



The Inherited Heart Rhythm Times

December 2015
Issue 2

Points of Interest

CASPER Wins 1st Prize in Venice, Italy:

'Whole-Exome Sequencing Identifies a Potentially Pathogenic Variant In a High Proportion of Idiopathic Ventricular Fibrillation Probands from the CASPER Registry'

Nice work Dr. Rafik Tadros
- Montreal Heart Institute.

Canadian Cardiovascular Congress (CCC) 2015

Three CASPER projects were presented by UBC fellows and residents.

Heart Rhythm Society (HRS) 2016 Submissions

'Genetic Analysis in the Evaluation of Unexplained Cardiac Arrest: From the Cardiac Arrest Survivors with Preserved Ejection Fraction Registry (CASPER)'
- Dr. Greg Mellor

'Dynamic 3-Dimensional Electrocardiographic Assessment in Victims of Unexplained Cardiac Arrest and Inherited Arrhythmia'
- Dr. Christian Steinberg

'Phospholamban (RI4del) Cardiomyopathy: A Canadian Perspective' & 'Mitral Valve Prolapse in Unexplained Cardiac Arrest'
- Dr. Chris Cheung

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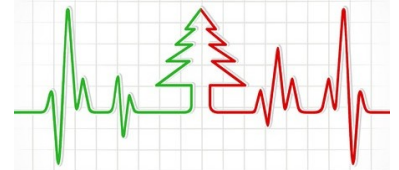
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Greetings from CASPER, ARVC, LQTS National registry headquarters in Vancouver:

As we finish off 2015 we look back on a great deal of good work and collaboration with our partners in the Canadian Genetic Heart Rhythm (CGHR) network. We are grateful to the coordinators for going the extra mile especially during the past month with the frenzy leading up to the Heart Rhythm Society (HRS) abstract deadline.

Together with those willing to share their data and donate a sample for bio banking, we will help to improve the understanding, care and treatment for all those affected by inherited heart rhythm conditions.



Growing Our Research Collaboration

It's been an exciting year as the CGHR network has received CIHR funding to support the National Long QT Registry and Bio bank project currently funded by the Heart and Stroke Foundation of Canada (HSF).

Our grant application for the inaugural Cardiac Arrhythmia Network of Canada (CANet) competition for the program titled: 'Innovative Strategies to Reduce the Risk of Sudden Death: Novel Clinical and Population Approaches' was successful and funded in the summer.

The program will build on the existing CGHR network to expand our patient engagement, promote public awareness and improve knowledge translation to reduce Sudden Cardiac Death in Canada.

The coordinating centre management team (Karen and Mike) have been slowly making their way to our friends at CASPER centres in Winnipeg, Hamilton, Ottawa, Quebec, St. Mike's, Sick Kids in Toronto and Victoria. Next stop is London, Ontario who is the highest enrolling centre for CASPER and a top centre for ARVC, and we are eager to see how they do it.

We have the pleasure of welcoming Dr. Anne Fournier from Sainte-Justine in Montréal, Québec, Dr. Joseph Atallah from Stollery Children's Hospital in Edmonton, Alberta and Dr. Jean-Marc Côté from Centre hospitalier de L'Université, Québec as our new pediatric centres to the CGHR Network. We hope to see more of them in 2016 in the enrolment column as they get up to speed, and at our collaborative meetings.

Welcome to the team!

Patient to Patient: Creativity and Stress

As heart patients, it is vital for us to find ways to manage our day-to-day stress and keep it to a minimum. Human beings, in general, are under much more stress now than they were 5 years ago, and it is taking its toll on our bodies and more importantly, our organs. Recent studies have shown that creativity is a very helpful way to help relieve stress, improve self esteem, increase awareness and help reme-

dy trauma. The beautiful part is that anyone can do it. Being creative helps us take our minds off what is stressing us out. It puts us in a "flow" state, likened to meditation, which is very beneficial to the central nervous system and that is very good for us.

Here are three easy ways to get started...

Adult colouring books: Yes, they are a thing. Get out your crayons, coloured

pencils or sharpie markers and colour until your heart's content.

Doodles: Grab some paper and a pencil or pen and start randomly drawing hearts, flowers, lines, stars or whatever your favourite doodle subject is, don't stop until your page is full. There, how was that?

Write a poem, a story or write in a journal: Get in touch with your inner author and put your thoughts down on paper. It's actually very cathartic.

Dedicate a few hours a week to doing something, anything creative and you will be on the road to zen in no time.

