

Supplementary Online Content

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Appendix. Hemangioma Severity Scale and Hemangioma Dynamic Complication Scale: instruction and training manual.

This supplementary material has been provided by the authors to give readers additional information about their work.

Appendix

Hemangioma Severity Scale and Hemangioma Dynamic Complication Scale Instructions and Training Manual



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Introduction

The **Hemangioma Severity Scale (HSS)** is a research tool developed to measure the clinical severity of an infantile hemangioma. The content of the scale and the numerical weight assigned to specific variables is based on medical literature regarding the morbidities associated with infantile hemangioma and the collective clinical experience of physicians with expertise in infantile hemangiomas.

An adjunctive **Dynamic Complication Scale (DCS)** is incorporated within the HSS and assigns grades of clinical severity to any hemangioma-related complications. The DCS grades will not be calculated into the overall HSS score, but will be used as a tool for measuring the clinical change in *hemangioma-related* complications.

Instructions

Prior to using the HSS or DCS, this entire manual should be carefully reviewed. The manual can serve as a reference for clarification when needed.

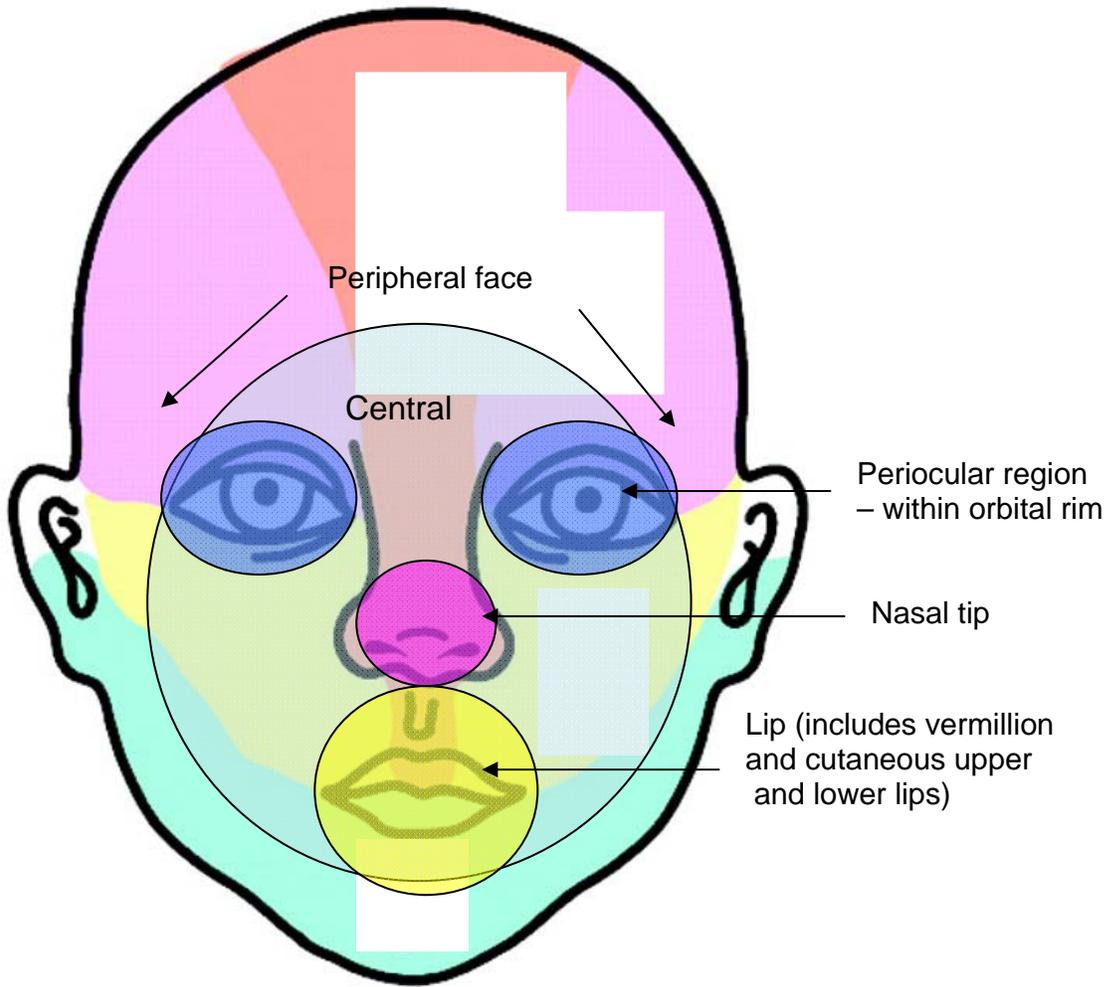
Clinical information regarding the child's clinical history and physical exam are required to complete the scale. Specifically, the **size, location, risk for underlying structural anomalies (PHACE/PELVIS/SACRAL/LUMBAR syndromes), complications, pain and risk of disfigurement** will be recorded. Each of these variables is discussed in detail below.

