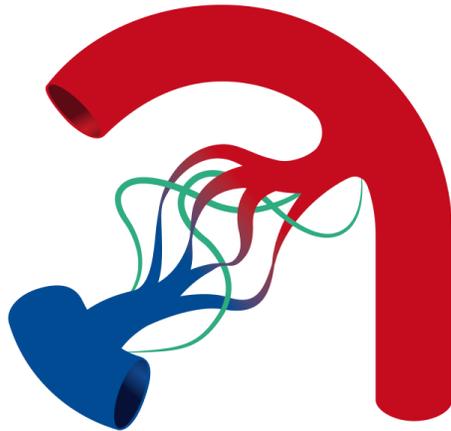




**European
Reference
Network**

for rare or low prevalence
complex diseases

 **Network**
Vascular Diseases
(VASCERN)



VASCERN DAYS 2018 PLENARY SESSION

10.35-12.00: **Feedback & discussion on
the Work Packages progress**
Rare Disease Working Groups Chairs (Prof
Claire Shovlin, Prof Julie De Backer, Dr Leema Robert,
Dr Robert Damstra, Prof Miikka Vikkula)
Transversal WG Chairs (Dr Alessandro Pini, Prof
Leo Schultze Kool), moderated by Marine Hurard

WP1 Sharing of experience: discussion of complex clinical cases on a secured Clinical Patient Management System

<https://vascern.eu/what-we-do/cpms-discussion-of-complex-clinical-cases/>



VASCERN CPMS Activity		
Registered Users	Active Users	Number of Panels
60 (6 guest users)	47	11

In September
2018:
35 active users!



Of the 11 panels opened for VASCERN: 1/11 have made it to the **sign-off stage**, 4/11 to the **outcome stage**, 7/11 to **Assessment stage**, 9/11 to **data completion stage**, all have made it to the **panel selection stage**.

Next steps:

- Resolve technical issues (videoconference tool tested by 4/5 RDWGs)
- Customisation of CPMS consultation form (per RDWG)
- IT helpdesk hired (CEF)

Feedback on experiences
by RDWG Chairs

WP2 Definition of (improved) patient pathways

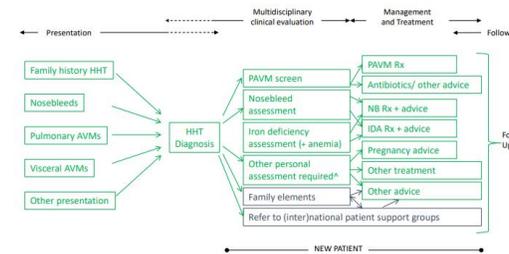


<https://vascern.eu/what-we-do/patient-pathways/>

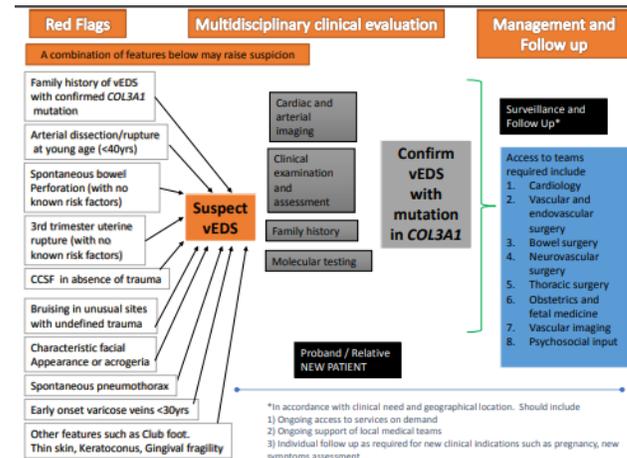
- HHT
 - 1st version published
- HTAD
 - Improved version published
- MSA
 - Improved version published
- PPL
 - Tested within WG
- VASCA
 - Several work ongoing



Hereditary haemorrhagic telangiectasia
Patient Pathway v2, 11/07/2017



*In accordance with clinical need and geographical location. Should include:
1) Ongoing access to services on demand
2) Ongoing support of local medical teams
3) Individual follow up as required for new clinical indications such as pregnancy, new symptoms assessment, and ~6 months post PAVM embolisation.
* As formalised through HHT WG Programmes

