



Alpha-1 Canada Community News



December 2010

What's New at
www.alpha1canada.ca

- Ontario withdraws funding for augmentation therapy, join the fight to reinstate
- Podcast with Durhane Wong-Rieger, PhD on the development and implementation of a Canada-wide regulatory framework for rare disorders

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Season's Greetings



PASS THE LIGHT

For the ones we love,
For the Alphas we know,
And the ones we don't.

CHERISH THE GIFT OF KNOWLEDGE

Please join Alpha-1 Canada in lighting our annual multipurpose Ecandle.



What better time of year to spread awareness, education and support about our disease locally, nationally and internationally in combination with your Season's Greetings. All you have to do is forward an Ecard, that you will receive on December 20th on behalf of Alpha-1 Canada, to your family and friends and ask them to do the same.

We thank you in advance for your participation and we look forward to making this simple activity a resounding, annual, global success.

Wishing You All the Blessings of the Season,
Staff and the Board of Directors of Alpha-1Canada



The development and implementation of a Canada-wide regulatory framework for rare disorders – Promises kept?

by Vanessa McLaughlin

On behalf of Alpha-1 Canada, I would like to thank our guest speaker on December 2, 2010 Dr. Durhane Wong-Rieger.



Dr. Wong-Rieger began her talk by mentioning that Alpha-1 Canada was the first

patient group that she had had an opportunity to update on the proposed changes to modernize drug regulations and the technical consultations held by Health Canada on a regulatory framework for rare diseases.

Specifically, Health Canada has begun to draft regulations for the development and implementation of a regulatory framework for rare disorders which would resemble the one in the European Union. This framework would provide incentives for the development of new drugs, guidance in conducting clinical trials and pathways to approval. It would also provide a definition of a rare disease as 5 in 10,000 (1 in 2,000) Canadians; Alpha-1 Canada would fall within this definition.

That being said, Dr. Wong-

Rieger reminded us that the regulatory framework that wouldn't go into effect until we have legislation – this could be a lengthy process. As such, we must continue to pressure Health Canada to focus on orphan drugs and a national framework for rare diseases separately from the modernization of drug regulations. Of course, you can help by continuing to write letters to your MP and asking him or her to commit to writing a letter to the Health Minister making such a framework a priority.

If you were unable to attend and would like to hear Dr. Wong-Rieger's talk on the development and implementation of a Canada-wide regulatory framework for rare disorders or if you would simply like to listen again, please go to the podcast on our website.

Local Events attracts Alpha-1 Exhibition booth

by Vanessa McLaughlin

The World COPD Forum, presented by the Sudbury Lung Disease Support Group, was held at the Howard Johnson Plaza Hotel on November 25, 2010 in Sudbury.

The venue was filled to capacity with upwards of 140 people in attendance. Topics discussed were stress and treatment of COPD, benefits of pulmonary rehabilitation and the role of oxygen therapy. Dr. Andrew Pipe of the Ottawa Heart Institute discussed smoking cessation. Along with a dozen exhibitions, 2 spirometry booths with respiratory therapists were on hand for lung testing.

Mr. Jerry Cunningham, Alpha-1 Canada's Treasurer

and Board Member, was in attendance and managed the Alpha-1 Canada exhibition booth with information pamphlets. A big thank you to Mr. Carmen Huot, who generously donated the beautiful gift basket. Based on the numerous ballots

that were placed in the raffle box, it is fair to say that many people passed by and stopped at our exhibition booth.

For more information about this event, please go to www.ldsg.ca.



PODCAST

Listen to our latest podcast with Durhane Wong-Rieger, PhD on the development and implementation of a Canada-wide regulatory framework for rare disorders : http://www.alpha1canada.ca/Educational_Podcasts

Local Events

If you have participated in a local event and would like to share it with our readers, please contact us.

Donations

Find out how you can give gifts that don't need to be wrapped and provide information, education and support to people who could really use your help. Contact Alpha-1 Canada at 1-888-669-4583 or vanessa.mclaughlin@alpha1canada.ca for more information.

We would like to take this opportunity to send out a hearty 'Thank you' to all of our donors and encourage those who can to join them.

Exercise your heart this holiday season!

The holidays will soon be here and many of us are trying to figure out what to give those hard to buy for people on our list. This year, why not make a contribution to Alpha-1 Canada by making a donation in the name of an outstanding parent, teacher or friend. Making a donation in someone else's name is also a great way to both support a charity and honor a deserving individual.

