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## BETWEEN MOLE AND MELANOMA: ATYPICAL MELANOCYTIC TUMORS

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### ATYPICAL NEVUS / DYSPLASTIC NEVUS

MELTUMP

SAMPUS

IAMPUS

AIMP

ATYPICAL SPITZOID TUMOR

STUMP

MELANOCYTOMA

**Pigmented skin lesions** can be **benign** or **malignant**. There are also **lesions that fall somewhere in between**, borderline cases where neither the dermatologist nor the pathologist can say with certainty whether it is a benign **mole** (nevus) or a malignant **melanoma**. These borderline cases are also referred to as **atypical melanocytic lesions**, but there are many other names in use, which are discussed in this brochure.



Usually, skin abnormalities that look suspicious are removed in their entirety (**excision**) and sent to the pathology laboratory. The pathologist examines the tissue under a microscope and makes a diagnosis (what it is) and whether it is a malignant abnormality.

This decision determines the further course of action. If it is a benign mole, no further action is required. If it is melanoma, the area must be operated on again (**re-excision**), and the area where it was located is then excised more extensively, with a safety margin of usually 1 cm around the scar. Sometimes the pathologist will say that there are some malignant characteristics, but not enough to make it melanoma, and the area has been completely removed. In that case, no further action is required. And sometimes the pathologist says that the area, which has malignant characteristics but is not melanoma, has not been completely removed. In that case, it is usually recommended to operate again to ensure that the abnormality has been completely removed, but not with a margin of 1 cm around it, but much smaller, for example a few millimeters, or 5 mm.

The final advice given about surgery and follow-up checks depends on the type of tumor and the pathologist's assessment. This advice is laid down in guidelines. There are Dutch, European, and international guidelines, and they are not all exactly the same. Guidelines also change over the years, becoming more flexible or more strict.

### ATYPICAL NAEVUS / DYSPLASTIC NAEVUS

A **normal mole** is called a **nevus** (nevus naevocellularis). Moles that are cause for concern can be assessed by a dermatologist or general practitioner. The clinical picture, or how it looks, determines whether the mole is suspicious. The size, any growth, the color, whether there are multiple colors or abnormal shapes, and the symmetry are all taken into account. The dermatologist will usually also use a dermatoscope, a powerful magnifying glass with special lighting.

The assessment may be that it is a **normal mole**, with nothing unusual to be seen. In that case, no action is required. The assessment may also be that it does not appear to be a normal mole, but an **abnormal mole** (medical term: **atypical nevus**), or perhaps a **melanoma**. In that case, the spot is usually removed in its entirety and sent to the pathologist.



The pathologist's assessment is sometimes available after a few days, but it can also take weeks, especially if it is difficult and a lot of additional staining is required.

There are then several possible outcomes:

1. It is a normal mole.
2. It is a melanoma.
3. It is a **dysplastic nevus**: not a normal mole, nor melanoma, but something in between. There are some atypical characteristics, unstable cells, but not serious enough to call it melanoma.
  - 3a. The lesion has been completely removed.
  - 3b. The lesion extends locally to one of the edges or to the bottom, is not completely removed.
4. It is something completely different.

A **dysplastic nevus (atypical nevus)** could eventually develop into melanoma. That is why it is recommended that the nevus be removed in its entirety. So if the pathologist writes that the lesion extends to one of the edges or to the bottom, then another operation is needed, more extensive or deeper.

Some people have a lot of atypical moles, which increases their risk of developing melanoma. They often have family members with many atypical moles or with one or more melanomas. This may be a hereditary condition called **dysplastic nevus syndrome**. Other names for this condition are **FAMMM syndrome** (Familial Atypical Multiple Mole/Melanoma syndrome) or **familial melanoma**. If familial melanoma is suspected, DNA diagnostics can be performed.

Instead of dysplastic nevus, the pathologist may also make other diagnoses, such as **MELTUMP**, **SAMPUS**, **IAMPUS**, **AIMP**, **atypical spitzoid tumor**, **STUMP**, or **melanocytoma**. The term melanocytoma is relatively new. In 2018, the WHO proposed using this term for all intermediate forms between moles and melanoma. However, the other terms are still in use as well.

