# A Multicentre, Prospective, Longitudinal, Observational Natural History Study to Evaluate Disease Progression in Subjects with Usher Syndrome type 1B (USH1B)

Published: 08-08-2018 Last updated: 11-04-2024

To evaluate the natural progression of disease over time in USH1B patients using visual field testing and best corrected visual acuity.

Ethical reviewApproved WMOStatusRecruitment stoppedHealth condition typeEye disorders NEC

**Study type** Observational non invasive

# **Summary**

#### ID

NL-OMON46147

#### Source

ToetsingOnline

#### **Brief title**

Natural history of Usher syndrome type 1B

#### **Condition**

Eye disorders NEC

#### **Synonym**

Usher syndrome type 1B

#### Research involving

Human

## **Sponsors and support**

Primary sponsor: Oogziekenhuis Rotterdam

Source(s) of monetary or material Support: EU

#### Intervention

**Keyword:** Natural history, Usher syndrome type 1B, Vision

#### **Outcome measures**

#### **Primary outcome**

Visual acuity & visual field.

#### **Secondary outcome**

**IOP** 

**ERG** 

OCT

**Fundusphotographs** 

VFQ-25

# **Study description**

#### **Background summary**

This natural history study is being conducted to understand the progression of disease in USH1B patients as measured by visual acuity and visual field testing and a number of other vision-related assessments.

#### Study objective

To evaluate the natural progression of disease over time in USH1B patients using visual field testing and best corrected visual acuity.

#### Study design

Multicenter, prospective, observational.

#### Study burden and risks

Electrodes for ERG measurements can cause superficial abrasions of the cornea which usually heal spontaneously within one day. Other risks are minimal.

Subjects do not benefit from participation. Due to the extensive measurements, participation can be somewhat exhausting; subjects are encouraged to indicate when they need a break. Study related time: 3 times two days.

## **Contacts**

#### **Public**

Oogziekenhuis Rotterdam

Schiedamse Vest 180 Rotterdam 3011 BH NL

#### Scientific

Oogziekenhuis Rotterdam

Schiedamse Vest 180 Rotterdam 3011 BH NL

## **Trial sites**

#### **Listed location countries**

**Netherlands** 

# **Eligibility criteria**

#### Age

Adolescents (12-15 years) Adolescents (16-17 years) Adults (18-64 years) Children (2-11 years) Elderly (65 years and older)

### **Inclusion criteria**

Informed consent
Diagnosed with USH1
Molecular diagnosis of USH1B due to MYO7A mutations (homozygotes or compound heterozygotes)

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Age >= 8 years Visual acuity >= 20/640 in at least one eye

#### **Exclusion criteria**

Unable to communicate with suitable verbal/auditory and/or tactile sign language Participation in clinical study with investigational drug in past 6 months Pre-existing ocular conditions that would interfere with the interpretation of study endpoints (e.g. glaucoma, corneal or significant lenticular opacities, cystoid macular oedema, macular hole) in both eyes

Complicating systemic diseases in which the disease itself, or the treatment for the disease, can alter ocular function

Prior ocular surgery within 3 months

# Study design

## **Design**

**Study type:** Observational non invasive

Masking: Open (masking not used)

Control: Uncontrolled

Primary purpose: Other

#### Recruitment

NL

Recruitment status: Recruitment stopped

Start date (anticipated): 12-12-2018

Enrollment: 10

Type: Actual

## **Ethics review**

Approved WMO

Date: 08-08-2018

Application type: First submission

Review commission: METC Erasmus MC, Universitair Medisch Centrum Rotterdam

(Rotterdam)

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# **Study registrations**

## Followed up by the following (possibly more current) registration

No registrations found.

## Other (possibly less up-to-date) registrations in this register

No registrations found.

## In other registers

Register ID

CCMO NL66164.078.18