



CIGNA MEDICAL COVERAGE POLICY

The following Coverage Policy applies to all health benefit plans administered by CIGNA Companies including plans formerly administered by Great-West Healthcare, which is now a part of CIGNA.

Subject Treatment of Cutaneous Hemangioma, Port Wine Stain and Other Vascular Lesions

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INSTRUCTIONS FOR USE

Coverage Policies are intended to provide guidance in interpreting certain **standard** CIGNA HealthCare benefit plans. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement (GSA), Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document **always supercedes** the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. Proprietary information of CIGNA. Copyright ©2011 CIGNA

Coverage Policy

Coverage for the surgical treatment of a cutaneous hemangioma, port wine stain , or other vascular lesion is dependent upon benefit plan language, may be subject to the provisions of a cosmetic and/or reconstructive benefit and may be governed by state mandates.

Under many benefit plans, the surgical treatment of a cutaneous hemangioma, port wine stain , or other vascular lesion is not covered when performed solely for the purpose of improving or altering appearance or self-esteem, or to treat psychological symptomatology or psychosocial complaints related to one's appearance.

Under many benefit plans formerly administered by Great-West Healthcare reconstructive services and surgery are covered when the reconstruction services are being performed for one of the following primary purposes: 1) to restore large skin defects due to a port wine stain; 2) to relieve severe physical pain caused by an abnormal body structure; 3) to treat a functional impairment caused by an abnormal body structure or to restore an individual's normal appearance, regardless of whether a functional impairment exists when the abnormality results from a documented illness that occurred within the preceding 12 months.

Please refer to the applicable benefit plan document to determine the terms, limitations and conditions of coverage.

If coverage is available for surgical treatment of a cutaneous hemangioma, port wine stain, or other vascular lesion, the following conditions of coverage apply.

CIGNA covers invasive treatment of a cutaneous hemangioma or other vascular malformation (i.e., venous, arteriovenous, lymphatic) as medically necessary for EITHER of the following indications:

- the lesion is compromising a vital structure (e.g., nose, eyes, ears, lips or larynx)
- the lesion results in **ANY** of the following:
 - bleeding
 - pain
 - ulceration
 - repeated infection
 - eating difficulty
 - swallowing difficulty

When the medical necessity criteria noted above for invasive treatment of a cutaneous hemangioma or other vascular malformation (i.e., venous, arteriovenous, lymphatic) have been met, CIGNA covers ANY of the following invasive treatments as either monotherapy or combination therapy:

- injectable corticosteroids
- sclerosing agents
- laser therapy
- surgical excision
- cryosurgery
- embolization
- radiotherapy

CIGNA covers laser treatment of a port wine stain as medically necessary for EITHER of the following indications:

- the lesion results in bleeding or painful nodules
- the lesion results in obstructed vision

General Background

Vascular lesions may be classified into two main categories: vascular tumors and vascular malformations. Vascular tumors are characterized by vascular endothelial cell hyperplasia and spontaneous involution. The most common vascular tumors are hemangiomas.

Vascular malformations are abnormalities in blood vessel formation; the lesions do not regress and slowly enlarge. The name of the malformation reflects the blood vessel forming the lesion: capillary, venous, arterial or lymphatic. A common capillary malformation, the port wine stain, is characterized by flattened endothelial cells with normal turnover. Venous malformations give a bluish color to the area under the involved skin or mucosa. Arterial malformations are rare, are often referred to as arteriovenous malformations and are direct connections of arteries to veins. Lymphatic malformations can involve either large (cystic hygroma) or small vessels (lyphangioma circumscriptum). Vascular malformations can also consist of combinations, such as with Klippel-Trenaunay Syndrome or Sturge-Weber Syndrome.

Vascular lesions may result in permanent disfigurement with the main goal of treatment aimed at improving cosmesis. However, some vascular lesions interfere with functioning of vital structures or result in symptoms, such as pain, ulceration and bleeding. Treatments are dependent on the type and severity of the lesion but generally include injectable medications, laser therapy, sclerotherapy, embolization, surgical debulking, and compression garments and in some cases radiotherapy.

