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

www.ai-online.info

60. Jahrgang | Juli/August 2019

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)

Behcet's disease

Biliary atresia

orphanaesthesia

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

SUPPLEMENT NR. 10 | 2019

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:

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

Projektleitung
Prof. Dr. Tino Münster, MHBA
Chefarzt
Klinik für Anästhesie und operative Intensivmedizin
Krankenhaus Barmherzige Brüder Regensburg
Prüfeninger Straße 86
93049 Regensburg,
Deutschland
Tel.: 0941 369-2350
E-Mail: Tino.Muenster@barmherzige-regensburg.de

DGAI
Deutsche Gesellschaft für Anästhesiologie & Intensivmedizin
www.dgai.de

A&I
ANÄSTHESIOLOGIE & INTENSIVMEDIZIN
www.ai-online.info

orphananesthesia

Anaesthesia recommendations for

Behcet's disease

Disease name: Behcet's disease

ICD 10: M35.2

Synonyms: Behcet's syndrome, BD

Disease summary: Behcet's disease is a multisystem inflammatory vasculitis of unknown aetiology, characterised by relapsing episodes of painful oral aphtous ulcers, genital ulcers, skin lesions, ocular lesions, neurological and vascular involvement [1] [2] [3]. The disease was initially described by the Turkish dermatologist Hulushi Behcet [4] and is mostly prevalent along the silk route from China to Mediterranean countries [5]. Although genetic and environmental factors are considered in its pathology, the symptoms and severity vary as per age and sex of the patient. One of the biggest contributors to morbidity and mortality is the predisposition for thrombosis and aneurysms which can occur at unusual sites such as the mesenteric and cerebral vasculature. The mainstay of the treatment involves the use of immunosuppression with or without anticoagulation in the setting of thrombosis.

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:** Gupta A: Behcet's disease. Anästh Intensivmed 2019;60:S434–S439.
DOI: 10.19224/ai2019.S434

Typical surgery

Ophthalmological procedures: Examination under anaesthesia, trabeculectomy in case of glaucoma.

Anaesthesia for MRI: MRI brain for parenchymal and nonparenchymal involvement [7].

Type of anaesthesia

General anaesthesia is usually the choice in paediatric population.

Regional anaesthesia is contraindicated in patients with CNS Behcet's disease.

Skin and mucosa puncture for nerve blocks and other regional procedures may predispose the patient to inflammation and nodule formation.

Necessary additional pre-operative testing (beside standard care)

Biopsy of the lesion for histopathological examination shows plasma and lymphocytic infiltration in epidermis and dermis with IgM and complement C3 deposit in dermis.

Ophthalmological examination for uveitis including dilatation, fundoscopy and slit lamp examination to assess extent of uveal tissue involvement.

Consider MRA or angiography to evaluate for aneurysms, especially involving the pulmonary and cerebral vasculature.

Particular preparation for airway management

To determine the correct size of endotracheal tube, USG confirmation of tracheal diameter may help [11].

Gentle laryngoscopy and intubation is advised as oral ulcers and inflamed gums may bleed while securing airway.

Avoid supraglottic airway device as its pressure on the airway may lead to post-operative ulcer and nodule formation.

Particular preparation for transfusion or administration of blood products

Avoid multiple pricks for intravenous access as patients will typically exhibit pathergy with evolution of papules over 24–48 hours.

© Anästh Intensivmed 2019;60:S434–S439 Aktiv Druck & Verlag GmbH

Particular preparation for anticoagulation

Lower extremity vein thrombosis (LEVT) is frequently followed by vena cava thrombosis, pulmonary artery aneurysm (PAA) and Budd-Chiari syndrome [12,13]. Anti-coagulant like warfarin is often started to avoid the risk of major vessel thrombosis, but there are no controlled data available to support it. Continuing immunosuppression is more effective than anticoagulants alone to prevent recurrent thrombosis [14]. Arterial involvement is seen as aneurysms. It is mostly found in pulmonary artery circulation and is also the leading cause of death in BD patients [15].

Particular precautions for positioning, transportation and mobilisation

Full padding of all the pressure points and joints, particularly eye care, is important as the presence of uveitis and glaucoma predisposes to raised intraocular pressure.

Interactions of chronic disease and anaesthesia medications

Steroid supplementation is to be continued till the day of surgery.

Colchicine – It enhances the effect of CNS depressants and sympathomimetics. Prolonged administration may lead to a depression of the medullary respiratory centre [16].

Azathioprine – The dose needs to be lowered in patients with kidney impairment. It also antagonises NDMRs and potentiates neuromuscular blockage by succinylcholine [17]. There is also a risk of bleeding due to thrombocytopenia.

