

Neuromotor Diseases and Hospice

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Focus

- Our goal in hospice is to incorporate a comprehensive palliative approach to pts with advanced Neuromotor disorders, especially PD (and its correlates), and others with Neurodegenerative Dz
- Recognize that end of life care and late palliative care calls forth our skills as clinicians with a whole-person perspective, and that attention to details is, in part, what helps to define us. We are, with these patients, dealing with persons, and their loved ones, who have struggled to stave off progressive losses in function and meaning, and are facing the difficult transition of embracing palliation and hospice.

QUESTION

QUESTION 1.

Any Neurologists, Neurosurgeons,
Neuro Nurses, Neuro Social Workers, or
Neuro Case Managers in this talk?

a. Yes b. No

Objectives

1. Review the classes of the Neurodegenerative Disorders, paying particular attention to the group now commonly accepted in classification as the Movement Disorders.
2. Compare and contrast the late clinical manifestations of various Movement Disorders.
3. Focus on the particularities that predispose disorders within this group to admission to hospice.
4. Identify palliative care skills, interventions, and principles geared to improve symptom management, coping, injury prevention, and care planning.
5. Provide some resources for further application in caring for this group of patients.

Question

QUESTION #3

If the average age of onset in PD is 63,
what percentage of PD patients present
with YOUNG ONSET PARKINSON'S?

a. 5%

b. 8%

c. 10%

d. 15%

Question

QUESTION #4

With newer treatment modalities available for treating Parkinson's Dz., how far have we impacted longevity?

a. 1 ½ yrs. b. 2 ½ yrs. c. 5 yrs. d. none of above

Introduction

- Progressive neurodegenerative disorders are common, partially understood, and poorly addressed in Palliative Care. Profound effect on Life Quality & Function. NO Curative Options.
- **Prevalence progresses markedly w age**
 - 1-2% >65
 - 2.6% >85
 - 50%+ >85 w Parkinsonism
- Over 1 million US citizens
- PD is most common Neuromotor dz.
- 5.2% LTC
- PD life expectancy slightly < avg. population
- Life expectancy greatly reduced other Neurodegenerative Dz's

Question

QUESTION #2

Parkinson's "Plus" Syndromes, or the Multi System Degenerations, are easily identified and treated, and typically respond well to Parkinson's Rx's?

a. True

b. False

Movement Disorders

Atypical Parkinson Syndromes

- Essential tremor-[most common Movement Disorder]
- Drug-Induced Parkinsonism
 - Neuroleptics, Meperidine, DA antagonists:
Haloperidol, metaclopramide,
 - DA depletors: reserpine, tetrabenazine
 - Others reported
- Vascular Parkinsonism
- Multiple BG infarcts
- Progressive Supranuclear Palsy
- Cortico-Basal Ganglionic Degeneration

Movement Disorders

Atypical Parkinson Syndromes

- Parkinsonism with Cerebellar and Autonomic Dysfunction: AKA “Parkinson Plus”
 - The Multiple System Atrophies (MSA)
 - Striatonigral Degeneration (SND)
 - Shy-Drager Syndrome (SDS)
 - Progressive pan-dysautonomia
 - symmetrical
 - Olivopontocerebellar Atrophy (OPCA)
 - Progressive cerebellar ataxia

Movement Disorders

Other Movement Disorders

Disorders with Dementia as Primary or Early

- Diffuse Lewy Body Disease
- ATD with Parkinsonism
- Parkinsonism-Dementia Complex of Guam
- Normal Pressure Hydrocephalus
- Crutzfeldt-Jacob Disease
- Pallidopontonigral degeneration & DDPAC mutations
- Early Onset Parkinsonism
- Dopa-responsive Dystonia
- Hemiparkinsonism-Hemiatrophy Syndrome

Toxin-Induced Parkinsonism:*Manganese,*MPTP

Neuromotor Classification By Molecular Defect

Dementias:

- Alzheimer's Disease w Parkinsonism
- Frontotemporal Dementia
- Spinocerebellar Ataxias
- Friedreich's Ataxia
- Parkinson Diseases (pleural)

Hereditary/Metabolic:

