Progressive Supranuclear Palsy (PSP): A Day Hospital Case Study

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St Finbarr’s Hospital
Background

Assessment & Treatment Centre

- The Community Liaison Support Service in the Day Hospital is an ANP / Nurse-led service.
- It provides person centred nursing care, specialist interventions, timely medical review, multidisciplinary input & responsive, co-ordinated community services.
<table>
<thead>
<tr>
<th>Specialist Clinic</th>
<th>Referral Criteria</th>
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</thead>
<tbody>
<tr>
<td>Medical Review</td>
<td>Comprehensive Geriatric Assessment Medical Review</td>
</tr>
<tr>
<td>Falls &amp; Syncope</td>
<td>Single or Recurrent Unexplained Falls requiring further investigation, Intervention.</td>
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<tr>
<td>Movement Disorder</td>
<td>Complex gait &amp; movement disorders requiring specialist intervention to aid diagnosis or treatment of Parkinsons.</td>
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<tr>
<td>Memory</td>
<td>Memory Loss, Cognitive changes impacting on function.</td>
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<tr>
<td>Continence</td>
<td>Urine Incontinence, Frequency, Retention Faecal Incontinence, Constipation.</td>
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<tr>
<td>Service</td>
<td></td>
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<tr>
<td>--------------------------------------------</td>
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<tr>
<td>Stroke Follow-Up</td>
<td></td>
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<tr>
<td>Infusions / Transfusions</td>
<td></td>
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<tr>
<td>Dexa / Bone Densitometry / Bone Health</td>
<td></td>
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<tr>
<td>Investigations ie. Holter, 24hr BP, Lumbar Puncture</td>
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<tr>
<td>Pre-Driving Skills Assessment</td>
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<tr>
<td>Falls &amp; Mobility &amp; Parkinsons Classes</td>
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<tr>
<td>Memory Intervention &amp; Support</td>
<td></td>
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<tr>
<td>Complex Case Management / Community Support</td>
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<tr>
<td>Therapy (Physio, OT, SLT, Dietician)</td>
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This case study is based in the day hospital & addresses the care of a man diagnosed with PSP in 2010, aged 73.

On initial presentation in 2012, Michael’s main issues were unsteady gait, recurrent falls, urinary incontinence, swallow impairment & general functional decline associated with marked upper limb rigidity.

Progressive Supranuclear Palsy (PSP), is a neurodegenerative disease, with approximately 270 patients in Ireland living with PSP (PSPA Ireland). This marginalised group often suffer in silence with inadequate supports.
Kaiser Model Chronic Disease Management

Day Hospital / Acute, Rehab, Respite, LTC beds & community supports.

Specialist Clinic for diagnosis / tx & Community supports.day, care, home help prn

Healthy Ageing Community Groups

<table>
<thead>
<tr>
<th>Level 1</th>
<th>Intensive care coordination</th>
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<tbody>
<tr>
<td>People with chronic diseases and complex needs who frequently use hospitals</td>
<td>• Care across the continuum</td>
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<td></td>
<td>• Tertiary and secondary prevention</td>
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<td></td>
<td>• Enrolled patient population</td>
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<tr>
<td></td>
<td>• Comprehensive assessment and care planning</td>
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<td></td>
<td>• Specialist medical and GP management</td>
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<td></td>
<td>• A package of services</td>
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<td></td>
<td>• Continuous, frequent interventions</td>
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<td></td>
<td>• Linkage to ongoing monitoring and maintenance</td>
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<td>Program example: HARP-CDM</td>
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<thead>
<tr>
<th>Level 2</th>
<th>Planned, managed and proactive care</th>
</tr>
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<tbody>
<tr>
<td>People with chronic diseases and complex needs who use hospitals and are at imminent risk of hospitalisation</td>
<td>• Assessment and care planning</td>
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<td></td>
<td>• GP care</td>
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<td></td>
<td>• Self-management interventions</td>
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<td></td>
<td>• Access to mainstream health services and community programs</td>
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<td></td>
<td>• Generic telephone advice</td>
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<td></td>
<td>• Planned review, recall and reminders</td>
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<td></td>
<td>• Ongoing management and monitoring</td>
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<td></td>
<td>Program example: EliICD, AHPACC Partnership</td>
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<table>
<thead>
<tr>
<th>Level 3</th>
<th>Prevention</th>
</tr>
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<tbody>
<tr>
<td>People with chronic diseases with/without complex needs or people with complex needs who may progress towards requiring hospitalisation in the medium to long term</td>
<td>Risk reduction, for example, obesity reduction and smoking cessation</td>
</tr>
<tr>
<td></td>
<td>Program example: Diabetes Prevention Program, AHPACC Partnership</td>
</tr>
</tbody>
</table>

| Level 4 | |
Person Centre Care

- Physical
- Psychological
- Social
- Gait/Balance
- Economic
- Environmental
- Cognitive
- Pharmacological
- Nutrition

Functional Status
POSTURE OF A MAN WITH PROGRESSIVE SUPRANUCLEAR PALSY

1. AXIAL rigidity is more prominent as compared to LIMB rigidity (in parkinson's limb rigidity is prominent). The person in figure has rigidity in neck & back muscle so that he leans backwards.

