EPIDEMIOLOGY AND CLASSIFICATION OF MOVEMENT DISORDERS

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To disseminate knowledge and promote research to advance the field of Movement Disorders

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- All member benefits *except* the print version of the journal *Movement* Disorders.
- Apply every 3 years to renew membership.

Task Force on Africa

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The Ambassador Program supports travel to underserved areas for education and scientific exchange.

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The Virtual Professor Program improves the availability of educational initiatives and training in underserved areas.

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Asynchronous Consultation for Movement Disorders (ACMD) Project

- Improve patient care in regions with limited access to specialists.
- Provide education on movement disorders for local physicians.
- Bring together doctors across different time-zones, with different levels of access to participate.

Contact: mguttman@movementdisorders.ca

Waived dues membership

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Learning Objectives

- To briefly describe the epidemiology of movement disorders from a global and African perspective
- To describe the current classification of movement disorders in general
- To describe the current classification of common movement disorders

Outline

- **Definition of movement disorders**
- **Epidemiology (Global data; Africa data)**
  - Parkinson disease
  - Essential tremor
- **Classification**
  - Broad subtypes
  - Parkinsonism
  - Dystonia
  - Tremor
  - Ataxias

What are movement disorders?

- Previously referred to as '*extrapyramidal disorders*' BUT…..
  - basal ganglia is not entirely extrapyramidal: intimate connections with pyramidal tract
  - other '*extrapyramidal'* non-basal ganglia tracts exist e.g. descending vestibulospinal and reticulospinal pathways
  - some disorders of abnormal movement are not associated with basal ganglia function (e.g. ataxia, myoclonus)

- Preferred term is that based on **clinical phenomenology** rather than anatomic location
- Coined by Stanley Fahn and Lewis P. Rowland in 1968.

Definition

Neurological syndromes in which there is

EITHER

an excess of movement

OR

a paucity of voluntary and automatic movements,

unrelated to weakness or spasticity

EPIDEMIOLOGY

Epidemiology: Global

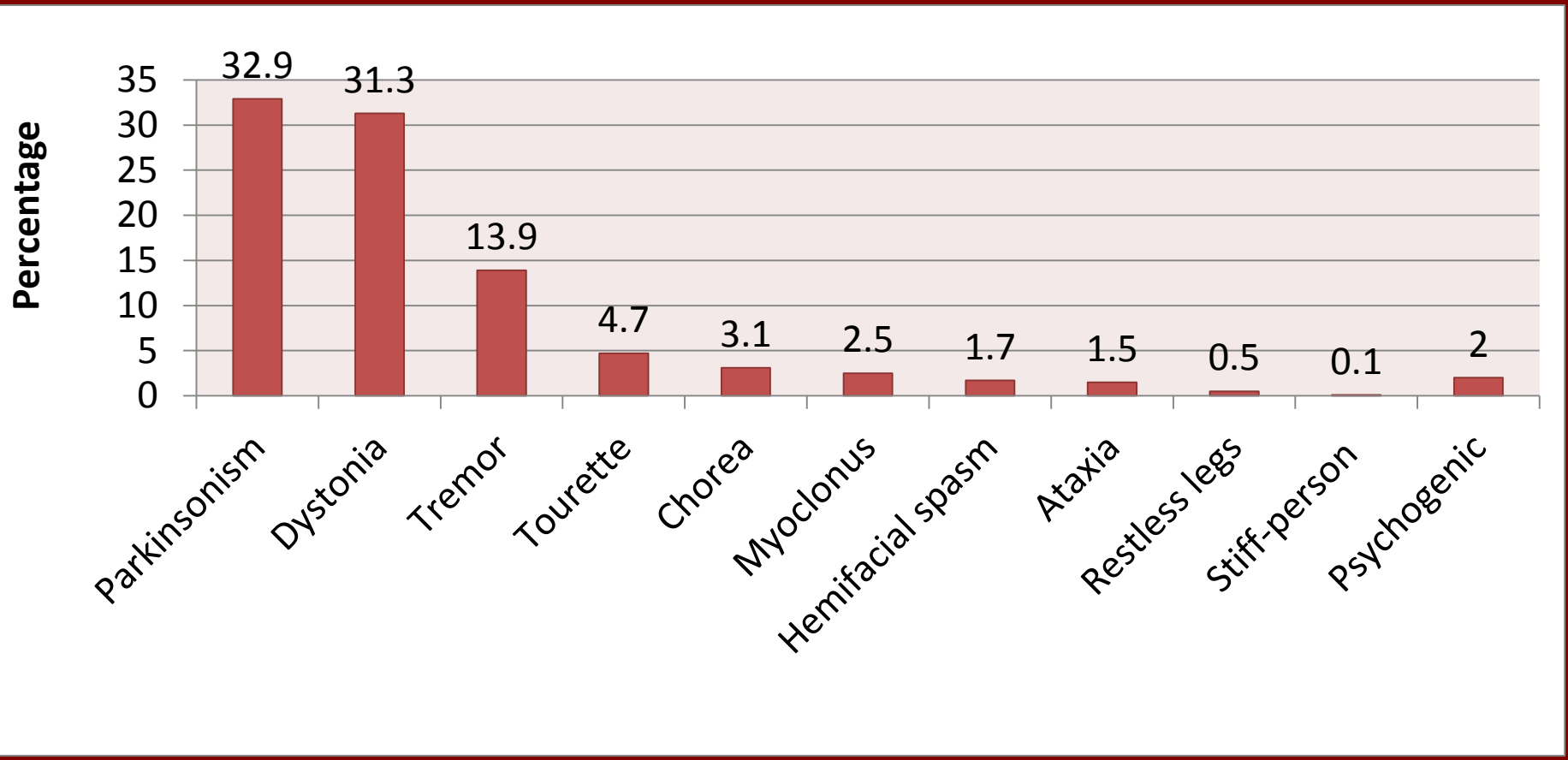
- Common neurological disorders
- Availability of population-based data for all movement disorders is hampered by the broad spectrum of conditions
- Most prevalence data refer to specific movement disorders
- Hospital-based data provide insight into more disabling MDs

