Full recommendations for childhood stroke

This document contains the full recommendations identified by the Royal College of Paediatrics and Child Health (RCPCH) Stroke in Childhood Guideline Development Group (GDG), which, if followed, will enhance the quality of stroke care in children and young people (aged 29 days to 18 years at time of presentation).

These recommendations have been extracted from the 2017 RCPCH Stroke in Childhood clinical guideline, which contains individual recommendations covering the diagnosis, management and rehabilitation of stroke in children and young people.

This document should not be read in isolation, and individuals should always consider the guideline in full. To access the main guideline, please visit 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.

- Do not apply the Recognition of Stroke in the Emergency Room (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 15 or AVPU ('Alert, Voice, Pain, Unresponsive') less than A) 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.

Diagnosis (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. This should include:
  - computerised tomography angiography (CTA) (covering aortic arch to vertex), 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.
- Consider primary imaging using magnetic resonance imaging (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 magnetic resonance angiogram (MRA) at the time of undertaking MRI; this should cover the aortic arch to vertex in arterial ischaemic stroke (AIS) and can be limited to the intracranial circulation in HS.

Referral pathways and further investigations (Chapter 4)

Referral and care pathway for childhood stroke (Chapter 4.1)

- 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. 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 paediatric intensive care (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.

Acute management (chapter 5)

Acute assessment (Chapter 5.1)

- 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 GCS or AVPU 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.

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.

- 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 International Classification of Functioning, Disability and Health (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.

Prevention, identification and management of complications (Chapter 5.3)

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

Complication	Detection	Management
Swallow dysfunction	• swallow safety for eating, drinking and saliva control should be assessed, ideally by a trained professional using validated screening methods. Monitor for aspiration	• refer to speech and language therapist if concern about impaired swallow safety
Aspiration	• if aspiration is considered likely undertake clinical assessment, monitor oxygen saturations, blood gases and obtain chest X ray	• consider the placement of a nasogastric tube • facilitate regular position changes in bed and early mobilisation • ongoing management of eating, drinking and swallow
Raised intracranial pressure	• provide regular clinical assessment sensitive to 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	• follow national and local guidelines for 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 CT or MRI will be required	• urgent referral to neurosurgical unit
Seizures	• assess clinically rather than routinely with electroencephalogram (EEG)	• seizures should be managed according to local and national treatment

	except when clinical assessment is not possible in paralysed and sedated patients or when distinction between seizure and movement disorder is not possible on clinical grounds	protocols • 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 paediatric dietitian
Deep vein thrombosis	• risk assessment including level of mobility, weight, 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

Arterial Ischaemic Stroke (AIS) (Chapter 6)

Risk factors for AIS and recurrent AIS (Chapter 6.1.1)

Risk factors for first AIS

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

Risk Category	Included factors/diagnoses
Arteriopathy	• focal cerebral arteriopathy of childhood • moyamoya • arterial dissection • central nervous system (CNS) vasculitis
Cardiac disease	• congenital cardiac disease • 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
Cardiac surgery/interventions	
Sickle Cell Disease	Additional factors in children and young people with SCD: • genotype (sickle haemoglobin (HbS) & HbSβ thalassaemia more than other genotypes) • abnormal transcranial Doppler studies • arteriopathy (intracranial & extracranial)

	• absence of alpha thalassaemia trait • acute anaemia • silent infarction • prior transient ischaemic attack (TIA) • high systolic blood pressure, acute chest syndrome • anaemia, high reticulocyte count
Infection	• 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.

Investigations to identify risk factors in AIS (Chapter 6.1.2)

- 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 CTA or 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 Varicella zoster virus (VZV)), immunisation, dysmorphic features, neurocutaneous stigmata, autoimmune disease and evidence of vascular disease in other organ systems.

Follow-up imaging in AIS (Chapter 6.1.3)

- 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.

- 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.

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.

- 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 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/young people with sickle cell disease (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.

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.

- 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 human leukocyte antigen (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 hematopoietic stem cell transplantation (HSCT).
- Monitor children with regular neurocognitive testing, MRI and transcranial doppler ultrasonography (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 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.

Silent cerebral infarctions (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. 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.

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 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.

Haemorrhagic Stroke (HS) (Chapter 7)

Risk factors for HS and recurrent HS (Chapter 7.1.1)

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:

Risk Category	Included factors/diagnoses
Vascular disorders	• arteriovenous malformation (AVM), especially with arterial phase aneurysms, varicosities or venous stenoses on the draining veins • cavernous malformations, especially Zabramski type 1 & 2 • cerebral arterial aneurysms • moyamoya
Clotting disorders	• severe platelet disorders/low platelet count • all severe inherited bleeding disorders • anticoagulation • severe vitamin K deficiency
Sickle Cell Disease	
Illicit drug use	• amphetamines • cocaine
Gender/ethnicity/age	• age 15 to 19 years • black ethnicity • male gender

- 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. Intranidal 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.

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

- 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 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 CTA or MRA, as well as formal CA

- 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).

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.

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

AVM	**Treatment and frequency**
Endovascular treatment	• CA to be performed at three to six months after obliteration
Surgical resection	• MRI and MRA and catheter angiogram to be performed at three to six months after resection
Stereotactic radiosurgery (SRS)	• MRI and MRA to be performed two years following treatment - if appears obliterated on MRI confirm with catheter angiogram - if still apparent, MRI/MRA & CA at three years to evaluate for further treatment
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.

Acute medical interventions for HS (Chapter 7.2.1)

- 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.

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 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 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 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 surgery or alternative-donor HSCT.

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 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

- 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 stereotactic radiosurgery (SRS) for unruptured lesions that are enlarging on serial imaging.
- Be aware that with current imaging a cure cannot be proven.

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.
- 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.

Rehabilitation (chapter 9)

Framework for assessing rehabilitation needs (Chapter 9.1)

- 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.

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, 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, Botulinum toxin A (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.

Sensory functions

- 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.

Dysphagia

- 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 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.
- Use SLT assessment as first line of investigation.
- Provide access to videofluoroscopy (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 TAC/F.
- Refer to a community paediatrician for consideration of medical/surgical interventions when there are parent/carer concerns regarding drooling.

Communication, speech and language functions

- 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 Augmentative and Alternative Communication (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.

Cognition

- 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.

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.

- 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 Acceptance & Commitment Therapy (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.

Interpersonal relationships and interactions/psychosocial (social relationships)

- 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.

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 (Children & Families Act (2014), Department for Education and Department of Health (2015)).
- 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.

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.
- 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 Cognitive Orientation to Daily Occupational Performance (CO-OP).
- Use standardised tools such as COPM and GAS to prioritise and evaluate self-care and independence interventions.

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).
- Consider using goal setting tools, such as Perceived Efficacy in Goal Setting (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.

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.
- 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 parent/carer version produced alongside this guideline).
- Be aware of other resources that may provide guidance (e.g. Children and Families Act 2014, Supporting Pupils with Medical Conditions at chool 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.

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.
- 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 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.

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

- Children with 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' (NG43).
- 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.