Kansas City

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Laser Treatment of Congenital Port Wine Stains and Hemangiomas

Policy Number: 7.01.40
Origination: 10/1988
Last Review: 3/2014
Next Review: 3/2015

Policy

Blue Cross and Blue Shield of Kansas City (Blue KC) may provide coverage for laser treatment of congenital port wine stains and hemangiomas when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered

Laser treatment may be considered **medically necessary** for congenital port wine stains when the lesions are located on the face and neck.

Laser treatment of port wine stains in the presence of functional impairment related to the port wine stains may be considered **medically necessary.**

Laser treatment may be considered **medically necessary** for congenital hemangiomas in patients over the age of three (3) with the presence of *ulceration/bleeding, blocking facial structures or GI/GU tract obstruction. (*Note, with involution, in normal presentation the surface of the lesions turns white/gray and may ulcerate. This normal presentation does not require intervention.)

When Policy Topic is not covered

Laser treatment for port wine stains and congenital hemangiomas is considered **cosmetic** when the criteria above are not met.

Treatment of port wine stains and congenital hemangiomas with lasers in combination with photodynamic therapy or topical angiogenesis inhibitors is considered **investigational.**

Considerations

Hemangiomas are benign vascular proliferations that rapidly enlarge during the first year of life and spontaneously involute by age 2 to 3 years. The lesions are asymptomatic and benign. Typical spontaneous involution leaves the best cosmetic results, and thus nonintervention in uncomplicated lesions is recommended. Less than 2% of hemangiomas require intervention. Individual consideration may be given to patients presenting with hemangiomas considered “complicated” as in the diagnosis of PHACE syndrome.

Performance of a prior test spot is necessary to select suitable candidates for treatment and to determine the degree of scarring that may occur.

- The size of the lesion may require more than 1 treatment.
- Treatment of an extensive area may require general anesthesia.

Description of Procedure or Service

Port wine stains are common vascular malformations that start as pink macules and, if untreated, tend to become darker and thicker over time. They usually occur on the face and neck, but can be located

elsewhere on the body. Treatment with lasers (including pulsed dye lasers, Alexandrite, Nd:YAG lasers and intense pulsed light [IPL]) is proposed.

Background
Port wine stains are the most common of the vascular malformations, affecting approximately 3 in 1,000 children. They are composed of networks of ectactic vessels and primarily involve the papillary dermis. Unlike many other birthmarks, port wine stains do not resolve spontaneously. In contrast, they typically begin as pink macules and become redder and thicker over time due to decreased sympathetic innervation. The depth of the skin lesions ranges from about 1 to 5 mm. Port wine stains are generally located on the face and neck but can occur in other locations such as the trunk or limbs.

Prior to the availability of laser treatment in the 1980s, there were no effective therapies for port wine stains. A laser is a highly focused beam of light that is converted to heat when absorbed by pigmented skin lesions. Several types of lasers have been used to treat port wine stains. Currently, the most common in clinical practice is the pulsed dye laser (PDL), which uses yellow light wavelengths (585-600 nm) that selectively target both oxyhemoglobin and deoxyhemoglobin. Pulsed dye lasers penetrate up to 2 mm in the skin. Newborns and young children, who have thinner skin, tend to respond well to this type of laser; the response in thicker and darker lesions may be lower. Other types of lasers with greater tissue penetration and weaker hemoglobin absorption are used for hypertrophic and resistant port wine stains. In particular, alternatives to the PDL are the long-pulsed 1,064 nm Nd:YAG and 755 nm pulsed Alexandrite lasers. The 1,064 nm Nd:YAG laser requires a substantial degree of skill to use to avoid scarring. Carbon dioxide and argon lasers are relatively non-selective; they were some of the first lasers used to treat port wine stains but were associated with an increased incidence of scarring and are not currently used frequently in clinical practice to treat port wine stains. Intense pulsed light (IPL) devices emit polychromatic high-intensity pulsed light. Pulse duration is in the millisecond range, and devices use an emission spectrum ranging from 500 to 1,400 nm. Compared to other types of lasers, IPL devices include both the oxyhemoglobin selective wavelengths emitted by PDL systems and longer wavelengths that allow deeper penetration into the dermis.

Regulatory Status
Several laser systems have been cleared for marketing by the U.S. Food and Drug Administration (FDA) through the 510(k) process for a variety of dermatologic indications, including treatment of port wine stains. Approved lasers for this indication include the Candela pulsed dye laser system (Candela Corp.; Wayland, MA), the Cynosure Photogenica pulsed dye laser (Cynosure Inc; Westford, MA), and the Cynosure Nd:YAG laser system. In addition, the Cynergy Multiplex Laser (Cynosure), a combined Nd:YAG and pulsed dye laser was approved by the FDA in 2005 for treatment of benign vascular and vascular dependant lesions, including port wine stains.