Prevalence of movement disorders

Disorder	Prevalence rate per 100,000 population
RLS	9800 (age 65 – 83)
Essential tremor	415
Parkinson disease	187
Tourette syndrome	29 – 1052/2990
Primary torsion dystonia	33
Hemifacial spasm	7.4 – 14.5
Blepharospasm	13.3
Hereditary ataxia	6
Huntington disease	6
Wilson disease	3
PSP	2 – 6.4
MSA	4.4

Fahn S, Jankovic J. Principles and Practice of Movement Disorders, 2007: page 5

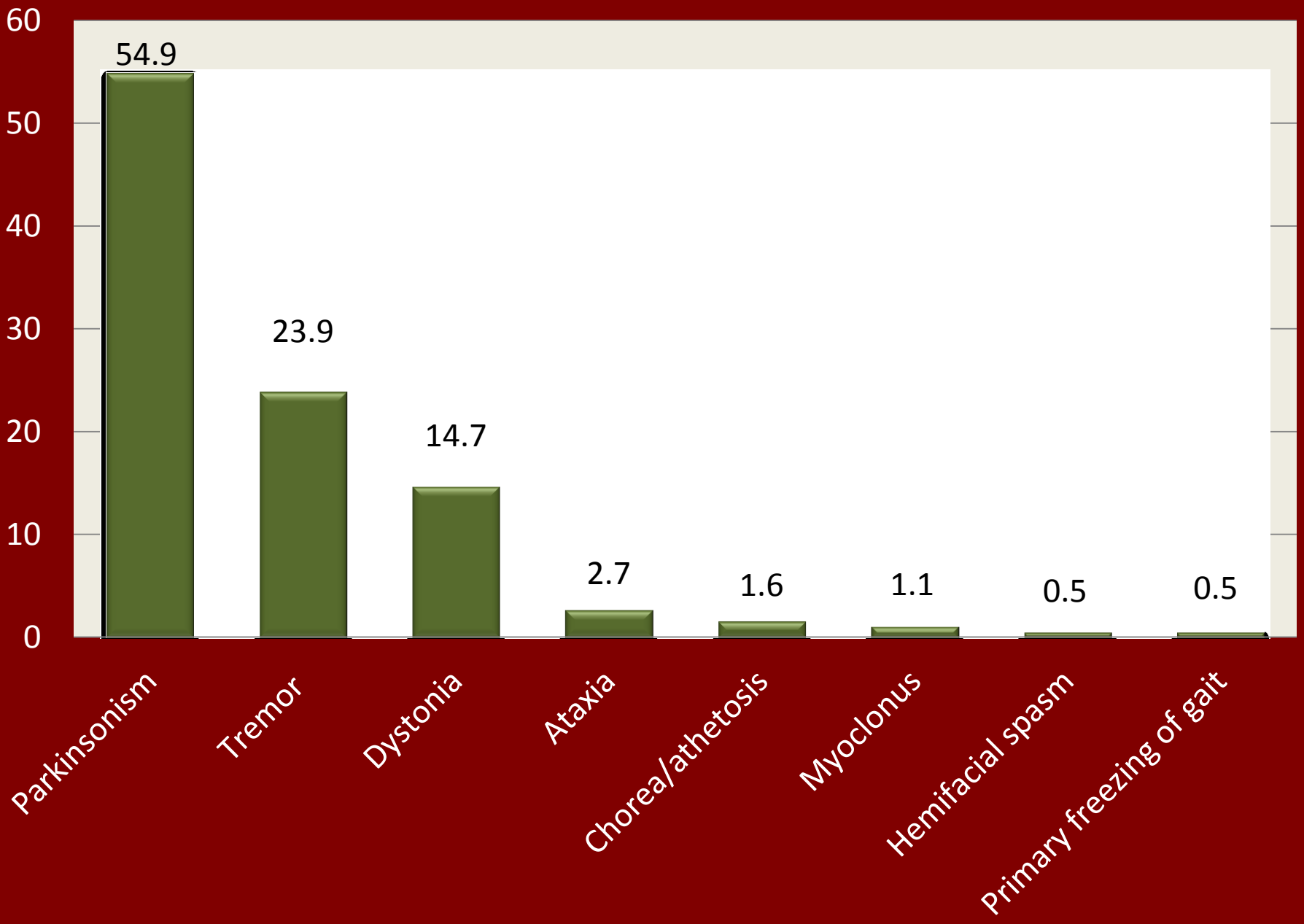
Hospital-based frequency of MD



Adapted from Fahn & Jancovic; data from 2 large MD clinics in USA 1996

Hospital-based frequency of MD: Africa

- ~5 – 15% of outpatient neurology consultations (*Bower JH et al, Ethiopia, 2005; Tegueu et al, Cameroon, 2013*)
- Hypokinesias – 55.4%; Hyperkinesias – 44.6% (*Okubadejo et al, 2012*)
- Most common – Parkinsonism/Parkinson disease
- **Predominant forms:** (similar to global MD clinic reports)
  - parkinsonism
  - other tremors
  - dystonia



