E09/S(HSS)/b

2013/14 NHS STANDARD CONTRACT
FOR VEIN OF GALEN MALFORMATION SERVICE (ALL AGES)

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – Service Specification

Service Specification No.	E09/S(HSS)/b
Service	Vein of Galen malformation service (All ages)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Vein of Galen malformation (VGAM) is an arteriovenous malformation which typically results in high output cardiac failure. Treatment of VGAM is considered a high risk procedure but, in experienced centres, offers a chance of good recovery for the child. Complications may include bleeding into the brain, stroke and passage of glue into the lungs (pulmonary embolism).

Recent publications include the Toronto experience from Li et al. (J Neurosurg Pediatrics 2011;7:3-10). Overall survival rate in this series was 76.9%. Mortality rate was 19%. Khullar et al (J Neurosurg Pediatrics 2010;6:444-451) published a review of the world literature up to 2010. Overall 84.3% of patients were found to have a good or fair outcome, with an overall mortality rate of 15.7%. Neonates had the worst outcome with a 35.6% mortality, and series treating higher proportions of neonates tend to show poorer outcomes.

Prior to a formal national service being available in the UK, children were often transferred for treatment at Hôpital Bicetre, in Paris, the centre with the largest experience of this condition and its treatment. A few were undertaken in the UK in various centres, sometimes in combination with colleagues from Paris. At the end of 1999 a UK service was re-established in Glasgow and a second centre was re-established at Great Ormond Street Hospital (GOSH) in 2004. The UK National Service commenced in 2006.

N McSweeney, S Brew, S Bhate, et al. (2010), *Management and outcome of vein of Galen malformation,* BMJ (December)

For revision Autumn 2016.

2. Scope

2.1 Aims and objectives of service

The overall aim of the service is to diagnose, treat and manage children with a Vein of Galen Aneurysmal Malformation (VGAM). The nationally commissioned service is based at Great Ormond Street Hospital in London.

Specifically the VGAM service aims to:
- provide accurate diagnosis;
- assess patients with regard to indications for, and timing of, treatment;
- deliver high quality holistic care from a multi-disciplinary team (MDT) that may include: interventional neuroradiology, foetal medicine, neonatal medicine, paediatric neurology, neurosurgery and cardiology, intensive care and nursing care;
- deliver high quality interventional treatment of patients with VGAM during the infant's first hospital admission for embolisation (blocking the vessels) and any subsequent admission (typically one or two) for further embolisation;
- optimise medical management to reduce risk of complications;
- maintain clinical and service audits (risk and governance) to ensure highest standards of safety, care and clinical effectiveness;
- work in partnership with local medical teams, primary care and the patient/carers to providing advice and optimising management of symptoms.
- provide longer term follow up until the child reaches adulthood;
- screen for and manage conditions associated with VGAM disease.

The UK VGAM service aims to diagnose, treat and manage all children with Vein of Galen malformation. This includes acute admission, sub-acute presentation and elective referrals.

Objectives

- provision of an exemplary and comprehensive service for all patients referred with VGAM;
- expert diagnosis of VGAM utilising the most up-to-date validated diagnostic tools and knowledge;
- expert management of patients with confirmed VGAM through the use of the most up-to-date clinical protocols;
- effective monitoring of patients to ensure optimal functioning for the patient with regards to their VGAM;
- operation of a rolling programme of clinical audit to test current practice and inform the evolution of care in VGAM;
- provision of care with a patient and family-centred focus to maximise the patient experience of care within the nationally designated providers;

- provision of expert advice for the diagnosis and management of VGAM within NHS;
- support local healthcare providers to manage patients with VGAM whenever it is clinically appropriate and safe to do so;
- provision of high quality information for patients, families and carers in appropriate and accessible formats and mediums;
- development of the experience, knowledge and skills of the MDT to ensure a high quality sustainable UK-wide service.

2.2 Service description/care pathway

The service aims to deliver high quality clinical care to patients with VGAM and provide central funding to cover the costs of this care.

