Hyperkinetic Emergencies
Pr François Tison, Bordeaux, France

Movement Disorders emergencies:
« Any movement disorder which evolves over hours to days, in which failure of appropriately diagnose and manage patients can result in morbidity or even mortality »
Poston and Frucht (J Neurol 2008;255:2-13)

- Parkinonism
- Hyperkinetic:
  dystonia, chorea, tics, myoclonus
General clinical approach (1)

1. To observe: localisation, speed, frequency, permanent or intermittent, repetitive or chaotic? rhythm and amplitude

   jerk     brief     slow
   myoclonic    choreic    dystonic

   duration

2. The phenomenology:
   - dystonia,
   - ballism,
   - chorea,
   - myoclonus,
   - tics

General clinical approach (2)

✓ **Ask:** time course ?, triggering/ameliorating factors ? continuous/paroxystic, wake/sleep ?

✓ **Review:** medical context, past and present medications, toxins, family history ?

✓ **Examine:** focal deficit, consciousness, meningitis, fever ?

✓ **Order:** lab tests (blood, CSF?), MRI
Dystonic emergencies

- « Status dystonicus »
- Acute drug-induced dystonia
- Breathing and swallowing dystonias

Pseudo-dystonic emergencies:
- Pseudo-torticollis (kids +++): Atlanto-axial subluxation inflammatory head and neck processes, posterior fossa and craniocervical junction tumors
- Tetanus
- Seizures (partial seizures with motor manifestations = frontal)
« Status dystonicus »


• Context of poorly controlled generalized idiopathic dystonia

• OR context of secondary dystonias: Wilson’s disease, post-traumatic, post-anoxic, PKAN …
Rethinking Status Dystonicus

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**Triggering factors:** Infection = 51.7%, drugs = 30%, surgery = 6.7%, metabolic disorder = 5%, DBS failure = 5%

No apparent cause = 32.6%
« Status Dystonicus » → life threatening

« movement disorder emergency characterized by severe episodes of generalized hyperkinetic movement disorders that had necessitated urgent hospital admission because of life-threatening complication regardless of the patient neurological condition at baseline »

- Stiffness and pain
- Rhabdomyolysis
- Dysphagia/aphagia
- Aspiration pneumonia
- Impaired ventilation
- Infections, fever
- Dehydratation
- Multi-organ failure (renal)
« Status dystonicus »

differential diagnosis

- Neuroleptic malignant syndrome (neuroleptics, fever, autonomic, CK +++)
- Serotonin syndrome (SSRI, myoclonus, confusion and agitation)
- Malignant hyperthermia (anesthesia, fever, rhabdomyolysis)
- Baclophen pump withdrawal

TABLE 2. Neurological emergencies and their comparison with status dystonicus

<table>
<thead>
<tr>
<th></th>
<th>Myoclonus</th>
<th>Rigidity</th>
<th>Tremor</th>
<th>Dystonia</th>
<th>Decreased LOC at onset</th>
<th>Hyper-reflexia</th>
<th>CK Increase</th>
<th>Autonomic dysfunction</th>
<th>Note</th>
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<tbody>
<tr>
<td>Status dystonicus</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+ (crisis)</td>
<td>-</td>
<td>+/−</td>
<td>+/−</td>
<td>+/−</td>
<td>Different triggers</td>
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<tr>
<td>Status epilepticus</td>
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<td>+/−</td>
<td>+/−</td>
<td>+/−</td>
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<td>Abnormal EEG</td>
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<td>Neuroleptic malignant</td>
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<td>-/−</td>
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<td>Neuroleptics use</td>
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<td>syndrome</td>
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<tr>
<td>Serotonin syndrome</td>
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<td>Serotoninergic drugs, hyperreflexia</td>
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<tr>
<td>Malignant hyperthermia</td>
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<td>Malignant hyperthermia, myoclonus, confusion and agitation</td>
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<tr>
<td>Acute Parkinsonism</td>
<td>+</td>
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<td>+/−</td>
<td>+/−</td>
<td>Acquired drugs, toxins, PD</td>
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<tr>
<td>Dystrophia myotonica</td>
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<td>+/-</td>
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<td>-/−</td>
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<td>Acquired CNS lesion</td>
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<tr>
<td>Cardiomyopathy</td>
<td>+</td>
<td>+/-</td>
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<td>-/−</td>
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<td>+/−</td>
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<td>PD patients</td>
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<tr>
<td>Parkinson-</td>
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<td>dystonia/myasthenia</td>
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<td>+/−</td>
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<td>PD patients</td>
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<tr>
<td>Instability with dystonia</td>
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<td>Acquired CNS lesion</td>
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<td>Intestinal ileusation</td>
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<td>Acquired CNS lesion</td>
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<tr>
<td>Acute dystonic reactions</td>
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<td>-/−</td>
<td>+/−</td>
<td>+/−</td>
<td>+/−</td>
<td>Acquired CNS lesion</td>
</tr>
</tbody>
</table>

