

What is a juvenile xanthogranuloma?

A juvenile xanthogranuloma (JXG) is a benign (non-cancerous) skin bump. JXGs can appear anywhere on the skin and at any age but most often develop within a child's first years of life. JXGs can be present at birth. While 90% of the time there is only one bump, some patients may have multiple JXGs. Rarely, especially when there are many JXGs, there could be involvement of organs other than skin. (SEE RARE CASES BELOW)

▼ WHAT CAUSES A JUVENILE XANTHOGRANULOMA?

JXGs are collections of immune cells in the skin. It is not currently known what causes JXGs to form.

▼ WHAT DOES A JUVENILE XANTHOGRANULOMA LOOK LIKE?

JXGs are non-painful, firm, round bumps. These start as a pink-red color and become more orange-brown-yellow over time. However, in skin of color they may appear more flesh-colored or dark red/brown. They vary in size but are usually less than 2 cm in diameter. Sometimes, the center of the bump might open up and become crusted or scab over. They typically do not cause pain or other symptoms.

▼ HOW IS A JUVENILE XANTHOGRANULOMA DIAGNOSED?

Your doctor can diagnose a JXG with a physical examination. They may use a handheld microscope to get a closer look at the bump. In some cases, the doctor might recommend a biopsy to confirm the diagnosis. This small procedure can be done in the doctor's office.

▼ WHAT IS THE TREATMENT OF JUVENILE XANTHOGRANULOMA?

JXGs usually go away and improve on their own so treatment is typically unnecessary. JXGs often disappear over 3-6 years. They can leave behind skin color changes or a scar after they are gone. Removal is not usually recommended. A provider should see your child if their JXG is causing issues such as pain, bleeding, or rapid growth.

RARE CASES

Although most JXGs do not usually cause any problems, there can be rare complications.

ULCERATION (SORE OR SCAB ON THE JXG)

Ulceration is higher risk in the rare "giant JXG" that is over 2 cm in size. Your doctor will help you care for your child if they develop an ulcer; this often involves wound care and pain relief if needed.

RARE CASES (CONT.)

ORGAN INVOLVEMENT

Internal involvement is higher risk in children with multiple JXGs. These growths are usually only present on the skin, but in rare cases may be associated with changes in the eye. Because of this, in certain situations, your doctor may recommend that your child see an ophthalmologist, which is a doctor who specializes in diagnosing and treating eye disease. Even more rarely, JXGs can form in the internal organs such as the liver and spleen. If that happens, your doctor may recommend an ultrasound to evaluate. For most patients this won't be necessary.

There are very rare reports of children with multiple JXGs developing leukemia. This is more common in children with a genetic disorder called neurofibromatosis. The vast majority of children with JXGs do NOT develop leukemia and monitoring for leukemia with blood work (labs) is not recommended. Rather, it is better to monitor by regular checkups and watching for unusual symptoms such as weight loss, consistent fevers, or easy bruising.



The Society for Pediatric Dermatology 8365 Keystone Crossing, Suite 107 Indianapolis, IN 46240 (317) 202-0224 www.pedsderm.net

The Society for Pediatric Dermatology and Wiley Publishing cannot be held responsible for any errors or for any consequences arising from the use of the information contained in this handout. Handout originally published in *Pediatric Dermatology*: Vol. 39, No. 1 (2022).

© 2022 The Society for Pediatric Dermatology

Contributing SPD Members: Irene Lara-Corrales, MD Christine Lauren, MD

Committee Reviewers: Heather Brandling-Bennett, MD Sheilagh Maguiness, MD

Expert Reviewer: Amy Paller, MD