## MELTUMP

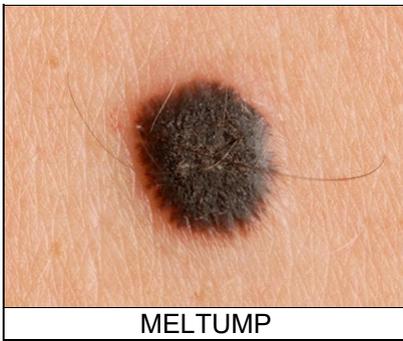
**MELTUMP** is an abbreviation for **melanocytic tumor of uncertain malignant potential**. **Melanocytic** means that the abnormality, like all other abnormalities discussed in this brochure, is composed of pigment cells (**melanocytes**). Tumor means lump or swelling. The term tumor does not mean that it is malignant. There are benign tumors and malignant tumors. Pigmented skin lesions are not always a lump or swelling; they can also be just a pigmented spot that is not raised. **Uncertain malignant potential** means that it could be a malignant tumor (a melanoma). The pathologist is then unsure whether it is benign or malignant.

## WHAT IS THE CAUSE OF A MELTUMP?

It is not known why a MELTUMP develops. A MELTUMP can develop anywhere on the skin. In principle, anyone can develop a MELTUMP, but they are more commonly seen in people with fair skin.

## WHAT DOES A MELTUMP LOOK LIKE?

A MELTUMP usually looks like a brown or black bump or spot. It actually looks like a mole. The diagnosis of MELTUMP can never be made based on how the spot looks. The diagnosis can only be made retrospectively by the pathologist, once the spot has been removed and sent for pathological examination. Lesions will be removed when there are reasons for this, such as growth, color differences, asymmetry, signs of unrest during examination with the dermatoscope, or itching, pain, or bleeding.



### HOW IS MELTUMP TREATED?

If the dermatologist thinks that a mole looks suspicious, the spot is usually removed in its entirety and sent to the pathologist. If the pathologist has diagnosed MELTUMP, the advice is to excise the area more extensively, with a safety margin of at least 5 mm around the scar, and if possible (in areas where there is plenty of space, not on the face) with a margin of 1 cm (as with melanoma).

### WHAT IS THE OUTLOOK FOR MELTUMP?

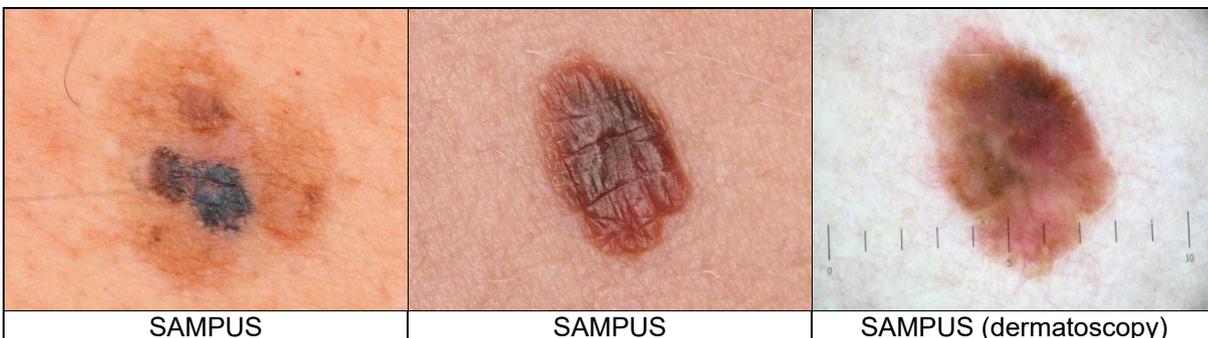
Once a MELTUMP has been completely removed, the treatment is complete. Follow-up checks are not necessary for MELTUMP. It is sufficient to check the skin yourself and, if new or changed areas appear, to visit your GP or dermatologist.

### WHAT ELSE CAN YOU DO YOURSELF?

Avoid sunburn. Check your skin regularly and see your doctor if you notice any new or suspicious spots.

### SAMPUS AND IAMPUS

**SAMPUS** and **IAMPUS** are actually exactly the same as MELTUMP, the only difference being that the lesions are not tumors (not raised or growing deep into the skin), but grow **superficially**. They usually look like slightly raised spots or completely flat pigmented spots. These are also diagnoses that can only be made retrospectively, by the pathologist, after the tissue has been examined.



**SAMPUS** stands for **superficial atypical melanocytic proliferation of uncertain significance**.

**IAMPUS** stands for **intraepidermal atypical melanocytic proliferation of uncertain significance**.

**Superficial** means that the abnormality continues to grow superficially and does not go deep anywhere.

**Intraepidermal** means that the abnormality is even more superficial, only in the upper layer of skin, the **epidermis**. The layer underneath is called the **dermis**. Between the epidermis and the dermis is a very thin but sturdy layer called the basement membrane. If tumor cells remain in the epidermis (**intraepidermal**) and do not penetrate that layer, this is good news because there is no risk of growth into the deeper layers or metastasis.

