Prolonged Disorders of Consciousness (PDOC): Overview of the National Clinical Guidelines and care pathway

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Prolonged Disorders of consciousness

Following catastrophic brain injury

- Patients may hover between coma and consciousness
  - Awake, but totally unaware
    - Vegetative state (VS)
  - Awake with minimal awareness
    - Minimally Conscious State (MCS)

For some, temporary stage on road to emergence

- For others life long
  - Young patients – otherwise fit
  - May live for years – or even decades
  - Need to manage both acute and life long care
RCP National Clinical Guidelines

- Sought to bring clarity on patient care
  - From diagnosis to death
  - **Define responsibilities**
    - Guidance on challenging aspects

- Patient centred focus
  - **Decision-making**
    - Centres on what they would want
  - **Families**
    - Hold the key to that information
      - Maintain open communication
  - **Team**
    - Highly specialist clinicians
      - With experience in management of PDOC
    - Shared care with local teams
Guidelines and the Law

Clinicians and patients families

- Sometimes come into conflict
  - Attempting to do what is best for the patient
- Usually resolved by discussion
  - May require resolution by the Court

The RCP guidelines

- Do not seek to challenge the existing law
  - Lay out best practice
    - Within the existing legal framework
  - To enable clinicians to fulfill their responsibilities
    - To the patient and their family
Why new guidelines were needed

“The Vegetative State” 2003
- RCP working party report
  - Not evidence based guidelines
- Relied on by Courts

Recent developments
- Medicine
  - Minimally Conscious State
  - Clinical assessment tools
- The Law
  - Mental Capacity Act 2005
  - Health and Social Care Act 2012
Main areas covered

6 sections

1. Defining criteria and terminology
2. Assessment diagnosis and monitoring
3. Care pathway
   • From acute to long term care management
4. Ethical and Medico-legal issues
5. End of life issues
6. Service organisation and commissioning
   • Future research
Tools for implementation

Series of electronic annexes:

- Detailed advice
  - Clinical assessment
    - Optimising conditions
    - Minimum requirements for assessor training and experience
  - Clinical management

- Checklists and forms
  - Formal record of diagnosis
  - Best interests decision-making

- Leaflets for families and friends
- Template for Advance decisions
Defining criteria and terminology

Diagnostic criteria for VS and MCS
- Factors that affect prognosis and recovery
- Conditions for diagnosis of
  - ‘Continuing’ VS and MCS
  - ‘Permanent’ VS and MCS
    - From which emergence is highly improbable.

Operational parameters
- For demonstrating reliable and consistent responses
  - That indicate emergence from MCS into full consciousness
    - Expansion of parameters from the Aspen working group
      - Proposed specific autobiographic questions
Prolonged Disorder of consciousness

These guidelines relate to

- Disorders of consciousness persisting > 4 weeks
- After severe sudden onset brain injury

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma</td>
<td>Direct impact, or diffuse axonal injury</td>
</tr>
<tr>
<td>Vascular event</td>
<td>Catastrophic intra-cerebral or subarachnoid haemorrhage, stroke</td>
</tr>
<tr>
<td>Hypoxic or hypo-perfusion</td>
<td>Due to cardio-respiratory arrest or profound hypovolaemia</td>
</tr>
<tr>
<td>Infection or inflammation</td>
<td>Encephalitis, vasculitis</td>
</tr>
<tr>
<td>Toxic or metabolic</td>
<td>Drug or alcohol poisoning, severe hypoglycaemia</td>
</tr>
</tbody>
</table>

PDOG also occur as a final stage of neurodegenerative disease

Including dementia, multi-infarct syndrome etc

Different requirements in a known deteriorating trajectory

Outwith these guidelines
Terminology

‘Low awareness state’
- Some patients are completely unaware
- US Taskforce “Disorder of consciousness”

