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# Vascular Birthmarks Foundation

## Facts About Vascular Birthmarks and Tumors

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## NOTES

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**Klippel-Trenaunay Syndrome**

- ❑ KT Foundation  
P.O. Box 205  
Lakeview, NC 28350  
Phone: 910-949-3054  
Email: [ktfoundation@earthlink.net](mailto:ktfoundation@earthlink.net)  
<http://www.ktfoundation.com>

**Other Online Organizations and Support Groups**

- ❑ Anomalie Vasculaire – French language site  
<http://www.anomalievasculaire.org>
- ❑ Cutis Marmorata Telangiectatica Congenita  
[http://www.cmtc.nl/HTML/eng\\_start.php](http://www.cmtc.nl/HTML/eng_start.php)
- ❑ Port Wine Stain Support Group  
<http://www.birthmarks.com/community.cfm#Secure>
- ❑ Sturge-Weber Syndrome Community  
Discussion Group  
<http://swscommunity.org/forum/index.php>
- ❑ Sturge-Weber Syndrome Community Parent  
Support Group – MSN Groups  
<http://groups.msn.com/SturgeWeberSyndromeCommunitySupport>
- ❑ Vascular Birthmarks Foundation Discussion Group  
<http://www.birthmark.org/board/index.php>
- ❑ Vascular Birthmark Support Group – MSN Groups  
<http://groups.msn.com/vascularbirthmarksupport>

# Vascular Birthmarks, Tumors and Related Syndromes

## Foundations and Organizations

**For more information on the conditions referenced in this publication, you may contact these foundations and organizations, or visit their websites.**

### Vascular Anomalies

- ❑ The Vascular Birthmarks Foundation  
P.O. Box 106  
Latham, NY 12110  
Phone: 877-823-4646  
Email: [HVBF@aol.com](mailto:HVBF@aol.com)  
<http://www.birthmark.org>

### Port Wine Stains

- ❑ Birthmarks.com  
5104 Beverly Hills Drive  
First Floor  
Austin, TX 78731  
Email: [editor@birthmarks.com](mailto:editor@birthmarks.com)  
<http://www.birthmarks.com>

### Sturge-Weber Syndrome

- ❑ Sturge-Weber Syndrome Community  
P.O. Box 24890  
Lexington, KY 40524-4890  
Phone: 859-272-3857  
Email: [swsc@swscommunity.org](mailto:swsc@swscommunity.org)  
<http://swscommunity.org>

## Arteriovenous Malformations

- ❑ Arteriovenous Malformations (AVM) are usually present at birth, but are sometimes not noticed until later in life. Sometimes they do not appear until adulthood. AVM's can also be acquired following direct trauma to the affected area.
- ❑ AVM is a complex mass of arteries and veins. Defective blood flow has been associated with these lesions. As the lesion ages, the vessels enlarge and thicken to compensate for the increased blood supply. There are two grades: low and high. Low grade grow slowly with the child, and high grade expand rapidly - growing faster than the child until the lesion may eventually become life threatening.
- ❑ An AVM is a firm compressible mass. Common sites are the lips and other head and neck areas. Mixed malformations include a combination of two or more vessel types (like arterial and venous vessels).
- ❑ The brain is also a common site for these lesions and they are almost always misdiagnosed as hemangiomas. AVM's may also occur in the brain stem or spinal cord.
- ❑ Symptoms associated with AVM in the brain can include headache, seizures, neurological problems, hemorrhage, paralysis, and loss of speech, memory or vision.
- ❑ AVM can often be differentiated from other vascular lesions by compressing the lesion and feeling for a pulse. The lesions usually feel soft and gel-like. Patients refer to feeling a heartbeat in the lesion and sometimes complain of severe pain in the affected area.

### **Arteriovenous Malformations continued**

- Treatment should only be attempted by a skilled surgeon. Removing an AVM is complicated and unless the Nidus is removed, the lesion will re-grow. The optimum treatment is a combination of embolization and surgical excision.
- Angiography is typically used to detect AVM.

