Matters arising

promise which helps us retain that distinction is “anaemic hypoxia.”

In summary, precise definition is important because, as Medalia, et al, point out, different aetiologies can result in different patterns of symptoms such as the immediate versus delayed encephalopathies associated with “pure circular collapse” versus carbon monoxide poisoning, respectively. We must also be alert to ways in which different aetiologies lead to overlapping pathologies and pathologies and pathophysiologies. The terminology which we have found helpful is stagnant hypoxia, which is divided into the states of ischaemic and oligemic hypoxia, and anaemic hypoxia. While these entities have clearly differing pathological 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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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 have a neurological syndrome 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 extent 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 fates, clearly indicated anterior horn cell disease. In particular, the absence of sensory changes, diffuse abolition of deep tendon reflexes, normal sphincter function and electromyographic changes extending to clinically normal muscles distant from the cervical region, all favour spinal muscular atrophy. The progressive extension of the disease in the frontal eye field depression and its occurrence in two siblings born 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 editorial on the process and limitations of establishing prognoses in patients with medical coma. Since we collaborated on the international non-traumatic coma study, 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 be of particular relevance to prognosis. 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 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 small subset 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 cases where we know that 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 mention the very damaging downstream 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 confers a 50%–60% likelihood of becoming 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 have lived as a permanent cripple, much less being a hopeless burden on the family”.

The humane decision of who and when to treat and discontinue treatment is 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.

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 it. The book entered the national bestseller list. The accompanying New York Times news story included comments from booksellers, journalists and potential patients that implied that physicians were increasingly unduly