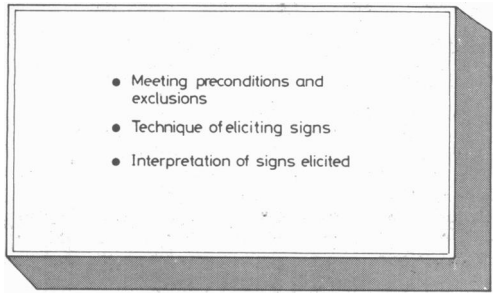
ABC of Brain Stem Death

CHRISTOPHER PALLIS

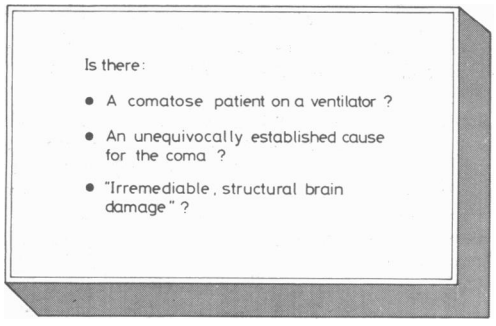
PITFALLS AND SAFEGUARDS

Pitfalls in diagnosis



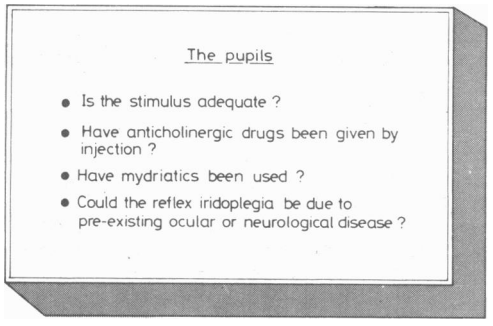
Criteria of brain stem death can be judged fairly only when they are correctly applied. The basic truth that we all make mistakes should not be allowed to blur the difference between fallible physicians and fallacious criteria. Recent controversies have, if anything, re-emphasised the soundness of the criteria used in the UK code. The adoption of similar criteria in other countries points in the same direction. Nevertheless, it is necessary to be aware of potential pitfalls. These fall into three broad categories: failure to ensure that preconditions and exclusions have been met; technical errors in testing; and mistakes in interpreting observed signs.

Failure to fulfil the preconditions



Failure to fulfil the preconditions is the commonest pitfall. To recommend that "it is almost (sic) always desirable to have an aetiologic basis for the diagnosis"[1] is asking for trouble. Testing for brain stem death should not be undertaken unless the cause of the coma has been established beyond all doubt. The UK code restricts the range of acceptable causes (by specifying which conditions should be excluded). It also emphasises the importance of timing.

