Stroke in childhood

Clinical guidelines for diagnosis, management and rehabilitation

Prepared by the Paediatric Stroke Working Group

November 2004

Clinical Effectiveness
& Evaluation Unit
ROYAL COLLEGE OF PHYSICIANS

The Clinical Effectiveness and Evaluation Unit

The Clinical Effectiveness and Evaluation Unit (CEEU) of the Royal College of Physicians has expertise in the develoment of evidence-based guidelines and the organising and reporting of multicentre comparative performance data. The work programme is collaborative and multiprofessional, involving the relevant specialist societies and patient groups, the National Institute for Clinical Excellence (NICE) and the Healthcare Commission. The CEEU is self-financing with funding from national health service bodies, the Royal College of Physicians, charities and other organisations.

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Foreword

Stroke, in both adults and children, used to be something that happened but which medicine could do, or chose to do, little about. Over the last decade there has been a revolution in stroke care for adults, with the advent of specialist stroke units and evolving treatments. Publication of the first edition of the *National clinical guidelines for stroke* in 2000 and the National Sentinel Audit of Stroke stimulated local units to consider the quality of the care they were delivering and put improvements in place. Rehabilitative care after stroke for adults is now considered the norm, and the collaboration between physicians, nurses, therapists and patients in these projects has led to true multidisciplinary working – much to the benefit of patients.

Similar improvements, however, have not be seen in the treatment of childhood stroke which, although less common than adult stroke, is still a serious problem and one which anecdotal evidence suggests is prone to an even more variable quality of care. Thus, when the national guidelines were being revised by the Intercollegiate Stroke Working Party, a subgroup was formed to consider the paediatric aspects of stroke care. During development it became clear that little evidence existed for many areas. The gaps were filled using the expertise of the group and the views of patients, parents and carers, but it is clear that there is a need for more formal research.

This guideline is aimed at healthcare staff in all parts of the NHS and related services, but much of it may also be of value to patients, parents and families. It is hoped that it will help childhood stroke services to emulate the improvement that has occurred, and is continuing to occur, in the care of adults, bringing a consistency and a knowledge of best practice to an area marked until now by dislocated care and uncertain standards.

To the casual reader wondering if this is important – I would urge you to begin by glancing at the italicised quotes from participants in the child and carer workshop which are scattered through the document. They plainly show how much more should and can be done to ensure good acute management, rehabilitation and secondary prevention, and to help these young people and their parents and families adjust to and cope with the effects of their stroke. Children have a lifetime ahead of them – any benefits from improved care will also last a lifetime.

November 2004

Professor Mike Pearson
Director, Clinical Effectiveness and Evaluation Unit,
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The child and family perspective

As part of the guideline development process a workshop was held for children affected by stroke and their families. It gave a sense of the issues children and families felt to be important. We particularly focused on these issues in developing the guidelines, and they are outlined here.

The child is part of a family and, therefore, any childhood illness will have effects on both the immediate and extended family. The diagnosis of stroke is an unexpected one in a child, which compounds the shock experienced by families at the time of diagnosis.

The lack of information for families of children affected by stroke was a strong theme in the workshop. Information on what has happened and what to expect could empower families to ensure that the immediate and long-term needs of their child can be met. Information for parents, carers and children should be designed to meet their specific needs. The same information pack is unlikely to be appropriate in all cases but the information leaflet provided with this document may form a useful starting point. It is important that the questions asked by parents, carers, and children are answered. The communication of information to children is often particularly neglected.

Parents and carers of children affected by stroke welcome support from those who have had similar experiences – they appreciate someone to talk to about what has happened and to help them look at the future. Children also welcome meetings with those in their own age group affected by stroke. Both parents and children explained that the diagnosis of stroke is traumatic and that they need help in adjusting. It is important for families to see that life goes on. At the end of this document there is a list of useful organisations, including support groups, which may be able to provide information and support.

Families commented on significant gaps in communication between the various professional teams involved in the care of a child affected by stroke, and, as discussed above, between professionals and the family. This is an area which is extensively addressed in the guidelines.

There are significant problems in accessing therapy following childhood stroke in some areas. It is important that there is a coherent plan for rehabilitation which takes into account all of the child's needs and which can be met within local resources. Gaps in service provision should be highlighted and brought to the attention of service planners.

Introduction

1 Introduction

1.1 Background

Childhood stroke is a neglected area, with both professionals and the general public lacking awareness of the problem and its potential consequences. Stroke affects several hundred children in the UK each year and is one of the top ten causes of childhood death (Fullerton *et al* 2002). Many children who have a stroke have another medical condition (such as a cardiac disorder or sickle cell disease) and, therefore, are already vulnerable to adverse neurodevelopmental effects (Lanthier *et al* 2000, Ganesan *et al* 2003). The prevalence of sickle cell disease varies widely within the United Kingdom. However, it is noteworthy that at least 10% of these children and young people will have a stroke during childhood. The burden of childhood stroke on the health services is, numerically, smaller than stroke in the elderly. However, the long-lasting physical, emotional and social effects of stroke on an individual near the beginning of their life affect not only the individual themselves, but also their family and society as a whole.

Many professional agencies can be involved in helping the affected child fulfil their potential and in providing support and advice to the family. These agencies may change in the course of the child's life and it is important that they are all aware of the consequences of childhood stroke, and that their efforts are co-ordinated. The child's cognitive, social and emotional needs are in constant evolution and the functional impact of childhood stroke may, as a consequence, vary over time.

