DEMENTIA **MEDICAL APPENDIX**

1. Various attempts have been made to formally classify psychiatric disorders, the two major systems being:

2. The **ICD-10 Classification of Mental and Behavioural Disorders** (World Health Organisation, Geneva) is part of the 10th edition of the International Classification of Disease. This appendix follows the common abbreviation of **ICD-10**. It is the international system used by the majority of clinical psychiatrists in Great Britain.

3. The **Diagnostic and Statistical Manual of Mental Disorders (fourth edition)** (American Psychiatric Association Washington DC). References to it in this appendix follow the common abbreviation of **DSM-IV**. It is a system devised mainly by and for workers in the USA, however UK psychiatrists were consulted in its formulation.

4. The two systems above have been in existence for many years but only in their current editions have they been closely comparable.

5. This appendix discusses some of the more significant features of normal ageing and then goes on to discuss the generally recognised clinical features and aetiology of the group of conditions which feature dementia as the main symptom. The appendix is generally based on the ICD-10 system with any major comparisons and distinctions with DSM-IV being discussed where relevant. The ICD-10 codes (numbers usually prefixed with F) are also provided.

Normal Ageing

6. With increasing age the brain decreases in weight by 5% between the age of 30 and 70, 10% by the age of 80 and 20% by the age of 90. The ventricles also gradually enlarge and the meninges become thickened. There is a reduction in the number of nerve processes and senile plaques increase, these being found in 80% of people over 70. In a smaller number there are neurofibrillary tangles and granulo-vacuolar degeneration. Ischaemic lesions are found in half of normal people over 65.

7. Tests of intellectual function show a general decline, however this varies considerably between individuals. Deterioration of short-term memory is also found in normal ageing this often being referred to as "benign senescent forgetfulness" or "age related cognitive decline". There is also a slowing of response, this appearing to be due to central rather than peripheral changes. Accompanying these changes there are important alterations in personality and attitudes such as increasing cautiousness, rigidity and a tendency to disengage from the outside world.

DEMENTIA

Definition

8. Dementia is a symptom of many different disease processes, usually of a chronic or progressive nature and is generally characterised by impairment of higher cortical functions including memory, thinking, orientation, comprehension, calculation, learning capacity, language and judgement. There is also deterioration in emotional control, social behaviour and personality. These changes must occur in the absence of impairment of consciousness.

DEMENTIA IN ALZHEIMER'S DISEASE F00

9. This is a primary degenerative disease with characteristic neuropathological and neurochemical features. It usually develops over a number of years, this period being as short as 2 years but more commonly, considerably longer.

10. The term "Alzheimer's disease" was originally used to indicate a pre-senile dementia. However it was recognised that the same clinical picture with similar neuropathological findings could be seen in those over 65: the condition was then termed "senile dementia, Alzheimer type". The current nomenclature is "Dementia in Alzheimer's disease with early onset" (when symptoms first occur before the age of 65) and "Dementia in Alzheimer's disease with late onset" (when symptoms begin after the age of 65).

11. When the disorder begins before 65 years the course is often more rapid with prominent features of parietal and temporal lobe damage, ie dysphasia, agraphia and dyspraxia. The later onset type is often much more slowly progressive with memory disturbance the most prominent feature. The mood state is very frequently one of anxious depression, sometimes accompanied by distressed agitation.

Pathology

12. There are characteristic changes in the brain including a marked reduction of neurons (especially in the hippocampus, substantia innominata, locus coeruleus and the temporo-parietal and frontal cortex), neurofibrillary tangles, neuritic plaques (largely progressive areas of amyloid, however non-amyloid areas also exist) and granulo-vacuolar bodies.

13. Neurochemical changes have also been found, in particular abnormalities in acetyl choline and choline acetyl-transferase.

Aetiology

14. The cause of Alzheimer's disease has not yet been identified. However the presence of a stronger family history in the early onset type has been noted and there is a possibility of abnormal genes on chromosomes 14 and 1. Patients with Down's syndrome also have a high risk of developing Alzheimer's disease.

