




Clinics in Dermatology

Volume 33, Issue 2, March–April 2015, Pages 170-182

Hemangiomas and the eye ☆

Allyson A. Spence-Shishido MD^a, William V. Good MD^b, Eulalia Baselga MD^c, Ilona J. Frieden MD^a  [Show more](#) ▾ Share  Cite<https://doi.org/10.1016/j.clindermatol.2014.10.009> ↗[Get rights and content](#) ↗

Abstract

Infantile hemangiomas are a common vascular birthmark with heterogeneous presentations and unique growth characteristics with early rapid growth and eventual self-involution. Hemangiomas that develop around the eye have the potential for inducing amblyopia by several mechanisms and may eventually result in permanent visual impairment in otherwise healthy infants. Segmental periocular hemangiomas carry the additional risk of associated structural anomalies and PHACE syndrome. In recent years, the treatment of periocular hemangiomas has been revolutionized by the serendipitous discovery of the effectiveness of beta-blockers (systemic and topical), with most experts viewing these as first-line therapies. The management of periocular hemangiomas should involve a close partnership between an ophthalmologist and dermatologist or other relevant specialists familiar with the unique clinical features, differential diagnosis, treatment approaches, and potential complications.

Introduction

Infantile hemangiomas (IH) are a common birthmark occurring in 4-5% of all infants, with female gender, prematurity, Caucasian race, and multiple gestation pregnancies as risk factors.[1], [2], [3] Hemangiomas of the head and neck are common,[4], [5] as is periocular involvement, but the exact incidence is unknown. One retrospective review found that 24.3% of all focal facial hemangiomas involved periocular sites.⁶ Additional data from the Hemangioma Investigator Group of 1096 consecutively enrolled patients with ≥1 hemangioma at any site,³ found that 12% had a periocular IH (E. Baselga, MD, personal written communication, October 2013). In contrast, a population-based cohort study estimated that periocular infantile hemangiomas occur in only 1 in 1586 live births, though the authors acknowledge that this may be an underestimate due to the retrospective design and possible incomplete data collection.⁷ Whatever the true incidence, the periorbital area is a relatively frequent site for IH and a particularly important one, because it can lead to permanent visual loss or distortion of anatomic landmarks in the area. Of the 1096 patients followed by the Hemangioma Investigator Group, 41% suffered some form of visual compromise (E. Baselga, MD, personal written communication, October 2013).

The timing of appearance of IH and the timing of its proliferative phase coincides with a critical time in the development of the visual axis, which includes integration of signals received by the retina, and processing the images in the central visual system.⁸ Abnormal visual development can result in abnormal vision at the level of the central nervous system, which cannot be later corrected as easily by simple intervention, such as the addition of glasses.⁹ The critical period for the development of the visual axis in humans is thought to be between birth and 9 years of age.[8], [9] Observation of monocular deprivation in humans suggests that younger age and longer duration of deprivation have more significant effect on vision⁸; however, studies done in kittens in 1970 revealed that even very brief 3-4 day periods of unilateral eye closure during the first few months of life result in irreversible changes in the visual axis.¹⁰ Similarly, early studies in humans showed that even after involution of a periocular hemangioma, associated refractive errors did not always resolve, suggesting permanent effects on the visual axis.¹¹ Given the potential for permanent visual impairment, physicians managing IH need to recognize worrisome clinical features, and be aware of when, and to whom, the patient should be referred.

This contribution reviews highlights of pathogenesis, clinical features, potential complications, differential diagnosis, and management options for periocular hemangiomas.

Section snippets

Pathogenesis

The pathogenesis of infantile hemangiomas is still incompletely understood. Several excellent recent review contributions have discussed recent advances in our understanding of hemangioma pathogenesis.[12], [13], [14] Numerous diverse hypotheses exist, including theories of placental origin,[15], [16] somatic gene mutation,[17], [18] hypoxia-driven events,¹⁹ and aberrant stem cells.[20], [21]

In 2000, it was shown that endothelial cells within infantile hemangiomas are glucose transporter 1...

Growth characteristics

IH have a characteristic and well-documented natural history: up to 65% of infants with superficial IH have a precursor sign at birth²⁸ (telangiectatic patches with a pale halo, erythematous patches, pale patches, bruise-like macules) followed by rapid proliferation, then slow involution.[2], [29] Hemangiomas grow most rapidly in the first 3.2 months of life, reaching an average of 80% of their final size during this time.³⁰ During these first 3 months of life, IH growth is most rapid between...

