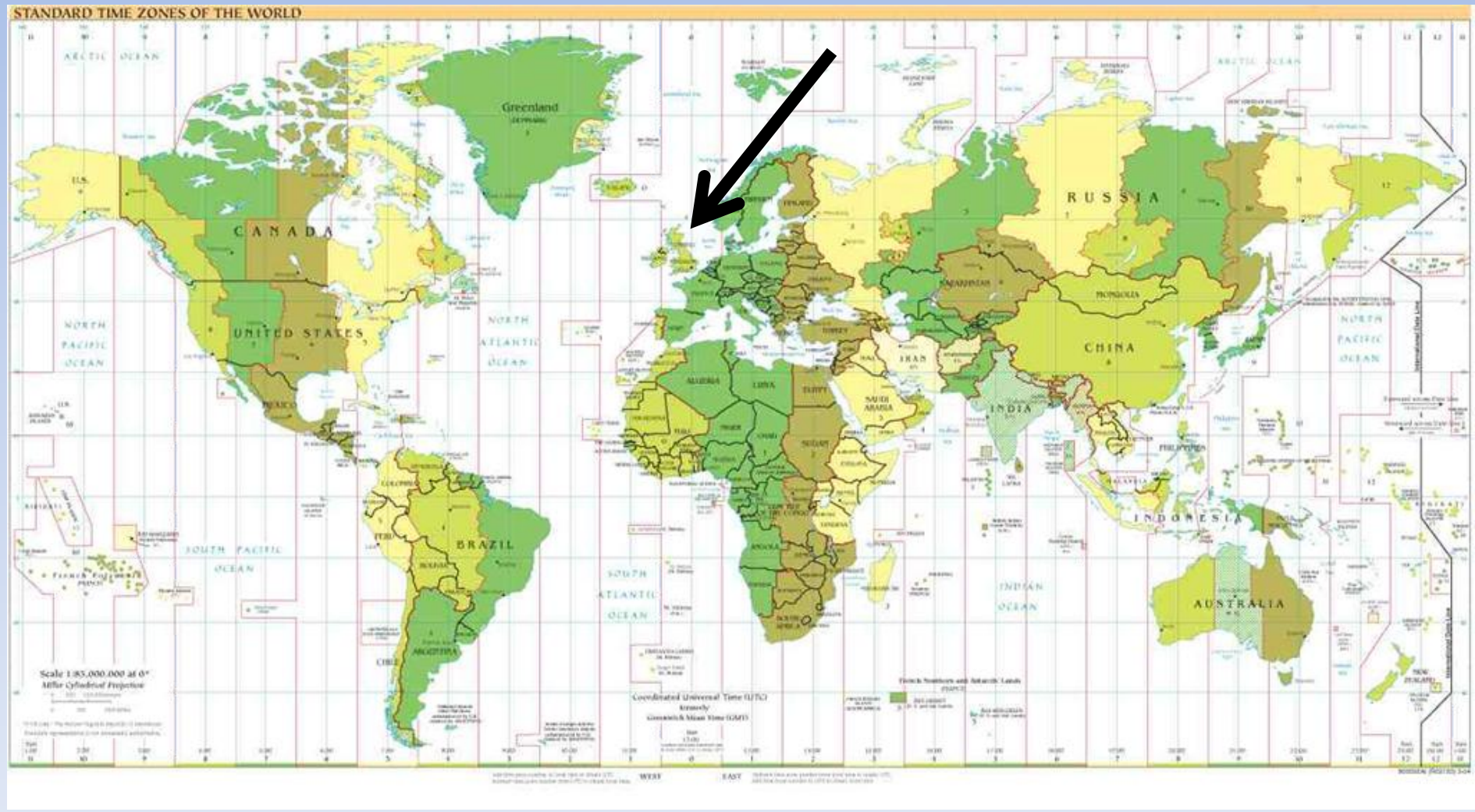
The Benefits of an Education Service for MND/ALS

R S Bestow, Education Officer MND Scotland

Where is Scotland?



- The United Kingdom of Great Britain and Northern Ireland consists of 4 countries

  Scotland, England, Northern Ireland and Wales
- The MND Association operates only in 3 of the 4 countries.
- MND Scotland is the only MND/ALS specific charity working in Scotland

Why Have an Education Service?

