Dystonia

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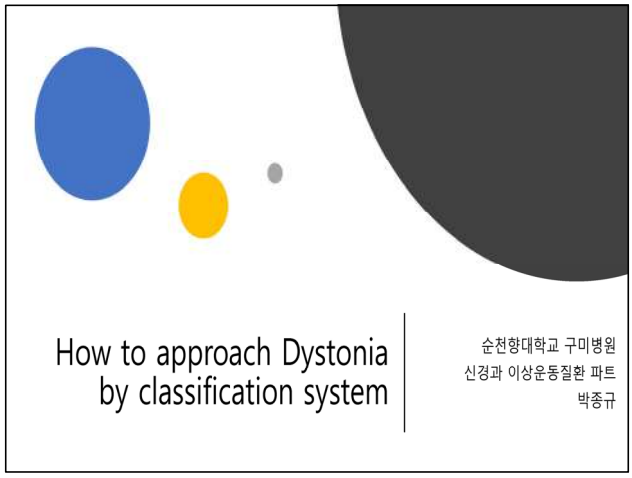

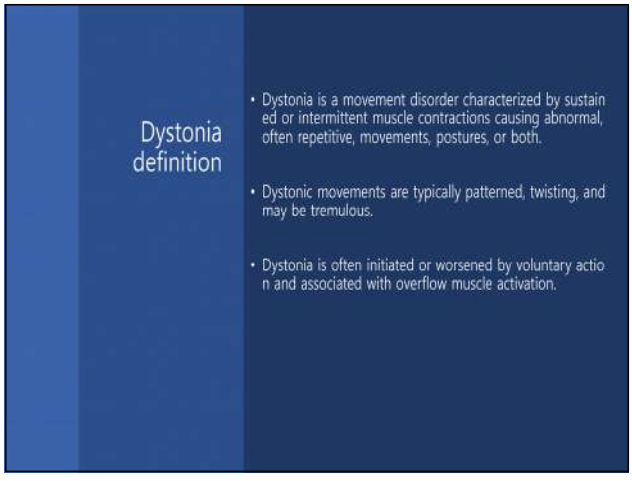

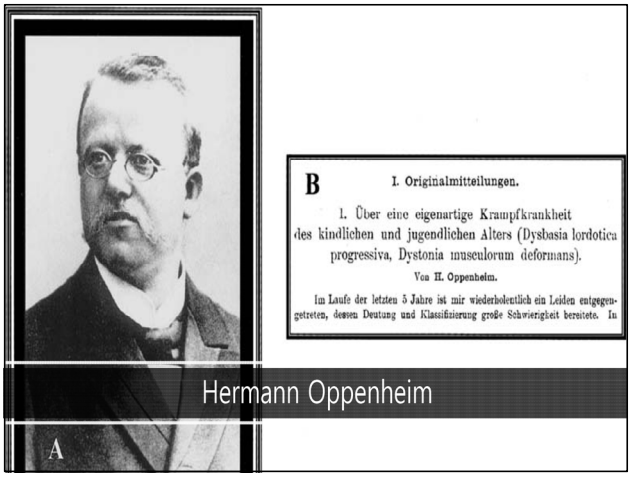

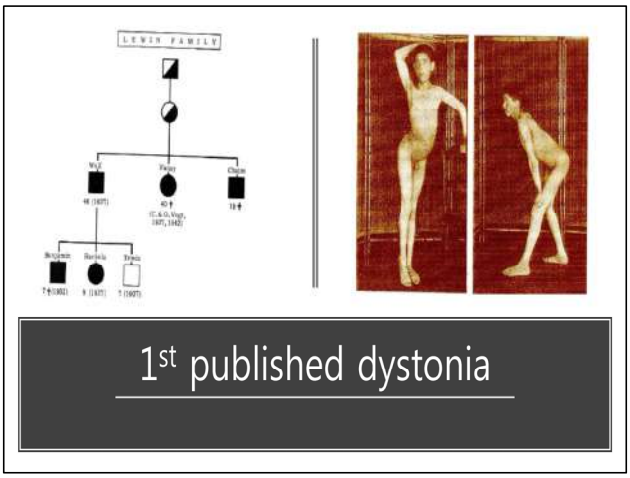

Dystonia described by Oppenheim

Hypotonia and tonic muscle spasm, on volitional movements

No paresis, no atrophy, no alteration of electrical excitability, no sensory, nor perceptive, nor sphincter disturbances, no significant psychological abnormalities

How to describe weird movement

- Level A: immediate impressions
- Level B: more observation
- Level C: longer observation

Level A: immediate impressions

- Rhythmic vs Arrhythmic
- Sustained vs Non-sustained
- Parxysmal vs Continual vs Continuous
- Sleep vs Awake

