

Withdrawal of care and the diagnosis of death in neurological illness in children

Colin Kennedy

Clinical Neurosciences

University of Southampton

crk1@soton.ac.uk

Case 1

- 8 month old previously health boy
- Febrile for 2 days
- Rythmical jerking movements of one arm 24 hours before arrive in A&E
- Increasingly lethargic
- Presented to A&E in status epilepticus
- Mother a family practitioner

Case 1 –progress in hospital

- Day 1
 - Seizures controlled promptly with IV diazemuls and phenytoin load
 - Treated empirically with high dose IV cefotaxime and aciclovir
 - CSF: 5 wbc, glucose <0.2 mmol/l; pneumococci seen on micro
 - Moving all limbs, some visual responsiveness; awareness of voice
 - MR: initially 'wnl'; on review, patchy basal ganglia low signal

Case 1 –progress in hospital (contd)

- Day 1
 - Seizures controlled promptly with IV diazemuls and phenytoin load
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 - CSF: 5 wbc, glucose <0.2 mmol/l; pneumococci seen on micro
 - Moving all limbs, some visual responsiveness; awareness of voice
 - MR: initially 'wnl'; on review, patchy basal ganglia low signal
- Day 2
 - Visually unresponsive; flexes L but extends R limbs to pain
 - Pneumococci confirmed on culture
 - Aspirin 5mg/kg/day commenced
- Day 3
 - Motor responses as before; no other responses
 - **Mother asks for care to be withdrawn**

questions

- What can be said about prognosis on day 1?
- What is the likely interpretation of MR changes?
- What is the role of aspirin?
- What further investigations would assist in discussing with mother her request for withdrawal of care?
- Would an examination of the brain stem reflexes form part of your assessment? Why or why not?

progress

- EEG: mixed delta, normal voltage, a few spikes and sharp waves
- Repeat MR: slight maturation of previously identified perforator artery infarcts
- Mother advised that baby remains seriously ill but may make a good recovery
- Child discharged home on day 10 with mild right hemiparesis. Four months later is taking independent steps, playing peek-a-boo and identifying objects by use

Case 2

- 8 year old boy with moderate learning disability
- Known to have epilepsy with occasional status epilepticus
- Parents woken by sounds from his bedroom
- Status epilepticus. Ambulance arrived promptly. Sz not controlled by buccal midazolam or dose of IV midazolam by paramedics
- Seizure terminated in A&E with phenytoin and IV midazolam at 25 minutes after first found. Lowest recorded pH 7.04
- Transferred to PICU sedated on morphine and midazolam.

Case 2 - progress in hospital

- CT – nil focal and no evidence of raised ICP
- Occasional breakthrough seizures controlled with additional half loads of phenytoin
- Day 2, 3 sedated – no response to noxious stimulation
- EEG – low voltage delta waves
- MR –? Poor white/grey differentiation. Nil focal.

Questions

- What is the prognosis?
- Are you sure: what special investigations might help you to feel confident of prognosis?
- What would you say to the child's mother over the course of the first week in hospital?

Subsequent course

- All sedation stopped on day 3
- Unresponsive on day 4. Stable. Mother advised that prognosis for multiple long term physical and learning disabilities in addition to pre-existing
- Electrolytes, temperature normal
- Day 5: no response to testing of brainstem reflexes except some BP and pulse change to suction. Blood levels of sedatives very low.
- EEG; 'low voltage delta'; on review, virtually flat.
- BAERs: no responses after wave 1.
- Situation re-discussed with mother and support of patient's usual doctor confirmed by phone.
- Day 6: transfer to hospice local to family at mother's request. Ventilation then discontinued.
- Day 10: apnoeic. Dies peacefully at local hospice.

A code of practice for the diagnosis and confirmation of death

UK Academy of Royal Colleges
2008

Google it for the pdf!

Purpose and scope

- To make practical recommendations ... where further intervention aimed at sustaining life can be of no further benefit to patients
.. acceptable to the relatives of the deceased, society in general and also to the medical , nursing and other professional staff involved
- Important to separate completely the diagnosis and confirmation of death from anything to do with the issues surrounding organ donation and transplantation.

Definition of death

- ‘Death entails loss of those essential characteristics to the existing of a living human person and, thus, the definition of death should be regarded as **the irreversible loss of the capacity for consciousness, combined with the irreversible loss of the capacity to breathe.** This may be due to a wide range of underlying problems in the body, for example, cardiac arrest.’