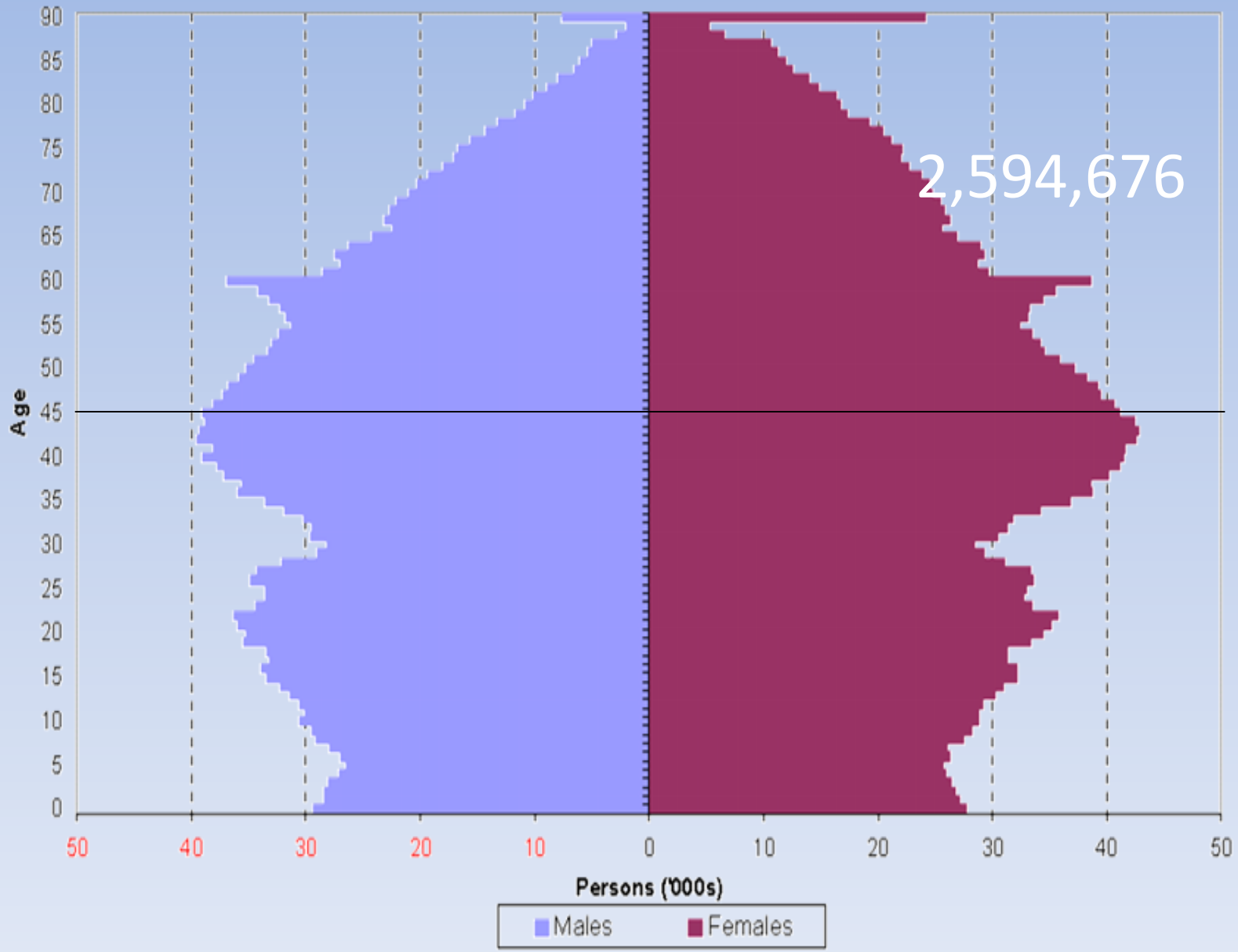
- Increasing numbers of requests for our MND specialist nurses to talk to professionals
  - Typical Family Doctor might see only one or two cases of MND/ALS in a career
  - Little detailed knowledge of the disease outside of specialists
- Allied Health Professionals treat the symptoms, but often don't understand the disease process
- Families live with the disease but know very little about it.

