

MATTERS ARISING

Hospital outpatient clinics, a neurology audit in South Catalonia

I read your letter about outpatient practice in Bristol, United Kingdom.¹ A similar audit in our hospital has some common aspects but differs greatly in others. The results are based on 245 new outpatient referrals to the consultant neurologist in Verge de la Cinta Hospital (Tortosa) between 1 January and 30 June 1991. This is a 202 bed district general hospital in the south of Catalonia, with a referral area population of 135 000 comprising the counties of Montsià, Baix Ebre and Terra Alta.

Fifty seven per cent (n = 139) of new outpatients were referred by the different departments of our own hospital, mostly internal medicine and traumatology, and 30% (73) by general practitioners. Four per cent (11) of those attending were categorised as "urgent" in referral letters, while 26% (64) were considered "preferent", an intermediate priority category, and 70% (170) "routine". Seventy per cent of total referrals were attending for a diagnosis, 27% for drug treatment or physical therapy and 3% for both reasons. The mean waiting time for the "urgent" group was two days, median 0.6, SD 2.7, range 0-8; mean waiting time for the "preferent" group was 14 days, median 11.6, SD 9.87, range 1-51; and mean waiting time for the "routine" group was 29.6 days, median 29.1, SD 11.40, range 7-61.

The preliminary diagnoses of new outpatients were similar in the three groups, without a significant relationship between priority category and presence or absence of a definite disease at consultation. The most common diagnoses, based on ICD classification, were migraine or headache (17% of total seen), disorders or peripheral nervous system (16%), mostly entrapment neuropathy and root lesion, epilepsy (13%), vague symptoms (12%), stroke (10%), Parkinson's disease (6%) and syncope (5%).

Forty four per cent (107) of new outpatients were discharged back to their referral source, while only 2% (5) were admitted to hospital after the consultation. Forty per cent (100) received outpatient specialised investigation, for example, 52 had a CT scan, 26 an electroencephalogram and 22 electromyography.

Only one patient of the "urgent" group was admitted to hospital after consultation, while the other four were of the "routine" group, one of them with myasthenia gravis.

Some of these aspects are similar to the audit mentioned previously, for example, the predominant diagnostic role of the neurological consultation, the main diagnoses, the small number of people admitted to hospital after consultation and some cases of inappropriate priority classification, but other results are very different. Our waiting time seems quite acceptable for the three priority categories, and this is probably the reason of the proportionately larger number of people in each a category and the small number considered "urgent". The short waiting time is explained by the number of sessions

devoted by the consultant to attending the outpatient clinic.

Another point is the high number of patients receiving specialised investigation. The main reason for this result is the highly restrictive criteria for hospital admission because of the problem of a shortage of beds. The great number of disorders of peripheral nervous system accounts for the EMG studies.

In conclusion, the results of this study are as follows: 1) A predominant diagnostic role of the neurological outpatient consultation; 2) A small proportion of patients with serious disease; 3) An acceptable waiting time, and 4) The considerable number of patients receiving specialised investigations.

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1 Wood VA, Hewer RL, Campbell MJ, Colley JRT. Hospital outpatient clinics: an efficient specialist service? A neurology audit. *J Neurol Neurosurg Psychiatry* 1991;54:370-1.

The Brain in schizophrenia

The excellent editorial by Ron and Harvey¹ notes that "to have forgotten that schizophrenia is a brain disease will go down as one of the great aberrations of twentieth century medicine". However, I think it is open to question as to whether schizophrenia can be considered as a brain disease in the same way as established brain diseases such as vital or atrophic disorders of the CNS. There may be more general reservations with the validity of the concept of schizophrenia itself,² but I have four specific reservations with calling schizophrenia a brain disease:

- 1 Unlike most brain diseases, there is as yet no diagnostic pre- or post-mortem biological or other physical marker for schizophrenia.
- 2 Compared with most brain diseases, there is no predictable pattern of deficit in sensory or motor functions or in "primitive" reflexes.
- 3 Unlike most brain diseases, psychological or psychosocial variables play a significant part in the aetiology and stability of outcome of many patients with schizophrenia.
- 4 The relationship between neurobiological features of patients with schizophrenia and the pattern or severity of psychiatric disturbance is much more equivocal than in the case of analogous relationships in brain diseases.

