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Stroke association

Implications of 2017 Childhood Stroke guidelines for children with Sickle Cell Disease

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PATRON HRH The Princess Royal

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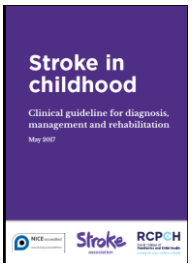
Outline & Background

- 2004 guidelines; parent role in making case
- Process – working party / scope
- Current Practice - NICE SCD guidelines; how children with stk are managed;
- The new guidelines
  - Typical pathway – screening onwards

Key differences c/w 2004

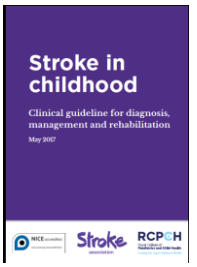


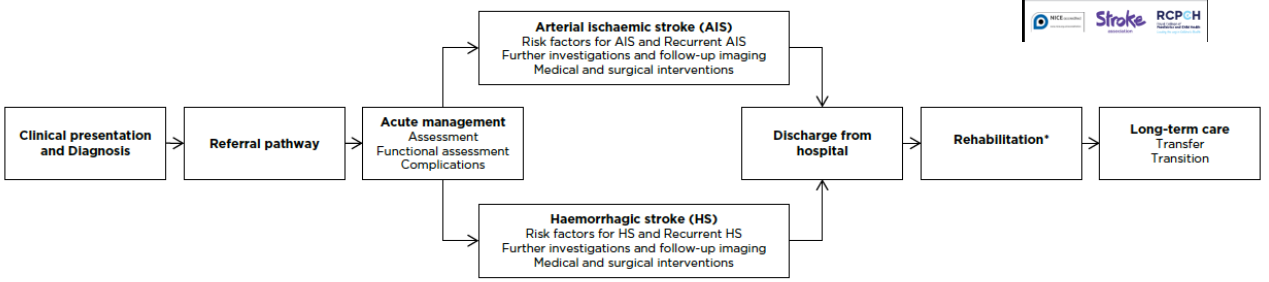
- Includes intracranial haemorrhage
- Delphi method used to reach consensus on contentious issues
- Recommendations for hyperacute AIS therapies
- Covers patient journey form pre-hospital phase to transition to adult services
- Recommendation that cases are managed by specialist neurovascular MDT convened at regional level

ASSOCIATION OF AMBULANCE CHIEF EXECUTIVES
NASMeD National Ambulance Service Medical Directors
BACCH British Association for Community Child Health
BACD British Academy of Childhood Disability
BAGP British Association of General Paediatrics
BRITISH ASSOCIATION OF STROKE PHYSICIANS BASP
BPNA British Paediatric Neurology Association
British Society of Paediatric Radiology
College of Occupational Therapists
PICS Paediatric Intensive Care Society
The Royal College of Emergency Medicine
Royal College of Nursing
Royal College of Physicians
ROYAL COLLEGE OF SPEECH & LANGUAGE THERAPISTS
SICKLE CELL & Young Stroke Survivors
UK Forum on Haemoglobin Disorders
London Ambulance Service NHS NHS Trust