VASCULAR DEMENTIA F01 (formerly termed arteriosclerotic dementia)

15. Dementia may occur when the blood supply to the brain is impaired. It is distinguished from dementia in Alzheimer's disease by the history, including onset, clinical features, and subsequent course and the presence of other manifestations of atherosclerotic disease, however this is not an invariable finding.

Vascular dementia of acute onset F01.0

16. This usually develops rapidly after a succession of strokes following cerebrovascular thrombosis, embolism or haemorrhage.

Multi-infarct dementia F01.1

17. This is a gradually developing picture of dementia sometimes showing a stepwise progression as the brain suffers a series of relatively minor ischaemic episodes. Typically there is a history of transient ischaemic attacks characterised by brief impairment of consciousness, fleeting limb weakness or visual loss.

Subcortical vascular dementia F01.2 (including Binswanger's encephalopathy)

18. In this form of dementia the sub cortical structures ie the white matter is affected, this being visible on CT scanning. The cortex is usually preserved and the clinical picture more closely resembles the dementia in Alzheimer's disease.

Aetiology

19. The vascular disorders causing dementia may be atherosclerotic, embolic or thrombotic and the aetiology of these dementias is therefore that of the underlying vascular disorder.

DEMENTIA IN PICK'S DISEASE F02.0

20. This is a progressive dementia which usually first manifests in the 6th decade and is characterised by slowly progressing changes in character with marked deterioration in social behaviour. This is followed by impairment of memory, intellect and language ability accompanied by euphoria or apathy and occasionally extrapyramidal signs such as rigidity and tremor. The person is often very disinhibited and loses sense of socially acceptable behaviour. The mood is often one of facile hilarity and sufferers are more commonly said to be unconcerned when compared to Alzheimer's sufferers.

Pathology

21. There is selective atrophy of the frontal and, to a lesser extent, the temporal lobes. There is no increase in neuritic plaques and neurofibrillary tangles greater than expected for a person of that age. There is no alteration in the acetyl choline related system, however some studies have shown muscarinic binding sites may be reduced.

Aetiology

22. There appears to be strong genetic element in some families although the majority of cases arise sporadically. Females are more commonly affected than males (2:1) and there appears to be a geographical variation in incidence, it being much more common in certain parts of the country than in others, for example, it is almost 20 times more common in Minnesota than in New York and most cases arising in Sweden occur around Stockholm whereas most cases of Alzheimer's occur around Göteborg.

DEMENTIA IN CREUTZFELD JAKOB DISEASE F02.1

23. This is a rapidly progressive dementia with prominent neurological symptoms, notably spastic paralysis of the limbs, extra-pyramidal tremor, rigidity and choreoathetoid movements. There are variants which include combinations of ataxia, visual failure, muscle fibrillation and upper motor neuron atrophy. A high proportion of cases also exhibit a characteristic "tri-phasic" electroencephalogram.

Aetiology

24. It is a subacute spongiform encephalopathy which is presumed to be caused by a transmissable agent, possibly a "slow virus", however this entity has not yet been isolated. Post mortem brain tissue removed from a sufferer can transmit the disease.

DEMENTIA IN HUNTINGTON'S DISEASE F02.2

25. This is a dementia which occurs as part of a widespread degeneration of the brain. Huntington's disease is characterised by insidious changes in behaviour with irritability, depression and anxiety. There is also choreoathetosis (abnormal fidgeting or writhing movements). Early in the disease there is difficulty in memory retrieval and judgement: as the disease progresses more severe memory deficits occur. The brain shows structural changes due to atrophy of the striatum.

Aetiology

26. This condition, which affects males and females equally, is a result of an autosomal dominant gene abnormality on the short arm of chromosome 4.

DEMENTIA IN PARKINSON'S DISEASE and LEWY BODY DEMENTIA F02.3

27. Dementia may develop in the course of Parkinson's disease especially when the condition is more severe. It is characterised by Parkinsonian features (tremor, rigidity and bradykinesia), fluctuation of cognitive deficits, including forgetfulness, difficulty in word finding, calculation and other cognitive skills and there is a high rate of visual hallucinations (up to 50% of patients) possibly indicating temporal lobe involvement.

