



Alpha-1 Canada Community News



April 2010

US Alpha-1 Association Conference in Orlando



The board of directors of Alpha-1 Canada is pleased to announce the awarding of three travel sponsorships to Canadian Alphas that will allow them to attend the US Alpha-1 Association Conference in Orlando, Florida this June.

The sponsorships were awarded to a liver-affected young person and his family from British Columbia, a recently diagnosed lung-affected man from Alberta and a lung-affected woman from Ontario.

The Canadian delegation will be rounded out by our Treasurer, Jerry Cunningham and our Executive Director, Jim Mundy who are both looking forward to meeting the sponsorship recipients in person.

Although it is too late to apply for these sponsorships now, it is not too late to register for and attend the conference June 11th – 13th. For more information about the conference visit the Alpha-1 Association website at <http://www.alpha1.org/education/nateduconf.php>

Here we grow again

After another successful year Alpha-1 Canada is growing again. As you may be aware, Alpha-1 Canada is run by one full-time Executive Director, Jim Mundy; and one virtually full time volunteer President and Chairperson, Mimi McPhedran.



To save money we do not have offices that we rent, rather Jim and Mimi work out of their homes.

Alpha-1 Canada is currently searching for a part time Administrative Assistant. This is a paid position and the successful applicant will be fluently bilingual (written and spoken) in English and French.

Like Jim and Mimi, this person will also work from their home for 20 hours per week. If you think you might be interested, please read the job posting here.

What's Inside

We still need your help 2

How drugs are approved and funded in Canada 2

Interesting Research:
Your Eyes 3
Your Balance 4
Your BMI 5

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We still need your help

Ontario Government Withdraws Funding for Augmentation Therapy



As you have read here before, Ontario has ceased approving funding of alpha-1 antitrypsin augmentation therapy (Prolastin®) for newly prescribed patients. This therapy is the only treatment available for lung-involved patients with alpha-1 antitrypsin deficiency.

This decision is wrong on clinical, economic, and more importantly, ethical grounds.

Please help us to convince the Ontario Government to reinstate funding for this lifesaving therapy. We are asking any patient or family member that is affected by this decision to immediately write to Ms. Helen Stevenson, Assistant Deputy Minister and Executive Officer, Ontario Public Drug Programs Division. Ms. Stevenson will make the ultimate decision on this funding, please help us convince her to do the right thing.

If you live in Ontario and are being reimbursed by a private insurance plan for the costs of your augmentation therapy you may want to check to ensure that your benefits do not run out at sometime in the future, for example when you or your spouse retire. If the Ontario government does not reinstate funding, you will have no other option when your private benefits cease.

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. Furthermore, private insurance companies frequently imitate drug reimbursement policies of provincial governments. If Ms. Stevenson decides not to reinstate funding in Ontario, the domino effect could well be felt across Canada resulting in a treatment availability disaster for the national alpha-1 community.

Please visit our website for contact information for Ms. Stevenson at <http://www.alpha1canada.ca/Ontario-Withdraws-Funding-for-Augmentation-Therapy>.

Although personal letters are more effective, we have added a form letter that you may use.

And please forward this to friends, family and colleagues encouraging them to also write.

“How Drugs Are Approved and Funded in Canada: What Every Patient Should Know”

The Canadian Organization for Rare Disorders (CORD) [<http://www.raredisorders.ca/>] and the Consumer Advocare Network [http://optimizinghealth.org/about_advocare.php] are hosting workshops entitled, “How Drugs Are Approved and Funded in Canada: What Every Patient Should Know” in Montreal and Toronto. Workshops have already been held in Vancouver, Edmonton and Halifax.

These workshops are specifically designed for patients and laypersons who want to better understand the processes related to drug research, development, approval, and availability through provincial drug plans. This training will also benefit patients who want to improve their capacity to



continued on page 3

advocate on behalf of themselves or their families.

Registration fees for patients and patient groups have been waived.

To receive the patient registration code, please send an e-mail request to angela@optimizinghealth.org

Travel scholarships are also available. For details please contact: Angela Covato at 877-302-7273, 416-969-7431 or by e-mail at angela@optimizinghealth.org

When

May 7 - 8, 2010, Montréal, Quebec (English)

11 et 12 juin, 2010, Montréal, Quebec (en français)

Interesting Research

Heel-First Walking Conserves Energy



Touching down toe-first is less economical in humans, research shows.

It takes far less energy to walk heel-first than to walk on the balls of your feet or on your toes, a new study has found.

Compared with heel-first walking, it takes 83 percent more energy to walk on your toes and 53 percent more energy to walk on the balls of your feet, according to the University of Utah researchers. They measured energy expenditure in 27 volunteers in their 20s, 30s and 40s as they walked or ran using the three different methods.

"Our study shows that the heel-down posture increases the economy of walking but not the economy of running. You consume more energy when you walk on the balls of your feet or your toes than when you walk heels first," study senior author David Carrier, a biology professor, said in a university news release.

To read the rest of this article at [healthfinder.gov](http://www.healthfinder.gov/news/newsstory.aspx?docID=635836), click here (English only) [<http://www.healthfinder.gov/news/newsstory.aspx?docID=635836>]

Eye disease in children with cholestatic disorders

K.T Fahnehjelm, B. Fischler, L. Martin and A. Nemeth of the Department of Clinical Neuroscience at the Karolinska Institutet in Stockholm, Sweden recently published, "Occurrence and pattern of ocular (eye) disease in children with cholestatic disorders.

