

# Anaesthesia recommendations for patients suffering from

## Behcet's disease

**Disease name:** Behcet's disease

**ICD 10:** M 35.2

**Synonyms:** Behcet's syndrome, BD

Behcet's disease is a multisystem inflammatory vasculitis of unknown etiology, characterised by relapsing episodes of painful oral aphthous ulcers, genital ulcers, skin lesions, ocular lesions, neurological and vascular involvement [1] [2] [3]. The disease was initially described by Turkish dermatologist Hulusi 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 varies 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 mesenteric and the cerebral vasculature. The mainstay of the treatment involves the use of immunosuppression with or without anticoagulation in the setting of thrombosis.

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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)**

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### **Typical surgery**

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Ophthalmological procedures: Examination under anaesthesia, trabeculectomy for glaucoma.

Anaesthesia for MRI: MRI brain for parenchymal and non parenchymal involvement [7]

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### **Type of anaesthesia**

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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 procedure may predispose the patient to inflammation and nodule formation.

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### **Necessary additional diagnostic procedures (preoperative)**

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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.

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### **Particular preparation for airway management**

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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 op ulcer and nodule formation.

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

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

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### **Particular preparation for anticoagulation**

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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 rather continuing immunosuppression is more effective than anticoagulants alone to prevent recurrent thrombosis [14]. Arterial involvement is seen as aneurysms which is mostly found in pulmonary artery circulation and is also the leading cause of death in BD patients [15].

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

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

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

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Steroid supplementation to be continued till the day of surgery.

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

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

Cyclosporine- there is a risk of increase neuromuscular blockage after NDMR, as a result doses needs to be reduced [18].

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

Anti TNF  $\alpha$  agents – induces P450 enzyme, resulting in decreased concentration of anaesthetic drugs requiring dose adjustments, namely induction agents, benzodiazepam and opioids.

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### **Anaesthesiologic procedure**

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Children can be premedicated by oral premedicant like midazolam, phenargan 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 edematous and inflamed tissue. These trauma may cause bleeding and further exacerbate nodule formation and ulceration in future.

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

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

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### **Particular or additional monitoring**

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

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### **Possible complications**

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In patients with thrombosis of larger veins there is a risk of pulmonary embolism and migration of thrombus. The usage of 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.

Prolong effect of muscle relaxant may lead to delayed recovery.

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

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### **Postoperative care**

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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.

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

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

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### **Ambulatory anaesthesia**

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Patients who need to be discharged on the same day, prolong neuromuscular weakness and respiratory depression due to drug interactions should be avoided. Inj atracurium and cis atracurium is the relaxant of choice. The patient should also be monitored for respiratory depression effect of opioid like fentanyl, morphine etc. Acetylamino phen and NSAIDS are the choice of analgesics for use in post-operative period.

## **Obstetrical anaesthesia**

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

## Literature and internet links

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**Peer revision 1**

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**Peer revision 2**

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*Please note that this guideline has not been reviewed by two anaesthesiologists, but by two disease experts instead.*

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