Alpha-1 Canada has a number of programs and services that can benefit from your support:

- ◆ the patient support hotline at 1-888-669-4583 is available to patients and their caregivers for information on both a regular and emergency basis
- ◆ the medical advisory board meets regularly providing the board of directors and staff with information on the latest advances in research and treatments
- ◆ the web site (www.alpha1canada.ca) is continually updated with the latest information
- ◆ the monthly newsletter keeps patients and their caregivers up-to-date on current events and offers stories about fellow Alphas
- ◆ in addition to the web site and newsletter, printed materials are distributed and are available to anyone, printing and postage are very expensive

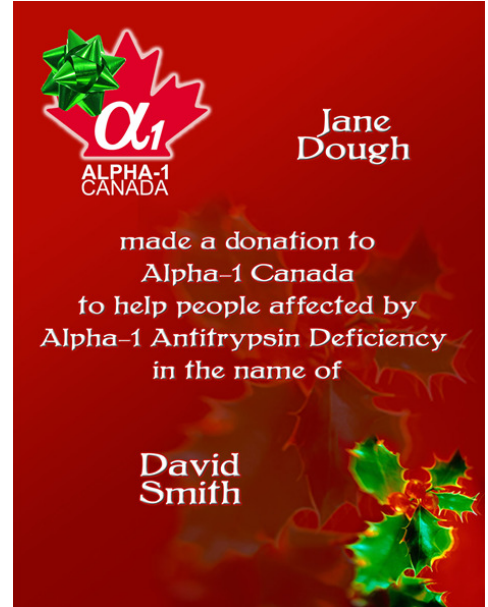
◆ patient support is offered through a variety of methods including e-mail, the hotline and a number of support groups

- ◆ through Alpha-1 Canada's membership in the Canadian Organization for Rare Disorders (CORD) we keep informed about government activities related to healthcare and drug coverages and participate in supporting those initiatives that will benefit the Canadian Alpha-1 Community.

These are just a few of the programs and services you can support and help others during this time of love and giving and receive a tax write-off in the process. We will send you a gift card for easy giving.

People often tell us that they can't afford to give much and that is certainly understandable, especially in these difficult economic times. But it doesn't take much to make a difference.

A donation of \$5 can pay the toll costs to make sure the parents of a newly diagnosed baby can talk to someone about what to expect and learn that although their family's lives will change, there is hope. Just \$10 can make sure two more doctors receive information on when to



suspect a COPD or asthma patient may have Alpha-1 and how to get testing done.

If two people were to donate \$100 each we could hold another Drop-in Meeting with a guest speaker to answer many questions and the recording of that meeting could help hundreds more who listen to the recording on our website.

Some of our donors use www.CanadaHelps.org to make their donations every month and multiply the good they do by twelve. CanadaHelps.org can accept credit card donations on our behalf on a one-time basis or even make those donations happen monthly without you having to log in over and over.

Knowing that a gift is a selfless contribution that made someone else's life brighter can put the season in perspective for you and the person in whose name you make a donation, perhaps even your own.

Interesting Research

The following are summaries of abstracts of recent studies of Alpha-1 and COPD. Because of copyright law we can only provide abstracts, if you want to read more check and see if your local library has these journals on their shelves.

The Role of Physical Inactivity in Increasing Disability Among Older Adults With Obstructive Airway Disease.

Katz P, Chen H, Omachi TA, Gregorich SE, Julian L, Cisternas M, Balmes J, Blanc PD. Department of Medicine, University of California, San Francisco.

Journal of Cardiopulmonary Rehabilitation and Prevention. 2010 Nov 29. [Epub ahead of print]

Abstract

PURPOSE: The independent contribution of physical inactivity to disability in obstructive lung disease (OLD) is difficult to study, partly because inactivity may reflect disease severity. We examined the relationship of physical inactivity to disability progression over a 1-year period among a group of older adults with OLD.

METHODS: A population-based cohort with self-reported physician-diagnosed emphysema, chronic obstructive pulmonary disease, or chronic bronchitis completed baseline interviews and in-person spirometry, with 1-year followup interviews. The Community Health Activities Model Program for Seniors physical activity questionnaire provided estimates of energy expenditure; we defined inactivity as no expenditure in moderate- or vigorous-intensity

activities. Disability was measured with the Valued Life Activity (VLA) disability scale; increased disability was defined as a 10% or greater increase in VLA disability score over 1-year follow-up. Logistic regression tested the relationship between baseline inactivity and disability increase, controlling for age, sex, baseline VLA disability, comorbidities, smoking, and pulmonary function.

RESULTS: 27% were physically inactive at baseline; 42.9% of individuals whose disability increased were inactive at baseline compared with 23.4% of those who did not experience a disability increase. With adjustment for covariates, increased disability after 1 year was significantly, more likely among individuals who were inactive at baseline.

CONCLUSIONS: Physically inactive individuals with OLD had more than double the odds of an increase in disability, even after controlling for baseline disability, lung function, and other covariates. These results provide strong support for the importance of maintaining physical activity among individuals with OLD.

Antibiotic Therapy for Exacerbations of Chronic Obstructive Pulmonary Disease (COPD).

Butorac-Petanjek B, Parnham MJ, Popovic-Grlje S.

Journal of Chemotherapy. 2010 Nov;22(5):291-7.

Abstract

Chronic obstructive pulmonary disease (COPD) is already the world's fourth most common cause of mor-

tality and likely to become the third in a few years' time. Because it is an inflammatory airway disease with altered host immune response, infectious complications are frequent. Acute exacerbations of COPD (AECOPD) significantly worsen the patient's general health, accelerating disability. Each exacerbation leads progressively to further deterioration of lung function. Among the various causes of AECOPD, including viruses, bacteria and air pollution, a bacterial etiology is most common (50-69%).

