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## Juvenile xanthogranuloma

Juvenile xanthogranuloma (JXG) has been previously called naevoxanthoendothelioma.

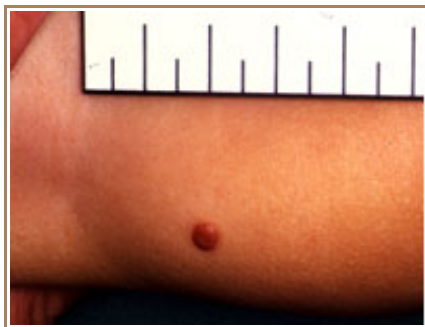
### What is juvenile xanthogranuloma?

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Juvenile xanthogranuloma is a type of [non-Langerhan's cell histiocytosis](#) (Class IIb). It presents as skin lesions in infants and young children, more often males, and is present at birth in 20% of cases. However, 10% of cases are adults. It is more common in Caucasians than in those of oriental origin. The cause is unknown.

At first the lesions are smooth and pink bumps, but later develop a yellowish appearance and may become scaly. Most are under 0.5 cm in diameter (papules), but giant nodules may be as large as 2 cm. They may arise on any site of the body, but more frequently appear on the trunk and upper extremities. Occasionally they also appear in the eye or internal organs.

### Juvenile xanthogranulomas



### Related conditions

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Juvenile xanthogranuloma is difficult to distinguish from several other conditions.

- Benign cephalic histiocytosis
- Generalised eruptive histiocytosis
- Papular xanthogranuloma

They are increasingly thought of as the same condition.

### Benign cephalic histiocytosis

- Usually occurs in infants less than one year of age
- Presents with multiple small reddish–brown bumps on the skin, mostly on the head and neck
- Bumps spontaneously resolve over months to years and often no visible mark is left
- Repeated attacks may occur

### Generalised eruptive histiocytoma

- Usually affects adults
- Presents with hundreds of reddish–brown, blue–red or skin–coloured papules (less than 1cm) on the skin of the trunk, face, upper arms and thighs
- Recurrent crops of lesions may appear and spontaneously resolve after several months
- May leave scars

### Papular xanthogranuloma

- Occurs in children and adults
- Papules occur both in the skin and the lining of the mouth
- Spontaneously resolves in children but may persist in adults
- There is no effective treatment at this time

## How are these disorders diagnosed?

The diagnosis of juvenile xanthogranuloma and related conditions can be confirmed by a [skin biopsy](#). The lesions are composed of collections of histiocytes. In older lesions, the histiocytes may appear foamy, filled with lipids (fats) or hugely enlarged (giant cells).

Despite the fat–filled histiocytes in the skin, the levels of lipids in the blood are quite normal and juvenile xanthogranuloma and related disorders are not usually associated with any serious abnormality. However 20% also have [café au lait birthmarks](#). Multiple café au lait birthmarks are associated with [neurofibromatosis type 1](#).

## Treatment

Unless they occur in the eyes, juvenile xanthogranulomas are harmless growths and disappear eventually over 2 to 3 years, usually without scarring.

Although individual lesions can be cut out, this will leave a scar. Removal is seldom necessary.

### Related information

#### On DermNet

- [Birthmarks](#)
- [Skin lesions](#)
- [Non–Langerhan's cell histiocytoses](#)
- [Histiocytoses](#)

#### Other websites:

- [Juvenile xanthogranuloma \(nevoxanthoendothelioma\)](#) – emedicine dermatology, the online textbook

#### Books about skin diseases:

See the [DermNet NZ bookstore](#)

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

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