- Vicki Pynn

Learn more:

Paul Brunes Foundation: <http://paulbrunesfoundation.org>

SADS Foundation: <http://www.sads.org/>

Parents Advocating for Cardiac Education (PACED): <http://www.paced.ca/>

Cardiac Arrhythmia Network of Canada (CANet): <http://www.canet-nce.ca/>

Canadian Genetic Heart Rhythm Network (CGHR): www.heartrrhythmresearch.ca

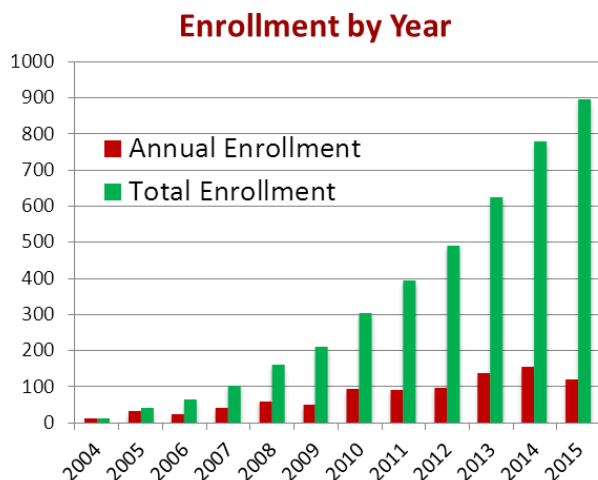
BC Inherited Arrhythmia Program: <https://sites.google.com/site/bciaprogram/>



Registry & Bio bank



Although a slow beginning to enrollment this year, we reached a milestone in the spring when we met our enrollment target of 800 patients. With the collective efforts of all centers, we are ending the year strong with a current total of 896 participants. We are excited and optimistic that we can reach

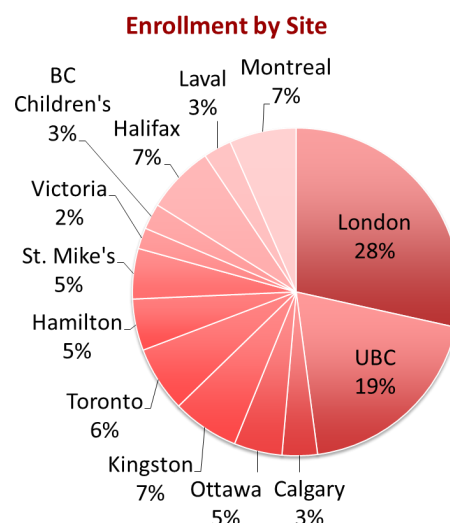


900 before 2016. The next challenge is to increase the enrollment of family members to help paint a clearer picture about how genetics plays a role in inherited heart rhythm conditions. With 376 Unexplained Cardiac

Arrest (UCA) probands and 474 family members of either UCA or Sudden Cardiac Death (SCD) victims, there is still plenty of room to grow and more to learn as we encourage centres to invite family member's to participate in research, and collect blood samples to detect the familial links needed to explain these deaths and near-miss cardiac arrests.

To date, we have collected bio bank samples from 30% of all

CASPER patients. The big five bio banking centres; Halifax, UBC, Kingston, Montreal and Ottawa have been responsible for half of all samples collected. Great work, and a challenge to all sites to increase sample collection!



Registry & Bio bank

Site	Enrollment
London	97
UBC	70
Calgary	34
Hamilton	71
Halifax	110
Winnipeg	1
BC Children's	1
Total	384

Six centres have paced CASPER in annual enrollment for most of 2015 with 79 patients this year. Currently we have 384 patients in the National registry with Halifax leading the pack with 110 patients enrolled.

The bio bank has seen 30 new samples collected in the last 6 months bringing the sample total to 94. Enrollment this year is down to 61% of the total enrollment for 2014. Increased recruit-

ment will bring us closer to our goals of improving our ability to identify and manage ARVC, learning about underlying genetic factors, improving patient care and ultimately preventing sudden cardiac arrest or death.

Congrats to Dr. Healey, Kai, Jenna and Angela in Hamilton who have recruited 17 new participants since the summer, the equivalent of 22% of total national enrollment this year. Thanks Hamilton team for your quality work!

