

Acquired immunological LOW prevalence and Complex Adult diseases of thE SKIN (ALLOCATE SKIN)



European
Reference
Network

for rare or low prevalence
complex diseases

 Network
Skin Disorders (ERN-Skin)



12 SUB THEMATIC GROUPS

**Epidermolysis
Bullosa**

**Ichthyosis &
Palmoplantar
Keratoderma**

**Ectodermal
Dysplasia**
including Skin
Fragility Disorders &
X-linked cutaneous
disorders

**Monogenic
Connective Tissue
Disorders**

**Cutaneous Mosaic
Disorders**

Nevi & Nevoid Skin
Disorders & Complex
Vascular Malformations
& Vascular Tumours

**Cutaneous
diseases related
to DNA Repair
Disorders**

**Autoimmune
bullous diseases
and severe
cutaneous drug
reactions**

**Follicular diseases
(HS, PAPA, PAPASH,
PASH, PASS,
SAPHO),
Vasculitis (ABD, DD),
Complement (AAE,
HAE)**

Photosensitivity

*Non bullous complex
autoimmune/inflam
matory cutaneous
diseases*

*Premature Skin
Ageing*

*Rare cutaneous
proliferation in
children and adults*

Subthematic groups to be included in the ERN-Skin expansion plan

Goal of *ALLOCATE SKIN*



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To improve health care and social support for patients with acquired immunological rare adult diseases of the skin:

- Rare follicular diseases [Hidradenitis suppurativa (HS; familial form), HS syndromes (PAPA, PASH, PAPASH, PASS, SAPHO)]
- Rare cutaneous vasculitis (Adamantiades-Behçet's disease, Malignant Atrophic Papulosis)
- Complement diseases (Hereditary and Acquired Angiodema)

Belgium (Brussels)
Croatia (Zagreb)
Denmark (Odense, Roskilde)
Germany (Dessau, Würzburg)
Hungary (Debrecen, Szeged)
Italy (Padova)
Lithuania (Kaunas)
Netherlands (Rotterdam)
Szech Republic (Prague)
UK (Cardiff, Leeds)

Diagnostic tools and services certified or accredited through National European / International Program



Detailed list of diagnostic technologies and services certified or accredited through National European, or International Programs in *each health care provider member of your sub-thematic groups and conditions of access for patients*

- ❖ Clinical diagnosis
- ❖ Mutation analysis of the nicastrin gene of the gamma-secretase complex
- ❖ Mutations in the PSTPIP1 gene
- ❖ Patchy test
- ❖ Skin sonography
- ❖ Skin thermography
- ❖ HLA typing
- ❖ C4
- ❖ C1 INH antigen
- ❖ C1 INH function
- ❖ FXII mutation

IT communication platform



Name of the responsible for
communication :
Petr ARENBERGER,
Andreas ALTENBURG,
Anette BYGUM
Dagmar PRESSER

- List and provide for each health care provider member of your sub-thematic group, the available information tools in the field of your sub-thematic group such as: website, newsletter, documents adapted to different public.

-Propose a strategy for producing information content adapted to different publics (Health Care Provider, patient representatives and general public)

List :

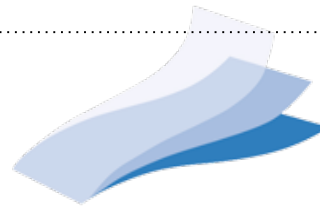
1. European Hidradenitis Suppurativa Foundation e.V.
www.ehsf.org
2. International Society for Adamantiades-Behçet's Disease
3. Deutsches Register Morbus Adamantiades-Behçet's Disease e.V.
www.behcet.de
4. Degos Disease Registry
www.degosdisease.com
5. Danish National HAE Registry
<https://open.rsyd.dk/OpenProjects/da/openProject.jsp?openNo=19>
6. Patient information materials

Registries for ALLOCATE SKIN diseases



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European Dermatology Forum



EURORDIS
Rare Diseases Europe



European
Hidradenitis
Suppurativa
Foundation e.V.