Question: How is consciousness assessed?

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Answer: Coma scales e.g. modified GCS for infants

| | | |
|-----------------|---|---|
| Eye opening | Spontaneous | 4 |
| | To verbal stimuli | 3 |
| | To pain only | 2 |
| | No response | 1 |
| Verbal response | Coos and babbles | 5 |
| | Irritable cries | 4 |
| | Cries to pain | 3 |
| | Moans to pain | 2 |
| | No response | 1 |
| Motor response | Moves spontaneously and purposefully | 6 |
| | Withdraws to touch | 5 |
| | Withdraws in response to pain | 4 |
| | Decorticate posturing in response to pain | 3 |
| | Decerebrate posturing in response to pain | 2 |
| | No response | 1 |

Death following loss of brain stem function

- ‘Cessation of brain-stem function... will produce this clinical state and therefore irreversible loss of the integrative function of the brain-stem *equates with* the death of the individual’
- NB important points:
 - irreversible LOC *by itself* (e.g chronic vegetative state) does not by itself entail death of the individual
 - death does *not* entail the cessation of *all* neurological function
 - there may be some residual reflex movement of the limbs after such a diagnosis and ‘these have no moral relevance to the diagnosis of death’

Death following cessation of cardiorespiratory function

- While dying is a process rather than an event, a definition of when the process reaches the point (death) at which a living human being ceases to exist is necessary
- Circumstances in which this may arise
 - Full attempts at reversal of any contributing cause
 - Resuscitation failed or withdrawn because of no benefit
- Death can be diagnosed when ... an appropriately qualified individual certifies the irreversible cessation of neurological (pupillary), cardiac and respiratory activity (= time of death)
- 5 minutes observation and confirmation of absence of pupillary & corneal reflexes and of response to supraorbital pressure

Death in a patient in coma:

diagnosis of death by neurological criteria (DNC)

- When the brain-stem has been damaged in such a way and to such a degree that its integrative functions (which include the neural control of cardiac and respiratory function and consciousness) are irreversibly destroyed, death of the individual has occurred and the heart will inevitably stop beating subsequently, although the time over which this occurs may vary considerably.*
- The appropriate course of action is then to consider withdrawal of mechanical ventilatory support, the ethical justification of which has passed, and to allow the heart to stop.
- Relatives, partners and carers should be kept fully informed by the local care team of the diagnosis, the inevitable outcome and the likely sequence of events.

*: 'This has been confirmed in all published series and has therefore been adequately validated'

Diagnosis

- Pupils fixed, no reaction to light
- No corneal reflex
- No oculo-vestibular reflexes*
- No responses within the cranial nerve distribution by adequate stimulation of any somatic area**
- No cough or gag reflex***

Only if all the above are absent, proceed to 'apnoea test':

- Absence of respiratory response to hypercarbia

*: visualize TM, head at 30 degrees, then 50 ml of ice-cold water over 1 min

** : includes supra-orbital stimulation

*** : tracheal suction; spatula to posterior pharynx

Testing respiratory response to hypercarbia

- Increase oxygen to 100%
- Check that ABG correlate with monitored values
- With oxygen sat >95%, decrease resp rate
- When PCO₂ >6kPa, check that ABG >6.0 and pH <7.40
- Maintain stable BP
- If chronic resp condition, increase CO₂ to >6.5kPa
- No resp response for 5 minutes; PCO₂ ↑ by >0.5kPa
- Reconnect ventilator to allow gradual return to normal

Repetition of Testing

- 2 medical practitioners, registered for > 5yrs,
- competent in conduct and interpretation of brain stem testing, **acting together**.
- at least one must be a consultant; no conflict of interest;
- neither may be a member of transplant team.
- two complete sets of tests i.e. only twice in total.
- no need for lengthy delay between tests.
- time of death = time of first set of tests.

