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EDITORIAL

Dementia of frontal lobe type[1]

Primary degenerative dementias can usually be classified using one of the conventional categories such as Alzheimer's disease or multi-infarct dementia but a significant proportion of cases do not fit standard diagnoses. In recent years the existence of a dementia characterized by specific impairments in frontal lobe function has been established. This has been designated as 'dementia of frontal lobe type' (Neary *et al.* 1988) or as 'frontal lobe degeneration of non-Alzheimer type' (Brun, 1987). A recent study identified dementia of frontal lobe type (DFT) in 10% of all cases of dementia (total 156) compared with only 2·5% for Pick's disease (Brun, 1987). Neary *et al.* (1988) in a series of 138 cases of dementia, found 26 (19%) patients met the clinical criteria for DFT.

In this editorial we ask whether DFT can be identified, clinically and neuropathologically, as a neurodegenerative disorder distinct from both dementia of the Alzheimer type (DAT) and Pick's disease, and we consider the relationship of DFT to 'Diogenes' syndrome', a psychiatric diagnosis which has historically been linked to personality disorder.

The onset of DFT is usually insidious with personality change, disinhibition, social misconduct, and lack of insight, followed by apathy and stereotypy being typical clinical features (Gustafson, 1987). Neary *et al.* (1988) described how a 49-year-old housewife who had previously been pleasant, well mannered and hard working, had progressively deteriorated in her personality and behaviour over three years. She became stubborn, lacking in motivation, neglectful of self-care, personal hygiene and domestic responsibilities. Her behaviour was socially embarrassing, inappropriate, disinhibited and she spent much of the day pacing restlessly, laughing and chattering inanely.

Neurological signs are uncommon, but primitive reflexes may occur. Neuropsychological testing on the other hand can be most important diagnostically. Performance on tests of verbal fluency, sequencing and the ability for abstract thinking, attentional shifting or category formation such as the Wisconsin Card Sorting Test (WCST), is impaired by frontal lobe lesions (Milner, 1963; McFie, 1975; Hagberg, 1987; Canavan *et al.* 1989; Owen *et al.* 1990). DFT patients also showed impaired verbal fluency and performed poorly on the WCST. On this latter test, both patient groups made a high percentage of perseverative errors (Milner, 1963; Neary *et al.* 1988). More recently, tests of planning and spatial working memory have been used to assess frontal lobe function by means of a microcomputer and a touch-sensitive visual display unit (Morris *et al.* 1987, 1988; Owen *et al.* 1990). During testing, patients with frontal lobe dysfunction tended to be distractible, think concretely, and made perseverative and rule-breaking errors. Neuroradiological studies have also shown a distinctive pattern of abnormality in DFT. Regional cerebral blood flow studies demonstrated a pathological blood flow pattern and a focal reduction in flow to the frontal lobes. This method had a 90% diagnostic accuracy in patients whose disorder was confirmed at autopsy (Risberg, 1987). Single Photon Emission Tomography (SPET) scans also showed selective reduction in tracer uptake in the anterior cerebral hemispheres (Neary *et al.* 1988). In addition, Positron Emission Tomography in five patients with DFT has demonstrated a significant reduction in glucose metabolism in the anterior frontal area, indicating hypofunction (Chase *et al.* 1987). Imaging studies, therefore, provide corroborative evidence that DFT is a frontal lobe syndrome.

[1] Address for correspondence: Dr Martin W. Orrell, MRC Social and Community Psychiatry Unit, Institute of Psychitary, De Crespigny Park, London SE5 8AF.

DEMENTIA OF FRONTAL LOBE TYPE – A VARIANT OF ALZHEIMER'S OR PICK'S DISEASE?

