

The Inherited Heart Rhythm Times

May 2015
Issue 1

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Where are we now?

Another May and preparations for Heart Rhythm 2015 are nearly complete.

Congratulations to Dr. Sanatani and Sonia at BC Children's Hospital for enrolling the 800th CASPER patient and a big thank you to all centres for their contributions.

We are excited about growing our network through the addition of two collaborating centres. We welcome

Dr. Colette Seifer and coordinator Rebecca Medeiros from St. Boniface Hospital in Winnipeg who are just beginning enrollment for CASPER and ARVC while building an Inherited Heart Rhythm clinic.

Pediatric enrollment should soon receive a boost as Dr. Robert Hamilton and Meena Fatah at SickKids in Toronto complete their regulatory process. We welcome both centres. ARVC newcomers are also Dr. Seifer

and Rebecca as well as some interest from Dr. Benoit Coutu in Montreal.

"Congratulations to

Dr. Sanatani and Sonia

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Hospital for enrolling

the 800th CASPER

patient..."



Data Registry and Biobank

Since its initiation, 330 probands and 472 family members have been enrolled in 13 centres across Canada. Of the 802 patients enrolled, we have been able to diagnose 494 patients. Thus, there is still an immense amount of knowledge to be discovered regarding sudden unexpected cardiac arrests.

CASPER Biobank sample collection

Sites Actively Biobanking

Halifax
Kingston
Montreal
Vancouver

since inception totals a heart-stopping 217. (That's 27% of all CASPER patients!) Since 2014, 96 samples have been collected across four sites; Halifax, Kingston, Montreal and Vancouver. These sites have been responsible for 44% of all CASPER samples collected.

Casper Enrollment by Site

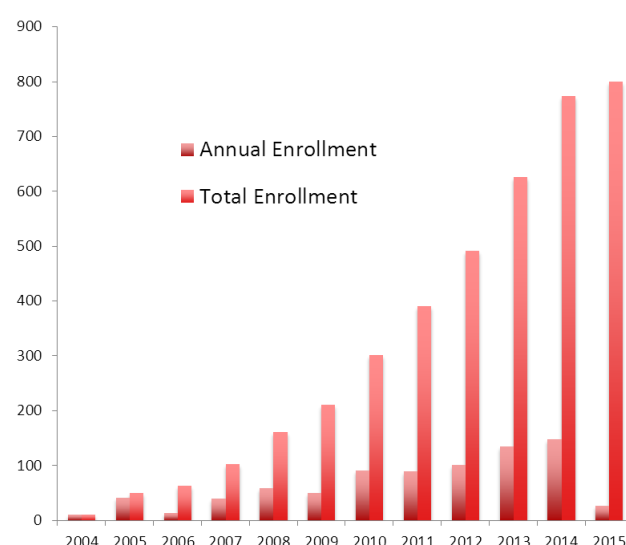
Site	Enrolled
London	246
UBC	149
Montreal	57
Toronto	56
Kingston	54
Halifax	53
Hamilton	37
St. Mike's	37
Ottawa	32
Calgary	28
Laval	23
BC Children's	18
Victoria	12

Heart Rhythm 2015 Presentations

- **PO01-109 A Novel SCN5A Mutation Associated With Epinephrine-Triggered Ventricular Arrhythmia**
May 13 6:00-7:30pm
EPicentre Exhibit Hall
- **PO03-196 Characterization of QT response to exercise in Anorexia Nervosa**
May 14 2:00-5:00pm
EPicentre Exhibit Hall
- **PO03-213 Post-Mortem Tissue Genetic Testing Identifies a KCNH2 Variant Not Previously Reported in Cases of SIDS**
May 14 2:00-5:00pm
EPicentre Exhibit Hall
- **PO05-201 Cost Analysis of Patients Referred to the Inherited Heart Rhythm Registry: Clinical Assessment and Diagnostic Evaluation**
May 15 2:00-5:00pm
EPicentre Exhibit Hall
- **PO05-205 Flecainide Monotherapy for CPVT**
May 15 2:00-5:00pm
EPicentre Exhibit Hall
- **PO06-84 Influence of Right Bundle Branch Block on ECG Morphology of RVOT Ventricular Tachycardia**
May 16 9:30am-12:00pm
EPicentre Exhibit Hall
- **CHRS Strategic Planning Meeting**
May 13 7:30am-5:00pm
The Colonnade Hotel

Year	Annual	Total
2004	10	10
2005	40	50
2006	13	63
2007	39	102
2008	58	160
2009	50	210
2010	91	301
2011	89	390
2012	101	491
2013	134	625
2014	148	773
2015	29	802

CASPER Enrollment by Year





Data Registry and Biobank

Current ARVC Sub-studies

- Distinguishing Phospholamban (PLN) from ARVC using Novel Techniques and Serial Evaluation—[Application]

Since inauguration in January 2013 the ARVC registry has grown to 319 participants across five centres nation-wide.

