DEMENTIA : MAKING A GOOD DIAGNOSIS

DR SHAN WILLIAMS
CONSULTANT OLD AGE PSYCHIATRIST

WHY DOES IT MATTER?

- Make an accurate diagnosis, and manage appropriately
- Allows dementia patient and carers to better deal with the disease, improve quality of life and make informed choices about care
- Save money
  - Evidence that the introduction of effective dementia services are cost effective in the long term
- Delay and prevent unnecessary admissions into care homes.
  - Care home placement of people with dementia costs the UK £7 billion per year and early provision of support can decrease institutionalisation by 22%

WHAT'S THE STORY?

I think there's something wrong with my memory

I think Mum/Dad's memory isn't too good – is it dementia?

There's nothing wrong with me, but I'm coming to please my daughter

She's accusing the neighbours of stealing her face cream

The neighbours have complained about the state of the property

He went to visit his son and ended up in a field….. etc.etc.

WHAT TOOLS DO I HAVE?

HISTORY

COGNITIVE ASSESSMENT

SCAN

STEP 1 – PHYSICAL CAUSES TO RESOLVE FIRST OR TAKE INTO ACCOUNT

Delirium? Exclude infection, pain and constipation, consider cholinergic burden of prescribed medication – **PINCH ME**

Drugs / alcohol? History of abuse? Current intake?

Physical health? Falls? Gait?

Bloods : FBC, $B_{12,}$ Folate, Plasma viscosity/CRP, Urea & electrolytes, Liver function tests, Thyroid function, Calcium, Cholesterol, Glucose.

Please check blood pressure, pulse and weight- consider BMI, check alcohol intake and smoking.

WHAT ARE THE CLUES TO LISTEN OR LOOK OUT FOR?

Are there any hints that this might be a dementia or can the symptoms be explained by something else - normal ageing, depression, epilepsy, Parkinson's disease, delirium, alcohol, metabolic syndromes, chronic psychiatric illness etc etc?

If you think there is a dementia, then what type is it?

TYPES

Alzheimers Disease - > 50%

Lewy body dementia - ? 20%

Parkinsons Disease dementia

Vascular dementia – 20%

Alcohol induced dementia – 10%

“reversible dementias” – 5%

- Tumours
- Normal pressure hydrocephalus
- Post traumatic
- Limbic encephalitis
- Metabolic

Pseudodementia – 5%

Fronto-temporal dementias– 3% - behavioural, semantic, PNFA

AIDS related dementia

Huntingtons Disease

Syphilis – GPI

CJD/nvCJD

Encephalopathic – Hashimoto’s encephalopathy, valproate encephalopathy

Etc.Etc.

HOW DO I TELL THE DIFFERENCE?

Some guidance may point you in the direction of one dementia or another

BUT, you often get a mixed picture

THERE IS NO WAY TO BE 100% CERTAIN WITHOUT A POST MORTEM – EVEN SCANS HAVE FALSE POSITIVES AND NEGATIVES, SO OUR DIAGNOSIS IS A BEST GUESS.

DON'T PANIC!

Exclude other reasons for memory loss

- History, examination, investigations including scanning

Decide if the dementia is affecting the front or back of the brain or both

- History, cognitive testing, scans

Start appropriate treatment

Refer to the memory clinic if you think the picture is very unusual, but not if you just want to check your own diagnosis – phone your practitioner and have a chat if you're unsure.

DEFINITION OF DEMENTIA (ICD10)

A syndrome usually of chronic or progressive nature in which there is disturbance of multiple higher cortical functions – memory, thinking, orientation, comprehension, calculation, learning capacity, language and judgement

AND

An appreciable decline in intellectual functioning and interference with personal activities of daily living.

HISTORY

Onset and course - insidious, sudden, gently progressive, stepwise, acute deteriorations

What problems is it causing?

Is it affecting daily functioning? If so, how? - must have some deterioration in function for a diagnosis of dementia, otherwise MCI

Has behaviour changed? If so, how?

Any other symptoms – hallucinations, misrecognition, psychotic symptoms, change in gait or mobility, aggression, appetite, speech, change in personality, change in handwriting, sleep etc?

What are the risks – driving, getting lost, aggression, cooking etc?

