or if a child could compensate for some of the loss. However, the ideal solution is to diagnose MLD and start treatment before problems present themselves."



*A dried blood spot specimen on a collection card, showing samples from the card that have been "punched out" to be tested in the laboratory. Dr. Rupar's lab team is perfecting how to efficiently and effectively screen for MLD from the punch card samples*

Enter newborn screening. Currently, in many parts of the world including Ontario, all infants undergo newborn screening for 29 diseases using a dried blood spot specimen card. MLD is currently not tested for, and there are important reasons for that. "Routine newborn screening can only happen after a valid screening test is developed and would be most effective if there were a treatment for a positive result," says Dr. Rupar. "Having a treatment in place would give parents the hope of stopping damage before it begins."

Cathy Regan in Dr. Rupar's lab is currently perfecting the technique to effectively screen for MLD through dried blood spot samples so that the testing will be ready as soon as the treatment is.



### 9th-Annual "Tiger in the Woods" Golf Tournament

On Monday September 17, 2007 owner Rick DeJager of Tiger Jacks Bar and Grill of London, Ontario hosted the 9th-Annual "Tiger in the Woods" Charity Golf Tournament in support of Bethanys Hope Foundation. As in the past, proceeds from this tournament benefited the research being conducted at Bethanys Hope Leukodystrophy Research Laboratory.



Golfers began the day at Echo Valley Golf Club with registration and a breakfast sandwich, allowing the participants to enjoy a trick shot show by the one-and-only Bob Martin and Ron Lampman from Spankit Golf. Then it was off for a wonderful day of golf where participants found many creative ways to spend some cash. The teams then made their way back to Tigers Jacks where they were treated to a wonderful buffet, a fabulous prize table, silent auction and live auction. This tournament is special in many ways, and one of the reasons is Rick's attention to detail - including the fact that no one leaves without a prize.



Once again, we are grateful for the enthusiasm of Brian Mortimer and Terry Provost as they worked the room as our auctioneers. Thank you to the many live and silent auction item donors, hole sponsors, gift table donors, volunteers and participants. Your generous donations of time, talent and funds enabled us to provide Dr. Rugar and his Medical Team with more than \$26,000 for their research program.

It is hard to find the words to describe the atmosphere at Tiger Jacks that night as the "Tiger in the Woods" participants opened their hearts and dug deep in their wallets to fund research. "Thank you" is simply not enough when it comes to expressing our gratitude to Rick, who spends countless hours each year planning every detail. His commitment and compassion to make a difference for the children and families of Leukodystrophy is inspiring, and we couldn't be more grateful. To Rick and the entire staff of Tiger Jacks: you are indeed our "Heroes of Hope." Congratulations Tiger Jacks Bar & Grill... you are the best!

### McHappy Day 2007

On Wednesday, May 9 local McDonald's Restaurants celebrated McHappy Day 2007.

This years event was an outstanding success. With your help more than \$28,402.88 was raised in support of Bethanys Hope Foundation and the London Ronald McDonald House.

This all adds up to a lot of good reasons to be "McHappy!" Thank you to McDonald's Restaurants and their dedicated crew, and the local community leaders.



### An Afternoon of Design 2007

On Saturday, March 31 over 430 guests celebrated another Afternoon of Design fundraising event. Included this year was a breakfast seminar affectionately called "Roast, Toast and Renos." The morning session featured presentations by three experts in the field of kitchen renos, gardening and window treatments. Breakfast was provided by our friends at McDonald's.



Doors opened at 10 a.m. so guests could visit with the 16 exhibitors and scout the silent auction table which included more than 160 items.

We were pleased to have Sarah St. Amand as the guest speaker. Guests of honour were Steven Sabados and Chris Hyndman, who had to make their way through the crowds of screaming women before hitting the stage. What a thrill!



It takes many hearts and hands to make an event like this happen. We appreciate the amazing sponsors that generously supported this event and the dedicated volunteers who assisted. Special thanks to the wonderful young men from the Acacia Fraternity at the University of Western Ontario and their president Ryan Gauss. Thank you for taking the time to make a difference... hope is in our hands.



### BETHANYS BUTTERFLIES • FROM PAGE 1



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Armed with this understanding of MLD, the team delivered presentations to homeroom classes. Their passion was infectious, and soon the school joined them in contributing to the Foundation. Through a week-long fundraising drive students, teachers, parents and members of the community joined the leadership class in raising more than \$1000.

"Every single part of it was about the one goal of supporting Bethanys Hope," says Shannon. "When we finally knew how much we raised, we knew we made our dream come true as a class...and this actually felt like the first time I'd come up with a goal and really surpassed it."

The class was particularly excited about their ability to band together around a single purpose, inspired by Bethany's story. "To see how my classmates and I can use everything inside us to make a change is so amazing," said classmate Rebecca Devries. "We all strove for the one goal and that one difference we could make."

Ben was truly overwhelmed by the efforts of his class and the support that the group generated throughout the school. "I got really excited when the class took it on as a project," he said. "I was surprised how many people knew about Bethany's Hope, and of course my parents were really excited."

Ms. De Bellis, of course, was also extremely proud of her students. "It's not easy for students to go and talk to a group of their classmates about a disease," she said. "But they were amazing and engaged the school. That's leadership, and it was inspired from a really moving story."





One person  
*can*  
make a  
*difference*

# "A Recipe for Hope"

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