# Hemangioma and its Management

**Sobhan Mishra[1], Subrat Padhiary[2], Bijayalaxmi Panigrahi[3], Shibasis Biswas[3]**

*[1]Professor & Head, [2]Professor, [3]Post Graduate Trainee, Department of Oral and Maxillofacial Surgery, Institute of Dental Sciences, Siksha 'O' Anusandhan (Deemed to be University), Bhubaneswar*

## Abstract

The highest frequent benign vascular neoplasms that occur quite often in neonates & infants are haemangiomas. Haemangiomas usually occur on the surfaces of the head and neck regions. Management of haemangiomasincludes wait and watch, drug therapy, sclerotherapy, cryotherapy, radiotherapy, laser therapy and surgical excision.Size, location, hemodynamic of the lesion and age of the patient are the important conditionsthat need to consider while planning for treatment of haemangiomas.

***Keywords:*** *Haemangiomas; Head and neck tumor; Management; Treatment.*

## Introduction

Haemangiomas are commonest benign vascular neoplasms that occur often in neonates, infants, and children. Microscopically thereisa proliferation of endothelial cells, which comprise a space comprising blood that is covered with an endothelial layer. Haemangiomas are classified into two types according to development,they are infantile and congenital haemangiomas.[1] Infant haemangiomas (IHs) seems to be the most common childhood tumor that occurs in 5-10 percent of children. These are vascular tumors associated with abnormal propagation of endothelial cells and anomalous architecture of the blood vessels. They can be differentiated from vascular malformations, which are structural anomalies and inborn vascular morphogenesis errors. These include capillary anomalies (includes port-wine stains), venous anomalies, lymphatic anomalies (also known as lymphangiomas or cystichygromas).[12] Infantile haemangiomasare benign, self-limited vascular endothelial neoplasms which commonly affect the maxillofacial and cervical region.[2] These tumorshave 3 phases' i.e. apparent neonates and grow rapidly to its maximum dimension during the first year of life, preceded over several years by stabilization and spontaneous involution. Spontaneous resolution occurs in 49% of patients within 5 years and 7 years of age 72% of cases resolved spontaneously. But few lesions can take up to 10 years to resolve. These tumors don't typically pose any functional issues; but in approximately 10-20 percent of cases, where there will be facial disfigurement, airway compromise due to pressure, loss of vision or ulceration, active intervention is required.[3]

**IH are classified based upon their depth of lesion.[4] It includes:**

1. "Superficial IH": bright red, thin, frequently paved/ pebbled, vascular papules/plaques, formerly known as "strawberry haemangiomas"
2. "Deep IH": blue/violaceous or skin-colored, compressible, subcutaneous papule/nodules, formerly known as "cavernous haemangiomas"
3. "Combined IH": both superficial and deep components, oftenfound as superficial IH overlying a deep IH component

**IH classified based upon anatomic configuration includes:**

1. "Focal IH": a solitary, isolated IH, frequently a papule/plaque/nodule

**Corresponding Author:**
**Dr. Bijaylaxmi Panigrahi**
Post Graduate Trainee, Department of Oral and Maxillofacial Surgery, Institute of Dental Sciences, Siksha 'O' Anusandhan (Deemed to be University), Bhubaneswar
e-mail: missdentist.rosy@gmail.com

2. "Segmental IH": IH involving larger region or whole body (i.e., hand, lower leg, hemi-face) in a well-demarcated area
3. "IH indeterminate": IH those does not fall uniformly into either category or that doesn't proliferate fully/ evenly[4]
4. "Multifocal IH": multiple, discrete focal IH not occupying a specific area or territory.

