

Diagnostic Approach of Neuromuscular Disorders



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신경근육계 질환의 특징



- 증상의 국소화 가치는 상대적으로 특이적이다
- 시간적 측면이 상대적으로 더 다양하다
- 다양한 전신적 질환과 연관되어 발생한다
- 종류는 많고 그 빈도는 낮다

Contents: broad + updated + common?

1. Clinical approach
 - 1) History
 - 2) N/Ex
2. Electrodiagnostic tests
 - 1) Conventional NCS
 - 2) RNS
 - 3) Needle EMG
 - 4) Others: QEMG, SFEMG, ANSFT, etc.
3. Laboratory studies
4. Imaging studies
5. Miscellaneous

1. Clinical approach

Take home message: Basic 4 steps



1. 무엇이 신경학적인 문제인가? *Recognition of Impaired Function*

	Sensory symptoms	Motor symptoms
Negative	Reduced or lack of sensation Numbness Postural instability*	Weakness Fatigability Atrophy or hypertrophy
Positive	Paresthesia Pain Burning, shooting, lancinating etc.	Cramps Fasciculations Myotonia Myokymia

Case 1. 55/M 은행원, 오른 팔 통증

Impairment	
Localization	
Etiology	
Ancillary	

Case 2. 22/M 군인, 팔을 못들어요

Impairment	
Localization	
Etiology	
Ancillary	

- 58/M 감지 못한다.
- Impression: paraplegia (X)



- 75/F 못일어난다.
- Impression: general weakness (X)



Atypical sensory symptoms

- 62/M 수전증.
- Impression: ET (X)



- 37/M 상처가 아물지 않는다.
- Impression: skeletal problem (X)



Positive motor phenomenon



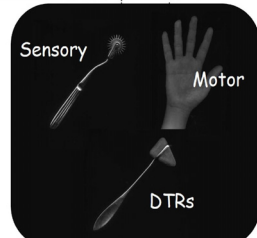
Clues for motor weakness



2. 국소화가 가능한가? 그렇다면 어디?

Localization of Lesion

1. Central lesion
2. Anterior horn
3. Roots / plexus
4. Peripheral mixed nerve
5. Peripheral sensory nerve
6. Peripheral motor nerve
7. Neuromuscular junctions
8. Muscle
9. Tendon



