

MOLAR TOOTH SIGN - JOUBERT SYNDROME

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The molar tooth sign is seen in very few conditions and is a very rare pediatric central nervous system congenital anomaly. Molar tooth sign is the result of cerebellar vermis hypoplasia, thick and maloriented superior cerebellar peduncles, and an abnormally deep interpeduncular fossa. In Joubert syndrome, this is seen in about 85% of patients. We present a case of a two-year old girl with flaccid paraparesis, regression of milestones and developmental delay. Magnetic resonance imaging (MRI) showed the characteristic molar tooth sign with apposition of cerebellar hemispheres, batwing-shaped fourth ventricle, cerebellar vermis agenesis and deep interpeduncular fossa consistent with the diagnosis of Joubert syndrome. *Acta Medica Medianae 2015;54(3):74-77.*

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