Journal of the Neurological Sciences, 1981 51: 311–327
Elsevier/North-Holland Biomedical Press

CLASSIFICATION OF EXTRAPYRAMIDAL DISORDERS

Currently used classifications of extrapyramidal disorders vary considerably and so do definitions of terminology. Certain authorities continue to dispute the distinction between extrapyramidal and pyramidal disease and the continuing debate reflects the controversial evolution of our understanding – still very far from complete – of the neurological diseases which affect the motor pathways and their underlying pathophysiology. For example, the anatomical demarcation and limits of the extrapyramidal system and it's diseases remains ill-defined and this gives rise to several practical difficulties in classification. Thus traditional clinical concepts of movement disorders – voluntary or involuntary – exclude spastic pareses and epileptic phenomena but include spinal, striatal and cerebellar origins. However, cerebellar syndromes are more readily classified in clinical and neuropathological terms than the extrapyramidal diseases. Where should the line be drawn? When clinical and neuropathological terminology has not been in accord it seemed preferable to give priority to clinical events. Thus, when certain cerebellar syndromes have been associated with involuntary movements these have been included under the rubric of extrapyramidal disease.

In recent years we have witnessed rapid advances in our knowledge of the neuropharmocology of the central nervous system with particular respect to neurotransmitter systems and these have proved to be particularly relevant to the functioning of the basal ganglia revealing opportunities for therapeutic development. These have stimulated worldwide and intensive research into the field of movement disorders which has involved neurologists, psychiatrists, neurosurgeons as well as those in the basic sciences of anatomy, pathology, neurobiology, physiology and pharmacology. During this phase of intense activity it seems particularly desirable to have a common frame of reference embracing the classification and terminology of extrapyramidal disorders which would be internationally acceptable and practical. With this aim in mind, Dr. Melvin D. Yahr, Chairman of the Research Committee on Extrapyramidal Disease of the World Federation of Neurology authorised the establishment of an ad hoc committee on nomenclature in November of 1977 with the following membership:

André Barbeau, M.D., F.R.C.P. C(C), F.R.S.C., Institut de Recherches Clinique de Montréal, Canada.

Roger C. Duvoisin, M.D., College of Medicine and Dentistry of New Jersey, U.S.A.

Prof. Dr. Franz Gerstenbrand, Neurologische Universitäts-Klinik, Innsbruck, Austria.

Dr. Johannes P. W. F. Lakke, Neurologische Kliniek, Academisch Ziekenhuis, Groningen, The Netherlands.

Prof. C. D. Marsden, Institute of Psychiatry, London, Great Britain.

0022-510X/81/0000-0000/$02.50

Gerald Stern, University College Hospital, London, Great Britain.

From the earliest discussions it was soon clear that classification should follow a pragmatic clinical approach and whenever possible we attempted to include the recommendations of recent authoritative reviews and contemporary opinions. There was much discussion and debate during the course of several meetings and subsequent correspondence, but it is chastening to concede that current concepts of basal ganglia disorders do not radically depart from the views of our predecessors at the turn of the century. We are aware that differences of opinion still exist within and beyond the committee and that some of these problems arose from our attempt to force complex and unresolved phenomena into unduly simple systems of classification. Nevertheless it proved possible to make preliminary proposals of classification, terminology and practical rating scales and these were reviewed by members of the Research Committee on Extrapyramidal Disease in Boston on 17 September, 1980. The present report includes the views and criticisms of members of the Research Committee. It attempts to place the known types of abnormal involuntary movements in classified form, to offer definitions of terminology that will be internationally acceptable and useful, systematically to group the disease entities known as extrapyramidal disorders and to recommend a set of rating scales which will facilitate the comparison of results from differing research centres. The publication of these proposals in advance of the 10th International Congress of the World Federation of Neurology in Kyoto, 1981, is intended to encourage improvements and constructive criticism. It would be particularly helpful if those with dissenting views would write directly to me before the Congress or have their proposals in a form suitable for projection during the satellite symposium on extrapyramidal diseases. The Committee's proposals will be open to general discussion and it is hoped that controversies will be satisfactorily resolved.