HISTORY HINTS

Check the course of memory problem

- When & what was the first symptom? Gradual? Fluctuating? Insight? Duration? Sudden onset? Relevant event? Patchy?
- What has changed?

Hallucinations? Delusions? Sleep disturbance? Fluctuation in awareness? Gait?

Personality change? Disinhibited? Apathy? Less empathy? Concrete thinking?

Speech? Word-finding difficulties? Comprehension difficulties?

Mental health? Depression? Anxiety? Psychosis?

Cognitive testing- GPcog., ACE III, Mini-ACE III, Mini Cog

Ecog (give to partner / carer) looks at functional change (needed to differentiate mild cognitive impairment from dementia)

ASK ABOUT THE RISKS

- What is particularly problematic for them? And their family? – repetition, apathy, aggression etc etc
- Any misrecognition
- Getting lost? At home? When out?
- Wandering – where do they go? How have they got back?
- Continence
- Mobility/falls
- Getting dressed
- Self care – washing/shaving
- Making meals – safe/fire risk
- Smoking
- Driving
- Managing money – lasting power of attorney?, Court of Protection?, Testamentary Capacity?
- Shopping
- Aggression/irritability
- Remembering to take medication

CASE HISTORY A

75 year old retired man

Family report him as becoming more forgetful over the last 3 years – insidious decline, can't retain any new information, repetitive, LTM less affected but can't always remember how many children he has

Unaware of his deficits, and seems quite happy with how things are

Recent difficulty with cooking, gardening, managing money and family have gradually had to support him more with this

Speech rather superficial and empty with some word finding difficulties

Preserved social skills but more apathetic and irritable

No change in personality, mobility or handwriting.

ALZHEIMER'S DISEASE

- Alzheimer's disease is the most common form of dementia
- Approximately 700,000 people in the UK have dementia. Estimated to increase to 1.4 million by 2038
- Cost of looking after people with dementia is going up; currently £17bn, over £50bn by 2038
- Estimated that at least 15 million people are affected worldwide