Cutaneous Hemangiomas

Cutaneous hemangiomas are characterized by rapid proliferation in early infancy and slow involution that may occur over several years. The mechanisms that control involution are not well understood. Some hemangiomas are present at birth as precursor lesions; rarely are they fully formed tumors at birth (AAD, 2010). More commonly, lesions become evident after birth, usually within two to four weeks. These hemangiomas frequently occur on the face and neck area but may also be located on areas such as the trunk and/or internal organ structures. Most hemangiomas do not require medical intervention, although a small number cause functional complications or disfigurement. Permanent disfigurement is more likely if lesions are present on the face; the nose, lip, and forehead are most vulnerable (Conlon and Drolet, 2004).

Several types of congenital hemangiomas have been described in the literature and include rapidly involuting, and noninvoluting, to name a few. Kasabach-Merritt syndrome is a complication of rapidly enlarging vascular lesions (hemangioma) and is characterized by hemolytic anemia, thrombocytopenia and coagulopathy. While these lesions are not hemangiomas of infancy, they result from a more aggressive proliferative vascular tumor that results in decreased platelets and other bleeding problems. Although the syndrome is rare it requires aggressive treatment and is often associated with a high mortality rate.

For a majority of hemangiomas no intervention is needed and lesions regress spontaneously. Some lesions result in untoward cosmetic changes that have no clinical significance. However, complications resulting from hemangiomas have been reported and are often related to the site of occurrence, with some requiring treatment. The most frequent complication associated with hemangiomas is ulceration which is often present during the proliferative phase. Periorbital hemangiomas may cause amblyopia, impaired vision and astigmatism and should be considered when a hemangioma involves the eyelids or periorbital tissue. Lesions located on the ear may result in auditory impairment and secondary speech delay. Subglottic hemangiomas may cause hoarseness and stridor leading to respiratory impairment. Many patients with subglottic hemangiomas also have cutaneous hemangiomas involving the lips, chin, and mandible. Hemangiomas may also be located on the cervicofacial area and lumbosacral spine. Pedunculated hemangiomas may be at risk for bleeding and irritation and have been associated with permanent cosmetic skin changes after involution such as fibrofatty tissue and excessive scarring.

The main goals of treatment include preventing permanent disfigurement and minimizing psychosocial distress, preventing functional complications, and treating ulceration. Several modalities have been used to treat hemangiomas and include the administration of steroid medications as the mainstay of treatment (e.g., topical, intralesional and systemic), pulsed dye laser therapy, and interferon. The pulsed-dye laser has been proven effective for the treatment of superficial hemangiomas, the superficial component of mixed hemangiomas and ulcerated hemangiomas. Efficacy is however limited by the depth of laser penetration. Several treatments may be necessary, and treatments have been associated with some risk of scarring (Rudolph, 2003). Other, less common treatments include: cryotherapy, other forms of laser surgery, embolization, and use of chemotherapeutic agents, such as vincristine and cyclophosphamide. Other forms of laser surgery have included the argon laser for hemangiomas, the Nd:YAG (neodymium: yttrium-aluminum-garnet) laser for deeper lesions, and carbon dioxide laser for lesions such as subglottic hemangiomas. Some of these devices have been associated with significant scarring. Surgical excision may be recommended for hemangioma lesions that are sharply demarcated and pedunculated, are ulcerated and bleeding, have not responded to other modalities of treatment, and those that are function-threatening (Rudolph, 2003).

Port Wine Stain

Port wine stains are a type of vascular malformation involving the superficial capillaries of the skin. They vary in size and location and are usually present at birth although not always clinically evident. In rare cases, a port wine stain may be referred to as "acquired" and become evident after injury to the skin or in association with hormonal influences (Legiehn, Heran, 2008). Most often, lesions are found on the face, neck, arms or legs. They may be related to other underlying conditions, such as Sturge-Weber syndrome. Sturge-Weber syndrome, also known as encephalotrigeminal angiomatosis, is characterized by a facial port wine stain in a trigeminal V1 (i.e., ophthalmic) distribution, leptomeningeal angiomatosis, and choroidal vascular malformation of the eye, which can lead to ipsilateral glaucoma and buphthalmos. Glaucoma occurs in 30% of patients with Sturge-Weber syndrome, and it develops before two years of age in 60% of these patients (Hussain, et al., 2004).

