orphanaesthesia

Anaesthesia recommendations for patients suffering from

Catecholaminergic polymorphic ventricular tachycardia

Disease name: Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

ICD 10: I47 (Paroxysmal tachycardia), I47.2 (Ventricular tachycardia), ORPHA3286

Synonyms: Catecholamine-induced polymorphic ventricular tachycardia, bidirectional tachycardia induced by catecholamines, familial polymorphic ventricular tachycardia (FPVT), stress-induced polymorphic ventricular tachycardia

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare genetic disease with an incidence of approximately 1:10,000 in European population and a very high mortality rate if left untreated, reaching 31% by the age of 30 years [1]. It is characterized by bidirectional or polymorphic ventricular tachycardias (VT) induced by an adrenergic triggering factor, such as emotional stress or physical activity, while some patients may also develop episodes of atrial fibrillation [1,2]. Stress-related syncope is the most common symptom in otherwise healthy children or adolescents, while sudden cardiac death due to degeneration of VT to ventricular fibrillation (VF) may also occur. Palpitations and dizziness during exercise represent more benign manifestations of the disorder [3].

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

Disease summary

A family history of syncopal episodes or sudden cardiac death exists in 30-35% of patients [4]. Sometimes the condition is misdiagnosed and mistreated as epilepsy in patients with syncope, especially if accompanied by incontinence [5].

Genetic mechanisms include mutations in the cardiac ryanodine receptor gene (RyR2) – calcium ions release channel – which are found in about 50% of CPVT patients and are responsible for the autosomal dominant inheritance of the disorder, known as CPVT1. On the other hand, mutations in cardiac calsequestrin gene (CASQ2) – a major calcium binding protein within sarcoplasmic reticulum – are the cause of the rare autosomal recessive type, described as CPVT2 [1,3,4,6]. In both types, there is abnormal release of calcium ions from the sarcoplasmic reticulum causing asynchrony in cardiac excitation and contraction [3]. As a result of calcium excess, delayed after-depolarizations and triggered activity constitute the pathophysiologic background of the disease [4].

Regarding diagnosis, patients with CPVT have normal resting electrocardiograms (ECGs), without QT prolongation or any other characteristic abnormality. Additionally, there are no echocardiographic or other imaging signs of cardiac structural abnormalities [1]. The tilt table testing is negative. Apart from the bidirectional VT with the alternating direction of QRS complexes from beat-to-beat, another distinctive marker of the disease is the conversion of premature complexes to malignant ventricular arrhythmias as the workload level increases; VT/VF appear frequently at heart rates of 110-130/min [2,4]. Induction and progressive deterioration of ventricular arrhythmias during stress testing or isoproterenol infusion are diagnostic hallmarks of CPVT [1,4]. Exercise stress tests and ECG Holter recordings (24h) represent invaluable diagnostic tools [3,4].

Treatment modalities, apart from avoidance of forceful physical activity, competitive athletics, and emotional stress, include beta blocker therapy (sotalol, nadolol, metoprolol) at doses titrated to achieve maximal efficacy [3]. Nadolol is the preferred beta blocker, due to its long duration of action [1]. Compliance to therapy is of vital importance, even for those with positive genetic but negative exercise tests, as missing doses may induce malignant arrhythmias [1,3]. Unfortunately, beta blocker therapy may not be enough to prevent cardiac events in a significant percentage of patients [1]. There is limited evidence on the advantageous addition of a calcium channel blocker, such as verapamil, while some data support the use of flecainide in addition to beta blocking therapy. Among invasive treatments, implantation of a cardioverter – defibrillator (ICD) represents a class I recommendation for CPVT diagnosed patients who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal drug therapy and/or left cardiac sympathetic denervation [7]. Nevertheless, cardiologists try to avoid ICDs in this population because of the potential for precipitating repetitive episodes of VT ("VT storm") in response to ICD shocks and consequent catecholamine release [7].