Scottish Statistics

- Approximately 5 million people
- Approximately ⅓ of the land area of the UK (7,710,00 hectares)

  = 65 people per square kilometre,
- Deaths ~ 57,500 ± 2,500 per year
- Deaths from MND/ALS ~ 120 per year
- Life risk of developing MND ~ 1 in 450

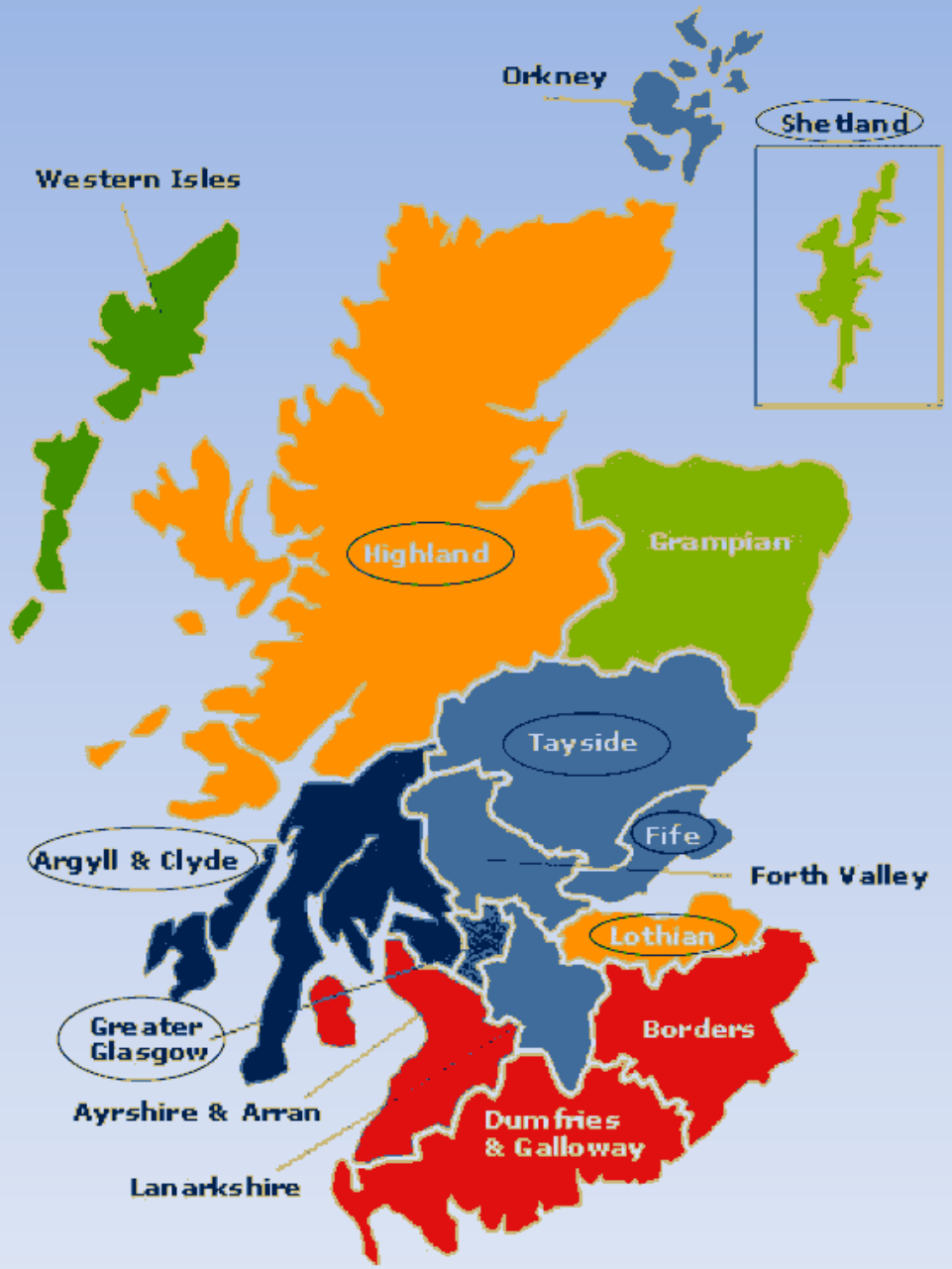
Figure 3 Estimated population by age and sex, 30 June 2007

