a guide to understanding

hemangiomas

a publication of children's craniofacial association

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This parent's guide to hemangiomas is designed to answer questions that are frequently asked by parents of a child with hemangiomas. It is intended to provide a clearer understanding of the condition for patients, parents and others.

how can children's craniofacial association (cca) benefit my family?

CCA understands that when one family member has a craniofacial condition, each person in the family is affected. We provide programs and services designed to address these needs. A detailed list of CCA's programs and services may be found on our website at www.ccakids.org, or call us at 800.535.3643.

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what are hemangiomas?

hemangiomas are the most common benign tumor of infancy. They occur in 1 out of every 10 Caucasian births,1 out of 71 African-American births and 1 out of 125 Asian births. In premature babies weighing less than a kilogram (2.2 pounds), hemangiomas may occur in 1 out of every 3 infants.

Most hemangiomas are not present at birth but appear within the first two weeks of life, most commonly on the head and neck area. Parents may notice a bright red spot resembling a scratch, but instead of disappearing, it gradually increases in size and volume. Superficial hemangiomas, which occur near the surface of the skin, are bright strawberry red, while deeper hemangiomas are bluish purple. Many children will have both components with a central red island sitting atop a bluish mound.

Parents are understandably concerned as the hemangiomas continue to enlarge and become increasingly noticeable. Hemangiomas usually continue to grow for 6 to 12 months, and then begin to shrink as the blood vessels stop growing and begin to shut down. The bright strawberry color begins to show patches of grey and white, and the hemangioma begins to soften as it becomes less engorged with blood. Most hemangiomas shrink very slowly over the course of 5 to 7 years. This is a process also called regression or involution. No one currently knows why hemangiomas occur nor why they stop growing and begin to regress.

Hemangiomas are at least 3 times more common in girls. There is no hereditary nor genetic cause of hemangiomas in the vast majority of cases. While 60% of hemangiomas are located in the head and neck area, they can occur anywhere on the body and also internally. Eighty percent of babies with hemangiomas will have only one, but 20% can have two or more. All hemangiomas eventually shrink, and about 70% will

regress so well that no treatment is necessary. Historically, pediatricians have advised patience and conservative observation. For most children this option is still appropriate; however, 30% of hemangiomas will leave behind a permanent deformity in the form of redundant (extra) skin, skin texture changes, bulky fatty scar tissue, and residual vessels. For this reason, many parents prefer to be proactive and seek various treatment options that will limit the growth and speed the resolution of a hemangioma during its early stages.

what kinds of vascular anomalies are there?

Although there are many kinds of vascular anomalies, they can be divided into two basic types: hemangiomas and vascular malformations. Hemangiomas occur far more frequently than vascular malformations. Because of this, some physicians tend to call all blood vessel conditions hemangiomas. It is important to distinguish between the two, because the clinical course, prognosis and treatment recommendations may be very different.

Hemangiomas are composed of blood vessels that are actively multiplying (a benign blood vessel tumor). Vascular malformations contain dilated vessels which are not actively reproducing (for example, varicose veins). Both are present in infancy, but vascular malformations may not be noticeable until later in life. Hemangiomas generally grow in the first year of life, then slowly shrink over a period of years. Vascular malformations tend to slowly dilate over time and do not have the ability to shrink.

how do our blood vessels work?

The heart pumps blood to all parts of the body through arteries. The arteries divide into smaller and smaller branches called arterioles. Eventually the blood vessels become extremely small as they pass through the tissues and are called capillaries. Oxygen and nutrients are delivered to the tissues, and eventually the blood passes into small veins called venules. These veins join together to form larger and larger veins, eventually returning to the heart. In addition to arteries, veins and capillaries, there is another type of vessel called a lymphatic. Lymphatics are similar to veins except they carry excess water instead of blood back to the heart. Lymphatics prevent swelling and are also part of the immune system.

when should treatment for hemangiomas be considered?

All physicians agree that hemangiomas located around the eyes or in the airway near the vocal cords are potentially dangerous and require urgent treatment. Hemangiomas that block vision can lead to permanent lazy eyes (amblyopia), and hemangiomas that press on the eye can distort the cornea. Hemangiomas in the airway can block breathing and are, therefore, life-threatening. Hemangiomas in the liver can become very large and stress the heart. In these cases conservative observation is not appropriate, and medical intervention, rather than surgery, is the first line of therapy.

