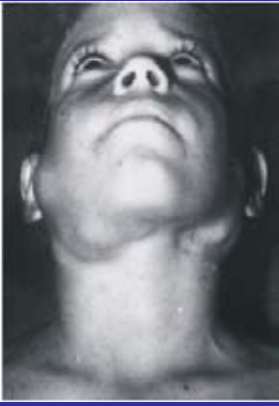


ROSAI-DORFMAN DISEASE



Rosai-Dorfman Disease occurs when normal histiocytes collect in the lymph nodes, typically of the cervical chain. This is a benign process unless it causes respiratory distress. A biopsy of an involved lymph node shows *sinus histiocytosis with massive lymphadenopathy (SHML)*. Treatment may be necessary if there is airway

compromise, and may consist of chemotherapy or steroids.

JUVENILE XANTHOGRANULOMA

Juvenile Xanthogranuloma (JXG) is a benign skin papule seen in infants & children. These lesions are typically orange or red-brown papules as shown in the picture above. They typically occur near the eyes, forehead and face, but can also be found in all locations. Treatment is not necessary unless the JXG compromises the function of the spinal cord or liver.



HOW TO REFER PATIENTS FOR HISTIOCYTOSIS

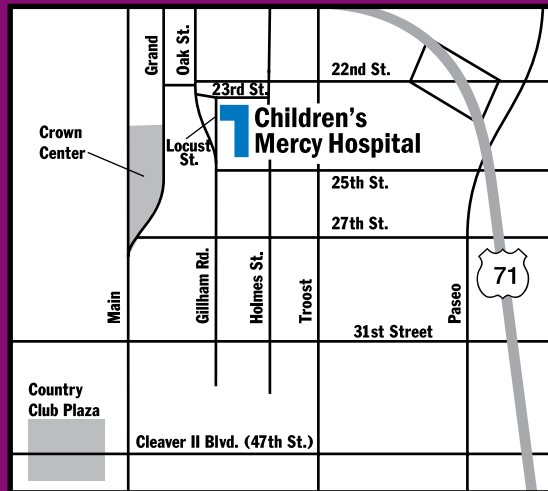
Referrals can be made by calling the Children's Mercy physician referral line:

1-800-GO-MERCY

or by simply calling (816) 234-3265 during business hours (9 a.m. to 5 p.m., Mon-Fri) for the Pediatric Hematology/Oncology Division.

Children's Mercy Hospital offers a wide variety of subspecialties, including Pediatric Hematology/Oncology, Pediatric Neurosurgery, Pediatric Dermatology and Pediatric Endocrinology

Children's Mercy Hospital
2401 Gillham Road
Kansas City, Missouri 64108
(816) 234-3000



REVIEW OF

histiocytosis

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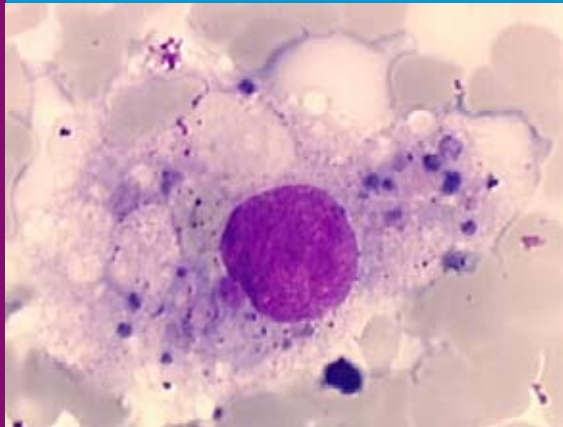
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02/09

review of histiocytosis

There are four main types of histiocytosis seen in children

1. Langerhans Cell Histiocytosis (LCH)
2. Hemophagocytic Lymphohistiocytosis (HLH)
3. Rosai-Dorfman
4. Juvenile Xanthogranuloma (JXG)



Normal Histiocyte in bone marrow

LANGERHANS CELL HISTIOCYTOSIS

Langerhans Cell Histiocytosis (LCH) has also been known as

- Histiocytosis X
- Eosinophilic Granuloma
- Letterer-Siwe disease
- Hans-Schuller-Christian syndrome

LCH may affect only one organ system, or multiple organ systems. There are three typical presentations for LCH: *single organ system disease*, *multi-system disease (low risk)* and *multi-system disease (high risk)*. Patients with **disease in the liver, bone marrow and/or spleen are considered high risk.**

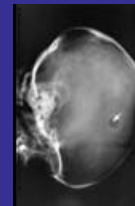
Single Organ System Disease

The two organ systems most often involved in single system LCH are the **skin** and **bone**. In general, single organ involvement has a *very high survival rate, and requires little if any treatment.*

The rash seen is typically red papules. It may present as a **diaper dermatitis that does not improve with standard topical therapy.**

This rash may occur in skin folds, on the entire trunk or behind the ears. It may also manifest as a **chronically draining ear or jaw pain with loose teeth.**

The bone involvement can be investigated with plain XR films of the affected area. The XR will show **lytic lesions** (arrow) and can be a single or multiple lesions. Treatment consists of curettage or steroids. The reactivation rate in single bones is <15 percent and in multiple bones is 25 percent. Survival for bone only LCH is 100 percent.



Multi-System Disease

High risk: organs affected

- Bone marrow: *anemia, neutropenia and thrombocytopenia on complete blood count*
- Liver: *abnormal liver function tests*
- Spleen: *splenomegaly*

Low risk: two or more organ systems without comprising 'risk' organs. Treatment consists of biopsy followed by chemotherapy. Reactivation of disease may occur in 30-50 percent of cases. Survival approaches 99 percent, with some incidence of diabetes insipidus (DI).

Diabetes Insipidus (DI) occurs due to LCH compressing the hypothalamic-pituitary axis causing decreased anti-diuretic hormone (ADH). Parents may report **polydipsia** and **polyuria**, but it is important to ask about these symptoms. Patients with DI may also develop growth hormone deficiency and short stature.

Please refer any questions or patients you think may have LCH to Children's Mercy Hospital, Division of Hematology/Oncology at (816) 234-3265.

HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

Hemophagocytic Lymphohistiocytosis (HLH) has been called familial erythrophagocytic lymphohistiocytosis and viral-associated hemophagocytic syndrome. HLH is a rare disease that occurs in an autosomal dominant inherited form, and an acquired form. Children with HLH have **abnormal blood counts, fevers, lymphadenopathy** and can present identical to those with septic shock. There are more specific tests for HLH like natural killer cell function and CD25 tests that support the diagnosis. Treatment consists of chemotherapy \pm bone marrow transplant.