How to describe weird videos

- Level A: immediate impressions
  - Rhythmic vs Arrhythmic
  - Sustained vs Non-sustained
  - Paroxysmal vs Continual vs Continuous
  - Sleep vs Awake

Level A: immediate impressions

Rhythmic	Irregular	Arrhythmic
Tremor	Cortical myoclonus	Chorea
resting	Minipolymyoclonus	Ballism
postural	Dystonic tremors	Dystonia
action	Epilepsia partialis continua	Akathitic movements
intention	Moving toes, fingers	Athetosis
Orthostatic tremor		Hemifacial spasm
Dystonic tremor		Hyperekplexia
Myoclonus, segmental		Arrhythmic myoclonus
Myoclonus, oscillatory		some Stereotypies
Myorhythmia		Tics
Periodic limb movements in sleep		
some Stereotypies		
classic Tardive dyskinesia		

Level A: immediate impressions

Sustained	Non-sustained
Stiff-person	
Rigidity	All others
Dystonia	
Oculogyric crisis	
Dystonic tics	
Congenital torticollis	
Orthopedic torticollis	
Neuromyotonia	
Sandifer syndrome	

Level A: immediate impressions

Paroxysmal	Continual	Continuous
Tics	Chorea	Abdominal dyskinesia
Paroxysmal kinesigenic dyskinesia	Ballism	Athetosis
Paroxysmal non kinesigenic dyskinesia	Dystonic movements	Tremor
Paroxysmal exertional dyskinesia	Myoclonus, arrhythmic	Dystonic posture
Episodic ataxia	Some stereotypies	Minipolymyoclonus
Episodic tremor		Myoclonus, rhythmic
Hypnogenic dystonia		Tardive stereotypy
Some Stereotypies		Myokymia
Akathitic movements		Tic status
Some Jumpy stumps		Moving toes, fingers
		Myorhythmia

Level B: more observation

Rest vs Action vs Overflow
Patterned vs Non-patterned
Combinations of movements

Level B: more observation

Rest Only	Action Only	Rest and Continues with action
Resting tremor	Ataxia	Chorea
Akathitic movements	Tremor: postural, action, intention	Ballism
Paradoxical dystonia	Action Dystonia	Athetosis
Restless legs	Action myoclonus	Minipolymyoclonus
Orthostatic tremor	Task specific dystonia	Moving toes, fingers
Posture specific tremors		Myokymia
		Tics
		Abdominal dyskinesias

Level B: more observation

Patterned (same muscle group)	Non-patterned
Dystonia	
Hemifacial spasm	All others
Moving toes, fingers	
Segmental myoclonus	
Myorhythmia	
Stereotypies	
Tardive dyskinesia	
Tremor	
Abdominal dyskinesias	

Level C: longer observation

Speed: slow vs fast
Amplitude: ballistic vs not ballistic
Force: powerful vs easy to overcome
Suppressibility
Vocalizations
Self-mulitation
Complexity of movements
Sensory component
Ocular movements

Level C: longer observation - speed

Fastest	Intermediate	Slowest
Minipolynmyoclonus	Chorea	Athetosis
Myoclonus	Ballism	Moving toes, fingers
Hyperekplexia	Tremors	Myorhythmia
Startle		Akathitic movements
Hemifacial spasm		Hemifacial spasm(tonic)

Level C: longer observation - amplitude

Large amplitude	Medium	Very Small
Ballism	Chorea	minipolymyoclonus
	All others	

Level C: longer observation - force

Powerful	Medium	Mild
Fixed postures of dystonia	Dystonia	All others
Jumpy stumps		
Stiff-person syndrome		

Classification of Dystonia

Mov Disord. 2013 Jun 15;28(7):863-73.

REVIEW

Phenomenology and Classification of Dystonia: A Consensus Update

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Motor phenomenology relevant to dystonia

- Voluntary action induced
- Dystonic tremor
- Overflow
- Sensory trick (gestes antagonistes)

List of pseudodystonias

- Dystonic (tonic) tics
- Head tilt (vestibulopathy, trochlear nerve palsy)
- Bent spine, camptocormia, scoliosis
- Atlanto axial and shoulder subluxation
- Arnold-Chiari malformation
- Soft tissue neck mass
- Congenital muscular torticollis
- Congenital Klippel-Feil syndrome
- Satoyoshi syndrome
- Dupuytren's contractures
- Trigger digits
- Neuromuscular causes (Isaacs syndrome, etc.)
- Spasms (hypocalcemia, hypomagnesemia, alkalosis)
- Orthopedic and rheumatological causes
- Sandifer syndrome
- Deafferentiation (pseudoathetosis

