Title: Polymicrogyria Overview *GeneReview* – Neuropathology of PMG
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Neuropathology

Gross neuropathologic examination reveals a pattern of complex convolutions to the cerebral cortex with multiple miniature gyri, often resulting in an irregular brain surface. The cortical ribbon can appear excessively thick as a result of the infolding and fusion of multiple small gyri.

Histology. Findings vary but always show abnormal cortical lamination with one or more festooning bands of neurons replacing the normal cortical architecture [Norman 1995]. This abnormal festooning of the cortical neuronal band can occur well before normal cortical folding is expected to start [Jansen et al 2016].

Both unlayered (also known as two-layered) and layered forms of PMG are described. Occasionally both forms are found in the same individual, suggesting that they may be variations of the same malformation [Judkins et al 2011].

PMG is commonly associated with pial disruption, regardless of etiology [Jansen et al 2016].

PMG is frequently associated with other brain malformations such as dysgyria or heterotopia.

Pathophysiology. The high prevalence of abnormalities of the brain surface overlying the polymicrogyric cortex is not surprising as this is the nexus between the radial glial endfeet, Cajal-Retzius cells, pial basement membrane, and leptomeningeal cells, all recognized to play a crucial role in cortical formation. Many recent animal studies have highlighted the frequency and importance of pial basement membrane and leptomeningeal abnormalities in a range of malformations of the cortex.

Literature Cited

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