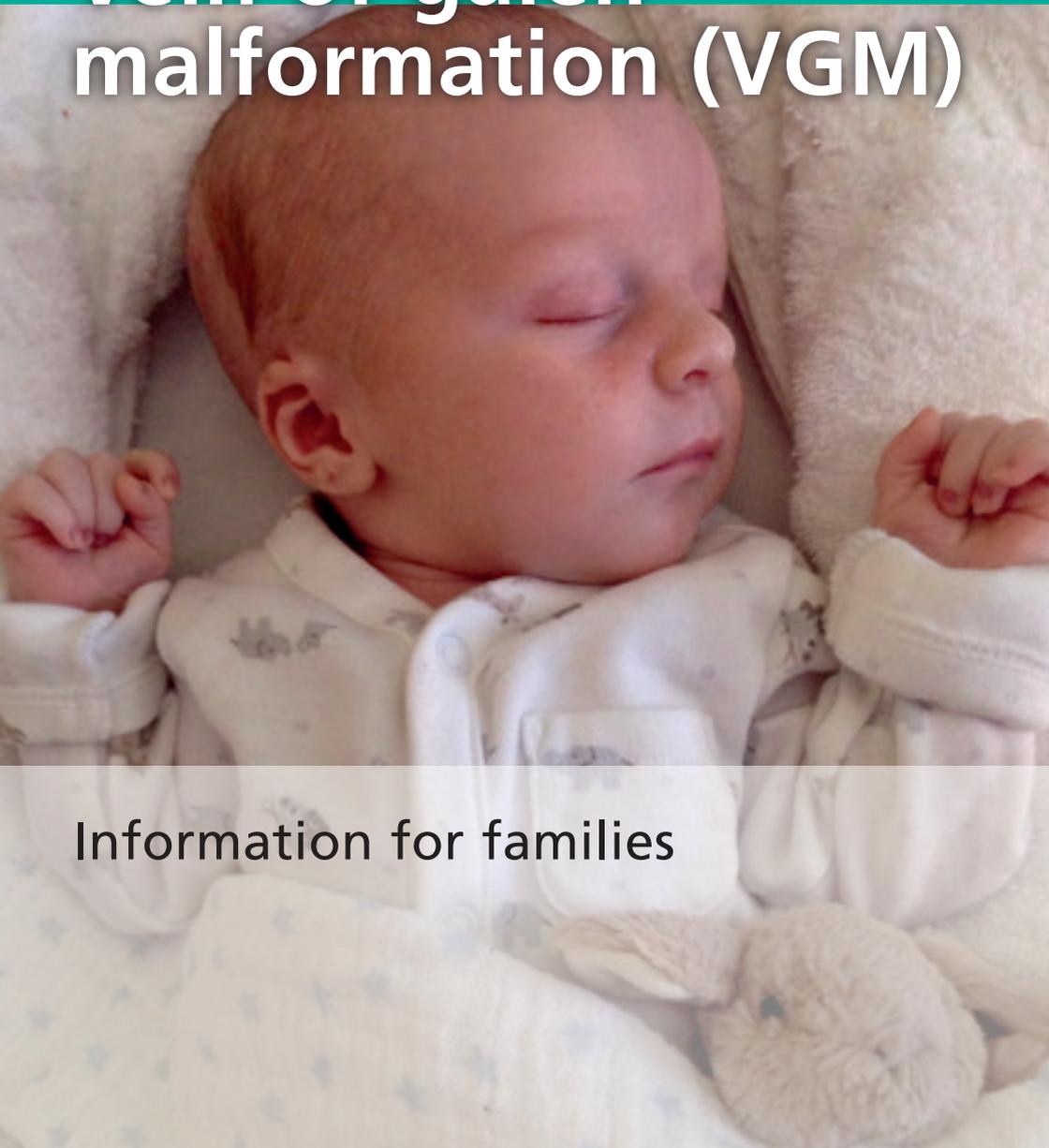


Vein of galen malformation (VGM)



Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This information sheet from Great Ormond Street Hospital (GOSH) explains the neurological condition Vein of Galen Malformation (VGM). It aims to help understanding the condition, how it can be treated and what to expect when your child is admitted to GOSH.

