Parkinsonism and Related Disorders 20S1 (2014) S13–S16

Contents lists available at SciVerse ScienceDirect

Parkinsonism and Related Disorders

journal homepage: www.elsevier.com/locate/parkreldis

Movement disorders in childhood

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ARTICLE INFO

Keywords:
Children
Movement Disorders
Tic
Dystonia
Tremor
Chorea
Stereotypy
Parkinsonism

SUMMARY

The aim of this article is to review movement disorders in children. They are common but have etiology and phenomenology different than in adults. Tics are the most common phenomena although in most instances they are mild and have a favorable long-term prognosis. Dystonia is the second most common phenomena but when present it is usually genetic or idiopathic and causes meaningful disability. Sydenham's chorea is the most common cause of chorea in children worldwide. Systemic lupus erythematosus is a much rarer cause of chorea but it is always to be ruled out given the lack of a specific diagnostic marker for Sydenham's chorea. Tremor, usually caused by drugs or essential tremor, is regarded as rather uncommon in children. Arguably, most pediatric patients with tremor do not seek medical attention because of the lack of disability. Stereotypies are relatively uncommon but their recognition is clinically relevant since they are usually associated with severe conditions such as autism and Rett syndrome. Parkinsonism is quite rare in children and either results from encephalitis or is a side effect of medications. Wilson's disease must be ruled out in all children with movement disorders.

1. Introduction

The aim of this article is to provide an overview of the phenomenology of the main causes of movement disorders in children, tackling them by order of frequency. The focus of the review is on conditions where movement disorders are isolated or the predominant phenomena. For this reason, Wilson's disease (WD), an autosomal recessive illness characterized by copper accumulation in the liver, cornea, brain and other organs, will not be described in further detail. Nevertheless, one must always bear in mind that WD needs to be ruled out in every single child with a movement disorder since if left untreated it inexorably leads to death [1].

In general, definitions will use a published consensus resulting from a meeting of specialists [2]. *Tics* are repeated, individually recognizable, intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement. It should be added that in addition to motor tics there are also phonic ones that have the same features although of course they are sounds rather than movements [2]. *Dystonia* is defined according to the International Parkinson and Movement Disorders Society criteria, i.e., a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both [3]. Although there are authors who characterize athetosis as a separate phenomenon, the present author's opinion is that it is merely a form of distal dystonia, often associated with cerebral palsy. The consensus meeting defined *tremor* as a rhythmic back-and-forth or oscillating involuntary movement about a joint axis [2]. The same group considers *chorea* as an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments. *Stereotypy*, a controversial topic, has recently been defined as a non-goal-directed movement pattern that is repeated continuously for a period of time in the same form and on multiple occasions, and which is typically distractible [4]. *Parkinsonism* is defined as a combination of bradykinesia and at least one more of the additional cardinal signs: rigidity, tremor and postural instability [5]. Children often present with *myoclonus* but this phenomenon is beyond the scope of this article since in the majority of childhood cases it has an epileptic origin.

2. Tics

Tics are the most common movement disorder in children. Several studies in different parts of the world report that near one fifth of students in elementary school have tics [6]. There is a clear association of tics with placement in special education: for instance, a study in Rochester, NY, found that 18.5% of students in regular education have tics whereas this figure increased to 23.4% among students of special education [7].

Tourette's syndrome (TS) and its variants, such as chronic motor or vocal tics as well as transient tic of childhood, account for almost all cases of tics in children. This group of conditions usually have onset around age 8 years and are more common in boys. TS is defined

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1353-8020/$ – see front matter

by the presence of multiple motor tics, at least one phonic tic, duration of at least one year and onset before age 21 years. The etiology remains to be determined although these conditions are thought to result from a combination of genetic predisposition and environmental factors. A feature rather unique to tics is the ability of patients to suppress them, although this is usually associated with the development of discomfort in the body area involved in the production of the phenomenon. The majority of patients with tics present with simple movements concentrated in the rostral part of the body. Examples are excessive blinking, grimacing, and head jerking. Phonic tics are also simple vocalizations, such as sniffing and throat clearing, in most patients. However, more severe cases, comprising a minority of subjects, may have tics with a more widespread topographic distribution, complex pattern (e.g., sequence of movements during gait) of contractions, not rarely causing pain or even compressive myelopathy [8]. Vocalizations may also be complex with emission of sentences as well as repetition of other individuals' statements (echolalia). A special case of complex phonic tics is coprolalia, emission of obscenities. It is not mandatory for the diagnosis of TS where it occurs in less than 25% of patients [9]. There is a strong association between TS and behavioral problems, particularly obsessive compulsive disorder and attention deficit and hyperactivity disorder. The long-term natural history of TS and related conditions is favorable, with spontaneous improvement or remission in up to 70% of individuals [10].

Rarely, tics may be secondary to an identifiable cause, such as structural brain lesions (encephalitis, trauma, stroke etc), toxic lesions, degenerative diseases and chromosomal disorders. In this case the term tourettism is used to describe the condition [11].

