orphanaesthesia

Anaesthesia recommendations for patients suffering from

Huntington's disease

Disease name: Huntington's disease

ICD 10: G10

Synonyms: Huntington's chorea

Disease summary:

Huntington's disease (HD) is a progressive neurodegenerative disease of the central nervous system, caused by an assumed toxic gain-of-function mutation of the Huntingtin gene on chromosome 4. This mutation, an increase in the number of cytosine-adenine-guanine (CAG) trinucleotide repeats in the gene's coding portion, creates a polyglutamine region in the Huntingtin protein. This region alters the protein's function, and this is believedto lead to neuronal degeneration, particularly affecting the basal ganglia. Greater than 40 CAG repeats produce fully penetrant disease. HD is inherited in an anticipatory autosomal dominant pattern; that is, the number of CAG repeats can increase during spermatogenesis, leading to earlier onset of disease and more rapid progression. Genetic anticipation in HD is therefore a paternal inherited feature. The effects of Huntington's disease include uncontrollable choreaoathetoid movements, dystonia, dysphagia, dysarthria, and psychiatric disease including depression, mania, obsessive-compulsive disorder, and dementia.. The disease is progressive, and the time from onset to death ranges from ten to 30 years.

Medicine 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

Typical surgery

There is no typical surgery for patients suffering from Huntington's disease. There are trials underway examining whether deep brain stimulation is of benefit for patients' motor symptoms. A case series from the Mayo Clinic included general surgical, orthopaedic, dental, thoracic, and ENT procedures. Some families may elect to place gastrostomy tubes as the patient's dysphagia worsens.

Type of anaesthesia

There are no absolute contraindications to any particular anaesthetic plan for patients with Huntington's. Regional anaesthesia may be challenging if the patient is unable to cooperate.

Case reports have associated HD with prolonged recovery from benzodiazepines and barbiturates, as well as with increased duration of paralysis after administration of both depolarizing and non-depolarizing neuromuscular blocking drugs (NMBDs).

At least one genetic study has found an increased incidence of atypical pseudocholinesterase in patients with HD.

However, a review of eleven patients with HD who underwent seventeen general anesthetics, by Kivela *et al* of the Mayo Clinic, did not find any atypical reactions to midazolam, sodium thiopental, succinylcholine, nor non-depolarizing NMBDs. Prolonged sedation observed after benzodiazepines and barbiturates in prior reports was attributed by these authors to relative overdosing of these drugs, not abnormal patient response.

While no abnormal response to NMBDs was observed, the authors recommended caution with succinylcholine given the association with atypical pseudocholinesterase. Succinylcholine may still be needed, however, due to the possible increased risk of aspiration in these patients.

Propofol sedation has been described as effective for monitored anesthesia care, though, again, aspiration risk and possible noncompliance with NPO guidelines must be considered.

Necessary additional diagnostic procedures (preoperative)

Diagnostic testing should be performed on a case-by-case basis based on the patient's medical history. Huntington-specific workup up includes genetic testing which confirms the diagnosis, genetic counselling for patient and family, brain imaging, psychiatric and neurological evaluations, and a swallow study when indicated. Aspiration should be investigated in the setting of acute worsening of respiratory status.

Particular preparation for airway management

Airway anatomy should be assessed as thoroughly as possible given the patient's ability to cooperate with an exam. If available, prior anaesthetic records should be reviewed. Some degree of aspiration risk may be present, which should be taken into consideration during induction and intubation.

Particular preparation for transfusion or administration of blood products

Patients with HD may receive blood products as appropriate given their medical, obstetrical, and prior transfusion histories. Patients with HD may donate blood without risk to potential recipients.

Particular preparation for anticoagulation

No particular reports exist regarding problems with perioperative anticoagulation for these patients. As the disease progresses and motility worsens, prophylaxis for venous thromboembolism can be provided.

Particular precautions for positioning, transport or mobilisation

Dystonia can make intraoperative positioning challenging, and care should be taken to minimize risk of peripheral nerve or musculoskeletal injury. Physical, occupational, and speech therapy play an important role in long-term management.

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

Common medications for patients with HD including antipsychotics, benzodiazepines, antiepileptics such as levetiracetam, and tetrabenazine, which help to suppress choreathetoid movements by depleting dopamine stores. Tetrabenazine also depletes serotonin, norepinephrine, and histamine stores, which could have variable effects on anaesthetic requirements and hemodynamic response to anaesthesia.

