RCPCH GUIDELINE APPRAISAL

Royal College of Physicians

Clinical Guidelines for Diagnosis and Management of Stroke in Children

The original guideline is NOT the work of the Royal College of Paediatrics and Child Health. This document represents the College's appraisal and summary of the authors' completed guidelines together with the main recommendations. Whilst only grade A & B recommendations have been appraised, other recommendations and good practice points (GPP's) are also reproduced. The British Paediatric Neurology Association initiated the guideline in collaboration with the Royal College of Physicians Intercollegiate group, in tandem with stroke guidelines for adults.

KEY POINTS

- Children with sickle cell disease should be screened annually from the age of 3 years using ultrasound Doppler; those with increased cerebral artery velocity should be offered long-term blood transfusion.
- The recommendations for acute management underline the importance of distinguishing between ischaemic and other categories of stroke.
- Children affected by stroke and their parents were involved in identifying the key themes to be addressed by the guidelines. Quotations are included as illustrations.
- No formal consensus process was used to formulate recommendations where evidence was lacking.
- Service organisation statements may have unforeseen cost implications and there may be other approaches that are equally or more appropriate: they do not have RCPCH endorsement, and have not all been included.

Guideline Scope

The scope of the guideline includes the prevention, acute management, rehabilitation and longer-term management of acute arterial ischaemic stroke arising beyond the neonatal period, as well as the organisation of services (the latter are not reproduced here).

Recommendations (***only grade A and B have been appraised by the RCPCH***)	**Grade**
ACUTE DIAGNOSIS OF ARTERIAL ISCHAEMIC STROKE IN CHILDREN	
Presentation and diagnosis	
All children with a clinical presentation of stroke should be under the care of a Consultant Paediatrician.	GPP
Cross sectional brain imaging is mandatory in children presenting with clinical stroke.	C
Brain MRI is recommended for the investigation of children presenting with clinical stroke.	C
Brain MRI should be undertaken as soon as possible after presentation. If brain MRI will not be available within 48 hours, CT is an acceptable initial alternative.	GPP
Brain imaging should be undertaken urgently in children with clinical stroke who have a depressed level of consciousness at presentation or whose clinical status is deteriorating.	GPP
Any new neurological symptoms or signs in children with sickle cell disease should be evaluated as potentially being due to stroke.	GPP
All children with clinical stroke should have regular assessment of conscious level and vital signs.	GPP
Investigations	
Imaging of the cervical and proximal intracranial arterial vasculature should be performed in all children with arterial ischaemic stroke.	C
Imaging of the cervical vasculature to exclude arterial dissection should be undertaken within 48 hours of presentation with arterial ischaemic stroke.	GPP
Transthoracic cardiac echocardiography should be undertaken within 48 hours after presentation in all children with arterial ischaemic stroke.	GPP

Recommendations (*only grade A and B have been appraised by the RCPCH*)	**Grade**
All children with arterial ischaemic stroke should be investigated for an underlying prothrombotic tendency. This should include evaluation for protein C protein S deficiency, activated protein C resistance, increased lipoprotein (a), increased plasma homocysteine, factor V Leiden, prothrombin G20210A and MTHFR TT677 mutations and antiphospholipid antibodies.	C
ACUTE CARE	
General care measures	
Temperature should be maintained within normal limits.	D
Oxygen saturation should be maintained within normal limits.	D
Care should be provided in an environment that is appropriate for the child's age and developmental level.	D
Specific medical treatments	
Aspirin (5mg/kg/day) should be given once there is radiological confirmation of arterial ischaemic stroke, except in patients with evidence of intracranial haemorrhage on imaging and those with sickle cell disease.	GPP
In children with sickle cell disease and arterial ischaemic stroke,	
(i) urgent exchange transfusion should be undertaken to reduce %HbS to <30% and raise haemoglobin to 10-12.5g/dl	GPP
(ii) if the patient has had a neurological event in the context of severe anaemia (e.g. splenic sequestration or aplastic crisis), or if exchange transfusion is going to be delayed for more than 4 hours, urgent top up blood transfusion should be undertaken.	GPP
Providing there is no haemorrhage on brain imaging, anticoagulation should be considered in children with,	
(i) confirmed extracranial arterial dissection associated with arterial ischaemic stroke	GPP
(ii) cerebral venous sinus thrombosis.	C
The decision to use anticoagulation in children with arterial ischaemic stroke who have a cardiac source of embolism should be discussed with a consultant paediatric cardiologist and paediatric neurologist.	GPP
Early neurological referral should be considered in children with stroke who have depressed or deteriorating conscious level or other signs of raised intracranial pressure.	GPP
Secondary prevention of arterial ischaemic stroke in childhood	
Patients with cerebral arteriopathy other than arterial dissection or moyamoya syndrome or those with sickle cell disease should be treated with aspirin (1-3mg/kg/day).	GPP
Anticoagulation should be considered,	
(i) until there is evidence of vessel healing, or for a maximum of 6 months, in patients with arterial dissection	GPP
(ii) if there is a recurrence of arterial ischaemic stroke despite treatment with aspirin	GPP
(iii) in children with cardiac sources of embolism, following discussion with the cardiologist managing the patient	GPP
(iv) until there is evidence of recanalisation or for a maximum of 6 months after cerebral venous sinus thrombosis.	GPP
In children with sickle cell disease,	
(i) regular blood transfusion (every 3 to 6 weeks) should be undertaken to maintain the HbS% <30% and the Hb between 10-12.5g/dl	C
(ii) transfusion may be stopped after 2 years in patients who experienced stroke in the context of a precipitating illness (e.g. aplastic crisis) and whose repeat vascular imaging is normal at this time	C
(iii) after 3 years a less intensive regime maintaining HbS <50% may be sufficient for stroke prevention	C
(iv) those who cannot receive regular blood transfusions because of allo-immunisation, auto-antibody formation, lack of vascular access or non-compliance with transfusion or chelation may be considered for treatment with hydroxyurea.	C
Children with moyamoya syndrome (including those with sickle cell disease) should be referred for evaluation to a centre with expertise in evaluating patients for surgical revascularisation.	D

