Movement Disorders in Different Ages

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إضطرابات الحركة فى مختلف الأعمار

رسالة مقدمة توطئة لنيل درجة الماجستير
في الأمراض العصبية والنفسية

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Introduction

Movement disorders in childhood are both similar and distinct from those occurring in adulthood. They both overlap in their definitions of specific movements and provision of diagnostic and therapeutic challenges. In contrast, there are several prominent differences with motor abnormalities in the pediatric population having an increased occurrence of hyperkinetic movements, rather than bradykinesia or rigidity, a major primary etiology of chronic motor dysfunction being residue of static encephalopathy, a greater likelihood that symptoms are secondary to hereditary metabolic disorders ***(Singer, 2007).***

Different age groups have been classified into infancy which are from 0-12 months, early childhood are from 1-3 years, middle childhood are from 3-7 years, late childhood are from 7-12 years, adolescence are from 12-20 years . Authors also classified early adulthood from 20-40 years, middle adulthood from 40-65 years ***(Thane, 1998)***. Young old are from 65-74 years, old are from 75-84 years, oldest old to those who are aged from 85 years and over ***(Timiras et al., 2007).***

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Classification of the abnormal movement is based on observed features (e.g. chorea, tic, stereotypy, dystonia, tremor, etc.), with subdivisions determined by etiological factors (e.g. metabolic, genetic, infectious, neoplastic, etc.) ***(Delgado and Albright, 2003).***

Only 13% of cases had onset before age of 10 years. Females were predominantly affected (Femal:Male ratio is 4:1). The most common types of movement disorders seen were dystonia (47%), tremor (40%), and gait disorders (13%) ***(Schwingenschuh et al, 2008).***

Movement disorders in children are more often secondary than primary, that is they are more often associated with lesions of the brain such as cerebral palsy, traumatic brain injury, or stroke than occurring as a primary disorder. The most common etiology of movement disorders in children is probably cerebral palsy, which affects 2.5 in 1000 live births in the United States ***(Delgado and Albright, 2003).***

Chorea is one of the major types of involuntary movement disorders originating from dysfunctional neuronal networks interconnecting the basal ganglia and frontal cortical motor areas. The syndrome is characterized by a continuous flow of random, brief, involuntary muscle contractions and can result from a wide variety of causes ***(Cardoso et al, 2006).***

Athetosis consists of nonpatterned, writhing movements that represent a form of "slow chorea" ***(Jankovic et al., 2009).*** Movement disorders experts use the term athetosis less and less, however it is still used to describe the combination of dystonia and chorea in the distal portions of limbs in patients with cerebral palsy. The phenomenology is characterized by a variable combination of dystonia, chorea, myoclonus, and spasticity ***(Cardoso, 2007).***

Ballismus is a form of severe coarse chorea that it is usually unilateral (hemiballismus) and often results from a lesion in the contralateral subthalamic nucleus and adjacent structures ***(Jankovic et al., 2009).***

Sydenham's chorea, the neurological manifestation of rheumatic fever, is the most common acquired chorea of childhood. This disorder is secondary to an autoimmune response against basal ganglia in the brain, induced by prior streptococcal infection. Chorea is a major criterion for the diagnosis of rheumatic fever and can also be the presenting feature of the disease ***(Palumbo, 2008).***

Huntington disease (HD) typically occurs during the fourth and fifth decades of life, however, onset occurs during childhood or adolescence in approximately 5% to 7% of affected patients. Patients with juvenile-onset HD develop

dystonia, ataxia, and seizures. Most of them have the akinetic-rigid syndrome termed the Westphal variant. Approximately one-fourth have the classic features of chorea seen in adults. Children also have more rapidly progressive disease than adults ***(Jankovic et al., 2009).***

Parkinson's disease (PD) is a progressive neurodegenerative disorder associated with a loss of dopaminergic nigrostriatal neurons. Parkinson's disease is recognized as one of the most common neurological disorders, affecting approximately 1% of individuals older than 60 years. Cardinal features include resting tremor, rigidity, bradykinesia, and postural instability ***(Hauser et al., 2009).***

Parkinsonism may be difficult to recognize particularly in young individuals, and may be overlooked for several months or years. Young-adults Parkinsonism which is defined as symptom onset before age of 40 years is comparatively rare, however an older patient reporting symptom onset in middle age is less rare as many as 12% of patients in some tertiary referral clinic populations date their symptom onset before age of 40 years. Idiopathic Parkinsonism that begins before age of 21 years is extremely rare ***(Jankovic, 2005).***

Patients with young-onset Parkinsonism have symptoms that are similar to those of older patients but have a higher incidence of dystonia, particularly in the lower extremities. Because dystonia as an isolated symptom of other diseases is unusual. Early Parkinsonism should be suspected in middle-aged individuals with an isolated dystonia in the upper or lower extremity ***(Jankovic, 2005).***

