

## THE STURGE-WEBER FOUNDATION POSITION PAPER ON THE NECESSITY FOR THE TREATMENT OF PORT WINE STAINS

Port wine stains are congenital vascular malformations of capillary sized blood vessels. They should be included in the class of birth defects such as cleft lip, malformed ear, clubfoot or other anomalies present at birth. Port wine stains are not normal skin. The skin affected by a port wine stain contains a massive excess of small blood vessels. Although many port wine stains begin as light pink, flat lesions, they are not static. Port wine stains lead to overgrowth and dysfunction of the affected area. In severe cases this may be present at birth, but in most cases it is a progressive deformity. Severe venous varicosities, venous stasis, superficial blebbing, pyogenic granuloma, bleeding and ulceration can also be associated with the hypertrophy. Even small port wine stains develop progressive nodularity, blebbing and bleeding. Treatment is performed with a vascular specific pulsed dye laser. This laser is FDA approved for this indication.

The treatment of port wine stains is not cosmetic surgery. We do not deny that treatment does improve appearance, but it is not cosmetic surgery. The American Medical Association (AMA) distinguishes between cosmetic and **reconstructive surgery**. The AMA defines cosmetic surgeries as procedures "performed to reshape **normal body structures** in order to improve the patient's appearance or self esteem". In contrast, the AMA defines **reconstructive surgery** as being "performed on **abnormal structures** of the body caused by **congenital defects**, development abnormalities, trauma, infection, tumors or disease". It is clear the treatment of port wine stains meets the definition for **reconstructive surgery**. Port wine stains are not normal. They are **congenital abnormalities**. Thus, treatment of port wine stain is not solely for the purpose of altering appearance.

In summary, port wine stains are congenital defects that if left untreated develop sometimes severe functional problems. Effective, FDA approved treatment is available. Treatment of port wine stains is not cosmetic surgery.



# The Medical Necessity for Treatment of Port-Wine Stains

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**BACKGROUND.** Port-wine stains are congenital vascular malformations that can be disfiguring and may lead to psychosocial as well as medical complications. The 585-nm pulsed dye laser is very effective in treating port-wine stains. Laser treatment is often viewed by insurance companies as a "cosmetic procedure" and not "medically necessary." Consequently many patients are denied coverage for treatment of their disfiguring birthmarks.

**OBJECTIVE.** To determine variability of insurance coverage for laser treatment of port-wine stains from state to state. Natural history, progression, and potential complications of port-wine stains are reviewed and rationale for consistent insurance coverage for laser treatment of port-wine stains is given.

**METHODS.** A questionnaire was mailed to 40 dermatologic surgeons in 22 states and the District of Columbia. We reviewed the literature regarding port-wine stains and their potential complications, and health care policy guidelines regarding "medical necessity" and "cosmetic procedures."

**RESULTS.** Insurance coverage for laser treatment of port-wine stains varies from state to state.

**CONCLUSION.** Based on current health care policy guidelines, laser treatment of port-wine stains should be regarded, and covered, as a medical necessity by all insurance providers. © 1997 by the American Society for Dermatologic Surgery, Inc. *Dermatol Surg* 1997;23:663-667.

A port-wine stain is a congenital vascular malformation of mature capillaries. These birthmarks occur with an estimated frequency of 0.3% in the general population.<sup>1</sup> Usually present at birth as a light pink to red macule, port-wine stains most commonly occur on the face and neck and persist into adulthood. Due to progressive ectasia of the abnormal capillaries, port-wine stains tend to darken in color to a reddish-purple and may develop thickening or nodularity as the lesion ages.<sup>2,3</sup> Because these lesions most commonly occur on the face and can be very disfiguring, many patients seek treatment for port-wine stains.

In the past, many different treatments have been attempted including excision with skin grafting, cryotherapy, ionizing radiation, dermabrasion, and tattooing, none of which were particularly effective. The development of laser therapy for port-wine stains has been a significant breakthrough. Many studies have shown that the flashlamp-pumped 585-nm pulsed dye laser is very effective at lightening these lesions without scarring even in very young children. Many consider pulsed dye laser the treatment of choice for port-wine stains.<sup>4-7</sup>

Unfortunately, many patients are denied coverage for laser treatment of disfiguring port-wine stains.

Some insurance companies classify laser treatment for port-wine stains as a cosmetic procedure that is not medically necessary. Given the psychosocial implications and potential medical complications of this congenital malformation, and the fact that safe, effective, proven therapy is now available, treatment of port-wine stains should be considered as medically necessary and covered as such by insurance companies. To evaluate insurance coverage for treatment of port-wine stains, we conducted a survey of dermatologic surgeons in various parts of the United States.

