Expression of hSPCA1 and ultrastructural analysis of the skin before and after laser therapy in Hailey-Hailey disease

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We aim to (1) study the expression of hSPCA1 in keratinocytes before and after laser therapy and (2) verify the loss of acantholysis by immunohistochemistry and electron microscopy of cell-cell adhesions before and after laser therapy.

Ethical review	Approved WMO
Status	Recruitment stopped
Health condition type	Skin and subcutaneous tissue disorders congenital
Study type	Observational invasive

Summary

ID

NL-OMON56274

Source
ToetsingOnline

Brief title
M. Hailey-Hailey: hSPCA1 expression and skin structure upon laser therapy

Condition

- Skin and subcutaneous tissue disorders congenital

Synonym
acantholytic dermatosis, bullous dermatosis

Research involving
Human

Sponsors and support

Primary sponsor: Medisch Universitair Ziekenhuis Maastricht
Source(s) of monetary or material Support: Ministerie van OC&W

Intervention

Keyword: desmosomes, Hailey Hailey, hSPCA1, Laser

Outcome measures

Primary outcome

Expression of the hSPCA1 calcium pump in keratinocytes before and after

ablative treatment.

Secondary outcome

- Ultrastructure of the skin before and after ablative treatment.

- Expression and localization of desmosomal proteins before and after ablative

therapy.

- Comparison of electron microscopy findings with those in subjects without M.

Hailey-Hailey.

Study description

Background summary

Hailey-Hailey disease is a genetic acantholytic dermatosis caused by a genetic defect in the hSPCA1 calcium pump. Calcium pumps are crucial for the processing of cell-cell adhesion proteins. Loss of function results in painful and burning erosion of the skin. Therapy has been mainly focused on symptom relief and prevention of secondary infection. However, following ablative laser therapy in the MUMC+ the skin remains clear form erosion in the months/years after treatment, regardless the existence of a germline mutation, suggesting that an epigenetic modification occurs in the process of wound healing.

Study objective

We aim to (1) study the expression of hSPCA1 in keratinocytes before and after laser therapy and (2) verify the loss of acantholysis by immunohistochemistry and electron microscopy of cell-cell adhesions before and after laser therapy.

Study design

Observational study with invasive measurements.

Study burden and risks

Participation does not require additional hospital visits and will take 30 min extra time. There is a limited risk of post interventional bleeding and secondary infection, which can be reduced by a single transcutaneous suture using a self-absorbing suture (vicryl 4.0).

AMENDMENT:
As for subjects without M. Hailey Hailey, the present study will be discussed at the end of a regular visit to the department of dermatology. Herefafter, patients will receive the patient information form and the informed consent form. After consent, at the next regular visit on our outpatient clinic, a 2mm skin biopsy for electron microscopy will be taken.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

- Clinically diagnosed M. Hailey-Hailey as confirmed by histopathology and/or genetic analysis.
- Indication for ablative laser therapy
- >18 years old
- Informed consent

Exclusion criteria

- <18 years old
- Treatment with tetracyclines or oral retinoids within the last 30 days preceding participation
- Lack of informed consent

Study design

Design

Study type:	Observational invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	Active
Primary purpose:	Basic science

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	31-01-2019

Enrollment:	20
Type:	Actual

Ethics review

Approved WMO Date:	21-12-2018
Application type:	First submission
Review commission:	METC academisch ziekenhuis Maastricht/Universiteit Maastricht, METC azM/UM (Maastricht)
Approved WMO Date:	03-08-2021
Application type:	Amendment
Review commission:	METC academisch ziekenhuis Maastricht/Universiteit Maastricht, METC azM/UM (Maastricht)
Approved WMO Date:	31-08-2023
Application type:	Amendment
Review commission:	METC academisch ziekenhuis Maastricht/Universiteit Maastricht, METC azM/UM (Maastricht)

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
ClinicalTrials.gov	NCT03849989

Register
CCMO

ID
NL64815.068.18