Pathology

28. The presence of Lewy bodies in the subcortical structures (notably the substantia nigra) has been well established in Parkinson's disease. When present in the cortex they have been thought to be the lesion responsible for the dementia associated with severe Parkinson's disease. However more recently it has been shown that nearly all Parkinson's sufferers show some cortical Lewy bodies: it appears that the concentration of the Lewy bodies determines the severity of the dementia.

Aetiology

29. Idiopathic Parkinson's disease ("paralysis agitans") is thought to be a result of degeneration of certain pigmented cells in the brain stem, particularly the substantia nigra. Some hereditary tendency is apparent with a dominant gene pattern of inheritance: 25% of carriers appear to manifest the disease. The same clinical picture may be induced by certain drugs such as reserpine, phenothiazines, butyrophenones and methyldopa. A similar syndrome was seen after a pandemic of encephalitis lethargica in the 1920s.

DEMENTIA IN HIV F02.4

30. The essential feature of the dementia in this case is a diffuse multi-focal destruction of the white matter and subcortical structures. The dementia is characterised by forgetfulness, slowness, poor concentration and difficulties with problem solving. Behavioural manifestations include social withdrawal and apathy. Physical signs include tremor, impaired rapid repetitive movements, imbalance, hypertonia and impaired saccadic eye movements (jerky sideways movements on following a moving object).

Aetiology

31. The condition is due to central nervous system infection with the human immunodeficiency virus.

NORMAL PRESSURE HYDROCEPHALUS F02.8

32. Under certain circumstances the normal flow of cerebro-spinal fluid becomes obstructed in the subarachnoid space. The CSF can leave the ventricles but is prevented from freely flowing upwards over the surface of the hemispheres for absorption in the superior sagittal sinus. The pressure of the CSF is often normal or even low. The result is a reversible dementia characterised by memory impairment, apathy (progressing to severe psychomotor retardation), difficulty thinking, reduced spontaneity and general impoverishment of psychic life. Insight is usually lost early but social comportment is usually preserved. The presenting symptom is most commonly a disturbance of gait which may eventually affect most body movements. There may be neurological features such as upper limb apraxia, sucking and grasping reflexes, tremors and nystagmus.

Aetiology

33. There may be a history of subarachnoid haemorrhage, posterior fossa surgery or meningitis but in many cases no antecedent cause can be found. More unusual causes include congenital aqueduct stenosis, basilar artery anomalies and tumours in the posterior fossa. In those without an identifiable cause the age of presentation is often in the older range ie 60-70 whereas in the others the age varies widely.

SYPHILIS F02.8

34. The "general paresis" stage of tertiary syphilis produces a dementia due to an encephalitic process which results in atrophy of the brain with accompanying hydrocephalus. Mental changes with impaired intellectual efficiency and memory lapses are apparent to others but not the patient. Eventually the dementia progresses to a severe degree and communication may be impossible. Some may be euphoric, anxious, irritable or violent. The physical symptoms are a coarse tremor of the lips, tongue and fingers.

Aetiology

35. The cause is a late manifestation of infection by treponema pallidum.

OTHER CONDITIONS CAUSING DEMENTIA

36. Various other conditions may **very rarely** produce dementia including hypercalcaemia, hypothyroidism, multiple sclerosis, polyarteritis nodosa, severe head injury, cerebral lipidoses, systemic lupus erythematosus, vitamin B12 deficiency and rare autosomal recessive genetic causes such as hepatolenticular degeneration, metachromatic leucodystrophy and the "myoclonic epilepsy of Unvericht" this being characterised by increasingly frequent attacks of myoclonic epilepsy in association with progressive dementia. The aetiology of these individual disorders is that of the underlying condition.

CONCLUSION

37. **Dementia** is a syndrome characterised by disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, language, calculation, learning capacity and judgement in the absence of clouding of consciousness. It is due to those conditions which may primarily or secondarily affect the brain. It is commonly chronic and progressive but in some cases may be reversed if the underlying cause is amenable to treatment, for example, hypothyroidism and normal pressure hydrocephalus.

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