

August 2010

What's New at  
[www.alpha1canada.ca](http://www.alpha1canada.ca)

- **New procedure for Alpha-1 testing**
- **2010 Annual General Meeting report from the board of directors**
- **Ontario withdraws funding for augmentation therapy, join the fight to reinstate**

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## Saving money or saving lives? The government of Ontario makes a fatal decision

by Jim Mundy

Alpha-1 Canada has recently learned the Ontario government will soon announce that they will no longer accept applications for funding for augmentation therapy (Prolastin®) for alpha-1 antitrypsin deficiency. The Ontario government has been denying funding to newly prescribed patients for over two years as they reviewed funding for augmentation therapy. This therapy is the only treatment available for lung-involved patients with the rare genetic disease called alpha-1 antitrypsin deficiency.

They will, we are told, continue to accept applications from patients currently on treatment; however, they will use new, yet to be developed, criteria.

Forty-four Ontarians are currently receiving this therapy (more than half funded through private insurance). As many as ten newly diagnosed Ontarians have been denied funding by Ontario.

Alpha-1 antitrypsin deficiency is a rare condition that is often misdiagnosed as asthma or COPD. In Canada, it takes on average 9.9 years from the time symptoms appear until an accurate diagnosis is obtained. Waiting this long to find out what is wrong and then being denied treatment because you can't afford it is a situation the Canadian healthcare

system was designed to prevent.

Augmentation therapy is approved and covered in British Columbia, Manitoba and Québec; more private



payers are covering it now than even one year ago. In Spain, Germany, Portugal, France and Slovenia it is covered and even Medicare and Medicaid in the US have covered it for years. These decisions outside of Ontario reflect the growing body of medical evidence that augmentation therapy not only reduces the rate of lung function decline (by conventional lung function monitoring) but preserves lung tissue (as measured by the latest in x-ray technology) and reduces mortality. When more and more private and public

payers are adding augmentation therapy to their lists of treatments covered it is disturbing that Ontario has moved in the opposite direction.

It is important to understand that although this situation is currently Ontario-specific, cost saving moves made in one province are often followed by other provinces and convincing other provinces to cover it will be that much more difficult if the Ontario decision is not reversed. Furthermore, private insurance companies frequently imitate drug reimbursement policies of provincial governments. The Ontario decision could very well cause a domino effect that will be felt across Canada resulting in a treatment availability disaster for the national Alpha-1 community.

But the people who made this decision have nothing to worry about. While the Ontario government denies coverage for this lifesaving treatment to Ontarians without private drug coverage, they continue to make it available through their benefit plan for politicians, public servants and their families.

Alpha-1 Canada will not allow this decision to go unchallenged. We will take whatever steps are necessary to shame the Ontario government into providing this lifesaving treatment.

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## Saving money or saving lives? The government of Ontario makes a fatal decision

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We are calling on the Alpha-1 Community to stand with us against this heartless government decision.

We especially need your help if you are one of the people directly affected. If you live in Ontario and were denied therapy by the Ontario government, please get in touch with us. If you live in Ontario and are currently receiving reimbursement from the Ontario government for augmentation therapy, we need to hear from you too. If you live anywhere in Canada and have a story about how augmentation

therapy has helped you, please tell us. If you simply want to help, we want to hear from you.

We also need to hear from physicians who prescribe augmentation therapy.

We know there is an abundance of expertise among the Alpha-1 community. If you or anyone you know has been involved with government issues such as this please contact us.

As we continue to develop our strategy, we will need all kinds of help, let us know how you can help and we will give you more details. We'll keep you

posted throughout this crisis. Reversing this decision will take a great deal of effort from many people and, unfortunately will cost money. To make a donation toward this effort, please make a cheque or money order payable to Alpha-1 Canada and send to: Alpha-1 Canada, 1638 Northway Avenue, Windsor ON N9B 3L9. If you prefer to make a credit card donation, please support us at [www.canadahelps.org](http://www.canadahelps.org).

We look forward to hearing from each and everyone one of you. Thank you for your continuous support and generosity.