Anaesthesiologic procedure

As described above, there are no absolute contraindications to any particular anaesthetics or anaesthetic techniques. Historically, questions have been raised related to a possible prolonged effect with succinylcholine, possibly due to an associated between HD and atypical pseudocholinesterase. A sensitive and careful approach must be taken, sometimes with the aid of the patient's family member, given the motor and psychiatric difficulties present later in the disease process. Patients may have difficulty remaining still and/or cooperating with the anaesthesiology team. Due to the frailty and poor nutritional status of these patients, anaesthetic requirements may be reduced.

Particular or additional monitoring

Additional monitoring is at the discretion of the anaesthesiology team, based upon the physiologic derangements anticipated due to the surgery, the presence or absence of dysautonomia, and patient's other underlying medical conditions.

Possible complications

An awareness of aspiration risk should be maintained due to uncertainty of adherence to fasting guidelines, as well as due to the frequency of dysphagia in advanced HD. Patients with HD can suffer from pronounced autonomic dysfunction, and medications should be available to manage swings in blood pressure while under anesthesia. Otherwise, risk assessment is based on the surgery and the patient's other underlying medical conditions.

Postoperative care

Routine postoperative monitoring and care is adequate, though patients may benefit from a calm setting with a family member at bedside to assist in redirection.

Information about emergency-like situations / Differential diagnostics

Vigilance must be maintained over the intermediate-term postoperative period with regards to signs and symptoms of pneumonia, as aspiration pneumonia can occur in the later stages of this disease.

Ambulatory anaesthesia

These patients may undergo ambulatory anaesthesia, keeping in mind the complexity of care required later in the disease's course. If they reside in a skilled nursing facility, careful coordination is required between that facility's medical team and the teams providing care for the procedure.

Obstetrical anaesthesia

There is not much literature available regarding obstetrical anaesthesia for patients with HD. A letter to the editor of the Journal of Clinical Anesthesia by Draisci et al, describing a single case of high spinal leading to hypotension in a patient with HD, suggests using caution with neuraxial techniques, though the patient described in this case had undergone an uneventful prior neuraxial anesthetic. It is possible that the autonomic dysfunction present in the later course of HD played a role in this patient's hypotension.

Literature and internet links

1. Huntington G. On chorea. Med Surg Rep 1872,26:317
2. Walker FO. Huntington's disease. The Lancet 2007,369:218
3. Rodrigo MRC. Huntington's chorea: midazolam, a suitable induction agent? Br J Anaesth 1987,59:388
4. Davies DD. Abnormal response to anesthesia in a case of Huntington's chorea. Br J Anaesth 1966,38:490
5. Whittaker M, Berry M. The plasma cholinesterase variants in mentally ill patients. Br J Psychiatry 1977,130:397
6. Kivela JE, Sprung J, Southorn PA, Watson JC, Weingarten TN. Anesthetic management of patients with Huntington disease. Anesth Analg 2010;110:515
7. MacPherson P, Harper I, MacDonald I. Propofol and remifentanil total intravenous anesthesia for a patient with Huntingtons Disease. J Clin Anesth 2004;16:537
8. Kaufman MA, Erb T: Propofol for patients with Huntington chorea? Anaesthesia 1990;45:889
9. Holland R. Huntington's chorea and anaesthesia. Anaesth Intensive Care 1992;20:256
10. Schramm BM, Orser BA: Dystonic reaction to propofol attenuated by benztropine (Cogentin). Anesth Analg 2002;94:1237
11. White T, Neustein S. Monitored anesthesia care for a patient with advanced Huntington's chorea. Middle Eastern Journal of Anesthesiology 2013 Jun;22(2):185-6
12. Draisci G, Sbaraglia F, Pinto R, Zanfini BA, Frassanito L, Catarci S. Does Huntington's disease enhance cephalad spread during neuraxial anesthesia for cesarean section? 2012 Sep;24(6):516-7
13. "Xenazine (tetrabenazine) Tablets, for Oral Use. Full Prescribing Information. Revised: 6/2015" (PDF). H. Lundbeck A/S. Retrieved 6 March 2017
14. Abildtrup, M., & Shattock, M. (2012). Cardiac Dysautonomia in Huntington's Disease. Journal of Huntington's Disease, 2(3), 251–261. http://doi.org/10.3233/JHD-130054.

Last date of modification: June 2017

This guideline has been prepared by:

Author
Taylor White, Anaesthesiologist, Icahn School of Medicine at Mount Sinai, New York, USA
taylor.white@mountsinai.org

Peer revision 1
Saleem Altaf Wani, Department of Anesthesia & Critical Care, Esi-Post Graduate Institute of Medical Science & Research, New Delhi, India
drsaleemwani@gmail.com

Peer revision 2
David G. Anderson, Consultant Neurologist, MBBCh MMED FC Neurol, University of the Witwatersrand, Donald Gordon Medical Centre, Johannesburg, South Africa
david@neuro.joburg