I would therefore at present feel comfortable in calling schizophrenia a brain *dysfunction*, but I do not think there is yet sufficient evidence to call it a brain *disease*. It is possible that the term *disease*, if commonly applied to schizophrenia, may in the perception of some clinicians limit the range of viable therapeutic options.

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1 Ron MA, Harvey I. The brain in schizophrenia. *J Neurol Neurosurg Psychiatry* 1990;53:725-6.

2 Boyle M. *Schizophrenia: a scientific delusion*. London: Routledge, 1990.

Harvey and Ron reply:

Dr Kapur's reservations about describing schizophrenia as a brain disease are doubtless shared by others and seem to depend as much on how one defines "disease" as on

our current knowledge of schizophrenia. Indeed, it would be interesting to survey whether physicians, including neurologists, would include all the conditions they diagnose and treat as cerebral diseases within the limits laid out by Dr Kapur. Nonetheless, whether the term "dysfunction" or "disease" is preferred should not interfere with the logical process of defining more clearly what might be abnormal about the brain in schizophrenia. In our opinion there seems little danger of psychological and social therapies falling into neglect simply because we understand more about any underlying organic deficits; that would be treating the dysfunction rather than the patient.

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The Neuropsychological sequelae of attempted hanging

Medalia *et al*¹ have made a valuable contribution to our understanding of the neuropsychological consequences of attempted hanging. However, their use of the terms "hypoxia" and "ischaemia" may inadvertently add to the semantic confusion already present in the literature.

Specifically, according to the authors, "In circumstances other than combined cardiac and pulmonary arrest a relatively pure hypotensive or hypoxic state may occur; perhaps the best examples are cardiac arrest while intubated and ventilated during general anaesthesia (pure ischaemia) and carbon monoxide poisoning without circulatory collapse (pure hypoxia)." We find difficulty with this model. Firstly, while hypoxia may occur without ischaemia in chronic obstructive pulmonary disease or other cases of low levels of oxygen saturation, ischaemia from cardiac arrest, for example, cannot occur without rapid parallel compromise of oxygen delivery to the affected tissue. The presence or absence of ventilatory support is essentially irrelevant if there is no blood flow. That is, there is a state of "ischaemia with hypoxia." A term that is less clumsy is stagnant hypoxia,² which emphasises decreased oxygen availability due to decreased or arrested circulation. A model of the time course of stagnant hypoxia has been proposed which we have found useful to our understanding of "watershed" lesions.³ For example, while total circulatory arrest produces what has been called ischaemic hypoxia, the periods of lower but not zero blood pressure and blood flow surrounding total arrest (called oligemic hypoxia) demonstrate the particular vulnerability of watershed areas. It is during the oligemic phases that ventilation, or the lack of, may make a difference in lesion severity since without ventilation the hypoxia produced by circulatory slow down is that much greater. Incidentally, the term "oligemic hypoxia" also alerts us to reperfusion phenomena.

Secondly, it is useful to make a distinction between hypoxia produced by a lower availability of oxygen and that produced by the reduction of circulating haemoglobin as in carbon monoxide poisoning, if only because encephalopathic symptoms vary between the two aetiologies, at least in time of onset. The term for the case of haemoglobin com-

promise which helps us retain that distinction is "anaemic hypoxia."⁴

In summary, precise definition is important because, as Medalia, *et al* correctly point out, different aetiologies can result in different patterns of symptoms such as the immediate versus delayed encephalopathies associated with "pure circulatory collapse" versus carbon monoxide poisoning, respectively. We must also be alert to ways in which different aetiologies lead to overlapping pathologies and pathologies and pathophysiological consequences. The terminology which we have found helpful is stagnant hypoxia, which is divided into the states of ischaemic and oligemic hypoxias, and anaemic hypoxia. While these entities have clearly differing pathophysiological implications our knowledge of their implications for neuropsychological symptoms is as yet incomplete. There is a greater chance of completing that side of the story if lesions are identified by standard nomenclature which accurately reflects the underlying pathophysiology.