All children admitted to the national service with VGAM will have a named consultant who will take responsibility for the clinical care of the patient. For children requiring urgent inpatient admission, this may be a neonatologist, intensive care consultant, cardiologist or a neurologist, depending on the child's needs.

The core MDTs at each centre may include:
- Interventional neuroradiologist
- Neonatologist
- Paediatric neurologist and neurosurgeon
- Paediatric cardiologist
- Paediatric intensivist
- Nursing staff

The clinician accepting the patient will advise on the optimal mechanism for transfer to the centre (for neonates in heart failure it is anticipated that specialist emergency medical transfer will be requested). Antenatal referrals should be directed to the foetal medicine team at University College Hospitals NHS Foundation Trust, London.

Note: retrievals are not funded by the service and are a local responsibility. However in some cases transfer costs will be funded by NHS England or National Services Division (NSD) Scotland, if there is an urgent need and delay would impact on the clinical outcome. In this instance the centres are to inform the commissioners of the transfer.

Management of children with VGAM requires a multi-disciplinary approach and will be guided by clinical assessment and radiological investigation. These will be performed expeditiously and the management plan discussed with the parents. Endovascular treatment will be performed as discussed above (see Section 1.3 Evidence Base).

Follow-up and subsequent clinical management will be under the care of the team detailed below.

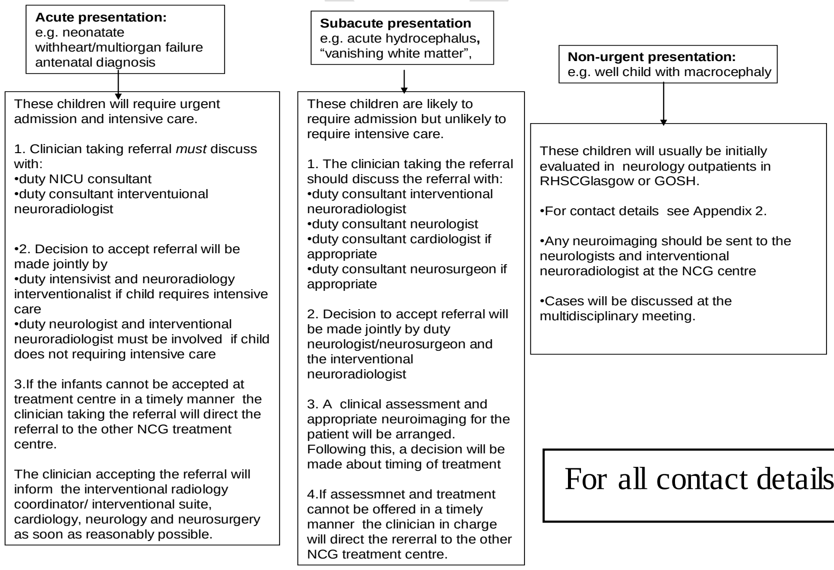
The designated centres will deliver the service in line with the following:

a) clinical protocols for the medical and endovascular management of patients with VGAM are to be continuously reviewed and developed at each centre;
b) the neuroradiological team is required to liaise with the neonatologist or paediatric neurologist to plan treatment and timing as per designated treatment protocols. Individuals work together with the same aims and clinical understanding of the condition and its management to create a multidisciplinary team approach in each centre.
c) inpatients are reviewed by consultants on daily ward rounds with input from the core MDT as clinically required. Care plans are clearly documented in the notes. Relevant investigations will be carried out. Any referred patients that are waiting for admission are discussed and the timing of admission is reviewed, with any actions required, updated;
d) clinical, service delivery and governance issues will be reviewed on a regular basis;
e) on-going audit programme aims to improve and enhance the delivery of clinical care;
f) patient and parent experience surveys are to be regularly undertaken. This will help in the continuing development of the service and improve information to parents.

Risk management

Care delivered by the VGAM service providers must be of a nature and quality to meet the care standards, specification and agreement for the service. It is the Health Board or trust (or equivalent) responsibility to notify the commissioner on an exceptional basis should there be any breaches of the care standards. Where there are breaches any consequences will be deemed as being the Health Board or trust's responsibility.