Johannes P. W. F. Lakke*
Chairman *ad hoc* Committee on Classification

* Address for correspondence: Dr. Johannes P. W. F. Lakke, Kliniek voor Neurologie, Academisch Ziekenhuis, Postbus 30.001, 9700 RB Groningen, The Netherlands.

The activities of the ad hoc committee would not have been possible without the very generous financial support of the following pharmaceutical industries and on behalf of the Committee members I wish to express our gratitude to: Ciba-Geigy B.V., Arnhem, The Netherlands; Ciba-Geigy Ltd., Basle, Switzerland; Hoffman-La Roche B.V., Mijdrecht, The Netherlands; Merck, Sharp & Dohme, B.V., Haarlem, The Netherlands; Sandoz Ltd., Basle, Switzerland.

CLASSIFICATION OF EXTRAPYRAMIDAL DISORDERS

Proposal for an International Classification and Glossary of Terms

A report by an ad hoc Committee established by the Research Group on Extrapyramidal Disorders of the World Federation of Neurology

INTRODUCTION

In November 1977 the Research Group on Extrapyramidal Disorders formed an ad hoc Committee under the chairmanship of Dr. Johannes P. W. F. Lakke to establish a standardized internationally acceptable glossary of terms and a clinical classification of disorders of the extrapyramidal system, and to suggest standard rating scales for abnormal involuntary movements and postures (AIMP).

In this pursuit the Committee's philosophy and approach stems from the systematic consideration of disorders of movement, posture and tone rather than a detailed consideration of extrapyramidal diseases. The Committee accepted entities only when there was unanimous agreement as to their existence or nosological identity. It was the intention to avoid eponyms, but this proved to be unavoidable. The members of the Committee recognized that all attempts at classification of extrapyramidal disorders are hampered by our limited knowledge of the basic pathological and etiological processess in the brain. Moreover, in extrapyramidal disorders the distinction between the various AIMP's is not always clear. Thus, the classification is only tentative and awaits further advance of knowledge. Yet, accepting these inadequacies, it is hoped that the Committee's effort has provided a classification to aid research and bring some measure of uniformity to the use of diagnostic terms.

In establishing this classification the Committee gave priority to symptoms and not to patho-physiology and distinguished between

primary – when the principal or predominant symptom under consideration was the initial manifestation of the intrinsic disease process, and *secondary* – when the symptom was a later manifestation of another disease or the consequence of an identifiable extrinsic process.

With respect to the rating scales, while the Committee is well aware that individual symptoms may vary considerably from time to time, it is recommended that only the situation at the time of assessment should be scored in contradistinction to retrospective evaluations. Rather than give a total score based upon summation of doubtfully weighed and relative components, we have preferred to offer separate profiles of mental, physical and social disability and of individual

0022-510X/81/0000-0000/$02.50

symptoms. We stress that these profiles should not be used for inter-patient comparison but for sequential analyses in a given patient. For each profile the actual examination technique is not specified but left to the discretion of the clinician. However, when a given profile is chosen, it should be used in its entirety.

The Committee recommends a choice of test batteries based, on the one hand, upon the differing needs of generalists, specialists and research workers, and on the other hand upon the necessity of obtaining the reasonable balance between the requirements of the regulatory agencies and the sometimes obsessive demands of drug companies. In the opinion of the members of the Committee these batteries represent a reasonably comprehensive method of assessment and we discourage, as a waste of effort and resources, more detailed or complicated requirements.

PART 1

CLASSIFICATION AND NOMENCLATURE OF TERMS USED IN THE CLINICAL DESCRIPTION OF EXTRAPYRAMIDAL DISORDERS

Accepting that movement is physiologically a change in posture and that posture is the position assumed at any moment by the various parts of the body acting in relation to each other and in relation to gravity, extrapyramidal symptoms can be clinically classified into 2 groups: (I) *disorders of movement*, and (II) *disorders of posture and tone*, whereby tone (tonus) is understood as the degree of resistance appreciated by an examiner during passive joint movements in a relaxed subject.

