Electronic Annex 2a
Full formal clinical assessment of people in prolonged disorders of consciousness

These recommendations apply to any formal comprehensive clinical evaluation of someone with a prolonged disorder of consciousness (PDOC). The purpose of this annex is to ensure that the neurological explanation of observed behaviour and responses is accurate, so that fully informed, appropriate clinical decisions are made.

The diagnostic assessment process should follow a structured approach and should consider:

- **causation**; what evidence is there relating to the reasons for the prolonged unawareness?
- **primary neurological pathways**; is there evidence that they are sufficiently intact to allow evidence of awareness to be detected?
- **awareness/responsiveness**; what is the behavioural evidence concerning level of awareness?

All investigations should be taken using normal good clinical decision-making processes, considering whether the investigation and its possible treatment consequences could alter the decision being made; if not it should not be undertaken.

Causation

It is essential to establish that there is a known cause for the PDOC, and that there are no reversible or treatable causes that are causing or exacerbating the reduced level of consciousness. Occasionally it is difficult to define the precise pathology but in those circumstances it is still important to establish that there are definite structural changes in the brain that could account for the clinical observations.

The following questions should be asked and evidence relating to the questions should be sought and documented.

What is the nature of the underlying neurological damage and dysfunction?

- Is it sufficient to cause the observed clinical state?
- Is it reversible or treatable (eg hydrocephalus or 'syndrome of the trephined') **and:**
  - is the extent of change likely to alter the decision made?
    (Because, if not, the investigation should not be carried out.)

Are there any additional or alternative causes that might account in part or in whole for the clinical state?

- drugs?
- complications of the original damage?
- another, unrelated disease?

Are any further investigations needed to establish the cause of the observed brain damage?

- These are generally only warranted for treatable or reversible factors that will alter outcome, which is increasingly unlikely for patents who remain in a prolonged disorder of consciousness more than 4 weeks after onset.

Primary neurological pathways

The level of awareness of the person is judged on the basis of their behaviours, and behaviour requires (a) some sensory input and (b) some motor output. It is important to establish that the person can receive some sensory input and can have some control over motor output.

For example, although in general primary pathways will be intact, it is always important to consider whether the person has:

- a severe critical illness neuropathy sufficient to limit motor function and/or sensory input
- spinal cord damage, which may reduce or prevent sensory input and/or the opportunities for motor output
- damage to specialised sensory organs (ears, eyes) or pathways limiting their ability to see and/or hear.

A systematic approach should be used to collect evidence, and one approach to the neurological examination is to ask the following questions.

Is there evidence that an intact primary visual pathway is present?

- Are direct and consensual pupillary reflexes to light present?
- Also consider:
  - is there any response to visual threat or very bright light?
  - does the person fixate on and track an object moving in their visual field?
  - does the person localise/look at a new object in their visual field?
- Investigations that may be used if there are no pupillary reflexes include visual evoked potentials and electro-retinograms, but these should only be used if essential.

Is there evidence that an intact primary auditory pathway is present?

- Is there a startle response or blink in response to a sudden loud noise?
- Also consider:
  - does the person wake (open eyes) in response to noise?
  - does the person localise sounds, looking towards sound?
  - are there any other behaviours obviously related to sound?
- Auditory evoked brainstem potentials may be studied if there is no response to sound, to establish whether or not the primary auditory pathway is intact.

Is there evidence that an intact primary somatosensory pathway is present?

- Are stretch reflexes present?
- Is there any response to painful stimuli (on each limb, and supra-orbitally)?
- Also consider:
  - is there any consistent response to touch or passive limb movement?
  - are there any responses to nursing and other care procedures?

- Electrophysiological measures may be used if evidence is completely lacking, and if severe neuropathy is a plausible explanation:
  - Nerve conduction studies can investigate peripheral nerves if a neuropathy is suspected.
  - Somatosensory evoked cervical or brainstem potentials can also be used to investigate primary sensory pathways, including spinal cord function.

Is there evidence that the primary motor output pathways are intact?

