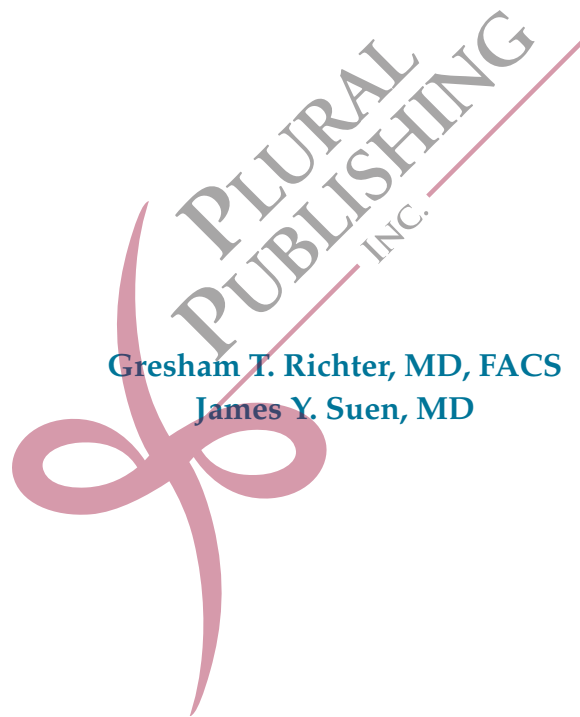


Head and Neck Vascular Anomalies

A Practical Case-Based Approach



Gresham T. Richter, MD, FACS
James Y. Suen, MD



Contents

<i>Preface</i>	<i>ix</i>
<i>Introduction to Vascular Anomalies by Lauren A. Kilpatrick, MD</i>	<i>xi</i>
<i>Acknowledgments</i>	<i>xvi</i>
<i>Contributors</i>	<i>xvii</i>
Chapter 1. Infantile Hemangiomas	1
Basic Tenants and Interventions	1
<i>Gresham T. Richter</i>	
Case Study 1–1. Anterior Neck Hemangioma	5
<i>Abby R. Nolder</i>	
Case Study 1–2. Infantile Hemangioma of the Eyelid	12
<i>David H. Darrow and Joel K. Lall-Trail</i>	
Case Study 1–3. Orbital Infantile Hemangioma	21
<i>Aaron Fay, Peter W. MacIntosh, and Milton Waner</i>	
Case Study 1–4. Glabella Hemangioma	27
<i>Gresham T. Richter and Venkata S. P. B. Durvasula</i>	
Case Study 1–5. Upper Lip Hemangioma	33
<i>Larry D. Hartzell</i>	
Case Study 1–6. Lower Lip Hemangioma	41
<i>Larry D. Hartzell</i>	
Case Study 1–7. Complex Facial Hemangioma	47
<i>Melanie Duval and J. Fredrik Grimmer</i>	
Case Study 1–8. Ulcerated Hemangiomas	52
<i>M. Taylor Fordham and Nancy M. Bauman</i>	
Case Study 1–9. Hemangioma of the Posterior Neck	59
<i>Lorelei Grunwaldt and Deepak Mehta</i>	
Case Study 1–10. Infantile Hemangioma of the Scalp	64
<i>Adnan Mir and Brandi Kenner-Bell</i>	
Case Study 1–11. Infantile Hemangioma of the Nasal Tip	69
<i>Marcelo Hochman</i>	
Case Study 1–12. Paranasal Hemangioma	76
<i>Robert H. Chun and Kristen E. Holland</i>	
Case Study 1–13. Segmental Hemangiomas	80
<i>Francine Blei</i>	
Case Study 1–14. Multifocal Infantile Hemangiomas “Hemangiomatosis”	86
<i>Denise M. Adams</i>	
Case Study 1–15. Parotid Infantile Hemangioma	90
<i>Teresa M. O and Milton Waner</i>	
Case Study 1–16. Subglottic Hemangioma	96
<i>Ian Jacobs</i>	

