

# JOHNS HOPKINS ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY PROGRAM

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Winter 2020

★ ★★ ★★★!!!HAPPY NEW YEAR!!!★★★ ★★ ★

A new year is upon us! What vision do you have for 2020? The Johns Hopkins ARVC Program is looking forward to new and continued collaborations as we work to improve our understanding of this rare condition. More importantly, we are excited to build and strengthen our partnership with those impacted by ARVC. We have learned a great deal over the past two decades, but many important questions remain such as safe exercise level, the best medications, progression, etc. We are thrilled to be enrolling participants in our first ever NIH sponsored clinical trial in ARVC. You can read more about the Flecainide Clinical Trial in this edition. This is an extremely important first step in clinical trials in ARVC. We absolutely need your support in these endeavors. From all of us in the ARVC Program, we wish you good health, happiness, and success in the coming year and always!

With the new year comes the announcement of our annual seminar! The 21<sup>st</sup> Patient and Family Seminar will be held the weekend of May 2<sup>nd</sup>, 2020 and we hope to see many of you here. Details are enclosed. Please register early and secure your hotel room, as they do go fast.

~ The ARVD/C Program

## 21<sup>st</sup> Annual ARVD/C Patient and Family Seminar

You and your family members are invited to join us for our annual ARVD/C Seminar! You won't want to miss this unique opportunity to meet and network with other families affected by ARVD/C and to learn the latest advances in the field. We have a great team of speakers lined up and will be posting the agenda shortly as soon as it is finalized. We are excited to welcome guest speakers, Professor Perry Elliott, MD, and Dr. Shannon Hourigan, PhD, MS. Professor Elliott is the Clinical Lead of the Inherited Cardiovascular Disease Unit at the Bart's Heart centre and the Professor of Cardiovascular Medicine at the University College London. Dr. Hourigan is a clinical psychologist specialist working with the Inherited Cardiac Arrhythmias Program at Boston Children's Hospital. In addition to our invited speakers, many from our Hopkins ARVC Program will be presenting on genetics, diagnosis, treatment strategies, heart failure, basic science research, as well as research opportunities.



Professor Perry Elliott, MD  
University College London



Shannon Hourigan, PhD, MS  
Boston Children's Hospital

Make plans to come into Baltimore early to attend a reception at the Homewood Suites by Hilton Friday evening (7pm-9pm). **Please register early!** There is no registration fee for this seminar, but we do appreciate early registration for planning purposes. Registration is required and if you need to cancel, please let us know ASAP. All are welcome!

**WHEN:** Saturday, May 2<sup>nd</sup>, 2020 8:00am-5:00pm

**WHO:** Patients and Families affected by ARVD/C, Healthcare Professionals

**WHERE:** PreClinical Teaching Building  
725 North Wolfe Street  
Baltimore, Maryland 21287

**REGISTRATION:** ALL participants must register! It is also helpful to list names of family members that will also be attending. **Please register all attendees separately!**

Register online by April 15<sup>th</sup>

<https://tinyurl.com/2020ARVCSeminar>

# 21<sup>st</sup> Annual ARVD/C Patient and Family Seminar

Presented by  
The Johns Hopkins ARVD/C Program

## **HOTEL ACCOMMODATIONS – RESERVE EARLY!!!**

Hotel rooms are available (limited) at the Homewood Suites by Hilton, Baltimore Inner Harbor (625 South President Street, Baltimore, Maryland 21202) at a special rate of \$189/night plus tax until March 30<sup>th</sup>, 2020 or as long as they are available.

Hotel reservations can be made through the Homewood Suites Central Reservations Line at **443-392-9013**. Request **Johns Hopkins ARVD/C 2020** group rate. Reservations can also be made through the online booking link:

<http://homewoodsuites.hilton.com/en/hw/groups/personalized/B/BALHWHW-JVD-20200429/index.jhtml>

The hotel front desk can be reached at 443-392-9013. Check-in 3pm / Check-out 11am. Self-Parking is available at a rate of \$23 and valet \$35 per day.

## **TRAVEL TIPS**

The Baltimore/Washington International (BWI) Thurgood Marshall Airport is the closest international airport to Johns Hopkins ([www.bwiairport.com](http://www.bwiairport.com)). It is about 30 minutes from the seminar location.

Two buses will be available to transport seminar participants to the seminar on Saturday, departing only from the Homewood Suites at TBD. The bus will leave the seminar at 5:00pm to return to the Homewood Suites.

