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

www.ai-online.info

59. Jahrgang | November 2018

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

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

Stüve-Wiedemann syndrome

Systemic sclerosis

orphanaesthesia

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

SUPPLEMENT NR. 17 | 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 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 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 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.

A survey of until now in A&I published guidelines can be found on:

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orphananesthesia

Anaesthesia recommendations for patients suffering from

Systemic sclerosis

Disease name: Systemic sclerosis

ICD 10: M34.0

Synonyms: Progressive systemic sclerosis, scleroderma, CREST syndrome

Systemic sclerosis (SSc), also called scleroderma, is a multisystem connective tissue disease characterised by the excessive production of collagen, glycosaminoglycans and fibrinonectins within the connective tissue. This results in the hardening and fibrosis of skin, mucus membranes, vasculature and internal organs. Clinical features include tightening and thickening of skin (skin sclerosis), Raynaud's phenomenon and involvement of various internal organs (particularly in the lungs). There are two major SSc phenotypes, a limited cutaneous and a diffuse cutaneous form, based on the extension of skin involvement.

The prevalence of scleroderma ranges from 4-489 cases per million worldwide, with an annual incidence of 0.6 – 122 million. The prevalence is higher in the US and Australia than in Europe and Japan. The ratio of women to men affected is 3:1 and it has a peak incidence in the fifth decade of life.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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

▶ **Citation:** Dempsey ZS, Rowell S, McRobert R: Systemic sclerosis. Anästh Intensivmed 2018;59:S644-S653. DOI: 10.19224/ai2018.S644

Typical surgery

Patients affected by systemic sclerosis may require surgery for any type of procedure, but will typically present for repeated oesophageal procedures, dental treatment and surgical management of vascular insufficiency including cervical, lumbar and digital sympathectomy in addition to amputation. In severe forms, lung transplantation may be considered due to severe interstitial disease or pulmonary arterial hypertension.

Type of anaesthesia

There is no definite recommendation for either general anaesthesia or regional anaesthesia and the choice of anaesthetic technique will depend on the type of surgery, an understanding of the pathophysiology of the disease and careful preoperative assessment of the patient.

General anaesthesia may be complicated by difficult intubation, high incidence of aspiration, due to gastro-oesophageal reflux disease (GORD) and significant respiratory disease.

Regional anaesthesia is a safe alternative to general anaesthesia and a useful adjunct in the treatment of postoperative pain and prevention of vasospastic crisis. Technical challenges exist in performing regional anaesthesia due to difficulties in positioning the patient and altered anatomy. There is the potential for prolonged sensory blockade with peripheral nerve blocks.

Necessary additional diagnostic procedures (preoperative)

There is a five- to eight-fold increase in mortality associated with SSc, particularly in patients with pulmonary hypertension and cardiac involvement.

To assess the extent of pulmonary disease, such as pulmonary fibrosis, patients should have a chest radiograph and pulmonary function tests to demonstrate any reductions in compliance, vital capacity and diffusion capacity. A risk factor for increased mortality is a forced vital capacity of less than 50% predicted. Pulse oximetry in air and arterial blood gases can be carried out to assess the degree of hypoxaemia. Consider cardiopulmonary exercise testing if available to assess functional ability.

Cardiac disease may present as pericarditis, pulmonary hypertension, congestive heart failure, cardiomegaly, systolic dysfunction, myocardial fibrosis, dilated or restrictive cardiomyopathy, conduction defects and arrhythmias. At-risk patients should have a baseline ECG (although only 19% of patients will exhibit an abnormal rhythm on continuous 24 ECG monitoring) and an echocardiogram.

Gastrointestinal disease may result in malnutrition, impaired absorption of vitamin K and electrolyte disturbance. All patients therefore require a full blood count, urea and electrolytes, liver function tests, bone screen and coagulation screen in addition to a group and hold or crossmatch depending on procedure.

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Particular preparation for airway management

Dermal fibrosis will lead to up to 70% of patients having a pinched face, atrophied nasal alae and restricted mouth opening, compounded by temperomandibular joint fibrosis. Limited neck extension may occur along with blunting of the angle of the mandible. Difficulties with intubation and mask ventilation should therefore be expected and access to difficult intubation equipment including jet ventilation should be made immediately available.

Fibre-optic, blind oral or retrograde intubation techniques may be considered. Patients are prone to mucosal telangiectasias, which may bleed profusely and therefore it is important to exercise careful airway manipulation techniques.