Patients with ICDs should continue beta blocker treatment for prevention of ventricular arrhythmias and any adverse/proarrhythmic effects of ICDs' discharges [1,3]. Left cardiac sympathetic denervation represents an invasive therapeutic approach which is reported to decrease significantly (>90%) the arrhythmic events [8]; it may be considered in patients with recurrent syncope or arrhythmias requiring frequent ICD shocks while on beta-blockers and in those who do not tolerate medication or continue to be symptomatic while on maximum drug doses (Class IIb) [7]. Catheter ablation is another invasive method which may be performed in patients with refractory arrhythmias; nevertheless, it is used far less often

than the other treatments, since its efficacy in CPVT has not been proved yet. Thus, catheter ablation for CPVT is not included in the relevant 2013 guidelines/recommendations [7].

Typical surgery

Placement of an ICD for prompt electrical termination of the arrhythmic episodes is used only in patients with repeated episodes of sustained VT in spite of the pharmacologic therapy outlined above. Also, left cardiac sympathetic denervation is a surgical therapeutic approach, performed via open thoracotomy or via video-assisted thoracoscopy [8, 9]. Less often, CPVT patients may undergo catheter ablation [7]. Apart from the surgical interventions performed for symptomatic treatment of the disease per se, paediatric or adult patients with CPVT may actually undergo any type of scheduled or emergency surgery due to other causes. For safety reasons, even minor surgical procedures should be performed in a hospital setting and under ECG monitoring [10].

Type of anaesthesia

General anaesthesia, with or without neuromuscular blockade and the use of an endotracheal tube or a supraglottic airway device, is typically administered for the invasive therapeutic interventions, such as left cardiac sympathetic denervation and ICD placement, respectively [2,9]. Although disease-specific data are lacking, ICD implantation could also be performed under local anaesthesia combined with IV anxiolysis, analgesia and sedation (monitored anaesthesia care), [11]. Nevertheless, there are some issues that probably render this option less attractive for CPVT patients, since it is of utmost importance that any adrenergic/stress factors are adequately suppressed, while deep sedation/sufficient analgesia should be achieved before device testing, because the shocks may be very painful [11].

General, neuraxial and local anaesthesia have all been used in patients with CPVT undergoing various surgical procedures [2,9,10,12,13]. Although the relevant existing data are quite limited, no significant anaesthesia-related complications have been reported. Regardless of the anaesthetic technique, the major concern of the anaesthesiologist should be the avoidance of factors that may trigger serious arrhythmias, such as sympathetic stimulation, tachycardia and beta agonists/sympathomimetic drugs [2,5]. Thus, anxiety, emotional stress and pain should be adequately suppressed or alleviated. Also, sufficient intraoperative anaesthesia/analgesia, maintenance of oxygenation, normocarbia, normothermia and normovolaemia are extremely important. In some cases, such as very stressed paediatric patients, general anaesthesia may be required even for minor surgery. Local anaesthesia can be used when indicated, i.e. for dental procedures, after sufficient anxiolysis and/or sedation [10]. When regional or local anaesthesia is applied, epinephrine – which is commonly used as adjuvant to local anaesthetics- should be omitted [5,10,13].

Necessary additional diagnostic procedures (preoperative)

The condition of the patient should be thoroughly investigated preoperatively, especially if there is adequate time, as before elective surgery. A multidisciplinary approach involving cardiac electrophysiologist, anaesthesiologist and surgeon is required [5]. The severity and frequency of cardiac events, along with the effectiveness of current treatment should be

assessed. A detailed informative history and a 12-lead ECG are mandatory. Treatment efficacy can be assessed by exercise stress testing and ECG Holter monitoring. Electrolyte levels and acid-base status should also be checked preoperatively [2,5,9].

Suspicious cases without definite diagnosis or patients with a positive family history should be investigated via ECG, Holter monitoring, exercise stress testing and also echocardiography to rule out any cardiac structural abnormality. It should be noted though, that only genetic screening will identify asymptomatic carriers of RyR2 mutations, who should also receive beta blockers even in the presence of a negative exercise test [7].