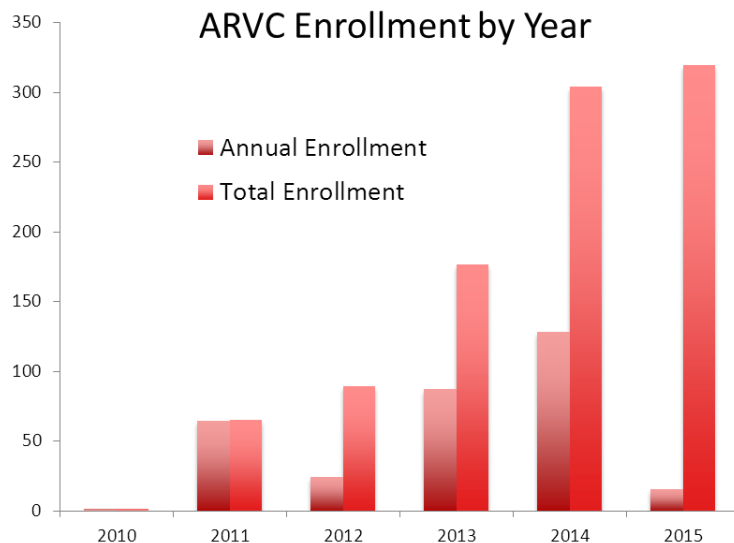
Biobanking at UBC Vancouver and Halifax began in 2013 and thus far 64 samples have been collected. Samples are currently being stored in Hamilton, ON, at the Population Health Research Institute.

Great work to Vicki Newman and Dr. Gardner at Halifax for collecting the most samples year-to-date.

ARVC Enrollment by Centre

Site	Enrollment
London	97
UBC	43
Calgary	26
Hamilton	55
Halifax	95
Total	319

ARVC Enrollment by Year



"We hope to expand enrollment across all CASPER sites"



Data Registry

With recent approval for funding, the national LQTS registry has started strong with 62 patients enrolled at UBC Vancouver.

We hope to expand enrolment across all CASPER sites.

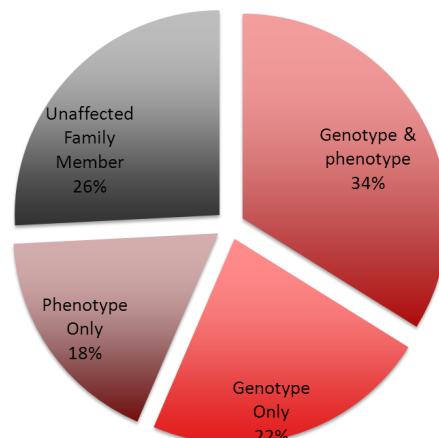
The LQTS study is currently fund-

ed by the Heart and Stroke Foundation of Canada with hopes that

CHIR approves a recent grant application.

