

RARE HISTIOCYTOSES COMMITTEE REPORT - JULY 2019

July 8, 2019

Members:The steering Committee consists of - Oussama Abla (Chair), Eli Diamond, Eric Jacobsen, Julien Haroche, Jean-Francois Emile, Jorge Braier, Benjamin Durham, Jennifer Picarsic, Zdenka Krenova, Akira Morimoto, Michael Girschikofsky & Ron Jaffe.

Activities:

1. International Rare Histiocytic Disorders Registry (IRHDR):The registry started enrolling patients with RHDs in April 2015 after ethics approval at the Hospital for Sick Children, Toronto, Canada. PI is Dr O. Abla & Co-PI is Dr Jim Whitlock. So far, 13 international participating centers are currently open for study enrollment. A total of 95 patients have been enrolled to study since the initiation, including 44 cases of JXG, 25 cases of RDD, 19 cases of ECD and 7 cases of malignant histiocytoses. Most diagnoses have been confirmed, but central pathology review is still ongoing on few of these cases. Many members of the committee have been actively enrolling patients. A biobank and the possibility of a telephonic consent have been recently approved by central IRB. Preliminary reports under the form of an abstract and a manuscript are planned for 2020.

2. Juvenile xanthogranuloma position paper:New paper similar to the ECD and RDD ones is currently underway. All members were invited to participate in the writing. Clinical figures, imaging and genome sequencing pictures were encouraged. The aim is to submit the paper by October 2019.

3. Malignant Histiocytosis position paper:the writing group consists of Dr E. Jacobsen, Dr J. Visser, Dr O. Abla, Dr J. Picarsic and Dr JF. Emile. The first draft of the paper is underway.

4. A Multicenter Treatment Trial with Sirolimus in patients with Rosai-Dorfman Disease:The committee discussed this proposal which was presented by Dr Abla in collaboration with Dr K. Nichols in February 2019. The proposal was well received by members of the committee, and was presented and favorably viewed at the NACHO group meeting in Memphis-April 2019. The future aim is to submit a grant to fund this multicenter treatment trial.

5. Other Projects:have been discussed by members of the committee including Pediatric Mixed Histiocytosis Study and an international Familial RDD case series. Other members of the Histiocyte Society will be sought to champion these projects.