Pontine tegmental cap dysplasia: A hindbrain malformation caused by defective neuronal migration
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Neurology 2010;74;1835
DOI 10.1212/WNL.0b013e3181e0f7f8

This information is current as of October 2, 2012

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http://www.neurology.org/content/74/22/1835.full.html
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A 13-month-old girl presented with developmental delay, generalized hypotonia, vertical pendular nystagmus, and cranial nerves IV, V, and VII paresis. MRI (figure) revealed pontine hypoplasia with ectopic dorsal transverse pontine fibers (tegmental cap). Concomitant profound sensorineural deafness and a butterfly Th2 vertebra led to the syndromic diagnosis of pontine tegmental cap dysplasia (PTCD).

PTCD has been described to date in several patients. It is suggested that many cases of pontocerebellar hypoplasia or Moebius syndrome should be revised for the features of PTCD. Pathomechanism of the condition involves defect in migration or navigation of axons of rhombencephalic neurons.

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Disclosure: The authors report no disclosures.

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ACKNOWLEDGMENT
The authors thank Prof. Bwee Tien Poll-The for the comments.

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