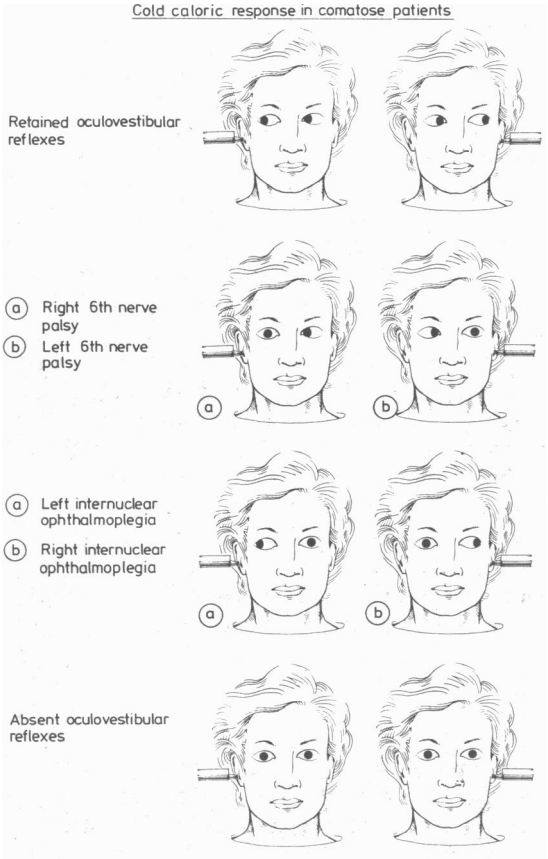
Eliciting and interpreting signs

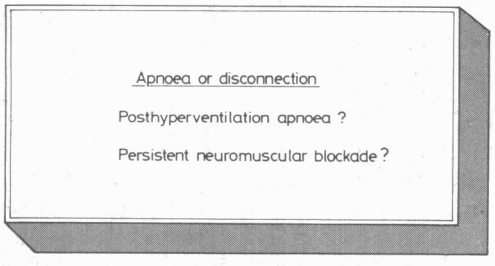


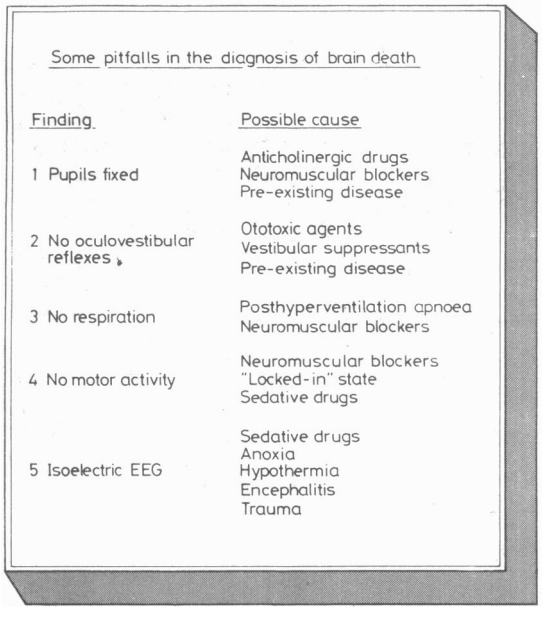
The pupils—Despite the requirements of several earlier codes, it is not necessary for the pupils to be dilated. Mydriasis is not a feature of a dead brain—as can readily be ascertained in any morgue.[2] When the brain stem is dead the pupils are usually in the mid-position. The important point is that they should show no response to light. A really bright light is needed: household torches or ophthalmoscopes should not be used as sources of light. It is also advisable to darken the room.

Widely dilated, unresponsive pupils may be caused by atropine, administered in the course of cardiac resuscitation. The effects may persist for several hours. Errors may also arise when topical mydriatics are instilled (to facilitate examination of the fundi) and the fact not recorded in the notes. Pre-existing ocular or neurological disease may account for the pupils failing to respond to light, as may local damage to the globe or nerves to the eye due to craniofacial injury.

Corneal reflexes—Testing corneal reflexes in patients with suspected brain stem death requires much firmer pressure than is used in conscious patients. Delicate dabbing with a wisp of cotton does not really provide an adequate stimulus. A sterile throat swab is more suitable.







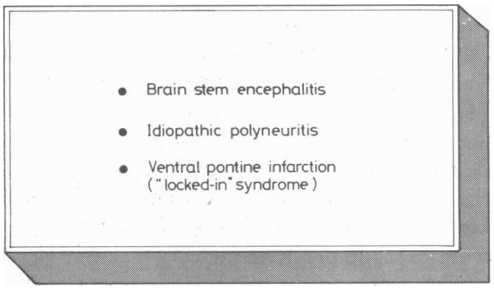
Caloric testing requires a wax-free external auditory canal, verified with an auroscope at the time of testing. The UK code recommends irrigation of the tympanic membrane with 20 ml of ice cold water. More may be used if the examiner thinks the whole volume has not been directed at the tympanum. If there is a large perforation—and the patient still has a functioning brain stem—a fall in blood pressure and bradycardia may occur. The stimulus should elicit no movement whatsoever in either eye. Tonic deviation towards the irrigated side, even if confined to one eye, implies that part of the brain stem is still alive. (Deviation confined to the ipsilateral eye indicates a contralateral internuclear ophthalmoplegia, and deviation confined to the contralateral eye suggests a sixth nerve palsy on the stimulated side.) In unconscious patients there is unlikely to be any fast "corrective" phase, away from the side of the irrigated ear. Occasionally, failure to elicit ocular movement in response to irrigation of the tympanic membranes with iced water is due to end-organ poisoning—for instance, from antibiotics such as gentamicin—or to end-organ disease. The central mechanisms responsible for the oculovestibular reflexes may be impaired or suppressed by drugs, including sedatives, anticholinergics, anticonvulsants, and tricyclic antidepressants.

Apnoea—The technical aspects of testing have already been described. Interpreting the results may occasionally cause difficulties. Excessive ventilation (resulting in a low $Paco_2$) is a potentially confusing cause of reluctance to breathe. Posthyperventilation apnoea is unlikely to cause diagnostic problems if the blood gases are measured before disconnection. At the time of disconnection the $Paco_2$ should be at least 5·3 kPa (40 mm Hg). If there are no facilities for blood gas determination the patient should (as previously described) be given 5% CO_2 in oxygen to breathe for a few minutes before disconnection, to ensure an appropriate "starting" level. Patients with chronic obstructive airways disease may depend on an anoxic stimulus to respiration and may fail to breathe at what would otherwise be appropriate levels of $Paco_2$. Great care is necessary in determining brain stem death in such cases. Prolonged apnoea may very occasionally occur when neuromuscular blocking agents are given to patients lacking the appropriate inactivating enzymes. The drugs will usually have been given to assist intubation or the management of emergency obstruction. The use of the nerve stimulator to recognise this state of affairs has already been mentioned.

Motor function in the limbs may be impaired (and the tendon reflexes reduced or abolished) as a result of neuromuscular blocking drugs or of large doses of sedatives. Either or both may have been prescribed by anaesthetists "to prevent the patient from struggling against the ventilator" (the very use of such terms suggests that the patient's brain stem is alive). Inquiries should always be made as to when the last dose of such drugs was given.