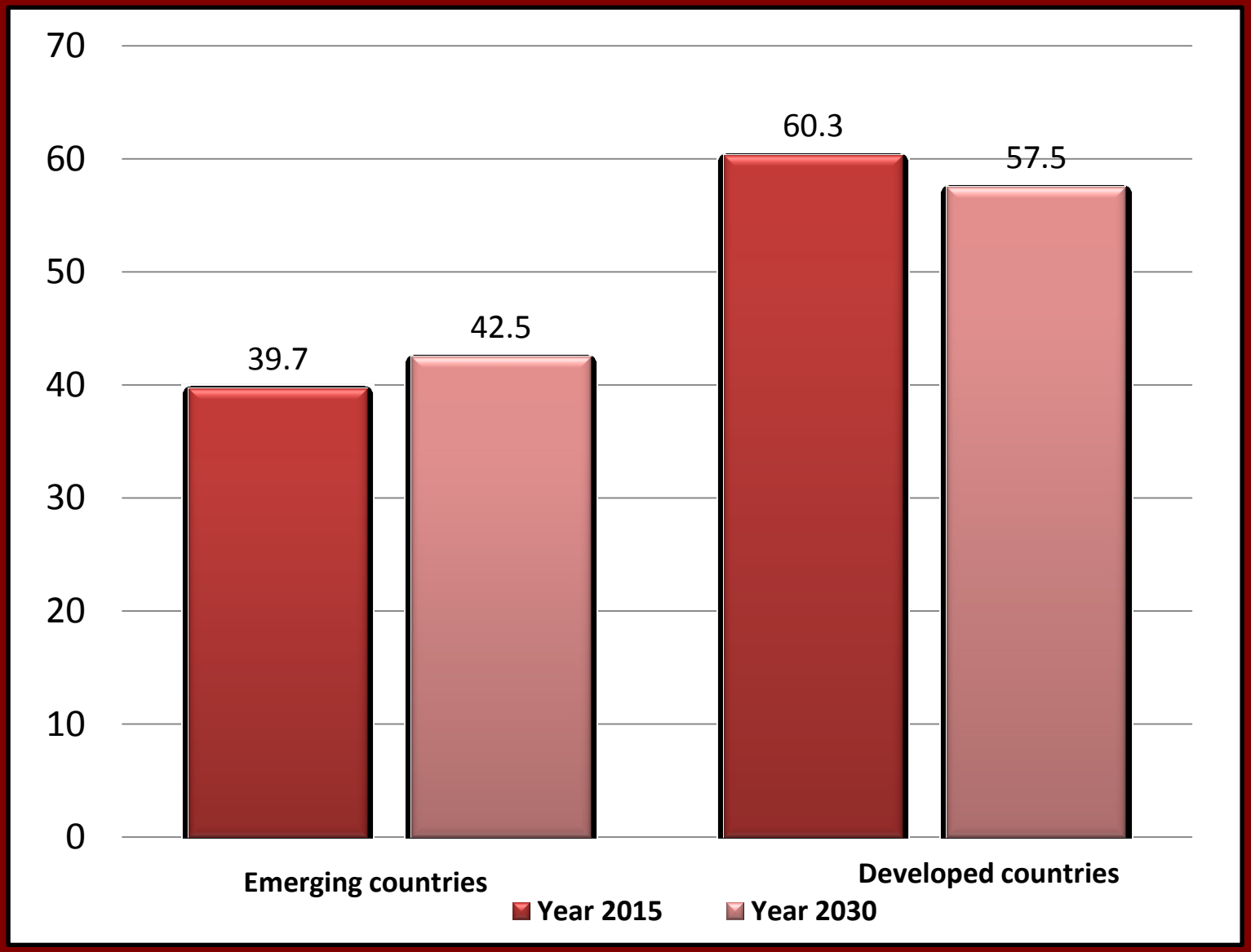
Okubadejo NU *et al*. Spectrum of movement disorders at the premier Lagos Movement Disorders Clinic in Nigeria: First year's experience(2012), Poster Presentations. Mov. Disord., 27: S1–S523. doi:10.1002/mds.25051

PARKINSON DISEASE

- One of the most frequent (top 10) neurological diseases encountered in primary care globally, including in Africa.
  - PD frequency in specialist care: 4th major neurological disease encountered in specialist care (46.2%) overall, 18.8% (6th in Africa) and 66.7% (4th in SE Asia).

- **Burden (population) of PD from 2015 to 2030**
  - Developing countries: 2.37 mill (39.7%) to 3.08 mill(42.5%)
  - Developed countries: 3.60 mill (60.3%) to 4.16 ml (57.5%)

Proportion (%) of PD in emerging and developed countries: 2015 to 2030.