Port wine stains appear as sharply demarcated pink-red patches that darken with time and do not proliferate; growth of the lesion is dependent upon growth of the child. As the child matures, the lesion may become raised and exhibit red-to-purple nodules and papules in adult years, leading to potential disfigurement (e.g., pebbly and

slightly thickened surfaces), and bleeding with trauma. Hypertrophy may develop in the soft tissue underlying the port wine stain. Early treatment may prevent the progression of development to hypertrophy and nodules in later years. It has been noted port wine stain lesions on the forehead or eyelids can be associated with ocular disorders and warrant frequent ophthalmology exams to prevent damage to the eye.

Laser devices such as the argon, carbon dioxide (CO₂), Nd:YAG, and copper vapor laser have been used to treat port wine stains. In many cases, these laser devices have been associated with poor cosmetic outcomes (Rothfleisch, et al., 2002). Pulsed dye laser therapy has been shown to be the most effective treatment for port wine stains; is associated with less adverse effects, including less post-operative scarring; and is considered the standard treatment of choice (Tucci, et al., 2009; Yang, et al., 2005; Schmults, 2005). Evidence in the published medical literature suggests efficacy is increased if lesions are treated in infancy, although size and location are also predictors of outcome (Conlon, Drolet, 2004). Nonetheless, while most port wine stains lighten after a series of pulsed dye laser treatments, some cannot be completely removed (Yang, et al., 2005).

Other Vascular Lesions/Malformations

Lymphatic (Lymphangioma): Lymphatic malformations consist of abnormally dilated lymphatic channels and commonly affect the head and neck area in children (lymphangioma). Most are present at birth although some may appear later in childhood as a result of infection or trauma. These lesions are either macrocystic or microcystic; microcystic are more difficult to treat and more often associated with complications. Aside from cosmetic concerns, depending on the size and location of the mass the lesion may be symptomatic. For example, when the oral and pharyngeal mucosa is involved there may be tongue swelling, tongue hypertrophy, mucosal bleeding, speech difficulty, and airway compromise. Common complications include disfigurement, infection and bleeding. Treatment is aimed at improving cosmesis, and alleviating any associated symptoms and involves surgical excision and/or sclerotherapy (Wetmore, Potsic, 2010; Tucci, et al, 2009; Morelli, 2007; Freiden, et al., 1997). Although used less frequently, other types of treatment such as scleroembolization and CO₂ laser have also been effective.

Arteriovenous: Arteriovenous malformations (AVM) of the skin are rare; however this type of lesion is a direct connection of artery to vein, bypassing the capillary bed. AVMs may appear at any time from birth to early adulthood and often remain stable for several years. They usually become noticeable at times of hormonal changes and at time smay suddenly enlarge following infection or trauma (Tucci, et al., 2009). If the lesions are asymptomatic treatment is not necessary, however if ulceration and/or bleeding develop treatment is warranted and consists of embolization and excision (Wetmore, Potsic, 2010).

Venous: Venous malformations include but are not limited to vein only malformations and angiokeratomas. These lesions vary in size and may be superficial, deep or a combination of both. The lesions grow as the child grows but have a tendency to enlarge after direct trauma or with hormonal change such as during puberty or pregnancy from progressive ectasia of the vascular structure (Tucci, et al., 2009). For most lesions treatment is not necessary. When treatment is warranted, such as with pain form enlargement, treatment for superficial nodular lesions is surgical excision; larger deeper lesions may be treated with sclerotherapy. Angiokeratomas are characterized by ectasia of the superficial dermal vessels with s hyperkeratosis of the overlying dermal layer (Freiden, et al., 1997). They appear as flat hemangiomas with an irregular surface, with surgical excision being the treatment of choice (Wetmore, Potsic, 2010). Although angiokeratomas are generally asymptomatic bleeding and itching may occur with trauma.

U.S. Food and Drug Administration (FDA)

Lasers are regulated by the FDA as Class II devices and receive approval through the 510(k) process. According to the FDA, pulsed dye lasers are indicated for use in the treatment of cutaneous vascular lesions such as port wine stains and hemangiomas, and benign cutaneous lesions such as warts, striae and some forms of psoriasis.