I. DISORDERS OF MOVEMENT

A. Hypokinesia – Akinesia

"Akinesia" is a disorder characterized by poverty and slowness of initiation and execution of willed and associated movements and difficulty in changing one motor pattern to another, in the absence of paralysis. This may include an inability to sustain repetitive movements and difficulty in performing simultaneous motor acts and may vary in severity from slight (sometimes called hypokinesia) to severe and complete immobility. The term bradykinesia should be reserved for slowness in the execution of movements.

1. Primary

(a) Akinetic form of Parkinson's disease

(b) As part of other degenerative disorders

(1) striato-nigral degeneration*

(2) Shy–Drager syndrome*

(3) olivo-ponto-cerebellar atrophy*

* Sometimes called Multiple System Atrophy (M.S.A.).

(4) progressive supranuclear palsy
(5) juvenile Huntington's chorea
(6) Westphal variant of Huntington's chorea
(7) Wilson's disease
(8) primary pallidal system atrophy
(9) Parkinson-dementia syndrome of Guam

2. Secondary

(a) Toxic
(1) drugs
(2) heavy metals (manganese, organic mercury)
(b) Traumatic
(1) apallic syndrome (sometimes called persistent vegetative state)
(c) Metabolic
(1) hypothyroidism
(2) Addison–Sholtz syndrome
(3) hepatic coma
(d) Infections
(1) postencephalitic parkinsonism
(e) Space-occupying lesions
(f) Diffuse brain atrophy

B. Hyperkinesia

B-1. Tremors

Tremor is a rhythmic, regular, oscillating movement of limbs, trunk, head or part thereof.

Physiological tremor: oscillations of limbs, at rest or in action under the influence of emotion, fatigue or physical exercise. The frequency varies throughout life and from one individual to another.

Rest tremor: this tremor is present in a distal part of the body at rest. It usually diminishes during voluntary movement. Its frequency varies between 4–8 c/s (the dominant tremor of Parkinson's disease).

Postural tremor: is most apparent when the muscles of a limb or trunk are activated to maintain a posture.

Intention tremor: appears during goal-directed movements and becomes more conspicuous approaching the target*.

"Rubral tremor": is a rotatory type of tremor of the head or hands accompanied by random movements of the fingers and often abnormal head posture. Often present at rest and made worse at movement.

"Wing beating tremor": is a proximal large amplitude, regular movement of the upper limbs best seen with the shoulder abducted and the elbows flexed. It occurs mainly in brainstem (dentato-rubral) lesions.

* Sometimes called incorrectly action tremor.

Flapping tremor: is seen in the outstretched hands and consists of a brisk extension of the wrist followed by a drop of the hand, giving it a jerking quality (asterixis). It occurs in metabolic and toxic disorders.

1. Primary

(a) Essential tremor
  (1) familial
  (2) senile
  (3) sporadic

(b) Cerebellar tremor (intention tremor) associated with degenerative disorders
  (1) olivo-ponto-cerebellar atrophy
  (2) other heredo-spino-cerebellar disorders

(c) Parkinsonian tremor
  (1) rest tremor of Parkinson's disease
  (2) postural tremor of Parkinson's disease

(d) Tremor associated with other degenerative disorders
  (1) hepato-lenticular degeneration (Wilson's disease)
  (2) heredo-spino-cerebellar disorders
  (3) dystonia musculorum deformans
  (4) Charcot–Marie–Tooth syndrome
  (5) Déjerine–Sottas syndrome

2. Secondary

(a) Metabolic disorders
  (1) hepatic (asterixis)
  (2) uraemic
  (3) hyperthyroidism
  (4) toxic

(b) Toxic disorders
  (1) lithium
  (2) heavy metals
  (3) alcoholic
  (4) beta-adrenergic agonists

B-2. Tics

Tics are sudden, stereotyped, complex, repetitive, normally coordinated but inappropriate movements, which can only be suppressed for a short period by an effort of will at the expense of increasing emotional tension. Tics may be single such as sniffing, vocalization, finger movements, or complex such as combinations of sniffing and grunting and generalized where complex tics are accompanied by impulsive, emotional behaviour.