### **Cutis Marmorata Telangiectatica Congenita**

- Cutis Marmorata Telangiectatica Congenita (CMTC) is a rare condition that affects blood vessels of the skin.
- CMTC is present at birth or soon after.
- Most infants are born premature, yet are large at birth (9 pounds or more). Most have large heads, especially the forehead (known as frontal bossing). Growth decreases by age two, and then most children affected are small for their age.
- Enlarged blood vessels show through the skin giving it the appearance of being marbled or blotchy (also referred to as looking mottled or having a fishnet appearance.)
- Blotchy patterns on the skin can increase, or veins beneath the skin can become more visible, when crying.
- Usually only part of the body is affected, or asymmetric, being that one side is more dramatically affected than the other (such as the head, face or limbs).

## **Resources**

## Surgery

- ❑ Surgery to remove a vascular lesion must be done by an expert. These lesions are very vascular and, therefore, prone to bleeding. Experts use a thermal scalpel that cauterizes as it cuts the tissue, but some also use a Bovie knife – which is a very precise and sharp knife that cuts very fine lines.
- ❑ Some surgeries are done in stages. These “serial” procedures are sometimes needed to allow good tissue to regenerate, so that future procedures can leave the most acceptable result. Some experts can remove a facial hemangioma in one procedure. Larger, more complicated hemangiomas and malformations, often take multiple procedures.
- ❑ Surgery to remove a vascular lesion should be utilized when the “skin expander effect” is maximized. The “good, unaffected tissue” often expands when a hemangioma or other focal vascular lesion is present and growing. This skin expander effect actually gives the surgeon good tissue to use for closing the skin once the lesion has been removed. There is an optimal time to take advantage of the skin expander effect. If a hemangioma is left to regress on its own, past the optimum time for using the skin expander effect, surgery could be more extensive to get a good cosmetic outcome. As soon as a hemangioma begins to regress, you begin to lose the skin expander effect. So, it is important to seek removal at the optimal time, if in fact, the lesion will leave a deformity that will eventually need to be removed.

## **Cutis Marmorata Telangiectatica Congenita continued**

- ❑ Symptoms vary from case to case, and are usually mild. In some cases sores can develop, as well as hypertrophy, hypotrophy, and problems with the teeth and vision. Developmental and speech delays can also be present.
- ❑ Marbling can reduce over time, but this is not always the case. The marbling pattern usually fades on its own, and no special treatment is needed.
- ❑ Other characteristics of CMTC can include wide-spaced and deep-set eyes, full cheeks, puffy hands and feet, wide space between first and second toes, fused toes, umbilical hernia, loose or stretchy skin, and skin that is very soft to the touch. In addition, glaucoma can be present, as well as hydrocephalus, enlarged organs, cysts, kidney blockage, epilepsy, and asthma.
- ❑ It is very difficult to differentiate CMTC from Klippel-Trenaunay syndrome. Experts agree that the conditions are likely “cousins” because they share many common symptoms or descriptors.
- ❑ X-rays, Computerized Tomography (CT), Magnetic Resonance Imaging (MRI), are used to detect and treat CMTC.

## Glaucoma

- ❑ There are 4 main types of glaucoma: primary open angle glaucoma (the most common), primary closed angle glaucoma, secondary open angle and secondary closed angle glaucoma, and developmental glaucoma.
- ❑ Primary open angle glaucoma (POAG), or chronic glaucoma, is the most common form of the disease. POAG develops slowly over time and is not painful. Most patients are unaware of a problem until permanent damage has been done. The drainage canals in the eye become clogged over a period of time, causing inner eye pressure to rise. This condition requires a lifetime of regular monitoring and treatment.
- ❑ Primary closed angle glaucoma (PCAG), or acute glaucoma, is more common in people of Chinese or Japanese descent. PCAG develops suddenly and can be very painful. Eye pressure can rise very swiftly, as the drainage canal is blocked or covered. In this condition, the iris and cornea is not as wide as it should be, resulting in the outer edge of the iris blocking the drainage canal when the pupil enlarges too much or very quickly. If immediate treatment is given, vision may be completely recovered, but if treatment is delayed permanent damage can occur.
- ❑ Secondary open angle and secondary closed angle glaucoma are caused by conditions secondary to primary conditions of the eye. The conditions of both the primary cause and the secondary cause may allow vision to return to normal, and need no further treatment. In some cases the eye may be permanently damaged and require lifetime treatment.