We have taken a child-centred approach to formulating these guidelines, working in partnership with children, families and support groups, specifically seeking the views of children and families, and centring the guidelines on issues raised by them. Throughout this document use of the term 'parents' is intended to encompass the child's parents and any other carers.

The large number of consensus statements and good practice points in these guidelines emphasise that research in the field of childhood stroke is urgently needed to provide definitive answers to many of the issues raised. There is an acknowledged need for multicentre collaboration in such research to enable the design of studies with sufficient power to produce definitive results. The networks necessary for this are beginning to be established and may lead to work which could provide a firmer evidence base for the care of children affected by stroke.

Participant in paediatric stroke workshop: *I am not a stroke, I have had one.*

1.2 Scope of the guidelines

These guidelines will primarily address the diagnosis, investigation and management of acute arterial ischaemic stroke in children beyond the neonatal period (aged one month to 18 years at time of presentation), including acute presentation and management, rehabilitation and longer-term care. Many of the issues covered here, in particular those relating to rehabilitation, will also be relevant to children with other causes of stroke (for example cerebral venous infarction, neonatal stroke or intracranial haemorrhage).

1.3 Purpose of the guidelines

These guidelines are aimed at professionals working in primary care, secondary level acute and community paediatrics, tertiary level paediatric neurology and neurodisability, education, and social services. The aim of the guidelines is to provide evidence-based recommendations for clinicians.

1.4 Methodology

These guidelines were formulated in accordance with the principles specified by the Appraisal of Guidelines Research and Evaluation (AGREE) collaboration (**www.agreecollaboration.org**)

Following the publication of the National Clinical Guidelines for Stroke in 2000 (Intercollegiate Working Party for Stroke 2000) – referred to hereafter, along with the second edition (Intercollegiate Stroke Working Party 2004), as the 'adult guidelines' – several parties approached the Royal College of Physicians inquiring about guidelines for childhood stroke. The British Paediatric Neurology Association instigated a working party to formulate guidelines. This work was done in collaboration with the Clinical Effectiveness and Evaluation Unit of the Royal College of Physicians. Potential members were identified through their recognised record of clinical and research activity in the field of paediatric stroke and also through their professional organisations. Representation was sought across a broad range of disciplines and two patient organisations (the Stroke Association and Different Strokes). The members of the working party are listed at the front of this book. Conflicts of interest were declared and monitored (and full statements held on file).

The working party began by constructing a list of headings using the existing adult stroke guidelines (Intercollegiate Working Party for Stroke 2000) as a reference. We also considered specific additional issues relevant to children (for example, return to school). For each area, the group decided on a list of specific questions that would be considered. Searches were done using key words relevant to these questions of available computerised databases from 1966 onwards: Medline, AMED, CINAHL and Embase. In addition, the Cochrane Collaboration database was searched and other national guidelines and publications were reviewed. Members of the working party brought their own expertise and knowledge of the literature, as well as information from their organisations and professional bodies.

Topics were divided and allocated to individual members for evaluation according to their expertise. These individuals had responsibility for appraising the evidence and drafting the recommendations. The Scottish Intercollegiate Guidelines Network (SIGN 50) guidelines appraisal checklists were used to assess the quality of published articles (**www.sign.ac.uk/guidelines**). Guidelines were written on the basis of the available evidence with grading of the strength of the recommendation and explanatory statements where necessary. All recommendations were then presented to the working party as a whole for discussion and agreement.

Selection of articles for inclusion was based on the following principles. Where evidence specifically relating to childhood stroke was available, this alone was used. However, such literature is extremely limited and, therefore, research from other paediatric neurological conditions was evaluated where these conditions were felt to be relevant to the issues being considered. If a recommendation was based on extrapolation from research in a different population to that covered by the guideline, the grade of recommendation was reduced by one level.

Where evidence from meta-analyses or randomised controlled trials (RCTs) was available, this was used. Where there was limited or no evidence from RCTs, then evidence from observational group studies or small-group studies was used. In general, evidence from single-case studies was not used, primarily because it is difficult to draw general conclusions from them. Where there was no evidence base to support guidelines in areas which were highly relevant to clinical practice, consensus statements from this working party, other working parties and professional bodies were used. Many recommendations are in line with those in the developing National Service Framework for Children (**www.dh.gov.uk/PolicyAndGuidance/HealthAndSocialCareTopics/ChildrenServices/fs/en**).

The strength of evidence and recommendations were graded using the scheme proposed by SIGN 50 and summarised in tables 1.1 and 1.2, overleaf. The 'Evidence' sections following guidelines give an indication of the nature and extent of the supporting evidence, together with key references. Lastly, for each topic, there is an evidence table or group of evidence tables giving further details of the main studies.

Children affected by stroke and their parents were invited to attend a structured workshop in order to identify areas they thought should be addressed within the guidelines. The findings were used to identify key themes, which were subsequently incorporated into the issues addressed by the guidelines.

All the guidelines have been peer reviewed by external reviewers, a group which included a range of stakeholders (see Appendix 1).

Table 1.1 Guideline strength: levels of evidence

Level of evidence	Type 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 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, eg case reports, case series
4	Expert opinion

Table 1.2 Guideline strength: grades of recommendation

Grade of recommendation	Evidence
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+
√	A tick after a guideline represents a 'good practice point' – the recommended best practice based on the clinical experience of the guideline development group

1.5 Context and use

These guidelines are intended to inform clinical decisions rather than to be rigidly applied.