Recommendations (*only grade A and B have been appraised by the RCPCH*)	**Grade**
Children with sickle cell disease who have had a stroke should be referred to a specialist centre for consideration of bone marrow transplantation. (**Original recommendation: Grade B**).	C
Advice should be offered regarding preventable risk factors for arterial disease in adult life, particularly smoking, exercise and diet.	D
Blood pressure should be measured annually to screen for hypertension.	GPP
Patients who are found to have a prothrombotic tendency should be referred to a haematologist.	GPP
EARLY DISABILITY ASSESSMENT AND MANAGEMENT	
As soon as possible after admission, all children following stroke should have an evaluation of,	
(i) swallowing safety	D
(ii) feeding and nutrition	GPP
(iii) communication	D
(iv) pain	D
(v) moving and handling requirements	D
(vi) positioning requirements	GPP
(vii) risk of pressure ulcers.	D
All children affected by stroke should have a multidisciplinary assessment within 72 hours of admission to hospital.	GPP
The medical, social, emotional and educational needs of the child affected by stroke should be considered early and systematically assessed in a co-ordinated manner when planning their subsequent care.	D
All members of the healthcare team should work together with the child and family, using an agreed therapeutic approach.	D
A key worker should be appointed to co-ordinate the package of care, ensure its delivery and to act as a central point of contact for the family. The key worker and their role should be explained to the family.	D
The professionals involved in the acute assessment and management of the child should initiate early liaison with their counterparts in the community to ensure a smooth transition of care.	GPP
APPROACHES TO REHABILITATION	
Children affected by stroke should be offered advice on, and treatment aimed to achieve, play, self-care, leisure and school related skills that are developmentally relevant and appropriate to their home, community and school environment.	D
Equipment which is appropriate in meeting rehabilitation aims should be assessed on an individual basis, provided in a timely manner, and regularly monitored by appropriate professionals.	D
SENSORIMOTOR REHABILITATION	
Underlying approach	
Sensorimotor therapies should be practiced within a neurological framework, and complement other interventions to improve functional skills.	GPP
Rehabilitation activities should be task orientated and relevant to the individual's life.	C
Therapy should be integrated into the child's daily home and school activities.	D
Delivery	
Children should be given as much opportunity as possible to practise skills.	C
Use of assessment measures	
The assessment tools selected should be appropriate for the child's age, developmental and functional level.	GPP
Standardised and validated assessment tools should be used where possible.	GPP