## Sclerotherapy

- ❑ Sclerotherapy is a non-surgical procedure where a chemical is injected into a vascular malformation using a small needle. The chemical is known as sclerosant (usually alcohol or hypertonic saline).
- ❑ More than one injection may be needed, depending on the size and shape of the malformation.
- ❑ The procedure generally takes about three hours to perform, and is most often performed under general anesthesia.
- ❑ The effects of sclerotherapy may not be seen for up to two months. Swelling or bruising usually occurs, and then decreases over the first week. However, it may take up to two months for these symptoms to completely diminish. Skin inflammation can also occur.
- ❑ Generally, aspirin and medications containing aspirin should not be taken prior to treatment, as aspirin can increase the risk of bleeding.
- ❑ Following the procedure, patients can resume normal activities, however, strenuous activity should be avoided for at least a week, or period of time specified by a doctor.



## Laser Surgery

- ❑ Light Amplification by the Stimulated Emission of Radiation, or Laser surgery, uses light beams to treat the skin.
- ❑ There are different types of lasers. The most commonly used lasers for vascular malformations are the pulse-dye laser, the ND: YAG laser, the CO<sup>2</sup> (carbon dioxide), argon laser, and Diode laser. Each laser works differently.
- ❑ Each layer of the skin reacts differently to the light beam of the laser.
- ❑ Treatment can be somewhat painful, and has been compared to the snapping of a rubber band. Sometimes general anesthesia is used. Many of the newer lasers have a cooling agent incorporated to reduce discomfort. There are also numbing creams available to help manage pain and discomfort associated with laser treatment.
- ❑ The number of treatments can vary, due to the thickness, location, and type of vascular anomaly.
- ❑ It is recommended to avoid exposure to sunlight, and the taking of aspirin prior to receiving laser treatment.
- ❑ Antibacterial ointments are used following laser surgery to help prevent infection of the treated area. After a couple of days, the skin can be cleansed normally, and sunscreen should be applied to the area.

## Glaucoma continued

- ❑ Secondary conditions can include trauma to the eye, inflammation, tumor, cataract, diabetes, or can be caused by the use of certain drugs like steroids.
- ❑ Developmental glaucoma is a rare condition present at birth. This condition occurs when the eye has failed to form correctly, or due to other developmental eye abnormalities. Surgery and eye drops are used to treat this type of glaucoma.

## Hemangiomas

- ❑ Approximately 30% of all hemangiomas are visible at birth. The remaining 70% become visible within one to four weeks after birth.
- ❑ Hemangiomas occur 5 times more often in females than in males and occur predominantly in Caucasians. Low birth weight infants (less than 2.2 pounds) have a 26% chance of developing a hemangioma.
- ❑ Approximately 83% occur on the head and neck area. The remaining 17% appear throughout the rest of the body (both externally and internally). In the early stages some appear either as bluish or reddish spots or flat patches. Rarely is a hemangioma fully grown at birth.
- ❑ Hemangiomas that are flat and appear reddish in color are called "superficial", and those that are deep beneath the skin and appear bluish in color are called "deep" hemangiomas. When a hemangioma is both deep and superficial it is called a "compound" hemangioma.

## Hemangiomas continued

- ❑ Hemangiomas can grow for up to 18 months and then begin a long slow regression known as involution. This involution can last from 3 - 10 years. While all hemangiomas eventually involute, the result is not always cosmetically acceptable. Early intervention has been shown to reduce the need for corrective surgery after involution has occurred; or to, at least, minimize extensive corrective surgeries in the future.
- ❑ In some cases, hemangiomas can be life threatening or severely problematic (interfering with eating, breathing, seeing, hearing, speaking, etc.) and require immediate, aggressive intervention.
- ❑ Secondary clotting can also occur in a large hemangioma, resulting in the trapping and destruction of blood platelets.
- ❑ Hemangiomas that grow internally can be very dangerous. They are difficult to detect and when they are detected, the infant is often in need of immediate intervention. Internal hemangiomas (referred to as visceral) occur in the liver, intestines, airway and brain. Infants who have what is referred to as hemangiomatosis (multiple hemangiomas) are suspect for internal lesions.
- ❑ When an infant has more than 3 hemangiomas, an ultrasound should be done of the entire body to rule out internal lesions. Jaundice may be a sign of liver hemangiomas, blood in the stool may be a sign of hemangiomas on the intestines, and stridor (croupy cough and difficulty breathing) may be a sign of airway hemangiomas.