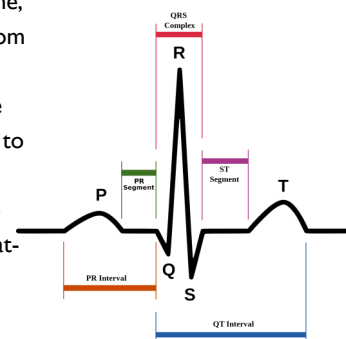
*"Far and away
the best prize
that life has to
offer is the chance
to work hard at
work worth doing."*
- Theodore Roosevelt



Registry & Bio bank

Funding from CIHR for the project titled 'Impact of Early Repolarization on Long QT syndrome' will supplement HSF funding and ensure prolonged support for enrollment and bio banking. The Long QT registry is 104 participants strong in one year of enrollment at St. Paul's/UBC, Vancouver. Newcomers to our CGHR network and the National LQTS registry

are Dr. Anne Fournier from St. Justine, Montreal and Dr. Jean-Marc Côté from Centre hospitalier de L'Université, Quebec. We need a strong presence from the other sites that committed to the project at the grant application stage so that we meet our target enrolment, which is well behind our stated goal.



Pedigree Pro (PDG) - A Single Common Database for all Registries

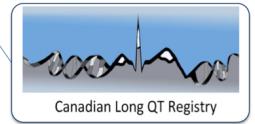
We aim to have all centres using PDG as it creates a simpler, consistent method of entering data into registries that also helps cross-reference patients with data.

PDG is a web-based central database system that is home to the three national registries where 14 centres contribute data. The UBC hosts the server which allows centres to enter and view identified information, however only de-identified data is collected in

the registries to abide by research ethics board (REB) and privacy requirements. We continue to strive for data quality and intuitive data entry processes.

Bio Specimens:

With the simplified bio specimen process implemented in CASPER and LQT we are confident the bio banked samples accrual will sky rocket in 2016 with more centres collecting and shipping samples.



Clarity to Commonly Asked Topics

Reason for referral: refers to the reason why the patient is being evaluated.

- (1) Symptomatic Patient: evaluated due to having symptoms common of inherited arrhythmias (at least 1 symptom checked).
- (2) Asymptomatic Family Member: evaluated due to another family member with an inherited arrhythmia but shows *no* symptoms themselves (no symptoms checked).
- (3) Symptomatic Family Member: evaluated due to another family member with an inherited arrhythmia and shows symptoms of common inherited arrhythmias (at least 1 symptom checked).
- (4) Asymptomatic primary referral: evaluated due to incidental findings of a possible inherited arrhythmia but shows no symptoms (no symptoms checked).

CASPER family ID: is the REG ID # of the first family member enrolled. This does NOT change, even if the affected family member is enrolled at a later date.

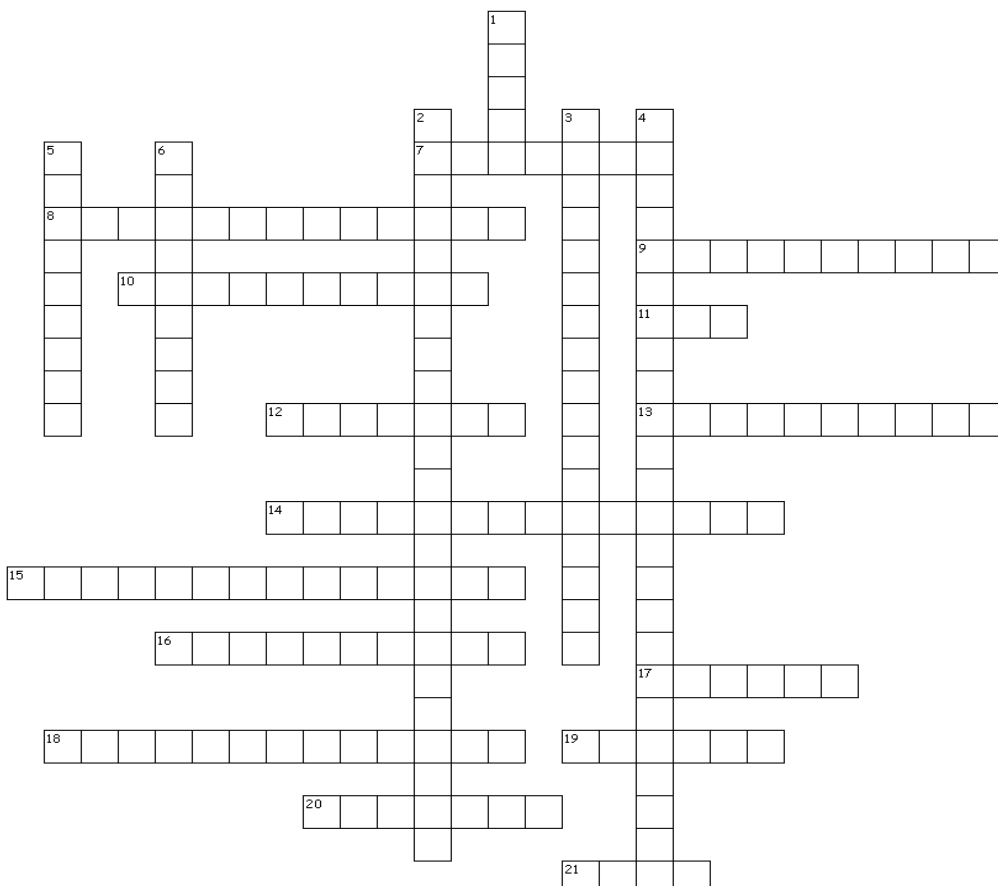
CASPER family proband: Refers to the first family member enrolled in CASPER. If multiple family members are enrolled at the same time, the family proband should be the UCA patient OR SCD victim. If the affected individual is not enrolled, arbitrarily assign one of the first enrolled family members as the 'family proband.' If the affected individual is enrolled at a later date, they will become the family proband, and the family proband patient type will change for all enrolled family members.