In 2003, the Lumenis family of intense pulsed light systems was approved by the FDA; indications for use include dermatologic applications. Subsequently, the NannoLight intense pulsed light system (Global USA Distribution) was approved by the FDA in 2008 and the Mediflash3 and Esterflash3 systems (Dermeo) were approved in 2010 for indications specifically including treatment of port wine stains.

Congenital hemangiomas are benign vascular proliferations that appear at or shortly after birth, usually within 4 to 6 weeks. They occur in 1% to 3% of newborns and in 10% to 20% of children by 1 year of age. From 15% to 30% of infants have multiple lesions. Hemangiomas are characterized by a 6- to 12-month period of proliferation during which they grow to a size of 2 to 20 cm. This is followed by a stationary or plateau period in which there is little change. A period of slow involution, or regression, begins at approximately 15 to 18 months of age and can last for several years. Complete regression occurs in approximately 50% of children by 5 years of age and 90% of children by 9 years of age. The superficial capillary, or strawberry, hemangioma accounts for approximately 50% to 60% of cases, while deep hemangiomas (also called cavernous hemangiomas) account for approximately 15%. Mixed hemangiomas, which contain both superficial and deep components, account for approximately 15% to 30% of lesions. Although they are heterogeneous in their appearance, hemangiomas frequently arise

as telangiectactic macules or blanched spots, or, rarely, as a small ulceration. They are located on the head or neck in 60% of cases, on the trunk in 25%, and on the extremities in 15%. Despite the fact that most hemangiomas resolve on their own, approximately 50% persist in school-age children and, even after involution, 20% to 40% leave behind residual skin changes. Hemangiomas can be complicated by bleeding, ulceration, or secondary infection, or may be located in areas of the body where they cause functional impairment. Some are potentially life threatening, such as those obstructing the respiratory tract. Low-risk hemangiomas are either left untreated or treated with intralesional corticosteroid injections, pressure occlusion, laser therapy, cryosurgery, or surgical excision. High-risk lesions, such as those that are large, potentially disfiguring, in a prognostically poor location, or causing functional impairment or life-threatening complications, are treated with systemic or topical corticosteroids, subcutaneous alpha-2a interferon, laser therapy, surgical excision, or cryosurgery.

Lasers are used to treat both PWS and hemangiomas. The flashlamp-pumped pulsed dye laser (PDL), introduced in 1985, and was developed specifically for the treatment of cutaneous vascular lesions. It emits one specific color, or wavelength, of light that can be varied in its intensity and pulse duration. The hemoglobin within dilated or enlarged blood vessels comprising cutaneous vascular lesions preferentially absorbs the energy from the PDL and generates heat, leading to the thermal destruction of the lesion, while sparing normal surrounding tissues. PDL therapy is administered in multiple sessions in an outpatient setting, with or without topical, local, or general anesthesia. Cryogen spray cooled PDL (CPDL) involves the application of a cryogen spurt to the skin surface milliseconds prior to laser irradiation. This cools the epidermis without affecting the deeper PWS blood vessels, and reduces the thermal injury sustained by the skin during laser treatment.

PHACE syndrome
PHACE Syndrome refers to posterior fossa anomalies, hemangioma, arterial lesions, cardiac abnormalities/aortic coarctation, and abnormalities of the eye. The syndrome is associated with an increased risk of neurological and congnitive impairments. There are both major and minor characteristics of PHACE syndrome defined by the published criteria. The criteria are classified by organ system affected. The organ systems include cerebrovascular, structural brain, cardiovascular, ocular, ventral or midline anomalies.

Major Criteria
- Cerebrovascular: Anomalies of the major cerebral arteries
- Structural Brain: Posterior fossa anomaly including Dandy Walker Anomaly
- Cardiovascular: Aortic arch anomaly, aberrant origin of the subclavian artery
- Ocular: Posterior segment abnormalities
- Ventral or Midline: Sternal defects.

Minor Criteria
- Cerebrovascular: Persistent embryonic arteries, intracranial hemangioma
- Structural Brain: Midline anomalies, neuronal migration disorder
- Cardiovascular: Verntricular septal defect, right aortic arch
- Ocular: Anterior segment abnormalities
- Ventral or Midline: Hypopituitarism

PHACE syndrome can be diagnosed in patients that present with a facial hemangioma >5cm in diameter plus 1 major criteria or 2 minor criteria.

Possible PHACE syndrome can be considered as a diagnosis in patients that present with hemangioma of the neck or upper torso plus 1 major or 2 minor criteria or in patients with no hemangioma plus 2 major criteria noted.