## Embolization continued

- ❑ Embolization combined with surgery is becoming the ideal approach to massive vascular lesions. The Interventional Radiologist blocks the blood from feeding the lesion and then the surgeon goes in and removes the lesion.
- ❑ Only an expert skilled in this type of treatment of vascular lesions should perform these procedures.

## Hydrotherapy

- ❑ Hydrotherapy is the use of water to treat various symptoms. Steam, ice, and cold or hot water can be used in conjunction with wraps and other devices and techniques.
- ❑ Hydrotherapy helps stimulate circulation and can aid in strengthening muscles.
- ❑ Swimming is very useful in the treatment of Klippel-Trenaunay syndrome, and can help reduce the stress and trauma on limbs experienced during traditional exercises.

### **Seizure Medications continued**

- Patients on seizure medications will need to have periodic blood testing to determine that the correct level of medication is in the patient's system.
- Depending on the type of medication used, varying side effects can occur. Please read all instructions carefully, paying attention to side effects, allergic reactions, and conflicts with other medications you might be taking.

### **Embolization**

- Embolization is a non-surgical procedure used to treat certain vascular malformations.
- Small blood vessels are clogged, blocking the flow of blood. A catheter is inserted through the femoral artery or vein (upper leg) into an abnormal artery or vein, where tiny foam-like particles are then injected. The blood vessel closes, thus preventing blood flow into the malformation.
- The procedure generally takes around three hours, and isn't considered painful. It is usually performed under general anesthesia.
- Following embolization, patients can resume normal activities. However, strenuous activities should be avoided for at least a week, or length of time recommended by a doctor.
- While embolization is considered safe, there can be side effects. Some side effects can include bleeding, bruising, clotting of an artery, or damage to normal tissue.

### **Hemangiomas continued**

- Multiple hemangiomas are not a concern for alarm if the infant is otherwise healthy. If problems are going to appear they usually occur within the first 4 or 5 months.
- Some internal lesions, like external lesions, require no intervention, as they will shrink in time. If the lesions are problematic, they can be treated with drug therapy, such as Vincristine or steroids.

### **Kasabach-Merritt Syndrome**

- Kasabach-Merritt syndrome (KMS) is sometimes referred to as Thrombocytopenia.
- KMS is a rare life threatening disease
- In KMS, a vascular tumor traps and destroys platelets (platelets in the blood help with clotting). The types of tumors associated with KMS are usually tufted hemangioma or hemangioendothelioma.
- Children less than 6 months old are usually affected.
- The hemangioma-type tumor is often found in the internal organs, head and neck area, or in the extremities. The tumor grows, trapping the platelets, thus decreasing the body's ability to clot. Spontaneous bleeding can occur, and the result can be fatal.
- There is no set treatment for KMS, and patients are often monitored in the hospital until the condition stabilizes.

### **Kasabach-Merritt Syndrome continued**

- ❑ Steroids and Interferon are typically used in the treatment of KMS. However, due to the potential development of spastic dysplasia from long-term use of Interferon, and developmental and immune suppression problems from long-term use of steroids, Vincristine (an old chemotherapy drug) is rapidly becoming the drug of choice for treatment.
- ❑ Embolization or surgical removal of the tumor might be necessary.
- ❑ Some lesions may go away with treatment, or some lesions may remain and the clotting problems diminish.

### **Klippel-Trenaunay Syndrome**

- ❑ Klippel-Trenaunay syndrome (KT) is a congenital disease. The cause is unknown and symptoms and their degree vary from patient to patient.
- ❑ The most common conditions present in KT are: a port wine stain, soft tissue and bone hypertrophy (excessive growth), venous malformations, and lymphatic abnormalities.
- ❑ Complications can include bleeding, cellulitis, venous thrombosis, and pulmonary embolism. Other conditions such as varicose veins, skin breakdown and ulceration, and secondary infection may also occur
- ❑ Other symptoms can include gigantism of the toes, hands, and feet, as well as lymphedema and involvement of the abdominal and pelvic organs, and fused fingers and/or toes.

