What is a “large (giant) congenital nevus”?
A Nevus (nē‘vəs) common skin growth composed of special pigment-producing cells called nevomelanocytes. Nevomelanocytes are related to pigment producing cells normally found in the skin, called melanocytes. A congenital nevus is a mole present at birth.

Since nevi can exist on any part of the body, and grow in proportion to body growth, it is difficult to find one specific definition for the term “large nevus” or “giant nevus”. There are at least four common ways a doctor may use to define a nevus as “giant” or “large”. None are universally accepted, and additional definitions do exist. Some doctors may even provide separate definitions for “giant” and “large” nevi, while in this document they are used interchangeably.

We have listed what we consider to be the most common definitions. Your doctor may use any or none of these in determining if you or your child has a large nevus.

• Some doctors define a large nevus as one that would take more than one surgical excision to remove. Surgeons, or others involved with the removal of nevi, sometimes use this definition.

• Another definition is that a nevus is large if it covers more than 2% of the patient’s total body surface area, or is larger than the palm of the patient’s hand.

• Others define a nevus as large if it measures over eight inches in diameter. If using this definition and measuring a nevus on a newborn or child, the nevus is large if it is expected to become over eight inches in diameter by the time the person is fully-grown. This definition provides a definite and exact number to large nevi.

• Another definition for a large nevus is one that covers a major portion of a major anatomical site. This definition provides allowances for nevi of the head, legs or arms to be categorized as large even if they do not meet the previous definition.

How common are congenital nevi?
One in every hundred babies is born with a congenital nevus. Congenital Nevi measuring four inches or greater in diameter, occur in approximately 1 in every 20,000 births. Larger nevi covering as much as 70% of the skin are less common, probably about 1 in 500,000.

How does “nevus skin” differ from “clear skin”?
Nevi are much more than simply “dark skin”. Besides producing pigmentation, the area covered by a nevomelanocytic nevus has a tendency to contain large hair follicles, often producing increased amounts of hair. Large nevi often produce thick, black hair over some or all of the nevus. Usually a person does no sweat within a large nevus because the mass of nevomelanocytes prevents the formation of sweat glands. Many nevi lack subcutaneous fat, a thin layer of fat just under the skin that pads the body everywhere. Some nevi will also have soft growth tumors, such as neurofibromas, or other neural tumors, within the nevus. These growths are not painful and are normally superficial. “Nevus skin” can be very thick, and have a folded appearance, or can be very thin and fragile. Nevi also have an increased number of mast cells, which makes them more susceptible to itching, scratching and infection.

Why are nevi important?
Nevi serve no function and most moles are harmless. Nevii are the most common abnormality of the skin. Most nevi are important because of their relationship to a potentially deadly skin cancer called melanoma. Also, children with large nevi may have a higher concentration of these same abnormal cells lining the meninges (outer covering) of the brain, this condition is known as neurocutaneous melanosis. The majority of people with large nevi have no neurological problems, other may develop seizures or hydrocephalus (“water on the brain”), and in extreme cases, these cellular deposits may develop melanoma or become so concentrated that they interfere with normal functioning of the brain.
**Why do moles form?**

Large nevi are caused by an unknown defect during embryologic development. There is no known method of prevention. Congenital nevi form in the womb very early in development, within the first twelve weeks of pregnancy. Drugs, diet, illness or injury have no known influence on the formation of moles. Nevi appear in either sex, in all races, and on all areas of the skin. It is important to recognize the difference between the terms “congenital” and “hereditary”. The term “congenital” is simply a synonym for “present at birth” and carries no additional significance. There is no data to support that large congenital nevi are hereditary in nature. There are several documented cases of identical twins, where one child was born with a large nevus and the other was not.

**Which moles may develop melanoma?**

Most moles are harmless, but some do have increased tendency to develop melanoma. Opinions vary as to the exact risk involved, but somewhere less than 10% of children born with large nevi develop melanoma over a lifetime. Any congenital nevus, small or large, may develop melanoma, but those of greatest concern are those that are very large, are dark in color, or variegated (speckled) in color. Studies indicate that satellite nevi very rarely, if ever, develop melanoma, and that nevi covering the trunk carry the highest risk of developing melanoma. Changes in size, color, surface texture, pain, bleeding, or itching are all of concern. Any such changes should be evaluated medically if they last longer than two weeks.