In practice these pitfalls are easy to avoid, and genuine diagnostic problems are rare. They arise only when several factors contribute to the overall clinical condition—for example, when patients with severe polyneuritis sustain anoxic insults from ventilatory accidents or when patients in drug-induced coma suffer cardiac arrest. Most other problems are of the hypothetical type beloved of ingenious students—but which nature seldom produces. The number of cases of subarachnoid haemorrhage occurring in gentamicin-intoxicated tabetics with chronic bronchitis must be very small.

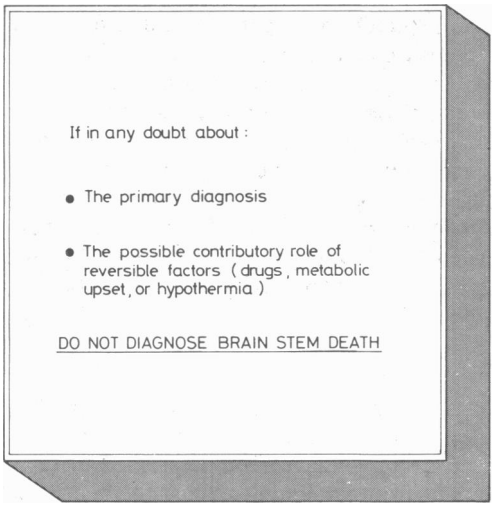
Three conditions will be mentioned, not because of any profound clinical resemblance to brain stem death but because they are repeatedly brought up in theoretical discussions of the subject.



Brain stem encephalitis[3] may result in severe external (and sometimes internal) ophthalmoplegia, facial diplegia, and bulbar palsy. The patients may be drowsy but are not in deep coma. Several brain stem reflexes could well be absent. But there is no significant motor or sensory deficit in the arms or legs, which are moved spontaneously or briskly withdrawn on stimulation. Ataxia is pronounced. Breathing is occasionally affected. The overall clinical picture is striking: "an apparently moribund patient who can use his limbs to operate suction apparatus to remove secretions accumulated in his throat."[3] In *idiopathic polyneuritis* (Guillain-Barré syndrome) cranial nerve involvement and respiratory paralysis are well recognised, although ophthalmolplegia is rare. The history will again be characteristic. *Ventral pontine infarction* may lead to the "locked-in syndrome." Bilateral lesions of the corticospinal and corticobulbar pathways render the patient tetraplegic and aphonic. Consciousness is retained, as are conjugate vertical gaze and (usually) the capacity to blink. Hearing is unaffected. The patients perceive pain normally and breathe spontaneously.

As repeatedly emphasised, the history and context are paramount in diagnosing brain stem death. Only when they are ignored, and signs of brain stem dysfunction are assessed in isolation rather than in context, can confusion conceivably arise. If the diagnosis of brain stem death is envisaged only in cases of known head injury, intracranial haemorrhage, or hypoxic encephalopathy there should be no diagnostic errors.

Safeguards for the patient



To diagnose as still alive someone who is already dead must sometimes be accepted, for a while. It is the price we pay for avoiding the opposite error. There must always be the most stringent safeguards for the patient and the benefit of any doubt must always be exercised in his or her favour.

No diagnosis of brain stem death should, in my view, be made if the physician in charge of the case still has any doubts about:

(*a*) the primary diagnosis;

(*b*) the possible contributory role of reversible causes of brain stem dysfunction (such as drugs or metabolic upset);

(*c*) the adequacy or completeness of the clinical testing.

There will be general agreement about the first two points, but argument may arise on the third. The patient may have a fractured skull base and cerebrospinal fluid otorrhoea, precluding caloric testing. Or there may be extensive facial injuries, rendering difficult the proper testing of the pupillary or corneal reflexes. More prosaically, the patient may have a glass eye. All or any of these may restrict the number of cranial nerves that can be tested. Is it permissible to diagnose irreversible loss of brain stem function on clinical grounds, on the basis of partial data? Some would argue that it is not.

Others would point out that there is sufficient redundancy in the tests to allow the diagnosis to be made, provided there is no doubt about the preconditions and provided apnoea has been rigorously confirmed. They would also emphasise that guidelines are no more than what they claim to be, that they are not edicts to be followed to the letter, that in a given context the answer is usually quite simple, and that all data—whether complete or not—should be interpreted, as in other areas of medicine, with common sense by experienced and humane physicians.

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The sixth illustration is based on a table from *The Diagnosis of Stupor and Coma* by F Plum and J B Posner (3rd ed) 1980 published by F A Davis and Co.

[1] Korein J. The problem of brain death: development and history. *Ann NY Acad Sci* 1978; **315**:19-38.
[2] Plum F, Posner JB. *The diagnosis of stupor and coma*. 3rd ed. Philadelphia: Davis, 1980.
[3] Al-Din AN, Anderson M, Bickerstaff ER, Harvey I. Brainstem encephalitis and the syndrome of Miller Fisher. A clinical study. *Brain* 1982;**105**:481-95.