Preoperative preparation

The clinical condition of CPVT patients should be optimized before a scheduled surgical procedure. The frequency of arrhythmic episodes should be minimized with optimization of pharmacotherapy and possible addition of invasive treatment, if indicated. It is of major importance that the beta blocker treatment is continued perioperatively, and not a single dose is omitted, since therapy discontinuation may facilitate the occurrence of lethal arrhythmia [1]. Any electrolyte abnormalities, such as hypokalaemia, hypomagnesaemia, hypocalcaemia and hypernatraemia, or acid-base disorders should be corrected [2,5,9]. Preoperative stress or anxiety that can possibly trigger catecholamine release should be adequately controlled. In this regard, reassurance and a sympathetic attitude during preoperative visit may be helpful. Also, per os premedication with a benzodiazepine, such as temazepam, the evening before and the morning of surgery is indicated for anxiolysis [2].

Particular preparation for airway management

There are no specific issues regarding possible difficulty with airway management. In this group of patients, the main problem is the catecholaminergic response to laryngeal instrumentation (laryngoscopy/tracheal intubation) which should be minimised as possible. Since supraglottic airway devices are less stimulating to the sympathetic nervous system (less invasive/better tolerated) [14], they should be preferred over tracheal tubes whenever possible (i.e. for ICD implantation).

Induction with a sufficient dose of a hypnotic agent to ensure deep anaesthesia and titrated doses of a short acting opioid (remifentanil, fentanyl, alfentanil) or a short acting beta blocker (esmolol) can be used to blunt the tachycardic response to airway instrumentation. Sympatholytic drugs, such as the centrally acting alpha-2 agonists clonidine or dexmedetomidine, may also be useful [15].

Particular preparation for transfusion or administration of blood products

No specific problems have been encountered with CPVT patients regarding transfusion or anticoagulation. The syndrome has not been associated with Hb abnormalities or coagulopathy. Transfusion of blood products and perioperative anticoagulation should be guided by the type of surgery and individual co-existing pathology.

Particular precautions for positioning, transport or mobilisation

There are no specific disease-related anatomical problems (bone, joints or skin) requiring particular precautions for positioning, transport or mobilisation. Nevertheless, changes in posture, especially if sudden, may precipitate paroxysmal arrhythmias [13]. Generally, patients should be handled gently and carefully in order to minimise the risk of hypotension/need for vasopressors in those receiving large doses of beta blockers/Ca++ channel blockers, or the stress and tachycardia in awake individuals. Intraoperatively, abrupt postural changes in the anaesthetised patient should be avoided (especially head up table tilt), while movement from one position to another (i.e from prone to supine or vice versa) should be gentle and coordinated, under close blood pressure monitoring.

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

The patients with CPVT who receive high doses of beta blockers/calcium channel blockers may have a relatively low or borderline blood pressure, which may be further reduced by general anaesthetics, especially during induction [16]. Similarly, the combination of beta-blockers with anesthetics may induce episodes of bradycardia [16]. Careful titration of anaesthetic drugs and esmolol -if used perioperatively- is required in order to achieve and maintain deep levels of anaesthesia while avoiding hypotension/bradycardia [17]. Additionally, positive pressure ventilation (IPPV) may reduce venous return/preload and thus, the tidal volume and positive end expiratory pressure (PEEP) should be optimized to maintain cardiovascular stability [2]. Regional modalities, especially subarachnoid anaesthesia, may also be complicated by significant hypotension [13].

In any case, mild to moderate hypotension should be initially managed with head down position and a bolus of IV fluids [2]. If severe hypotension occurs, the minimal effective dose of a pure alpha adrenergic agonist, such as phenylephrine, should be used [2,5]. Symptomatic bradycardia (i.e causing significant hypotension) should be managed with atropine or cardiac pacing, if persistent [2]. Finally, glucagon IV represents the last resort for treatment of a resistant beta blockade due to overdose.