The Role of Parents

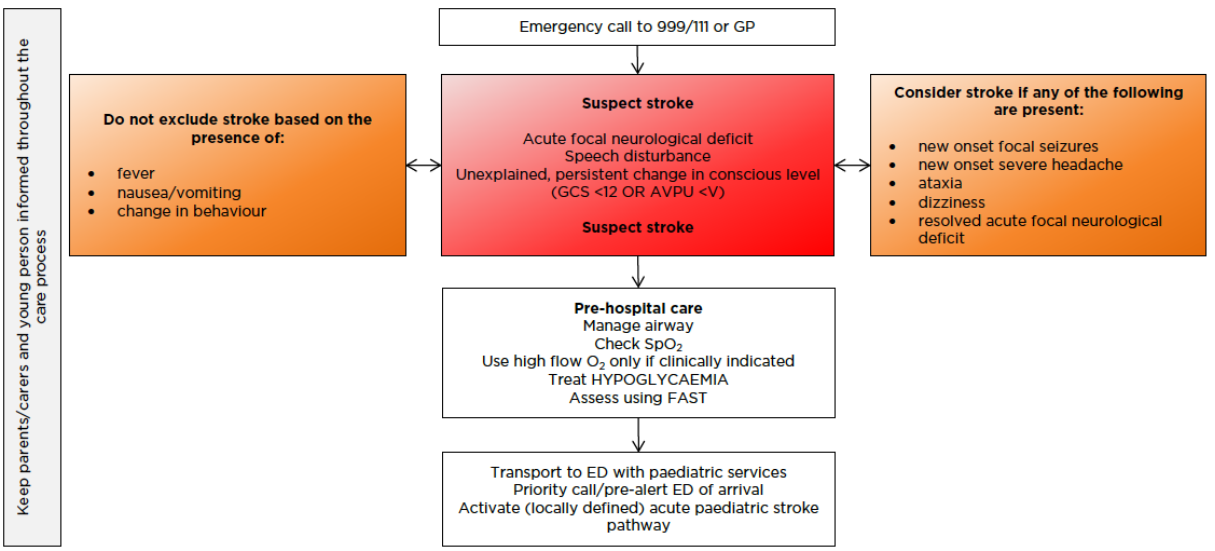
- Lobbied the Stroke Association and MPs
- Informed debate in House of Lords
- Active members of working party; informed scope & parent version
- Workshops to develop consensus statements re communication; information; transition
- Contributed to and reviewed parent version of guidelines

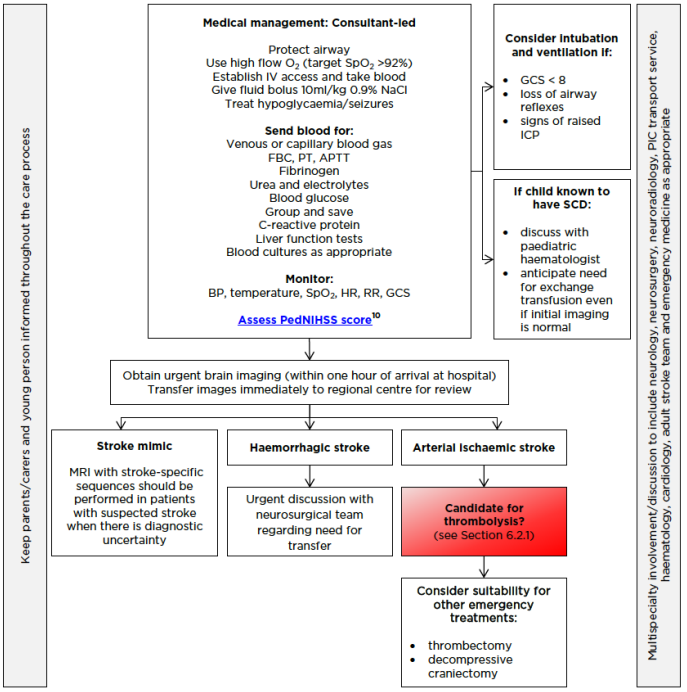




Risk factors for AIS

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
	• prior transient ischaemic attack (TIA) • high systolic blood pressure, acute chest syndrome • anaemia, high reticulocyte count





Imaging the child w “brain attack”

- Potential for hyperacute treatment of AIS means that imaging within 1 hour mandated – but all patients could benefit from this
- CT & CTA if MRI not possible
- In SCD exchange should not be delayed to await imaging if the child has a clinical deficit
- Image vasculature from aortic arch to circle of Willis

