

Wednesday 3 April 2019

ERN

GUARD-Heart

Gateway to Uncommon And Rare Diseases of the Heart



ERN GUARD-HEART BIMONTHLY NEWSLETTER

YEAR 2019 NUMBER 2

5th Board Meeting of the ERN GUARD-Heart

Paris, 13 February 2019

The 5th ERN GUARD-Heart Board meeting was held on 13 February 2019 at Hôpital Universitaire Pitié-Salpêtrière. The topics discussed at the meeting included a summary of the ERN activities in 2018, update on the regulations and plans for extension of existing ERNs, translation of patient information leaflets in official European languages and creation of new disease-specific leaflets, the organization of national topic meetings for patients, educational work plans of ERN, design of cross-border patient pathways, update on the European patient advocacy

groups (ePAGs) currently active in the ERN, the ERN continuous monitoring system and the 18 common ERN indicators the monitoring system is based on, update on the CPMS activities of each ERN member, update on ERN registries, evaluation of benchmark studies and plans for new studies, and the general plans for the third ERN year (from 1 March 2019 till 29 February 2020). Notes of the meetings have been produced by the project management office and disseminated among the representative members of the 24 member centres. ♥



Representative members of the ERN GUARD-Heart at the 5th Board Meeting in Paris (France) on 13 February 2019.



ERN Meetings

7th Meeting of ERN Coordinators Group

Brussels, 25 March 2019

The 7th meeting of the ERN Coordinators Groups and the European Commission took place on 25th March in Brussels. The activities of the 24 ERNs in the CPMS, IT platform, and other communication channels (website, Newsletter) and the activities of different Coordinators Working Groups were presented and discussed.

In addition, an update on the current status of the enlargement of the ERNs was provided by the European Commission. The call for inclusion of new healthcare providers (HCPs) is expected to open in the 2nd half of 2019. In the meanwhile, the EU has offered ERNs to extend by including associated national centres. Associated national centres are not official ERN members, but form links to ERNs for countries who do not have a ERN member yet. They act as entry points to ERNs for patients, shall participate in the CPMS for patients that they have referred to the ERN, and may at later stage become full members. However, they will lose their partnership if a HCP from the same Member State joins the ERN as a full member. ♥

ERN Hospital Managers' Meeting

Paris, 16 May 2019

A meeting organized by the European Commission for Hospital Managers will take place on 16 & 17 May 2019 in Hospital Saint-Louis, Paris, France. The Commission believes that the role of Hospital Managers is an essential key for the development of the ERN system in Europe. This ERN Hospital Managers Meeting is expected to help preparing further actions:

- To better understand the benefits and impact of the ERNs and its Members in the Hospitals.
- To align goals and activities in a win-win patient centred strategy of ERNs, Member States and Hospital managers.
- To discuss future actions, links and next steps and the feasibility of creating a structured ERN Hospital Managers Group.

Earlier meetings for the Hospital Managers took place in Rotterdam and Brussels & at the 4th ERN Conference. ♥



Rare Disease Models and Mechanisms-Europe registry [open for registration](#)

"Solve-RD - Solving the unsolved Rare Diseases" is a large European research project funded by the EU for five years (2018-2022) under Horizon 2020. The consortium is collaborating with ERNs to improve the diagnosis of rare diseases. Solve-RD is about to establish a European Rare Disease Models & Mechanisms Network (RDMM-Europe) using the successful Canadian blueprint as a role model. The goal of RDMM-Europe is to link clinicians discovering new genes in patients with rare diseases with basic scientists, who can functionally analyse equivalent genes and pathways in model organisms. Solve-RD will provide 50 Seeding Grants (20.000 EUR each) to fund projects that will allow rapid confirmation of potentially disease-causing genes and elucidation of the underlying molecular disease mechanisms. For more information please check the [Solve-RD website](#) or contact Sanja Hermanns (sanja.hermanns@med.uni-tuebingen.de).



ERN News

ERN Monitoring System: first data submitted

Amsterdam, 1 April 2019

The 1st phase of the ERN Monitoring System (with 18 ERN common quality indicators) to assess the performance of ERNs has been initiated. The 18 indicators have been circulated among the ERN GUARD-Heart members & will be discussed in the next Board meeting. DG SANTE Data Collection Platform to collect the information and data related to the 18 indicators is available online.