### **Steroids Continued**

- ❑ During steroid treatment, some patients can experience symptoms of gastric irritation, vomiting, moon face or swelling of the face, behavioral issues, mood swings, high blood pressure, or an increase in appetite.
- ❑ Topical steroid creams are on the rise for treating superficial hemangiomas. These steroids should be altered one week on and one week off and discontinued after several months, so as not to cause tissue atrophy.

### ***Seizure Medications***

- ❑ Seizure activity is also referred to as epilepsy or seizure disorder.
- ❑ There are many types of medicines used to control or alleviate the symptoms of seizure activity. Based on the age of the patient, the severity and type of seizure, and any other medical problem, a specific medication or combination of medications can be used.
- ❑ Generic forms of seizure drugs may be used, however, not all generic forms are exactly the same as the brand name drug. Sometimes the generic form is not as strong, and seizures may increase while on generic forms of seizure medications. Your physician should be consulted before changing to a generic drug. If a patient has seizures that are hard to control, switching from a brand name drug to a generic drug is not recommended.
- ❑ Patients should always check their medication before leaving a pharmacy, to make sure the proper medication and dosage are given.

## Drug Therapy

Drugs are used to treat a wide range of diseases, including those related to vascular anomalies and related syndromes. Here are few of the most commonly used drug therapies.

### *Antibiotics (oral and topical)*

- Oral and topical antibiotics are used to treat infection. Antibiotics can be taken orally, intravenously, or topically.
- Antibiotics can reduce the swelling and inflammation associated with bacterial infections.
- Topical antibiotics are used for infections on or near the surface of the skin. They are directly applied to the affected area.

### *Steroids*

- Steroids are often used to reduce the growth of hemangiomas. Typically, steroids are only used during the rapid growth stage of hemangioma development. Corticoid-steroids are the type most commonly used.
- Some rapid growing hemangiomas can create health problems, and should be treated aggressively. Hemangiomas affecting the face, mouth, eye, nose, lips, ear, and the airway are usually most often treated with steroids. Hemangiomas between the fingers, toes, and in the genital area can also be treated with steroids.
- When hemangiomas affecting the eye fail to respond to typical steroid treatment, Interferon Alpha-2a is usually used.
- During the use of steroids to treat a hemangioma, live virus immunizations should not be given to children.

## **Klippel-Trenaunay Syndrome continued**

- Associated conditions, such as arteriovenous fistulae, can result in heart failure if left untreated.
- If venous malformation is the dominant condition, the following symptoms may be present to varying degrees: painful thrombosis (clotting), muscle cramps, joint pain, pain when walking, lower intestinal involvement which can result in rectal bleeding, or bladder involvement which can result in blood in the urine.
- If lymphatic abnormalities are dominant the following symptoms may be present to varying degrees: soft tissue swelling and enlargement.
- KT is usually present in only one limb. However, it may occur in multiple limbs, head, trunk area, or internal organs.
- Treatment for KT is generally conservative. Compression therapy by the use of orthopedic elastic garments or intermittent pneumatic compression pumps is used to help with swelling in the limbs. Elastic garments also help protect the limbs from injury. Laser surgery is used to treat the port wine stain. Surgery is used to debulk excessive tissue. Sclerotherapy is used to restrict the flow of blood. Antibiotics, pain medication, and elevation of the affected limb(s) are used to treat symptoms of cellulitis. Hydrotherapy, such as swimming 2-3 times a week is highly recommended for extremity KT cases.
- Anticoagulants are used to prevent blood clotting. Vein ligation where veins are clamped off, or vein stripping and vein resection might be necessary in some cases.

### **Klippel-Trenaunay Syndrome continued**

- ❑ Heel inserts may be used where the difference in the legs are less than 1 inch. Amputation may be necessary in very extreme cases.
- ❑ Magnetic Resonance Imaging (MRI) studies are usually performed to help in the diagnosis of KT. Other imaging studies such as Doppler ultrasonography and angiography may also be used.

