

JOHNS HOPKINS ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY PROGRAM

Volume 9, Issue 1

Winter 2019

★ ★★ ****!!!HAPPY NEW YEAR!!!**** ★★ ★

From all of us here at the ARVD/C Program, we hope you and your families had a wonderful Holiday Season. We wish you good health, happiness, and success in the coming year and always!

We also want to take this time to thank YOU! Please know that your continued support both financially as well as through your research participation does not go unnoticed. Thank you for volunteering your records, your time, and your ideas. Advances that have been made in the field of ARVD/C are all because of your dedication. We truly value this partnership and look forward to improving our understanding of ARVD/C together. We cannot thank you enough!

As we mentioned in our last newsletter, The Johns Hopkins ARVD/C Program has been recognized as a Precision Medicine Center of Excellence. We have continued to work on expanding access to multidisciplinary expert care, optimizing early detection through genomic, imaging, and epidemiological approaches, and developing patient-specific therapies for those at highest risk of sudden cardiac death and/or heart failure. This will be an ongoing partnership not only between our program and you, the patients and research participants, but also our international collaborators. This is a very exciting time for our program and for scientific discovery as it pertains to ARVC.

We are beyond excited to announce the 20th anniversary of our Program this year! Our Annual Patient and Family Seminar will be held the first weekend in May, and we hope to see many of you here as we celebrate this important milestone. Details are enclosed.

~ The ARVD/C Program

20th 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 some amazing speakers lined up and will be posting the agenda shortly as soon as it is finalized.



Adalena Tsatsopoulou
Naxos, Greece



Peter van Tintelen
Utrecht, The Netherlands



Mario Delmar
NYU Langone Health



Andre La Gerche
Melbourne, Australia



Frank Marcus
University of Arizona

You will never see this much history of ARVD/C all in one place again! It will be exciting to share the collaborative efforts all around the world in solving the mysteries of ARVD/C.

Make plans to come into Baltimore early to attend a reception at the Hilton Garden Inn Friday evening (7pm-9pm). **Please register early!** There is no registration fee for this seminar, but given our anniversary event we anticipate being at capacity. You must register and if you need to cancel, please let us know ASAP. All are welcome!

WHEN: Saturday, May 4th, 2019 8:00am-5:00pm

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

WHERE: Johns Hopkins Hospital - Chevy Chase Conference Center Auditorium
Main level of Sheikh Zayed Tower - 1800 Orleans Street
Baltimore, Maryland 21287

REGISTRATION: ALL participants must register! It is also helpful to list names of family members that will also be attending.
Register online by April 15th

<https://tinyurl.com/2019ARVCSeminar>

20th Annual ARVD/C Patient and Family Seminar

Presented by
The Johns Hopkins ARVD/C Program

HOTEL ACCOMODATIONS – 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 \$174/night plus tax until April 1st, 2019 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 2019 ARVD/C Seminar** group rate. Reservations can also be made through the online booking link:

https://homewoodsuites.hilton.com/en/hw/groups/personalized/B/BALHWHW-JHR-20190501/index.jhtml?WT.mc_id=POG

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 closet international airport to Johns Hopkins (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, parking is available at your own expense (max \$12) in the Orleans Street Garage. There is a bridge that connects the garage to the main level of Sheikh Zayed Tower.

SPECIAL EVENT

Join us for a Meet 'n Greet Reception, **7:00-9:00pm**, on **Friday, May 3rd, 2019** in the Great American Grill at the Hilton Garden Inn. 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, May 2nd and Friday, May 3rd 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 at 410-502-7161 or ctichnell@jhmi.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 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.

Tele – Genetic Counseling

We continue to take steps to expand our clinical services via telemedicine for those unable to travel to Baltimore. This service currently consists of the genetic counseling/genetic testing appointment only. We hope to be able to offer second opinion consultations with our physicians in the future. Telemedicine appointments are currently not billable to your insurance company and require payment out of pocket. This service may not be available in certain states. To discuss your eligibility or to schedule an appointment, contact Crystal.

Pediatrics

The world's only specialized Pediatric ARVD/C Clinic began in the Fall 2015 under the direction of pediatric electrophysiologist, Dr. Jane Crosson. It is with mixed emotions that we share news of Dr. Crosson's retirement. Dr. Crosson has been a great collaborator over the years working with our ARVC families and will be greatly missed. We are also excited for her to start this new chapter of her life in retirement.

We are thrilled to introduce Dr. Caridad de la Uz, pediatric electrophysiologist, who has a particular interest in inherited cardiomyopathies and will be taking over the ARVD/C Clinic. This is a monthly clinic staffed by a genetic counselor, Brittney Murray, along with Dr. de la Uz. We will offer second opinions/consults for both patients possibly affected and also cardiac testing associated with screening for family history of ARVD/C. Contact Crystal for information regarding upcoming dates of this specialty clinic.

