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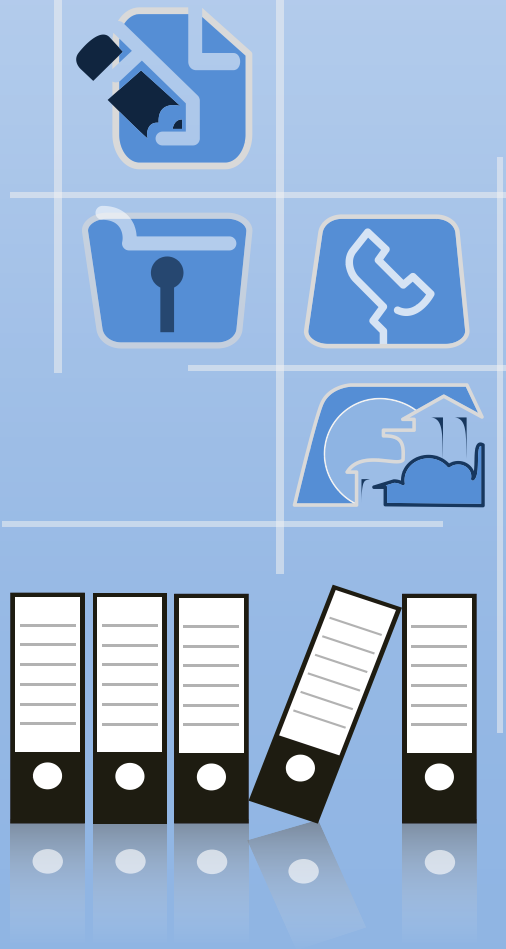
Rosai Dorfman Disease



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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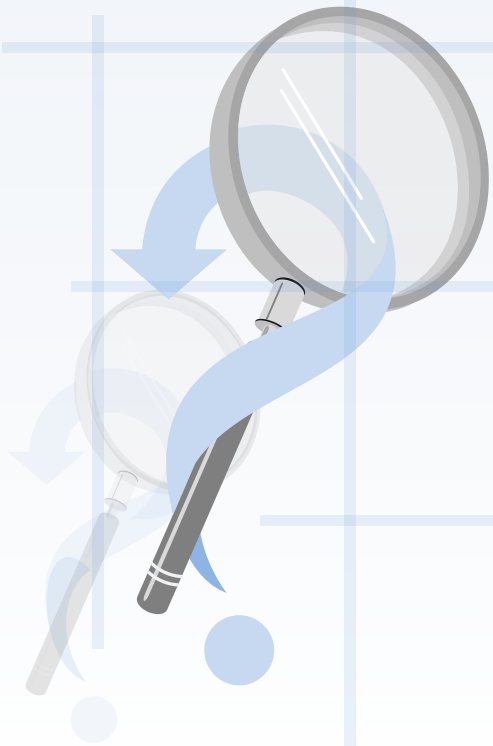

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What Is Histiocytosis

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.

Sinus Histiocytosis with Massive Lymphadenopathy (SHML)

Rosai-Dorfman disease (RD), also known as sinus Histiocytosis with massive lymphadenopathy (SHML), is a rare histiocytic disorder which involves the over-production of a type of white blood cell called non-Langerhans-cell sinus histiocytes. These cells then accumulate, most often in the lymph nodes but sometimes in other areas of the body and can lead to organ damage. The reason for over production of these cells is not known, although many possibilities have been considered, including viral, bacterial, infection, environmental, and genetic causes.

In 1969, two pathologists, Juan Rosai and Ronald Dorfman, reported a distinct histiocytic disorder in several patients with massive enlargement of the lymph nodes, as well as other symptoms. They named this condition sinus histiocytosis with massive lymphadenopathy, and the disease has since come to be known as Rosai-Dorfman disease.

The true number of RD cases is not known, although it does occur worldwide and seems to affect equal numbers of males and females. It is most commonly seen in the first 10 years of life, but it also occurs in adult patients.

Because this disease is so rare, no large studies have been performed, and there is no established, widely-accepted treatment. However, RD is usually not life-threatening, and many patients do not require treatment.

FAQ – Rosai Dorfman

Rosai-Dorfman Disease.

1. What causes Rosai-Dorfman?

Rosai-Dorfman involves over-production of a type of white blood cell called a non-Langerhans-cell sinus histiocyte. The cause of this over-production is not yet known.

2. Is there a cure for Rosai-Dorfman?

While many patients go into remission and live normal lives with or without treatment, we usually do not use the term “cure.” There is no established period of inactive disease before RDD is considered cured.

3. What are the different therapies/treatments commonly used to treat Rosai-Dorfman?

Many Rosai-Dorfman patients do not require treatment. Some patients with severe or persistent disease may need treatment with surgery, steroids, and/or chemotherapy. Rarely, radiation therapy is used.

4. Can an infant be tested at birth for Rosai-Dorfman?

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.

5. What causes chronic pain in adults with Rosai-Dorfman?

Some pain and cramping can be a side effect of treatment, such as vinblastine and steroids. Pain may also be directly related to active disease. In cases of more chronic pain, some researchers suspect that cytokines, which are a type of messenger, stimulate white blood cells to release inflammatory molecules that produce pain.

Possible Side Effects

What are the side effects of vinblastine?
Side effects include:
a. Low blood counts (with higher risk of infection)
b. Mild nausea/vomiting/constipation
c. Easily sunburned
d. Skin irritation at site of injection
e. Thin or brittle hair
f. Fatigue
g. Bone pain
h. Hoarseness
i. Seizures
j. Shortness of breath
k. Nerve damage (especially in adults) with tingling, numbness and/or pain of the hands and feet

What are the side effects of prednisone?
Side effects include:
a. increase in blood sugar
b. Increase in appetite
c. Heartburn
d. Bloating/fluid retention/weight gain
e. Difficulty sleeping
f. Mood/behavior/personality changes
g. Higher risk of infection
h. Slow wound healing
i. Muscle weakness
j. Loss of bone calcium
k. Increased hair growth
More unusual side effects may include:
a. Problems with vision/eye pain
b. Seizures
c. Confusion
d. Muscle twitching
What are the side effects of methotrexate?
Side effects include:
a. Mouth sores/swollen, tender gums
b. Nausea/vomiting/diarrhea/decreased appetite
c. Low blood counts
d. Dizziness/drowsiness
e. Headache
More unusual side effects may include:
a. Blurred vision or loss of vision
b. Seizures
c. Confusion
d. Weakness/difficulty moving one or both sides of the body
e. Loss of consciousness
f. Lung damage
g. Allergic reactions

What are the possible side effects of 6-MP (mercaptopurine)?
More common signs/symptoms include:
a. Low blood counts (red cells, white cells, and clotting cells)
b. Nausea/vomiting/decreased appetite
c. Headache
d. Weakness/fatigue/achiness
e. Rash/darkening of the skin