Differential diagnosis

The characteristic appearance and growth characteristics (with early rapid proliferation and later slow involution) help to make the clinical diagnosis of infantile hemangioma; therefore, the diagnosis may be made, or refined, over the course of several clinic visits. The differential diagnosis of a periocular hemangioma includes other vascular tumors and malformations. In early superficial hemangiomas, especially before proliferation occurs, one should consider a capillary malformation, such...

Management

Because IH resolve spontaneously, treatment is not needed in all patients; however, with periocular involvement, active treatment should be considered in all cases. Treatment decisions depend on many factors, including location, size, findings on eye exam (proptosis, strabismus, globe displacement, lid margin change, visual axis obstruction), patient age, perceived growth potential, and parental preference. Figure 12 proposes an algorithm for management.

If a patient is very young (<4 weeks old) ...

Conclusions

Periocular IH are very heterogeneous, with varied morphologies and locations, both within the skin and within the orbit. They also display unique growth characteristics, including rapid early growth. Given the potential for complications, including vision loss, dermatologists, ophthalmologists, pediatricians, and other relevant specialists should have a working knowledge of IH and an approach to management. Open communication, and a multidisciplinary approach to the exam and management are in...

[Special issue articles](#) [Recommended articles](#)

References (109)

K.N. Kanada *et al.*

[A prospective study of cutaneous findings in newborns in the united states: Correlation with race, ethnicity, and gestational status using updated classification and nomenclature](#)

J Pediatr (2012)

R.M. Robb

[Refractive errors associated with hemangiomas of the eyelids and orbit in infancy](#)

Am J Ophthalmol (1977)

Y. Yu *et al.*

[Endothelial progenitor cells in infantile hemangioma](#)

Blood (2004)

P.E. North *et al.*

[GLUT1: A newly discovered immunohistochemical marker for juvenile hemangiomas](#)

Hum Pathol (2000)

W.A. Ahrens *et al.*

[GLUT-1 expression in mesenchymal tumors: An immunohistochemical study of 247 soft tissue and bone neoplasms](#)

Hum Pathol (2008)

K. Tambe *et al.*

[Relationship of infantile periocular hemangioma depth to growth and regression pattern](#)

J AAPOS (2009)

J. Dubois *et al.*

Orbit and eyelid hemangiomas: Is there a relationship between location and ocular problems?

J Am Acad Dermatol (2006)

J.A. Shields *et al.*

Vascular tumors of the conjunctiva in 140 cases

Ophthalmology (2011)

B.G. Haik *et al.*

Capillary hemangioma (infantile periocular hemangioma)

Surv Ophthalmol (1994)

R.L. Garcia *et al.*

Occlusion amblyopia secondary to a mixed capillary-cavernous hemangioma

J Am Acad Dermatol (1984)

 View more references

Cited by (20)

Vascular Anomalies and Other Cutaneous Congenital Defects

2023, Avery's Diseases of the Newborn

Periocular infantile hemangioma masquerading as dacryoceles

2020, Journal of AAPOS

Citation Excerpt :

...Periocular hemangiomas are frequently associated with a variety of ophthalmic sequelae, including amblyopia due to induced astigmatism and/or visual axis obstruction due to ptosis, nasolacrimal duct obstruction, mass effect leading to compression of orbital structures, and ulceration.^{5,6} Infantile hemangiomas tend to grow most rapidly in the first 3 months of life, followed by involution, which is complete by approximately age 4 years, leaving behind some degree of residual hemangioma.⁵ Systemic or topical beta blockers are used in the treatment of periocular hemangiomas.^{5-7...}

Show abstract 

Conjunctival hemangioma: From atypical presentation to topical beta-blocker treatment

2018, Journal Francais d'Ophtalmologie

Cutaneous Congenital Defects

2018, Avery's Diseases of the Newborn: Tenth Edition

Show abstract 

Cutaneous Congenital Defects

2017, Avery's Diseases of the Newborn, Tenth Edition

Show abstract 

Palpebral malformations in children

2016, Annales de Chirurgie Plastique Esthetique

[Show abstract](#) 

[View all citing articles on Scopus](#) 

- ☆ Drs. Frieden and Baselga disclose that they are consultants for Pierre Fabre. Dr. Baselga is also a principle investigator in the HEMANGIOL study, which was sponsored by Pierre Fabre.

[View full text](#)

Copyright © 2015 Elsevier Inc. All rights reserved.



All content on this site: Copyright © 2024 Elsevier B.V., its licensors, and contributors. All rights are reserved, including those for text and data mining, AI training, and similar technologies. For all open access content, the Creative Commons licensing terms apply.

