

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

Severe/Rare Infantile
Hemangiomas (IH)

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).

Ulcerated IH

Sometimes IH may ulcerate and may bleed or become infected

- Recommendations

- Contact the doctor as soon as possible (dye laser or propranolol may be indicated)
- Apply a silicone sheet or polyurethane foam to the skin surface in order to maintain the wound moist
- Use disposable gloves and sterile garments at dressing changes to avoid contaminating the wound
- Apply topical antiseptics, vaseline gauzes and zinc oxide in the diaper area

- What you should not do

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

Appearance of crusts on IH

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

- Recommendations
 - 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
 - To avoid bleeding, do not forcibly remove the crusts

Bleeding of an ulcerated IH

Ulcerated hemangiomas may bleed, causing anemia. Sometimes bleeding may be severe

- Recommendations

- Apply a compressive dressing using a thick layer of gauze secured with an elastic outer layer which is comfortable for the patient
- Remove the dressing only if bloodstains appear on the dressing
- In case of recurrent and significant bleeding, alert the doctor in order to check for anemia
- Attend an emergency department if the bleeding does not stop

- What you should not do

- Avoid removing the dressing to check for cessation of bleeding
- The bandage should not be so tight that it restricts blood flow to a limb

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

- Recommendations

- If your baby has an hemangioma on the face, or a large hemangioma, ask the general practitioner for an hemangioma specialist

- What you should not do

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

Very large IH

Very large IH may cause problems in the development of the child and functional impairment depending on the location of the hemangioma

- What to do?
 - Ask for a referral to a hemangioma specialist?
 - Alert the doctor in case of:
 - Development delay
 - Growth retardation
 - Functional impairment (physiotherapy may be an option)

Living with a different appearance

Living with a different appearance due to large IHs in visible areas or sequelae of IHs, may be difficult for the child that needs to interact with other children to be part of a group. These difficulties sometimes reflect on the whole family and can cause psychological problems for both the child and the parents

- What to do?
 - Share your experience with other parents with a similar condition
 - Ask the doctor or search for a patient association
 - can help them in tips and tricks from other parents/caregivers
 - important in dealing with having a baby with a different appearance in a difficult environment
 - In case of signs of depression because of constant negative/hurtful comments from the environment ask for a psychological and psychosocial support
 - You may contact the European Patients Advocacy Groups (ePAG) of [VASCERN](#) or [ERN SKIN](#)
- What you should not do
 - Do not isolate yourself due to feelings of shame or frustration

Treatment with propranolol

Propranolol carries a risk of hypoglycemia in small children

- What to do?
 - Give the medication after feeding or immediately before feeding (if the child feeds regularly and without difficulties)
 - Pause the medication if the child has a gastrointestinal upset or is not feeling well
 - Further educational videos may be found on hemangiomaeducation.org
- What you should not do?
 - Do not give propranolol in cases of poor appetite, poor nutrition, cough, high fever, diarrhoea, vomiting
 - Do not give another dose if the child regurgitates or vomits the medication straight after eating

General anesthesia during propranolol

Propranolol reduces adrenergic reactions, response to epinephrine and worsens hypoglycemia

- Recommendations

- Ask the hemangioma specialist whether to pause giving propranolol before general anaesthesia

- What you should not do

- Do not fast during propranolol treatment

Occurrence of coughing or bronchospasm during respiratory infection and propranolol treatment

Propranolol can induce bronchospasm in predisposed child in particular during respiratory infections

- What is recommended
 - Interrupt propranolol
 - Inform the doctor immediately
 - Restart propranolol only after authorisation by the doctor once the bronchospasm and infection has been resolved
- What you should not do
 - If bronchospasm occurred in the absence of respiratory infection, do not restart propranolol

Sequelae of IH / residual IH after propranolol or spontaneous regression

After propranolol treatment or after spontaneous regression some sequelae may last and potentially cause psychological problems

- What is recommended
 - Ask for evaluation with a multidisciplinary approach (dermatologist, plastic surgeon)
 - Treat abnormalities with potential psychological impact before school age
- What you should not do
 - Do not ask for treatment of minor sequelae without potential psychological impact before school age in order to avoid unnecessary general anaesthesia/sedation

PHACES syndrome follow up

Phace syndrome may be associated with several problems apparently not related with the IH. It is important to alert the doctor if specific symptoms appears

- What to do?
 - Alert the doctor if you suspect
 - Hearing loss and speech-language delay
 - Swallowing disorders (dysphagia)
 - Growth abnormalities
 - Headache
 - Dental anomalies
 - Psychological or psychosocial impact of the disease