The research so far indicates that DFT may well be relatively common. It seems unlikely however that it has arisen *de novo* over the last two decades. As dementia of the Alzheimer type (DAT) is more common, perhaps it could be simply an unusual variant of DAT. However, the early clinical features of DAT and DFT are very different, with changes in learning and memory occurring in the former (Corkin, 1982) and changes in personality and social behaviour in the latter. Moreover, early changes in personality are not common in DAT. Neurological signs are much more common in DAT, and the EEG is often abnormal in the early stages, in contrast to DFT. Genetic data also suggest they may be different disorders. Gustafson (1987) reported a positive heritability for dementia in 50% of DFT cases compared with only 30% in a reference group with early onset DAT. Significantly, in post-mortem studies, the typical pathological changes of plaques and tangles in DAT were absent in the DFT group (Brun, 1987). Given the above evidence, it is reasonable to conclude Alzheimer's disease and DFT are different disorders.

It is more difficult to establish DFT as a separate entity from Pick's disease. As Lishman (1987) pointed out, the most distinctive clinical feature of Pick's disease is a tendency to begin with changes indicative of frontal lobe damage. Thus, early changes are of character and social behaviour, including diminished drive, lack of self restraint and ill-judged social conduct. This is in marked contrast to the early changes in new learning and memory seen in DAT (Lishman, 1987; Sahakian *et al.* 1988). Neary (1988) has suggested that DFT may represent a form of Pick's disease and this view is supported by the clinical findings (Robertson *et al.* 1958). Gustafson (1987) reported only small and non-systematic clinical differences between the two conditions. Further similarities include: the average age of onset (53–56 years), the average survival (8 years for DFT, 10·5 years for Pick's) and the estimated heritability of 50% for each illness. The characteristic neuropathological changes seen in Pick's disease are 'swollen' neurons with displaced nuclei (Pick's cells), and round globular argentophilic intraneuronal inclusions known as Pick bodies (Slaby & Wyatt, 1974; Cummings & Benson, 1983) which have been shown to react with a monoclonal antibody against neurofilament proteins and antitubulin antisera (Munoz-Garcia & Ludwin, 1984). However, this pattern of neuropathology distinguishes Pick's disease from DFT which shows neither change, but only non-specific degeneration of the grey matter. Subcortical areas such as the head of the caudate nucleus can also be affected (Brun, 1987). In contrast, the frontal-lobe white-matter changes in Pick's and DFT appear to be similar (Englund & Brun, 1987), consisting of astrocytic gliosis and myelin loss. Efforts to separate Pick's and DFT through differences in neuropathology have been further complicated because not all cases of Pick's disease show the typical changes described above. This problem has even led to controversy with respect to a pathological diagnosis of the disorder. For example, Pick bodies arise in only 20 to 30%, and inflated cells in only 60%, of patients who otherwise typify Pick's disease (Jervis, 1971; Constantinidis *et al.* 1974). Because of this discrepancy Neumann (1949) proposed that Pick's disease should be divided into types I and II according to whether the characteristic neuronal changes were present or not. It was later suggested that they could even represent distinct diseases, and the term progressive subcortical gliosis (because of the extensive white-matter gliosis) was coined to delineate the type without Pick bodies or inflated cells (Neumann & Cohn, 1967). This classification has not found wide acceptance but the term progressive subcortical gliosis (PSG) remains in use (Verity & Wechsler, 1987; Morita *et al.* 1987).

IS 'DIOGENES SYNDROME' REALLY DEMENTIA OF FRONTAL LOBE TYPE?

If DFT is not a new condition it is likely that it has been previously described in the literature by another name but with a very similar clinical picture. One possibility for this is the diagnosis of senile self neglect or 'Diogenes' syndrome' (Clark *et al.* 1975), a term that has found some acceptance internationally (Klosterkotter & Peters, 1985). This was originally called 'senile breakdown' by Macmillan & Shaw (1966) whose classic study describes a series of elderly patients