1. Size Scoring: The size of the hemangioma refers to longest dimension as measured in centimeters (cm). The numerical point value of each size is specified on the scale and **differs for facial/ear and nonfacial/scalp/neck locations**. For segmental lesions, the measurement should be of the entire segmental hemangioma, even if the segmental hemangioma is composed of papules and plaques that are noncontiguous (have skip areas). *Record only one single value for this category.*

2. Location Scoring: The location refers to where the hemangioma is found on the body. If a patient has a hemangioma encompassing more than one area, the location receiving the highest point value should be recorded. For example, a large or multisegment hemangioma on the face encompassing the periorbital, temporal and lateral cheek should receive a point value of 6 designated for periorbital because this represents the location of highest point value involved. Another example is, if there is a hemangioma that spans both the central face and the peripheral face, the point value of 4 for central face should be assigned. The diagram on page 4 should be used for reference.

The nasal tip, lip and periocular areas are shown on the figure. Note the lip includes the vermillion and cutaneous upper and lower lips. The periocular area is within the orbital rim.

Record only one single value for this category.



3. Risk of Associated Structural Anomalies Scoring: Those hemangiomas that fulfill clinical criteria that prompt investigation for internal structural anomalies are scored in this section. These criteria include: facial hemangiomas that are 5 cm or greater in the longest diameter, lumbosacral midline hemangiomas that are at least 2.5 cm in diameter, and segmental perineal/perianal/genital hemangiomas. *Record only one single value for this category.*

4. Complication Scoring:

- All complications **must be hemangioma-related** and NOT due to associated structural anomalies. For example, if a child suffers a cardiac complication secondary to a coexisting coarctation in the setting of PHACE, this would not be considered directly related to the hemangioma itself.
- For all the variables, the presence or absence of the variable determines whether or not points are assigned.
- **For any complications that are present, complete the Dynamic Complication Scale (DCS) for that complication. Under each complication below, there are specific guidelines that will be helpful when scoring.**

Infection is defined as the clinical presence of bacterial infection (erythema, exudate, impetigization manifested by honey-colored crusting and scale). Bacterial culture for confirmation is not required.

Ulceration is defined as a disruption in the epidermis and/or dermis. For the HSS, the presence of any size and depth of ulceration will allow points to be scored for ulceration. For the DCS, the depth of the ulcer, impact on daily living, treatment required and presence of vital structure loss are important.

Feeding difficulties include difficulty sucking/latching, swallowing, or chewing secondary to the direct presence of the hemangioma. Additionally, bleeding during feeding secondary to mechanical trauma is also considered a feeding difficulty.

Note that grade 2 of the DCS includes weight loss OR lack of adequate weight gain – because infants normally are gaining weight quite rapidly, even a lack of adequate weight gain is significant.

Torticollis is a cervical dystonia due to the presence of the hemangioma manifested by either abnormal posturing of the head/neck or spastic movements of the head and neck related to the presence of the hemangioma. For the DCS,

there is a distinction made between intermittent and persistent abnormal head posture.

