Motor Neurone Disease…

…..and the role of specialist palliative care

Kate Grundy, July 2015

Format

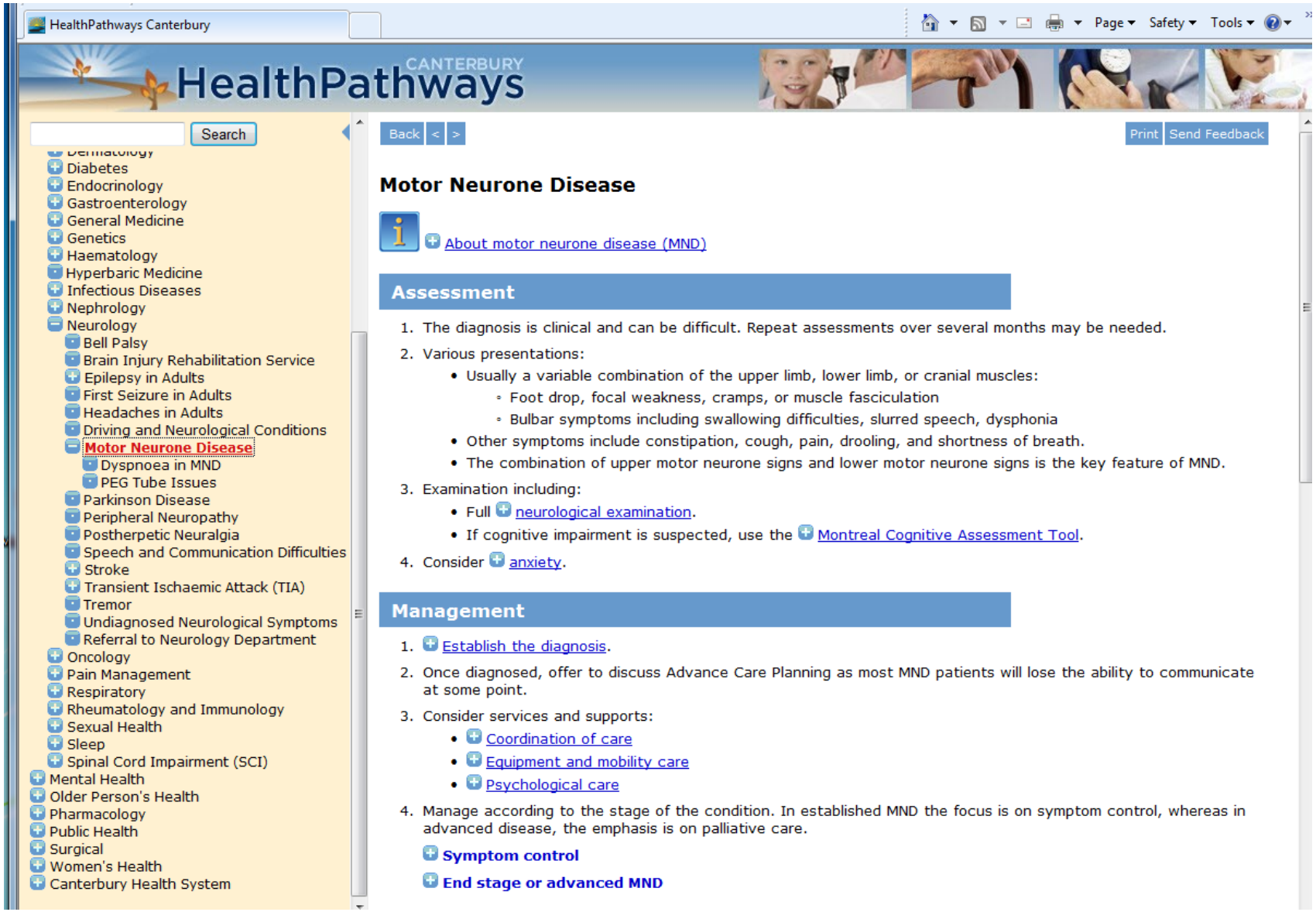
- Overview of the MND service in Canterbury
  - HealthPathways (CDHB)
  - MND co-ordinator (Canterbury Initiative)
- What is MND?
  - 3 case vignettes
- Literature overview / Hot topics
- Role of the MDT and of palliative care
- What is a good outcome?
- Questions……