Service model and Care Pathways - The patient pathway and entrance criteria:



Acute presentation:
e.g. neonatate withheart/multiorgan failure antenatal diagnosis

These children will require urgent admission and intensive care.

1. Clinician taking referral *must* discuss with:
•duty NICU consultant
•duty consultant interventuional neuroradiologist

•2. Decision to accept referral will be made jointly by
•duty intensivist and neuroradiology interventionalist if child requires intensive care
•duty neurologist and interventional neuroradiologist must be involved if child does not requiring intensive care

3.If the infants cannot be accepted at treatment centre in a timely manner the clinician taking the referral will direct the referral to the other NCG treatment centre.

The clinician accepting the referral will inform the interventional radiology coordinator/ interventional suite, cardiology, neurology and neurosurgery as soon as reasonably possible.

Subacute presentation
e.g. acute hydrocephalus, "vanishing white matter",

These children are likely to require admission but unlikely to require intensive care.

1. The clinician taking the referral should discuss the referral with:
•duty consultant interventional neuroradiologist
•duty consultant neurologist
•duty consultant cardiologist if appropriate
•duty consultant neurosurgeon if appropriate

2. Decision to accept referral will be made jointly by duty neurologist/neurosurgeon and the interventional neuroradiologist

3. A clinical assessment and appropriate neuroimaging for the patient will be arranged. Following this, a decision will be made about timing of treatment

4.If assessmnet and treatment cannot be offered in a timely manner the clinician in charge will direct the rererral to the other NCG treatment centre.

Non-urgent presentation:
e.g. well child with macrocephaly

These children will usually be initially evaluated in neurology outpatients in RHSCGlasgow or GOSH.

•For contact details see Appendix 2.

•Any neuroimaging should be sent to the neurologists and interventional neuroradiologist at the NCG centre

•Cases will be discussed at the multidisciplinary meeting.

For all contact details see Appendix 2

Elements of care provided for children with VGAM:

1.	**Referral**	To interventional neuroradiologists, fetal medicine, neonatologists, paediatric neurologists, cardiologists or PICU
2.	**Transfer to centre**	Ideally via specialist neonatal/paediatric transport services
3.	**Radiology**	• **Interventional radiologist sessional time** • **Anaesthetic sessions (imaging, embolisation)** • **Diagnostic radiologist sessional time to report on scans**
4.	**Radiographer**	Radiographer time
5.	**Consumables**	Pre- and post- embolisation imaging (magnetic resonance imaging (MRI) / Magnetic resonance angiography (MRA) / diffusion / perfusion)
6.	**Neonatal Intensive care**	**In patient episode**
7.	**Paediatric Intensive Care**	**In patient episode**
8.	**Paediatric Cardiology**	• **Usually consultation, may be inpatient episode** • **Echocardiography**
9.	**Paediatric Neurology**	• **Inpatient and outpatient episodes** • **Non-radiology investigations (e.g. Electroencephalography (EEG))** • **Administration** • **Clinical nurse specialist**

Days/hours of operation - 24 hours a day, 365 days a year

The acute service is available on a daily basis. Non-acute services are managed through the appropriate paediatric neurology or interventional neuroradiology service in London.

Discharge planning:

Criteria for discharge from inpatient care:
- no further investigation required;
- no adverse outcomes anticipated;
- patient is safe, post-embolisation and intensive care;
- clinically appropriate arrangements for local care and VGAM service follow-up have been discussed and agreed by all relevant parties;
- parents/carers have demonstrated competence in any care they will be required to provide in relation to VGAM;
- parents/carers understand and have the necessary information to contact their nationally designated VGAM provider.

All discharge planning will be managed by the paediatric neurologist or other consultant in charge of the case with local health and social care providers being fully informed of the patient's condition and any responsibilities they will have to assume. This will be formalised in written communication to the patient's GP and all other relevant parties.

