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#### DEPARTMENT OF NEUROLOGY

# CLASS 7: Extrapyramidal syndromes: Parkinson's syndrome and other extrapyramidal syndromes.

Extrapyramidal motor disorders are associated mainly with damage of the basal ganglia. These disorders cause:

- A change of the muscle tone (rigid hypertonia or hypotonia);
- Retardation of motor activity (bradykinesias);
- Postural disorders;
- Disorders of involuntary movements (hyperkinesias);

**Hypokinetic disorders** are due to increased activity of the indirect pathways and reduced activity of the direct extrapyramidal pathways, which in turn is caused by reduced dopamine in the striatum. This leads to increased inhibitory activity of basal ganglia outputs to the thalamus, which leads to reduced activation of cortical motor neurons by excitatory thalamocortical afferentations.

- 1. Parkinson's syndrome clinical symptoms:
- Akinesia and bradykinesia;
- Rigidity muscle tone;
- "Cogwheel phenomenon";
- Tremor at rest; Parkinson's tremor is rhythmic (with frequency 4-6Hz), bilaterally asymmetric, and distal;
- Postural instability;
- Hypomimic face;
- The speech becomes hypophonic and monotonous;
- Micrographia;
- The gait is retarded, in little steps, and without physiological synkineses of the arms during walking; They have difficulties to start walking;
- Pulsion phenomena can be observed;
- Typical posture with flexed head and torso. Arms attached to the body and bent in the elbow joints.

**Hyperkinetic disorders** are involuntary movements which can involve various parts of the body. They result from damage to various structures and connections in the extrapyramidal system. In hyperkinetic disorders, the balance is skewed to the direct extrapyramidal pathway. Consequently, there is reduced inhibitory activity of basal ganglia outputs. The main hyperkinetic disorders such as chorea, dystonia, and balismus could result from reduced output activity, which decreases the inhibitory effect of the thalamus, and thus promotes the

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activity of excitatory thalamocortical projectionds. There following hyperkinetic disorders can be distinguished:

#### 1. Chorea

The manifestations of chorea are involuntary, arrhythmic, disordered hyperkinesias affecting various muscle groups of the face, limbs and head. They are abrupt and sudden, brief, fast and explosive. Most often they are observed in the distal parts of upper limbs, torso, face, tongue and larynx. Hyperkinesias in the arms disrupt usual activities such as dressing, eating washing, etc. The gait is 'dancing'. There is a muscle hypotonia, which promotes the rapidity, suddenness and wide amplitude of the movements.

#### 2. Athetosis

Athetotic hyperkinesias are involuntary and tonic, but slow and of greater amplitude compared to choreic ones. They feature coarse, worm-like movements. The condition is characterized by inability to keep the fingers, thumb, tongue and other parts in a single position. The maintained position is disrupted by slow worm-like, tonic movements, which tend to concatenate in seamless series. Athetosis affects primarily distal muscles, such as those of the fingers, face, tongue and neck. The muscle tone changes frequently.

#### 3. Balismus

This form of hyperkinesia tipically affects the half of the body, hence it is referred to as hemibalismus. The patients present coarse, disorderly, sudden sideways jerks, which are much more violent than in chorea. Typically, it affects the proximal muscles of the pelvic and pectoral girdle. Balismus can be combined with muscular hypotonia.

## 4. Dystonia

Dystonia is involuntary movement characterized by prolonged muscular contractions that result in twists, rotational movements and abnormal posture. They involve larger parts of the body than athetosis and produce bizarre postures and distortions due to the excessively increased muscle tone in certain muscle groups.

Depending on the affected parts of the body, dystonia can be generalized, focal or segmental:

- Torsional dystonia
- Cervical dystonia
- Blepharospasmus

### 5. Myoclonic disorders

Myoclonic disorders are sudden, short-lived, involuntary contractions of muscles, parts of muscles or muscle groups, regardless of any functional connection between them. The movements can be ad-hoc or repeated and are similar to those elicited by

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muscular stimulation. Myoclonic disorders can be rhythmic or arrhythmic, synergistic or asynergistic, topical or diffuse.

## 6. Tics

Tics are fast, stereotypical, repetitive movements localized in a particular muscle group. Most often they affect the face, neck, and shoulder shrugs, or head jerks. Tics are often seen in infant age and their genesis is mainly neurogical.