There is also a form of melanoma that remains in the epidermis (intraepidermal) and does not pass through the basement membrane. This is called **lentigo maligna**.

## AIMP

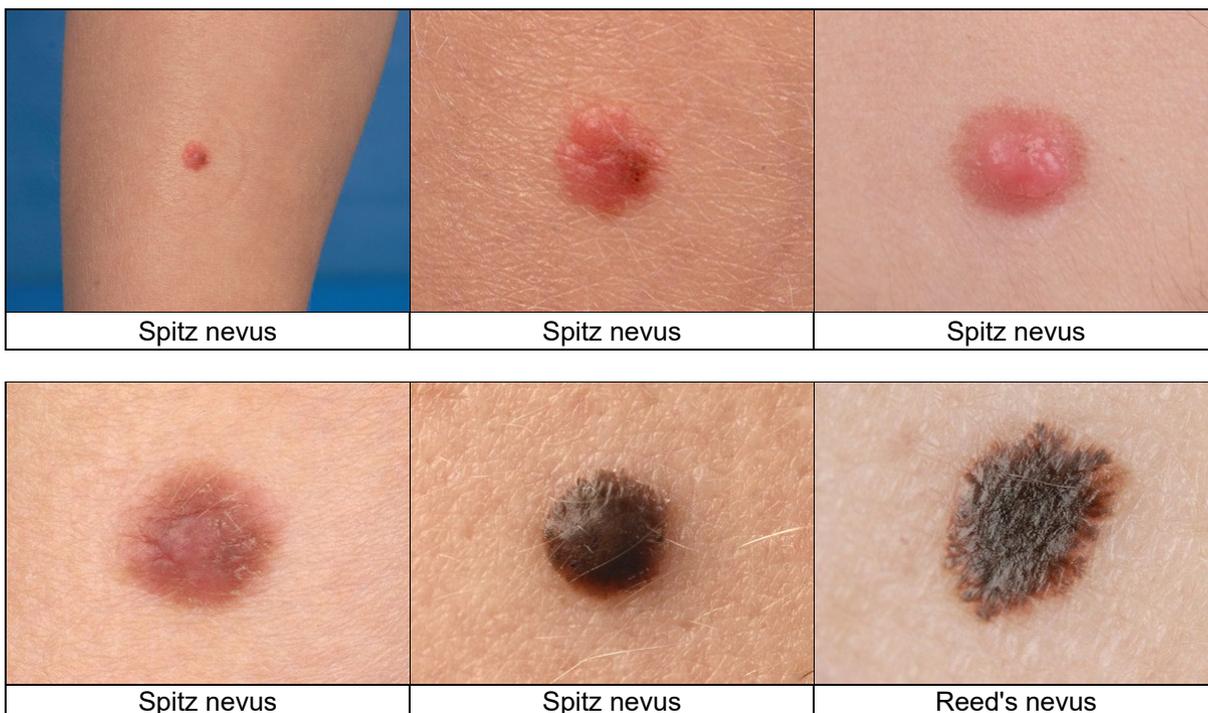
**AIMP** stands for **atypical intraepidermal melanocytic proliferation**. This is a descriptive term used only by pathologists. It means that there is an increase in atypical (not entirely normal) melanocytes in the epidermis.

## STUMP

**STUMP** stands for **Spitz tumor of uncertain malignant potential**. To understand what a STUMP is, we first need to explain what a Spitz tumor is. There are three different types of Spitz tumors: a **Spitz nevus**, an **atypical Spitz nevus**, and a **Spitz melanoma**.

### Spitz nevus

A **Spitz nevus** is a mole composed of **melanocytes** that have a distinctive **elongated shape**. These are also called spindle cell melanocytes. The name comes from the dermatologist Spitz, who first described the mole. A Spitz nevus is a benign, rare mole that occurs in children and young adults. A Spitz nevus looks like a pink, red, or reddish-brown, symmetrical, round bump measuring approximately 5 mm. Spitz nevi are mainly found on the face, neck, or legs. There is also a rare variant in which several Spitz nevi are grouped together in one area. There is also a variant that is very dark brown to black in color and flat. This is also known as a **Reed's nevus**. The nevus of Reed is usually found on an arm or leg. Its size can vary, up to a maximum of 1 cm in diameter.



