CONgenital CORvitia

CONCOR www.CONCOR.net

National registry and DNA-bank of adults with congenital heart disease

Number of participating hospitals: 103
Number of included patients: 10,942

Project group: BJM Mulder, AMC Amsterdam; ET van der Velde, LUMC Leiden; J Vis, ICIN; CJM Engelfriet-Rijk, ICIN; I Harms, ICIN; S Mantels, ICIN

Steering committee: BJM Mulder, chairman, ICIN; ET van der Velde, LUMC Leiden; ECM Mariman, AZM Maastricht; FJ Meijboom, Radboud UMC Nijmegen; HW Vliegen, LUMC Leiden; G Veen, VUMC Amsterdam; PG Pieper, UMC Groningen; HWM Plokker, St Antonius Hospital Nieuwegein; APJ van Dijk, Radboud UMC Nijmegen; JLM Stappers, AZM Maastricht; JW Roos-Hesselink, EMC Rotterdam; JGP Tijssen, AMC Amsterdam; RMF Berger, UMC Groningen; GJ Sieswerda, UMC Utrecht

Figure 1. CONCOR research nurses recently visited the OLVG hospital in Amsterdam; 54 patients were included in CONCOR.

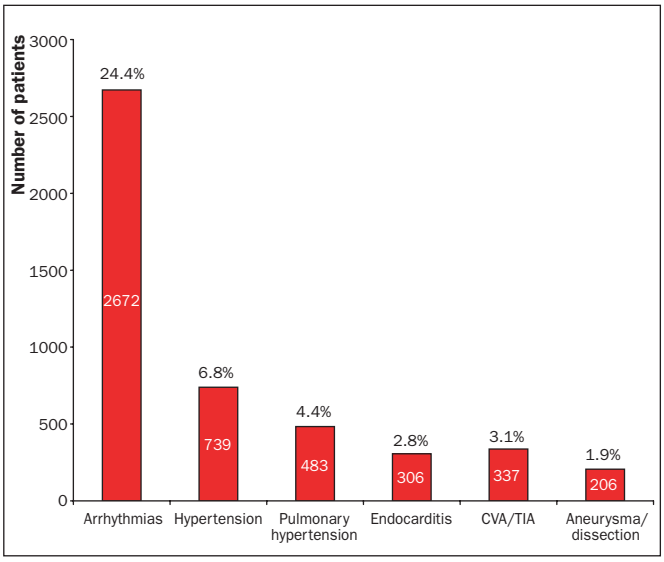


Figure 2. Late complications occur frequently in CONCOR.

GENetic CORvitia

GENCOR Cardiogenetische databank

National registry for patients and families with a familial heart disease

www.gencor.nl

Number of included patients: 1548
28 July 2009

Project leaders: Y.M. Pinto, ICIN; A.A.M. Wilde, ICIN

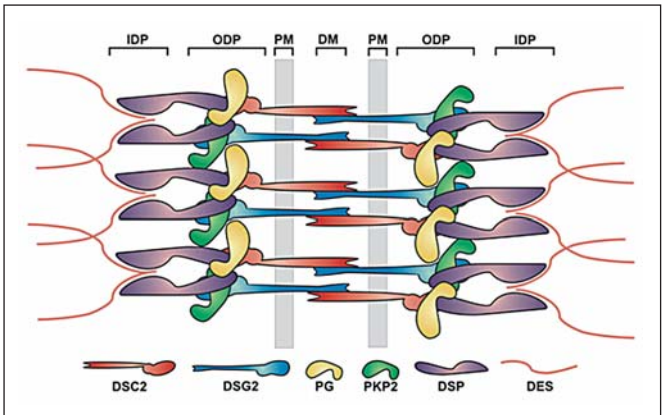
Project coordination: J.F. Hermans-van Ast, ICIN

Steering committee: Y.M. Pinto, chairman, ICIN; A.A. Wilde, chairman, ICIN; I.M. van Langen, AMC Amsterdam; J.P. van Tintelen, UMC Groningen; D.E. Atsma, LUMC Leiden; Y.H. Arens, MUMC Maastricht; C.L. Marcelis, UMCN Nijmegen; M. Michels, EMC Rotterdam; J.J. van der Smagt, UMC Utrecht; A.C. Houweling, VUMC Amsterdam

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterised by fibrofatty replacement of cardiomyocytes. About 50% of patients with ARVC have affected family members, suggesting a genetic cause. In recent years, progress has been made in elucidation of the genes underlying these inherited forms of ARVC. Mutations in the genes encoding proteins of the cardiac desmosome, a structure important for cell-cell interaction, have proved to be the major player in the pathogenesis of ARVC. To date, mutations have been identified in more than 50% of cases, in particular in the PKP2, DSG2, and DSC2 genes. Interestingly, some patients carry more than one disease-causing variant in one or more genes. Because a substantial number of mutations have yet to be identified, one of the GENCOR studies will focus on patients without known mutations. This study will try to elucidate whether mutations are missed with standard sequencing techniques. Moreover, candidate genes important for cell structure or adipogenic pathways will be studied. It is hoped that this will lead to a higher yield of genetic screening and to a better understanding of the pathogenesis of ARVC. ■

Reference

1 van Tintelen JP, Hofstra RM, Wiesfeld AC, van den Berg MP, Hauer RN, Jongbloed JD. Molecular genetics of Arrhythmogenic Right Ventricular Cardiomyopathy: emerging horizon? Curr Opin Cardiol. 2007;22:185-92.