### **Lymphatic Malformations**

- ❑ Lymphatic Malformations used to be called cystic hygroma, hemangiolympangioma, or lymphangiomas. The lymphatics serve as a collection and transfer system for tissue fluids. When something disturbs this system, a lymphatic malformation is formed. The excess fluid accumulates and the affected lymphatic vessels enlarge and you see a mass.
- ❑ Research indicates that an absence of the correct number of lymphatics is the cause of the lymphatic malformation.
- ❑ Lymphatic malformations are sponge-like masses of abnormal channels and spaces containing clear fluid. Leakage from the skin can occur. This can further lead to cellulitis.
- ❑ If the lymph vessels in the face are affected, the face swells because the normal active transport mechanism has been disturbed.

### **Compression Therapy**

- ❑ Various types of compression garments and pumps are used to alleviate symptoms of some types of vascular anomalies.
- ❑ Compression stockings and hose are used to reduce swelling and to assist with blood flow. These elastic garments help protect the limbs. In addition, lymphatic or intermittent pneumatic compression pumps are also used to help with swelling in the extremities.
- ❑ Elastic wraps, compression wraps, and compression pumps with sleeves that cover the affected limb can aid the symptoms of edema and cellulitis.
- ❑ The use of compression stockings can help prevent venous leg ulcers.
- ❑ Compression therapy has also been used for large, trunk hemangiomas that do not respond to drug therapy. The compression literally “squeezes” the blood out of the tumor causing reduction that is significant enough to enable the child to undergo surgical removal of the lesion. This has also been used on extremity lesions with some positive response.

### **Lymphatic Malformations continued**

- ❑ These lesions can occur anywhere but are common in the head and neck area. These lesions may be superficial or deep (superficial ones are seen in the mouth area and look like frogs eggs). These lesions increase or grow with the individual. They may enlarge following an upper respiratory infection.
- ❑ Lymphatic malformations are either micro-cystic or macro-cystic. Lesions that are macro-cystic (large spaces in the lesion) appear as a soft, clear mass under normal or bluish skin and may respond to a drug called OK-432. This treatment should be discussed with a physician to determine if the patient is a candidate; and should only be attempted if no prior surgery has been performed on the lesion. Micro-cystic malformations are small raised lesions containing clear fluid. These superficial lesions appear like small, clear bubbles, sometimes turning dark red due to bleeding.
- ❑ It should be noted that only a skilled surgeon should operate on a lymphatic lesion. By removing some lymphatic vessels during surgery, you can actually cause enlargement of the lesion and further growth period.
- ❑ MRI and CAT scan are used to diagnose lymphatic malformations. Laser treatment, sclerotherapy, and surgery are used to treat or remove these lesions.

## Port Wine Stains

- ❑ Venular Malformations (Port Wine Stains) are always present at birth and can range from pale pink to dark purple in color. They are sometimes referred to as nevus flammeus. In the past these lesions were erroneously called "capillary hemangiomas."
- ❑ Port Wine Stains (PWS) occur in .3% of births and occur equally among males and females.
- ❑ Laser therapy, which is used to remove a port wine stain, in most cases is incomplete. Since the deficiency is in the circulatory system, in time the blood will re-pool in the affected area and the birthmark will once again appear, although to a lesser degree
- ❑ Once a Port Wine Stain is lasered, it is important at the first sign of reoccurrence to have one or two treatments to keep it faded. The individual will need to have maintenance laser treatments for life.
- ❑ Because Port Wine Stains can be progressive, treatment should be done early to prevent cobbling of the skin, thickening, and darkening of the stain. These lesions vary from low-grade to high-grade, pale to dark. Low-grade lesions progress at a slower rate than high-grade. Thickening of the gums and lips can also result from growth of a port wine stain. Debulking of the lip may be necessary.
- ❑ Port wine stains occur most often on the face, but can be on any part of the body. Port wine stains covering the eyelid (or very near the eye or on the forehead) can be associated with elevated eye pressure, causing glaucoma.

## Treatments for Vascular Birthmarks and Tumors

As with any disease, a wide range of treatments and therapies may be used to treat, correct, or to alleviate symptoms. Here are some of the most commonly used for the treatment or removal of vascular anomalies. Some of these treatments can be used in conjunction with others.