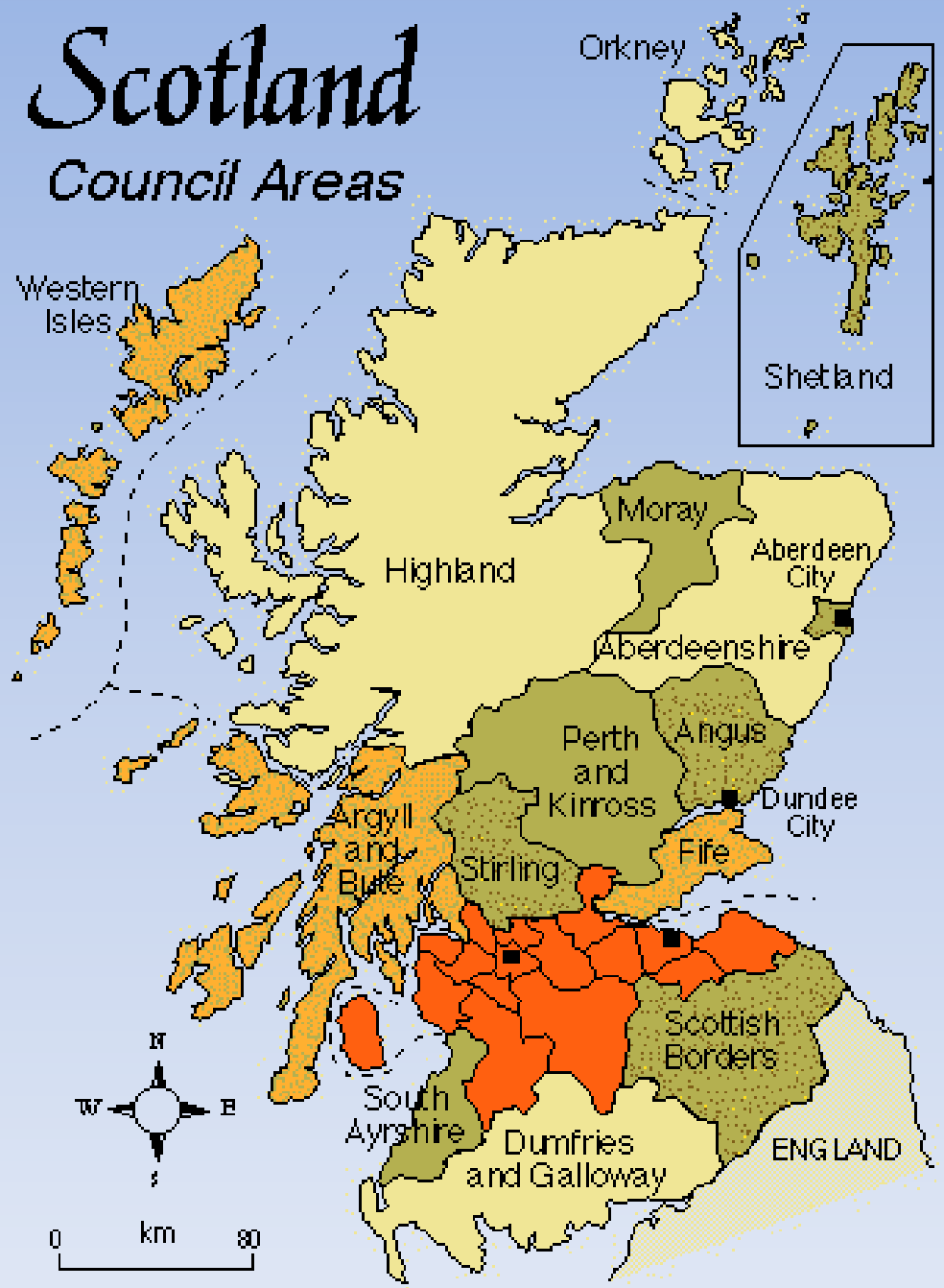


How are Health Services Provided?