## Methods

A questionnaire was mailed to 40 dermatologic surgeons in 22 states and the District of Columbia.

## Results

Table 1 summarizes the insurance coverage for port-wine stains in 18 states. A total of 23 surveys were returned, and all were utilized, giving a response rate of 58%. A total of 18 states were represented. None of the 18 states had a state law mandating insurance coverage for the treatment of port-wine stains. Of the 23 respondents representing 18 states, most stated that the majority of insurance providers in their state would cover laser treatment of port-wine stains, but determination was made on a case-by-case basis, with the majority requiring preauthorization. The percentage of requests approved for coverage varied from 50% to 100%. Ten

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Table 1. Eighteen-state Survey of Insurance Coverage for Laser Treatment of Port-Wine Stains

State	State Law	Do Most Insurance Co.'s Cover RX?	% Co.'s Requiring Preauthorization	% of Requests Approved	Covers Rx Only if Functional Problem Exists	Covers Rx Only if pt. <1 Year Old
AR	No	Yes	100%	—	Yes	No
CA	No	No	—	—	Yes	—
CA	No	Yes	100%	—	No	No
CA	No	Yes	100%	50%	Yes	No
CO	No	Yes	100%	75%	Yes	No
CT	No	Case by case basis	100%	—	Yes	No
FL	No	Yes	100%	—	—	—
IN	No	Yes	100%	70%	Yes	Occasionally
LA	No	Yes	100%	—	—	No
MA	No	No	100%	—	—	No
MO	No	Yes	—	85%	Yes	No
NC	No	Yes	100%	95%	No	No
NJ	No	*	—	—	—	—
NJ	No	Yes	100%	95%	No	No
NM	No	Yes	100%	80%	Yes	No
NY	No	Yes	70%, HMO's 100%	—	—	No
NY	No	Yes	<50%	—	No	No
NY	No	Yes	—	70%	Yes	No
SC	No	+	Many	—	—	—
TX	No	Yes	75%	100%	No	No
VA	No	Yes	None	—	No	No
WA	No	Yes	Most	—	No	No
WI	No	Yes	100%	95%	Yes	No

\* Small, private insurers deny more than major carriers.

† Often insurance denies coverage, especially adults. More receptive to younger patients.

respondents stated some insurance providers would cover treatment only if a functional impairment existed. One of the 23 respondents reported that insurance coverage for laser treatment of port-wine stains was restricted to patients less than 1 year of age.

## Discussion

### Insurance Coverage for Laser Treatment of Port-Wine Stains Varies from Company to Company and State to State

The flashlamp-pumped 585-nm pulsed dye laser offers safe and effective treatment for port-wine stains, however, insurance coverage for this treatment often varies between states and even within states. Some insurance companies view laser treatment as a noncovered cosmetic procedure and deny coverage to many people with disfiguring port-wine stains. While the 18 states represented in our survey had no state laws regulating treatment of port-wine stains, the state of Minnesota does. In 1993, after hearing heart-wrenching testimony from adults and children afflicted with port-wine stains and the emotional trauma they endure, the state of Minnesota passed a law that requires all regular health insurance, HMO plans, and Blue Cross and Blue Shield to cover the elimination or maximum feasible treatment of "port-wine stains." The bill initially was introduced

with mandatory coverage limited to children under 18 years of age. The age limitation was removed after hearing testimony from adults with port-wine stains about the social and employment discrimination they experience, that treatment later in life when it becomes a medical necessity is at a considerable cost, and that it is not merely a cosmetic problem but a serious medical condition. Given the psychosocial implications and potential medical complications of this congenital malformation, and the fact that safe, effective, proven therapy is now available, treatment of port-wine stains should be considered a medical necessity and covered as such by insurance companies. This coverage should be widespread and consistent within and between states.

