Movement Disorders mimicking Epileptic Seizure

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Movement Disorders and Epilepsy

- Semiological mimicking
  - paroxysmal
  - episodic
  - myoclonic
- DDx. with diagnostic tools
  - Electrophysiological (EEG, SEP)

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MDs mimicking Epileptic Seizures

- Epilepsia Partialis Continua (EPC)
- Paroxysmal Kinesigenic dyskinesia (PKD)
- Vascular induced MDs (Moyamoya disease)
- Psychogenic MDs

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from DY Kwon in KUMC

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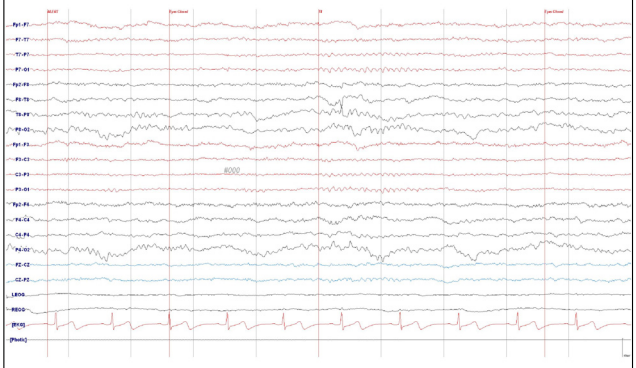

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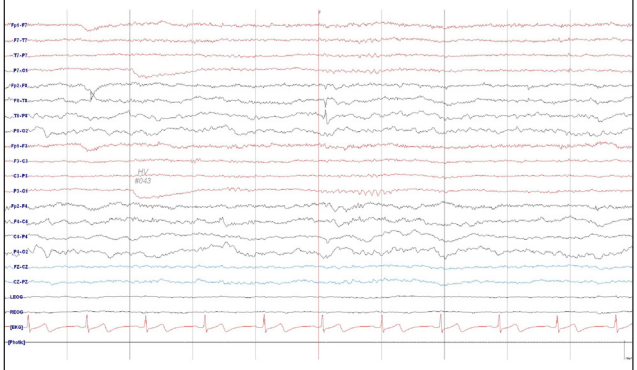

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- Mental status: Alert
- Background rhythm: 10-11Hz
- Classification: Abn III
- 1. Spike, Rt. temporal region (T8 max), 0.5-1/min
- 2. Intermittent theta to delta slow, Rt. hemisphere, moderate amount
- Conclusion: These 32-channel digital EEG findings are suggestive of partial seizure disorder arising from Rt. temporal region and mild cerebral dysfunction in Rt. hemisphere.

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Novel Movement Disorder of the Lower Lip: Is It Epilepsia Partialis Continua? Clues From a Secondary Case

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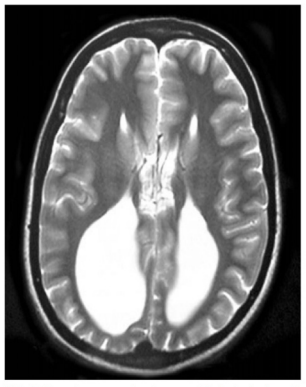

FIG. 1. Magnetic resonance imaging scan of the brain showing evidence of agenesis of the corpus callosum as revealed by widely separated frontal horns of the lateral ventricles, raised third ventricle, and posterior colpocephaly.

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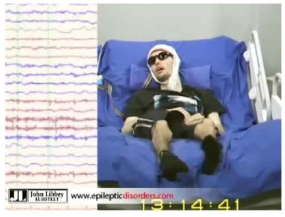

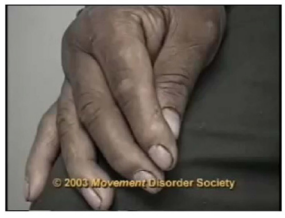

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Epilepsy Partialis Continua (EPC)

- **Definition**

1. Clinical (semiological) grounds
   - spontaneous regular or irregular clonic muscular twitching affecting a limited part of the body
   - sometimes aggravated by action or sensory stimuli
   - occurring for a minimum of one hour, and recurring at intervals of no more than seconds

2. additional electrophysiological evidence
   - epileptiform EEG abnormalities
   - giant SSEPs

➔ "Cortical myoclonus"??

