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Research Details :

Research Title	:	*Misdiagnoses in Children With Doparesponsive Dystonia* *Misdiagnoses in Children With Doparesponsive Dystonia*
Descriptipn	:	Dystonia is a state of continuous contraction of groups of agonist and antagonist muscles resulting in a sustained abnormal posture. Dopa-responsive dystonia was first described in 1976 by Segawa. Patients typically have diurnal variation of their symptoms with worsening at the end of the day and a dramatic response to low-dose L-dopa. This report presents five consecutive children with dopa-responsive dystonia who were misdiagnosed initially as spastic diplegic cerebral palsy, intractable epilepsy, hereditary spastic paraplegia, or a neurodegenerative disorder. There were two males and three females aged 3-13 years (mean 8.6 years). They were monitored for up to 2 years (mean 14.8 months). One had focal, one axial, one segmental, and two generalized dystonia. The dystonia was paroxysmal in two (tiptoe walking and opisthotonus), and all had a progressive course. All children responded dramatically to L-dopa (mean 200 mg/day), including three who were wheelchair-bound for several years. The difficulties in early diagnosis, variability of clinical presentation, and dramatic response to L-dopa will be illustrated. To conclude, dopa-responsive dystonia should be considered in any child who presents with paroxysmal or progressive hypertonia of unknown etiology, because it responds so dramatically to L-dopa. © 2004 by Elsevier Inc. All rights reserved.
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File Name	Type	Description
Dystonia.pdf	pdf	مشاهدة المقالة العلمية كاملة