### Natural History of Port-Wine Stains and Resultant Complications

Unlike other types of vascular birthmarks, port-wine stains do not fade with age. Initially a light pink to red macule at birth, the natural progression of a port-wine stain is to a deeper reddish-purple lesion that may become hypertrophic and nodular due to progressive vessel ectasia.<sup>2</sup> A review of 415 patients at New York University Medical Center revealed nearly two-thirds developed nodularity or hypertrophy of their port-wine stains by the fifth decade of life.<sup>3</sup> These mature lesions

are more prone to spontaneous or trauma-induced bleeding. This potentially could serve as a nidus for infection, which could be hazardous to a patient's health. Pyogenic granulomas have an increased tendency to occur within port-wine stains as they mature. A giant proliferative hemangioma arising in a port-wine stain has been reported.<sup>8</sup> Mature port-wine stains are more prone to bleeding and infection. Underlying hypertrophy of the involved area can occur even without any other extracutaneous manifestations. Hypertrophy can lead to asymmetric facies or extremities sometimes requiring orthopedic intervention. Progressive hypertrophy or modularity of a port-wine stain may block ear canals, nasal passages, field of vision, or closure of the mouth, resulting in interference with vital functions such as hearing, breathing, sight, and eating or talking.

While the natural progression of port-wine stains may lead to medical complications, the major morbidity of port-wine stains is psychosocial. Most port-wine stains occur on the face and neck. The value our society places on facial appearance is high and many studies have shown that physical attractiveness plays a major role in psychologic development and social interactions. Attractive persons are often judged to possess more desirable personalities, to be more intelligent, to be more successful in occupations and marriages, and to have a higher social status than others.<sup>9-11</sup> The negative psychosocial impact of a port-wine stain occurs throughout a person's lifetime. Port-wine stains affect parent's perceptions and interactions with their children from birth.<sup>12</sup> Affected school children are often ostracized by their peers. Schoolteacher's expectations and treatment of a child may also be affected by their physical attractiveness.<sup>10</sup> Affected individuals may experience feelings of stigmatization, embarrassment, anxiety and depression, and often have low self-esteem and impaired social interactions.<sup>12-15</sup> Port-wine stain patients may become reclusive, develop difficulties with speech and drooling, and have life adjustment problems. They may have difficulty finding employment. Psychologically, an affected individual must also deal with the progressive changes which occur in their port-wine stain as it matures. The progressive darkening and hypertrophy that a port-wine stain undergoes makes it increasingly difficult to camouflage with makeup.

#### *Pulsed Dye Laser Is Effective Treatment for Port-Wine Stains*

Because of the potentially devastating psychosocial effects, as well as the medical complications, port-wine stains should be treated. Laser therapy has been a major advancement in the treatment of these lesions. The ar-

gon laser used initially has been supplanted by the flashlamp-pumped 585-nm pulsed dye laser. Many studies have cited the effectiveness and safety of this therapeutic modality even in young children.<sup>4-6</sup> Early treatment in infancy is recommended by many because of increased effectiveness, smaller surface area of lesion, fewer number of treatments needed, and the avoidance of medical and psychosocial complications along with their higher treatment costs.

#### *Medical Necessity*

Even in view of the fact that a port-wine stain is a congenital malformation with many studies revealing major psychosocial as well as medical complications, many insurance companies view treatment of port-wine stains as a noncovered cosmetic procedure that is not medically necessary. This denies treatment to a large number of patients who must live with the psychosocial and medical complications of their disfiguring birthmark even though safe and effective treatment is available.

To determine whether something is medically necessary we must first define "medical necessity." Medical necessity is a vague term with no universally accepted definition among insurance companies, courts of law, or even the medical community. Yet reimbursement decisions based on this term are made on a daily basis. Without a universally accepted definition, individual health insurance plans selectively determine what defines "medical necessity" and therefore what services they will cover for their plan subscribers. Medicare's definition of "medical necessity," defined by the Social Security Act, Section 1862(a)(1)(A), states that "items and services which are not reasonable and necessary for the diagnosis or treatment of illness or injury, or to improve the functioning of a malformed body member" are not medically necessary and are excluded from coverage.<sup>16</sup> Medicare also excludes medically unproven, experimental, or investigational procedures.<sup>17</sup> Medicare has no universal regulations, however, to help determine what constitutes "medically necessary" treatment. Medicare contracts with independent insurance contractors locally to determine what defines medical necessity and therefore payment for services.<sup>18</sup> Medicare coverage therefore may vary from state to state. In an attempt to improve consistency in contractor coverage decisions, the Health Care Financing Administration states its interpretation of "reasonable and necessary" to mean that an item or procedure is safe and efficacious as well as widely accepted by the medical profession.<sup>18</sup> State administered Medicaid programs likewise have no nationally accepted definition of what constitutes "medically necessary care." Congress has neither a definition of or a procedure for determining what is