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Epilepsy Partialis Continua (EPC)

- **Semiology of EPC**
  - combination of the repetitive myoclonic jerks with hemiparesis
  - monomorphic, simple, brief excursions of the affected limb
  - regular or irregular occurrence of the jerks
  - involvement of distal rather than proximal muscle groups
  - physical exercise, sensitive stimulation or psychic exertion may increase the amplitude and frequency of the myoclonic jerks
  - more frequent involvement of the upper than the lower half of the body
  - more often continues during sleep
  - a mean frequency of 90 jerks per minute (1.5 Hz)

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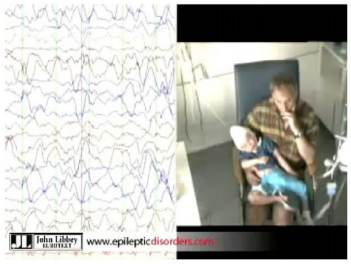
Epilepsy Partialis Continua (EPC)

- Differential diagnosis

1. tremor
2. myoclonic jerks
3. Parkinson's disease



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Paroxysmal dyskinesia (PD)

- Genetically and clinically heterogenous
- Seven different forms

1. Paroxysmal kinesigenic choreoathetosis (PKC)
2. Paroxysmal dystonic choreoathetosis (PDC)
3. Paroxysmal exertion-induced dyskinesia (PED)
4. nocturnal hypnogenic paroxysmal dyskinesia
5. paroxysmal choreaoathetosis and spasticity (CSE)
6. infantile convulsions and paroxysmal choreoathetosis (ICCA)
7. rolandic epilepsy, paroxysmal exercise-induced dystonia and writer's cramp (RE-PED-WC)

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Are Paroxysmal Dyskinesias Epilepsies?

- Simmilarity

1. recurrent episodes of abnormal brain function that manifest as stereotyped
2. clinical responsiveness to medications that prevent repetitive neuronal discharge, the central pathophysiologic event in cortical seizure
3. the presence of an aura for many patients
4. normalization of neurologic exam between events
5. resemblance to supplementary motor cortex seizures
6. familial association

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Yes, it's an epileptic disorder.

- Based on

1. occasional reports of interictal EEG abnormalities in patients with PMD
2. by the response of some PMD to antiepileptic medication

- 8 yrs old PKC Pt. with normal EEG
  → prolonged seizure on postictal EEG ➔ Reflex epilepsy *(Beaumanoir)*
- 18 yrs old PKC Pt. with 5Hz spike on ictal EEG
  → 3/300 PKC Pts : EEG abnormality *(Hirata)*

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No, it's a non-epileptic disorder

- Based on,

Epilepsy
; paroxysmal occurrence of specific, usually brief, stereotyped events.
; accompanied by interictal epileptiform discharges on EEG

PMD
; stereotyped motor events but not evolve into tonic-clonic seizures, not associated with epileptiform discharges.

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No, it's a non-epileptic disorder

- In PMD,
  1. the attacks are paroxysmal, non-progressive.
  2. most EEG
     : no seizure activity that attacks do not evolve into generalized or focal convulsions
  3. there are no LOC.

     → support a subcortical focus

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Clinical Analysis of Paroxysmal Kinesigenic Dyskinesia

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Background : To define the pathophysiology of paroxysmal kinesigenic dyskinesia(PKD), we analyzed detailed clinical features. **Methods** : We studied characteristics of the attack, family history, response to the treatment and clinical courses of 30 patients with PKD. **Results** : Twenty-six of the 30 patients were men and four were women. Thirteen patients had a family history of PKD. There were no patients who had symptomatic PKD. In three-fourths of our patients, the attacks ameliorated within 10 seconds and two-thirds experienced one to ten attacks per day. They showed dystonia much more frequently than chorea. In all patients,sudden movements of the legs while standing precipitated the attacks. The attack occurred very rarely during driving or swimming. Sudden movements of the arm did not precipitate the attacks. **Conclusions** : We suggest that neuronal system maintaining standing posture and strong afferent inputs delivering sudden high velocity movements of the legs to the spinal cord are involved in the genesis of PKD.
J Korean Neurol Assoc 20(3):000~000, 2002

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Table 1. Clinical characteristics of our patients