2. This patient will also have vertical supranuclear gaze palsy which is more for downward gaze then upward gaze.

These two factors will make it difficult for him to look on the floor while walking so he will frequently trip over bumps on the way or will fall backwards while going downstairs.

POSTURAL INSTABILITY
<table>
<thead>
<tr>
<th></th>
<th>PSP</th>
<th>Parkinson’s Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gait &amp; station</td>
<td>Early falls</td>
<td>Normal gait initially</td>
</tr>
<tr>
<td>Speech</td>
<td>Spastic speech</td>
<td>Soft speech</td>
</tr>
<tr>
<td>Face</td>
<td>Startled</td>
<td>Masked</td>
</tr>
<tr>
<td>Tremor</td>
<td>&lt;10%</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Rigidity</td>
<td>Axial</td>
<td>Limb</td>
</tr>
<tr>
<td>Symmetry</td>
<td>Symmetric</td>
<td>Asymmetric (unilateral) onset</td>
</tr>
<tr>
<td>Response to L-DOPA</td>
<td>Little</td>
<td>Excellent</td>
</tr>
</tbody>
</table>
Parkinsons Plus Syndromes: PSP, Multiple System Atrophy, Alien Limb, Important: Observe Presentation, Gait, Patient History .......
Appendix.
Progressive Supranuclear Palsy (PSP) Rating Scale and Staging System

Medical Advisory Board of the Society for Progressive Supranuclear Palsy (SPSP)—Lawrence J. Golbe, MD, Chair
For more copies or for information on PSP, contact the SPSP at 1-800-457-4777/www.spso.org

History (from patient or other informant)
□ 1. Withdrawal (relative to baseline personality)
  0 = None.
  1 = Follows conversation in a group, may respond spontaneously but rarely initiates exchanges.
  2 = Rarely or never follows conversation in a group.
□ 2. Aggressiveness
  0 = No increase in aggressiveness.
  1 = Increased, but not interfering with family interactions.
  2 = Interfering with family interactions.
□ 3. Dysphagia for solids
  0 = Normal; no difficulty with full range of food textures.
  1 = Tough foods must be cut up into small pieces.
  2 = Requires soft solid diet.
  3 = Requires pureed or liquid diet.
  4 = Tube feeding required for some or all feeding.
□ 4. Using knife and fork, buttoning clothes, washing hands and face (rate worst)
  0 = Normal.
  1 = Somewhat slow but no help required.
  2 = Extremely slow; or occasional help needed.
  3 = Considerable help needed but can do some things alone.
  4 = Requires total assistance.
□ 5. Falls (average frequency if patient attempted to walk unsaid)
  0 = None in the past year.
  1 = 1 to 4 per month.
  2 = 5 to 30 per month.
  3 = >30 per month.
□ 6. Urinary incontinence
  0 = None or a few drops less than daily.
  1 = A few drops staining clothes daily.
  2 = Large amounts, but only when asleep; no pad required during day.
  3 = Occasional large amounts in daytime; pad required.
  4 = Consistent requiring diaper or catheter before and after sleep.
□ 7. Sleep difficulty
  0 = Neither 1° nor 2° insomnia (ie, falls asleep easily and stays asleep).
  1 = Either 1° or 2° insomnia; averages >5 hours sleep nightly.
  2 = Both 1° and 2° insomnia; averages >3 hours sleep nightly.
  3 = Either 1° or 2° insomnia; averages <3 hours sleep nightly.
  4 = Both 1° and 2° insomnia; averages <3 hours sleep nightly.