The management of AECOPD remains extremely challenging and places a heavy economic burden on health care institutions. The decision to administer antibiotics in AECOPD is multifactorial, the most important considerations being severity of the COPD stage and patient performance status, clinical symptoms (increased dyspnea, sputum volume and sputum purulence), severity of current and previous exacerbations, comorbidity and current smoking. Exacerbations which require hospital admission are associated with significant in-patient mortality. AECOPD patients presenting with worsening dyspnea, increased sputum volume and purulence should be offered antimicrobial therapy. If treating with antibiotics, treatment must include coverage for *Haemophilus influenzae*, *Streptococcus pneumoniae* and *Moraxella catarrhalis* in all cases, but other bacteria (such as Gram-negatives) may need to be covered depending on the condition of the patient. Antibiotics, particularly macrolides and

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Tip of the month

Over 50? Get an extra serving of milk or alternative (yogurt, cheese, soy milk) daily to help with bone health.

To read full article: http://morethanmedication.ca/en/article/index/nutrition_aging

Interesting Research

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fluoroquinolones, when administered under suitable conditions, shorten the clinical course and prevent severe deterioration. Possible complications resulting from untreated severe AECOPD surpass the potential risks from the use of antibiotic therapy. Additional anti-inflammatory and immunomodulatory actions of some antibiotics may contribute to their efficacy in AECOPD.

Pulmonary rehabilitation reduces depression and enhances health-related quality of life in COPD patients--especially in patients with mild or moderate disease.

Bratås O, Espnes GA, Ranestad T, Walstad R. Faculty of Nursing, Sør-Trøndelag University College, Trondheim, Norway. *Chronic Respiratory Disease*. 2010;7(4):229-37.

Abstract

The first objective of the study was to evaluate a 4-week inpatient pulmonary rehabilitation program on exercise capacity, health-related quality of life (HRQL) and psychological distress in patients with COPD. The second objective was to investigate the influence of gender, age, disease severity, co-morbidity, anxiety and depression on improved HRQL after rehabilitation.

The study comprised 136 consecutive patients from baseline to follow-up with mild-to-severe COPD. Exercise capacity was measured by the 6-min walking distance test, disease severity by spirometric tests, HRQL by The St. George's Respir-

atory Questionnaire and psychological distress by the The Hospital Anxiety and Depression Scale. Variables on socio-demography and co-morbidity were self-reported. Exercise capacity was improved from baseline to follow-up with a score difference of +44 metres ($p = 000$). Except for the activity score, HRQL was significantly improved: a change of -3.5 for the symptom score ($p = 014$), -3.1 for the total score ($p = 003$) and a clinical significant change of -4.0 for the impact score ($p = 002$). The anxiety score did not change significantly after rehabilitation (-0.1 , $p = 545$), though there was a significant reduction of the depression score (-0.8 , $p = 002$) and a 10.4% reduction in the prevalence of possible depression cases ($p = 017$). Patients with forced expiratory volume in 1 second $\geq 50\%$ predicted were 4.2 times more likely to achieve a clinical significant improved HRQL after rehabilitation than patients with forced expiratory volume in 1 second $< 50\%$ predicted (95% confidence interval [CI] 1.7-10.3, $p = 002$).

A 4-week inpatient rehabilitation program improves HRQL and exercise capacity and reduces depression in COPD patients. Patients with mild or moderate disease are more likely to achieve an improved HRQL after rehabilitation than patients with severe or very severe disease.

Long-term outcome in 42 pediatric liver transplant patients with alpha 1-antitrypsin deficiency: a single-center experience.

Hughes MG Jr, Khan KM, Gruessner AC, Sharp H, Hill M, Jie T, Kandaswamy R, Humar A, Payne WD, Gruessner RW. University of Minnesota, Minneapolis, Medical University of South Carolina, Charleston, SC University of Arizona, Tucson, AZ, USA.

Clinical Transplantation. 2010 Nov 16. doi: 10.1111/j.1399-0012.2010.01371.x. [Epub ahead of print]

Abstract

Introduction: We examined the long-term outcome of transplantation for alpha 1-antitrypsin deficiency (A1ATD).

Method: Data were reviewed on 42 transplants in 35 children with A1ATD over 42 yr and compared with 129 transplants in 116 children with biliary atresia (BA). Results: Over 50% of patients were followed up for > 10 yr. A1ATD were older than BA at transplantation and transplanted earlier in the course of liver failure. Patient survival was greater in A1ATD than BA: one-yr post-transplant, 82.7% vs. 67.9%; five yr, 76.5% vs. 60.2%; and 10 yr, 76.5% vs. 55.9%. Death-censored graft survival was similar: one-yr post-transplant, 68.4% vs. 66.2%; five yr, 68.4% vs. 55.8%; and 10 yr, 68.4% vs. 52.5%. Deaths were from infection, hemorrhage, and graft failure < 6 months post-transplant. The age at transplantation and the degree of liver dysfunction were related to the differences in graft and patient survival between A1AT and BA.

If you know of any research, 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

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

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