

# A&I

## ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

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



**Stress cardiomyopathy**  
**Sturge-Weber syndrome**

orphan**a**nesthesia

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

**SUPPLEMENT NR. 15 | 2018**

## 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](http://www.ai-online.info) veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

## OrphanAnesthesia –

### a common 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 recommendations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via [www.ai-online.info](http://www.ai-online.info). As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

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

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# orphananesthesia

## Anaesthesia recommendations for patients suffering from **Stress cardiomyopathy**

**Disease name:** Stress cardiomyopathy

**ICD 10:** I42

**Synonyms:** Ballooning cardiomyopathy, broken heart syndrome

Stress cardiomyopathy (SCM) is a transient clinical condition, which mimics myocardial infarction in patients with no coronary heart disease.

Typical SCM affects the apex of the left ventricle and causes ballooning of apex. This is popularly known as "Takotsubo cardiomyopathy" or "Broken heart syndrome". SCM wherein there is no involvement of the apical segment of the heart is called as atypical or variant SCM.

The occurrence of SCM in the perioperative period is no longer rare. It is being reported regularly even immediately prior to surgery in patients with excessive anxiety. The prevalence of SCM is estimated to be 1.2-2.2%, and the atypical SCM constitutes 40% of these cases.

Even though the aetiology of SCM is considered to be exposure to excessive levels of catecholamines in genetically susceptible individuals, the exact mechanism is not yet proven. The pathophysiology could be explained by (1) enhanced sympathetic activity originating from the central nervous system, (2) catecholamine-induced microvascular endothelial dysfunction, or (3) coronary vasospasm.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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**Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)**

► **Citation:** Halemani KR: Stress cardiomyopathy. AnästH Intensivmed 2018;59:S586-S595.  
DOI: 10.19224/ai2018.S586

### Disease summary

Atypical SCM constitutes four types depending on the patterns of LV involvement: (1) inverted/ reverse, (2) mid-ventricular, (3) focal and, (4) global type. Recently, a fifth type 'reverse mid-ventricular' has been described, in which the apex and base are akinetic/hypokinetic and the mid-left ventricle is hyperdynamic.

There are case reports of involvement of both ventricles or the right ventricle alone. The difference in the distribution, density and sensibility of adrenergic receptors over the myocardium determines the area of hypokinesis while the cause remains the same. The area with a highest density of adrenergic receptors is typically the most affected region. The most common of these atypical forms is the inverted type with hyperdynamic apex and akinetic base of the LV.

The density of adrenergic receptors is relatively high at the apex in the elderly population compared to younger patients. Hence, elderly patients, especially postmenopausal women, are more prone for classical SCM whereas younger patients are more prone to the atypical variety. The atypical variety presents with a lower prevalence of T-wave inversion. The release of troponin is higher, which is the consequence of the larger muscle region involved. Basic natriuretic peptides (BNP) are more elevated in apical and mid-ventricular patterns, which is evident by more severe symptoms. The rise in cardiac markers and ECG changes may be similar to acute coronary syndrome but a rise in BNP is much higher in SCM.

The most common complications described are cardiogenic shock (10% of patients), ventricular arrhythmias, formation of apical thrombus, stroke, obstruction of the left ventricular (LV) outflow tract, LV rupture and sudden cardiac death. Some might even develop pleural effusion and mitral valve regurgitation (resulting from chordal tethering as well as a systolic anterior motion of the mitral valve apparatus).

Diagnosis is made by coronary arteriography, left ventriculography, and echocardiography. Differential diagnosis includes acute coronary syndrome and other types of transient ventricular dysfunction such as seen in sepsis or myocarditis.

The condition is transient and the main stay of treatment is supportive.  $\beta$ -adrenergic blockers, angiotensin converting enzyme inhibitors, statins, diuretics and heparin with or without clopidogrel or aspirin are the mainstay of treatment.

Patients who develop hypotension must be evaluated for a dynamic intra-ventricular pressure gradient in the left ventricular cavity and left ventricular outflow tract. Echocardiography or left heart catheterisation can be used for this evaluation.

Dynamic intra-ventricular obstruction in patients with the syndrome is managed by administration of beta-blockers (to increase diastolic ventricular filling time and left ventricular end-diastolic volume), administration of phenylephrine (to increase afterload with subsequent reduction of the intra-ventricular gradient) and administration of fluid resuscitation if pulmonary congestion is not present. However, beta-blockers and phenylephrine would not be recommended for the treatment of dynamic intra-ventricular obstruction in patients with documented epicardial coronary vasospasm. In this situation, a nondihydropyridine calcium-channel blocker, such as diltiazem or verapamil, could be considered.

If there is no dynamic intra-ventricular obstruction, inotropes of choice are levosimendan or milrinone. If cardiogenic shock is persisting, intra-aortic balloon pump (IABP) or ventricular assist devices (VAD) can also be used. Use of extracorporeal membrane oxygenation has also been reported.

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### Typical surgery

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All elective cases with SCM should be postponed.