*present; −: absent; +/- may or not be present.
LOC: level of consciousness; Dyst: dystonia; Myo: myoclonus; CK: creatine kinase; EEG: electroencephalogram; DRB: dopamine receptors blockade; PD: Parkinson’s disease; CNS: central nervous system.
« Status Dystonicus » = management

Deep sedation

Mariotti P et al. Movement Disorders 2007; 22(7):963-968
« Status Dystonicus » = management

- Bilateral GPI DBS
Acute drug-induced dystonia

- **Neuroleptics +++** (typical: 6%, atypical: 1-2%) including anti-emetics
- Introduction or dose increase (usually <24h, 90% within 5 days)
- Young males (tardive dyskinesias and parkinsonism in older women, acute choreic reactions in children)
- Self-limiting but distressing may be life-threatening (laryngeal dystonia)
Acute drug-induced dystonia

More frequent clinical forms: head and neck!

- **Oculogyric crisis:** eye deviation/head rotation, opisthotonus, rigidity, autonomic symptoms, dysarthria, anxiety
- **Oculo-cephalic dystonia:** trismus, torticollis, blepharospasm, laryngeal dystonia
- Any form of dystonia

**Management:**
- Parenteral anticholinergics (trihexyphenidil diphenylhydramine 25-50 mg, benztropine 1-2mg often repeated + 5-7 day oral course),
- Clonazepam or diazepam if resistant
Breathing and swallowing dystonias

- **Spasmodic dystonia**: usually no airway obstruction unless botulinum toxin-induced weakness

- **Adductor laryngeal dystonias** (Gerhardt’s syndrome), adductor spasms with stridor in focal dystonias, X-linked-dystonia parkinsonism (Lubag) and MSA

- **Tardive dystonias** (laryngeal spasms)

- **HD and chorea-acanthocytosis**
Choreic emergencies


- Vascular (50%)
- Drug-induced (16%)
- Metabolic (14%)
- AIDS-related (12%)
- Infectious and inflammatory (8%)

- Hemichorea-Hemiballism
- Severe levodopa induced-dyskinesias
- Acute generalized chorea (secondary causes)
Hemichorea-Hemiballism

- **Hemiballism**: Acute onset, dramatic flinging rotatory movements of proximal muscles on one side (arm and/or leg)
  Increased by action, stress, absent during sleep
- **Hemichorea**: less ample, more distal
Hemichorea-Hemiballism

- Share the same pathophysiology: historically a lesion of the controlateral STN (15%) but anywhere within the BG (GPI, putamen, caudate) and adjacent WM (85%).

  *Postuma RB and Lang AE
  *Lancet Neurology 2003;2:661-68*

- Any type of lesion, stroke far more common (>75%)

- Postpump chorea following CP bypass (1.2%) in childrens
Hemichorea-Hemiballism

- Distressing, exhausting, self-injuries, dehydration and rhabdomyolysis in most severe cases
- Usually subsides within hours or days, but may be prolonged in a minority
- Long-term prognosis depends on the prognosis of the underlying disease
- Management: of the cause (stroke, hyperglycaemia), protection of limbs (pads), rehydration
- If severe and/or prolonged consider neuroleptics (haloperidol) or tetrabenazine
- GPi or thalamus DBS (or lesion)
Hemichorea-Hemiballism