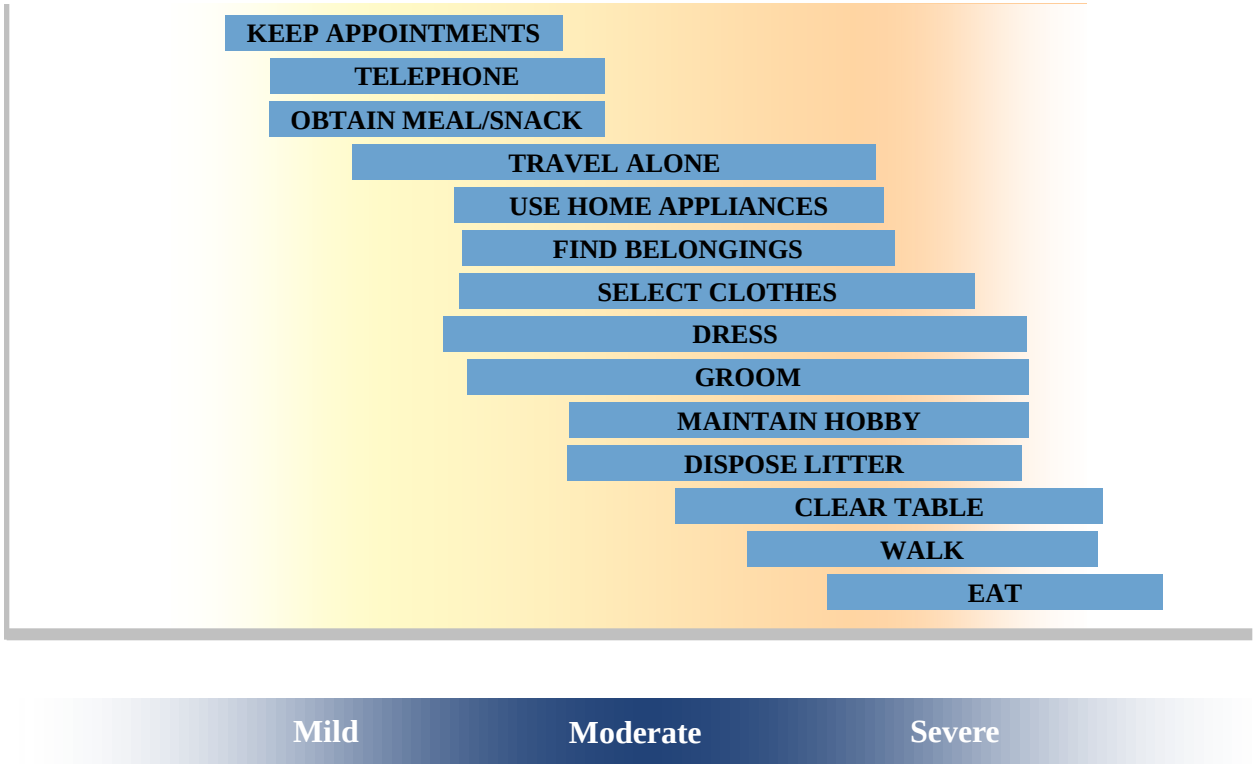
MEMORY CHANGES IN EARLY AD COMPARED TO NORMAL AGEING

Someone with AD symptoms	Someone with normal age-related memory changes
Forgets entire experiences	Forgets part of an experience
Rarely remembers later	Often remembers later
Is gradually unable to follow written/spoken directions	Is usually able to follow written/spoken directions
Is gradually unable to use notes as reminders	Is usually able to use notes as reminders
Memory loss interferes with activities of daily living	No significant impairment of activities of daily living
Is gradually unable to care for self	Is usually able to care for self

EARLY FUNCTIONAL SIGNS OF AD

- An important sign of AD is change over time in levels of function
- People have reduced levels of functioning years before clinical diagnosis of AD
- As AD progresses, ability to carry out every day functions reduces

NORMAL ACTIVITIES WORSENS AS AD PROGRESSES



POINTERS TO LOOK OUT FOR - AD

Slow gradual onset and progression

Forgets having been asked to remember items – even with prompting

Lack of insight – no problem with me!

Global impairment across multiple domains of cognitive assessment

May not be able to draw a clock, but may be able to copy one

Medial temporal atrophy on CT scan

CASE HISTORY B

86 year old lady who lives alone with some support from her family

Aware that her memory has been less reliable since a Right total hip replacement 6 years ago and sometimes she has more muddled and vague episodes

Family report that her memory can vary, some days it's pretty good, other days not so good, but that some parts of it are unchanged

Overall they feel that her memory has gradually declined over the last 6 years.

More apathetic at times, but able to cook her own lunch (now always fishfingers), get washed and dressed (always in the same clothes), manage her own finances (but family have set up direct debits for all the bills, and do her shopping for her).

No change in personality but much more apathetic – just sits and watches TV most of the time, but will dog sit for her daughter's small dog

No other symptoms – no misrecognition, getting lost, risks, hallucinosis, change in mobility etc.

History of copd and aortic stenosis

VASCULAR DEMENTIA

Less common in it's pure form – usually mixed with AD – so prescribe!

Embolic aetiology usually so risk factors are

hypertension

Cigarette smoking

Heart disease

Hyperlipidaemia
alcohol consumption

May see lots of infarcts and ischaemic changes on scans – but these can occur with AD too!

POINTERS TO LOOK OUT FOR – VASCULAR COGNITIVE IMPAIRMENT

An acute onset eg after a stroke, or other less obvious cerebrovascular event, followed by a STEPWISE decline. May have variable deficits.

May see gradual decline too however, can present with apathy (can misrecognise as depression but sleeping and eating well, and remains interested in things, just unmotivated to participate)

Patchy deficits with areas of preserved function on cognitive assessment – better function in areas not affected by damage – can surprise you! May remember items some minutes after you've moved on. Remembers being asked to remember items.

Personality is often preserved until late

Depression and anxiety is often prominent – so treat as usual with antidepressants (may need bigger doses due to vascular damage to the brain)

Insight is often intact – I know my memory isn't so good. Risk of suicide.

