Stroke in childhood

Clinical guideline for diagnosis, management and rehabilitation

May 2017
Endorsement

Association of Ambulance Chief Executives
Association of Paediatric Chartered Physiotherapists
British Association for Community Child Health
British Academy of Childhood Disability
British Association of General Paediatrics
British Association of Stroke Physicians
British Paediatric Neurology Association
British Society of Paediatric Radiology
College of Occupational Therapists
London Ambulance Service NHS Trust

Paediatric Intensive Care Society
Royal College of Emergency Medicine
Royal College of General Practitioners
Royal College of Nursing
Royal College of Physicians
Royal College of Speech & Language Therapists
Sickle Cell & Young Stroke Survivors
UK Forum on Haemoglobin Disorders

The principles of this document are supported by The Royal College of Radiologists [June 2017]
Foreword

Stroke is a devastating disease, whether it happens in adulthood or childhood. In adults it has only been in the last few decades that it has been regarded as a disease that can be treated and this change has happened because of research showing that well organised specialist care is effective both acutely and in the rehabilitation stages of the illness. It is also mainly thought of, by the public and professionals, as a disease of old people. These guidelines are therefore important in raising awareness that stroke does happen to children and young people and in showing that there are interventions that can make a difference.

The first edition of these guidelines was published over 12 years ago, and one of the major conclusions then was a desperate need for more research into stroke in childhood. It is therefore disappointing that there has been so little progress in so many areas of stroke management. We are still in a position where most of the recommendations are based on expert consensus or weak evidence. The most important factor in stroke care in adults is being managed on a stroke unit by staff with specific expertise and interest in the disease. Stroke units have never been tested for childhood stroke and as a result are not even mentioned in these guidelines. Clot busting treatment for ischemic stroke, or clot removal, are becoming mainstays of care in adults, yet there is very little evidence for these treatments in children because research has not been done.

These guidelines are the first stage in beginning to take stroke in childhood seriously and to encourage the clinicians and research funding organisations to invest time and money in improving care for children and young people and their families.

The authors are to be congratulated on their achievement in producing this document but I hope that in another 12 years the recommendations will be more extensive and based on a much stronger evidence base.

Professor Tony Rudd FRCP CBE
Professor of Stroke Medicine, King’s College London
National Clinical Director for Stroke, NHS England
From speaking with families, I am all too aware of the huge impact a stroke in childhood can have, not just for the child but for the whole family. Too often families tell us that it took too long to get a diagnosis; this is because too few professionals know that stroke can occur in children, and that it is a constant fight to ensure that their child gets the long-term support they need. This long-term support may include physiotherapy, speech and language therapy, support with education, or help with accessing statutory benefits. Families also tell us that there is little emotional or practical support available for them, and they don’t know which health professionals they should be in touch with or what support is available. A key contact would greatly help with this.

I welcome the publication of this new guideline which gives recommendations on how to support a child affected by stroke, as well as their family, all the way from diagnosis through to long-term support. Quick diagnosis is vital and I am pleased to see this guideline clearly states that a scan should be carried out within one hour of arrival at hospital for every child with a suspected stroke.

It is particularly good news that a parent and carers’ version accompanies this guideline to help push up standards of care and ensure that families are informed and aware of what to expect and when. We need to ensure that every child with stroke receives the best possible treatment, care and support.

Thank you to everyone who has helped to develop these guidelines, especially the families affected by childhood stroke whose personal experience has been invaluable. We now need to share these guidelines widely so that good practice is uniformly adopted and more people recover and live a full and enriching life after stroke.

**Mrs Juliet Bouverie**
Chief Executive Officer, Stroke Association
Preface

It is my pleasure and privilege to write the preface for this important guideline, which gives recommendations on how best to support children who had a stroke and also importantly their family. The parent and carer input is a very welcome edition.

This clinical guideline is the most comprehensive and up to date document on how stroke care should be provided, covering the whole care pathway from identification, diagnosis and management of children and young people with arterial ischaemic stroke and haemorrhagic stroke until their transition to adult care. It is aimed at professionals working in primary care, secondary level acute paediatrics and tertiary level paediatric neurosciences, as well as those within the ambulance sector, paediatric intensive care unit, community paediatrics, neurodisability, education, and social services. It is also for those with responsibility for commissioning stroke services.

It is acknowledged that there are still many areas where the recommendations are based on expert consensus, and as such this guideline highlights the need for further research. This guideline also aims to help researchers and funding bodies to identify the key questions and areas which still need to be answered through research.

The guideline has been achieved by excellent input from professionals, researchers, and the families affected by childhood stroke. The Royal College of Paediatrics and Child Health are extremely grateful to the Stroke Association's supporters, in particular The Thompson Family Charitable Trust. We would like to say a huge thank you to all those who have been involved in producing such an important clinical guideline.

Professor Anne Greenough
Professor of Clinical Respiratory Physiology, King’s College London
RCPCH Vice President for Science & Research
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The Stroke Association is registered as a charity in England and Wales (211015) and in Scotland (SC037789); also registered in Northern Ireland (XT33805), Isle of Man (945) and Jersey (NPO 369). The Stroke Association is a ‘restricted Fund’ of the RCPCH, which is registered as a charity in England and Wales (1057744) and in Scotland (SCO38299).

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Association of British Neurologists
Association of Paediatric Chartered Physiotherapists
Association of Paediatric Emergency Medicine
British Association of General Paediatrics
British and Irish Orthoptic Society
British Academy of Childhood Disability
British Aphasiology Society
British Association for Community Child Health
British Association of Stroke Physicians
British Congenital Cardiac Association
British Dietetic Association
British Paediatric Neurology Association
British Paediatric Neurosurgical Group
British Psychological Society
British Society of Haematology
British Society of Neuroradiologists
British Society of Paediatric Radiology
Child Brain Injury Trust
College of Occupational Therapists
College of Paramedics
Connect*
Different Strokes
EKS Childhood Strokes
Fighting Strokes*
General Paediatrics College Specialty Advisory Committee
Hemihelp

Intercollegiate Stroke Working Party
London Ambulance Service NHS Trust
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National Institute for Health Research Stroke Research Network
National Stroke Nursing Forum
NHS England Programme of Care Board for Children
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Paediatric Intensive Care Society
Paediatric Neurodisability College Specialty Advisory Committee
Royal College of Emergency Medicine
Royal College of General Practitioners
Royal College of Nursing
Royal College of Physicians
Royal College of Radiologists
Royal College of Speech and Language Therapists
Scottish Stroke Allied Health Professionals Forum
Scottish Stroke Nurses Forum
Sickle Cell & Young Stroke Survivors
Society for Research in Rehabilitation
Stroke Association
The Children's Trust
UK Forum on Haemoglobin Disorders
UK Swallowing Research Group
Welsh Association of Stroke Physicians
World Stroke Organisation

*Fighting Strokes and Connect are no longer active charities; however, did participate in the consultation of the guideline scope in 2015.

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## Acronyms and abbreviations

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<thead>
<tr>
<th>Acronym</th>
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<tr>
<td>A&amp;E</td>
<td>Accident &amp; Emergency</td>
</tr>
<tr>
<td>AAC</td>
<td>Augmentative and Alternative Communication</td>
</tr>
<tr>
<td>ABI</td>
<td>Acquired brain injury</td>
</tr>
<tr>
<td>ACLA</td>
<td>Anticardiolipin antibody</td>
</tr>
<tr>
<td>ACT</td>
<td>Acceptance &amp; Commitment Therapy</td>
</tr>
<tr>
<td>AHA</td>
<td>Assisting Hand Assessment</td>
</tr>
<tr>
<td>AIS</td>
<td>Arterial ischaemic stroke</td>
</tr>
<tr>
<td>APLS</td>
<td>Antiphospholipid syndrome</td>
</tr>
<tr>
<td>aPTT</td>
<td>Activated partial thromboplastin time</td>
</tr>
<tr>
<td>ARE</td>
<td>Adverse radiation effects</td>
</tr>
<tr>
<td>ARUBA trial</td>
<td>A Randomised trial of Unruptured Brain Arteriovenous malformations</td>
</tr>
<tr>
<td>AT</td>
<td>Antitrypsin</td>
</tr>
<tr>
<td>AVM</td>
<td>Arteriovenous malformation</td>
</tr>
<tr>
<td>AVPU scale</td>
<td>‘Alert, Voice, Pain, Unresponsive’ scale</td>
</tr>
<tr>
<td>BoNTA</td>
<td>Botulinum toxin A</td>
</tr>
<tr>
<td>BP</td>
<td>Blood pressure</td>
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<tr>
<td>CA</td>
<td>Catheter angiography</td>
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<tr>
<td>CAF</td>
<td>Common Assessment Framework</td>
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<tr>
<td>CASP</td>
<td>Critical Appraisal Skills Programme</td>
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<tr>
<td>CBT</td>
<td>Cognitive Behavioural Therapy</td>
</tr>
<tr>
<td>CCC</td>
<td>Comprehensive Care Centre</td>
</tr>
<tr>
<td>CIMT</td>
<td>Constraint Induced Movement Therapy</td>
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<tr>
<td>CINAHL</td>
<td>Cumulative Index of Nursing and Allied Health Literature</td>
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<tr>
<td>CNS</td>
<td>Central nervous system</td>
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<tr>
<td>CO-OP</td>
<td>Cognitive Orientation to Daily Occupational Performance</td>
</tr>
<tr>
<td>COPM</td>
<td>Canadian Occupational Performance Measure</td>
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<tr>
<td>CP</td>
<td>Cerebral palsy</td>
</tr>
<tr>
<td>CQC</td>
<td>Care Quality Commission</td>
</tr>
<tr>
<td>CT</td>
<td>Computerised tomography</td>
</tr>
<tr>
<td>CTA</td>
<td>Computerised tomography angiography</td>
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<tr>
<td>DECIMAL</td>
<td>Decompressive craniectomy in malignant MCA infarcts</td>
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<tr>
<td>DGH</td>
<td>District General Hospital</td>
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<tr>
<td>DESTINY</td>
<td>Decompressive surgery for the treatment of malignant infarction of the MCA</td>
</tr>
<tr>
<td>DLA</td>
<td>Disability Living Allowance</td>
</tr>
<tr>
<td>DSA</td>
<td>Digital Subtraction Angiography</td>
</tr>
<tr>
<td>ECG</td>
<td>Electrocardiogram</td>
</tr>
</tbody>
</table>
ED  Emergency Department
EDAS  Encephalo-duro-arterio-synangiosis
EEG  Electroencephalogram
EHCP  Education, Health and Care Plan
EMG  Electromyographic
ENERCA  European Network for Rare and Congenital Anaemias
EPG  Electopalatography
FAST tool  ‘Face, Arms, Speech Time’ tool (indicators)
FBC  Full blood count
FCA  Focal cerebral arteriopathy
FES  Functional electrical stimulation
FLAIR  Fluid attenuation inversion recovery
FVL  Factor V Leiden
GAS  Goal Attainment Scaling
GCS  Glasgow Coma Scale
GDG  Guideline development group
GOS  Glasgow Outcome Scale
GRADE methodology  Grading of Recommendations Assessment, Development and Evaluation
HAMLET  Hemicraniectomy after MCA infarction with life-threatening edema trial
HASU  Hyperacute stroke unit
HbS  Sickle haemoglobin
HCY  Homocystinuria
HR  Heart rate
HLA  Human leukocyte antigen
HS  Haemorrhagic stroke
HSCT  Hematopoietic stem cell transplantation
ICA  Internal carotid artery
ICF  International Classification of Functioning, Disability and Health
ICH  Intracerebral haemorrhage
ICIDH  International Classification of Impairments, Disabilities and Handicaps
ICP  Intracranial pressure
ICU  Intensive care unit
INR  International normalised ratio
ITP  Immune Thrombocytopenic Purpura
IV thrombolysis  Intravenous thrombolytics
LMWH  Low molecular-weight heparin
Lp(a)  Lipoprotein(a)
<table>
<thead>
<tr>
<th>Abbreviation</th>
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<tr>
<td>MCA</td>
<td>Middle cerebral artery</td>
</tr>
<tr>
<td>MDT</td>
<td>Multidisciplinary team</td>
</tr>
<tr>
<td>MI</td>
<td>Myocardial infarction</td>
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<tr>
<td>ModASPECTS</td>
<td>Modified Alberta Stroke Program Early CT Score</td>
</tr>
<tr>
<td>MRA</td>
<td>Magnetic resonance angiogram</td>
</tr>
<tr>
<td>MRI</td>
<td>Magnetic resonance imaging</td>
</tr>
<tr>
<td>mRS</td>
<td>Modified Rankin Scale</td>
</tr>
<tr>
<td>NCCPC-PV</td>
<td>Non-Communicating Children’s Pain Checklist – Post-operative Version</td>
</tr>
<tr>
<td>NDT</td>
<td>Neurodevelopmental therapy</td>
</tr>
<tr>
<td>NHS</td>
<td>National Health Service</td>
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<tr>
<td>NICE</td>
<td>National Institute for Health and Care Excellence</td>
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<tr>
<td>NIH</td>
<td>National Institutes of Health</td>
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<td>NIHSS</td>
<td>National Institutes of Health Stroke Scale</td>
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<td>NMS</td>
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<tr>
<td>ODN</td>
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<td>PACNS</td>
<td>Primary Angiitis of the Central Nervous Dystem</td>
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<td>PedNIHSS</td>
<td>Paediatric National Institute of Health Stroke Scale</td>
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<tr>
<td>PICANet</td>
<td>Paediatric Intensive Care Audit Network</td>
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<td>PICO framework</td>
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<td>PICU</td>
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<td>Partial thromboplastin time</td>
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<td>Royal College of Paediatrics and Child Health</td>
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<tr>
<td>ROSIER</td>
<td>Recognition of Stroke in the Emergency Room</td>
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<td>RR</td>
<td>Respiratory rate</td>
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<td>RRQ</td>
<td>Recovery and Recurrence Questionnaire</td>
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<td>SAH</td>
<td>Subarachnoid haemorrhage</td>
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<td>Sickle cell anaemia</td>
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<td>Sickle cell disease</td>
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<td>Silent cerebral infarctions</td>
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<td>Scottish Intercollegiate Guidelines Network</td>
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<tr>
<td>Abbreviation</td>
<td>Full Form</td>
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<tr>
<td>SLT</td>
<td>Speech and language therapy</td>
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<td>SMART goals</td>
<td>‘Specific, Measurable, Agreed, Realistic and Time-bound’ goals</td>
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<tr>
<td>SRS</td>
<td>Stereotactic radiosurgery</td>
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<tr>
<td>SSNAP</td>
<td>Sentinel Stroke National Audit Programme</td>
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<tr>
<td>STOP</td>
<td>Stroke Prevention Trial in Sickle Cell Anaemia</td>
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<td>SWiITCH trial</td>
<td>Stroke With Transfusions Changing to Hydroxyurea trial</td>
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<td>Team Around the Child/Family</td>
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<td>TBI</td>
<td>Traumatic brain injury</td>
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<td>TCD</td>
<td>Transcranial Doppler ultrasonography</td>
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<td>TIA</td>
<td>Transient ischaemic attack</td>
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<tr>
<td>TIPS trial</td>
<td>Thrombolysis in Pediatric Stroke trial</td>
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<td>TMS</td>
<td>Transcranial magnetic stimulation</td>
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<tr>
<td>tPA</td>
<td>Tissue plasminogen activator</td>
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<td>UKHCDO</td>
<td>United Kingdom Haemophilia Centre Doctors' Organisation</td>
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<td>VF</td>
<td>Videofluoroscopy</td>
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<td>VR</td>
<td>Virtual Reality</td>
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<td>VZV</td>
<td>Varicella Zoster Virus</td>
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<td>WHO</td>
<td>World Health Organization</td>
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<tr>
<td>WISC</td>
<td>Wechsler Intelligence Scale for Children</td>
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Key recommendations

This section contains 83 of the total 261 recommendations that the RCPCH Stroke in Childhood Guideline Development Group (GDG) have developed and felt key. If followed, these key recommendations will enhance the quality of stroke care in children and young people (aged 29 days to 18 years at time of presentation).

This section should not be read in isolation, and individuals should always consider the guideline in full. A downloadable list of the full guideline recommendations can be found at www.rcpch.ac.uk/stroke-guideline.

Acute diagnosis of stroke in childhood (Chapter 3)

Clinical presentation (Chapter 3.1)

- Use the FAST (‘Face, Arms, Speech Time’) criteria to determine stroke in children and young people, but do not rule out stroke in the absence of FAST signs.

- Undertake urgent brain imaging of children/young people presenting with symptoms (e.g. acute focal neurological deficit, aphasia, or a reduced level of consciousness).

To access full recommendations, see Chapter 3.1 here.

Diagnosis (imaging) (Chapter 3.2)

- Ensure that a cranial computerised tomography (CT) scan is performed within one hour of arrival at hospital in every child with a suspected stroke; including computerised tomography angiography (CTA), if the CT scan does not show haemorrhage, OR CTA limited to intracranial vascular imaging, if haemorrhagic stroke (HS) is demonstrated.

- Initial scan images should be reviewed on acquisition and if necessary transferred immediately to the regional paediatric neuroscience centre for review.

To access full recommendations, see Chapter 3.2 here.

Referral pathways and further investigations (Chapter 4)

Referral and care pathway for childhood stroke (Chapter 4.1)

- Children and young people seen by ambulance clinicians, or primary care providers outside hospital with the sudden onset of acute focal neurological symptoms should be screened for hypoglycaemia with a capillary blood glucose, and for stroke using a simple screening tool such as FAST. Where these are normal or negative, but stroke is still suspected, the acute stroke pathway should be used.

- Children and young people with persisting neurological symptoms who screen positive using a validated tool (or who screen negative, but in whom stroke is
suspected) should be transferred to an emergency department with paediatric services urgently.

To access full recommendations, see Chapter 4.1 here.

Acute management (chapter 5)

Acute assessment (Chapter 5.1)

- Use the Paediatric National Institute of Health Stroke Scale (PedNIHSS) and age-appropriate Glasgow Coma Scale (GCS) or AVPU (‘Alert, Voice, Pain, Unresponsive’) to assess the child’s neurological status and conscious level respectively.

To access full recommendations, see Chapter 5.1 here.

Framework for early functional assessment (Chapter 5.2)

- Provide clinical assessment of a child’s body structures and functions and activities, by members of the relevant hospital multidisciplinary team (MDT) (including occupational therapists, physiotherapists, speech and language therapists), as soon as possible during hospital admission (within 72 hours), with consideration of the child’s age and developmental abilities.
- Initiate early liaison with community-based medical, nursing, occupational therapists, physiotherapists, psychologists, orthoptists, speech and language therapists and other allied health professionals to establish links with local networks.

To access full recommendations, see Chapter 5.2 here.

Prevention, identification and management of complications (Chapter 5.3)

- Be aware of possible complications after arterial ischaemic stroke (AIS)/HS, as listed in the full recommendations.

To access full recommendations, see Chapter 5.3 here.

Arterial Ischaemic Stroke (Chapter 6)

Risk factors for AIS and recurrent AIS (Chapter 6.1.1)

Risk factors for first AIS

- Be aware that certain conditions/factors are associated with an increased risk of AIS in children/young people, as listed in the full recommendations.
Risk factors for recurrent AIS

- Be aware of increased risk of recurrence in children/young people with AIS and the following risk factors: arteriopathy, moyamoya, arteriopathy in sickle cell disease, congenital heart disease, thrombophilia, low birthweight.

To access full recommendations, see Chapter 6.1.1 here.

Follow-up imaging in AIS (Chapter 6.1.3)

- Be aware that magnetic resonance imaging (MRI) is the modality of choice for follow-up imaging of children and young people with AIS as it provides the best assessment of the extent of any permanent structural damage and of the cerebral circulation without using ionising radiation.
- Catheter angiography (CA) should be undertaken in children and young people with occlusive arteriopathy, who are being considered for revascularisation; if surgery is undertaken CA should be repeated a year after surgery.

To access full recommendations, see Chapter 6.1.3 here.

Acute medical interventions for AIS (Chapter 6.2.1)

Use of thrombolysis or anti-thrombotic therapy

- Prescribe and deliver 5mg/kg of aspirin up to a maximum of 300mg within 24 hours of diagnosis of AIS in the absence of contraindications (e.g. parenchymal haemorrhage). After 14 days reduce dose of aspirin to 1mg/kg to a max of 75mg.
- The off label use of tissue plasminogen activator (tPA) could be considered in children presenting with AIS who are more than eight years of age and may be considered for children aged between two and eight years of age on a case by case basis when the criteria detailed in 6.2.1 have been met.

Acute AIS treatment in children/young people with sickle cell disease (SCD)

- Treat children/young people with sickle cell disease (SCD) and acute neurological signs or symptoms urgently with a blood transfusion, to reduce the sickle haemoglobin (HbS) to less than 30%, and increase the haemoglobin concentration to more than 100–110g/l. This will usually require exchange transfusion.
- Provide a small top up transfusion to bring Hb to 100g/l to improve cerebral oxygenation if the start of the exchange is likely to be delayed by more than six hours.

To access full recommendations, see Chapter 6.2.1 here.
Interventions to prevent recurrence of AIS (Chapter 6.2.2)

Medical interventions to prevent recurrence of AIS

- Continue antithrombotic treatment initiated acutely in children and young people with AIS. Reduce dose of aspirin from 5mg/kg to 1mg/kg after 14 days.
- Treat all children and young people with AIS with aspirin, unless they have SCD or are receiving anticoagulation e.g. for a cardiac source of embolism.
- Maintain adequate levels of hydration in patients with occlusive arteriopathies including moyamoya, especially when fasting or during intercurrent illness.

AIS recurrence prevention in SCD

- Start regular blood transfusions as secondary stroke prevention in children and young people with SCD, aiming to keep the pre-transfusion HbS less than 30% and keeping the pre-transfusion haemoglobin above 90g/l. This can be done with either exchange or simple top-up blood transfusion.
- Monitor children with regular neurocognitive testing, MRI and transcranial doppler ultrasonography (TCD); frequency should be determined on a case-by-case basis.
- Hydroxycarbamide should be considered as part of a secondary stroke prevention programme when suitable blood (e.g. multiple alloantibodies or hyperhaemolysis) is not available, or when continued transfusions pose unacceptable risks (uncontrolled iron accumulation).

SCI progression prevention in SCD

- Discuss the possible benefits of transfusion with children/young people and families if silent cerebral infarctions (SCI) are identified on MRI. See 6.2.2 for factors favouring the implementation of a treatment program involving regular blood transfusions.

To access full recommendations, see Chapter 6.2.2 here.

Surgical and endovascular interventions for AIS (Chapter 6.2.3)

Indications for referral to neurosurgery in children and young people with AIS

- Discuss any impairment of conscious level or decline in Pediatric National Institutes of Health Stroke Scale (PedNIHSS) in a child with AIS with a neurosurgical team.
- Consider decompressive hemicraniectomy in children/young people with middle cerebral artery (MCA) infarction under the circumstances listed in 6.2.3.

Indications for referral to interventional neuroradiology

- Patients with acute AIS causing a disabling neurological deficit (NIHSS score of 6 or more) may be considered for intra-arterial clot extraction with prior intravenous thrombolysis, unless contraindicated, beyond an onset-to-arterial puncture time of five hours if a) PedNIHSS score is more than six, b) a favourable profile on
salvageable brain tissue imaging has been proven, in which case treatment up to 12 hours after onset may be appropriate.

To access full recommendations, see Chapter 6.2.3 here.

Haemorrhagic Stroke (Chapter 7)
Risk factors for HS and recurrent HS (Chapter 7.1.1)
Risk factors for first HS

- Be aware that certain factors/conditions are associated with an increased risk of HS in children/young people, as listed in the full recommendations.

Risk factors for recurrent HS

- Be aware of increased risk of recurrence in children/young people with HS and the following risk factors: arteriovenous malformation (AVM), cerebral arterial aneurysms, cavernous malformations, moyamoya, SCD, all severe bleeding disorders, ongoing anticoagulation, illicit drug use.
- Be aware that in arteriovenous malformations, which have already bled, the greatest risk of a rebleed is from the part of the malformation which was responsible for the initial haemorrhage. Intratidal or perinidal aneurysms and venous varicosities/stenoses are sinister features.

To access full recommendations, see Chapter 7.1.1 here.

Investigations to identify underlying risk factors in HS (Chapter 7.1.2)

- If the child is known to have SCD, additional tests should include TCD and an extended blood group phenotype (e.g. ABO, Rh C, D and E, and Kell).

To access full recommendations, see Chapter 7.1.2 here.

Follow-up imaging in HS (Chapter 7.1.3)

- Discuss the modality and timing of imaging in children and young people with HS within a MDT; this will be influenced by factors relating to the individual patient and the lesion.
- Offer all children and young people with a previously treated brain AVM and angiographic confirmation of obliteration a final catheter angiogram at 16 to 18 years of age, prior to transition to adult services, to exclude AVM recurrence or a de novo lesion.

To access full recommendations, see Chapter 7.1.3 here.
Acute medical interventions for AIS (Chapter 7.2.1)

- Take blood for the measurement of routine coagulation parameters (prothrombin time (PT), partial thromboplastin time (PTT), Clauss fibrinogen) and full blood count (FBC) in all children and young people presenting with HS. Abnormal results should be discussed with a paediatric haematologist in order that appropriate investigations can be carried out urgently to ascertain whether a coagulation abnormality is primary or secondary.
- Treatment should be focussed on maintaining normal levels of the appropriate coagulation factor for a period of intense treatment and then prophylactic treatment to prevent recurrence.

To access full recommendations, see Chapter 7.2.1 here.

Interventions to prevent recurrence of HS (Chapter 7.2.2)

Medical interventions to prevent recurrence of HS

- Refer all children and young people with inherited bleeding disorders to a children’s comprehensive care centre (CCC) as the management of all inherited bleeding disorders is highly specialised. They will be registered on the United Kingdom Haemophilia Centre Doctors’ Organisation’s (UKHCDO) National Bleeding Disorders database.

HS recurrence prevention in SCD

- Provide anti-sickling treatment to children and young people with SCD and HS, and either a regular blood transfusion or a haematopoietic cell transplantation from a human leukocyte antigen (HLA)-matched sibling (or alternative donors in rare circumstances).
- Provide regular blood transfusions if there is clear evidence of arteriopathy (e.g. occlusive lesions or aneurysms) to keep HbS less than 30%.
- Follow up children and young people with HS in SCD, long-term with repeat neurocognitive testing, MRI and TCD to assess evidence of progressive cerebrovascular disease.

To access full recommendations, see Chapter 7.2.2 here.

Surgical and endovascular interventions for HS (Chapter 7.2.3)

Neurosurgical management of HS

- Children and young people with HS should always be cared for in conjunction with a neurosurgical team.
• Do not routinely evacuate intracerebral haemorrhage (ICH) in children and young people, except in cases where there is a rapidly deteriorating age-appropriate GCS score.

**Interventional neuroradiology**

• Discuss patient’s cases with acute HS and vascular lesions in a neurovascular MDT including an interventional neuroradiologist.

**Stereotactic radiosurgery**

• Stereotactic radiosurgery (SRS) may be considered as a treatment option for vascular lesions and should be included in the discussion of the case in the MDT.

**The safety and efficacy of surgical, radiosurgical and endovascular interventions in the treatment of ruptured in comparison to unruptured vascular lesions**

• Consider active management more readily in children and young people with diagnosed unruptured AVM than in adults due to the higher cumulative risk of rupture attributable to the projected longer life span.
• Consider treatment options such as no treatment, surgical resection or stereotactic radiosurgery in the discussion of the case within the MDT.
• Consider micro-surgical resection or stereotactic radiosurgery for unruptured lesions that are enlarging on serial imaging.

*To access full recommendations, see Chapter 7.2.3 here.*

**Discharge from hospital (Chapter 8)**

• Plan discharge with input from the child or young person and their family and the MDT (medical, nursing and allied health professionals including education staff, occupational therapists, physiotherapists, orthoptists, psychologists, speech and language therapists) prior to discharge from hospital. If the child has been admitted for an extended period, this may involve more than one meeting and should occur in a time-frame that allows all necessary support to be in place on discharge.
• Provide a named key worker or a core group model (such as Team Around the Child/Family (TAC/F)). This can be effective in ensuring that the family has easy, personalised access to appropriate services as required, and is made aware of anticipated timelines and who is accountable for certain actions.

*To access full recommendations, see Chapter 8 here.*
Rehabilitation

Framework for assessing rehabilitation needs (Chapter 9.1)

- Provide a comprehensive multidisciplinary assessment of needs, taking into account all domains of the ICF, using appropriate measures considering the child or young person and family priorities/preferences as well as the age and developmental stage of the child or young person.
- Consider using quality of life measures to support evaluation of rehabilitation outcomes, and note that tools such as the Canadian Occupational Performance Measure (COPM) or Goal Attainment Scaling (GAS) may assist with identifying individual targets for intervention and evaluating outcome.
- The MDT should work in active partnership with the child/young person and family in a) formulation and agreement of individualised goals across health domains to develop a unified and coordinated approach across disciplines; b) goal setting and decision making around intervention plans; c) identification of priorities when considering rehabilitation options.
- Identify a named key worker or key point of contact for families, who will remain a key point of contact through transfer from hospital to community or specialist rehabilitation services, and including starting/re-entering school. This named key worker/contact may vary as appropriate as the child progresses through different life stages.

To access full recommendations, see Chapter 9.1 here.

Rehabilitative interventions (Chapter 9.3)

Motor function and mobility

- Provide rehabilitation that fits within a neurological and developmental framework; individual therapies should complement each other to maximise functional skills.
- Deliver rehabilitation intervention focussed on what the child or young person and family need to, want to, or are expected to do. Motor interventions should be focussed on functional goals and undertaken with consideration of the whole child and their needs and abilities across all domains of health.
- Time since stroke should not be a barrier for the consideration of intensive training.
- Offer motor skills rehabilitation interventions based on the principles of motor learning with sufficient intensity, repetition and functional relevance to support lasting change.

Sensory functions

- Assess vision and hearing as part of the multidisciplinary assessment.
- Treat all pain actively, using appropriate measures including positioning, handling and medication.
Dysphagia

- Refer for speech and language therapy (SLT) assessment and advice if parents/carers have concerns about coughing or choking on eating and drinking, frequent chest infections, or failure to move through the typical stages of eating and drinking development.

Communication, speech and language functions

- Offer neuropsychological assessment (by educational, clinical or neuropsychologist) for children and young people when starting or returning to school/not meeting their attainment targets. Refer for more detailed SLT assessment, including the use of formal testing, where there are specific concerns about speech, language or communication limitations.
- Offer referral to SLT when there are parental or professional concerns about communication skills, language understanding, expressive language or poor intelligibility due to persisting motor speech disorders (dysarthria and dyspraxia), dysfluency or voice disorders.

Cognition

- Provide neuropsychological assessment and advice to schools and affected families throughout formal education.
- Train and involve parents/carers of children who have suffered stroke in delivery of interventions to support cognitive functioning in their child's daily life activities.

Mental health

- Refer children, young people and their families to local children and young people's mental health services or paediatric psychology services within hospitals for psychotherapeutic interventions.
- Develop acquired brain injury specific adaptations to support local children and young people's mental health services to provide appropriate input.

Interpersonal relationships and interactions/psychosocial

- Refer children, young people and families to psychology services when there are concerns about social relationships.
- Include parent/carer, child/young person, and teacher reports using standardised questionnaires in assessment and monitoring of family and peer relationships.
Learning and applying knowledge

- Teach factual knowledge (e.g. word reading, maths facts) through Precision Teaching with Direct Instruction. Direct Instruction refers to systematic scripted lesson plans. Use the principles of Precision Teaching which is a well-established method of teaching involving high levels of repetition of specific material e.g. high frequency words, typically involving daily assessment of progress.

- Provide a Special Educational Needs and Disabilities Co-ordinator (SENCo) or equivalent to act as a keyworker/named coordinator once the child is attending school. This individual should liaise with parents/carers and professionals as per the Special Educational Needs and Disability (SEND) code of practice: 0 to 25 years.

- Health professionals should provide regular consultation to educators, including both advice and brain injury training. This should be with a professional with experience of both education and acquired brain injury.

Self-care/independence

- Assess the child's ability to perform self-care tasks, household tasks, tasks in major life areas such as school, play, and community life.

- Involve an occupational therapist in provision of intervention in this area if difficulties are identified.

- Consider goal directed, functional training with home programmes where appropriate.

Goal setting

- Discuss areas of functional difficulty and intervention priorities with children, young people and families.

- Create goals/principles which follow the general principles of being SMART (Specific, Measurable, Agreed, Realistic and Time-bound).

- Review goals and priorities at least annually. This should be done with the child/young person and their family and health and education professionals.

To access full recommendations, see Chapter 9.3 here.

The needs of the family during the planning of care/rehabilitation (Chapter 9.4)

- Inform, as relevant for the individual child or young person and family, the potential or actual role of health, education and social care systems in providing support and care. Include information and education about assessment processes.

- Consider the impact of stroke on the health, social and economic wellbeing of family members and make onward referrals as necessary to support the broader family.
• Provide regular opportunities for the child or young person and family to access support from professionals from health, education and social care as needed; this should include (with parent/child or young person consent) communication between care agencies including the family and child or young person and documented integrated planning.

To access full recommendations, see Chapter 9.4 here.

Long-term care: transfer and transition (Chapter 10)
Managing educational and social-care transition (Chapter 10.1)

• Ensure regular, effective collaboration and communication between the child, young person and family and health, education, and social care professionals throughout the child’s schooling to identify and respond to their specific needs and disabilities. This can include meetings, joint assessments and sharing of relevant knowledge and skills to optimise and personalise the provision of learning support.

• Be aware that children and young people with stroke may require a flexible, holistic, integrated approach in supporting them, ranging from targeted therapy or educational interventions for particular difficulties, to a comprehensive Education, Health and Care Plan (EHCP).

To access full recommendations, see Chapter 10.1 here.

The transition of a young person into adult health care (Chapter 10.2)

• Consult the National Institute for Health and Care Excellence (NICE) guideline on ‘Transition from children’s to adults’ services for young people using health or social care services’ (NG43).

• Inform young people and their parents/carers about the professionals involved in future management and how to gain access to them.

To access full recommendations, see Chapter 10.2 here.
1. **Introduction**

1.1. **Overview**

Stroke is an important childhood disorder and is at least as common as brain tumours\(^2\), affecting several hundred children and young people in the UK each year. Stroke was found to be in the top 10 causes of death in children and young people in America\(^3\); however, more recent studies have reported relatively lower mortality rates\(^2\), perhaps related to improvements in critical care support for these acutely sick children\(^4\). At least half of survivors have some long-term impairment\(^5\); of note, the full impact of stroke on the developing brain may only emerge over time, with increasing demands on neurocognitive functions, and on educational and social roles, resulting in widespread and long-lasting personal, family and societal consequences.

Over the last 15 years there has been a dramatic increase in the recognition of both the occurrence and consequences of childhood stroke, mirroring the transformation of medical and societal attitudes and treatment pathways in adults. However, in comparison with stroke in adults, that is now considered a medical emergency, with the attendant development of hyperacute recognition, investigation and management pathways, childhood stroke management lags behind. Challenges in treating children and young people with stroke include delays in recognition, the logistic challenges of timely investigation and imaging, a wide differential diagnosis, diverse stroke aetiologies and the lack of trial-based evidence for hyperacute treatments. Although there have recently been substantial developments in the evidence base for paediatric rehabilitation interventions in other populations of children\(^6\), those affected by stroke still face barriers accessing timely best-evidenced rehabilitation to enhance recovery and long-term development of independence skills.

