DEMENTIA

6.1 DEMENTIA

IDENTIFICATION

Epidemiology: >65 years 5%, >80 years 20%.

REFERRED

Carer, liaison, forensic, General Practitioner, community health centre, self.

ISSUES

Diagnosis, management, carers supports, insight, difficulty, history of behaviour and memory loss, issues pertinent to stage in life cycle, issues pertinent to stage of dementia.

PRESENTING COMPLAINT

Memory loss, behaviour problems, personality, decrease in functioning

Use mnemonic *MAJOR PD*

Memory loss – decreased short term and long term memory
Abstract thought decreased, Aphasia, Apraxia, Agnosia – four As
Judgement decreased
Organicity presumed
Rule out depression and
Personality disorder
Decline in functioning at work and relationships

No neurological signs – degenerative disorder, e.g. Alzheimer's disease

Neurological signs – vascular dementia?

RELEVANT NEGATIVES

Exclude cerebrovascular accident, transient ischaemic attacks, increased blood pressure, alcohol, psychosis, depression, delirium, drug effects (3Ds – Depression, Delirium, Drugs)

ASSESSMENT

- Onset – sudden or gradual?
- NB sudden may be unmasked by death of partner
- Progression – stepwise, abrupt or progressive – depending on cause
- Current difficulties

CONSEQUENCES

- Affect – depressed symptoms, labile, angry, irritable
- Behaviour – disinhibited, aggressive, at risk of abuse
- Problems – relationships at work, social, forensic, navigational
- Severity – current level of functioning – enquire in detail, self care activities, continence
- Find appropriate level of impairment
- Abstract, complex tasks
- Activities of daily living: basic, instrumental, complex

FAMILY HISTORY

Dementia, senility, CVA, multiple sclerosis, psychiatric disorder, depression, drug and alcohol, deliberate self-harm, movement disorders, Huntington's disease, Downs, apolipoprotein E

PERSONAL HISTORY

Usually brief. Ask about exposure to heavy metals, occupation, head injuries, boxing, sexually transmitted diseases.

PAST PSYCHIATRIC HISTORY

Previous admissions, depressive episodes, deliberate self harm.

PAST MEDICAL HISTORY

- Vascular risk – smoke, increased blood pressure, myocardial infarction, atrial fibrillation
- Parkinson's disease, head injury, Downs, iatrogenic, thyroid, gastrectomy (low B_{12}), hypoglycaemia

DRUG AND ALCOHOL HISTORY

Alcohol and possible use of benzodiazepines and anticonvulsants.

PREMORBID PERSONALITY

Coarsening, exaggeration or flattening of traits.

MENTAL STATE EXAMINATION

- Appearance and behaviour – attitude
- Level of activity, apathy, agitation
- Posturing, stereotypies, mannerisms
- Talk, poverty of content, perseveration
- Affect, mood, denial, neglect, psychotic, cognitions consistent with depression, delirium
- Catastrophic reaction
- Failure of new learning

Use mnemonic *CALM FACE*

Consciousness – alert, lethargic, stupor, coma, drowsy
Attention – digits, serial 7s, WORLD
Language – speech naming, repetition, comprehension, reading and writing
Memory – short-term memory, orientation, remote, personal
Frontal lobe tests – alternate hand movements, frontal release signs (palmomental, grasp, snout)
Apraxia, agnosia, astereognosis, aphasia
Construction – figures
Executive functions – general knowledge, abstract, proverbs

ON EXAMINATION

- Cardiovascular system, retina, carotid bruit, blood pressure
- Movement – Huntington's, Pick's (primitive reflexes), Parkinson's (tremor, rigidity, akinesia)
- Thyroid – myxoedema
- Central nervous system – frontal lobe release signs, parietal, upper motor neurone, extrapyramidal side effects
- Primitive reflexes – snout, pout, grasp, glabella, palmomental
- Liver/alcohol stigmata
- Calcium ring
- B_{12} – SACD
- Steele–Richardson syndrome – impaired upward gaze

DIFFERENTIAL DIAGNOSIS

- Psuedodementia (depression, dementia + depression, schizophrenia, anxiety, mania)
- Benign senescent forgetfulness, AAMI age-associated memory impairment
- Delirium – can be demented and delirious
- Amnestic disorder, frontal lobe syndrome, parietal lobe syndrome (Gerstmann)
- Dementia due to general medical condition
- Substance intoxication, withdrawal – causing multiple cognitive deficits
- Mental retardation
- Malingering, factitious

ISSUES

Safety

- Admit only for complications and occasionally for diagnostic clarification.
- Consider deliberate self harm, comorbid depression.