Taxi Services – For Taxi services call Sun Cab at 410-235-0300 or Yellow Cab at 410-752-1096. Uber and Lyft are plentiful in Baltimore as well.

## **PARKING AT THE SEMINAR**

If you are not taking the bus from the hotel, FREE parking (Saturday from 7am to 7pm) is available at the Washington Street Garage (701 Washington Street, Baltimore, MD 21205), just one block from the seminar location.

## **SPECIAL EVENT**

Join us for a Meet 'n Greet Reception, **7:00-9:00pm**, on **Friday, May 3<sup>rd</sup>, 2019** in the Great American Grill at the Homewood Suites. Please register for this event when you register for the seminar or contact Crystal.

## **CLINIC CONSULTATIONS – REQUEST YOUR APPOINTMENT NOW!!!**

Dr. Hugh Calkins and Dr. Hari Tandri and the genetic counselors will be available Thursday, April 30<sup>th</sup>, Friday, May 1<sup>st</sup> and Monday, May 4<sup>th</sup> for consultations. Diagnostic tests can also be arranged if necessary. We ask that if you live locally to please consider arranging your appointment at another time to allow new patients and patients traveling from a distance an opportunity to schedule. These appointments will be billed to your insurance. Please contact Crystal **ASAP** to schedule as these appointments go quickly.

## **QUESTIONS**

Contact Crystal Tichnell, MGC, RN at 410-502-7161 or [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu)

### Effects of Flecainide on Cardiac Arrhythmias in ARVC Patients

Johns Hopkins IRB00197430

Funded by The National Institute of Health (NIH)  
Principal Investigator: Hugh Calkins, MD  
Enrolling Site Coordinator: Crystal Tichnell, MGC, RN

The purpose of this study is to assess the effect of the antiarrhythmic drug, flecainide, on cardiac arrhythmias in individuals with ARVC. Participation in this study will last for about 10 weeks. This is a randomized, double-blinded, crossover study which means for part of the study you will be given the study drug, flecainide, and for the other part of the study you will be given a placebo. After 28 days you will switch, so everyone will be given the study drug at some point in the study. Neither you nor your enrolling physician will know if you are given the study drug or placebo first and that process is randomized.

During the study you will be asked to wear a monitor for 7 days on 2 occasions. We will also obtain ECGs and blood draws to assess flecainide levels through a home visit. Remote device interrogations will be obtained by your enrolling center. This study requires **one** in-person visit to discuss your enrollment and to sign the consent form.

#### **To participate in this study you must:**

- Be 18 years of age
- Meet the diagnostic criteria for ARVC
- Have a minimum of 500 PVCs on a recent 24 hour Holter monitor
- Have an implantable cardioverter defibrillator (ICD) with remote interrogation capability
- Be on a beta-blocker such as metoprolol, propranolol, atenolol, nadolol, carvedilol, unless contraindicated
- If prescribed, be willing to discontinue sotalol, quinidine, procainamide, propafenone, disopyramide, dronedarone, phenytoin, or mexilitene for 5 days with subsequent repeat 24 hour Holter
- Agree not to use any another antiarrhythmic medication during the 10 weeks of participation, unless for the management of life-threatening arrhythmias
- Agree to use medically acceptable contraceptive measures during participation unless documented as surgically sterile or post-menopausal

If you would like to learn more about this study or to discuss your eligibility, please contact Crystal Tichnell, MGC, RN at [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu) or 410-502-7161. This is an important first clinical trial in ARVC and will pave the way for future clinical drug trials.

## Research Opportunities at Johns Hopkins

### Why Participate in a Clinical Trial?

Clinical trials are so important in discovering new treatments and the safety of those treatments, particularly of rare conditions. The decision to participate in a clinical trial is a personal one and only you can decide if it's the right choice for you. As you think about whether to participate, please take a minute to consider the impact your participation or non-participation might have on overall clinical trials in ARVC. As we all know, ARVC is a rare condition and there isn't much funding for research. So, whatever funding we do get, we need to make the most of it. We need to prove to big funding agencies that individuals with ARVC are interested and willing to participate in new discovery research, otherwise, there will be huge delays in new discoveries, new treatment options, and ultimately finding a cure.

<https://www.nih.gov/health-information/nih-clinical-research-trials-you/basics>

### Relative Safety, Efficacy, and Patient Satisfaction of Standard ICDs versus the Sub-Cutaneous ICD

**Who can participate?** Patients diagnosed with ARVD/C and have:

- ✓ S-ICD implanted
- ✓ Transvenous ICD implanted after January 2013

**What do I have to do?** Contact Crystal ([ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu)).

You will need to sign a consent form, send us your medical records, and complete online questionnaires.