Personal interest

- Established Christchurch Hospital Palliative Care Service in 1999
  - MND referrals infrequent
  - Occasional use of Hospice services
- Gradually developed expertise in the area
  - Especially when developed an interest in ACP around 2007
- Began working collaboratively with a Respiratory Physician and a Neurologist
  - We started to think of ourselves as a "virtual clinic"
  - Close liaison with our SI MND Association Field Officer
  - Realised the wealth of community allied health interest and expertise
  - Came together occasionally – imagining how much better things could be if our ideas could be realised!

HealthPathways

- Well known across the South Island – started in the CDHB
- HP is one stream of work within the Canterbury Initiative
  - *"Working together at the primary-secondary interface"*
- Perfect for MND where complex care occurs predominantly in the community
- Working group established 2011 with cross sector participation
  - Facilitated by the Canterbury Initiative (Planning and Funding)
  - Incredible energy from the outset – lots of views and opinions!
  - Pathway went live in October 2012 (making the diagnosis, symptom management and end stage disease)
  - Referral information to relevant services e.g. allied health, respiratory department, genetic testing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Motor Neurone Disease
About motor neurone disease (MND)
Assessment
1. The diagnosis is clinical and can be difficult. Repeat assessments over several months may be needed.
2. Various presentations:
• Usually a variable combination of the upper limb, lower limb, or cranial muscles:
◦ Foot drop, focal weakness, cramps, or muscle fasciculation
◦ Bulbar symptoms including swallowing difficulties, slurred speech, dysphonia
• Other symptoms include constipation, cough, pain, drooling, and shortness of breath.
• The combination of upper motor neurone signs and lower motor neurone signs is the key feature of MND.
3. Examination including:
• Full neurological examination.
• If cognitive impairment is suspected, use the Montreal Cognitive Assessment Tool.
4. Consider anxiety.
Management
1. Establish the diagnosis.
2. Once diagnosed, offer to discuss Advance Care Planning as most MND patients will lose the ability to communicate at some point.
3. Consider services and supports:
• Coordination of care
• Equipment and mobility care
• Psychological care
4. Manage according to the stage of the condition. In established MND the focus is on symptom control, whereas in advanced disease, the emphasis is on palliative care.
Symptom control
End stage or advanced MND

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Symptom control

- Analgesia
- Bladder problems
- Carer fatigue
- Cognitive changes and associated problems e.g., difficulties with work or finances
- Constipation
- Coughing and choking
- Dysarthria and dysphonia
- Dysphagia
- Dyspnoea
- Joint stiffness
- Secretions and drooling
- Muscle fasciculation and cramps
- Oedema – causes include restricted activity or posture, DVT, or other intercurrent disease
- Pain
- Pressure management
- Spasticity or jaw spasm
- Weight loss and consideration of percutaneous endoscopic gastrostomy (PEG) insertion.

End stage or advanced MND

Dyspnoea in MND

1. Provide explanation with reassurance of ongoing support. Discuss treatment options early and plan ahead for management of progressive or acute dyspnoea.
2. Clinical history and examination.
3. Consider referral to:
    - occupational therapy, physiotherapy, and/or speech language therapy for advice, equipment, breathing exercises, and monitoring.
    - the Respiratory Department if considering oxygen use or ventilatory support including overnight bilevel positive airway pressure ventilation (BPAP or BiPAP) mask. These are cumbersome but can give significant symptom relief.
4. Consider discussing and documenting patient wishes regarding ventilation in an acute event, and other end of life decisions.
5. Chronic phase medication.