Perhaps the most challenging recommendations for the acute healthcare sector in this guideline relate to hyperacute interventions for childhood arterial ischaemic stroke (AIS). These will be contentious as they are not evidence-based; however, with the collapse of the multi-centre Thrombolysis in Paediatric Stroke study (TIPS)\(^7\), it seems unlikely that trial-based evidence for hyperacute paediatric AIS therapy will be forthcoming anytime soon. Given that hyperacute thrombolysis and now thrombectomy are the accepted standards of care in adults, the issue of how to manage children and young people with acute AIS is a challenge for paediatricians in day-to-day clinical practice. While accepting the lack of evidence, the guideline development group (GDG) followed a formal Delphi consensus method (referred to as [Delphi] within the recommendations) to suggest what might be considered a reasonable approach recommended by experts; this will, however,
be open to challenge and it should be recognised that these are guidelines and not protocols.

These guidelines will challenge both commissioners’ and clinicians’ established attitudes and pathways relating to the care of childhood stroke in the UK. The timely, aggressive treatment of the small number of children and young people with stroke due to AIS who might be candidates for hyperacute therapy will have knock-on effects on the pathway for all children and young people with ‘brain attack’ (an acute clinical deficit that may or may not represent an AIS or haemorrhagic stroke (HS)). It is important to emphasise that the GDG has included representation extending from the pre-hospital sector to tertiary centres and, crucially, has engaged with parent input throughout. It is suggested that these guidelines lead to the development of a funded national registry which will enhance both data capture and critical analysis of cases considered for hyperacute treatments, and inform need for and access to rehabilitation services.

The International Classification of Functioning, Disability and Health (ICF)\(^8\) has been used as the framework within this guideline to define and describe functioning and disability and support identification of targets for intervention. This model describes functioning in terms of a dynamic interaction between an individual’s health condition and environmental and personal factors. For ease of reading, in some cases ICF terms have been replaced with terms more commonly used in UK clinical practice. Where these first appear, both the ICF term and the replacement term are noted. In using the term ‘rehabilitation’ in this guideline, the concepts of habilitation (support for ongoing acquisition of developmental and functional skills) and rehabilitation (recovery of skills) are referred to as both or either of these approaches may be relevant when working with a developing child or young person.

1.2. Current practice in the UK

Acute stroke

The structure of this guideline describes the clinical approach to a child presenting with an acute clinical deficit that may or may not represent an AIS or HS. It is recognised that 30 to 50% of children and young people with such presentations will have a non-vascular diagnosis, but in order to detect vascular stroke acutely, the clinical approach needs to be over-inclusive as well as discriminatory. In constructing the recommendations, the acute recognition and diagnostic investigations of AIS and HS (as they would be indistinguishable prior to diagnostic imaging) have been combined. Risk factors and management have been considered separately, while generic recommendations have been made regarding rehabilitation. The term HS is used here to refer to all non-traumatic intracranial haemorrhage in children and young people, with the exception of neonatal intraventricular haemorrhage.
This guideline provides examples of how referral and management pathways might be improved going forward. These begin from the point of referral or the activation and response of emergency medical services and go through to long-term community care, covering the management pathway from the acute setting to the more elective management of neurovascular disorders in children and young people. The guideline also discusses the activation of regional retrieval services to expedite time-critical imaging and transfer to a facility that will hasten the hyperacute management of stroke.

The use of the FAST tool⁹ (Face Arms Speech Time) is now widespread in the adult population and is an acronym comprising common signs accompanying stroke:

- **Face**: the face, mouth or eye may have drooped on one side of the face and the person may not be able to smile.
- **Arms**: the person with suspected stroke may not be able to lift their arms due to arm weakness or numbness.
- **Speech**: speech may be slurred, or the person may not be able to talk at all despite appearing to be awake.
- **Time**: it is time to dial 999 immediately if any of these signs/symptoms are present.

It is hoped that with increased awareness, pre-hospital and emergency room professionals will recognise that most children and young people with stroke can also be identified using the FAST tool and that there is an opportunity to extend awareness of the significance of FAST symptoms into the paediatric population.

The management of stroke in adults is well commissioned and has led to pathways that allow early recognition and rapid transfer directly to stroke centres. In light of the current organisation of paediatric services within the UK, sick children and young people will be taken to the nearest acute paediatric unit. This is appropriate to ensure early triage and that the child is in a place of safety, but must be combined with a general raising of the level of awareness in the community and amongst healthcare professionals to facilitate early recognition and appropriate onward care of children and young people with suspected stroke. Whilst care in specialist stroke units has been shown to improve outcomes in adult patients, it is unrealistic to deliver care for children in a specialist stroke unit. The needs of these children are best managed within secondary and tertiary services that are used to dealing with neurologically sick children; this guideline aims to supplement this more generic care by providing guidance on disease-specific aspects of investigation and management.
In summary, crucial areas in the childhood care pathway are:

- Rapid access to diagnostic imaging from emergency departments (ED), specifically computed tomography (CT) followed by CT angiography (CTA) if AIS is proven within one hour of presentation to secondary care (ED or the paediatric ward).
- Familiarity with standardised stroke severity assessments such as the paediatric version of the National Institutes of Health (NIHSS) Stroke Scale\textsuperscript{10} in secondary and tertiary centres.
- Involvement of regional paediatric acute transport teams in coordinating case discussion between the relevant secondary and tertiary teams regarding the diagnosis, management, and onward transport modality/destination.
- Recognition that acute cases of childhood stroke need rapid transfer to regional neuroscience centres, but that this should not delay starting hyperacute treatment.
- Ability to deliver hyperacute therapies for AIS in a timely manner in the secondary centre; this will necessitate local protocols for dosing and monitoring and real-time conferencing between different professionals in different centres.
- Developing an interface with hyperacute adult stroke teams in district general hospitals (DGH), to support the delivery of hyperacute therapy to eligible children and young people.

Clinical experience and audit suggests that each UK regional paediatric neuroscience centre would encounter fewer than five children annually who would meet the criteria stipulated in this guideline for hyperacute intravenous thrombolysis for AIS. Each DGH is therefore likely to see less than one eligible child per year, and management of these cases will be highly challenging, both to the generalist and to the neurologist advising from a distance. Even more challenging will be the identification and transport of children and young people who might benefit from thrombectomy.

**Management of neurovascular disease in regional centres**

This section relates to the longer-term, less acute planning of management of neurovascular diseases that may be associated with AIS (e.g. moyamoya) or HS (e.g. arteriovenous malformations (AVM) or aneurysms) in children and young people. In general, these issues will be dealt with in regional neuroscience units, rather than secondary level centres.

An increasingly recognised principle of good clinical practice is that these cases should be discussed and managed in multidisciplinary teams (MDT), with input from neurologists, neurosurgeons (including those familiar with stereotactic radiosurgery (SRS)), and diagnostic and interventional neuroradiologists; in subsequent sections this group is
referred to as a multidisciplinary neurovascular team. In reality, it is unlikely that such a team will be totally quorate in each regional centre. This guideline strongly advocates that local MDTs develop networking arrangements where a specific area of expertise is lacking, and develop locally agreed practices for management of specific pathologies. Children, young people and families should have access to all potential therapeutic modalities for vascular malformations (surgical excision, endovascular treatment, SRS or conservative management) in order to make an informed choice. Discussion with the local adult neurovascular team could be helpful, especially as some pathologies (e.g. aneurysms) are much more commonly encountered in adult practice. Counselling about the risks and benefits of treatment are complex, particularly in the instance of unruptured AVM, and again the recommendations here reflect consensus opinion.

These guidelines provide an opportunity to use the recommendations as a skeleton to develop national multidisciplinary networks to standardise childhood stroke care and to audit outcomes. The National Health Service (NHS) provides a unique opportunity for such an approach and there is real potential to systematically synthesise data to improve care.

It is important to recognise the biological differences between children and adults when considering the aetiology and management of stroke. The paucity of high quality research evidence relating to childhood stroke will be apparent throughout this document and therefore in many cases it has been necessary to exercise clinical judgement in interpreting research evidence in adult populations for use in this guideline. There are also major differences in the structure of clinical care for adults and children. The efficacy of stroke units in improving outcomes in adult stroke is clearly established; however, given the low frequency of childhood stroke it seems reasonable to tap into existing networks of hospital and community care for children with acquired neurological disorders without losing sight of issues specific to stroke.

**Longer term care and rehabilitation**

This section of the guideline relates to care from the time that a child is medically stable, usually described as the sub-acute phase of recovery, through to long-term support and care in the years following diagnosis. This phase of care often involves the broader multidisciplinary team working closely with the child/young person and family, and may extend to the involvement of education and social care services as well as healthcare. The scope includes all domains of the ICF (body structures and functions, activity and participation). Environmental factors, in particular the impact on families and consideration of their needs, are included but environmental interventions are not reviewed.

The wide variation in duration of acute hospital admission of children and young people with stroke means that there are significant variations in the location and coordination of
care. There is a need for rehabilitation (including habilitation) to commence during hospital admission, and for a small proportion of children and young people to receive intensive input in an inpatient or residential specialist rehabilitation facility. In the majority of cases, however, rehabilitation is provided by general community teams, through community child health centres, and care is usually delivered at home or in educational settings.

In order to receive the best available intervention, there is a need for coordination and planning to start from the point of stroke diagnosis, and for the broader multidisciplinary team to be involved to plan and prepare for discharge and educational reintegration.

The research evidence on the emergence of the functional consequences of stroke over time is now clear. However, surveillance and the opportunity for routine review of functional daily life abilities is not routinely available for all children and young people. Young people may present in the months and years after a stroke with new or changed needs for rehabilitation to support independence skills.

Communication, coordination and planning across healthcare providers and with educational and social care professionals are key to effectively supporting children, young people and families. The need to identify a named key worker/key contact has been strongly voiced by the parents/carers, family and young people involved in the development of this guideline, to help families remain informed of services, and to help them navigate the health and care systems.

In addition to transfer from an acute setting to local hospital care, rehabilitation unit or home, the key transitions considered in the guideline are entering school, moving through school and on to work or tertiary education, and the transition from child to adult healthcare services. Children and young people who have a stroke will grow into adults living with the consequences of an acquired brain injury, which may evolve and change over time as the demands of life change. Access to review by the MDT in the long-term may be necessary to meet and support children and young people as their needs evolve and change in daily life over time.

This guideline aims to support these individuals and their families in developing the skills to live as full and independent lives as possible. Development of skills in self-management and self-advocacy are important considerations for all individuals, regardless of their circumstances.

The active engagement of parents and children and young people in two-way communication is a key element to all phases of the care pathway in informing priorities.
for targeting intervention. The parent and young person workshops held to inform these guidelines identified six areas of priority to be considered in care delivery:

- timely, individualised information including the opportunity for the child and young person to raise questions and meet with professionals separately to parents (if preferred)
- support that is consistent
- regular personalised review
- transfer and transition planning, which is agreed in advance and actively involves the child, young person and family
- coordinated care, ideally with a named key contact to provide consistent support and who is knowledgeable about brain injury, and
- access to health professionals with knowledge and experience of acquired brain injury and stroke; and child and family-centred communication that is proactive, sensitive to needs, and allows time for asking questions.

1.3. Clinical need for the guideline

Current guidelines on the diagnosis and management of childhood stroke are based on recommendations published in 2004 by the Royal College of Physicians (RCP), which come from the clinical guideline, *Stroke in Childhood: Clinical Guidelines for Diagnosis, Management and Rehabilitation*. These guidelines may no longer reflect best and most up to date clinical practice, and as such the guideline required urgent updating to ensure it utilises the most up to date evidence.

1.4. Aims and objectives

This guideline is an update and expansion of the 2004 RCP Stroke in Childhood guideline and provides guidance on the identification, diagnosis and management of children and young people (aged 29 days to 18 years at time of presentation) with AIS (arterial ischaemic stroke - an acute focal neurological disorder with imaging evidence of cerebral infarction in a corresponding arterial distribution) and HS until their transition to adult care.

Consideration has also been given to the management of unruptured at risk vascular malformations (arteriovenous malformations, cavernous malformations, cerebral aneurysms and arteriovenous fistulae).

The guideline addresses the entire patient pathway, from presentation, to acute care and longer-term management of medical issues and rehabilitation.
An upper age limit of 18 years has been set, although it is recognised that adult pathways might be more applicable to older teenagers and that some clinical guidance for young people extends to 25 years of age. Of note, in this iteration the scope of the guideline has been extended to include HS, defined as non-traumatic intracranial haemorrhage, but not cerebral venous thrombosis, nor stroke in newborns.

See Appendix 1 for further details on the scope, including more information on what has been and not been covered in terms of population.

1.5. **Using the guideline**

This guideline is aimed at professionals working in primary care, secondary level acute paediatrics and tertiary level paediatric neurosciences, as well as those within the ambulance sector, paediatric intensive care unit (PICU), community paediatrics, neurodisability, education, and social services. It may also be of use and interest to professionals working with young people transitioning into adult care.

It is intended for use by all UK paediatricians and other healthcare professionals involved in the regulation or practice of the care of children and young people who have had or are suspected of having a stroke, as well as non-healthcare professionals involved with educational/social services. While sections may also be relevant to education and social care professionals, it is intended to inform clinical decision making.

This guideline is set in the context of the current legal framework in the UK governing the provision of services, and is not intended to overrule such regulations; it should be considered in conjunction with such regulations, such as the Care Act (2014) in England. Within this framework, the intention for the guideline is to facilitate practice not only in health services but also in social services and other organisations. It is assumed that clinicians will be operating within the recognised standards of practice laid out by their professional and regulatory bodies.

In order to make the structure of the guideline relevant to the patient journey, the initial sections on recognition and diagnosis consider AIS and HS together; specifics of targeted investigations, management and prevention of recurrence consider AIS and HS separately; and rehabilitation is considered without distinction (see Diagram 1.1). Where relevant, other published guidance has been cross-referenced, such as adult stroke or sickle cell disease.
Diagram 1.1. Structure of guideline

*Please note that while Diagram 1.1 follows the structure of the guideline, it is important to highlight that rehabilitation starts within the acute setting through to community care.
2. **Methodology**

2.1. **Introduction**

The guideline aims to produce evidence-based guidance on the diagnosis, management and rehabilitation of stroke in children and young people aged 29 days to 18 years. It is based partly on the 2004 Royal College of Physicians (RCP) guideline, *Stroke in Childhood: Clinical Guidelines for Diagnosis, Management and Rehabilitation*.

The guideline update has been funded by the Stroke Association, which is registered as a charity in England and Wales (211015) and in Scotland (SC037789); also registered in Northern Ireland (XT33805), Isle of Man (945) and Jersey (NPO 369). With many thanks to the Stroke Association's supporters, in particular The Thompson Family Charitable Trust, whose generous donation enabled the Stroke Association to fund these guidelines.

The Royal College of Paediatrics and Child Health (RCPCH), which is registered as a charity in England and Wales (1057744) and in Scotland (SCO38299), in collaboration with the Stroke Association and a guideline development group (GDG) comprised of clinicians from a range of relevant specialities and lay members, have carried out this update in accordance with the RCPCH standards for the development of clinical guidelines in paediatrics and child health. Further details on the guideline scope can be found in Appendix 1.

2.2. **Developers and conflicts of interest**

A GDG was convened to oversee the development of the guideline, and included representatives nominated by their stakeholder organisations.

The RCPCH Clinical Standards Team led on the development of the guideline, funded by the Stroke Association, carried out the systematic searches, critical appraisal and data extraction of publications, and oversaw the Delphi consensus method with input from the GDG.

The guideline was drafted in consultation with the GDG, who met every two to three months during the development of the guideline. All conflicts of interest were declared by the GDG at different stages throughout guideline development and are recorded in Appendix 2a. Types of interest to declare include personal pecuniary interest, non-personal pecuniary interest, personal non-pecuniary interest and/or personal family interest; the Chair of the GDG declared no pecuniary competing interests. A copy of the conflicts of interest form can be found in Appendix 2b.
2.3. **Editorial independence**

All GDG members declared any conflicts of interests prior to the guideline development starting, and periodically throughout the development of the guideline (see Appendix 2a).

The Stroke Association did not influence the GDGs decisions or the guideline recommendations other than through its role as a stakeholder.

2.4. **Developing the clinical questions**

The GDG identified the clinical areas to be covered by the guideline remit, and the RCPCH Clinical Standards Team formulated these into structured questions.

The review questions were developed based on a framework of Population, Intervention, Comparison and Outcome (PICO). A protocol was prepared which guided the literature search, critical appraisal and synthesis of evidence, and facilitated the development of recommendations by the GDG.

2.5. **List of review questions**

**Arterial ischaemic stroke (AIS) and haemorrhagic stroke (HS)**

- In childhood, which conditions/factors are associated with higher risk of development of AIS/HS?
- In childhood, which conditions/factors are associated with higher risk of recurrence of AIS/HS?
- What are the presenting clinical signs and symptoms for suspecting AIS/HS diagnosis in children and young people?
- In children and young people, what is the role, modality and timing of brain imaging in diagnosis, of AIS/HS?
- In children and young people, what is the role, modality and timing of imaging in:
  - assessment?
  - monitoring of AIS/HS?
- What is most appropriate referral pathway/course of action for children and young people:
  - diagnosed with acute AIS/HS?
  - suspected clinically with acute AIS/HS?
  - incidentally diagnosed with silent AIS/HS?
  - diagnosed with unruptured, at risk vascular lesion?
- In children and young people with AIS/HS, what are the most appropriate investigations to identify underlying risk factors?
• In childhood, what is the risk of future haemorrhage from a diagnosed vascular lesion:
  - previously haemorrhaged?
  - previously not haemorrhaged?
• In children and young people with acute/recurrent HS, what are the most appropriate investigations to identify underlying risk factors?
• In children and young people with AIS/HS, what is the optimal way to assess:
  - neurological status?
  - stroke severity including key elements to be assessed?
• Does the setting where children and young people with AIS/HS receive acute care affect mortality, morbidity and complications?
• What are the key elements of acute medical care management of children and young people with AIS/HS?
• Who are the key individuals to be involved in the acute management of children and young people with AIS/HS?
• In children and young people with acute AIS/HS, what is the framework for early functional assessment including: nutrition and hydration, communication difficulties, swallowing difficulties, cognition, and mobility?
• What are the likely complications during the acute and sub-acute phase of recovery from AIS/HS?
• What are most effective ways to detect, prevent and minimise AIS/HS complications in children and young people?
• Which elements of the information and care provided by key health professionals and other sectors are most important for AIS patients and their families at the acute stage? (including environmental factors)
• What is the safety and efficacy of thrombolytic agents/anticoagulants/antiplatelet agents for the acute treatment of children and young people with AIS?
• What is the safety and efficacy of coagulation factor replacement for the acute treatment of children and young people with HS?
• What is the safety and efficacy of blood transfusion in the treatment of children and young people with HS?
• What is the safety and efficacy of blood transfusion in the treatment of sickle cell disease (SCD) in children and young people with AIS?
• What is the safety and efficacy of medical interventions to prevent recurrence of AIS/HS? (AIS subgroups of importance: SCD and moyamoya; HS subgroups of importance: SCD and moyamoya. Congenital/acquired vascular anomalies: arteriovenous malformations, cavernous malformations, cerebral aneurysms and arteriovenous fistulae, inherited platelet and coagulation disorders)
• In children and young people with acute/chronic AIS/HS, what are the indications for referral to neurosurgery?
• What is the safety and efficacy of surgical interventions in the treatment of acute AIS/HS in children and young people?
• What is the effectiveness of surgical interventions in the prevention of recurrence of AIS/HS in children and young people?
• In children and young people with acute/chronic AIS/HS, what are the indications for referral to interventional neuroradiology?
• What is the safety and efficacy of gamma knife intervention in the treatment of acute HS in children and young people?
• Is there a difference in the safety and efficacy of surgical, radiosurgical and endovascular interventions between treating ruptured and unruptured at-risk vascular lesions?

Rehabilitation and long-term care

• What is the most appropriate framework for the evaluation of rehabilitation needs in children and young people with stroke?
• What are the components and effectiveness of interventions for:
  - motor functions/mobility?
  - sensory functions including pain?
  - communication and speech and language functions?
  - dysphagia?
  - mental functions/education/cognition/executive function?
  - interpersonal relationships and interactions/psychosocial?
  - learning and applying knowledge?
  - self-care/independence?
  - goal setting?
  - mental health?
• What are the needs of families and the role of voluntary sector during the planning of care and rehabilitation for children and young people with stroke?
• What environmental factors (equipment, adaptations, educational support, access to community and social life, support and relationships) are the most important for stroke patients in rehabilitation and in the long-term?

Discharge and transition

• What are the elements to consider when planning the discharge of children and young people with stroke from acute hospital care to rehabilitation care and to long-term community care?
• What is the most effective way of managing educational and social care transfer through various educational stages (nursery, primary and secondary school, college/work) for children and young people after stroke?
• For young people who have had a stroke, how should the transfer to adult healthcare be managed?

2.6. Identifying the evidence

Review questions were grouped to develop systematic review protocols. These protocols formed the starting point for the systematic reviews of the relevant evidence, defining the inclusion and exclusion criteria as well as subgroups of importance and other considerations (see Appendix 3).

All literature searches were conducted on core databases, including MEDLINE, Embase, Cochrane Library, Cumulative Index of Nursing and Allied Health Literature (CINAHL) and PsycInfo. Searches were limited to the English language. There was no searching of grey literature, nor was hand searching of journals undertaken.

AIS searches were carried out on literature published from January 1995 to December 2015 and HS searches were carried out on literature published from January 1995 to February 2016. Full inclusion/exclusion criteria, search terms and search strategies can be found in Appendix 4a.

2.7. Reviewing and synthesising the evidence

The initial title screening of the electronic search results were initially done by the RCPCH Clinical Standards Team. Relevant abstracts were then screened further and assessed by two members of the GDG, against predefined data extraction forms (see Appendix 4b); disagreements were resolved by a third person. If the reviewer was uncertain about the appropriateness of rejecting the article, the full text article was retrieved.

Full text studies were assessed for inclusion against predefined inclusion and exclusion criteria to identify studies that addressed the review questions in the appropriate population and reported outcomes of interest. Data related to each included study’s population, methods and results were extracted using a proforma and confirmed by the RCPCH Clinical Standards Team.

The quality of individual studies was assessed using validated critical appraisal checklists developed by the Scottish Intercollegiate Guidelines Network (SIGN) (i.e. Randomised Controlled Trials (RCT), case-control and cohort studies) and Critical Appraisal Skills Programme (CASP) (i.e. qualitative studies). Studies were categorised as being of low,
moderate or high quality depending on the number of the checklist items they met and their risk of bias. Studies meeting all of the predefined inclusion criteria and marked with a low risk of bias were categorised as high quality. Those meeting at least 80% of checklist items and with a moderate or low risk of bias were categorised as moderate quality, and those meeting less than 80% of checklist items with low, moderate or high risk of bias were categorised as low quality. If a study met the required number of checklist items but had a higher risk of bias than allowed by a quality category then it was downgraded. Studies were, however, not downgraded based on their design. This was because the vast majority of included studies were case series or case-control design, and so downgrading based on study design would have created a floor effect where studies that were otherwise well conducted could not achieve a high quality rating because of their design.

The quality of the body of evidence included for each clinical question was then assessed and rated following the principles of the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach\textsuperscript{15}, namely risk of bias, heterogeneity, indirectness, imprecision and publication bias.

Data were tabulated in evidence tables (see Appendix 4c) and used by the GDG members to develop recommendations, which were then reviewed and agreed by the GDG as a whole. A list of all included studies can be found in Appendix 4d.

2.8. Developing recommendations

The GDG was split into two groups (acute care, and longer-term care and rehabilitation), according to the expertise of the members, and meetings were held to discuss the evidence and formulate recommendations around these elements of the care pathway.

Recommendations were derived and explicitly linked to the evidence that supported them. In the first instance, individual members or pairs from each group developed short clinical evidence statements using a proforma. This proforma asked the individual to summarise the evidence, provide clinical and non-clinical considerations that arose from these studies and then develop short clinical evidence statements. These were presented to the group for discussion and then further refined and presented to the group again for approval. For some clinical questions the evidence provided by studies carried out in adult patients was also considered. This was done only when paediatric evidence was missing and the GDG felt that adult studies provided information that was applicable to children and young people. In particular, some recommendations to clinical questions were informed by the recently published RCP guideline for adult stroke\textsuperscript{12}. Where adult data had been used in this way it is indicated in the description of the evidence.
Where the evidence base to formulate recommendations were lacking, an expert consensus was necessary. In several specific and contentious areas, the GDG felt that using a formal consensus methodology and engaging with professionals outside the GDG would improve the strength of recommendation (see Section 2.9).

For those recommendation retained from the 2004 publication, the wording was updated in line with that of the National Institute for Health and Care Excellence’s (NICE) and with the recommendations presented in this guideline.

2.9. Delphi consensus method

The evidence base to formulate recommendations was lacking in many areas and expert consensus was often necessary. In these instances, and as indicated in the methodology, a three-round online Delphi consensus method was used to derive recommendations. This involved the participation of 70 healthcare professionals from different medical specialities including general paediatrics, neurology, neuroradiology, haematology, and neurosurgery. Where recommendations have been developed via the Delphi consensus method, these have been referenced [Delphi]. For further details of how Delphi Panel members were recruited, see Appendix 5.

Delphi statements were developed by a sub-group and agreed by the full GDG. The Delphi participants rated the statements using a 1 to 10 point Likert scale (with 1 being strongly disagree and 10 being strongly agree), with an option to select ‘not my area of expertise’. In addition, the Delphi panel was given the opportunity to add comments during the survey, which were then reviewed by the GDG and used to amend statements where consensus was not reached before being presented to the panel once more. Consensus was considered reached when at least 75% of the ratings received were between 7 and 10 on the Likert scale, indicating a high level of agreement.

The defining rules of the Delphi consensus method were as follows:

- The panel should be multidisciplinary and include at least eight representatives from each speciality.
- A 10-point Likert scale was used for panellists to provide their responses to statements.
- Consensus agreement defined as 75% of panellists who responded selecting 7, 8, 9 or 10 on the Likert scale.
- Consensus agreement should be calculated based on the number of respondents for that round, excluding those who did not answer individual statements or who answered ‘not my area of expertise’.
• There should be a minimum of two rounds.
• Any recommendations whose underpinning Delphi statements failed to reach consensus will be made explicit and all Delphi results can be found in Appendix 5.

The Delphi panel survey was conducted online with panellists being contacted via e-mail. The Delphi panel voted on a total of five statements in ‘round 1’. The Delphi panel voting is summarised in Table 2.1:

Table 2.1. Delphi panel voting

<table>
<thead>
<tr>
<th>Delphi round</th>
<th>Number of statements</th>
<th>Number of respondents</th>
<th>Response rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Round 1</td>
<td>5</td>
<td>69</td>
<td>99%</td>
</tr>
<tr>
<td>Round 2</td>
<td>3</td>
<td>56</td>
<td>80%</td>
</tr>
<tr>
<td>Round 3</td>
<td>2</td>
<td>54</td>
<td>77%</td>
</tr>
</tbody>
</table>

Following ‘round 1’ voting any statements that reached consensus (75% or more votes indicating strong agreement) were used to shape the guideline recommendations. Where there was no consensus, Delphi panel voting and comments were reviewed by the GDG and statements were revised following ‘round 1’ in order to improve their clarity or to bring them in line with current practice as suggested by Delphi panel comments. These revised statements were then sent out for ‘round 2’ voting.

The GDG reviewed the Delphi findings from ‘round 2’, accepting statements that received consensus as recommendations and amending and recirculating those that did not for a ‘third round’ of Delphi in ‘round 2’. For statements which did not reach consensus in the ‘third round’, the GDG considered the Delphi findings and comments and consensus was agreed within the group. Voting for the Delphi statements is given as a percentage of panellists agreeing with the statement (voting 7, 8, 9 or 10). Full details of Delphi statements and results are given in Appendix 5.

2.10. Stakeholder involvement

Due to the breadth of the scope, input from a wide variety of specialities was required in all stages of the guideline development.

The GDG included representatives from stakeholder organisations, and stakeholders were invited to comment on the draft scope and draft guideline (for a full list of stakeholders see Acknowledgments section).

2.11. Parent, carer and patient participation

Guideline development was designed to involve parents, carers and young people from the outset and who were involved at every stage, and the GDG included three parents of
affected children and young people. The GDG and stakeholder representatives also included parent, carer and patient information charities and organisations/associations.

The parent representatives from the GDG reviewed in detail both the clinical guideline and associated parent/carer guideline.

2.12. **External peer review**

Initially a stakeholder consultation on the guideline scope took place between August and September 2015. During this time, stakeholders were given the opportunity to comment on the scope. All comments received were collated and reviewed for consideration and discussion.

Following the evidence review and agreement on the content by the GDG, there was a period of external peer review during which key stakeholders reviewed the guideline.

The stakeholder consultation took place between December 2016 and January 2017, and during this time stakeholders were given the opportunity to comment on the guideline. Relevant changes were made to the guideline draft after careful consideration and discussion of the stakeholder feedback (see Appendix 6) with the GDG. Full details of stakeholders can be found in the Acknowledgements section.

2.13. **Parent and young person engagement in formulation of recommendations**

To allow recommendations to be made which encompassed the views of parents and young people who had suffered a stroke, two parent/carer and young people engagement workshops were conducted.

The workshops were designed and facilitated by a member of the GDG, and were supported by the RCPCH and Stroke Association. The workshops use focus groups to systematically capture data that could be thematically analysed to support development of recommendations and to help shape the wording. The process followed is outlined in Appendix 7.

The content of the workshops focussed on the following domains:

1. Information needs for families both at the time of diagnosis and afterwards.
2. Support needs for the family as a whole and during transfer from hospital to home.
3. Information and support needs for transfer through education stages and transition into adult healthcare.

The resulting recommendations were then discussed at the GDG meetings, agreed, and inserted into the appropriate sections of the guideline.

2.14. Quality assurance

The guideline draft was independently appraised in line with the Appraisal of Guidelines for Research & Evaluation tool (AGREE II)\textsuperscript{16}, which is an international tool used to assess the quality and reporting of practice guidelines. This was done to ensure all methodological requirements were met in order to display the NICE accreditation mark.

The RCPCH holds the prestigious NICE accreditation for the development process used to produce clinical guidelines. The accreditation will remain valid until 2020 and applies to clinical guidelines produced using the methods and processes described in the RCPCH ‘Setting Standards for Development of Clinical Guidelines in Paediatrics and Child Health’\textsuperscript{14} publication. Further information on NICE accreditation can be found at http://www.nice.org.uk/About/What-we-do/Accreditation.

2.15. Guideline update

It is recommended that this guideline is reviewed, with a view to update or partially update, within the next three years (2020) so that clinical recommendations take into account important new information.

The evidence should be checked and healthcare professionals and patients views should be sought to assess whether all or part of the guideline requires updating. If important new evidence is published at other times, which is likely to influence the recommendations, it may be decided that a more rapid update of some recommendations is necessary.
3. Acute diagnosis of stroke in childhood

The presentation of arterial ischaemic stroke (AIS) and haemorrhagic stroke (HS) in children and young people can be non-specific, and include symptoms such as isolated vomiting or fever. Early recognition depends on both awareness and consideration of the diagnosis in order to expedite appropriate investigation and management. It is impossible to distinguish between AIS and HS and to differentiate symptoms of real stroke from ‘stroke mimics’ on clinical grounds, and therefore imaging is key to diagnosis. Predisposing risk factors should be considered, and, if present, should increase suspicion of the diagnosis of stroke.

Given the low prevalence of stroke in children and young people and the frequency of non-specific symptoms, the diagnosis of stroke can be elusive. It is important to emphasise that the majority of children and young people with stroke (AIS and HS) will have an acute focal neurological syndrome, and would meet adult criteria as assessed with the FAST tool9 (‘Face, Arms, Speech, Time’ indicators). The clinical significance of an acute focal neurological deficit in children and young people is frequently not recognised by healthcare professionals, which is a key factor in diagnostic delay; whereas, parents would rapidly identify any concerns.

Review questions

- What are the presenting clinical signs and symptoms for suspecting AIS/HS diagnosis in children and young people?
- What is the role, modality and timing of brain imaging in diagnosis of AIS/HS in children and young people?

3.1. Clinical presentation

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified 46 studies2,17-61 which reported data on the clinical signs and symptoms present in AIS in children and young people.

Of these, 11 were cohort studies2,17,20,22,25,37,40,45,53,55,57, 13 case series21,24,27-30,46,47,50-52,54,61, one cross sectional study31, three prospective studies32,43,44, 10 retrospective case series33,38,39,48,58-60, two retrospective chart reviews34,36, one retrospective study42, one retrospective case notes review26 and four retrospective cohort studies35,41,49,56. The quality of evidence was classified as high in two studies2,45, moderate in 16 studies17,25,31,34,41,43,44,47,48,50,52,53,56,58,60,61 and low in 28 studies18-24,26-30,32,33,35-40,42,46,49,51,54,57,60,62.
Sample size ranged from seven to 287 children, and study populations were reported from a number of regions including Europe (ten studies\textsuperscript{23,24,26,27,30,32,46,55,57}), America (nine studies\textsuperscript{17,18,25,31,34,35,39,52,61}), the UK (four studies\textsuperscript{2,29,45,48}), Australia (four studies\textsuperscript{9,43,59,60}), Canada (three studies\textsuperscript{22,38,51}), China (four studies\textsuperscript{42,56,58,63}), Saudi Arabia (two studies\textsuperscript{21,28}), Africa (two studies\textsuperscript{40,49}), India (two studies\textsuperscript{20,37}), Turkey (three studies\textsuperscript{33,36,50}), Brazil (one study\textsuperscript{47}), Korea (one study\textsuperscript{41}) and a mixed population (one study\textsuperscript{44}) which included samples from Australia, Canada, Chile, China, Georgia, Germany, Malaysia, Thailand, the UK, and the USA. For the studies where applicability to the UK population was less certain, the results were approached with caution.

Sixteen studies\textsuperscript{30,58,64-77} described the signs and symptoms at presentation in HS. Of these, there were six retrospective case series\textsuperscript{30,65,66,71,72,77}, two cohort studies\textsuperscript{68,74}, and eight case series\textsuperscript{58,64,67,69,70,73,75,76}. The quality of evidence was moderate in seven studies\textsuperscript{65-67,72-74,77} and low in nine studies\textsuperscript{30,58,64,68-71,75,76}.

Sample size ranged from 10 to 249 children, and study populations were reported from a number of regions including China (four studies\textsuperscript{58,68,71,77}), Europe (four studies\textsuperscript{30,67,74,75}), America (two studies\textsuperscript{65,72}), India (two studies\textsuperscript{70,73}), Egypt (one study\textsuperscript{69}), Korea (one study\textsuperscript{64}), Thailand (one study\textsuperscript{76}), and Turkey (one study\textsuperscript{66}).

