Invitation and Introduction to the Arrhythmogenic Right Ventricular Dysplasia (ARVD) Children's Project — A Letter of Invitation from the Principal Investigator

Dear ARVD Patients, Families and Friends:

My name is Li Zhang. I am a research cardiologist at the Main Line Health Heart Center in Wynnewood, Pennsylvania.

I would like to provide you some background on my medical training and research experience as well as the ARVD Children's project. I hope that you and your children will be interested in working together with me on this important medical research project on ARVD.

Background

After practicing medicine for 7 years in a major Chinese medical school/teaching hospital where I trained as a cardiologist, I followed my husband to the United States when he began his PhD studies in the late 1980s. Instead of practicing medicine, I have applied my medical knowledge and experience in research and authored/coauthored more than 100 research publications, mostly in top ranked cardiology journals, over the last 15 years. I also served as the director for multi-national/multi-center ECG studies in sudden arrhythmic death disorders at LDS Hospital of Intermountain Healthcare.

My primary interest is to identify electrocardiographic (ECG or EKG) patterns characteristic of heritable heart rhythm disorders, then use this information to guide genetic and clinical testing for these disorders. Our research outcomes have begun to improve the diagnostic accuracy for these disorders, which may lead to improved treatment and prevention of unexpected, sudden arrhythmic death.

The research project

Arrhythmogenic right ventricular dysplasia (ARVD) is largely an inherited disease of the heart muscle often associated with abnormal heart rhythms. Disease development is concealed in the early stages, and sudden death can be the first symptom. Currently, early diagnosis is difficult, costly, and inaccurate. In the last several years, I have devoted my expertise and efforts in ARVD ECG studies. After two years of preliminary investigations guided by Dr. Guy H. Fontaine, a world pioneer ARVD investigator, I have identified several ARVD-related ECG patterns that are likely predictive of the disease. Recently I received the Scientist Development Grant, awarded by the American Heart Association (AHA) for this project, entitled "Sudden Death Prevention: Identifying the ECG Markers in the Early Stage of Arrhythmogenic Right Ventricular Dysplasia", to further this work.

The research team

As the Principal Investigator, I will be working with a group of distinguished medical experts to investigate the ECG changes in ARVD adults and their children.

Collaborating Investigators
Guy H. Fontaine – Research Director and world-renowned expert who first described ARVD
Jeffrey A. Towbin – Pediatric Cardiology Professor and one of the leading U.S. experts in genetic studies of ARVD
Hugh Calkins – Pediatric and Adult Cardiology Professor and one of the leading U.S. experts in genetic studies of ARVD
Dayi Hu - Professor and Director of Chinese National ARVD Registry
Jielin Pu – Professor and Research Director of Center for Arrhythmias, Beijing Fuwai Hospital, China

Consultants
Frank Marcus - Professor Emeritus, and Principal Investigator, the NIH Study of ARVD
Robert L. Lux – Professor Emeritus, Cardiovascular Research and Training Institute, University of Utah.

If you are eligible to participate in this study this is what you will need to do:

All adults and children who are diagnosed with ARVD, and children with a biologic parent diagnosed with ARVD are eligible for this study.

If you are eligible and you give written consent for participation in this clinical investigation, you will be invited to **send us copies of your previously completed ECG studies and/or your child's previous ECG studies.** After we receive the ECGs, your involvement is minimal, as the investigators will mainly examine the serial ECGs.

In addition to the previous ECGs, we will pay special attention to the ECG changes called T wave evolution in enrolled children. T wave evolution in children diagnosed with ARVD and children from ARVD families will be studied prospectively for up to 4 years. **Thus we need parents to send us yearly ECGs of their children after the enrollment.**

Research funds have been set aside to reimburse the cost of obtaining this yearly ECG for enrolled children.

In addition, a small group of eligible study subjects will be invited to Main Line Health Heart Center for a 2 day ECG study. If you or your child is eligible for this more extensive evaluation, some limited compensation is available for study related expenses.

Every ECG is important, and your contribution(s) will help researchers learn to recognize the subtle, progressive ARVD ECG changes in children who may have this condition.

Detecting ARVD-related ECG abnormalities by cost effective screening methods and improving the early diagnosis for the prevention of sudden unexpected death in young people with ARVD are the goals of this project. There is no guarantee, however, of personal benefit for participating in this investigation. Long-term benefits may be realized by enhanced ability

to identify ARVD-related ECG changes that may lead to earlier diagnosis of ARVD once the study results are analyzed and validated in the future.

We cannot provide personal care, diagnosis or treatment. We hope to help our study participants, but all their primary medical care must come from their personal physicians.

Your enrollment in this study is voluntary. Every study subject has the right to discontinue or withdraw anytime without penalty.

If you are interested in participating in this ARVD project, please contact me at (484) 222-1876 or Fax (610) 896-0643. Alternatively, you can contact me by email at ldlzhang@gmail.com

Thank you,

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