Any surgical procedure which has to be done on an emergency basis in patients who are exposed to severe mental or physical stress, for example, emergency craniotomy because of intracranial bleed, post poly trauma surgeries, postpartum bleed and explorations, might have to be accepted with due risk of perioperative cardiac failure, ventricular arrhythmias, left ventricular rupture and sudden cardiac death.

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### Type of anaesthesia

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Peripheral nerve blocks can be used wherever possible like upper limb vascular injuries, orthopaedic open fracture fixations or wound debridements.

Use of a laryngeal mask airway can be useful if the procedure is short and patient is not having severe hypotension, pulmonary oedema or pleural effusions. This avoids the stress response to intubation.

General anaesthesia (GA) with endotracheal intubation is the choice if the patient is haemodynamically unstable, if there is pulmonary oedema or pleural effusion, surgical risk is higher, or there are other indications for GA.

A decrease in systemic vascular resistance secondary to spinal or epidural anaesthesia might not be well tolerated. In addition, the patient may be on anticoagulants, which could prevent the use of neuraxial anaesthesia.

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### Necessary additional diagnostic procedures (preoperative)

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ECG, Trans thoracic or transoesophageal echocardiogram.

Cardiac enzyme markers, coronary angiogram to rule out acute coronary syndrome. Cardiac enzymes help not only in diagnosing the disease, their increasing trend indicates ongoing ischemic damage and poor myocardial function.

Chest X-ray to rule-out pulmonary congestion.

Serum electrolytes, liver and renal function tests and coagulation profile.

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### Particular preparation for airway management

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Care to prevent stress response to laryngoscopy and intubation.

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**Particular preparation for transfusion or administration of blood products**

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Intravenous fluid administration should be done with caution and preferably guided by transesophageal echocardiogram (TEE). Use of CVP, pulmonary artery catheter is not much useful in the presence of LV failure. Systolic volume variation (SVV) can be another useful guide for fluid therapy. Blood product administration should follow general guidelines of maintaining adequate blood volume and haemostasis without causing a volume overload.

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**Particular preparation for anticoagulation**

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Patient may be on anticoagulants. Due care should be taken.

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**Particular precautions for positioning, transport or mobilisation**

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Invasive monitoring should be instituted preferably in the intensive care unit itself. Patient should be continuously monitored while shifting. Rapid changes in the body position should be avoided as the heart might not be able to compensate. Sudden head low or head up might not be well tolerated as the heart cannot cope with sudden increase or decrease of preload with these positions, respectively.

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**Probable interaction between anaesthetic agents and patient's long-term medication**

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Patient might have been started on  $\beta$ -adrenergic blockers, angiotensin converting enzyme inhibitors (ACEIs), diuretics and heparin.

Continue with beta-blockers and, if possible, stop diuretics and ACEIs at least one day prior to surgery. If it is not possible to stop, we should be aware that an inhibition of the renin angiotensin aldosterone system (RAAS) by ACEIs can increase the risk of intraoperative hypotension. This can be treated with phenylephrine or vasopressin.

As these patients usually come for emergency procedure, the cardiologist can delay starting clopidogrel until surgery is over. However, heparin can be started if there is time to stop it 6 hours prior to surgery, especially if the surgery involves brain, spine or intraocular structures. Reversing the heparin effect with protamine might cause a LV thrombus. Pros and cons should be considered.

### Anaesthesiologic procedure

Management strategies are similar to managing a case of acute cardiac failure patient coming for emergency surgeries.

#### Intra-operatively:

Anaesthetic goals would be:

- A. Maintain sinus rhythm
- B. Avoid sudden increases in preload like rapid fluid boluses or sudden head low.
- C. Maintain normal after load: High or low will not be tolerated
- D. Monitor and maintain optimum contractility of the heart.

Before induction of anaesthesia, along with standard non-invasive monitors, the arterial line has to be inserted under local anaesthesia if it is not there already. A good peripheral vascular access should be obtained if there is no central line. Attach defibrillator pads to the chest and apply pneumatic compression stockings to the lower limbs.

Poor ventricular function can cause severe hypotension on induction with propofol. Etomidate in titrated doses or opioid-based induction is well tolerated. Avoid any stress responses during intubation. Positive pressure ventilation and positive end expiratory pressures (peak pressure < 30 cm of H<sub>2</sub>O) are beneficial to improve oxygenation.

Cardiac output monitoring with pulmonary artery catheter or Pulse index Continuous Cardiac Output (PiCCO) monitor can be used, but is not routinely required. Pulmonary capillary wedge pressure might not be useful for intravenous fluid management due to poor ventricular compliance. Hence a trans-oesophageal echocardiogram (TEE) is recommended in such cases. In addition, a TEE can also give information regarding ventricular wall motion, presence or absence of dynamic ventricular outflow obstruction and valvular functions.