Patient No.	Gender	Onset Age (year)	Type of dyskinesia	Duration	Frequency (per day)	Treatment	Response
1	M	13	D	3-7s	10-20/d	PHT	Poor
2	M	25	D	5-6s	1-2/d	PHT	Good
3*	M	13	D	10s	4-5/y	CBZ	Good
4*	M	6	D	10s	3-4/d	PHT	Poor
5*	M	6	D	5s	10-30/d	PHT	Good
6	M	4	D	2-3s	10/d	CBZ	Good
7	M	7	CD	10s	10-20/d	PHT	Good
8	M	12	D	2-3m	2-3/y	PHT	Good
9	M	15	CD	5-10s	3-5/d	uk	
10	M	9	CD	10s	3-4/d	PHT	None
11*	M	12	D	20-30s	0-10/d	PHT	Good
12	M	13	D	10s	4-5/d	PHT	Good
13*	M	9	D	60s	3-5/d	PHT	None
14	M	11	D	3-5s	20-30/d	uk	
15	M	11	D	5-10s	5-6/d	uk	
16*	F	8	D	1-2s	40-50/d	uk	
17	M	14	D	3s	0-5/d	uk	
18	M	12	D	10s	10-20/d	PHT	Good
19*	M	5	D	5s	3-4/d	uk	
20*	M	12	D	30s	0-5/d	PHT	Poor
21	M	15	D	20s	10/d	uk	
22*	F	8	D	60s	30/d	uk	
23	M	15	D	10s	0-10/d	uk	
24*	F	10	D	20-30s	10/d	uk	
25	M	16	D	1-2m	30/d	uk	
26	M	14	D	3s	0-10/d	PHT	None
27*	F	13	D	10s	1-6/d	PHT	Good
28*	M	11	D	5s	1-3/d	PHT	Good
29*	M	15	D	5s	1-2/d	None	Self limited
30	M	12	D	5s	10-20/d	PHT	Good

* positive family history; D. Dystonia; CD.Choreodystonia; uk.Unknown; PHT.Phenytoin; CBZ.Carbamazepine; s.Second; m.Minute

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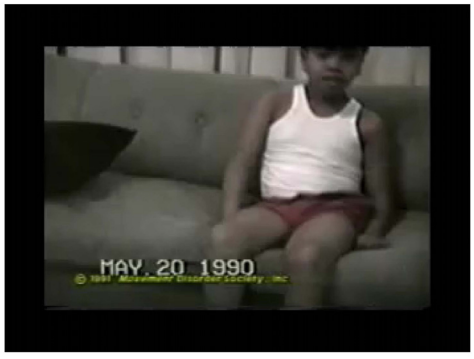

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Movement Disorders
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Movement Disorders Associated with Moyamoya Disease: A Report of 4 New Cases and a Review of Literatures

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Abstract: The aim of this study was to define the clinical characteristics of patients who developed movement disorders in association with moyamoya disease (MMD). Using PubMed and medical records of our hospital from 1985 to 2008, we searched for patients who developed movement disorders in association with MMD. This study included 38 patients described in previous studies and 4 patients found in the medical records. The onset of movement disorders was thought to be sudden. In 13 patients, the movement disorders were precipitated by hyperventilation or emotional stress. Twenty-seven of the 42 patients developed chorea, 4 patients developed dystonia, and 4 developed a mixture of both. The movement disorders of the remaining 7 patients were described as dyskinesia. A third of the 42 patients developed bilateral movement disorders, and their mean age was younger than that of those with unilateral movement disorders. In 37 of the 42 patients, brain imaging studies showed ischemic lesions, but the remaining 5 patients showed no parenchymal lesions. Cerebral perfusion studies showed hypoperfusion in the basal ganglia and in the cerebral cortical areas. Most patients improved whether they were treated or not. MMD must be included in the differential diagnosis of the sudden onset of dyskinesias, particularly chorea and focal dystonia. Even in patients with no parenchymal lesion in brain imaging studies, cerebral angiography and cerebral blood perfusion studies must be performed, if they develop a sudden onset or recurrent movement disorders preceded by emotional stress or hyperventilation. © 2010 Movement Disorder Society

Key words: movement disorders; moyamoya disease; chorea; dystonia; dyskinesia

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TABLE 1. *Demographic and clinical characteristics of MD-MMD*

Total patients	42
Women	31 (73.8%)
Men	11 (26.2%)
Asian	32 (76.2%)
Non-Asian	10 (23.8%)
Mean age at onset of MD	21.4 yr (1–62)
Mean duration for follow up	18.9 mo (3-48)
MD symptom[a]	
Chorea	31 (67.4%)
Arm only	3 (9.7%)
Arm and leg	18 (58.1%)
Arm and face	2 (6.5%)
Arm, leg and face	8 (25.7%)
Dystonia	8 (17.4%)
Hand	2 (25%)
Foot	2 (25%)
Arm and leg	2 (25%)
neck	2 (25%)
Dyskinesia	7 (15.2%)
arm	5 (71.4%)
arm and leg	2 (28.6%)
Mode of symptom onset	
Constant	27 (64.3%)
Paroxysmal	15 (35.7%)
Laterality	
Bilateral	14 (33.3%)
Unilateral	28 (66.7%)
Treatment	30
Medication	6 (20.0%)
Surgery	21 (70.0%)
both	3 (10.0%)
No treatment	10
Prognosis	40
Subside	32 (80%)
Improved	4 (10%)
Persist	4 (10%)

MD, movement disorders; MMD, moyamoya disease.
[a]Including four patients who have overlap symptoms with both chorea and dystonia.