"medically necessary".<sup>19</sup> Therefore, every state may have a different determination of what constitutes "medically necessary" care. Private insurance providers also have varying definitions of "medical necessity" leading to variable coverage of services from plan to plan. Many insurance providers have adopted similar guidelines for the determination of "medical necessity" as those of Section 1862(a)(1) of the Social Security Act. That is, to be considered medically necessary under the Act, items and services must be: 1) consistent with the symptoms or diagnosis of the illness or injury under treatment; 2) necessary and consistent with generally accepted professional medical standards (ie, not experimental or investigational); 3) not furnished primarily for the convenience of the patient, the physician or supplier; and 4) furnished at the most appropriate level that can be provided safely and effectively to the patient.<sup>22</sup>

### *Cosmetic Surgery*

Many insurance companies classify treatment of port-wine stains as a "cosmetic procedure." As with "medical necessity," there is no universally accepted definition of a "cosmetic procedure." This also leads to variable insurance classifications and coverage for certain services from plan to plan. The AMA Policy Compendium defines cosmetic surgery as that which is performed to reshape normal structures of the body in order to improve the patient's appearance and self-esteem, and reconstructive surgery as that which is performed on abnormal structures of the body, caused by congenital defects, developmental abnormalities, trauma, infection, tumors, or disease. It is generally performed to improve function, but may also be done to approximate a normal appearance.<sup>19</sup> Medicare's definition of a cosmetic surgery includes any surgical procedure directed at improving appearance, except when required for the prompt repair of accidental injury or for the improvement of the functioning of a malformed body member.<sup>20</sup>

### *Rationale for Insurance Coverage for Laser Treatment of Port-Wine Stains*

Unfortunately there are no adequate, easily applicable standard definitions for "medical necessity," or "cosmetic vs reconstructive procedures" that can be used by all health care providers. However, based on the current guidelines and definitions in use, the treatment of port-wine stains with the 585-nm pulsed dye laser should be regarded, and covered, as a medically necessary noncosmetic procedure by all insurance providers. Based on the AMA Compendium, laser treatment of port-wine stains is not a cosmetic procedure, but is a reconstructive procedure: a port-wine stain is an abnor-

mal structure of the skin caused by a congenital defect, and pulse dye laser treatment is used to approximate a normal appearance and, in some cases, may be needed to improve function if the port-wine stain is interfering with a vital function or is causing asymmetry and distortion. Using Medicare's definition of "medical necessity" and the Health Care Financing Administration's interpretation of a "reasonable and necessary" procedure, laser treatment of port-wine stains again fits the criteria. The 580-nm pulsed dye laser has been shown to be safe, efficacious, and widely accepted by the medical profession for the treatment of port-wine stains. It is a proven treatment that is neither experimental or investigational. Viewed as the current treatment of choice, laser therapy is both reasonable and necessary in the treatment of port-wine stains and, as stated above, may improve functioning, or may actually prevent malfunctioning and medical complications of a body member in the first place. Many insurance companies provide coverage for the repair of cleft lip even if there is no underlying cleft palate or other associated functional defect. A port-wine stain is analogous to a cleft lip. They are both congenital malformations, not within the normal range, that can have devastating psychosocial complications. Unlike a cleft lip, which is relatively static, however, a port-wine stain progressively darkens and thickens with age with the possibility of resultant complications discussed earlier. Breast reconstruction immediately following a mastectomy is considered by Medicare and many other providers as a relatively safe and effective noncosmetic procedure. This procedure is appropriately covered for these psychologically devastated women, but only to approximate a normal appearance and not to improve function. The treatment of port-wine stains should likewise be covered.

A determination of medically necessary care should carefully consider two competing interests: the insured's interest in obtaining the medical treatment and the insurer's interest in maintaining the financial integrity of the company.<sup>21</sup> Preventive medical care is cost effective. Insurance companies are willing to provide coverage for psychotherapy and medications needed for depression, low self esteem, social dysfunction, etc or therapy of medical complications due to port-wine stains. However, many do not provide coverage for early treatment of port-wine stains to prevent complications and suffering when treatment is most effective and least costly. Early treatment of port-wine stains will save health care dollars in the long run. Treatment when lesions are small and flat takes fewer treatment sessions and is more effective in clearing the lesion. This would also save on the treatment of potential medical and psychosocial problems arising as the lesions mature, not to mention probably the most significant and worthwhile savings of all—the removal of psychosocial

stigma an affected individual endures for a lifetime. The adage of "pay a little now, or pay a lot later" applies to the treatment of port-wine stains.