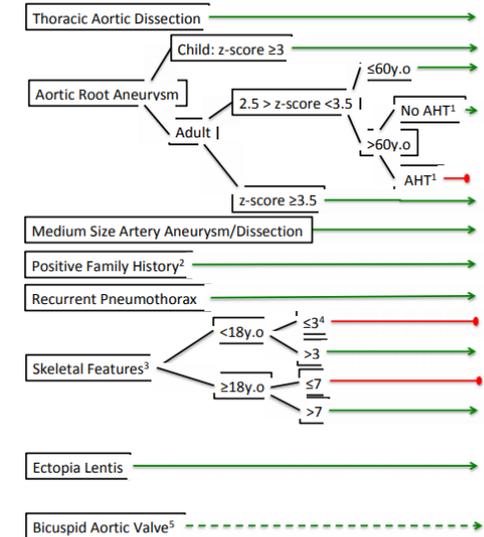


*In accordance with clinical need and geographical location. Should include:
1) Ongoing access to services on demand
2) Ongoing support of local medical teams
3) Individual follow up as required for new clinical indications such as pregnancy, new symptoms assessment...



Vascular EDs

Final approved patient pathway
28/09/2018





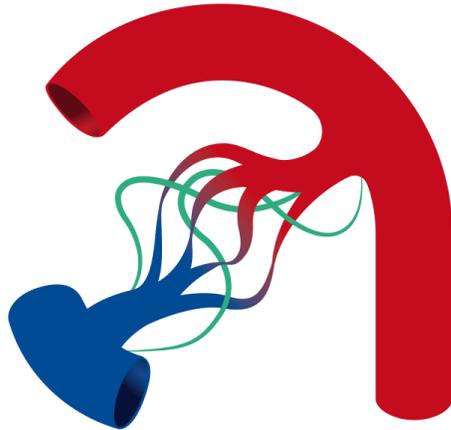
**European
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complex diseases



Network

Vascular Diseases
(VASCERN)

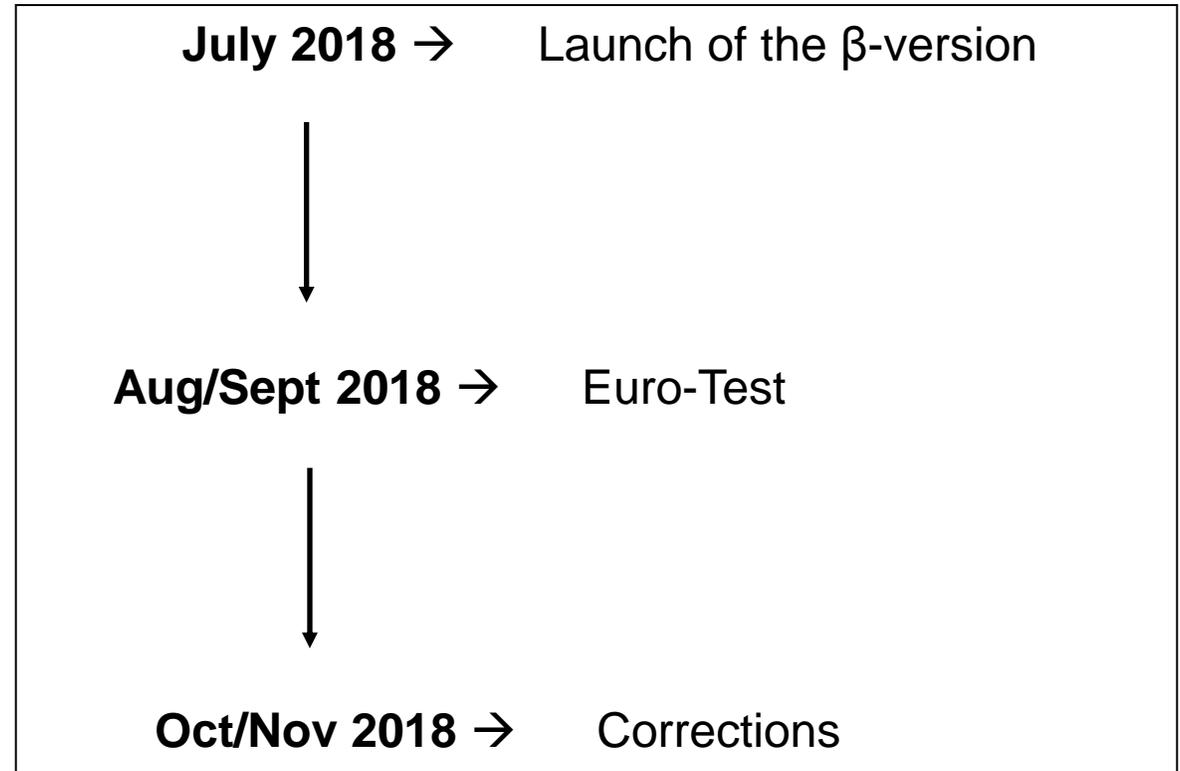
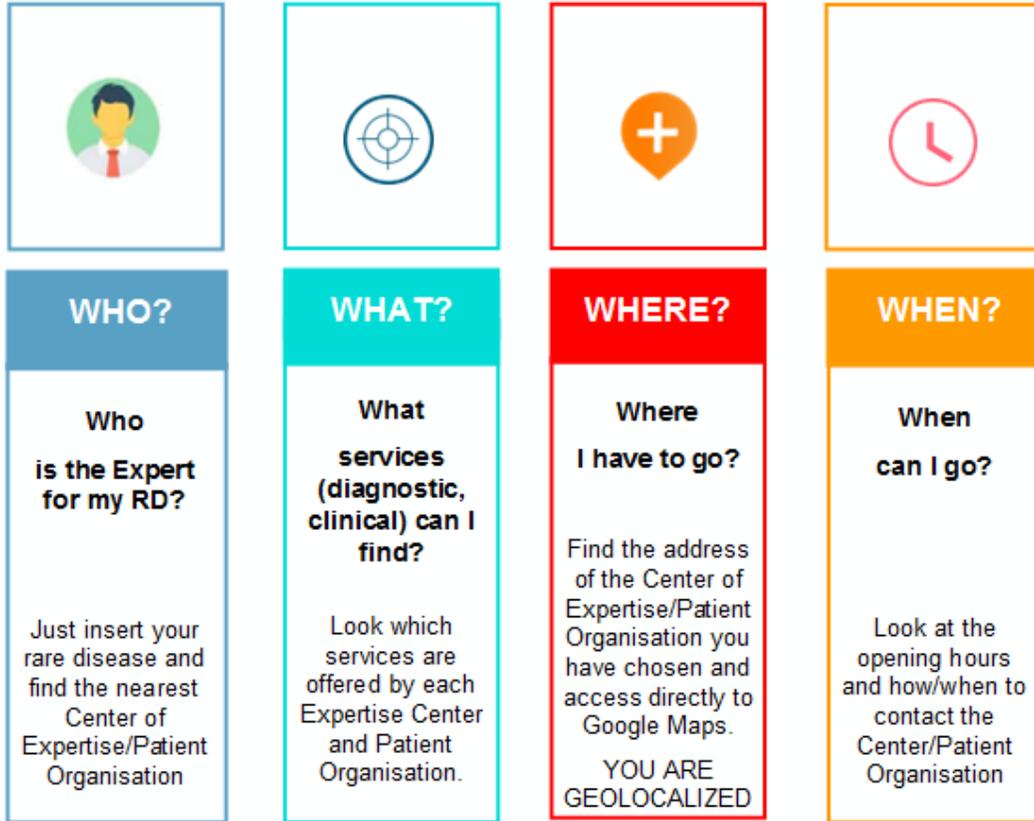