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MOTOR IMPAIRMENT	
Muscle strengthening	
Muscle strengthening should be used as part of the therapy programme to prevent or reverse contractures.	D
Muscle strengthening activities should be used to improve functional activity.	D
Management of Spasticity	
If spasticity results in functional limitation or discomfort, botulinum toxin injection may be trialled to reduce muscle tone and improve joint motion. (**Original recommendation:** If spasticity results in functional limitation or discomfort, botulinum toxin injection may be used to reduce muscle tone and improve joint motion.)	B
The most effective dose for gastrocnemius injection is 20µg/kg to reduce the dynamic component of muscle shortening and increase active length.	C
Botulinum toxin should not be used in isolation from other therapy interventions.	GPP
Ankle foot orthoses	
A rigid AFO could be considered to aid standing balance, swing through in gait and prevention of foot and ankle contractures.	GPP
A hinged or posterior leaf spring AFO should be used to facilitate normal gait patterns.	D
SENSORY IMPAIRMENT	
Somatosensory impairment	
Rehabilitation professionals should consider the presence of somatosensory impairment and integrate this in planning and implementing rehabilitation.	GPP
Hearing & vision impairment	
Vision and hearing should be assessed as part of the multidisciplinary assessment.	GPP
Pain	
Children affected by stroke should be assessed for the presence of pain using a validated paediatric pain tool.	D
All pain should be treated actively, using appropriate measures including positioning, handling, and medication.	GPP
In cases of intractable pain, the child should be referred to health professionals with specialist expertise in pain management.	GPP
LANGUAGE AND COMMUNICATION	
Professionals working with children affected by stroke should be aware that language and communication skills may be affected.	D
If parents, professionals or the child's educational assessment raise concerns regarding language or communication, the child should be referred to a specialist speech and language therapist.	D
A detailed assessment of the child's communication abilities should be carried out in collaboration with the child, parents/carers, teachers and other therapists to identify the child's strengths and weaknesses and plan intervention that aims to increase functional capabilities.	D
A collaborative approach to the management of communication difficulties that includes working with educational psychologist, other therapists, teachers and social workers should aim to equip the child with a language for life.	D
COGNITIVE EFFECTS	
Professionals working with children affected by stroke should be aware that cognitive function may be affected, both immediately and in the longer term.	C
A detailed psychological assessment of the child's cognitive and functional abilities together with any wider family concerns should be carried out in collaboration with the child, parents/carers and teachers to identify any special educational needs.	D
Cognitive assessment should take account of the presence of any visual or hearing deficits.	GPP

Recommendations (*only grade A and B have been appraised by the RCPCH*)	**Grade**
MOOD AND BEHAVIOUR	
Families and professionals should be aware that stroke may have effects on mood and behaviour.	D
The psychological assessment of the child should include evaluation of mood and behaviour, including wider family concerns. This should be undertaken in conjunction with cognitive assessment.	D
Mood and behaviour should be assessed if there is a change in the child's functioning in the home or school environment.	GPP
If mood or behaviour problems are identified and are having an impact on the child's functioning, the child should be referred to professionals with expertise in treating such problems, such as the local Child and Adolescent Mental Health Team.	GPP
ACTIVITIES OF DAILY LIVING	
Therapists working with a child affected by stroke should assess the child's ability to perform daily living activities.	GPP
An occupational therapist should be involved in identification of therapeutic need in self-care, work/school and leisure activities and provision of intervention in this area if indicated.	GPP
LONGER TERM AND COMMUNITY CARE	
Return to school	
Child health services, usually community child health services, should take responsibility for informing the Local Education Authority of children who may have special educational needs as soon as possible after the stroke.	D
The child's key worker should liaise with the Special Educational Needs Coordinator at the child's school prior to school return.	D
A collaborative meeting should be undertaken to plan educational provision with appropriate assessment or support.	D
Health and school staff should agree procedures for communicating information.	GPP
For children presenting with mobility difficulties, the school environment should be assessed prior to return to school, ideally by an occupational therapist.	GPP
It is recommended that all children affected by stroke are placed on a minimum of School Action* as many difficulties remain latent. **See full guideline*	GPP
Transition between paediatric and adult services	
Paediatric general and speciality clinics and Child Development Services should have a local policy on transition to adult services, which should be the responsibility of a named person.	D
A named professional should take responsibility for arranging an introduction to adult health services.	GPP
A flexible approach to the timing of this transfer needs to be considered which takes into account the young person's readiness, current health status and links to other social transitions such as leaving school.	D
A multi-agency transition plan should be formulated for young people with special educational needs, with input from health, education, social services and the young person to plan transition into further education, training or employment.	D
A named professional should take responsibility for co-ordinating this transition plan and ensuring delivery of services.	D
PRIMARY PREVENTION	
Children with haemoglobin SS or Sβ° thalassaemia should be screened yearly from the age of 3 years for internal carotid artery or middle cerebral artery velocity >200cm/s using appropriately trained personnel and transcranial Doppler ultrasound.	B
Children with sickle cell disease who have internal carotid artery/middle cerebral artery velocity >200cm/s should be offered long-term blood transfusion.	B
TERMINOLOGY AND THEORETICAL FRAMEWORK	
Each team should use a consistent framework and terminology in providing care to child affected by stroke.	GPP
It is recommended that the World Health Organisation's International Classification of Functioning (ICF) terminology is used.	GPP