Clarify diagnosis

- Observations and disability
  Inpatient observation – look for depression, diurnal mood variation (typically, am: worse depression, pm: worse dementia), interaction with other patients, activities of daily living
  Look at patients behavioural ability to perform tasks to look after self and person
- Exclude differential diagnosis
- Exclude reversible causes

ASSESSMENT OF MEMORY LOSS IN THE ELDERLY

Memory loss

Benign age-associated.

Pathological memory loss

Functional, depression, mania, anxiety, schizophrenia, hysteria, malingering.

Organic

Acute ?cause delirium, dementia.

Chronic

- Focal
- Dysmnestic syndrome
- Frontal lobe syndrome
- Parietal lobe syndrome
- Dysphasia

Cause of dementia

Potentially reversible

- Thyroid ↓ (or ↑)
- Calcium ↓ (or ↑)
- Folate ↓
- B_{12} ↓
- Tumour – intracranial
- Space-occupying lesion
- Normal pressure hydrocephalus
- Syphilis, AIDS

Irreversible

Cortical

- Degenerative
- Alzheimer's
- Frontotemporal including Picks
- Diffuse Lewy Body disease

Subcortical

- Degenerative
- Vascular dementia
- Parkinson's disease
- Steele–Richardson syndrome

Other

- Alcohol
- Head injury

Nature of deficits

- Language
- Praxis
- Personality

Other pathology

- Sensory
- Physical
- Psychiatric

Patient's assets

Effects on family

SPECIAL INVESTIGATIONS

To exclude ... I would do ...

Mandatory, FBC, B_{12}, folate, ESR, UE, CA, LFT, TFT, CT (without contrast).

If indicated: glucose, ANF, VDRL, HIV, ECG, EEG, toxicology.

Specialist centres.
MRI SPECT.
Neuropsychological assessment.

CT

- May show cerebral atrophy, focal brain lesions, hydrocephalus
- Help in aetiology of movement disorders, frontal lobe atrophy in Pick's
- Does not give Alzheimer's diagnosis but excludes other causes

EEG

- Slow frontal wave pattern and cortical atrophy on CT in Alzheimer's
- Lesions in the white matter on CT and periventricular white matter hyperintensities with MRI in vascular dementias
- Enlarged ventricles but normal sulci with normal pressure hydrocephalus
- Subcortical degeneration in Parkinson's disease, Huntington's disease

- Collateral history from other sources to verify and provide additional information, e.g. family, other notes, doctors
- Role of other professionals – to do what?
  Domicillary assessment, occupational therapist, physiotherapist, activities of daily living, social work, family therapy, speech therapy

  Care giver support – respite, home helps

Establish therapeutic alliance (if possible)

Decrease symptoms using a biopsychosocial model

SHORT-TERM MANAGEMENT

Aim to keep at home:

Biological

Treat reversible causes, e.g. comorbid depression with:

- SSRI's, SNRI, MAOIs/RIMA, TCA with least anticholinergic side effects, e.g. nortriptiline/desipramine
- Low dose of medication in elderly
- Treat psychiatric symptoms of dementia – antipsychotics

- Consider side effects – use new antipsychotics, e.g. risperidone
- High potency antipsychotics, e.g. haloperidol 0.5–5 mg daily, side effects akathisia and psuedoparkinsonism
- Low potency antipsychotics, e.g. thioridazine 25 mg nocte, side effects: postural hypotension and sedation
- Treatment of dementia with tacrine (TCA) – side effects: nausea, vomiting, diarrhoea, blood dyscrasias. Monitor LFTs and FBC
- Acetylcholinesterase inhibitors – donepezil, rivastigmine, metrifonate

Psychological

Break news, counselling, offer hope, memory aids, support carer, psychoeducation, behavioural strategies.

Social

Occupational therapy, meals on wheels, domiciliary, day centre, mobilise support, carers, financial counselling, enduring power of attorney, wills, general practitioner, social services, work, driving.

MEDIUM-TERM TREATMENT

Biological

Review comorbidity, treatment of behavioural symptoms, psychosis/depression.