- Huntington's Disease
- Wilson's Disease
- Machado-Joseph Disease

Neuromotor Classification By Molecular Defect

Other Idiopathic Neurodegenerative Disorders

- Motor Neuron Diseases
 - Amyotrophic Lateral Sclerosis/MND
- Spinal Muscular Atrophy
- Spinal and Bulbar Muscular Atrophy
- Familial Spastic Paraparesis

Other Movement-Affected Disorders

- Muscular Dystrophies
- NMJ Disorders
- Demyelinating Diseases
 - MS
- Cerebrovascular Dz & Neoplastic Dz

Common Problems – Gen'l Group

- Fatigue
- Pain
- Nausea and Vomiting
- Dysphagia
- Spasticity
- Communication Impairment
- Seizures
- Gastrointestinal Symptoms
- Respiratory Symptoms
- Psychiatric Symptoms
- Respiratory Symptoms

Clinical Nonmotor Symptoms: PD

- Craniofacial
- Cognitive
- Autonomic
- Sensory
- Musculoskeletal
- Dermal
- Psychiatric
- Other

Clinical Nonmotor Symptoms: PD

- Craniofacial:
 - Excess Salivation
 - Masked facies
 - Decrease blinking
 - Olfactory hypo-functioning.
 - Dysarthria
 - Blurred vision

Clinical Nonmotor Symptoms: PD

- Cognitive:
 - 7-85% {20%}
 - Dementia: cortical & “subcortical”
 - Bradyphrenia
- Visio-spatial, attention, & executive deficits

Clinical Nonmotor Symptoms: PD

- Psychiatric:
 - 4-70% reported incidence
 - Depression: 40-50% at some time, 31% at any given time.
 - Anxiety: 25-40% prevalence.
 - Multiple overlapping sx's
- Psychosis: 15-40%.
 - Delusions, disruption, paranoia, hallucinations, phobias, apathy
 - Potential SE of Rx as well

Common Symptoms – ALS

Direct

- Weakness and Atrophy
- Fasciculations & mm. cramps
- Spasticity
- Dysarthria
- Dysphagia
- Dyspnea
- Pathologic Laughing/Crying (Pseudo-bulbar Affect)
 - Up to 50%
 - Related to frontal lobe damage (35%)

Common Symptoms – ALS

Indirect

- Sleep Disturbance
- Constipation
- Drooling
- Thick mucous
- Chronic Hypoventilation Pain

Causes of Sleep Disturbance

- Psychological Disturbances, Anxiety, Depression, Nightmares
- Fasciculations & Muscle Cramps
- Dysphagia w Aspiration
- Respiratory Insufficiency
 - Hypoxia, Dyspnea
- Insecurity
- Inability to change body position

Question

QUESTION # 5

Of the following medications, which are contraindicated in treating sx's in Parkinson's patients?

- a. Reglan (metoclopramide)
- b. Compazine (prochlorperazine)
- c. Haldol (haloperidol)
- d. Phenergan (promethazine)

Contraindication Rx in Parkinson's

Possible/Potential

BP	Alpha-methyldopa
SZ	Phenytoin
Mood	Lithium
Anxiety	Buspiron

Contraindication Rx in Parkinson's

AntiPsychotic	Haloperidol (Haldol)
	Perphenazine (Trilafon)
	Chlorpromazine (Thorazine)
	Trifluoperazine (Stelazine)
	Flufenazine (Prolixin)
	Thioridazine (Mellaril- HighDoses)
	Thiothixene (Navane)

Contraindication Rx in Parkinson's

Depression	Perfenazine/Amitriptyline
N/V	Prochlorperazine (Compazine)
	Metoclopramide (Reglan)
	Thiethylperazine (Torecan)
Misc	Reserpine
	Tetrabenazine

Specific Symptom Rx

Fasciculations & mm Cramps

Vit E	400 IE bid
Mg+	400 mg qd-qid
Quinine S	220 mg bid
Carbamazepine	200mg bid
Phenytoin	100 mg qd-tid
Diazepam	2.5-5 mg tid

Specific Symptom Rx

Spasticity

Baclofen	10-80 mg/day & Intrathecally
Tizanidine	4-24 mg/day
Memantine	10-60 mg/day
Clonazepam	0.5-6 mg/day
Diazepam	7.5-30 mg/day