The features of AIS or HS are presented in Table 3.1 together with corresponding studies:

**Table 3.1.** Presenting features of AIS or HS.

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Type of stroke</th>
<th>AIS</th>
<th>HS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal neurological deficit, including lateralising weakness</td>
<td></td>
<td>31–100</td>
<td>13–60</td>
</tr>
<tr>
<td>and/or cranial nerve palsy and/or sensory loss</td>
<td></td>
<td>2,17-21,23,24,26-28,30-32,34,36,37,39-61</td>
<td>20,21,26,28,30,3</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td>5–85</td>
<td>14–91</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4,17-31,33,34,36,37,40-53,55,56,58,61</td>
<td>20,21,23,28,30,3</td>
</tr>
<tr>
<td>Headache</td>
<td></td>
<td>4–64</td>
<td>20–77</td>
</tr>
<tr>
<td></td>
<td></td>
<td>19,20,22-24,26,32-35,37,39,41,44,46,50-55,57-59,76</td>
<td>20,21,26,30,34,3</td>
</tr>
</tbody>
</table>

21
<table>
<thead>
<tr>
<th>Condition</th>
<th>Pages</th>
<th>Reference Numbers</th>
<th>CI/Range</th>
<th>Reference Numbers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aphasia</td>
<td>3–45</td>
<td>17, 21, 26, 27, 30-33, 34, 43, 44, 50, 51, 53, 57, 59, 60</td>
<td>4–39</td>
<td>21, 30, 60, 65, 68</td>
</tr>
<tr>
<td>Altered conscious level including transient loss of consciousness or coma</td>
<td>3–71</td>
<td>2, 18, 20, 21, 23, 24, 26 - 29, 32, 36, 37, 40, 44 -48, 50-53, 58, 60, 61, 69, 77</td>
<td>3–63</td>
<td>20, 28, 30, 37, 58, 65, 67, 68, 70, 74, 76</td>
</tr>
<tr>
<td>Altered mental status</td>
<td>5–48</td>
<td>34, 39, 41, 43, 56, 57 - 54, 60, 64, 72, 75</td>
<td>53–57</td>
<td>34, 39, 60, 64, 72, 75</td>
</tr>
<tr>
<td>Ataxia, vertigo or dizziness</td>
<td>2–31</td>
<td>17, 29, 27, 29, 30, 37, 4, 34, 51, 53-56</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>Nausea or vomiting</td>
<td>7–58</td>
<td>21, 26, 32, 33, 35, 41, 4, 35, 57, 58</td>
<td>4–70</td>
<td>21, 26, 58, 65, 66, 6, 9, 70, 72, 74, 76, 77</td>
</tr>
<tr>
<td>Neck pain</td>
<td>3–5</td>
<td>25, 24, 29</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>Fever</td>
<td>5–43</td>
<td>18, 20, 34, 35, 41, 42, 5, 0, 56</td>
<td>9–46</td>
<td>20, 34, 66</td>
</tr>
</tbody>
</table>

There is moderate evidence to suggest that clinical presentation does not distinguish between AIS and HS. In both stroke sub-types, the most common symptoms at onset were:

- Acute focal neurological deficit
- Seizure
- Headache

In addition, there is moderate evidence that children less than one year old are more likely to present with seizures than older children (e.g. 32 to 85% versus 9 to 26% reported) and that headache at presentation is common in stroke in children with sickle cell disease.

One retrospective case series of 46 children investigated the applicability of adult stroke scores to stroke in children, and found that 78% of children diagnosed with stroke had at least one positive variable on the FAST criteria and 81% had a positive score of one or greater on the Recognition of Stroke in the Emergency Room (ROSIER) scale. Further details on each study are provided within the evidence tables, in Appendix 4c.
Linking the evidence to the recommendations

The majority of evidence is drawn from studies in children with confirmed diagnosis of stroke. It is therefore difficult to comment on the prevalence of stroke among children presenting with individual symptoms, such as seizures. Whilst the combination of clinical features listed above is highly suggestive of stroke, there is insufficient data on the frequency of this diagnosis in children with similar presentation from alternative causes. Moreover, it is clear that stroke cannot be excluded in children with non-specific presentations. This is especially important in high risk groups, such as children with sickle cell disease, in whom the threshold for considering a stroke diagnosis should be low. The recommendation for a child entering the ‘acute paediatric stroke pathway’, with a need for urgent neuroimaging, was therefore based upon the clinical experience of the scenarios where a stroke is likely to occur.

The ROSIER scale has not been validated in children and although it may be reasonable to apply the positive criteria, the prevalence of seizures and possible loss of consciousness in the context of childhood stroke makes it inappropriate to apply the negative scoring criteria and hence its use is not recommended. While there are no validated diagnostic stroke scores in children; application of the FAST is reasonable, although absence of FAST criteria does not exclude stroke. A further advantage of using FAST is that it is now a tool very familiar to professionals in the pre-hospital and emergency setting.

Recommendations

Professionals in health and education services should be aware of the possibility of stroke in children and young people at higher risk (e.g. sickle cell disease (SCD), congenital heart disease). Carrying written or other alerting materials (e.g. medical alert bracelets) should be discussed with the parents/carers, family and child/young person.

- Use the FAST criteria to determine stroke in children and young people, but do not rule out stroke in the absence of FAST signs.

- Do not apply the ROSIER scale for identifying stroke in children and young people.

- Undertake urgent brain imaging of children and young people presenting with one or more of the following symptoms:
  - Acute focal neurological deficit
  - Aphasia
  - Reduced level of consciousness (age-appropriate Glasgow Coma Scale
(GCS) less than 12 or AVPU (‘Alert, Voice, Pain, Unresponsive’) less than V) at presentation

- Consider urgent brain imaging for children and young people presenting with the following symptoms which may be indicative of stroke:
  - New onset focal seizures
  - New onset severe headache
  - Altered mental status including transient loss of consciousness or behavioural changes
  - New onset ataxia, vertigo or dizziness
  - Sudden onset of neck pain or neck stiffness
  - Witnessed acute focal neurological deficit which has since resolved

- Be aware that the following non-specific symptoms can be present in a child presenting with stroke:
  - Nausea or vomiting
  - Fever

- Be aware that acute focal neurological signs may be absent, and that attention should be given to parental or young person concerns about the presentation of unusual symptoms.

### 3.2. Diagnosis

**Evidence summary**

A systematic review was conducted for the relevant clinical questions and identified eleven studies\(^45,46,51,78-85\) which compared and demonstrated the superiority of magnetic resonance imaging (MRI) over computerised tomography (CT) scan in terms of sensitivity and specificity for the initial diagnosis of AIS in children and young people. However, many studies were retrospective and described heterogeneous study populations with different imaging pathways.

Of the eleven studies, there were two retrospective cohort review studies\(^78,83\), one cohort study\(^45\), one prospective and retrospective case series\(^79\), one retrospective case review\(^81\), one retrospective review\(^80\) and five case series\(^46,51,82,84,85\). The quality of evidence was classified as high in two studies\(^45,80\), moderate in two studies\(^79,83\) and low in seven studies\(^46,51,78,81,82,84,85\).
Only six of the included studies assessed HS as a separate entity, separately or together with AIS\textsuperscript{39,65,67,77,86,87}. Of these studies, two were case series\textsuperscript{67,86}, one was a database review\textsuperscript{39}, one was a prospective case series\textsuperscript{87}, and two were retrospective case series\textsuperscript{65,77}. The quality of evidence was classified as moderate in four studies\textsuperscript{65,67,77,87} and low in two studies\textsuperscript{39,86}.

In the studies discussing HS, the modalities of imaging included: MRI, magnetic resonance angiogram (MRA), CT, computed tomography angiography (CTA), catheter angiography (CA), and digital subtraction angiography (DSA). A CT scan was explicitly used to diagnose haemorrhage in three of these studies\textsuperscript{67,77,86}, and described a shorter time from arrival at the emergency department (ED) to first scan than MRI\textsuperscript{39}. In HS, MRA was shown to be able to detect clinically relevant vascular pathologies when compared with CA\textsuperscript{86} and was found to be effective at visualising a range of arteries at all ages\textsuperscript{87}.

The sample sizes across the AIS and HS studies ranged from 16 to 204 children, and study populations were reported from a number of regions including America (three studies\textsuperscript{65,85,88}), Australia (one study\textsuperscript{83}), South Africa (one study\textsuperscript{86}), Canada (one study\textsuperscript{51}), China (one study\textsuperscript{77}), Europe (seven studies\textsuperscript{46,67,78,80,82,84,87}), Argentina (one study\textsuperscript{79}), and the UK (two studies\textsuperscript{45,81}). For the studies where applicability to the UK population was less certain, the results were approached with caution.

Two more studies of low quality from France and America demonstrated that CA has a role in the identification of possible dissection in posterior circulation AIS and also in anterior circulation AIS where CTA/MRA is normal or questionable and to assess severity or progression of arterial disease\textsuperscript{84,85}.

It is commonly acknowledged that if HS is detected on the initial scan, an underlying vascular lesion should be actively sought as these are the commonest cause of childhood HS. Such lesions include arteriovenous malformations (AVM), aneurysms and cavernous malformations. Given the wide range of available modalities, with variable spatial resolution and radiation burden, the imaging strategy in HS should be determined by clinical presentation and likely underlying lesion, in discussion within a neurovascular multidisciplinary team (MDT). Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

As in adult stroke, minimising the time from symptom onset to diagnosis is essential for the initiation of successful hyperacute therapy in AIS, and to facilitate timely transfer to a neurosurgical centre in HS. Rapid diagnosis in HS is key to ensuring any underlying vascular lesion is adequately assessed to render it safe from early recurrence of
haemorrhage. Using MRI allows more specific information to be obtained regarding anatomical features (i.e. site, extent and vascular involvement with MR angiography). Despite the greater sensitivity and specificity of MRI, the time critical nature of stroke diagnosis means that unless MRI is immediately available following initial emergency clinical assessment, it has not been recommended for primary imaging in this guideline. Instead, it has been recommended that a CT scan should be performed in the first instance, which will detect any associated haemorrhage and exclude other major ‘stroke mimics’, and that an MRI is performed as soon as possible afterwards. The superior temporal and spatial resolution of MRI, and the radiation burden associated with CT scans, are acknowledged in this guideline but the recommendation aims to be pragmatic and to facilitate rapid diagnosis.

The MR imaging protocol for childhood stroke should include standard brain sequences, diffusion-weighted imaging, susceptibility-weighted imaging and MRA of the intracranial circulation (and cervical vessels in AIS). Studies show good correlation between the results of catheter angiography (CA) and non-invasive vascular imaging techniques such as CTA and MRA in the context of both AIS and HS.

The current clinical role of perfusion imaging and/or techniques such as arterial spin labelling is unclear in this population.

**Recommendations**

- Ensure that a cranial CT scan is performed within one hour of arrival at hospital in every child with a suspected stroke. This should include:
  - CTA (covering aortic arch to vertex), if the CT scan does not show haemorrhage **OR**
  - CTA limited to intracranial vascular imaging, if HS is demonstrated.

- Initial scan images should be reviewed on acquisition and if necessary transferred immediately to the regional paediatric neuroscience centre for review.

- Consider primary imaging using MRI in suspected stroke only if it is available within one hour of arrival at hospital.

- Provide MRI in a clinically timely manner for both AIS and HS patients for improved diagnostic resolution, if not obtained in/at the initial imaging investigation.
• Provide MRI within 24 hours if initial CT is negative and stroke is still suspected.

• Consider adding MRA at the time of undertaking MRI; this should cover the aortic arch to vertex in AIS and can be limited to the intracranial circulation in HS.
4. Referral pathways and further investigations

Childhood stroke is an emergency and amenable to acute interventions that can potentially impact on morbidity and mortality. However, relative rarity and lack of recognition means that children and young people are often not diagnosed and triaged within a time frame when hyperacute interventions could be delivered. Interventions include intravenous (IV) thrombolysis and endovascular recanalization therapy, both of which are proven benefit in the early treatment of adult arterial ischaemic stroke (AIS).

Recognition by community healthcare providers and rapid transfer to a local hospital with appropriate acute paediatric services is a vital first step if a pathway similar to adult stroke is to be initiated. The management of child stroke is likely to involve multiple medical specialties in most secondary and tertiary centres.

This guideline aims to heighten awareness of stroke and improve the recognition of signs and symptoms by parents, triage services and pre-hospital teams. A pre-alert from the pre-hospital team should trigger an appropriate team based response in the emergency department and is recognised as being an important component of the pre-hospital care of a child with a suspected stroke.

At present, there is often a delay in the diagnosis of acute stroke in children and young people due in part to the high rate of ‘stroke mimics’ but also to the lack of an agreed referral and management pathway. This chapter aims to address this by recommending an acute stroke pathway that will allow for the same prioritisation of childhood stroke as that in adults.

The suggested acute stroke pathway involves urgent cross sectional imaging, ideally within one hour of arrival at hospital, and the urgent electronic transfer of images to a paediatric neurosciences centre for neuroradiological review. It is recognised that there are regional variations in the organisation of relevant services but local arrangements involving the general paediatric service, regional neuroscience service and paediatric intensive care (PIC) network should be considered analogous to the now robust arrangements that exist for paediatric neurotrauma, where some elements are time-critical, some elements require transfer to a tertiary centre on a more or less urgent basis and that clinicians come to a locally implementable decision about the appropriate route of care for the specific child.

Neuroscience and PIC Operational Delivery Networks (ODN’s) or the regional paediatric
The role of the PIC transport service in coordination with the hyperacute multidisciplinary response to children presenting to a district general hospital (DGH) with acute haemorrhagic stroke (HS) or AIS

Pathways for children with AIS:
- who are otherwise physiologically stable after thrombolysis
- who are otherwise physiologically stable but who are not eligible for thrombolysis, and
- who have middle cerebral artery (MCA) infarcts and clinical features that qualifies them for decompressive hemicraniectomy

The pathway for children with HS:
- who are otherwise physiologically stable, and
- who are otherwise physiologically unstable (time-critical)

Such pathways should identify:

- Definitions for time-critical transfers in children with acute stroke
- Locally developed protocols that will allow DGH personnel to perform these transfers if more clinically appropriate than PIC transport team
- The destination in the neuroscience centre for children in each of these groups (paediatric intensive care unit (PICU) versus neuroscience ward)
- Ideally a single number (e.g. the PIC transport service) that will allow hyperacute mobilisation of the appropriate multidisciplinary assessment of
  - the clinical situation
  - cross-sectional imaging
  - suitability for hyperacute treatment
  - consent for novel interventions

The guideline development group (GDG) note that a potential advantage of using the PIC network as a hyperacute paediatric stroke receiving area is that this would allow accurate data capture of all children with HS and AIS (assessed for suitability and either receiving or being declined hyperacute therapies) using the Paediatric Intensive Care Audit Network (PICANet), which is an existing national database that records data on all PIC admissions; this database already has additional subspecialty datasets (e.g. for children receiving renal replacement therapy). It is therefore feasible that a stroke dataset could be developed at
no additional cost to allow robust data capture for all children presenting with acute stroke.

**Review questions**

Referral and care pathways

- What is most appropriate referral pathway for children and young people:
  - diagnosed with acute AIS/HS
  - suspected clinically with acute AIS/HS
  - diagnosed with clinically silent AIS/HS
  - diagnosed with unruptured, at risk cerebrovascular lesion
- Does the setting where children and young people with AIS/HS receive acute care affect mortality, morbidity and complications?
- What are the key elements of acute medical care management of children and young people with AIS/HS?
- Who are the key individuals to be involved in the acute management of children and young people with AIS/HS?

**4.1. Referral and care pathway for childhood stroke**

**Evidence summary**

A systematic review was conducted for the relevant clinical questions and identified eight studies10,37,39,80,83,89-91 which explored the referral and care pathways and best course of action for children and young people with stroke. The quality of evidence was classified as high in one study80, moderate in four studies10,83,89,91 and low in three studies37,39,90.

Study populations were reported from a number of regions including America (five studies10,39,89-91), Switzerland (one study80), Australia (one study83) and India (one study37). Study sample sizes ranged from 79 to 10,236 children.

Bernard and colleagues89 detailed the set-up of the Thrombolysis in Paediatric Stroke trial (TIPS) and list individuals and facilities needed to deal with an emergency stroke patient. The specialities comprising the stroke team detailed in this study were as follows: paediatric stroke neurologist, haematologist, neuroradiologist, neurosurgeon, interventional neuroradiologist, cardiologist, rheumatologist, neuropsychologist, rehabilitation specialist, psychologist, geneticist (vascular), and social worker. The study assessed centres’ readiness to deal with an acute paediatric stroke patient based on the availability of an established system for stroke triage, a 24/7 stroke team, emergency department (ED) and PICU stroke protocols, and the availability of 24/7 magnetic
resonance imaging (MRI) with the option for sedation. The study showed that after preparation for this trial there was a greater readiness for treating acute childhood stroke in the centres taking part; thus suggesting that recommendations for hyperacute intervention included in this guideline have the potential to improve readiness for treatment.

Two studies\textsuperscript{37,90} suggest pathways for the initial evaluation of a child presenting with a stroke. Gumer and colleagues\textsuperscript{90} presented a pathway for the initial diagnostic imaging of a child presenting in an ED with a stroke based on the most common aetiologies of HS and AIS. They suggested that computerised tomography (CT) imaging should be used initially to differentiate between AIS and HS especially as it can be performed quickly and often without sedation. Following CT, magnetic resonance angiogram (MRA) and haematological tests are recommended for both HS and AIS with catheter angiography (CA) for a final evaluation if no aetiology has been found. Similarly, Kalita and colleagues\textsuperscript{37} presented a protocol for identifying the underlying aetiologies and risk factors in children presenting with stroke which included patient history, initial investigations and imaging.

Ichord and colleagues\textsuperscript{10} presented the evaluation of the paediatric version of the National Institutes of Health Stroke Scale (PedNIHSS) which is a freely available assessment tool for stroke related acute neurological deficit, the adult version of which is predictive of stroke outcome. This study showed the PedNIHSS to have excellent inter-rater reliability.

Ladner and colleagues\textsuperscript{39} implemented a rapid assessment and imaging protocol for suspected paediatric stroke. There was a high rate of stroke or clinically important ‘stroke mimics’ and they recommended implementing ‘paediatric stroke alerts’ to improve the time course to diagnosis and, ultimately, to treatment.

Finally, three studies\textsuperscript{80,83,91} discussed the factors that influence time to diagnosis and care received, including the location of the child/young person at symptom onset and during treatment.

**Linking the evidence to the recommendations**

It is apparent that the identification and management of childhood stroke leaves much room for improvement. Some of these areas that need to be addressed include awareness of the possibility of stroke and recognition of signs and symptoms by the whole spectrum of clinical practice and subsequent triage and urgent transfer to appropriate emergency departments supported by acute paediatric services, emergent imaging, referral to and advice from a regional paediatric neuroscience centre facilitated by a regional paediatric intensive care transport service. A pre-alert or priority call to trigger an appropriate team-
based response in the emergency department is recognised as being an important component of the pre-hospital care of a child with a suspected stroke.

Reorganising the care of acute childhood stroke in this way will be challenging at many levels, but the use of networks already in existence might facilitate this. Key to change is an attitudinal shift in the approach of clinicians to childhood stroke. The pathway proposed, involving many clinicians, liaison between secondary and regional centres and rapid acquisition and transfer of neuroimaging, coordinated by a regional paediatric transfer service, already operates for paediatric head injury. A challenging aspect of the pathway proposed in this guideline is the identification of children who can benefit from hyperacute IV thrombolysis without delay, and initiation of treatment in the emergency department or general paediatric unit.

It is acknowledged that not every element of the care pathway proposed will be available at every DGH. The proposed pathway aims to act as a framework and local protocols will need to be developed to deliver the care goals set out. This will need to include consideration of which personnel need to be involved (including how they should be contacted), the care setting and other specifics such as clinical monitoring and drug dosing.

The literature provided some examples of elements of pathways and tools that were used as starting points for the acute paediatric stroke pathway laid out in this section. In addition, parents/carers and young people in the workshops reported the importance of early communication with clinicians from the time of presentation to hospital. It was felt that there was the need for regular updates about their child’s condition, care processes and investigations. The opportunity to ask questions of clinicians and be involved in decision making wherever possible was stressed as important from this early stage of care onwards. The findings of the literature, the workshops and the expertise of the GDG members all contributed to the formation of the following recommendations. Due to the lack of published evidence regarding the pre-hospital care of children and young people, appropriate recommendations were also taken from the Royal College of Physicians (RCP) national clinical guideline for stroke12.

**Recommendations**

- Community medical services and ambulance services (including call handlers, telephone triage and advice services such as National Health Service (NHS) 111 and primary care reception staff) should be trained to recognise children and young people with symptoms suggesting an acute stroke as an emergency
requiring urgent transfer to hospital.

- Children and young people seen by ambulance clinicians, or primary care providers outside hospital with the sudden onset of acute focal neurological symptoms should be screened for hypoglycaemia with a capillary blood glucose, and for stroke using a simple screening tool such as FAST (‘Face, Arms, Speech Time’). Where these are normal or negative, but stroke is still suspected, the acute stroke pathway should be used.

- Children and young people with persisting neurological symptoms who screen positive using a validated tool (or who screen negative, but in whom stroke is suspected) should be transferred to an emergency department with paediatric services urgently.

- The possibility of stroke should still be considered in children and young people where there is a clear history of an acute neurological deficit which has since resolved.

- The pre-hospital care of children and young people with suspected stroke should minimise time from call to arrival at hospital and should include a hospital pre-alert to expedite specialist assessment and treatment.

- The acute paediatric stroke pathway, according to a locally agreed protocol, should be triggered upon arrival at the emergency department (see Diagrams 4.1 and 4.2).

- Care should be consultant delivered at the earliest opportunity, involving a multi-specialty team according to the child’s clinical need.

- If the child has sickle cell disease (SCD), paediatric haematologists should also be involved in acute management.

- Local protocols should be developed to coordinate liaison between specialties at the secondary and regional centres (including acquisition and transfer of images) and to facilitate clinically appropriate and time-sensitive transfers between centres. This could involve the PIC transport network, or use local arrangements already in existence for management of other paediatric neurological emergencies, e.g. acute neurotrauma.
• Parents/carers and young people should be regularly informed and updated throughout the care process. This should include age-appropriate and multi-format information for the child or young person as well as the parent/carer about the condition/suspected condition, investigation plans and findings, and management plans.

• Where possible and appropriate, the young person and parents/carers should be actively involved in decision making.
Diagram 4.1. Pre-hospital pathway for the management of suspected childhood stroke.
Diagram 4.2. Acute paediatric stroke pathway (according to locally agreed arrangements for delivery).
5. Acute management

Delay in diagnosis and treatment of stroke or failure to recognise early complications such as cerebral oedema may adversely affect the outcome. The initial assessment of a child with suspected stroke should be focussed on distinguishing symptoms of real stroke from ‘stroke mimics’, and haemorrhagic (HS) from arterial ischaemic (AIS) stroke. Recognising potential complications and stroke severity is crucial so appropriate management, including whether or not intensive care or neurosurgical management is needed, can be determined.

Children and young people with acute stroke are likely to present to or be taken to their local Accident and Emergency (A&E) Department or Paediatric Service; however, these departments may not have specialist children’s services (e.g. intensive care, neurosurgery and neurology services), usually located at regional centres. As such the initial medical assessment and care will often be provided in non-specialist settings, with continuing treatment given following transfer to specialist services in regional units. It is essential to consider how regional and local services work together to allow appropriate assessment and management of children and young people with suspected stroke.

An added difficulty is that the pathogenesis of adult stroke differs from that which predominates in children, the possibility of a non-stroke aetiology accounting for an acute focal neurological deficit is greater in children than in adults. Investigation and treatment approaches appropriate in adults might not therefore always be applicable to children and stroke pathways, and services set up for adults are not generally appropriate settings for children and young people.

Review questions

- In children and young people with AIS/HS, what is the optimal way to assess:
  - neurological status?
  - stroke severity?
- In children and young people with acute AIS/HS, what is the framework for early functional assessment including: nutrition and hydration, communication difficulties, swallowing difficulties, cognition, and mobility?
- What are the likely complications during the acute and sub-acute phase of recovery from AIS/HS?
- What are most effective ways to detect, prevent and minimize AIS/HS complications in children and young people?
• Which elements of the information and care provided by the key health professionals and other sectors are most important for AIS/HS patients and their families at the acute stage (including environmental factors)?

5.1. **Acute assessment**

**Evidence summary**

**Clinical assessment**

A systematic review was conducted for the relevant clinical questions and identified 12 studies\(^\text{10,27,37,42,92-99}\) relevant to the assessment of children and young people with AIS. Of the 12 studies, there were four cohort studies\(^\text{10,37,95,97}\), three cross sectional studies\(^\text{92,93,98}\), two retrospective reviews\(^\text{42,94}\), two case series\(^\text{27,96}\) and one retrospective case series\(^\text{99}\).

The quality of evidence was classified as high in two studies\(^\text{96,97}\), moderate in seven studies\(^\text{10,92-95,98,99}\) and low in three studies\(^\text{27,37,42}\). Study populations were reported from a number of regions including America (five studies\(^\text{10,92,93,95,98}\), Australia (one study\(^\text{94}\)), Europe (three studies\(^\text{27,96,99}\), India (one study\(^\text{37}\), China (one study\(^\text{42}\)), and a mixed population (one study\(^\text{97}\)) which included samples from America, Asia, Australia, Canada, Europe, and South America.

For HS, the systematic review identified seven studies\(^\text{100-106}\), of which five were retrospective case series\(^\text{100,101,103-105}\) and two were case series\(^\text{102,106}\). The quality of evidence was classified as high in one study\(^\text{103}\), moderate in two studies\(^\text{100,106}\) and low in four studies\(^\text{101,102,104,105}\). Study populations for HS were reported from a number of regions including India (two studies\(^\text{103,104}\), China (two studies\(^\text{102,106}\), Canada (one study\(^\text{101}\)) and Europe (two studies\(^\text{100,105}\)).

There was one moderate quality multicentre prospective study which validated the use of the Paediatric National Institutes of Health Stroke Scale (PedNIHSS) for objective assessment of neurological status and as an indicator of stroke severity\(^\text{10}\) in AIS and another which showed that the PedNIHSS could be used retrospectively\(^\text{92}\), but there was no evidence on the neurological assessment of children presenting with HS.

There was evidence from four studies suggesting increased mortality and neurological morbidity in children with reduced level of consciousness at the time of presentation with stroke\(^\text{27,37,42,97}\) and evidence of worse outcome in children with fever\(^\text{42,96}\) or hypertension\(^\text{94}\) at presentation from three studies.

Radiological features associated with a poorer outcome in AIS were larger stroke volume which predicted a higher risk of haemorrhagic conversion\(^\text{93}\) and the presence of
arteriopathy or posterior circulation stroke which both predicted a higher risk of recurrence of stroke\textsuperscript{98,99}.

There was low and high quality evidence correlating outcome with preoperative Hunt and Hess grade and Fisher grade\textsuperscript{102-105} at presentation with outcome in children with aneurysmal subarachnoid haemorrhage. Similar data was found regarding cavernous malformation and AVM bleeding using two non-validated scales, the Zabramski classification and Spetzler-Martin grade\textsuperscript{100,106}.

The findings of the parent/carer and young person workshops suggest that the priorities around the time of diagnosis and in the early stages of care are centred on early and regular contact with clinicians. Clinicians should communicate in a way that is family-centred, proactive and sensitive and provide enough time to ask questions. Care processes and reasons for investigations should be explained in detail. A named key contact during hospital admission can help to keep families updated on what to expect, and the child/young person’s progress, on a regular basis. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

PedNIHSS has good inter-rater reliability and predictive value in terms of stroke severity to justify using it routinely in assessment of children and young people with AIS. As such PedNIHSS is recommended.

The evidence does not suggest that any of the scales used in HS assessment offer additional value over the Glasgow Coma Scale and routine observations.

Level of consciousness is the best predictor for serious outcome in both AIS and HS. Brain imaging can estimate stroke size, identify the presence of arteriopathy and help to guide treatment decisions, such as the use of anticoagulation or neurosurgical interventions (external ventricular drain and craniectomy). The use of brain imaging is essential when considering the appropriateness of thrombolytic therapy.

**Recommendations**

- Monitor blood pressure, temperature, oxygen saturation, heart rate and respiratory rate in all children and young people presenting with a clinical diagnosis of stroke (see Diagram 4.2.).

- Use the PedNIHSS and age-appropriate Glasgow Coma Scale (GCS) or AVPU...
(‘Alert, Voice, Pain, Unresponsive’) to assess the child’s neurological status and conscious level respectively.

- Withhold oral feeding (eating and drinking) until the swallow safety has been established.

- Maintain normal fluid, glucose and electrolyte balance.

- Target oxygen saturations above 92%.

- Treat hypotension.

- Consider the cause and necessity of treating hypertension in HS on a case-by-case basis.

- Children and young people with AIS should only receive blood pressure-lowering treatment in the following circumstances:
  - in patients who are otherwise eligible for intravenous (IV) thrombolysis but in whom systolic blood pressure exceeds 95th percentile for age by more than 15%
  - hypertensive encephalopathy
  - end organ damage or dysfunction, e.g. cardiac or renal failure.

- Parents/carers and young people should be actively involved in decision making. This may require modification of information to meet the communication needs of the individual child or young person, with the support of a speech therapist and/or occupational therapist.

- Maintain regular contact with parents/carers and young people from the time of presentation in order to explain investigations, processes and what to expect. Allow time for questions, and provide age-appropriate and multi-format information for the child or young person as well as the parent/carer.

- Consider using a named key worker for the family as a central point of contact for questions, updates and coordination of multidisciplinary care.
5.2 Framework for early functional assessment

In this section assessment soon after diagnosis by members of the broader multidisciplinary team (MDT) is referred to. Typically this assessment first takes place in the acute phase of recovery once the child or young person is medically stable.

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified four studies\textsuperscript{107-110} which reviewed the framework for early functional assessment in a child with stroke. Of the four identified studies, one was a retrospective review\textsuperscript{107}, one was a case series\textsuperscript{110}, one was a cohort study\textsuperscript{109} and one was a systematic review\textsuperscript{108}. The quality of evidence was moderate in two studies\textsuperscript{108,109} and low in two studies\textsuperscript{107,110}. Study populations were reported from a number of regions including Australia (two studies\textsuperscript{107,110}), Europe (one study\textsuperscript{109}) and the UK (one study\textsuperscript{108}).

While the review identified three\textsuperscript{107,109,110} studies that informed the recommendations, only one study\textsuperscript{108} assessed children who had experienced a stroke. As a result, studies including children with acquired brain injury and diseases which can result in similar functional impairments (e.g. cerebral palsy) were included.

Levels of acute speech, language and swallowing abilities were assessed\textsuperscript{110} using a number of standardised and non-standardised tools. These assessment tools included Ranchos Los Amigos Cognitive Scale, Verbal Motor Production Assessment of Children, Frenchay Dysarthria Assessment, Schedule of Oral Motor Assessment, Paramatta Hospitals Assessment of Dysphagia, and a non-standardised feeding trial. Impairments were identified in some children when using all of the tools, but no discussion on the usefulness or usability of the tools was included.

Gordon\textsuperscript{108} conducted a review of studies reporting functioning and disability outcomes of AIS. The ICF framework was used to highlight gaps in knowledge about the relationship between impairments in body structure and functions, limitations in activity and participation and the impact of environmental factors. The study highlighted the Pediatric Stroke Outcome Measure (PSOM) as the only tool found that was validated for use in the children with stroke.

Massaro and colleagues\textsuperscript{109} explored the use of three dedicated pain scales by caregivers and nursing staff in children with cognitive impairments and limited verbal communication. All pain scales involved behavioral observation. The scales varied in inter-rater reliability, and the need for professionals to consult caregivers when making judgments about children’s pain. This study found that the Non-Communicating Children’s Pain Checklist
(Post-operative Version (NCCPC-PV)) was the easiest pain assessment to use, appropriately rated the child’s pain, and recommended its use in clinical settings.

With regard to measuring the Assisting Hand Assessment (AHA) to determine its reliability in evaluating unilateral impairment, Davis and colleagues\textsuperscript{107} found good and excellent inter-rater reliability in most domains with the exception of the ‘Chooses Assisting Hand’ and ‘Moves Forearm’ domains. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Limited evidence was identified to help guide recommendations regarding children’s likely functional difficulties and intervention needs in the acute period following stroke. However, a small number of studies were found which identified impairments and assessment tools in the acute rehabilitation period in children with acquired brain injury/traumatic brain injury and other neurological disorders. Assessment tools have some relevance in highlighting the possible consequences of neurological impairment in AIS and HS, their intervention needs, and methods of assessment. Knowledge of functional skills can assist families and professionals in determining goals and outcome measures. The recommendations for this section were formed by taking into consideration the evidence identified and through discussion with the guideline development group (GDG) members.

As described in Section 1.1, the World Health Organization (WHO) ICF is the framework recommended throughout this guideline to inform habilitation and rehabilitation practice from early sub-acute assessment through to long-term management. The ICF may be applied to early functional assessment soon after diagnosis with a view to identifying impairments in body structures and functions, and activity limitations, and designing individualised interventions. In using the term ‘rehabilitation’ in this guideline, reference to the concepts of habilitation (support for ongoing acquisition of developmental and functional skills) and rehabilitation (recovery of skills) are made as both or either of these approaches may be relevant when working with a developing child or young person.

Acute paediatric services vary across the UK in terms of access to MDTs and allied health professionals. There is also variation in workforce capacity and competencies. Hospitals treating acute childhood stroke require a close collaborative approach to multidisciplinary communication and management, in collaboration with tertiary specialist centres, community child health and specialist rehabilitation providers.

The findings of the parent/carer and young person workshops suggest that the priorities around the time of diagnosis and in the early stages of care are centred on early and
regular contact with clinicians. Clinicians should communicate in a way that is family-centred, proactive and sensitive and provide time to ask questions. Care processes and reasons for assessment/investigations should be explained. A named key contact during hospital admission can help to keep families updated on what to expect, and the child/young person’s progress, on a regular basis. The possible impact of the stroke and needs including rehabilitation should be discussed as early as possible to aid preparation.

**Recommendations**

- Provide clinical assessment of a child’s body structures and functions and activities, by members of the relevant hospital multidisciplinary team (including occupational therapists, physiotherapists, speech and language therapists), as soon as possible during hospital admission (within 72 hours), with consideration of the child’s age and developmental abilities.

- Adoption of a collaborative approach to working with children, young people and families is important in supporting identification of priority areas for assessment and intervention.

- Involve the parents/carers, family and child/young person as key participants in the assessment of activities (e.g. mobilising, dressing, eating), and where clinically appropriate in the identification of early rehabilitation priorities.

- Parents/carers and young people should be regularly informed and updated throughout the care process. This should include age-appropriate and multi-format information for the parent/carer and child/young person about their condition/suspected condition, investigation plans and findings, and management plans including rehabilitation.

- Use the ICF framework to identify domains for assessment and intervention, including impairments in body structure and functions and activity limitations.

- Consider the use of both clinical and instrumental methods to assess body structures and function. Key areas to consider include:
  - swallow safety (ingestion)
  - hydration and nutrition
  - pain
  - motor function (muscle functions, movement functions)
  - vision (seeing) and hearing
- sleep
- sensation and perception
- fatigue

- Assess activity limitations, using clinical and instrumental methods as appropriate. Key areas to consider are:
  - mobility and gross motor activities (walking and moving, changing and maintaining body position)
  - eating and drinking (ingestion)
  - self-care (washing, dressing, toileting)
  - communication, including language understanding and expressive skills (receiving, producing and conversation)
  - social interaction (interpersonal interactions and relationships)
  - behaviour and emotion (general tasks and demands, including handling stress and other psychological demands, and managing one’s own behaviour)
  - cognition (learning and applying knowledge)
  - play and fine motor activities

- Assess the communication, information and support needs of the parents/carers, family and child/young person during early functional assessment.

- Explain the purpose of assessments to the parents/carers, family and child/young person.

- Consider the use of both functional and developmental assessments to describe and monitor any change in children’s abilities and limitations.

- Undertake at least weekly multidisciplinary review of abilities and rehabilitation needs during the inpatient stage.

- Initiate early liaison with community-based medical, nursing, occupational therapists, physiotherapists, psychologists, orthoptists, speech and language therapists and other allied health professionals to establish links with local networks.

- Consider the use of technology to support the exchange of information and maintenance of communication.
5.3 Prevention, identification and management of complications

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified four studies\textsuperscript{58,111-113} which discussed possible complications following AIS or HS and the methods used to detect and prevent or minimise them. Of these, one was a cohort study\textsuperscript{111}, one was an observational study\textsuperscript{112}, one was a retrospective case series\textsuperscript{113} and one was a case series\textsuperscript{58}. The quality of evidence was classified as moderate in three studies\textsuperscript{111-113} and low in one study\textsuperscript{58}.

Sample size ranged from 63 to 77 children, and study populations were reported from a number of regions including America (three studies\textsuperscript{111-113}) and Taiwan (one study\textsuperscript{58}).