IDP=inner dense plaque, ODP=outer dense plaque, PM=plasma membrane-bound, DM=dense midline, DSC2=desmocollin-2, DSG2=desmoglein-2, PG=plakoglobin, PKP2=plakophilin-2, DSP=desmoplakin, DES=desmin.

Figure 1. Schematic representation of the cardiac desmosome.[1]

ZAHARA II

Pregnancy in adult congenital heart disease

A prospective, multi-centre cohort study to examine the cardio-uterine interaction in pregnancy of women with congenital heart disease (CHD)

Start of study: 1 March 2008
Planned inclusion: 160 patiens, 60 controls
Number of included patients: 101
Number of included controls: 11
5 May 2009

Study coordination: A Balci, ICIN; Department of Cardiology, UMCG, Groningen, the Netherlands; a.balci@thorax.umcg.nl

Project leaders: PG Pieper, Department of Cardiology, UMCG, Groningen
JG Aarnoudse, Department of Obstetrics & Gynaecology, UMCG, Groningen

Principal investigators: http://www.studies-obsgyn.nl/home/page.asp?page_id=803

For information or inclusion of patients, please contact us at: a.balci@thorax.umcg.nl. ■

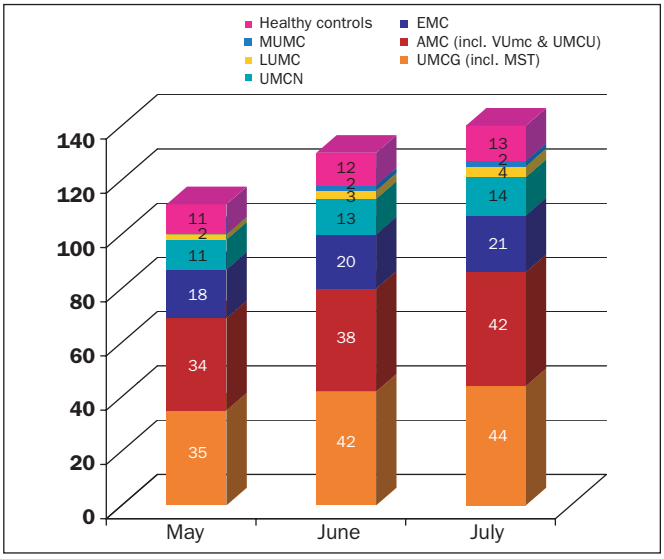


Figure 1. Inclusion of patients and controls ZAHARA II.

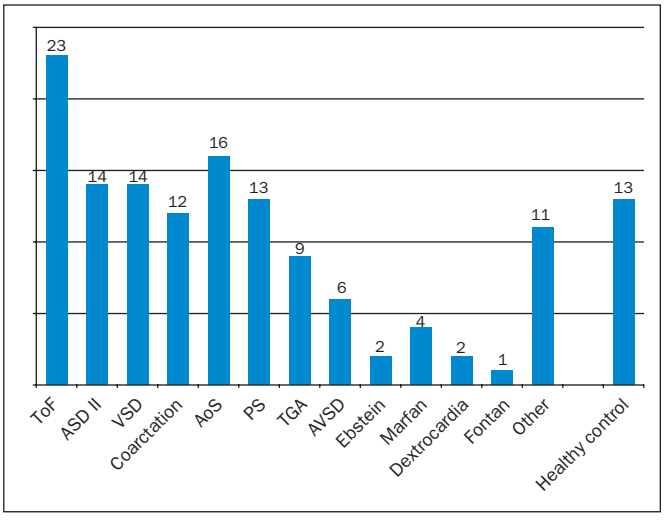


Figure 2. Distribution of main CDH-diagnoses in ZAHARA II.

Registry for Heart Disease and Pregnancy

In cooperation with the ESC and the AEPC

Total inclusion Europe: 551
Total inclusion Netherlands: 58
July 2009

International Expert Committee: Jörg Stein, Austria; Roger Hall, UK; Jolien Roos-Hesselink, the Netherlands
National coordinators for the Netherlands: Jolien Roos-Hesselink, Department of Cardiology, ErasmusMC, Rotterdam; Els Pieper, Department of Cardiology, UMCG, Groningen

Study summary

Heart disease is a major cause of mortality in pregnant women. The most common causes of mortality are aortic pathology, cardiomyopathy and ischaemic heart disease.

This registry will focus on complications during pregnancy, method of delivery and medication (anticoagulation) during pregnancy. All patients with structural heart disease who gave birth after 1 January 2007 can be included in this electronic, web-based registry.

Inclusion

- All patients with structural heart disease (valvular, congenital, or ischaemic heart disease, disease of the aorta or cardiomyopathy) who were pregnant and/or gave birth after 1 January 2007.

