# Ajmaline testing for early detection of Arrhythmogenic Cardiomyopathy (ACM)

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Ajmaline induces more pronounced electrical and mechanical dysfunction of those myocardial areas that are affected in the early stage of ACM. Therefore, this ajmaline provocation and electrocardiographic imaging could identify those mutation...

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|---|---|
| **Ethische beoordeling** | Niet van toepassing |
| **Status** | Anders |
| **Type aandoening** | - |
| **Onderzoekstype** | Interventie onderzoek |

## Samenvatting

### ID

NL-OMON29255

**Bron**
Nationaal Trial Register

**Aandoening**

Arrhythmogenic cardiomyopathy, ARVC, carriers of pathogenic PLN and PKP2 mutations

### Ondersteuning

**Primaire sponsor:** UMC Utrecht
**Overige ondersteuning:** Hartstichting, e-DETECT consortium

### Onderzoeksproduct en/of interventie

### Uitkomstmaten

**Primaire uitkomstmaten**

Mean changes in Activation Time Duration before and during ajmaline administration between the subtricuspid area and LV/RV/RVOT as calculated by ECG imaging.

# Toelichting onderzoek

### Achtergrond van het onderzoek

Rationale: Arrhythmogenic Cardiomyopathy (ACM) is a disease with a genetic origin and involves cardiac desmosomes dysfunction and fibrofatty replacement of the myocardium. Clinically, patients present with ventricular arrhythmias or sudden cardiac death. Genetic testing in family members of patients with ACM shows incomplete penetrance of the disease. Earlier studies have shown that the electromechanical interval and RV deformation imaging is abnormal in the subtricuspid area of the right ventricle (RV) even in the early stage of disease. We hypothesize that ajmaline induces more pronounced electrical and mechanical dysfunction of those myocardial areas that are affected in the early stage of ACM. Therefore, this ajmaline provocation could identify those mutation carriers who are at risk for the development of ACM, arrhythmias and/or sudden cardiac death.

Objective: Describe the electrocardiographic changes and areas of late myocardial activation using electrocardiographic imaging in PLN and PKP2 mutation carriers during ajmaline provocation.

Study design: Multicentre, diagnostic trial, cohort study

Study population: All patients who are diagnosed with ACM due to an PLN and PKP2 mutation and their asymptomatic family members who are PKP2 and PLN mutation carriers known in the UMC Utrecht, UMC Groningen and Amsterdam Medical Centre. Patients with structural normal hearts (determined by echocardiography or cardiac MRI) who are referred for ajmaline provocation to exclude Brugada syndrome.

Intervention: ajmaline (class 1c sodium channel blocker, with a short half-life) infusion in fractions of 10mg every minute up to a target dose of 1mg/kg.

Main study parameters/endpoints: difference in mean activation time duration (ATD) of the subtricuspid area before and after ajmaline provocation using electrocardiographic imaging (ECGI).

### Doel van het onderzoek

Ajmaline induces more pronounced electrical and mechanical dysfunction of those myocardial areas that are affected in the early stage of ACM. Therefore, this ajmaline provocation and electrocardiographic imaging could identify those mutation carriers who are at risk for the development of ACM, arrhythmias and/or sudden cardiac death.

**Onderzoeksopzet**

T0 selection

T1 ajmaline provocation

T2 bloedtest liver parameters

T3 Regular follow up

**Onderzoeksproduct en/of interventie**

Ajmaline provocation

# Contactpersonen

## Publiek

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## Wetenschappelijk

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# Deelname eisen

## Belangrijkste voorwaarden om deel te mogen nemen (Inclusiecriteria)

Patients or asymptomatic carriers of pathogenic PLN mutation, pathogenic PKP2 mutation or patients without structural heart disease who are referred for ajmaline provocation to exclude Brugada syndrome.

New York Heart Association functional class ¡Ü 1.

## Belangrijkste redenen om niet deel te kunnen nemen (Exclusiecriteria)

Severe hepatic impairment (Child-Pugh class C)

Severe renal dysfunction (eGFR <30 ml/min/kg)

Symptomatic heart failure, NYHA ¡Ý 2

Women who are currently pregnant

Known intolerance or contraindication to Ajmaline

Sick sinus syndrome, second or third degree AV block without pacemaker implantation

Recent myocardial infarction

Known strong allergic reaction to ECG electrodes

# Onderzoeksopzet

## Opzet

| | |
|---|---|
| Type: | Interventie onderzoek |
| Onderzoeksmodel: | Anders |
| Toewijzing: | Niet-gerandomiseerd |

**Controle:** Actieve controle groep

### Deelname

Nederland

| | |
|---|---|
| Status: | Anders |
| (Verwachte) startdatum: | 01-06-2018 |
| Aantal proefpersonen: | 65 |
| Type: | Onbekend |

## Ethische beoordeling

Niet van toepassing

| | |
|---|---|
| Soort: | Niet van toepassing |

## Registraties

### Opgevolgd door onderstaande (mogelijk meer actuele) registratie

Geen registraties gevonden.

### Andere (mogelijk minder actuele) registraties in dit register

Geen registraties gevonden.

### In overige registers

| Register | ID |
|---|---|
| NTR-new | NL6861 |
| NTR-old | NTR7039 |
| Ander register | EudraCT: 2018-000752-18 : METC nummer: 17-924, ABR nummer: 65196 |

## Resultaten