Two studies identified seizures as a complication of both AIS and HS\textsuperscript{112,113} and reported that seizures and recurrent seizures did not vary by stroke type and were more likely in children and young people where there was cortical involvement in the stroke. Singh and colleagues\textsuperscript{112} also found that status epilepticus occurred in 17\% of patients, was more common in infants when there was cortical involvement, and did not vary by stroke subtype.

Another study\textsuperscript{111} of moderate quality found that larger stroke volume (greater than 5\%) as identified on brain imaging predicted a higher risk of haemorrhagic conversion.

Wang and colleagues\textsuperscript{58} reported that visual impairments, ataxia, and mental or psycho-motor retardation were possible complications following AIS and HS, however none of these occurred in more than a quarter of the included children and this small study only reported the numbers experiencing each complication without any analysis of the relative likelihood of each complication occurring.

Due to the lack of high quality studies examining complications other than seizures and haemorrhagic conversion, the following recommendations, except those for seizure and haemorrhagic conversion, are based on the consensus of the GDG members. Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

Due to a lack of published evidence, the majority of these recommendations were formed through discussion and consensus between the GDG members drawing on clinical experience, incorporating where appropriate recommendations for adult stroke care.
The GDG agreed that, similar to the recommendation presented in the Royal College of Physicians' (RCP) national clinical guideline for stroke\textsuperscript{12}, position or posture and early mobilisation should also apply to children and young people, and as such have formulated recommendations based on this. Considering the potential risk of recurrence, early mobilisation, particularly in first 24 hours, should only be carried out when the child or young person is medically stable and following discussion with medical and surgical teams in order to minimise risk, particularly in HS.

Clinical experience suggests that problems with tissue viability and pressure areas or ulcers are rare following childhood stroke; however, for dependent children, it is recommended to follow the National Institute for Health and Care Excellence (NICE) quality standard on pressure ulcers\textsuperscript{114} to ensure all children and young people have regular positional changes. In adult stroke patients, a six hour position change is recommended, and this should also apply as a maximum duration prior to reposition in children and young people.

**Recommendations**

- Be aware of the following possible complications after AIS/HS, as tabulated:

<table>
<thead>
<tr>
<th>Complication</th>
<th>Detection</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Swallow dysfunction</td>
<td>swallow safety for eating, drinking and saliva control should be assessed, ideally by a trained professional using validated screening methods. Monitor for aspiration</td>
<td>refer to speech and language therapist if concern about impaired swallow safety</td>
</tr>
<tr>
<td>Aspiration</td>
<td>if aspiration is considered likely undertake clinical assessment, monitor oxygen saturations, blood gases and obtain chest X ray</td>
<td>consider the placement of a nasogastric tube</td>
</tr>
<tr>
<td></td>
<td></td>
<td>facilitate regular position changes in bed and early mobilisation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>ongoing management of eating, drinking and swallow</td>
</tr>
<tr>
<td>Raised intracranial pressure</td>
<td>provide regular clinical assessment sensitive to</td>
<td>follow national and local guidelines for</td>
</tr>
</tbody>
</table>
| the detection of raised intracranial pressure (ICP); this should include monitoring of conscious level, pupil size and response, blood pressure and heart rate  
• hypertension and bradycardia reduction in conscious level are ominous, even if transient  
• consider ICP monitoring | management of acutely raised intracranial pressure |
|---|---|
| Hydrocephalus • suggestive symptoms or signs include accelerated rate of head growth in infants/babies, vomiting, declining conscious level, impaired upgaze, abducens palsy, bradycardia and hypertension  
• when hydrocephalus is suspected urgent brain imaging is mandatory; cranial ultrasound may be acceptable when the anterior fontanelle remains patent, in all other children computerised tomography (CT) or magnetic resonance imaging (MRI) will be required | urgent referral to neurosurgical unit |
| Seizures • assess clinically rather than routinely with electroencephalogram (EEG) except when | seizures should be managed according to local and national treatment protocols |
Clinical assessment is not possible in paralysed and sedated patients or when distinction between seizure and movement disorder is not possible on clinical grounds.

- New onset seizures may signal further stroke or haemorrhagic conversion and repeat brain imaging should be considered.
- There is no evidence to support the use of prophylactic anticonvulsant medication.

### Endocrine derangement
- Serum electrolytes (sodium and glucose) should be measured at presentation with the frequency of repeat measurements determined individually.
- Maintain sodium and glucose levels within normal range.
- Administer 3mls/kg 3% NaCl if the sodium is less than 125mmol/L.

### Deranged coagulation
- Monitor coagulation and platelet count.
- Correct abnormalities in discussion with haematology.

### Nutrition
- Monitor weight.
- All children admitted to a hospital setting require a nutritional assessment, monitoring of weight, and referral to a paediatric dietitian.

### Deep vein thrombosis
- Risk assessment including level of mobility, weight, and family history should be considered.
- Pneumatic compression boots should be considered.
- Do not routinely administer prophylactic low molecular weight heparin.
- Early mobilisation should be encouraged.
6. **Arterial Ischaemic Stroke**

6.1. **Conditions and factors associated with a risk of AIS or recurrence**

Stroke is an increasingly recognised cause of significant morbidity in childhood and beyond and has widespread personal, familial, economic, and social consequences. Previous studies have identified a wide range of associated factors, commonly termed as risk factors, although, with the exception of antecedent varicella infection, lack of case-control data means that these are presumptive rather than definite risk factors\(^{115,116}\).

The current literature suggests that risk factors for childhood stroke are distinct from those in adults\(^{117}\), and it is important to know if childhood stroke has a similar risk factor profile across the world and to also understand the potential and limitations of international collaborative studies. Neonatal stroke has distinct characteristics, but the variability in risk factors across age groups beyond the neonatal period is not clear.

In arterial ischaemic stroke (AIS) the risk factors that predominate in children and young people are distinct from adults. These include arteriopathies such as moyamoya, cardiac disorders such as congenital heart disease, haemoglobinopathies such as sickle cell disease, chronic systemic conditions, infections, acute and chronic head and neck disorders (including trauma leading to extra-cranial arterial dissection), acute systemic conditions, and prothrombotic states (PTS). Of note, atheroma, the most common risk factor for adult AIS is not implicated in paediatric AIS, which reinforce the need to have different approaches between adults and children. It is important to appreciate that risk factors commonly co-exist in individuals, modifying recurrence risk and mandating a comprehensive approach to investigation.

The question of recurrence after AIS is a major concern to parents/carers and its prevention is a major focus of medical management. The risk of recurrence will depend on the risk factors identified and secondary prevention will be directed by these.

**Review questions**

- In childhood, which conditions/factors are associated with higher risk of development of AIS?
- In childhood, which conditions/factors are associated with higher risk of recurrence of AIS?
Further investigations & follow up imaging

- What are the most appropriate investigations to identify underlying risk factors in AIS?
- What are the role, modality and timing of brain imaging in assessment and follow-up of childhood AIS?

Acute medical interventions

- What is the safety and efficacy of thrombolytic agents/anticoagulants/antiplatelet agents for the acute treatment of children and young people with AIS?
- What is the appropriate management for AIS in children and young people with sickle cell disease?

Medical interventions to prevent recurrence

- What is the safety and efficacy of medical interventions to prevent recurrence of:
  - AIS
  - AIS in sickle cell disease (SCD)
  - progression of silent cerebral infarctions (SCI) in SCD

Surgical & endovascular interventions

- In children and young people with acute/chronic AIS, what are the indications for referral to neurosurgery?
- What is the safety and efficacy of surgical interventions in the treatment of acute AIS in children and young people?
- What is effectiveness of surgical interventions in the prevention of recurrence of AIS in children and young people?
- In children and young people with acute/chronic AIS, what are the indications for referral to interventional neuroradiology?

6.1.1. Risk factors for AIS and recurrent AIS

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified 42 studies, which reported data on the risk factors for AIS and recurrent AIS in children and young people. Of the 42 studies, two were systematic reviews, four cohort studies, 22 case-control studies, and 14 case series. The quality of evidence was classified as high in five studies, moderate in 21 studies, and low.
in 16 studies\textsuperscript{123,124,127,128,130,132,138,139,141-144,146,147,150,155}. Sample size ranged from 10 to 3.25 million children.

Study populations were reported from a number of regions including America (14 studies\textsuperscript{115,120-122,125-129,132,133,136,138,156}), Europe (15 studies\textsuperscript{123,124,135,139-143,145-147,151,153-155}) and the UK (five studies\textsuperscript{2,48,119,130,148}). For the studies where applicability to the UK population was less certain\textsuperscript{118,131,134,137,144,149,150,152}, the results were approached with caution.

Blood disorders including thrombophilia and sickle cell disease, and infection were the subject of the greatest number of studies\textsuperscript{48,115,118-121,124,128-130,134-140,143,144,147,149-151,153}. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Most studies were retrospective in design and thus identified risk factors for AIS in children and young people who had already experienced AIS. There was little attempt to estimate the risk of having an AIS if the individual has been diagnosed with one of the risk factors. It therefore remains undetermined as to which of the risk factors is the most significant, and as such the list of the risk factors which increase the risk of AIS in children and young people has been compiled from the findings of the included studies; this is not indicative of the magnitude of the increase in risk. As well as risk factors identified from the literature review, the guideline development group (GDG) recommended consideration of other factors that are important in clinical practice but that do not widely feature in the literature.

In the formation of these recommendations, the GDG considered the characteristics of the children described in the studies, including age at the time of AIS, presenting symptoms, lesion location and investigation findings.

A particularly contentious area with regard to AIS risk factors was genetic thrombophilia, of which there are many studies often with differing findings. Some studies do indeed report an increased incidence of thrombophilia associated with AIS, and hence inclusion of thrombophilia in the recommendations. Moreover, much of the childhood AIS literature relates to non-UK cohorts, with a reduced applicability of this data to a UK population.

Current clinical practice in the UK for genetic thrombophilia testing varies widely, both between centres and groups of healthcare professionals. Of note, the GDG and expert panel were unable to reach consensus on the clinical necessity for genetic thrombophilia testing in a child with AIS. Testing is expensive and identification of genetic thrombophilia and may have implications for other family members. The clinical relevance of genetic thrombophilia in the genetics of stroke remains contentious and does not mandate altered
management, as such identification may generate disproportionate concern. In the absence of consensus, this area remains open for individual clinical discretion.

While no evidence was identified on the importance of Fabry disease in childhood AIS, the GDG agreed this as is a treatable condition and implicated in young adult AIS. It should therefore be considered in children, pending better evidence.

Feedback from parent/carers and young people revealed the importance of early individualised information, provided in direct conversation and supported where possible in multiple format for later reflection; such as web-based, written and graphic formats.

**Recommendations**

**Risk factors for first AIS**

<table>
<thead>
<tr>
<th>Risk Category</th>
<th>Included factors/diagnoses</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arteriopathy</td>
<td>• focal cerebral arteriopathy of childhood</td>
</tr>
<tr>
<td></td>
<td>• moyamoya</td>
</tr>
<tr>
<td></td>
<td>• arterial dissection</td>
</tr>
<tr>
<td></td>
<td>• central nervous system (CNS) vasculitis</td>
</tr>
<tr>
<td>Cardiac disease</td>
<td>• congenital cardiac disease</td>
</tr>
<tr>
<td></td>
<td>• additional risk factors in children and young people with cardiac disease: Right to Left shunt, increased, Lipoprotein(a) (Lp(a)), anticardiolipin antibody (ACLA), combined prothrombotic disorders</td>
</tr>
<tr>
<td>Cardiac surgery/interventions</td>
<td></td>
</tr>
<tr>
<td>Sickle Cell Disease</td>
<td>Additional factors in children and young people with SCD:</td>
</tr>
<tr>
<td></td>
<td>• genotype (sickle haemoglobin (HbS) &amp; HbSβ thalassaemia more than other genotypes)</td>
</tr>
<tr>
<td></td>
<td>• abnormal transcranial Doppler studies</td>
</tr>
<tr>
<td></td>
<td>• arteriopathy (intracranial &amp; extracranial)</td>
</tr>
<tr>
<td></td>
<td>• absence of alpha thalassaemia trait</td>
</tr>
<tr>
<td></td>
<td>• acute anaemia</td>
</tr>
<tr>
<td></td>
<td>• silent infarction</td>
</tr>
</tbody>
</table>
| Infection | • prior transient ischaemic attack (TIA)  
| • high systolic blood pressure, acute chest syndrome  
| • anaemia, high reticulocyte count  
| | • varicella zoster  
| • upper respiratory tract infections  
| • multiple infections  
| Gender/Ethnicity | • black ethnicity  
| • Asian ethnicity  
| • male gender  
| Thrombophilia | • genetic: Factor V Leiden (FVL), PT20210, MTHFR c677T, protein C deficiency, increased lipoprotein(a) (Lp(a)), more than 2 genetic thrombophilia traits, high homocystinuria (HCY)  
| • acquired: antiphospholipid syndrome (APLS)  
| Miscellaneous | • iron deficiency anaemia  
| • radiotherapy  
| • high alpha 1 antitrypsin (AT), trauma  
| • under-vaccination  
| • multiple risk factors  

- Consider the following conditions which are linked with childhood AIS and may be clinically important in relevant cohorts (although have not been scrutinised in case-control analyses):
  - trisomy 21
  - neurofibromatosis
  - malignancy and long-term effects of treatment for malignancy (especially cranial radiotherapy)
  - auto-immune diseases, e.g. systemic lupus erythematosus
  - illicit drugs and other recreational drugs (e.g. cocaine)

- The importance of Fabry disease in children and young people has not been investigated but is treatable and implicated in young adults and until this is resolved should be considered in the work-up.

- Take these factors into account when considering a need for counselling in high-risk groups.

- Information on risk factors should be delivered in face-to-face conversation
with parents/carers and young people (where appropriate) and supported
where possible with web-based or written materials for later reference. The
information provided should be age-appropriate and multi-format, and relevant
to the child or young person as well as the parent/carer.

Risk factors for recurrent AIS

- Be aware of increased risk of recurrence in children and young people with AIS
  and the following risk factors:
  - arteriopathy (especially if progressive on interval imaging)
  - moyamoya
  - arteriopathy in sickle cell disease
  - congenital heart disease (especially if either infection was present at
    sentinel stroke or there is a thrombotic state)
  - thrombophilia (e.g. homozygosity for MTHFR mutation, protein C and/or
    protein S deficiency)
  - low birthweight

- Take these factors into account when considering a need for counselling in high
  risk groups.

- Information on risk of recurrence and how to minimise risk should be delivered
  in face to face conversation with parents/carers and young people (where
  appropriate) and supported where possible with web-based or written
  materials for later reference. The information provided should be age-
  appropriate and multi-format and relevant to the child or young person as well
  as the parent/carer.

6.1.2. Investigations to identify risk factors in AIS

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified ten
studies, which reported data on investigations to identify risk factors in AIS. Of
the ten studies, one was a cohort study, two were case-control studies and seven
were case series designs. The quality of evidence was classified as high in
two studies, moderate in two studies, and low in six studies. Sample
sizes ranged from 29 to 144 children.
Study populations were reported from a number of regions including America (two studies\textsuperscript{18,160}), the Middle East (two studies\textsuperscript{21,158}), Australia (two studies\textsuperscript{94,161}), Korea (one study\textsuperscript{159}), Europe (two studies\textsuperscript{33,157}), and the UK (one study\textsuperscript{162}).

Tests used in studies to identify underlying risk factors included imaging such as computerised tomography (CT) and magnetic resonance imaging (MRI)/magnetic resonance angiogram (MRA)\textsuperscript{21,159,162}, blood tests such as lipid and glucose levels, thrombophilia markers, plasma total homocysteine, D-dimer levels, immunologic profiles\textsuperscript{18,21,33,157,158,160,161}, and blood pressure\textsuperscript{94}. These studies presented little evidence to support the use of many of the tests and no data was presented on sensitivity/specificity of the imaging modalities.

**Linking the evidence to the recommendations**

Due to the lack of informative literature, the recommendations are based on the clinical experience of the members of the GDG.

Although Right to Left (R to L) intracardiac shunts are associated with AIS in young people, their role in the aetiology of childhood AIS is unclear; however, clinical experience suggests that they may be relevant in a minority of cases. The necessity for a bubble study in addition to a standard echocardiogram should be discussed with the cardiology team in light of the patient’s clinical and radiological presentation. If a R to L intracardiac shunt is detected, further management should be decided on a case-by-case basis and should involve discussion between the treating neurologist and cardiologist. Factors to consider are stroke subtype, other risk factors and features of the R to L shunt.

**Heritable thrombophilia**

There are numerous case series which demonstrate an increased prevalence of the heritable thrombophilia mutations (protein C and S deficiency, antithrombin deficiency, Factor V Leiden, the prothrombin gene mutation and the MTHFR mutation) in children with AIS\textsuperscript{48,140,143,144,146-154}, but there is no evidence to support the hypothesis that the presence of any of these mutations has an impact on morbidity, mortality or long-term outcome.

In addition, there is some discrepancy between the results of different case series which may reflect the low frequency of some of these mutations. In adult thrombosis and stroke, it is now recognised that the MTHFR mutation is not clinically relevant and that previous studies suggesting it was significant were flawed due to the background rate of heterozygosity in the European population being 45%.
Due to the lack of evidence to suggest that the results of tests for thrombophilia mutations impact on acute care for AIS, some members of the GDG felt it should be recommended that they are not to be carried out routinely. It was recognised, however, that this recommendation would be a departure from current practice and so without published evidence to support this and without consensus within the GDG, a statement was added to the Delphi survey to assess expert opinion on the matter. When this statement was presented to the Delphi panel, consensus could not be reached and therefore consensus was sought on whether the results of these tests affect the way in which a child with AIS is managed without a directive on whether they should be carried out or not. The Delphi panel agreed (83%) that the results of these tests did not affect management.

In light of the result of the Delphi survey the GDG decided that testing for heritable thrombophilia mutations would not be routinely recommended, but would remain a matter for individual clinician discretion. The rationale for not recommending testing all children and young people with AIS for heritable thrombophilia included:

- The result is very unlikely to change initial management nor will it influence outcome.
- There remains uncertainty about the meaning of some of the tests, especially MTHFR mutation, and they are not routinely recommended in any adult thrombotic setting including AIS.
- The majority of acute hospitals will give an adult normal range for the protein C, S and AT which is likely to be misleading in younger children.
- It is possible that the results of protein C, S and AT at the time of AIS will be unreliable, as acute illness can alter the results. As a consequence it is recommended that testing for heritable thrombophilia in adults, where appropriate, is carried out at a distance from the acute event, in order to avoid false positive and negative results.
- Given the gene frequency of some of the heritable thrombophilia mutations (1:20 of the Northern European population are heterozygous for Factor V Leiden and around 1:100 for the prothrombin gene mutation), it is inevitable that positive results will arise frequently and lead to requests for family screening and the possibility of disproportionate anxiety for families where another child carries one of these very common mutations.

The recommendations are not related to heritable thrombophilia mutations testing and were formed through discussion of the evidence from the literature review and consensus within the GDG.
Lumbar puncture (LP)
The role of LP in the evaluation of children with AIS is contentious. Bedside clinical assessment normally guides whether active CNS infection (meningitis/encephalitis) needs to be considered and when assessing safety of undertaking a LP in an acutely sick child, particularly in light of the potential for brain swelling after AIS. Whilst clinical experience suggests that in some cases it may be useful to assess other biomarkers of infection (e.g. Varicella zoster virus (VZV) antibody ratio in blood:cerebrospinal fluid), there is currently no evidence to guide at which point in time this should be undertaken, nor how cases with a positive result should be managed. Many children and young people with stroke would already have been started on anti-thrombotic therapy, and in such instances, the risk:benefit ratio of LP (or stopping antithrombotic therapy to enable LP) is debatable. Whilst the GDG were unable to provide clear guidance on the role of LP in investigating AIS, it was felt that this could be considered by the bedside clinician in selected cases, perhaps during the subacute period, and in children or young people in whom there was no clinical contra-indications.

Recommendations

- Carry out the following investigations in children and young people with a diagnosis of AIS:
  - haematological investigations, including full blood count, iron status (e.g. iron, ferritin, total iron binding capacity) and haemoglobinopathy screen
  - biochemistry tests, including total plasma homocysteine, alpha galactosidase, fasting blood sugar, fasting cholesterol, and Lipoprotein(a)
  - lupus anticoagulant and ACLA, and discuss beta 2GP1 testing with haematology if necessary
  - cardiac evaluation: electrocardiogram (ECG), echocardiogram (to identify structural lesions and R to L shunts)
  - cerebrovascular imaging from the aortic arch to vertex, with computed tomography angiography (CTA) or magnetic resonance angiogram (MRA) at the time of CT or MRI respectively
  - transcranial Doppler in patients with SCD

- Clinically evaluate all patients for history of prior infection (especially VZV), immunisation, dysmorphic features, neurocutaneous stigmata, autoimmune disease and evidence of vascular disease in other organ systems.
6.1.3. Follow-up imaging in AIS (role, timing, investigations)

Evidence summary

Whilst there are a number of studies addressing the issue of acute AIS imaging, only ten described strategies for assessment of AIS or follow-up imaging\(^78,84,93,163-169\). The quality of evidence was classified as high in two studies\(^{165,168}\), moderate in two studies\(^{93,163}\), and low in six studies\(^{78,84,164,166,167,169}\). Studies were reported from a number of regions including America (two studies\(^{93,163}\)), the UK (three studies\(^{164,165,168}\)), Europe (four studies\(^{78,84,166,169}\)), and Australia (one study\(^{167}\)).

Two studies\(^{93,163}\) described methods to estimate the volume of an infarction. Beslow\(^93\) used the modified Alberta Stroke Programme Early CT Score to establish infarct volume, modified Alberta Stroke Program Early CT Score (ModASPECTS) of more than or equal to five had sensitivity of 80% and specificity of 87% for predicting large infarcts which are increased risk of haemorrhagic transformation. Filippi\(^{163}\) compared automated volume assessment to manual volume assessment and found that automated was equivalent to manual but significantly faster.

There is a clinically important rate of clinically silent radiological AIS recurrence\(^{165}\), highlighting the importance of surveillance imaging to identify this and any modifiable risk factors. Three studies\(^{84,167,168}\) discussed the accuracy of MRA in comparison to other imaging modalities. MRA was found to overestimate the degree of stenosis\(^{84}\), but this is not currently relevant in children. Catheter angiography (CA) could provide more information over MRA and could identify abnormalities that were not detected by MRA and which could mandate a change in management\(^{167}\). A small subset of patients had no evidence of arteriopathy, even when they experience recurrent AIS\(^{168}\).

Although a majority of the studies found discussed the different imaging modalities, the merits of one over the others was rarely done comprehensively.

Linking the evidence to the recommendations

Due to the lack of systematic and comprehensive comparisons of different imaging modalities, and the lack of information around the timing and frequency of imaging, the evidence found in the literature review served as a starting point. The recommendations are also based on the clinical experience of and consensus among the GDG members. These recommendations are based on clinical rather than resource considerations.
Recommendations

- Be aware that MRI is the modality of choice for follow-up imaging of children and young people with AIS as it provides the best assessment of the extent of any permanent structural damage and of the cerebral circulation without using ionising radiation.

- Consider the clinical circumstances and the presence of conditions predisposing to recurrence (e.g. moyamoya or other arteriopathy) when considering the frequency and duration of follow-up imaging in childhood AIS.

- CA should be undertaken in children and young people with occlusive arteriopathy, who are being considered for revascularisation; if surgery is undertaken CA should be repeated a year after surgery.

6.2. Medical and surgical interventions

The literature related to the treatment of paediatric stroke is characterised by a lack of randomised controlled trials and studies with small sample sizes. It is important to distinguish between treatment (i.e. reverse or limit brain injury) and secondary prevention (prevent another stroke). Trial evidence has shown that hyperacute thrombolysis and mechanical thrombectomy/clot retrieval improve outcomes in adult stroke; while intravenous (IV) thrombolysis is now widely available, the practicalities of implementing of recanalisation therapies are currently under consideration in the UK. The Thrombolysis in Paediatric Stroke study (TIPS) was designed to assess the efficacy and safety of hyperacute thrombolysis in the paediatric population had to be abandoned due to lack of patient recruitment. In spite of difficulties in diagnosis in time for acute interventions, it is recognised that some children and young people do benefit from thrombolysis and possibly recanalisation therapies.

There is a small body of low quality evidence which shows that small numbers of children and young people, particularly adolescents, are already being treated; however, due to the methodological shortcomings of this research, recommendations could not be formed based on its conclusions. As such a Delphi consensus method was used to arrive at the recommendations that follow.

Prompt identification and treatment of complications, e.g. raised intracranial pressure, also limit brain injury and the importance of managing homeostasis cannot be over-emphasised. Most of the recommendations in this chapter are based on expert consensus gained through GDG discussions and the Delphi consensus method.
Anti-thrombotic treatment has been recommended in previous guidelines for acute AIS treatment in children and young people. The recommendations that follow incorporate updated data on safety and efficacy of these treatments.

There is a lack of evidence regarding the use of decompressive craniectomy for large infarcts in children and young people. The recommendations that follow are based on the evidence from adult data and on the clinical experience of the GDG.

Transfusion therapy is well established in the acute management of stroke related to sickle cell disease but whether exchange transfusion is better than simple top-up transfusion is not established. Recommendations have been made after assessing the limited evidence.

The parents/carers and young people workshops raised the importance of families being informed in a timely way about the reasoning for interventions, what to expect, having the opportunity to exercise choice where possible, and ask questions. A named key contact is a helpful support to families in maintaining contact with the clinicians and being updated regularly.

**Review questions**

**Acute Medical interventions**

- What is the safety and efficacy of thrombolytic agents/anticoagulants/antiplatelet agents for the acute treatment of children and young people with AIS?
- What is the appropriate management for AIS in children and young people with sickle cell disease?

**Medical Interventions to prevent recurrence**

- What is the safety and efficacy of medical interventions to prevent recurrence of:
  - AIS
  - AIS in SCD
  - progression of SCI in SCD

**Surgical & endovascular interventions**

- In children and young people with acute/chronic AIS, what are the indications for referral to neurosurgery?
- What is the safety and efficacy of surgical interventions in the treatment of acute AIS in children and young people?
- What is effectiveness of surgical interventions in the prevention of recurrence of AIS in children and young people?
In children and young people with acute/chronic AIS, what are the indications for referral to interventional neuroradiology?

6.2.1. Acute medical interventions for AIS

Evidence summary

Use of thrombolysis or anti-thrombotic therapy

A systematic review was conducted for the relevant clinical questions and identified 18 studies. All were non-experimental and the quality of evidence was classified as high in two studies, moderate in five studies, and low in 11 studies. Study populations were reported from a number of regions including America (13 studies), India (one study), Egypt (one study), and Europe (three studies). These studies provided some evidence on the safety of various antithrombotic agents but no clear evidence on dose or effectiveness.

Antithrombotic therapies included unfractionated heparin, low molecular weight heparin, aspirin, and clopidogrel. Clopidogrel was used in three studies either on its own or in combination with aspirin and on occasions when aspirin was contraindicated. Rates of symptomatic intracranial haemorrhage were low with anticoagulation therapy and one moderate quality prospective study reported a symptomatic intracranial haemorrhage rate of 4%. In general, rates of complications were low following antithrombotic, anticoagulant and antiplatelet treatments.

Two studies which reported the use of acute thrombolysis were identified in the systematic review. All were non-experimental and had major limitations. No conclusions can be drawn from these studies on the safety and efficacy of hyperacute thrombolysis in children and young people with AIS.

One study with a retrospective analysis of a hospital inpatient database confirmed a trend of increasing use of thrombolysis but results on safety were conflicting.

Acute AIS treatment in children and young people with SCD

There are no trials examining acute management of AIS in sickle cell disease. There is one study comparing exchange to top-up transfusion demonstrating superiority of exchange transfusion. Further details on each study are provided within the evidence tables, in Appendix 4c.
Linking the evidence to the recommendations

Use of thrombolysis or anti-thrombotic therapy

Antithrombotic therapy appears safe in the acute treatment of AIS in a number of different settings but there is no evidence about whether this limits brain injury or is associated with better outcomes (as opposed to secondary prevention). It would, therefore, be appropriate to commence antithrombotic treatment at presentation of AIS in the absence of contraindications. There is insufficient evidence to recommend antiplatelets (aspirin and clopidogrel) and anticoagulants (unfractionated heparin, low molecular weight heparin (LMWH) as there is no comparative data. Use of aspirin would be least expensive, would require least monitoring, and there is extensive clinical experience of safety and tolerability.

Although some studies reported the use of thrombolysis in paediatric AIS, these did not provide evidence of sufficient quality on which to base recommendations. As it is unlikely that randomised control trial (RCT) data will be available to make any recommendations for or against thrombolysis (IV or mechanical) in the near future, a consensus statement using a Delphi consensus method was used to provide guidance on the use of hyperacute thrombolysis (see Appendix 5).

Acute AIS treatment in children and young people with SCD

No RCT or large studies assessing the acute management of stroke in SCD were identified by the systematic review; therefore, the following recommendations are based on the consensus of the GDG.

Acute management is focused on reducing the percentage of HbS in the blood and correcting anaemia. Exchange transfusion is usually necessary to achieve this, unless the patient is severely anaemic, when a simple transfusion can be adequate.

For the majority of patients with sickle cell disease an acute stroke, whether it is haemorrhagic or ischaemic, is likely to represent an exacerbation of cerebral vasculopathy secondary to sickle cell disease. It is standard practice, therefore, to decrease the percentage of sickle haemoglobin to less than 30% and keep the haemoglobin at a level that will provide adequate cerebral oxygenation without the increased viscosity seen at higher levels of haemoglobin i.e. haemoglobin 100 to 110g/l. There are no studies in adults or children looking at other thrombolysis, antiplatelet agents or anticoagulation, and these are not routinely used because the underlying vasculopathy may increase the risk of bleeding. However, there may be specific circumstances in individual cases that make these treatments appropriate. The choice of different exchange transfusion modalities will likely be based on resources and venous access. However, whichever is chosen, volume
shifts are likely to be harmful and therefore isovolaemic procedures should be chosen. If there is likely to be a long-delay before exchange transfusion can be completed, it may be appropriate to consider a simple transfusion to correct anaemia whilst this is being organised.

It is important to work closely with the blood bank to ensure that appropriate blood is available as soon as possible. The senior biomedical scientist in the blood bank should be contacted by the haematology team as soon as a stroke is suspected. They should be given the full transfusion history and asked to order 80ml/kg of packed red cells that are ABO, Rh and Kell compatible.

Two vascular access points need to be established for an exchange procedure, one for the blood to go ‘IN’ and one for it to go ‘OUT’. An arterial line can be used in a manual exchange as the ‘OUT’ access with appropriate support and monitoring. The red cell exchange can happen on a paediatric ward with the appropriate support, or the patient may be transferred to a high-dependency unit, depending on their clinical state.

**Recommendations**

**Use of thrombolysis or anti-thrombotic therapy**

- Prescribe and deliver 5mg/kg of aspirin up to a maximum of 300mg within 24 hours of diagnosis of AIS in the absence of contraindications (e.g. parenchymal haemorrhage). After 14 days reduce dose of aspirin to 1mg/kg to a max of 75mg.

- Delay administering aspirin for 24 hours in patients where thrombolysis has been given.

- Aspirin should not be routinely given to children and young people with SCD presenting with AIS.

- In children and young people with cardiac disease presenting with AIS, make a multidisciplinary decision (including haematologists, paediatric neurologists and cardiologists) regarding the optimal antithrombotic therapy (antiplatelet versus anticoagulation) with assessment of the risk-benefit in individual cases.

- The off label use of tissue plasminogen activator (tPA) *could* be considered in children presenting with AIS who are more than eight years of age and *may* be considered for children aged between two and eight years of age on a case-by-case basis. 
case basis when the following criteria have been met:

- AIS has occurred as defined by:
  - an acute focal neurological deficit consistent with arterial ischaemia **AND**
  - Paediatric National Institute of Health Stroke Scale (PedNIHSS) more than or equal to 4 and less than or equal to 24 **AND**
  - treatment can be administered within 4.5 hours of known onset of symptoms

  - **AND** intracranial haemorrhage has been excluded:
    - CT and CTA demonstrates normal brain parenchyma or minimal early ischaemic change **AND** CTA demonstrates partial or complete occlusion of the intracranial artery corresponding to clinical or radiological deficit
    - OR MRI and MRA showing evidence of acute ischaemia on diffusion weighted imaging plus partial or complete occlusion of the intracranial artery corresponding to clinical or radiological deficit
    - **PROVIDING** that there are no contraindications [DELPHI]

• Begin thrombolysis irrespective of patient location at the point of AIS diagnosis and when above criteria are fulfilled; this will usually be in the secondary receiving centre emergency department or paediatric ward.

### Acute AIS treatment in children and young people with SCD

• Treat children and young people with SCD and acute neurological signs or symptoms urgently with a blood transfusion, to reduce the HbS to less than 30%, and increase the haemoglobin concentration to more than 100–110g/l. This will usually require exchange transfusion.

• Provide a small top up transfusion to bring Hb to 100g/l to improve cerebral oxygenation if the start of the exchange is likely to be delayed by more than six hours.

• Provide other standard supportive stroke care.
Prioritise exchange transfusion over thrombolysis.

Additional information

This section provides additional information to the recommendations for the use of thrombolysis or anti-thrombotic therapy.

As previously stated, the TIPS trial of hyperacute IV tPA in children closed due to non-recruitment and is therefore unlikely that a robust evidence base establishing efficacy of this treatment will become available. The role of IV therapy is a major area of concern for the clinical care of children with AIS and therefore the GDG felt it was important to make a recommendation around this, with ratification via Delphi consensus process (see Section 2.9). The GDG felt it reasonable to base the following contraindications to thrombolysis on the exclusion criteria which is specified in the protocol of the TIPS study⁷.

Contraindications to thrombolysis

Exclusion criteria for IV tPA (based on the Thrombolysis in Pediatric Stroke Study⁷, for further information see http://stroke.ahajournals.org/content/46/3/880/tab-figures-data).

- Unknown time of symptoms onset
- Pregnancy
- Clinical presentation suggestive of subarachnoid haemorrhage (SAH), even if brain imaging is negative for blood
- Patient who would decline blood transfusion if indicated
- History of prior intracranial haemorrhage
- Known cerebral arterial venous malformation, aneurysm or neoplasm
- Persistent systolic blood pressure more than 15% above the 95th percentile for age while sitting or supine
- Glucose less than 2.78mmol/L or more than 22.22mmol/L
- Bleeding diathesis including platelets less than 100 000, prothrombin time (PT) more than15s (international normalised ratio (INR) more than 1.4), or elevated activated partial thromboplastin time (aPTT) more than upper limits of the normal range
- Clinical presentation consistent with acute myocardial infarction (MI) or post-MI pericarditis that requires evaluation by cardiology before treatment
- Prior stroke, major head trauma, or intracranial surgery within the past three months
- Major surgery or parenchymal biopsy within 10 days (relative contraindication)
- Gastrointestinal or urinary bleeding within 21 days (relative contraindication)
• Arterial puncture at non-compressible site or LP within seven days (relative contraindication). Patients who have had a cardiac catheterization via a compressible artery are not excluded
• Patient with malignancy or within one month of completion of treatment for cancer
• Patients with an underlying significant bleeding disorder. Patients with a mild platelet dysfunction, mild von Willebrand disease, or other mild bleeding disorders are not excluded
• Stroke related exclusion criteria:
  - Mild deficit (Paediatric National Institute of Health Stroke Scale (PedNIHSS) less than 4) at start of tPA infusion or at time of sedation for neuroimaging, if applicable
  - Severe deficit suggesting large territory stroke, with pre-tPA PedNIHSS more than 24, regardless of the infarct volume seen on neuroimaging
  - Stroke suspected to be because of subacute bacterial endocarditis, moyamoya, sickle cell disease, meningitis, bone marrow, air, or fat embolism
  - Previously diagnosed primary angiitis of the central nervous system (PACNS) or secondary central nervous system (CNS) vasculitis. Focal cerebral arteriopathy of childhood is not a contraindication
• Neuroimaging related exclusions:
  - Intracranial haemorrhage on pre-treatment head CT and MRI
  - Intracranial dissection (defined as at or distal to the ophthalmic artery)
  - Large infarct volume, defined by the finding of acute infarct on MRI involving one-third or more of the complete middle cerebral artery (MCA) territory involvement
• Drug-related exclusions:
  - Known allergy to recombinant tissue plasminogen activator
  - Patient who received heparin within four hours must have activated partial thromboplastin time (aPTT) in normal range
  - Low molecular-weight heparin (LMWH) within past 24 hours (aPTT and INR will not reflect LMWH effect)

6.2.2. Interventions to prevent recurrence of AIS

Evidence summary

Medical interventions to prevent recurrence of AIS

A systematic review was conducted for the relevant clinical questions and identified two studies that discussed the prevention of AIS recurrence on children other than those with SCD. The first study by Ganesan and colleagues (2006) was a retrospective case series which examined the rates and risk factors for clinical and radiological recurrence of
AIS. The children included in this study suffered a range of vascular abnormalities for which many were receiving prophylactic treatment, however, the efficacy of specific treatments was not evaluated compared with no treatment. The second study by Pandey and colleagues was a case series carried out in America. This study described 42 children with craniocervical arterial dissection, two of which experienced a recurrence while on antiplatelet and anticoagulation therapy.