2 Terminology and theoretical framework

A factor interfering with delivery of good stroke care for children is the lack of a widely accepted framework of care and universal vocabulary and terminology. Health professionals have widely varying experiences of childhood stroke, but the problem is exacerbated by the variation in expertise and preferences for intervention in the absence of an accepted framework. The revised World Health Organization (WHO) classification (International Classification of Functioning, Disability and Health (ICF)) is intended to include all aspects of the health of an individual throughout life. This is designed to replace the former ICIDH (International Classification of Impairments, Disabilities and Handicap), which was widely accepted and used in describing health.

2.1 The International Classification of Functioning, Disability and Health

ICF classifies health and health-related states. The unit of classification is 'categories' within health and health-related domains. It is important to note, therefore, that in the ICF persons are not the units of classification; that is, ICF does not classify people, but describes the situation of each person within an array of health or health-related domains. Moreover, the description is always within the context of environmental and personal factors. This interaction can be viewed as a *process* or a *result* depending on the user. For an example of how the ICF might be used in practice, see Appendix 3.

Overview of ICF components

In the context of health:

- **body functions** are the physiological functions of body systems (including psychological functions)
- **body structures** are anatomical parts of the body such as organs, limbs and their components
- **impairments** are problems in body function or structure such as a significant deviation or loss
- **activity** is the execution of a task or action by an individual
- **participation** is involvement in a life situation
- **activity limitations** are difficulties an individual may have in executing activities
- **participation restrictions** are problems an individual may experience in involvement in life situations

- **environmental factors** make up the physical, social and attitudinal environment in which people live and conduct their lives.

An overview of these concepts is given [in the table below]. As the table indicates:

- ICF has two *parts*, each with two *components:*

  Part 1. Functioning and Disability

  (a) Body Functions and Structures

  (b) Activities and Participation

  Part 2. Contextual Factors

  (a) Environmental Factors

  (b) Personal Factors

- Each component can be expressed in both *positive* and *negative* terms...

An overview of ICF

	Part 1: Functioning and Disability		**Part 2: Contextual Factors**	
Components	Body Functions and Structures	Activities and Participation	Environmental Factors	Personal Factors
Domains	Body functions Body sructures	Life Areas (tasks, actions)	External influences on functioning and disability	Internal influences on functioning and disability
Constructs	Change in body functions (physiological) Change in body structures (anatomical)	Capacity Executing tasks in a standard environment Performance Executing tasks in the current environment	Facilitating or hindering impact of features of the physical, social and attitudinal world	Impact of attributes of the person
Positive Aspect	Functional and structural integrity	Activities Participation	Facilitators	not applicable
	Functioning			
Negative Aspect	Impairment	Activity limitation Participation restriction	Barriers / hindrances	not applicable
	Disability			

Source: World Health Organization (2001) *ICF: International Classification of Functioning Disability and Health*. WHO: Geneva. Available at: **www3.who.int/icf/icftemplate.cfm**

Guidelines

1 Each team should use a consistent framework and terminology in providing care to the child affected by stroke √

2 It is recommended that the World Health Organization's International Classification of Functioning (ICF) terminology is used √

Evidence

1 & 2 Working party consensus

The guidelines

3 Service organisation

Children affected by stroke make use of all levels of health, education and social services in the United Kingdom. From a medical perspective the patient journey can be considered in terms of acute medical care, and both acute and longer-term rehabilitation. Rehabilitation should be integrated with the child's educational, social and emotional needs.

In contrast to adult stroke, where the model is to develop specialist stroke services, the relative rarity of childhood stroke means that existing primary, secondary and tertiary systems of child health will – appropriately – be involved. These services and their potential roles are outlined in Table 3.1, overleaf. Services for the rehabilitation and longer-term needs of children with any acquired brain injury, including stroke, are relatively underdeveloped in the United Kingdom; in fact, their care challenges services and processes in both health and education, which are typically built around the needs of children with much more stable, slowly changing requirements (eg those with cerebral palsy). Although the development of more accurately targeted services is being proposed, the following section will describe the potential roles, and involvement in the management of children affected by stroke, of services as they are currently structured. The wide variation in the nature and potential severity of the long-term effects of stroke means that it is difficult to propose a single approach which would be suitable for all children, and the applicability of each recommendation to the specific child and family should be considered.

It is our view that all children affected by acute stroke should be referred to a consultant paediatric neurologist. However, it may not always be appropriate for the child to be transferred to an acute paediatric neurology unit. If this is the case, the child's management should be discussed with the tertiary level paediatric neurology service. At present many, but not all, tertiary paediatric neurology units have multidisciplinary teams with expertise in the evaluation of children with acquired neurological problems. However in other tertiary centres, and many secondary centres, formal acute-based teams do not exist and are convened on an *ad hoc* basis. Where specialist expertise is not available locally professionals are encouraged to liaise with, and obtain advice from, colleagues in specialist centres. The needs of the child must remain central to the consideration of which professionals to involve.