Case Study 1–17. Circular Excision and Purse-String Suture for Infantile Hemangiomas <i>Dov C. Goldenberg, Patricia Y. Hiraki, and Raphael Manzini</i>	101
Case Study 1–18. Cheek Hemangiomas <i>Jessica L. Hootnick, Stephen R. Hoff, Julia F. Corcoran, and Jeffrey C. Rastatter</i>	106
Chapter 2. Other Vascular Tumors	111
Case Study 2–1. Rapidly Involuting Congenital Hemangioma and Noninvoluting Congenital Hemangioma (RICH and NICH) <i>Ravindhra G. Elluru, Kashif Mazhar, and Manish N. Patel</i>	111
Case Study 2–2. Pyogenic Granuloma <i>Tara L. Rosenberg</i>	116
Case Study 2–3. Kaposiform Hemangioendothelioma <i>Lauren A. Kilpatrick</i>	119
Chapter 3. Capillary Malformations	127
Basic Tenants and Interventions <i>Rachel A. Giese and Gresham T. Richter</i>	127
Case Study 3–1. Nevus Simplex: Medial Fronto-Facial Capillary Malformations <i>Venkata S. P. B. Durvasula and Gresham T. Richter</i>	129
Case Study 3–2. Port-Wine Stains in Association With Underlying Syndromes <i>Arisa E. Ortiz and J. Stuart Nelson</i>	133
Chapter 4. Venous Malformations	141
Basic Tenants and Interventions <i>Fang Hou</i>	141
Case Study 4–1. Venous Malformation of the Larynx <i>Gresham T. Richter</i>	144
Case Study 4–2. Venous Malformation of the Scalp <i>Kashif Mazhar, Manish N. Patel, and Ravindhra G. Elluru</i>	150
Case Study 4–3. Buccal Space Venous Malformation <i>Amir Pezeshkmehr and Leah Braswell</i>	154
Case Study 4–4. Masseteric Venous Malformations <i>Amir Pezeshkmehr and Leah Braswell</i>	160
Case Study 4–5. Upper Lip Focal Venous Malformation <i>Patrick D. Munson</i>	166
Case Study 4–6. Venous Malformation of the Head and Neck <i>Yan An Wang, Jia Wei Zheng, Han Guang Zhu, and Zhi Yuan Zhang</i>	171
Case Study 4–7. Venous Malformation of Cheek and Upper Lip <i>Steven L. Goudy and Joshua R. Mitchell</i>	178
Case Study 4–8. Venous Malformation of the Oropharynx-Hypopharynx <i>James Y. Suen</i>	186
Case Study 4–9. Extensive Venous Malformation of Face and Neck <i>Behfar Eivazi</i>	192