In March 2019, the project management office collected all the data needed. This included the total number of new patients referred to HCPs with diseases/conditions that fall within the scope of the ERN (N = 14.777 for 2017 and 15.927 for 2018 for all 24 HCPs in the ERN GUARD-Heart). The data were entered in the online platform and submitted to the European Commission March 30, 2019.

From April 2019, ERNs will start to collect the information corresponding to the first quartile of 2019. From then on, data should be collected at least every three months although it would be possible to upload the information monthly. ♥

UK withdrawal from the EU: preparation & contingency measures

Amsterdam, 13 March 2019

The UK will withdraw from the EU & be no longer a Member State as from the intended withdrawal date. Presently, there is no certainty on whether a Withdrawal Agreement between the EU & the UK will be ratified by that date. The Commission has highlighted the need for ERNs to prepare for this event. The Commission has also focussed on the contingency measures needed in a no-deal scenario. One aspect that needs to be implemented is the *"disconnection and adaptation of databases & IT systems & other platforms for communication & information exchange to which the UK should no longer have access"*. The ERNs work with such platforms for communication and information exchange (e.g., IT Platform, CIRCABC, and CPMS). The Commission has requested ERN Coordinators to assess what accesses need to be removed in ERNs in each of the 2 scenarios, with or without a ratified Withdrawal Agreement. In general, access of any expert with UK nationality to IT platforms should be removed as from the withdrawal date if the Withdrawal Agreement is not ratified. ♥



EUROPEAN JOINT PROGRAMME RARE DISEASES

The **European Joint Programme on Rare Diseases (EJP RD)** brings over 130 institutions from 35 countries to create a comprehensive, sustainable ecosystem allowing a virtuous circle between research, care and medical innovation.

EJP RD will foster joint transnational calls for collaborative research projects resulting in funding encompassing various research fields of rare diseases in continuation of previous **E-Rare** calls.

The EJP RD funding organizations intend to open the 1st EJP RD Joint Transnational Call (JTC 2019) for funding multilateral research projects on rare diseases together with the European Commission (EC) under the **EJP-COFUND** mechanism.

For more information visit: <http://www.ejprarediseases.org/>



Sports & Disease



Amsterdam, 1 April 2019

Currently, evidence-based guidelines for patients with long QT syndrome (LQTS) who engage in (competitive) sports are lacking. ERN GUARD-Heart has established an advisory panel of experts who can provide advice on patients with LQTS who participate in (competitive) sports and who are referred to the ERN for consultation through CPMS. ERN experts who have agreed to participate in such a advisory panel include Elijah Behr, Lia Crotti, Antoine Leenhardt, Anita Hiippala, Pier Lambiase, Jacob Tfelt-Hansen, and Arthur Wilde. Consultations with regard to sports activity in LQTS cases can be entered into CPMS and experts (*as listed above*) can be invited as panel members. The cases will be collected in a database for future research. The enrolment of patients will start on 1 May 2019.

In addition, a 2nd panel of experts will be available for consultations with regard to sports activity in pre-symptomatic patients with a genetic defect associated with arrhythmogenic right ventricular cardiomyopathy (ARVC), i.e. those without manifestation of the disease'. These experts include: Barbara Bauce, Juan Kaski, Pablo Garcia-Pavia, Françoise Hidden-Lucet, and Elijah Behr.

When cases are entered in CPMS for consultation with regard to their participation sports in (competitive) sports, the referring physician should at least upload a 12-lead ECG for a LQTS case and imaging data (echocardiography and/or MRI) for a pre-symptomatic patient with ARVC. In addition, the following questions should be addressed:

- Which sports does the athlete primarily perform?
- Does the athlete perform any secondary sports? (>3 hours per week) If yes, which?
- How many hours per week does the athlete train and compete?
- How many years has the athlete been training at a comparable level?
- The athlete trains at the following level (multiple choice):
 - Recreational
 - Recreational with competitions
 - Competitive national
 - Competitive international
- What is the maximal heart rate of patient during a maximal exercise test?
- What is the maximal capacity of patients during a maximal exercise test? (*answer numbers and unit, i.e. Watts, km/h, VO₂max, and METS*)

Further details will follow in due course. For any question regarding this project you can contact ERN GUARD-heart management office. ♥





Cardiac Patient Trial & Registry Tool (Cardiac Patrol)

*now available in
app stores*

*sponsored by
ERN GUARD-Heart*

Cardiac Patrol

Cardiac Patient Trial and Registry Tool. The app contains relevant scientific studies and database information (registries) in the field of rare and congenital heart diseases.