CASE HISTORY C

64 year old builder

Denies any problems

Family report 1-2 years of progressive personality change – he is less motivated, has erratic mood swings, lacks empathy about wife's diagnosis of breast cancer, tendency to crack infantile jokes and make suggestive comments to female shop assistants

His memory is good

Well orientated to time and place

Normal speech

FRONTAL LOBE DEMENTIA

Basically three main syndromes (although overlap syndromes exist)

- Behavioural variant – which can be apathetic or overactive
- Nonbehavioural (language) variant – primary progressive aphasia
    - Progressive non-fluent aphasia – effortful, non-fluent speech
    - Semantic dementia – asks the meaning of familiar words, fluent empty speech, generalities

Often younger

More often have a family history (in 50%)

Slow and insidious onset – may present initially with psychotic or depressive symptoms

Watch out for frontal syndromes – as part of chronic psychiatric illness, alcohol related cognitive impairment, vascular damage affecting the frontal lobe, advanced AD. This may not indicate FTD.

POINTERS TO FTD

FEATURES

- **Repetitive behaviours, more obsessional**
- **Disinhibition – reduced social awareness, Lack of judgement**
- **Lability of affect**
- **Shallowness, lack of empathy**
- **Apathy**
- **Inappropriate jocularity**
- **Reduced spontaneity of language**
- **Incontinence**
- **Primitive reflexes**
- **Memory loss is variable**
- **Change in appetite**
- **Distractible**
- **Agnosias and dyspraxias are less common**
- **Change in sensitivity to pain or temperature**

CASE HISTORY D

- 67 YEAR OLD RETIRED SHOP ASSISTANT
- C/O PROBLEMS WITH MEMORY AND CONCENTRATION
- 6/12 H/O RAPID DECLINE IN MEMORY FROM NORMAL BASELINE, TOGETHER WITH CHANGE IN PERSONALITY (MORE APATHETIC AND DISINTERESTED)
- MORE IRRITABLE
- APPETITE REDUCED, SLEEP DISTURBED (EMW)
- DENIES FEELING DEPRESSED BUT SAYS HE CAN'T CONCENTRATE VERY WELL ALTHOUGH THIS IMPROVES AS THE DAY GOES ON
- WELL ORIENTATED TO TIME AND PLACE

OVERLAPPING SYMPTOMS OF DEPRESSION AND AD

- Depression and AD can be hard to distinguish because of overlapping signs and symptoms[1]
- Neuropsychological deficits can be present in both but will improve with treatment of depression!
- Overlapping signs can include[1-3]
  - Apathy
  - Disturbance in concentration
  - Loss of interest
  - Social withdrawal
  - Self-neglect
  - Irritability
  - Anxiety

DEPRESSION VS DEMENTIA

Depression -

Relatively acute onset

Low mood (+ psychomotor retardation in some)

Biological symptoms

Diurnal variation in cognitive impairment

Past or family psychiatric history

CASE HISTORY E

- 78 YEAR OLD MAN
- FAMILY REPORT HIS MEMORY IS SOMETIMES LESS GOOD, BUT NOT CONSISTENTLY. SOMETIMES HE APPEARS QUITE VAGUE, AT OTHER TIMES HE IS QUITE ALERT.
- HOWEVER THEY HAVE NOTICED HE IS STRUGGLING TO SHOP, AND RECENTLY GOT LOST ON THE WAY TO THE NEWSAGENT (HE HAS BEEN VISITING THE SAME SHOP FOR THE LAST 50 YEARS)
- HE HAS BEEN LESS STEADY ON HIS FEET AND FELL IN THE GARDEN – HE WAS SURE HE HAD SEEN SOME CHILDREN IN THE GARDEN AND WENT OUT TO INVESTIGATE. GENERALLY HE SEEMS TO BE A LITTLE STIFFER, BUT HE DOES SUFFER FROM ARTHRITIS AND THE FAMILY ARE WONDERING IF THIS IS THE PROBLEM.
- HE IS OTHERWISE WELL, BUT HIS ELDERLY WIFE REPORTS THAT HE HAS BEEN KEEPING HER AWAKE AT NIGHT THRASHING AROUND IN HIS SLEEP.

LEWY BODY DEMENTIA

Is this a different disease from dementia associated with Parkinson's Disease?

Often a question of which came first

FEATURES

- dementia often looks like Alzheimer's type but you may get a clue with the clock drawing task

- MORE LIKELY TO HAVE

  - Confusional states which fluctuate leading to fluctuating impairment

  - Hallucinations – which are recurrent, vivid, usually visual and well formed

  - Memory loss may be very mild or variable

  - Motor disorder – mild stiffness, rigidity, slowing of movement, gait disturbance, mask like face

  - REM behaviour sleep disorder

IT'S IMPORTANT TO TRY AND IDENTIFY THESE PATIENTS AS THEY ARE VERY VERY VERY SENSITIVE TO NEUROLEPTICS, SO AVOID THEM IF YOU CAN.

