

STROKE • 1/2

This guideline is intended for use in the neonatal period in babies with suspected perinatal stroke

- Defined as a group of heterogeneous conditions with focal disruption of cerebral flow secondary to arterial or venous thrombosis, embolisation or haemorrhagic events between 20 weeks gestation and 28th postnatal day, and confirmed by neuroimaging studies. Includes:
 - perinatal arterial ischaemic stroke (PAIS)
 - cerebral sinovenous thrombosis (CVST)
 - haemorrhagic infarct
 - periventricular haemorrhagic infarction
- Prevalence in term and near term babies estimated 6–17 per 100,000; approximately 80% ischemic stroke and 20% CVST and haemorrhagic stroke

RISK FACTORS

Exact cause of neonatal stroke unknown but risk factors include:

- Neonatal
 - cardiac lesions or procedures
 - coagulation disorders
 - polycythaemia
 - intrauterine growth restriction
 - infection
 - trauma
 - metabolic conditions
 - hypoxic ischaemic encephalopathy
- Maternal
 - primiparity or history of infertility
 - chorio-amnionitis
 - oligohydramnios
 - premature rupture of membranes
 - vacuum extraction
 - emergency caesarean section
 - coagulation disorders
 - pre-eclampsia
 - medications
 - substance misuse (cocaine)/toxins
- Prothrombotic disorders involving protein C, protein S, anti-thrombin III, Factor V Leiden mutation, prothrombin mutation, methyltetrahydrofolate reductase (MTHFR) mutation, antiphospholipid antibody or homocysteine defect may be contributory in 40–70% of babies with PAIS; can also occur in CVST

ACUTE PRESENTATION

- Most common presentation is seizure (typically focal) involving 1 extremity
- occurs in 70–90% of cases
- typically presents within first 3 days of life
- Approximately 80% of cases involve left hemisphere
- May manifest with:
 - features of encephalopathy (irritability, lethargy, increased or decreased muscle tone)
 - feeding difficulties
 - apnoeic episode

LONG-TERM PRESENTATION

- Subtle signs may not be obvious in newborn period
- As child grows, most common sign is weakness or decreased movement on one side of body
- parents commonly report one-handedness or hand preference aged <1 yr
- Delayed/missed developmental milestones

INVESTIGATIONS

Initial investigations

- As majority of babies with stroke will present with seizure, initial investigations for stroke is similar to first line investigations for seizures (see **Seizure** guideline)
- Placental histology

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- Cranial ultrasound scan (USS)
- PAIS – typical triangular or wedge-shaped echo-density in region of middle cerebral artery may be seen but typically takes several days (up to end of first week) to evolve on USS
- difficult to diagnose CVST on USS due to high false negatives
- MRI
- diffusion-weighted imaging with apparent diffusion co-efficient considered most sensitive measure for identifying infarct in neonatal brain
- location and extent of lesions best assessed 2–4 days after onset of stroke when apparent co-efficient of diffusion reaches its nadir
- MR venogram
- may be indicated where venous thrombosis suspected to confirm patency or thrombosis within sinuses

Second line investigations where stroke diagnosis strongly suspected

- Echocardiography to assess for cardiac problems, especially if:
 - abnormal cardiac examination
 - multifocal infarcts on scans
- Thrombophilia screen
- discuss with tertiary paediatric haematologist before conducting this
- limited utility in neonatal period due to decreased levels of protein C, protein S, antithrombin, and Factor XI (30% of adult levels)
- if carried out too early in neonatal period repeat testing after 3–6 months may be required to confirm diagnosis
- no longer routinely indicated in neonates except:
 - positive family history of venous thromboembolic disease – perform factor V Leiden
 - maternal history suggestive of antiphospholipid syndrome – antiphospholipid antibodies test

MANAGEMENT

- Admit to NNU
- Ensure ABC of resuscitation; avoid hyperventilation
- Seizure control
- see **Seizures** guideline and liaise with neurology team if necessary
- once seizures stopped aim to discontinue treatment if possible, due to long-term effects on the developing brain
- seizures rarely persist beyond neonatal period in babies with stroke
- Treat underlying infection if suspected – avoid hyperthermia
- If too unstable to feed start IV fluids
- Correct electrolyte/glycaemic derangement or dehydration (common in CSVT)
- Discuss antithrombotic agents for CSVT with tertiary paediatric haematologist (not shown to be useful in PAIS treatment)

Haemorrhagic stroke

- Correct platelet or clotting factor deficiencies if present; extra dose of vitamin K may be required
- Urgent neurosurgical discussion if confirmed on scan

LONG-TERM OUTCOMES

- Dependent on type of stroke and extent and location of infarction
- MRI has important role in predicting motor outcome especially in PAIS
- involvement of basal ganglia, cerebral hemisphere and posterior limb of internal capsule is highly predictive of contralateral spastic hemiplegic cerebral palsy
- Approximately two-thirds of children with PAIS have poor long-term outcomes, ranging from mild to severe neurologic disability. Deficits can be variable and recognised early in infancy (i.e. delayed motor milestone or early handedness), or later into childhood and adolescence. Hence these babies require long-term follow-up in neonatal clinic or with a neurologist (where available)
- Physiotherapy follow-up required on discharge