Conditions necessary for the diagnosis of death

- **No doubt that patient's condition is due to irreversible brain damage of known aetiology**
- Exclusion of potentially reversible causes
 - No evidence state is due to depressant drugs
 - Primary hypothermia excluded (T>34 Celsius)
 - Reversible circulatory, metabolic and endocrine disturbances excluded **as the cause**
 - Reversible causes of apnoea (e.g. n/m block) excluded

Other tests in diagnosis of death

- Not included in criteria for diagnosis
- Consider if full neuro exam not possible (e.g. maxillofacial injury; high cord injury; residual sedation; ‘some cases of paediatric hypoxic injury’)
- ‘All such investigations are prone to artifice and each has attracted its own literature defining false negative and false positive rates’

The vegetative state (VS)

- Defined as a clinical condition of unawareness of self and environment in which the patient breathes spontaneously, has a stable circulation and shows cycles of eye closure and opening which may simulate sleep and waking
- Absence of brain stem reflexes is **not** part of the VS

Diagnosis of death by neurological criteria (DNC) in children (according to 2008 AoMRC report)

- Age >2 months: same criteria as in adults
- 37/52 to 2 months: 'rarely possible confidently to make diagnosis of death as a result of cessation of brain stem reflexes'
- <37/52: 'criteria cannot be applied'

But this is changing in the UK in 2015 – see next 3 slides

Infants aged less than 2 months

2014/15 DNC working group of RCPCH

Preconditions

- The working group recommends that the preconditions detailed for [older children and adults] should be fulfilled before diagnosing DNC:[see previous slides for detail]
- In post-asphyxiated infants, or those receiving intensive care after resuscitation, whether or not they have undergone therapeutic hypothermia, **a period of at least 24 hours of observation, during which the preconditions necessary for assessment for DNC should be continuously present**, should elapse before clinical testing for DNC. (This is an 'additional precautionary measure' to be taken in young infants.)
- If there are concerns about residual drug-induced sedation, then **this period of observation may need to be extended**

Infants aged less than 2 months

2014/15 DNC working group of RCPCH

Clinical diagnosis of DNC

- **The diagnosis of DNC can be confidently used in infants from 37 weeks gestation to less than two months of age** using the clinical examination criteria used to establish death in adults, children and older infants.
- **a stronger hypercarbic stimulus** is used to establish respiratory unresponsiveness. Specifically, there should be a clear **rise in PaCO₂ levels of >2.7kPa (>20 mm Hg) above the base line** with no respiratory response at that level. This is a precautionary measure.
- **Interval time between tests need not be prolonged** (as stated in 2008 AoMRC Report).

Infants aged less than 2 months

2014/15 DNC working group of RCPCH

Ancillary tests

- The diagnosis of DNC in young infants from 37 weeks gestation corrected age (i.e. post conceptional) and less than two months of age **is a clinical diagnosis** with certain preconditions
- **ancillary tests do not help in this diagnosis.**

APPENDIX 1

PROCEDURE FOR THE DIAGNOSIS AND CONFIRMATION OF CESSATION OF BRAIN-STEM FUNCTION BY NEUROLOGICAL TESTING OF BRAIN-STEM REFLEXES

Diagnosis is to be made by two doctors who have been registered for more than five years and are competent in the procedure. At least one should be a consultant. Testing should be undertaken by the doctors together and must always be performed completely and successfully on two occasions in total.

Patient Name: _____ **Unit No:** _____

Pre-conditions

Are you satisfied that the patient suffers from a condition that has led to irreversible brain damage?

Specify the condition:

Dr A: _____ **Dr B:** _____

Time of onset of unresponsive coma:

Dr A: _____ **Dr B:** _____

Are you satisfied that potentially reversible causes for the patient's condition have been adequately excluded, in particular:

| | DR A: | DR B: |
|-------------------------------------|-------|-------|
| DEPRESSANT DRUGS | | |
| NEUROMUSCULAR BLOCKING DRUGS | | |
| HYPOTHERMIA | | |
| METABOLIC OR ENDOCRINE DISTURBANCES | | |

| TESTS FOR ABSENCE OF BRAIN-STEM FUNCTION | 1 ST SET OF TESTS | 2 ND SET OF TESTS | 1 ST SET OF TESTS | 2 ND SET OF TESTS |
|---|------------------------------|------------------------------|------------------------------|------------------------------|
| DO THE PUPILS REACT TO LIGHT? | | | | |
| ARE THERE CORNEAL REFLEXES? | | | | |
| IS THERE EYE MOVEMENT ON CALORIC TESTING? | | | | |
| ARE THERE MOTOR RESPONSES IN THE CRANIAL NERVE DISTRIBUTION IN RESPONSE TO STIMULATION OF FACE, LIMBS OR TRUNK? | | | | |
| IS THE GAG REFLEX PRESENT? | | | | |
| IS THERE A COUGH REFLEX? | | | | |
| HAVE THE RECOMMENDATIONS CONCERNING TESTING FOR APNOEA BEEN FOLLOWED? | | | | |
| WERE THERE ANY RESPIRATORY MOVEMENTS SEEN? | | | | |