Treatment is not mandatory but should be strongly considered for problematic hemangiomas in other areas, especially if they are likely to result in significant cosmetic deformity. Hemangiomas are of particular concern in the facial area, where loose skin and bulky scar tissue are extremely common in the nasal, lip and cheek areas.

Ulcerated hemangiomas occur when the fragile overlying skin breaks down and an open wound results. In most patients bleeding can be stopped with pressure, and the wound can be treated with an antibiotic ointment. However, ulcerated hemangiomas can be excruciatingly painful, particularly in the genital area where stool and urine constantly irritate the wound. Occasionally some ulcerated hemangiomas will bleed heavily from an open artery, requiring sustained pressure, combined with suture placement or urgent surgical intervention.

Hemangiomas can also occur in the salivary gland in front of the ear – the parotid gland. This can lead to blockage of the ear canal as the hemangioma grows. If both sides are affected, medical treatment is usually indicated to avoid problems with hearing loss.

how are hemangiomas diagnosed?

The vast majority of hemangiomas can be diagnosed on the basis of their medical history and physical examination alone. No further diagnostic testing is necessary. The history of an enlarging red or purple bump that began shortly after birth, and the appearance of a strawberry-colored patch of skin on examination is usually enough evidence to make the proper diagnosis.

Hemangiomas located underneath the skin are harder to diagnose. Sometimes a series of examinations over time will be enough to make the proper diagnosis. This is especially true as the hemangioma stops growing and begins to shrink after the first year of life. In problematic cases, an MRI scan is usually the preferred diagnostic test (rather than a CT scan or ultrasound). The MRI will give the most information about the extent and flow

characteristics of the mass without subjecting the child to radiation. The MRI scan will also help to distinguish a hemangioma from a vascular malformation or a rare solid tumor. Large hemangiomas in the facial and neck region are also best studied with an MRI scan to rule out airway involvement, possible distortion of the eye structures or rare brain abnormalities.

Hemangiomas located over the lower back and pelvis may be associated with a tethered spinal cord or other pelvic abnormalities, thereby warranting a diagnostic MRI scan. Neurosurgery may be necessary if there is spinal cord compression.

Arteriography (placement of a catheter into the feeding arteries of a hemangioma) is rarely indicated, except in the case of large hemangiomas causing heart failure. Injecting clotting materials into the feeding artery may be very useful to decrease excessive flow through the hemangioma.

Ultrasound may be used to watch the progress of a deeper hemangioma. For example, when there is a hemangioma in the liver, monthly ultrasound exams may be helpful to document the response to medical treatment such as steroid therapy. Ultrasound may also be useful as a screening tool to look for cardiac or arterial abnormalities in patients with large facial hemangiomas. These patients may have a set of conditions or symptoms that fall into a pattern that have been given the term "PHACES":

Posterior cranial fossa (brain) abnormalities,

Hemangiomas,

Arterial abnormalities,

Coarctation (a narrowing of the aorta) and cardiac abnormalities,

Eye abnormalities, and

Sternal clefts (a split in the breastbone).

what are the treatment options for hemangiomas?

there are basically four ways to treat hemangiomas:

Conservative observation – The growth and regression (shrinkage) of the hemangioma is monitored by periodic office visits with a pediatrician, pediatric dermatologist or pediatric plastic surgeon. The majority of hemangiomas can be safely monitored since 70% will not cause significant problems during the growth phase and will adequately regress or shrink over an average of 5 years. The vast majority of patients are followed by their pediatricians and do not require the additional expertise of a pediatric dermatologist or plastic surgeon.

Laser treatment – Lasers use light energy to selectively injure the blood vessels of a hemangioma. Lasers that treat abnormal blood vessels target the blood itself. The laser light passes through the outer skin and is absorbed by the chemical oxyhemoglobin in the red blood cells. The blood heats up, causing the vessels to either burst or shrink, depending upon the type of laser. The hemangioma may show initial regression or shrinkage, but often rebound growth occurs within a few weeks. In this case periodic additional laser treatments are performed until permanent regression occurs. The laser light penetrates only about a millimeter into the skin, therefore lasers are most effective for early flat hemangiomas. Deep or thick hemangiomas do not respond well to lasers designed to treat superficial skin vessels. Some physicians use a laser with a glass fiber that is passed into a thick hemangioma to damage the abnormal blood vessels with heat, with variable results. Otolaryngologists (ear, nose and throat doctors) may elect to laser hemangiomas that partially block the airway, in addition to giving steroids.