- Local Councils (Local Authorities/Government) provide non-medical “Social Services”

  e.g. Occupational Therapy, Social Work, Home Care Services
- National Health Service Health Boards provide medical care, e.g. Doctors, Dietitians, Speech and Language Therapists, District Nurses


Orkney
Shetland
Western Isles
Highland
Grampian
Tayside
Fife
Argyll & Clyde
Forth Valley
Lothian
Borders
Greater Glasgow
Ayrshire & Arran
Dumfries & Galloway
Lanarkshire


Scotland
Council Areas
Orkney
Shetland
Western Isles
Highland
Moray
Aberdeen City
Aberdeenshire
Angus
Perth and Kinross
Dundee City
Argyll and Bute
Stirling
Fife
Scottish Borders
South Ayrshire
Dumfries and Galloway
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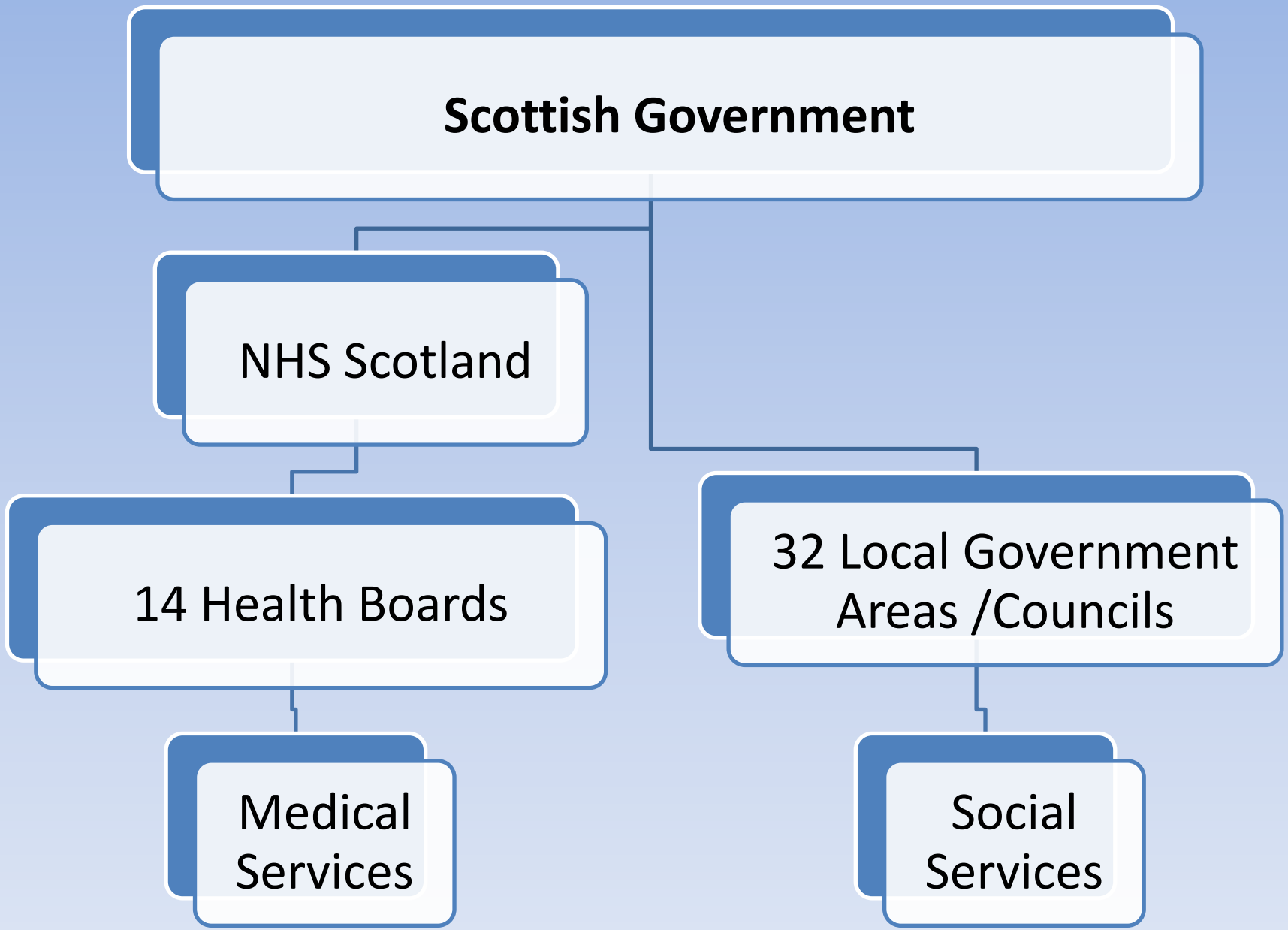
80