presenting with self-neglect, often in the absence of any obvious psychiatric illness or intellectual decline. Abandon of accepted standards of behaviour frequently occurred, and a history of heavy drinking was not uncommon (although only 4% could be defined as alcoholic). The previous personality was described as hostile and quarrelsome, but because of the other findings it seems that the personality deteriorated over a period of one to ten years. Prior to their admission to hospital many had rejected offers of social support and other help. In the remaining patients, a variety of help and services had been quite unable to cope. The later study by Clark *et al.* (1975) examined the typical personality in more detail, describing the group of Diogenes' patients as more aloof, suspicious, aggressive, and with a 'tendency to distort reality'. All were of average to high IQ, poverty was not evident, and many had had business or professional careers and good family backgrounds. Because it appears to affect such people, who have been previously stable, it has been suggested that it is a reaction late in life to stress in a certain type of personality (Clark *et al.* 1975). Post (1982) viewed it as the end-stage of a personality disorder. Neither of these suggestions is compelling and to date they remain unsubstantiated. Evidence against Diogenes' syndrome being a personality disorder is that the personality appears to deteriorate, whereas it is generally thought to stabilize or even improve with age in personality disorders. Even if some psychiatric disorder is present it is not sufficient to explain the decline. However, the symptoms do bear close resemblance to those of frontal lobe dysfunction. Irritability, aggression, paranoid ideas, lack of motivation and loss of initiative can occur in frontal lobe dysfunction, the patient may even need help and supervision to care for his or her own appearance (Lishman, 1987). By comparison, a history of drug and alcohol abuse appears to be more common in DFT (Gustafson, 1988). Indeed, a recent report (Orrell *et al.* 1989) on a case of severe self-neglect with drug and alcohol abuse but without apparent psychiatric illness, showed neuropsychological and CT changes compatible with DFT. There are no genetic or neuropathological studies of Diogenes' syndrome to our knowledge. In many ways the early stages of DFT mirror the presentation of Diogenes' syndrome. Commonly in DFT there is neglect of personal hygiene, social breakdown and personality change with lack of concern, loss of initiative and insight, and often paranoid symptoms. Gustafson (1987) describes how the insidious onset can make it difficult for close relatives to date the start of the illness. However, there may be an age difference in the onset of DFT compared to Diogenes' syndrome. Although there is some overlap, the latter is more common in the over 70 age-group whereas DFT generally has an onset between 55 and 65 years of age.

CONCLUSION

In conclusion, there is little evidence that dementia of frontal lobe type is a form of Alzheimer's disease. As has been illustrated, the clinical picture, neuropsychological, radiological and neuropathological changes are sufficiently different to define DAT and DFT as separate disorders. It is far more difficult to distinguish Pick's from DFT as they may be clinically indistinguishable. The question as to whether Pick's should be divided into types I and II depending on the presence or absence of the characteristic pathology, and the alternative suggestion that it should be divided into two entities, Pick's and progressive subcortical gliosis for the same reason, further confuses the picture. The question is, are they all the same disorder? Because of the clinical similarities it seems pragmatic to group them as DFT, retaining the term Pick's disease as a diagnosis to be made by post-mortem neuropathology.

Considering the striking similarities in the clinical picture, it is possible that a proportion of those patients diagnosed as 'Diogenes' syndrome' were in fact cases of DFT. The lack of long-term follow-up and the assumption that the patients suffered from a personality disorder discouraged the collection of the necessary neuropsychological and neuropathological evidence which may have resolved this question. Bergmann (1988) and his colleagues (Orrell *et al.* 1989) first pointed out the possible link between Diogenes' syndrome and frontal lobe dysfunction. Accordingly, 'dementia of frontal lobe type' may be the appropriate diagnosis for linking this form of social breakdown to a presumptive pathology.

Self-neglect is one of the symptoms which best correlates with an admission to a psychiatric hospital (Mezzich *et al.* 1984). Marked personality change, self-neglect, social breakdown, or other signs of frontal lobe dysfunction, presenting in a middle-aged or elderly patient without obvious medical or psychiatric disorder, must raise the possibility of dementia of frontal lobe type. Early diagnosis should improve management strategies and may prevent some of the most serious consequences in the family such as divorce, economic problems and suicide (Gustafson, 1987). These outcomes might be avoided if the family and health professionals understood that the patient suffers from a dementing illness and not merely an unruly personality.

MARTIN W. ORRELL AND BARBARA J. SAHAKIAN

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