Cartilage may be damaged by hemangiomas on the nose or ear. If the cartilage is anyway distorted or disfigured, HSS points should be assigned. Please note that nasal tip hemangiomas with a deep component are assumed to distort the cartilage. (see figure) For the DCS, the presence of distortion, focal destruction and major destruction determine which grade is assigned. For those lesions where surgical correction is likely, it should be considered major destruction.



Figure. Superficial and Deep Nasal Tip Hemangioma. The deep component will cause distortion of the nasal cartilage.

Airway compromise refers to hemangioma in the airway including the epiglottic, glottic and subglottic regions. To be assigned 3 points, the airway hemangioma must be narrowing the airway (any % obstruction will qualify for the 3 points). Symptoms such as stridor or hoarse cry imply narrowing of the airway and would indicate there is airway compromise. For the DCS, the presence of symptoms, respiratory distress, and treatment required are important variables for assigning grades.

Visual compromise refers to any detrimental effect of the hemangioma on vision including astigmatism, amblyopia, and visual axis occlusion as found by an ophthalmologist. For the DCS, eyelid distortion should be assumed if there is a deep component present on the affected eyelid.

Hypothyroidism refers to an abnormality of thyroid function tests with or without symptoms of hypothyroidism. The hypothyroidism must be a direct result of hemangioma and not associated pituitary anomalies, structural or otherwise. For the DCS, the treatment required is important for assigning grades.

Anemia secondary to bleeding from a hemangioma (cutaneous or visceral) is characterized by decreased hemoglobin compared to age-matched normal values. For the DCS, the presence of symptomatic anemia and the treatment required are important distinguishing features.

Congestive heart failure as confirmed by a cardiologist suggesting decreased cardiac function directly related to the hemangioma. Congestive heart failure may manifest as peripheral edema, abdominal distention, tachycardia, and fatigue. For the DCS, the presence of symptoms, response to intervention and treatment used are important considerations in grading.

Gastrointestinal bleed (GI bleed) refers to hemangiomas (or a single hemangioma) in the GI tract that are complicated by bleeding. For the DCS, the presence of symptoms and treatment used are important considerations in grading.

Hepatic dysfunction refers to visceral hemangiomas in the liver causing alteration in liver function tests, synthetic dysfunction and/or vascular flow abnormalities in the liver. For the DCS, the presence of hepatomegaly with or without hepatic dysfunction are important.

5. Pain Scoring: Pain will be scored according to severity and severity is implied by the level of treatment the child requires to control the pain. Children having intermittent pain not requiring any pain medications will receive a score of 1, with progressively higher scores assigned as the level of pain medicine advances from over-the-counter to prescription to inpatient pain control.

Record only one single value for this category.

6. Risk of Disfigurement Scoring: The risk of disfigurement on the HSS is divided by location into facial/ear and nonfacial/scalp/neck. *Record only one single value for risk of disfigurement.*

- The risk of disfigurement is related to the hemangioma subtype, location, and extent of proliferation.
- Hemangiomas that are barely elevated with a gentle slope noted at the edges are likely to leave only **minimal residual telangiectasia or textural change**. (See Figure)
- Hemangiomas that are more exophytic with a steeper slope and more prominent superficial component are more likely to leave **significant fibrofatty tissue and a resultant scar**. Hemangiomas that are exophytic and likely to leave fatty residua but that lie on a planar surface such as the cheek should not be considered to distort anatomic landmarks.
- All hemangiomas on the scalp should be expected to produce a **relative alopecia that should be considered a scar**.
- Hemangioma that, even after involution, are likely to distort anatomic boundaries are those that have ulcerated in cosmetically sensitive areas such as the lip or those lesions that will **leave residual distortion of anatomic landmarks** such as those that affect the nasal tip or ear cartilage. (See Figure). Some examples of distortion of anatomic landmarks include: philtrum distortion, nasal crease or nasolabial fold distortion, contour of nasal tip, or permanent eyelid distortion resulting in asymmetry. Non-facial examples include distortion of breast anatomy.
- Hemangiomas that are **deep and involve the nasal tip should be assumed to be associated with permanent deformity** of normal anatomic landmarks.

Record only one single value for this category.