ERHS

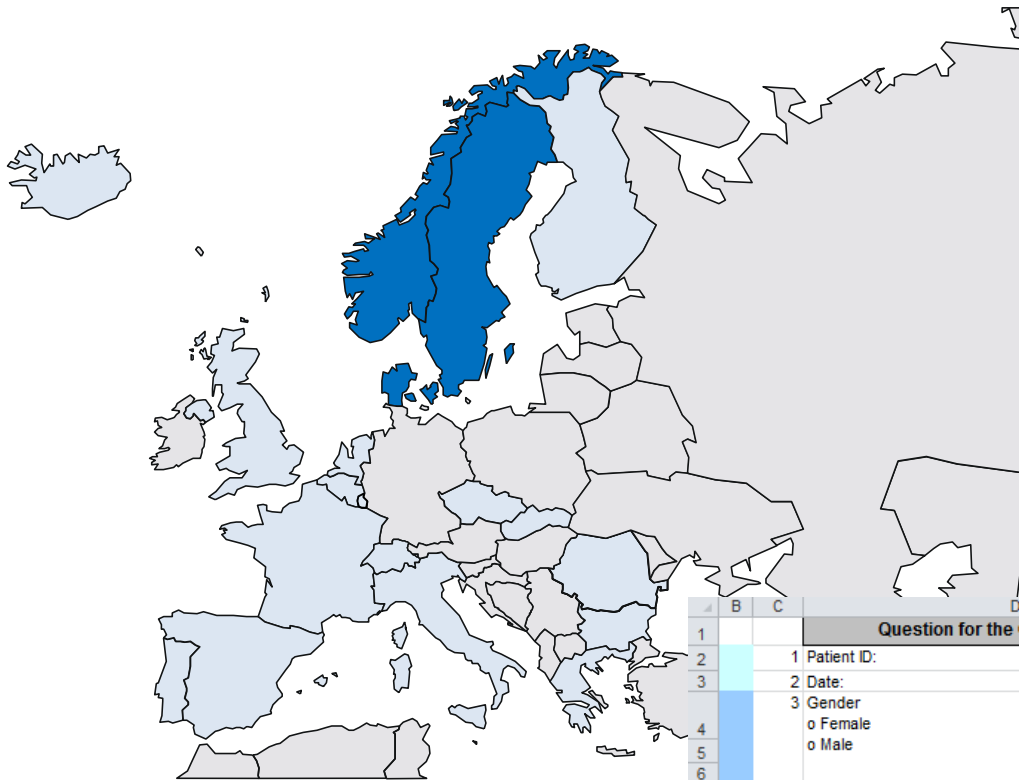
*Deutsches Register
Morbus Adamantiades-Behçet e.V.*



Degos Disease

THE DEGOS DISEASE SUPPORT NETWORK

European Registry of Hidradenitis Suppurativa / acne inversa



HISREG

↑
**compatible
complementary
cooperation**
↓

ERHS

	B	C	D	E	F
			Question for the Openclinica CRF	Country	How to ask
1			1 Patient ID:		
2			2 Date:		
3			3 Gender o Female o Male	Belgium	Sex : o Female o Male
4				Norway	/
5				England	o Female o Male
6				Netherlands	Sex : M / F
7				Spain	Gender? 1. Man 2. Woman
8				France (Pasteur)	/
9				France (Mondor)	Sex : o F o M
10			4 Date of birth: _____ (MM/YYYY)	Belgium	How old are you ?
11				Norway	/
12				England	Age?(yrs)
13				Netherlands	Date of birth? Age?
14				Spain	Date of birthday? (DD/MM/YY)
15				France (Pasteur)	Date of birthday?
16				France (Mondor)	Date of birthday? (DD/MM/YYYY)
17			5 Weight : _____Kg	Belgium	What is your weight?
18				Norway	Weight?
19				England	Weight? (Kg)
20				Netherlands	What is your weight today? (Kg)
21				Spain	Weight? (Kg)
22				France (Pasteur)	Weight? (Kg)
23				France (Mondor)	Weight: (Kg)
24			6 Waist circumference : _____cm	Belgium	/
25				Norway	Waist circumference? (cm)
26				England	/
27				Netherlands	/
28				Spain	Abdominal obesity?(Men > 94 cm, Women > 80cm) 1. Yes 2. No
29				France (Pasteur)	Obesity: o Yes o No o Unknown
30				France (Mondor)	/
31					

New diagnostic and classification criteria for *ALLOCATE SKIN* diseases



International Hidradenitis Suppurativa Severity Score System (IHS⁴)

- **Active, inflammatory lesions**

- Nodule

(raised, 3D, round, infiltrated lesion >10 mm)

1 point

- Abscess

(tender but fluctuating mass >10 mm surrounded by an erythematous area)

2 points

- Draining Tunnel

(Sinus/Fistula; raised, tender but fluctuating longitudinal mass of variable length and depth, ending at the skin surface, and sometimes oozing a fluid)