**Do I have to travel to Johns Hopkins?** No

**Once enrolled, please remember to check your email for reminders to complete the online questionnaires. It is really important that we continue to collect this follow up data. Thank you for your participation.**

Sponsored by Boston Scientific  
PI: Hugh Calkins, MD  
Johns Hopkins IRB NA\_00042471 (*Predictors of Implantable Cardioverter Defibrillator (ICD) Firing in Right Ventricular Dysplasia*)

### Clinical and Genetic Investigations of Right Ventricular Dysplasia (ARVD/C Registry)

**Who:** Children and Adults with ARVD/C

**What:** Collection of pertinent past medical records and continued collection for 5 years. A blood sample for DNA for genetic mapping of ARVD/C genes will also be collected.

**How to Join:** Contact Crystal at 410.502.7161 or [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu).

### Have you had an epicardial ablation?

We are looking for people with ARVD/C who have had an epicardial ablation to join our Registry. Help us discover how this new technique affects the course of ARVD/C! Contact Crystal at 410-502-7161 or [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu)



## 2019 Publications

Carruth ED, Young W, Beer D, James CA, Calkins H, Jing L, Raghunath S, Hartzel DN, Leader JB, Kirchner HL, Smelser DT, Carey DJ, Kelly MA, Sturm AC, Alsaïd A, Fornwalt BK, Haggerty CM. **Prevalence and Electronic Health Record-Based Phenotype of Loss-of-Function Genetic Variants in Arrhythmogenic Right Ventricular Cardiomyopathy-Associated Genes.** *Circ Genom Precis Med.* 2019 Nov;12(11):e002579.

Sharma A, Assis F, James CA, Murray B, Tichnell C, Tandri H, Calkins H. **Misdiagnosis of ARVC leading to inappropriate ICD implant and subsequent ICD removal - lessons learned.** *J Cardiovasc Electrophysiol.* 2019 Oct;30(10):2020-2026.

Cadrin-Tourigny J, Bosman LP, Tadros R, Talajic M, Rivard L, James CA, Khairy P. **Risk stratification for ventricular arrhythmias and sudden cardiac death in arrhythmogenic right ventricular cardiomyopathy: an update.** *Expert Rev Cardiovasc Ther.* 2019 Sep;17(9):645-651.

van Lint FHM, Murray B, Tichnell C, Zwart R, Amat N, Lekanne Deprez RH, Dittmann S, Stallmeyer B, Calkins H, van der Smagt JJ, van den Wijngaard A, Dooijes D, van der Zwaag PA, Schulze-Bahr E, Judge DP, Jongbloed JDH, van Tintelen JP, James CA. **Arrhythmogenic Right Ventricular Cardiomyopathy-Associated Desmosomal Variants Are Rarely De Novo.** *Circ Genom Precis Med.* 2019 Aug;12(8):e002467.

Chelko SP, James C, Tichnell C, Murray B. **The Johns Hopkins ARVC International Symposium.** *Eur Heart J.* 2019 Aug 1;40(29):2387-2389.

Cadrin-Tourigny J, Bosman LP, Nozza A, Wang W, Tadros R, Bhonsale A, Bourfiss M, Fortier A, Lie ØH, Saguner AM, Svensson A, Andorin A, Tichnell C, Murray B, Zeppenfeld K, van den Berg MP, Asselbergs, FW, Wilde AAM, Krahn AD, Talajic M, Rivard L, Chelko S, Zimmerman SL, Kamel IR, Crosson JE, Judge DP, Yap SC, van der Heijden JF, Tandri H, Jongbloed JDH, Guertin MC, van Tintelen JP, Platonov PG, Duru F, Haugaa KH, Khairy P, Hauer RNW, Calkins H, Te Riele ASJM, James CA. **A new prediction model for ventricular arrhythmias in arrhythmogenic right ventricular cardiomyopathy.** *Eur Heart J.* 2019 Jun 14;40(23):1850-1858.

Assis FR, Krishnan A, Zhou X, James CA, Murray B, Tichnell C, Berger R, Calkins H, Tandri H, Mandal K. **Cardiac sympathectomy for refractory ventricular tachycardia in arrhythmogenic right ventricular cardiomyopathy.** *Heart Rhythm.* 2019 Jul;16(7):1003-1010. doi: 10.1016/j.hrthm.2019.01.019. Epub 2019 Jan 21.