These studies were relatively small and non-experimental and as such the recommendations were based on the clinical experience of the GDG.

**Medical interventions to prevent recurrence of AIS in SCD**

The systematic review further identified 10 studies which discussed medical interventions to prevent recurrence of AIS in SCD. The quality of evidence was classified as high in one study, moderate in three studies, and low in six studies. Study populations were reported from a number of regions including America (seven studies), Europe (one study), and the rest of the world (two studies).

There are no RCTs directly comparing observation/placebo and regular blood transfusion in secondary prevention of AIS in SCD, although one RCT showed blood transfusion to be superior to hydroxycarbamide (‘hydroxyurea’). Two small, retrospective studies showed progressive cerebrovascular disease in the majority of children despite transfusion. One small, retrospective study suggested that recurrent strokes were more common in those treated with top-up transfusion versus exchange transfusion.

The only primary data came from the Stroke with Transfusions Changing to hydroxycarbamide (SWiTCH) study, and hydroxycarbamide+chelation for secondary stroke prevention. This study was stopped early because no differences in liver irons were reported; however, there were 7/67 strokes in the hydroxycarbamide arm and 0/66 strokes in transfusion arm. One systematic review explored the Stroke Prevention Trial in Sickle Cell Anemia (STOP) Trials and the SWiTCH trial together concluded that blood transfusions significantly reduced the risk of recurrent stroke in children and that this effect was greater than that of hydroxycarbamide. Another four small, uncontrolled, non-randomised pilot and cohort studies of hydroxycarbamide as secondary stroke prevention suggest that hydroxycarbamide is similar in efficacy to blood transfusion and better than nothing.

One uncontrolled study reported on the stabilisation of cerebrovascular disease in children with SCD following haematopoietic stem cell transplantation (HSCT) from an human...
leukocyte antigen (HLA) identical sibling with no secondary AIS or SCI in 28 patients followed for several years (mean of 3.2 years (0.6 to 7.3 years))\textsuperscript{187}.

**Medical interventions to prevent progression of SCI in SCD**

One randomised controlled study\textsuperscript{189} involved children aged five to 15 years old with sickle cell anaemia (SCA) and SCI with normal transcranial Doppler ultrasonography (TCD) velocities. Of 99 children transfused, six (6\%) had an event (one stroke, five new/enlarged SCIs) while of 97 children observed, 14 (14\%) had an event (seven strokes, seven new/enlarged SCIs) \textit{P}=0.04\textsuperscript{189}. Although this was a positive result in favour of transfusion, its meaning is unclear as significance came from unexpectedly high numbers of overt strokes in the control arm.

**Linking the evidence to the recommendations**

**Discussion and considerations for medical interventions to prevent recurrence of AIS**

The lack of published evidence identified by the systematic review meant that the recommendations are the consensus view of the GDG.

**Secondary AIS prevention in SCD**

Following the review and discussion of the evidence, the GDG agreed the following conclusions. Blood transfusion is almost universally used, where safely available, as secondary prevention of AIS. Historical data suggests that second stroke rates of up to 90\% occur without secondary prevention. Standard practice is to maintain the pre-transfusion HbS level at less than 30\%, whilst keeping the pre-transfusion haemoglobin above 90g/l. There is no firm evidence to recommend exchange over simple transfusion in children with SCD, although iron accumulation may be less with exchange transfusion.

HSCT from an HLA identical sibling seems an effective method of secondary stroke prevention, although only about 10\% of children have a suitable donor. Transplantation in children is likely to result in subfertility, and has an approximately 95\% success rate, with 1\% chance of procedure-related death.

Hydroxycarbamide is inferior to regular blood transfusions in secondary stroke prevention but probably offers some efficacy over supportive care only.

There is no evidence to support the routine use of anticoagulation or anti-platelet agents, although few/no studies have been performed. This may be appropriate in certain circumstances such as arterial dissection.
For further reading see the National Institute for Health and Care Excellence (NICE) Medical Technology guidance\textsuperscript{190}, which has recommended automated exchange for the management of long-term transfusion therapy in patients in SCD.

**Prevention of SCI progression in SCD**

While the randomised controlled trial\textsuperscript{189} found in the systematic review for this question confirms the beneficial effects of transfusion in general, it does not provide adequate evidence for either screening for SCIs or routinely transfusing all children with SCIs. Many SCIs occur before the age of five, and there are difficulties using MRI as a screening test because of the current need for general anaesthesia in young children.

This study\textsuperscript{189} provides some evidence in favour of starting transfusions in children with SCIs. Approximately 30% children with SCD in the UK are now taking hydroxycarbamide, although its role in the prevention of SCIs is unknown. The study only included children with SCA (HbSS and HbS/b\textsuperscript{0}thalassaemia). The same considerations and approach would usually be applied to children with HbSC disease and other types of SCD although specific evidence is lacking.

**Recommendations**

**Medical interventions to prevent recurrence of AIS**

- Continue antithrombotic treatment initiated acutely in children and young people with AIS. Reduce dose of aspirin from 5mg/kg to 1mg/kg after 14 days.

- Treat all children and young people with AIS with aspirin, unless they have SCD or are receiving anticoagulation e.g. for a cardiac source of embolism.

- In patients with cardiac disease the choice of antithrombotic agent should be decided on a case-by-case basis following discussion between the treating neurologist and cardiologist.

- Duration of antithrombotic treatment should be considered on a case-by-case basis depending on risk factors identified.

- Maintain adequate levels of hydration in patients with occlusive arteriopathies including moyamoya, especially when fasting or during intercurrent illness.
AIS recurrence prevention in SCD

- Start regular blood transfusions as secondary stroke prevention in children and young people with SCD, aiming to keep the pre-transfusion HbS less than 30% and keeping the pre-transfusion haemoglobin above 90g/l. This can be done with either exchange or simple top-up blood transfusion.

- Ensure that all children and young people with SCD and their siblings are HLA typed. Children and young people with HLA-identical siblings and recurrent stroke or worsening vasculopathy despite optimum haematological treatment should be referred for discussion of HSCT.

- Monitor children with regular neurocognitive testing, MRI and TCD; frequency should be determined on a case-by-case basis.

- Intensify treatment if there is evidence of progressive cerebrovascular disease, if identified through either TCD or magnetic resonance angiography. Options may include:
  - intensified transfusion with lower HbS target
  - the addition of hydroxycarbamide or antiplatelet agents during red cell transfusions
  - consideration of surgical revascularisation (in the presence of arteriopathy)
  - referral for alternative-donor HSCT

- Children and young people’s cases should be discussed in an appropriate multidisciplinary team (MDT) with experience of managing children and young people with SCD prior to referral for either surgery or alternative-donor HSCT.

- Hydroxycarbamide should be considered as part of a secondary stroke prevention programme when suitable blood (e.g. multiple alloantibodies or hyperhaemolysis) is not available, or when continued transfusions pose unacceptable risks (uncontrolled iron accumulation).

- Hydroxycarbamide may be used as an alternative to blood transfusion if transfusion is genuinely unacceptable to the parents/carers and child. It is imperative that the decision to stop transfusions and switch to hydroxycarbamide is taken by a MDT.
Consider using anticoagulation or antiplatelet agents only when there are other risk factors for cerebrovascular disease that justify their use.

SCI progression prevention in SCD

- Discuss the possible benefits of transfusion with children, young people and families if SCI are identified on MRI. Factors favouring the implementation of a treatment program involving regular blood transfusions include:
  - impaired cognitive performance
  - progressive deterioration in cognitive function
  - evidence of increase in size or number of SCIs on serial MRIs
  - evidence of intracranial or extracranial vasculopathy on MRA
  - other co-existent morbidities of SCD which may benefit from regular blood transfusions, including frequent episodes of acute pain, progressive pulmonary damage, and progressive renal impairment.

- Consider haematopoietic stem cell transplantation in children and young people starting transfusions.

- Consider starting hydroxycarbamide as an alternative therapy if repeated transfusions are declined or contra-indicated.

6.2.3. Surgical and endovascular interventions for AIS

Evidence summary

Neurosurgical intervention for AIS

Decompressive Craniectomy

A systematic review was conducted for the relevant clinical questions and identified three case series studies, of low and moderate quality primarily due to their very small numbers. This suggested that selected paediatric patients benefit from decompressive craniectomy and that recovery with good quality of life is possible even with extensive infarction of the dominant MCA territory. Decompressive craniectomy may also be beneficial following posterior fossa infarction in children and young people.

Revascularisation procedures in moyamoya

The systematic review further identified five case series of low and moderate quality which described direct vessel anastomoses and indirect synangiosis procedures, including encephalo-duro-arterio-synangiosis (EDAS) and multiple burr holes, are both clinically effective and improve angiographic vascularisation with a low risk of
complications in paediatric patients. One large case series of 143 patients\textsuperscript{198} suggested that pial synangiosis continues to protect against stroke over longer term follow-up.

**Interventional neuroradiology**

There were no randomised controlled trials identified comparing endovascular treatment of arterial ischaemic stroke in paediatric patients against medical treatment with IV thrombolysis. However, the GDG noted that current adult studies indicated superiority of recanalisation therapy (following IV thrombolysis) over IV thrombolysis alone.

The limited evidence that does exist was comprised of three small case series of low quality\textsuperscript{199-201}. Two studies\textsuperscript{199,200} used mechanical thrombectomy and showed favourable outcomes on the modified Rankin scale (mRS) (0 to 3) at follow-up. Both studies, however, only presented the results in four children. A third study\textsuperscript{201} reported a partial or complete recanalization rate of 74\% and a complication rate of 29\% in children undergoing thrombolytic therapies.

**Linking the evidence to the recommendations**

**Neurosurgical intervention for AIS**

**Decompressive Craniectomy**

In the adult literature, three European RCTs (DECIMAL (decompressive craniectomy in malignant MCA infarcts), DESTINY (decompressive surgery for the treatment of malignant infarction of the MCA) and HAMLET (hemicraniectomy after MCA infarction with life-threatening edema trial)) of decompressive craniectomy were carried out within 48 hours of stroke onset in patients with malignant (space occupying) infarction of the middle cerebral artery territory aged 18 to 60 years old. The trials showed reduced mortality and an increased number of patients with a favourable functional outcome was shown\textsuperscript{202}.

Strokes of this nature are uncommon in paediatrics, perhaps fewer than 2\% of cases of paediatric AIS. No randomised trials of this nature have been carried out in the paediatric age group but there is no reason to suppose that paediatric outcomes would be any worse on the basis of the limited information published from small case series.

Smith and colleagues\textsuperscript{191} emphasised that walking and talking outcomes were possible following extensive MCA territory infarcts in the dominant hemisphere and for patients in coma but it seems likely that for the best results a degree of case selection and early decision to decompress are important. They suggested that ‘any change in the level of consciousness in a patient with large middle cerebral artery infarction should prompt immediate consideration of decompressive craniectomy’.
Revascularisation procedures in moyamoya

Over 75% of the patients who had a transient ischaemic attack (TIA) experienced a reduction in attacks following surgical revascularisation therapies (whether direct or indirect) and the surgical complication rate overall was low\textsuperscript{196}.

Patients with demonstrated recurrence of clinical ischaemia and/or radiological evidence of infarction, but also with structurally intact cerebral tissue within the target vascular territory, can reasonably be considered for surgical revascularisation\textsuperscript{196}. Several other groups have emphasised the low complication rates, improvement in angiographic appearances\textsuperscript{195} and long-term effectiveness of direct bypass procedures\textsuperscript{194} as well as pial synangiosis\textsuperscript{197,198} and multiple burr hole procedures\textsuperscript{193}. None of the various techniques are clearly superior to the others as reported in the literature.

The indication for surgical revascularisation for patients who are asymptomatic is less clear. Some of these patients may be experiencing more subtle signs of ischaemia such as headaches and poor school performance. In the adult literature, imaging signs other than obvious progressive moyamoya, such as the ivy sign of increased signal in sulci on fluid attenuation inversion recovery (FLAIR) imaging may be helpful in predicting symptomatic patients who are at higher risk of progressing\textsuperscript{203}.

Interventional neuroradiology

The limited available evidence retrieved in the literature review on thrombectomy and thrombolysis is made up of small case series of low quality and therefore the recommendations were shaped largely by the clinical opinion of the GDG and the evidence from adult patients.

Recommendations
Indications for referral to neurosurgery in children and young people with AIS

- Discuss any impairment of conscious level or decline in PedNIHSS in a child with AIS with a neurosurgical team.

- Consider decompressive hemicraniectomy in children and young people with MCA infarction under the following circumstances:
  - neurological deficit indicates infarction in the MCA territory
  - surgical treatment can be given less than or equal to 48 hours after the onset of stroke
  - a decrease in the level of consciousness to a score of 1 or more on item 1a of the PedNIHSS
- PedNIHSS score of more than 15
- while not validated in children, signs on CT of an infarct of at least 50% of the MCA territory with or without additional infarction in the territory of the anterior or posterior cerebral artery on the same side.

- If a patient meets the above circumstances and is not already in a neurology unit, they should be ventilated and neuroprotected and moved to the neurological unit as time critical transfer.

- Consider performing decompressive craniectomy in vascular infarctions in other territories, e.g. posterior fossa infarction.

- Refer children and young people with moyamoya to a paediatric neurosurgical centre with expertise in surgical revascularisation.

- Consider surgical revascularisation in patients with moyamoya and ongoing ischaemic symptoms or other risk factors for progressive disease.

**Indications for referral to interventional neuroradiology**

- Patients with acute AIS causing a disabling neurological deficit (NIHSS score of 6 or more) may be considered for intra-arterial clot extraction with prior IV thrombolysis, unless contraindicated, beyond an onset-to-arterial puncture time of five hours if:
  - PedNIHSS score is more than six
  - a favourable profile on salvageable brain tissue imaging has been proven, in which case treatment up to 12 hours after onset may be appropriate.
7. Haemorrhagic Stroke

7.1. Conditions and factors associated with a risk of HS or recurrence

Haemorrhagic stroke (HS), like arterial ischaemic stroke (AIS), in children and young people has a different risk factor profile to adults; in particular, structural causes are more common (e.g. arteriovenous malformation (AVM), aneurysms, cavernous malformations, arteriopathy with moyamoya, sickle cell disease (SCD)). Bleeding disorders may also present with childhood HS.

The question of recurrence after HS is a major concern to parents/carers and its prevention is a major focus of medical management. The risk of recurrence will depend on the risk factors identified and secondary prevention will be directed by these.

Review questions

- In childhood, which conditions/factors are associated with higher risk of development of HS?
- In childhood, which conditions/factors are associated with higher risk of recurrence of HS?

Further investigations & follow up imaging

- What are the most appropriate investigations to identify underlying risk factors in HS?
- What are the role, modality and timing of brain imaging in assessment and follow-up of childhood HS.

Acute medical interventions

- What is the appropriate management for acute HS in children and young people with sickle cell disease?
- What is the safety and efficacy of coagulation factor replacement for the acute treatment of children and young people with HS?

Medical interventions to prevent recurrence

- What is the safety and efficacy of medical interventions to prevent recurrence of:
  - HS
  - HS in SCD
Surgical & endovascular interventions

- In children and young people with acute/chronic HS, what are the indications for referral to neurosurgery?
- What is the safety and efficacy of surgical interventions in the treatment of acute HS in children and young people?
- What is effectiveness of surgical interventions in the prevention of recurrence of HS in children and young people?
- In children and young people with acute/chronic HS, what are the indications for referral to interventional neuroradiology?
- What is the safety and efficacy of gamma knife intervention in the treatment of acute HS in children and young people?
- Is there a difference in the safety and efficacy of surgical, radiosurgical and endovascular interventions between treating ruptured and unruptured at risk vascular lesions?

7.1.1. Risk factors for HS and recurrent HS

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified 42 studies, which reported data on the risk factors for HS in children and young people. Of the 42 studies, two were systematic reviews and 40 were case series. The quality of evidence was classified as high in four studies, moderate in 17 studies and low in 21 studies.

Study populations were reported from a number of regions including America (18 studies), Europe (nine studies) and the UK (one study). For the studies where applicability to the UK population was less certain, the results were approached with caution.

There is some evidence that male gender, older age and black ethnicity increase the risk of HS from studies of low and moderate quality in samples that are applicable to the UK. The quality of evidence for haematological risk factors was classified as moderate (four studies) and low (four studies). The strongest evidence is for neurovascular disease, where 29 studies, including the four high quality studies, report on a range of neurovascular malformations as a risk factor for haemorrhagic stroke. Further details on each study are provided within the evidence tables, in Appendix 4c.
Linking the evidence to the recommendations

From the 42 included studies, 13 risk factors for HS in childhood were identified (see recommendations). Traumatic brain haemorrhage was excluded. The guideline development group (GDG) took into account the characteristics of the children and young people described in the included studies, such as age, presenting symptoms, lesional factors and investigation findings. The GDG also considered the risk factors for recurrence in the context of their clinical experience and this was especially informative for the risk factors that were only reported in a small number of the included studies.

The heterogeneity of the studies and the frequent descriptive rather than statistical presentation of the results, means that it is not possible to estimate the magnitude of risk. This is particularly challenging in the context of vascular malformations where the estimated risk of future haemorrhage could critically inform the risk:benefit ratio of different interventional strategies. Estimation of risk needs to be individualised and is likely to be informed to some extent by angioarchitectural features. Some diagnoses, such as hereditary haemorrhagic telangiectasia, have implications for other family members. Wider family counselling and investigation (e.g. brain imaging) should be undertaken with consideration of the clinical implications, especially for asymptomatic individuals, and it would be appropriate to involve a specialist neurovascular team in this.

Parent/carers and young people workshops revealed the importance of early individualised information around risk and how to minimise risk. Preferences identified at these workshops were for information around risk to be provided in direct conversation and supported where possible in multiple formats for later reflection (e.g. web-based, written, graphic formats).

Recommendations

Risk factors for first HS

- Be aware that the following factors/conditions are associated with an increased risk of HS in children and young people, as tabulated:

<table>
<thead>
<tr>
<th>Risk Category</th>
<th>Included factors/diagnoses</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vascular disorders</td>
<td>• AVM, especially with arterial phase aneurysms, varicosities or venous stenoses on the draining veins</td>
</tr>
<tr>
<td></td>
<td>• cavernous malformations, especially Zabramski type 1 &amp; 2</td>
</tr>
<tr>
<td></td>
<td>• cerebral arterial aneurysms</td>
</tr>
</tbody>
</table>
• Take these factors into account when considering a need for counselling in high risk groups.

• Information on risk factors should be delivered in face-to-face conversation with parents/carers and young people (where appropriate) and supported where possible with web-based or written materials for later reference. The information provided should be age-appropriate and multi-format.

**Risk factors for recurrent HS**

• Be aware of increased risk of recurrence in children and young people with HS and the following risk factors:
  - AVM
  - cerebral arterial aneurysms
  - cavernous malformations
  - moyamoya
  - SCD
  - all severe bleeding disorders
  - ongoing anticoagulation
  - illicit drug use e.g. amphetamines and cocaine

• Be aware that in arteriovenous malformations, which have already bled, the greatest risk of a rebleed is from the part of the malformation which was responsible for the initial haemorrhage. Intratinal or perinidal aneurysms and venous varicosities/stenoses are sinister features.
• Take these factors into account when considering the need for counselling in high-risk groups.

• Information on risk of recurrence, and how to minimise risk, should be delivered in face-to-face conversation with parents/carers and young people (where appropriate) and supported where possible with web-based or written materials for later reference. The information provided should be age-appropriate and multi-format.

7.1.2. Investigations to identify underlying risk factors in HS

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified three studies\(^{21,73,157}\) which discussed investigations used to identify underlying risk factors for HS. The quality of evidence was classified as high on one study\(^{73}\) and low in two studies\(^{21,157}\).

The quality of evidence was classified as high in one study\(^{73}\) and low in two studies\(^{21,157}\). Study populations were reported from a number of regions including Europe (one study\(^{157}\)), India (one study\(^{73}\)) and Saudi Arabia (one study\(^{21}\)).

Tests used in the studies to identify underlying risk factors included imaging such as CT and magnetic resonance imaging (MRI)/magnetic resonance angiogram (MRA)\(^{21,73}\) and blood tests\(^{157}\). Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

The GDG discussed the evidence found in the literature review and formulated the recommendations based on this and clinical experience.

The evidence base for an increased risk of HS in haematological disorders (thrombocytopenia\(^{206}\)), inherited\(^{220}\) and acquired\(^{225}\) bleeding disorders and haemoglobinopathies is poor, but the absence of good evidence does not equate with the absence of risk. It is well recognised by clinicians treating these patient groups that HS is a risk and much of the management of these conditions is aimed at preventing this grave complication. In some instances HS may be this first evidence of a bleeding disorder in a baby or young child and identification of the disorder with appropriate treatment may be life-saving.
The presence of an inherited or acquired bleeding disorder can be detected in the vast majority of cases by carrying out a full blood count and a coagulation screen (when the latter is taken from a free flowing venous sample and not a capillary sample). There are three severe inherited bleeding disorders that would not be excluded by a normal coagulation screen and full blood count, namely factor XIII deficiency, Glanzmann’s thrombasthenia and alpha 2-antiplasmin deficiency. These are extremely rare autosomal recessive conditions and should be considered in a baby or young child with HS whose parents are consanguineous; such consideration should be discussed with a paediatric haematologist who can advise on appropriate testing where necessary. In an older child, these conditions would likely have manifested with other haemorrhagic symptoms prior to the HS.

**Recommendations**

- Carry out the following investigations in children and young people diagnosed with HS:
  - **Haematological investigations:**
    - coagulation screen including activated partial thromboplastin time (aPTT), prothrombin time (PT), fibrinogen (ideally by Clauss method) (taken by a free-flowing venous sample), full blood count (FBC), haemoglobinopathy screen.
    - discuss any abnormality of these haematological tests with a paediatric haematologist so that they can advise on further testing including specific clotting factor assays.
    - establish whether the parents are consanguineous as there are some rare severe recessive bleeding disorders that cannot be ruled out with a normal blood count and coagulation screen.
  - **Imaging investigations:**
    - discuss the child’s case in a neurovascular multidisciplinary team (MDT) to plan further investigations to identify/exclude underlying vascular malformation and to plan any interventional treatment; such investigations may include non-invasive angiography such as computed tomography angiography (CTA) or MRA, as well as formal catheter angiography (CA)

- If the child is known to have SCD, additional tests should include transcranial Doppler ultrasonography (TCD) and an extended blood group phenotype (e.g. ABO, Rh C, D and E, and Kell).
7.1.3. Follow-up imaging in HS (role, timing, modality)

**Evidence summary**

Whilst there are a number of studies addressing the issue of imaging of paediatric stroke on presentation and in the acute setting, a systematic review identified only one high quality prospective case series which addressed the strategies for follow-up imaging of HS. This study explored the use of MRA in children and found that MRA was able to visualise the internal carotid arteries at all ages. The primary branches of the posterior circulation and the anterior and posterior communicating arteries could be visualised inconsistently but this improved up to age six. The authors also described how MRA was used following treatment for AVM and that no discrepancy was found between MRA and digital subtraction angiography (DSA) findings.

**Linking the evidence to the recommendations**

In the absence of evidence, the recommendations were based on GDG discussions and a Delphi Panel consensus. Two of the recommendations were formulated through a Delphi survey (which are clearly indicated). For a full description of the Delphi consensus method see Appendix 5.

**Recommendations**

- Discuss the modality and timing of imaging in children and young people with HS within a MDT; this will be influenced by factors relating to the individual patient and the lesion.

- Consider the following commonly used follow-up imaging paradigms for treated vascular malformations:

<table>
<thead>
<tr>
<th>AVM</th>
<th>Treatment and frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endovascular treatment</td>
<td>• CA to be performed at three to six months after obliteration</td>
</tr>
<tr>
<td>Surgical resection</td>
<td>• MRI and MRA and catheter angiogram to be performed at three to six months after resection</td>
</tr>
<tr>
<td>Stereotactic radiosurgery (SRS)</td>
<td>• MRI and MRA to be performed two years following treatment</td>
</tr>
<tr>
<td></td>
<td>- if appears obliterated on MRI confirm with catheter angiogram</td>
</tr>
<tr>
<td></td>
<td>- if still apparent, MRI/MRA &amp; CA at three years to evaluate for further treatment</td>
</tr>
</tbody>
</table>
### Aneurysm Treatment and frequency

| Endovascular treatment | • MRI and MRA to be performed every three to six months, for two years  
• follow-up thereafter should be determined on a case-by-case basis with giant or partially treated aneurysms likely to require more intensive follow-up |

| Surgical | • one catheter angiogram demonstrating exclusion of the aneurysm is sufficient; however, there should be recognition that in children presenting with cerebral aneurysms, de novo aneurysms may develop in subsequent years |

- Offer all children and young people with a previously treated brain AVM and angiographic confirmation of obliteration a final catheter angiogram at 16 to 18 years of age, prior to transition to adult services, to exclude AVM recurrence or a de novo lesion. [DELPHI]

- Consider surveillance imaging in children and young people with a single or multiple untreated cavernous malformations for the first two years following diagnosis, with further follow-up imaging offered if there are new or changing clinical symptoms which could be attributable to the cavernous malformations. [DELPHI]

- If no cause of HS is identified acutely, follow-up should be undertaken with a MRI and MRA at six months as a minimum and consideration should be given to catheter angiography thereafter.

### 7.2. Medical and surgical interventions

The literature around the treatment of paediatric HS is characterised by a lack of randomised controlled trials and small sample sizes. It is important to distinguish between treatment (i.e. reverse or limit brain injury) and secondary prevention (prevent another HS).

Most HS in children and young people is secondary to a structural vascular lesion. The lesions underlying haemorrhagic stroke in children and young people are thought to be around 40 to 50% AVM, 10 to 15% aneurysm, and 5 to 10% cavernous malformation.
Management options include endovascular, surgical, radiosurgical and conservative approaches that need to be patient and lesion oriented.

The parents/carers and young people workshops raised the importance of families being informed in a timely way about reasoning for interventions, what to expect, having the opportunity to exercise choice where possible, and ask questions. A named key contact is a helpful support to families in maintaining contact with the clinicians and being updated regularly.

**Review questions**

**Acute medical interventions**

- What is the appropriate management for acute HS in children and young people with sickle cell disease?
- What is the safety and efficacy of coagulation factor replacement for the acute treatment of children and young people with HS?

**Medical interventions to prevent recurrence**

- What is the safety and efficacy of medical interventions to prevent recurrence of:
  - HS
  - HS in SCD

**Surgical & endovascular interventions**

- In children and young people with acute/chronic HS, what are the indications for referral to neurosurgery?
- What is the safety and efficacy of surgical interventions in the treatment of acute HS in children and young people?
- What is effectiveness of surgical interventions in the prevention of recurrence of HS in children and young people?
- In children and young people with acute/chronic HS, what are the indications for referral to interventional neuroradiology?
- What is the safety and efficacy of gamma knife intervention in the treatment of acute HS in children and young people?
- Is there a difference in the safety and efficacy of surgical, radiosurgical and endovascular interventions between treating ruptured and unruptured at risk vascular lesions?
7.2.1. Acute medical interventions for HS

Evidence summary

Use of coagulation factors replacement for the acute treatment of children and young people with haemorrhagic stroke

Evidence for the management of children and young people with HS secondary to underlying haematological conditions is of poor quality generally. A systematic review was conducted for the relevant clinical questions and identified four studies \(^{66,69,229,230}\). The quality of evidence was classified as moderate in two studies \(^{66,229}\) and low in two studies \(^{69,230}\). Study populations were reported from a number of regions including Egypt, America, and Turkey.

Using a small retrospective case series, Aydinli and colleagues \(^{66}\) described the management of haemorrhagic disease of the newborn secondary to vitamin K deficiency, this is not relevant to UK practice as all babies are given vitamin K at birth in the UK unless parents/carers choose otherwise. The study by Nakar and colleagues \(^{230}\) describes the management of boys with severe haemophilia complicated by antibodies to FVIII with recombinant factor VIIa, but as this is a very specialist area it cannot be extrapolated to more general HS. The third study described the treatment of intra-cranial haemorrhage in children with immune thrombocytopenic purpura \(^{69}\); however, the very small number of children in this study and the lack of a comparison group meant that it did not provide any evidence for the recommendations.

Heffren and colleagues \(^{229}\) published a small case series on the use of Nimodipine in both aneurysmal and non-aneurysmal subarachnoid haemorrhage (SAH) in children. Nimodipine is the standard of care in adult SAH patients to prevent the effects of cerebral vasospasm. This study showed that clinical outcomes appeared favourable compared to the adult population but there was a significant incidence of hypotension requiring intervention, dose titration or discontinuation.

Linking the evidence to the recommendations

As the studies identified in the systematic review provided very little evidence that could be used to form recommendations for this question, the following discussion and recommendations are based on the consensus of the GDG.

Children and young people with HS often develop an acquired coagulopathy (disseminated intravascular coagulation) secondary to a significant intracerebral haemorrhage and may need support with blood products to minimize any additional bleeding that might otherwise result. Abnormal coagulation results at presentation in a
previously well child should be discussed with a haematologist so that appropriate investigations can be carried out urgently to ascertain whether the coagulation abnormality is primary or secondary. Severe haemophilia A and B and several severe rare bleeding disorders (most notably severe FX and FXIII deficiency) can present with an intracerebral bleed as the first event and delay in measuring the relevant clotting factors and starting treatment could have a grave impact on outcome.

**Recommendations**

- Take blood for the measurement of routine coagulation parameters (PT, partial thromboplastin time (PTT), Clauss fibrinogen) and FBC in all children and young people presenting with HS. Abnormal results should be discussed with a paediatric haematologist in order that appropriate investigations can be carried out urgently to ascertain whether a coagulation abnormality is primary or secondary.

- Be aware that coagulation abnormality can be corrected to allow neurosurgery, if surgery is deemed appropriate.

- Discuss coagulation management options with the haematology team if the child/young person has a known underlying inherited or acquired bleeding disorder; treat the child without delay with the relevant coagulation factor replacement; this could either be supplied by their family or their treatment centre.

- Transfer children and young people with an underlying inherited bleeding disorder (such as severe haemophilia) who have an intracerebral bleed in HS to a Paediatric Haemophilia Comprehensive Care Centre (CCC) as soon as possible.

- Treatment should be focussed on maintaining normal levels of the appropriate coagulation factor for a period of intense treatment and then prophylactic treatment to prevent recurrence.

- Consider Nimodipine (mean starting dose 1mg/kg every four hours) to prevent the effects of vasospasm in children and young people with subarachnoid haemorrhage.
7.2.2. Interventions to prevent recurrence of HS

Evidence summary

Medical interventions to prevent recurrence of HS

A systematic review was conducted for the relevant clinical questions and identified one study, which was of moderate quality\textsuperscript{231}. This study discussed the effectiveness of cryoprecipitate and fibrogammin in treating and preventing the recurrence of intracerebral haematomas (ICH) in patients with FXIII deficiency. It also reported that 94.7% of patients had a good prophylaxis response without recurrent haemorrhage but the sample described was small and there was no control group.

Where there is an underlying haematological condition causing HS, ongoing management to prevent recurrence is based on replacement of a deficient coagulation factor or treatment of thrombocytopenia. The evidence for clotting factor replacement is essentially non-existent other than in historical controls. Before the advent of safe, effective clotting factor replacement, children with severe bleeding disorders simply died following HS and in the developing world this remains the most likely outcome, as treatment is generally not available because of cost.

GDG consensus determined that on-going clotting factor replacement, or prophylaxis, is the standard of care in this clinical situation in the UK and should be managed in a Paediatric Haemophilia CCC.

Management of HS in children with immune thrombocytopenic purpura (ITP) is also a very specialist area and can be managed in a tertiary children’s hospital, the treatment strategy being based on maintaining an adequate blood platelet count in order to prevent recurrence. The detail of how this may be achieved is outside the scope of this document\textsuperscript{232}.

Medical interventions to prevent recurrence of HS in SCD

One study describes seven children with SCD and spontaneous intracranial haemorrhage\textsuperscript{225}. All were assessed for intracranial aneurysms and one underwent coiling of multiple aneurysms. Three patients had small intracranial aneurysms not suitable for intervention and were followed closely, and one of these were enlarged and required coiling. Two patients started on the medication hydroxyurea (‘hydroxyurea’) and four on regular transfusions.
Linking the evidence to the recommendations

A summary of discussion and considerations for medical interventions to prevent recurrence of HS

Due to the small amount of published evidence and the methodological shortcomings of the study, the recommendations are the result of discussion and consensus of the GDG.

Secondary HS prevention in SCD

Following review and discussion of the evidence, the GDG came to the following conclusions. Acute management should be the same as for other children with HS, although urgent exchange transfusion is often performed to reduce the risk of cerebral vaso-occlusion secondary to vasospasm, raised intracranial pressure among other symptoms, and to reduce the risk of any future neurosurgical procedures.

Neurosurgical management should be the same as for other children or young people, although it should take into account that the child has an underlying vasculopathic process and further aneurysms may occur in the future. Multidisciplinary discussions should include a physician with expertise in paediatric SCD. Exchange transfusion should be performed prior to any neurosurgical procedures if time permits.

The increased risk of HS in children or young people with SCD is likely to be related to the same vasculopathic process responsible for the increased risk of AIS, and be primarily related to the pathological processes which flow from polymerisation of deoxygenated sickle haemoglobin (HbS). It is therefore reasonable to use measures for the prevention of AIS in SCD to prevent recurrence of HS, including blood transfusions, hydroxycarbamide and haematopoietic stem cell transplantation.

These studies only included children and young people with sickle cell anaemia (SCA) (HbSS and HbS/b^thalassaemia). The same considerations and approach would usually be applied to children or young people with HbSC disease and other types of SCD although specific evidence is lacking.

Recommendations

Medical interventions to prevent recurrence of HS

- Refer all children and young people with inherited bleeding disorders to a children’s CCC as the management of all inherited bleeding disorders is highly specialised. They will be registered on the United Kingdom Haemophilia Centre Doctors’ Organisation’s (UKHCDO) National Bleeding Disorders database.
The management of intracranial haemorrhages associated with these disorders is outside the scope of this document. Preventative strategies for re-bleed will relate to prophylactic coagulation factor replacement of the relevant protein.

**HS recurrence prevention in SCD**

- Perform neuroimaging as recommended for other children and young people with acute HS.