## One of our Alphas makes national headlines

by Vanessa McLaughlin

Whether you picked up a copy of a Canadian newspaper last month, be it the National Post or the Globe and Mail, or were reading an online news source, such as [cbc.ca](http://cbc.ca), the name Michael Cunningham should be a familiar one. He is the 52 year-old Edmonton Alpha who successfully underwent a double lung transplant despite having H1N1. Even more remarkable was that his new lungs also tested positive for H1N1. It is unclear in the articles whether the donor had H1N1, however, Dr. Atul Humar, director of transplant infectious diseases at the University of Alberta in Edmonton, reported that the donor was tested and did not have the virus. Michael says that he "cannot say for sure whether the donor had H1N1," but does not believe so. It is possible that

the virus moved into his new lungs.

What most headlines neglected to mention was that Michael has alpha-1 antitrypsin deficiency. He jokes that perhaps "it [was] too hard to spell" for journalists. According to his family, Michael had all the symptoms for a few years and even after the diagnosis of alpha-1 antitrypsin deficiency, he ignored his condition until he was driven to the hospital by a roommate after a few days of progressive illness. Diagnosed with pneumonia, his hospital stay at the University of Alberta Hospital in Edmonton turned into a full month. He became known as 'the Alpha guy.' The hospital not only had one of the finest and busiest transplant centres in Canada but they were very



Photo from National Post

knowledgeable about Alpha-1. During his stay, Michael learned that he could possibly be eligible for a double lung transplant and was registered at the Edmonton General Hospital's Respiratory Rehab Program. He was placed on the official transplant list in August 2009 and a donor was found in November 2009, just three months later. "I consider myself quite lucky in regard to the waiting list," said Michael, but he also admits that he "was very reluctant to sign up

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### Tip of the month

#### Love your lungs!

**Deep breathing increases lung function, gives your body more oxygen and helps reduce fatigue.**

**Dr. Mohammed Al Aklabi of the cardiac surgery division of the University of Alberta in Edmonton and his colleagues reported their findings of Michael's transplant in last month's issue of the *Journal of Heart and Lung Transplantation*.**

**For the abstract of the paper, please see our 'Interesting Research' section on page 5.**



### Create Awareness

**On behalf of Alpha1 Canada we strongly encourage you to join CORD's Annual Action Day on Parliament Hill on September 30, 2010.**

**Joining with other rare disease patient advocates will provide a strong common voice to advocate for health policy and a healthcare system that works for those with rare diseases.**

**It will also give a voice to Alphas and create an opportunity to meet other Canadians, from coast to coast, for support and information.**

**If you also plan to attend CORD'S Annual Conference, do not forget to apply for a travel scholarship.**

**Please do get in contact with us to share your experience should you have the opportunity to attend. We'll look forward to hearing from you.**

## One of our Alphas makes national headlines

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for the program and get on the list. Stubbornness and fear aside I knew something had to change."

Prior to surgery, Michael affirms that his health was in rapid decline: "The last lung function test gave me a FEV-1 of 12% and FVC of 38%." Simple tasks - such as getting up, getting dressed, eating and doing laundry - became chores as they required too much effort. Michael, in fact, had end-stage lung disease.

"The call came on a cold and icy winter day in November," remembers his family. Michael had not been feeling well and his tests showed positive for H1N1. You will remember that the H1N1 flu outbreak in 2009 presented additional health risks to those with Alpha-1. As such, Michael's doctors faced an additional challenge. Despite this, the operation went ahead as Michael was told that time was of the essence and an ideal match had been found. Doctors also told his family that lungs like these would not come around for another 5 years. Most believed Michael did not have 5 years to live without the transplant.

He was treated with the anti-viral drug Tamiflu prior to and after the transplant and was placed in ICU isolation. The operation was a success; Michael had the perfect lungs and match. "Ten hours after surgery he was awake and complaining about the vent. One day post op, he was breathing on his own. He was sitting up in bed with a Cheshire cat grin and could

then eat real food and talk and laugh (sort of)," recounts his family.