If a nevus is suspected to be malignant, a small incisional biopsy can be taken. This is normally done in a dermatologist’s office.

**What should be done about a congenital nevus?**

You should consult a physician soon after the birth of an affected child. Because of the rarity of this condition as well as the lack of knowledge about its cause, there is a rather wide range of professional opinions regarding the proper course of action with respect to treatment of large congenital nevi. However, there is a reasonable consensus on the first few diagnostic steps. Work closely with your pediatrician, who may refer you to both a pediatric neurologist and a dermatologist specializing in pigmented skin lesions. These evaluations may lead to other tests such as a skin biopsy. The usefulness of an MRI (Magnetic Resonance Imaging) is controversial when nevi involve the head, neck, or midline back. In all but the most severe cases, surgical removal of the nevus can be considered.

Treatment is very individualized and depends on the age and health status of the child, as well as the size, location, appearance, and growth history of the mole. While most plastic surgeons are trained in the techniques for removal of nevi, their approaches and proficiencies with respect to your child’s particular case can vary. Your own decisions and preferences will be critical in determining how your child’s condition is followed and/or treated.

**What methods or removal are available?**

Smaller moles can be removed by single surgical excision. Excision of larger moles requires “replacement” of the excised skin in one form or another. This can be accomplished by transferring skin from another area of the body (skin graft), replacement with cultured skin cells from the child, or stretching adjacent “good” skin (skin expansion). The first two methods can be done in a relatively short period of time, but leave substantial scarring in both the donor and recipients sites. Skin expansion and serial excision, on the other hand, can bring skin with near normal smoothness, color, and sweat gland activity into the excised region. These procedures are typically done over a much longer period of time and require numerous surgeries under general anesthesia. They, too, involve scarring, but whenever feasible, they are the preferred method from a cosmetic standpoint.

There are also several techniques that do not remove the mole but can sometimes provide temporary aesthetic benefits. Chemical peels can be used to lighten nevi with varying success. Lasers can also be used to temporarily lighten nevi, and to remove hair from a nevus. Laser treatments do not reduce melanoma risks and results are unpredictable and usually temporary.

Treatments vary based on individual circumstances and different doctors will often have different preferences for treatment of a nevus. You should consult your doctor to find out about specific options for you.

**Why remove a nevus?**

The purpose of removing a nevus is to reduce the melanoma risk. It is often not possible to remove every cell of a large congenital nevus. The goal is to remove as many cells as possible while at the same time to preserve function and to leave as small of a scar as possible. Nevi cannot be removed without leaving a scar.

Additional reasons to remove the nevus are to improve the appearance, strength, texture, sensation, and sweat gland activity of the affected area, if feasible. These reasons,
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Nevus Outreach, Inc. is proud to list its professional advisory committee:

A. James Barkovich MD
Neuroradiologist
University of California at San Francisco

Bertrand Chretien-Marquet, MD
Plastic Surgeon
Paris, France

Ilona Frieden, MD
Pediatric Dermatologist
University of California at San Francisco

Ashfaq Marghoob, MD
Clinical Dermatologist
Stony Brook, New York

Miguel Reyes-Mugica, MD
Pediatric Pathologist
Yale-New Haven Medical Center

Carolyn Russo, MD
Pediatric Neuro-Oncologist
University of California at San Francisco

Lewis Strauss, MD
Pediatric Hematologist-Oncologist

Frank Vicari, MD
Pediatric Plastic Surgeon

Childrens Memorial Hospital in Chicago

although of much less medical importance than reducing the risk of malignant melanoma, may be profoundly important to the child’s psychological development.

**How often should a congenital nevus be examined if not excised?**

Moles should be examined at home once a month and by a physician once or twice a year. A mole’s color may stay the same, lighten slowly, or darken slowly over time. Hair in a mole is not helpful in judging whether a mole is malignant or benign. Any rapid or persistent change in color, texture, or outline, or the development of a new nodule, is cause for immediate examination by your doctor.

**If you have further questions** after reviewing this document, please contact your doctor or Nevus Outreach. Nevus Outreach can be reached toll free at 1-877-4-A-NEVUS or visit our web site at www.nevus.org

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