

JOHNS HOPKINS ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY PROGRAM – A PRECISION MEDICINE CENTER OF EXCELLENCE

Volume 10, Issue 2

Summer 2020

Typically, this issue of the newsletter reflects on the annual patient and family seminar held in the Spring at Johns Hopkins. Unfortunately, due to the COVID-19 pandemic, our annual event had to be cancelled, something we have never had to do in 20 years! We were beyond disappointed, but certainly your well-being was in the forefront of our minds.

We are so grateful we were able to celebrate our 20th anniversary of the ARVD/C Program at Johns Hopkins last year. If you haven't had a chance to review the presentations from both the patient and family seminar as well as the physician seminar, please take a look at our website. We had over 60 physicians, scientists, and genetic counselors from 12 different countries share their experiences and ideas related to ARVD/C. It truly was a memorable event!

What was your vision for 2020? I bet you weren't counting on having to quarantine, not being able to find toilet paper or hand sanitizer, endless celebrations being canceled, schools closing and moving to online learning, many losing their jobs, some of you may have lost family and friends to the virus, businesses closing (some for good), universal mask wearing, etc. The list goes on and on.

The COVID-19 pandemic forced us as a country, as a community, as a family, and individuals to slow down for a minute. For some, that meant doing some deep cleaning in your home, maybe starting some home projects on the to-do list, reading a good book, maybe getting a new puppy like Dr. Calkins! For others, it meant more time for minds to wander, worrying about family and friends and even ourselves. Please take time to care for yourself during these unprecedented times and let us know if we can be of any assistance. Take some time to enjoy walks outside or practice some mindfulness meditation. Try to focus on the things that you can control. While we may not be in the same boat, we are in this together and can help one another through these challenging times. Be careful not to emotionally distance yourself too much from family and friends – remember you can stay connected while maintaining physical distance.

There have been many changes to our worklife here at Hopkins. We are primarily working remotely, but we are just as available to address your needs. Many clinic visits can be conducted via telemedicine when eligible.

Procedures are ramping back up. We are also available for in-person cardiac testing and in-person consultations. Research lab studies are getting back up and running as well. Some things look different around Hopkins, but we're taking all of the necessary precautions to keep our patients and staff safe and healthy. Please remember the importance of not putting off your routine follow up, so please reach out to us!



COVID-19 and ARVC

A new coronavirus causing COVID-19 was first identified in Wuhan China and has since spread to the United States. We are not aware of any severe complications among our ARVC patients that we follow. Please let us know if you test positive so we can learn more and provide better management recommendations. We believe that our patients who are most at risk of severe illness are those who have progressed to heart failure. We are happy to discuss your specific risks at an upcoming appointment. While researchers and clinicians are learning more about this virus each day, there are still many uncertainties. Here are some things we believed to be true based on current evidence:

- There are many types of coronavirus – some cause the common cold
- The type of coronavirus causing COVID-19 has never been seen in humans before
- Spreads through droplets from person to person when an infected person coughs, sneezes, or talks
- Symptoms typically appear 2-14 days after exposure
- Symptoms include fever, cough, shortness of breath, fatigue, muscle/body aches, headache, new loss of taste/smell, sore throat, congestion or runny nose, nausea or vomiting, diarrhea
- Some people with COVID-19 develop no symptoms or only minor symptoms, yet they can still spread the virus to others
- Most people who get COVID-19 will be able to recover from home
- Older adults and people with underlying medical conditions are at higher risk for severe illness
- Children are at lower risk for severe illness

What can you do to reduce your risk of getting COVID-19 as an ARVC patient?

- Take your medications as prescribed
- Eat a balanced diet
- Follow recommendations for exercise - Remember light to moderate exercise is acceptable
- Maintain physical distancing (6 feet)
- Wear a mask when in public or when you are around people that don't live with you
- Wash your hands often
- Monitor your health daily – watch for fever, cough, shortness of breath, etc
- Keep up with your regularly scheduled healthcare appointments

Reliable Sources of information

- Centers for Disease website: [cdc.gov](https://www.cdc.gov)
- Johns Hopkins Coronavirus (COVID-19) Information and Updates: hopkinsmedicine.org/coronavirus
- Johns Hopkins Coronavirus Resource Center: coronavirus.jhu.edu



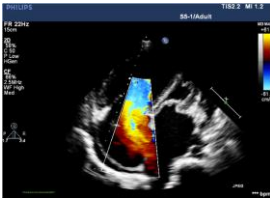
Clinical Services at Johns Hopkins

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 genetic testing, routine ICD management, as well as 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. She will help determine whether an in-person visit or telemedicine visit is most appropriate.

Please review the Hopkins visitor policy prior to your visit:

<https://www.hopkinsmedicine.org/coronavirus/visitor-guidelines.html#policy>



Telemedicine

Thanks to the COVID-19 pandemic, telemedicine at Johns Hopkins transformed overnight! Telemedicine has been a great option to be able to check in with our patients, but of course, cannot include complete cardiac testing. In many cases, we have been able to offer cardiac testing in-person on one day with a follow up telemedicine visit when results are available. While you may have had no problem having telemedicine visits with physicians within your own state, when you are seeing an out-of-state provider, it becomes a bit more complicated.