Of course his recovery did not come without difficulties for himself or his family. His medication induced psychosis which left him



unaware of his actions or speech. "He even tried to escape. He was either hoarding things or throwing them out. He had his cookies packed for one of his escapes but forgot where he had put them," recounts his family. Michael thankfully never made it past security as the staff was aware and diligent. Family, of course, feared that Michael would remain like that but once the medication was reduced, Michael was back to being himself. Interestingly he then felt the need for walking aids, but only for a short time.

Michael's life post-surgery next consisted of daily clinics and physiotherapy at the University of Alberta Hospital. Then tragedy struck the family when Michael's 19 year-old niece was killed in a car accident. "Amongst the flurry of the funeral preparations and out of town family and visitors, the Noro Virus was introduced. Nobody was left untouched. Michael was immediately admitted to hospital to combat dehydration and to be moni-

tored. He suffered less than any of the others: the big, the healthy, the fit and the young all ravaged by dehydration," recalls his family. Another sign of his determination.

Michael continued with his clinic and rehab at the hospital until his clinic days were only one day a week. "His work outs included what he could do in his building - he would walk/run down 13 stories and climb up 3, many times over. When the weather turned friendlier, he walked for miles, up and down the river valley," said family. This is remarkable considering he had been dependent on an oxygen tank prior to surgery.

Michael truly had a miraculous recovery and is enjoying life to the fullest with his new lungs. He continues to attend the rehab facility at the Edmonton General Hospital and has a new lease on life: "At present I feel fantastic. Better than I have in the last ten years. I can do things physically that I felt had been denied to me. Obviously being a lot more health conscious now, I try to eat right, get plenty of exercise and stay away from sick people."

A big thank you to Michael for sharing his story with our readers and also to his caring siblings Teryle, Jack, Jan and Vicky for their contribution to this article. We wish Michael sustained success with his recovery.

*If you know of any other Alpha making 'headlines,' including yourself or if you would like to make ours, please contact us.*

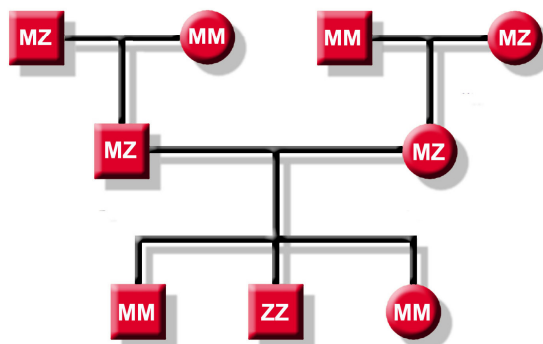
## Genetics are a family affair

by Vanessa McLaughlin

Michael Cunningham's story highlights the genetics of alpha-1 antitrypsin deficiency and the importance of being tested. His family suspected that he had A1AD as he had all the symptoms. A1AD is suspected in individuals with evidence of pulmonary disease (i.e., emphysema, asthma, persistent airflow obstruction, and/or chronic bronchitis); and/or evidence of liver disease at any age, including obstructive jaundice in infancy and in individuals with bronchiectasis. A1AD is also observed rarely in individuals with Wegener granulomatosis and necrotizing panniculitis. The World Health Organization, the American Thoracic Society, the European Respiratory Society and Alpha-1 Canada recommend that everyone with COPD be tested for A1AD.

Teryle Read, Michael's sister, also has A1AD. Doctors assumed she had asthma for years until a Respirologist took a family history. Suddenly things started to make sense: "Mom had bronchiectasis and her younger sister had part of her lung removed as a teenager." However, Teryle's mom did not tell her sister about her bronchiectasis until Teryle started talking about it. When this information was shared with the Respirologist, "the doctor said that I might have what was then a long name. No way did I think it was more than a pimple to be fixed," recalls Teryle. Blood tests confirmed that Teryle did in fact have A1AD.