1. Primary

(a) Elementary tics
  (1) mimic tics
  (2) blinking tics
  (3) nose tics, including sniffing

(4) oral tics, including tooth grinding tic, swallowing tic and regurgitation tic
(5) respiratory tics
(6) vocal tics, such as snouting, grunting, barking, swearing
(7) stuttering tic
(8) neck tics
(9) shrugging tics
(10) trunk tics
(11) leg tics

(b) Complex tics
(1) fixed combinations of (a) such as mimic and sniffing and jumping

(c) Generalized tics
(1) without vocal tics
(2) Gilles de la Tourette disease

2. Secondary

(a) Toxic
(1) drugs
(2) intoxications like "Myriachit" of Siberia

(b) Infection
(1) encephalitic
(2) postencephalitic

B-3. Myoclonus

Myoclonus is characterized by sudden shock-like contractions of a muscle or a group of muscles. This may or may not be accompanied by joint movement and may occur at rest or during action and may be isolated, repetitive or rhythmic, as in palatal or diaphragmatic myoclonus.

1. Primary

(a) Hereditary conditions
(1) Ramsay Hunt's dyssynergia cerebellaris myoclonica
(2) Unverricht–Lundborg's myoclonus epilepsy
(3) Friedreich's paramyoclonus multiplex
(4) inborn errors like lysosomal enzymopathies
(5) Startle syndromes (hyperekplexia)
(6) Jumping Frenchmen of Maine

(b) Non-hereditary syndromes
(1) so-called essential myoclonus
(2) myorhythmia including palatal myoclonus
(3) opsoclonus

2. Secondary

(a) To the epilepsies
(1) so-called myoclonic epilepsy
(2) epilepsia partialis continua of Koshevnikov

(b) To infections
    (1) virus encephalopathy
    (2) subacute sclerosing panencephalitis
    (3) Creutzfeld–Jakob disease
(c) To metabolic disorders
    (1) anoxic (Lance–Adam syndrome)
    (2) hepatic
    (3) uraemic
    (4) intoxications
(d) To spinal cord lesions (spinal myoclonias)

B-4. Chorea

Chorea is a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. It may vary in severity from restlessness with mild intermittent exaggeration of gesture and expression, fidgeting movements of the hands, unstable dance like gait to a continuous flow of disabling, violent movements.

1. Primary

(a) Hereditary dominant
    (1) Huntington's chorea
    (2) hereditary non-progressive chorea of early onset
    (3) chronic chorea of late onset
(b) Hereditary sex-linked
    (1) Lesch–Nyhan syndrome
(c) In association with other degenerative or metabolic disorders
    (1) Canevan spongy degeneration
    (2) hepato-lenticular degeneration (Wilson's disease)

2. Secondary

(a) To infectious disease
    (1) virus encephalitis (measles and mumps)
    (2) neurosyphilis
    (3) pertussis
    (4) Sydenham's chorea (plus chorea gravidarum)
(b) To space-occupying lesions
    (1) meningioma, angioma, etc.
(c) To focal or multi-focal lesions (vascular and traumatic)
    (1) polycythaemia vera
    (2) vascular hemi-chorea
    (3) cerebral concussion
(d) To auto-immune disorders
    (1) systemic lupus erythematosus
    (2) ataxia telangiectasia (Louis–Barr)
(e) To metabolic disorders
    (1) hyperthyroidism
    (2) hypocalcaemia

(3) angiokeratoma corporis diffusum (Fabry's disease)

(f) To intoxications

(1) phenothiazines

(2) mercury poisoning

(3) oral contraceptives

(4) lithium

(5) levodopa and agonists

B-5. Ballism

When involuntary movements of the limbs are unilateral, proximal, violent and flinging in character, they are described as hemiballistic; some consider this to be a variety of chorea. The usual localisation of the lesion is in or near the subthalamic nucleus.

1. Primary

(a) Hereditary bilateral ballism

2. Secondary (usually hemiballism)

(a) Space-occupying lesions

(b) Trauma

(c) Vascular (thrombotic and embolic)

(d) Infection

(e) Intoxication

B-6. Athetosis

Athetosis is characterized by irregular, slow, forceful, writhing movements generally of the extremities, very often characterized by finger movements.