Progressive or Acute Dyspnoea

1. Plan ahead:
    - Establish out-of-hours medical / nursing contact.
    - Consider use of relaxation therapy and other complementary therapies.
2. Facilitate air movement by open window, fan, or oxygen (if already in use).
3. A food bolus could be the cause requiring use of the Heimlich manoeuvre. Re-educate family members.
4. Explain and prescribe drugs:
    - Morphine:

Clinical caseload

- Acute admissions referred to palliative care
- Clinic appointments (in Oncology) for selected patients
- Liaison with Hospice and community palliative care
- Stats available since 2005
  - Last 4 years average about 15 referrals per year
- In 2011, started a Hospice MND clinic with the Nurse Maude dietitian
  - Supported her excellent work with patients, often in advance of a formal community palliative care referral
  - Helped to demystify Hospice for many patients and families
  - Promoted our integrated model for palliative care in Canterbury
  - 86 patients seen in clinic since its inception

MND Coordinator

- By 2012, a clear case had been made for a coordinator
  - Established as a contractor in July 2013 – Heather Brunton (RN)
  - Covers CDHB, WCDHB, SCDHB
- All patients seen at diagnosis - All neurologists supportive
  - 74 referrals to date, 34 have died
- Heather took over organising the Hospice MND clinic
  - Acts as gate-keeper and is able to generate referrals
  - Approx. 2/3 patients have been referred, 22 active patents at present
  - Close liaison with allied health, GP and Neurology
- Evaluations have been extremely positive
  - From CI and from patients and families
  - Permanent employment arrangement being finalised
  - Very steep learning curve!

What is MND?

- Aligned very closely with Amyotrophic Lateral Sclerosis (ALS) and first described in the mid 1800's
  - NOT a single disease entity – best considered as a syndrome [1]
  - Has been a huge improvement in understanding over last 15 years
  - Overlap with the clinical spectrum of frontotemporal dysfunction which highlights possible pathology and also impacts on prognosis
- Characterised by progressive weakness of limbs and bulbar and respiratory muscles due to loss of upper and lower motor neurones
- Prevalence of 6-7:100,000 - Survival is poor (approx. 2 to 5 years)
  - Riluzole confers modest benefit only and effect on Q of L is unknown
- 20 years ago, discovered that a dominant mutation of SOD1 accounts for 15% familial cases
  - Since then, over 100 distinct SOD1 mutations have been identified with huge variability in phenotype
  - Approx. 2/3 familial and 10% sporadic have a genetic mutation

Palliative care and MND

- Feared and often long-awaited diagnosis
  - Patients have often consulted the internet before the diagnosis is confirmed by a Neurologist and can already be significantly disabled
- Physical symptoms are extremely common
  - Weakness, fasciculations, cramps
  - Dysphagia, weight loss, dyspnoea, dysphasia, dysarthria, drooling
  - Pain, constipation, sleep problems, cough
  - Pathological laughing and/or crying
- Holistic care is paramount
  - Psychological and social concerns
  - Spiritual issues and existential suffering
  - Family support
- Focus is on “living with” not “dying from” MND

Symptoms on Hospice Admission[2]

	Patients with MND	Patients with Cancer
Constipation	65%	48%
Pain	57%	69%
Cough	53%	47%
Insomnia	48%	29%
Breathlessness	47%	50%

BMJ, 1992

Case vignettes

- Variability of presentation and disease course
- Complex interface between many healthcare professionals
- Accumulated experience over time
- Being prepared for the unexpected

Dean

- Aged 44. Lives with flatmate. Supportive parents
- June 2011 – Resp review – "probable asthma"
- July 2011 – Neurology review with cramps, spasms, drooling and unintentional weight loss
  - "Probable MND"
- Admitted for PEG insertion Oct 2011
  - Downplayed his symptoms O/A and went into florid respiratory failure post sedation
  - Ventilated on ICU
  - Home on BiPAP – friend became fulltime carer after 2 failed discharges
- Eventually transferred to HLC in April 2012
  - Did very well but requested withdrawal from BiPAP in September
  - Died very peacefully within 24 hours