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- 1 Medalia AA, Merriam AE, Ehrenreich JH. The neuropsychological sequelae of attempted hanging. *J Neurol Neurosurg Psychiatry* 1991;54:546-8.
- 2 Brierley JB, Graham DI. Hypoxia and vascular disorders of the central nervous system. In: Adams JH, Corsellis JAN, Duchon LW, eds. *Greenfield's neuropathology*. New York: John Wiley, 1984:125-56.
- 3 Sobota WL, Evans JE, Rowe RJ. Selective vulnerability of visual-integrative functioning after cardiopulmonary arrest: overview and case report. *Int J Clin Neuropsychol* 1988; 4:145-55.
- 4 Brierley JB, Graham DI. Hypoxia and vascular disorders of the central nervous system. In: Adams JH, Corsellis JAN, Duchon LW, eds. *Greenfield's neuropathology*. New York: John Wiley, 1980:125-56.

A predominantly cervical form of spinal muscular atrophy

We have reservations about the paper by Dr Goutieres *et al*,¹ though not the concept itself. Documenting the clinical and histological findings in five infants, they describe a condition "A predominantly cervical form of spinal muscular atrophy". Cervical spinal muscular atrophy (SMA) is a real entity and has been reported previously.² It is probably due to an infarct or malformation of the lower cervical segments of the spinal cord resulting in muscle wasting and contractures of the upper extremities and normal lower limbs.

They have not mentioned any necropsy findings. They need other sources of confirmation since they attempt to describe a newly defined clinical condition. In atypical presentations such as they describe the authors should have ruled out a possible cervical hydromyelia or syringomyelia with amyotrophy or even a congenital cervical spinal tumour,³ and familial syringomyelia is well-known.⁴ For this purpose a myelo-CT or an MRI of the cervical spine should have been done. Imaging spinal areas by MRI has almost revolutionised our concepts. We have seen two children with cervical spinal pathol-

ogies, one with a dermoid tumour and one with suspected syringomyelia (unoperated upon as yet, so no tissue diagnosis is available). Both cases had symptoms mimicking SMA. In each case, MRI was the greatest help and was the most straightforward method. In conditions running a more benign course such as monomelic amyotrophy, studies with MRI or metrizamide CT have demonstrated atrophy of the related spinal cord.⁵

Muscle biopsy findings in the cases of Dr Goutieres *et al* were all consistent with "neurogenic fascicular atrophy". In the absence of giant type 1 fibres which are typical for SMA in early infancy,⁶ muscle biopsy revealed the presence of denervation, the extent of reinnervation and the chronicity of the process, but did not conclusively disclose the precise anatomical localisation of the denervating event.⁷ There is no comment on whether there were any giant type 1 fibres in the muscle biopsies.

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- 1 Goutieres F, Bogicevic D, Aicardi J. A predominantly cervical form of spinal muscular atrophy. *J Neurol Neurosurg Psychiatry* 1991; 54:223-5.
- 2 Darwish H, Sarnat HB, Archer C, *et al*. Congenital cervical spinal muscular atrophy. *Muscle Nerve* 1981;4:106-10.
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- 4 Jackson M. familial lumbo-sacral syringomyelia and the significance of developmental errors of the spinal cord and column. *Med J Aust* 1949;4:433-9.
- 5 Biondi A, Dormont D, Weitzner I. Jr, Bouche P, Chaîne P, Bories J. Imaging of the cervical cord in juvenile amyotrophy of distal upper extremity. *AJNR* 1989;10:263-8.
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- 7 Sarnat HB. *Muscle pathology and histochemistry*. Chicago: ASCP Press, 1983.

