Aktiv Druck & Verlag GmbH | ISSN 0170 - 5334 I 02330

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63. Jahrgang | Dezember 2022

# A&I

# ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

Offizielles Organ: Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V. (DGAI)
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Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

## Tuberous sclerosis complex

## Vein of Galen malformation

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

## SUPPLEMENT NR. 15 | 2022

## OrphanAnesthesia –

### ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

**Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:**

**www.ai-online.info/Orphsuppl**
**www.orphananesthesia.eu**

## OrphanAnesthesia –

### a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

**Find a survey of the recommendations published until now on:**

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Anaesthesia recommendations for

# Vein of Galen malformation

**Disease name:** Vein of Galen malformation

**ICD 10:** Q28.2

**Synonyms:** Great cerebral vein, great vein of Galen, vein of Galen malformation, vein of Galen aneurysmal malformations (VGAMs)

**Disease summary:** Vein of Galen malformation is a large intracranial arteriovenous shunt that develops during the 1st trimester of pregnancy. The origin is failure of the embryonic median vein of prosencephalon to obliterate. Multiple arteries feed directly to a large median venous sac resulting in a low resistance intracranial circulation. Most studies report an equal male to female preponderance. It is a rare congenital anomaly (<1/25,000 live births), although the exact incidence is not known. The genetic basis is heterogeneous. Diagnosis may occur in the 3rd trimester of pregnancy if routine ultrasound is undertaken at this stage. Post-natal diagnosis is usually early due to the development of cardiac failure soon after birth. Blood flows preferentially through the low-resistance intracranial AVM causing volume overload and failure of the right ventricle with pulmonary hypertension, and multi-organ failure. Neonates can be shocked at presentation and may need to be intubated on NICU with inotropic support. In less severe cases, post-natal diagnosis is made later with hydrovenous features, namely mild cardiac failure, failure to thrive, increased head circumference and/or developmental delay.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

**Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net**

▶ **Citation:** Lewis H: Vein of Galen malformation. Anästh Intensivmed 2022;63:S391–S398.
DOI: 10.19224/ai2022.S391

## Typical surgery

Embolisation will be required in the neonatal period for severe lesions or during infancy. Endovascular embolisation is the first-line treatment performed in interventional radiology by neuroradiologists. Glue or coils are used to embolise the arterial 'feeders' of the AVM. Patients often require >1 procedure (2–3 staged embolisations is common).

Hydrocephalus can develop as a hydrovenous complication of the arteriovenous malformation (AVM). An external intraventricular drain (EVD) or ventriculoperitoneal (VP) shunt may be required to treat the hydrocephalus. However, the AV shunt should be treated with embolisation 1st line, as otherwise there is a high risk of over-drainage of CSF.

The Bicetre score is one method used to determine treatment options. This is a 21-point scale and its calculation is based on the severity of symptoms and signs of cardiovascular, pulmonary, neurological, renal and hepatic dysfunction. The maximum score is 21. A score of 8–12 identifies neonates most likely to benefit from emergent embolisation. A score of >12 allows time for medical management until around 5 months when embolisation can be more safely performed. A score of <8 is considered too unstable for emergent embolisation and has a poor prognosis.

## Type of anaesthesia

General anaesthesia will be required as procedures take place in the neonatal period or infancy.

Anaesthetic management is particularly challenging due to the complex physiology of a vein of Galen malformation, cardiac failure and pulmonary hypertension.

Specific considerations for neonatal surgery apply – glucose control, temperature management (underbody forced-air warmer and warmed infusion fluids).

Patients may be on diuretic therapy or inotropes to treat heart failure. The combination of starvation and diuretics can make intravenous access difficult and blood pressure labile.

Femoral arterial puncture should be avoided to ensure that an access remains for endovascular procedures.

## Necessary additional preoperative testing (beside standard care)

Full blood count (including platelets) and coagulation should be performed prior to interventional radiology procedures.

Urea and electrolytes should be performed, especially if the patient is on diuretic therapy, and renal function should be checked, particularly if cardiac output and systemic perfusion are low.

An echocardiogram should be performed – high output, right sided heart failure develops as blood preferentially flows through the low-resistance intracranial arteriovenous shunt (from the left ventricle to the carotids) and back to the right side of the heart, causing volume overload and failure. There is often a degree of pulmonary hypertension due to high flow



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from the right side of the heart through the pulmonary circulation and from the persistent pulmonary hypertension of the newborn (PPHN).

Cranial ultrasound (trans-fontanelle) should be made to diagnose and assess the extent of the AVM.

Patients may have had additional imaging – MRI / MRA / CT scan. A practical issue, when feasible, is to combine brain imaging and embolisation under the same anaesthetic. Some centres are able to perform a CT scan on the table prior to waking the patient.

## Particular preparation for airway management

Vein of Galen malformation, as an isolated lesion, is not associated with a difficult airway (bag mask ventilation and/or intubation).

Considerations are for neonatal / infant airway management.

Intubation is required. There may be limited access to the airway during the procedure and the patients are often neonates or young infants.

The patient may already be intubated on the neonatal intensive care unit (NICU) prior to the procedure and on inotropes if haemodynamically unstable from a low cardiac output.

## Particular preparation for transfusion or administration of blood products

Normal transfusion thresholds should apply. Uncomplicated procedures are unlikely to have significant blood loss. A group and save sample should be available.

The requirement for platelets and other clotting products should be discussed with interventional radiology and haematology in advance of the procedure. Term neonates have normal platelet values and function. Pre-term neonates can have low platelets. Prothrombin time (PT) and activated partial thromboplastin time (APTT) can be prolonged in the neonatal period, or if coagulopathic secondary to liver involvement.

