Guideline for the management of a child aged 0-18 years with a decreased conscious level

Appendix D

Stakeholder Groups

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1. Enrolling Stakeholder participation

All potential users of the guideline should be consulted during the guideline development phase. The comments received from these “stakeholders” will affect the implementation of the recommendations once they have been written.

An invitation was sent to the chairs of all the relevant national bodies of the potential users of the guideline. The chair was asked to be a link person between the guideline and the group they represent. If the chair was unable to perform this task, then this role was delegated to another member of that stakeholder group.

The link person was charged with commenting on draft versions of the guideline, having first sought the views of other members of their professional group. The link person was provided with all the details of the guideline development process and invited to contribute throughout. Any contributions received were raised with the Guideline Development Group and acknowledged.

2. Stakeholder groups participating during development

The following groups made representations to the Guideline Development Group:

Stakeholder group	Link representative
The Association of Paediatric Anaesthetists of Great Britain and Ireland	Dr Charles Stack
British Association of Emergency Medicine	Dr Stephen Nash
Association of Paediatric Emergency Medicine	Dr Lisa Goldsworthy
Royal College of Paediatrics and Child Health (RCPCH) British Association of General Paediatricians	Dr Andrew Boon
Paediatric Intensive Care Society	Dr Charles Stack
The British Inherited Metabolic Disease Group	Dr Graham Shortland
Royal College of Pathologist – Clinical Biochemistry	Dr Mario Marrero-Feo
Royal College of Pathologists – Special Advisory Committee on Medical Microbiology	Dr Clive Graham
Royal College of Pathologists – Special Advisory Committee on Medical Virology	Dr Marie Ogilvie
UK Clinical Virology Network	Prof. Paul Griffiths
RCPCH Nephrology Interest Group	Dr Heather Maxwell
British Paediatric Neurology Association	Dr Colin Kennedy
The Society of British Neurological Surgeons	Mr James Leggate
The British Society for Clinical Neurophysiologists	Dr Zenobia Zaiwalla
Royal College of Nursing	Fay Valentine
Royal College of Pathologists – Special Advisory Committee on Paediatric Pathology	Dr Christopher Wright
RCPCH Pathology Interest Group	Dr Michael Ashworth
Neonatal and Paediatric Pharmacy Group	Sharon Conroy
Royal College of Radiologists	Dr Kieran McHugh
The British Society of Paediatric Radiology	Dr Karl Johnson
Children Living with Inherited Metabolic Diseases (CLIMB)	Pam Davies
The Encephalitis Society	Elaine Dowell
National Reyes Syndrome Foundation	Dr John Glasgow

3. Contributions of stakeholders

Below are the contributions of the Stakeholder Groups received by the Guideline Development Group. Responses to these contributions are provided in the minutes of the Guideline Development Group meetings (Appendix C).

Comments from Association of Paediatric Emergency Medicine

have just noted down anything I spotted as I went along so please excuse any perception of abruptness.

I'd give high flow oxygen to anyone who's sick – the dkas will get it even if sats ok – I would suggest oxygen anyway and MUST be given if sats < 95%. I tend to say it improves the dissolved 0_2 so that more might be available to the shocked tissues at the peripheries etc – don't know if there's evidence to support this though.

"Papillary" in the lumbar puncture bit
Few will leap to LP anyone with reduced conscious level and no obvious cause.
Will need to reference the "no one dies and the diagnostic assistance is worth it" to get people to follow.

Shock
Prolonged cap refill but with increased heart rate? HR does not get a mention – the urine output is rarely if ever measured - have to wait an hour and that's after you've catheterised them
Beware of saying that giving fluid causes pulmonary oedema – can it be worded differently? –" if >40ml/kg given and there is no response then the child is very unwell and may need ITU and central access for inotropic support – and so consideration should be given to inducing anaesthesia and ventilating the child via ET tube. If the pathology involves capillary leakage then this will help control ensuing pulmonary oedema."

Must cross reference the LP in sepsis for contra indications – as otherwise it looks like they don't apply.

What guidance on recording cGCS? A lot of medically unwell's will be V or less unless you wake them up properly first – I know that sounds trite but there is a danger of overestimating the reduction in conscious level if taken at face value.

There's an italic line missing in HSV enceph – and the format changes on others too – if this is because some are essential and some are advisory then should that be explained?

Can write what amounts of blood in what bottles for the hypoglycaemia – if as an appendix? And which need to be frozen etc – for the frantic moment it's really helpful

We're so used to seeing abnormal lactates that aren't that bad – is there any recommendation that above a certain level is truly abnormal but lower ones may be a 'normal' response to illness?

What's the evidence for the Ca and Mg in under ones only? Anecdotal but I've certainly seen hypocalcaemic fits present in a 12 year old (years ago and I can't remember the underlying cause)
In 16 the Ix change from normal to italic – any reason?