Dr Goutieres et al reply:

We appreciate the interest of Dr Yalaz *et al* in our paper. We do not think that the cases we reported had much in common with those described by Darwish *et al*. In their three patients, muscle atrophy was already present at birth and the condition remained static, in marked contrast with the progressive disease with a postnatal onset seen in our patients.

We agree that MRI of the cord is desirable but was not available to us at the time we saw these patients. We did not feel it justifiable to perform contrast myelography as the clinical features, clearly indicated anterior horn cell disease. In particular, the absence of sensory changes, diffuse abolition of deep tendon reflexes, normal sphincter function and electromyographic evidence of denervation extending to clinically normal muscles distant from the cervical region, all favour spinal muscular atrophy. The progressive extension of the disease in the form of isolated denervation and its occurrence in two siblings born to consanguineous parents are also strong arguments against a malformative, vascular or tumoural process. Indeed, the late clinical picture in our patients was very similar to that of the classic types of spinal muscular atrophy. Our aim was to draw attention to an unusual clinical variant rather than describe a new disease entity.

Defining prognosis in medical coma

I appreciated David Bates' well written editorial¹ on the process and limitations of establishing prognoses in patients with medical coma. Since we collaborated on the international non-traumatic coma study,² perhaps I may make one or two points not strongly emphasised by Dr Bates.

The editorial generally summarises accurately the international study's results but omits emphasising a dimension which may not be self-evident. That study excluded all patients whose coma did not result either from known organ failure, or from known exogenous causes such as a deprivation of oxygen supply or an excess insulin dose. All cases with self-induced coma-causing drug poisoning as well as all cases in which aetiological diagnosis was uncertain were automatically excluded. The reasons are straightforward: nearly all such patients survive intact with intensive care including some with a flat EEG and fixed pupils lasting for a day or more.

I regret that I disagree with Dr Bates in his contention that since only a study cohort of several thousand patients can reduce the theoretical error of 5% in predicting poor outcome, one cannot make decisions based on unfavourable early signs, however bad they may be. What about the 95 to 98% who, if they survive are doomed to severe disability? Most Americans are aware of the meaning of probability odds. Given 20:1 odds they might be willing to bet on a horse which had never won a race, but they certainly would not do so if they risked having to witness and indefinitely support a pained and crippled being if that was the cost of losing the bet.

Dr Bates does not consider this potentially damaging downside statistical feature anywhere in his editorial. Many Americans are becoming increasingly apprehensive about being rescued from an early death by critical care measures only to face lives permanently blighted by intractable pain, severe physical disability, cognitive impairment or some combination of all three. When we advise patients or their families on day 1, 3, 5 or later that if they continue to receive maximal care, they or their loved ones may have a 2% or 5% statistical chance of a good recovery we also tell them that continued survival also carries at least a 50-60% chance of being associated with permanent severe disability. Facing such choices, a few will say, "Please do everything, doctor". In my own experience, however, most will urge, "please be merciful—he/she couldn't stand living as a permanent cripple, much less being a hopeless burden on the family".

The humane decision of who and when to treat and for how long is necessarily delicate, difficult and sometimes painful for the physician; it is an even greater burden for the family. At least by the US Constitution, the doctor is neither the only party nor the major decider in this situation, the patient is. Evidence in this country, is that we physicians are under-fulfilling our responsibility on this critical matter.

Last year, Derek Humphrey was the author of a small monograph entitled "Final Exit", which offers direct advice on how to commit suicide for those who, for whatever reason, wish to consider that act. The book topped the national bestseller list. The accompanying New York Times news story included comments from booksellers, journalists and potential patients that implied that physicians often overtreat or unduly