Crystal Tichnell, MGC ctichnell@jhmi.edu 410-502-7161

Johns Hopkins at AHA 2018

The Johns Hopkins ARVD/C Program was well represented this year at the American Heart Association Scientific Sessions, held November 10-12 in Chicago. Our recently graduated research fellow Weijia Wang, MD presented his exercise interview findings and explained why

“Exercise Restriction is Protective in Genotype Positive Family Members of Arrhythmogenic Right Ventricular Cardiomyopathy Patients”.

Sithu Win, MD shared results of echocardiogram analysis which showed **“Right Ventricular Strain Predicts Structural Disease Progression in Patients with Arrhythmogenic Right Ventricular Cardiomyopathy”.** Our colleague from Geisinger, Chris Haggerty, PhD, showed that the risk of ARVC is low in people who have a desmosome mutation found through a large genetic testing research study of the general population, **“Prevalence and Electronic Health Record-Based Phenotype of Loss-Of-Function Genetic Variants in Arrhythmogenic Cardiomyopathy-Associated Genes in 61,000 Individuals.”**

Meanwhile Cindy James, ScM, PhD shared our recent data and approaches to **“Risk Stratification in ARVC: Substrates and Triggers.”** We look forward to sharing these results with you at the seminar, in clinic, and in future publications.



American
Heart
Association®

Research Opportunities at Johns Hopkins

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).

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.

Flecainide Trial

The Johns Hopkins ARVC program will be participating in a pilot study focusing on the antiarrhythmic effects of flecainide and its safety among ARVC patients. Medication research within the ARVC population has been difficult given that our patients tend to be on various "cocktails" to find that right combination to manage your arrhythmias. This will be an opportunity to solely look at one medication for treatment of arrhythmias and assess its efficacy and safety. This is an exciting opportunity and will pave the way for additional clinical trials. Stay tuned for more information regarding your eligibility to participate.

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

As we've mentioned before, we are now offering tele-genetic counseling! As part of this, we asked many of you to fill out surveys about your experience with genetic counseling to compare video tele-counseling to in person, face-to-face genetic counseling. Our results will provide some of the first evidence of genetic counseling outcomes in cardiology clinics. We were happy to see that genetic counseling makes a difference! Patients receiving genetic counseling had decreased fear after their GC visit, and also felt more empowered. The empowerment variable was a complex construct that included facets of feeling in control of your diagnosis, knowing next steps, feeling hopeful for the future, and understanding the diagnosis and risks. This data supports a nationwide push for genetic counselors to be involved in the multidisciplinary care of patients with ARVC and other inherited heart disease, and it supports our clinic policy that a genetic counselor is involved in the care of every patient that visits our center.