For the flow chart – I tried to change the format of the oxygen intubation box- if 0_2 and intubation were the words in white that would be clearer. And I'd love to play with the boxes and uniformity – but couldn't work out how you had formatted the group of objects!

Well done – looks good!

Lisa G

Comments from The Association of Paediatric Anaesthetists of Great Britain and Ireland

Thank you for sending me this guideline. I have sent to a few of my colleagues here and on APA council and PICS council.

I have as yet not had many replies but I will put the collective comments below. I am effectively away from now until 11/04/05 and will therefore put any other comments I receive after that to you then or if none I will let you know.

Algorithm
Airway box

Everyone should receive oxygen initially not only if they have sats less than 95%

it was felt that starting with airway causes for intubation should come first ie in APLS order: ABCD

this would be simpler if it was "airway obstructed"

Will read better if bullet points used

Main guideline
General comments
This is written for junior medical and nursing staff.

I feel it would look better in places if bullet points were used eg Section 5 following intubation would remove the "theirs"

Also there are a lot of "should be" which could be replaced by more positive comments such as "will"

I know how difficult it is to make a punchy yet thorough guideline but I still think that it is overly long and perhaps some things such as the pathologists input at the end is necessary. Firstly it is unlikely that they will see this guideline, it is not relevant to most junior staff and in general I would be very surprised if any pathologist would need telling this especially any involved in paediatric post mortem. it would be more sensible to check through the Royal College of Pathologists about this and whether they already have appropriate protocols.

Specific comments

2. I was rather surprised that not everybody agreed that observations were not collected a s a norm. They would be in any sick patient in a paed A&E

5.Call anaesthetist or intensivist if considering intubation

I would expect to be considering intubation after 40ml/kg of resuscitation fluid

6. all children should receive oxygen initially

7. under lumbar puncture should include as contraindication
on anti-coagulants
suspected DIC or septic shock as these can occur in patients who have not got meningococcal sepsis

8. Capillary refill time should be central
using CRT alone can lead to over treatment
(last para bar 1) mention inotropes not drugs
consider intubation after 40ml/kg

9 LP if not contraindicated

11. ICP

treatment needs to start before CT scan
any patient with reduced level of consciousness is at risk in a CT scanner from clinical deterioration which is not observed and the presence of an anaesthetist or intubation and ventilation prior to a scan is more sensible and probably usual practice

PaCO2 should be kept 4.5 to 5.0
I would be wary personally of using that much mannitol in one go as a dose of 0.25 or 0.5 g/l is often effective and the total daily dose is 1.5 g/l

need to keep normovolaemia. I would use 80% maintenance. The replacement fluid is much more important and should not be 0.18% saline

12. insert comment that antibiotics are often prescribed at higher doses for meningitis

17 excellent points but could be one sentence

Hope these are useful

I will contact you if I receive any more comments when I am back from holiday in about 3 weeks

Charles Stack

Comments from Royal College of Nursing

Have just read the attached guidelines on reduced consciousness. I showed it to our high dependency unit sister and she was impressed with it. No faults found with it and it might change some of the treatments here ! very good!
It covers every eventuality. Thanks Jane

Comments from The British Society of Paediatric Radiology

Sorry for the delay. Thanks well done
Only one issue sect 18 PM radiography - can you insert
... full post mortem Xray skelatal survey to BPSR standards (www.bspr.org.uk) including skull films.
Many people omit skull films if there has been a premortem CT. Skull fractures are often missed by CT (which is used to detect intracranial complications NOT the fracture) - a hairline skull fracture is useful info to define before the pathologist opends the skull!

Comments from Neonatal and Paediatric Pharmacy Group

A)
Should the dose of mannitol be written as '5ml/kg of 20% which is equivalent to 1g/kg' to reduce the calculations needed?

Similarly should magnesium sulphate read 50mg/kg (=0.5ml/kg of 10% solution)?

Nurses (& some doctors) are not good at converting percentages to mg or grammes & vice versa.

Also how do you prepare the 3% sodium chloride (is this by diluting 30% injection or do you have a special product available?). If it is the first brief details would be helpful to those who don't do it very often.

Many thanks for inviting NPPG to contribute to this important document.
Best wishes,
Sharon

Sharon Conroy
Lecturer in Paediatric Clinical Pharmacy
Academic Division of Child Health
(University of Nottingham)
The Medical School,
Derbyshire Children's Hospital

B)
12. My understanding is that there is only evidence of benefit for dexamethasone in specific types of meningitis and that its use could be problematic in suspected tuberculous meningitis or if CSF samples have not been taken to subsequently identify an organism and its antibiotic resistance pattern.

Is there evidence or even expert opinion that ALL patients with suspected bacterial meningitis should receive dexamethasone?

14. Dose of Mg should be in mmol or micromole and not milligram.

I haven't had chance to check the doses. Are they the same as MfC?