- Each Health Board is independent of all of the others
  - NHS Scotland acts as a unifying force
- Each Council is independent of all the others
- The Scottish Government has devolved authority from Westminster for matters related to Health, Civic Government and other issues such as education.


Scottish Government
NHS Scotland
14 Health Boards
Medical Services
32 Local Government Areas /Councils
Social Services

Objectives

- To raise awareness of MND/ALS amongst all health and social care professionals who come into contact with people affected by the disease.
  - Provide an understanding of the disease process
  - Provide examples and resources to encourage the development of best practice
- To give families an understanding of how the disease might develop.

Targeted Audiences

- Occupational Therapists
- Speech and Language Therapists
- Physiotherapists
- District Nurses
- Social Workers
- Care Home Staff
- Palliative Care Staff/Hospices
- Family Doctors
- Families

Materials and Methods

- PowerPoint presentations for specific audiences
- Case Studies
- General study Days
  - e.g. Multidisciplinary Care for MND/ALS
- Specialist Study Days
  - e.g. MND and Dementia
- Family Information Sessions
- Food Preparation Demonstrations (for swallowing difficulties)


MND Factsheet 24
Powers Of Attorney
MND Factsheet 25
Guardianship and
MND Factsheet 27
Coping With MND
MND Factsheet 28
Complementary
MND Factsheet 39
Lithium
MND Factsheet 13
Coping after
Bereavement
MND Factsheet 16
Stem Cells
MND Factsheet 17
Motoring and MND
MND Factsheet 29
Telling Children
About MND
MND Factsheet 18
Clinical Trials
MND Factsheet 5C
Getting the Best
From Physiotherapy
MND Factsheet 5D
Aids to Maintain
Mobility
MND Factsheet 6
Feeding Tubes
MND Factsheet 7
Bereavement
And Children
MND Factsheet 8
Care Planning and
End-of-Life Issues
MND Factsheet 2
Inherited MND
MND Factsheet 3
Introducing Genetics
MND Factsheet 3B
Disease Inheritance
MND Scotland
Patient Information
Folder

MND Scotland

Integrated Care Pathway

Although traditionally regarded as unrelated entities, it is increasingly clear that MND and frontotemporal lobar degeneration (FTLD or FTD) are neurodegenerative conditions with overlapping clinical and neuropathological features. That there is an association between Amyotrophic Lateral Sclerosis (ALS), the most common form of MND, Cognitive Change and Behavioural Change is now well established. Up to 15% of ALS patients meet the criteria for Frontotemporal Dementia and a further 35% have a mild cognitive impairment or other behavioural change.

At the time of writing two genes, TDP-43 and FUS/UTL, are implicated in both ALS and FTLD. TDP-43 positive pathology, where TDP-43 deposits are found within cytoplasmic and nuclear inclusions is present in virtually all patients who have FTLD associated with ALS. .However, the deposits of the protein made by this gene are found in different sub-cellular locations of the neurone in FTLD when compared to those cases of ALS in which the gene has been implicated as a causative agent. Cognitive & Behavioural change in MND is generally agreed to form a spectrum ranging from mild changes in perception occurring later in the illness, often detectable only by specific testing, through aphasia, which can occur at almost any stage of the illness, to a more severe familial condition in which a dementia that begins with behavioural changes can precede the onset of ALS symptoms by six months or longer.

One model, (see, for example, Lillo and Hodges J. Clin. Neurosc. 16: pp1131-1135, 2009) proposes that FTLD in MND can present as either a behavioural variant (bvFTD) or a language-onset variant and that the language onset variant can manifest as either Semantic Dementia (SD) or Progressive Nonfluent Aphasia (PNFA).