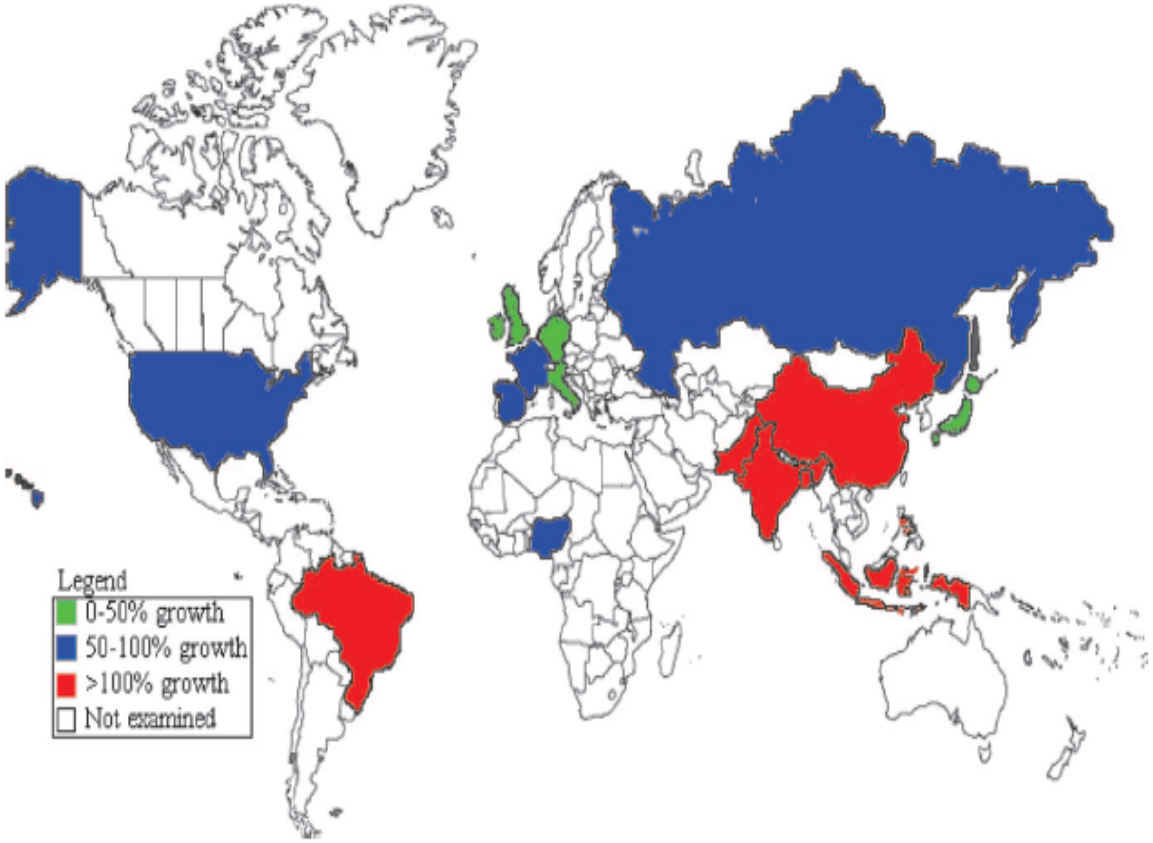


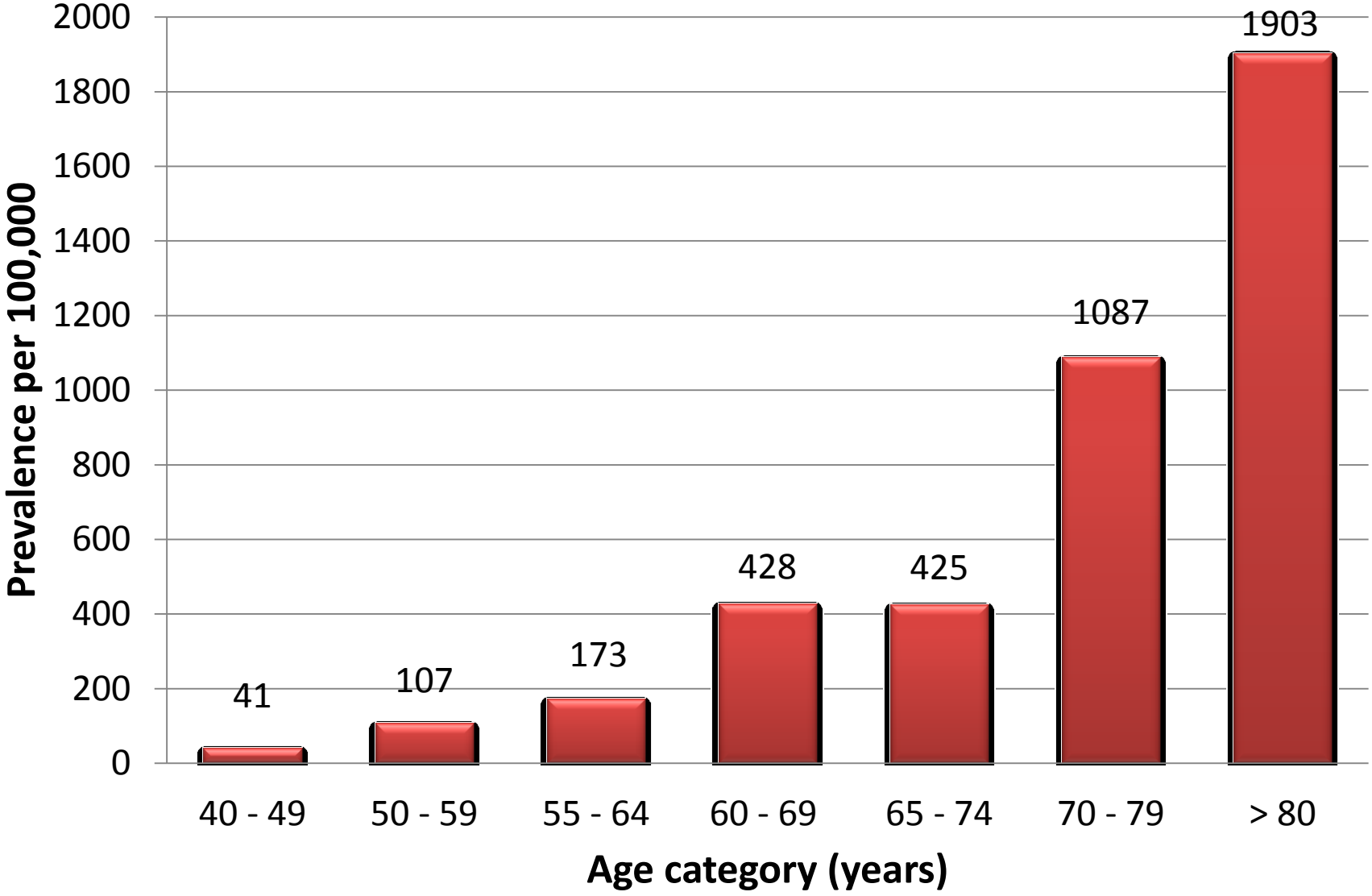
Figure 2. Projected growth rates in number of individuals over 50 with Parkinson disease in the most populous nations in Western Europe and the world from 2005 to 2030.

