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GUARD-Heart

Gateway to Uncommon And Rare Diseases of the Heart



ERN GUARD-HEART BIMONTHLY NEWSLETTER

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Update Exchange Programme

Amsterdam, 26 November 2021

The response on the survey on the ERN Exchange Programme survey revealed a nice indication of the interest of the members and affiliated partners in the Programme. The goal of the Exchange Programme is to stimulate collaboration between healthcare professionals in ERNs and to promote knowledge sharing. All members of multidisciplinary teams can make use of the programme. At this moment, plans for 8 of the available packages (pre-) have been approved, and the first visit took place recently.

Due to the 4th wave of the Covid-19 pandemic and the steep rise in the infection rates, an increasing number of exchange visits have had to be cancelled or postponed. Therefore, in close consultation with the European Commission (HaDEA and DG SANTE), the implementation of the Exchange Programme has been suspended until the situation improves. Hopefully the activities can be resumed from March 2022 onwards – this will be reviewed and confirmed with the European Commission based in February.

Any visits already planned for the winter period (1 Dec 2021 – 28 Feb 2022) will have to be postponed until the reactivation of the programme.

It is still possible to send us requests for an exchange in spring- and summertime of 2022. If there are any questions, please contact the management office (n.hofman@amsterdamumc.nl). ♥

Internal ERN-coordinators-meeting Strasbourg

Strasbourg, 21 October 2021

On 21 October, a delegation of 6 coordinators went to the European Parliament in Strasbourg to meet with Members of European Parliament (MEPs) to discuss crucial issues with regard to the future of European Reference Networks.

On 22 October, all 24 ERN coordinators and project managers gathered together to discuss the future of the ERNs, in particular the funding of ERNs, CPMS, ERN registries, monitoring and guidelines. Regarding the funding for the future and the introduction of the new full members to the ERNs, there are a lot of uncertainties at this moment. Further information will be disseminated as soon as available. ♥

ERICA Project

Amsterdam, 25 November 2021

The European Rare disease research Coordination and support Action (ERICA) project has started its activities in the last year. ERICA project gives ERNs a platform for a systematic approach to rare disease research. Work packages deal with data integration and data sharing (for example a standard data sharing agreement for ERN patients registries), bio-banking, patient-centred research, clinical trial support, translation and innovation and integration and dissemination. There will soon be an interactive web-based platform for ERN-based research. This could be a central driver for ERN research in the near future. More information is available on [the website](#). ♥





Current status of overall patient registries in the 24 ERNs

Amsterdam, 25 November 2021

Patient registries in the 24 ERNs differ by design, use different types of platforms, and differ in the number of data fields. In terms of patients enrolled, 16 have less than 100, four have up to 1000, while four have more than 1000 patients. ERKNet-registry (the overarching patient registry of the ERN for rare kidney diseases) is one of the largest and most successful ERN registries, started in 2019, and has passed the 10.000 enrolled patients' landmark, with 47 sites across Europe.

The Heart-Core registry, the overarching patient registry in the ERN GUARD-Heart, started in 2020. The goal of this registry (in REDCap) is to monitor the number of patients in individual patient registries and, ideally, to monitor the number of patients with rare cardiac diseases per healthcare provider (HCP) in the ERN. The registry data will not be used for scientific research, but only for epidemiological and monitoring purposes within the ERN GUARD-Heart. Therefore, only very limited data will be entered, i.e., common data elements provided by the European Commission's Joint Research Centre (see: [Set of common data elements](#)). Based on technical and legal issues, and after discussions in the ERN Board, it was decided to enter patient data in the Heart-Core registry in 2 different ways: completely anonymized data from existing patient registries in the ERN (in that way no consent is required as patients cannot be identified) or pseudo-anonymized data from patients that have been discussed in the CPMS (and where consent for data collection in the ERN registry has been given via the standard ERN consent form). So far, Amsterdam UMC, the coordinating centre of the ERN GUARD-Heart, has entered completely anonymized common data elements for LQTS and CPVT patients in the Heart-Core registry. Other registry leaders and HCPs in the ERN GUARD-Heart are cordially invited to participate in the Heart-Core registry. ♥



Activities of EJP-RD

Amsterdam, 25 November 2021

Besides the ERN patient registries, there is work ongoing with the European Joint Programme on Rare Diseases (EJPRD). This multinational, multi-site project supported from Horizon Europe is devoted to providing infrastructure tools for rare disease research. A virtual platform connecting all kinds of catalogues, databases and tools is being developed. It should be possible to query other knowledge bases to get information on where patients data or samples are, and ERN registries can play a central role. For this to happen registries must be made interoperable. FAIRification Stewards (Findable, Accessible, Interoperable, Reusable) are there to help make databases become 'FAIR'. Current activities include development of the generic informed consent form for all registries. These are currently being reviewed and should be available soon (and translated into all European languages).