Medical treatment – A number of studies have shown that a medication called Propanolol, which in the past was used to treat high blood pressure and certain types of heart disease, may be effective at speeding up the rate of hemangiomas involuting (getting smaller). This medication can be given orally or applied topically. Some doctors will only use this medication in selected instances.

Surgical intervention – Surgery is performed to remove or reduce a hemangioma, or revise the remaining deformity left by a hemangioma. There is a strong temptation for parents to seek surgical treatment as an immediate cure for a disfiguring hemangioma. Timing of surgical intervention is an area of intense debate among pediatric surgeons. Some surgeons favor aggressive removal of all hemangiomas at an early stage. Others recommend postponing surgery until regression (shrinkage) is complete, usually at 5-7 years of age. 7 Parents should realize that up to 70% of hemangiomas regress adequately without surgical intervention and that all surgical procedures leave permanent surgical scars. Therefore, an aggressive approach may mean that some children will undergo unnecessary surgery. Some of these children may end up with surgical scars that are worse than natural shrinkage. Each child should be individually evaluated. Surgical recommendations must take into account the risk. In some cases surgery may be the preferred treatment option and may provide additional psychological benefits to the patient and parents. These cases include situations when surgery can be performed with acceptable blood loss and minimal complications. Additional considerations would include cases where the anticipated surgical scar is better than the expected deformity associated with natural regression (shrinkage).

The timing of surgery can be roughly divided into four stages:

1. **Proliferative phase** – The hemangioma is still growing and is engorged with blood.
2. **Early involutional phase** – The hemangioma has reached maximum size and shows signs of regression (shrinkage) but still has a significant blood supply. If compressed, it still fills rapidly with blood.
3. **Mid involutional phase** – The hemangioma has lost much of its blood supply and turgor (puffiness) and a superficial hemangioma may demonstrate grayish-purple skin with a spongy consistency. If compressed, it still fills with blood, although less rapidly.
4. **Later involutional phase** – The hemangioma has very little or no residual blood flow. The skin is flesh-colored, often loose, with a crepe-like or spongy consistency when compressed, with no refill.

Patients seeking treatment in the late involutional stage (stages of shrinkage) are excellent candidates for surgical debulking (reduction) or hemangioma scar revision because additional improvement is not expected. The surgery is relatively bloodless, because the hemangioma is composed mostly of fatty scar tissue. Most of these children, however, are already in school since the average patient is 5-7 years of age, raising issues of appearance and psychological embarrassment, particularly for facial hemangiomas.

There is a strong movement among parents of hemangioma support groups to push for early surgery. In a limited number of patients with small or mushroom-shaped hemangiomas or ulcerated, bleeding hemangiomas, surgery can be performed during the proliferative (growing) phase. Surgery is appropriate in these hemangiomas, because they will almost certainly leave redundant skin or scars that will eventually benefit

from surgical intervention. However, there are cases where early surgery may be inappropriate. One case is in large hemangiomas that cannot be easily removed and may present a significant operative risk during the proliferative (growing) stage. Another is where excessive bleeding, facial nerve injury and a significant surgical scar may be reasons not to have early surgery. Surgery during the early to mid involutional (shrinking) phase is becoming increasingly common. By that time it is more predictable which hemangiomas are highly likely to leave residual deformity. With early surgery, patients and parents are not subjected to a psychologically painful delay while awaiting complete involution. Parents should be aware that more than one surgical procedure may be necessary for optimal results. A second procedure addresses residual asymmetries and deformities. All parents must understand from the beginning that surgery always leaves a scar of some kind and that surgery is appropriate only if the expected outcome is an improvement over natural involution (shrinkage).

There are risks involved with the treatments described in this booklet Always discuss risks with your physician before selecting a treatment option.

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