

Thursday 02 April 2020

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GUARD-Heart



Gateway to Uncommon And Rare Diseases of the Heart

ERN GUARD-HEART BIMONTHLY NEWSLETTER

YEAR 2020 NUMBER 2

ERN GUARD-Heart in the Time of COVID-19

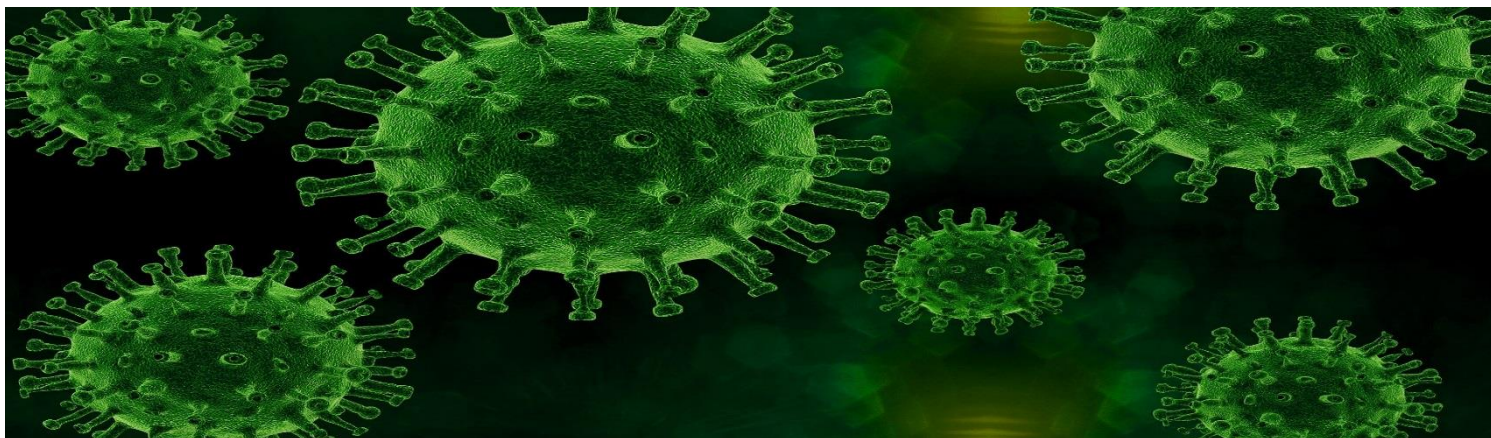
Amsterdam, 2 April 2020

Dear friends,

Although we are the first to realize that our ERN GUARD-Heart is currently not at the top of the priority list, we thought it of importance to keep you informed about the current developments. And that does include some issues related to the SARS-CoV-2 pandemic, which currently hits our respective countries, in particular Italy and Spain so terrible. We truly hope all of you as well as your close family are safe and well and we wish you all strength and good health in these difficult weeks.

The Corona-related issues include a recently released manuscript on "SARS-CoV-2/COVID-19 interaction with inherited arrhythmia syndromes" that was prepared by several GUARD-Heart members in a very short time. This manuscript is published by Heart Rhythm (see page 2); the European Heart Journal was first attempted but their response time was very slow. A second issue is the first patient-information session (through Zoom) in the Netherlands on this issue. We all encourage you to organize such sessions on short notice, patients do need guidance. Keep strong and stay healthy!

All the best from the management team, Arthur, Amin & Nynke. ❤





ERN activities



SARS-CoV-2, COVID-19 and inherited arrhythmia syndromes

Amsterdam, 1 april 2020

Last week the first ERN GUARD-Heart COVID-19 article was published online in Heart Rhythm.

Abstract: Ever since the first case was reported at the end of 2019, the SARS-COV-2 virus and associated lung disease COVID-19 has spread throughout the world and has become a pandemic. In particular, the high transmission rate of the virus has made it a threat to public health globally. Currently, there is no proven effective therapy against the virus, and the impact on other diseases is also uncertain, especially inherited arrhythmia syndrome.

Arrhythmogenic effect of COVID-19 can be expected, potentially contributing to disease outcome. This may be of importance for patients with an increased risk for cardiac arrhythmias, either secondary to acquired conditions or co-morbidities or consequent to inherited syndromes. Management of patients with inherited arrhythmia syndromes such as Long QT syndrome, Brugada syndrome, Short QT syndrome and Catecholaminergic Polymorphic Ventricular Tachycardia in the setting of the COVID-19 pandemic may prove particularly challenging. Depending on the inherited defect involved, these patients may be susceptible to pro-arrhythmic effects of COVID-19-related issues such as fever, stress, electrolyte disturbances and use of antiviral drugs. We here describe the potential COVID-19 associated risks and therapeutic considerations for patients with distinct inherited arrhythmia syndromes and provide recommendations, pending local possibilities, for their monitoring and management during this pandemic. ♥