WP3 VASCERN App

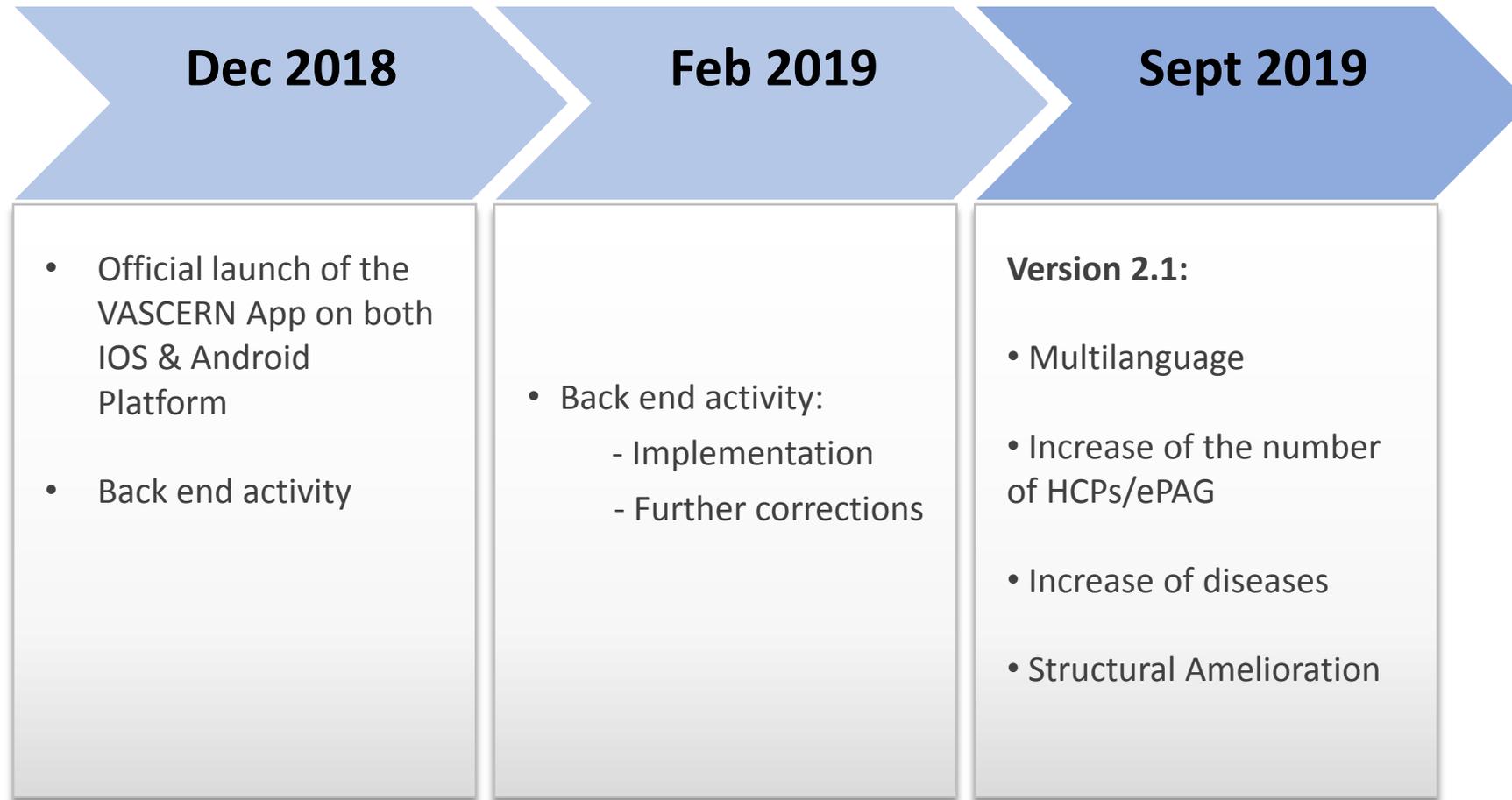
<https://vascern.eu/what-we-do/mobile-app/>

Dr Alessandro Pini
eHealth, Training &
Education WG Chair

WP3 Cross-Border pathway: Mobile Application



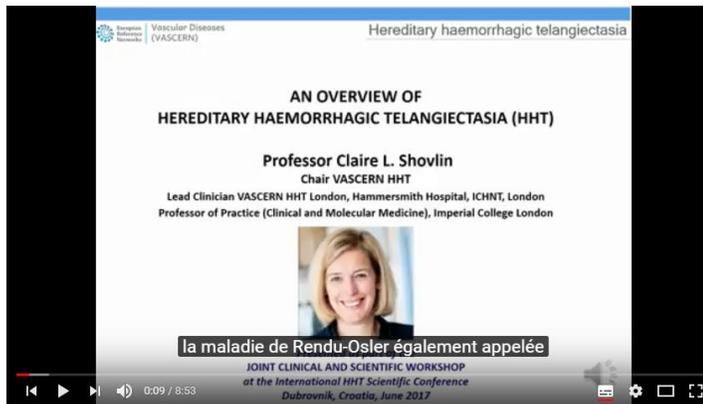
WP3 Cross-Border pathway: Mobile Application



WP4 Pills of Knowledge

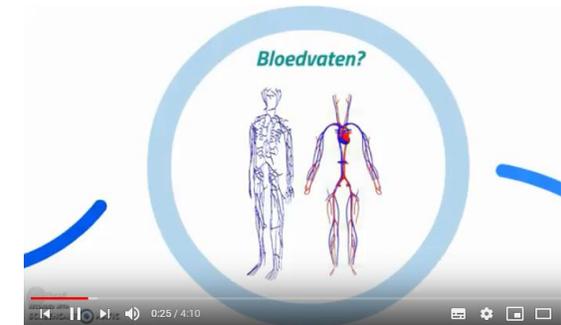
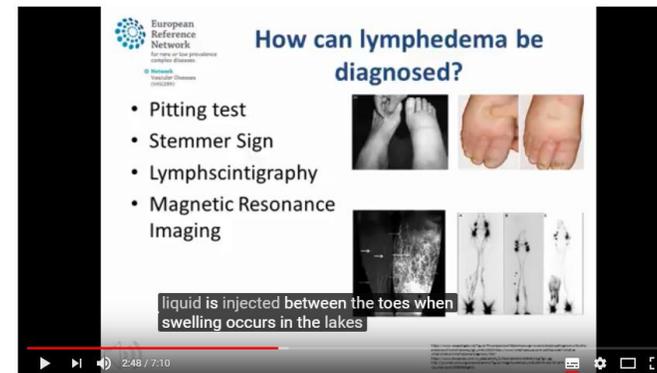
<https://vascern.eu/what-we-do/pills-of-knowledge/>