‘Vegetative state’
- Some families dislike the term - Others find it helpful
- Liege group
  - “Unresponsive Wakefulness syndrome”
    - may be responsive, albeit at reflexive level

Until an internationally agreed term emerges:

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coma</td>
<td>Absent wakefulness, absent awareness</td>
</tr>
<tr>
<td>Vegetative state (VS)</td>
<td>Wakefulness, with absent awareness</td>
</tr>
<tr>
<td>Minimally conscious state (MCS)</td>
<td>Wakefulness with minimal awareness</td>
</tr>
</tbody>
</table>

Quite distinct from ‘locked in syndrome’ or ‘brain stem death’
Pre-conditions for diagnosis of VS/MCS

- Cause of condition known
  - eg brain injury

- Reversible causes excluded:
  - Influence of drugs – eg sedative medications
  - Metabolic causes – eg hypoglycaemia
  - Treatable structural causes – eg hydrocephalus

- Careful clinical examination
  - By trained assessors
    - Expert in management of PDOC
  - Under appropriate conditions
    - Positioning, environment, medically stable etc
  - Using validated tests
    - Repeated observation over time
Structure assessment tools

Seel et al (Arch Phys Med Rehabil 2010; 91:1795-1813)

- Review of available tools for assessing PDOC (n=13)
  - 5 recommended with mild-moderate reservations
- 3 Commonly used in the UK
  - WHIM (Wessex Head Injury Matrix)
  - SMART (Sensory Modality Assessment and Rehabilitation technique)
  - CRS-R (Coma Recovery Scale - revised)

Guidelines

- One or more of these should be used during evaluation
  - Choice of tool depends on degree of certainty required for diagnosis

- Assessment should be undertaken
  - Under suitable conditions (including medical stability)
  - On at least 10 occasions, over a minimum of 2-3 weeks
  - At several different times of day
Tools

- **CRS-R**
  - Widely used in the US – less so in the UK

- **WHIM**
  - Simple hierarchical tool – 62 items
    - Can be applied by any member of the team
      - Or even by family members
    - Useful for monitoring trajectory of change over time
    - Suitable also for more generalist settings

- **SMART**
  - Most detailed and comprehensive tool
    - Tests responsiveness across 5 sensory modalities
    - Stringent training and accreditation of assessors
      - Cost and time implications
    - Useful for confirming diagnosis when high level of certainty required
Criteria for diagnosis of VS

Essential criteria

– No evidence of:

- Awareness of self or environment
- Sustained purposeful or voluntary behaviours
  – Either spontaneously or in response to stimuli
- Language, comprehension / meaningful expression

Compatible features may include:

- Spontaneous movements
  Eg chewing, teeth-grinding, grimacing, smiling, shedding tears etc
- Reflexive movements
  Eg bite, grasp, startle, swallowing, groaning, withdrawal reflex etc
- Eyes may turn fleetingly
  Eg towards sound, or to fix on a target
  But do not track movement

Incompatible features

- Evidence of discriminative perception
- Localising or purposeful actions
- Anticipatory actions
- Communicative acts

So smiling spontaneously is compatible with VS
Smiling in response to arrival of a friend/relative is not
Criteria for diagnosis of MCS

- Limited but clearly discernible evidence of:
  - Awareness of self or environment
  - Reproducible, but inconsistent behaviours

Aspen criteria (Giacino et al 2002)

