TRIM71 (Tripartite Motif Containing 71) is a Protein Coding gene. Diseases associated with TRIM71 include Hydrocephalus, Congenital Communicating, 1, and Congenital Communicating Hydrocephalus. Among its related pathways is Class I MHC mediated antigen processing and presentation and the Innate Immune System. Gene Ontology (GO) annotations related to this gene include ligase activity and MicroRNA binding. An important paralog of this gene is TRIM3.

Integrating human brain transcriptomics with whole-exome sequencing of 483 patients with congenital hydrocephalus (CH), Duy et al. found convergence of CH risk genes in embryonic neuroepithelial stem cells. Of all CH risk genes, TRIM71/lin-41 harbors the most de novo mutations and is most specifically expressed in neuroepithelial cells. Mice harboring neuroepithelial cell-specific Trim71 deletion or CH-specific Trim71 mutation exhibit prenatal hydrocephalus. CH mutations disrupt TRIM71 binding to its RNA targets, causing premature neuroepithelial cell differentiation and reduced neurogenesis. Cortical hypoplasia leads to a hypercompliant cortex and secondary ventricular enlargement without primary defects in CSF circulation. These data highlight the importance of precisely regulated neuroepithelial cell fate for normal brain-CSF biomechanics and support a clinically relevant neuroprogenitor-based paradigm of CH [1)].

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