

# Study hints at regeneration of nerve insulation to treat CHARGE birth defects

February 29 2016

---

Research in *Nature Neuroscience* suggests the possibility of treating a group of genetic birth defects with molecular therapy that would regenerate malformed nerve insulation in the central nervous system.

Scientists from Cincinnati Children's Hospital Medical Center report their findings in the journal's Feb. 29 online edition. The study focuses on a genetic condition called CHARGE syndrome, which leads to multiple life-altering birth defects that include craniofacial malformations, neurological dysfunction and growth delay.

The researchers discovered that a gene associated with CHARGE (CHD7) serves as a central molecular control point in the production of a substance called myelin. Myelin forms a protective sheath around the nerves to allow speedy communication between neurons.

Defects in CHD7 disrupt a large number of molecular pathways that researchers identified in the current study. Those pathways control over a dozen other genes that help form [myelin sheath](#) in nerves and make bones and other organs.

"Our findings could provide a molecular framework for identifying signaling pathways and molecules as therapeutic targets to promote myelin regeneration in patients with CHARGE and other demyelinating diseases," said senior author Richard Lu, PhD, scientific director of the Brain Tumor Center in the Division of Experimental Hematology and Cancer Biology at Cincinnati Children's.

The study involves a multi-institutional research team that includes co-senior author Carlos Parras PhD, of Sorbonne Universités (University of Pierre and Marie Curie) in Paris and Donna Martin, MD, PhD, at the University of Michigan. Researchers conducted genome-wide gene analysis and biological tests with laboratory mouse models of CHARGE syndrome (which means coloboma, heart defect, atresia choanae, retarded growth and development, and ear abnormalities including deafness).

Their analyses show that CHD7 works in unison with a regulatory protein called Sox10 to trigger the onset of myelin formation. Sox10 is important to the development of nerve cells, including oligodendrocytes for myelin formation.

In laboratory mice either having or lacking functional CHD7, the authors found CHD7 and Sox10 also work together to prompt myelin to resume formation (a process called re-myelination) after it has initially been disrupted.

Researchers continue their research by testing various molecular strategies involving CHD7, Sox10 and other genes for reforming the myelin coating on nerves in different types of neurological diseases with myelin defects.

**More information:** "Chd7 cooperates with Sox10 and regulates the onset of CNS myelination and remyelination," *Nature Neuroscience* (2016). [DOI: 10.1038/nn.4258](https://doi.org/10.1038/nn.4258)

Provided by Cincinnati Children's Hospital Medical Center

Citation: Study hints at regeneration of nerve insulation to treat CHARGE birth defects (2016,

February 29) retrieved 1 May 2024 from <https://medicalxpress.com/news/2016-02-hints-regeneration-nerve-insulation-birth.html>

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.