<table>
<thead>
<tr>
<th>Compatible features may include:</th>
<th>Eg:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Following simple commands</td>
<td>• Crying, laughing, smiling</td>
</tr>
<tr>
<td>• Yes/no responses</td>
<td>• In response to linguistic/visual content</td>
</tr>
<tr>
<td>• gestural or verbal</td>
<td>• Vocalising / gestures</td>
</tr>
<tr>
<td>• Intelligible verbalisation</td>
<td>• In response to comments/questions</td>
</tr>
<tr>
<td>• Purposeful or discriminating behaviours</td>
<td></td>
</tr>
<tr>
<td>• In relation to environmental stimuli</td>
<td></td>
</tr>
<tr>
<td>• Not due to reflexive activity</td>
<td>• Reaching for objects</td>
</tr>
<tr>
<td></td>
<td>• Moving towards location of object</td>
</tr>
<tr>
<td></td>
<td>• Sustained visual pursuit</td>
</tr>
<tr>
<td></td>
<td>• Tracking of objects or people</td>
</tr>
<tr>
<td></td>
<td>• Differential responses</td>
</tr>
<tr>
<td></td>
<td>• To preferred objects or people</td>
</tr>
</tbody>
</table>
Sub-categorisation of MCS

Bruno et al 2011

- Broad spectrum of MCS – subclassified
  - MCS-Minus – low level
    - Only non reflexive movements to noxious stimuli
    - Pursuit eye movements
  - MCS-Plus – more complex behaviours
    - Eg following commands

GDG’s view: Helpful to define upper and lower range

- Could be based on several parameters eg
  - Complexity of behaviours
  - Consistency of response
- More research is required to define these levels
  - Insufficient evidence for prognostic significance
## Emergence from MCS

Recovery of reliable and consistent responses

- **Parameters extended**

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>Operational parameters</th>
</tr>
</thead>
<tbody>
<tr>
<td>Functional use of objects</td>
<td>Generally appropriate use of at least two different objects</td>
</tr>
<tr>
<td></td>
<td>• on two consecutive evaluations</td>
</tr>
<tr>
<td>Discriminatory choice making</td>
<td>Consistently indicates the correct choice from two pictures</td>
</tr>
<tr>
<td></td>
<td>• On 6/6 trials on two consecutive occasions.</td>
</tr>
<tr>
<td></td>
<td>• (Use at least 3 different picture pairs)</td>
</tr>
<tr>
<td>Functional interactive communication</td>
<td></td>
</tr>
<tr>
<td>Evidence of awareness of their environment</td>
<td>Gives correct Yes/No responses</td>
</tr>
<tr>
<td></td>
<td>• to 6/6 basic situational questions</td>
</tr>
<tr>
<td></td>
<td>• on 2 consecutive evaluations</td>
</tr>
<tr>
<td>Evidence of awareness of self</td>
<td>Gives correct Yes/No responses</td>
</tr>
<tr>
<td></td>
<td>• to 6/6 autobiographical questions</td>
</tr>
<tr>
<td></td>
<td>• on 2 consecutive evaluations</td>
</tr>
</tbody>
</table>
Prognosis for recovery

Updated literature review

- For both VS and MCS
  - Likelihood of significant functional recovery reduces over time
- The majority of cases will have emerged
  - From VS by 12 months
  - From MCS by 5 years
- Isolated reports of very late recovery
  - Remain profoundly disabled

Strongest determinants of recovery

- Cause of brain injury
  - Traumatic vs non-traumatic (e.g., hypoxia)
- Trajectory on serial testing
Continuing and permanent VS

Previous guidelines
- Defined ‘Persistent’ and ‘Permanent’ VS (PVS)

New terminology - ‘Continuing VS’ – instead of ‘persistent’
- Continues to remain in VS for > 4 weeks

‘Permanent VS’ – (emergence is highly improbable)
- Remaining in VS for
  - >6 months - following hypoxic brain injury
  - >1 year - following traumatic brain injury
- But take into account
  - Nature and severity of injury – and trajectory over time
  - Diagnosis can only be made when the patient is medically stable
    - Cases of genuine uncertainty may require a further period of targeted monitoring
Continuing and permanent MCS

‘Continuing MCS’
- Continues to remain in MCS for > 4 weeks

‘Permanent MCS’
- Further long term studies are required
  - Before definitive advice given on criteria for diagnosis of ‘permanent MCS’
  - The point at which emergence may be regarded as ‘highly improbable’
- Likely to depend on a number of factors
  - The duration of MCS
  - Nature and severity of injury
  - Patient’s general condition / comorbidities
  - Level of responsiveness
  - Any observed trajectory for recovery on serial testing
Interim advice