- 6 PoK video on YouTube, with subtitles added in different languages (ongoing process)
 - Ex: 6 languages + English subtitles added for Julie De Backer's PoK on the diagnosis of Marfan Syndrome
- 13 to be filmed during VASCERN days 2018!



An Overview of Hereditary Haemorrhagic Telangiectasia (HHT) by Prof Claire Shovlin

770 vues



Marfan Syndrome - Diagnosis by Prof Julie De Backer

196 vues



10-11 October 2018



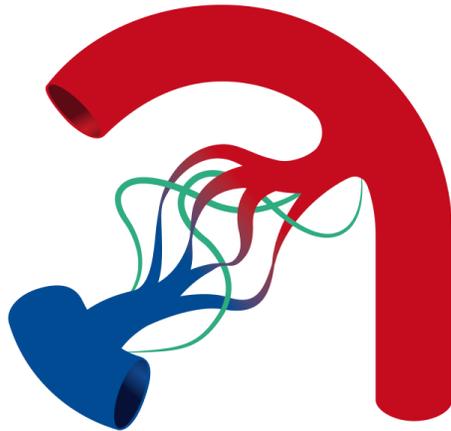
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complex diseases



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Vascular Diseases
(VASCERN)



WP5

Patient Registries

<https://vascern.eu/what-we-do/patient-registries/>

**Prof Leo Schultze
Kool, Patient Registry WG
Chair**



WP6 Clinical trials & Research

- HHT
 - Research project ongoing, multicentric clinical trial proposal
- HTAD
 - F101G
- MSA
 - European vEDS RaDiCo Cohort project ongoing
- PPL
 - Work on collecting research projects
- VASCA
 - Several research projects and clinical trials ongoing



WP7 Availability of videos on YouTube

https://www.youtube.com/channel/UC1sI4_jnqiaLhjNhktiN7ZA



- Currently **75 videos**
- Submit/ share with us if you are filmed
- RDWG Chairs and eHealth WG Chair

• Video collection

- Medical content: Pills of Knowledge + meetings
 - Discussion about content:
 - Transversal WG (e-health + training education)
 - RDWGs validation

• VASCERN related

- Interviews during the year (new scientific publications, patient representatives, etc.)
- Meetings filmed

WP8 Definition of clinical outcomes

<https://vascern.eu/what-we-do/clinical-outcomes-measures/>



- HHT
 - Shovlin, Buscarini et al, European Reference Network for Rare Vascular Diseases (VASCERN) Outcome Measures for Hereditary Haemorrhagic Telangiectasia (HHT)
- HTAD
 - Ongoing
- MSA
 - Ongoing
- PPL
 - Ongoing
- VASCA
 - Ongoing

Shovlin et al. *Orphanet Journal of Rare Diseases* (2018) 13:136
<https://doi.org/10.1186/s13023-018-0850-2>

Orphanet Journal of Rare Diseases

POSITION STATEMENT **Open Access**

 CrossMark

European Reference Network For Rare Vascular Diseases (VASCERN) Outcome Measures For Hereditary Haemorrhagic Telangiectasia (HHT)

Claire L. Shovlin^{1,2*}, Elisabetta Buscarini³, Anette D. Kjeldsen⁴, Hans Jurgen Mages⁵, Carlo Sabba⁶, Freya Droege⁷, Urban Geithoff^{8,11}, Sara Ugolini⁹ and Sophie Dupuis-Grod^{10*}

Abstract

Hereditary haemorrhagic telangiectasia (HHT) is a multisystemic vascular dysplasia that leads to nosebleeds, anaemia due to blood loss, and arteriovenous malformations (AVMs) in organs such as the lungs, liver and brain. HHT is estimated to affect 85,000 European citizens, but most health care providers have limited prior HHT exposure or training. Outcome Measures were developed and implemented by the HHT Working Group of the European Reference Network for Rare Vascular Diseases (VASCERN), in order to maximise the number of patients receiving good care. The measures specifically target areas where optimal management reduces morbidity and mortality in HHT patients, and were designed to be robust to emerging new evidence. Thresholds are the percentage of patients in particular settings who have been recommended screening, or provided with written advice. The 5 Outcome Measures cover (1) pulmonary AVM screening, (2) written nosebleed advice, (3) assessment of iron deficiency; (4) antibiotic prophylaxis prior to dental and surgical procedures for patients with pulmonary AVMs, and (5) written advice on pregnancy. They are not a blueprint for detailed HHT management, but are suitable for all clinicians to be aware of and implement. In summary, these 5 Outcome Measures provide metrics to identify healthcare providers of good care, and encourage care improvement by all healthcare providers.