Patients are at risk of aspiration due to GORD, which can be severe. Rapid sequence induction should be undertaken with caution due to the risk of failed/difficult intubation. Sellick's manoeuvre may also be ineffective due to fibrosis or the oesophagus and impair view at laryngoscopy further.

In particularly difficult situations it may be necessary to consider awake tracheotomy with local anaesthesia.

Particular preparation for transfusion or administration of blood products

There is no definite recommendation for transfusion; administration of blood products will depend on the type of surgery, patient symptoms and advice from senior haematology clinicians.

Particular preparation for anticoagulation

In rare cases, scleroderma patients have antiphospholipid antibodies and are at higher risk of vascular thrombosis, however, there is no definite recommendation for anticoagulation; administration will depend on type of surgery, patient symptoms and senior clinical advice.

Particular precautions for positioning, transport or mobilisation

Due to flexion contractures, positioning should ideally be guided by the awake and co-operative patient. Due to vascular insufficiency, pressure areas should be carefully padded and checked regularly. A vacuum mattress should be considered for patient transportation.

During the surgical procedure, Trendelenburg position may favour pulmonary aspiration and should therefore be avoided unless the airway is secure.

The patient's temperature should be maintained at all times to prevent vascular crisis and digital ischemia. Sweating is hindered and therefore care should also be taken not to overheat the patient, which may present as malignant hypertension.

Patients are prone to developing dry eyes which may be compounded by scarring of the eyelids preventing complete closure. Eyes should be carefully lubricated and padded to avoid corneal abrasions.

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

Immunosuppressants are the mainstay of treatment increasing the risk of postoperative infective complications. An additional steroid cover should be provided in patients on glucocorticoids.

Angiotensin–converting enzyme inhibitors are the first line antihypertensive agents in patients with Systemic sclerosis and may produce refractory hypotension post induction of anaesthesia.

Anaesthesiologic procedure

In case general anaesthesia is required, endotracheal intubation is advocated to decrease the risk of aspiration.

Ventilation may be challenging due to reduced lung compliance and protective lung strategies should be employed to prevent barotrauma.

Induction agents, volatiles, depolarising and non-depolarising muscle relaxants, and reversal agents are all safe in Systemic sclerosis.

Use of vasopressive amines can worsen Raynaud's phenomenon and is associated with an increased risk of digital ischaemia, treatment with iloprost can be proposed in accordance with the patient's haemodynamic status and before necrotic lesions occur.

There is a strong association of renal and gastrointestinal disease and therefore non-steroidal anti-inflammatories should be avoided. Patients can also be sensitive to opioids. Where possible, opiate sparing techniques should be used, including the use of regional anaesthesia, which may also be considered as a safe alternative to general anaesthesia in high-risk cases.

Regional anaesthesia may be challenging due to patient positioning, altered fascial planes and prolonged sensory blockade. The use of ultrasound to identify structures and spread of local anaesthesia is therefore recommended. The spine is frequently spared in systemic sclerosis and many of the challenges and complications associated with regional anaesthesia are therefore reduced by neuroaxial approaches. Marked hypotension can occur secondary to anaesthesia-induced vasodilatation and may be refractory to inotropes. Excessive fluid administration may result in pulmonary oedema once the vascular tone is restored. Techniques that enable a gradual or incremental adjustment of the block height, such as epidural or combined epidural spinal anaesthesia, are therefore preferable.

Particular or additional monitoring

Routine monitoring (as per Association of Anaesthetists Great Britain and Ireland guidelines) is advocated in all SSc patients. Dermal thickening, flexion contractures and vasoconstriction may make it difficult to obtain intravenous access and non-invasive blood pressure readings. This may necessitate the use of invasive monitoring and central venous access.

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Radial arterial cannulation can precipitate Raynaud's phenomenon and even subsequent necrosis. Moreover, some patients have a macroangiopathy with radial artery thrombosis. It is important to alternate pulse oximeter probes between digits during surgery, as failure to do so can result in precipitation of ischaemic damage. Patients with severe cardiac disease and pulmonary hypertension may benefit from cardiac output monitoring, although the presence of oesophageal fibrosis, aortic disease and altered vascular performance may affect the accuracy of newer cardiac output monitors.

Possible complications

Patients with systemic sclerosis are at increased risk of failed or difficult intubation and aspiration.

Patients with pulmonary disease will have a reduced oxygen reserve and an impaired pulmonary compliance. They may therefore desaturate suddenly, particularly during airway manoeuvres and may be difficult to ventilate with risk of barotrauma. They might be sensitive to opiates and have a high risk of postoperative respiratory failure especially in the presence of a severe disease (vital capacity of less than 1 litre).