Tui

- Age 67, very rich and active life
  - Family history of frontotemporal dementia
  - Married, 2 children from previous marriage
- Diagnosed with MND of (progressive bulbar palsy type) Oct 2013
  - Referred to MND clinic April 2014 for advance care planning and psychological support
  - Strongly expressed her support of euthanasia and her intention to withdraw from treatment in future at a time of her choosing
- PEG tube inserted but complicated by bowel perforation
  - Terrible constipation requiring Gastro consult and Hospice admission
- Journey characterised by courage, humour, honesty and respect
  - Out of the blue, requested to be allowed to stop eating and drinking
  - Died at home within 1 week

Mary

- Intermittently under my care since 2007, diagnosed in 2005
  - Then able to walk with a stick but increasingly using a wheelchair
  - Seen in clinic – "feel hopeless, feel like giving up"
  - Determined not to have strangers in the house
- Married with 4 children aged 12 to 24 – 3 still at home
  - She now has 6 grandchildren!!
  - Very strong faith
- Many, many hurdles to overcome since then, the main one being respiratory failure requiring a tracheostomy Sept 2012
  - Fully ventilated since then….at home
  - PEG, suprapubic catheter, cholecystitis with gallbladder necrosis (4/14) – CRP 375!!
  - Still able to go on family holiday every summer to Wanaka!
- At last appointment started to discuss end of life care planning……

The MDT

- GP team (+/- MND coordinator)
- Neurologist (or Geriatrician)
- MND Assoc Field Officer
- Dietitian
- Gastro (PEG) nurse +/- Gastroenterologist
- Speech Language Therapist
- Occupational Therapist
- Physiotherapist
- Respiratory Physician
- Lifelinks/ Enable/ WINZ
- Social worker / Needs Assessor

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- Respiratory Physician
- Lifelinks/ Enable/ WINZ
- Social worker / Needs Assessor
- Family support/counsellor
- Health Care Assistants
- District Nurses
- ARC staff
- Sleep technician
- Cultural / Spiritual support
- Orthotics
- Dentist
- Talklink (Assistive Technology)
- Botox clinic
- ENT (trachy) nurse
- Urologist
- Specialist Palliative Care

Specialist palliative care (UK survey[3])

- Majority of specialist palliative care services in the UK are involved in the care of patients with MND
  - Often only in the terminal stages
  - Major decisions re interventions have already occurred
  - Early involvement appears to be reducing (18% of units in 1999, 9% in 2003)
- Many Palliative Medicine consultants had limited knowledge about interventions and the likelihood of improving both quantity or quality of life
- Different physicians (palliative care, neurology, rehab etc.) indicated that there was contact between the specialties but limited understanding of each others roles
- Clear that achieving appropriate holistic care in combination with appropriate interventions was ideal[4]
  - Overall aim of maintaining Q of L - despite disease progression

What affects the disease course?

- Nutritional care
- Respiratory care
- Presence of absence of FTD
- Multidisciplinary support
  - Improves survival
  - Needs to be coordinated and complimentary
  - ? Role of telemedicine

Nutritional care

- Inadequate nutrition and weight loss are common in MND and if present they shorten survival
  - Causes include dysphagia, impaired motor function (bulbar and extremity) and ? Hyper-metabolic state
- Interventions
  - Dietary input (consistency, calorific content, hydration)
  - OT (braces, utensils, home modifications)
  - Physio (seating, strength maintenance, range of movement)
  - SW (financial support, carers, emotional health)
  - Respiratory (adequate ventilation, secretions e.g. botox)
- No large studies but overall it is accepted that the combination of all relevant interventions above does improve BOTH quality and quantity of life[1]
- Tip; actively enquire as to the burden of oral feeding as it can take up to an hour to eat a small plate of food….

