

## Congenital Melanocytic Nevus

A congenital melanocytic nevus, is a melanocytic nevus that is already present at birth, distinguishing it from 'ordinary' melanocytic nevi. A congenital nevus can cause symptoms and complaints. Here, we provide information on what a congenital nevus means for your child.

### **What is a congenital melanocytic nevus?**

A congenital melanocytic nevus (CMN), is unlike a 'ordinary' melanocytic nevus, present at birth. Congenital melanocytic nevi occur in 1-2% of the population. The larger the melanocytic nevus, the rarer.

### **Different forms of congenital melanocytic nevi**

The melanocytic nevus can occur anywhere on the body and can vary in size. The melanocytic nevus can be small, with a size (in adulthood) of under 1.5cm. However, the melanocytic nevus can also be very large, with a size (in adulthood) of more than 60cm.

The melanocytic nevi can also vary in numbers. It can be a single melanocytic nevus, several medium-sized melanocytic nevi, or one very large melanocytic nevus with multiple small melanocytic nevi (satellites).

Congenital melanocytic nevi can also vary in color (from very light brown to black, pink, blue, gray) and texture (smooth to very folded). Swellings may also be present on the skin and/or under the skin, and hair can grow from the melanocytic nevus.

### **Causes of congenital melanocytic nevus**

We think that a CMN arises from a change in the genetic material of the precursors of pigment cells early in the development of an embryo. As a result, pigment cells tend to nestle excessively in the skin and the central nervous system.

The timing of the change in genetic material during the development of the embryo likely determines the spread of the melanocytic nevus. If the change occurs early in the development, it leads to a greater dispersion of melanocytic nevus cells. The melanocytic nevi are then larger, and the cells may also infiltrate the nervous system. If the change happens later in development, it only results in smaller melanocytic nevus without satellites.

Because the change occurs after fertilization (and thus not through a mutation in the sperm or egg cell), a CMN is not hereditary.

### **Symptoms and consequences**

- The skin of a CMN has a different structure than normal skin. This can sometimes cause complaints such as dry skin, itching, hair follicle inflammation.
- Because sweat glands are most times not present in melanocytic nevi, this can result in overheating of the person with a larger melanocytic nevus. Under the heading 'overheating,' you can learn how to recognize overheating and what you can do about it.
- There may be less fat in the skin under the melanocytic nevus. As a result, the CMN may lie lower than the rest of the skin. Also, an arm or leg may be thinner due to the melanocytic nevus.

- Subcutaneous swellings in a CMN are usually present from birth. These usually remain the same, but sometimes they can also occur during childhood. If that happens, they often grow rapidly. These are called proliferative nodules. They can be soft or hard and can range from a few millimeters to a few centimeters in size. It can sometimes be difficult to see or feel the difference between a CMN and a melanoma (skin cancer). Usually, we need to cut out a piece of proliferative nodules for further examination.

### *Neurological issues*

Sometimes, the melanocytic nevus cells are not only present in the skin but also in the central nervous system. If melanocytic nevus cells are present in the central nervous system (referred to as neurocutaneous melanosis), there is a risk of neurological problems, such as hydrocephalus, epilepsy, or neurological failure symptoms. Under the heading 'when to contact us,' you will find more information about what to look for in case of neurological problems.

The risk of severe neurocutaneous melanosis appears to be around 2% in patients with a melanocytic nevus expected to be larger than 20cm in adulthood. In children with larger congenital melanocytic nevus (>40cm expected size in adulthood and multiple smaller CMNs), this risk is higher. The exact extent of this increased risk is not yet known.

### **Overheating**

People with CMN can become overheated. This is because the skin structure is different in the melanocytic nevus, causing most people with a CMN to have difficulty sweating.

Tips to prevent heat emergencies:

1. Minimize movement during warmer parts of the day; prefer activities during cooler times.
2. Use air conditioning during the hottest times of the day if possible. If not, use a fan, stay out of the sun, avoid warm environments, and rest as much as possible.
3. Drink plenty of water, more than you would normally. Avoid alcohol and reduce coffee intake, as both contribute to fluid loss.
4. Use a small pool, foot bath, or garden sprinkler for cooling. A plant sprayer with ice water can also be helpful.
5. Wear light and breathable clothing, preferably in light colors. Natural materials feel cooler than synthetic fabrics.
6. Various manufacturers offer cooling neck wraps, wristbands, and vests.
7. Seek medical help for the following symptoms:

Early signs of overheating:

- Dizziness
- Fatigue
- Headache
- Muscle cramps
- Nausea
- Thirst
- Feeling of weakness and lightheadedness

Later signs of overheating:

- Cold, pale skin
- Dilated pupils
- Headache
- Nausea and vomiting

- Incoherent behavior
- Unconsciousness

Symptoms of heatstroke:

- Dry, warm, and very red skin
- Fever
- Dark urine
- Confusion
- Rapid and shallow breathing
- Fast and weak pulse
- Small pupils
- Epileptic seizures
- Unconsciousness

First aid for overheating:

1. Take the person to a cool space where they can lie down, and elevate their feet a bit.
2. Apply cool wet cloths to the skin or use cool water and a fan to cool down. Place cold compresses on the neck, groin, and armpits. Avoid using alcohol.
3. Provide cold water, sports drink, or a lightly salted drink. Avoid alcohol or coffee.
4. Seek medical help if the person shows signs of loss of consciousness, signs of shock (blue lips and nails, decreased consciousness), or during an epileptic seizure.