If inotropic support is needed, a calcium sensitiser such as levosimendan may be a good choice if it is available. It is an inodilator and improves myocardial contractility without increasing the intracellular levels of calcium and hence there is no increase in myocardial oxygen demand. Milrinone is reported to be of benefit in one case in which the patient had acquired SCM following a head injury. Adding a vasopressor like phenylephrine to counter vasodilatation caused by these drugs can be helpful. Excessive inotropy may worsen LV outflow obstruction and the inotropes should therefore be carefully titrated.

In case catecholamines have to be used, avoid using them in high doses. But there are always chances of developing tachyarrhythmia.

A close watch on urine output, core body temperature, airway pressures and acid base status will help in better management.

Dosage of levosimendan: 18-36ug/kg bolus followed by 0.05 to 0.2 µg/kg/min. as a continuous infusion. Totally avoiding or using a reduced bolus dose of 6-24 µg/kg can be considered as bolus dose can cause a precipitous drop in blood pressures. Levosimendan should not be administered to children or adolescents under 18 years of age. It is not advised for more than 24hours.

Dosage of milrinone: 50 mcg/kg loading dose by IV push over 10 minutes, then 0.375-0.5 mcg/kg/min. IV. Bolus dose can be avoided.

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**Particular or additional monitoring**

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Five lead ECG, cardiac output (PICCO), transesophageal echocardiogram and SVV

Keep defibrillator, pacemaker, IABP or VADs as standby.

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**Possible complications**

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Cardiogenic shock, ventricular arrhythmias, formation of apical thrombus, stroke, obstruction of the LV outflow tract, LV rupture and sudden cardiac death.

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**Postoperative care**

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Postoperative extubation depends upon the severity of heart failure and type of surgery.

Continued monitoring in intensive cardiac care unit.

Adequate anxiolytics and good pain relief.

Continue all the preoperative medications.

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**Information about emergency-like situations / Differential diagnostics**

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Hypotension and arrhythmias are the main complications to be expected. TEE will help in early diagnosis and appropriate management. Arrhythmias can be managed as per standard protocols and life-threatening hypotension needs IABP or VADs.

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**Ambulatory anaesthesia**

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Not advised.



### **Obstetrical anaesthesia**

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General anaesthesia with all the safety precautions is advisable. Invasive arterial line inserted prior to induction is a must. A modified rapid sequence induction with etomidate, fentanyl and succinyl choline can be acceptable. The insertion of central line is better done prior to induction. If surgery is emergency at least the external jugular vein should be accessed. Inotropes should be loaded and kept ready.

Use of a TEE or PA catheter should be decided on a case to case basis. Neonatologists should be well informed about the possibility of respiratory depression in newborn. They should keep an intubation trolley and naloxone ready. Fluid management should be done very cautiously as parturients have very delicate fluid requirements.

Hypotension on induction can be managed with phenylephrine. Oxytocin should be administered carefully as it causes tachycardia and reduces systemic vascular resistance. Post-delivery, there is a possibility of pulmonary oedema because of an abrupt inflow of large volumes of blood into the systemic circulation as a consequence of uterine contraction. A small dose of frusemide can be given after the delivery of the baby. Sometimes the patient might need postoperative mechanical ventilation.

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**Last date of modification: March 2017**

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## Verlag & Druckerei

### Aktiv Druck & Verlag GmbH

An der Lohwiese 36 |  
97500 Ebelsbach | Deutschland  
www.aktiv-druck.de

### Geschäftsführung

Wolfgang Schröder | Jan Schröder |  
Nadja Schwarz  
Tel.: 09522 943560 | Fax: 09522 943567  
E-Mail: info@aktiv-druck.de

### Anzeigen | Vertrieb

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Tel.: 09522 943570 | Fax: 09522 943577  
E-Mail: anzeigen@aktiv-druck.de

### Verlagsrepräsentanz

Jürgen Distler  
Roritzerstraße 27, 90419 Nürnberg  
Tel.: 0171 9432534 | Fax: 0911 3938195  
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### Herstellung | Gestaltung

Manfred Wuttke | Stefanie Triebert  
Tel.: 09522 943571 | Fax: 09522 943577  
E-Mail: ai@aktiv-druck.de

### Titelbild

Dipl.-Designerin Monique Minde,  
Nürnberg

### Erscheinungsweise 2018

Der 59. Jahrgang erscheint jeweils zum  
Monatsanfang, Heft 7/8 als Doppelausgabe.

### Bezugspreise (inkl. Versandkosten):

- Einzelhefte 30,- €
- Jahresabonnement:
  - Europa (ohne Schweiz) 258,- €  
(inkl. 7 % MwSt.)
  - Schweiz 266,- €
  - Rest der Welt 241,- €

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prüft werden. Gleiches gilt für berufs-  
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und Empfehlungen.

# CONTACT US

Please do not hesitate to contact us. We will be glad to answer and provide further information to you at any time.

.....  
Name

.....  
First Name

.....  
Department / Hospital

.....  
Place

.....  
Telephone

.....  
E-Mail

.....  
Date / Signature

Please contact me for further information

I would like to participate in the project

## ADDRESS

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