In common with the other symptoms of MND there is no fixed pattern of development of the symptoms associated with Cognitive and Behavioural Change that is applicable in all cases. Instead most ALS-FTD patients will develop some of the features of one of the subsets of ALS-FTD (above) while very few will develop all of the features of that subset.

It is important to consider that patients with unaddressed respiratory deficiencies may show signs of confusion, lack of concentration, listlessness and poor memory due to the cumulative effects of weeks of sleep disturbances as a result of sleep hypoxia. It is very probable that such respiratory deficiencies will exacerbate any symptoms arising from cognitive or behavioural changes. In consequence it is important to eliminate respiratory insufficiency as a contributing factor to the person's cognitive or behavioural changes before attempting any psychological assessment.

Manifestation	Presentation	Psychosocial Considerations	Potential Risks	Interventions	Supportive Resources
Behavioural Changes	These fall within the range typical of FTD and may include apathy, restlessness, impulsiveness, self-centredness, reduced concern for the feelings of others, perseveration (fixed on one activity or routine), social disinhibition, increased irritability, emotional blunting, food cramming (especially sweet foods), stereotypical behaviours and compulsive behaviour (taking advice or suggestions as imperatives).	Reluctance to become involved in activities. Unpredictability of actions (impulsivity); may make inappropriate actions or comments, may develop fixed behavioural routines and insist others should also comply with these. May appear demanding or controlling.	Alienation of friends, family and others who may be offended due to not understanding the origins of these behaviours. Weight gain if food cramming. Social problems, e.g. Police, due to disinhibited and sometimes sexually overt behaviours or comments.	Education of all involved of the nature and scope of these behavioural changes. Distraction of the patient from the troublesome or concerning behaviour. Manage the environment to remove "triggers" for the behaviour.	MND Scotland Care Team, Community Psychiatric Nurse, GP, DN, MND Scotland's "Problem Solving Approach" booklet.
Semantic Aphasia	Loss of names for things and impaired understanding of, concepts, words, objects, or facial expressions. The patient frequently can't "find the right word" and has fluent, empty speech with substitutions such as "*thingy*", and "*what's it called*", but the grammatical aspects are preserved. Naming is impaired with errors (such as "animal" or "horse" for zebra). Patients fail to understand less frequent words and fail on a range of tasks such as matching words to pictures and matching pictures according to their meaning. Repetition of words and phrases is normal even though patients are unaware of their meaning. Day-to-day memory (episodic memory) with good visuospatial skills and non-verbal problem-solving ability are relatively preserved, at least in the early stages unlike in Alzheimer's disease. MRI imaging in semantic aphasia shows a typically asymmetric pattern of anterior and inferior temporal lobes (left greater than right).	Difficulties describing where the person has been, what they have done, or what they want to do. Frustration or embarrassment due to recognition by the person themselves that they are unable to name objects or places. May fail to recognise the emotional state of others and not moderate themselves accordingly. Difficulty in understanding what is being said, particularly when being instructed in new ways of doing things, e.g. how to operate	Inappropriate responses arising from frustration, e.g. withdrawal. Misunderstandings by others of what the person is trying to communicate. Difficulty in learning new ways of doing things or operating new equipment due to not understanding instructions if unfamiliar words are used.	Education of all involved with the person of the nature and scope of language changes. Consider how MND can progress when making suggestions for interventions for other symptoms so that initial interventions are built upon later, rather than being replaced by new and unfamiliar instructions, equipment or techniques which require complete relearning.	MND Scotland Care Team, GP, DN, MND Scotland's "Problem Solving Approach" booklet.

Benefits of an Education Service

- Allows Care Team staff to concentrate on patients and their needs
- Can develop specific resources to meet specific needs
- Can act as a central point of contact for disease specific information for professionals and families
- Can cultivate contacts amongst Local Authority and NHS Training Managers

Benefits

- Health Boards, Local Authority staff and Private Sector Staff now approach us asking for information when they have an MND/ALS case to deal with
- Palliative Care Doctors in general hospitals are now also approaching us for disease specific advice