Port-wine stains are progressive lesions with potentially devastating psychosocial and medical complications. It is for these reasons, treatment of port-wine stains is a medical necessity. Widespread and consistent insurance coverage should exist, not just for some, but for all.

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# The Necessity for Treatment of Childhood Port-Wine Stains

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The pulsed tunable dye laser has recently become available for the treatment of children with port-wine stains. Parental reactions and psychosocial implications of untreated port-wine stains in children are reviewed. It is recommended that children with port-wine stains be treated as early as possible to prevent adverse sequelae to their psychological development.

**T**he tunable dye laser has advanced the treatment of port-wine stains. Recent studies have demonstrated significant lightening of the lesions with minimal scarring or epidermal changes following treatment with the tunable dye laser.<sup>1-3</sup> This form of treatment has been recently applied to children with port-wine stains. Tan et al<sup>4</sup> treated children ranging in age from three months to fourteen years with the flashlamp pulsed tunable dye laser. After an average of 6.5 laser treatments to the lesions, all of the children had complete clearing of the port-wine stains. The availability of this treatment has significant implications for the psychosocial development of children with port-wine stains.

Prior to the introduction of laser technology, there was no consistently effective treatment for port-wine stains. In most cases, this congenital vascular malformation progressed over time from a pale pink macule at birth to a dark red-purple raised lesion by middle age.<sup>5</sup> For parents who have sought treatment for their children's port-wine stains, cosmetic makeup has been frequently recommended by physicians to cover the lesions.<sup>6-8</sup> It has also been suggested that parents refer to the child's port-wine stain as a "beauty mark" instead of a birthmark.<sup>9</sup> These recommendations have minimal utility for parents who are concerned about the cosmetic aspects of the port-wine stains.

## Reaction of Parents to Children with Port-Wine Stains

There have been no systematic investigations of parents' views of port-wine stains or the effect of the port-wine stain on child-parent interactions. Adults with port-wine stains have been interviewed about their views of their parents' reactions to their birthmarks. One man reported that his mother insisted on putting makeup on his face before he went out to play. His mother was oversolicitous and did not allow him to participate in many activities. A woman reported that when she was born with a port-wine stain, her father thought she should be protected and hidden. However, her mother inter-

vened and encouraged her to become involved with others.<sup>10</sup>

A parent's reaction to a child born with a port-wine stain may consist of two major components, both of which will affect the parent-child relationship. One aspect is the acute awareness of potential social difficulties the child will encounter. The port-wine stain singles the child out from other children and may serve as a focus of negative comments by other children. A parent who anticipates this situation may develop a style of interaction with the child ranging from overprotection to the encouragement of fierce independence. The parent may shelter the child from life experiences or encourage the child to stand independently in the face of adversity. Either of these extremes are attributable to the effect of the port-wine stain on the relationship between the parent and the child.

Another more subtle component, perhaps unconscious, that affects the interaction with the child is the degree of the parent's narcissistic vulnerability. Since children are direct biological products of parents, parents generally have a narcissistic investment in their children. For parents, children serve as a direct reflection of themselves. In the case of a child with a port-wine stain, there is a conspicuous defect present from birth. If a parent's self-esteem (narcissism) is intricately tied to the attractiveness of the child, the presence of the port-wine stain will be a traumatic situation. The parent may handle this dilemma by emotionally distancing himself from the flawed child. That way, the child is no longer a direct reflection of the parent. Another solution, although opposite, is for the parent to minimize the port-wine stain. If the reality of the port-wine stain is denied, the parent's self-esteem can remain intact. This solution, however, compels the child to avoid discussion of the port-wine stain with the parent. The child's opportunity to obtain coping skills from the parents is limited by this approach.

## Reaction of Children to Port-Wine Stains

There have been no research studies focused on children's reactions to port-wine stains. A recent study involved adults with port-wine stains who were interviewed about their childhood experiences.<sup>10</sup> The sample was divided into adults with large port-wine stains and adults with small ones. As children, the group with large port-wine stains were acutely aware of the reaction of others to this birthmark. Most of them were teased at school by other children and often found themselves asked intrusive questions by strangers. One person with an extensive port-wine stain admitted that he was treated as if he had psychiatric problems. As children, they experienced sadness, withdrew from interactions, and tended to comment about the birthmark before being asked inevitable questions. From a psychological perspective, the situation was similar for those adults with small port-wine stains. As children, they were self-conscious about the birthmark even if no comments were directed at them. They believed that other people noticed the stain but chose not to comment on it. They felt embarrassed interacting with strangers. Attention from their families was directed at covering up the port-wine stain.