Patients may have a severe cardiac disease resulting in systolic dysfunction, conduction defects and arrhythmias. They have a relatively reduced intravascular compartment and may become profoundly hypotensive as a result of an anaesthesia-induced vasodilatation and tolerate dehydration and blood loss poorly. Rebound pulmonary oedema may occur on restoration of vascular tone.

Patients are at increased risk of cerebrovascular events. Uraemia and malignant hypertension may also cause seizures.

Stress, pain, dehydration, hypothermia and vasoconstrictors therapy may induce vasospastic crisis leading to peripheral ischaemia and ulceration.

Sweating is hindered and patients are at risk of hyperthermia.

Wound healing can be impaired due to poor peripheral perfusion and patients may be prone to pressure sores.

Malnutrition and immunosuppressant therapy may increase the patients' susceptibility to infection.

There is potential for prolonged sensory blockade with peripheral nerve blocks, although there is no evidence that patients are at increased risk of permanent nerve injury following regional anaesthesia, and full sensory function is usually returned within 24 hours.

Postoperative care

Postoperative care will depend on type of surgery and disease severity. Postoperative ventilation may be required given the high risk of postoperative respiratory failure, and admission to High Dependency or the Intensive Care Unit may be indicated.

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Continuous ECG monitoring or invasive monitoring is indicated in the postoperative period in those with cardiac manifestations of the disease. It is important to maintain euvolaemia throughout the peri- and postoperative period to avoid renal crises or pulmonary oedema.

Patients are at significant risk of developing scleroderma renal crisis (SRC) in the postoperative period. This may present with an acute onset and or a progressive worsening of arterial hypertension (>150/85 mmHg confirmed by at least 2 different measurements) and oligo/anuria. They may go on to develop thrombotic microangiopathy, thrombocytopenia, and haemolytic anaemia. Patients with diffuse cutaneous scleroderma lasting less than 5 years are of particular risk and, if suspected, the diagnosis may be confirmed by measuring serum haptoglobin and schizocyte levels following discussion with haematology.

Mobilisation can prove difficult due to contractures, malnutrition and prolonged sensory blockade from regional anaesthesia and additional assistance may be required.

Thromboembolic stockings should be avoided due to peripheral vascular disease and risk of ischaemia.

Postoperative analgesia should avoid non-steroidal anti-inflammatory drugs and opiates should be used with caution.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease

Vasospastic crisis may be triggered by stress, pain, dehydration, exposure to cold and use of inotropic drugs. It can present with severe pain, hypertension, tachycardia and peripheral ischaemia. Calcium channel blockers are usually the first-line treatment, although iloprost and sildenafil may also be effective. Regional anaesthesia can provide good pain relief in addition to vasodilatation. A sympathectomy of the affected area may be required.

All anaesthetists anaesthetising patients with SSc should be familiar with difficult and failed intubation protocols.

Ambulatory anaesthesia

Availability of ambulatory anaesthesia will be guided by severity of disease, surgical procedure and local guidelines. It is unlikely to be appropriate in any but the mildly affected.

Obstetrical anaesthesia

Systemic sclerosis does not usually affect fertility, but there is a high incidence of miscarriage, stillbirth and premature labour. The disease is accelerated in 50% of cases and women with widespread multi-organ involvement may be counselled against continuation of pregnancy.

Renal scleroderma may present as pre-eclampsia, is differentiated by raised plasma renin, and treated with angiotensin-converting enzyme inhibitors.

Pregnant women with systemic sclerosis should have experience in obstetric lead care and multidisciplinary team involvement. Early epidural anaesthesia is recommended for labour as there is a high risk of obstructive labour and need for expedient operative delivery.

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Last date of modification: August 2015

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Herausgeber

DGAI
Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V.
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Verlag & Druckerei

Aktiv Druck & Verlag GmbH
An der Lohwiese 36 |
97500 Ebelsbach | Deutschland
www.aktiv-druck.de

Geschäftsführung
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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) (inkl. 7 % MwSt.) 258,- €
  Schweiz 266,- €
  Rest der Welt 241,- €

Mitarbeiter aus Pflege, Labor, Studenten und Auszubildende (bei Vorlage eines entsprechenden Nachweises)
Europa (ohne Schweiz) (inkl. 7 % MwSt.) 94,- €
Schweiz 90,- €
Rest der Welt 94,- €

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