Tony

Tony Nunn
Clinical Director of Pharmacy
Royal Liverpool Children's NHS Trust
Eaton Road
Liverpool

C)
Dear Richard,
As requested, I have had a look at the draft guidelines for the management of reduced conscious level in children. Can I say, it looks good and congratulations on all your obvious hard work.
I have only a few comments:
(section 12)
In bacterial meningitis, you are recommending starting dexamethasone (0.4mg/kg) before or with the first dose of antibiotics.
How did you decide upon this dose?
Should you state a length of dexamethasone course?
Different doses of dexamethasone have been used. A dose of 0.4mg/kg given every 12 hours for a total of 2 days proved to be safe and as efficacious as 0.15mg/kg given every six hours for four days. (Schaad UB et al.Lancet 1993:342:457-61)
However, the more recent Cochrane Review (Corticosteroids in acute bacterial meningitis, Beck et al 2004) recommends: a four day regimen of dexamethasone (0.6mg/kg/day) in effect, equivalent to 0.15mg/kg every six hours.

(section 14) Magnesium sulphate infusion. Would it be prudent to say that the infusion should be diluted to 10% strength? There is a 50% injection available which should be diluted before IV administration.

I am not sure if the colours in the draft document are the final choice but section 11 (in dark blue) is very difficult to read. It may also be worth noting that not everyone has a colour printer, and other sections may be difficult to read if printed from a black & white printer.

You have used abbreviations such as AVPU on the algorithm and in the main text. It would be useful to state what this is in the text (as you have done for GCS)

Colleagues have commented on the algorithm layout and found it to be very 'busy'. I am not sure whether this is avoidable or not, given the amount of information you are trying to convey on one sheet.

Hope these comments are helpful
Best wishes
Gill Hinson
Sheffield Childrens Hospital

D)
Hi Sharon
Was having a look at these this morning (have lots of unopened e-mails!), I don't know if you've looked at it yet?

Looks like it should be a very helpful, comprehensive document. Just a couple of thoughts about the electrolyte infusions mentioned on p12.

Hyponatraemia - it says to give a 3% saline infusion. We have guidelines for SCBU which advise max conc of 1.8% peripheral infusion.

I think this is because of the highly irritant nature of strong saline and am fairly sure Maxine found guidance that 1.8% is the max that should be given. Appreciate this is an acute situation but just thought it was worth mentioning - if they asked for 3% to be made here, we'd probably suggest 1.8% (and the ressies would be thrown by a different conc!). Will investigate further if you want me to

Magnesium - don't have much experience with this but it recommends an infusion of 50mg/kg over 1 hour. No suggestion of whether neat or diluted but apparently we always dilute it here. Haven't seen for kids but adults use for low Mg levels, cardiac patients and pre-eclampsia - all get it diluted, either in infusion bags or in syringes. Can find out more, just thought i'd write it down while I thought about it!

Speak to you soon

Julie

Comments from RCPCH British Association of General Paediatricians

1) We are not sure what other material on this subject will be available for the front-line staff, but we both wondered about the point where one should pause and take a history after establishing the ABC.
Clearly there are important points in the history that are specific to children presenting with disordered consciousness and which could aid diagnosis-when for example should there be a reminder about the vitally important bits of the history like medications, foreign travel etc?. As it stands, history is only mentioned under trauma.

2) On the subject of other material – is there to be a list of differential diagnoses of the patient with disordered consciousness?

3) We are not clear what criteria determine whether the patient coming through the door actually gets entered into the algorithm by having their GCS/AVPU assessed.

4) Is there an age limit on its use –we are thinking of older children and teenagers coming to an A&E at a general hospital.

5) Is the material designed for general front-line A&E medical and nursing staff or for junior paediatricians or both? We were not sure whether the setting for its use is just the A&E or the paediatric admission ward as well, given that some of the recommendations involve evaluations over a period of time; also some of the management recommendations, eg of raised ICP, would presumably be undertaken in intensive care, so from the front-line point of view, having suspected it, should there be a recommendation to consult the relevant intensivist and transfer the patient to the nearest PICU/ICU?

6) We rather felt that recognition and management of "classic" RS is in danger of being overlooked. Clearly it is very low on the list of differentials these days but, as you know, I am concerned that its current low incidence may lead to its being forgotten altogether, which could be disastrous in the setting of a flu epi/pandemic. I am concerned that RS will be missed in teenagers and older children who self medicate with aspirin either because they don't read the warning or think that because they are over 16 it doesn't apply!

The history would give particularly important diagnostic clues (including aspirin exposure - although its absence should not exclude a consideration of RS); also combativeness is an important feature and we are not sure where this fits in to the coma assessment scales –in the teenager it can appear very much like drug intoxication, which is partly why we raised the issue of the age limit on the guideline.