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## Psychosocial Implications of Port-Wine Stains

An important consideration regarding the psychosocial implications of the port-wine stain is its location. Most port-wine stains occur primarily on the face and neck. This is unfortunate since it is the face that a child presents to the social environment. Our society places emphasis on the affectionate description of children's faces through the use of terms such as adorable, cute, or cherubic. It is significant that there is no endearing term to describe the faces of children with port-wine stains.

A number of studies provide evidence for the importance of the face in psychological development. Of a list of twenty-four body characteristics (including items such as chest, legs, weight, body build), adolescents selected facial complexion as the most important physical trait.<sup>11</sup> Facial appearance was rated by adolescents as a significant predictor of self-concept for girls and effectiveness for boys.<sup>12</sup> College students rated the face as more strongly correlated with sexuality than body build.<sup>13</sup> Moreover, there is evidence that personality determinations may be based on the face of a person.<sup>14</sup> In one study, attractive persons were judged to possess more desirable personalities and were predicted to have better marriages, occupations, and social status than less attractive persons.<sup>15</sup> In an epidemiologic survey of children with chronic dermatologic disorders affecting their appearance, afflicted children were judged to have behavioral problems, social isolation, and psychological problems.<sup>16</sup>

The previously cited studies demonstrate the emphasis that is placed on facial appearance during childhood and adolescence. A child's view of his degree of physical attractiveness has impact on his self-concept and self-esteem. For a child with a port-wine stain, there is often a pronounced facial defect. This defect will be incorporated into the identity of the developing child. A potential outcome is the occurrence of feelings of low self-esteem and self-worth. This negative self-concept may subsequently interfere with the development of peer relationships. Overall, the child may feel insecure and isolated by others.

There are other critical areas of development that may be adversely effected for the child with a port-wine stain. It has been clearly demonstrated that physical attractiveness can affect teacher expectations of a child. In one study, teachers were given a standardized fifth grade report card and an attached photograph of either an attractive or an unattractive child. Despite identical information about the children's scholastic and social performance, the attractive children were judged by the teacher to be more intelligent, more popular with peers, more likely to progress in school, and to have parents with more interest in education than the less attractive children.<sup>17</sup> It is well known that teacher expectation can have a significant impact on a child's academic performance. Given the findings of this study, children whose appearance is marred by a port-wine stain may have academic limitations imposed upon them based on misconceived teacher expectations.

Similarly, children's physical attractiveness has been related to the evaluation of their transgressions. A transgression committed by an attractive child is characteristically judged as less socially undesirable than the same act committed by an unattractive child. Furthermore, unattractive chil-

dren who transgressed were viewed as more dishonest and more unpleasant than attractive children in the same circumstance.<sup>18</sup>

## Conclusion

Port-wine stains have the potential for lasting detrimental effects on children's psychological, social, interpersonal, and cognitive development. In addition, children with port-wine stains may find it difficult to develop positive relationships with their parents.

The availability of the pulsed tunable dye laser for treatment of port-wine stains in childhood is a psychosocial windfall to both children and their parents. From a psychological perspective, it is recommended that children with port-wine stains be treated at the earliest possible age to reduce the likelihood of permanent adverse sequelae to their development.

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# The Medical Necessity of Evaluation and Treatment of Port-Wine Stains

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LASERS

*New lasers and improved laser delivery systems have allowed for the safe and effective treatment of port-wine stains in patients of all ages. The satisfactory results obtained by laser treatment have increased the number of patients seeking consultation regarding their birthmarks. It is imperative that physicians recognize the various medical syndromes and problems associated with port-wine stains. A review of 415 patients with facial port-wine stains has revealed hypertrophy and/or nodularity in 65% of patients by the fifth decade of life, which increases significantly the morbidity of these lesions. It is believed that laser treatment will minimize the medical and psychologic complications that result from the natural evolution of port-wine stains. J Dermatol Surg Oncol 1991; 17:76-79.*

**R**ecent developments in the treatment of port-wine stains have allowed physicians to re-evaluate the opportunity and necessity for treatment of these lesions. Port-wine stains occur in 0.03% of the general population and are noted to occur most commonly on the face over the areas of the first and second trigeminal nerves.<sup>1,2</sup> In the past several years, the development of the flashlamp-pumped pulsed-dye laser, continuous-wave tunable-dye yellow-light laser, copper-vapor laser and the use of other continuous-wave lasers in conjunction with special scanning devices, have all led to improved results in the treatment of these lesions.<sup>3-6</sup> One of these lasers (the flashlamp-pumped pulsed-dye laser) has even allowed for the treatment of infants and young children.<sup>7</sup> Thus, physicians and patients afflicted

with this congenital anomaly are faced with the decision of whether or not to seek treatment with any of these lasers, where previously treatment may have been unsatisfactory or contraindicated.