Case 3. 43/M, 근육이 꿈틀거린다

Impairment	
Localization	
Etiology	
Ancillary	

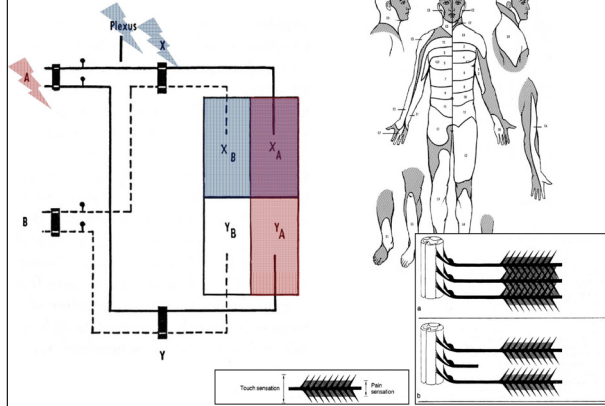
Case 4. 37/M, 발 모양이 이상하다

Impairment	
Localization	
Etiology	
Ancillary	

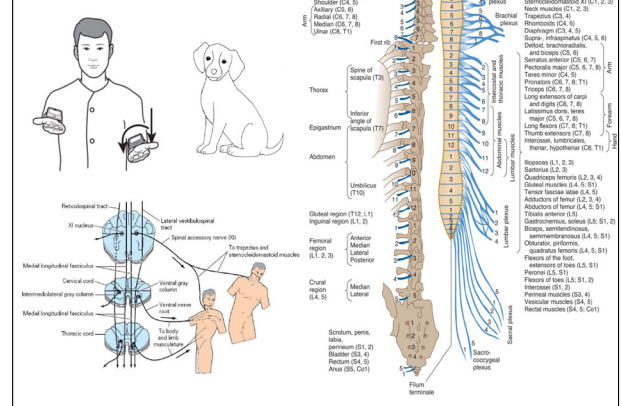
Basic principle of localization

	Sensory	Weakness	DTRs*
Cord	Present (sensory level) Different modality	Distal / symmetric Usually paraplegic	Hyperreflexic
Root	Present (root level)	Distal / Wasting > weakness Late development	Abolished, even early
Nerve	Present	Distal / Wasting > weakness	Abolished, even early
Anterior horn	Absent	Distal / Wasting > weakness	Variable
NMJ	Absent	Proximal / symmetric Fluctuation(+)	Variable
Muscle	Absent	Proximal / symmetric Proportional to wasting	Proportional to weakness

Sensory: pitfalls



Motor: pitfalls

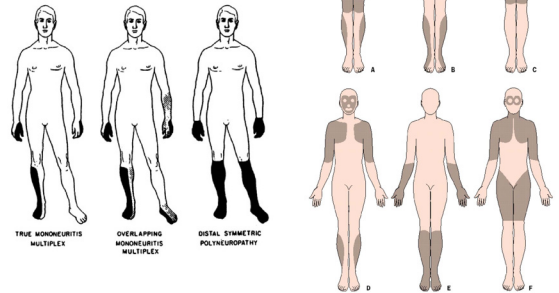


Manual Muscle Power Examination																
Upper Limb																
Movements	EMN	Reflex	Muscle	Nerve	Cord	Division	Trunk	Root								
					L	M	P	A	P	U	M	L	C5	C4	C3	C2
Scapula elevation			Trapezius	Accessory												
Scapula medial rotation			Rhomboid	Dorsal scapular												
Scapula lateral rotation			Serratus anterior	Long thoracic												
Shoulder lateral rotation			SSB	Suprascapular	(+)											
Shoulder medial rotation			Delto	Axillary	(+)											
Elbow flexion	++	+	Biceps	Musculospiral												
			Brachioradialis	Radial	(+)											
Elbow extension	+	+	Triceps													
Forearm pronation			PZ	Median												
			PQ	ADON												
Forearm supination			Supinator	Radial												
Wrist extension	+		ECR longus	Radial												
			ECU	ULN												
Wrist flexion			FCR	ULN												
			CU	Median												
Finger extension	+		EDC	PCN												
Finger flexion	+		FPL, FDP (2+, 3+)	ADON												
			FDP (4+, 3+)	Ulnar												
			DD	Median												
Finger abduction	++	+	FDI, ADM	Ulnar												
	++		APB	Median												

Lower Limb																
Movements	EMN	Reflex	Muscle	Nerve	Nerve	Nerve / plantar	L1	L2	L3	L4	L5	S1	S2			
Hip flexion	++	+	Iliopsoas	Femoral		P. dev.										
Hip adduction			Adductor*	Obturator		A. dev.										
Hip abduction			G medius*	S. gluteal		P. dev.										
Hip extension			G maximus	S. gluteal		P. dev.										
Knee flexion			Hamstring	Tibial peroneal	Sciatic	A. P. dev.										
Knee extension	++	+	Quadriceps	Femoral		P. dev.										
Ankle dorsiflexion			TA	D. peroneal	C. peroneal sciatic	P. dev.										
Ankle eversion			Peronei*	S. peroneal	C. peroneal sciatic	P. dev.										
Ankle plantarflexion	+	+	G, Soleus	Tibial	Sciatic	V. dev.										
Ankle inversion			TP	Tibial	Sciatic	V. dev.										
Big toe extension			EHL	D. peroneal	C. peroneal sciatic	P. dev.										
Toe plantarflexion			FDL	Tibial	Sciatic	V. dev.										