INVESTIGATIONS

Laboratory investigations include:

- **FBC, B_{12}, Folate, U&E, LFT's, Ca, Glucose, Cholesterol, TFT's, CRP, (syphilis serology?)**

Neuroimaging:

- **CT scan on all patients – I know there are problems at the moment!**
- MRI, SPECT, DAT when appropriate

Other investigations - as clinically indicated

- ECG, Peak Flow, CXR, EEG

COGNITIVE ASSESSMENT

Conscious level – alert, fluctuating,

Validated cognitive assessment (lots available)– don't just focus on the score, try to get a sense about how the person approached the task, put it in the context of their premorbid ability

(Frontal lobe battery – may be part of the test or done separately)

General knowledge – how aware are they of history, current events, what's happening in Emmerdale?

Autobiographical knowledge - ?confabulates, ?aware of important dates – marriage, date of birth, children's names

COGNITIVE ASSESSMENTS

MMSE

AMT

6-CIT

Felix Post

ADAS-COG

ACE-R

Mini ACE

TYM

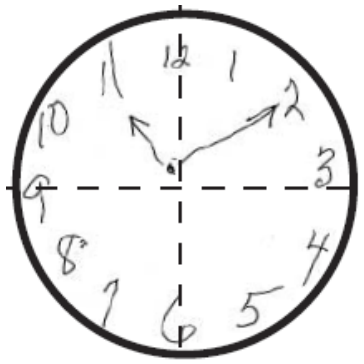
MOCA

CLOX 1 and 2

etc etc etc

THE CLOCK DRAWING TEST (CDT)

- The CDT is a simple tool that can be used to test for AD in primary care
- The test takes only a few minutes and has high sensitivity for detecting cognitive difficulties and may provide a pointer towards some of the other dementias
- The person undergoing testing is asked to:
  1. Draw a clock
  2. Put in all the numbers
  3. Set the hands at ten past eleven
- Several scoring systems are available
- Copying a clock may provide additional info



1 point for circle

1 point for numbers 1-12

1 point for correct distribution within the circle

1 point for hands placed correctly

WHAT DOES THE TEST ASSESS?

Visuospatial and executive function

Elements of praxis, concentration, memory, planning and orientation involved in the task

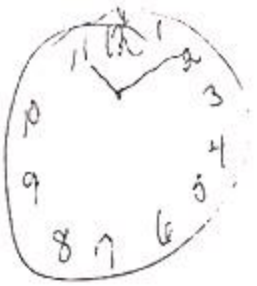
Use in collaboration with other cognitive tests

Can be used to measure disease progression

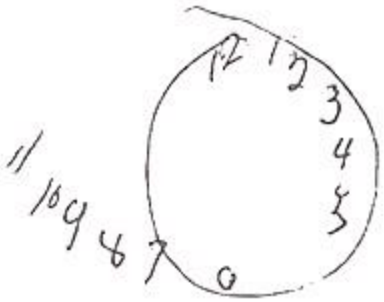
Easy to administer and can show problems even when other tests are scoring well