The lack of structured paediatric rehabilitation services could be attributed to i) a lack of research regarding long-term outcomes of acquired brain injury in childhood, ii) a misplaced optimism regarding the plasticity of the child's brain and the potential for recovery, iii) a lack of appreciation of the developmental context, and the fact that effects not apparent immediately may emerge with time, iv) a lack of recognition of the 'invisible' consequences of brain injury (for example, cognitive or emotional effects). These factors all need to be taken into account when considering the services available to children affected by stroke. The

Table 3.1 Composition of current paediatric services in the United Kingdom and potential roles in the care of children affected by stroke

Service	Professionals	Roles
Tertiary care services		
Specialist children's hospital	– Paediatric neurologist – Nursing staff – Allied health professionals (occupational therapist, physiotherapist, speech and language therapist) – Clinical psychologist – Social worker – Other tertiary level paediatric specialists	– Establish diagnosis – Acute medical (or surgical) treatment – Early disability assessment and treatment during inpatient period – Liaison with secondary acute and community services (including provision of advice and support to secondary services after discharge)
Specialist children's rehabilitation unit	– Paediatrician with neurodisability or rehabilitation training – Nursing staff – Allied health professionals – Educational psychologist – Teacher – Child and adolescent mental health professionals	– Assessment of impairment and disability – Rehabilitation – Plan for transition to community services – Liaison with secondary acute and community services (including provision of advice and support to secondary services after discharge)
Secondary care services		
Acute paediatrics	– Consultant paediatrician – Nursing staff – Allied health professionals – Social worker – Teacher	– Establish diagnosis – Acute medical treatment – Early disability assessment and treatment during inpatient period – May take on longer-term rehabilitation depending on availability of local services – Liaison with tertiary hospital and community services
Community child health	Child development service usually includes (*Standards for child development services* (RCPCH 1999)): – Consultant paediatrician – Community nurse – Allied health professionals – Clinical psychologist – Social worker – Portage worker – Teacher – Child psychiatrist – Educational psychologist	– Assessment of impairments and disabilities – Set up and deliver long-term package of care – Liaison with educational and social services, secondary and tertiary hospital paediatric services
Primary care services		
General practice	– General practitioner – Health visitor – Community nurse	– Ongoing developmental surveillance – Management of general medical issues – Liaison with secondary and tertiary care services as required

proposed national working group to develop a paediatric rehabilitation policy would be highly relevant to children affected by stroke. At present there are only 50 specialist paediatric rehabilitation beds in the UK. This means that the rehabilitation of the majority of children affected by stroke will take place either in the community or on general paediatric wards.

Multidisciplinary assessment and co-ordination, and the provision of long-term care, are usually undertaken by community child health services, most often by the child development service. It is also at this level that ongoing liaison between health, social and education services should occur. In many areas there will be a specific team, usually based in a child development centre, responsible for children aged five and under with disabilities (*Standards for child development services* (RCPCH 1999)). The community child health service, alongside professionals in the education services, will also be involved in the management of school age children. It is important that services are not duplicated and that all those involved are clear on who is taking the lead.

Primary care services are usually involved in general health issues, and the child's general practitioner should be routinely and regularly informed by tertiary and secondary services of a child's health and the services they are using. The health visitor may play an important co-ordinating role within the multidisciplinary team.

Effective multi- and inter-agency working is essential to ensure comprehensive care in the rehabilitation of children with acquired brain injury. This is also emphasised in several documents, for example *Together from the start* (DfES 2002), and the *Standards for child development services* (RCPCH 1999). Developments in information sharing resources, such as the forthcoming Integrated Children's System (DfES) should facilitate multi- and interagency working.

The aim of team-working is to provide a smooth, coordinated and integrated service for children and their families. A 'team' is defined as a group of people working towards a single goal or set of goals, but it is important that this is an interactive effort. The aim of the multidisciplinary team is to provide a holistic perspective of the child and family in planning or providing interventions, and to stop any duplication of questions, assessments or services.

The model of having a key worker for the child and family is controversial and has not been researched in this group of children. However, documents relating to the management of children with disability (for example *Together from the start* (DfES 2002) and *Standards for child development services* (RCPCH 1999)) as well as the recent green paper, *Every child matters* (DH 2003b), advocate such a model. Given the complex and evolving nature of the potential consequences of childhood stroke and the multitude of agencies which could be involved, we feel that a key worker is likely to increase the likelihood of delivering a co-ordinated care package. A key worker is defined as a person who 'works in partnership with the family, with the function of co-ordinating service provision and serving as a point of reference for the family' (*Together from the start*, DfES 2002). A further critical aspect of this role is that the key worker takes responsibility for ensuring delivery of the package of care. Any professional could take on the role of key worker, but it is likely to be most appropriate that this is a member of the secondary level team. Additional factors which should influence the choice of the key worker are the preference of the child and family and the key worker's competencies. The family should be given clear information about the identity and role of their key worker.