I don't know whether anyone on the Guideline Development Group has had clinical experience in managing RS, but if not perhaps it would be worthwhile getting someone who has, to look at the algorithm. From my reading, a good outcome does seem to depend on getting 10% glucose into the patient as soon as possible, but hypoglycaemia on admission is not universal in RS so if the algorithm was followed as it is, these patients might not be given glucose. Also, I don't know what "Reyeologists" would think about treating the hyperammonaemia with benzoate –it isn't mentioned in the literature on management of RS. Perhaps it wasn't available and perhaps it would be a good thing. At the Workshop Rob Tasker made the point that if classic RS does re-emerge there are all sorts of modern management techniques which could improve outcome and which were not available when it was prevalent in the 70s and early 80s (this is why I suggested to the DoH that it be put on the research agenda for the pandemic flu plan).

At this point I will hand over to David!

Dear Richard,

I hope you will not mind receiving a few additional comments from an ageing bureaucrat. I looked through your algorithm with Sue and was very impressed with all the work that you have put into this. I think the end product will be very useful.

I wonder if you have spoken to Dr Mike Stein, who is working on something called the Map of Medicine. I do not know exactly how this works but I think there are similarities to the kind of sophisticated approach you have been taking -he is trying to produce something that has several layers of information which I believe can be tailored according to local circumstances. The whole enterprise involves a collaboration with the National Electronic Library for Health. He is not a very good correspondent but you will eventually get him on his mobile phone 07970 830714.

Just a few minor points occurred to me from the clinical side.

I know it is implicit in your chart, but perhaps it should be absolutely explicit that one should not administer intravenous glucose on the basis of a capillary sample before drawing venous blood for a proper laboratory determination. I think the bedside measurement of glucose is significantly better than it used to be but I believe it is still thought to be somewhat unreliable at the bottom end of the range. On several occasions I have been irritated when panicking house officers have neglected this precaution and one is left uncertain as to whether or not there really was hypoglycaemia.

Having supported the Doncaster Workshop with great enthusiasm and been pleased about the discussion regarding ammonia, one nevertheless does wonder what effect these recommendations will have on laboratory services and we wondered whether there is any plan to test this out in the field and see just what is the increasing workload that might result.

Of course an algorithm cannot be a textbook, but the points about interpreting the pupils, doll's eye movements etc are probably poorly understood by many otherwise well-trained people. In the far-off days when I was a neurology registrar, I carried round with me a wonderful book called "the diagnosis of stupor and coma" by Plum and Posner. There have probably been several editions since then but it was a superb book about the neurology and physiology of coma and had many practical applications. If you can get hold of a copy I think you would enjoy it

I imagine that on most occasions, by the time a child entered an algorithm for depressed consciousness, s/he would already have the classic meningococcal rash, but there are now several papers on the pitfall of the blanching rash which precedes the classic purpuric one. Should that perhaps have a brief mention?

There was no mention of measuring blood pressure specifically looking for *high* blood pressure as in hypertensive encephalopathy. I also wondered about a mention of haemorrhagic shock encephalopathy syndrome though it seems to be less common than it used to be, but presumably is still around, and there is also the question of toxic shock syndrome.

What is the current teaching about measuring urine output? There always used to be a debate about the merits and hazards of a bladder catheter that would allow one to do this but I do not know what the current thinking is in the kind of situation envisaged in this algorithm.

I think the question of TB meningitis is a very difficult one. It is a pet hobby horse of mine because I saw a lot of this in South Africa and it was generally agreed that unless you had a very specific clue it was often necessary to make a presumptive diagnosis and start treatment immediately, in the full knowledge that as the situation became clear over the next few days you might need to stop treatment. South African clinicians who I worked for were completely relaxed about this and insisted that unless you took this course of action you would miss the boat because by the time the diagnosis became clear your chance of good outcome would be very much less. The important clues to diagnosis were usually a slightly more chronic history than a classic acute bacterial meningitis, a greater degree of drowsiness and yet less evidence of acute toxicity, a variety of localising signs like odd cranial nerve pulses, and very helpfully an abnormal chest x-ray. A positive household contact was also helpful although there was so much TB around that a negative did not mean much.

In my years at St George's I saw only two cases of children with TB meningitis, both in native born white Tooting children. One of them had an aunt who was known to be TB patient but the contact tracing had been seriously delayed, in the other there was no immediate history but the child had a slightly stiff neck, a few white cells in the CSF and a solitary fourth nerve palsy and the father who was quite well except from a cough had obvious TB in his chest x-ray.

I know this is a long rambling comment in relation to the rarity of the problem but I think the important point is to pursue vigorously and urgently the slightest possibility of TB meningitis, at least by getting a chest x-ray of the child and any immediate adult contacts and not relying either on the classic laboratory CSF examination, which is amazingly variable notwithstanding what the books say, or indeed the newer immunological TB tests which are currently being evaluated. TBM is an emergency in spite of looking "chronic". With our diverse population, and very variable standards of contact tracing when adult cases are diagnosed, we may see more in the coming years.

Anyway, use these comments as you will. We look forward with great interest to seeing the next edition!

