MYOCLONUS, STEREOTYPIES AND TICS

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Myoclonus

In this portion of the course I will discuss the major clinical features of **myoclonus** (with accompanying video examples) and discuss some more common and less common etiologies for this movement disorder. I will discuss a basic diagnostic approach and potential therapies for myoclonus as well.

Background:

Myoclonus is common in clinical practice and some forms of myoclonus are a normal physiological phenomenon we all experience (hiccups and hypnic jerks). Myoclonus is characterized by sudden, brief, shock-like involuntary movements caused by contraction or inhibition of a muscle or a group of muscles. Typically, myoclonus is classified into two major forms: *positive myoclonus* with a muscle jerk or twitch or brief lapses of contraction known as *negative myoclonus*.

The etiology of myoclonus varies widely and may be due to demyelinating disease, Parkinsonian syndromes, epilepsy, serotonin toxicity, Huntington's disease, encephalitis, narcotics, genetic mutations, renal failure, liver disease, and many other causes. In some cases, myoclonus is due to relatively benign, reversible causes, and in others myoclonus can be associated with terminal neurodegenerative disease (e.g., Creutzfeldt-Jakob disease).

Myoclonus may originate from the cortex, subcortical structures, spinal cord or even nerve roots or peripheral nerves and further characterization and localization relies on electromyography (EMG) and electroencephalography (EEG).

Treatment of myoclonus is perhaps best discussed in the context of where the myoclonus is originating from. Antiepileptics such as levetiracetam, valproic acid, and zonisamide have potential utility. Clonazepam and sodium oxybate can also be useful.

Suggested Reading

Caviness JN. Treatment of myoclonus. Neurotherapeutics. 2014;11:188-200.

Kojovic M, Cordvari C, Bhatia K. Myoclonic disorders: a practical approach for diagnosis and treatment. Ther Adv Neurol Disord 2011;4:47-62

Lozsadi D. Myoclonus: a pragmatic approach. Practical Neurology 2012;12:215-224.

Zutt R et al. A novel diagnostic approach to patients with myoclonus. Nat Rev Neurol 2015;11:687-697.

Stereotypies

In this portion of the talk I will discuss a very commonly seen movement that can occur in both normal individuals and in association with disease states; stereotypies. I will present video examples of stereotypies and discuss the common clinical settings where these movements occur. I will briefly discuss potential therapies for these movements when they become intrusive.

Background:

Stereotypies are repetitive, predictable, purposeful but purposeless movements that occur in a child with normal developmental background (*primary stereotypies*) or in the setting of autism, mental retardation, or schizophrenia or other conditions (*secondary stereotypies*).

These movements can be motor or vocal in nature and can inhibit socialization and learning in some individuals. Motor stereotypies may consist of hand flapping, head nodding, waving of the arms and rocking back and forth or more complex movements. Patients may also have vocal stereotypies in the setting of autism and other diseases. More ominous causes of stereotyped movements include frontotemporal lobar degeneration (FTLD) and encephalitis (anti-NMDA receptor encephalitis and others)

Treatment of stereotypies when they become bothersome or intrusive is largely behavioral. So far, pharmacotherapy has been less than satisfactory.

Suggested Reading:

Baizabal_Carvallo JF et al. The spectrum of movement disorders in children with anti-NMDA receptor encephalitis. Mov Disord 2013;28:543-547.

Lydon S, et al. A systematic review and evaluation of inhibitory stimulus control procedures as a treatment for stereotyped behavior among individuals with autism. Dev Neurorehabil. 2016;23:1-11.

Rapp JT, Vollmer TR. Stereotypy I; a review of behavioral assessment and treatment. Res Dev Disabil 2005;26:527-547.

Singer HS. Stereotypic movement disorders. Handb Clin Neurol. 2011;100:631-639.

Tics

Tics are very common and underdiagnosed in clinical practice. In this portion of the course I will discuss the clinical features, potential etiology, and treatment of tic disorders including Tourette syndrome (TS).

Background:

Tics are very commonly seen in practice in both adults and children. Tics are spontaneous, repetitive movements that occur in the setting of an urge to do these movements. Tics are classically delineated into either vocal or motor phenomenologies and can be quite simple (eye blinking) or complex (coprolalia). Tourette syndrome consists of motor and vocal tics occurring at least a year starting before the age of 21 years, but typically beginning in early childhood.

While Tourette syndrome is often autosomal dominantly inherited, tics can occur in the setting of many other diseases (secondary causes) as well: Huntington's disease, neuroleptic exposure and antiepileptic therapy, etc. Despite a large inherited component for many with Tourette syndrome, identification of the genes responsible for these kindreds has been slow in coming.

There is significant psychiatric comorbidity in those with Tourette syndrome and tic disorders in most cases. Obsessive compulsive behavior, ADHD, depression, self-injurious behavior, anxiety and socialization issues are common and perhaps respond best to an interdisciplinary approach.

Therapy for tics typically focuses on α2-adrenoreceptor agonists, dopamine blocking agents and dopamine depleting agents. ADHD responds to stimulants without worsening of tics in many instances. SSRIs may be useful for obsessive-compulsive features and depression and anxiety. Deep brain stimulation has emerged as a promising therapy for the most difficult to treat cases.

Suggested Reading:

Ganos C, Martino D. Tics and Tourette syndrome. Neurol Clin 2015;33:115-136.
Ganos C et al. The functional anatomy of Gilles de la Tourette syndrome. Neurosci Biobehav Rev 37:1050-1062.
Jankovic J. Therapeutic developments for tics and myoclonus. Mov Disord. 2015;30:1566-73.
Robertson MM. A personal 35-year perspective on Gilles de la Tourette syndrome: prevalence, phenomenology, comorbidities, and coexistent psychopathologies. Lancet Psychiatry 2015;2:68-87.