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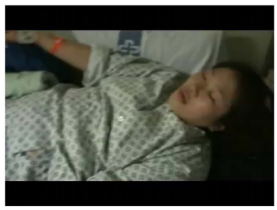


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Diagnostic Dilemma of MUS and PMD

- Absence of reliable diagnostic test
- Both have wide spectrum
- Bizarre presentation
- Possibility of co-occurrence (organic + psychogenic)

10-15% in Movement disorders
10-37% in Epilepsy

Coexistent organic movement disorder		
Never	27	6
1–5%	175	38
6–10%	136	30
11–20%	91	29
21–40%	26	6
>40%	4	1
Coexistent organic neurologic disorder		
Never	56	12
1–5%	198	43
6–10%	115	25
11–20%	70	15
21–40%	17	4
>40%	5	1

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Clues Suggesting a PMD

Table 1 Clues suggesting psychogenic cause

Historical	General examination
Abrupt onset (symptoms often maximal at that time)	Movement inconsistent
Static course	Variability over time (frequency, amplitude, direction/distribution of movement)
Spontaneous remissions/cures	Distractibility reduces or resolves[a], attention increases movement
Paroxysmal symptoms (generally nonkinesigenic)[b]	Selective disability
Psychiatric comorbidities[c]	Entrainment (especially with tremor)
Secondary gain (often not apparent)	Movement incongruous with organic movement disorders
Risk factors for conversion disorder[d]	Mixed (often bizarre) movement disorders
Psychological stressors[e]	Paroxysmal attacks (including pseudoseizures)
Multiple somatizations/undiagnosed conditions	Precipitated paroxysms (often suggestible/startle)
Employed in allied health professions (infrequent)	Suggestibility[f]
	Effortful production or deliberate slowness (without fatiguing) of movement
	Self-inflicted injury (caution: tic disorders)
	Delayed and excessive startle response to a stimulus
	Burst of verbal gibberish or stuttering speech[g]
	False (give-away) weakness
	Nonanatomical sensory loss or spread of movement
	Certain types of abnormal movements common in individuals with PMDs[h]
	Functional disability out of proportion to examination findings

[a] Distractibility should be tested both with mental and motor tasks. Although most often organic movement disorders are not suppressed, organic tics or akathisia can be suppressible, and recently it was shown that diaphragmatic tremor was suppressed by simple motor tasks, perhaps by interference with central nervous system circuitry [9].
[b] Separation from organic paroxysmal dyskinesias can be challenging, particularly if they occur infrequently with prolonged symptom-free periods.
[c] Psychiatric diseases can also coincide with organic illness or present as part of the organic movement disorder.
[d] Sexual and physical abuse, trauma.
[e] Often initiated by injury (often minor) or motor vehicle accident associated with litigation or compensation.
[f] Application of pressure with finger or tuning fork may reduce symptom. With paroxysmal symptom, suggestibility and placebo trial may not be helpful, unless repeated reversals with placebo are documented when symptom otherwise is frequent and attacks are prolonged.
[g] Particularly if the entire word is repeated (typically broken up into syllables, each repeated), rather than the initial syllable.
[h] Such movements include dystonia that begins as a fixed posture (particularly if abrupt onset, painful, and early contractures are seen); bizarre gait; twisting facial movements that move mouth to one side or the other (organic dystonia of the facial muscles usually does not pull the mouth sidewise).
Adapted from [8].

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Placebo in PMD

- For many reasons, it is better to avoid the use of placebos a nd strive to obtain successful results without them *(Ford et al., 1995)*
- If really in doubt about the diagnosis, it can aid in making t he correct diagnosis and thereby lead to proper treatment. *(Tan et al., 2004)*

Segment 1

Psychogenic Paroxysmal Dyskinesia: The Role of Placebo in the Diagnosis and Management

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Video

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.....We used a placebo as both a diagnostic and therapeutic tool in our patient. Based on previous reports and our experience, *placebo treatment can serve as a good diagnostic and therapeutic tool in PMD, especially in psychogenic PKD, like pseudoseizure.* **(Baik et al., 2009)**

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SUMMARY

1. EPCs: Rare type of focal status epilepticus
2. Characteristic semiological features of EPCs
   : to be diagnosed and distinguished from other movement disorders or myoclonic symptoms
3. Paroxysmal dyskinesias
   : subset of the hyperkinetic movement disorders likely reflecting episodic abnormal activity involving the basal ganglia
4. "Are some paroxysmal dyskinesias partial seizures?"
   → necessary further studies of the pathophysiologic basis of paroxysmal dyskinesias for answer this question.
5. Vascular induced MD and psychogenic MD also may differentiate with Epileptic seizure.

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