Family Tree for  
Alpha-1 Antitrypsin Deficiency



Alpha-1 antitrypsin deficiency is a genetically transmitted disorder. It is something one is born with. However, there are varying degrees of disorder depending on what genes are passed on from parents to children. The range is from normal to severely deficient. MM is normal, ZZ is severely deficient and the others (MZ, MS, SS, SZ, etc.) are intermediate. If a child inherits one normal gene and one defective gene, for example MZ or MS, each gene will express its specific protein that can be identified in the blood (codominance). In order to inherit full-blown A1AD (usually ZZ), an individual must inherit two defective Z genes, one from each parent. Although MZ individuals carry a deficient gene, they usually do not develop disease. Also, people who are diagnosed as ZZ have been known to never develop clinical disease. So, while inheriting two ZZ genes means that a person may be predisposed to develop A1AD, it is not a simple cause and effect equation.

Two of Teryle and Michael's sisters volunteered to be tested. "One brother seemed more resistant but after me sending him back to his doctor a couple of times with firm direction of what needed to be done, he tested MZ," remembers Teryle. Then her mother, one sister, and two brothers, one of them Michael, were tested with the finger-tip kits she received from Florida. When Michael was tested, he was a ZZ, just like Teryle. Two of his brothers as well as a sister tested MZ, which is a slight increased risk for decreased lung function. The other sister tested MM. The siblings who tested MZ are not symptomatic. "Our concern was that my siblings were all having grandchildren and we didn't want this to be forgotten about until it appeared a few generations later to baffle and challenge or perhaps sadden another family," said Teryle.

Thank you Teryle for sharing your diagnosis and family testing with our readers.

If you suspect that a family member has A1AD, please ask them to get their doctor to determine their AAT level. If the level is low (1.5 g/L or less, or below the normal mean for the testing laboratory) his or her doctor can call the bilingual call-centre at 1-877-3 ALPHA1 (1-877-325-7421), to order a test kit.

Once your doctor receives the kit, three dry blood drop samples are taken then mailed to a testing facility at the University of Florida. Within two to three weeks the doctor will receive the results through the mail.

This vital information could not only save their lives, but in some cases could help them avoid transplantation and encourage them to eat healthy and exercise to enjoy life to the fullest.

Join the Canadian Registry

The Alpha-1 Canadian Registry was established in 1999 to gather information on Alpha-1 in order to study the natural history of the disorder and to conduct studies and clinical trials.

All information in the registry is strictly confidential. The registry is open to both patients and carriers.

Consent to become part of the registry does not mean consent for clinical studies. Individuals are free to withdraw their names from the registry at any time, should they so choose.

So if you haven't already joined the registry, consider doing so today.

Contact the registry by clicking [here](#).

## The new food allergies and asthma

by Vanessa McLaughlin  
Many Alphas have asthma. Those with asthma have allergies that make it worse. Although seasonal triggers seem to peak in the fall, some allergies, such as food allergies, can be a year round danger. One of the best ways to manage your asthma is to figure out your food triggers. Thankfully, priority food allergens are required to be declared on food products in Canada so that consumers with food allergies can avoid the products.

Those who have made the priority allergen list include the usual suspects: peanuts, milk, eggs, shellfish, and wheat. However, you might be surprised to learn that sesame has been on the list since 2000 and

mustard seed is in the process of being added. Mustard seeds were asked to be put on the list by 22% of people who responded to Health Canada's public consultation regarding pro-



posed amendments to the list in 2008. Garlic and onions were also popular choices by respondents; however, they didn't make the cut.

An article entitled "The Allergens to Watch" in Allergic Living (Summer 2010) highlights these changes and also suggests future foods to make the list could

include kiwi, mangos, lentils and chickpeas. The reasons for the shifts in allergies in Canada are unclear. Some attribute it to the influence of immigrant cuisines and ingredients and our penchant for the got to have it healthy and trendy food of the month. As such, our diets are changing. Others believe this shift is simply due to the fact that doctors are more aware of it and are better able to diagnose it. What we do know is that food allergies are on the rise and are much more prevalent than in years past.

So, if you think you have a food allergy, avoid the food and consult your physician. Although most food allergies only produce mild symptoms, some people do experience life-threatening reactions.