PEGs and RIGs

- Ideally encourage joint patient/family education from the PEG nurse and dietitian
  - Address dietary issues and feeding regimes (bolus v pump)
- Important to consider before loss or respiratory function (FVC < 50%) and before weight loss is marked
  - Ideally commence as a supplement not a full feed
  - RIGs can be inserted more safely in respiratory failure but tend to be less satisfactory
  - No evidence that they improve survival (on their own) but they do improve quality of life[5]
- Insertion come with risks and need to consider how to manage nutrition without a PEG if that is the persons request
- Allow medication delivery and hydration
  - Not just about food
  - Does not mean being fully fed until the end

Respiratory Care

- Breathlessness is common
- Management improves quality of life and may minimise hospital admissions
- Sialorrhoea can affect breathing – address the fear patients may have of "drowning" in one's own secretions
- Opioids in titrated doses may improve quality of life[7]
- Pre-emptive education and routine monitoring of respiratory function tests improves uptake and understanding of NIV
  - FVC and overnight pulse oximetry (SoO_2)
  - Can also use the Sniff Nasal Inspiratory Pressure test (SNIP)[8]
- Respiratory failure can develop insidiously as well as acutely

Non invasive ventilation (NIV)

- NIV improves both quality and quantity of life
  - median survival is 48 days longer for patients treated with NIV (219 v 171 days) and there was enhanced quality of life[9]
- The survival and quality of life benefit was much more in the subgroup with normal to moderately impaired bulbar function (compared to having poor bulbar function)
  - median survival was 205 days longer (216 days in NIV group versus 11 days in standard care group)
- NIV >4hrs /night improves survival where there is minimal or no bulbar symptoms[6]
- Planning for the end of life care includes discussing the issues of progression and withdrawal of ventilation[8]
  - Advance care planning/advance directives
  - Needs a very proactive approach, especially in non specialist or community settings
  - These cases can stay with you a very long time…..

Frontotemporal Dementia (FTD)[10]

- 10-50% all patients with MND have evidence of subtle cognitive decline
- 5-10% have an overt FTD with neurobehavioral dysfunction
  - Personality change
  - Irritability
  - Poor insight
  - Executive dysfunction
  - Poor levels of empathy
  - Reduced survival (3.3 years med survival vs 4.3 years) with rapid decline[1]
- Poor survival is partly related to reduced efficacy of life-prolonging therapies (esp. NIV) with reduced compliance and reduced tolerance
- How best should we evaluate FTD?
  - Should testing be routine?

Allied Health Team & Pall Care

- Talk, listen, ask for (and offer) help and advice
  - Active collaboration
  - Copy individual team members on letters
  - Support them with funding applications (a Specialist letter can open doors)

- Numerous studies have shown that MDT care improves Quality of Life
  - What does MDT mean?
  - Who are we referring to?
  - Encourage innovation and perseverance
  - *Sometimes it's the little things that make all the difference…*

Pain

- Very common, up to 76% in the dying phase
- Cause include
  - Cramps/spasticity (neuronal degeneration)
  - Musculoskeletal (muscle atrophy, joint stiffness)
  - Skin pressure (immobility)
- Also, feet pain, shoulder pain and bladder spasm
- Medications
  - Baclofen
  - Paracetamol
  - NSAIDS
  - Opioids
  - Benzodiazepines
- Supportive measures are crucial

Constipation

- Very common problem
- Multiple causes
  - Dietary changes over time
  - Reduced fluids (in an attempt to minimise need for toileting)
  - Gradually worsening mobility
  - Reduced strength
  - Privacy concerns
  - Difficulty getting into a correct position on the toilet
  - Anticholinergic agents for drooling
    - No wonder patients are not keen to have opioids!
- My approach…
  - Name the problem
  - Address the easy stuff
  - Consider community nursing referral
  - May require scheduling of rectal interventions

Challenges

- Advance care planning
  - Whose role is it?
  - Don't get too specific
  - Need to be clear even without an ACP if patient does not want ventilatory support
- Bureaucracy and delays
  - e.g. Equipment and funding
- Respite
  - Complex care needs – *"no one does it as well as we do"*
  - Hospice v Aged Residential Care
  - Burden v Duty
- Younger patients

What is a good outcome?

For patient/family

- Survival
- Strength
- Function
- Well being

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For patient/family

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For us[11]

- "Just one step ahead"
  - What might be needed, how best to get it and when to step in
- Up to date expertise
  - Timely and focussed education
- "Bespoke Communication"
  - Patient and Carers
  - Care team
- Being able to manage complexity and change
  - Trust allows planning

Questions?

A "mo" ment of light relief!

Tony Gilchrist
2015

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