Cheng-I Wu, Pieter G. Postema, Elena Arbelo, Elijah R. Behr, Connie R. Bezzina, Carlo Napolitano, Tomas Robyns, Vincent Probst, Eric Schulze-Bahr, Carol Ann Remme, Arthur A.M. Wilde
<https://doi.org/10.1016/j.hrthm.2020.03.024>

Registry grant and ERN GUARD-Heart registries

Rome, 21 February 2020

Recently, the ERN GUARD-Heart has received a 200K grant for registries. Besides the continuation and expansion of the disease-specific registries, the general objective of this grant/project, in line with the criteria of the call, is to build an overarching registry for the ERN GUARD-Heart (the Heart-Core Registry), following the standards and tools provided by the European Platform on Rare Disease Registration (EU RD Platform). In the Heart-Core Registry, a minimal core dataset will be collected prospectively of the new patients in the disease- or gene-specific registries of ERN GUARD-Heart. The minimal core dataset includes the Set of Common Data Elements for Rare Diseases Registration provided by the European Commission's Joint Research Centre (JRC). Each participating HCP will have only access to its own patient. Patients should have provided consent with an ERN consent form. Updates on this topic will be provided in the upcoming newsletters.

Furthermore there are direct plans for building two extra disease specific registries in REDCAP:

- An international registry of patients with a CACNA1C mutation (F. Drago & A. Baban, Rome)
- An international registry of patients with a FHL1 mutation (R. Jurcut, Bucharest)

In the meantime, ERN GUARD-Heart members are invited and requested to include their patients in the existing registries. ♥



7th Board Meeting of ERN GUARD-Heart

Rome, Friday 21 February 2020

The 7th board meeting of ERN GUARD-Heart was held in Rome, Italy, on 21 February 2020. Rome was sunny, and beautiful and it's hard to believe how fast Italy and thereafter the rest of Europe was overwhelmed by the Corona-virus. On the evening before there was a delicious dinner organized by Fabrizio Drago in Restaurant Taberna de'Gracchi. The meeting was attended by 27 participants out of 20/24 HCP's of the Network. The notes of the meeting were disseminated last month by the project-management.





ERN activities

National online patient-meeting with COVID-19 related questions

Amsterdam, 1 April 2020

In the Netherlands a first online patient-meeting for COVID-19 related questions has been organized for the 2nd of April. LQTS- and CPVT-patients are invited to join an online meeting where they can ask live-questions to an adult and a pediatric cardiologist. This can be followed by similar meetings for patients with other diseases. Our Dutch E-PAGs were involved in the plans and communication. Please share your national activities on this topic with the project-management for publication in the next newsletter.♥

Assessment ERN applications

Amsterdam, 1 April 2020

The call for HCPs to join existing ERNs was closed on 30 November 2019. There are 32 applications for the ERN GUARD-Heart. After the eligibility check by the European Commission, the proposed delegation of the ERN GUARD-Heart Board (Arthur Wilde, Carlo Napolitano, Georgia Sarquella-Brugada, Philippe Charron, Eric Schülze-Bahr and the project managers) started online assessment sessions on the applications to provide an official opinion. The complete procedure will take about (at least) one year. Due to the current situation with the COVID-19 virus, the deadlines will probably be extended. ♥

Latest ERN GUARD-Heart Publications

1. Lieve KVV, Dusi V, van der Werf C, Bos JM, Lane CM, Stokke MK, Roston TM, Djupsjöbacka A, Wada Y, Denjoy I, Bundgaard H, Noguer FRI, Semsarian C, Robyns T, Hofman N, Tanck MW, van den Berg MP, Kammeraad JAE, Krahn AD, Clur SB, Sacher F, Till J, Skinner JR, Tfelt-Hansen J, Probst V, Leenhardt A, Horie M, Swan H, Roberts JD, Sanatani S, Haugaa KH, Schwartz PJ, Ackerman MJ, Wilde AAM. Heart Rate Recovery After Exercise Is Associated With Arrhythmic Events in Patients With Catecholaminergic Polymorphic Ventricular Tachycardia. *Circ Arrhythm Electrophysiol.* 2020 Mar;13(3):e007471. doi: 10.1161/CIRCEP.119.007471. Epub 2020 Feb 16. PMID: 32063070
2. Postema PG, Schwartz PJ, Arbelo E, Bannenberg WJ, Behr ER, Belhassen B, Brugada J, Brugada P, John Camm A, Casado-Arroyo R, 't Hoen E, Hollak CEM, Käb S, Lambiase PD, Leenhardt A, Priori SG, Probst V, Stunnenberg BC, Tfelt-Hansen J, Van Engelen BGM, Veltmann C, Viskin S, Wilde AAM. Continued misuse of orphan drug legislation: a life-threatening risk for mexiletine. *Eur Heart J.* 2020 Feb 1;41(5):614-617. doi: 10.1093/eurheartj/ehaa041. No abstract available. PMID: 32006435

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