Specific Symptom Rx

Drooling

Glycopyrrolate	0.1-0.2 mg sq/im tid
Glycopyrronium Bromide	??0.3-0.8 po tid
Scopolamine Transdermal	1-2 patches
Amitryptiline	10-150 mg/day
Atropine/benzotropine	0.25-0.75 mg/1-2 mg per d
Clonidine	0.1-0.3 mg/day

Specific Symptom Rx

Thick Mucous

Guiafenesin	600-2400 mg/day
Glycopyrrolate	inhaled via updraft
N-acetylcysteine +/-	
>Hydration	
>Humidification	

Specific Symptom Rx

Pseudo-bulbar Sx's

Amitryptiline	10-150 mg/day
Fluvoxamine	100-200 mg/day
Citalopram	40-60 mg/day
Lithium	400-800 mg/day
I-dopa	500-600 mg/day

Specific Symptoms/Problems

- Spasticity
- Dysphagia
- Communication Difficulties
- Dyspnea
- Cough

SPASTICITY

- Any defect of the Pyramidal (long corticospinal) tract causes spasticity.
- Chronic pain is common with spasticity
 - Primary (poorly understood)
 - Neuropathic
 - Associated with phasic flexor spasms and mass reflexes
 - Musculotendinous and joint
 - Contractures – later
- Physical Therapy is key early on and midway
- Rx Muscle Relaxants, (& Limited use botulinum toxin)
- Future possible canniboids

DYSPHAGIA

Cooperation is a MUST

Exercises with initial instruction are key

- Lip closure, tonization of cheeks, mastication, velar movement
- Tongue movements, triggering swallowing reflex, improving rate of swallow

Compensatory measures

- Positioning, postural changes, dietary modifications, swallowing techniques,
- Adaptations,

Safety Strategies

PEGs

Rx limited to mucolytics and anticholinergics

COMMUNICATION DIFFICULTIES

Assess:

Hearing and sight, mentation

Oral-pharyngeal function

Laryngeal function

Respiratory function

Wishes and needs

Tx:

Positioning & comfort

Environmental conditions (light, sound)

Facilitating techniques

Augmentive & Alternative Communication

Anticipation and Education, Patience

Chronic Respiratory Insufficiency

Daytime Fatigue & Sleepiness

<Concentration

Sleep Disturbances

Poor sleep quality

Poor hypnosis

Nightmares

Morning HA

Tremor, nervousness, Sweating, Tachycardia

Depression & Anxiety

Dyspnea, Tachypnea, <Phonation

Wt. Loss, <Appetite, Gastritis

Edema, Cyanosis

Dizziness, Syncope

Visual Disturbances

Pain – often diffuse , varies w stage, H&N&back most

Dyspnea Treatment

1. Treat Reversible Causes

2. Non-pharmacological Measures

3. Intermittent Dyspnea

Rx Anxiety, Rx Opioids, If severe = Rx
midazolam, Thorazine

4. Constant Dyspnea

Rx Opioids, Rx Benzos, If terminal = consider
infusions (sq/iv)

5. Oxygen: only helps if hypoxemic

Cough

- Etiology:
 - Excessive bronchial secretions
 - Aspiration
 - Abnormal stimulation of airway receptors
- Three types:
 - Ineffective cough
 - Secretion plagued cough
 - Chronic cough
- !st goal = eliminate causative factors

Cough

Ineffective Cough

Assisted coughing techniques (man or mech)

IPPB

Secretions

Mucolytic Rx ?, Hydration, Airway humidification

Vibration tx, postural drainage, Suction +/-,

Caution with anticholinergics

Chronic

Think aspiration, infection, inflammation, secretions,
dryness or wetness

Can be debilitating, esp. in the weakened pt.

Inhaled Bronchodilators, local anaesthetics

Rx GER

Mechanical Ventilation

“Mechanical ventilation is a mixed blessing as it's potential good is not always good enough. While offering hope of prolonged life, mechanical ventilation has drastic implications for the quality of life.”

-seniors-site.com

Question

Since ALS is a diagnosis with “automatic” Medicare admission (along with CRF), Medicare pays for all of the following except:

- a. Prescriptions through Medicare D, and through Hospice
- b. Ventilators at 80% through Medicare A, or through Hospice
- c. Caregivers through Medicare A & B
- d. Motorized wheelchair, with supplies

Mechanical Ventilation

External:

CPaP, BiPaP;

Inability to protect upper airway relative
contraindication

Negative pressure to thorax or abdomen
(little used)

Internal:

Intermittent PiP,

Bi-level (inspiratory/expiratory)

Causes of Death – ALS

Respiratory failure	86%
Heart Failure	6%
Pneumonia	5%
Suicide	0.5%
Other	2.5%

Death – ALS

Terminal Stages - ALS

Evidence that the majority do NOT choke to death.

