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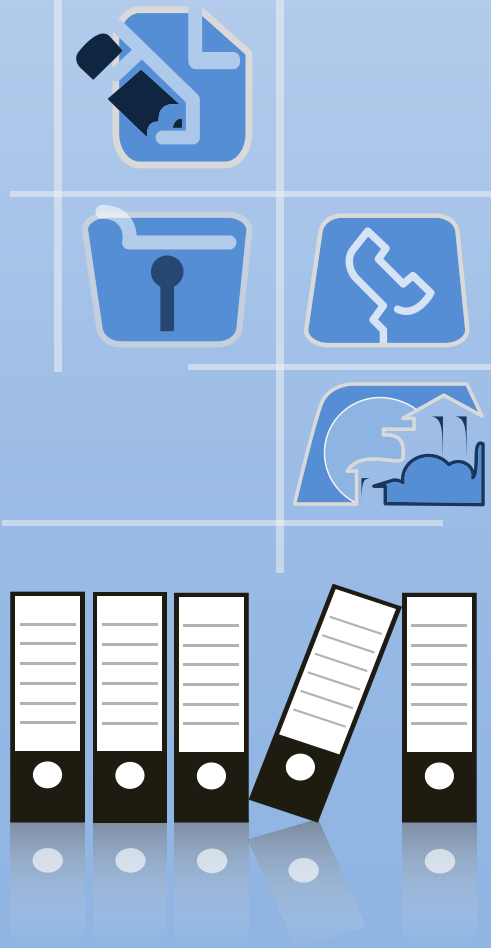
Juvenile Xanthogranuloma JXG



Histiocytosis UK

Introduction

Despite the misery it causes, Histiocytosis is too rare a disease to have generated substantial research in medical circles. Unfortunately, for every child or adult fighting for his or her life, the pain and suffering are just as severe for children and adults afflicted with other better known disorders receiving funding.

For the children and adults battling these illnesses, there is now reason to hope. To ensure the research continues, we ask for your help, to complete the funding puzzle.

Our awareness and research programmes provide a beacon of hope for the many children and adults battling Histiocytosis, to ensure this research continues we ask you to pledge your support.

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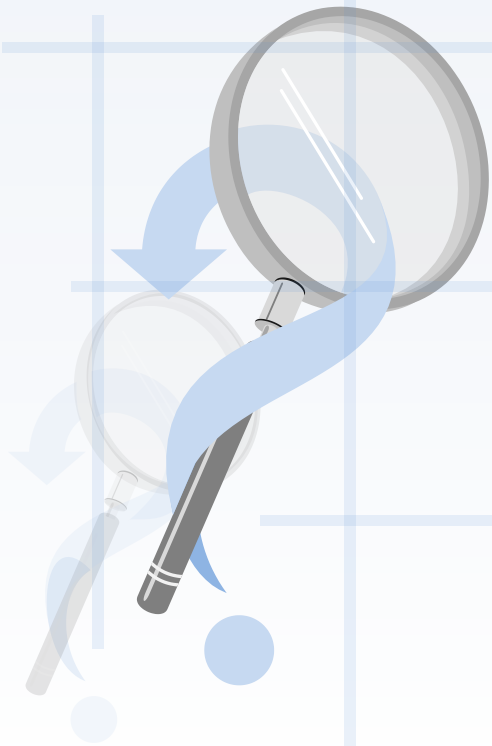

What is Histiocytosis?

Histiocytosis is an umbrella term applied to a group of rare diseases, characterised by increased numbers of white blood cells called histiocytes in the blood and tissues. In all forms of histiocytosis, these cells, which are part of the protective immune system, begin to attack the body, targeting many organs of the body including the bone marrow, liver, spleen, lungs, skin, bone and brain.

The prognosis for patients varies greatly depending on the form of histiocytosis.

Please be advised that all the information you read in this document is not a replacement for the advice you will get from your consultant and their team.

Who we are?

Histiocytosis UK is a registered charity dedicated to promoting and funding scientific research into uncovering not only the causes of all histiocytic diseases, which include Langerhans Cell Histiocytosis and Haemophagocytic Lymphohistiocytosis, but also ensuring early diagnosis, effective treatment and a cure.

The Charity aims to support patients and their families by means of information and awareness as well as raise public and professional awareness of histiocytic disorders. Its team of Trustees include the UK's leading paediatric LCH and HLH specialists.

Juvenile Xanthogranuloma JXG

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 but 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.

This disease may have been first reported by Rudolf Virchow in 1871 and again in 1905 by H.G. Adamson. In 1954, it was named juvenile xanthogranuloma to reflect the appearance of the cells under a microscope.

JXG mainly affects infants and small children with an average age of 2 years, although it can also occur in adults of all ages. Usually it presents as a single skin lesion, which varies in size, but 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.

Skin lesions are self-limited and rarely require treatment in most patients. Those with large abdominal masses, liver, bone marrow, or central nervous system involvement may do well with treatment such as chemotherapy similar to that used for Langerhans Cell Histiocytosis. Because this disease is so rare, no large studies have been performed, and there is no established, proven treatment for the more complicated cases.

Juvenile xanthogranuloma (JXG)

The questions below specifically relate to Juvenile xanthogranuloma (JXG).

1. What causes juvenile xanthogranuloma (JXG)?

JXG involves the over-production of a kind of histiocyte called a dendritic cell. What triggers these cells to accumulate is not known.

2. Is there a cure for JXG?

We usually do not use the term "cure" with this disease, although most patients with only skin or soft tissue JXG have spontaneous remission over time without treatment. Children with liver, bone marrow, CNS involvement and masses in the abdominal cavity usually survive with chemotherapy treatment. There is no established period of inactive disease before JXG is considered cured.

3. What are the different therapies/treatments commonly used to treat JXG?

Patients with a single lesion or just a few lesions, as well as children with skin-only JXG often require no therapy. Surgical removal may be required if the mass is causing organ dysfunction. A small percentage of patients with rapidly growing disease may require treatment with chemotherapy or low-dose radiation, however there is no agreed standard. Steroids have been used to treat eye lesions and in some cases low dose radiation has been effective in preventing visual loss.

4. Can an infant be tested at birth for JXG?

A biopsy of the affected tissue, rather than a blood test, is required for diagnosis and would therefore not be appropriate as a routine test unless this disease is suspected.