Anaesthesiologic procedure

Sympathetic stimulation and tachycardia should be avoided during the whole perioperative period. Thus, before anaesthetic induction, the environment should be quiet, without loud noises/voices or stressful behaviours. After the placement of an IV line under standard monitoring, premedication with a benzodiazepine such as midazolam would be helpful [5,9]. Diazepam has also been used uneventfully [12].

Among IV anaesthetic agents, propofol has a favourable profile and has been used uneventfully for induction and maintenance of anaesthesia in patients with CPVT [2]. While thiopental has also been used without adverse effects [12], it should be mentioned that the drug has been associated with increased noradrenaline plasma concentration [15]. On the other hand, etomidate is an agent charaterized by haemodynamic stability, but data regarding its use in CPVT are lacking. Finally, ketamine is a drug that should be better avoided because of its sympathomimetic properties [5,9,15]. Among volatile agents, isoflurane and sevoflurane have been safely administered to patients with CPVT [12]. Desflurane may induce sympathetic stimulation when used for inhalational induction due to airway irritation, while fast increases in inhaled concentrations (i.e from 3% to 10%) also result in sympathetic activation and increased concentrations of epinephrine and

norepinephrine in plasma [15].Ventricular premature beats and bigeminy have been reported during enflurane anaesthesia [12]. Even though there are no relevant clinical data, halothane should be probably avoided because it has been associated with arrhythmogenicity, especially in the presence of other triggering factors [15]. This is further supported by findings suggesting that halothane increases Ca++ release from cardiac sarcoplasmic reticulum [18].

Regarding intraoperative analgesia, both fentanyl and alfentanil have been used in patients with CPVT without adverse effects [12]. Remifentanil depresses the sinus node function and atrioventricular conduction [15], while its ultra short action allows rapid and precise dose titration. It has been successfully used during induction and maintenance of anaesthesia in CPVT [2] and represents an attractive choice due to its favorable properties.

Most neuromuscular blockers, such as atracurium, vecuronium, rocuronium, alcuronium and pancuronium have been used in patients with CPVT without complications [12] The majority of non-depolarizing agents have minor direct effects on cardiac electrophysiology and could be considered as relatively safe [5]. The only exception is probably pancuronium which should be better avoided due to its vagolytic effects and subsequent risk of inducing tachycardia. Rocuronium has a mild vagolytic action which is probably not clinically significant. Cis-atracurium is possibly the agent with the safest profile, since it lacks any autonomic activity [15]. On the contrary, succinylcholine, may exert a positive or negative chronotropic action via adrenergic or muscarinic receptors of the sinus node. There is a possibility of sympathoadrenal activation and increase of noradrenaline plasma levels following administration of succinylcholine [15], while the accentuation of electrolyte disorders may represent an additional risk factor for patients susceptible to arrhythmias. Even though it has been used without complications in patients with CPVT, there is one report describing the development of VT which rapidly degenerated to VF after administration of 65 mg succinylcholine in a patient with undiagnosed CPVT undergoing electroconvulsive therapy [19]. Thus, if needed, succinylcholine should be used with caution.

It should be clarified that succinylcholine and volatiles do not affect significantly cardiac intracellular Ca++ in CPTV patients with defects in cardiac ryanodine receptor (RyR2), [12]. These individuals are not at risk of developing malignant hyperthermia, since this condition is associated with disorders in the skeletal muscle ryanodine receptor (RyR1), [5]. In this regard, the abovementioned agents are not contraindicated in CPVT patients.

