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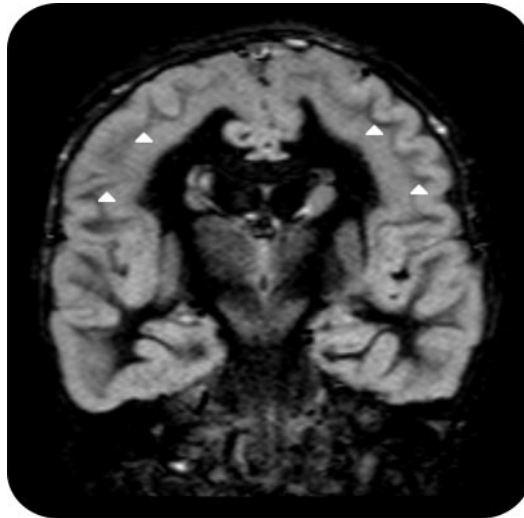
Double cortex

“Two” is too many

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Figure 1 Dual inversion recovery MRI



Coronal gray matter dual inversion recovery image shows thin white matter layer (arrowheads) separating the 2 layers of gray matter.

A 6-year-old ambidextrous girl born to unrelated parents presented with complex partial seizures and mental retardation. Her sister and mother had seizures and tested positive for the doublecortin gene mutation (DCX). EEG showed epileptiform discharges in the right occipital area and spikes over right frontal and left temporal areas. MRI showed subcortical band heterotopias, consisting of a linear band of gray matter below cortex (figure 1). She tested positive for the DCX mutation. Doublecortin is important for neuronal migration.¹ In males, all neurons are affected, leading to lissencephaly, a smooth-appearing brain due to absent or abnormally thick gyri with neuronal disorganization and rudimentary cortex. In females, affected neurons migrate

Figure 2 Axial T1 MRI



Axial T1 image shows band heterotopia (arrow) with white matter separating the band from cortex and thickened gray matter (arrowheads) which consists of cortex and underlying band heterotopia.

to the subplate region, forming band heterotopia (figure 2), while unaffected neurons migrate to form the cerebral cortex.² There can be a continuum between band heterotopia and lissencephaly depending on the degree of gene expression.

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