Dorsey ER, et al. Projected number of people with Parkinson disease in the most populous nations, 2005 through 2030. Neurology. 2007;68(5):384-6.

PD epidemiology ii

- Affects ~ 1% population 65 – 85 yrs; 4.3% >85 yrs
- Prevalence rates vary widely: *methodological, geographic, genetic, environmental differences*
- In metanalysis, steady increased prevalence with age
- Age-standardized DALY rates rank (Global Burden of Disease Study 2015) for all neurologic dx (14 categories)
  - Global: 11th
  - North, central SSA, western SSA: 11th
  - Eastern SSA: 12th
  - Southern SSA: 10th

Global prevalence of PD



Based on data from systematic review by Pringsheim T. *et a*l (2014). Movement Disorders. 29(13):1583 - 1590

Parkinson's Disease in Africa: A Systematic Review of Epidemiologic and Genetic Studies

Njideka U. Okubadejo, MD,[1,2] James H. Bower, MD,[1*] Walter A. Rocca, MD, MPH,[1,3] and Demetrius M. Maraganore, MD[1]

Movement Disorders. 2006;21(12):2150 - 2156

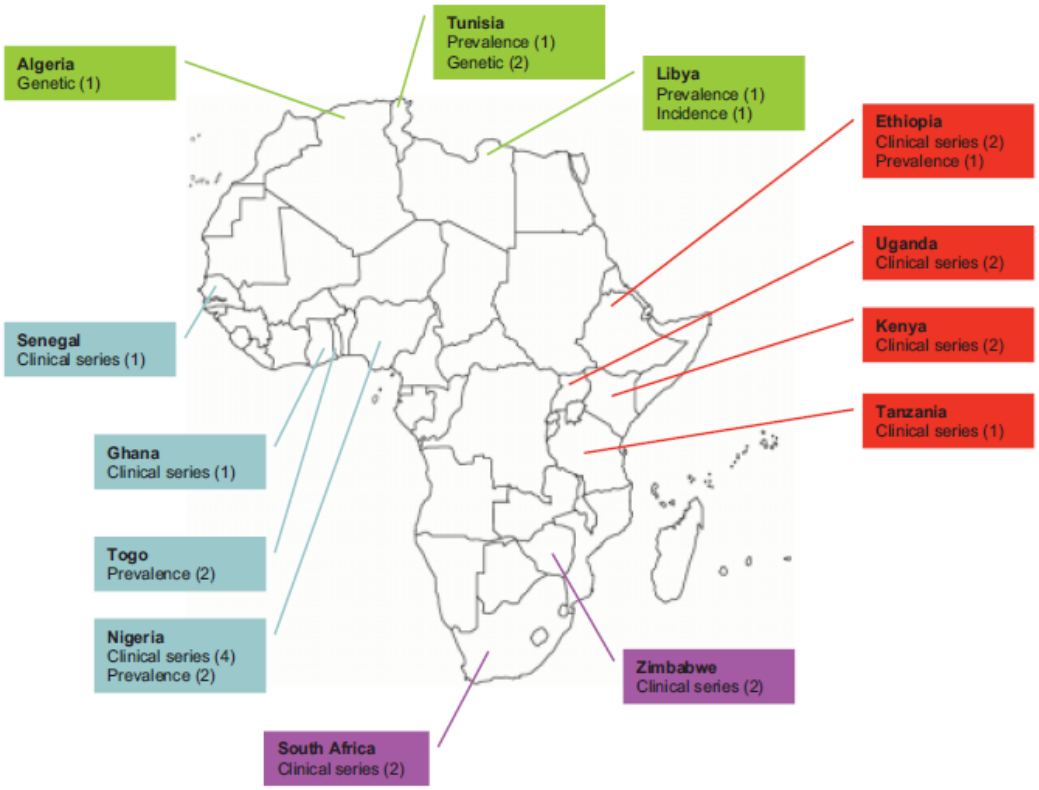


FIG. 1. Distribution of epidemiologic and genetic studies of Parkinson's disease in Africa by country. The number of published reports for each type of study (clinical series, prevalence, incidence, or genetic) is reported in parentheses. Red, East Africa; Blue, West Africa; Green, North Africa; Violet, South Africa. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

PD epidemiology iii: incidence

- Based on most recent systematic review and metanalysis (*Hirsch L, et al. Neuroepidemiologu 2016;46:292 – 300*)
- Overall incidence (females >40): 37.55 per 100,000 person-years
- Overall incidence (males > 40): 61.21 per 100,000 person-years
- Increased incidence with age in both sexes

BMC Neurology

BioMed Central

Research article

Open Access

Clinical profile of parkinsonism and Parkinson's disease in Lagos, Southwestern Nigeria

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* Corresponding author

BMC Neurology 2010;10:1 (doi:10.1186/1471-2377-10-1

- Descriptive study of General Neurology Clinic based PD cases: 1996 – 2006
- 124 parkinsonism – 98 (79.0% PD) and 26 (21.0% others)
- Mean time from onset to diagnosis: **24.6 months (median 12 mo)**
- Severity at presentation (Hoehn and Yahr stage 2)