Cyclosporine – There is a risk of increased neuromuscular blockage after NDMRs. As a result, doses need to be reduced [18].

Cyclophosphamide – The action of succinylcholine is increased because cyclophosphamide acts as pseudocholinesterase inhibitor [19].

Anti-TNF-α agents – Induce P450 enzymes, resulting in a decreased concentration of anaesthetic drugs requiring dose adjustments, namely induction agents, benzodiazepine and opioids.

Anaesthetic procedure

Children can be premedicated with an oral premedicant like midazolam, promethazine to avoid separation anxiety and crying. It is better to avoid succinylcholine as it raises IOP in children presenting with ocular symptoms with glaucoma.

Care should be taken during mask ventilation and intubation as there is risk of trauma to oedematous and inflamed tissue. These traumata may cause bleeding and further exacerbate nodule formation and ulceration in the future.

There is no special consideration regarding the usage of inhalational agents or induction agents, but NDMRs may need dose adjustments in patients on azathioprine and cyclosporine.

Regional procedure including nerve blocks is not preferred in Behcet's disease.

© Anästh Intensivmed 2019;60:S434–S439 Aktiv Druck & Verlag GmbH

Particular or additional monitoring

Neuromuscular monitoring can be used to guide the dose of NDMRs.

Possible complications

In patients with thrombosis of the larger veins, there is a risk of pulmonary embolism and migration of thrombus. The usage of a tourniquet should be guarded and be done only after ruling out DVT by Doppler studies.

Difficult airway is a possibility in paediatric population with oral ulcers and inflammation. Bleeding in oral cavity during laryngoscopy needs to be taken care of while securing the airway.

A prolonged effect of muscle relaxant may lead to delayed recovery.

Long-term complications of BD like nodule, ulcer formation and healing problems may result in post-operative mucosal fibrosis and narrowing of orotracheal tract [20].

Post-operative care

Effective pain control to avoid crying and rise in IOP patients must be given.

DVT prophylaxis to be continued in patients with risk of lower extremity venous thrombosis.

Disease-related acute problems and effect on anaesthesia and recovery

Desaturation and hypoxia may be due to pulmonary thromboembolism, other causes which need to be ruled out are endotracheal disconnection, kinking, mucosal plug and dislodgement.

Ambulatory anaesthesia

With patients who need to be discharged on the same day, prolonged neuromuscular weakness and respiratory depression due to drug interactions should be avoided. Injection of atracurium and cisatracurium is the relaxant of choice. The patient should also be monitored for respiratory depression effect of opioid like fentanyl, morphine etc. Acetaminophen and NSAIDs are the analgesics of choice to be used in the postoperative period.

Obstetrical anaesthesia

Since the antenatal period is a hypercoagulable state, the risk of thromboembolism and placental insufficiency gradually increases until the time of delivery. Non-compression stocking, steroid therapy etc., is to be continued until delivery. In some cases, the coagulation profile needs to be assessed to guide the choice of anaesthesia [21].

