

Wednesday 7 June 2023

# ERN

## GUARD-Heart

Gateway to Uncommon And Rare Diseases of the Heart

EUROPEAN REFERENCE NETWORKS  
FOR RARE, LOW-PREVALENCE AND COMPLEX DISEASES

Share. Care. Cure.



ERN GUARD-HEART BIMONTHLY NEWSLETTER

YEAR 2023 NUMBER 4

### Reimbursement for completed CPMS cases

**Amsterdam, 01-06-2023**

CPMS activity is an important indicator within the ERNs. During the current grant-period (until September 2023) there is budget available for a (small) fee for completed CPMS-cases. As presented during the ERN GUARD-Heart board-meeting last year in Barcelona, we propose that each HCP presents **at least 5 cases in CPMS (before September 2023)** and when these cases are completed (outcome report available) the HCP can reimburse 200 EURO per case. In principle there is not a maximum number of reimbursable cases per HCP. This money is only available for the HCP, and cannot be transferred to private accounts. To support you for the reimbursement procedure, a template was created for your CPMS-invoices. This form can be found at the ERN-website: [Template-CPMS-230323.pdf](https://ern-net.eu/Template-CPMS-230323.pdf) ([ern-net.eu](https://ern-net.eu))  
→ Please send us the CPMS-invoices for the completed cases preferably before 15-07-2023. ❤

### Grant submission completed

**Amsterdam, 06-06-2023**

Yesterday, the grant for the next period (1 september 2023 – 1 september 2027) was submitted, for the continuation of the work in the ERN. The workpackages and plans will be presented during the next board meeting. ❤

### ERN GUARD-Heart board meeting in Amsterdam

**Amsterdam, 01-06-2023**

As announced already by e-mail, the next board meeting of ERN GUARD-Heart will be held in Amsterdam again, this time because of the combination with the ESC-congress. The hybrid meeting will be held at **Monday 28<sup>th</sup> of August**, in the Rosarium, Amstelpark 1, in Amsterdam, which is on a 15 minutes walking distance (1,2 km) from the ESC (Amsterdam RAI) and a nearby trainstation to the airport. There will be lunch available from 12:15 PM, and the meeting will start at 13:00 (closure around 17:00). To be able to organize this as structured as possible, we herewith ask all the HCP representatives to fill in their plans and availability, in the link below:

<https://nl.surveymonkey.com/r/7F8KLJG>

→ Please fill in before 20-06-2023. ❤



Rosarium, Amsterdam



## Who are our ePAG's?

**Amsterdam, 01-06-2023**

As second in the series of ePAGs who introduce themselves we are grateful to **Ester Costafreda**, for sharing her story with us:



*My name is Ester Costafreda, cofounder of SAMS Association, a Spanish association of patients and families with inherited cardiomyopathies and channelopathies.*

*On March 12th 2014, my life changed forever. My son Angel who was 15 years old died of sudden death while he was sleeping without having had any symptoms before of cardiomyopathy.*

*We couldn't save our son's life but in 2017 we founded SAMS Association to work in decreasing the number of sudden deaths with activities of awareness, psychological support, networking, and prevention by training the population on Pulmonary Cardiac Resuscitation (PCR) and use of Automated External Defibrillator (AED).*



*Our family  
in  
August 2013*

*In August 2017, the European Patient Advocacy Group (Epag) in the ERN Guard-Heart was founded to represent the voice and advocate for patients. SAMS was one of the first 4 patient organizations of the group and I was appointed as co-chair for one year and as chair in 2018. Our main goal was to increase the number of patient organizations in our Epag making sure all countries in the ERN were represented to be able to exchange knowledge between patients and different countries and to work side by side with health care providers sharing our knowledge in living with these inherited heart diseases. Currently I am again co-chair of the Epag.*

*One of our main activities in SAMS Association is the psychological support. We cannot cure these diseases but at least we can care about the patients. In some cases, our patients must face the death of a young person in their families, the diagnosis of a young member who must have an ICD implanted, must stop doing sports, must take medication or be in list for a heart transplant.*

*All these experiences, questions, doubts, and fears require a professional support for patients, their families, and a peer-to-peer support group to be able to live with these diseases feeling part of a community. We also give this support to cardiologists who unfortunately are not always able to save their patients.*

*In January 2022 after 18 years, I quit my previous job to dedicate exclusively to the Association. One year before we had begun our cardio protected program in schools and companies which was having a big success. In Spain only 10% of the population knows what to do in front of a sudden death or a cardiac arrest. After two years we have already trained more than 2500 students teaching them how to save lives with their hands doing a PCR and using an AED.*

*One of our main awareness activities is the sponsoring of our SAMS day organized by Georgia Sarquella Brugada, head of the Arrhythmia Unit in Sant Joan de Déu Hospital with the participation of a big list of expert cardiologists, geneticists, gynaecologists with an attendance of more than 400 people. Last edition was in October 2022. It is a very important day for patients and their families where we also organize PCR and health food workshops.*



*Another of our main activities is networking. We are members of several national, European, and international organizations such as FEDER (Spanish Federation of Rare Diseases), Eurordis where we are trained on medical research and development as well as communication skills and where we were invited to participate in the Rare Disease Week last February to meet with Spanish Members of the European Parliament in the EU to advocate for a European Rare Disease action plan.*

*We are also one of the members of the Cardiomyopathy council in Global Heart Hub where we organized an awareness campaign on cardiomyopathies last year and we are now working on a Genetics campaign on June this year that will be translated into 11 languages.*

*Currently there are no borders Cardiologists are the experts in rare diseases of the heart and we are the experts in living with these diseases. Rare cardiac diseases are a global problem, and we need to create synergies worldwide and work together, collaborate and share knowledge and experiences practicing medicine from the heart. ♥*



*25th anniversary of Arrhythmia Unit in Sant Joan de Déu Hospital (April 2023)*

<https://www.samsasociacion.com/>



## ERICA survey on ERNs Clinical Research

This survey aims to collect information on the status quo on ERNs and Clinical Research. It is developed by ERICA WP4 Clinical Trial Support on the basis of the previous survey delivered in 2018.