PD: environmental risk factors

- Variable evidence for specific risk factors
- Rural living, agricultural occupation, well water consumption
- Pesticides – paraquat, rotenone, organochlorines, organophosphates, permethrin, benomyl
- Solvents
- Polychlorinated biphenyls
- Mild to moderate head injury
- Toxins: MPTP
- Behavioural risk factors (***reduced risk***): caffeine and cigarette smoking

PD: genetic factors

- **Monogenic causes of PD/parkinsonism**
  - Classic PD: PARK-*SNCA*, PARK-*LRRK2*, PARK-*VPS35* (all autosomal dominant)
  - Early onset PD: PARK-*parkin*, PARK-*PINK1*, PARK-*DJ1* (all autosomal recessive)
  - Parkinsonism: PARK-*ATP13A2*, PARK-*FBX07*, PARK-*DNAJC6*, PARK-*SYNJ1*

- **Genetic risk variants (GWAS):** > 26 genetic risk variants consistently associated

ESSENTIAL TREMOR

- Most common tremor
- Worldwide prevalence: 4.9 – 39.2 per 1000
- Highest prevalence >60: 13.0 – 50.5 per 1000

Okubadejo *et al. BMC Neurology* 2012, **12**:110
http://www.biomedcentral.com/1471-2377/12/110

BMC Neurology

RESEARCH ARTICLE Open Access

Prevalence of essential tremor in urban Lagos, Nigeria: a door-to-door community-based study

Njideka U Okubadejo[1*], Idowu A Bankole[2], Oluwadamilola O Ojo[1], Frank I Ojini[1] and Mustapha A Danesi[1]

- Prevalence in Lagos: 23.8 per 1000 (age-adjusted to WHO New world population)
- Consistent increase in prevalence with advancing age

CLASSIFICATION

Broad subtypes of MD

- Hypokinesias
- Hyperkinesias

Hypokinesias

1. **Parkinsonism**

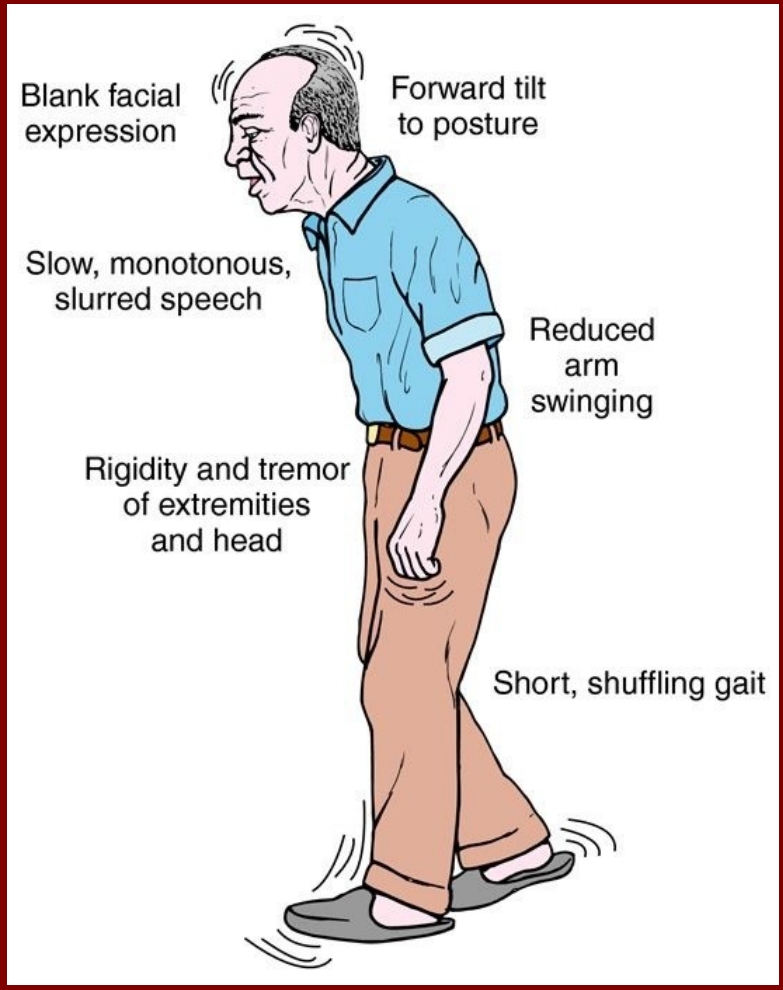
(Akinesia/bradykinesia)

2. Apraxia
3. Cataplexy and drop attacks
4. Freezing
5. Rigidity
6. Blocking (holding) tics
7. Catatonia, psychomotor depression, and obsessional slowness
8. Hesitant gaits
9. Hypothyroid slowness
10. Stiff muscles

Hyperkinesias

1. **Tremors**
2. **Dystonia**
3. **Chorea/Athetosis/ Ballism**
4. **Ataxia/dysmetria**
5. **Myoclonus**
6. **Hemifacial spasm**
7. **Tics**
8. Hyperekplexia
9. RBD
10. Myokymia and synkinesis
11. Restless legs
12. Moving toes/fingers
13. Paroxysmal dyskinesia
14. Abdominal dyskinesias
15. Akathisia
16. Hypnogenic dyskinesias
17. Jumpy stumps
18. Myorhythmia
19. Stereotypy