- Cranial nerves
  - Are there spontaneous eye movements?
  - Is there spontaneous blinking?
  - Are there spontaneous facial movements?
  - Are there facial movements in response to pain?
  - Are there spontaneous jaw movements (teeth grinding, yawns etc)?
  - Are there grunting noises, moans or other vocalisations?
  - Is saliva being swallowed (ie absence of drooling, without suction)?
  - Are there any other movements in response to pain or other stimuli?

- Limbs
  - Are there any spontaneous limb movements?
  - Are there reflex movements in response to pain, stretch reflexes, sudden noise etc?
  - Can the limbs be moved passively through a range sufficient for movement to be seen?

Is there evidence that the spinal cord is intact?

- Does limb pain cause facial or cranial movement?
- Does noise cause startle movement in limbs?
- Does facial pain or tracheal suction cause limb movement?

Hydrocephalus

In all patients with a prolonged disorder of consciousness, there will be generalised cerebral atrophy, which may be more obvious in some locations depending upon the initial damage. Some patients, for example those who had subarachnoid haemorrhage or intracranial infection, are at risk of hydrocephalus. Further investigation is only warranted if the outcome will alter management decisions, and requires formal consideration of best interests before being started.

In patients whose level of consciousness deteriorates or fluctuates to an unusual degree, the possible development of hydrocephalus should be considered. Post-traumatic hydrocephalus may develop many months after injury. If a brain scan (computed tomography (CT) or magnetic resonance imaging (MRI)) reveals ventricular dilatation, timely referral to neurosurgery should be considered for advice as to further investigation and a possible cerebrospinal fluid (CSF) diversion procedure. Units specialising in the management of PDOC should establish cooperation with a neurosurgical service that has the appropriate expertise (eg intracranial pressure monitoring, CSF infusion studies, extended lumbar drain test) and resources for managing this potentially high-risk group of patients.[1] It is not appropriate to undertake invasive procedures such as high-volume CSF removal via a lumbar puncture in a rehabilitation setting. Such interventions should always be undertaken under direct neurosurgical supervision.

Similar considerations apply to patients who might develop a syndrome of the trephined after decompressive craniotomy.[2]

Assessing level of responsiveness and awareness

The third aspect of the assessment is to determine the level of responsiveness and awareness. This depends upon observations made of behaviour and it is vital to distinguish between the actual behaviour observed and the interpretations made from or attributions placed on the behaviour.

Three types of behavioural observations may contribute:

- spontaneous behaviours, not requiring external stimuli
- behaviours occurring in response to normal incidental stimuli
- behaviours occurring when using structured, planned stimuli.

When assessing responsiveness several complementary sources of behavioural observations should be used:

- routine observations recorded within notes, made by staff; patient records should be reviewed carefully
- observations made by relatives and friends, usually obtained by questioning them but potentially also recorded by them;
    - this may include asking them to interpret their observations, in terms of degree of awareness., for example using the Wessex Head Injury Matrix (WHIM) or the JFK Coma Recovery Scale-Revised (CRS-R) as a framework for structured interview

    - many relatives video the patient, and it is always worth asking to see the videoclips they have.
- observations made specifically by trained staff using a structured assessment protocol to investigate observed behaviours at rest and level of responsiveness.

A formal structured observational assessment should always be one part of the overall assessment, but it should never be the only part (see main guidelines).

Documentation

All documentation should record observed behaviours, how often the behaviour was seen and by whom; interpretation should be given separately. If behaviours are thought to be related to external stimuli or events, the context and stimuli should be described.

- Whenever a formal diagnostic assessment is undertaken, the responsible clinician or team should document carefully and fully the evidence considered and the reasoning behind the conclusions drawn.

Notes:

- The diagnostic process is clinical, and depends upon the clinician having sufficient neurological knowledge to allow him or her to reach a conclusion.
- Investigations should not be undertaken as a routine, but if an investigation is requested, with the justification or reasoning being given, then it must be undertaken just as it would for any other patient.

References

1 Pickard JD, Coleman MR, Czosnyka M. Hydrocephalus, ventriculomegaly and the vegetative state; a review. *Neuropsychol Rehab* 2005;15:224–36.

2 Joseph V, Reilly P. Syndrome of the trephined. *J Neurosurg* 2009;111:650–2.

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