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Hospital outpatient clinics, a neurology audit in South Catalonia

I read your letter about outpatient practice in Bristol, United Kingdom. A similar audit in our hospital has some common aspects but differs 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 "preferred", 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 0-6; mean waiting time for the "preferred" group was 14 days, median 1.6, 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 of 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 longer than 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.

### The Brain in schizophrenia

The excellent editorial by Ron and Harvey's note 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,


The Neuropsychological sequelae of attempted hanging

Medalia et al. 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 those with 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 anaesthetics (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 partial 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 viable than "stagnant hypoxia," which emphasizes 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 in our understanding of "watershed" lesions. For example, while total circulatory arrest produces what has been called ischaemic hypoxia, the periods of low 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 etiologies, at least in time of onset. The term for the case of haemoglobin com-
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promise which helps us retain that distinction is "anaemic hypoxia"."

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 pathophysiologically. 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 neuro-psychological 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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A predominantly cervical form of spinal muscular atrophy

We have reservations about the paper by Dr Goutieres et al., 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.1 It is probably due to underdevelopment 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 atlantoaxial region, it described at least in thirty cases and the authors should have ruled out a possible cervical hydromyelia or syringomyelia with myotrophic or even a congenital cervical spinal tumour, 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 pathologies, one with a dermoid tumour and one with suspected syringomyelia (unoperated upon yet, so no tissue diagnosis is available). Both children 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 monomeric amyotrophy, studies with mRNA or meritizamide CT have demonstrated atrophy of the related spinal cord.

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, muscle biopsy revealed the presence of denervation, the extent of reinnervation and the chronology of the process, but did not conclusively disclose the precise anatomical localisation of the denervating event. There is no comment on whether there were any giant type 1 fibres in the muscle biopsies.

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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 described had at least in any way at all, a condition 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 findings extending to clinically normal muscles distant from the cervical region, all favour spinal muscular atrophy. The progressive extension of the disease in the face of ongoing denervation and its occurrence in two siblings born to consanguineous parents are also strong arguments against a malformative, vascular or tumoral 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's well written editorial on the process and limitations of establishing prognoses in patients with medical coma. Since we collaborated on the international non-traumatic coma study,4 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 have special relevance for our mid-Atlantic community. The 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 aetiologic 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 one study showed that 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 5% survival/long term disability which we so highly value? If they survive are doomed to severe disability? Most Americans are aware of the meaning of probability odds. Given 20:1 odds they are 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 take into account any 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 advice 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 means a 98% or 95% association 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 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 decision maker in this situation, the patient is. Evidence in this country, is that we physicians are under-fulfilling our responsibility on this critical matter.

In September of last year, Derek Humphry was the author of a small monograph entitled "Final Exit", which offers direct advice on how to commit suicide for those who, for whatever reasons, wish to consider it. The book topped the national bestseller list. The accompanying New York Times news story included comments from bookellers, journalists and potential patients that implied that physicians often oversanct and unduly