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Myxoma syndrome

What is myxoma syndrome?

Myxoma syndrome is the name given to a group of diseases that are characterised by the presence of atrial myxomas (tumours of heart tissue) and [lentiginos](#) (pigmented spots on the skin). Often other abnormalities are associated with the different disorders and this has in some cases given rise to the naming convention of these conditions.

Myxoma syndromes include:

- LAMB (lentiginos, atrial myxomas, mucocutaneous myxomas, and blue naevi)
- NAME (naevi, atrial myxomas, myxoid neurofibroma, and ephelides (freckles))
- Carney syndrome (atrial, cutaneous and mammary myxomas, lentiginos, blue naevi, endocrine disorders and testicular tumours)

It has been suggested that these conditions may all be part of a spectrum of manifestations of the same disorder.

What are the clinical features of myxoma syndrome?

The clinical features of myxoma syndrome differ between the different types according to the associated abnormalities.

Myxoma syndrome	Clinical features
LAMB	<ul style="list-style-type: none"> • Lentiginos – most commonly occur on lips, face, sclera and vulva. Lesions are brown and may be up to 1cm in diameter. • Atrial myxomas – tumours of heart tissue that often originate in the atria (upper chamber of the heart). These may obstruct blood flow through the heart and cause fainting, shortness of breath, chest pain and other cardiac type symptoms. • Mucocutaneous myxomas – appear as papules or nodules at various sites on the body, including the breasts, shoulders, oral mucosa and tongue. • Blue naevi – dark blue moles appearing anywhere over the body
NAME	<ul style="list-style-type: none"> • Similar features to those in LAMB syndrome. Additional feature is myxoid neurofibroma, which is usually a solitary lesion occurring on the extremities. • Ephelides (freckles) usually appear at birth and become more obvious in the summer. Lesions vary from pale to dark brown and most commonly found around the neck, trunk and thighs.
Carney	<ul style="list-style-type: none"> • Cutaneous myxomas often found on the eyelids, nipples, scalp, face, ears, neck, oral mucosa, trunk and limbs. • Lentiginos are brown to black and 0.2–2mm in diameter. They are widespread throughout the body and often join together to form brown patches. • Mammary involvement includes gynaecomastia (enlarged breasts) and myxomatous enlargement of the stroma

- Atrial myxomas are either single or multiple and often result in fibrosis or calcification
- Other features include testicular tumours and endocrine overactivity (e.g. pituitary adenomas and Cushing syndrome).

What causes myxoma syndrome?

Some cases of myxoma syndrome are inherited in an autosomal dominant manner, which means that if one parent is affected there is a 50% chance of passing this mutated gene on to their offspring. Mutations in the PRKAR1A gene are described. This encodes the type 1A regulatory subunit of protein kinase A. This molecule appears to have a role in cell cycle regulation, growth, and/or proliferation.

Many cases of myxoma syndrome are sporadic and do not appear to have this gene mutation. The exact cause is unknown.

What treatments are available?

A team of specialist doctors including a cardiologist, endocrinologist, and dermatologist should manage myxoma syndromes.

If necessary, cutaneous lesions (lentigines, naevi, skin tumours) can be removed through the use of [chemical peels](#), [cryotherapy](#), [laser treatments](#) or [surgical excision](#). For some patients, treatment with [topical retinoids](#) and [hydroquinone](#) cream may be helpful.

Atrial myxomas can be surgically removed. The surgery is safe, with a mortality rate of 2.2%. Occasionally part of the tumour can break off during surgery and get stuck in other blood vessels, causing other problems such as stroke or pulmonary embolism.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Lentigo](#)
- [LEOPARD syndrome](#)
- [Brown spots and freckles](#)

Other websites:

- [Atrial myxoma](#) - emedicine, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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