The superior results now offered by lasers in the treatment of port-wine stains have also allowed for the influx of patients with this congenital malformation to seek medical attention. Thus, it is important that those physicians consulting these patients recognize the associated medical problems that may be found with port-wine stains. One important example is the Sturge-Weber syndrome, which is commonly found among those patients with port-wine stains involving the distribution of the first branch of the trigeminal nerve.<sup>8-10</sup> The Sturge-Weber syndrome has a high incidence of congenital glaucoma of the ipsilateral eye,<sup>11</sup> calcification and vascular anomalies of the brain, associated seizure disorders, and, in some cases, mental retardation.<sup>8-10</sup> Ophthalmologic and neurologic evaluation in these patients are recommended early in life. Other congenital syndromes associated with port-wine stains include the Klippel-Trenaunay-Weber syndrome with the associated hypertrophy of skeletal tissue,<sup>12</sup> as well as the Cobb syndrome, which may be a sign of underlying arteriovenous malformation of the spinal cord.<sup>13,14</sup> It is also not uncommon to see a port-wine stain overlying an arteriovenous malformation, arterial malformation, or venous malformation,<sup>15,16</sup> therefore requiring the physician to look beyond the skin for any underlying problem.

Port-wine stains begin as macular lesions and become progressively ectatic over time.<sup>17,18</sup> This progressive ectasia can result in significant darkening of the lesion as well as the development of nodularity and occasional tumors. A review of 415 patients presenting at the laser section of the Skin and Cancer Unit of the New York University Medical Center has revealed that approximately two-thirds of all patients with port-wine stains develop nodularity or hypertrophy in the fifth decade of life (Table 1).

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Table 1. Percent of Patients Showing Thickening or Hypertrophy of Port-Wine Stains in Relation to Age

Age (y)	N	Patients showing hypertrophy or thickening (%)
≥ 0 ≤ 14	197	0
≥ 15 ≤ 25	72	15.3
≥ 26 ≤ 35	80	32.5
≥ 36 ≤ 45	34	35.3
≥ 46 ≤ 55	20	65.8
≥ 56 ≤ 65	9	66.7
≥ 66 ≤ 75	3	66.7

These changes are gradual in their development, with the mean age of hypertrophy noted to be 37 years. The progression of the port-wine stain from its macular stage to the hypertrophic stage may take place at any point in the development of an individual's lifetime; however, the most dramatic changes are noted as the patient has become older (Figures 1 and 2). Associated with this nodularity and hypertrophy is the risk of spontaneous bleed-

Figure 1. Port-wine stain in a 15-year-old girl.

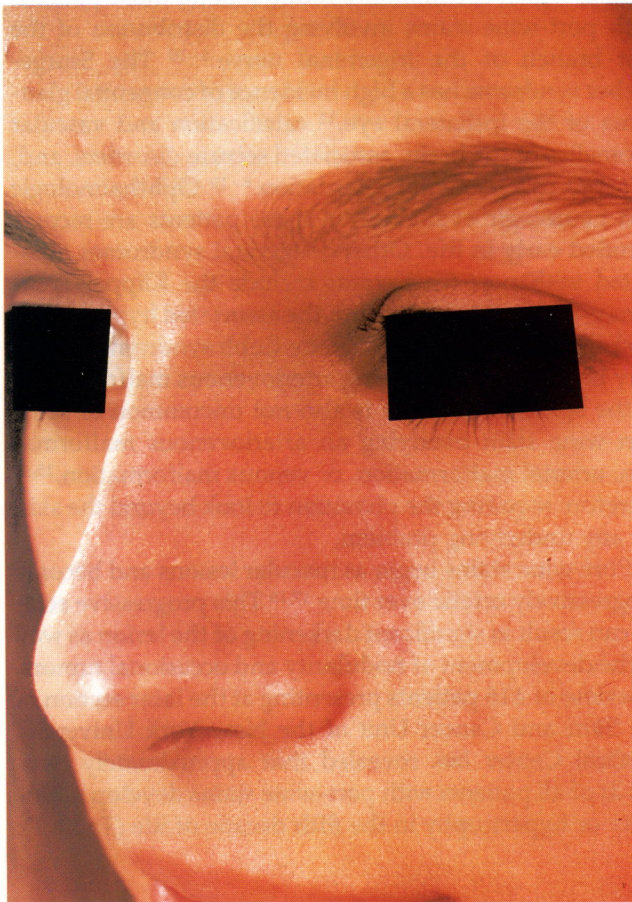


Figure 2. Facial thickening and nodularity noted 20 years later in the patient from Figure 1.