1. Primary

(1) status dysmyelinisatus (Vogt)

(2) Pelizaeus–Merzbacher disease

(3) familial paroxysmal choreo-athetosis (Mount Reback)

2. Secondary

(a) Perinatal ischaemic disorders as status marmoratus, athétose double

(b) Toxic

(1) carbon dioxide poisoning

(2) carbon disulfide

(3) drugs (phenothiazine)

(c) Metabolic

(1) kernicterus

(2) ataxia telangiectasia

(3) Lesch–Nyhan syndrome

(d) To focal lesions (vascular and traumatic)

(1) anterior thalamic syndrome

(2) Benedikt–Nothnagel syndrome

(e) To severe proprioceptive defects (pseudo-athetosis)

B-7. Akathisia

Abnormal compulsion to move, of which the patient is painfully aware,

parts of the limb or the whole body, in itself not an abnormal movement, sometimes stereotyped.

1. Primary

(1) Parkinsonism

2. Secondary

(1) levodopa and agonists

II. DISORDERS OF POSTURE AND TONE

A. Dystonia

Dystonic states are abnormal postures produced by slow sustained muscle contractions which distort limbs, trunk, neck, face or mouth into characteristic postures; they may be distal (athetoid), proximal (torsion spasm) generalized, unilateral or focal.

Typical dystonic postures include the hyperpronated forearm with flexed wrist and extended fingers, the inverted plantar-flexed foot, the trunk curved to one side (scoliosis) or backwards (lordosis), the neck rotated to one side (torticollis), forward (antecollis) or backwards (retrocollis).

1. Primary

(a) Hereditary torsion dystonias

(1) autosomal dominant torsion dystonia
(2) autosomal recessive torsion dystonia
(3) X-linked recessive torsion dystonia
(4) dystonia with marked diurnal variation
(5) paroxysmal kinesiogenic dystonia (or choreo-athetosis)
(6) paroxysmal dystonic choreo-athetosis

(b) Associated with other hereditary neurological syndromes

(1) hepato-lenticular degeneration (Wilson's disease)
(2) Huntington's chorea
(3) Hallervorden–Spatz disease
(4) primary pallidal system atrophy
(5) olivo-ponto-cerebellar atrophy
(6) supranuclear palsy
(7) ataxia telangiectasia (Louis–Barr)
(8) Lesch–Nyhan syndrome
(9) Pelizaeus–Merzbacher disease

(c) Sporadic

(many of these cases may turn out to be isolated examples of group (a). Many are also focal dystonias

(1) focal, axial or segmental dystonias which may affect:

eyes : (blepharospasm)*

---

* Sometimes called Meige's or Breugel's syndrome.

mouth, jaw : (oro-mandibular dystonia)
neck : (torticollis, retrocollis, anterocollis)
trunk : ("peacock trunk")
abdominal : ("belly dancer's dystonia")
arm : (dystonic writer's cramp, crampe des écrivains)
leg : ("ballet-dancer's foot")
phonation : (vocal spasmodic dystonia)

2. Secondary

(a) Symptomatic of brain damage due to
(1) perinatal cerebral lesion (status marmoratus)
(i) kernicterus
(2) encephalitis, e.g. encephalitis lethargica
(3) brain trauma
(4) focal cerebrovascular lesion
(5) brain tumor
(6) intoxication, e.g. manganese, organic mercury
(7) vascular malformations
(b) Drug-induced dystonias (generalized, localized, complex)

B. Torsion spasm

Torsion spasms are characterized by slow, forceful, twisting movements at the hip, shoulder, trunk and neck and are proximal manifestations of dystonia.

C. Cogwheel phenomenon

The cogwheel phenomenon is characterized by regular repetitive interrupting resistance to passive movements.

D. Hypertonia

D-1. Rigidity

Rigidity is a form of hypertonia characterized by a constant uniform increase in resistance to passive movement, throughout the range of joint displacement, while the patient attempts to relax. This should be distinguished from spasticity. This is a form of hypertonia characterized by an initial increase in resistance to active and passive movements followed by a decrease in tone in the same muscles occurring immediately (clasp-knife phenomenon) except in marked spasticity when repetitive movements may be required.