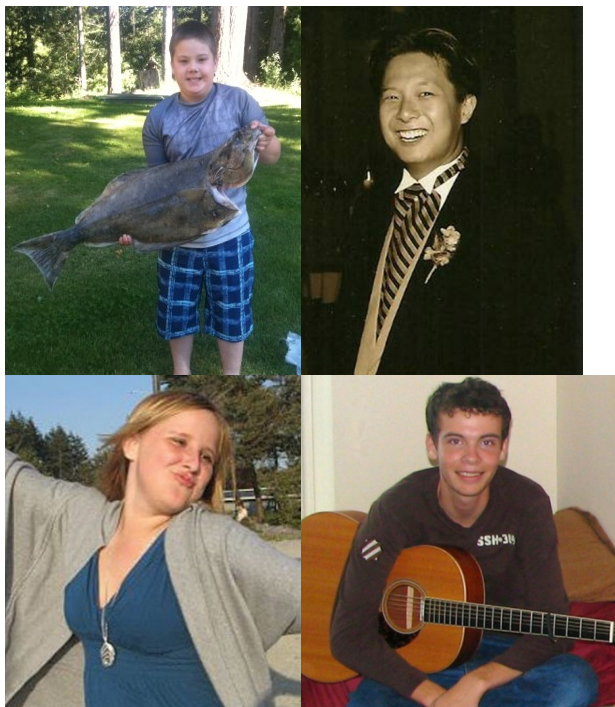
LQTS Registry Enrollment

Patient Type	Enrolled
Genotype & Phenotype Positive	21
Genotype Only	14
Phenotype Only	11
Unaffected Family member	16
Total	62



In Memory of Our Patients

Word Search



H I Z G Q I C K Z P X P B N U D E P Y T O N E G Z K A W W H
X K C Y F W R F Q E Q H U P J K J E N X X G P O I V U B S D
Z S J T O R A O C U R Y S C M F G C X X O Y Z Z F L Z W D Y
N A K L L C I H T J F E X D T S I Z X I Q E V S D Z N E P N
X R E Q L N A F Z A H A P F N T C I H P R O M Y L O P I D W
E K V Q O M Q S N J N Q M S O G N L B C I N E G O H T A P S
E D H Q W E P V P A G I G I A G C H C M X O D L C A G A U S
H G Q C U B I I H E V K D M L C N N L Y J T V C S U C A X B
N M O S P I W P M C R V Q R J Y O K V C G E D S T R E S S G
D N A B O R P U U R F Z J O C H Q N J P L G I Q A A A N C
F O K A A R X W T S I A A E Q O S I U C V W N Q L S Q W R J
S G B I L H A T L V C V E M B Z C F S R C D W F V Y W K M I
J G B N O A H V Y K F R T S I Z B X A T V G G X Q M R J W K
A G I O W L D M L N Y T A R E L J F F M O I Y I X P H U P N
T E O I I Y C A Y A X P E P B R Y J I Y W R J O X T Y C O Y
T A B T Y E Y I T F V Y E U H D O H N U Z N Y V A O M C N L
L N A A J M E C T A K B T M L E N L V C I A X V Q M S E A I
H V N T R O L Z M A E N C H F P N G E I Y N Z C P A H Q Q X
Q K K U Q R K N C G M N F M U Z E O S R J E N V R T J H O A
Y Q W M T D D W Y X T O T H S G F D T V H K I R D I H X W T
D U E R A N I I A C Z D T R N F Q L I Y R S Z A U C R T A A
C A N Q L Y Q N Q Y P V W P Y Z C G G G P L C B E Q U Q N N
X I L A P S O J B T R V R P M H N K A O R E X I G E N H P H
C A R D I A C A R R E S T G S Y Q V T Z G E R C T R R E G H
V Y S W Q D X R I N L I M R Q K S Q O B D T E B A E C Q L Z
L M R O C A G V L U C T V Q I H V L R F I I B P Y B N Z B Z
B H X U F G T N E M L L O R N E G Y G Z U X A V R R T E O I
Z B G H R U B A C U R G L R U N T Y U N M H X P V O E V G R
X B M P R R W J Q F R S F Z W G Y C H D B S G S S E Q U U K
V T C O B B I R S C R Q R I I U P N S N A Y H P B U K J Z S

CASPER
ARVC
LQTS
RESEARCH
BIOBANK
ECG
STRESS
ASYMPTOMATIC
PROBAND
CASPERFAMILY
CPVT
BRUGADASYNDROME
CARDIACARREST
PHENOTYPE
GENOTYPE
MUTATION
PATHOGENIC
VUS
POLYMORPHIC
GENETICS
FAMILYHISTORY
UCA
FOLLOWUP
SYMPTOMATIC
INVESTIGATOR
COORDINATOR
DATAENTRY
PEDIGREEPRO
ENROLLMENT

The Database & Data Entry

FAQs:

1. **What is the family ID?**
2. **What does "Reason for Referral" mean?**
3. **Where can we get Biobank kits for CASPER?**
4. **How do we differentiate strength of diagnosis?**
5. **What is the difference between critical fields and required data fields?**
6. **How should we prioritize data entry?**

Pedigree Pro (PDG) is home to the National registries for CASPER, ARVC and LQT. It is a site-based webpage that acts as a central research database where patient data entered can be selectively placed to each of the studies. This secure site allows local entry of identified patient information, however only de-identified data is uploaded to the national registries to abide by re-

search ethics board and privacy requirements.