ing and hemorrhaging upon injury to the affected area. Although the exact risks have not been established in the literature, they are clearly associated with the degree of hypertrophy and the location of the lesion. Men with port-wine stains on the face in the bearded area run significant risks of excessive bleeding upon shaving, as would a woman with a port-wine stain in the hair-bearing area where epilation is desired. The development of hypertrophy or nodularity in a periorbital area may inhibit the visual field (Figure 3) and, similarly, these changes around the nose or mouth may also interfere with breathing or eating.

It has become clear that treatment of a port-wine stain in its macular stage will prevent the development of the hypertrophic component of the lesion. Postoperative biopsies upon laser treatment of a port-wine stain reveal that the existing blood vessels are smaller and fewer in number compared with pretreatment biopsies.<sup>19</sup> Thus, the opportunity for the progression of these lesions to a more ectatic state is less likely to occur. Not uncommonly, the development of pyogenic granulomas (Figure 4) in



Figure 3. Hypertrophy and nodularity seen in the seventh decade of life.

patients with port-wine stains can occur and these pyogenic granulomas have the potential to also bleed spontaneously. In our experience, the incidence of pyogenic granulomas is significantly lower in those patients who have been treated with laser therapy.

The psychological problems associated with a congenital birthmark of this type may have short- and long-term implications for the patient. Detailed studies have demonstrated lower self-esteem and problems with social interaction for both the patient and family alike.<sup>20-22</sup> Successful treatment of a port-wine stain with any modality helps minimize psychological problems that occur as a result of the presence of the port-wine stain. Although the treatment of these lesions has improved dramatically over the past several years with the development of newer techniques and lasers, many patients are denied the opportunity for treatment of these lesions by the unfortunate classification of this treatment as a cosmetic procedure. It has been demonstrated clearly that these safe treatments can minimize the chance of nodularity, spontaneous bleeding, and the development of pyogenic granulomas. The psychologic benefits of removing port-wine stains in these patients are of considerable value to their mental health and may, in fact, obviate the need for

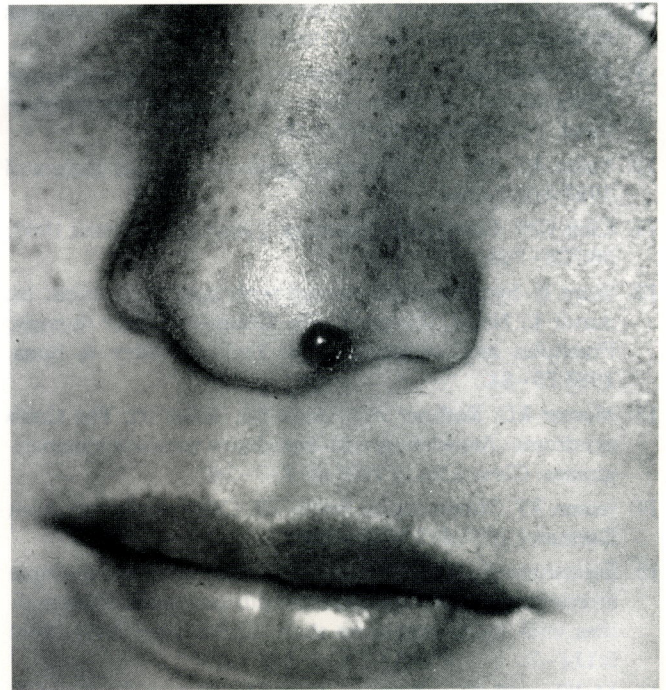


Figure 4. Pyogenic granuloma arising within a port-wine stain on the nose of a 12-year-old boy.

psychological counseling at a later date. It is incumbent upon those physicians who evaluate patients with port-wine stains and insurance carriers who evaluate their claims to realize the medical benefits in the treatment of these lesions. Failure to recognize the treatment of port-wine stains as a medical necessity may, in fact, be harmful to the medical and psychologic well-being of the patient.

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