James CA, Calkins H. **Arrhythmogenic Right Ventricular Cardiomyopathy: Progress Toward Personalized Management.** *Annu Rev Med.* 2019 Jan 27;70:1-18.



## Clinical Services

The Johns Hopkins ARVD/C Program provides a variety of clinical services. We see patients for second opinion consultations to discuss diagnosis and management, genetic counseling and testing, routine ICD management and family member screening. We can also arrange concurrent cardiac testing.

Patients are seen in consultation with Dr. Hugh Calkins or Dr. Hari Tandri and our clinical genetic counselor, Brittney Murray, to discuss test results, family history, and to provide guidance regarding further management. We see all of our patients for genetic counseling to discuss the diagnosis, the psychosocial impact of living with ARVD/C and with an ICD, as well as to discuss the benefits and limitations of appropriate genetic testing. In selected cases we also offer catheter ablation as a treatment for difficult to manage ventricular tachycardia. Appointments with our heart failure specialist, Dr. Nisha Gilotra can also be arranged. These appointments are billed to your health insurance.

To schedule an appointment, contact Crystal at [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu) or 410-502-7161

## ARVD/C Program Info

### ***ARVC Program Staff***

Hugh Calkins, MD—Director

Harikrishna Tandri, MD—Co-Director

Cynthia James, ScM, PhD—Genetic Counselor

Brittney Murray, MS—Genetic Counselor

Crystal Tichnell, MGC, RN—Genetic Counselor, Nurse

Emily Krupa—Genetic Counselor Assistant

Christal Holmes-Igwebike—Clinic Coordinator

Julia Agafonova—Research Program Coordinator

Apurva Sharma—Research Fellow

**Keep us informed of your most up-to-date contact info!**

**Please send any changes and updated medical records to Crystal via the contact info below.**

**Thank you!**

### **Contact Us**

Johns Hopkins Hospital  
600 North Wolfe Street, Blalock 545  
Baltimore, Maryland 21287  
P: 410-502-7161, F: 443-873-5073  
Website: [www.ARVD.com](http://www.ARVD.com)

We would like to welcome and introduce our new financial clearance and scheduling coordinator, Christal Holmes-Igwebike. Christal has been working at Johns Hopkins since July 2011 when she first started working in the gift shop. Over the years, she found herself working in a variety of positions ranging from patient transporter to patient service coordinator to medical office coordinator. She worked as a senior patient access specialist for 8 years before starting her new position as a financial clearance and scheduling coordinator within the Center for Inherited Heart Disease. We are thrilled to have her join our team as she helps schedule your appointments and works on the insurance aspect of your visit.



## How You Can Help

### Your Support of the Johns Hopkins ARVD/C Program Ensures Our Success

As a charitable, tax-exempt organization, Johns Hopkins Medicine relies on donations to make a difference in the lives of our patients. Supporters of Dr. Calkins, Dr. Tandri, and their team of experts in the ARVD/C Program, become part of our mission to provide exceptional personalized care and to find better ways to diagnose and treat our patients. Here are some of the many ways that you can help:

#### Make a Donation

Donations of all sizes, whether they're one-time or recurring, make a difference and can be made online at [www.arvd.com](http://www.arvd.com), by phone at **443-287-7382**, or by mail (information listed below). There are a variety of ways to make a gift to support our efforts in the ARVD/C Program:

- Make an outright gift of cash or securities
- Become a monthly donor
- Give in honor or in memory of a loved one
- Give through IRA's, wills and trusts
- Leverage matching gifts through your workplace

#### Fundraising

There are many opportunities to become personally involved in raising awareness and much-needed funds on behalf of the Johns Hopkins ARVD/C Program:

- Create an online giving page and leverage social media
- Ask friends to make contributions in lieu of gifts
- Host your own event or auction
- Plan a fundraising event in your community or school
- Contribute a portion of your company's sales

#### The Johns Hopkins Heart and Vascular Institute Development Office is here to help!

We welcome your questions, concerns, ideas, and feedback. Please contact **Adrienne Rose**, Senior Associate Director of Development, at 443-287-7382 or [arose25@jhmi.edu](mailto:arose25@jhmi.edu), for more information.

