



## WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you understand more about primary localised cutaneous amyloidosis. It tells you what this condition is, what causes it, what can be done about it, and where you can find out more about it.

## WHAT IS PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS?

Primary localised cutaneous amyloidosis (PLCA) is a rare skin condition that causes an abnormal build-up of proteins called amyloids in the skin. The normal amyloid proteins start to change or mutate, forming twisted clumps that can build up in different organs, including the skin, and cause damage. This can lead to a range of symptoms and conditions.

Some forms of amyloidosis can affect many organs and tissues in the body: they are known as "systemic". Other forms only affect specific areas and are known as "primary" and "localised". Primary localised cutaneous amyloidosis (PLCA) mainly affects the skin.

There are three main forms of PLCA: **macular**, **lichen (papular)** and **nodular** (the rarest form) amyloidosis.

In macular and lichen amyloidosis, the abnormal amyloid protein is produced by skin cells and deposits occur only in the skin. In the rarer condition, nodular amyloidosis, the abnormal protein is produced by immune cells and can be associated with amyloid deposits in other body organs and myeloma (a form of bone marrow cancer).

PLCA more commonly affects people from Asia, South America and the Middle East. It usually starts in adult life and tends to persist for many years.

## WHAT CAUSES PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS?

The cause of PLCA is not fully understood. The most common trigger is repeated scratching and rubbing of the skin such as in chronic eczema. It is thought that frequent rubbing of the skin damages skin cells, causing them to release proteins which form amyloid in the skin.

## IS CUTANEOUS PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS?

PLCA does not usually run in families, but for about 1 in 10 people there is a history of the skin condition passing from one parent to a child. In rare cases of inherited PLCA, mutations in two genes have been found. These are called oncostatin M receptor-beta and interleukin (IL)-31 receptor-alpha, which are known to be related to the sensation of itching. Although it is not exactly known how this causes PLCA, these genes are important for producing and maintaining normal skin cells.

## WHAT DOES PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS FEEL AND LOOK LIKE?

- **Macular amyloidosis** is most often seen on the upper back in a symmetrical distribution (both sides evenly) as greyish-brown, slightly thickened skin. The skin may have a rippled appearance. Macular amyloidosis is usually very itchy.
- **Lichen amyloidosis** appears as clusters of small skin or dark coloured scaly bumps, which can merge together to form raised thickened areas, especially on the shins and lower limbs. The arms and back can also be affected. Lichen amyloidosis is usually very itchy.

- **Nodular localised cutaneous amyloidosis** can affect any part of the body in the form of one, a few, or many bumps ranging from a few millimetres to centimetres wide. They can appear as red, pink or brown raised lumps. Sometimes, several bumps fuse together to form raised areas of skin (plaques). Nodular amyloidosis is not usually itchy.

## HOW IS PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS DIAGNOSED?

When a diagnosis of cutaneous amyloid is suspected, your healthcare professional may request a skin biopsy. This means that a tissue sample will be taken under local anaesthetic and examined under a microscope. Pathologists can do special tests on the skin sample to check for the characteristic appearance of amyloid deposits which are found in the skin.

Other tests, including blood and urine tests may be carried out if it is necessary to investigate whether someone has an associated disease, especially myeloma.

## CAN PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS BE CURED?

Currently, there is no cure for PLCA. Treatments aim to manage the symptoms.

## HOW CAN PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS BE TREATED?

Many different treatments have been tried, but large studies are lacking and there is no preferred therapy.

For lichen amyloidosis and macular amyloidosis, the treatments are similar to those used for [eczema](#). They include antihistamine tablets, [corticosteroid creams and ointments](#), vitamin D ointment (e.g. calcipotriol) and narrowband ultra-violet B-light ([NB-UVB](#)). For thicker lesions covering the skin with a hydrocolloid dressing after application of a steroid ointment or cream

can improve the effectiveness, reduce irritation and flatten the skin lesions.

Oral medication options include [acitretin](#), [colchicine](#), [ciclosporin](#) (an immune-suppressive drug) and low doses of amitriptyline (an antidepressant drug).

Lesions of nodular amyloidosis can be removed surgically if they are causing symptoms, but they often re-grow. Removal, curettage and cautery (scraping the lump off and stopping the bleeding with heat) or destroying skin with 'ablative' lasers require a local anaesthetic injection. [Cryotherapy](#) (extreme cold to freeze and remove the abnormal tissue) can be done without local anaesthetic. Steroid injections may help to shrink the lumps.

## SELF-CARE (WHAT CAN I DO?)

The most important but hardest thing to do is to stop scratching.

- Anything you can do to take your mind off the itch will help reduce or stop the scratching. Try distraction techniques, keep your hands occupied or wear mittens / cotton gloves to reduce scratching, especially at night.
- You may find that laying a cool damp flannel on the skin can reduce the feeling of itch. A cooled [emollient](#) which has been kept in the refrigerator, or has added menthol, may also help.
- Keep the temperature in your bedroom cool (18-20°C) and use light and breathable bedclothes. If you feel hot and itchy during the day, a fan can help to cool the skin down. If possible, discuss work adjustments with your employer.
- Wearing layers of cotton clothing can be better than heavy, scratchy or synthetic fabrics. It is advisable to avoid wool or polyester clothing. These materials do not absorb sweat as easily, leading to overheating and itching. For more information on how to care for itchy skin,



visit <https://www.nhs.uk/conditions/itchy-skin/>

## WHERE CAN I GET MORE INFORMATION ABOUT PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS?

*Information on how to care for itchy skin:*

BAD patient information leaflet on pruritus - <https://www.bad.org.uk/pils/pruritus-itching/>

NHS advice - <https://www.nhs.uk/conditions/itchy-skin/>

*Web links to other relevant sources:*

<https://dermnetnz.org/topics/cutaneous-amyloidosis>

<http://emedicine.medscape.com/article/1102770-overview>

*Please note that the British Association of Dermatologists (BAD) provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.*

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

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*This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel*

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## BRITISH ASSOCIATION OF DERMATOLOGISTS

### PATIENT INFORMATION LEAFLET

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