## Particular preparation for anticoagulation

Heparin use is decided by the interventional radiologist on a case-by-case basis to prevent thrombosis during endovascular embolisation.

It is prudent to ensure if vitamin K has been given at birth.



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## Particular precautions for positioning, transportation and mobilisation

The endotracheal tube and ventilator tubing should be positioned to one side of the patient so it does not interfere with the imaging during the procedure.

Monitoring on the head should be avoided as it can interfere with the imaging. Saturation monitors and blood pressure cuffs should be on the upper limbs, not lower limbs. This avoids interference from the femoral sheath. A rectal temperature probe is preferred.

Usually, one intravenous cannula is sufficient, ideally in the upper limbs, so it is easy to access.

Transfer to and from intensive care with full monitoring (saturations / NIBP or invasive BP / ECG and ETC02 if intubated), emergency drugs and airway equipment.

## Interactions of chronic disease and anaesthesia medications

Vein of Galen malformation is not associated with other chronic diseases. It does cause a high output, right sided heart failure and patients may be on diuretic therapy / inotropes.

## Anaesthetic procedure

Endovascular embolisation generally takes place in the neonatal period and infancy. A general anaesthetic is necessary. Induction in this age group is usually inhalational with 100 % oxygen and sevoflurane. An intravenous induction is not contraindicated although caution should be exercised. Most intravenous induction agents, except ketamine, can cause further haemodynamic instability. This can be a problem if the patient already has a low cardiac output state. Muscle relaxation; usually either 0.5 mg/kg atracurium or rocuronium (depending on preference), is given to aid intubation and to ensure a still operating field. Maintenance is usually with an oxygen / air mixture and volatile agent. Controlled ventilation is important during endovascular embolisation as the procedure necessitates frequent ventilator ‘on’ and ‘off’ time. Furthermore, spontaneous breathing causes small amounts of head movement which is magnified on the operative screen. Blood pressure control can be an issue during the endovascular embolisation. A high blood pressure can cause the glue to embolise further than intended resulting in cerebral ischaemia. Once the shunt is closed, there is a risk of re-perfusion injury and stroke from increased cerebral flow. A low blood pressure reduces this risk. The patient should be well paralysed and sedated. Sevoflurane 2 % and a short-acting opioid such as fentanyl or remifentanil can help to reduce the blood pressure if needed. Usually the blood pressure is already low as a result of the low cardiac output state. Following embolization, the blood pressure, especially the diastolic, will improve as systemic vascular resistance increases.

Although transcatheter embolisation is essential for the treatment, inhaled nitric oxide was helpful as a bridge treatment to reduce right-to-left shunt before the initial emergency embolisation in a neonate with congestive heart failure and pulmonary hypertension.

Endovascular embolisation is not particularly painful and simple analgesics with paracetamol and local anaesthesia to the puncture site is usually sufficient. An opioid sparing technique avoids the increased risk of postoperative apnoea in neonates. If opioids are used for blood pressure control, they should be short-acting. Postoperative nausea and vomiting is low risk and anti-emetic prophylaxis is not required.



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After the first embolisation, patients should go to NICU/PICU for at least 24 hours. There is usually a period of instability as the left ventricle adjusts to the increased systemic vascular resistance.

Following subsequent embolisations, the patient can be extubated and go to the ward if appropriate.

In older patients without contraindications or airway concerns, extubating deep is beneficial to prevent coughing and straining. This reduces the risk of bleeding and a groin haematoma at the puncture site.

Other reasons for anaesthesia are EVD/VP shunt insertion or imaging. The anaesthetic considerations are the same. Patients are often more stable if they have had a previous embolisation.

## Particular or additional monitoring

An arterial line is not always necessary. In some centres, if required, the femoral sheath can be transduced for invasive blood pressure monitoring and blood gas sampling.

## Possible complications

Intraoperatively, there is a risk of cerebral ischaemia/re-perfusion injury/AVM rupture/lower limb ischaemia/vessel perforation.

There is a risk of groin haematoma from the femoral arterial puncture site. Anaesthetic technique should minimise coughing and straining, particularly at extubation.

Failure of the left ventricle can occur in the immediate postoperative period due to the increased workload from the higher systemic vascular resistance.

## Postoperative care

NICU/PICU is required after the 1$^{st}$ embolisation. Left sided heart failure can develop due to the sudden increase in vascular resistance increasing the workload of the left ventricle.

The 2$^{nd}$ embolisation is usually done 1–2 months after the first. The patients are often haemodynamically more stable and off cardiac medications. It may be suitable for them to go to an appropriately designated ward postoperatively.

## Disease-related acute problems and effect on anaesthesia and recovery

Apnoea monitoring will be needed for term infants <44 weeks post conceptual age (PCA) and pre-term infants <60 weeks PCA.



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## Ambulatory anaesthesia

This is not relevant.

## Obstetrical anaesthesia

There are no specific reports of pregnant women with vein of Galen malformation previously diagnosed and submitted to anaesthesia.

The pre-natal diagnosis can be made in the 3rd trimester of pregnancy. Delivery should take place in a specialist centre with appropriate neonatal expertise.



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**Date last modified:** **December 2021**

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**Disclosure** The author has no financial or other competing interest to disclose. This recommendation was unfunded.

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**Disclosure** The reviewers have no financial or other competing interest to disclose.



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Paumgartnerstraße 28 | 90429 Nürnberg
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**Erscheinungsweise 2022**
Der 63. Jahrgang erscheint jeweils zum Monatsanfang, Heft 7/8 als Doppelausgabe.

**Bezugspreise** (inkl. Versandkosten):

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  Europa (ohne Schweiz) (inkl. 7 % MwSt.) 258,- €
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