Regardless of aetiology

- A patient who has remained in MCS for 5 years
  - with no trajectory for improved responsiveness
- May be classified as ‘Permanent MCS”

Threshold for diagnosis of Permanent MCS

- May reduce to 3-4 years or even below
  - in certain circumstances which include:
  - Severe diffuse injury
    - Eg anoxic injury with prolonged ‘down time’
    - Consistently low level of responsiveness
    - No trajectory for improvement is seen on serial testing

On-going monitoring is essential

- Using structured assessment tools
Role of Hi-tech Assessment

- Hi-tech assessments
  - Imaging - fMRI, DTI etc
  - Electrophysiology – EEG etc

- Important area of research and development
  - Not yet sufficiently developed
    - For inclusion as routine clinical practice
  - Further exploration recommended
    - But only as part of an established research programme

- Reliant on behavioural assessment
Key messages 1

Consistent approach to evaluation

- Variability is a hallmark of PDOC
  - Expert observation with consistent measurement
  - Over a sufficiently long period
    - On-going surveillance

- Accurate diagnosis
  - Repeated assessment over time
    - Detailed clinical evaluation
    - Structured assessment tools
      - Systematically applied and documented
  - Trajectory of change in responsiveness
WHIM Trajectory of Change

Vegetative state

Emerging

Date of assessment

Date of assessment
WHIM Trajectory of Change 2

Minimally conscious state - stable at lower level

Minimally Conscious state - fluctuating
Key time points for assessment

The timing for assessment for a diagnosis of permanent MCS will depend on
- the nature and severity of the injury,
- the level of responsiveness and
- any observed trajectory to improved responsiveness on serial testing
Care pathway for patients with PDOC

Acute care
ITU
Neurosurgical/ orthopaedic

Hospital ward
Multi-disciplinary rehabilitation

Specialist PDOC neurorehabilitation service
In-patient admission for assessment / management of PDOC in designated centre (usually 3-4 months)

Specialised PDOC outreach support
Multidisciplinary monitoring

If DOC continues – involvement of specialist neurorehab team:
- After 96 hrs: Assessment for interim advice
- After 2 wks: Review and evaluation to eliminate treatable causes
- After 4 wks: Referral to specialist neurorehab team for PDOC management

‘Roving door’ policy if showing signs of change

‘Slow stream’ rehabilitation
Active management + ongoing assessment
In a specialist nursing home or equivalent environment
For up to 1 year post injury

Formal assessment / review
VS - At 6 months (non-TBI) or 12 months (TBI)
MCS – annual review to 5 years

If remains in VS / MCS
At formal review point.
Refer to Court of Protection for
Formal consideration of Best Interests Management

Long term care
Long term support under NHS continuing care
In specialist nursing home (or own home)

End of life care
Specialist support for end of life palliative care
Joint between Specialist DOC and palliative care

Hospital
Community

Acute Injury / illness
Tracking of patients

- There is a dearth of information
  - How many patients in PDOC
  - Long term prognosis
    - Which patients will emerge
  - Lack of consistent approach to evaluation

- Recommendation:
  - Development of a national clinical register
    - Linked to UKROC (The UK Rehabilitation Outcomes Collaborative)
      - National clinical database for specialist rehabilitation (application in process)
  - Longitudinal register for outcomes and research
    - Tracking over time – where do they go?
    - Monitoring – which patients emerge
      - For those who remain in PDOC – a concerted plan of long term care
Care pathways for assessment

- **NHSE service spec for specialist rehabilitation**
  - Includes PDOC evaluation
    - Outreach to support local care teams (in some areas)
    - Provision is still very patchy

- **Formal service designation in some regions**
  - Expertise somewhat variable
  - Inconsistent application of measures