1 LOGIN

Create an account or login with your BIG-number

2 LIST OF CONDITIONS

Click the disease your patient has been diagnosed with and enter their age

3 STUDY INFORMATION

Select 'line of treatment' and see if there is an 'open trial'

4 LINE OF TREATMENT

Review the study scheme or enter the in- and exclusion criteria and see which hospitals participate

5 REFERRALS

Refer the patient to the researcher(s)



TO ADD A REGISTER OR STUDY TO THE APP, THE RESEARCHER PROVIDES THE FOLLOWING COMPONENTS

- Disease
- Study name
- Study status
- Study start date
- Study completion date
- Principal Investigator name
- PI email address
- PI hospital + city
- Eligible ages
- Line of treatment
- Picture of study arms
- Participating sites + local PI
- Inclusion criteria
- Exclusion criteria



Hart4Onderzoek is the first platform that brings together scientific research in the field of congenital heart and vascular diseases, both nationally and within the EU.
www.hart4onderzoek.nl

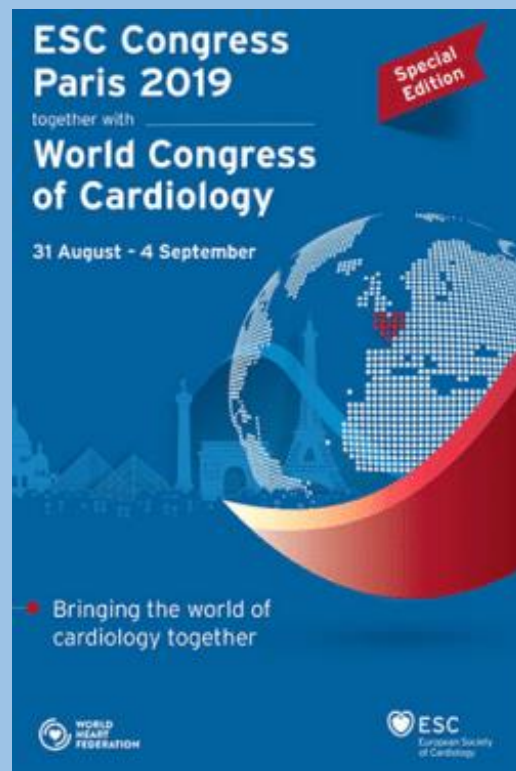


Upcoming Events

SAVE THE DATE

6th ERN GUARD-Heart Board Meeting

3 September 2019



UPCOMING CARDIOLOGY EVENTS

EuroPrevent	11 April – 13 April 2019	Lisbon - Portugal
EuroCMR	02 May – 04 May 2019	Venice - Italy
Heart Rhythm Society Sessions	8 May – 11 May 2019	San Francisco – California
27th Nordic-Baltic Congress of Cardiology	10 June – 12 June 2019	Helsinki - Finland
Basic Science Summer School	16 June – 20 June 2019	Sophia-Antipolis - France
European Society of Cardiology	31 August - 4 September 2019	Paris – France
Update in Clinical Cardiology	16 October – 18 October	Boston - Massachusetts



27th NORDIC-BALTIC CONGRESS OF CARDIOLOGY

June 10-12, 2019
Finlandia Hall, Helsinki/Finland

www.nbcc2019.org / [@nbcc2019](https://twitter.com/nbcc2019)



Follow ERN GUARD-Heart on:
(click on logo to proceed)



ERN GUARD-Heart
Project
Management
Office



Arthur A.M. Wilde
Network Coordinator
Academic Medical Center
Amsterdam, Netherlands
Email: a.a.wilde@amc.nl
Phone: +31205662904



Nynke Hofman
Project Manager
Academic Medical Center
Amsterdam, Netherlands
Email: n.hofman@amc.nl
Mobile: +31622046435



Ahmad S. Amin
Project Manager
Academic Medical Center
Amsterdam, Netherlands
Email: a.s.amin@amc.nl
Mobile: +31614471713