2.3 Population covered

This service covers patients registered with an English General Practitioner, resident in Scotland, resident in the European Union and eligible for treatment in NHS under reciprocal arrangements. Patients from Wales, Channel Islands and Northern Ireland are not part of this commissioned service and the trust must have separate arrangements in place.

2.4 Any acceptance and exclusion criteria

Referral criteria, sources and routes

Referrals are accepted from:

- Consultant paediatricians and neonatologists
- Consultant paediatric neurologists
- Consultant neurosurgeons

All referrals should are made directly to Great Ormond Street. Direct GP referrals are not accepted. Please see appendix for referral pathways and cover arrangements

Exclusion criteria

This service is available for children and presenting with evidence of VGAM.

Response time & detail and prioritisation

Most VGAM patients present as emergencies and are seen acutely. Health Board and trust policies will be followed for non-acute referrals.

The VGAM service is accessible to all families. The designated centre at Great Ormond Street Hospital for Children NHS Trust in London should have trust policies in place to ensure that the needs of individual patients and their families are catered for. This may include assistance with family accommodation and the use of interpreters.

The service is accessible to all patients with a suspected VGAM regardless of sex, race, or gender.

Providers may require staff to attend training on equality and diversity, and the facilities provided should offer appropriate disabled access for patients, family and carers. When required the providers will use translators. Printed information is available in multiple languages.

The provider has a duty to co-operate with the commissioner in undertaking Equality Impact Assessments as a requirement of race, gender, sexual orientation, religion and disability equality legislation

2.5 Interdependencies with other services

Internally, the VGAM team will link into multiple clinical and administrative teams as a result of the composition of the MDT as outlined above.

External to this, the nationally designated VGAM service providers are the leaders in NHS for patient care in this area. They provide a direct source of advice and support when other clinicians refer patients to the service. This support will continue until the patient is transferred to the nationally designated service provider or it becomes apparent that the patient does not have a VGAM.

The nationally designated providers also provide education within NHS to raise and maintain awareness of VGAM and its management.

The national providers will form a relationship with local health and social care providers to help optimise management. This may include liaison with consultants, GPs, community nurses or social workers.

Relevant networks and screening programmes

The national service, provided by Great Ormond Street Hospital for Children NHS Trust, London, is required to work with local/regional paediatric neurological networks.

3. Applicable service standards

3.1 Applicable national standards e.g. NICE, Royal College

The nationally designated VGAM providers must be fully integrated into their Health Board or trust's corporate and clinical governance arrangements. The commissioners and service will conduct a formal Joint Service Review at least every six months. There is an expectation that practitioners will participate in continuous professional development and networking. There should be on-going monitoring of risk incidents.

Also see applicable NHS England VGM Service Standards.

4. Key service outcomes

Outcome monitoring is complex, because of small numbers and varying severity at presentation. Treatment outcomes monitored include:

- overall survival and morbidity;
- developmental assessment.

Additional data gathered includes:

- number of treatments;
- technical complications;
- patients seen but not suitable for treatment;
- patients treated by age of presentation (antenatal, neonatal, infant or child).

5. Location of provider premises

The service is delivered across England, Scotland, Wales and Northern Ireland by the designated service based at Great Ormond Street Hospital for Children NHS Foundation Trust, London.

Appendix 1 - Contact information for VGAM referrals

GOSH:	
Consultant Paediatric Neurologist	Via GOSH switchboard 0207 405 9200 ask for on call consultant paediatric neurologist, Secretary 0207 405 9200 ext 5819 (for non urgent queries)
Consultant Paediatric Neurosurgeon	Mobile via GOSH switchboard 0207 405 9200
Consultant Interventional Neuroradiologist	Mobile via GOSH ((switchboard 0207 405 9200

Enquiries for:

NHS patients should be directed to Dr Bhate's secretary in the first instance on extension 5819 Fax number: 0207 813 8279.

Mr. James (consultant neurosurgeon) is also available to assist with queries and should be contacted for all queries relating to International or Private Patients.

All non-urgent queries should come through to Dr Bhate's secretary.

Acute referrals should be made to the duty consultant paediatric neurologist via GOSH switchboard on: 0207 405 9200.