Keywords: Anaemia, Antibiotic prophylaxis, Epistaxis, Iron deficiency, Nosebleeds, Pulmonary arteriovenous malformations, Pregnancy

Background

Development and implementation of Outcomes Measures are an effective part of Quality and Safety Frameworks that lead to Service Improvements. More specifically, if the Outcome Measures are carefully selected, their dissemination and implementation can directly improve patient care, including that from health care providers with limited prior exposure or training on the specific disease. Therefore simple, clinical practice-based Outcome Measures are particularly important for rare multisystemic conditions.

The current statement refers to one specific rare disease, hereditary haemorrhagic telangiectasia (HHT; Online Mendelian Inheritance in Man® #187300), which is a multi-systemic vascular dysplasia that leads to telangiectasia and arteriovenous malformations (AVMs) in visceral and mucocutaneous vascular beds [1]. Based on a conservative population prevalence of 1 in 6000 [2–4], HHT is estimated to affect approximately 85,000 European citizens. The main goal of management is to maximise the number of affected individuals receiving safe and effective preventative strategies in order to limit the number and severity of HHT complications. The reason this is

* Correspondence: c.shovlin@imperial.ac.uk; elisabetta.buscarini@vascern.eu; sophie.dupuis-grod@chu-lyon.fr
¹Respiratory Medicine, and VASCERN HHT European Reference Centre, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
²Haematology Department, and VASCERN HHT European Reference Centre, Maggiore Hospital, ASST Crema, Crema, Italy
³Trispolino, Culla de Lyon, Hôpital Femme-Mère-Enfant, Service de Génétique, and VASCERN HHT European Reference Centre Centre de Référence pour la maladie de Rendu-Osler, F-69677 Bron, France
Full list of author information is available at the end of the article

 **BMC**

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WP9 Writing Clinical Practice Guidelines (expert consensus statements, recommendations)

<https://vascern.eu/what-we-do/clinical-guidelines/>



- HHT
 - Shovlin, Buscarini et al, European Reference Network for Rare Vascular Diseases (VASCERN) Outcome Measures for Hereditary Haemorrhagic Telangiectasia (HHT)
- HTAD
 - ACTA2 finalised, to be submitted to scientific journal
- MSA
 - Work on pregnancy in vEDS
- PPL
 - Collection of guidelines, new projects?
- VASCA
 - Not prioritised for Y2. Working first on VASCA classification

Shovlin et al. *Orphanet Journal of Rare Diseases* (2018) 13:136
https://doi.org/10.1186/s13023-018-0962-2

Orphanet Journal of Rare Diseases

POSITION STATEMENT Open Access

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¹Department of Vascular Medicine and VASCERN HHT European Reference Network, Copenhagen University Hospital, Rigshospitalet, Copenhagen, Denmark
²Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
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⁶Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
⁷Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
⁸Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
⁹Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
¹⁰Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK
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¹²Department of Vascular Medicine and VASCERN HHT European Reference Network, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK

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VASCERN Consensus Statement: ACTA2-Related Vasculopathy

VASCERN HTAD working group Members: Ingrid van de Leear¹, Eloise Arbustini², Kalman Benke, Erik Bjorck, Maarten Groenink, Marlies Kempers, Yskert von Kodolitsch, Bart Loeys, Barbara Mulder, Lise Murphy, Guglielmina Pepe, Alessandro Pini, Leema Robert, Jolien Roos-Hesselink, Zoltan Szabolcs, Guillaume Jondeau, Julie De Becker

Cooperating Guests: Arturo Evangelista, Roxandra Jurcut, Gisela Teixeira

* Corresponding Author: Dr Ingrid van de Leear, E-mail: ingrid.van.de.leear@erasme.it

VASCERN coordination team: vascern@erasme.it



VASCERN
The European Reference Network on Rare Vascular Diseases
Co-funded by the Health Programme of the European Union

WP10 Do's and Don'ts factsheets for rare vascular disease patients facing frequent situations

