orphanaesthesia

Anaesthesia recommendations for patients suffering from

Arrhythmogenic right ventricular dysplasia

Disease name: Arrhythmogenic right ventricular dysplasia

ICD 10: I42.8

Synonyms: arrhythmogenic right ventricular cardiomyopathy

Disease summary: Arrhythmogenic Right Ventricular Dysplasia (ARVD), also known as arrhythmogenic cardiomyopathy, is an inherited cardiomyopathy. It is disproportionately highly represented in cases of unexplained death under anesthesia, but there are few published reports of the safe anaesthetic management of patients with ARVD.Arrhythmogenic right ventricular dysplasia is a form of genetic cardiomyopathy that is a well-recognised cause of ventricular tachycardia and sudden death, particularly in athletes and the young. ARVD is also one of the leading causes of death during anaesthesia. In one reported analysis of forensic autopsies, eighteen out of fifty perioperative deaths in ASA 1 patients undergoing surgical procedures, usually associated with extremely low mortality, have previously been found at autopsy to show histological features of ARVD. None of these patients had any prior cardiac history. Four of the patients died at induction of anaesthesia, nine during the surgical procedure and five within 2 hours after surgery was complete.

The estimated prevalence of ARVD in the general population is about 1 in 5,000. Young men are affected approximately three times more frequently than women, with a higher prevalence (up to 0.8%) reported in certain parts of Italy (Veneto) and Greece (Naxos Island). At a prevalence rate of 1 in 5,000, most anaesthetists will encounter patients with ARVD, in some of whom the diagnosis may not have been established. The histological demonstration of myocyte atrophy and transmural fibro-fatty replacement of right ventricular myocardium is typical for this condition. Immunohistochemistry for plakoglobin may also add to the accuracy of this diagnosis. Interestingly, in 70% of the cases the left ventricle is also involved. ARVD is caused by mutation in the desmosomal proteins Plakophillin 2 (PKP2), Desmoglein 2 (DSG2), Desmocollin 2 (DSC2), Desmoplakin (DSP), and Plakoglobin (JUP), and others.

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

Typical surgery

Patients can present for any type of surgery, from minor dental and gynaecological conditions to major vascular and cardiac operations.

Type of anaesthesia

Both general and regional anaesthesia have been described for management of these patients. There is scarcity of data on the safest anaesthetic techniques for these patients. Avoidance of particular anaesthetic agents has been recommended, although the evidence base for these recommendations has been anecdotal. The use of agents like succinylcholine and volatile anaesthetics has been discouraged. However, the maintenance of optimal haemodynamic conditions for myocardial perfusion may be the best protective strategy against ventricular fibrillation in these patients.

Use of invasive monitoring perioperatively has been recommended. Pulmonary artery catheter (PAC) placement may lead to ventricular fibrillation due to mechanical stimulation of the right ventricular structures and the risk versus benefit should be carefully considered.

Necessary additional diagnostic procedures (preoperative)

It has to be highlighted that in many of the patients with ARVD presenting for surgery the diagnosis may not be known pre-operatively, and hence no specific investigations done. Patients with known and symptomatic ARVD often have implantable defibrillator (ICD) devices in-situ. It is important to continue anti-arrhythmic medication perioperatively.

Particular preparation for airway management

Not reported.

Particular preparation for transfusion or administration of blood products

Not reported.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Not reported.

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

Continue anti-arrhythmic medication perioperatively. If an ICD or pacemaker is known to be in-situ, ensure that its function is checked pre-operatively and that the device is switched to a mode compatible with the use of intra-operative diathermy.

Anaesthesiologic procedure

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Particular or additional monitoring

Invasive blood pressure monitoring as well as the usual non-invasive monitoring for surgery e.g. ECG are considered mandatory.

External defibrillation and pacing equipment should be available and connected via defibrillation / external pacing pads to the patient prior to induction of anaesthesia.

Possible complications

Ventricular arrhythmias.

Postoperative care

Postoperative ECG and invasive haemodynamic monitoring is recommended until cardiovascular stability has been established for an adequate period post-operatively, commensurate with the duration and type of surgical procedure performed.

Information about emergency-like situations / Differential diagnostics

Patients with known ARVD should always have a consultation with a cardiologist specialising in electrophysiology pre-operatively. The most likely perioperative complication is ventricular arrhythmia and treatment protocols for ventricular tachycardia or ventricular fibrillation should be available and followed.

Ambulatory anaesthesia

Due to the high incidence of morbidity in patients with known ARVD undergoing surgery, ambulatory anaesthesia is not recommended; these patients should be admitted as in-patients and monitored post-operatively as discussed above.

Obstetrical anaesthesia

Similar to other types of surgery.

Literature and internet links

1. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes. N Engl J Med 1998; Aug 6;339(6):364-9
2. Staikou C, Chondrogiannis K, Mani A. Perioperative management of hereditary arrhythmogenic syndromes. Br J Anaesth 2012; May 108(5):730-44
3. Alexoudis AK, Spyridonidou AG, Vogiatzaki TD, Iatrou CA. Anaesthetic implications of arrhythmogenic right ventricular dysplasia/cardiomyopathy. Anaesthesia 2009 Jan;64(1):73-8
4. Valchanov K, Goddard M, Ghosh S. Anesthesia for heart transplantation inpatients with arrhythmogenic right ventricular dysplasia. J Cardiothorac Vasc Anesth 2014; Apr 28(2):355-7.

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