Case Study 4–10. Orbital Venous Malformation <i>Ming Lin, Jia Wei Zheng, and Xianqun Fan</i>	198
Case Study 4–11. Cheek Venous Malformation <i>Teresa M. O and Milton Waner</i>	204
Case Study 4–12. Venous Malformation of the Temporal Bone <i>Jumin Sunde and John L. Dornhoffer</i>	209
Case Study 4–13. Venous Malformation of the Tongue <i>James Y. Suen</i>	215
Case Study 4–14. Vascular Malformation of the Geniculate Ganglion and Internal Auditory Canal <i>James C. Wang and Jennifer J. Shin</i>	220
Case Study 4–15. Laser Therapy for Venous Malformations <i>H. Peter Berlien</i>	224
Chapter 5. Lymphatic Malformations	229
Basic Tenants and Interventions <i>Rachel A. Giese and Gresham T. Richter</i>	229
Case Study 5–1. Mixed Cheek Lymphatic Malformation <i>Robert S. Glade and G. Paul Digoy</i>	233
Case Study 5–2. Complex Large Neck Macrocystic Lymphatic Malformation <i>Adva Buzi and Steve Sobol</i>	238
Case Study 5–3. Parotid Lymphatic Malformation <i>Kris R. Jatana, William E. Shiels, and Gregory Wiet</i>	243
Case Study 5–4. Cervicothoracic Macrocystic Lymphatic Malformation <i>Deidre Wyrick, Gresham T. Richter, and Richard Jackson</i>	251
Case Study 5–5. Upper Lip Microcystic Lymphatic Malformation <i>Jeffrey C. Rastatter and Stephen R. Hoff</i>	258
Case Study 5–6. Bilateral Stage Four Lymphatic Malformation of the Neck <i>Jonathan A. Perkins</i>	266
Case Study 5–7. Lymphatic Malformation of the Mandible <i>James Y. Suen</i>	273
Case Study 5–8. Orbital Lymphatic Malformation <i>Srinivasan Paramasivam, Peter W. MacIntosh, Alejandro Berenstein, Teresa M. O, and Aaron Fay</i>	278
Case Study 5–9. Laryngeal Microcystic Lymphatic Malformation <i>Gresham T. Richter</i>	286
Case Study 5–10. Ex Utero Intrapartum Treatment Procedure for Large Cervical-Mediastinal Lymphatic Malformation <i>Kris R. Jatana, William E. Shiels, and Gregory Wiet</i>	293
Case Study 5–11. Deep Cervical/Supraclavicular Lymphatic Malformation <i>Charles A. James and Amir Pezeshkmehr</i>	300
Case Study 5–12. Tongue Microcystic Lymphatic Malformation <i>Gresham T. Richter</i>	307

Chapter 6. Arteriovenous Malformations	315
Basic Tenants and Interventions	315
<i>James Y. Suen, Rachel A. Giese, and Gresham T. Richter</i>	
Case Study 6–1. Focal Arteriovenous Malformation of the Lower Lip	318
<i>James Y. Suen</i>	
Case Study 6–2. Midfacial Arteriovenous Malformation Involving the Upper Lip	322
<i>Behfar Eivazi and J. A. Werner</i>	
Case Study 6–3. Facial Arteriovenous Malformations	328
<i>Patricia E. Burrows</i>	
Case Study 6–4. Extensive Cervicofacial Arteriovenous Malformation	334
<i>Javier Couto and Arin K. Greene</i>	
Case Study 6–5. Auricular Arteriovenous Malformation	339
<i>Wayne F. Yakes and Alexis M. Yakes</i>	
Case Study 6–6. Scalp Arteriovenous Malformations	343
<i>Dov C. Goldenberg, Patricia Y. Hiraki, and Andrea Koga</i>	
Case Study 6–7. Periorbital Arteriovenous Malformation	351
<i>Xindong Fan and Lixin Su</i>	
Case Study 6–8. Multicentric Arteriovenous Malformation of the Face	358
<i>Adewumi Amole</i>	
Case Study 6–9. Arteriovenous Malformation of the Upper Lip	367
<i>James Y. Suen</i>	
Case Study 6–10. Hereditary Hemorrhagic Telangiectasia-Related Epistaxis	374
<i>Angela C. Paddack and Marcus W. Moody</i>	
Case Study 6–11. Dural Arteriovenous Fistula	380
<i>Mary E. Meek</i>	
Case Study 6–12. Arteriovenous Malformation of the Tongue	384
<i>James Y. Suen</i>	
Case Study 6–13. Midline Arteriovenous Malformation of the Upper Lip in a Child	390
<i>Milton Waner and Teresa M. O</i>	
Case Study 6–14. Arteriovenous Malformation of the Mandible	396
<i>Tara L. Rosenberg, Gresham T. Richter, and James Y. Suen</i>	
Case Study 6–15. Auricular and Parotid Arteriovenous Malformation	402
<i>Jonathan M. Grischkan, Andrew J. Rabe, and Kris R. Jatana</i>	
<i>Index</i>	409

Preface

A better understanding of the nature and source of vascular anomalies has vitalized an interest in this field among numerous disciplines. The language used to describe these lesions is now coherent across specialties and allows for treatment algorithms to be unified. However, each vascular tumor and malformation has a unique management profile based on its type, size, and location as well as disciplines involved. Head and neck vascular anomalies are no exception to this rule and are the subsequent motivation behind this text.