CASPER family proband patient type: Refers to the patient type of the current CASPER family proband.

Relationship of patient to sudden death victim: The wording of this field may be interpreted in different ways. For consistency, it should be filled out according to the perspective of the patient being entered. The patient is the _____ of the sudden death victim.

Date of first assessment: refers to the date closest to consent date that a patient was seen by a physician regarding a inherited heart condition.

Crossword Puzzle



Down:

1. A type of cold or rhythm.
2. A condition in which there is uncoordinated contraction of the cardiac muscle of the ventricles in the heart, making them quiver rather than contract properly. (2 words)
3. ECG.
4. UCA. (3 words)
5. A small device that's placed in the chest or abdomen to help control abnormal heart rhythms.
6. Cardiac channelopathies are a group of _____ that affect the electrical system of the heart.

Across:

7. This wave can be seen on the ECG of some ARVC patients.
8. CPVT is an example of a _____.
9. The awareness of an unusually fast heart beat.
10. A name for abnormal heart rhythms.
11. Automated External Defibrillator.
12. This syndrome was named after the three brothers who first described the syndrome in 1992.
13. These are put on your chest, arms and legs for an ECG.
14. Main types include dilated, hypertrophic and restrictive.
15. Ultrasound of the heart.
16. An example of a beta blocker.
17. A friendly ghost or study name.
18. 24 hour heart test. (2 words)
19. A treadmill test is also called a _____ test.
20. Where is the CCC being held this year? (Hint: Answer can be found within this newsletter).
21. This syndrome takes its name from its characteristic appearance on an ECG (acronym).



Our Collaborators

We have the pleasure of working with some of the greatest research teams and individuals across Canada and had the pleasure of meeting many of the research staff and physicians at other centres this past year during site visits, conferences and meetings. Here are some of the great people who work tirelessly to contribute to the CASPER, ARVC and LQT registries with the common goal of learning more about inherited heart rhythms.

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NEW

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SickKids, Toronto, ON

**Dr. Colette Seifer &
Rebecca Medeiros**
St. Boniface Hospital, MB



Centre of Attention: Hamilton, Ontario



Population Health Research Institute
Kai Fan, Angela Frechette, Dr. Jeff Healey, Jenna Spears

Dr. Jeff Healey has an amazing research team in Hamilton, Ontario committed to the CASPER and ARVC registries along with the many other studies they are involved with. Our visit with them in June 2015 strengthened a relationship between centres in different parts of Canada and gave us insight to their successes and roadblocks in enrolling patients. Over the summer Jenna Spears, a medical student, immensely improved the data quality in Hamilton. Hamilton currently has 45 patients enrolled in CASPER and 71 in ARVC with 13 ARVC bio bank samples collected. Thank you all for your work and dedication!

Dr. Andrew Krahn



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CANet
Cardiac Arrhythmia Network of Canada
Réseau canadien sur l'arythmie cardiaque



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Vancouver Coastal Health
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Questions, suggestions or comments?

We would like to hear from you.

Please email Mike Thai at mthai@providencehealth.bc.ca