The results of the two surveys will be compared to provide a more comprehensive state-of-the-art of ERNs and clinical research.

The survey is addressed both to ERN Coordinators and their ERN HCP members

Please fill in the survey here:

[https://ec.europa.eu/eusurvey/runner/ERICA\\_survey](https://ec.europa.eu/eusurvey/runner/ERICA_survey)

It will be available until 15 June. ♥







## CardioMYopathy Awareness Week

Join us (June 26–July 2)!

MYself. MYfamily. MYlegacy.  
Is CardioMYopathy in MY family?

Please mark your calendars and join us during Cardiomyopathy Awareness Week (June 26 to July 2) to drive awareness of cardiomyopathy. The objective of the MYCardiomyopathy campaign is to raise awareness of cardiomyopathy and the importance of getting genetically tested to protect families and help save lives.

The Global Heart Hub together with the Cardiomyopathy Patient Council have built the foundation and we look forward to sharing our campaign with you in the coming weeks. We hope you will join us in sharing the materials (including videos and social media posts) with your network and on your social media channels. MYCardiomyopathy materials will be available to share in multiple languages in the coming weeks.

We look forward to your support in raising awareness of cardiomyopathy together during this important week. To join the campaign or for more information, please email [info@globalhearthub.org](mailto:info@globalhearthub.org).

The awareness campaign is led by the Cardiomyopathy Patient Council of the Global Heart Hub. The Cardiomyopathy Patient Council was formed in November 2021 and brings together patient organisations across the world to increase awareness and understanding of cardiomyopathy. The Patient Council advocates for improved diagnosis of the condition, access to appropriate testing, counselling and treatments. In addition, the Council aims to raise awareness and improve understanding of the disease, with a view to enabling people to live well with the condition.

This campaign has been developed in collaboration with the US Genetic Cardiomyopathy Awareness Campaign ([www.geneticcardiomyopathy.org](http://www.geneticcardiomyopathy.org))



Supported by





# ERN evaluation reports

**Amsterdam, 01-06-2023**

The Andalusian agency for healthcare quality started to share the results of the 5 –years evaluation of the ERNs. In our ERN, all 21 HCPs which are included in the ERN since the beginning, completed the self-evaluation and the supporting documentation. Five HCPs had an onsite visit as well, those centres will receive the outcome report at or after the 15<sup>th</sup> of June. The final outcome will be 'satisfactory' or 'needs improvement'. Please contact us if any questions or any help is needed. We are happy to share the general outcome of the Network evaluation which was: 'satisfactory'. ❤



## Latest ERN GUARD-Heart Publication(s)

1. Miles C, Boukens BJ, Scrocco C, Wilde AAM, Nademanee K, Haissaguerre M, Coronel R, Behr ER. Subepicardial Cardiomyopathy: a disease underlying J-Wave syndromes and idiopathic ventricular fibrillation. *Circulation*. 2023; 23:1622-1633. PMID 37216437.
2. Krasemann T, Butera G, Thomson J, Chessa M, Bokenkamp R. AEPC recommendations for training in interventional catheterization for CHD. *Cardiol Young* 2023;22:1-8. PMID 37212061.
3. Stroeck SLVM, Hellebrekers D, Claes GRF, Krapels IPC, Henkens MHTM, Sikking M, Vanhoutte EK, Helderma-van den Enden A, Brunner HG, van den Wijngaard A, Verdonchot JAJ. Diagnostic and prognostic relevance of using large gene panels in the genetic testing of patients with dilated cardiomyopathy. *Eur J Hum Genet*. 2023; may 17; online ahead of print, PMID 37198425.
4. Nademanee K, Chung FP, Sacher F, Nogami A, Nakagawa H, Jiang C, Hocini M, Behr E, Veerakul G, Jan Smit J, Wilde AAM, Chen SA, Yamashiro K, Sakamoto Y, Morishima I, Das MK, Khongphatthanayothin A, Vardhanabhuti S, Haissaguerre M. Long-term outcomes of Brugada Substrate Ablation: a report from BRAVO (Brugada ablation of VF substrate ongoing multicenter registry). *Circulation* 2023;147:1568-1578. PMID 36960730.
5. Giannetti F, Barbieri M, Shiti A, Casini S, Sager PT, Das S, Pradhananga S, Srinivasan D, Nimani S, Alerni N, Louradour J, Mura M, Gnechi M, Brink P, Zehender M, Koren G, Zaza A, Crotti L, Wilde AAM, Schwartz PJ, Remme CA, Gepstein L, Sala L, Odening KE. Gene- and variant-specific efficacy of serum/glucocorticoid-regulated kinase 1 inhibition in long QT syndrome types 1 and 2. *Europace* 2023; 25 (5): euad094. PMID 37099628.

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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**  
Network Manager  
Academic Medical Center  
Amsterdam, Netherlands  
Email: n.hofman@amc.nl  
Mobile: +31622046435



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