- May also be metabolic: « hyperglycaemic hemiballism »
- Women >65 y, diabetes type 2, more frequent in Asians
- Severe hyperglycaemia, subsides after correction
- Characteristic = high signal on T1 MRI sequences in the putamen
  
  Postuma RB and Lang AE
  Lancet Neurology 2003; 2:661-68
Drug-induced chorea/ballism

More frequent = L-Dopa in PD:
L-Dopa dose increase or ICOMT introduction
- Risk of dehydration and rhabdomyolysis
- Management: decrease dosage or transient withdrawal, rehydration, sedation (diazepam)

Other drugs: lithium, anti-epileptic drugs (lamotrigine), opioids and methadone
Acute/subacute generalized choreas

- Choreas of rapid onset, often associated with other neurological or neuropsychiatric features:
- **Disclose secondary causes +++**
  - Auto-immune diseases = Post-streptococcal neurological disorders (Sydenham chorea), Systemic Lupus erythematosus (SLE) and anti-phospholipid syndrome (APLS)
  - Metabolic = glucose, thyreotoxicosis
  - Encephalitis
Myoclonic emergencies

Generalized myoclonus and/or asterixis (« negative myoclonus ») are common in the setting of metabolic, toxic and drug-induced encephalopathies:

- Liver, kidney and respiratory failure
- Serotoninergic drugs « serotoninergic syndrome », tricyclic anti-depressants, lithium
- Opiates and benzodiazepine withdrawal, amphetamine, cocaine ecstasy
- Post-anoxic myoclonus
Opsoclonus-myoclonus

Subacute Triad:
- Opsoclonus
- Myoclonus: Craniocervical and trunk, extremities (adults)
- Ataxia

- **Paraneoplastic** (Anti Ri, HU, others)
  Neuroblastoma in childrens
  Others (Melanoma, non-Hodgkin
- **Auto-immune** (NMDA, AMPA, GLUR5, GABA-B, GAD…)
- **Infections** (VZV, EBV, Coksackie, West Nile, Lyme …)

Traitement: IVIg, corticosteroids, plasmapheresis immunosuppressants
Tics

- Brief paroxysmal movements and vocalizations accompanied by a premonitory urge to move.
- Rare at the emergency ward:
  - Exacerbation (stress, fatigue, infections, drugs (stimulants, AD, BZD withdrawal)
  - Behavioural/compulsive/self-mutilation crisis in Gilles de la Tourette
Psychogenic Jerks and tremors

Context:
- Psychiatric illness (rare)
- History of psychogenic disorder
- Physical or emotional stress/trauma

Clinical picture:
- variable, complex,
- inconsistent phenomenology,
- suggestibility, distractibility,
- Absence of urge to move

Explorations?
- Muscle activation pattern
- Bereitschaftspotential preceding the movement
... and two diagnosis and therapeutic emergencies

#1 Wilson’s disease

- Any emergent movement disorder in the young (< 40 y+++): tremor (any type, midbrain+++), dystonia (face +++ risus sardonicus) and choreo-athethosis

- Urgent to diagnose: serum and urine copper, plasma ceruleoplasmin, slit-lamp examination (KF ring), brain MRI.

- Urgent to treat by decoppering drugs: irreversible brain and liver damage!
#1 Wilson’s disease

Chelation drama and Liver grafting « miracle »
… and two diagnosis and therapeutic emergencies

#2 Whipple’s disease

- Oculo-masticatory or oculo-facio-skeletal myorhythmias
- Progressive supranuclear palsy and/or cognitive and behavioral changes
- Jejunal biopsy for histological and PCR analysis
- PCR (T. Whippelii) in the CSF
Whipple’s disease:
to treat (sulfamethoxazole-trimethoprim and/or ceftriaxone) and cure
Conclusion

- **Recognize:**
  - the movement disorder
  - the emergency situation

- **Diagnose the more common causes:**
  - cerebrovascular diseases
  - acute drug reactions

- **Urgent to « cure »:** the « 2W »
  - Wilson’s disease, Whipple’s disease
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Humana Press, 2005