**"Congenital haemangiomas (CH)"can be classified as below:**

1. "Rapidinvolution congenital haemangioma" (RICH),
2. "Partiallyinvolution congenital haemangioma" (PICH) and
3. "Non-involution congenital haemangioma" (NICH)[3]

**Figure 1**

**Initial Evaluation & Management:** Haemangioma's management must be planned based on the patient's age, the diameter & extent of the lesion, growth rate, complexity (superficial, deep or segmental),complications (like ulceration or vision loss, or vital organ dysfunction) and location. There is a 4% increase in the need for intervention for approximately every 10 cm rise in lesion diameter.[5] Two systems were proposed to help clinicians evaluate the lesion extension and the response to the therapy. Haemangioma Severity Scale (HSS), developed in 2012, was designed to classify patients based upon complications and help in guiding treatment needs. Patients scoring above 10 usually needed beta-blocker therapy with the HSS. Higher HSS scores were associated with a raised presence of an ulcer, structural abnormalities, and everlasting disfiguration. Hemangioma Activity Score (HAS) had been proposed in 2011, based in large measure on IH lesion color. One prospective study conducted by the HAS system's creators compared HSS to HAS and concluded that the HAS system was preferable in assessing the response of therapy.[2] Generally, lesions that are segmental, large in number, or peri-orbital should be considered complicated. Additionally, lesions larger than 5 cm in size may raise skeptical for extensive underlying defect.[6]

Phace Association: PHACE syndrome is a rare condition diagnosed in 1996 and with limited research data. Following conditions are associated with PHACE syndrome:

Malformations of posterior fossa of the brain: Haemangioma, Anomalies of the artery, Aortic coarctation/other cardiac problems, Malformation of the eye.

The white female neonates with massive haemangiomas of the face are at a risk for PHACE syndrome must always be screened for extreme aortic coarctation using echocardiography or cardiac magnetic resonance imaging. Systemic beta-blockers cannot be given to these patients as coarctation is a contraindication for b-blocker usage. Before treatment selection, MRI of the head and neck with angiography is suggested as vascular malformations involved with PHACE syndrome can lead patients to hypotension condition.[7-11]

**Non-operative Management:** Medical intervention usually not necessary for Small, uncomplicated, localized, superficial capillary haemangiomas. Management involves reassurance, teaching, and the setting of expectations with sequential followups.[2] Many IHs are operated by observation because 90 percent are minimal, localized, and do not include areas of aesthetic or functional significance. In the proliferative stage, infants are closely monitored regularly if a lesion has the potential to cause obstruction, damage, or ulceration that needs intervention. Once the IH has stabilized in growing, patients are followed annually during the involving process, if intervention in childhood may still be necessary.[12, 13]

B Adrenergic Blockers: Propranolol is a nonselective β-blocker commonly recognized as a first-line treatment for complexhaemangiomas. Following the serendipitous discovery of the dramatic action of propranolol on lesions in 2008 in patients having steroid-induced cardiomyopathy under treatment for haemangiomas.[14] The efficacy has been verified by several broader research and randomized clinical trials. B-blockers action on haemangiomas is not clear still. Propranolol is believed to limit the development of the lesion by blocking proangiogenic activation of the vascular endothelial growth factor and the essential fibroblast growth factor. Apoptosis is also believed to be aggravated in the late phase of therapy.[15]

Since 2014, the US Food and Drug Administration has approved Propranolol for the proliferation of infant haemangiomas requiring systemic treatment. Complete or near full resolution of the infantile haemangioma is normal after 3-6 months of therapy during the proliferative stage. Patients of cardiogenic shock, chronic sinus bradycardia, chronic hypotension, 2nd or 3rd-degree cardiac obstruction, heart failure, H/O reactive airway dysfunction, aortic coarctation, preemies under 5 weeks and PHACE syndrome-associated vascular anomalies (predisposing to stroke in hypotension setting), a systemic beta-blocker is contraindicated.[16]

Dose: It is advised to initial treatment with 0.5-1mg/kg/day oral propranolol in 2-3 divided dosages with feed. The dosage may be raised progressively by 0.5 mg/kg/day over 2 weeks to a target dose of 2 mg/kg/day, with 2-3 divided dosages. Considering the possible side effect of hypoglycemia, it is highly advised to medication with feed.

**Topical β-Adrenergic Blockers:** Topical b-blocking agents are usuallyused to managetiny, superficial, uncomplicated,and ulcerated haemangiomas. 92 percent and 77 percent of patients having uncomplicated haemangiomas reported a substantial recovery in the color and dimension of the lesions, respectively, after 6-9 months of topical timolol therapy in a study. All topical timolol and propranolol found to have similar efficacy.

