




**European  
Reference  
Network**

for rare or low prevalence  
complex diseases

 **Network**  
Vascular Diseases  
(VASCERN)

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

# Severe/Rare Infantile Hemangiomas (IH)

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# VASCERN

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

VASCERN currently gathers 48 expert teams from 39 highly specialized multidisciplinary HCPs, plus 6 additional Affiliated Partner centers, coming from 19 EU Member States, as well as various European Patient Organisations, and is coordinated in Paris, France.

Through our 6 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: [www.vascern.eu](http://www.vascern.eu)  
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# Table of Contents

## Severe/Rare Infantile Hemangiomas (IH)

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List of abbreviations.....	4	Treatment with propranolol.....	11
Introduction.....	5	General anaesthesia during propranolol.....	12
Ulcerated IH.....	6	After spontaneous regression or propranolol treatment of IH, residual skin changes may persist.....	13
Appearance of crusting on IH.....	7	PHACES syndrome follow up.....	14
Bleeding of an ulcerated IH.....	8	Editorial Board/Contributions.....	15
Large IHs or IH growing on the face.....	9		
Living with a different appearance.....	10		

# Abbreviations

**IH:** Infantile Hemangiomas

**PHACE:** Posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies, and eye anomalies

**LUMBAR:** Lower body hemangioma, Urogenital abnormalities / ulceration, Myelopathy, Bony deformities, Anorectal malformations / arterial anomalies, and Rectal anomalies

**PELVIS:** Perineal hemangioma, external genitalia malformations, lipomyelomeningocele, vesicorenal abnormalities, imperforate anus, and skin tag

**SACRAL:** Spinal dysraphism, anogenital, cutaneous, renal and urologic anomalies, associated with an angioma of lumbosacral localization

**VASCA-WG:** Vascular Anomalies Working Group

# Introduction

This document is directed to parents of children affected with severe and rare infantile hemangiomas (IHs). The aim is to educate them about medical situations where specific care is required and about relevant symptoms and how to act when they occur.

Severe IH may be life-threatening or cause functional impairment based on the localization, severe pain, and/or significant and permanent disfigurement.

When large IHs are located on the face, they may permanently distort anatomic landmarks (nose, lips, ears, philtrum) and cause significant psychological consequences in childhood and adolescence.

Moreover IH may include syndromic variants associated with extracutaneous abnormalities (PHACE and LUMBAR/PELVIS/SACRAL syndromes).

## Severe/Rare Infantile Hemangiomas (IH)

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# Ulcerated IH

Sometimes IH may ulcerate and may bleed or become infected.



### WHAT IS RECOMMENDED

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- Seek medical advice from a doctor as soon as possible (dye laser or propranolol may be indicated).
- Apply an appropriate wound dressing (silicone sheet or polyurethane foam) (change every 1-3 days) to the skin surface in order to maintain the wound moist.
- Wash your hands (can use disposable gloves) in a clean environment at dressing changes to avoid contaminating the wound.
- If located in the diaper area, frequently change the diaper, and apply topical antiseptics (non-alcoholic, non-stinging, non-iodine), vaseline gauzes and zinc oxide.



### WHAT YOU SHOULD NOT DO

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- Do not leave the wound directly open to the air.
- Do not apply dressings that would stick to the bottom of the wound e.g. cotton gauze.
- Do not apply local propranolol or timolol on ulceration.

# Appearance of crusting on IH

The first manifestation of ulceration is the appearance of crusts on the IH.



### WHAT IS RECOMMENDED

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- Apply an oily ointment on crusted lesions.
- Apply an antiseptic ointment if there is purulent discharge.
- Ask the doctor to consult a hemangioma specialist to consider systemic antibiotics if deeper infection is suspected.



### WHAT YOU SHOULD NOT DO

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- Do not forcibly remove the crusts to avoid bleeding.

# Bleeding of an ulcerated IH

Ulcerated hemangiomas may bleed. Bleeding is rarely severe.



### WHAT IS RECOMMENDED

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- Stay calm, most of the bleeds are mild with just a few drops, without consequences.
- To stop the bleeding, apply pressure with gauze or clean towel /disposable tissue for 10-15 minutes.
- Remove the dressing only if bloodstains appear on the dressing.
- In case of recurrent and significant bleeding, alert the doctor.
- Attend an emergency department if the bleeding does not stop.



### WHAT YOU SHOULD NOT DO

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- Do not remove the dressing to check for cessation of bleeding.
- Do not apply the bandage too tight to impair blood circulation.



# Large IHs or IH growing on the face

Most of hemangiomas on the face should be treated with propranolol. Large IH sometimes may be associated with other abnormalities.



### WHAT IS RECOMMENDED

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- If your baby has a hemangioma on the face, or a large hemangioma, ask to see a hemangioma specialist.
- Consider checking the hemangioma referral score (IHReS: <https://www.ihscoring.com/>).



### WHAT YOU SHOULD NOT DO

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- Do not wait for a late referral, since starting propranolol early - if indicated - is associated with better outcomes.

# Living with a different appearance

Living with a different appearance due to large IHs in visible areas or sequelae of IHs, may make socializing difficult. These difficulties sometimes reflect on the whole family and can cause psychological problems for both the child and the parents.



### WHAT IS RECOMMENDED

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- Reach out to other families with a similar condition.
- Ask the doctor or search for a patient association.
  - can help you with tips and tricks from other parents/caregivers
- Ask for psychological and psychosocial support in challenging situations or signs of depression.
- You may contact the European Patients Advocacy Groups (ePAG) of [VASCERN](#) or [ERN SKIN](#)



### WHAT YOU SHOULD NOT DO

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- Do not isolate yourself and/or your child.

# Treatment with propranolol

Propranolol may lower blood sugar and cause wheezing in small children.



## WHAT IS RECOMMENDED

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- Give the medication with feeds.
- Pause medication in case of poor appetite, poor nutrition, high fever, diarrhoea, or vomiting.
- Pause medication and contact your doctor in case of cough, wheezing or difficulties in breathing.
- Further educational videos may be found on [hemangiomaeducation.org](http://hemangiomaeducation.org).



## WHAT YOU SHOULD NOT DO

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- Do not give another dose if the child regurgitates or vomits the medication straight after eating.
- Do not use a syringe different from the one supplied by the manufacturer.

# General anesthesia during propranolol

Propranolol reduces body reactions to stress, response to certain drugs and lower blood sugar levels.



### WHAT IS RECOMMENDED

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- Pause propranolol treatment for IH during fasting before general anaesthesia.
- Inform the anesthesiologist about the propranolol medication and when the child has received the previous dose.



### WHAT YOU SHOULD NOT DO

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- Do not give propranolol while the child is fasting.

## Severe/Rare Infantile Hemangiomas (IH)

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After spontaneous regression or propranolol treatment of IH, residual skin changes may persist



### WHAT IS RECOMMENDED

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- Ask for evaluation with a multidisciplinary team for treatment options before school age.



### WHAT YOU SHOULD NOT DO

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- Do not delay asking for a consultation before school age.

# PHACES syndrome follow up

PHACES syndrome may be associated with rare problems apparently not related with the IH. It is important to alert the doctor if specific symptoms appear.



## WHAT IS RECOMMENDED

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- Alert the doctor if you suspect
  - Hearing loss and speech-language delay
  - Visual impairment
  - Swallowing disorders
  - Growth abnormalities
  - Headache
  - Dental anomalies
  - Psychological or psychosocial impact of the disease

# Editorial Board/ Contributors

This document resulted from the collaboration of the VASCA-working group (VASCERN) and the Cutaneous Mosaic Disorders thematic group (ERN SKIN).

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