Guidelines

1 All children with acute stroke should be referred to, or have their management discussed with, a consultant paediatric neurologist √

2 Where specialist expertise is not available locally, professionals from all disciplines are encouraged to liaise with, and obtain advice from, colleagues in specialist centres regarding the acute assessment and management of the child affected by stroke √

3 Care should be provided in an environment that is appropriate for the child's age and developmental level (**D**)

4 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**)

5 All members of the healthcare team should work together with the child and family, using an agreed therapeutic approach (**D**)

6 The longer-term management of the child affected by stroke should be co-ordinated by a consultant paediatrician √

7 A multidisciplinary team with expertise in the care of children with neurological conditions should be involved in the management of the child affected by stroke. Whilst this may initially be at tertiary level, it is essential that the relevant secondary level child development service is involved from an early stage √

8 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 (**D**). The key worker and their role should be explained to the family

Evidence

1 Consensus of working party

2 Consensus of working party

3 Recommendation 18 of *Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal infirmary 1984–1995* (**www.bristol-inquiry.org.uk**); Children's National Service Framework (**www.dh. gov.uk**) (**4**)

4 *Together from the start* (DfES 2002) (**4**); *Standards for child development services* (RCPCH 1999) (**4**)

5 *Standards for child development services* (RCPCH 1999) (**4**)

6 Consensus of working party

7 Consensus of working party

8 *Together from the start* (DfES 2002) (**4**); *Standards for child development services* (RCPCH 1999) (**4**)

4 Children and their families

4.1 Consent

Children, whatever their age, have a right to be consulted and informed about any proposed treatment. The UN Convention on Children's Rights recognises the right of children to make informed decisions. Information (either verbal or written) needs to be accessible to children. Their dignity, self-respect, and rights to self-determination and non-interference should be preserved (**www.unicef.org/crc/crc.htm**).

The Children Act 1989 and European Association for Children in Hospital charter (**www.each-for-sick-children.org/charter.htm**) require that children and parents participate in decision-making. Children's feelings and wishes should be sought and taken into account, and any reasons for not following them should be explained. Religious persuasion, racial origin, culture and language should also be considered. Children should be protected from unnecessary treatment and interference. The Gillick Judgement (*Gillick* v *West Norfolk Health Authority* 1985) requires that consent be given by a child if they have 'sufficient understanding and intelligence to enable understanding fully what is proposed, even if under the age of consent'.

4.2 Families and carers

The child and family perspective

'I wanted to know… if I could use my hand normally, how long it would take to heal' (participant in paediatric stroke workshop).

Parents of affected children had experienced *'not being told what is going on'* and *'being kept in the dark'*. Parents and carers also reported feelings of helplessness, distress and guilt from witnessing their children's pain and fear and being unable to help them: *'I felt helpless, I couldn't do anything for her', 'I rushed … to the hospital, all I could hear was my niece screaming … I cried, saying to myself she's only 15 years old', 'Seeing your child suffering and feeling guilty'.*

Parents found it hard when they felt that their knowledge of their child was ignored: *'The staff at hospital who did not listen to mother', 'Not having your mothering/fathering instincts listened to'*. Parents suggested that it would be helpful if doctors could talk to them using more accessible language when explaining what is wrong with their child: *'Getting doctors to explain the child's condition in layman terms, not "doctor speak"'*.

Parents also reported significant emotional problems following their child's stroke and

suggested that meeting other parents would be very beneficial: *'We have at times both seriously contemplated suicide', 'Parents benefit from meeting other parents'.*

The majority of parents felt that they have to constantly fight public services so their child can receive the care and treatment that they should be getting: *'Having to constantly fight for the justice of your child', 'Having to fight/ask for help when you are so vulnerable'.*

All quotes are from participants in the paediatric stroke workshop.

Communication with the child and their family

The recognition that the child is part of a family is central to paediatric care. Any childhood illness has an impact on the whole family, including parents, siblings and grandparents. Childhood stroke has been shown to have an adverse impact on parents' emotional and physical health (Gordon *et al* 2002).

Stroke is a completely unexpected illness in a child and parents and children feel emotionally devastated by the diagnosis. This is compounded by the lack of awareness of childhood stroke amongst professionals, which means that it is often left to the child and family to pursue treatment, rehabilitation and appropriate educational support. All professionals should be aware of the stress associated with a diagnosis of stroke on the child and family from the outset. The importance of emotional support and sensitive and comprehensive communication at the time of diagnosis of a disorder with potential long-term developmental consequences is emphasised in *Together from the start* (DfES 2002) and the *Standards for child development services* (RCPCH 1999).

The Stroke Association and Different Strokes (see Appendix 4 for contact details) provide information and support for children affected by stroke and their families. Children, family members or carers need both factual and practical information at various stages, presented in a format appropriate to their needs (Rushforth 1999; Helps *et al* 2003). It should be recognised that parents have particular knowledge of their child and, therefore, their concerns should be addressed in planning the child's care and educational placement.

Guidelines

1 Families/carers should be given factual information about their child's condition as soon as possible after diagnosis (**D**). This should be simple and consistent, avoiding technical terms and jargon

2 Written information should be provided to the child and family regarding the child's health and the statutory and voluntary services available (**D**)

3 Children should be given information about their condition at an appropriate level (**D**)

4 The child and family should be involved in making decisions about the child's care, including rehabilitation and education (**D**)

5 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**)

Evidence

1 Paediatric stroke workshop (**4**); *Together from the start* (DfES 2002) (**4**); *Standards for child development services* (RCPCH 1999) (**4**)

2 *Together from the start* (DfES 2002) (**4**); Paediatric stroke workshop (**4**)

3 Rushforth 2002 (**4**)

4 Report of Bristol enquiry (**www.bristol-inquiry.org.uk**) (**4**); *Together from the start* (DfES 2002) (**4**); *Standards for child development services* (RCPCH 1999) (**4**); *Children's National Service Framework* (DH 2003a) (**www.dh.gov.uk**) (**4**)

5 *Consensus of working party* (**4**); Mukherjee *et al* 1999 (**4**)