4 points

- **Score classification**

- Mild

1-3 points

- Moderate

4-10 points

- Severe

≥ 11 points

Clinical outcomes for *ALLOCATE SKIN* diseases



Name of the responsible for
clinical outcomes :

*Eva REMENYIK,
Skaidra VALIUKEVICIENE*

List specific clinical outcomes
(based in the *ERN skin Road Map*)

List specific clinical outcomes :

1. Observational comparative effectiveness research (CER)
2. Special methodology (Core Outcome Measures in Effectiveness Trials (COMET), Outcome Measures in Rheumatology (OMERACT)
3. Hidradenitis Suppurativa CORE outcomes set International Collaboration (HISTORIC)
4. International Dermatology Outcome Measures organization (IDEOM)
5. Patient reported outcomes in HAE

Guidelines for *ALLOCATE SKIN* diseases



Name of the responsible of the guidelines :

*John INGRAM,
Gregor B.E. JEMEC,
Christos C. ZOUBOULIS*

- List and provide for each HCP member of your sub-thematic group the available guidelines at the local, national, european and international level*
1. HS: National and International (actualization 2018)
 2. Syndromic HS: -
 3. Adamantiades-Behçet disease: National
 4. Malignant Atrophic papulosis: -, international upcoming (2018)
 5. HAE: International
 6. AAE: -

e-health tools for patient follow-up in *ALLOCATE SKIN*



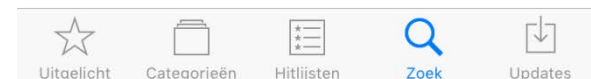
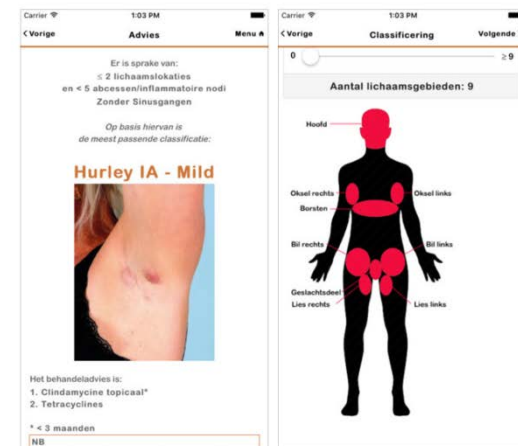
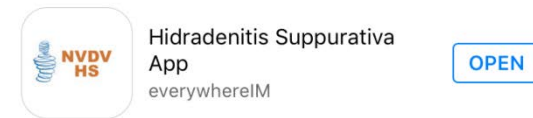
Name of the responsible for e-health :

Veronique DEL MARMOL,
Thomas WILD,
Réka KOVÁCS

List and describe for each health care provider member of your sub-thematic group, the available e-health/connected tools for patient follow up in the field of your sub-thematic group

List :

1. EHSF App
2. Danish HAE App
3. more to be developed



Training resources in *ALLOCATE SKIN* diseases



- **Follicular diseases**

7th International Conference of the EHSF e.V., 7.-9.2.2018, Rotterdam (NL)

6th International Conference of the EHSF e.V., 8.-10.2.2017, Copenhagen (DK)

3rd International Conference on Sebaceous Gland Diseases, 7.-9.9.2016,
Shanghai (CH)

- **Adamantiades-Behçet's disease**

18th International Conference of the ISBD, 13.-15.9.2018, Rotterdam (NL)

17th International Conference of the ISBD, 15.-17.9.2016, Matera (IT)

- **Köhlmeier-Degos disease (Malignant Atrophic Papulosis)**

3rd International Conference, 28.4.2018, NIH, Bethesda (USA)

- **Day of Rare Diseases**

8th Annual Conference of Patients Education, 24.2.2018, Dessau (GE)

7th Annual Conference of Patients Education, 25.2.2017, Dessau (GE)

Cooperation with patient groups in *ALLOCATE SKIN* diseases



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HS research is in rapid development. New knowledge, therapies and stronger cooperation between researchers, therapists and patient associations give hope to you and all those with HS.

The ambition of this publication is to gather and communicate the insights from both patients and specialists, since both perspectives are of major importance for HS understanding and treatment.

We hope that this publication can contribute to a more nuanced understanding of HS among patients and relatives, and in this way, strengthen the cooperation and partnership between patients and physicians.

KNOWLEDGE ON HS

- UNDERSTAND MORE, AND IMPROVE YOUR LIFE LIVING WITH HS

News from Hidradenitis Suppurativa
research for patients and their relatives