Examples of Clock Drawing Test

Early Alzheimer's Disease

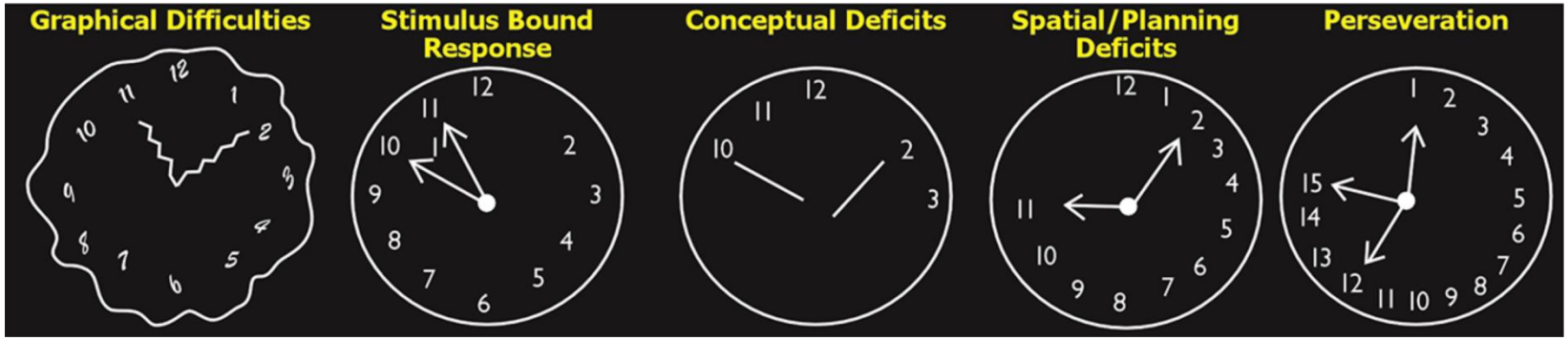


Moderate Alzheimer's Disease



Severe Alzheimer's Disease

VARIOUS TYPES OF ERRORS



ADDED EXTRAS

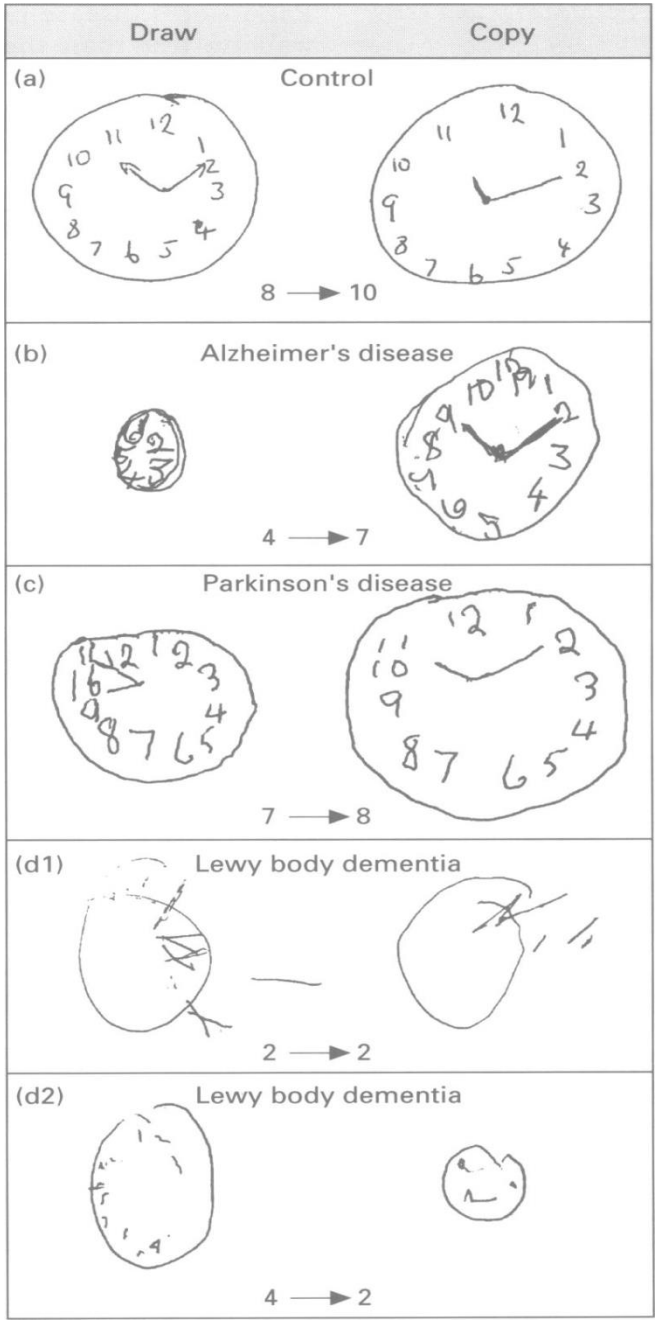
Look for hemi neglect - ?VaD

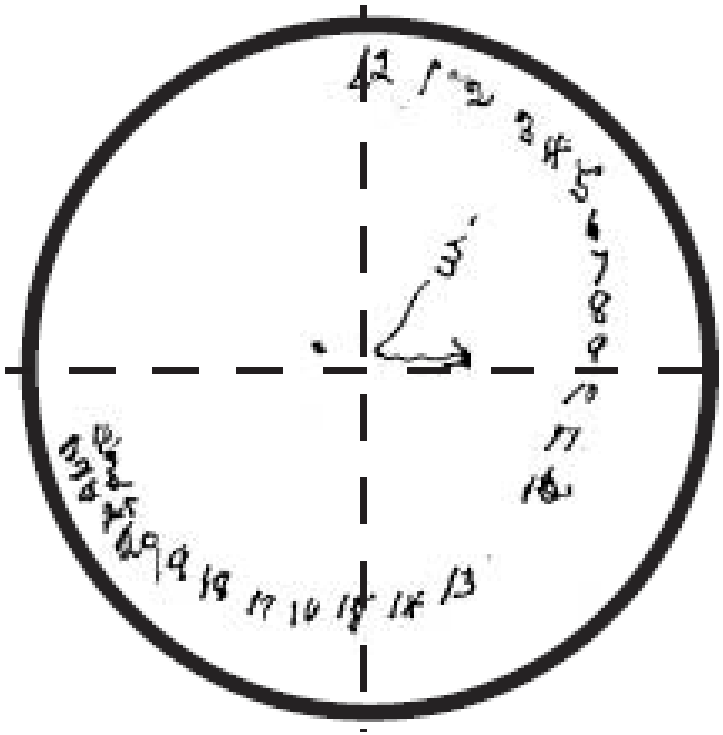
Micrographia - ?PDD,

Perseveration? - ?FTD

If they can't draw can they copy? If not ?LBD

FTD – generally score better, fewer visuospatial errors, fewer stimulus bound responses, fewer conceptual deficits


Draw
Copy
(a)
Control
8 → 10
(b)
Alzheimer's disease
4 → 7
(c)
Parkinson's disease
7 → 8
(d1)
Lewy body dementia
2 → 2
(d2)
Lewy body dementia
4 → 2



OVERALL MANAGEMENT STRATEGY

- Promote exercise, healthy diet and lifestyle:
    - Minimise risk of adult onset diabetes, obesity and high blood pressure (BP)
- Promote social interaction and emotional wellbeing
    - Avoid stress, anxiety and depression
- Promote healthy sleep habits
- Promote mental agility
    - Avoid mental stagnation (e.g. enrol in social activities at the nearest day centre)
- Avoid smoking and excessive alcohol consumption
- Control comorbid conditions
- Consider memory enhancing medication if appropriate
- Non-stat organisations
- Consider referral to social services

SPECIFIC MANAGEMENT I

ALZHEIMER'S DISEASE

- **Cholinesterase inhibitors – donepezil, galantamine, rivastigmine**
  - Watch pulse (must be >60), peptic ulceration, epilepsy, resp problems