Red flags on history and examination

Abnormal birth/perinatal history
Developmental delay
Seizures
Previous exposure to drugs e.g. dopamine receptor blockers
Continued progression of symptoms
Prominent bulbar involvement by dystonia
Ununual distribution of dystonia given age of onset (e.g. leg dystonia in an adult)
Hemidystonia
Presence of another movement disorder
Additional neurologic symptoms (pyramidal signs, cerebellar signs, cognitive decline)
Other systems affected (e.g. organomegaly)

Classification of Dystonia

01

Axis I. Clinical characteristics

02

Axis II. Etiology

Axis I. Clinical characteristics

Age at onset

- Infancy: birth to 2 yeas
  - Inherited metabolic disorder, poor prognosis
- Childhood: 3 – 12 years
  - Dystonic cerebral palsy
- Adolescence: 13 – 20 years
  - Dopa-responsive dystonia
- Early adulthood: 21 – 40 years
- Late adulthood: > 40 years
  - Sporadic focal dystonia

Axis I. Clinical characteristics

Body distribution

- Focal
  - Blepharospasm
    - video
  - oromandibular dystonia
    - video
  - cervical dystonia
    - video
  - Laryngeal dystonia
    - video
  - Writer's cramp
    - video

Axis I. Clinical characteristics

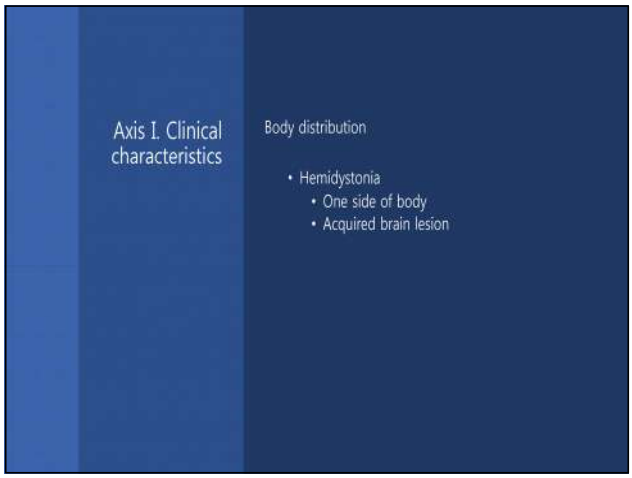
Body distribution

- Segmental
  - Cranial dystonia: Blepharospasm with lower facial and jaw or tongue involvement
  - Bibrachial dystonia
- Multifocal

