CONDITION

Giant congenital nevus

A giant congenital nevus is a very large, usually dark birthmark formed by pigment-producing cells before birth.

WHAT IT IS
A very large pigmented birthmark present from birth

BODY SYSTEM
Skin and sometimes nervous system

MAIN SYMPTOMS
Large birthmark, coarse hair, raised areas, nearby smaller spots

WHAT CAUSES IT
A random genetic change before birth

HOW IT IS DIAGNOSED
Skin examination, sometimes dermoscopy, biopsy, or MRI

MAIN TREATMENT
Monitoring, surgery, skin care, and emotional support

CAN IT BE CURED?
No - removal may not eliminate risk

IS IT AN EMERGENCY?
Sometimes - a first seizure needs emergency care

LONG-TERM OUTLOOK
Most people live full lives with ongoing surveillance

Overview

A giant congenital nevus, also called a giant congenital melanocytic nevus, is a large birthmark made up of melanocytes, the cells that produce skin pigment. It is present at birth or appears soon afterward.

The mark is usually brown or black but can vary in color and texture. It may be flat, raised, uneven, or covered with coarse hair. Smaller marks called satellite nevi may occur elsewhere.

Congenital nevi are classified by the size they are expected to reach in adulthood. A projected adult size greater than about 40 centimetres is commonly considered giant, although definitions vary. The nevus grows as the child grows.

A giant congenital nevus is not skin cancer. However, very large or numerous congenital nevi

are associated with a higher risk of melanoma. In some people, pigment cells are also present around the brain or spinal cord. These risks make skin and, when appropriate, neurological follow-up important.

The nevus may also affect comfort, movement, self-image, and social experiences. Treatment decisions consider these effects alongside the risks and burden of surgery.

Symptoms

The main feature is a large area of differently colored skin, often on the back, trunk, scalp, face, or a limb.

Features may include:

Congenital nevi may become thicker, lighter, darker, or hairier as a child grows, especially during puberty. Many nodules are benign, but a new or changing lump should be assessed.

Most people have no neurological symptoms. When pigment cells affect tissues around the brain or spinal cord, symptoms can include seizures, repeated headaches, vomiting, developmental difficulties, weakness, or balance problems. These require medical assessment.

- Brown, tan, black, reddish, or mixed coloring
- Thick or coarse hair
- A flat, folded, pebbled, or raised surface
- Firm bumps or nodules
- Smaller satellite nevi elsewhere
- Dryness, itching, tenderness, or reduced sweating

Causes and risk factors

A giant congenital nevus begins during early development before birth. It is linked to a genetic change in a developing pigment cell, often in a gene that controls cell growth.

This is usually a mosaic change, meaning it is present in only some of the body's cells. It generally happens by chance after conception and is not inherited from either parent.

Nothing a parent did during pregnancy is known to cause the nevus. It is not caused by diet, infection, medication, or sun exposure.

A very large projected adult size, many satellite nevi, or a nevus over the scalp or spine may be associated with a greater chance of nervous-system involvement or melanoma. These

features guide assessment but do not predict what will happen to an individual.

Diagnosis

Diagnosis is usually based on the nevus's appearance, location, and presence at or soon after birth. A clinician may measure it, estimate its adult size, and record satellite nevi.

Dermoscopy is a painless examination using magnification and light to view structures below the skin surface. Standardized photographs can provide a baseline for detecting meaningful changes.

A biopsy is not routinely needed. It may be recommended if an area grows unusually, becomes firm, ulcerates, bleeds without a clear injury, or is otherwise difficult to assess. A small tissue sample is then examined under a microscope.

A specialist may discuss MRI of the brain and spine for some babies with a giant nevus, many congenital nevi, or other higher-risk features. MRI uses a strong magnet rather than radiation and can detect pigment-producing cells or related changes around the nervous system. Its usefulness and timing are considered individually; not everyone needs an MRI.

Treatment and management

There is no single best treatment. Care may involve dermatology, plastic surgery, neurology, psychology, and other specialists.

Regular monitoring may include skin examinations, photographs, and assessment of new or changing areas. Reviews are often more frequent during infancy or periods of rapid change, but the schedule is individualized.

Surgery can remove part or, less commonly, all of the visible nevus. Complete removal of a giant lesion may be impossible or require several operations. Options include staged excision, skin grafts, skin flaps, or tissue expansion. Tissue expansion gradually stretches nearby skin before reconstruction.

Surgery leaves scars and carries risks including anesthesia complications, infection, bleeding, wound problems, and altered sensation. Large procedures may require substantial recovery. Decisions should consider function, comfort, appearance, emotional effects, surgical burden, and the person's or family's preferences.