We designed *Head and Neck Vascular Anomalies: A Practical Case-Based Approach* with the goal to provide hands-on, step-by-step, management algorithms for specific vascular anomalies of the head and neck encountered in daily practice. This is a condensed, multidisciplinary, practical guide for both simple and complex lesions. Our colleagues in otolaryngology, dermatology, pediatric surgery, plastic surgery, oncology, and interventional radiology have all contributed amazing cases with clinical detail, scientific evidence, and therapeutic options.

In each chapter, the initial steps to diagnose a vascular lesion are followed by a recommended treatment in a case-based format with photographs, radiographic imaging, and alternative therapies. All cases are based upon current literature with the aim to give state-of-the-art information on the major-

ity of head and neck vascular anomalies. Medical, radiographic, and surgical techniques for frequently encountered and more difficult vascular anomalies are described.

This text is designed to be a reference guide. As you will see, each case follows a consistent and relatively rigid presentation outline. This style is meant to provide clarity, brevity, and simplicity to the reader. As a result, redundancies may be encountered for similar anomalies. For this we apologize, but frankly, we did not design the text to be read from cover to cover. Actually, we hope the reader can simply turn to a chapter and capture a complement of knowledge required to help their specific patient.

Of note, we also did not filter out any author or discipline bias in the chapters. In essence, the authors were allowed to express their opinion and therapeutic approach to their assigned case with the requirement to provide treatment alternatives. This decision was made to maintain the authenticity of opinion that is frequently found in the multidisciplinary field of vascular anomalies.

We, thereby, humbly submit to you *Head and Neck Vascular Anomalies: A Practical Case-Based Approach*. With an increasing number of vascular anomaly centers, patients, and interest in the field, we hope you find this text important to your everyday practice and a valuable aid for your patients.

—Gresham T. Richter and James Y. Suen

1

Infantile Hemangiomas

BASIC TENANTS AND INTERVENTIONS

Gresham T. Richter

Basic Tenants

Infantile hemangiomas (IHs) are the most common vascular tumor. They are composed of proliferating immature endothelial cells that express histologic marks found on placental blood vessels (GLUT-1, Lewis Y Antigen, FcγRII, and merosin).¹ IHs are thought to be sporadic events although family lineage has been reported.² Coincidentally, IHs are also the most common tumor of infancy and are present in approximately 5% of the population.³ They have a higher prevalence in females, Caucasians, and

premature, and low birth weight infants.⁴ They also occur more frequently in infants from mothers with early trimester bleeding, preeclampsia, and placental anomalies.

Infantile hemangiomas are rarely present at birth but early blanching or macular erythema of the skin may be a precursor to their later development. They may present anywhere on the body but involve the head and neck in over 60% of cases. Eighty percent of IH grow within the first 3 months of life and continue to grow up to 1 year of age.⁵ IHs undergo predictable proliferative, quiescent, and involution

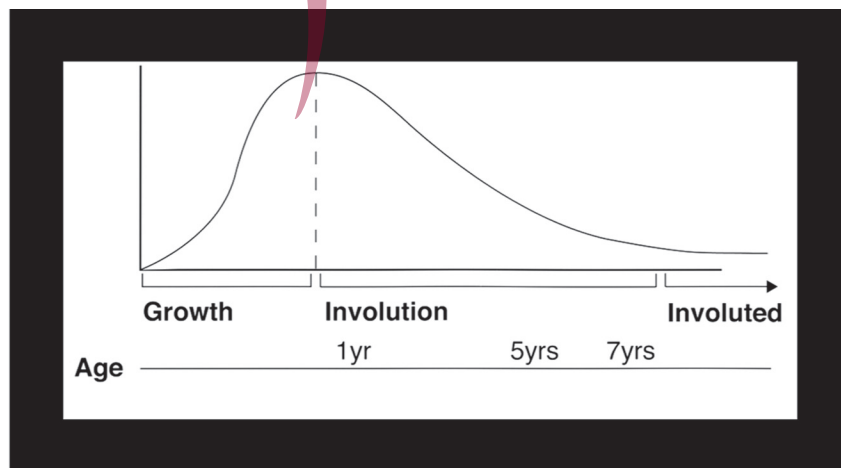


Figure 1-1. Typical growth phases of infantile hemangiomas.