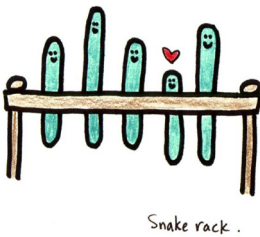
**Special issue in localization:
categorization**

- Mono- vs polyneuropathy
- Myopathy: distribution

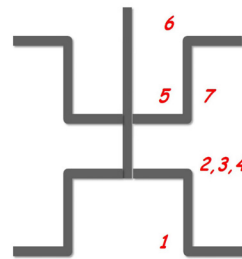


Polyneuropathy

- [Distal symmetric (length-dependent) axonal] polyneuropathy → "glove & stocking" pattern



DTRs



Grade	Description
0	Absent
1+ or +	Hypoactive
2+ or ++	"Normal"
3+ or +++	Hyperactive without clonus
4+ or ++++	Hyperactive with clonus



3. 원인질환의 성격은 무엇일까?

- **Temporal profiles**
 - Acute (episodic) /subacute/chronic (progressive)
 - Monophasic/relapsing
- **Mechanical or focal events**
 - Trauma
 - Compression / entrapment
- **Clues for immunological process**
 - Previous infection
 - Previous vaccination
- **Coexisting medical / genetic conditions**
- **Etc. etc. ...**

Case 5. 21/M, 안면마비?

Impairment	
Localization	
Etiology	
Ancillary	

Case 6. 27/M, 주기성 마비?

Impairment	
Localization	
Etiology	
Ancillary	

2. Electrodiagnostic tests

Diagnostic criteria of ALS (rEEC, Awaji)

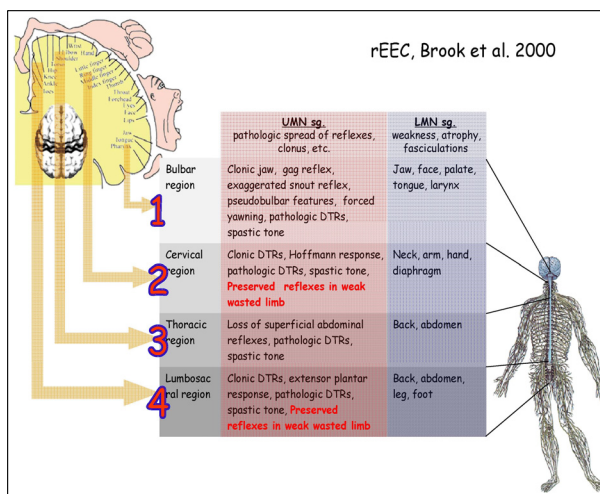
Presence

- LMN degeneration by clinical, electrophysiological or pathological exam.
- UMN degeneration by clinical exam.
- Progressive spread

Absence

- Evidence of other ds. such as, electrophysiological or neuroimaging fx.

rEEC, Brook et al. 2000



Split-hand index for the diagnosis of amyotrophic lateral sclerosis

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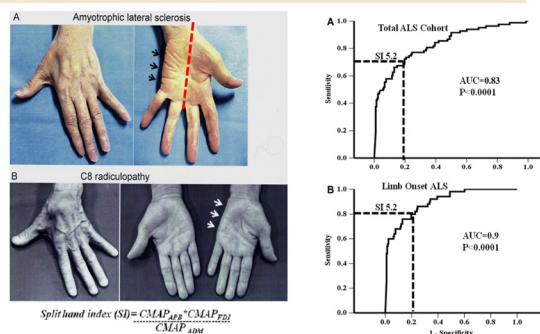
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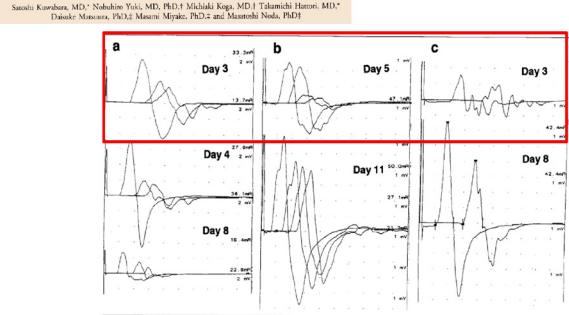


Is ALS an neocortical disease?



Reversible conduction failure of node

IgG Anti-GM1 Antibody Is Associated with Reversible Conduction Failure and Axonal Degeneration in Guillain-Barré Syndrome
Kuwabara et al. Ann Neurol (1998)



AMAN: major form of Korean GBS?

Guillain-Barré syndrome in Asia

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