- Consider administering a transfusion to decrease HbS less than 30% prior to direct intra-arterial injection of contrast for catheter angiography.

- Provide anti-sickling treatment to children and young people with SCD and HS, and either a regular blood transfusion or a haematopoietic cell transplantation from a human leukocyte antigen (HLA)-matched sibling (or alternative donors in rare circumstances).

- Provide regular blood transfusions if there is clear evidence of arteriopathy (e.g. occlusive lesions or aneurysms) to keep HbS less than 30%.

- Ensure that all children and young people with SCD and their siblings are HLA typed. Children and young people with HLA identical siblings and recurrent stroke or worsening vasculopathy despite optimum haematological treatment should be referred for discussion of haematopoietic stem cell transplantation (HSCT).

- Consider children and young people with HS and isolated small aneurysms and no other cerebral vasculopathy for treatment with hydroxycarbamide or regular blood transfusions in addition to evaluation for endovascular or surgical treatment.

- Follow-up children and young people with HS in SCD, long-term with repeat neurocognitive testing, MRI and TCD to assess evidence of progressive cerebrovascular disease.

- Children and young people’s care should be discussed in an appropriate MDT with experience of managing children with SCD prior to referral for either
7.2.3. Surgical and endovascular interventions for HS

Evidence summary

Neurosurgical intervention for HS

A systematic review was conducted for the relevant clinical questions and identified 38 case series which outlined the outcomes of surgical treatment either alone or in combination with other modalities of treatment for dealing with specific vascular lesions which underlie HS. These are mostly single institution studies and few of them report on more than 50 patients. The quality of evidence was classified as high in four studies, moderate in 16 studies, and low in 18 studies. Study populations were reported from a number of regions including America (17 studies), Europe (five studies), and the rest of the world (16 studies).

These complex series of studies are summarised in accordance with those making general points about HS, rate of re-bleeding, management of vascular malformations, and surgical, endovascular and SRS treatments.

HS (general)

Kumar and colleagues reported on the presentation of clinical features, underlying aetiological lesions and outcome of 50 consecutive paediatric patients with HS managed in one neurosurgical centre. AVM was reported to be the leading cause of HS (44%), but with a wide range of underlying aetiologies. 74% of patients had either a good outcome or moderate disability and 26% a poor outcome. Cerebellar bleed and late presentation were associated with poorer outcomes and age was not found to be associated with outcome. These results are typical for paediatric series of this nature.

Levy and colleagues described 93% of children return to their previous activity levels and found no treatment related morbidity nor mortality.

Arteriovenous malformations

In the studies presenting mortality data from a primarily surgical series of AVM there was a range of mortality rates from 3.7% to 21%. Summarising the literature relating to AVM within the various studies of all treatment modalities haemorrhage was reported to have occurred in the target AVMs in 52% to 71.5% of cases.
Aneurysm

Within the included studies, haemorrhage had occurred in the target aneurysms in up to 95.4% of cases\textsuperscript{205}.

Cavernous malformation

Within the various studies identified, haemorrhage had occurred in the target cavernous malformations in 20% to 100% of cases. The mortality rate percentage in Abla and colleagues\textsuperscript{210} series of brainstem cavernous malformations was 2.5%; otherwise no mortality data were reported in the surgical series. A post-operative worsening of neurological function in 48% of the cases, of which 23% resolved, was also reported.

Bilginer and colleagues\textsuperscript{212} reported no permanent morbidity with temporary neurological deficit and new onset seizures of cavernous malformation patients. While the reported outcome in this study showed 66.7% of patients scoring five on the Glasgow Outcome Scale (GOS), 27.8% showing four and 5.5% showing three, 90.9% presenting with seizures were seizure free (Engel class 1) post-operatively. Nine out of 15 (60%) patients with pre-operative neurological deficits had resolved and the other six had improved.

Another series of a case study describing brainstem cavernous malformation patients reported surgical related morbidity in 48.1% of patients\textsuperscript{219}. There was a neurological worsening in 17 patients (32.7%) with 10 of these recovering to pre-operative levels.

Re-haemorrhage rates

Arteriovenous malformations

A variety of studies reported re-haemorrhage rates from 2.4% to 23\textsuperscript{213,214,218,223,224,237,238}\textsuperscript{239}. For all forms of treatment there is a clear finding that AVMs completely obliterated on follow-up CA do not have an incidence of re-haemorrhage, although recurrent AVMs are well described in the paediatric population.

In all studies where surgery was the sole treatment or was performed in combination with other modalities, there was no re-haemorrhage rate when the AVM was completely excised. The rates of AVM haemorrhage after SRS were reported as 0%, 1.6%, 2.4%, 5.5% and 10%.

Cavernous malformation

Abla and colleagues\textsuperscript{210} reported a post-operative haemorrhage rate of 5.25% per year in their retrospective case series, with all of these being brainstem lesions with post-operative residual. Li and colleagues\textsuperscript{106} reported a post-operative annual haemorrhage rate of 0.5% with a significant difference between the rate for residual lesions 2.5% per year versus complete excision of 0.3% per year.
Management of vascular malformations (multimodality treatment)

Darsaut and colleagues\textsuperscript{238} reported a 24 year experience of 120 children with AVMs, of which many of them had been managed surgically. Zheng and colleagues\textsuperscript{240} described a similarly large series of 127 children with AVMs with over half of that group managed with interventional radiological procedures and with many having multimodality treatment involving microsurgery, radiosurgery or a combination. This approach was common in several other reported case series.

Furthermore, one case\textsuperscript{238} suggested that initial single-modality therapy led to AVM obliteration in 76\% low-grade AVMs, improving to 87\% with further treatment. For high grade AVM, however, cure rates were considerably lower at 9\% after single modality therapy and 35\% after multiple modalities. Strategies of surgery alone or endovascular treatment followed by surgery were highly effective in low grade AVM (95\% and 100\% cure rate respectively). The authors emphasised the value of a delayed post-treatment angiogram three to six months after treatment in confirming whether obliteration had genuinely been achieved even if the immediate post treatment angiogram suggested it had.

Aneurysm

Hetts and colleagues\textsuperscript{221} described 77 paediatric patients with 103 aneurysms managed over a 27 year period with a range of treatment strategies in 59 patients including selective clipping, surgical trapping, selective coiling, and endovascular parent artery occlusion. Eighteen patients with long segment vascular dysplasias, none of whom had presented with bleeding, were managed conservatively to good effect with none of them bleeding over the follow-up period. Other author’s present series of more than 50 paediatric patients with aneurysms treated with a range of modalities\textsuperscript{103,215,241}.

Single modality literature

Arteriovenous malformations

Surgery has the benefit of immediate negation of haemorrhage risk if the AVM is completely excised. However, mortality and morbidity may be higher than other treatments as this is clearly linked to size, location and angio-architecture of the AVM.

The only study to describe morbidity specific to a surgical series was carried out by Di Rocco and colleagues\textsuperscript{100}, whereby a 24.1\% rate of new neurological deficit, 8.9\% of which were permanent, was reported.

The success rate of a complete radiological obliteration of the AVM by surgical treatment varied between studies depending upon the distribution of the grade of the lesions\textsuperscript{100,238}.
Aneurysm

Mortality rates following surgical intervention were presented as 0%\textsuperscript{239}, 2%\textsuperscript{215}, 5%\textsuperscript{102,103}, 8.77%\textsuperscript{241} and 11%\textsuperscript{104}. Aryan and colleagues\textsuperscript{215} reported 20% neurological complication rate and 4% other complication rate, Kalani and colleagues\textsuperscript{239} reported a 14.3% neurological and 14.3% other complication rate, Mehrotra and colleagues\textsuperscript{241} reported a GOS of five in 69.2% and GOS of four in 15.4%, Proust and colleagues\textsuperscript{205} reported a GOS of five in 63.6% and GOS of three in 13.6%, while Sharma and colleagues\textsuperscript{103} reported a 7.5% neurological and 10% other complication rate.

Cavernous malformation

Bilginer and colleagues\textsuperscript{212} presented data on 36 paediatric patients with cavernous malformations showing good outcomes for surgical resection, and noted that none of their patients, whether operated on or managed conservatively, re-haemorrhaged during their follow up period (mean 6.9 years).

Li and colleagues\textsuperscript{106} on the other hand described surgery related morbidity in the short term at least in 25 of 52 of patients undergoing cavernous malformation resections in the brainstem including postoperative worsened neurological function in 17 although most of those recovered to baseline levels over time.

Endovascular treatment

Arteriovenous malformations

Two studies\textsuperscript{211,240} found a 0% mortality rate when assessing endovascular therapies for AVM.

Six studies were also identified in the review\textsuperscript{208,218,223,224,236,237}, four presenting a 0% mortality rate following SRS\textsuperscript{208,223,224,237}, and two presenting a mortality rate of 0.7% and 1.3\textsuperscript{218,236}. Endovascular and surgical therapy combined is described in two studies and in both the mortality rate was 0\textsuperscript{100,238}. Bristol and colleagues\textsuperscript{213} further studied mortality with a variety of treatments in relation to the Spetzler-Martin grade and reported the following grade one – 0%, grade two– 12.5%, grade three – 11.7%, grade four – 6.2%, and grade five – 0%. Endovascular only cohorts describe complete obliteration rates of 4% and 21.2% decreasing to 19.7% if treatment is delayed\textsuperscript{238,240}.

Endovascular AVM cohorts were presented by Bristol and colleagues\textsuperscript{213} where they described a 12.1% new neurological deficit rate in patients, 3% of which were permanent. While Zheng and colleagues\textsuperscript{240} reported a 7.3% deficit rate, 2.4% of which were permanent, Soltanolkotabi and colleagues\textsuperscript{235} reported an overall complication rate of 26.3%, some of which were clinically silent and none of which were permanent.
Aneurysm

Two studies presented mortality rates following endovascular intervention and reported the mortality rate between 0%73 and 4%207. While Saraf and colleagues73 reported a GOS of five in 87%, GOS four in 4.3%, GOS three in 4.3% and GOS two in 4.3%, Lv and colleagues207 reported a GOS of five in 92% and GOS of four in 4%.

Hetts and colleagues221 reported the development of new aneurysms in four children and enlargement of unruptured aneurysm in two children.

Stereotactic radiosurgery

Arteriovenous malformations

SRS has been utilised in the UK since 1985 to treat AVM. It is a fully accepted, safe and proven primary treatment modality for AVM alone or in combination with endovascular or surgical intervention100,238,240.

In an earlier, larger case series study, 200 patients were described with a total obliteration rate of 59% after one or more SRS procedures were reported223. A post-radiosurgery haemorrhage was seen in 2.5% of children and 1% of patients suffered adverse radiation effects.

The third study reported a total obliteration rate of 30%208. None of the children who achieved total obliteration underwent staged SRS and total obliteration was associated with a higher dosage compared to those with partial or no obliteration. The haemorrhage rate after radiosurgery was 17%.

Potts and colleagues reported218 84% of patients having primary SRS. Thirty-one percent of patients required further treatment for their residual AVM consistent of either further SRS, microsurgery, endovascular intervention or a combination of those. 78% were seen to have good outcomes.

Zabel-du Bois and colleagues237 analysed the clinical outcome of SRS, early and late toxicity and post-treatment bleed rate. Complete obliteration was observed in 75% with doses of 18 Gy or above. There was no increased bleeding risk after SRS in patients who had bled before. The annual bleeding risk observed following SRS was 9.1% in the first and 13.6% in the second year. Large AVM over 3cm diameter showed intracranial haemorrhage in 50%, while no intracranial haemorrhage was reported in AVM under 3cm. No new onset of neurological or visual deficits was seen during the follow-up period. No significant side effects were observed, no focal necrosis and no radiation induced malignancy.
A radiosurgical case series study carried out by Kano and colleagues reported a total obliteration rate of 81% after one or more radiosurgery procedures. The effects were latent and post radiosurgical haemorrhage was observed in 6%. No haemorrhages were observed after obliteration of the AVM was achieved. Adverse radiation effects were observed in 6% of patients, 1.5% of which were permanent deficits, and these were more likely in eloquently located lesions and lesions with a higher Pollock-Flickinger score.

Obliteration rates were higher with smaller AVM size (both volume and diameter) and larger marginal dose. In other case study series, a 68% and 34% obliteration rate was reported. Potts and colleagues further highlighted a lower marginal dose was utilised, which may explain the lower obliteration rate.

Aneurysm

SRS does not have a role in the management of aneurysms.

Cavernous malformation

In the study by Lee and colleagues, eight patients were described undergoing radiosurgery for cavernous malformation, of which one patient died of re-bleeding one day after the radiosurgery. The lesions otherwise were stable or smaller at follow up.

Management of unruptured vascular lesions

No randomised controlled studies compared endovascular treatment of ruptured and unruptured vascular lesions. Agreement on a recommendation was therefore achieved following a Delphi consensus method.

SRS is a safe and efficacious treatment in ruptured and un-ruptured AVMs. The treatment effect is latent and full obliteration is not expected before a time interval of at least two years. Residual AVMs might require further radiosurgery treatment after such interval.

First time haemorrhage or recurrent haemorrhage can be observed in this interval period and need to be considered when evaluating all treatment modalities, their risks and benefits and overall morbidity/mortality free cure rates and outcomes. Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

Neurosurgery: indication, safety, efficacy including prevention of recurrence

Whilst the spectrum of underlying aetiologies and the location and distribution of HS is likely to differ in children compared to adults, all children with HS should be discussed with
neurosurgeons, partly to allow assessment of any potential benefit to them from clot removal, and also as part of the process of evaluating the underlying lesion responsible for the haemorrhage and its best management.

The following summary and recommendations were formed with a combination of the evidence described above, the consensus reached with a Delphi survey and clinical experience of the GDG.

Decision-making in patients with HS secondary to a vascular lesion needs to be undertaken in a multidisciplinary neurovascular team involving neurosurgeons, neurologists, neuroradiologists/interventional neuroradiologists and nurse specialists. Consideration of all treatment modalities and combination of modalities must be discussed and presented to the patient (if appropriate for their age) and their families. Discussions for these complex decisions may need to take place over multiple meetings.

**Arteriovenous malformations**

Imaging evaluation of AVM in the acute and follow-up period has been outlined previously. Specific ‘high risk’ factors which indicate a higher risk of re-bleeding and guide the urgency of treatment include:

- Associated intranidal or pedicular aneurysm
- Deep venous drainage
- Stenosis of a draining vein
- Periventricular or intraventricular location

While the literature on multi and single modality treatments has also been mentioned, in clinical practice multidisciplinary discussion is key to mapping out a person- and lesion-specific treatment strategy, with a clear end point defined where possible. Such end points may include focal treatment of high-risk features or attempting to achieve angiographic cure.

The recent ARUBA (A Randomised trial of Unruptured Brain Arteriovenous malformations) trial has challenged treatment for unruptured AVM in adults\(^{242}\); however, it is questionable whether these findings apply to children\(^{91}\) who have a longer projected life-span\(^{91}\). The issue of whether the treatment threshold for intervention in paediatric as opposed to adult AVM should be different was presented to the Delphi panel.

**Aneurysm**

Aneurysms are heterogeneous lesions, with often undefined wall substrate and natural history, amenable to a variety of approaches, and requiring a specific plan in each case.
While clipping and endovascular approaches are both possible, the GDG note that clipping of the aneurysm in younger patients (with longer expected life span) may be a superior treatment due to the need to retreat coiled patients over time.

**Cavernous Malformation**

Xia and colleagues\(^{222}\) presented results suggesting that complete microsurgical removal of cavernous malformations should be feasible in almost all cases where is necessary, even if noted as multiple and a staged approach is required. Morbidity is low in modern series making use of neuronavigation and intraoperative ultrasonography. As opposed to conservative management, it is harder to define what constitutes grounds for attempting surgical removal or other interventions for patients with cavernous malformations.

As with AVMs, complete surgical excision of cavernous malformations will immediately negate further haemorrhage risk and will usually have a beneficial effect on seizures caused by the lesion. Associated mortality and morbidity is dependent on lesion location and size and is low outside of the brainstem.

Indications to consider microsurgical removal would include a single symptomatic haemorrhage from a lesion in a non-eloquent area where risk of resection was felt to be low. In addition, multiple episodes of symptomatic haemorrhage from any cavernous malformation would prompt consideration of surgical resection\(^{212}\).

**Endovascular treatment: indication**

Given the evidence the GDG felt that endovascular treatment of haemorrhagic vascular lesions should be considered as part of a MDT approach to treatment.

**Stereotactic radiosurgery intervention: safety and efficacy**

SRS is a proven technique in the treatment of patients with vascular pathology and haemorrhagic stroke. It is minimally invasive and due to the focussed nature of the radiation, and is generally safe to deliver and attractive for lesions located in deep, eloquent parts of the brain\(^{236}\).

AVM totally obliterated by SRS does not recur, and until obliteration is complete a bleeding risk persists. It is a technique that can be repeated should a residual AVM persist after an interval of two to four years. Combination treatment with open surgery or endovascular treatment can be considered.
Cure rates of more than 80% can be observed; however, larger and eloquent lesions with higher Pollock-Flickinger score are associated with a reduced success rate, higher adverse radiation effects (ARE) and might require a staged treatment. Whilst the procedure is typically carried out under local anaesthetic, children under 13 years of age are typically treated under general anaesthetic in one of the two commissioned supra-centres.

The safety and efficacy of surgical, radiosurgical and endovascular interventions in the treatment of ruptured in comparison to unruptured vascular lesions

A Delphi consensus was reached concerning the management of unruptured vascular lesions. A recommendation that more active management should be considered more readily in children than in adults due to the higher cumulative risk of rupture reached consensus.

**Recommendations**

**Neurosurgical management of HS**

- Children and young people with HS should always be cared for in conjunction with a neurosurgical team.

- Do not routinely evacuate ICH in children and young people, except in cases where there is a rapidly deteriorating age-appropriate Glasgow Coma Scale (GCS) score.

- The management of any structural vascular lesions underlying an ICH (most commonly AVM, aneurysm or cavernous malformation) must be discussed at a neurovascular MDT.

- Treat lesions at higher risk of early re-bleeding urgently (i.e. on the initial admission), such as:
  - ruptured aneurysms
  - arteriovenous malformations with high risk features.

**Interventional neuroradiology**

- Discuss patient’s cases with acute HS and vascular lesions in a neurovascular MDT including an interventional neuroradiologist.
**Stereotactic radiosurgery**

- SRS may be considered as a treatment option for vascular lesions and should be included in the discussion of the case in the MDT.

**The safety and efficacy of surgical, radiosurgical and endovascular interventions in the treatment of ruptured in comparison to unruptured vascular lesions**

**Arteriovenous Malformations (AVM)**

- Consider treating both ruptured and un-ruptured AVM.
- Consider active management more readily in children and young people with diagnosed unruptured AVM than in adults due to the higher cumulative risk of rupture attributable to the projected longer life span. [DELPHI]
- Discuss all cases in a neurovascular MDT when considering treatment options.

**Aneurysm**

- Consider treating both ruptured and un-ruptured aneurysms.
- Be aware that new aneurysms in patients with cerebral aneurysms may develop during childhood.

**Cavernous Malformation (CM)**

- Consider treatment options such as no treatment, surgical resection or SRS in the discussion of the case within the MDT.
- Consider micro-surgical resection or SRS for unruptured lesions that are enlarging on serial imaging.
- Be aware that with current imaging a cure cannot be proven.
8. Discharge from hospital

Discharging or transferring a child or young person from an acute hospital setting to a rehabilitation unit or the community can be a challenging experience for the child or young person and their family. Parents/carers and young people describe leaving hospital as a time of uncertainty and therefore being informed with a clear plan, an understanding of what to expect and from whom, can greatly help this process. Families may lack knowledge about their child’s prognosis; also it may be unclear how long and how well rehabilitation will be managed in the community, and in educational and social settings. Early communication by the acute hospital team with local health professionals, the child’s educational setting and social care services, and including the family in these discussions have been found to be useful.

Members of the local healthcare team, especially from education and social care sectors, may have limited understanding of the child or young person’s long-term needs and disabilities after suffering a stroke. Professionals must be aware that these needs do not always depend on the severity of the stroke.

Stroke can adversely impact on the child or young person’s physical health, and bring about or worsen behavioural, mood, and cognitive issues. As such, assistance may be required to deal with issues relating to the educational setting, family financial issues, and to support the family in general. Early discussions between the acute healthcare team and local teams increase the local team’s understanding of the child or young person’s health and well-being needs and the degree of support required, as an essential part of ensuring an effective discharge and transfer.

Review questions

- What are the elements to consider when planning the discharge of children and young people with stroke from acute hospital care to rehab care and to long-term community care?

8.1. Discharge

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified four studies, which reported data on discharge from hospital, out of which one was a systematic review, another was a cross sectional study based on qualitative interviews, and two were studies based on qualitative interviews. The quality of
evidence was classified as moderate in two studies\textsuperscript{244,247} and low in two studies\textsuperscript{245,246}. Sample size ranged from eight to 350 children.

Study populations were reported from a number of regions including Australia\textsuperscript{247}, America\textsuperscript{244}, the UK\textsuperscript{246} and from a review of studies worldwide\textsuperscript{245}.

A study reporting discharge from hospital was considered by staff as a key point for the exchange of information between health and education professionals; however, this communication was often inadequate\textsuperscript{246}. As reported, the need for improved training for education staff, including raising awareness of the changing needs of children as they develop, was also identified as a key theme for professionals working with children with an acquired brain injury (ABI).

With regard to the needs of families of children and young people with traumatic brain injuries (TBI) and ABI, two studies used interviews and questionnaires to understand experiences of transitioning from hospital back in to a school setting\textsuperscript{244,247}. Glang and colleagues\textsuperscript{244} highlighted that schools rely upon hospital staff and parents to help them to understand what is needed when the child leaves hospital. The link between hospital and school was critical in identifying the need for special education support. Sharp and colleagues\textsuperscript{247} reported that early and ongoing communication between the child and young person's care team, family and school teachers to plan the return to school is needed; the education of teachers and the young person's peers in the academic and social impact of ABI was also found to assist the return to school.

The systematic review by Lindsay and colleagues\textsuperscript{245}, which aimed to gather evidence on the components of effective hospital to school reintegration programs found important types of school reintegration interventions included information provision, problem solving, behavioural and cognitive support, and family support. The common components of interventions which successfully facilitated reintegration included one-on-one sessions provided by a trained clinician or educator, as well as homework activities and parental involvement. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

The evidence found supports the belief that it is beneficial for families to be fully informed and involved at all stages in the child or young person's journey and that the particular needs of each individual is considered.

Through clinical experience and the findings of the parent and young person workshops as detailed in Appendix 7, the guideline development group (GDG) agreed that the time that
a child or young person spends in the hospital environment should be as short as possible and limited to the period when the child or young person is acutely unwell or has outstanding diagnostic issues. Equally, discharge should not occur until a coordinated community care package is in place. This requires assessment, as early as possible and when safe for the child and young person, of the components of their functioning, and the personnel and interventions needed to deliver the necessary care.

Links with the community should be established from diagnosis and may involve one or more multidisciplinary discharge planning meetings. These meetings should include key family members as well as those specifically identified professionals from health (both acute and community), education and social care who will have a continuing role in supporting the child or young person in their ongoing treatment, rehabilitation and reintegration into home and school life. Discussions at these meetings should include the anticipated future needs for support and rehabilitation. A named professional to coordinate the discharge should be allocated and contact details for key professionals given to the young person and parents/carers. Health professionals working in specialist children’s hospitals can have a role in supporting community-based clinicians in providing specialist advice on management and rehabilitation beyond discharge.

From discussions at the parent/carer and young person workshops, it was suggested that a named professional is normally identified to coordinate the discharge plan, and that contact details for key professionals who are to see the child or young person in their next setting (rehabilitation facility or community) are provided. Parents/carers need to understand their child’s condition as fully as possible before going home, as they are the key advocate for their child’s ongoing health and care. A conversation with a medical professional prior to discharge can be helpful in providing the opportunity to discuss any concerns and clarify needs and expectations, and a checklist or action plan can help all involved to monitor progress in preparing for going home and going through the early stages at home and rehabilitation.

With regard to the child or young person’s educational setting, this needs to be prepared in terms of the level of care and any medical action plans. There may need to be adaptations to the environment both at home and at school. Social care involvement for financial (e.g. Carer’s Allowance and Disability Living Allowance (DLA)) and practical support (e.g. respite and help with activities of daily living) may also be required.

It can be helpful for educational professionals to make use of Government resources such as the Special Educational Needs and Disability (SEND) code of practice and Supporting pupils with medical conditions at school, as well as the Royal College of Paediatrics and Child Health (RCPCH) Disability Matters online resource.
Recommendations

- Plan discharge with input from the child or young person and their family and the multidisciplinary team (MDT) (medical, nursing and allied health professionals including education staff, occupational therapists, physiotherapists, orthoptists, psychologists, speech and language therapists) prior to discharge from hospital. If the child has been admitted for an extended period, this may involve more than one meeting and should occur in a timeframe that allows all necessary support to be in place on discharge.

- Plan a discharge meeting as soon as possible after discharge for children and young people who have had a short admission, and include all key family members and specifically identified professionals from health (both acute and community), education and social care who will have a continuing role in supporting the child in their ongoing treatment, rehabilitation and reintegration into home and school life.

- Provide a named key worker or a core group model (such as Team Around the Child/Family (TAC/F)). This can be effective in ensuring that the family has easy, personalised access to appropriate services as required, and is made aware of anticipated timelines and who is accountable for certain actions.
9. Rehabilitation

The clinical sequelae of childhood stroke is highly variable and is influenced by many factors, including lesional, the individual’s age, developmental level and cognitive abilities, pre-injury, family environment, and access to resources. While children and young people do not necessarily recover from stroke better than adults, they may appear to ‘grow into’ their problems as they struggle to acquire new skills. An early, persistent and flexible multidisciplinary rehabilitation is essential.

The International Classification of Functioning, Disability and Health (ICF) framework was introduced earlier within the guideline to define and describe functioning and disability, and to support identification of targets for intervention. Within this biopsychosocial framework functioning, there is an interaction between an individual’s health condition and environmental and personal factors.

These guidelines have used the framework and language to describe the domains of health and functioning relevant to outcome and rehabilitation. In classifying functioning and disability, the framework does not distinguish between health conditions or aetiology. Therefore, the focus in the outcome and rehabilitation sections of the guideline is on the impact on daily life with the recognition that abilities may be impacted by a range of factors, with acquired brain injury (ABI) being one of those factors. The classification could be used to describe both the positive and negative aspects of functioning.

The World Health Organization (WHO) recognises that the functioning and disability of an individual occurs in a context, which is representing in the ICF with the inclusion of a list of environmental factors including equipment, adaptations, educational support, access to community and social life, attitudes, policies, support and relationships.

The spectrum of difficulties that children and young people experience after suffering from either arterial ischaemic stroke (AIS) or haemorrhagic stroke (HS) is broad, with reported deficits in all domains of the ICF. The extent and severity of impact across multiple domains may not be evident in the short term; however, body structures and functions may be affected such as altered tone or weakness, sensory impairments, dysphagia, problems with speech and voice, and mental functions. Activity and participation difficulties have also been reported and include physical problems with mobility, gross and fine motor skills, communication, and learning and applying knowledge.

Child and family-centred care forms the basis for paediatric rehabilitation. Goals should focus on the expressed needs and priorities of the parents/carers or child/young person and framed within the context of participation in daily life roles. The ongoing active
engagement of the child or young person and family can support the achievement of functional outcomes. Consideration of the impact of a stroke on the child/young person, parents/carers and also the wider family and friends is important in building relationships and meeting the needs of the child in the context of their family.

Paediatric rehabilitation following ABI aims to harness opportunities to remodel neuronal connections, to adapt and learn, and to compensate for missing skills\textsuperscript{258}. The environment (physical, social and attitudinal) is a key consideration in identifying factors that may support or impede response to intervention in the short and longer term\textsuperscript{259}.

Childhood stroke occurs within the context of a family and the needs of the whole and extended family must be considered throughout the recovery journey. These needs should be a factor in the planning and delivery of rehabilitation including the demands on the child and family in selection of modality, intensity, environment and location of intervention.

Throughout this chapter the term rehabilitation will encompass both habilitation and rehabilitation (see section 1.1. for a definition of rehabilitation).

**Review questions**

- What is the most appropriate framework for evaluation of rehabilitation needs in children and young people with stroke?
- What are the components and effectiveness of interventions for:
  - motor functions/mobility
  - sensory functions including pain
  - communication, speech and language functions
  - dysphagia
  - mental functions/education/cognition and executive function
  - interpersonal relationships and interactions/psychosocial
  - learning and applying knowledge
  - self-care/independence
  - goal setting
  - mental health
- What are the needs of families and the role of voluntary sector during the planning of care and rehabilitation for children and young people with stroke?
- What environmental factors (equipment, adaptations, educational support, access to community and social life, support and relationships) are most important for stroke patients in rehabilitation and in the long-term?


9.1. **Framework for assessing rehabilitation needs**

**Evidence summary**

A systematic review was conducted for the relevant clinical questions and identified 18 studies\textsuperscript{96,107,253,257,260-273}, which reported frameworks for assessing rehabilitation needs in a child with stroke. Of the 18 studies, nine were cohort studies\textsuperscript{107,253,262,265,268,269,271-273}, three were case series studies\textsuperscript{96,257,266}, three were cross-sectional studies\textsuperscript{260,261,264}, one study was a systematic review\textsuperscript{267}, one study was a case control\textsuperscript{270}, and one study was a randomised controlled trial\textsuperscript{263}. These studies included children with stroke as well as ABI and mixed aetiologies. The quality of evidence was classified as high in two studies\textsuperscript{96,269}, moderate in eight studies\textsuperscript{253,260,262,263,265,266,268,272}, and low in eight studies\textsuperscript{107,257,261,264,267,270,271,273}. Sample size ranged from 18 to 814 children.

Study populations were reported from a number of regions including America (five studies\textsuperscript{261,265,269,270,272}), Australia (three studies\textsuperscript{107,257,260}), Brazil (one study\textsuperscript{263}), Canada (two studies\textsuperscript{268,271}), Europe (four studies\textsuperscript{96,253,264,266}) and the UK (two studies\textsuperscript{262,273}). One study did not specify the study population\textsuperscript{267}.

In relation to stroke specific outcome measures, one study illustrated the use of outcome measures for paediatric stroke\textsuperscript{267}. A wide variety of measures for paediatric stroke have been used, including the Wechsler Intelligence Scale for Children (WISC) and the Paediatric Stroke Outcome Measure (PSOM); however, only the PSOM (a standardised neurological exam/impairment level assessment tool) has been specifically validated with established reliability for infants (less than two years) and children and young people (two to 16 years) with AIS. In comparison, one study concluded that overall the modified Rankin Scale (mRS) was easier to acquire than the PSOM and includes an important measure of cognitive outcome. This corresponds better with the doctor's impression of outcome as well as highlighting function rather than deficit\textsuperscript{264}.

When considering assessment framework, one study investigated the impact of age and environmental factors on cognitive/behavioural outcomes and highlighted which outcome predictors should be taken into account at time of assessment\textsuperscript{260}. Focal insults before the age of three years were associated with poorer outcomes across all domains, with outcome predictors including lesion location and extent, laterality of seizures, and social risk factors such as family function.

One study evaluated readiness for participation in a variety of settings (e.g. home, school and community life) at discharge from inpatient rehabilitation in children with ABI\textsuperscript{261}. The findings emphasised the need to consider the readiness to leave inpatient rehabilitation,
and suggested a robust framework for assessing this readiness prior to discharge as well as a need to establish ongoing rehabilitative input post discharge.

One study highlighted the need for a multidisciplinary team (MDT) approach and the importance of age in predicting outcomes. It was noted that children improved more in self-care, mobility and cognition if they were older than seven years of age, and had lower measures in each specific area at admission, and if they received more occupational therapy, physiotherapy and speech and language therapy (SLT) respectively.

One study identified the most significant risk factors for a poor cognitive outcome in children with AIS (at two years post insult) to be young age, seizures, combined lesion location (cortical and subcortical), and considerable neurological impairment. They advocate for neuropsychological assessment to ensure implementation of appropriate interventions and environmental adjustments as early as possible. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

The guideline development group (GDG) agreed that a framework for the assessment of rehabilitation needs relies on valid and appropriate outcome measures to identify problems across all domains of the ICF and also to assess change during recovery, rehabilitation and growth. The only validated outcome measures for paediatric stroke are the PSOM and Recovery and Recurrence Questionnaire (RRQ); however, these have their own limitations, focus on body functions and do not assess activity and participation. The mRS is a tool developed for use to classify functional disability following adult stroke and has been applied in a small number of child stroke studies; however, it does not have established validity or reliability for use with children or young people and the content has not been devised to consider developmental factors. The wide number and variety of outcome measures that have been applied to children and young people with stroke reflects the wide age and breadth of impact of stroke on domains of health. It may also reflect individual clinician preference and knowledge in the absence of measures specifically validated for use with children and young people with stroke.

The GDG considered that with the domains of the ICF central to practice, tools should be selected based on the domain of health to be evaluated, taking into account preferences and personal circumstances. Consideration should also include awareness of body structures and functions (e.g. the distribution of motor and sensory impairment, brain lesion characteristics, swallowing function). As such, it is recommended that clinicians have a working knowledge of the psychometric properties of measures to guide tool selection in terms of the age and abilities of the child. While there is no evidence to support specific measures of activity or participation in paediatric stroke, reviews of the psychometric
properties and applications of tools used in allied populations may provide a useful reference for clinicians. It is worth noting that some tools have a training validation/purchase cost, whilst others are freely available. This will undoubtedly influence the availability and selection of the tool, and the clinical utility would need to be balanced against availability of the resource.

It was recognised and agreed by GDG consensus that there will be a wide variation in location, environment and intensity of provision; however, the following principles can be applied to a range of settings including universal (community-based) services, district general hospital (DGH), tertiary centres and specialist rehabilitation facilities.

**Recommendations**

- Baseline assessments should be undertaken before initiating any intervention.

- Avoid delay before commencing baseline assessment of functioning. Depending on the child's individual circumstances the initial focus may be on body structures and functions as well as activity and participation. Where possible, use tools with established robust psychometric properties.

- Consider the need for assessment for hearing and vision on an individual basis.

- Consider liaising with and referral to a tertiary centre for advice regarding appropriate assessment frameworks.

- Provide a comprehensive multidisciplinary assessment of needs, taking into account all domains of the ICF, using appropriate measures considering the child or young person and family priorities/preferences as well as the age and developmental stage of the child or young person.

- Consider using quality of life measures to support evaluation of rehabilitation outcomes, and note that tools such as the Canadian Occupational Performance Measure (COPM) or Goal Attainment Scaling (GAS) may assist with identifying individual targets for intervention and evaluating outcome.

- Consider individual factors (developmental abilities and social, family and educational demands) when planning the timing, the intensity and the nature of rehabilitation intervention.
• The MDT should work in active partnership with the child or young person and family in:
  - the formulation and agreement of individualised goals across health domains to develop a unified and coordinated approach across disciplines,
  - goal setting and decision making around intervention plans, and
  - the identification of priorities when considering rehabilitation options.

• Identify a named key worker or key point of contact for families, who will remain a key point of contact through transfer from hospital to community or specialist rehabilitation services, and including starting/re-entering school. This named key worker/contact may vary as appropriate as the child progresses through different life stages.

• Given the wide variation in the evolution of the sequelae following stroke, consider repeat evaluation across domains of the ICF to capture changing needs. This is particularly important at key stages of transition e.g. entering school, moving from primary to secondary school and into tertiary education or the workforce.