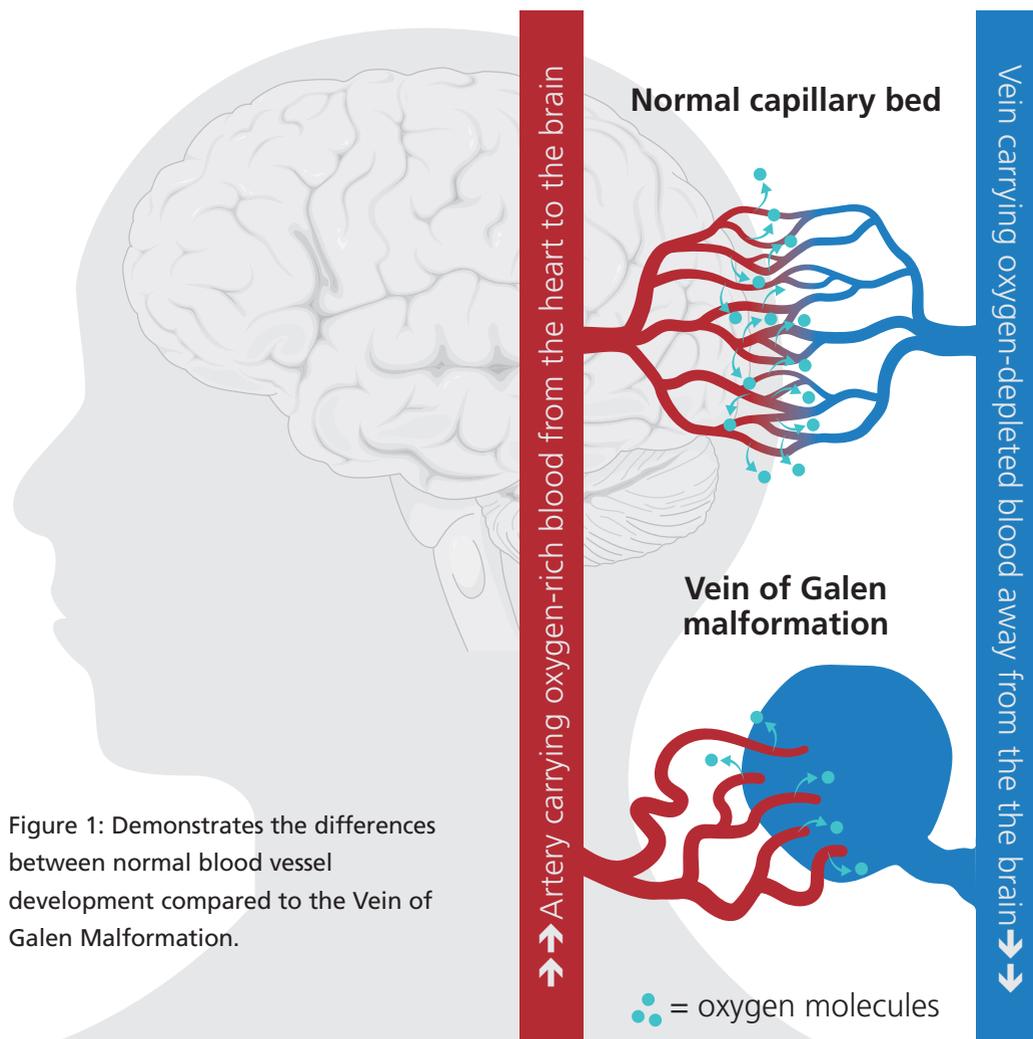


Figure 1: Demonstrates the differences between normal blood vessel development compared to the Vein of Galen Malformation.

What is a Vein of Galen Malformation (VGM)?

Vein of Galen Malformation (VGM) is a rare condition which occurs during pregnancy and results in abnormal connections between blood vessels within the brain.