## Interesting Research

*The following is a summary (abstract) of a recent study of Alpha-1.*

*Because of copyright law we can only provide an abstract, if you want to read more check and see if your local library has this on their shelves.*

### **Successful bilateral lung transplantation in a patient with end-stage lung disease and positive novel influenza virus (H1N1)**

Mohammed M. Al Aklabi MDa, Justin G. Weinkauf MDb, Atul Humar MDc and Nitin Ghorpade MDa, University of Alberta, Edmonton, Canada.

The Journal of Heart and Lung Transplantation, Volume 29, Issue 8, August 2010.

In 2009, a pandemic novel influenza virus (H1N1) outbreak was declared by the World Health Organization and resulted in significant worldwide illness. This report describes a 50-year-old male with end-stage lung disease secondary to  $\alpha$ 1-anti-trypsin deficiency and chronic obstructive pulmonary disease. He was admitted for potential bilateral lung transplantation when suitable organs became available. Incidentally, he was found to have some non-specific symptoms, including malaise and myalgias. These findings were attributed to killed-virus H1N1 vaccine given 48 hours earlier.

However, as a safety measure, a nasopharyngeal swab was taken, and anti-

viral therapy with oseltamivir (Tamiflu) was started empirically. He underwent bilateral lung transplantation on the same day of admission. In the immediate post-operative period his nasopharyngeal swab came back positive for H1N1 influenza virus. Then, post-operatively, two consecutive bronchoalveolar lavage samples from the transplanted lungs were found to be positive for H1N1 virus. He received three-weeks of antiviral treatment post-operatively and he had uneventful procedure with favourable outcome.

*If you know of any re-search, articles or other publications that would be of interest to our readers please contact us.*

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Visit us on the web  
at [alpha1canada.ca](http://alpha1canada.ca)

Our website is continuously updated with useful information for Alphas, their caregivers and healthcare providers, as well as news on promising research. Make a habit of checking our website regularly so you won't miss out on exciting updates and always read our monthly newsletter from top to bottom.

Help us spread awareness by sharing this newsletter with your family and friends.

If you would like to receive this newsletter by e-mail, please contact us at 1-888-669-4583 or [vanessa.mclaughlin@alpha1canada.ca](mailto:vanessa.mclaughlin@alpha1canada.ca)

This newsletter is designed to support, not replace, the relationship that exists between you and your physician. It is not the intention of this newsletter to provide specific medical advice but rather to provide the Canadian Alpha-1 Community with information to better understand their health and their diagnosed disorder.

Specific medical advice will not be provided and Alpha-1 Canada urges you to consult with a qualified physician for diagnosis and for answers to your personal questions.

**Alpha-1 Canada**  
**Making a difference in the lives of Alphas**

## We are on Facebook

by Vanessa McLaughlin

In Canada alone, close to 16 million of us have Facebook accounts – that is half the population! The fastest growing age groups on Facebook are women 35 to 50, and people over 55. While Facebook certainly has its critics, there is no denying the appeal of this social networking site as a way to keep up with your favourite non-profit organization: Alpha-1 Canada. Our online community is getting bigger and represents a unique way for Canadian Alphas to keep in touch. As of mid-July, our Alpha-1 Antitrypsin Deficiency Canada Inc. page had 103 fans. It is also a

place where you will find a link to our newsletters, the latest news and information and an opportunity to network with Alphas internationally, such as in the UK. We also have a discussion forum where you can introduce yourself and tell fellow Alphas your story.

So, if you are one of almost half the Canadian population on Facebook, please join our site and help make it another wonderful resource to inform, educate and support Canadians affected by alpha-1 antitrypsin deficiency as well as to generate an international awareness of this disease.

Special thanks to our Alpha-1 Canada member, Mr. Tim Blades, who volunteers his time to manage the site. We couldn't do it without you Tim and are very grateful for your support and dedication. On behalf of Alpha-1 Canada, we applaud you for a job well done. Keep up the good work!

**On Facebook? Join us.**



**Joignez-nous sur Facebook!**