Generation of Zic4 Mutant and Zic1/4 Double Mutant Mice as a Model for Dandy-Walker Malformation

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Technology description

Summary

Temporary stub for UCHI 1743

Description

Dandy-Walker Malformation (DWM) is a congenital brain malformation that is defined by hypoplasia and upward rotation of the cerebellar vermis and cystic dilation of the fourth ventricle. The incidence of DWM in the United States has been estimated as 1 case per 25,000 live births. Symptoms often occur early in infancy and include slow motor development and progressive enlargement of the skull. Affected individuals also often present with motor deficits. About half of all individuals affected by DWM also have mental retardation and some have hydrocephalus. DWM is also frequently associated with disorders of other areas of the central nervous system including absence of the corpus callosum. The low emperic recurrance rate for nonsyndromic DWM suggests a polygenic basis for this disorder, although the genes involved are largely unknown. The innovators have found that the genes encoding the Zinc finger proteins Zic1 and Zic4 are involved in DWM. In order to investigate the underlying mechanism of Zic1 and Zic4 in the pathology of DWM, the innovators have generated Zic4 mutant and Zic1/4 double mutant mice. The Zic1+/- Zic4+/- mice display a phenotype that closely resembles DWM and represent a novel model system for investigating DWM as well as the role of the Zic family proteins in brain development.

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