Removing visible skin does not guarantee that melanoma will be prevented. Nevus cells may remain in deeper tissue, other skin, or the nervous system, so follow-up may continue after

surgery.

Curettage, dermabrasion, or laser treatment may lighten or smooth selected areas but usually does not remove deeper nevus cells. Color may return, and surface treatment can make later skin checks more difficult.

Gentle cleansers, fragrance-free moisturizers, and protection from friction may reduce dryness or itching. Hair may be carefully trimmed or shaved. Counseling and peer support can help with body image, unwanted attention, school experiences, or treatment decisions.

Complications

Melanoma is the most serious possible complication. Risk is higher than in the general population, particularly with very large nevi and numerous satellite nevi, but most affected people do not develop melanoma.

Melanoma may begin in the skin, deeper tissue, or nervous system. Detection can be difficult because congenital nevi naturally vary in color, texture, and thickness. A persistent or clearly unusual change needs prompt assessment.

Some people have melanocytes in tissues around the brain and spinal cord. This is often called neurocutaneous melanosis or melanocytic neurocutaneous syndrome. Possible effects include seizures, increased pressure around the brain, movement problems, and developmental difficulties. An abnormal MRI does not always lead to symptoms.

Other complications include fragile or itchy skin, reduced sensation, restricted movement where a thick nevus crosses a joint, and problems related to repeated surgery.

Visible differences may also affect confidence and mental health. Staring, questions, teasing, or pressure to hide the nevus can be distressing and deserve support.

Prevention

A giant congenital nevus cannot currently be prevented. It results from a chance genetic change during development, not something done before or during pregnancy.

Sun protection cannot eliminate the melanoma risk associated with the nevus, but it prevents sunburn and reduces additional ultraviolet damage. Use shade, clothing, hats, and age-appropriate sunscreen.

Monitoring cannot prevent every complication, but knowing the nevus's usual appearance and attending planned reviews may help identify concerning changes earlier.

Living with it and what to expect

A giant congenital nevus grows with the body. Its color, thickness, hair, and surface may change over time, making baseline photographs and planned reviews useful.

Some people choose observation, while others have one or more operations. Surgery may reduce the visible area or improve movement and comfort, but it replaces the birthmark with scars. Goals may change as a child becomes old enough to express preferences.

Most people can attend school, work, exercise, form relationships, and take part in everyday activities. Individual restrictions may depend on skin sensitivity, surgery, or neurological findings.

Follow-up may continue into adulthood, including after extensive removal. It usually covers remaining skin, scars, new symptoms, and emotional well-being. People with neurological findings may need additional developmental or neurological care.

Children benefit from simple, honest explanations and appropriate control over photographs, clothing, hair removal, and how they answer questions. Adults may also benefit from counseling or contact with others who have congenital nevi.

When to get medical help

Arrange a prompt, non-emergency assessment for:

Routine appointments are appropriate for planned skin checks, skin or hair care, surgery discussions, and emotional support.

Seek urgent medical advice for repeated headaches with vomiting, new balance problems, developmental loss, weakness, or other new neurological symptoms.

Call emergency services for a first seizure, a seizure lasting several minutes, repeated seizures without recovery, difficulty breathing, sudden severe weakness, or loss of consciousness.

- A new lump that keeps enlarging
- A firm area that differs from nearby tissue
- Repeated bleeding, crusting, or an open sore
- Persistent pain, itching, or tenderness
- A rapid or unusual change in color, shape, or texture
- A surgical wound becoming increasingly red, swollen, painful, or wet

Common questions

Is a giant congenital nevus cancer?

No. It is a birthmark made from pigment-producing cells. It does carry a higher melanoma risk than ordinary skin, so ongoing awareness and follow-up are recommended.

Will it turn into melanoma?

Most giant congenital nevi do not become melanoma. Risk is influenced by factors such as size, location, and the number of satellite nevi. No examination or operation can reduce the risk to zero.

Can a giant congenital nevus be completely removed?

Sometimes all visible involvement can be removed, but this is often impractical for a giant nevus. Several operations may be needed, and cells can remain in deep tissue or the nervous system. Follow-up may still be necessary.

Why might a baby need an MRI?

MRI can identify pigment cells or related changes around the brain and spinal cord. It may be considered for a giant nevus, many satellite nevi, or other higher-risk features, but it is not needed for every congenital nevus.

Can shaving the hair make the nevus cancerous?

No. Cutting or shaving hair does not cause melanoma or make the nevus grow. Use gentle methods to avoid irritation. Have scars, open areas, or changing nodules assessed before using cosmetic procedures over them.

Can I pass a giant congenital nevus to my children?

Usually not. Most result from a mosaic genetic change that occurred by chance during early development and is not inherited in the usual way.

Sources

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