

Fact Sheet

Juvenile Xanthogranuloma Disease

What is Juvenile xanthogranuloma disease?

Juvenile xanthogranuloma, also known as JXG, is a rare, non-Langerhans cell histiocytosis that is usually benign and self-limiting. It occurs most often in the skin of the head, neck, and trunk; it can also occur in the arms, legs, feet, and buttocks. JXG can affect the eye, most commonly in young children with multiple skin lesions. Less commonly, JXG may involve locations such as the lung, liver, adrenal gland, appendix, bones, bone marrow, pituitary gland, central nervous system, kidney, heart, small and large intestines, and spleen.

JXG involves the over-production of a kind of histiocyte called a dendritic cell (not a macrophage). These cells then accumulate and lead to various symptoms, depending on location. The cause of this disease is not known.

JXG mainly affects infants and small children with an average age of 2 years; however, it can also occur in adults of all ages. Most frequently, it presents as a single skin lesion which varies in size; children less than 6 months of age are more likely to have multiple lesions. It occurs at birth in about 10% of patients, and more males are affected than females. When JXG occurs in adults, it tends to be more complicated and is not known to spontaneously improve. The total number of patients with JXG is not known, but it may be higher than reported since this disease is sometimes misdiagnosed or may spontaneously improve in children.

What are the symptoms of Juvenile xanthogranuloma disease?

Symptoms of JXG may include:

- » Reddened, yellowish or brownish, slightly raised, and rubbery bumps on the skin
- » Abnormalities in blood counts
- » Abnormalities in blood liver tests
- » Elevated inflammation level (sedimentation rate) in the blood
- » Potentially diabetes insipidus

What is the treatment for Juvenile xanthogranuloma disease?

Patients with minimal lesions usually need no therapy. Surgical removal may be undertaken for several reasons: to obtain a biopsy for diagnosis; when there is an organ-function problem because of disease; for cosmetic reasons; or to remove scar tissue. Apart from these reasons, skin-only JXG in children is usually observed without therapy. For patients who have symptomatic or rapidly growing disease, treatment with chemotherapy or low-dose radiation has been reported, although there is no standard treatment that is agreed upon. With eye involvement, steroids may be applied to the surface of a lesion, injected, or taken in pill form. Rarely low dose radiation treatment may be given to prevent visual loss.

Histiocytosis Association

The Histiocytosis Association is a global nonprofit organization dedicated to addressing the unique needs of patients and families dealing with the effects of histiocytic disorders while leading the search for a cure. It is the only organization of its kind, connecting the patient and medical communities to:

- » Grow and share knowledge of histiocytic disorders,
- » Provide critical emotional and educational support to patients and families, and
- » Identify and fund key research initiatives that will lead to a world free of histiocytic disorders.

Histiocytic disorders affect fewer than 200,000 people. It is thus considered an "orphan disease" and, as such, does not receive substantial government funding for research. The Association relies on contributions from corporations, foundations and individual donors to fund critical research, build awareness and conduct community outreach initiatives.

For more information, contact:

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