Mental Exam
Items 8-11, use this scale:
□ 8. Disorientation
  0 = Clear
  1 = Equivocal or minimal
  2 = Clearly present, but not interfering with activities of daily living (ADL)
  3 = Interfering mildly with ADL
  4 = Interfering markedly with ADL
□ 9. Bradynphrenia
  0 = None
  1 = 1°
  2 = 2°
□ 10. Emotional incontinence
  0 = None
  1 = 1°
  2 = 2°
□ 11. Gropping/limitative/utilizing behavior
  0 = None
  1 = 1°
  2 = 2°

Bulbar Exam
□ 12. Dysarthria (ignoring palilalia)
  0 = None.
  1 = Minimal; all or nearly all words easily comprehensible (to examiner, not family).
  2 = Definite, moderate; most words comprehensible.
  3 = Severe; may be fluent, but most words incomprehensible.
  4 = mute; or a few poorly comprehensible words.
□ 13. Dysphagia (for 30-50 cc of water from a cup, if safe)
  0 = None.
  1 = Fluid pools in mouth or pharynx. Or swallows slowly, but no choking or coughing.
  2 = Occasionally coughs to clear fluid; no frank aspiration.
  3 = Frequently coughs to clear fluid; may aspirate slightly; may expectorate frequently rather than swallow secretions.
  4 = Requires artificial measures (oral suctioning, tracheostomy, or feeding gastrostomy) to avoid aspiration.

Supranuclear Ocular Motor Exam
Items 14-16, use this scale. Rate by inspection of saccades on command from the primary position of gaze to a stationary target.
□ 14. Voluntary upward saccades
  0 = None.
  1 = 1°
  2 = 2°
  3 = 3°
  4 = 4°
□ 15. Voluntary downward saccades
  0 = None.
  1 = 1°
  2 = 2°
  3 = 3°
  4 = 4°
□ 17. Eyelid dysfunction
  0 = None.
  1 = Blink rate decreased (<15/min) but no other abnormalities.
  2 = Mild inhibition of opening or closing of mild blepharo-spasms.
  3 = Moderate lid-opening inhibition or blepharospasm causing partial visual disability.
  4 = Functional blindness or near-blindness because of involuntary eyelid closure.

Limb Exam
□ 18. Limb rigidity (rate the worst of the four)
  0 = Absent.
  1 = Slight or detectable only on activation.
  2 = Definitely abnormal. But full range of motion possible.
  3 = Only partial range of motion possible.
  4 = Little or no passive motion possible.
□ 19. Limb dystonia (rate the worst of the four; ignore neck and face)
  0 = Absent.
  1 = Subtle or present only when activated by other movement.
  2 = Obvious but not continuous.
  3 = Continuous but not disabling.
  4 = Continuous and disabling.
□ 20. Finger tapping (if asymmetric, rate worst side)
  0 = Normal (>14 taps/5 s with maximal amplitude).
  1 = Impaired (5-14 taps/5 s with moderate loss of amplitude).
  2 = Barely able to perform (0-5 taps/5 s or severe loss of amplitude).
**SOAP vs. SBAR**

**S** = **Subjective**: Patient's complaints, date started, what relieved the complaint, previous medical treatment, family history, etc.

**O** = **Objective**: Vital signs and physical examination findings. Results of diagnostic/laboratory testing

**A** = **Assessment**: The physician’s analysis of the problem based on the patient’s complaints and physical examination

**P** = **Plan**: Treatment plan including follow up

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**S** = **Situation**: What is going on with the patient. A concise statement of the problem.