(Alpha-1 antitrypsin deficiency in children is a cholestatic disorder.)

The following is an abstract of their research:

Purpose: To describe visual function and ocular manifestations in patients with onset of cholestasis during the neonatal period.

continued on page 4

Methods: Patients with neonatal cholestasis, either transitory or chronic, who came for assessment to our tertiary referral centre were included in a cross-sectional study and underwent ophthalmological examinations including fundus photography.

A total of 57 patients (24 girls and 33 boys), aged 0.4-18.0 years, were included. Of these, 28 patients had biliary atresia, 11 had Alagille's syndrome, five had progressive familial intrahepatic cholestasis and nine had different



disorders such as pituitary insufficiency, alpha-1-antitrypsin deficiency, mitochondriopathy, congenital infections or cholestasis caused by unknown reasons.

Results: Visual dysfunction and/or one or several ocular manifestations occurred in 39 out of 57 patients. Major ocular malformations occurred in five patients.

Out of three patients with biliary atresia, one patient had severe visual impairment caused by microphthalmia and chorioretinal coloboma, one patient with Cat Eye syndrome had bilateral uveochoioretinal coloboma and one patient had Rieger's anomaly. Two patients, both with pituitary deficiency and transient cholestasis, had severe unilateral visual impairment caused by optic nerve hypoplasia.

Conclusion: The majority (68%) of the patients with cholestasis had ocular manifestations. Although the severity of ocular complications varied with diagnosis, and was most apparent among patients with biliary atresia or pituitary deficiency, no conclusion can be drawn regarding the connections between these conditions from the present study. Nevertheless, ocular assessment is important for diagnostic purposes and for early intervention in patients with cholestasis.

People with chronic obstructive pulmonary disease have reduced control of balance in the mediolateral (from side to side) direction

M.D. Smith, A.T. Chang, H.E. Seale, J.R. Walsh and P.W. Hodges of the Clinical Centre of Research Excellence in Spinal Pain, Injury and Health at The University of Queensland, Australia recently published "Balance is impaired in people with chronic obstructive pulmonary disease."

The following is an abstract of their research:

Background: It has recently been suggested that people with chronic obstructive pulmonary disease have an increased risk of falls. Although falls risk is multifactorial, impaired balance may contribute. The primary aim of this study was to compare balance between people with and without chronic obstructive pulmonary disease and the secondary aim was to determine if balance deteriorates when respiratory demand is increased by upper limb exercise.

continued on page 5

Methods: Twelve people with chronic obstructive pulmonary disease and 12 healthy control subjects participated in this study. Participants stood on a force plate to record centre of pressure displacement during a range of conditions that challenge balance.

Lumbar spine and hip motion were measured with inclinometers. Balance trials were performed before and after participation in upper limb exercise that increased respiratory demand in those with chronic obstructive pulmonary disease.

Findings: People with chronic obstructive pulmonary disease had increased mediolateral (from side to side) centre of pressure displacement and increased angular motion of the hip compared to healthy controls.

Mediolateral centre of pressure displacement was further increased in people with chronic obstructive pulmonary disease following exercise, but unchanged in controls. Anteroposterior (front to back) centre of pressure displacement did not differ between groups.

Interpretation: People with chronic obstructive pulmonary disease have reduced control of balance in the mediolateral direction. This may contribute to an increased risk of falls in this population.

Obesity and Mortality in Obstructive Lung Disease

J.G. Jordan Jr, and J.R. Mann of the Department of Family and Preventive Medicine, University of South Carolina recently published, "Obesity and Mortality in Persons with Obstructive Lung Disease Using Data from the NHANES III."

The following is an abstract of their research:

Objectives: The objective of this study was to determine the relationship between obesity and all-cause mortality among participants with obstructive lung disease in the National Health and Nutrition Examination Survey (NHANES III).

Methods: Public data from a retrospective cohort of the 33,994 participants in NHANES III was analyzed to determine the relationship between obesity and all-cause mortality among participants with obstructive lung disease.

Results were analyzed using proportional hazard models and controlled for age, sex, race, smoking status, current oral corticosteroid use, and severity of airway obstruction.

Secondary analysis considered time until death from respiratory disease or time until death from chronic lower respiratory disease (excluding asthma).

Results: The subset used in the analysis consisted of 2,439 persons with 844 documented deaths. Extreme obesity (body mass index [BMI] >40) was significantly associated with increased respiratory disease mortality (hazard ratio [HR] 5.78; 95% confidence interval {CI} [1.09 to 30.61]) and chronic lower respiratory disease mortality (HR 13.69; 95% CI [1.45 to 129.29]).

In addition, underweight status (BMI <18.5) was significantly associated with increased all-cause mortality (HR 2.42; 95% CI [1.31 to 4.46]), respiratory disease mortality (HR 7.10; 95% CI [1.94 to 26.00]) and chronic lower respiratory disease mortality (HR 14.80; 95% CI [2.24 to 97.99]).



continued on page 5

Conclusion: Underweight adults had increased risk of death from all causes and respiratory conditions, compared to class I obese adults. Extreme obesity was associated with increased risk of death from respiratory conditions, but not all-cause mortality. Additional research is needed to explain the complex relationship between BMI and specific causes of mortality in the context of pulmonary disease.

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 jim.mundy@alpha1canada.ca

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

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.

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