1. Primary

(1) in Parkinson's disease
(2) rigid form of Huntington's chorea (Westphal variant)
(3) rigid form of hepatolenticular degeneration (Wilson's disease)
(4) Parkinson-dementia syndrome of Guam
(5) supranuclear palsy with nuchal rigidity

2. Secondary

(1) cerebellar atrophy
(2) senile encephalopathy
(3) "normotensive" hydrocephalus
(4) chronic intoxication
(5) metabolic disorders
  (i) Fahr's syndrome
  (ii) hepatic encephalopathy
  (iii) storage disease

3. Other forms of rigidity

(1) stiff man syndrome
(2) rigid spine syndrome
(3) progressive encephalomyelitis with rigidity
(4) continuous muscle activity (Isaac's syndrome)

D-2. Gegenhalten

Reactive hypertonia elicited by passive movement of the limb, causing increased resistance in the limb in parallel with the intensity of the stimulus (so-called Gegenhalten and rigidity are oppositional).

E. Hypotonia

Hypotonia is a decrease in tone such as found in lower motor neuron damage, in cerebellar or in choreic disorders.

Note: Many of the above phenomena may occur in combination spontaneously or iatrogenically, to form clinically defined syndromes. Some phenomena can change during maturation in others like e.g. athetosis of an infant in dystonia of a pre-school child.

PART 2

RATING SCALES FOR MOVEMENT DISORDERS

INTRODUCTION

Rating scales for movement disorders pose a difficult problem, because there is often a marked oscillation in performance, depending upon, amongst other factors, the time of day, the attitude of the experimenter, test circumstances, time-dose medication dependence and psychological factors.

Moreover, all scales were unweighted, e.g., not all items have equal value and thus the total sum does not accurately represent individual disabilities. Accepting the inherent inadequacies of "objective" testing, the following recommendations are proposed:

(1) Tests should only be used for comparing a patient with himself; between patients comparisons are not advised.

(2) Rating scales should be simple, short, easily applicable, yet sufficiently flexible to be used for simple rating purposes as well as for more sophisticated research projects.

(3) That proposed test batteries should be appropriate and sufficient for drug research and that more elaborate test procedures would be unnecessary.

(4) International acceptance of identical test procedures markedly increase the value of these tests and makes research more easily comparable.

(5) As far as possible, rating scales should be limited to 4 points whereby 0 is normal.

(6) The possibilities of doctor-rating, nurse-rating and self-rating are recognized.

From a wide variety of possibilities the Committee selected and recommended the following scales:

(1) Disability profiles: including mental, physical and social state, to be considered as a baseline evaluation for the adequate interpretation of the other tests.

(2) Movement profiles: including abnormal involuntary movements and postures (AIMP).

(3) A repertoire of basic objectives, mainly as a tool for objective evaluation of the interference by AIMP of circumscribed motor tasks.

(4) Staging: in order to indicate the severity of a patient group, the Committee recommends for Parkinsonism, Hoehn and Yahr's system and for Huntington's chorea the system of Shoulson and Fahn. It should be recognized that staging is only feasible for motor disorders which are progressive, therefore, as an example, torticollis can be excluded. The Parkinson scale seems also applicable for Wilson's disease, dystonia, etc. If one of the staging systems cannot be applied, then a simple global scale is recommended, using three stages: slight, moderate and severe.

The Committee is aware of the empiricism and limitations of the above tests, and emphasize the great importance of accepting certain standards for comparison purposes.

I. DISABILITY PROFILE

Disability should be considered in relation to the patient's surroundings and can be considered under three aspects: mental, physical and social. The state of disability is a prerequisite in the proper evaluation of other tests, therefore certain minimal features of the patient should be known. It should be reemphasized that the following profiles are non-parametric, non-comparable and should not be summated.