Rationale

The policy was created in 1996 and was on “no further review” status from 2003 to 2010, at which time it returned to active review. The most recent literature search was for the period April 2012 through

April 25, 2013. Following is a summary of the key literature to date on laser treatment of port wine stains.

Laser treatment monotherapy

In 2011, a Cochrane review of trials on light or laser sources for treating port wine stains was published by Faurschou and colleagues. (1) The review included randomized controlled trials (RCTs) comparing any laser or light source to any comparison intervention. Five RCTs with a total of 103 participants met inclusion criteria. The investigators reported that interventions and outcomes were too heterogenous for a meta-analysis of study findings. All studies used a within-participant (e.g., split-side) design and none of them included a sham treatment or no treatment group. Interventions in all of the trials were between 1 and 3 treatment sessions and all trials followed patients for less than 6 months' follow-up. A primary efficacy outcome of the review was reduction in redness; investigators judged that a reduction of more than 20% would represent a clinically relevant effect. In all of the 5 trials, treatment with the pulsed dye laser (PDL) resulted in more than 25% reduction in redness in 50-100% of participants, depending on setting of the laser device. In addition, intense pulsed light (IPL) and the Nd:YAG laser also led to a reduction in redness in 1 trial each. The trials found that long-term adverse effects of laser and light treatment were rare; only 1 participant in 1 trial experienced scarring of the skin and this person had a too-high dose of the Nd:YAG laser. The authors concluded that the evidence supports the use of the PDL as the treatment of choice for port-wine stains.

Representative RCTs included in the Cochrane review and published more recently that evaluated laser treatment of port wine stains are described below:

In 2009, Faurschou and colleagues in Denmark published a study with 20 patients with port wine stains. (2) Port wine stains were on the face (n=15), trunk (n=4), or extremities (n=1). Eight (40%) had previously untreated lesions, and the remainder were previously treated, but with types of lasers not under investigation in the study. Patients received one treatment with a PDL on a randomly selected side of the lesion (left/lower or right/upper) and intense pulsed light (IPL) treatment on the other side. Blinded assessment 12 weeks' post-treatment found a median of 65% percentage lightening on the PDL side and 30% on the IPL side ($p<0.0003$). Fifteen (75%) of 20 patients had more than 70% lightening with PDL treatment compared to 6 (30%) in the IPL group; this difference was also statistically significant, $p=0.014$.

A 2010 study in Germany by Babilas and colleagues was a split-face comparison of PDL and IPL treatment in 25 patients; 11 (40%) had previously untreated port wine stains, and the other 14 had received previous laser treatment. (3) Port wine stains were located in the face and neck region in 18 patients, the trunk in 3 patients, and the extremities in 4 patients. The previously untreated patients were treated with IPL, short-PDL (585 nm and 0.45-millisecond pulse duration), and long-PDL (584-600 nm and 1.5-millisecond pulse duration). Patients who previously failed either short- or long-PDLs did not receive that type of treatment. Blinded outcome assessment was done 6 weeks after treatment. In the treatment-naïve group, assessors rated lightening as excellent (at least 75%) or good (51-75%) in at least one test spot in 7 of 11 (64%) patients after IPL treatment, 5 of 11 (45%) after long-PDL, and 1 of 11 (9%) after short-PDL (between group p value was not reported). In the group that had been previously treated, lightening was rated as excellent or good in at least one test spot in 4 of 14 (29%) patients after IPL treatment, 1 of 14 (7%) after long-PDL treatment, and 0 (0%) after short-PDL treatment.

In 2012, Klein and colleagues in Germany published findings of an RCT evaluating treatment with a diode laser augmented by the dye indocyanine green. (4) The study included 31 patients with port wine stains. Two areas of 2 by 2 cm were selected in each patient's port wine stain. The areas were randomly assigned to receive treatment with a PDL or with an indocyanine green-augmented diode laser (ICG + DL). The cosmetic appearance of the lesions was assessed using a 5-point Likert-type scale with 0=no clearance to 4=excellent clearance. Three months after treatment, the mean investigator-rated clearance score was 0.89 (standard deviation [SD]: 0.99) for lesions receiving PDL

treatment and 1.30 (SD: 1.29) for lesions receiving ICG + DL treatment. The difference in scores between groups was not statistically significant, p=0.11. At 3 months, patients rated the clearance level as a mean of 0.89 (SD: 0.88) after PDL treatment and 1.71 (SD: 1.24) after ICG + DL, p=0.004. Two patients in the diode laser treatment group experienced adverse events. There was one case of site-specific pain during ICG + DL treatment (8 on a 10-point scale) and 1 case of an atrophic scar measuring 5 mm in diameter. Other side effects were burning (PDL: 58%, ICG + DL: 68%), edema (PDL: 3%, ICG + DL: 10%) and purpura (PLD: 71%, ICG + DL, 42%).

