striatal levels of dopamine generated. Patients with fluctuating and dyskinetic motor complications show a short duration and high amplitude peak of striatal dopamine following medication. Abnormal craving for levodopa relates to excessive production of ventral striatal dopamine.
In asymptomatic Huntington's disease gene carriers PET can detect striatal microglial activation and loss of dopamine D2 receptors suggesting that inflammation may drive neuronal loss in this disorder. Dystonia patients who carry DYT1 or DYT6 genes show a characteristic pattern of altered glucose metabolism and striatal D2 receptor downregulation even when premanifest suggesting these are trait markers for the disorder.
PET has been used to as an imaging biomarker to study the efficacy of putative neuroprotective agents in PD and HD. Implants of fetal midbrain cells, infusions of growth factors such as GDNF, and innoculation of engineered viruses into striatum can all reliably increase dopamine storage in PD, however, clinical responses have been variable and PET cannot currently be used as a surrogate marker.

Video Session December 15
Primary and secondary dystonias
15:00–16:30 Hall VI

O.072
Dystonia

S. Schneider. *Sobell Department, Institute of Movement Neuroscience, Institute of Neurology, UCL, London, UK*

Dystonia is a hyperkinetic movement disorder defined by involuntary sustained muscle spasms resulting in twisting movements and abnormal posturing of one or more body parts. Etiologically, dystonic syndromes can be broadly divided into primary and secondary forms, dystonia-plus syndromes and heredodegenerative forms. Diagnosis, particular when the clinical picture is complex, can be challenging for the clinician. Here, clinical clues and syndromic associations may be helpful in the approach to a patient, some of which we will be pointed out in this session.
The session will introduce the concept of dystonia and the classification schemes. Primary dystonias will be discussed, including DYT1 and DYT6 dystonia, both inherited in an autosomal dominant fashion. The phenomenon of clinical heterogeneity as well as the recent genetic insights will be discussed. In the second part, recessively inherited dystonia syndromes will be focussed on, some of which may present with a rather complex phenotype, including those associated with mutations in *PANK2*, *PLA2G6* or *ATP13A2*. The term of syndromes of neurodegeneration with brain iron accumulation (NBIA) will be introduced.

December 16, 2009

Breakfast Session December 16
Current understanding of myoclonus
7:15–8:00 Hall I

O.073
Current understanding of myoclonus

J. Caviness. *Mayo Clinic, Scottsdale, AZ, USA*

Myoclonus is defined as sudden, brief, shock-like, involuntary movements caused by muscular contractions or inhibitions. Numerous conditions and disease may give rise to myoclonus. As a result, classification of myoclonus presentations is necessary in order to determine a diagnosis and treatment. Etiological classification organizes the myoclonus disorders and provides major categories of clinical presentation. This classification consists of:
1. Physiologic (normal),
2. Essential (mostly monosymptomatic, stable),
3. Epileptic (part of a seizure disorder), and
4. Symptomatic (secondary to defined neurologic or medical illness).

However, the physiological classification scheme categorizes myoclonus according to its source. The electrophysiology characteristics under each category provide insight about pathophysiology. The five main physiological classification categories are:
1. Cortical,
2. Cortical-Subcortical,
3. Subcortical-Suprasegmental,
4. Segmental, and
5. Peripheral.

Each myoclonus "source" is believed to have particular neuronal circuitry characteristics. The best strategy for symptomatic treatment is derived from defining the pathophysiology via physiological classification pertaining to the source.

O.074
Current understanding of myoclonus

M. Hallett. *Human Motor Control Section, NINDS, NIH, Bethesda, MD, USA*

Myoclonus is a simple, quick, involuntary movement. There are many different types of myoclonus, and often it is difficult to tell them apart. The reason for this is that the process of excitation–contraction coupling is relatively slow and EMG bursts from 30 ms to 300 ms might look visually similar even though the pathophysiology differs. Hence, the clinical neurophysiological assessment is extremely valuable, perhaps the most valuable for any type of involuntary movement. The first question to answer is whether the myoclonus is a fragment of epilepsy or not. Epileptic fragments have brief EMG bursts, typically 30 to 50 ms, synchronous in antagonist muscles and associated with an EEG discharge. In the most common type of epileptic myoclonus, cortical myoclonus, the EEG discharge can be identified by backaveraging and is a positive–negative potential about 20 ms prior to the myoclonus. The P1–N2 component of the SEP is also "giant" in size, and like the backaveraged potential from spontaneous jerks reflects a hyperactive, synchronous discharge from the sensorimotor cortex. Another feature often seen in epileptic myoclonus is a C-reflex, a myoclonic burst at short latency produced by a sensory stimulus. Myoclonus that is not an epileptic fragment is characterized by longer bursts of EMG, 50 to 300 ms, synchronous or asynchronous in antagonist muscles and, in general, no EEG correlate. One important entity to identify is psychogenic myoclonus. The electrophysiological correlates are long EMG bursts, long and variable latencies to C-reflexes and an EEG backaverage that looks like a normal Bereitschaftspotential.

Video Session December 16
Atypical parkinsonism
7:15–8:00 Hall III

O.075
Atypical parkinsonism

A.E. Lang[1], W.J. Weiner[2]. [1]*Movement Disorder Clinic, Toronto Western Hospital, Toronto, ON, Canada;* [2]*Department of Neurology, University of Maryland School of Medicine, Baltimore, MD, USA*

We will present numerous examples of "atypical parkinsonism" to the participants and will try to answer the question of what is atypical? "Classic" examples of Multiple System Atrophy (MSA), Progressive Supranuclear Palsy (PSP) and Corticobasal Syndrome (CBS) will be demonstrated. We will also demonstrate a wide variety of "atypical" features which do not easily fit into our diagnostic categories. Lively discussion will be encouraged.