HISTIOCYTOSIS ASSOCIATION™
A Rare Community

Fact Sheet

Rosai-Dorfman Disease

What is Rosai-Dorfman disease?

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 sinus histiocyte. These cells accumulate and can lead to organ damage. This occurs most-often in the lymph nodes, but may occur in other areas of the body as well. The reason that these cells over-produce 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 name 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 research 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.

What are the symptoms of Rosai-Dorfman disease?

Symptoms of RD may include:

- Fever
- Paleness/anemia
- Weakness
- Weight loss
- Headaches
- Shortness of Breath
- Lymph node enlargement
- Nosebleeds
- Blockage or discharge of the nose

What is the treatment for Rosai-Dorfman disease?

It is believed that 70% to 80% of patients have spontaneous improvement of symptoms without treatment, although they may have alternating episodes of worsening and relieving of symptoms for a long period of time. Some patients with severe or persistent disease or cases where organ function is threatened (such as breathing obstruction or kidney failure) may require treatment with surgery, steroids, and/or chemotherapy. In rare cases, radiation therapy may be used.

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:

Histiocytosis Association
332 North Broadway | Pitman, NJ 08071 USA

Phone: +1 856-589-6606 | Fax: +1 856-589-6614
www.histio.org | info@histio.org
Federal Tax ID # 22-2827069