Work is ongoing on identifying domain-specific common data elements, which could be available in more than one registry. Facilitating registry access to European Patient Identity Management (EUPID) is also ongoing. A series of webinars is being held every Tuesday to inform everyone involved in ERN registries. ♥

PRKAG2-registry renewed

Amsterdam, 26 October 2021

The PRKAG2 registry has been renewed and has a new registry leader and coordinator: Pablo Garcia Pavia* and his colleague Fernando Dominguez* (Hospital Universitario Puerta de Hierro Majadahonda, Madrid). They have updated the set of the data elements in the REDCap platform, hosted by the Amsterdam UMC. These changes have been processed and now the new registry is available for adding patients. It would be great if all members and affiliated partners of the ERN add their PRKAG2 patients to the registry. *PMID:32646569.

To add your patients please contact: fdominguezr@salud.madrid.org ♥



En route: LMNA-registry

Amsterdam, 24 October 2021

A collaboration between the LMNA research groups from Paris (Karim Whabi*, Gisèle Bonne*, and Philippe Charron*), Amsterdam (Yigal Pinto and Arthur Wilde), ERN EURO-NMD (ERN for Neuromuscular Disease, coordinated by Teresinha Evangelista) and the newly founded patient advocacy group (LMNACardiac.org network) has led to a new REDCap-based ERN registry of patients with a LMNA-related disease. Existing files will be translated and the CRF will be build in consensus with the pre-mentioned registry leaders. As soon as possible, further announcements will be done where after ERN members and affiliated partners that are interested will be able to participate and enter their patients in the registry.

*PMID: 31155932 ❤️



LMNA related heart disease?



Please join us at
lmacardiac.org to take part in
our network of patients and
specialists

Latest ERN GUARD-Heart Publications

- Task Force for the management of COVID-19 of the European Society of Cardiology:** Colin Baigent, Stephan Windecker, Daniele Andreini, Elena Arbelo, Emanuele Barbato, Antonio L Bartorelli, Andreas Baumbach, Elijah R Behr, Sergio Berti, Héctor Bueno, Davide Capodanno, Riccardo Cappato, Alaide Chieffo, Jean-Philippe Collet, Thomas Cuisset, Giovanni de Simone, Victoria Delgado, Paul Dendale, Dariusz Dudek, Thor Edvardsen, Arif Elvan, José R González-Juanatey, Mauro Gori, Diederick Grobbee, Tomasz J Guzik, Sigrun Halvorsen, Michael Haude, Hein Heidbuchel, Gerhard Hindricks, Borja Ibanez, Nicole Karam, Hugo Katus, Fredrikus A Klok, Stavros V Konstantinides, Ulf Landmesser, Christophe Leclercq, Sergio Leonardi, Maddalena Lettino, Giancarlo Marenzi, Josepa Mauri, Marco Metra, Nuccia Morici, Christian Mueller, Anna Sonia Petronio, Marija M Polovina, Tatjana Potpara, Fabien Praz, Bernard Prendergast, Eva Prescott, Susanna Price, Piotr Pruszczyk, Oriol Rodríguez-Leor, Marco Roffi, Rafael Romaguera, Stephan Rosenkranz, Andrea Sarkozy, Martijn Scherrenberg, Petar Seferovic, Michele Senni, Francesco R Spera, Giulio Stefanini, Holger Thiele, Daniela Tomasoni, Lucia Torracca, Rhian M Touyz, Arthur A Wilde, Bryan Williams. European Society of Cardiology guidance for the diagnosis and management of cardiovascular disease during the COVID-19 pandemic: part 1—epidemiology, pathophysiology, and diagnosis. *Eur Heart J.* 2021 Nov 16:ehab697. doi: 10.1093/eurheartj/ehab696.
- Task Force for the management of COVID-19 of the European Society of Cardiology (see author list above)** ESC guidance for the diagnosis and management of cardiovascular disease during the COVID-19 pandemic: part 2—care pathways, treatment, and follow-up. *Eur Heart J.* 2021 Nov 16:ehab696. doi: 10.1093/eurheartj/ehab697.
- Knops RE, van der Stuijt W, Delnoy PPHM, Boersma LVA, Kuschyk J, El-Chami MF, Bonnemeier H, Behr ER, Brouwer TF, Kaab S, Mittal S, Quast ABE, Smeding L, Tijssen JGP, Bijsterveld NR, Richter S, Brouwer MA, de Groot JR, Kooiman KM, Lambiase PD, Neuzil P, Vernooy K, Alings M, Betts TR, Bracke FALE, Burke MC, de Jong JSSG, Wright DJ, Jansen WPJ, Whinnett ZI, Nordbeck P, Knaut M, Philbert BT, van Opstal JM, Chiccos AB, Allaart CP, Borger van der Burg AE, Clancy JF, Dizon JM, Miller MA, Nemirovsky D, Surber R, Upadhyay GA, Weiss R, de Weger A, Wilde AAM, Olde Nordkamp LRA. Efficacy and Safety of Appropriate Shocks and Antitachycardia Pacing in Transvenous and Subcutaneous Implantable Defibrillators: An Analysis of All Appropriate Therapy in the PRAETORIAN trial. *Circulation.* 2021 Nov 14. doi: 10.1161/CIRCULATIONAHA.121.057816.

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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