phases as seen in Figure 1–1. The majority of IHs are thought to involute completely by 7 years of age. This natural history can help the clinician differentiate IHs from other congenital lesions and guide management decisions.

The classification of IHs is rather complex. They are first determined to be either focal or segmental. Focal IHs have discrete borders and further characterized as either superficial, deep, or compound. Early nomenclature has been supplanted by this new terminology to describe the majority of IHs (Table 1–1). Superficial and compound hemangiomas present with dark red cutaneous staining in a cobblestone pattern. Compound hemangiomas contain a subcutaneous component whereas deep hemangiomas do not involve the skin and present as a protuberance with an overlying blue skin discoloration (Figure 1–2). It is extremely rare for a focal IH to involve muscle or penetrate beyond subcutaneous fat. An exception is a parotid IH, the most common nonepithelioid tumor of the gland, which is frequently deep.

Problematic focal IHs typically involve the lip, eyelid, orbit, and subglottis where aesthetic and

functional problems occur during the rapid proliferative phase. Sixteen percent of infants with 5 or more focal IHs will also have hepatic involvement and should undergo abdominal ultrasound.⁶ Segmental IHs have a more complex growth pattern than their focal counterpart. In the head and neck, segmental IHs follow a trigeminal nerve (V) distribution. They are diffuse, compound, and maintain irregular borders. More than one facial subunit is frequently involved. They usually penetrate into deep fascial planes of the head and neck. The beard distribution IH (V3) is most commonly described.⁷ These involve the lower lip, chin, neck, and preauricular areas and are frequently accompanied with ulceration. Sixty-three percent of segmental beard distribution will involve the subglottis and require airway endoscopy. All patients with segmentally distributed IHs should undergo systematic evaluation for PHACES (posterior fossa malformations, hemangiomas, arterial lesions, cardiac abnormalities, eye abnormalities, sternal cleft) syndrome.

The cause of IHs remains unclear but is postulated to either be ectopic placental tissue or an endo-

Table 1–1. Old and New Nomenclature for Infantile Hemangiomas

Old Nomenclature	New Nomenclature
Strawberry or Capillary Hemangioma	Superficial Hemangioma
Cavernous Hemangioma	Deep Hemangioma
Capillary Cavernous Hemangioma	Compound (Mixed) Hemangioma



Figure 1–2. Focal hemangiomas described as superficial, compound, or deep (left to right).

thelial progenitor stem cell.⁸ IHs are not associated with increased morbidity or mortality except in the very large hemangiomas that may rarely cause high output heart failure.

Intervention

Because of their natural involution, IHs were historically managed with observation alone. Although many resolve spontaneously others will cause significant functional and disfiguring consequences. Problematic hemangiomas are defined as those leading to significant events affecting the future life of the child. Most problematic events from IHs occur during the proliferative phase and include ulceration, bleeding, pain, vision disturbance, airway compromise, and feeding difficulties. However, late and deforming sequelae also occur to include scarring, telangiectasias, and fibrofatty residuum (Figure 1–3). Many cease to improve after 4 years of age and up to 69% of IHs will leave residual lesions.⁹ At least 10% of IHs persist beyond 9 years of age. The age of self-recognition occurs around 4 years of age and must be considered in the treatment of hemangiomas during their early phase of growth. Although it is difficult to predict future consequences for each lesion, early observation for rapid growth, protuberance, segmental disease, and functional compromise will help guide appropriate therapy.