#### Gifts by Mail:

The Johns Hopkins Heart and Vascular Institute  
600 North Wolfe Street, Blalock 536B  
Baltimore, MD 21287

**\*Indicate the "ARVD/C Program" on the memo line**

## Support Resources

### Looking for support resources?

#### **FACEBOOK Groups (private):**

- ARVD/C Youth Society
- Hope for ARVD
- ARVC can't stop me from...



#### **PLN Heart Disease Foundation:**

<https://www.plnheartdiseasefoundation.org>

Information for those of you affected by PLN or phospholamban type ARVC.

#### **Precision Medicine Website:**

Read “Feeling the Beat”: Stories from the ARVC Community here:

<http://www.hopkinsmedicine.org/inhealth/precision-medicine-centers/arvc>

#### **ARVD/C Mentor Program:**

Connect with a mentor who has navigated the challenges of life with ARVD/C.

Contact Nancy Bogle at [nbstjohn@gmail.com](mailto:nbstjohn@gmail.com) or visit [ARVDHEARTANDSOUL.org](http://ARVDHEARTANDSOUL.org)

#### **Published Personal Stories:**

##### ***Racing Heart: A Runner's Journey of Love, Loss and Perseverance***

by Kate Mihevc Edwards can be purchased from Amazon or through [katemihevcedwards.com](http://katemihevcedwards.com)



*Kate was a passionate runner for years and lived for endurance sports. Until the day doctors told her she'd have to stop—or possibly die. After being diagnosed with ARVC, Kate was forced to mourn the loss of a lifestyle she loved and face a very scary question: What now? With the grit and determination that made her such an accomplished runner, Kate embarked on a journey that taught her to let go of her former self and claim a new life filled with strength, gratitude, and peace. **Racing Heart** is Kate's testament to this journey as she walks readers through her transition from a runner eager to finish the race to a woman who values the preciousness of the present. Written for anyone facing a major life change or hungering for a life beyond the mundane, **Racing Heart** is the inspirational reminder that it is often from the most painful experiences that true joy and passion emerge.*

##### ***Farther Than 26.2 Miles*** by Cheryl Collins Gatons

can be purchased from Amazon



*This is a true story of Cheryl, who went from running simply for the fun of it to qualifying for the Olympic marathon trials, thrusting her into the world of competitive running. It was in that world that Cheryl met another competitive runner, Kevin, and the two fell in love. Their shared passion for running brought the pair together, but it also tore them apart, as Kevin passed away (later determined to be from ARVD) navigating the state cross-country course with an athlete he coached. Cheryl shares her difficult journey in **Farther Than 26.2 Miles**—how running brought her to the love of her life, how it took him away from her, and how it helped her survive after he was gone.*



## Support Resources

We are happy to announce that Jennifer Cory of HEART Initiative will be providing a free, live webinar for our ARVD/C and SADS communities on January 28<sup>th</sup>. Jen will talk about the impact of cardiac disease on quality of life, and discuss key, evidenced-based strategies for reducing anxiety and increasing peace of mind and well-being.



The HEART Initiative is a social impact organization dedicated to substantially improving quality of life for people living with chronic disease. It was founded by Jennifer Cory, a psychotherapist, international speaker, and expert in the field of applied positive psychology. She has also been living with an inherited cardiac disease since 2010.

Jennifer presented at the Johns Hopkins ARVD/C Family Seminar in 2018, and was a keynote speaker and workshop leader at the 2019 International SADS Conference in Atlanta. Through the HEART Initiative, Jennifer offers programs and resources for people living with chronic disease, with a particular emphasis on cardiac diseases. Her work is derived from the science of human flourishing and she teaches resilience building strategies to help people better navigate life with chronic diseases.

Examples of comments Jennifer has received for previous live and online presentations:

*"I can't thank you enough for allowing me to attend the [...] program. It has made a huge, positive impact of my life. I have been coping so much better since then, and my symptoms feel much more under control. I feel I have regained so much of my independence. Thank you from the bottom of my heart for creating this program." (8/19)*

*"Jennifer Cory's break out session was amazing! It was absolutely my favorite and had potential life changing implications for me!" (10/19)*

Registration is required to attend this webinar. Those who register will receive a recording of the session as well. For more information or to sign up for the January 28<sup>th</sup> webinar contact HEART Initiative by following this link: [webinar.heartinitiative.net](https://webinar.heartinitiative.net)



If you have a topic or information you'd like to see discussed in the next newsletter, please reach out to Crystal Tichnell at [ctichnell@jhmi.edu](mailto:ctichnell@jhmi.edu)

Thanks for reading!