## Venous Malformations

- ❑ Venous Malformations is an abnormality of the larger, deep veins and is often called a "hemangioma." Some actually look like hemangiomas but a good history will reveal whether or not the lesion is a hemangioma or vascular malformation.
- ❑ A venous malformation can be deep or superficial, localized or diffused. The closer the vessels are to the surface, the deeper the color to the eye. A very deep lesion will have no color but will show a protruding mass.
- ❑ The jaw, cheek, tongue and lips are common sites for a venous malformation. These lesions are soft to the touch; the color disappears and empties as the lesion is compressed. When the child cries or is lying down, the lesion expands, the vessels fill and the color becomes more intense.
- ❑ A Venous Stain is a type of flat birthmark that is bluish in color and is comprised of enlarged venular vessels. There is often an underlying malformation. Sometimes blebs occur on these lesions, called angiokeratomas or granulomas. These can pop and bleed spontaneously.
- ❑ The natural history is a slow-steady enlargement. Regardless of how small it is upon detection, it will grow. Certain things can cause them to grow more rapidly such as serious sickness, trauma, infection, and hormone changes (puberty, pregnancy, or menopause). Partial removal is not recommended since these lesions will just grow back.

## Port Wine Stains continued

- ❑ Port Wine stain on the face can also indicate brain involvement and an MRI should be performed to rule out associated diseases (like Sturge-Weber syndrome).
- ❑ Lesions that are know as “geographic” with good skin peeking through parts of the stain, almost always respond well to laser therapy. Lesions that are more concentrated and dense will require more laser treatments.
- ❑ Most experts agree that the first 7-8 treatments will achieve the maximum results. After these first initial treatments, the subsequent treatments will generally produce marginal results. Following maximum results, the patient should probably take a break of one year and then try more laser treatment for further PWS lightening. New lasers are coming out every year that are getting deeper vessels, which are, in turn, getting more clearance.

## Sturge-Weber Syndrome

- ❑ Sturge-Weber Syndrome (SWS) is a congenital disease and the cause is unknown. Not all cases of SWS are identical and symptoms and their severity can vary.
- ❑ A port wine stain (PWS) on the face is usually present at birth. The PWS most often covers the forehead and eyelid, but can include a larger portion on one or both sides of the face and head, and may also extend into the throat, tongue, gums, or ear canal. However, the patterns and severity of PWS will vary on a case-to-case basis. There are reported cases of SWS where no visible PWS is present.

### Sturge-Weber Syndrome continued

- ❑ Abnormalities of the brain are common. The PWS is present on the outer layer of the brain causing calcification of the brain, and atrophy of the brain tissue.
- ❑ Seizures are a common symptom of SWS and are usually a result of the calcification process. Most seizure activity can be controlled or modified by the use of medication. In the most severe cases, a hemispherectomy (where one hemisphere of the brain is removed) is used as a “last ditch” effort to stop uncontrolled seizure activity.
- ❑ Glaucoma may be present at birth, or can appear months or years later. This disease affects the eye, resulting in vision loss due to damage to the optic nerve. It is recommended that patients have a yearly ophthalmology exam for glaucoma.
- ❑ Some patients have been given the diagnosis of SWS when they only have glaucoma and a PWS, but brain involvement is typically the defining factor for SWS.
- ❑ Most cases of SWS are diagnosed by age three. If there is no sign of brain involvement by age three, experts agree that the patient will likely not develop SWS. However, close monitoring of the eyes for glaucoma should continue throughout life.
- ❑ Early treatment of the PWS is recommended, as the PWS can thicken over the years and develop nodules. Laser treatments are now begun on infants, and can greatly improve the appearance and reduce effects of the PWS in years to come.

### Sturge-Weber Syndrome continued

- ❑ Also, early treatment of areas like the gums and lip are important, as these can become problem areas as they become engorged and grow larger.
- ❑ Affected gums can result in dental problems, and the lip area may require surgery to be de-bulked. The development of laser treatment has been greatly advanced over the years, and it is recommended that adults, who have never been treated or had little treatment, seek the advice of a skilled laser surgeon.
- ❑ Mild to severe retardation can also be a result of SWS. Learning disability can be another factor in SWS. Symptoms similar to ADD/ADHD can be also present.
- ❑ SWS can affect the brain by being bilateral or unilateral (affecting one or both sides of the brain). Hemiparesis, the weakening or loss of use on the side of the body opposite the port wine stain, can also be present. In rare cases SWS can affect other organs in the body
- ❑ Magnetic Resonance Imaging (MRI) studies are typically used in the diagnosis of SWS.