### Atypical Spitz nevus

An **atypical Spitz nevus** (atypical Spitzoid tumor) is a mole composed of the same elongated melanocytes, but the difference is that these cells are **atypical, irregular**. The melanocytes exhibit characteristics that point toward malignant behavior, but not enough to diagnose melanoma. The pathologist can also distinguish between mildly atypical and severely atypical.



If the diagnosis is atypical Spitz nevus, it is usually recommended to remove the entire nevus, if this has not already been done, with a safety margin of a few millimeters to 5 mm.

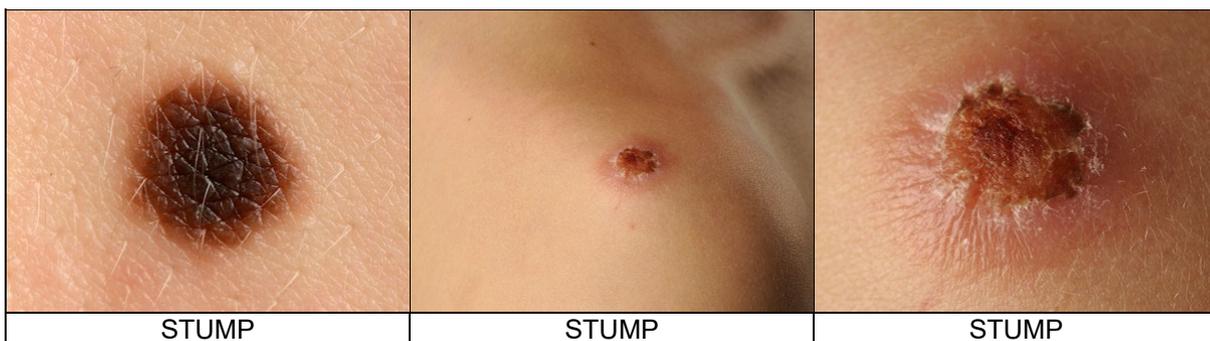
### Spitz melanoma / Spitzoid melanoma

A **Spitz melanoma** is also made up of elongated spindle-shaped melanocytes, but in this case, **malignant cells** are clearly present. It is a variant of melanoma and must also be treated as melanoma; the site must be removed again with a safety margin of 1 cm around the scar.

### STUMP

A **STUMP** (**Spitz tumor of uncertain malignant potential**) is a tumor composed of elongated melanocytes, in which the pathologist, as with MELTUMP, cannot clearly distinguish between benign and malignant. A STUMP could therefore be a Spitz melanoma and must therefore be treated in the same way as a melanoma. However, there is a difference: extensive research has shown that with STUMP, it is not necessary to maintain a large safety margin of 1 cm; 5 mm is sufficient.

Research is currently being conducted to determine whether re-excision of melanomas with a safety margin of 1 cm, and sometimes even 2 cm, is really necessary. It is possible that the guidelines will change on this point.



### MELANOCYTOMAS

In 2018, the WHO proposed **melanocytoma** as a new term for skin lesions that fall between benign moles and malignant melanoma. This term is not yet widely accepted. The old terms are still in use. However, it is possible that in the future, the term melanocytoma will be used instead of atypical or dysplastic nevus. The WHO also distinguishes between melanocytomas with few atypical features (**low-grade melanocytoma**) and melanocytomas with many atypical features (**high-grade melanocytoma**). For low-grade melanocytoma, the recommendation is to excise the lesions with a margin of a few millimeters (1-3 mm). For high-grade melanocytoma, the recommendation is to excise the lesions with a margin of at least 5 mm (5-10 mm).

### Spitz melanocytoma

The term melanocytoma is also increasingly used for **Spitz tumors**. The current term for abnormalities that fall between a **benign Spitz nevus** and a **malignant Spitz melanoma** is **atypical Spitz nevus** or atypical Spitzoid tumor. The WHO proposes replacing this term with **Spitz melanocytoma**, which can be further subdivided into **low-grade Spitz melanocytoma** and **high-grade Spitz melanocytoma**.

This new classification also has new consequences. What has not changed is the policy that suspicious pigmented skin lesions must always first be removed in their entirety with a narrow margin of 2 mm and sent to the pathology lab. If the result is Spitz nevus, no further action is required, as this is a benign mole. If the result is low-grade Spitz melanocytoma, no further action is required, as the 2 mm margin is sufficient. If the result is high-grade Spitz melanocytoma, the recommendation is to excise the area again with a margin of at least 5 mm (5-10 mm).

### Other melanocytomas

There are other types of melanocytomas, such as **pigmented epithelioid melanocytoma**, **deep penetrating melanocytoma**, and **BAP1-inactivated melanocytoma**, but these are all very rare.