### **As your child grows older**

Congenital melanocytic nevi change as your child gets older, especially during the early years of life. They often become darker, more raised, hairs come out, and swelling may develop. However, melanocytic nevi can also become lighter, especially in individuals with a light skin type. A CMN grows in proportion to the growth of the skin.

### **Risk of melanoma (skin cancer)**

The risk of melanoma is not or nearly not higher than normal (less than 1%) for smaller congenital melanocytic nevi (under 20cm in size in adulthood). When congenital melanocytic nevi are larger (more than 20cm in adulthood), the risk of melanoma is 2 to 3%. Most melanomas occur in melanocytic nevi that are larger than 40cm in adulthood. The melanoma can be present in both the skin and the central nervous system and sometimes metastasize to other parts of the body.

In particular, children with a CMN with an expected size of more than 60cm in adulthood, seem to have an increased risk. The exact magnitude of this risk is not yet known.

### **Care**

You should protect the CMN well from the sun. This means that you should apply sunscreen with a sun protection factor of 50 every 2 hours and after the melanocytic nevus has become wet.

If the skin of the melanocytic nevus feels dry or itchy, you can apply a nourishing, oily base ointment or cream. If the itchiness persists, the dermatologist will prescribe a dermatocorticosteroid ointment. Sometimes, the hair follicles in the melanocytic nevus get inflamed. If there are many of them and/or you are bothered by them, you can inform your dermatologist.

Hair in the melanocytic nevus may only be cut short. You may not shave/epilate/wax/resin or using depilatory cream on the hair, as this can damage the skin of the melanocytic nevus.

### **Self-Examination**

Because there is a (small) chance that CMN can become malignant (skin cancer, or melanoma), it is important to regularly check the melanocytic nevus for changes. The working group recommends doing this monthly. In doing so, it is helpful to take photos (annually) and regularly compare the melanocytic nevus with them.

During self-examination, pay attention to the following characteristics by looking and feeling:

#### **Observing**

##### *Growth*

During childhood, the melanocytic nevus will grow along with the skin. In adulthood, a melanocytic nevus essentially no longer grows. If the melanocytic nevus grows, or if it grows faster during childhood than the part of the body where it is located, then contact the treating dermatologist (expedited).

##### *Color*

The color of the melanocytic nevus may change over time (darker, lighter). If this color change is not symmetrical and occurs very quickly, contact the treating dermatologist (expedited).

##### *Shape*

If the melanocytic nevus changes shape, the edges become irregular, or it develops extensions, contact the treating dermatologist (expedited).

##### *Symptoms*

Congenital melanocytic nevi may sometimes cause itching. If itching increases, the melanocytic nevus becomes painful, bleeds easily without clear trauma, or develops crusts, contact the treating dermatologist (expedited).

#### **Feeling**

##### *Lumps*

Bumps may develop in a CMN. If the bumps become hard, grow rapidly, or are painful, contact the treating dermatologist (expedited).

### **Medical Consultations**

At the Center for Pediatric Dermatology/Center of Rare Skin Diseases at Erasmus MC Sophia, a multidisciplinary outpatient clinic for congenital melanocytic nevi takes place monthly for children and adults with congenital melanocytic nevi. During this outpatient clinic, a (pediatric) dermatologist, pediatrician, (pediatric) plastic surgeon, and (pediatric) neurologist are present. Typically, you will be initially examined by a dermatologist in training, after which the involved specialists will also evaluate your condition. The frequency of follow-up appointments depends on the size of the melanocytic nevus and the risk of melanoma or neurological abnormalities.

### **Treatment**

During the consultation, we will discuss potential treatment options with you. Whether excision or laser treatment is possible depends on the size and location of the melanocytic nevus. Treating the CMN does not reduce the risk of developing melanoma.

## **When to contact us**

### *Neurological Symptoms*

If you notice that your child is beginning to behave differently from others or from what you are used to, please contact us.

Other examples of neurological symptoms in which you should contact us include:

- Headaches: severe and increasing headaches or vomiting may indicate increased pressure in the head. There could be other explanations as well.
- Seizures: episodes of loss of consciousness, convulsions, or stiffness may indicate epilepsy. 1 in 7 children will experience a febrile seizure or epileptic attack at some point in their lives. Children with neurocutaneous melanosis have a higher risk of epilepsy.
- Loss of skills: loss of strength, coordination, continence, or other skills that your child previously possessed is a reason to contact your doctor.
- Developmental delays: for example falling behind in developmental milestones, falling behind in school

## **Contact Information**

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