PARKINSONISM



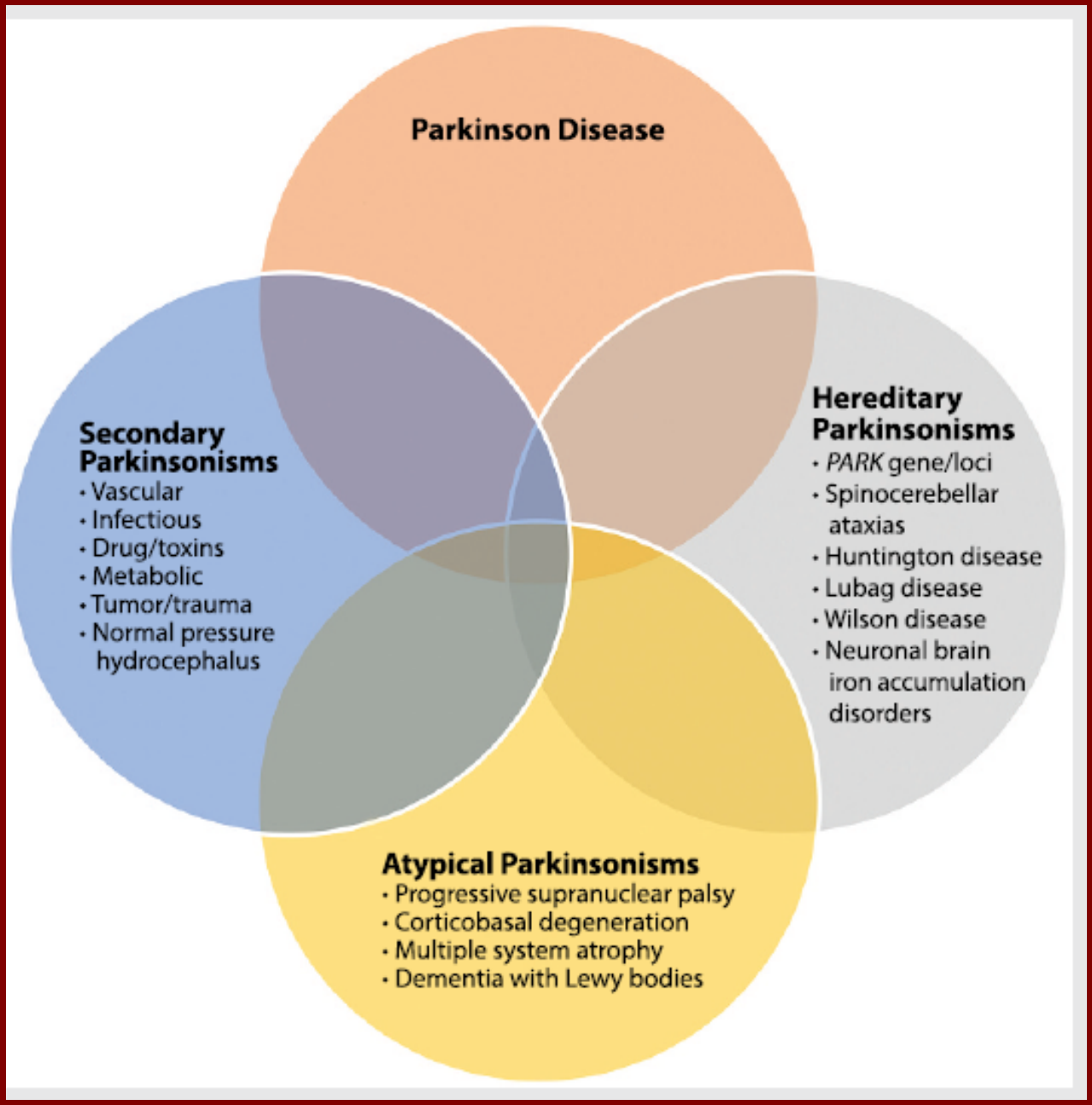
Parkinsonism: classification

I. **Primary (Parkinson disease, idiopathic PK)**

II. **Atypical parkinsonism:** *MSA, PSP, CBD, parkinsonism with dementia*

III. **Heredodegenerative disorders**

I. **Secondary:** *infections, toxins, drugs, tumor, trauma, vascular, metabolic*



Continuum. 2016;22(4):1117-1142

Classification of PD (motor subtypes)

i. Tremor dominant

ii. Postural impairment and gait disturbance (PIGD)

Multiple system atrophy (subtypes)

i. MSA - cerebellar

ii. MSA – parkinsonian

Progressive supranuclear palsy subtypes/phenotypes

- Classic PSP (Richardson disease)
- PSP – parkinsonism
- PSP – pure akinesia with freezing of gait
- PSP – corticobasal syndrome
- PSP – behavioural variant of frontotemporal dementia
- PSP – cerebellar ataxia
- PSP – primary lateral sclerosis

Corticobasal degeneration (CBD) subtypes/phenotypes

- Corticobasal syndrome (classic CBD)
- CBS – frontal behavioural variant
- CBS – posterior cortical atrophy syndrome
- CBS – progressive non-fluent /agrammatic aphasia
- CBS – PSP syndrome

DYSTONIA

Dystonia – Axis I: Clinical characteristics

- **Age at onset**
  - Infancy (birth – 2 years)
  - Childhood (3 – 12 years)
  - Adolescence (13 – 20 years)
  - Early adulthood (21 – 40 yrs)
  - Late adulthood (> 40 years)
- **Body distribution**
  - Focal
  - Segmental
  - Multifocal
  - Generalized (with or without leg involvement)
  - Hemidystonia
- **Temporal pattern**
  - Disease course: Static or Progressive
  - Variability: persistent, action-specific, diurnal, paroxysmal
- **Coexistence of other movement disorders**
  - Isolated dystonia ($\pm tremor$)
  - Combined (with another MD e.g. myoclonus, park)
- Other neurological manifestations (complex)
- Other systemic features

Movement Disorders, Vol. 28, No. 7, 2013

Dystonia – Axis II: Aetiology

- **Nervous system pathology**
  - Evidence of degeneration
  - Evidence of structural (often static) lesion
  - No evidence of degeneration or structural lesion

