FOR IMMEDIATE RELASE:

Erdheim-Chester Disease Declared a Histiocytic Neoplasm
The World Health Organization reclassifies the rare disease

DeRidder, LA - May 18, 2016 - The World Health Organization headquartered in Geneva, Switzerland has reclassified Erdheim-Chester Disease as a histiocytic neoplasm. This means the rare disease is now considered a slow-growing blood cancer that may originate in the bone marrow or a precursor cell. The March announcement represents a significant step toward improving the support for suffers.

The reclassification comes after expert findings that the majority of ECD patients harbor mutations in the MAPK/ERK pathway. These genetic mutations lead to targeted therapeutic treatments. Research shows that the lesions and tumors associated with this disease have acquired mutations, with no known inherited abnormalities.

The WHO announcement has been published in Blood Journal's March 2016 edition in an article entitled, "The 2016 revision of the World Health Organization (WHO) classification of lymphoid neoplasms" by Steven Swerdlow, et. al.

Prior to the reclassification, ECD was an unclassified disorder and commonly described as a non-Langerhans histiocytosis. ECD is still considered to have a strong inflammatory component, common with histiocytoses.

ECD results in the overproduction of histiocytes, a type of white blood cell created in the bone marrow. These cells collect in different organs resulting in ECD lesions and tumors that can manifest in several organs of the body including the brain, heart, lungs, kidneys, and long bones amongst others.

With no FDA approved treatments for ECD, all treatments are considered off-label.

Some insurance plans decline to pay for treatments used off-label on the grounds that these uses are "experimental" or "investigational", even when indications strongly point to their effectiveness.

However, in the US, legislation requires coverage of medically appropriate cancer therapies including off-label uses recognized by peer-reviewed literature. The new Blood Journal article will support ECD patients in this way.

Current trials and studies associated with ECD treatments are showing great promise and are being published in peer-reviewed articles. Some research shows that targeted cancer-fighting drugs result in dramatic improvements in those afflicted with ECD, placing ECD at the forefront of precision medicine.

The announcement that ECD is a histiocytic neoplasm will make it easier for many patients to receive treatments as insurance plans will find it harder to deny payment.

ECD remains a largely overlooked disease, and increased recognition by clinicians and pathologists is necessary for effective diagnosis and treatment.

In addition to insurance support, being classified as a type of cancer should allow ECD patients to obtain additional help from cancer support organizations.

Learn more about the WHO classification by reading the abstract for "The 2016 revision of the World Health Organization (WHO) classification of lymphoid neoplasms".

Erdheim-Chester Disease is an ultra-rare condition with no known cause. It is a rare non-Langerhans histiocytic disorder that is challenging to diagnose and treat. It is considered an inflammatory histiocytic neoplasm. The accumulation of histiocytes in tissue and organs characterizes ECD. Histiocytes are cells that normally fight infections. The infiltration of the histiocytes causes tissue and organs to become dense and fibrotic, leading to organ failure unless finding a successful treatment.

The ECD Global Alliance is a 501(c)(3) non-profit organization dedicated to awareness, support, education, and research related to Erdheim-Chester Disease.

To donate, please send a check to ECD Global Alliance, P.O. Box 775, DeRidder, LA 70634. Make an online donation at the ECD Global Alliance Razoo site. Thank you for your support!

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