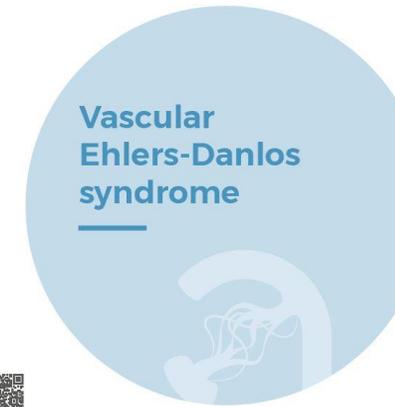


<https://vascern.eu/what-we-do/dos-donts-factsheets-for-rare-vascular-disease-patients/>

- HHT
 - 12 Do's and Don'ts Factsheets published
- HTAD
 - 23 Do's and Don'ts Factsheets published
- MSA
 - 17 Do's and Don'ts Factsheets published
- PPL
 - 13 Do's and Don'ts Factsheets finalised, ready for graphical layout, publication by the end of the year
- VASCA
 - Work in progress



VASCERN DO'S AND DON'TS FACTSHEETS
FOR RARE VASCULAR DISEASE PATIENTS
FACING FREQUENT SITUATIONS



VASCERN DO'S AND DON'TS FACTSHEETS
FOR RARE VASCULAR DISEASE PATIENTS
FACING FREQUENT SITUATIONS



VASCERN DO'S AND DON'TS FACTSHEETS
FOR RARE VASCULAR DISEASE PATIENTS
FACING FREQUENT SITUATIONS



WP11 Communication and project management



ERN COLLABORATIVE PLATFORM [MEMBERS AREA] CLINICAL PATIENT MANAGEMENT SYSTEM [MEMBERS AREA]

ERN Newsletter #22 - September 2018

European Reference Network **VASCERN** (Gathering the best expertise in Europe to provide accessible cross-border healthcare to patients with rare vascular diseases)

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Diseases WGs Transversal WGs ePAG (Patient Advocacy) Newsletters

Page	Total Page Likes	Engage...
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2 Epi CARE Epicare ERN	182	19
3 Ern-Eye	162	15
4 ERN for rare adult solid ...	127	0
5 MetabERN	109	0

Latest News

<http://vascern.eu/>

Newsletter #22 - September 2018

View this email online

Do's and Don'ts Factsheets for HTAD, MSA and HHT now available!

We are very proud to present the VASCERN Do's and Don'ts Factsheets for Rare Vascular Disease Patients Facing Frequent Situations. These factsheets were developed in order to assist patients and the...

Hiring: IT Helpdesk/End-User Support & Data Manager

The VASCERN coordination team, located in Paris, France, is looking for a full-time IT Helpdesk/End-User

This Month:

Followers 387 ↑25

Tweet impressions 16.4K ↑66.7%



<https://twitter.com/vascern>

43 Total d'abonnés

<https://www.linkedin.com/company/vascern/>

Abonnés actuels 49

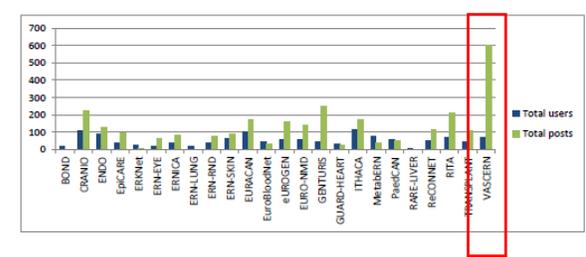
+ 5 au cours des 28 derniers jours

https://www.youtube.com/channel/UC1sI4_inqiaLhJNhktiN7ZA

Monthly virtual meetings & F2F meetings

VASCERN Days 2018

TOTAL USERS REGISTERED/ERN AND TOTAL POSTS/ERN



ERN Collaborative Platform (ECP): internal communication

WP11 Communication and project management

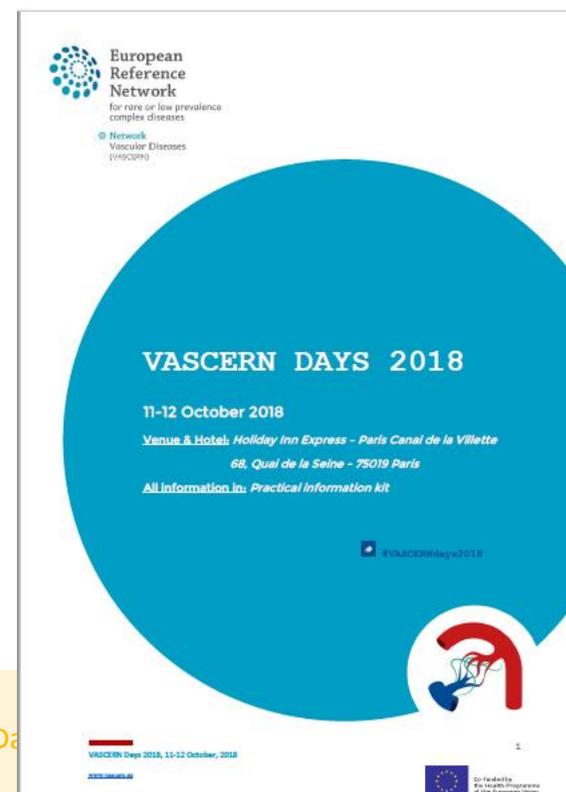
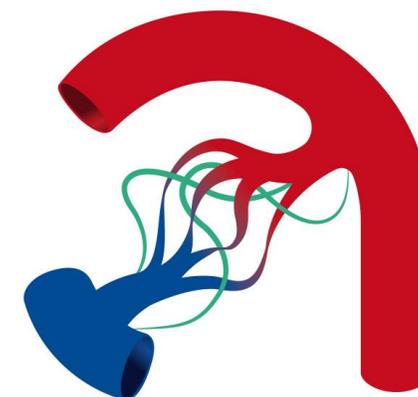


European
Reference
Network

for rare or low prevalence
complex diseases

 **Network**
Vascular Diseases
(VASCERN)

- New specific VASCERN visual identity
 - Visual « vascular » added to the official ERN logo
 - Templates (PPT, word, letter)
 - Graphical chart – additional colours
- Material will be sent to members for their VASCERN presentations



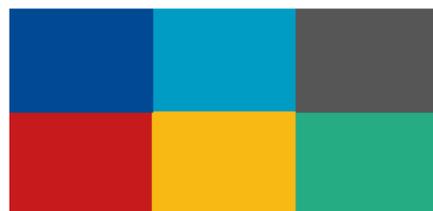
Primary and secondary colours

Primary colours

Ern Colour 1
CMYK: C 100 - M 70 - Y 0 - K 10
RGB: R 5 - G 77 - B 144
WEB: 054C9D

Ern Colour 2
CMYK: C 100 - M 0 - Y 20 - K 0
RGB: R 0 - G 154 - B 191
WEB: 009ABF

Ern Grey
CMYK: C 0 - M 0 - Y 0 - K 80
RGB: R 88 - G 88 - B 88
WEB: 5B5B5B



Secondary colours

Red
CMYK: C 20 - M 100 - Y 100 - K 0
RGB: R 197 - G 26 - B 27
WEB: C5A1B

Orange
CMYK: C 0 - M 30 - Y 95 - K 0
RGB: R 251 - G 186 - B 0
WEB: F8B400

Green
CMYK: C 75 - M 0 - Y 60 - K 0
RGB: R 35 - G 174 - B 132
WEB: 22A084

Main typography

The typography used is **Montserrat**
It can be used on all branded, printed and digital communication media.
<https://fonts.google.com/?query=montserrat&category=Serif&sort=Sort/Display/monospace>



Aa

Light / *Light italic*
Regular / *Italic*
Medium / *Medium Italic*
Bold / *Bold Italic*

Alphabet

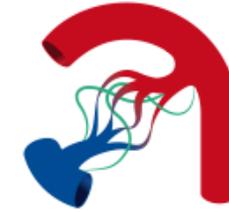
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European
Reference
Network

VASCERN

Gathering the best expertise in Europe
to provide accessible cross-border healthcare
to patients with rare vascular diseases



VASCERN, the European Reference Network on Rare Multisystemic Vascular Diseases, is dedicated to gathering the best expertise in Europe in order to provide accessible cross-border healthcare to patients with rare vascular diseases (an estimated 1.3 million concerned). These include arterial disease (affecting aorta to small arteries), arterio-venous anomalies, venous malformations, and lymphatic diseases.

VASCERN currently consists of 31 highly specialised multidisciplinary Healthcare Providers (HCPs) from 11 EU Member States and of various European Patient Organisations and is coordinated in Paris, France.

Through our 5 Rare Disease Working Groups (RDWGs) as well as several thematic WGs and the ePAG - European Patient Advocacy Group, we aim to improve care, promote best practices and guidelines, reinforce research, empower patients, provide training for healthcare professionals and realise the full potential of European cooperation for specialised healthcare by exploiting the latest innovations in medical science and health technologies.

More information available at: <https://vascern.eu>

Follow us on [Twitter](#), [Facebook](#), [YouTube](#) and [LinkedIn](#)



Co-funded by
the Health Programme
of the European Union