A. Mental state

1. 4 Educational levels: low grade – high school – college – university
2. Emotional state: normal – labile – depressed – manic or euphoric
3. Vigilance: normal – diminished
4. Orientation: normal – abnormal (confused)
5. Memory: normal – decreased
6. Other abnormal cognitive function:...
7. Psychiatric manifestations: present – absent

B. Physical state

1. Overall degree of physical independence for daily living: totally independent — partially independent — totally dependent.
2. For a more precise evaluation, items of the North West Disability Scale can be used in a 4-point scale whereby 0 = normal.
   - (a) Walking
     - (i) Unsupported in the house, outside the house partially independent, help of another person
     - (ii) In the house partially aided by others, outside mechanical or personal support
     - (iii) Does not walk
   - (b) Dressing
     - (i) Takes more time
     - (ii) Costs considerable time
     - (iii) To be dressed by others
   - (c) Eating
     - (i) Eats slower than usual, but uses fork and spoon
     - (ii) Eating takes considerable time, food must be specially prepared
     - (iii) Be fed by others
   - (d) Hygiene
     - (i) Washes independently, but uses more time
     - (ii) Needs help for specific hygienic actions
     - (iii) Must be washed
   - (e) Speech
     - (i) Some impairment, no melody, usually understandable
     - (ii) Decrease in volume, only partially understandable
     - (iii) Can not or only minimally be understood

C. Social state

It is recognized that assessment of social impairment depends on many factors such as cultural differences, employment and morale. Employability depends not only on working capacity but also on drive, motivation and social circumstances. Assessment of social activity should take cognizance not only of the ability to cope at work but also make due allowance for household and normal retirement

occupations; evaluation of social impairment should reflect significant individual differences between actual and anticipated circumstances.

1. Description of the normal previous and prevailing social employment situation.
2. Degree of activity
   (a) full-time at previous level (normal)
   (b) part-time at previous level
   (c) full-time at decreased level
   (d) part-time at decreased level
   (e) unemployed

II. MOVEMENT PROFILES

Abnormalities in movement are difficult to assess accurately because of great diurnal variability and distribution. When feasible, cinéphotography and videotape can be used, but for the overall assessment of hyperkinesias a simple 4-point scale is advised: normal – slight – moderate – severe.

A. Hypokinesia

1. Rigidity, to be judged separately in the 4 limbs, head and trunk

0 – absent, normal tonus; 1 – slight (synkinetic manoeuvres as Noïka–Froment or contralateral activation can be employed); 2 – moderate, resistance is unequivocally felt; 3 – severe

2. Akinesia, to be considered for left and right separately

0 – absent, normal social behaviour; 1 – slight – bradykinesia, poverty and slowness of movement; 2 – moderate – hypokinesia, amplitude of movement decreases; 3 – severe – akinesia, frozen attitude

B. Hyperkinesia

1. Tremor. To be rated separately the head and part thereof (chin and/or tongue are included in the heading head) and the 4 limbs

0 – absent; 1 – slight or occasionally present, such as upon contralateral activation; 2 – moderate or continually present but decreases upon willed movements; 3 – severe, marked amplitude, or not to be influenced

2. Various motor patterns.

Hyperkinesia. Whatever hyperkinetic movement is concerned, distinguish if possible between unilateral and bilateral and state which limbs should be rated. 0 – absent; 1 – slight or infrequent; 2– moderate or occasional; 3 – severe and/or frequent.

Dyskinesia. Instead of judging frequency and/or amplitude as in hyperkinesia, in dyskinesias, degree of incapacity should be evaluated (only for general simple use).

0 – normal; 1 – slightly incapacitated; 2 – moderately incapacitated; 3 – severily incapacitated

III. REPERTOIRE OF OBJECTIVE TESTS

These should be limited to an assessment of objective motor disabilities, e.g., interference of the movement abnormalities with the required motor performance, two more complex tests pertaining to visual spatial organisation and an assessment of intellectual capacities.

A. Tapping test

Based on the execution of alternating movements over a joint, correctly using agonist and antagonist muscles. Tapping with finger, wrist elbow and shoulder (left and right), then foot – ankle, knee, hip. This makes up for 14 tests, usually executed in 30 seconds each. Number of tappings is counted (usually with mechanical counting device).

B. Time-dotting test (Gerstenbrand, Grünbaum) [*Nervenarzt*, 44 (1973) 428–433].

The number of dots, placed in small squares within a certain time-limit (30 s) for the left and right hand separately.

