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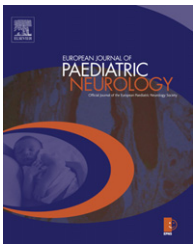

Invited papers presented at the EPNS 2011 Cavtat meeting

Movement disorder emergencies in childhood

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ABSTRACT

The literature on paediatric acute-onset movement disorders is scattered. In a prospective cohort of 52 children (21 male; age range 2mo-15y), the commonest were chorea, dystonia, tremor, myoclonus, and Parkinsonism in descending order of frequency. In this series of mainly previously well children with cryptogenic acute movement disorders, three groups were recognised: (1) Psychogenic disorders ($n = 12$), typically >10 years of age, more likely to be female and to have tremor and myoclonus (2) Inflammatory or autoimmune disorders ($n = 22$), including N-methyl-D-aspartate receptor encephalitis, opsoclonus-myoclonus, Sydenham chorea, systemic lupus erythematosus, acute necrotizing encephalopathy (which may be autosomal dominant), and other encephalitides and (3) Non-inflammatory disorders ($n = 18$), including drug-induced movement disorder, post-pump chorea, metabolic, e.g. glutaric aciduria, and vascular disease, e.g. moyamoya. Other important non-inflammatory movement disorders, typically seen in symptomatic children with underlying aetiologies such as trauma, severe cerebral palsy, epileptic encephalopathy, Down syndrome and Rett syndrome, include dystonic posturing secondary to gastro-oesophageal reflux (Sandifer syndrome) and Paroxysmal Autonomic Instability with Dystonia (PAID) or autonomic 'storming'. Status dystonicus may present in children with known extrapyramidal disorders, such as cerebral palsy or during changes in management e.g. introduction or withdrawal of neuroleptic drugs or failure of intrathecal baclofen infusion; the main risk in terms of mortality is renal failure from rhabdomyolysis. Although the evidence base is weak, as many of the inflammatory/autoimmune conditions are treatable with steroids, immunoglobulin, plasmapheresis, or cyclophosphamide, it is important to make an early diagnosis where possible. Outcome in survivors is variable. Using illustrative case histories, this review draws attention to the practical difficulties in diagnosis and management of this important group of patients.

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1. Introduction

The literature on recognition and management of acute-onset movement disorders in adult practice is scattered[1] and is very limited for paediatric practice, except for reviews on drug-induced neuroleptic malignant syndrome which typically has clinical features of paroxysmal autonomic instability with dystonia (PAID), and may evolve into life-threatening status dystonicus.[2,3] Some paediatric cases within the spectrum of status dystonicus/PAID but unrelated to drug treatment have been published under the term 'neuroleptic malignant syndrome'. Chorea, dystonia, tremor, myoclonus, and Parkinsonism can all present acutely, and since some may be treatable, timely recognition and diagnosis is important to prevent morbidity.

Disorders with a psychological component are relatively rare in paediatric movement disorder clinics, but are common acute presentations in practice.[4] Patients are more likely to be female, particularly if >13 years of age, and to have tremor, dystonia or myoclonus, often after a triggering traumatic or infectious illness.[4,5] Rehabilitation by experienced physiotherapists and occupational therapists is usually successful provided that the adolescent and family engage with psychological input in addition and the search for organic pathology ceases. It is important to remember this from a practical point of view despite the increasing evidence for overlap with organic disorders.[6] It is not harmful (and may even be helpful) to consider low risk treatment strategies such as antibiotics,[7] provided that the family continues to engage with the multidisciplinary team. Rehabilitation and psychological support are also important for children where there is convincing evidence for an organic pathology.

In a prospective[4] cohort of 52 children (21 male; age range 2mo-15y), the commonest movement disorders were chorea, dystonia, tremor, myoclonus, and parkinsonism (Table 1) in descending order of frequency. In this series of mainly previously well children with cryptogenic acute movement disorders, three groups were recognised:

(1) Psychogenic disorders ($n = 12$)
(2) Inflammatory or autoimmune disorders ($n = 22$), including N-methyl-D-aspartate receptor encephalitis (Fig. 1A), Sydenham chorea and other post-streptococcal movement disorders (Figs. 1B, 1C1–3) opsoclonus-myoclonus (which may be paraneoplastic in relation to Neuroblastoma, Fig. 1D), systemic lupus erythematosus, acute necrotizing encephalopathy (which may be autosomal dominant), and other encephalitides (Table 2) and
(3) Non-inflammatory disorders ($n = 18$), including drug-induced movement disorder, post-pump chorea and Tourette syndrome (Fig. 1E), metabolic, e.g. Leigh's disease (Fig. 1F) glutaric aciduria,[8] and vascular disease, e.g. moyamoya (Fig. 1G).[9,10] Other important non-inflammatory movement disorders, typically seen in symptomatic children with underlying aetiologies such as trauma, severe cerebral palsy[11], epileptic encephalopathy, post-hemispherectomy, after shunt dysfunction in hydrocephalus,[12] Down syndrome and Rett syndrome