Both surgical and medical interventions are available in the treatment of IHs. These include surgical excision, laser therapy, topical therapy, intralesional corticosteroids, systemic corticosteroids, systemic beta-blockers, and vincristine chemotherapy. Each of these therapeutic modalities is discussed in the following case presentations. Every IH has a unique profile that governs its treatment and is typically based on location and risk of aesthetic and functional compromise. Management during the proliferative phase generally will lead to the best final outcome. However, many IHs require multimodal therapy of which the final treatment occurs during the involution period. Absolute indications for early intervention include an impact on vital structures, active or impending functional impairment, the possibility of permanent scarring, large segmental facial hemangiomas, and ulcerative lesions.



Figure 1–3. Focal scalp hemangioma at 4 months and seen again, untreated, at 3.5 years with resultant residuum that will require intervention.

References

1. North PE, Waner M, Mizeracki A, Mihm MC, Jr. GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. *Hum Pathol.* 2000;31:11–22.
2. Blei F, Walter J, Orlow SJ, Marchuk DA. Familial segregation of hemangiomas and vascular malformations as an autosomal dominant trait. *Arch Dermatol.* 1998;134:718–722.
3. Dickison P, Christou E, Wargon O. A prospective study of infantile hemangiomas with a focus on incidence and risk factors. *Pediatr Dermatol.* 2011;28:663–669.

4. Haggstrom AN, Drolet BA, Baselga E, et al. Prospective study of infantile hemangiomas: demographic, prenatal, and perinatal characteristics. *J Pediatr.* 2007;150:291–294.
5. Chang LC, Haggstrom AN, Drolet BA, et al. Growth characteristics of infantile hemangiomas: implications for management. *Pediatrics.* 2008;122:360–367.
6. Horii KA, Drolet BA, Frieden IJ, et al. Prospective study of the frequency of hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas. *Pediatr Dermatol.* 2011;28:245–253.
7. Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous hemangiomas in a “beard” distribution. *J Pediatr.* 1997;131:643–646.
8. Yu Y, Flint AF, Mulliken JB, Wu JK, Bischoff J. Endothelial progenitor cells in infantile hemangioma. *Blood.* 2004;103:1373–1375.
9. Luu M, Frieden IJ. Haemangioma: clinical course, complications and management. *Br J Dermatol.* 2013;169:20–30.



CASE STUDY 1–1. ANTERIOR NECK HEMANGIOMA*Abby R. Nolder***Representative Case**

A 2-month-old, former 26-week preterm newborn male was referred to the pediatric otolaryngology clinic for evaluation of middle ear pathology following a failed newborn hearing screen. During that visit, the patient's mother expressed concern about a growing mass under the child's chin. It was not present at birth but had been rapidly progressing over the last several weeks. He had a history of intubation for 2 days in the neonatal intensive care unit but had no associated airway symptoms. He was having some feeding difficulties that seemed to be worsening as the mass increased in size.

On physical examination, he was found to have a 4-cm, soft, mobile, cystic appearing submental neck mass with faint blue discoloration of the overlying skin (Figure 1–4). He also had a 3 × 4-cm compound, pedunculated hemangioma on the right posterior

scalp without bleeding or ulceration (Figure 1–5). No other lesions were discovered elsewhere on his body. He had mild stertor at rest without significant retractions or increased work of breathing; however, work of breathing increased during bottle feeding resulting in spillage of formula from his mouth.

Overview

Hemangiomas are common vascular tumors, occurring in up to 10% of children.¹ They grow rapidly during the first year of life and depending on the anatomic location can cause significant functional and cosmetic impairments. Hemangiomas of the neck should be managed based on the size and symptoms (eg, ulceration, bleeding) of the lesion. Rapidly growing tumors of the anterior neck can cause compression and result in airway and feeding difficulties in young infants; therefore, prompt



Figure 1–4. Midline anterior neck hemangioma.