Psychological

Ongoing support, carer, remotivation, reminiscence, reality orientation, reinforcement, validation.

Social

Respite day care, behavioural modification.

LATE MANAGEMENT

- Nursing home placement, grief issues, memory loss, cognitive techniques
- Self help Alzheimer's Association

DIAGNOSTIC CRITERIA FOR DEMENTIA OF THE ALZHEIMER'S TYPE

A. The development of multiple cognitive deficits manifested by both

1. Memory impairment (impaired ability to learn new information or to recall previously learned information).
2. One (or more) of the following cognitive disturbances:

- Aphasia (language disturbance)
- Apraxia (impaired ability to carry out motor activities despite intact motor function)
- Agnosia (failure to recognise or identify objects despite intact sensory function)
- Disturbance in executive functioning (i.e. planning, organizing, sequencing, abstracting)

B. The cognitive deficits in (A1) and (A2) cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C. The course is characterized by gradual onset and continuing cognitive decline.

D. The cognitive deficits in (A1) and (A2) are not due to any of the following:

1. Other central nervous conditions that cause progressive deficits in memory and cognition (e.g. cardiovascular disease, Parkinson's disease, Huntington's disease, subdural haematoma, normal pressure hydrocephalus, brain tumour).
2. Systemic conditions that are known to cause dementia (e.g. hypothyroidism, vitamin B_{12} or folic acid deficiency, niacin deficiency, hypercalcaemia, neurosyphilis, HIV infection).
3. Substance-induced conditions.

E. The deficits do not occur exclusively during the course of a delirium.

F. The disturbance is not better accounted for by another axis 1 disorder, e.g. major depressive disorder, schizophrenia.

Early onset <65years

Late onset >65 years

Specify if:

With Delirium
With Delusions
With Depressed Mood
Uncomplicated
With Behavioural Disturbance

AETIOLOGY OF DEMENTIA

DEGENERATIVE

Alzheimer's, Pick's, Huntington's, Creutzfeldt–Jakob, Parkinson's, multiple sclerosis, Wilson's, progressive supranuclear palsy, progressive multifocal leucoencephalopathy.

VASCULAR

Multi-infarct dementia, subarachnoid haemorrhage, ischaemic encephalopathy (Binswanger's disease).

TOXIC

Alcoholic dementia, dialysis, metal toxicity, aluminium, poisons, drugs, e.g. psychotropic drugs and anticholinergic, methyldopa, clonidine, propanolol, anticonvulsants, cimetidine and amantidine.

ANOXIC

Cardiac, respiratory, anaemia, post anaesthesia, carbon monoxide poisoning, post-cardiac arrest.

INFLAMMATORY

Encephalopathies, cranial arteritis, SLE, neurosyphilis, encephalitis.

SPACE-OCCUPYING LESION

Tumour, abscess, haematoma.

METABOLIC

Hepatic, renal.

ENDOCRINE

Myxoedema, hypopituitarism, Addison's, hypo- and hyperparathyroid.

VITAMIN

$B_{1,6,12}$ folate.

NORMAL PRESSURE HYDROCEPHALUS

Ataxia, incontinence, dementia.

TRAUMA

Head injury, punch drunk, child abuse.

EPILEPSY

DISTANT NEOPLASIA

HIV

PREVENTION

No drug has been proven to prevent Alzheimer's disease. However, long-term use of anti-inflammatory drugs and oestrogen replacement therapy in post-menopausal women may have a protective effect. Those at risk of developing Alzheimer's disease (strong family history) should weigh up the risks and benefits of drug treatment.

PROGNOSIS

- Alzheimer's – gradual decline over 8–10 years, progressive disease
- Vascular dementia – onset is sudden, greater preservation of personality, course is stepwise and patchy although some patients have clinical course similar to Alzheimer's
- 90% of Alzheimer's will have psychiatric or behavioural complications during the course of the dementia
- 30% delusions, paranoid ideation, 16% hallucinations, 30% misidentifications syndromes, 5–20% major depressive episode, 20% physical aggression, 20% wandering
- **NB** when diagnose dementia there is a second patient, the family carer. The burden of caring for the patient with dementia is immense. This can be manifest physically, financially, social isolation or psychological distress eg depression, somatic or anxiety symptoms