### 9.2. General principles for rehabilitation

It is difficult to provide clear recommendations to clinicians regarding the optimal rehabilitation approaches and long-term care of childhood stroke due to lack of strong evidence. Child stroke outcome research suggests the impact of stroke and concerns of families change over time. An individualised approach is indicated, eliciting and acting upon the concerns of young people and their families, and recognising that priorities and needs may change as the child or young person progresses through the life span. At all times the child or young person and family should be central to goal setting and decision-making. In providing rehabilitation there should be an emphasis on developing a positive collaborative working relationship with the child or young person and family in conjunction with delivery of the intervention modality.

Although there is evidence from adult stroke to suggest that more therapy improves the rate of recovery and outcome within the first six months after stroke\textsuperscript{275,276}, the optimal dosage and timing of specific therapies is an emerging area of research\textsuperscript{276}. Chen and colleagues\textsuperscript{265} reported that in a mixed population of children with ABI, who were admitted for rehabilitation, bigger improvements in self-care, mobility and cognition were found in those who started at lower baselines and received more occupational therapy, physical therapy, and SLT.
In the recently published Royal College of Physicians (RCP) national clinical guideline for stroke\textsuperscript{12}, it was recommended that individuals should accumulate at least 45 minutes of each appropriate therapy every day, at a frequency that enables them to meet their rehabilitation goals, and for as long as they are willing and capable of participating and showing measurable benefit from treatment. In children, tolerance of rehabilitation varies according to factors including cognitive, behavioural, communication and motor functioning and developmental age and abilities. Other factors such as fatigue can also impact engagement in intensive intervention. While the GDG agreed that the principle of identifying a target dose is desirable, it is difficult to prescribe this across all age ranges and abilities. Intervention that targets identified areas of priority for the child and family, at a frequency that enables rehabilitation goals to be met is desirable, however there are a number of key considerations. These include the focus on daily life activities and participation (i.e. integration of rehabilitation in the context or home and school life), the coordination and agreement between professionals of intervention targets informed by the priorities of the child or young person and their family, and their willingness and ability to actively participate. The identification of a target dose and intensity for a rehabilitation programme is an important component in prescribing intervention to maximise outcomes.

Key components and general principles to optimise recovery include the following:

- Individualised and patient-centred therapy strategies including consideration of the context in which the intervention takes place.
- Identifying and assessing the rehabilitation needs of the child/young person and understanding that these needs are likely to change over time.
- The practice and frequent repetition of goal-directed, task specific, purposeful activities.
- Holistic multidisciplinary working which includes the child and the family. This MDT should include healthcare professionals from the following domains:
  - medical personnel
  - nursing
  - physiotherapy
  - occupational therapy
  - speech and language therapy
  - dietetics
  - clinical neuropsychology/clinical psychology
  - social work
  - orthoptics
  - play specialist
• There should be access to pharmacy, orthotics, specialist seating, assistive technology and information, advice and support for people with stroke and their family/carers.
• Addressing all dimensions of the child’s health and wellbeing as reflected in the ICF.
• Working towards pre-defined SMART (Specific, Measurable, Agreed, Realistic and Time-bound) goals/principles that have been set with the child/young people and their family and are meaningful and relevant to them.
• The use of outcome measures to measure function, monitor progress and determine change.
• The recognition of the need to support the child/young person and family in adjusting to changed abilities and circumstances.

9.3. **Rehabilitative interventions**

9.3.1. **Motor function and mobility**

Motor deficits are common in children and young people following AIS and HS, with a reported incidence between 50 to 80%\(^5,254,277,278\). Impairments may include muscle weakness and loss of dexterity, disorders of muscle tone, and of quality and coordination of movement, and the distribution of these impairments often varies. Typical presentations include hemiparesis and hemidystonia; however, deficits may not be exclusively unilateral, particularly after HS.

Current rehabilitation of children and young people with motor/mobility impairments follows recognised principles of motor learning, moving away from the treatment of impairments in isolation. Motor learning follows a distributed model of training with variation in type and duration of task practiced to achieve a specific goal, and requires sufficient repetition, intensity and duration\(^279\). However, the optimal dosages of motor training are yet to be established and the evidence for lasting functional gain, and transference of benefit to untrained tasks is variable across domains. In common with any type of rehabilitative intervention, family and parental engagement is key\(^280,281\).

Evidence to support traditional neurodevelopmental therapy (NDT) for paediatric rehabilitation in neurological conditions is weak\(^282\). Motor interventions that may be applicable to child stroke rehabilitation include Constraint Induced Movement Therapy (CIMT), bi-manual therapy, Electromyographic (EMG) triggered neuromuscular stimulation (NMS), functional electrical stimulation (FES), robotic interactive therapy, and virtual reality\(^279,282\).
Evidence summary

A systematic review was conducted and identified 17 studies\textsuperscript{281,283-298}, which reported data on the rehabilitative interventions to improve motor function and mobility. Of the 17 studies, two were systematic reviews\textsuperscript{292,293}, four were randomised-control trials\textsuperscript{285,289,294,296}, three were cohort studies\textsuperscript{287,288,291}, six were case series\textsuperscript{281,284,286,290,295,297}, one was a case-report\textsuperscript{298}, and one was a literature review\textsuperscript{283}. The quality of evidence was classified as high in two studies\textsuperscript{292,293}, moderate in six studies\textsuperscript{281,286,287,289,294,297}, and low in nine studies\textsuperscript{283-285,288,290,291,295,296,298}. Sample size ranged from one to 45 children. Feasibility studies were common amongst novel interventions such as the use of robotics to facilitate reach or walking\textsuperscript{283,288,290}.

Study populations were reported from a number of regions including America (eight studies\textsuperscript{284,285,288,289,294-296}), Canada (one study\textsuperscript{283}), Australia (three studies\textsuperscript{286,292,293}), Europe (four studies\textsuperscript{287,290,291,295}), and the UK (one study\textsuperscript{281}). Few studies included children with AIS or HS only with most presenting a mixed population\textsuperscript{284,294,296}; including children who had unilateral presentations, similar to those typically found in AIS, but also found in children with cerebral palsy (CP) or perinatal stroke. Although the search did not include terms such as CP, the evidence was considered and recommendations were informed by these studies with mixed populations. Eliasson and colleagues\textsuperscript{287} showed that the location of the lesion influences the response to treatment in children with similar presentations. Therefore, whilst the findings from the studies that used mixed populations are valuable they should be interpreted with caution when looking at the children and young people affected by stroke. Further details on each study are provided within the evidence tables, in Appendix 4c.

Evidence to support upper limb interventions

CIMT has the strongest evidence for efficacy of a motor upper limb intervention\textsuperscript{281,285,287,288,294,296}. The use of an arm restraint in isolation was supported by a statistically significant effect size in two studies\textsuperscript{287,296}.

This approach is targeted at individuals with hemiparesis and involves applying a constraint to the less affected arm and hand, and intensive highly repetitive task-based practice with the more affected hand and arm. A protocol for the use of a combination of CIMT with bimanual training has been published, however, the study results are not yet available\textsuperscript{299}. This follows the motor learning principles of task variety and repetition, and builds on studies of combined training in other patient populations.
The use of upper limb robotics to improve reach has weak strength of evidence based on study quality and effect size\(^\text{290,298}\). Fasoli and colleagues\(^\text{288,298}\) demonstrated a positive effect on upper limb coordination and quality of movement, but limited impact on strength and spasticity. Frascareli and colleagues\(^\text{290}\) also reported clinical improvement in reach control and coordination, however were unable to determine which training variables; including timing, intensity, task type and duration; have the greatest impact on recovery.

Bloom and colleagues\(^\text{284}\) demonstrated a positive effect on upper limb use with biofeedback. However, small sample size supports the consideration of biofeedback in conjunction with conventional therapy only. Similarly, Corn and colleagues\(^\text{286}\) investigated the effect of Lycra™ on upper limb function. The sample was characteristic of a small case series, of four children, with mixed results.

Treatment with spasticity modifying agents, specifically Botulinum toxin A (BoNTA), was supported by two studies of moderate\(^\text{289}\) and low quality\(^\text{295}\).

Mirror Therapy has been explored by Gygax and colleagues\(^\text{291}\). Feasibility of this intervention is supported alone and in combination with bimanual training in a mixed group of children with hemiplegia. However, it was a small sample of convenience (10 children) and efficacy of this modality remains unknown.

There is limited but promising evidence of the benefit of Virtual Reality (VR) for improving motor performance and physical activity\(^\text{292}\). The systematic review adds some weight to the benefit of environments to maximize practice opportunity in this way.

Morgan and colleagues\(^\text{293}\) conducted a systematic review and metaanalysis of enriched environments versus standard care for children at risk of developing CP (n=150). The evidence is promising, however, there is insufficient description in the studies of what defines an enriched environment and what defines standard care.

Another study suggested that the new technique of Kinesio Tape used in rehabilitation programs to treat upper-arm/hand pain may be associated with improved upper extremity control and function\(^\text{297}\).

**Evidence to support lower limb and mobility interventions**

Yang and colleagues\(^\text{283}\) supported the feasibility of using intensive lower limb treadmill training (repetitive movement, supported stepping and standing balance activities), in young children (less than two years) with neonatal stroke. The Gross Motor Function Measure-66 (GMFM66) showed big effect sizes suggesting this training is effective at this age.
Linking the evidence to the recommendations

Through experience, the GDG support the use of the ICF to enable clinicians to focus on deficits across domains. Motor interventions need to address not only impairments (motor functions), but also consider activity domains (mobility and gross motor skills) and potentially improve participation. This will help to ensure there is better long-term skill performance, and together with environmental modification and use of equipment will help to ensure transferability across environments into activities of daily living.

There is evidence in allied brain injured populations to support the efficacy of both CIMT and bimanual training in children including combined models281,285,287,294,296; however, there is no clear evidence to support the use of any of the other interventions instead of another in stroke. In practice, it may be dependent on resource availability, clinical experience, or to the personal preference of both the clinician and the child/young person.

Whilst the other interventions with strong positive evidence to support their use in children with CP have not been explored in stroke specifically, the clinical expertise of the GDG supports the consideration of tone management and maintenance of joint range and muscle length. Further support can be gained from adult stroke literature, splinting in neurology, and the National Institute for Health and Care Excellence (NICE) paediatric spasticity guidelines to support splinting, particularly at the ankle in weight bearing, and medical tone management.

When considering CIMT, bimanual training, or a hybrid model as an intervention for children with unilateral motor disorders after paediatric stroke, a range of intervention intensities and types of restrain have been shown to be beneficial; however, clinical experience and published evidence is clear that a specific protocol, either intensive (e.g. 60 hours over two weeks) or distributed practice (e.g. 60 hours over six weeks) and mode of constraint should be clearly defined in advance and monitored along with evaluation of objective outcomes.

Most studies of CIMT and bimanual training are primarily focused on school-aged children with spastic hemiparesis; however, younger children and children presenting with dystonia and/or spasticity should not be excluded. A suggested framework for supporting clinicians in selecting CIMT versus bimanual intervention has been published281. Be aware that intensive training combined with constraint is not appropriate for all children and clinicians should consider that goal directed therapy has been shown to have a clear benefit in other similar populations282.
The use of other emergent therapies such as the use of transcranial magnetic stimulation (TMS) as a primer for repetitive practice, or use of dynamic splints and robot facilitated movements for repetitive practice, only have evidence to support their use in paediatric neurology if used in the context of goal directed training, context focused therapy, or as a component of CIMT and bimanual training. Lycra™ should be considered on a case-by-case basis but has a very weak level of supporting evidence.

**Recommendations**

- Provide rehabilitation that fits within a neurological and developmental framework; individual therapies should complement each other to maximise functional skills.

- Deliver rehabilitation intervention focussed on what the child or young person and family need, want, or are expected to do. Motor interventions should be focussed on functional goals and undertaken with consideration of the whole child and their needs and abilities across all domains of health.

- Time since stroke should not be a barrier for the consideration of intensive training.

- Incorporate into standard care the provision of an environment in which a child lives and develops, and which maximises their potential development and recovery.

- Offer motor skills rehabilitation interventions based on the principles of motor learning with sufficient intensity, repetition and functional relevance to support lasting change.

- Offer each appropriate therapy discipline daily, depending on the age of the child and their tolerance of treatment, at a frequency that enables the child to meet their rehabilitation goals, and for as long as they are willing and capable of participating and showing measurable benefit from treatment.

- Offer children and young people with unilateral presentations either constraint induced movement therapy, bimanual training or hybrid protocols of sufficient intensity where developmental, behavioural and cognitive abilities, and external circumstances including family factors, support engagement with the protocol.
Consider splinting and medical management of disorders of muscle tone, BoNTA in combination with other active treatments. In the upper limb BoNTA is a therapy adjunct and should be prescribed with accompanying task-based occupational therapy intervention. Consider referral to a movement disorder service for advice.

- Consider using muscle strengthening, treadmill training and repetitive gait practice as adjunctive modalities of intervention.

- Consider other interventions alongside standard care, such as virtual reality, enriched environments and biofeedback.

9.3.2. Sensory functions

Childhood stroke can result in a range of sensory impairments, including hearing and vision loss, with vision impairment affecting various functions of central and/or peripheral vision and eye movement disorders. Children and young people can also experience lost or altered sensation of a limb.

Experience of pain may also persist and include headaches, neuromuscular pain, and pain associated with increased tone. Such difficulties may influence movement performance and sensorimotor rehabilitation, and they may also influence body awareness and have implications for safety in functional daily activities.

Evidence summary

A systematic review was conducted and identified no evidence relating to the sensory functions of children and young people following a stroke.

Linking the evidence to the recommendations (considerations)

Due to the lack of evidence describing the sensory functions of children and young people following a stroke, and recognition of the importance of this as an important clinical consideration, the following recommendations are based on consensus of the GDG.

While there is little to guide intervention, it was agreed by consensus that a baseline assessment of the child should be performed. It was noted that the presence of sensory impairments can influence the selection of sensori-motor interventions, and have an impact on rehabilitation outcomes. Interventions that use slow, organised touch, movement or massage can relieve discomfort and could be considered on a case-by-case
basis, in conjunction with more conventional therapy/standard care; however the GDG were not aware of any evidence to support repetitive sensory stimulation.

Assessment and intervention by an orthoptist, optometrist and ophthalmologist supports the wider clinical professions to understand the impact and changing presentation of visual deficits, and justifies the use of specialist resources which may be available to support children with visual impairment. Similarly, assessment by an audiologist should be considered on an individual basis. The GDG were conscious that sensory issues may impact on rehabilitation outcomes.

It is worth noting that pain tools, validated for other paediatric client groups, may be useful in assessing and managing pain in children and young people following a stroke. Children and young people may not be able to verbalise that they are in pain; however, it can have a powerful negative effect on their ability to participate in daily life or any form of therapy.

**Recommendations**

- Consider the presence of sensory impairments and pain, and integrate this into the planning and implementing of rehabilitation.

- Assess vision and hearing as part of the multidisciplinary assessment.

- Consider that an individual’s sensory functions (e.g. hearing and vision) may change over time and therefore require reassessment.

- Be aware that children with sensory impairments may benefit from specialist support services, e.g. vision impairment teacher support and hearing impairment support.

- Consider the impact of sensory impairments at a functional level and how they may impact on a child’s participation.

- Be aware of sensory impairments when selecting the most appropriate interventions for other domains, such as sensorimotor interventions.

- Be aware that tactile stimulation can be used in conjunction with task based motor intervention to support a young person’s orientation to an affected limb with altered sensation.
• Treat all pain actively, using appropriate measures including positioning, handling and medication.

• Refer children and young people with intractable pain to health professionals with specialist expertise in pain management.

9.3.3. Dysphagia

There is wide variation in eating, drinking and swallowing outcomes in childhood stroke after the acute phase. Most children and young people will have no limitations in this area beyond the acute phase; however, children and young people with a severe motor disability caused by stroke may also have significant difficulties with drinking, chewing and swallowing.

Evidence summary

A systematic review was conducted and identified three studies\(^{110,300,301}\), two of which reported data on the management of dysphagia\(^{110,300}\) and one which reviewed dysphagia interventions\(^{301}\). Of these studies, one was a case series\(^{110}\), one was a systematic review\(^{301}\) and one was a non-randomised trial\(^{300}\). The quality of evidence was classified as high in one study\(^ {301}\), moderate in one study\(^ {300}\), and low in one study\(^ {110}\). Sample size ranged from three to 30 children.

Study populations were reported from Australia (two studies\(^ {110,301}\)) and America (one study\(^ {300}\)).

Describing acute dysphagia following traumatic brain injury (TBI), not including stroke, one study\(^ {110}\) highlighted that eating, drinking and swallowing were influenced by motor and cognitive status, as well as the complementary roles of both clinical and instrumental investigations in assessment.

One study detailed a pilot study on the effects of carbonated liquids compared with non-carbonated liquids on swallow function in ABI\(^ {300}\). The small number of stroke participants showed no change on the intervention, however, the numbers were too small to draw conclusions.

Another study, a systematic review of dysphagia intervention, included a qualitative synthesis of two studies including children with CP and one study of myotonic dystrophy type one\(^ {301}\). Two interventions focused on oral sensori-motor programmes and one on lip strengthening. These studies have very limited applicability to the stroke population. Further details on each study are provided within the evidence tables, in Appendix 4c.
Linking the evidence to the recommendations

Due to the limited evidence describing dysphagia in children and young people following a stroke, the recommendations proposed are largely based on the consensus of the GDG, with due recognition of the importance of managing dysphagia after adult stroke on clinical outcomes. Intervention studies that target only oral motor skills have limited application to children with dysphagia.

In clinical practice, management of dysphagia typically involves multiple professionals and addresses multiple aspects of eating, drinking and swallowing simultaneously. Interventions include positioning and postural management, modification of food and fluid textures, feeding techniques and behavioural strategies to reduce risk of aspiration, improve eating and drinking efficiency, promote the development of oral motor skills and address emotional and behavioural issues that impact on family functioning.

Good quality studies of multi-component interventions, focusing on important outcomes such as growth and weight gain, nutritional status, respiratory health and the impact of activity limitations on the mealtime experience of children and their caregivers are lacking across neurodisability. However, there is widespread agreement by professional bodies and Government departments that multidisciplinary management of swallowing disorders is good practice.\textsuperscript{302,303} Interventions developed for children with CP and other neurological conditions are relevant to children with severe functional disabilities following stroke.

Most areas in the UK provide specialist SLT and dietetics services for children with dysphagia, although not all have dedicated MDTs. Access to instrumental investigations such as videofluoroscopy (VF) is variable. Administration and interpretation of VF in children requires specific paediatric competencies.\textsuperscript{303} Increasing the number of radiology departments that can provide multidisciplinary paediatric VF would have resource implications.

Recommendations

- Be aware that eating, drinking and swallowing difficulties (dysphagia) can result from stroke in the short or longer term, particularly in children and young people with severe motor and cognitive disabilities.

- Refer for dietetic assessment and advice if a child is failing to follow expected patterns of growth and weight gain (over or underweight) or showing signs of nutritional compromise.
- Refer for SLT assessment and advice if parents/carers have concerns about coughing or choking on eating and drinking, frequent chest infections, or failure to move through the typical stages of eating and drinking development.

- Use SLT assessment as first line of investigation.

- Provide access to VF with a specialist paediatric team, including speech and language therapist and radiographer, if there is uncertainty about swallow safety after clinical assessment.

- Include health and nutritional status, developmental skills, and parent/carer reported activity and participation in measures of outcome.

- Offer access to specialist multidisciplinary feeding services when children and young people have complex dysphagia (including when there is consideration of non-oral feeding).

- Provide a coordinated approach to management of eating, drinking and swallowing, with collaboration between families, medical and allied health professionals, teachers and other members of the Team Around the Child/Family (TAC/F).

- Refer to a community paediatrician for consideration of medical/surgical interventions when there are parent/carer concerns regarding drooling.

### 9.3.4. Communication, speech and language functions

There is a wide spectrum of speech, language and communication outcomes in childhood stroke, from children and young people with very subtle difficulties to children and young people with very limited language understanding or spoken language.

**Evidence summary**

While a systematic review was conducted and identified no studies for communication, speech and language interventions specific to stroke, one case series study\(^{304}\) reporting data on three children from the UK with TBI and speech dysarthria (motor speech disorder) was identified. The quality of evidence was classified as low.
This study suggested the use of electropalatography (EPG) improved intelligibility on parent-reported measures\textsuperscript{304}. The study size, diagnoses of the participants and lack of established reliability of the outcome measures significantly limits the application of the study to the stroke population. The use of EPG in the treatment of dysarthria is rare in UK clinical practice.

Receptive or expressive aphasia, dysarthria, dysfluency and dysphonia have been described in studies of outcomes of childhood stroke. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Due to the lack of evidence describing communication, speech and language interventions of children and young people following a stroke, the recommendations in this section are based on the GDG experience.

Speech and language therapists typically differentiate between speech, language and communication functioning. While speech disorders can reflect motor impairments or voice disorders, language is a cognitive function which is also influenced by social skills. Language skills may be described in terms of syntax (grammar), semantics (vocabulary and using language to express specific meanings) and pragmatics (use of language in interaction). Communication however is a broader concept, encompassing the use of speech, language and non-verbal skills for social interaction.

Standardised assessments to explore the different aspects of communication functioning are often used by speech and language therapists, with these then being devised into individualised programmes in order to address impairments and activity limitations either through direct therapy or through training a child’s or young person’s communication partners in the use of strategies to support functional communication. In the absence of stroke-specific programmes, speech, language and communication interventions from other areas of clinical practice such as learning disability, specific language impairment, autistic spectrum disorders, or CP may be applicable depending on a child’s individual profile.

In children with apparently good speech, language and communication skills, high level processing difficulties (such as understanding inferential or idiomatic language) have been noted after stroke, but it is unclear whether these are responsive to intervention has yet to be explored.

Augmentative and Alternative Communication (AAC), including the use of signing, pictures, graphic symbols, and speech-generating devices, is well-established clinical
practice in the UK for children and young people with significant difficulties in receptive or expressive language. Robust intervention studies are lacking across the field, with none specific to stroke. In practice, interventions are largely tailored to each individual’s functional speech, language and communication profile rather than aetiology.

Community and school-based speech and language therapists may lack specific experience of childhood stroke. Knowledge and therapy techniques from other clinical groups are likely to be appropriate. Access to speech and language therapists in tertiary services may be required to guide intervention in children with complex speech, language, and communication disorders related to childhood stroke.

Recommendations

• Be aware that children and young people who have apparently unaffected language skills may have high level language processing difficulties that will impact on educational performance, communication and socialisation/social participation.

• Offer neuropsychological assessment (by educational, clinical or neuropsychologist) for children and young people when starting or returning to school/not meeting their attainment targets. Refer for more detailed SLT assessment, including the use of formal testing, where there are specific concerns about speech, language or communication limitations.

• Be aware that a child or young person’s needs may evolve or change over time necessitating reassessment and review of any statutory supports in place, such as the Education and Health Care Plan (EHCP).

• Offer referral to SLT when there are parental or professional concerns about communication skills, language understanding, expressive language or poor intelligibility due to persisting motor speech disorders (dysarthria and dyspraxia), dysfluency or voice disorders.

• Use standardised assessments of speech, language and communication functioning, alongside measures of activity and participation, to establish a baseline prior to intervention, and to evaluate the impact of interventions.

• Outcome measures should include parent/carer and school report as well as clinical or instrumental findings.
• Offer referral to AAC services where children and young people have significantly impaired language understanding and/or expressive speech/language that are contributing to activity and participation limitations, such as Communication Matters, where information on UK-wide AAC assessment services (including Specialist Commissioning in England) can be found (see http://www.communicationmatters.org.uk/page/assessment-services).

• Provide an individualised, coordinated approach to management of speech and language (and communication) difficulties, with collaboration between families, allied health professionals, teachers and other members of the team around the child.

9.3.5. Cognition

Children and young people who have had a stroke will frequently present with problems of cognitive function that have the potential to impact upon social, educational and occupational achievement. Prognosis for cognitive and educational recovery is poor, particularly in younger children, and often requires long-term monitoring.

Evidence summary

While a systematic review was conducted and identified seven studies the majority of the children included in these studies had not suffered a stroke, consequently limiting the applicability of these studies to the childhood stroke population. Of the seven studies, four were randomised control trials and three were systematic reviews. The quality of evidence was classified as high in three studies, moderate in three studies, and low in one study. Sample size ranged from 11 to 87 children.

Study populations were reported from a number of regions including America (one study), Brazil (two studies), China (one study), Europe (two studies) and the UK (one study).

The evidence for interventions improving general cognitive functioning is generally weak. One systematic review found an improvement in attention problems, after TBI in children, through cognitive/behavioural training and the use of medication. This highlighted that cognitive training appears beneficial when targeting attention and memory specifically rather than in combination with other behavioural/cognitive interventions.
A second systematic review provided details of psychological interventions aimed at alleviating cognitive difficulties in children with ABI, and concluded that while high quality evidence for effective interventions for outcomes (including attention, memory and learning difficulties) is limited, there was evidence to support that interventions can alleviate internalising symptoms (e.g. depression, anxiety, withdrawal and in parent–child interactions) was available for psychosocial outcomes\textsuperscript{311}.

There is evidence that memory and cooperative learning are improved through teaching metacognitive techniques (e.g. reasoning, decision-making, and ability to show insight and awareness)\textsuperscript{280,309}. One study described the problem-solving training approach, which emphasises metacognitive principles, led to improvements in goal-directed functional behaviour\textsuperscript{309}.

One randomised controlled trial (RCT) highlighted that cooperative learning helped children with ABI develop metacognitive strategies and improve self-concept\textsuperscript{280}. While instruments such as the WISC are not designed to measure specific cognitive areas, as it is an IQ test, this study showed improvements in WISC scores over a year-long intervention\textsuperscript{280}. Another RCT, by the same author, demonstrated general cognitive improvement, as well as functional independence, in children who had received indirect family-support rehabilitation rather than clinician delivered therapy\textsuperscript{263}.

Despite this, one systematic review found that there was an absence of evidence and no effect on attention or executive function from cognitive training programmes. However, the effect on aspects of memory was more positive\textsuperscript{310}. Such findings were supported by other studies\textsuperscript{312}. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Despite the lack of evidence specific to childhood stroke, the GDG considered that the findings of the studies identified did have some relevance to stroke in children and young people. As such the recommendations are based both on the literature described and clinical experience.

It is potentially unrealistic to expect rehabilitation to show demonstrable improvement on general measures of cognitive function such as IQ scales; however, while evidence of generalisation appears limited, cognitive rehabilitation programmes may be able to produce improvement in specific areas of cognitive function, such as attention. It may well be that basic cognitive functions such as attention, can be directly re/trained, while more complex cognitive functions require teaching of techniques to remediate for the acquired cognitive difficulties. Teaching of metacognitive/thinking skills appears to have more
potential benefit in terms of complex cognitive functions, and there is evidence for promoting active engagement of parents in rehabilitation programmes.

A significant issue in the delivery of cognitive rehabilitation strategies is which profession should provide the input, and at present there is typically some input for children and young people within educational settings. There is limited delivery of cognitive rehabilitation to children from the National Health Service (NHS), beyond advice to parents and children given by neuropsychologists within tertiary hospitals. Access to neuropsychological expertise is typically limited to regional paediatric neurology services.

**Recommendations**

- Provide neuropsychological assessment and advice to schools and affected families throughout formal education.

- Be aware that a child’s need for intervention related to cognition is likely to change according to demands, and particularly at transition points, e.g. from primary to secondary education.

- Train and involve parents/carers of children who have suffered stroke in delivery of interventions to support cognitive functioning in their child’s daily life activities.

- Teaching staff and allied health professionals should teach metacognitive skills, methods encouraging the ability of the child/young person to problem solve within the home, school and community.

- Consider education for the child/young person and their family on the impact of identified cognitive weakness on daily life activities and appropriate compensatory strategies.

- Consider skills training in a functional context to improve daily life abilities impacted by cognitive impairment.

**9.3.6. Mental health**

The impact of ABI on mental health function is well established\(^3\)\(_{13}\), with recent studies focusing on the family and social factors which mitigate against the development of mental health difficulties\(^3\)\(_{14}\). Clinical experience also suggests that fatigue can impact daily life activities in both the short and long-term after stroke diagnosis.
Evidence summary

A systematic review was conducted and identified four studies reporting data on rehabilitative interventions to improve mental health in children after a stroke. All of the studies were randomised controlled trials and from populations in the United States. The quality of evidence was classified as high in one study, moderate in two studies, and low in one study. Sample size ranged from 17 to 20 children.

The studies showed the emerging evidence which demonstrates that direct training of cognitive functions produces some improvements in mental health presentation. There are also now indications that demonstrably successful programmes used in paediatric mental health services (Problem-Solving approaches, Cognitive Behavioural Therapy, Positive Parenting, Acceptance & Commitment Therapy (ACT)) can be adapted to be used with families where a child has an acquired brain injury. Such programmes include the I-Interact programme for training positive parenting which shows some promising effects. Two studies by Wade and colleagues reported the effects of internet based interventions in children and teenagers. These studies showed increased parental understanding of their child’s condition and achievement of set goals in the intervention groups. Furthermore, behavioural problems decreased in both the child and teenage participants with younger participants reporting that they felt supported and understood. Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

The GDG considered that there is an issue across the UK about who takes responsibility for providing mental health support to children with brain injuries and their families. Often local services such as child and young people’s mental health services will decline to offer interventions and refer back to tertiary services. This perception that only experts in brain injury can offer mental health interventions to this population needs to be challenged, as the evidence reviewed suggests that, with some adaptations, commonly used interventions can be effective.

Recommendations

- Refer children, young people and their families to local children and young people’s mental health services or paediatric psychology services within hospitals for psychotherapeutic interventions.

- Tertiary services such as regional neuropsychology or paediatric psychology services should train, supervise and support local services.
• Treat behavioural difficulties with adaptations to existing parenting programmes such as Triple P, Signpost and ACT.

• Consider the use of technology to support and deliver family interventions as part of a package of individualised treatment, rather than only as a method to provide information.

• Develop acquired brain injury specific adaptations to support local children and young people’s mental health services to provide appropriate input.

• Consider individualised Cognitive Behavioural Therapy (CBT) based approaches for treatment of mood related problems.

• Consider the presence and impact of fatigue on daily life abilities and mental health. Strategies including graded activities to balance demands across the day.

• Consider the benefits of pharmacological treatment in conjunction with other treatment.

9.3.7. Interpersonal relationships and interactions/psychosocial (social relationships)

Children with all forms of ABI have the potential to experience significant disruption to their social relationships\textsuperscript{318}.

Evidence summary

A systematic review was conducted and identified five studies\textsuperscript{280,309,311,317,319}, which reported data on interpersonal relationships and interactions. Of the five studies, three were randomised control trials\textsuperscript{280,309,317}, one was a cohort study\textsuperscript{319} and one was a systematic review\textsuperscript{311}. The quality of evidence was classified as high in two studies\textsuperscript{280,311}, moderate in two studies\textsuperscript{309,319}, and low in one study\textsuperscript{317}. Sample size ranged from 11 to 20 children.

Study populations were reported from a number of regions including Australia (one study\textsuperscript{319}), America (one study\textsuperscript{317}), Brazil (one study\textsuperscript{280}), China (one study\textsuperscript{309}) and the UK (one study\textsuperscript{311}).

There is emerging evidence that teaching metacognitive skills can improve aspects of social interactions. While two of the identified studies showed improvements on self-
report\textsuperscript{280,309}, this has not been demonstrated by improvements on teacher or parent ratings of improvements in social relationships.

More recently, there has been positivity around the use of the internet to foster social participation. One study\textsuperscript{319} found improvements in children with disabilities through supported use of social media. Another study\textsuperscript{317} suggested that an online cognitive-behavioural approach can improve child adjustment after TBI, with Ross and colleagues\textsuperscript{311} highlighting that children of lower socio-economic status and those injured older than 11 years old may benefit most from online family intervention. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

The recommendations within this section were formed as a result of combining clinical experience within the GDG and the literature reviewed.

The evidence for effective psychosocial benefits remains modest and suggests that delivering rehabilitation in the appropriate setting is important, within the home for family interventions and within school for peer interventions. Access to appropriate packages for the delivery of programmes of psychosocial rehabilitation is limited in both mental health and educational services. As with cognitive rehabilitation, the resources for developing such provision and the professional time required in direct therapy and training of others is very limited.

Teaching and training delivered via the internet appears to be a viable approach for many families, and the supported use of social media is increasingly important for children and young people.

**Recommendations**

- Refer children, young people and families to psychology services when there are concerns about social relationships.

- Tertiary services such as regional neuropsychology or paediatric psychology services should train, supervise and support local services.

- Include parent/carer, child/young person, and teacher reports using standardised questionnaires in assessment and monitoring of family and peer relationships.
• Involve the family in interventions aimed at improving social relationships between parent/carer and child/young person.

• Involve peers in interventions aimed at improving peer relationships.

• Consider teaching metacognitive strategies to support improvement in social interactions.

• Consider individual tuition to help children and young people to use the internet safely and effectively for social interaction. Train and support families to be confident in supervising their child's use of social media.

• Promote the development of acquired brain injury specific training packages within schools, to include development of social relationships.

9.3.8. Learning and applying knowledge

ABI is known to have a significant impact on a child or young person’s cognitive abilities, and as such has the potential to impact on an individual's academic achievement\textsuperscript{320-322}.

Evidence summary

A systematic review was conducted and identified no studies which directly pertained to the learning, application of knowledge and teaching of children and young people following a stroke.

Linking the evidence to the recommendations

Due to the lack of evidence describing the learning, application of knowledge and teaching of children and young people following a stroke, the following recommendations are based on the clinical experience and expertise of the GDG. The recommendations are reliant on the extensive experience and work on using Precision Teaching/Direct Instruction in children and young people with learning difficulties, and around support to the educational sector.

Precision teaching with Direct Instruction has been established as an effective method of teaching in children and young people with general and specific learning difficulties and is based on sound theoretical principles including errorless learning, repetition and planned generalisation. As such, through agreement by consensus the GDG used these principles when forming the recommendations.
The application of Precision Teaching and Direct Instruction is likely to involve increased demand on teaching staff and teaching assistants. It may be necessary to support families in using statutory legislation to enable the child to access additional teaching resources.\textsuperscript{251,323}

**Recommendations**

- **Teach factual knowledge (e.g. word reading, maths facts) through Precision Teaching with Direct Instruction.** Direct instruction refers to systematic scripted lesson plans. Use the principles of Precision Teaching which is a well-established method of teaching involving high levels of repetition of specific material e.g. high frequency words, typically involving daily assessment of progress.

- **Provide a Special Educational Needs and Disabilities Co-ordinator (SENCo) or equivalent to act as a keyworker/named coordinator once the child is attending school.** This individual should liaise with parents/carers and professionals as per the Special Educational Needs and Disability (SEND) code of practice: 0 to 25 years (Children & Families Act (2014))\textsuperscript{323}, Department for Education and Department of Health (2015)\textsuperscript{251}.

- **Treating hospital and community-based therapeutic staff should collaborate with the SENCo to ensure that interventions are communicated, appropriately planned and delivered.**

- **Health professionals should provide regular consultation to educators, including both advice and brain injury training.** This should be with a professional with experience of both education and acquired brain injury.

- **Be aware that a child or young person's needs may evolve or change over time necessitating reassessment and review of any statuary supports in place e.g. the EHCP.**

**9.3.9. Self-care/independence**

Children and young people may have a reduced ability to carry out self-care tasks as a result of motor, sensory and cognitive impairments. Such activities may include dressing, bathing, toileting and feeding, and the ability to move around the home or school environment, play and access the school curriculum.
Self-care is important to children, young people and families\textsuperscript{324}, and depending on the age of the child or young person, along with their family and cultural environment, the affected individual will have established a level of independence pre-injury. Self-care is age dependent and a lower age has been found to be indicative of greater difficulty in self-care in children affected by stroke\textsuperscript{108}.