Dystonia (from Greek, means altered muscle tone) refers to a syndrome of involuntary sustained or spasmodic muscle contractions involving co-contraction of both the agonist and the antagonist. The movements are usually slow and sustained. They often occur in a repetitive and patterned manner, but they can be unpredictable and fluctuate. The frequent abnormal posturing and twisting can be painful and functionally disabling ***(Albanese et al., 2006).***

Infantile dystonia begins before age 2 years. Childhood dystonia begins at age 2-12 years. Juvenile dystonia begins at age 13-20 years. Adult dystonia begins after age 20 years ***(Albanese et al., 2006).***

Age at onset of primary dystonia is bimodally distributed, with modes at 9 years (early onset) and 45 years (late onset). Age at onset is closely related to anatomic distribution. Early-onset dystonia usually first affects legs or

arms and less commonly starts in neck, vocal cords, or other cranial muscles. Late onset primary dystonia commonly affects neck or cranial muscles and is less likely to begin in a limb ***(Geyer and Bressman, 2006).***

Tics, patterned movements distinct from stereotypies, myoclonus and other hyperkinetic movements are quite common in children particularly among those with developmental and psychiatric disorders. Thus, tics can indicate the presence of atypical neurodevelopment or broader difficulties with cognition or mood. Tics are also the cardinal feature of Tourette syndrome. A childhood onset neurobehavioral disorder is characterized by a chronic inability to suppress or an urge to perform patterned repetitive movements. Patients with Tourette syndrome most commonly have in addition to tics, symptoms of inattention, hyperactivity, obsessiveness, or anxiety ***(Gilbert, 2006).***

Myoclonus is defined as sudden, brief, jerky, shocklike, involuntary movements arising from the central nervous system and involving extremities, face, and trunk. Most myoclonic jerks are caused by abrupt muscle contraction (positive myoclonus), but abrupt movements are also caused by sudden cessation of muscle contraction associated with the silent period

of the electromyographic (EMG) discharges (negative myoclonus) ***(Shibasaki, 2007).***

Onset is usually in the first month of life with myoclonus persisting for several months rarely into early childhood ***(Singer, 2007).***

Essential tremor affects up to 5% of the general population after the age of 60 years. Essential tremor is often inherited in an autosomal dominant. The age of onset may be as early as the first or second decade of life, but the tremor may be delayed until the mid-60s. Patients first become aware of a mild postural and action tremor in hands, which is indistinguishable from an enhanced physiologic tremor and may result in little functional impairment for many years until it gradually interferes with activities. Older patients with large-amplitude, lower-frequency tremors can have a resting component that is often misdiagnosed as Parkinson's disease ***(Long, 2007).***

The term drug-induced movement disorder (DIMD) refers to a variety of phenomenologically distinct, treatment-emergent, involuntary motor symptoms including akathisia, dyskinesia, dystonia, and parkinsonism. DIMDs, also pervasively referred to as extrapyramidal adverse reactions

remain a significant iatrogenic burden among selected patient populations ***(Claxton et al., 2007).***

Aim of the work

The aim of this work is to highlight the impact of age on clinical presentation in movement disorders to refine our diagnosis and to achieve better management.

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مقدمـــة

إن إضطرابات الحركة فى مرحلة الطفولة مماثلة للإضطرابات التى تحدث فى مرحلة البلوغ ولكن يتداخل الأمرين فى التشخيص والعلاج. وفى المقابل هناك العديد من الإختلافات البارزة مثل زيادة عدد الحركات غير الطبيعية عند الأطفال بدلا من التيبس وبطء الحركة والسبب الرئيسى فى خلل الحركات المزمن هو بقايا إعتلال المخ وهناك إحتمال كبير أن تكون هذه الأعراض ثانويه لإضطرابات الأيض الوراثية.

يتم تصنيف الأعمار إلى مرحلة الرضاعة والتى تبدأ منذ الولادة حتى السنة الأولى، ثم مرحلة الطفولة المبكرة والتى تكون من سن 1–3 سنوات ويليها مرحلة الطفولة المتوسطة والتى تكون من 3-7 سنوات، ثم مرحلة المراهقة والتى تكون من 7-20 سنة. بينما تكون مرحلة النضوج من 20–65 سنة، ثم مرحلة الشيخوخة المبكرة والتى تكون من 65–74 سنة، وهناك أيضاً مرحلة الشيخوخة والتى تكون من 75–84 سنة، ويستخدم البعض مصطلح الهِرم لهؤلاء الذين تذيد أعمارهم عن 85 سنة.

تصنيف الحركات الغير طبيعية يتم بناء على ملاحظات مميزة مثل الحركات المتلازمة والحركات النمطية والأرتعاشات وخلل توتر العضلات والتى تحددها العوامل المسببة للمرض كالعوامل الوراثية والأمراض المعدية وخلل التمثيل الغذائى والأورام.

تظهر أعراض المرض قبل سن العاشرة فى 13% فقط من الحالات، والإناث هم الأكثر تضرراً من هذه الإضطرابات حيث تبلغ نسبة إصابة الإناث إلى الذكور أربعة إلى واحد على التوالى والأنواع الأكثر شيوعا فى إضطرابات الحركة هى خلل