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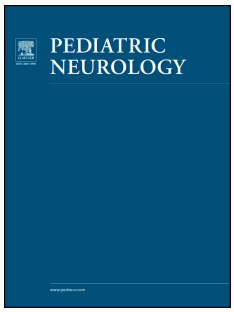

JUVENILE HUNTINGTON DISEASE Pediatric neurologist: look at the neuroimage

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JUVENILE HUNTINGTON DISEASE
Pediatric neurologist: look at the neuroimage

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A 15-year-old female presented with ataxia, rigidity and developmental impairment for 5 years, developing a tendency to opisthotonos, progressive inability to walk and behavior modification for the last 6 months. She was the only child of non-consanguineous parents and her earlier development was unremarkable. Brain MRI showed bilateral caudate and putamen atrophy suggestive of juvenile Huntington disease (Figure) and genetic analysis revealed 80 CAG repeats in the *HTT* gene. The diagnosis, initially hindered by an absence of family history, was later corroborated by the information that both the father and paternal grandfather had developed involuntary movements late in life.

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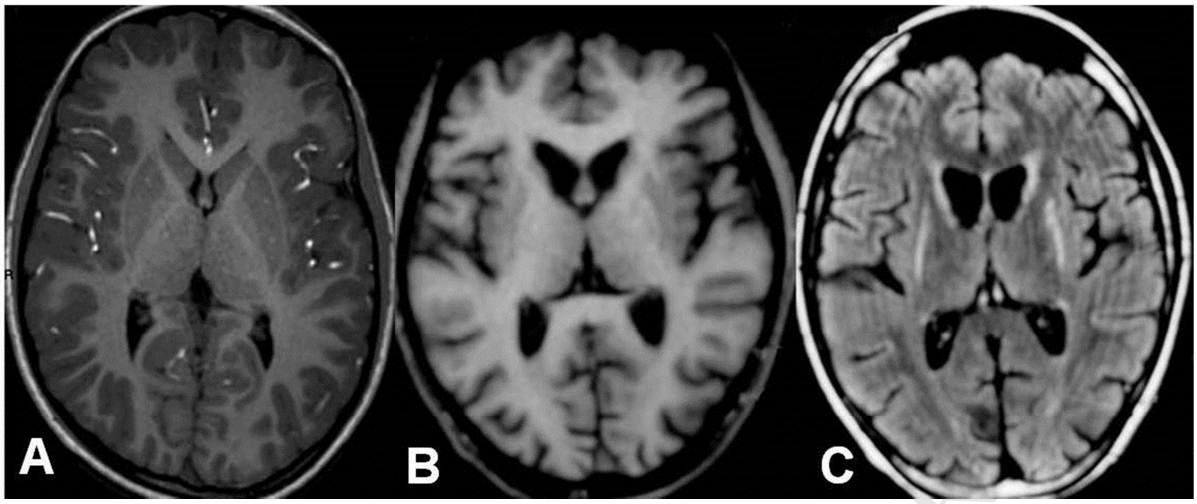

Figure: Magnetic resonance axial images. (A) Axial T1-weighted image: normal anatomy. Observe the head of the caudate nucleus shaping the frontal horn of the lateral ventricles; (B) T1-weighted images (patient): dilation of the frontal horns caused by atrophy of the caudate nuclei; (C) FLAIR (patient): putaminal atrophy and hyperintensity.