DISTINGUISH BETWEEN DEMENTIA AND PSUEDODEMENTIA

PSUEDODEMENTIA

- Family aware of history, dysfunction, severity
- Aware of onset of severity
- Date onset with relative precision
- Rapid progression of symptoms and then plateau
- Previous psychiatric dysfunction common
- Cognitive loss complained of
- Detailed complaints of cognitive dysfunction
- Disability emphasised
- Highlight failures
- Communicate sense of distress
- Affective change pervasive
- Loss of social skills
- Behaviour incongruent with severity of cognitive dysfunction
- Answer 'don't know'
- Memory loss, gaps
- Variable performance on tasks

DEMENTIA

- Family often unaware of dysfunction and severity
- Onset known only in broad limits
- Symptoms of long duration
- Symptoms of long course
- Cognitive loss rarely complained of
- Conceal disability
- Delight in accomplishments
- Struggle to perform tasks
- Rely on notes and calendars
- Unconcerned, labile, shallow affect
- Social skills retained
- Attention and concentration faulty, near miss answers frequent
- Memory loss for recent events more severe than remote
- Consistent poor performance on tasks

SIGNS OF CORTICAL DEMENTIA

- Aphasia
- Apraxia
- Agnosia
- Amnesia
- Acalculia

SIGNS OF SUBCORTICAL DEMENTIA

- Slow thinking
- Attention decreased
- Arousal decreased
- Concentration decreased, forgetful, decreased ability to manipulate knowledge
- Motor signs, gait, weakness, incoordination, tremor, reflexes increase, primitive reflexes, posture bowed and extended, gait, tremor, weakness
- Personality change
- Language and parietal lobe function spared

EXTENDED COGNITIVE EXAMINATION

- Attention, concentration (digit span, serial sevens, world backwards), orientation (name day of week, month, year, location)
- Cognitive skills
- Memory
- Reasoning and problem solving

FRONTAL LOBE SIGNS

- Apathy
- Euphoria

- Irritability
- Social inappropriateness
- Lack spontaneity
- Decrease mental and physical activity
- Intellectual impairment
- Poor concentration
- Inability to carry out plans
- Attention deficit
- Sequencing – decreased, slow mental processing
- Executive role, personality, planning, abstraction
- Behavioural control, appropriate
- Disinhibited and euphoric – basal orbital lesion
- Apathy, decreased drive, improved planning – dorsal lateral convexities

FRONTAL LOBE TESTS

- Generativity – test with verbal fluency (<13 'c' words in 90 seconds abnormal), write a sentence
- Abstract, proverbs
- Shift sets – pattern reproduction, alternating sequences e.g. fist/palm/side
- Judgement – social setting
- Wisconsin card sort test, trail making tests, frontal release signs, primitive reflexes
- Planning, draw a house, clock
- Primitive reflexes

TEMPORAL LOBE

- Naming objects, e.g. key, – dominant temporal lobe
- Word finding, reading
- Conversation during interview – receptive language – dominant temporal lobe
- Repetition of words, apple, table, penny – immediate recall – temporal lobes and frontal lobes
- Recall of words – recent memory, name and address, Babcock sentence – hippocampus, thalamus, fornix, mamillothalamic tract
- Long term memory – history, four presidents, last war

DOMINANT PARIETAL LOBE

Gerstmann's syndrome:

- Finger agnosia
- Dyscalculia – simple sums
- Dysgraphia – dictate a sentence and ask patient to write it down
- Right/left disorientation
- Ideomotor apraxia – blow out match, comb hair, simple tasks
- Ideational apraxia – mime putting stamps on letters etc

NON DOMINANT PARIETAL LOBE

Construction apraxia – copy outline of objects.

Dressing apraxia

OCCIPITAL LOBES

- Visual perception defect – cannot name object when camouflaged
- Useful tests – Luria Nebraska, Halstead Reitan, Trail Making test, Bender Gestalt, WAIS R, Wisconsin Card Sorting Test, Ravens Matrices

PERSEVERATION

Occurs in:

- Prefrontal cortex
- Head injury
- Cerebrovascular accident
- Tumours
- Dementias
- Schizophrenia – catatonic

ACTIVITIES OF DAILY LIVING

Basic:

- Bath
- Dress
- Toilet, continence
- Transfer

Instrumental:

- Telephone
- Finances
- Medication management
- Shopping
- Laundry
- Cooking
- Housework

Complex:

- Self direction
- Interpersonal relationships
- Social transactions
- Learning
- Recreation