**Corticosteroids:** Previous to the introduction of oral propranolol, systemic corticosteroids had been the mainstay for IH management. The usage of systemic corticosteroids in managing haemangiomas is currently restricted to patients contraindicated to systemic b-blockers. The mode of action of corticosteroids on haemangiomas is not known but it has been shown that corticosteroids impede vasculogenesis by inhibiting the factor-A expression of vascular endothelial development.

Dose- While steroid dose had been variable, oral prednisolone at 2-3 mg/kg/day is a normal starting dosage every morning. Depending on the lesion response,usually, the course of treatment lasts for 6-12 months. Once the treatment response is achieved, a steroid taper should commence. Oral corticosteroids could result in adrenal crisis or rebound generation of haemangiomas if discontinued abruptly.[17]

Intra-lesional and Topical corticosteroids: In resistant IHs with mild ulcerations, highefficacy topical corticosteroids, such as clobetasol cream are used. Intralesional triamcinolone acetonide injections are restricted for tiny, localized, deep haemangiomas that are resistant to treatment. Dosage ranges between 10 and 40 mg/mL and shouldn't more than 3 mg/kg/day. Clinical lesion reaction is usually noticed initially in the first 2 weeks of injection, with further improvements over the subsequent months.

**Laser Therapy:** In fewer cases, pulse-dye laser (PDL) can be used to treat IH. While its use has been limited to ulcerated IH in the superficial or very early lesions where Pulse Dye Laser therapy tends to promote re-epithelialization and analgesia. It is also used subsequently for residual telangiectasias in life after IH involution. In combination with other treatments, like topical timolol or systemic treatment (propranolol or corticosteroids), PDL is quite effective when used for treating ulcerated IH.

**Surgical Management:** Surgical excision of infantile haemangioma is reserved for fibro-fatty residues, post involution phase scars, or deformed zones for reconstruction. Surgery could frequently be considered in patients who are unresponsive to conservative management in the proliferative stage.[18]

**Sclerotherapy:** Sclerosing agents' intralesional injection, such as bleomycin, may be useful in small lesions and only 49 percent of thorough resolution is recorded in literature. Sclerotherapy has been extensively used in the management of minorextraoral lesions. However, after the injection of the sclerotic agent into intraoral lesions, the pressurized bandage cannot be applied, thereby restricting the efficacy for intraoral use. Cutaneous necrosis and neural impairment are the commonadverse effects of sclerotherapy. Systemic complications are uncommon and linked to alcohol's absorption into the systemic circulation. These can involve hemolysis and cardiac arrest with possible renal toxicity. There must beno extravasation during injection.[19]

**Embolization:** Vascular anomalies already in advanced stages are generally best managed by preoperative embolization followed by surgically, particularly if not responsive to medicines. The efficacy of embolizing vascular lesion in normalizing function and bleeding prevention with decent cosmetic outcomes as demonstrated by Churojana et al. Nonetheless, extended intubation or tracheostomy must be considered in the condition of involving the tongue, oropharynx and laryngotracheal region to protect the airway from extreme inflammation or significant postoperative bleeding as claimed by them. Besides, the embolization procedure can have some possible disadvantages. The risk of iatrogenic embolization through majorcarotid arteries has been identified on intracranial vessels. Therefore, this procedure needs to be performed by an experienced radiologist, and temporary flow blockage is required.[3]

**Radiotherapy:** Radiotherapy especially forintraosseoushaemangiomas, has been described as a beneficial option. Though, its negative effects, like damage to the regular development of adjacent tissues, residual scarring, and neoplasm, are known and unjustified for benign neoplasms such as haemangiomas and vascularmal formation.[3, 20]

## Conclusion

Although most haemangiomas could be observed without treatment, medical or surgical intervention will benefit others. While surgical excision remains the favored treatment, it can be appropriate for curettage, sclerotherapy, embolization, or a combination of those method. Additionally, routine follow-ups should be carried out due to the possible risk of complications and recurrence. [12]Where complications are possible or management is unclear, it maybe advisable to refer to an expert or multidisciplinary vascular anomaly center.

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