- **Insufficient capacity**
  - Especially for tracheostomy patients

- **Specialist nursing homes gaining expertise**
  - Many now routinely using the WHIM
How often to review

- Depends on phase of recovery
  - Early on
    - 2-3 times per week
  - Later – if condition static
    - Every 1-2 weeks
  - After 6 months
    - Every 2-3 months
  - After 1 year
    - Every 6 months - annually
When to refer to specialist units

 Depends on the time scale

  - Initially
    - If in PDOC after 4 weeks
      - Or when medically stable after this
    - Initial assessment – 3-4 months

  - Later – for review
    - At critical decision-making periods
      - 6 months for VS in non traumatic BI
      - 1 year for VS in traumatic BI

  - Or when something changes
    - Start to wake up
    - Or for BI decision-making
Which patients to refer

- For confirmation of diagnosis
  - VS/ MCS

- For management
  - Seating / 24 hour positioning
  - Communication assessment
  - Tracheostomy weaning
  - Nutritional management / swallowing assessment
  - Family distress
  - BI decision-making – Court application
  - End of life care
Pain and Depression

- MCS may be a worse condition than VS
  - VS patients – unaware
    - Evidence that MCS patients experience pain
      - No differently from normal controls
    - Presumably also emotional disturbance

- Should be vigilant for symptoms including
  - Pain
  - Depression

- Guidelines provide screening tools
  - Based on behavioural observation
## Pain

<table>
<thead>
<tr>
<th>ITEMS</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>SCORE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Negative vocalization</td>
<td>None</td>
<td>Occasional moaning or groan.</td>
<td>Loud moaning or groaning. Crying</td>
<td></td>
</tr>
<tr>
<td>Facial expression</td>
<td>Smiling or inexpensive</td>
<td>Sad, frightened, frown, mild facial grimacing</td>
<td>Marked facial grimacing in response to presumed painful stimuli</td>
<td></td>
</tr>
<tr>
<td>Body language</td>
<td>Relaxed / calm</td>
<td>Tense. Fidgeting</td>
<td>Rigid. Marked tonal posturing</td>
<td></td>
</tr>
<tr>
<td>Consolability</td>
<td>No need to console</td>
<td>Distracted or reassured by voice or touch</td>
<td>Unable to console, distract or reassure</td>
<td></td>
</tr>
<tr>
<td>Physiological change</td>
<td>Normal</td>
<td>Mild increase in vital signs (temperature, pulse, BP etc)</td>
<td>Marked increase in vital signs, or sweating, flushing/pallor</td>
<td></td>
</tr>
<tr>
<td>Presence of painful conditions</td>
<td>None</td>
<td>Mild changes eg marked skin, previous healed injuries, mild contractures</td>
<td>Marked changes eg broken skin, active arthritis or heterotopic ossification, severe arthritis / contractures</td>
<td></td>
</tr>
<tr>
<td>TOTAL</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Depression

<table>
<thead>
<tr>
<th>Question</th>
<th>Yes / No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Does the patient sometimes look sad, miserable or depressed?</td>
<td></td>
</tr>
<tr>
<td>Does the patient ever cry or seem weepy?</td>
<td></td>
</tr>
<tr>
<td>Does the patient seem agitated, restless or anxious?</td>
<td></td>
</tr>
<tr>
<td>Does the patient seem withdrawn, showing little interest in the surroundings? (This may include evidence of deliberate withdrawal from interaction, eg eye closure when approached by staff)</td>
<td></td>
</tr>
<tr>
<td>(Score 1 for ‘yes’ and 0 for ‘no’)</td>
<td>Total Score</td>
</tr>
</tbody>
</table>
Summary

- Guidelines provide helpful advice
  - Tools and resources

- Shared care
  - Find your local PDOC assessment unit
    - Out-reach support
      - Develop shared care arrangements

- Familiarise yourself with common tools
  - WHIM