Sue and David Hall

Comments from UK Clinical Virology Network

A)
I have a couple of comments on section 12 (b):

(i) The statement "HSE should be suspected clinically in a child with reduced conscious level if ..... the child has or has been in contact with herpetic lesions" Does the presence or absence of herpetic lesions, or a history of contact with herpetic lesions, make any difference to the likelihood that a child has HSE? Herpetic lesions may arise in a patient debilitated for any reason, and therefore their presence doesn't help in the differential diagnosis. I doubt whether there is a positive history of contact with herpetic lesions in most patients with HSE. The problem with leaving this statement in is that it may mislead junior clinicians into believing that if their patient doesn't have evident herpetic lesions, or doesn't give a history of contact, then HSE becomes a much less likely diagnosis - which isn't correct.

(ii) Under the heading "Treatment", can I suggest that aciclovir should be administered "as soon as the diagnosis is suspected", rather than "without waiting to perform an LP if an LP is contraindicated" - which isn't the same thing at all. Indeed, the phrasing as it currently stands suggests that if an LP is to be performed, then it is OK to delay the aciclovir until after the LP has been performed - and I would argue that that is absolutely wrong. Aciclovir must be given as soon as anyone thinks about the diagnosis, full stop. Once the aciclovir has been started, then proceed to confirm or refute the diagnosis.

i would like to see detail in the section on post mortem sampling ie what specimens (tissues, liquids etc) and what tests. Since introducing PCR for everything ,we are finding all sorts of infection in kids who die suddenly and this ought to be investigated to a national algorithm

B)
It has been pointed out that HSVE is extremely rare and although it is a rare disease HSE is the commonest non-epidemic (sporadic) viral encephalitis in industrialised countries. It may well be that the incidence is an under-estimate and some minor cases may be unrecognised but still leave patients with residual problems. The broad brush approach is difficult because the clinical picture differs among neonates and older persons, e.g. neonates do not have febrile convulsions although many clinicians fail to appreciate this. It is possible that with the rise in sexually transmitted diseases, more cases of neonatal encephalitis will occur, mostly infected by HSV2. It is also important to appreciate that there are not recommendations for treatment with aciclovir for longer than 10 days, e.g. 14-21 days and with higher doses, e.g. 60 mg/kg per day. A recent study showed that morbidity may be due to intermittent reactivation of herpes simplex in the months, or even years, after neonatal treatment.

Comments have been made that the rarity of HSE must be balanced against the cost of treatment. However, it must be born in mind that cases in which have been demonstrated that there was a delay in recognising HSE with subsequent and usually severe brain damage, is very costly, usually of the order of about £1million or more. For example, I have four large and multiple lever arch files for neonatal HSE which have yet to be settled, although not occurred in the same year. I have one or two for adults, but usually two or three new cases are sent to me each year.

Comments from Royal College of Pathologists: Clinical Biochemistry

Core Investigations
The 10ml of urine to be saved for later analysis should be collected in a plain bottle not a borate bottle

8. Shock
Would this section be better called Shock/Circulatory Failure?
Should tachycardia be mentioned as a sign?

9. Sepsis
Surely blood culture should be considered as a first line test in a child with a clinical diagnosis of sepsis

12a. Bacterial Meningitis
Is it worth mentioning that in young children conventional features of meningitis are often absent?

13a. Hyperglycaemia
Are the NICE guidelines for the management of type 1 diabetes applicable to children?

13b. Hypoglycaemia
Measurement of growth hormone is of very limited value.
It is probably worth suggesting that an extra sample of plasma should be collected during hypoglycaemia and stored frozen in case further investigations are required (ACTH, IGF-I etc).

14. Prolonged Convulsion
What is the logic of waiting 10 minutes before treating a child who is convulsing on arrival?

Comments from Royal College of Pathologists: Paediatric Pathology

I circulated your guideline to our committee and have received a number of replies which I have collated. The basic points made were:

- although very desirable, it is not always possible for a post mortem to be performed within 24 hours, and by a paediatric pathologist. The coroners system is often not flexible enough to move this quickly (even allowing for weekends and bank holidays) and as you probably know paediatric pathologists are in short supply and not always immediately available. This highlights the point that if there is to be any delay, thought should be given to taking appropriate initial specimens (such as skin biopsies and blood) as soon as possible after death, on the ward or in casualty, with the consent of the coroner. Relevant specimens are listed in the Table 1 of the recent Kennedy Report on infant death.
- it is difficult to be prescriptive in setting out a post mortem protocol in these cases, given the differing ages of the children and large number of possible aetiologies. The types of specimen taken at post mortem examination will to a great extent be determined by the circumstances of an individual case, the results of investigations made in life, and also the local requirements of particular laboratories. For the sudden death of an infant, most pathologists would now follow the guideline given in Appendix III of the Kennedy Report. It might be better for your protocol to simply specify that the pathologist should undertake screening for infection and metabolic disease, and also to consider toxicology as indicated.
- how rapidly neuropathology is undertaken will again depend on the nature of the case and other factors such as referral for an expert neuropathological opinion. It is not always possible (or desirable) to complete it in a few days.
- full skeletal survey may not be indicated in every case. Radiologists with expertise in NAI are scarce and it may only be possible to obtain a provisional report from a local radiologist before starting the post mortem.