- **Inherited or acquired**
  - Inherited: AD, AR, X-linked recessive, Mitochondrial
  - Acquired: Perinatal brain injury, infection, drug, toxic, vascular, neoplastic, brai injury, psychogenic
  - Idiopathic: Sporadic, Familial

Movement Disorders, Vol. 28, No. 7, 2013

TREMOR

Tremor: classification by phenomenology

A) Rest tremor

B) Action tremor

i. Postural tremor

i. Position-independent postural tremor

ii. Position-dependent postural tremor

ii. Kinetic tremor

a) Simple (non – goal directed)

b) Target-directed (intention)

iii. Task-specific kinetic tremor

iv. Isometric tremor

Tremor: syndromic classification

a) Physiologic tremor
b) Enhanced physiologic tremor
c) Essential tremor
d) Dystonic tremor
e) Parkinsonian tremor
f) Cerebellar tremor

g) Holmes' tremor
h) Palatal tremor
i) Drug/ Toxin-induced
j) Tremor in periph. neuropathy
k) Psychogenic tremor
l) Unclassified

ATAXIAS

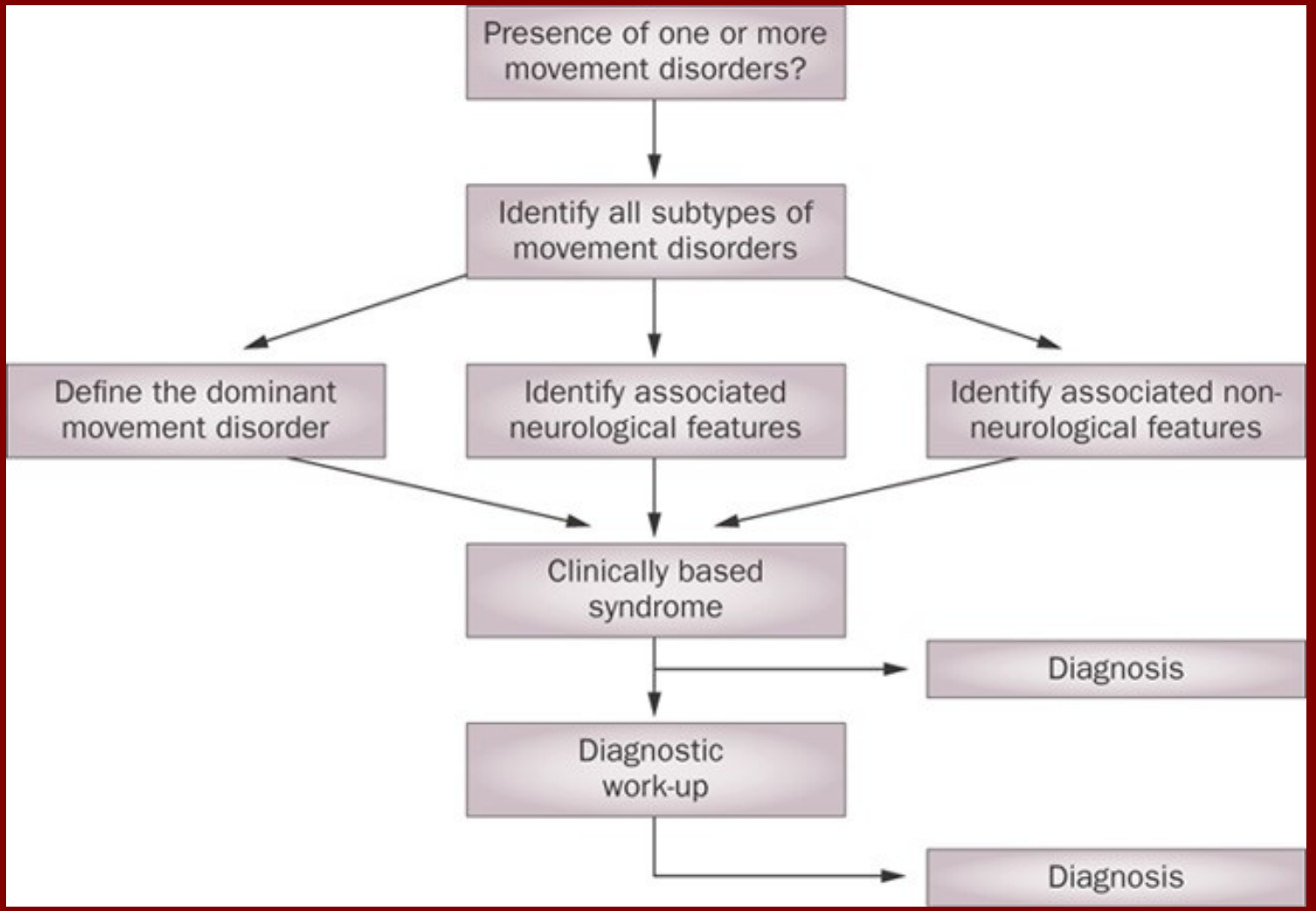
Classification

- Age of onset
- Time course: *acute, subacute, chronic, episodic*
- Anatomic involvement
- Distribution: *focal or generalized/symmetrica*l
- Cause: acquired or inherited or sporadic

Hereditary ataxias

- Autosomal dominant: *Spinocerebellar ataxias (SCAs), DRPLA, episodic ataxias 1 & 2*
- Autosomal recessive: e.g. *Friedreich ataxia, Ataxia telangiectasia*
- X-linked
- Mitochondrial e.g. *MERRF, MELAS*

Systematic Approach to Diagnosis of Movement Disorders



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