2018 Publications

- Bosman LP, Sammani A, James CA, Cadrin-Tourigny J, Calkins H, van Tintelen JP, Hauer RNW, Asselbergs FW, Te Riele ASJM. **Predicting arrhythmic risk in arrhythmogenic right ventricular cardiomyopathy: A systematic review and meta-analysis.** Heart Rhythm. 2018 Jul;15(7):1097-1107. doi: 10.1016/j.hrthm.2018.01.031. Epub 2018 Feb 3.
- Orgeron GM, Te Riele A, Tichnell C, Wang W, Murray B, Bhonsale A, Judge DP, Kamel IR, Zimmerman SL, Tandri H, Calkins H, James CA. **Performance of the 2015 International Task Force Consensus Statement Risk Stratification Algorithm for Implantable Cardioverter-Defibrillator Placement in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy.** Circ Arrhythm Electrophysiol. 2018 Feb;11(2):e005593. doi: 10.1161/CIRCEP.117.005593.
- Wang W, James CA, Calkins H. **Diagnostic and therapeutic strategies for arrhythmogenic right ventricular dysplasia/cardiomyopathy patient.** Europace. 2018 Apr 23. doi: 10.1093/europace/euy063.
- Murray B, Hoorntje ET, Te Riele ASJM, Tichnell C, van der Heijden JF, Tandri H, van den Berg MP, Jongbloed JDH, Wilde AAM, Hauer RNW, Calkins H, Judge DP, James CA, van Tintelen JP, Dooijes D. **Identification of sarcomeric variants in probands with a clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC).** J Cardiovasc Electrophysiol. 2018 Jul;29(7):1004-1009. doi: 10.1111/jce.13621. Epub 2018 May 21.
- Wang W, Cadrin-Tourigny J, Bhonsale A, Tichnell C, Murray B, Monfredi O, Chrispin J, Crosson J, Tandri H, James CA, Calkins H. **Arrhythmic outcome of arrhythmogenic right ventricular cardiomyopathy patients without implantable defibrillators.** J Cardiovasc Electrophysiol. 2018 Oct;29(10):1396-1402. doi: 10.1111/jce.13668. Epub 2018 Jun 27.
- Haggerty CM, Murray B, Tichnell C, Judge DP, Tandri H, Schwartz M, Sturm AC, Matsumura ME, Murray MF, Calkins H, Fornwalt BK, James CA. **Managing Secondary Genomic Findings Associated With Arrhythmogenic Right Ventricular Cardiomyopathy: Case Studies and Proposal for Clinical Surveillance.** Circ Genom Precis Med. 2018 Jul;11(7):e002237. doi: 10.1161/CIRCGEN.118.002237.
- Hoorntje ET, Posafalvi A, Syrris P, van der Velde KJ, Bolling MC, Protonotarios A, Boven LG, Amat-Codina N, Groeneweg JA, Wilde AA, Sobreira N, Calkins H, Hauer RNW, Jonkman MF, McKenna WJ, Elliott PM, Sinke RJ, van den Berg MP, Chelko SP, James CA, van Tintelen JP, Judge DP, Jongbloed JDH. **No major role for rare plectin variants in arrhythmogenic right ventricular cardiomyopathy.** PLoS One. 2018 Aug 30;13(8):e0203078. doi: 10.1371/journal.pone.0203078. eCollection 2018.
- Zghaib T, Bourfiss M, van der Heijden JF, Loh P, Hauer RN, Tandri H, Calkins H, Nazarian S, Te Riele ASJM, Zimmerman SL, Velthuis BK. **Atrial Dysfunction in Arrhythmogenic Right Ventricular Cardiomyopathy.** Circ Cardiovasc Imaging. 2018 Sep;11(9):e007344. doi: 10.1161/CIRCIMAGING.117.007344.
- Zghaib T, Ghasabeh MA, Assis FR, Chrispin J, Keramati A, Misra S, Berger R, Calkins H, Kamel I, Nazarian S, Zimmerman S, Tandri H. **Regional Strain by Cardiac Magnetic Resonance Imaging Improves Detection of Right Ventricular Scar Compared With Late Gadolinium Enhancement on a Multimodality Scar Evaluation in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy.** Circ Cardiovasc Imaging. 2018 Sep;11(9):e007546. doi: 10.1161/CIRCIMAGING.118.007546.
- Orgeron G, Bhonsale A, Migliore F, James CA, Tichnell C, Murray B, Bertaglia E, Cadrin-Tourigny J, De Franceschi P, Crosson J, Tandri H, Corrado D, Calkins H. **Subcutaneous Implantable Cardioverter-Defibrillator in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia: A Transatlantic Experience.** J Am Heart Assoc. 2018;7:e008782. DOI: 10.1161/JAHA.118.008782.
- James CA, Calkins H. **Arrhythmogenic Right Ventricular Cardiomyopathy: Progress Toward Personalized Management.** Annu Rev Med. 2018 Oct 24. doi: 10.1146/annurev-med-041217-010932. [Epub ahead of print]
- Brosnan MJ, Te Riele ASJM, Bosman LP, Hoorntje ET, van den Berg MP, Hauer RNW, Flannery MD, Kalman JM, Prior DL, Tichnell C, Tandri H, Murray B, Calkins H, La Gerche A, James CA. **Electrocardiographic Features Differentiating Arrhythmogenic Right Ventricular Cardiomyopathy from an Athlete's Heart.** JACC Clin Electrophysiol. 2018 Dec;4(12):1613-1625. doi: 10.1016/j.jacep.2018.09.008. Epub 2018 Nov 1.
- Aliyari Ghasabeh M, Te Riele ASJM, James CA, Chen HSV, Tichnell C, Murray B, Eng J, Kral BG, Tandri H, Calkins H, Kamel IR, Zimmerman SL. **From an Athlete's Heart.** JACC Clin Electrophysiol. 2018 Dec;4(12):1613-1625. doi: 10.1016/j.jacep.2018.09.008. Epub 2018 Nov 1. **Epicardial Fat Distribution Assessed with Cardiac CT in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy.** Radiology. 2018 Dec;289(3):641-648. doi: 10.1148/radiol.2018180224. Epub 2018 Aug 21.

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, 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

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

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

Anna Burton—Genetic Counselor Assistant

Giovanna Jimenez—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

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Baltimore, Maryland 21287
P: 410-502-7161, F: 443-873-5073
Website: www.ARVD.com



**M
a
x**

In the Fall of 2018 we welcomed baby Max Murray to the ARVD/C family. Both he and mom are doing well and we are happy to have Brittney back in the office!

Congratulations to the Murray Family!

Support Group Info

Looking for a support group?

FACEBOOK Groups (private):

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

Precision Medicine WebPage:

Read "Feeling the Beat": Stories from the ARVC Community here:

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

ARVD/C Mentor Program:

Get matched with an ARVD/C mentor!

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

Contact Nancy Bogle at nbstjohn@gmail.com for more information and visit ARVDHEARTANDSOUL.org