Date and time of first set of tests:

Date and time of second set of tests:

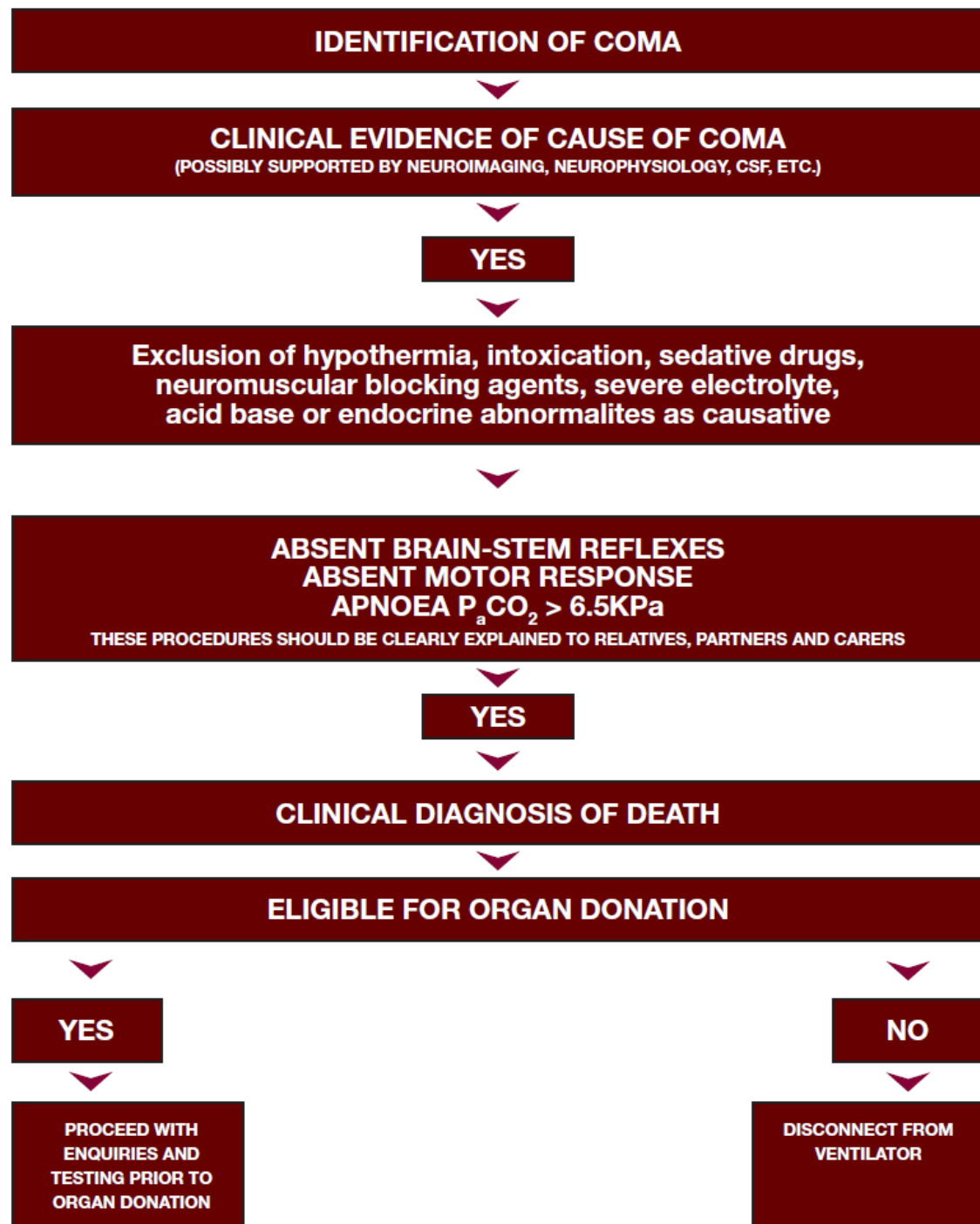
Dr A Signature:

Dr B Signature:

Status:

Status:

Diagnostic
and
management
algorithm



Summary

- Clinical circumstances require neurologists to offer advice on value or otherwise of continuing care
- The question of the diagnosis of death, which may arise in giving such advice, almost always occurs in highly charged circumstances: clarity inspires confidence.
- Clarity on how to assess neurological function in coma also has a much wider applicability in management of coma
- Read 'a code of practice for the diagnosis of death'
- Know what the practice is in your country