We are working to reduce the workload for sites by asking them to upload tests and test reports rather than enter the data on the test page itself. It is simpler to de-identify, scan and upload a test than to chase the investigator for test data. The Coordinating centre is continuously working to make data collection and

completeness as effective and painless as possible.

We will also start sending data queries quarterly and ask that sites send invoices quarterly to create consistent payment schedules.

If you have any questions regarding the database or your site's data, please contact Mike Thai at

604-682-2344 ext. 66860 or mthai@providencehealth.bc.ca

Answers:

1. A family ID links individual patients who are related together. The ID number itself corresponds to the registry ID number of the first patient enrolled in that family.
2. "Reason for Referral" indicates why a patient was seen. If they show symptoms then they would be a symptomatic patient or a symptomatic family member (of a patient previously seen). If a patient does not have symptoms then they would be an asymptomatic patient (primary referral) or an asymptomatic family member (of a patient previously seen).



RESEARCH

"...reduce workload

for sites by asking

them to upload

tests and test

reports..."



3. If you are running low on CASPER Biobank kits, let us know and we will ship kits to your site. Please also take note of the expiry dates on the collection tubes.
4. Strength of diagnosis is defined in three categories; (i) definite: clinical & genetic testing both positive or clinical testing clearly positive, (ii) probable: clinical or genetic testing is positive, and (iii) possible: where one or more clinical test is borderline but not conclusive.
5. Critical fields contain a red ' ? ' and must be addressed whereas required data fields are required to reach baseline data entry completion and meet payment criteria. These fields can vary between studies.
6. Although all the data is important, we ask that sites put more emphasis on uploading de-identified tests and reports.

Strength in Numbers

We could not have achieved what we have thus far without the hard work and contributions of our collaborators across Canada.

This section is dedicated to them and all their efforts.

London Health Sciences Centre

Investigators:

George Klein
Jason Roberts

Coordinators:

Lynn Nymann
Kate Relou

University of British Columbia

Investigator:

Andrew Krahn

Project Manager:

Karen Gibbs

Toronto Hospital for Sick Children

Investigator:

Robert Hamilton

Coordinator:

Meena Fatah

University of Calgary

Investigators:

Robert Sheldon
Brenda Gerull

Kingston General Hospital

Investigator:

Chris Simpson

Coordinator:

Sharlene Hammond

University of Ottawa Heart Institute

Investigators:

David Birnie
Martin Greene

Coordinator:

Karen MacDonald

Toronto University Health Network

Investigator:

Vijay Chauhan

Coordinators:

Adrian Susko
Khurana Mamta

Hamilton Health Sciences Centre

Investigators:

Jeffrey Healey

Coordinator:

Kai Fan

St. Michael's Hospital

Investigator:

Paul Angaran

Coordinator:

Katherine Allan

Victoria Jubilee Hospital

Investigators:

Lawrence Sterns
Rick Leather

Coordinators:

Fritha Munday
Deb Parfett

BC Children's Hospital

Investigator:

Shubhayan Sanatani

Coordinator:

Sonia Franciosi

Queen Elizabeth II Health Sciences Centre

Investigator:

Martin Gardner

Coordinators:

Lisa Hebert
Victoria Newman

Institut Universitaire de Cardiologie et de Pneumologie de Québec

Investigators:

Jean Champagne
Francois Phillipon

Coordinator:

Paule Banville

Montreal Heart Institute

Investigator:

Mario Talijic

Coordinator:

Marie-Josée Billo

St. Boniface Hospital

Investigator:

Colette Seifer

Coordinator:

Rebecca Mediros



Meet The UBC Vancouver Team

The CASPER, ARVC and now LQTS Vancouver coordinating team is growing. You may have met most of the team already.

Inherited Heart Rhythm Research

St. Paul's Hospital
Rm 220, 1033 Davie St
Vancouver, BC
V6E 1M7

604-682-2344 ext. 63260
kgibbs@providencehealth.bc.ca



Dr. Andrew Krahn
MD, FRCPC, FHRS

Dr. Krahn is the UBC Head of Cardiology and the Director of our great team.

Dr. Christian Steinberg
Christian will be starting his research fellowship with us in September.