3. Dystonia

Primary dystonias, defined as those without a structural lesion of the nervous system, are usually idiopathic or have a genetic cause [3]. They account for at least two thirds of the cases of dystonia among children. Although a large number of genetic forms of dystonia has already been identified [12], in this article focus will be given to Oppenheim's dystonia (DYT1) and dopa-responsive dystonia (DYT5).

DYT1, or Oppenheim's dystonia, an autosomal dominant condition with variable penetrance and expression, is the most common cause of generalized dystonia in patients under the age of 26 years. Typically, the onset is in the first decade of life with unilateral dystonia of the foot induced by gait. This is followed by gradual spreading of the movement disorder with involvement of the contralateral lower limb, trunk and even upper limbs, neck and face. Severe forms leading patients to become wheelchair bound are rare but may occur [3,12]. With the availability of the molecular diagnosis of DYT1 after the discovery that the gene is located on chromosome 9q [13], there is a better understanding of the phenomenology of this condition. Indeed, it is more common among Ashkenazi Jews and there is no obvious neurological abnormality other than dystonia (i.e., it is a primary dystonia). However, it is now known that in many subjects it has a mild presentation with focal and segmental forms of dystonia.

Dopa-responsive dystonia (DYT5) is also known as Segawa's disease after the Japanese physician who originally described it. Similarly to DYT1, it is an autosomal dominant condition with greater expression in females, with patients developing action dystonia of the foot during the first decade of life. Its prevalence is about 1/10 of DYT1 and many patients notice fluctuation of the symptoms, with increasing severity throughout the day and after exercising. Some subjects may have parkinsonian findings such as rigidity, bradykinesia and mild tremor. The presence of this additional phenomenology justifies the labeling of DYT5 as belonging to the group of dystonia-plus conditions, although this term has often been criticized [3]. The unique distinguishing feature is the remarkable response to low-dose L-dopa that leads to complete disappearance of dystonia. Because it is a metabolic disease with impairment of the metabolism of dopamine and not a degenerative disease like Parkinson's disease (PD), patients with DYT5 do not develop the long-term complications of the use of L-dopa seen in the latter [12,13]. Because of this response and the common difficulty in differentiating DYT5 from other forms of childhood-onset dystonia, it is mandatory to treat all these patients with L-dopa for at least 60 days.

The most common form of secondary dystonia in children is cerebral palsy (CP). Unlike the two dystonias described in the preceding paragraphs, patients with CP display a myriad of movement disorders and other neurological findings: dystonia, chorea, tremor, myclonus, ataxia and spasticity. Unlike primary dystonias, the severity of dystonia in CP is greater in the upper half of the body and patients commonly have severe dysarthria. As already mentioned, many authors still use the term athetosis to describe the distal dystonic movements displayed by DP patients [14].

4. Chorea

The most common cause of acute chorea in children worldwide is Sydenham's chorea (SC) [15,16]. It is a major manifestation of rheumatic fever, presumably caused by antibodies induced by β-hemolytic streptocci that cross-react with brain antigens [17]. The average age at onset is 9 years and it is more common in girls. In addition to chorea, that can be limited to one half of the body in 20% of patients, there are other motor and non-motor features: decreased muscle tone, vocalizations, emotional lability, obsessions and compulsions, attention deficit and hyperactivity, carditis, and others. It is worth mentioning that a cardiac lesion is the most important source of disability in patients with SC [15]. In at least one quarter of patients with SC, the disease has a protracted course, lasting two or more years, called persistent SC [18]. It is also well recognized that patients with a previous history of SC may develop recurrence later in life when becoming pregnant (*chorea gravidarum*) or if treated with oral contraceptive agents [19]. As there are no specific biological markers for SC, the diagnosis relies on the presence of typical clinical findings and ruling out alternative diseases. The most common source of diagnostic confusion is chorea associated to systemic lupus erythematosus (SLE), where up to 2% of patients may develop chorea. From a clinical point of view, the majority of subjects with this condition will have other non-neurologic manifestations such as arthritis, pericarditis, and other serositis as well as skin abnormalities. Moreover, the neurologic picture of SLE tends to be more complex and may include psychosis, seizures, other movement disorders, and even mental status and consciousness level changes. Only in rare instances will patients with SLE have chronic isolated chorea with a tendency for spontaneous remissions and recurrences. The difficulty in distinguishing lupus from SC is increased by the finding that up to 20% of patients with the latter display recurrence of the movement disorder. Eventually, patients with SLE will develop other features, meeting diagnostic criteria for this condition [20]. Primary antiphospholipid antibody syndrome is differentiated from SC by the absence of other clinical and laboratory features of rheumatic fever as well as the usual association with repeated abortions, venous thrombosis, other vascular events, and the presence of typical laboratory abnormalities [15]. It is important to emphasize that Huntington's disease (HD) is not a differential diagnosis of SC since the childhood or juvenile form of HD (the Westphall variant) is characterized by a rigid-akinetic syndrome and not chorea.