Nuclease Immune Mediated Brain and Lupus-like conditions (NIMBL): natural history, pathophysiology, diagnostic and therapeutic modalities with application to other disorders of autoimmunity

From 2010-06-01 to 2013-11-30, closed project

Objective

Nuclease Immune Mediated Brain and Lupus-like (NIMBL) conditions, comprising Aicardi-Goutieres Syndrome, Retinal Vasculopathy with Cerebral Leukodystrophy and some cases of Systemic Lupus Erythematosus, are devastating genetic disorders resulting in greatly reduced quality of life, high mortality especially in children, and significant risks of recurrence within affected families. NIMBL conditions are rare, but under diagnosed. No effective treatments or cures exist at present. To enable optimum patient care in Europe and worldwide, a better understanding of the natural course of these disorders and their underlying pathological basis is essential. In the NIMBL project, European and U.S. clinical and basic scientists at the forefront of NIMBL-related research have united to develop a translational approach to these problems. A registry of patients will reveal the natural history of the NIMBL diseases, and the efficacy of current treatments. Investigation of patients together with the use of existing and novel cellular and animal models will precisely define their pathogenesis and identify potential drug targets. These same platforms will then be used to test existing and new drug treatments, thus providing unique opportunities to European companies. The NIMBL project will build on very recent discoveries of the cell-intrinsic initiation of autoimmunity that we have described, and which have major implications for our understanding of the discrimination of self from non-self. This new biological paradigm involves cytosolic sensors that detect accumulated, endogenous nucleic acids, and induce the body to mount an immune response against its own cells. Thus, the investigation of NIMBL diseases will not only improve the health and well-being of NIMBL patients and their families, it will also lead to better treatments of much more common autoimmune disorders including lupus.

Related information

Result In Brief
New therapeutic concepts for NIMBL disorders

Report Summaries
Final Report Summary - NIMBL (Nuclease Immune Mediated Brain and Lupus-like conditions (NIMBL): natural history, pathophysiology, diagnostic and therapeutic modalities with application to other disorders of autoimmunity)
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Subjects
Medical biotechnology - Medicine and Health

Last updated on 2017-05-25
Retrieved on 2019-07-16

Permalink: https://cordis.europa.eu/project/rcn/94538_en.html
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