**B** = **Background**: What is the clinical background information that is pertinent to the situation

**A** = **Assessment**: What did you find? Analysis and considerations of options

**R** = **Recommendation**: What action/recommendation is needed to correct the problem. What do you want?
<table>
<thead>
<tr>
<th>Symptoms of PSP</th>
<th>Michaels Presentation</th>
<th>Treatment / Intervention</th>
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<tbody>
<tr>
<td>Vertical Supranuclear Palsy, Diplopia, blurred vision, Burning sensation</td>
<td>Loss of vertical gaze &amp; voluntary eye movements, upper eyelid retraction, Photophobia, Dry Eyes, Fixed Stare</td>
<td>Positioning, Eye lubrication, Sunglasses / Hat</td>
</tr>
<tr>
<td>Bradykinesia, Postural Instability</td>
<td>Slowing Down, Backward Leaning, Recurrent Falls End stage: Wheelchair Dependent</td>
<td>Assisted Transfers, Physiotherapy</td>
</tr>
<tr>
<td>Muscle rigidity with progressive dyskinetic movements due to disordered tonicity of muscles, Tremor (rare)</td>
<td>Increased muscle rigidity, hypertonia, Dystonic neck extension, trunkal malalignment, spasticity of joints</td>
<td>Occupational / Physiotherapy, Seating / Positioning Aids, Passive Exercise Programme, Medical / Nursing Input, Analgesia, Paracetamol, Amitryptline</td>
</tr>
<tr>
<td>High Risk of Pressure Ulcers</td>
<td>Grade 2 pressure ulcer scapula and coccyx, healed with dressings and individualised seating &amp; pressure relief</td>
<td>Nursing Management</td>
</tr>
<tr>
<td>Pseudobulbar Palsy / Dysphagia</td>
<td>Difficulty Swallowing, Progressive Dysphagia requiring modified texture diet &amp; thickened fluids, Increasing high risk of aspiration, with distressing coughing episodes, Increasing Weight loss in late stage.</td>
<td>Speech &amp; Language Therapy, Modified Texture Diet &amp; Grade 2-3 fluids, Increasing risk requiring SLT input, Weight decrease significant in last 6 months, Dietician, Medical Review / Nursing Support, URTI / Aspiration Pneumonia, Palliative Care Involvement</td>
</tr>
<tr>
<td>Speech Disturbance, Dysarthria</td>
<td>Progressive deterioration of Speech, Initial presentation bulbar speech, Aphasic in the latter phase &amp; non-verbal communication limited due to physical limitations.</td>
<td>Speech &amp; Language Therapy, Advance Care Planning, Sign Language</td>
</tr>
<tr>
<td>Mood &amp; Emotional Lability, Depression, Agitation / Irritability / Disinhibition</td>
<td>Remained cheerful despite deteriorating condition &amp; limited communication with masked expression &amp; no hand movements, Outbursts of uncontrolled laughing, Rare periods of agitation associated with pain / discomfort, No irritability or dis-inhibition.</td>
<td>Person Centred Care, Individualised Approach, Family Support, Relationship with Carers</td>
</tr>
<tr>
<td>Constipation / Impaction</td>
<td>Regular bowel function until terminal phase when Laxatives required.</td>
<td>High fibre diet, Increased Fluids as tolerated, Laxatives PRN</td>
</tr>
<tr>
<td>Urinary Frequency / Retention Common</td>
<td>Initial Urinary Frequency, Nocturia with Urge Incontinence, Urinary Retention</td>
<td>Individual Assessment &amp; Continence promotion, Supra-pubic Catheter inserted, Recurrent UTI’s</td>
</tr>
<tr>
<td>Cognitive impairment, poor executive function, Eventually Dementia</td>
<td>Memory impairment difficult to assess in the latter phase however MMSE 28/28 Sept 2013 Responsive to Family &amp; MDT.</td>
<td>Sensory Stimulation, Family Interaction, Family brought Michael to Church every Sunday</td>
</tr>
<tr>
<td>Sleep Disturbance / Insomnia / Delayed somnolence</td>
<td></td>
<td>Night Sedation</td>
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Michaels disease progression concords with Golbe & Ohman-Strickland (2007) who found that progression of PSPRS score in 3 age onset groups (n = 162) reconverge at the end stage of illness with usual life expectancy 6 years. Michael lived 5.5 years post diagnosis but he is likely to have had symptom presentation 6 months earlier.

In the last year of his life, weekly attendances provided much needed support until his death in 2015 due to aspiration pneumonia.

Outside of 4 - 6 weeks respite in the year, Michael was not admitted to hospital in the last 3 years of his life.

Michael scored 60/100 on the PSPRS in 2012 & 75/100 in 2015.

His emotional lability and spontaneous outbursts of laughter were silenced by disease progression; recurrent falls gave way to wheelchair dependence.

By 2015, Michael had marked rigidity. Specialised seating was required to reduce risk of pressure ulcers. Time lag in approval of funding was a source of frustration for the family.

Mealtimes were particularly challenging with high risk of aspiration, and despite modified diet, assistance at mealtimes and nutritional supplements, weight loss was significant in the last 6 months, prompting family to explore the possibility of gastrostomy peg feeding.

The support of healthcare professionals was required to address quality of life issues, assist decision making, and manage family expectations around end of life care. Specialist palliative care services were engaged. Weekly Day Hospital attendance provided valued support (see comments below from family).

Michael was cared for at home by family, supported by a homecare package & regular respite. Despite complex issues, Michael was not admitted to hospital during the last 3 years of his life.
Weekly attendance reduced burden of care & gave me time out when his management was too complex for the day centre.

You gave us the confidence to look after him at home & enabled us to face the challenges of coping with a debilitating disease.

You cared for him as if he was a family member & I knew that I could ring at any time if I ran into difficulty.

Your ongoing assessment supported our case for additional home supports.

We had access to healthcare professionals who provided symptom relief & valued advice.
Conclusion

• This case study demonstrates patient & family resilience in the face of a debilitating neurodegenerative disease when offered support in the community.

• The disease trajectory requires a co-ordinated person centred palliative care approach which can be facilitated in community liaison support services in the Day Hospital.

• This case study demonstrated the value of ongoing monitoring & timely intervention of the multidisciplinary team & ongoing communication with patient & family. As a nurse led clinic, no additional resources was required to support this complex case management, however referrals require additional resources.

• ANP role avoids unnecessary admission to hospital & longterm care & sustains care-giving in the home.