MPDZ (multiple PDZ domain protein) is a gene that encodes a protein involved in various cellular processes, particularly those related to cell-cell adhesion and signaling. The MPDZ protein contains multiple PDZ domains, which are important for protein-protein interactions. These domains allow MPDZ to interact with various other proteins, playing a crucial role in maintaining the integrity of tight junctions between cells, especially in the brain.

Mutations in the MPDZ gene have been linked to congenital hydrocephalus. The protein encoded by MPDZ is important for the function of the ependymal cells that line the ventricles of the brain. These cells play a key role in the movement of cerebrospinal fluid (CSF) and maintaining the blood-brain barrier. When MPDZ is dysfunctional due to mutations, it can lead to the disruption of these processes, resulting in the accumulation of CSF within the brain's ventricles, which characterizes hydrocephalus.

Specifically, MPDZ mutations are often associated with non-obstructive hydrocephalus, where the problem lies not in the physical blockage of CSF flow but in the failure of CSF to be properly absorbed or circulated due to the impaired function of the ventricular lining or associated structures. The condition underscores the importance of cellular and molecular mechanisms in maintaining the homeostasis of CSF and brain fluid dynamics.