Exclusion

- Non-structural heart disease, e.g. arrhythmias occurring in the context of a normal heart
- Inclusion in the ZAHARA II study
- Miscarriage / abortion <20 weeks of gestation. ■

Please join the Registry!

Contact the EuroHeartSurvey: ehs@escardio.org or contact us at: j.roos@erasmusmc.nl *or* p.g.pieper@thorax.umcg.nl.

Table 1. Inclusion in the Netherlands

Rotterdam	Erasmus MC	41
Utrecht	Wilhelmina Children's Hospital	7
The Hague	MC Haaglanden	4
Groningen	University Medical Center	1
Lelystad	IJsselmeer Hospitals	1
Leiden	Leiden University Medical Center	1

RACE 3

Planned start inclusion:
1 June 2009

This study is supported by: Astra Zeneca, Biotronik, Boehringer Ingelheim, Boston Scientific, Medtronic, Sanofi Aventis, and St. Jude Medical.

Steering committee: Isabelle C. Van Gelder, UMC Groningen; Marco W. Alings, Amphia Hospital Breda; Harry J.G.M. Crijns, Maastricht UMC; Raymond Tukkie, Kennemer Gasthuis Haarlem; Johan Brügemann, UMC Groningen; Joep R.L.M. Smeets, UMC Nijmegen; Charles J.H.J. Kirchhof, Rijnland Hospital, Leiderdorp; Hans L. Hillege, UMC Groningen; Jan G.P. Tijssen, AMC Amsterdam; Dirk J. Van Veldhuisen, UMC Groningen
Study coordination: Marcelle D. Smit, UMC Groningen; PhD Student Ismaël D. Achekar, UMCG Groningen

Endpoints in the Race 3 study

The primary endpoint of the Race 3 study is sinus rhythm after one year of follow-up, defined as sinus rhythm during ≥6/7th of assessable time of continuous seven-day Holter monitoring during the last week of follow-up *and* the patient is still following a rhythm-control strategy.

Secondary endpoints include:
1. Sinus rhythm on end-of-study ECG;
2. Sinus rhythm after one year of follow-up *without* the need for class III ion-channel antiarrhythmic drugs;
3. Sinus rhythm after one year of follow-up *without* the need for pulmonary vein isolation;
4. Permanent atrial fibrillation;
5. Number of electrical cardioversions;
6. Number of radiofrequency ablations;
7. AV node ablation;
8. Left ventricular function;
9. Exercise capacity;
10. Hospitalisation for heart failure;
11. Hospitalisation for other cardiovascular reasons;
12. Mortality;
13. Biomarkers and genetics;
14. Quality of life;
15. Cost and cost-effectiveness. ■

HEBE III

A prospective, randomised, clinical study to examine the effects of a single bolus of erythropoietin on left ventricular function in patients with a myocardial infarction

Number of included patients: 529
1 August 2009

Funded by the Netherlands Heart Foundation

Study coordination: A. Belonje

Principal investigators: A.A. Voors, F. Zijlstra, D.J. van Veldhuisen

Participating centres: University Medical Center Groningen, University Medical Center Leiden, Medical Centre Alkmaar, Isala Clinics Zwolle, Amphia Hospital Breda, Academic Medical Center Amsterdam, St. Antonius Hospital Nieuwegein

Last patient out, time to analyse…

As we mentioned in the July/August issue of this journal, we stopped including patients on 1 June. The final patient was seen at the outpatient clinic in July and now we are gathering all the data in all the participating centres. In the upcoming issues of the Netherlands Heart Journal we will keep you informed about the latest results of the first analyses of the HEBE III trial. In this issue we give you the results of the input of each centre to the HEBE III inclusion (figure 1). ■

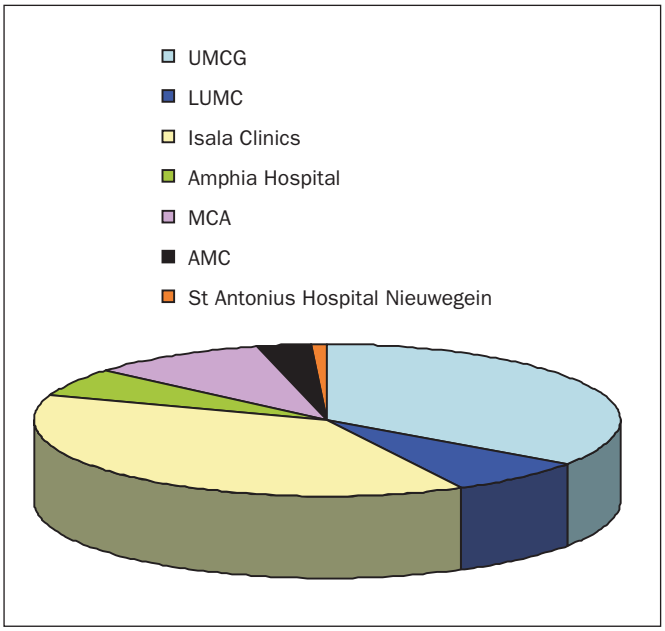


Figure 1.