These comments are intended to be constructive and I hope they are helpful. Please get in touch if you would like to discuss this further.
Yours sincerely,

Dr Chris Wright
Chairman of the RCPath Paediatric Pathology Specialist Advisory Committee.

Comments from British Paediatric Neurology Association

General comments:
My overall impression is that the guidelines are generally sound and fairly comprehensive.

1) I think that they are too long and the desire to be comprehensive has perhaps led to the inclusion of details which may obscure the overall message. For example it is surely not necessary to have detailed descriptions of the management of: shock, sepsis, meningitis, herpes simplex encephalitis, abscess, hypoglycaemia, hyperammonaemia etc. If one of these conditions is suspected, then the reader could be referred to a different guideline as has been done for trauma and DKA. The danger of having all this detail on other conditions is that the neurological aspects of the condition may be forgotten once a different algorithm is being followed
2) Related to the above is the way in which **raised intracranial pressure** is presented (between trauma and bacterial meningitis) as though it were a separate condition, rather than a complication that could develop in any of the specific conditions mentioned. I think some sort of general statement about the possibility of raised intracranial pressure being present in any child with depressed conscious level, should be included. Likewise it should be emphasized that the commonest sign of raised ICP is a depression of conscious level

Specific comments:
Section 1.1: The specific coma score to be used should be mentioned e.g. Tatman et al 1997 and somewhere there should be mention of the need for continuous training and evaluation for staff, in use of coma scores.
Section 1.2: These observations are an integral part of "neuro obs" and should be mandatory.
I don't think the criteria for deciding on whether to do GCS every 15 minutes should only be "a GCS of 12 or less". If the history suggests a recent deterioration of conscious level GCS every 15 minutes may be appropriate, whatever the current GCS is.
Section 5: as above, a GCS of 8, or a deteriorating GCS, is a potential sign of raised ICP in its own right. Maybe at this point signs of "raised ICP" should be spelt out(see below).
Section 7: The list of suspected conditions should include **poisoning/toxic** causes.
The use of the word "convulsions" is problematic. In this section of the guideline it would be better to simply put **epileptic seizures**. The problem with "convulsions" is two fold. Firstly it does not specify what abnormal movement it is referring to: does it mean tonic, clonic, tonic-clonic, dystonic, focal or generalized etc.? The real danger of this is that tonic movements due to herniation are misinterpreted as epileptic seizures and treated as such.
Secondly, the implicit assumption is that "convulsions" are epileptic in nature. This may lead to mis diagnosis of nonepileptic disorders as above, or to the failure to recognize that non convulsive epileptic seizures can cause depression of conscious level(eg. nonconvulsive status)
In the next section " Core investigations" I would replace the phrase in brackets; "except those patients within one hour post convulsion" with the phrase; "except those patients within one hour following a generalized tonic-clonic seizure".
Section 7, Lumbar puncture: It would be helpful to highlight the phrase in the opening sentence "when no contraindications exist"
There is a typo; papillary dilation, should read pupillary dilatation as should papillary reaction.
Presence of papilloedema should also be included.
"signs of raised ICP" need to be specified. (See below)
Section 10. The importance of considering NAI as a cause of acute encephalopathy in infancy should be emphasized.
Section 11. This is the section I was least happy with. As discussed above I think there needs to be a section earlier in the guidelines about the symptoms and signs of raised ICP. Specific points about this section are:
The section on the relation of ICP to CPP seems to be out of context and a bit irrelevant without a more detailed discussion of the pathophysiology of raised ICP. I would remove it.
Im not sure what the assertion that " 2 or more of the following signs" is based on. This seems potentially dangerous as any of these signs alone could be due to raised ICP.
Reduced conscious level of **any** degree could be due to raised ICP. I don't think GCS < 9, or the imprecise term "unrousable" should be used.
Abnormal respiration of any pattern could be due to raised ICP
Abnormal posture and tone, with abnormal flexion or extension would be preferable to "decorticate or decerebrate"
Presence of papilloedema should be included.
Bradycardia, or hypertension or both should be included.

Treatment: The dose of mannitol is high. I use 0.5g/kg by repeated doses if necessary. If high doses are going to be used there needs to be some warning about the risks of a hyperosmolar state, and not giving further mannitol if the osmolality is greater than 320.
It would be practically useful to specify doses to be given eg. 1g/kg mannitol is equivalent to: 5mls/kg of 20% mannitol.
I am unhappy about the statement: "maintenance fluids should not be hypotonic". There is always a risk of hyponatraemia and worsening cerebral oedema in an acute neurological disorder. I would specify that maintenance fluids should be 0.45% saline/5% dextrose, unless there is a specific clinical reason to use a different fluid.
I am not sure that there is good evidence to recommend routine fluid restriction for patients with raised ICP/coma, but as above avoidance of overhydration is important.