A Note from Karen:

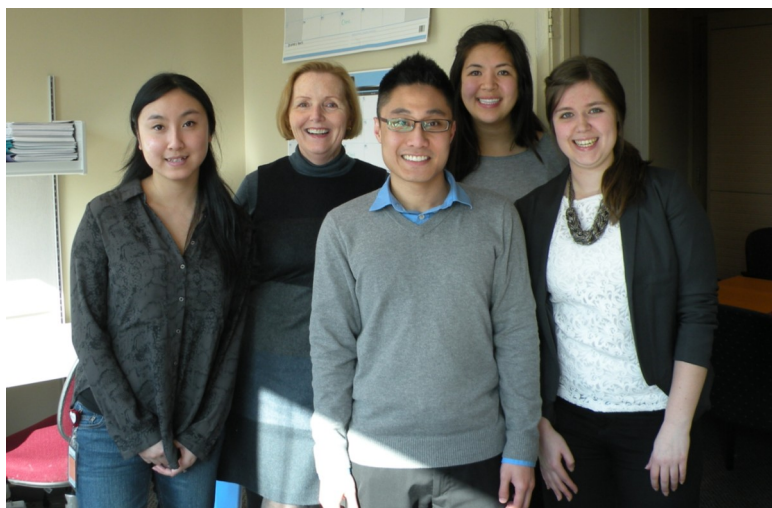
Thanks to all the coordinators for such dedication to recruitment, consenting and data entry during changes to the protocols and data entry system over the last two years. Since taking on the role of Project Manager from Laura and Bonnie before her, it has been my greatest pleasure to get to know all of you.

I began as a CASPER coordinator 10 years ago while working in Electrophysiology (EP) research at BC Children's heart centre I can still remember asking Bonnie what on earth is CPVT and really you want us to give kids epinephrine and procainamide just to see how it affects their heart rhythm? Little did I know of the world of Inherited Heart Rhythm research that was opening to me.

Since coming to work with Dr. Krahn just before the start up of the BC Inherited Arrhythmia Clinic (BCIAP) many things once done for CASPER are now considered standard clinical practice. The clinical care of the BCIAP is based on sound research in the area of inherited heart rhythms from CASPER.



Karen Gibbs, RN CCRP
Research Manager



Brianna Davies
Research Assistant

Mikyla Janzen
MSc. Graduate Student

Jennifer Lin
Research Assistant

Michael Thai
Research Assistant

Congratulations to Brianna upon just receiving her BSc. from UBC this month and from also being a successful candidate for the UBC Genetic Counseling Masters Program.

Brianna started with us as a volunteer in her third term in the UBC program, then working full time with us last summer and will be returning this month full time until her program starts this coming September (YAY!)

Brianna, if we gave you any time off for fun what would you like to do this summer?

"I plan to visit New York City on vacation this summer."

Known to most of you as Mika, she came to us as a summer student two years ago and has graciously continued working part time as she completed her BSc. She has been accepted into the UBC Experimental Medicine program beginning in July 2015. We are thankful to have her not too far away utilizing the CASPER and ARVC data for her research projects as she continues to work through her program.

Congratulations Mika, any travel plans for this summer?

"I will be heading to Liberia to volunteer with the Universal Outreach Foundation."

Jennifer started working with us in 2013 helping with mostly administrative tasks in the office and has recently become our #1 recruiter/consenter for all our local patients who meet inclusion criteria for the national registries.

Her administrative talents and her pleasant manner with patients make her a much appreciated part of the team.

What are your plans for summer fun this year Jen?

"I will be busy planning my upcoming wedding this summer and trying to get outdoors to enjoy the Vancouver sunshine whenever possible."

Once upon a time Mike started working with us one day a week when he wasn't busy at the bank, to help with our bookkeeping and data entry. His economics background has been useful in keeping the study funds well organized.

Starting with us full time this past February he is quickly becoming our "go to" for all things finance, budget and data related. Look for Mike's data queries and payment schedules quarterly starting very soon.

Mike, we hear rumors of a Vegas trip for you in the near future?

"It will be my last trip before I'm taken off the market and tie the knot."

What would you like to see in the newsletter?
Questions, suggestions or comments? We would like to hear from you. Please send an email to Mike Thai at mthai@providencehealth.bc.ca

*information contained in this newsletter is as of May 5, 2015



HEART & STROKE
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