  - SE – nausea, agitation, headaches, insomnia, irritability etc etc
- **Memantine**
  - Watch blood pressure, epilepsy, pulmonary embolism
  - SE – drowiness, sedation, impaired mobility, seizures, hypo or hypertension, constipation
- **General healthy living advice**

VASCULAR DEMENTIA

General advice

Treat cardiovascular risk factors

MIXED VaD/AD

Treat as for AD

SPECIFIC MANAGEMENT 2

FRONTOTEMPORAL DEMENTIA - REFER

LEWY BODY DISEASE - REFER

ANYTHING ELSE – REFER

if the presentation is young, refer to neurology to exclude something more unusual which will require full neuro workup including LP, antibody levels Etc (also consider this if the presentation is unusual at any age)

WHO CAN I ASK IF I'M NOT SURE?

All areas of Bristol have a specialist Dementia Wellbeing hub with Specialist Dementia practitioners and navigators to support you and your service users.

Each practice has a named practitioner and navigator who will meet with you regularly to discuss any cases you want to raise, or you can ring your practitioner up for a discussion about any service users you are not sure of, or who is already in the service.

If the history is not typical, or if patients don't fit an obvious pattern of dementia (and you have excluded any other possible cause for their memory difficulties) then refer in for a specialist opinion.

GIVE DWS A RING OR DISCUSS AT THE NEXT MEETING WITH YOUR DEMENTIA PRACTITIONER AND NAVIGATOR!