PedNHISS definitions	Scale definition
1a. Level of Consciousness:	0 = Alert; keenly responsive. 1 = Not alert, but rousable by minor stimulation 2 = Not alert, requires repeated stimulation to attend, or is obtunded and requires strong or painful stimulation to make non-stereotyped movements. 3 = Responds only with reflex motor or autonomic effects or totally unresponsive
1b. LOC Questions:	0 = Answers both questions correctly. 1 = Answers one question correctly. 2 = Answers neither question correctly.
1c. LOC Commands:	0 = Performs both tasks correctly 1 = Performs one task correctly 2 = Performs neither task correctly
2. Best Gaze:	**0 =** Normal 1 = Partial gaze palsy 2 = Forced deviation/complete gaze palsy
3. Visual: Visual field testing by tested by finger counting ***(for children > 6 years)*** or visual threat ***(for children age 2 to 6 years).***	0 = No visual loss 1 = Partial hemianopia 2 = Complete hemianopia 3 = Bilateral hemianopia (including cortical blindness)
4. Facial Palsy:	0 = Normal symmetrical movement 1 = Minor paralysis (flattened nasolabial fold, asymmetry on smiling) 2 = Partial paralysis (total or near total paralysis of lower face) 3 = Complete paralysis of one or both sides
5 & 6. Motor Arm and Leg:	**5a. Left Arm,5b. Right Arm, 6a Left Leg, 6b Right Leg** 0 = No drift for full 10s 1 = Drift ≤ 10s 2 = Some effort against gravity 3 = No effort against gravity 4 = No movement 9 = Amputation
7. Limb Ataxia (assessed by reaching for or kicking a toy)	0 = Absent 1 = Present in one limb 2 = Present in two limbs
8. Sensory: Grimace to pin prick or withdrawal from noxious stimulus	0 = Normal; no sensory loss. 1 = Mild to moderate sensory loss. 2 = Severe to total sensory loss
9. Best language: ≥ 6 years describe picture 2-6 years observe speech and comprehension	0 = Normal 1 = Mild to moderate aphasia 2 = Severe aphasia. 3 = Mute, global aphasia

In children presenting with AIS THROMBOLYSIS USING tPA:
could **be considered if ≥ 8 yrs and**
may **be considered if 2-8 yrs**

IF ALL OF THE FOLLOWING ARE TRUE

PedNIHSS ≥4 and ≤24
tPA can be administered ≤4.5 hours of symptom onset
CT has excluded intracranial haemorrhage;
CTA demonstrates normal brain parenchyma or minimal early ischaemic change
CTA demonstrates partial/complete occlusion of the intracranial artery corresponding to clinical/radiological deficit

OR

MRI and MRA showing evidence of acute ischaemia on diffusion weighted imaging + partial/complete occlusion of the intracranial artery corresponding to clinical/radiological deficit

PROVIDING THAT THERE ARE NO CONTRAINDICATIONS

Alteplase:

Total dose = 900 micrograms/kg (max. 100mg)

10% (90 mcg/kg) to be delivered as IV bolus

Rest of dose as IV infusion over 60 minutes

Acute AIS management: differences in SCD

- Urgent exchange, don't wait for scan
- Isovolaemic exchage
- Do not routinely give aspirin
- Role of thrombolysis/anti-thrombotics not established – not routine but *may* be appropriate in some cases

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.

Assess for and prevent complications

- Secondary complications: raised ICP, hydrocephalus, seizures
- Swallowing/nutrition
- Endocrine
- Coagulation
- DVT

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.

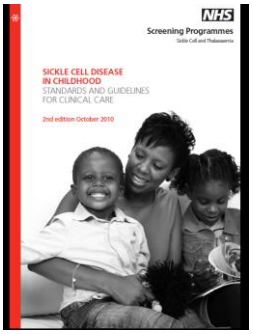
- Consider using anticoagulation or antiplatelet agents only when there are other risk factors for cerebrovascular disease that justify their use.

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.

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 surgery or alternative-donor HSCT.