Even though specific data on reversal of neuromuscular blockade in patients with CPVT are lacking, the administration of anticholinergic/anticholinesterase may result in a transient rise of the heart rate. Significant increases (i.e HR >110-130 bts/min) are not desirable in patients with CPVT. On the other hand, sugammadex seems to be devoid of significant cardiovascular side effects, since it exerts minimal or no actions on cholinesterase, nicotinic or muscarinic receptors [15]. Although there are no relative published reports, the use of rocuronium/suggamadex for induction/reversal of neuromuscular blockade seems a reasonable choice in patients with CPVT. Regarding other drugs, beta adrenergic agonists, such as isoproterenol, are clearly contraindicated in CPVT. On the contrary, supplementation of patient's beta blocker treatment may be helpful perioperatively; esmolol IV is an attractive choice due to the rapid and accurate titration to patient's needs, considering the already existing beta blockade. Also, the centrally acting alpha-2 adrenergic agonists clonidine and dexmedetomidine decrease the central sympathetic outflow and norepinephrine release and may represent useful adjuvants. On the other hand, although the uneventful administration of N2O has been reported in patients with CPVT [10,12], it should probably be used with caution because of its sympathomimetic effects and reported potential to facilitate the arrhythmogenic action of other agents, such as epinephrine and halothane [5].

Uneventful regional anaesthesia has been reported in a parturient with CPVT undergoing caesarean delivery [13]. It should be kept in mind that central neuraxial blocks, especially subarachnoid anaesthesia, may be complicated by bradycardia and hypotension requiring vasoactive drugs, depending on the height and extent of sympathetic blockade. Patients with CPVT who receive large doses of beta blockers are more prone to such cardiovascular adverse effects. Considering these risks, an epidural or a combined subarachnoid/epidural technique which allow dose titration, controlled and gradual establishment of the block are probably more advantageous over the single shot spinal [13].

Particular or additional monitoring

For safety reasons, after placement of the patient on the operating table, standard monitoring should be attached, including electrocardiogram, non-invasive blood pressure measurement and pulse oximeter. Any intervention, such as placement of a peripheral venous catheter, should be performed under monitoring. Prior use of a local anaesthetic cream for minimization of puncture pain and possible sympathetic response can be helpful. An arterial line should be better placed before induction of anaesthesia, but preferably after adequate IV premedication (i.e midazolam). Intraoperatively, the depth of anaesthesia can probably be assessed more reliably by the use of specific monitors, such as Bispectral Index or Entropy. Additionally, given the necessity of providing adequate analgesia to prevent excessive sympathetic stimulation, monitors of nociceptive/anti-nociceptive balance (e.g. pupillometry) may also be useful.

Regarding particular/additional monitoring, a 5-lead electrocardiogram and invasive arterial blood pressure measurement should be used for reliable, accurate and prompt identification and treatment of arrhythmias with haemodynamic compromise. Also, the pads of an external defibrillator should be attached to the patient, while any existing ICDs should be inactivated in order to avoid any interference with diathermy or electromagnetic devices [5,9].

Possible complications

Adrenergic-induced tachyarrhythmias, in the form of bidirectional or polymorphic VT with haemodynamic compromise, syncope or VF and cardiac arrest are disease-related complications that may occur perioperatively. Tachycardia exceeding 120-130 bpm should be treated promptly with a beta-blocker such as esmolol or labetalol IV [2]. Among other pharmacological agents, magnesium IV may also be helpful (drug of choice in Torsades de Pointes), while prompt electrical cardioversion should be applied in case of VT with a pulse, or defibrillation in case of pulseless VT/VF according to the Advanced Life Support Algorithm [2,20]. If significant hypotension without arrhythmia occurs, a vasopressor with pure alpha-agonistic action, such as phenylephrine, should be administered [5]. Any factor causing or facilitating haemodynamic compromise, such as electrolyte, metabolic abnormalities, or drugs, should be corrected or discontinued immediately.

Postoperative care

Documentation and stabilisation:

Psychiatric and behavioural problems such as hyperactivity, depression, self-injurious behaviour and bipolar disorder.

Ventriculomegaly and hydrocephalus usually due to aqueductal stenosis are common. Different degrees of trigeminal anaesthesia leading to recurrent corneal and facial scarring.

Parietal or parieto-occipital alopecia.

Postoperative care:

After patient's smooth recovery from anaesthesia, the ICDs should be reactivated and reset at their preoperative mode. It is suggested that haemodynamic monitoring and close clinical observation should be continued for at least 24 hours in a High Dependency Unit [5,13].