As noted I think there should be a prominent box or section listing possible signs and symptoms of raised ICP eg.
Symptoms of raised ICP: headache, vomiting, diplopia or visual obscurations
Signs of raised ICP: **altered conscious level**, pupillary abnormalities, papilloedema, eye movement abnormalities, bradycardia, hypertension, respiratory abnormalities. Extensor or flexor posturing.
Somewhere in the guidelines, perhaps in the section on "convulsions" and certainly on the algorithm, there should be a prominent statement to the effect that: repeated extensor or flexor stiffening/posturing(tonic posturing, tonic "seizures") in a child with an altered conscious level, are signs of tentorial herniation until proven otherwise and they are not likely to be epileptic seizures.
Section 12. I find the "clinical decision rule" almost incomprehensible and would strongly resist recommending such an approach to the diagnosis of bacterial meningitis.
In clinical practice, unless an overt cause for reduced conscious level is rapidly apparent, then CNS infection is potentially present and should be treated until it is ruled in or out. Certainly any child with febrile encephalopathy should be treated with antibiotics and anti virals
Again, the guidelines are not about how to diagnose herpes encephalitis, but the management of reduced conscious level and the main management points are: 1) starting acyclovir in any child with an unexplained encephalopathy and 2) appropriate investigations to make a diagnosis of HSE, which should include paired serology as well as CSF PCR.
There are many other causes of encephalitis and this should be recognized.
The dose of acyclovir given as mg/kg is probably too low. eg a 30 kg child with a surface area of $1m^2$ would require 500mg/dose which is equivalent to 16mg/kg.
Some centres (including ours) routinely also add treatment with erythromycin or clarithromycin to cover the possibility of mycoplasma encephalitis, when CNS infection is suspected.
Secrtion 13: The section on hypoglycaemia suggests that an infusion of 10% dextrose solution is administered. Because of the risks of hyponatraemia with electrolyte free solutions it should be spelt out that this should be combined with a saline containing fluid (0.45% NaCl for example)
Section 14: As discussed above, I think a preferable heading would be: prolonged epileptic seizure or, prolonged tonic-clonic seizure
Section 15: As above this should either be headed: **post ictal state**, or **depression of consciousness due to an epileptic seizure.**
The first sentence should specify that it is referring to an epileptic seizure. eg. "After a generalized tonic clonic seizure, a child will often have a period of reduced consciousness, the "post-ictal state"
"The post-ictal state will last for less than 1 hour in the majority of children. After 1 hour if they have not recovered consciousness the possibility of an acute symptomatic cause for the seizure needs to be considered, and appropriate investigations performed"
Section 16: An EEG should be part of the routine investigations of a child with an unexplained reduced conscious level. I would recommend an EEG even when the diagnosis is clear as it gives useful information about: presence or absence of seizures, background and focal abnormalities that might suggest the need for imaging if this has not already been done.
As noted above my practice is to start blind erythromycin as well as acyclovir and broad spectrum antibiotics, when the cause is unknown.

Additional comments: Somewhere in the guideline, perhaps in the section on raised ICP, or in section 16, there should be a brief discussion of the management pathway for a child who has presented acutely with a space occupying lesion such as tumour, abscess, intracranial haemorrhage eg . ABC, intubate ventilate, give mannitol, CT scan then urgent referral to neurosurgeons.
There should also be specific mention of children with VP shunts or ventriculostomies where a depressed conscious level should be interpreted as shunt/ventriculosomy malfunction until proven otherwise. These children need urgent referral to neurosurgeons and this should be emphasized.
Perhaps in section 4 under "history" a sentence could be added: **do they have a VP shunt/ventriculostomy.]**

Algorithm: As discussed above, I think raised ICP should not be regarded as a separate cause of reduced conscious level, but rather a potential complication of any of the possible aetiologies. This needs to be incorporated into the algorithm.

Comments from RCPCH Nephrology Interest Group

A)
1. How can hypertensive encephalopathy be distinguished from hypertension secondary to raised intracranial pressure in children with reduced consciousness?

Presence of features suggesting a longer duration of the hypertension: hypertensive retinopathy, left ventricular hypertrophy.

Presence of findings suggestive of a secondary hypertension due to renal causes e.g. renal scarring, haematuria / proteinuria

2. How would you define the diagnosis of hypertensive encephalopathy in terms of BP and other specific features? (unfortunately headache, vomiting etc are all pretty non-specific and could be signs of raised ICP also)

The problem is that hypertensive encephalopathy occurs at different severities of hypertension, depending of the patient and presumably the rate of rise of blood pressure. I would not want to give a specific cut off of blood pressure. The answer to this question perhaps lies within the answer to question 1.