In order for us to provide care to patients who are in a state other than Maryland, we have to make sure we are licensed in the state where the patient is residing. Given the current emergency, many states have established waivers that essentially allow a physician in any other state, as long as they are licensed, to conduct telemedicine visits with patients in their state. Other states require a temporary license before you can provide telemedicine consultations. Still others have limits as to the type of patient (new or follow up) or no waiver at all. In addition, the waiver time frames are constantly changing, so we're doing our best to accommodate our patients in the best way possible. We are here to see you whether it is via telemedicine or in-person!



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 mexiletine 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 or 410-502-7161. This is an important first clinical trial in ARVC and will pave the way for future clinical drug trials.

COVID-19 update: This clinical trial is currently on hold until we obtain approval to resume. We are also looking into a remote consent option which would eliminate the need for an in-person visit. Stay tuned!

Research at Johns Hopkins

Relative Safety, Efficacy, and Patient Satisfaction of Standard ICDs versus the Sub-Cutaneous ICD (S-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).

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!

**Email Crystal to
discuss your
eligibility and
enrollment**

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.

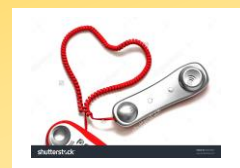
How to Join: Contact Crystal at 410.502.7161 or ctichnell@jhmi.edu.

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

Outcomes of genetic counseling for arrhythmogenic cardiomyopathy: A comparison of face-to-face and tele-genetic counseling

We are excited to be able to expand this study to include Cardiogenetics centers around the country. Given the current circumstances, we have decided to include tele-genetic counseling sessions as well. We are taking this opportunity to determine whether genetic counseling services offered via telephone/videoconference differs from the traditional face-to-face genetic counseling in achieving three key cardiac genetic counseling outcomes: reducing cardiac-specific anxiety, increasing disease-specific genetic understanding, and enhancing patient empowerment. Data will be collected by completing questionnaires 2 weeks preceding and 2 weeks following a genetic counseling session for an arrhythmogenic cardiomyopathy indication. Results of the study will provide some of the first evidence of genetic counseling outcomes in cardiology clinics, and also contribute outcome data of alternative methods to expand genetic counseling services. **THANK YOU TO EVERYONE WHO HAS PARTICIPATED IN THIS STUDY!!!**



Featured Manuscript

Exercise Restriction Is Protective for Genotype-Positive Family Members of Arrhythmogenic Right Ventricular Cardiomyopathy Patients (Europace 2020 Jun 23)

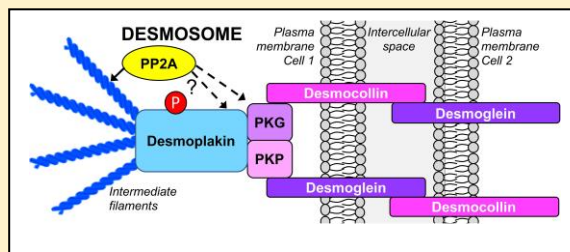
Weijia Wang, Crystal Tichnell, Brittney A Murray, Julia Agafonova, Julia Cadrin-Tourigny, Stephen Chelko, Harikrishna Tandri, Hugh Calkins, Cynthia A James.

Aims: In arrhythmogenic right ventricular cardiomyopathy (ARVC) patients, exercise worsens disease course, so limiting exercise is recommended. However, recommendations for gene-positive ARVC family members is incompletely resolved. We aimed to provide evidence for exercise recommendations for gene-positive ARVC family members.

In this study, gene positive family members of ARVC probands participated in an interview that asked them about their exercise history since the age of 10 years. Each specific type of exercise was carefully characterized by the duration and intensity. One hundred and one family members were included, of which 44 met diagnostic criteria for ARVC diagnosis and 16 experienced a sustained ventricular arrhythmia. Individuals who developed ARVC had higher average exercise duration and dose (duration*intensity). Only one individual who exercised below the American Heart Association recommended minimum (650 metabolic equivalent of task-hours/year) had ARVC or a sustained arrhythmia. In conclusion, ARVC family members who carry a pathogenic desmosomal variant should avoid endurance, high intensity type activities.

Next Research Initiative

Currently, we are actively compiling data on all of our registry-consented patients and family members who carry a pathogenic variant in the desmosomal gene, desmoplakin (*DSP*). We hope to better characterize our population of *DSP* carriers based on their arrhythmias and extent of structural involvement. In addition, we are collecting exercise interviews, as well as pregnancy history to assess whether exercise or pregnancy play a role in overall progression and/or prognosis. We will also be assessing how this group of patients fit into the current ARVC Diagnostic Task Force Criteria and whether modifications for this subgroup of ARVC patients may be necessary. If you are not currently enrolled in our registry, please reach out to Crystal (ctichnell@jhmi.edu) to become involved in this important effort.



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 or visit 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*



*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.*

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Supporting Our Program

Your Support Makes Breakthroughs Possible

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, 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, for more information.

Gifts by Mail:

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

**Please confirm the mailing
address before mailing
during the COVID-19
pandemic**

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

ARVD/C Program Info

ARVC Program Staff

Hugh Calkins, MD—Director

Harikrishna Tandri, MD—Faculty

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

Don't forget to keep us informed of your most up-to-date contact info!

Please send any changes and updated medical records to Crystal at ctichnell@jhmi.edu Thank you!

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www.ARVD.com

Contact
Us

Pre-COVID19 Gathering at our 20th Anniversary Seminar in 2019



Stay Safe and Healthy!