5 Acute diagnosis of arterial ischaemic stroke in children

5.1 Definition

The World Health Organization defines stroke as 'a clinical syndrome typified by rapidly developing signs of focal or global disturbance of cerebral functions, lasting more than 24 hours or leading to death, with no apparent causes other than of vascular origin' (World Health Organization 1978). This definition is a *clinical* one and such a presentation has many potential underlying causes in childhood. Brain imaging is mandatory for accurate diagnosis, subsequent referral and, in particular, to exclude conditions requiring urgent neurosurgical intervention. Arterial ischaemic stroke, which is the main focus of these guidelines, can be defined as 'a clinical stroke syndrome due to cerebral infarction in an arterial distribution'. Transient ischaemic attacks (TIAs) (where the neurological deficit resolves within 24 hours) may also occur in children. Although clinical symptoms may be transient, a significant proportion of children with this presentation have cerebral infarction. Terms such as 'acute infantile hemiplegia' are clinical descriptions, which do not identify the underlying aetiology; they should, therefore, be avoided.

The following sections will deal, firstly, with guidelines for establishing a diagnosis in a child presenting with an acute clinical stroke syndrome (section 5.2), and then discuss the further investigation of children with a diagnosis of arterial ischaemic stroke in order to establish underlying aetiology (section 5.3). It may be pragmatic to combine the initial (diagnostic) and subsequent investigations, especially in the case of imaging, and both sections should, therefore, be considered together.

5.2 Presentation and diagnosis

At the time of stroke children and families reported feelings such as 'frightened', 'annoyed', 'angry', 'confused', 'devastated'.

Parents reported feeling concerned and frightened at the amount of time they had to wait for diagnosis, treatment and information about their child's condition: 'Sitting for hours in the emergency department, with ———, before it was finally acknowledged she had had a stroke' (parent participant in paediatric stroke workshop).

Recognition of clinical stroke may be difficult, particularly in infants and young children, and especially as neurological signs may be relatively subtle. If there is doubt, the child should be examined by a senior paediatrician. The most common clinical presentation of clinical stroke in childhood is with acute hemiparesis. Focal signs may be absent in neonates or young infants, in whom seizures may be the only manifestation of clinical stroke. Clinical symptoms and signs of arterial ischaemic stroke may be particularly subtle in children with sickle cell disease, and may be difficult to distinguish from painful crisis or the effects of treatment, for example treatment with opiates. Advice should be sought from a tertiary centre if there is concern about the acquisition and interpretation of imaging studies in a child with clinical stroke.

Guidelines

1 All children with a clinical presentation of stroke should be under the care of a consultant paediatrician √

2 Cross-sectional brain imaging is mandatory in children presenting with clinical stroke (**C**)

3 Brain magnetic resonance imaging (MRI) is recommended for the investigation of children presenting with clinical stroke (**C**)

4 Brain MRI should be undertaken as soon as possible after presentation. If brain MRI will not be available within 48 hours, computed tomography (CT) is an acceptable initial alternative √

5 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 √

6 Any new neurological symptoms or signs in children with sickle cell disease should be evaluated as potentially being due to stroke √

7 All children with clinical stroke should have regular assessment of conscious level and vital signs √

Evidence (Tables 1 and 2)

1 Consensus of working party

2 Ganesan *et al* 2003 (**2+**)

3 Bryan *et al* 1991 (**2+**); Kucinski *et al* 2002 (**2+**); Barber *et al* 1999 (**2–**); Lansberg *et al* 2000 (**2–**)

4 Consensus of working party

5 Consensus of working party

6 Consensus of working party

7 Consensus of working party

5.3 Investigations

This section aims to provide some guidance about investigations in the evaluation of a child with arterial ischaemic stroke; it is not, however, intended to be comprehensive. There are many potential risk factors for arterial ischaemic stroke in children and the diagnostic process should be directed towards identifying as many of these as possible. The proportion of patients in whom no risk factors are identified has decreased as understanding of aetiology and investigation methods have improved. There is little information on the diagnostic sensitivity of individual investigations. Although the investigations discussed below should be undertaken in all cases, other investigations may be indicated in individual patients, and should be considered on a case-by-case basis. For a more complete discussion of this topic see Kirkham 1999. A clerking checklist is provided to highlight important aspects of the clinical history and examination.

Transfer to a tertiary centre may be necessary if facilities for definitive imaging or other investigations (eg echocardiography) are not available locally. As mentioned in relation to brain imaging, advice should be sought from a tertiary centre if there is concern about the acquisition and interpretation of paediatric echocardiography.

Non-invasive cerebrovascular imaging with techniques such as MR angiography (Husson *et al* 2003), CT angiography, ultrasound with Doppler techniques or a combination of such modalities can be applied in the first instance, and may be adequate. The existing research on paediatric arterial ischaemic stroke only includes studies limited to visualisation of the arterial vasculature between the distal common carotid artery and the circle of Willis. The value of imaging the aortic arch and its proximal main branches is unknown. It is acknowledged that, in some cases, non-invasive angiographic techniques alone will not provide sufficient information to enable the planning of subsequent management, and in these cases catheter cerebral angiography may also be required. Due to the lack of specific research evidence we have not made more detailed recommendations regarding imaging sequences but a helpful discussion of these can be found in the review article by Hunter (Hunter 2002).

If there are unusual features to the identified infarct, such as the anatomical location, the presence of excessive brain swelling and then the possibility of venous infarction or haemorrhage should be considered. More specific venous imaging investigations may then be applied by the radiologist as necessary. In the first instance, non-invasive options such as MR venography or CT venography are preferred over catheter angiography.