3. What further investigations would you like performing to help determine the cause of the hypertension? (specifically tests which can be requested in the first hour or so of management)

Biochemistry assessing renal function, specifically serum creatinine.
Urinalysis looking for blood and protein.
Renal ultrasound
Upper and lower limb bp to exclude coarctation.

4. What treatment would you recommend as a first line therapy? (the "treatment" can include contact the nearest paediatric nephrologist for advice)

Management on a paediatric iCU with paediatric nephrology input.
My first line drug treatment for hypertensive encephalopathy would be intravenous sodium nitroprusside. Use of this drug requires intra-arterial bp monitoring. An alternative is intravenous labetalol, but this is less potent. Treatment should aim to bring the blood pressure down rapidly to bring seizures under control. From that point, blood pressure should be normalized over a minimum of 72 hours.

If you know of any research in this area that would be useful (I've done a comprehensive review and only come up with isolated paediatric case reports and adult data on SL nifedipine).

SL nifedipine would not be used in an encephalopathic patient. I don't know of any trials.

B)
1. How can hypertensive encephalopathy be distinguished from hypertension secondary to raised intracranial pressure in children with reduced consciousness?

There has been a manuscript on this in Critical Care Medicine. Hypertension associated with hypertensive encephalopathy is constantly raised, unless of course treament has been given to decrease it.

The hypertension of raised intracranial pressure is episodically raised (plateau waves) and it is associated with bradycardia and other features of raised ICP. Second the level of BP is important - the Cushing's triad BP does not go up to 250 mm Hg. Third, the context is important - seizures (non convulsive) and head injury are instances where we see raised BP that is not hypertensive encephalopathy. Fourth there is the eye examination - the fundus of hypertensive encephalopathy does not look the same as papilloedema - there is more. Last, both of these states (raised ICP, and possible hypertensive encephalopathy) have to be distinguished from the sympathetic storm at the time of tentorial hernaition.

2. How would you define the diagnosis of hypertensive encephalopathy in terms of BP and other specific features? (unfortunately headache, vomiting etc are all pretty non-specific and could be signs of raised ICP also)

There is literature on this - look under <urgent> and <emergent>. The symptoms are not necessary the key thing here. The facial nerve and eye findings are what most people look at.

3. What further investigations would you like performing to help determine the cause of the hypertension? (specifically tests which can be requested in the first hour or so of management)

Context again. The main priority is treament and saving the right samples for later tests (VMA, renin, adrenaline, etc) that will help to sort out what is going on. The tests that you could get a result from early are clinical (has the patient got endocrine disease, coarctation etc), renal ultrasound scan and Doppler (looking for renal artery compression, renal artery stenosis) etc.

4. What treatment would you recommend as a first line therapy? (the "treatment" can include contact the nearest paediatric nephrologist / intensivist for advice)

If were talking hypertensive encephalopathy we're going for broke - labetolol, and then GTN - dropping systolic by 5 mm Hg in the first 24 hrs if this has been chronic. Really need to have context - uregent or emergent. There are lots of oncology kids we see - not encephalopathic but with BP 180 systolic (age 6 yrs) who start on propranol and nifedipine.

Regards
Robert

C)
I just picked up your round robin e-mail circulated by Heather Maxwell with your queries about hypertensive encephalopathy as a cause of altered consciousness.

My brief comments to the 4 questions would be:

1) Hypertensive encephalopathy is usually distinguished from hypertension secondary to raised intracranial pressure by the presence usually of severe sustained hypertension, ie 220/160, as opposed to more moderate hypertension with raised ICP. Blood pressures are invariably above the 99th centile and usually the urinalysis shows proteinuria and possibly blood. Most cases have renal impairment and several cases will have fundoscopy changes of hypertensive retinopathy.

2) Under core investigations I would specify U&E and creatinine, LFTs including calcium and phosphate. Obviously an elevated phosphate and creatinine may point towards renal impairment with renal parenchymal damage (due to reflux scarring) being one of the commonest causes for hypertensive encephalopathy.

3) Your boxes such as raised intracranial pressure don't include fundoscopy and in most cases of hypertensive encephalopathy there is usually associated retinopathy. Fundoscopy is, to my mind, an imperative clinical examination before CT scans!
The therapy for the hypertensive crisis can be obtained from the 6th edition of Forfar & Arneil which I have revised twice (p638 Table 16.14).

Cheers

4. Public Open Day for Stakeholder groups

A Public Open Day was held at Queens Medical Centre, Nottingham on 7th October 2005. This was an open forum for stakeholders to review the guideline as it reached its final draft version. Invitations were sent out to all the Stakeholder representatives and Delphi panellists. A national advert was placed in The Royal College of Paediatrics and Child Health evidence-based bulletin publication “CHERUB”, to invite members of the RCPCH. Electronic invitations were sent to hospitals locally.

A satisfactory turn out ensured that stakeholders contributed to the wording and format of the final guideline versions.