

Teaching Video NeuroImage: Congenital Hemidystonia–Hemi-midbrain Atrophy Syndrome

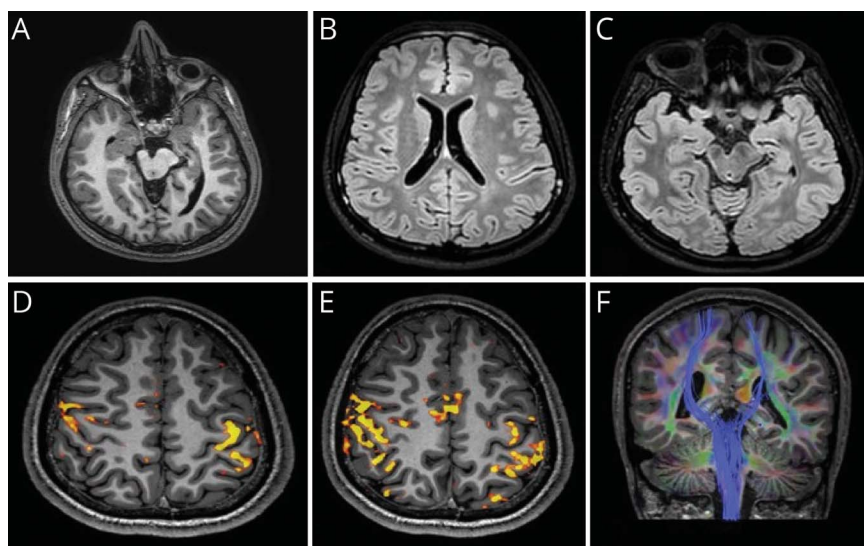
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Figure MRI of the Brain (T1 and T2 FLAIR), fMRI, and MR Tractography Findings



(A–C) Right-sided hemiatrophy of the midbrain with slight right hemispheric white matter volume loss. (D and E) Bilateral cortical activation on fMRI with right and left finger taps, respectively. (F) Magnetic resonance tractography shows normal decussation of corticospinal tracts.

An 18-year-old man with normal birth and development and a negative family history complained of nonprogressive clumsiness and posturing of his left hand since early childhood. His left limbs were shorter than his right but showed no evidence of atrophy (eFigure 1, A–C, links.lww.com/WNL/B902). Examination revealed dystonic posturing with mirror dystonia of the left hand and subtle mirror movements in his feet (Video 1). MRI of the brain showed right hemiatrophy of the midbrain, bilateral cortical activation with unilateral finger taps on fMRI, and normal corticospinal tract decussation (Figure, A–F). Levodopa response was present, suggesting dopaminergic denervation due to a congenital hemi-midbrain insult.¹ This case represents hemidystonia-hemiatrophy syndrome.² Subtle limb shortening and mirror dystonia can aid in diagnosis.

MORE ONLINE

Video

Teaching slides

links.lww.com/WNL/B902.

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