C. Koh's block design test

Cultural independent testing visual spatial organisation.

D. 10-Piece puzzle completion test

Gives indication for global mental performance and mental akinesia. Number of correct pieces after the first, second and third minute are noted.

E. Selected as single motor tests, easily placed in records:

Archimedian spiral, triangle (for ataxia) } both hands

word: minimum

F. Walking test: time and number of steps during 6 meter distance (or 10 feet) are measured.

G. Self-rating 100 mm test: Patient determines his position (left bad, right good) upon a 100-mm line in accordance with his mood, motor handicap, etc. This to be chosen by the experimenter.

IV. STAGING OF PATIENT GROUPS

A. Parkinsonism (Hoehn and Yahr)

Stage I. Unilateral involvement only, usually with minimal or no functional impairment.

Stage II. Bilateral or midline involvement, without impairment of balance.

Stage III. First sign of impaired righting reflexes. This is evident in unsteadiness as the patient turns or is demonstrated when he is pushed from standing equilibrium with the feet together and eyes closed. Functionally the patient is somewhat restricted in his activities but may have some work potential depending upon the type of employment. Patients are physically capable of leading independent lives, and their disability is mild to moderate.

Stage IV. Fully developed, severely disabling disease; the patient is still able to walk and stand unassisted but is markedly incapacitated.

Stage V. Confinement to bed or wheelchair unless aided.

B. Huntington's disease (Shoulson and Fahn)

Stage I refers to a diagnosed HD patient who shows no functional deficits in regard to activities of daily living (i.e., eating, dressing, bathing), family interactions or employability. Despite signs and/or symptoms referable to HD, this individual remains employable in his present occupation, whether it be a professional job, unskilled labor or household responsibilities. Furthermore, this patient continues to be a fully functioning member of the family unit and is independent in managing financial affairs.

Stage II designates those HD patients who remain employable, but at a lower level of capacity. Such individuals are able to perform occupational or household roles, but not necessarily in the manner to which they have been accustomed. Despite some difficulty, these affected patients retain the capacity to manage daily financial affairs such as shopping and short-term budgeting. In addition, patients in the early intermediate phase continue to be completely independent in activities of daily living.

Stage III is applied to patients who are no longer employable in any capacity or can no longer manage household responsibilities. Patients may be able to handle daily financial affairs, but require a considerable degree of assistance or supervision. Activities of daily living may be marginally impaired but these individuals usually require only minimal assistance in such functions.

Stage IV indicates that those HD patients who are no longer independent in activities of daily living but are capable of being supported by the family within the home environment or by minimal professional assistance in an extended care facility.

Stage V characterizes those patients who can no longer function independently, usually necessitating professional health assistance. In this stage, patients require complete support in activities of daily living.

Journal of the Neurological Sciences, 1981, 51: 328
Elsevier/North-Holland Biomedical Press

News and Notes

14. DONAU-SYMPOSIUM FÜR NEUROLOGISCHE WISSENSCHAFTEN
(Vienna, Austria, 29–31 October, 1981)

The Symposium will be held under the auspices of the World Federation of Neurology and the Research Group on Neuropathology of the WFN.

Program: (1) The occipital lobe and the visual system; (2) Petit mal and its problems; (3) Free lectures.

Information can be obtained from: Sekretariat des Donau-Symposiums für Neurologische Wissenschaften, c/o Wiener Medizinische Akademia, Alser Strasse 4, A-1090 Vienna, Austria, Tel. (0222) 427165, Cables: Medacad Wien.

INTERNATIONAL MEETING ON "THE IMMUNOLOGY OF NERVOUS SYSTEM INFECTIONS"
(Medical Society of London, London, Great Britain, 12–13 November, 1981)

Invited speakers include over 40 international authorities. The meeting will provide a comprehensive overview of modern aspects, including therapy and directions of future research.

The contents will be published as an edited volume in the series *Progress in Brain Research* by Elsevier/North-Holland Biomedical Press.

For further information contact Dr. P.O. Behan, Institute of Neurological Sciences, Southern General Hospital, 1345 Govan Road, Glasgow G51 4TF, Scotland, U.K.