The yield of investigating children with arterial ischaemic stroke for thrombophilia is variable and will depend on factors such as ethnicity. Protein C deficiency and elevated lipoprotein(a) have been shown to be associated with an increased risk of recurrence (Strater *et al* 2002). Although the appropriate preventative treatment in affected patients is unknown, identification of a prothrombotic tendency may have other implications for the child's more general health, for example risk of venous thrombosis. Additional specific investigations to be included when screening for thrombophilia should be discussed with the local haematology service, with consideration of the local prevalence of specific thrombophilia.

The importance of more conventional childhood stroke risk factors in children with sickle cell disease has not been evaluated. The clinical experience of the working party is that these may play a role in some patients and therefore we would not exclude children with sickle cell disease from the recommendations below.

Guidelines

1 Imaging of the cervical and proximal intracranial arterial vasculature should be performed in all children with arterial ischaemic stroke (C)

2 Imaging of the cervical vasculature to exclude arterial dissection should be undertaken within 48 hours of presentation with arterial ischaemic stroke √

3 Transthoracic cardiac echocardiography should be undertaken within 48 hours after presentation in all children with arterial ischaemic stroke √

4 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)

Evidence (Tables 3 and 4)

The papers cited here provide information about risk factors associated with childhood arterial ischaemic stroke, but do not all provide direct information about sensitivity or specificity of specific diagnostic tests in the context of childhood arterial ischaemic stroke.

1 Levy *et al* 1994 (**2+**); Ganesan *et al* 2002 (**2+**); Husson *et al* 2003 (**2+**); Ganesan *et al* 1999 (**3**)

2 Consensus of working party

3 Consensus of working party

4 Nowak Gottl *et al* 1999 (**2+**); deVeber *et al* 1998b (**2+**); Strater *et al* 2002 (**2+**); Subcommittee for Perinatal and Pediatric Thrombosis of the Scientific and Standardization Committee of the International Society of Thrombosis and Haemostasis (**4**)

Evidence

1 Extrapolation from *National clinical guidelines for stroke: second edition* (Intercollegiate Stroke Working Party 2004) (**4**)

2 Consensus of working party; *National clinical guidelines for stroke: second edition* (Intercollegiate Stroke Working Party 2004) (**4**)

6.2 Specific medical treatments

There are no studies specifically examining the efficacy of acute treatments for arterial ischaemic stroke in children. The following recommendations are based on the consensus opinion of the working party.

The use of anticoagulation in children with cardiac embolism is controversial as it involves balancing the risk of precipitating haemorrhagic transformation of the infarct with the potential to prevent further embolic events. The decision may be influenced by the cardiac pathology, time elapsed after the stroke and by neurological and imaging findings. In the absence of any evidence, we were unable to make a general recommendation, but felt that individual patient management should involve senior clinicians in paediatric cardiology and neurology.

The efficacy and optimal dose of aspirin in the treatment of children with acute arterial ischaemic stroke is unknown. The lowest dose recommended for treatment of other paediatric conditions, such as Kawasaki disease, in the paediatric formulary *Medicines for children* (Royal College of Paediatrics and Child Health 2003) is 5 mg/kg/day. This would approximate to the dose (300 mg) recommended for acute treatment of ischaemic stroke in adults (*National clinical guidelines for stroke: second edition*, Intercollegiate Stroke Working Party 2004) and therefore has been recommended below. The lowest effective dose for long-term prophylaxis may be lower, as discussed in the following section. Although children with sickle cell disease have been excluded from the first guideline, aspirin or anticoagulation may need to be considered if other risk factors, for example arterial dissection, are identified in individual patients.

There is currently no evidence to support use of thrombolytic agents such as tissue plasminogen activator (tPA) in the acute treatment of arterial ischaemic stroke in children.

Guidelines

1 Aspirin (5 mg/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 √

2 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 10–12.5 g/dl √

ii if the patient has had a neurological event in the context of severe anaemia (eg splenic sequestration or aplastic crisis), or if exchange transfusion is going to be delayed for more than four hours, urgent top-up blood transfusion should be undertaken √

3 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 √

ii cerebral venous sinus thrombosis (C)

4 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 √

5 Early neurosurgical referral should be considered in children with stroke who have depressed or deteriorating conscious level or other signs of raised intracranial pressure √

Evidence (Table 5)

1 Working party consensus

2 Working party consensus

3 i) Working party consensus; ii) Extrapolation from Stam *et al* 2003 (**1–**) (level of recommendation downgraded from B to C due to small number of studies in Cochrane review (n = 2) and based on adult data)

4 Working party consensus

5 Working party consensus

6.3 Secondary prevention of arterial ischaemic stroke in childhood

Child in paediatric stroke workshop: *'What happens if you get two strokes?'*

Stroke recurrence is a major concern for children and their families. Arterial ischaemic stroke recurs in between 6% and 20% of all children and in over 60% of children with sickle cell disease. The risk of recurrence is increased in children with multiple risk factors (Lanthier *et al* 2000) and in those with protein C deficiency, increased levels of lipoprotein (a) and vascular disease (Sträter *et al* 2002). At present, there is very little evidence regarding the efficacy of secondary prevention strategies (Sträter *et al* 2001) but this is likely to change as the need for multicentre trials is gaining momentum. Thus we would emphasise that the consensus statements below are not long-term recommendations and will need to be updated as new evidence emerges.