Professional Societies/Organizations

The American Academy of Dermatology (AAD) published a guideline of care for hemangiomas of infancy (Freiden, et al., 1997). It appears the guideline has not been modified since the initial publication. According to the AAD guideline, treatment of hemangiomas is dependent upon the size, location and severity of the tumor, the age of the patient, and the rate of involution. The guidelines support treatment for the following conditions:

- hemangiomas affecting vision, laryngeal involvement, nasal and auditory canal obstruction, Kasabach-Merritt syndrome, hepatic hemangiomatosis, cardiac failure, and skin ulceration
- hemangiomas that are likely to be disfiguring (e.g., located on the nose, lips, ear)
- hemangiomas that are very large with prominent dermal component, with or without subcutaneous component (e.g., facial hemangiomas)
- pedunculated hemangiomas

Summary

Cutaneous congenital hemangiomas, port wine stains and other vascular malformations such as venous, arteriovenous, or lymphangioma, can result in an undesired appearance. Elective treatment in such cases is often aimed at improving the individual's appearance, and is considered cosmetic. However, these lesions can also require medically appropriate treatment when they cause problems such as ulceration, bleeding, and recurrent infections, or when they are located in areas that can compromise function of vital structures such as eyes, ears, or vocal cords.

Coding/Billing Information

Note: This list of codes may not be all-inclusive.

Hemangiomas and Other Venous Malformations

Covered when medically necessary for the treatment of a cutaneous hemangioma or other vascular lesion (i.e., venous, arteriovenous, lymphatic):

CPT* Codes	Description
11400	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter 0.5 cm or less
11401	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter 0.6 to 1.0 cm
11402	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter 1.1 to 2.0 cm
11403	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter 2.1 to 3.0 cm
11404	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter 3.1 to 4.0 cm
11406	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), trunk, arms or legs; excised diameter over 4.0 cm
11420	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter 0.5 cm or less
11421	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter 0.6 to 1.0 cm
11422	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter 1.1 to 2.0 cm
11423	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter 2.1 to 3.0 cm
11424	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter 3.1 to 4.0 cm
11426	Excision, benign lesion including margins, except skin tag (unless listed elsewhere), scalp, neck, hands, feet, genitalia; excised diameter over 4.0 cm
11440	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter 0.5 cm or less
11441	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter 0.6 to 1.0 cm

11442	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter 1.1 to 2.0 cm
11443	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter 2.1 to 3.0 cm
11444	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter 3.1 to 4.0 cm
11446	Excision, other benign lesion including margins, except skin tag (unless listed elsewhere), face, ears, eyelids, nose, lips, mucous membrane; excised diameter over 4.0 cm
11900	Injection, intralesional; up to and including seven lesions
11901	Injection, intralesional; more than seven lesions
17106	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); less than 10 sq cm
17107	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); 10.0 to 50.0 sq cm
17108	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); over 50.0 sq cm
61626	Transcatheter permanent occlusion or embolization (eg, for tumor destruction, to achieve hemostasis, to occlude a vascular malformation), percutaneous, any method; non-central nervous system, head or neck (extracranial, brachiocephalic branch)

ICD-9-CM Diagnosis Codes	Description
228.01	Hemangioma of skin and subcutaneous tissue
228.1	Lymphangioma, any site
747.60	Congenital anomalies of circulatory system, peripheral vascular, unspecified
759.6	Other congenital hamartosis, not elsewhere classified

Port Wine Stains

Covered when medically necessary for the treatment of port wine stain:

CPT* Codes	Description
17106	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); less than 10 sq cm
17107	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); 10.0 to 50.0 sq cm
17108	Destruction of cutaneous vascular proliferative lesions (eg, laser technique); over 50.0 sq cm

ICD-9-CM Diagnosis Codes	Description
757.32	Congenital vascular hamartomas

***Current Procedural Terminology (CPT®) © 2010 American Medical Association: Chicago, IL.**

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Policy History

<u>Pre-Merger Organizations</u>	<u>Last Review Date</u>	<u>Policy Number</u>	<u>Title</u>
CIGNA HealthCare	4/15/2009	0313	Treatment of Cutaneous Congenital Vascular Lesions

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