- Annual TCD scans should be performed on all children with SCD from aged 2 years in accordance with the TCD Standards and Guidelines attached in Appendix 10. For those children who are considered to be "high risk", the risks and benefits of starting regular blood transfusions and/or other treatments should be fully discussed by an appropriate multidisciplinary team with parents/carers (A)
- The symptoms and signs of stroke should be discussed with parents/carers in the first two years of life and information given on what action to take should the child develop neurological symptoms. (C)
- Appropriate imaging studies to assess the extent of cerebrovascular disease should also be arranged if there is evidence of cerebral vessel narrowing on TCD, learning difficulties, atypical symptoms such as unusual behaviour during acute pain, frequent headaches, fits or other unexplained neurological, psychiatric or psychological symptoms. (C)
- Blood pressure should be measured and recorded annually. (C)
- Overnight oxygen saturation monitoring should be recorded if there is a history of snoring, nocturnal enuresis after the age of 6 and low steady-state oxygen saturations on air (<95%). (C)
- Children should have access to a neuropsychologist to assess cognitive function, learning and behavioural difficulties. (C)
- Transfusion therapy should be offered throughout childhood for the secondary prevention of stroke. (B)

Outstanding issues

- Define role of neuroscience clinicians in management of SCD
  - Acute stroke
  - Interpretation of imaging
  - Risk counseling
- Management of acute stroke is not usually in a neuroscience centre
  - Access to MDT & community services
- Different aspects of care are fragmented (e.g. neurology/neurosurgery vs. transfusion vs BMT)
- Relative weighting of different treatments may not be considered in the round (e.g. BMT vs. revascularisation)
- Access to specialist neurovascular MDT
- Role of surgical revascularisation in patients with severe occlusive disease

Habilitation & Rehabilitation

- WHO International Classification of Functioning Framework
- Identify domains for assessment and intervention across ICF
- Early involvement of MDT (within 72 hours)
- Weekly MDT reviews
- Early community liaison
- *Key worker / key contact*

- Active partnership with families:
  - Involve parents and young people in assessment; identification of rehab priorities; regularly inform and update; provide choice where possible
- Assess communication, information and support needs during early functional assessment

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.

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.

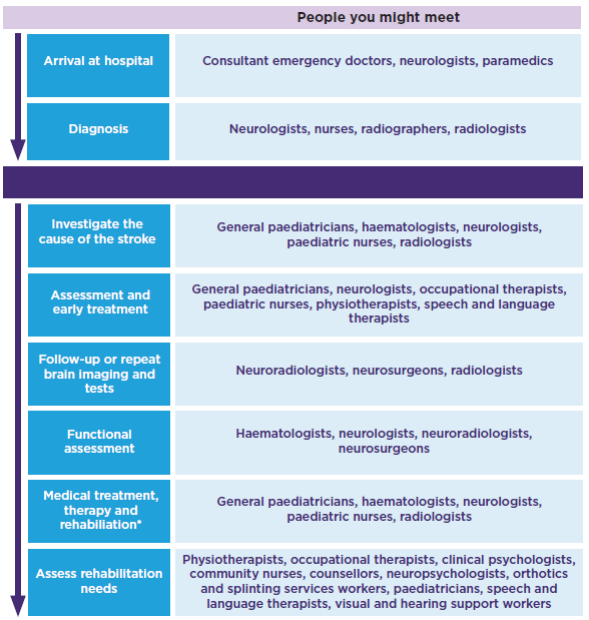
The needs of the family during the planning of care/rehabilitation (Chapter 9.4)

- 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.

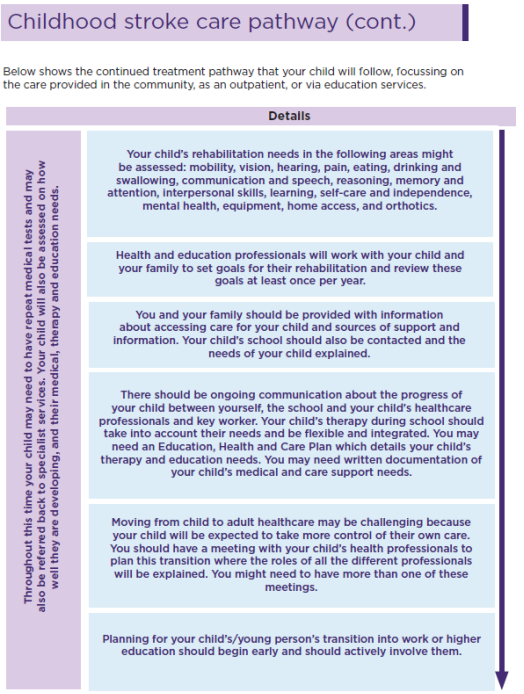
Long-term care: transfer and transition (Chapter 10)

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.