Although widely used, the dose of aspirin to be used for secondary prevention of ischaemic stroke is undefined in childhood. Doses between 50–300 mg/day are recommended for adults (*National clinical guidelines for stroke: second edition* Intercollegiate Stroke Working Party 2004) and doses between 1–3 mg/kg/day have been recommended for secondary prevention in children (Nowak-Gottl *et al* 2003). Given the lack of evidence we have not been able to recommend a specific dosage; however, complications such as bruising may limit the dose which can be tolerated by the child.

Guidelines

1 Patients with cerebral arteriopathy other than arterial dissection or moyamoya syndrome or those with sickle cell disease should be treated with aspirin (1–3 mg/kg/day) √

2 Anticoagulation should be considered:
   - i until there is evidence of vessel healing, or for a maximum of six months, in patients with arterial dissection √
   - ii if there is recurrence of arterial ischaemic stroke despite treatment with aspirin √
   - iii in children with cardiac sources of embolism, following discussion with the cardiologist managing the patient √
   - iv until there is evidence of recanalisation or for a maximum of six months after cerebral venous sinus thrombosis √

3 In children with sickle cell disease:
   - i regular blood transfusion (every three to six weeks) should be undertaken to maintain the HbS% <30% and the Hb between 10–12.5 g/dl (C)
   - ii transfusion may be stopped after two years in patients who experienced stroke in the context of a precipitating illness (eg aplastic crisis) and whose repeat vascular imaging is normal at this time (C)
   - iii after three 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)

4 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)

5 Children with sickle cell disease who have had a stroke should be referred to a specialist centre for consideration of bone marrow transplantation (B)

6 Advice should be offered regarding preventable risk factors for arterial disease in adult life, particularly smoking, exercise and diet (D)

7 Blood pressure should be measured annually to screen for hypertension √

8 Patients who are found to have a prothrombotic tendency should be referred to a haematologist √

Evidence (Tables 6, 7, 8 and 9)

1 Consensus of working party

2 Consensus of working party

3 i) Powars *et al* 1978 (**2–**); Portnoy & Herion 1972 (**2–**); Balkaran *et al* 1992 (**2–**); Wilimas *et al* 1980 (**2+**); Russell *et al* 1984 (**2+**); Wang *et al* 1991 (**2+**); Cohen *et al* 1992 (**2+**); De Montalambert *et al* 1999 (**2–**); Rana *et al* 1997 (**2–**); Pegelow *et al* 1995 (**2–**); Scothorn *et al* 2002 (**2+**); Dobson *et al* 2002 (**2+**); ii) Dobson *et al* 2002 and Scothorn *et al* 2002 (**both 2+**); iii) Cohen *et al* 1992 (**2+**); iv) Ware 1999 (**2+**); Sumoza 2002 (**2+**)

4 Golby *et al* 1999; Olds *et al* 1987; George *et al* 1993; Ishikawa *et al* 1997; Matsushima *et al* 1992; Fryer *et al* 2003 (**all 3**)

5 Vermylen *et al* 1998; Bernaudin 1999; Walters *et al* 2000 (**all 2++**)

6 Consensus of working party; Extrapolation from *National clinical guidelines for stroke: second edition*, Intercollegiate Stroke Working Party 2004 (**4**)

7 Consensus of working party

8 Consensus of working party

6.4 Early disability assessment and management

The philosophy of paediatric medicine is for care to be delivered, where possible, in the child's home environment and to minimise the time a child spends in hospital. The inpatient period may be limited to the time when the child is acutely unwell or when there are outstanding diagnostic issues. It is, therefore, important that links to community child health services should be made from the outset in order for a package of care to be set up, and that the child is not discharged until this is in place. Ultimately this package may be delivered in a variety of settings, as discussed in chapter 3. Parents (and children when possible and appropriate) should be involved in planning long-term care.

Guidelines

1 As soon as possible after admission, following stroke all children should have an evaluation of:

   i swallowing safety (**D**)

   ii feeding and nutrition √

iii communication (D)

iv pain (D)

v moving and handling requirements (D)

vi positioning requirements √

vii risk of pressure ulcers (D)

2 All children affected by stroke should have a multidisciplinary assessment within 72 hours of admission to hospital √

3 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 √

Evidence (Table 10)

1 i) Swallowing guidelines of the Royal College of Speech and Language Therapists (RCSLT 2004) (**4**); Morgan *et al* 2003 (**3**); ii) Working party consensus; iii) RCSLT guidelines on assessment of acquired childhood aphasia from the RCSLT's *Communicating quality (2)* (RCSLT 2001) (**4**); iv) Royal College of Nursing's *Clinical Practice Guidelines: The recognition and assessment of acute pain in children* (RCN 2001) (**IV**); v) Royal College of Nursing's *The guide to the handling of patients: fourth edition* (RCN 1999) (**4**); vi) Working party consensus; vii) NICE guideline on pressure ulcer risk management and prevention (Guideline B) (**4**); *Working together to prevent pressure ulcers – a guide for patients and their carers* (**www.nice.org.uk/pdf/clinicalguidelinepressuresoreguidancenice.pdf**) (**4**); *Pressure relieving devices – CG7* (**www.nice.org.uk/pdf/clinicalguidelinespressuresorespatleafletenglish.pdf**) (**4**)

2 Working party consensus

3 Working party consensus