**Evidence summary**

While the systematic review found no specific literature on the effect of interventions to address childhood stroke on self-care/independence, the review identified one cohort study\textsuperscript{325} which reported data on a structured cognitive approach, known as Cognitive Orientation to Daily Occupational Performance (CO-OP) showing significant improvements of functional ability in a population of six children and young people from Canada. The quality of evidence was classified as moderate.

This study reported that a structured CO-OP had a significant positive change in functional ability in children with a mixed ABI aetiology\textsuperscript{325}. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Children and young people's ability in self-care and independence will undoubtedly be affected by other impairments such as sensory, cognitive, motor, behavioural challenges, fatigue, as well as by age. There is a body of knowledge which includes age-related self-care and independence in children and young people with brain injury resulting from aetiologies other than stroke that it will be important to consider in order to manage expectations accurately.

While the evidence was limited, the GDG considered that children and young people may seek assistance with personal care tasks so as to conserve energy for school work. Individuals may have a preference of who supports them with such tasks, with some adolescents may prefer professional support rather than being bathed by a parent, while others may only wish a parent/family member to assist. It is therefore imperative to seek individual views and tools, such as the COPM and GAS, as these may help to facilitate a child or young person and family focussed approach.

In terms of supporting self-care training, while it is not described in the evidence, there is a body of knowledge regarding generic training techniques such as task analysis, backward chaining and repeated practice.

CO-OP is an emerging occupational therapy intervention approach that appears to be transferable to a variety of patient groups, and would seem appropriate for children and
young people with stroke. It is worth noting that both training and formal accreditation is required for CO-OP at time of publication, and that training is available at a cost.

Gaining skills in self-care and independence can provide children and young people with additional benefits. These may include improved mood, self-efficacy, engagement with peers and social interactions. The GDG support this, along with the role of the occupational therapist in goal directed, functional training for self-care and independence.

Recommendations

- Assess the child’s ability to perform self-care tasks, household tasks, tasks in major life areas such as school, play, and community life.
- Involve an occupational therapist in provision of intervention in this area if difficulties are identified.
- Work in partnership with child, parent/carer and school staff.
- Offer child and family-centred care.
- Be aware of developmental norms for self-care tasks, household tasks, tasks in major life areas such as school and play and community life.
- Consider goal directed, functional training with home programmes where appropriate.
- Use specific learning techniques and repeated practice in context.
- Consider structured approaches to intervention such as CO-OP.
- Use standardised tools such as COPM and GAS to prioritise and evaluate self-care and independence interventions.

9.3.10. Goal setting

Goal setting with children, young people and families is encouraged in rehabilitation following childhood stroke and collaborative goal setting is considered an essential component of family-centred care, and is a key standard of the National Service Framework: children, young people and maternity services. Centres that deliver commissioned rehabilitation in England should carry out goal setting as a requirement,
using a particular type of goal setting known as GAS which can be used as an outcome measure\textsuperscript{327}.

**Evidence summary**

A systematic review was conducted and identified one randomised control trial\textsuperscript{324}, which reported data on goal setting in a population of 26 children from Australia. The quality of evidence was classified as low.

The study found that goal setting tools such as Perceived Efficacy in Goal Setting (PEGS) and COPM can be useful for children with stroke and their families in order to highlight areas of concern across a number of domains\textsuperscript{324}. Further details on each study are provided within the evidence tables, in Appendix 4c.

**Linking the evidence to the recommendations**

Despite the limited evidence on goal setting, the GDG acknowledge that not only does communication of goals and priorities to the team around the child or young person help to support the coordination of an intervention plan, but goal setting is an important area of intervention and teams should take time to develop goal setting skills.

Children and young people may present with a range of difficulties and there may be number of competing priorities which can be overwhelming for children, young people and their families. It is helpful to know personal priorities in order to help focus intervention and decide on the pacing of the interventions.

Further to this, the GDG agreed by consensus that personal goals and priorities may change over time. It may be useful to use formulation tools in order to support the team around the child in defining areas of concern and outlining potential goals to discuss with the family.

While PEGS and COPM were deemed useful tools\textsuperscript{324}, there are other tools available, such as GAS and Child Occupation Self-Assessment, which may also be of use. It is worth noting that while GAS is freely available at the time of publication, other resources are commercially available with some at a cost.

It is to be noted that there is a wide body of goal setting literature in allied fields, such as education, that identify principles of goal setting; however, goals should be written following SMART principles.
Recommendations

- Discuss areas of functional difficulty and intervention priorities with children, young people and families.

- Create goals/principles which follow the general principles of being SMART (Specific, Measurable, Agreed, Realistic and Time-bound).

- Consider using goal setting tools, such as PEGS, COPM and GAS.

- Agree and coordinate the timing and delivery of interventions across the MDT.

- Health and education professionals should define interventions, associated goals and accountability.

- Communicate goals in a range of formats to the child/young person, wider family, carers, school and other professionals.

- Review goals and priorities at least annually. This should be done with the child/young person and their family and the health and education professionals.

9.4. The needs of the family during the planning of care/rehabilitation

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified While the systematic review identified no evidence to address the question in relation to the needs of the family during the planning of care and rehabilitation, there was evidence classified in terms of which environmental factors are the most important for stroke patients in rehabilitation and in the long-term.

The systematic review identified 14 studies outlining which environmental factors are the most important for stroke patients in rehabilitation and in the long-term. Of the 14 studies, three were randomised control trials and one was non-randomised, three were systematic reviews, two were cross sectional studies, two were based on interviews, two were cohort studies, and one was a descriptive case study. The quality of evidence was classified as high in three
studies\(^{293,331,335}\), moderate in six studies\(^{315,329,333,336,337}\), and low in five studies\(^{246,324,328,330,332}\). Sample size ranged from eight to 427.

Study populations were reported from a number of regions including America (five studies\(^{315,334-337}\), Australia (two studies\(^{293,324}\)), Russia (one study\(^{328}\)), Sweden (one study\(^{329}\)), Canada (two studies\(^{330,332}\)) and the UK (two studies\(^{246,331}\)). While one study did not specify the study population\(^{333}\).

While the studies identified were not specific to children with stroke (e.g. covering children with TBI\(^{315,328,333,334}\), ABI\(^{246,329,331,332}\), CP\(^{293}\), physical disabilities\(^{330}\), as well as families/carers\(^{324,335}\). These studies highlight the need for support with both emotional and psychological issues\(^{328}\), as well as the need for education and information about the brain and the consequences of a brain injury for children, young people and families\(^{332}\), using telehealth if appropriate\(^{333}\).

Participation in everyday life may be reduced following a stroke in childhood for a number of reasons, including demographics and the age of child or young person. One study\(^{330}\) of children with neurodisability identified that participation was lower in children over 12 years and families with a lower income, while another study\(^{331}\) suggested that children with TBI are more likely to have reduced Quality of Life (QoL), specifically with difficulties in cognitive fatigue and behavioural problems, and be prone developing persistent clinical problems.

In addition to supporting children, young people, and families during care planning and long-term support, one study\(^{293}\) suggested that the promotion of an enriched environment, such as parent training or coaching in parent-infant interaction or in various stimulation activities, can enhance the outcome of motor intervention for a group of children with CP. Whilst this is a different population to that in question within this guideline, the GDG agreed by consensus that the motor interventions would be analogous for stroke. Another study demonstrated the importance of family involvement in psychological interventions\(^{337}\).

While training and coaching in parent interactions has been suggested in addition to education regarding modifying and adapting the environment, one study\(^{328}\) concluded that two complementary directions have been defined in the psychological rehabilitation of children with a brain injury: emotional and psychological support to the child or young person from the parents, and psychological work with the child or young person whose state of consciousness has been seriously altered. Another study\(^{336}\) suggested that parents preferred web-based coaching in comparison to other more traditional interventions. Further details on each study are provided within the evidence tables, in Appendix 4c.
Linking the evidence to the recommendations

A timely discharge from a rehabilitation setting may be delayed and children may take longer to return to home if adaptations are required. If children have a short admission to hospital or rehabilitation there may not be time to initiate a thorough discharge planning. The GDG agreed by consensus that not only may providers be reluctant to issue costly equipment if the child or young person is still making changes during the recovery phase, but that cultural issues in healthcare and educational settings could present attitudinal barriers.

From evidence generated by the parent/carer and young person workshops it was suggest that having a documented plan of care, with named professionals, is important. The focus of the intervention should be on rehabilitation and maximising outcome(s), not on how to live with the consequences or compensate for changed abilities.

The provision of repeated opportunities to access support, particularly counselling, is helpful as the needs of young people and family member’s changes over time. Routine follow-up that considers information and support needs as a component of rehabilitation practice is suggested, and guidance about how to navigating the health, social care and education systems is often needed by parents and carers. Where a child is school-aged at the time of the stroke it is important that the school maintains contact with the child or young person and family. For young people leaving school and entering employment, support on how to access work opportunities is needed.

Rehabilitation should be centred on what the child or young person needs and what they would like to achieve. This, in conjunction with an understanding by health professionals that stroke can impact the entire family, helps a family to feel supported.

Recommendations

- Inform, as relevant for the individual child or young person and family, the potential or actual role of health, education and social care systems in providing support and care. Include information and education about assessment processes.

- Communicate the priorities of the child, young person and family to health care education and social care professions.

- Consider the impact of stroke on the health, social and economic wellbeing of family members and make onward referrals as necessary to support the
broader family.

- Refer children and young people to the Children with Disabilities social care team within the local authority. This should be a part of a MDT plan, and discharging clinical teams should be responsible for initiating the referral.

- Discuss and disseminate clinically age-appropriate information/correspondence with parents/carers and the child/young person. Consider the use of technology (e.g. emails, apps and websites).

- Provide regular opportunities for the child or young person and family to access support from professionals from health, education and social care as needed; this should include (with parent/child or young person consent) communication between care agencies including the family and child or young person and documented integrated planning.

- Provide school with age-appropriate information about stroke with the consent of the parent/carer and child/young person (see the associated parent/carer guideline).

- Be aware of other resources that may provide guidance (e.g. Children and Families Act 2014, Supporting Pupils with Medical Conditions at school and the local authority ‘Local Offer’).

- Consider education resources such as the Disability Matters online resource to increase disability awareness in the community and wider society.

- Provide children and young people with access to additional resources within the education sector according to individual need, such as an educational psychologist, SENCo, and social support.

- Assess physical, social, academic, attitudinal and environmental factors that may impact on the child/young person with stroke.

- Consider the role of the charitable and voluntary sectors in ongoing support and care. This may include independent advocacy for a young person and family.
10. Long-term care: transfer and transition

As mentioned in Chapter 8, members of the local healthcare team, especially education and social care sectors, may have limited understanding of the child or young person’s long-term needs after a stroke. Professionals must be aware that the degree of disability and long-term need does not always depend on the severity of childhood stroke.

Stroke can adversely impact on a child or young person’s physical health and bring about or worsen behavioural, mood, and cognitive issues. As such, assistance may be required to deal with issues relating to educational settings, family financial issues and to support the family in general. Early discussions between the acute healthcare team and local teams should aim to increase local team understanding of the child or young person’s disability and the degree of support required as an essential part of ensuring an effective discharge and transfer.

Transition from paediatric to adult services can be equally challenging for the young person and their family. Moving into adult healthcare services requires clear planning involving all members of the team, including the child or young person and their family.

Transition also has additional issues such as dealing with the young person’s further education or employment, housing, money management, relationships and mental health. During transition, the young person must be given the opportunity to voice their opinion about their future and to take responsibility and set goals for their future, as appropriate.

Primary care professionals, particularly general practitioners (GP), have a key role in coordinating and supporting joined up care across health education and care sectors. The GP should be actively engaged in the process of transition planning for a young person moving from child to adult healthcare services.

Voluntary and charitable organisations, both locally and nationally, may have a role to play in discharge into the community and all life’s transitions. Reference to the National Institute for Health and Care Excellence (NICE) guidelines about transition from children’s to adult services for young people using health or social care services is recommended.

Review questions

- What is the most effective way of managing educational and social care transfer through various educational stages (nursery, primary and secondary school, college/work) for children and young people after stroke?
• For young people who had stroke, how should transfer to adult healthcare be managed?

10.1. Managing educational and social-care transition

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified two studies\(^{339,340}\) which reported data on the transition of children and young people from school to employment. Of these studies, one was a prospective longitudinal study\(^{339}\) and the other was a qualitative study\(^{340}\). The quality of evidence was classified as high in one study\(^{340}\) and moderate in the other study\(^{339}\), with sample size ranging from 14 to 200 children.

Study populations were reported from Australia (one study\(^{339}\)) and America (one study\(^{340}\)), with the latter comprising white American (74%), African-American (13%) and Hispanic (11%) ethnicities with 2% of the study population classified as ‘other’.

Parents of children and young people with acquired brain injury (ABI) have reported high levels of stress related to their child’s education and future employment prospects coupled with a lack of information and support regarding post-school rehabilitation and employment services\(^{339}\). Similarly, many young people reported a lack of support when dealing with both current and future work and school related issues.

Wehman and colleagues\(^{340}\) highlighted that almost three quarters of young people with a traumatic brain injury (TBI) had been employed after high school and employment had been achieved if the young person had had goals to transition into work after secondary education. Furthermore, active student participation in transition planning and the support of outside individuals and organisations was recommended. Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

The identified studies\(^{339,340}\) confirmed that good cooperation and communication at all stages, and between all the necessary agencies, are key factors in identifying needs and improving transition through education stages and into adult life. Equipping the individual and their family with information, support and coping strategies can improve outcomes.

While the evidence did not reveal specific measures that should be adopted, goal setting was supported. The limited evidence identified was considered in combination with the findings of the parent/carer and young people engagement workshops (see Appendix 7).
The guideline development group (GDG) discussed options on how to support schools in identifying subtle cognitive deficits in children with stroke. It was agreed by consensus that there is different provision of educational psychologist input and school support throughout the UK. Generic documents relating to supporting children with educational and medical needs throughout school and on exiting education and other children’s services should be consulted or referred to.

**Recommendations**

- Ensure regular, effective collaboration and communication between the child, young person and family and health, education, and social care professionals throughout the child’s schooling to identify and respond to their specific needs and disabilities. This can include meetings, joint assessments and sharing of relevant knowledge and skills to optimise and personalise the provision of learning support.

- Ensure health and education professionals have access to information about child stroke.

- Establish channels of communication between school and the family from school entry/re-entry.

- Consider the individual communication needs of young people after brain injury.

- Be aware that children and young people with stroke may require a flexible, holistic, integrated approach in supporting them, ranging from targeted therapy or educational interventions for particular difficulties, to a comprehensive Education, Health and Care Plan (EHCP).

- Integrate therapy interventions into the child’s educational provision where possible, to minimise school absence and promote inclusion.

- Ensure the early, active participation of the young person in planning their transition from school to higher education or work.

- The creation of a long-term condition passport can support information sharing and reduce repetition.
• Provide the young person with appropriate support in planning for adult life and building life skills in the context of their health condition.

• Consider the provision of a named key worker to support the young person and family in transitioning into and through education.

10.2. The transition of a young person into adult health care

Evidence summary

A systematic review was conducted for the relevant clinical questions and identified two studies\(^{88,341}\) which reported data on the transition of a young person into adult healthcare. Of these studies, one was a qualitative study\(^{341}\) and one was a cross-sectional survey\(^{88}\). The quality of evidence was classified as high in one study\(^{341}\) and low in the other study\(^{88}\), with sample sizes ranging from 30 (of which 15 were classified as young people) to 71 children. Study populations were reported from Canada (one study\(^{341}\)) and America (one study\(^{88}\)).

Transition from paediatric to adult services can be a difficult time for any adolescent with a chronic illness or disability. Not only will the individual need to assume some primary responsibility for their condition (depending on cognitive ability) but they will also need to cope with a change of care providers. One of the greatest reported concerns is that new healthcare providers may not understand their individual needs. As described in one of the studies\(^{88}\), a transition clinic which is run alongside an adult clinic, where the families can meet professionals from adult services, may lead to improved feelings regarding transition and help with the general transitioning process.

A lack of information about the process, lack of trust regarding the knowledge and ability of adult healthcare providers, feelings of uncertainty and a need for greater support are all factors that have been identified as important factors during the transition to adult healthcare services\(^{341}\). Further details on each study are provided within the evidence tables, in Appendix 4c.

Linking the evidence to the recommendations

The limited evidence identified was considered in combination with the findings of the parent/carer and young people engagement workshops (see Appendix 7).

The GDG agreed by consensus that transition can be a positive experience to both service users and providers if the barriers identified in the transition process are addressed\(^{88,341}\).
While the identified studies provided no specific evidence around transition of a young person into adult health care, it is understood that functional difficulties experienced by the young person, as a consequence of stroke, may become more problematic as they become increasingly independent and the higher level of support sometimes available to children is withdrawn. Consideration therefore needs to be made for the functional difficulties that may be less obvious, including social, higher level executive functioning, psychological and language difficulties. Emotional difficulties associated with the expectations of adulthood (e.g. relationships) may be apparent, particularly if there is inadequate planning and support.

**Recommendations**

- Children with sickle cell disease (SCD) on long-term transfusion for prevention of stroke should be referred to an adult unit where transfusion therapy can continue to be provided and support is given to continue transfusion during and after the transitional period.

- Be aware that higher levels of support may be needed during the transition to adulthood and adult services, and that paediatric and adult stroke services should have a written protocol for transfer.

- Consult the National Institute for Health and Care Excellence (NICE) guideline on ‘Transition from children’s to adults’ services for young people using health or social care services’.

- Inform young people and their parents/carers about the professionals involved in future management and how to gain access to them.

- Ensure clear delineation of roles and responsibilities through face-to-face meetings during the transition process.

- Identify one named key worker either from social care, health or education who can be the main point of contact for the young person and family during the transition process.

- Provide clear written information about the professionals involved, their role and the transition process.

- Be aware of the issues a young person recovering from stroke may face. This
includes health related issues, mobility and self-help skill difficulties, neuro-behavioural issues, mental health issues, educational difficulties, and issues relating to reintegration with social life.

- Transition between previous and future care givers must be discussed with the young person and their family in a dedicated transition session, either in a health or education setting, or in the young person’s house.

- Be aware that there may be the need to meet multiple times to ensure both young person and their family clearly understand the transitional process and to address the issues that the young person may face.
11. Implications for practice

11.1. Facilitators and barriers

Within the context of current National Health Service (NHS) pathways for children affected by stroke many of the recommendations outlined within this guideline are aspirational and will require additional financial, workforce and organisational resources at both national and local levels. These will likely be needed at all stages of the care pathway from additional training or materials to raise awareness of childhood stroke symptoms, to increased rapid access to diagnostic imaging and specialist clinicians, and support and advocacy services for stroke survivors and their families/carers. A major consideration in implementing these recommendations is the balance between the rarity of childhood stroke, the high frequency of ‘stroke mimics’, and the relative complexity of the care pathways described. Inevitably resource limitations will have an impact on some of the goals that are set out. It is anticipated that this guideline will set the standards for clinical care, and in turn will increase the impetus to improve resource availability.

The organisation of services at secondary and tertiary centres varies from hospital to hospital within the NHS. Rather than proposing a ‘one size fits all’ model, the guideline development group (GDG) emphasise that it is useful to identify and exploit existing areas of relevant clinical expertise, such as within paediatric intensive care (PIC) transport teams, adult acute stroke services or community paediatric services, and to define local deliverable operational policy, as the pathways discussed will involve interaction between multiple clinical teams and centres. It is important that all stakeholders have the opportunity to ratify local arrangements. It may also be helpful to interface with other existing clinical networks, such as paediatric neuroscience or haemoglobinopathy.

Important areas for training across all healthcare and education sectors include recognition of the clinical features of childhood stroke, the necessity of seeking urgent medical advice and activating the relevant clinical pathway. It is also important that public awareness of childhood stroke is improved and children and young people should be included in public health campaigns aimed at increasing awareness of stroke in general.

While many implications highlighted in the 2016 Royal College of Physicians (RCP) national clinical guideline for stroke\(^\text{1}\) remain true for this guideline, the following factors are felt to potentially act as facilitators or barriers to implementation of this guideline.
### Training and education

<table>
<thead>
<tr>
<th>Facilitators</th>
<th>Barriers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Through an increase in education, an improved awareness of childhood stroke in trainees and qualified personnel (in medicine, nursing, allied health, education and social services) can be achieved.</td>
<td>There is a limited awareness of the treatment options available to children experiencing an arterial ischaemic stroke (AIS).</td>
</tr>
<tr>
<td>Knowledge of the neurorehabilitation of paediatric stroke patients could draw on the evidence from allied populations, such as children with acquired brain injury.</td>
<td>Limited awareness of the urgent imaging requirements of suspected paediatric stroke patients and the necessity of urgent imaging for a number of the treatment modalities.</td>
</tr>
<tr>
<td>Sub-specialty interest groups, such as the British Paediatric Neurology Association (BPNA) Cerebrovascular Special Interest Group, could assist in the development of clinical networks and facilitate the sharing of knowledge/expertise on childhood stroke.</td>
<td>Health professionals will need to be made aware of the novel nature of the pathways presented in this guideline.</td>
</tr>
<tr>
<td>An increased awareness of childhood stroke among both healthcare professionals and the public could be achieved via the Stroke Association, and other organisations/charities endorsing this guideline. Incorporation of children in public health campaigns to improve awareness of stroke as a medical emergency.</td>
<td>Training of allied health professionals to recognise the changing and emerging needs that are seen after childhood stroke.</td>
</tr>
<tr>
<td>Linking with the Paediatric Intensive Care Audit Network (PICANet), for example, could utilise existing data capture networks.</td>
<td>Training and assessment tools for childhood stroke require development and validation, as they are not currently comprehensive.</td>
</tr>
<tr>
<td>Training and education</td>
<td>Need for training in taking consent from patients and their families for novel AIS treatments, e.g. IV tissue plasminogen activator (tPA) should be provided to health professionals.</td>
</tr>
</tbody>
</table>
| Awareness should be raised of the applicability of FAST (‘Face, Arms, Speech
and paediatrics/health professionals should be trained in its use for children and young people.

**Financial**

<table>
<thead>
<tr>
<th>Facilitators</th>
<th>Barriers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Published tools such as FAST, paediatric National Institute of Health Stroke Scale (PedNIHSS) and Goal Attainment Scaling (GAS) are freely available to healthcare professionals, and awareness of these published tools should be promoted.</td>
<td>Necessary funding to extend established treatments in new populations is needed, e.g. IV tPA/thrombectomy for paediatric AIS.</td>
</tr>
<tr>
<td></td>
<td>There is a lack of funding for rapid paediatric neuroimaging.</td>
</tr>
<tr>
<td></td>
<td>Funding is needed for training materials and releasing staff to attend relevant training.</td>
</tr>
<tr>
<td></td>
<td>Funding is a major issue for paediatric neurorehabilitation (e.g. in terms of training, access to assessment tools, equipment and residential or specialist services).</td>
</tr>
</tbody>
</table>

**Resource**

<table>
<thead>
<tr>
<th>Facilitators</th>
<th>Barriers</th>
</tr>
</thead>
<tbody>
<tr>
<td>There is an opportunity to learn from the structure and organisation of adult stroke services in the UK, and apply these to stroke services for children and young people.</td>
<td>There is a limited availability of rapid transfer resources.</td>
</tr>
<tr>
<td>The emergence of paediatric emergency medicine as a sub-specialty has the ability to act as a facilitator to delivering hyperacute pathways.</td>
<td>There is a limited availability of acute care beds.</td>
</tr>
<tr>
<td>Development of specialised paediatric neurovascular services will generate the multidisciplinary neurovascular teams proposed.</td>
<td>Standardisation of access to specialist services across the UK, including paediatric neuroscience centres, is needed.</td>
</tr>
<tr>
<td>There is an opportunity to engage with</td>
<td>Fragmentation of multidisciplinary ongoing</td>
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</tbody>
</table>
There is an opportunity to promote and disseminate this guideline via various social media channels.

There is currently a lack of licences for therapies in paediatric patients.

11.2. Implementation tools and advice

To help implement this guideline, a number of tools and resources have been developed by the Royal College of Paediatrics and Child Health (RCPCH):

Full guideline
- hosted and available freely on the RCPCH and Stroke Association websites

Parent/carer guideline
- hosted and available freely on the RCPCH and Stroke Association websites
- provides a summarised lay version of the guideline
- aims to provide parents/carers with information and sign-post them to relevant support agencies for organisations working with children who have had a stroke and their families

Quick reference guide
- a concise list of the key recommendations to help busy clinicians find clinical guidance easily in order to guide those responsible for commissioning and evaluating those services

Pre-hospital and in-hospital algorithms for the management of suspected childhood stroke
- hosted and available freely on the RCPCH and Stroke Association websites
- provides a downloadable user-friendly algorithm poster to support and aid guideline implementation

To ensure this guideline, and accompanying materials, reach the end user all confirmed stakeholders will be approached for direct publication on their website, or link to the RCPCH site.
To help implement this guideline, the following suggestions have been made by the GDG:

- The guideline should be read by all healthcare professionals who are involved in the care of a child who is suspected of having had a stroke, or where the diagnosis has been confirmed, from pre-hospital to tertiary care settings.
- The guideline should be read by adult stroke professionals who may be asked to provide advice or support to staff involved in the care of a child or young person who is suspected of having had a stroke or where the diagnosis has been confirmed.
- Clear local multidisciplinary guidelines for implementation are needed and should be developed.
- Local written protocols need to be maintained and updated regularly. The care setting needs to be defined, along with arrangements for networking between secondary and tertiary centres.
- Training in the use of the following assessment tools should be provided:
  - FAST (www.stroke.org.uk/FAST)
  - PedNIHSS (http://stroke.ahajournals.org/content/42/3/613)
  - Clinical investigation (see Diagrams 4.1 and 4.2)
  - Specific medication and/or intervention protocols (e.g. aspirin, thrombolysis, thrombectomy and decompressive surgery - see 6.2.1 and 6.2.2).

11.3. Guideline audit

National audit will be critical to evaluate the implementation and efficacy of the recommendations proposed within the guideline.

Experience from the RCP national stroke programme suggests that a robust national audit process such as the Sentinel Stroke National Audit Programme (SSNAP) will make it more likely that the recommendations of the RCPCH guideline have an effective impact on the experience of children and young people with stroke and their families. As such, an audit would ideally cover the whole breadth of the care pathway from pre-hospital alert to rehabilitation.

The existence of an adult stroke audit programme presents an opportunity to mirror an already successfully established programme, or even to combine efforts in auditing adult and paediatric stroke services. Such an audit could provide evidence for a number of key areas where evidence is missing, in particular for the treatment of AIS.

Such an audit will need to be taken forward by the relevant stakeholders and appropriate funding will need to be identified to set up and maintain the collection of audit data. As a
minimum, it is suggested that a compulsory registry of all childhood stroke cases be established to collect data on novel treatments and their outcomes.

The audit process could be considered in three stages:

1. An organisational audit of services in each provider service with links to regional systems of care
2. A snapshot audit of a group of unselected cases
3. A National individual patient audit of all cases (this should build on the RCP SSNAP\textsuperscript{342}).

Below is a list of suggested criteria on which the implementation of this guideline could be audited.

### Audit points

#### Point 1: Organisational

<table>
<thead>
<tr>
<th>Description</th>
<th>信任应概述他们为支持转诊和治疗路径所拥有的资源。</th>
</tr>
</thead>
</table>
| Data items |资源类型记录：
- 顾问及其专业性质
- 儿童医疗士
- 放射科医生（包括进入神经放射学的专科医生）
- 护理
- 疗法
- 营养师
- 职业治疗师
- 物理治疗师
- 心理学家
- 语言治疗师
- 一个书面的急性儿科中风路径 |

#### Point 2: Patient information

<table>
<thead>
<tr>
<th>Description</th>
<th>应记录家庭是否在初次入院时获得了书面信息</th>
</tr>
</thead>
</table>
| Data items | 提供信息：
- 儿童中风？
- 法定和志愿机构？
- 文档化出院计划在出院前与儿童/年轻人及其家人讨论过吗？
- 指定联系人，供家庭/看护者/儿童/年轻人在有问题时联系？ |
**Point 3: Clinical**

**Description**  
Acute presentation/diagnosis of risk factors (e.g. SCD, cardiac and prothrombotic)

**Data items**
- What is the interval between symptom onset and arrival in hospital?  
- What was the time from admission to cross sectional neuroimaging?  
- What was the time of image transfer to the regional paediatric neuroscience centre?  
- What was the PedNIHSS score pre-treatment and after a defined period post-treatment?  
- What was the interval between presentation and consultation with a multidisciplinary clinical team?  
- What is the interval between presentation and transfer to regional paediatric neurosciences unit?  
- What is the involvement of the PIC transport team?  
- What is the time from arrival at hospital to first brain scan?  
- Was computerised tomography angiography (CTA) or magnetic resonance angiogram (MRA) performed at the time of the first brain scan?

**Point 4: Patient – Intravenous (IV) thrombolysis**

**Description**  
IV thrombolysis patients

**Data items**
- What was the time from onset to door at the secondary receiving centre?  
- What was the time from door to computerised tomography (CT) scan?  
- What was the time between symptom onset and admission to needle time for IV tPA?  
- Were there any complications?  
- Was the case discussed retrospectively in a neurovascular multidisciplinary team (MDT)/involvement of adult stroke service?

**Point 5: Malformations**

**Description**  
Vascular malformations (arteriovenous malformations (AVM)/aneurysm/cavernous malformation)

**Data items**
- Was the case discussed in a neurovascular MDT?  
- What elements were present in the MDT?  
- What treatment modalities were considered/offered?

**Point 6: Rehabilitation**

**Description**  
Longer-term care and rehabilitation

**Data items**
- Is the child under the care of a consultant paediatrician?  
  - If so, have they been seen in the last 12 months?  
- Was a meeting held between professionals in
<table>
<thead>
<tr>
<th>Question</th>
<th>Answer</th>
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</thead>
<tbody>
<tr>
<td>Health/education services and the child’s parents prior to return to or</td>
<td>- If so, has this been reviewed in the last 12 months?</td>
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<td>commencement of school?</td>
<td>• Was the family provided with the details of community/specialist</td>
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<td></td>
<td>• Was a rehabilitation plan documented?</td>
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<td></td>
<td>• Were rehabilitation goals documented at the beginning of intervention</td>
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<td></td>
<td>• Was a key named contact provided for the child, young person and</td>
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<td>• Were rehabilitation goals agreed with input from parents/teachers to</td>
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<td></td>
<td>• Further outcomes, such as:</td>
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12. Research recommendations

It will be apparent in reading this guideline and as highlighted in Professor Rudd’s foreword that the evidence base underpinning guidance on the management of childhood stroke remains sparse and almost all areas discussed in this document are ripe for well-designed research. A major issue in this field is the incidence of stroke, which signifies that research will need to have a multi-centre design in order to provide meaningful conclusions.

The International Paediatric Stroke Study\textsuperscript{343} consortium is an international collaboration of childhood stroke researchers and has been key to providing large datasets for analysis and, more recently, the infrastructure to support well designed adequately powered studies. Within the UK, the British Paediatric Neurology Association (BPNA) has recently formed a cerebrovascular special interest group that aims to improve clinical and research networks. This group, and special interest groups in other professional organisations, will be key to rolling out the acute care recommendations outlined here.

The collapse of the Thrombolysis in Pediatric Stroke (TIPS) trial\textsuperscript{7} illustrated the challenges of childhood stroke research; even with robust funding and infrastructure. It seems unlikely that another hyperacute treatment trial will be funded in the near future. Given the recommendations herein regarding hyperacute treatment, centralised data collection on children assessed for and receiving such treatment is strongly recommended so that this data can be systematically evaluated.

While this guideline has emphasised the differences in stroke affecting children and adults, there are clear overlaps and potential opportunities for collaboration. This guideline already flags the opportunity to improve awareness of childhood stroke in FAST (‘Face, Arms, Speech Time’) campaigns. In research terms there are potential studies or trials where it would be relevant to recruit children or adolescents. In particular, the recommendations taken from the Royal College of Physicians (RCP) national clinical guideline for stroke\textsuperscript{12} should be considered to be tested in children.

It is now recognised that research priorities must involve those affected by the condition, as well as professionals. Thus, many areas for potential research are apparent within the scope of this guideline and the guideline development group (GDG) recommend that the key research priorities in paediatric stroke should be identified within a priority setting partnership exercise involving professionals, parents and children and young people. Such an exercise would lend weight to the importance of the priority areas identified and provide a uniform focus between funders and researchers.
The GDG identified the following areas in which more research is needed and has made some specific recommendations for future studies to feed into guidelines and improve patient care.

- **Networks, collaborations and registries in furthering research into stroke in childhood.**
  - Use multi-centre collaborations to explore the longer-term impact of stroke and interventions to address deficits in children and young people is needed.
  - Establish what the key areas for future research should be with key stakeholder groups (e.g. childhood stroke survivors, families, clinicians, allied health professionals).

- **Training and assessment tool development and validation.**
  - Develop and assess standardised training materials for non-clinical professionals who have frequent contact with children to recognise the signs and symptoms of stroke, such as teachers.
  - Validate standardised assessments in the paediatric population, e.g. FAST.

- **Stratification of the importance of known risk factors.**
  - Investigate the additional risk that specific known factors may bring to the baseline risk of paediatric stroke. Most studies describe the risk factors present in children who have already had a stroke. By studying the numbers of children with known risk factors that go on to have a stroke, risk factors could be ranked and any differences between the risks for arterial ischaemic stroke (AIS) and haemorrhagic stroke (HS) could be established.
  - Investigate the risk of future haemorrhage in vascular malformations with a view to creating a method of estimating risk in individual patients (e.g. algorithm) to inform risk:benefit ratio considerations of different intervention strategies.

- **Diagnostic value of signs and symptoms**
  - Establish the frequency with which the signs and symptoms of paediatric stroke laid out in the guideline are associated with a stroke diagnosis and with other non-stroke diagnoses.
  - Establish the predictive power of signs/symptoms (and combinations thereof) of paediatric stroke.

- **Imaging effectiveness and timing**
  - Review, systematically describe, and critically compare the different imaging modalities for the detection and follow-up of AIS and HS.
• **Effectiveness of treatments and therapies**
  - Assess the proposed pathway's ability to identify children who would benefit from hyperacute thrombolysis and facilitate their timely treatment.
  - Investigate whether antithrombotic therapy is associated with better outcomes and limited brain injury.
  - Compare the effectiveness of exchange versus simple transfusion for the secondary prevention of AIS in children with SCD.
  - Compare the long-term outcomes of children with aneurysms treated by clipping or coiling the aneurysm.
  - Investigate the impact of heritable thrombophilia mutations on mortality, morbidity and long-term outcomes.

• **Frequency and nature of complications**
  - Review the complications that children experience following AIS and HS, such as medical, physical, emotional, or social.

• **Rehabilitation needs of paediatric stroke patients**
  - Characterise the deficits in sensory functioning that children experience following a stroke.
  - Characterise the presence and impact of fatigue after stroke on daily life and assess the effectiveness of strategies to support children impacted by fatigue.
  - Assess the effectiveness of rehabilitation interventions specifically in children affected by stroke.
  - Develop and trial multi-component interventions for dysphagia following paediatric stroke that measure outcomes such as growth, weight gain, nutritional status, respiratory health, and the impact of activity limitations on mealtime experience of children and caregivers.
  - Develop and trial communication, speech, and language interventions specifically in children who have suffered a stroke which include a component which helps the child/young person to understand the change in their ability to communicate and adjust to this loss or change. Such interventions should also train family and peer group members to be aware of and adapt to these changes.
  - Investigate the acute functional difficulties that children experience following a stroke and the most appropriate tools to use to identify these difficulties.
  - Investigate and identify the optimal dosage and modality of rehabilitation interventions, in line with the emerging evidence of motor, social, behaviour and communication sequelae following stroke.
• **Long-term outcomes**
  - Assess the long-term outcomes regarding education, employment, independence, and support needs. Explore the relationship between outcome and the support the young person received during earlier stages of transition.
References