Sufficient postoperative analgesia is mandatory; wound infiltration at the end of the surgical procedure can be helpful [2], while opioids have also been administered uneventfully. Specifically, tramadol has been given for moderate pain in CPVT and provided adequate analgesia without adverse effects [2]. Pethidine has also been used without complications [12], but its atropine-like effect should be kept in mind. There are no reports on other analgesics, but based on their pharmacodynamic profile, paracetamol, ketorolac and morphine represent relatively safe choices. Regarding morphine, it exerts a direct effect on sinoatrial and atrioventricular node (negative chronotropic and dromotropic action) and could be a useful drug for analgesia after major surgery [5].

Antiemetics for prevention or prompt treatment of postoperative nausea/vomiting should also be administered to these patients, since this condition may lead to significant stress. Ondansetron [2] and droperidol [12] have both been given to CPVT patients without complications; interestingly, droperidol has been found to exert a preventive action on catecholaminergic arrhythmias [5]. Also, dexamethazone has no cardiovascular actions and could be used for antiemesis.

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

As CPVT typically presents as syncopal episodes, differential diagnosis includes epilepsy, vagal hyperactivity, drug induced bradycardia (beta blockers, antiarrhythmics, anaesthetic drugs), electrolyte or metabolic disorders (i.e hypoglycaemia), cardiomyopathy, or arrhythmia associated with other syndromes such as long QT, Brugada, Wolff-Parkinson-White. Syncope associated with adrenergic stimulation (i.e stress, tachycardia), family history of sudden cardiac deaths, absence of characteristic ECG signs (i.e long QT) or structural cardiac abnormalities suggest the possibility of CPVT and necessitate further investigation of the disease. Since epilepsy represents a common misdiagnosis in patients with CPVT, anaesthesiologists should be suspicious and alert in cases characterised as epileptics, but without a definite diagnosis based on thorough workup [2,5].

If a sudden cardiac death occurs perioperatively, postmortem examination should be performed, and if CPVT is suspected, the family should undergo genetic screening [5].

Ambulatory anaesthesia

Even for short procedures, the patients should preferably remain in hospital for 24h under cardiac monitoring and close clinical observation [2,10]. During that time, they should be kept comfortable and calm, in a quiet environment with promptly available drugs and equipment for cardiopulmonary resuscitation. They should receive their analgesics as needed, adequate hydration and antiemetics for prevention/treatment of nausea and vomiting.

Obstetrical anaesthesia

In obstetric cases with CPVT, neuraxial techniques are advantageous because risks associated with general anaesthesia, such as difficult endotracheal intubation, aspiration and sympathetic response to airway instrumentation, are avoided. Uneventful combined spinal-epidural anaesthesia for caesarean delivery has been reported in a parturient with CPVT: a small dose of bupivacaine 0.5% (1 ml) and fentanyl 25 µg were injected intrathecally and a further dose of bupivacaine 0.5% (5 ml) was given epidurally in order to achieve the desired level of block [13]. Compared to single shot spinal, both epidural and combined low dose spinal-epidural anaesthesia offer the advantage of titration, in terms of providing a successful block while maintaining haemodynamic stability. Additionally, the epidural catheter can be used for high quality postoperative analgesia. In any type of regional technique, supplementation of local anaesthetic with epinephrine is contraindicated.

If general anaesthesia is administered, it should be kept in mind that the use of large doses of lipophilic opioids (i.e. fentanyl, sufentanil) for prevention of excessive sympathetic response to intubation may produce neonatal respiratory depression possibly requiring short-term ventilator support and/or naloxone. The risk is less with remifentanil [21], but still, neonatal depression and assisted ventilation may be needed. The neonatologist should be informed accordingly and experienced staff is required to provide neonatal care. Beta blockers, such as propranolol and labetalol, are relatively safe, should be received throughout pregnancy and also continued perioperatively [13]. If an IV beta blocker is given peripartum as supplementation, fetal monitoring is recommended because acute fetal bradycardia may occur [13].

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