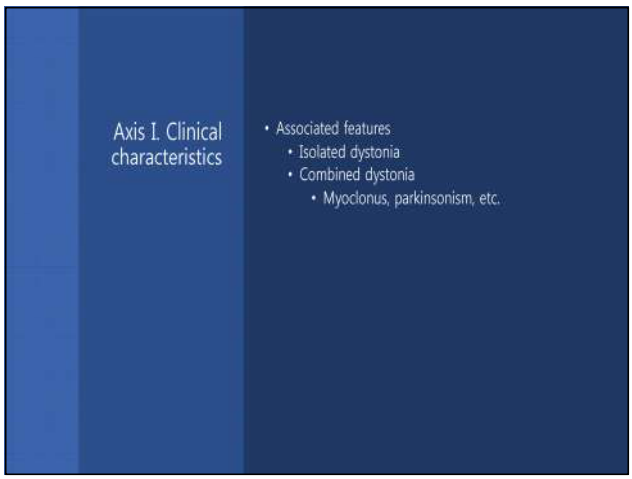
Axis I. Clinical characteristics

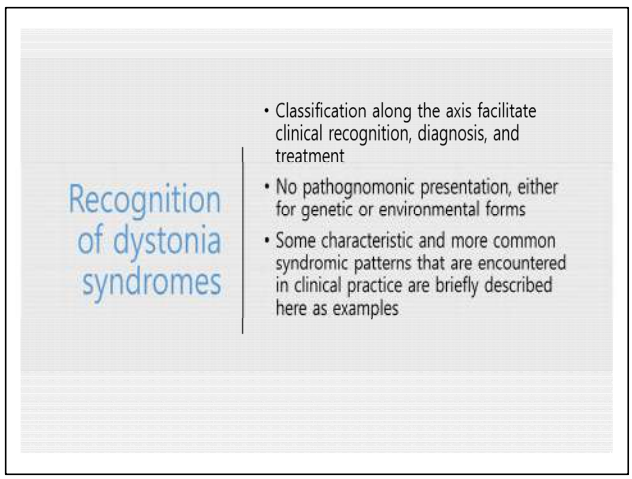
Body distribution

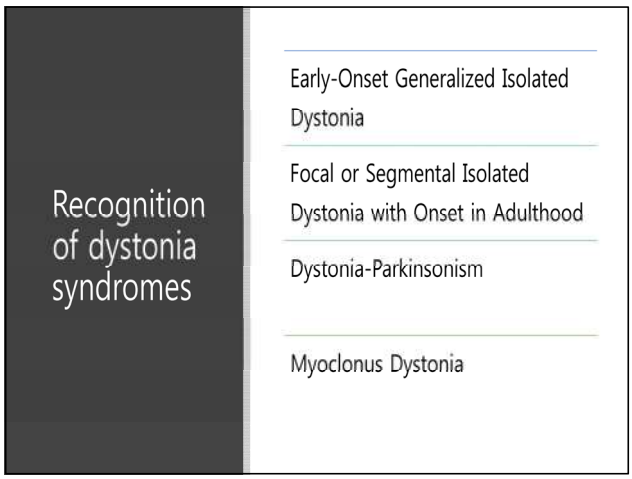
- Generalized
  - Trunk and at least 2 other sites
  - Video
- Lower limb dystonia in children may be an initial symptom of generalized dystonia
- Lower limb dystonia in adults – identifiable cause

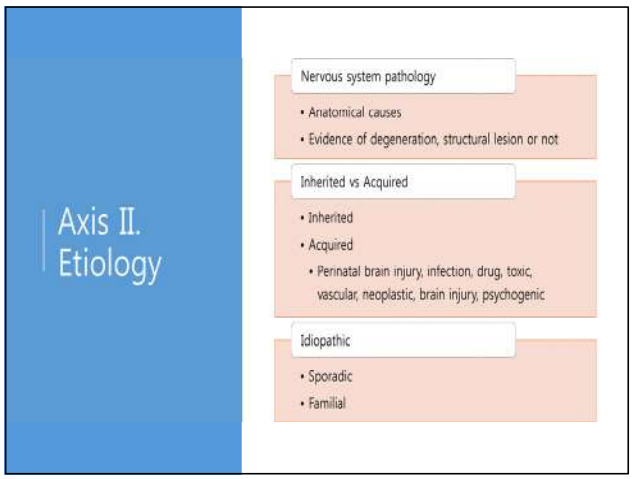

Axis I. Clinical characteristics
Body distribution
• Hemidystonia
• One side of body
• Acquired brain lesion

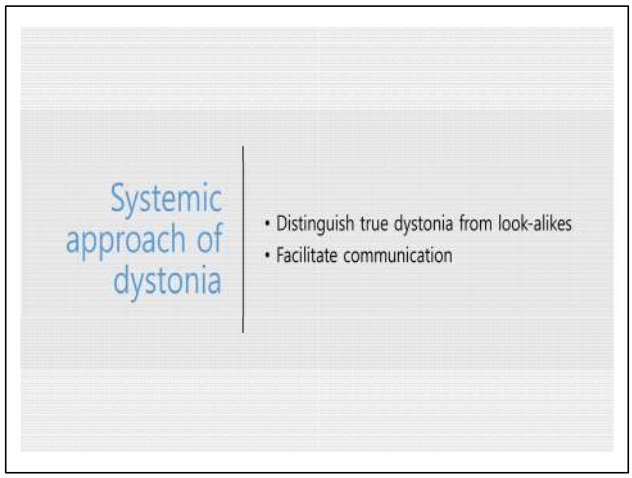

Axis I. Clinical characteristics
Temporal pattern
• Disease course
• Static vs Progressive
• Variability
• Persistent
• Action-specific
• Occurs only during a particular activity
• Diurnal fluctuations
• Circadian variations
• Paroxysmal
• Sudden self limited episodes


Axis I. Clinical characteristics
• Associated features
• Isolated dystonia
• Combined dystonia
• Myoclonus, parkinsonism, etc.


Recognition of dystonia syndromes
• Classification along the axis facilitate clinical recognition, diagnosis, and treatment
• No pathognomonic presentation, either for genetic or environmental forms
• Some characteristic and more common syndromic patterns that are encountered in clinical practice are briefly described here as examples


Recognition of dystonia syndromes
Early-Onset Generalized Isolated Dystonia
Focal or Segmental Isolated Dystonia with Onset in Adulthood
Dystonia-Parkinsonism
Myoclonus Dystonia


Axis II. Etiology
Nervous system pathology
• Anatomical causes
• Evidence of degeneration, structural lesion or not
Inherited vs Acquired
• Inherited
• Acquired
• Perinatal brain injury, infection, drug, toxic, vascular, neoplastic, brain injury, psychogenic
Idiopathic
• Sporadic
• Familial


Systemic approach of dystonia
• Distinguish true dystonia from look-alikes
• Facilitate communication