These abnormal connections are between the arteries (the vessels used to carry blood to the brain from the heart) and the deep veins, which are used to drain the blood away from the brain back to the heart. This draining occurs in the Vein of Galen, which is located in the base of the brain. To ensure that the brain receives a good blood supply to continue developing, the heart needs to work harder in order to account for this additional requirement.

What causes VGM?

There is currently no known cause for the Vein of Galen Malformation, but it is NOT due to anything you did or did not do in pregnancy.

How common is a VGM and who does it affect?

The Vein of Galen Malformation appears to affect both sexes and races equally but requires more research to confirm this. It roughly affects one in one million children born – 10 to 12 in the United Kingdom each year.

What are the signs and symptoms?

Every child is different therefore symptoms vary from child to child and are often subtle and difficult to detect. Some babies will develop symptoms straight after birth, but others will exhibit no signs or symptoms so it may be identified close to discharge after birth or during a routine cranial or heart ultrasound. A small number of babies will be discharged home and the symptoms will be noticed by the midwife or health visitor during a routine check-up.

The VGM can cause pressure on the brain, an enlarged head (hydrocephalus), and prominent veins on the face and dark circles under the eyes. The heart may also be affected by the increased workload and become enlarged. If untreated, this may lead to heart failure.

Newborn presentation of VGM

Newborn babies tend to have multiple arteries attached to the Vein of Galen Malformation, resulting in up to 25 per cent of their blood volume passing through these arteries, causing high pressure in the heart. Clinical signs of this can include tachypnoea (breathing fast), tachycardia (fast heart rate), failure to thrive, cyanosis (blue tinge around the lips) and lethargy or tiredness.

How is a Vein of Galen diagnosed?

Diagnosis of the VGM differs in each individual baby, as some VGMs can be identified when having a routine ultrasound before birth. The majority of babies are identified soon after birth when they start to show signs and symptoms. However, in extremely rare situations, some babies can be discharged home and the problem is suspected when the health visitor/midwife does a routine visit. If the baby's heart is able to cope with the increased pressure, the VGM may not be identified until later childhood.

When a baby is suspected of having a VGM then there are three standard tests which can be used to diagnose the condition:

■ MRI (Magnetic Resonance Imaging)

- A type of scan that uses strong magnetic fields and radio waves to produce detailed images of the inside of the body.

■ Cranial Ultrasound (CUSS)

- A type of ultrasound scan which uses soundwaves to form an image of the brain through the fontanelle (the soft spot on the baby's head).

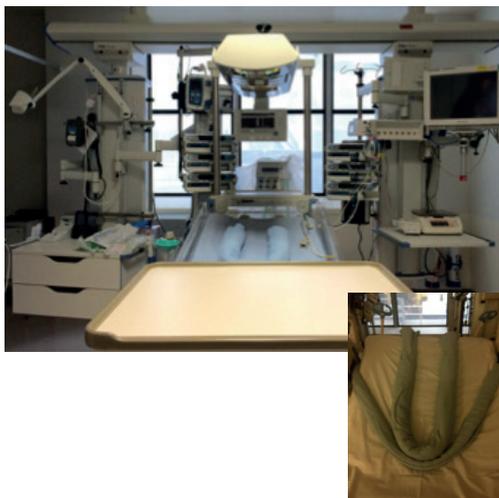
■ Echocardiogram (ECHO)

- A type of ultrasound scan of the heart which measures the extent of any strain or failure.

Initial stabilisation

VGM is a rare condition so is best treated at a specialist centre where several teams can work together to plan and carry out treatment. The exact treatment your child will need and when this is carried out will depend on how sick they are at diagnosis and any other problems present. Children may need to stay on one of our intensive care units – Neonatal Intensive Care Unit (NICU) if they are a baby or the Paediatric Intensive Care Unit (PICU) if they are older.

A complication of the VGM is heart failure due to the extra pressure placed on the heart to supply the brain with enough blood. If your child is in heart failure then the first aim of treatment is to stabilise their heart function. Your child will need breathing support from a ventilator connected to a tube in their mouth going down into their lungs. They will also be on a number of drug infusions to support their heart function.



Once your child has been diagnosed with a VGM, they will be transferred to a specialist unit. Currently GOSH is the only specialist hospital in the UK treating VGM.