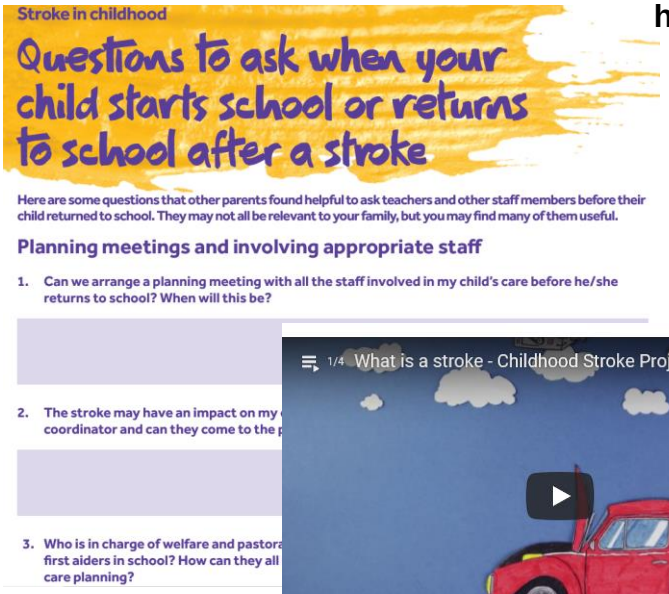


Process



Current practice

- Meeting communication and information needs of CYP and family
- Continuity of care
- Communication between health, education and social care
- Flexibility in delivering changing needs
- Accessibility of services
- Expertise & availability
- Importance of signposting
- Utilisation of charitable sector support


Stroke in childhood

Questions to ask when your child starts school or returns to school after a stroke

Here are some questions that other parents found helpful to ask teachers and other staff members before their child returned to school. They may not all be relevant to your family, but you may find many of them useful.

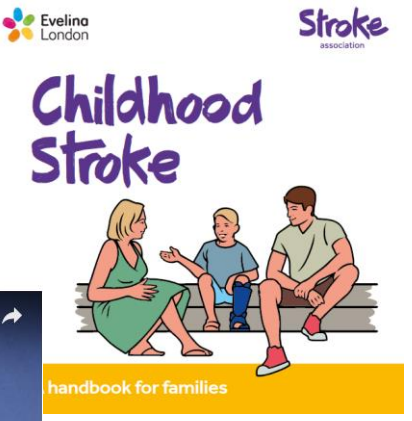
Planning meetings and involving appropriate staff

1. Can we arrange a planning meeting with all the staff involved in my child's care before he/she returns to school? When will this be?

2. The stroke may have an impact on my
coordinator and can they come to the

3. Who is in charge of welfare and pastora
first aiders in school? How can they all
care planning?

https://www.stroke.org.uk/



Key Pointers for Haematologists managing children with SCD

- Early rehabilitation
- Parent communication
- Planning support and guidance from neuroscience clinicians
- Who can support / guide care – what should families and clinicians expect from services
- Families can use guidelines to self-advocate
- Child Stroke Support Service via The Stroke Association

Key Points

- Gaps in guidelines – questions for clinicians; joined up care after diagnosis; support for at-risk patients
- What could be done now – joined up pathway btw neuroscience and haematology that could be audited against
- Role of charitable sector to support parent/YP engagement
  - (Visibility of children and stroke within existing charities??)
- Generalisability of findings to ABI groups