Great Variation in terminal phase

Most with gradual deterioration, then sudden change

Hours to few days

Following URI/ Silent aspiration

1/2 to 2/3 die rapidly within 24 hrs

Increased Hypercapnea

Drowsy . . to . . sleep . . to . . Coma

MUCH DIFFERENT IF ON VENT; then ALL THAT CHANGES

Terminal Phase-Neuromotor

Characteristic Warning Signs:

Profound Weakness

Bedbound

Drowsy for extended periods

Disorientation

Severely limited Attention

>Disinterest in food/fluids

>Difficulty taking meds

Terminal Phase

Sx's in Last 48 Hours of Life

Noisy & moist breathing

Pain

Restlessness & Agitation

Urinary incontinence

Dyspnea

Urinary Retention

Nausea &/or vomiting

Sweating

Jerking, twitching, or plucking'

Confusion

Managing the Dying Patient

Prevalence of Sx's

ALS:

Weakness
Immobility
Constipation
Pain
Impaired Speech
Insomnia
Dyspnea
Drooling

MS:

>> Cognition
Pain
Spasticity
Ataxia
Incontinence

Managing the Dying Patient

Recognition/Tx of Dying-

Often, new sx's at the end days

Likely unfamiliar and frightening

Respiratory Sx's

Breathlessness

Choking

Restlessness/Agitation

Must distinguish from myoclonus

Family

Question

If today I were diagnosed with ALS, would I ultimately want to be placed on a ventilator?

- a. Yes b. No

Ventilator Withdrawal

Why Pt/Families Chose Ventilator Withdrawal:

- Expressed desire to be withdrawn
 - Advanced Directives
 - No hope for recovery
- The patient not want to live this way
- Extremely difficult decision to make.
 - “Feels like playing God”

Ventilator Withdrawal

Key Elements

- Staff selection/preparation mandatory
 - Psychosocial involvement is critical
- Team Conference should occur early
 - Explore gut feelings
- Family meeting for preparation and support

Ventilator Withdrawal

Choosing Location:

Home

LTC

GIP

Hospital

Ventilator Withdrawal

Assessments

- Nursing: Pt Summary, AD's, Vent Info, Converse w Pt/Family
- Spiritual: Pt and family, Enlist personal clergy
- Psychosocial: Pt wishes, Surrogacy, ACP, Family Dynamics, Family wishes,
- Physician: Plan if pt survives, Assure adequate care during transitions, Pt/Family preparation
- All: Policies & Procedures at Facilities & Med transport

Ventilator Withdrawal

Preparatory Conference:

- Include hospital/LTC staff
 - Multi-disciplinary
 - Family
 - Surrogates
 - ACP Review
 - Goal-directed
 - Plan established
 - Rx's
 - DME
 - Transportation

Ventilator Withdrawal

Equipment

Respiratory:

- Oxygen
- Masks
- AmbuBag
- Suction
- Nebulizer
 - Rx
 - NS

- Infusional
- Needles
- Tubing
- Fluids
- Pumps
- Syringes

- Electrical

- RX's

Ventilator Withdrawal

Medications

Transitional:

- Morphine - IV/SQ
 - 0.1-5 mg/hr +
- Haldol - IV/SQ
 - 0.1-2 mg/hr +
- Lorazepam or Midazolam - IV
- Updraft: A&A, Steroid, racemic epi

• Afterword

- Morphine - IV drip +
 - Convert
- Haldol - IV drip +
 - Convert
- Lorazepam or Midazolam +
 - Convert
- Steroid - po, topical, updraft
- A & A updraft
- Atropine or scopolamine

Ventilator Withdrawal

Symptoms

- Breathlessness
 - Anxiety
 - Pain
- Secretions
 - Stridor
 - Wheezes

Ventilator Withdrawal

Method

Wean vs. Immediate

NO Established agreement as to which is optimal, although H&PM literature shows preference for immediate withdrawal w extubation

Physician is Always Present