You will meet lots of different people from different specialties at the specialist centre.

As a rough guide, you should expect to meet the following people:

- **The ICU clinicians** – These are the doctors who will be responsible for your child while they are staying in the Intensive Care Unit.
- **The Specialist ICU nursing staff** – Your child will have a bedside nurse looking after them for each shift. They are there to support you as well so please ask questions.
- **The Neurology team** – You will have a Consultant Neurologist who will also be heavily involved with your child's care. Once your baby is ready to leave ICU, they will go to a neurology ward.
- **The Interventional Radiology (IR) Team:** These are the specialist doctors who will carry out treatment of the VGM.
- **Koala neurology ward staff:** Once your child is stable and any heart failure is responding to treatment, they will be transferred to the specialist neurology ward.

How is a VGM treated?

Embolisation works by blocking off blood vessels using special 'glue' or coils so that blood flow through the VGM is reduced. This reduces the work the heart has to do to pump blood around the body by ensuring enough blood reaches the brain so it can grow and develop.

The neuroradiologist and the specialised team in the Intervention Radiology (IR) department will carry out the procedure. They will explain the procedure, including the risks of the procedure to you fully before asking you to give permission by signing a consent form.

A catheter (narrow plastic tube) will be inserted into a blood vessel in the groin area and fed through other blood vessels until it reaches the VGM. This procedure takes place with x-ray guidance and close monitoring. The neuroradiologist will inject the substance to block part or all of the VGM.

Your child may return to the ICU with a number of infusions to stabilise their blood pressure. They will also have infusions for pain relief and to limit movement while they recover from the procedure and anaesthetic. Your child will be nursed on a bed raised at an angle to reduce swelling following the procedure.

Risks

With any procedure there are risks. The risks associated with the embolisation increase if your child is very sick or very small. The neuroradiologists who carry out this procedure are specialists in this field, who have had years of experience and will endeavour to minimise the chance of any complications.

The embolisation is carried out under general anaesthetic, and although every anaesthetic carries a risk, this is extremely small. There is only a small risk of infection because no incisions or cuts are necessary.

There is always a chance that the 'glue' or coils used to block the blood vessel will flow to another blood vessel and block it, but the risk of this occurring is very small. The effect of this varies, but the blood supply to this area of the brain will be cut off and this may result in permanent effects. There is also a small chance of bleeding in the brain after treatment.

Your child may bleed from the area where the catheter was inserted, but this can be minimised by applying pressure for a few minutes after the procedure. He or she may develop a bruise where the catheter was inserted.

Recovery from the procedure

Your child will be closely monitored over the next two to three days or so, with support from breathing machines and infusions reducing as they are no longer needed. A specialist physiotherapist will also visit your child to assess their breathing and carry out chest physiotherapy as needed.

Once your child is stable and does not require breathing and blood pressure support, discussion with the neurology ward will begin. Once your child no longer needs intensive care, they will be moved to our specialist neurology ward to recover further. They will continue to be monitored during their stay and, when well enough, can be transferred to a hospital local to your home or if possible directly home.

Support during admission

Everybody recognises this is a very stressful anxious time for families. Being away from home and other family members with the emotions experienced during your stay can be difficult to manage. Every family is different and will require different support at different times.

We offer a number of additional support services that will be offered during your stay. You can also ask for support at any time during your admission.

- Specialist VGM team – Neurovascular Nurse Specialist, ICU coordinator, ICU medical team, Neuromedical team
- Chaplaincy
- Psychologist
- Social work department
- Neonatal Nurse Advisor/
Breastfeeding link team
- Midwifery service – can be arranged if your child was transferred to GOSH soon after birth.
- Family Liaison Sisters will see all ICU parents.

Further information and support

You can contact the **Vein of Galen Support Group** by calling 01963 34393 or visiting their website at www.veinofgalen.wordpress.com

There is also a support organisation in the USA – visit their website at www.vogmparents.org. They also have a Facebook™ page – search for VOGM parents.



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Compiled by the Neonatal Intensive Care Unit and Neurology department
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