© Anästh Intensivmed 2019;60:S434–S439 Aktiv Druck & Verlag GmbH

References

1. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet 1990;335:1078–1080
2. International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD). The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol 2014;28:338–347
3. Koné-Paut I, Shahram F, Darce-Bello M, Cantarini L, Cimaz R, Gattorno M, et al. Consensus classification criteria for paediatric Behçet's disease from a prospective observational cohort: PEDBD. Ann Rheum Dis 2015. DOI:10.1136/annrheumxdis-2015-208491
4. Behçet H. Über rezidivierende, aphthöse, durch ein Virus verursachte Geschwüre am Mund, am Auge und an den Genitalien. Dermatol Wochenschr 1937;105:1152–1157
5. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, Akhlaghi M, et al. Behcet's disease: from East to West. Clin Rheumatol 2010;29:823-833
6. Mendes D, Correia M, Barbedo M, et al. Behçet's disease – a contemporary review. J Autoimmun 2009;32:178–188
7. Akman-Demir G. Behcet hastalığında nörolojik tutulum. Turkderm 2009;43 (Supplement 2): 61–64
8. Riera-Mestre A, Martínez-Yelamos S, Martínez-Yelamos A, Ferrer I, Pujol R, Vidaller A. Clinicopathologic features and outcomes of neuro-Behçet disease in Spain: a study of 20 patients. European J Intern Med 2010;21:536–541
9. Mizuki N, Inoko H, Ohno S. Pathogenic gene responsible for the predisposition to Behcet's disease. Int Rev Immunol 14;1:33–48
10. Kaya IT. Genetics of Behcet's disease. Pathol Res Int 2012;2012:912589. DOI: 10.1155/2012/912589
11. Shibasaki M, Nakajima Y, Ishii S, Shimizu F, Shime N, Sessler DI. Prediction of pediatric endotracheal tube size by ultrasonography. Anesthesiology 2010;113:819–824
12. Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-term mortality and morbidity of Behcet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. Medicine (Baltimore) 2003;82:60–76. DOI: 10.1097/00005792-200301000-00006
13. Melikoglu M, Ugurlu S, Tascilar K, et al. Large Vessel Involvement in Behcet's Syndrome: A Retrospective Survey. Ann Rheum Dis 2008;67(Suppl II):67
14. Ahn JK, Lee YS, Jeon CH, Koh EM, Cha HS. Treatment of venous thrombosis associated with Behcet's disease: immunosuppressive therapy alone versus immunosuppressive therapy plus anticoagulation. Clin Rheumatol 2008;27;2:201–205
15. Seyahi E. Behçet's disease: How to diagnose and treat vascular involvement. Best Pract Res Clin Rheumatol 2016;30:279–295
16. Balek RW, Kocsis JJ, Geiling EM. Potentiation of several hypnotic and anesthetic agents by colchicine. Arch Int Pharmacodyn Ther 1957;111:182–190
17. Dretchen KL, Morgenroth VH, Standaert FG, et al. Azathioprine: effects on neuromuscular transmission. Anaesthesiology 1976;45:604–609
18. Wood CG. Cyclosporine-vecuronium interaction. Can J Anaesth 1989;36:358–366
19. Koseoglu V, Chiang J, Chan KW. Acquired pseudocholinesterase deficiency after high-dose cyclophosphamide. Bone Marrow Transplant 1999;24:1367–1368
20. Turner ME. Anaesthetic difficulties associated with Behçet's syndrome. Case report. Br J Anaesth 1972;44:100–102
21. Martineau M, Haskard DO, Nelson-Piercy C. Behçet's syndrome in pregnancy. Obstet Med 2010;3:2–7.

Date last modified: October 2017

This recommendation was prepared by:

Author(s)

Anurag Gupta, Associate consultant anaesthesiology, VPS Rockland Hospital, New Delhi, India
guptaanurag80@gmail.com

Disclosure(s) The authors have no financial or other competing interest to disclose. This recommendation was unfunded.

This recommendation was reviewed by:

Reviewer 1

Jan AM van Laar, Section of Clinical Immunology, Departments of Internal Medicine and Immunology, Erasmus Medical Centre, Rotterdam, The Netherlands
j.vanlaar@erasmusmc.nl

Reviewer 2

William Johnson, Internal Medicine Resident, Lankenau Medical Center, Wynnewood, PA, USA
williamjo@pcom.edu

Disclosure(s) The reviewers have no financial or other competing interest to disclose.

Please note that this recommendation has been reviewed not by an anaesthesiologist and a second disease expert, as usual, but by two disease experts instead.

© Anästh Intensivmed 2019;60:S434–S439 Aktiv Druck & Verlag GmbH

Herausgeber

DGAI
Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V.
Präsident: Prof. Dr. R. Rossaint, Aachen

BDA
Berufsverband Deutscher Anästhesisten e.V.
Präsident: Prof. Dr. G. Geldner, Ludwigsburg

DAAF
Deutsche Akademie für Anästhesiologische Fortbildung e.V.
Präsident: Prof. Dr. F. Wappler, Köln

Korrespondenzadresse: Roritzerstraße 27 | 90419 Nürnberg | Deutschland
Tel.: 0911 9337812 | Fax: 0911 3938195
E-Mail: anaesth.intensivmed@dgai-ev.de

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
Pia Engelhardt
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
E-Mail: jdistler@bda-ev.de

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 2019
Der 60. 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,- ¡

Für Mitglieder der DGAI und/oder des BDA ist der Bezug der Zeitschrift im Mitgliedsbeitrag enthalten.

Allgemeine Geschäfts- und Lieferbedingungen
Die allgemeinen Geschäfts- und Lieferbedingungen entnehmen Sie bitte dem Impressum auf www.ai-online.info

Indexed in **Current Contents®/Clinical Medicine, EMBASE/Excerpta Medica; Medical Documentation Service; Research Alert; Sci Search; SUBIS Current Awareness in Biomedicine; VINITI: Russian Academy of Science.**

Die Beiträge aus der A&I finden Sie online unter: **www.ai-online.info**

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

German Society of Anaesthesiology and
Intensive Care Medicine
Ursula Homberg
Roritzerstrasse 27 | 90419 Nuremberg | Germany
Tel.: +49-911-9337828 | Fax: +49-911-3938195
Email: uhomberg@orphananesthesia.eu