Recommendations (*only grade A and B have been appraised by the RCPCH*)	**Grade**
CHILDREN AND THEIR FAMILIES	
Families/carers should be given factual information about their child's condition as soon as possible after diagnosis. This should be simple and consistent, avoiding technical terms and jargon.	D
Written information should be provided to the child and family regarding the child's health and statutory and voluntary services available.	D
Children should be given information about their condition at an appropriate level.	D
The child and family should be involved in making decisions about the child's care, including rehabilitation and education.	D
The multidisciplinary health team at secondary level should provide co-ordinated care and liaise closely with Education and Social services through the key worker.	D

The full guideline may be downloaded via the following website: **http://www.rcplondon.ac.uk**
The guideline appraisal can be accessed via: **http://www.rcpch.ac.uk/publications/clinical_docs.html**

LEVELS OF EVIDENCE/DERIVATION OF GRADES OF RECOMMENDATIONS

The levels of evidence used throughout are those derived from SIGN guideline 50.

Levels of evidence

1++ High quality meta analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+ Well conducted meta analyses, systematic reviews of RCTs, or RCTs with a low risk of bias
1 - Meta analyses, systematic reviews of RCTs, or RCTs with a high risk of bias
2++ High quality systematic reviews of case-control or cohort or studies. High quality case-control or cohort studies with a very low risk of confounding, bias, or chance and a high probability that the relationship is causal
2+ Well conducted case control or cohort studies with a low risk of confounding, bias, or chance and a moderate probability that the relationship is causal
2 - Case control or cohort studies with a high risk of confounding, bias, or chance and a significant risk that the relationship is not causal
3 Non-analytic studies, e.g. case reports, case series
4 Expert opinion

Grades of recommendation

A At least one meta analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or a systematic review of RCTs or a body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results
B A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 1++ or 1+
C A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 2++
D Evidence level 3 or 4; or extrapolated evidence from studies rated as 2+
GPP Recommended best practice based on the clinical experience of the guideline development group.

SUMMARY OF 'AGREE' FINDINGS

The methods used to identify the evidence
Medline, AMED, CINHAL and Embase were searched.

Which professionals were involved
The guideline development team consisted of Vijeya Ganesan, Senior Lecturer in Paediatric Neurology, Kling Chong, Consultant Neuroradiologist, Jane Evans, Consultant Paediatric Haematologist, Anne Gordon, Research Occupational Therapist, Dianne Gumley, Consultant Clinical Psychologist, Fenella Kirkham, Reader in Paediatric Neurology, Janet Lees, Consultant Speech and Language Therapist, Donal O'Kelly & Keith Wood, Different Strokes, Dominic Thompson, Consultant Neurosurgeon, Terry Pountney, Senior Physiotherapist, Susan Rideout, Clinical Specialist Paediatric Neurology, Beth Ward, Clinical Nurse Specialist, Sue Wayne & Eoin Redahan, The Stroke Association, Andrew Williams, Consultant Community Paediatrician, Kofie Anie, Consultant, Lola Oni, Nurse Director/Lecturer.

Involvement of parents &/or children
Children affected by stroke and their parents were invited to attend a structured workshop in order to identify areas that they thought should be addressed within the guideline.

Consensus method used
Previous versions have used an informal consensus process.

Clinical audit
A number of audit criteria are included.

Overview
This publication presents evidence-based information. Guidelines are "systematically developed statements to assist decisions about appropriate care for specific clinical circumstances" based on systematic reviews of the research literature. Guidelines are not intended to restrict clinical freedom, but practitioners are expected to use the recommendations as a basis for their practice. Local resources and the circumstances and preferences of individual patients will need to be taken into account. Where possible, recommendations are based on, and explicitly linked to, the evidence that supports them. Areas lacking evidence are highlighted and may form a basis for future research.

The Role of the Royal College of Paediatrics and Child Health
In order to raise awareness about the existence of the original guideline and to ensure its relevance for children's health, the College (through its Quality of Practice Committee) appraised the original guideline against the 'AGREE' checklist laid out in its 'standards' document. Having established the quality of the guideline's methodology in this way, the College Clinical Effectiveness team examined the recommendations presented in the guideline document in the context of the original research papers from which they were derived. Their findings are presented here. Where discrepancies between their findings and the originals exist, both recommendations have been included. The shaded boxes indicate these areas of discrepancy.

Acknowledgements: The members of the QPC who oversaw the process of the review: Dr Harry Baumer (Chairman), Mrs Linda Haines, Dr Monica Lakhanpaul, Miss Samantha Love, Professor Neil McIntosh, Dr Ian Maconochie, Dr Richmal Oates-Whitehead, Dr Bob Phillips, Dr Martin Richardson, Dr Aung Soe, Mrs Morwenna Stewart, Dr Kate Verrier Jones, Dr William Whitehouse.

The College's appraisal should not be considered valid beyond January 2006, and new evidence at any time could invalidate these recommendations.