Combination treatment

One RCT of combined treatment was identified. In 2012, Tremaine and colleagues published findings from a trial evaluating pulsed-dye laser treatment with and without the addition of imiquimod cream. (5) The study included 24 individuals with port wine stains. All patients initially received 1 session of laser treatment. Previous treatment with the PDL was not an exclusion criterion. Five patients enrolled in the study twice, with a washout period of at least 4 weeks before re-enrollment. Patients were randomized to receive additional treatment with either 5% imiquimod cream or placebo cream, to be applied 3 times a week for 8 weeks, beginning the day following laser treatment. Chromameter measurements were taken at baseline and at 8 weeks after laser treatment. The primary outcomes were change in erythema (defined as red/green color saturation with values ranging from +60 green to -60 red) and overall change in 3 color dimensions (reflected light intensity, green/red color saturation, and blue/yellow color saturation). Two patients were excluded from analysis due to chromameter malfunction. The mean change in erythema was 0.43 (SD: 1.630 for the PDL plus placebo sites and 1.27 (SD: 1.76) for the PDL plus imiquimod sites. The difference between groups was statistically significant (p=0.03) and favored combined treatment. Similarly, the mean change in overall color was 2.59 (SD: 1.54) for PDL plus placebo and 4.08 (SD: 3.39) for PDL plus imiquimod, p=0.04.

A prospective case series on combined laser treatment was published by Alster and Tanzi in 2009. The study included 25 patients who had incomplete clearance after at least 11 PDL treatments. (6) The patients received treatment with the Cynergy device (combination of 595 PDL and 1,064 Nd:YAG laser). Nineteen patients had port wine stains in a trigeminal location, and 6 had extremity involvement. Patients received a mean of 3.8 Cynergy treatments on the face and 4.9 on the extremities. Moderate clinical improvement (25-50%) was observed in 12 (48%) patients, and mild improvement (1-25%) was observed in 13 (52%) patients.

Summary

Studies have generally found that laser treatment can be effective at lightening port wine stains. The preponderance of evidence is on the pulsed dye laser (PDL); there is insufficient evidence from comparative studies that one type of laser results in more lightening than another. In terms of combination treatment, there is one small randomized controlled trial which found that treatment with PDL combined with a topical angiogenesis inhibitor is superior to PDL treatment alone. Additional studies evaluating specific combinations of treatments are needed before conclusions can be drawn about safety and efficacy.

Practice Guidelines and Position Statements
None identified.

Medicare National Coverage
No national coverage determination.

References

1. Faurschou A, Olesen AB, Leonardi-Bee J et al. Lasers or light sources for treating port-wine stains. Cochrane Database Syst Rev 2011; (11):CD007152.

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3. Babilas P, Schreml S, Eames T et al. Split-face comparison of intense pulsed light with short- and long-pulsed dye lasers for the treatment of port-wine stains. Lasers Surg Med 2010; 42(8):720-7.
4. Klein A, Szeimies RM, Baumler W et al. Indocyanine green-augmented diode laser treatment of port-wine stains: clinical and histological evidence for a new treatment option from a randomized controlled trial. Br J Dermatol 2012; 167(2):333-42.
5. Tremaine AM, Armstrong J, Huang YC et al. Enhanced port-wine stain lightening achieved with combined treatment of selective photothermolysis and imiquimod. J Am Acad Dermatol 2012; 66(4):634-41.
6. Alster TS, Tanzi EL. Combined 595-nm and 1,064-nm laser irradiation of recalcitrant and hypertrophic port-wine stains in children and adults. Dermatol Surg 2009; 35(6):914-8; discussion 18-9.

Billing Coding/Physician Documentation Information

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

Additional Policy Key Words

N/A

Policy Implementation/Update Information

10/1/88	New policy.
5/1/00	No policy statement change.
5/1/01	No policy statement change.
5/1/02	No policy statement change.
5/1/03	Policy archived.
5/1/05	Policy removed from archives. No policy statement change.
5/1/06	No policy statement change.
5/1/07	Policy statement changed from indicating this treatment is medically necessary to indicating this treatment is effective.
5/1/08	No policy statement changes.
7/15/09	The policy statement was clarified to indicate criteria for lesions for which the effective treatment would be considered medically necessary vs. cosmetic.
5/1/10	No policy statement changes.
5/1/11	Policy statement revised to include combined treatment as investigational.
10/1/11	Considerations section updated to add that individual consideration may be given to patients presenting with hemangiomas considered “complicated.”
5/1/12	Policy statement revised to include laser treatment of port wine stains (not limited to face and neck) in the presence of functional impairment to be medically necessary.
9/1/13	No policy statement changes.
3/1/14	No policy statement changes.