Perioperative management of patients with pre-excitation syndromes

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Abstract

Patients with pre-excitation abnormalities are at a high risk for life-threatening perioperative arrhythmias. In Wolff-Parkinson-White syndrome, the anaesthetics used for invasive diagnostic testing/ablation, should not affect cardiac electrophysiology; propofol, sevoflurane, fentanyl, sufentanil, alfentanil are suitable. In non-ablative surgery, propofol, sevoflurane, isoflurane, fentanyl, alfentanil, sufentanil have been used safely. Among neuromuscular blockers, cis-atracurium, rocuronium and vecuronium are good choices. Ketamine, pancuronium and pethidine should be avoided because of their sympathomimetic actions. Anticholinergic/anticholinesterase combinations for neuromuscular block reversal should preferably be omitted, while sugammadex seems more attractive. In regional anaesthesia, addition of epinephrine and high sympathetic blocks should be avoided. Hypotension should be treated with pure alpha-adrenergic agonists. Other pre-excitation abnormalities associated with different accessory pathways are the Mahaim Fiber and Lown-Ganong-Levine syndrome. Sympathetic activation should be avoided. Total intravenous anaesthesia with propofol probably represents the safest option. A careful anaesthetic plan and close cooperation with cardiologists are mandatory for successful management.

Keywords: Pre-excitation; Wolff-Parkinson-White syndrome; Mahaim fiber syndrome; Lown-Ganong-Levine syndrome; anaesthesia

Introduction

In 1893, the physiologist S. Kent reported the existence of atrio-ventricular (AV) pathways in mammalian hearts [1], while in 1930, the cardiologists Wolff, Parkinson and White described an “unusual cardiac mechanism” manifested as paroxysmal tachycardia or atrial fibrillation (AF) and characterised by a bundle-branch block and a short PR interval on the electrocardiogram (ECG) [2]. Since then, this electrophysiological abnormality has attracted the interest of investigators [3, 4], while the term “pre-excitation” was first used by Öhnell in 1944 [5].

Normally, the cardiac electrical impulses are generated in the sino-atrial (SA) node and then are spread to the right and left atrium. After passing through the AV node and His bundle, the stimuli reach the Purkinje fibers and propagate across the ventricular myocardium. The action potential of the AV node depends on slow inward calcium current which delays AV conduction as the heart rate (HR) increases, thus preventing fast supraventricular rhythms to reach the ventricles. However, abnormal AV muscular connections may serve as accessory pathways (APs) allowing atrial impulses to bypass the critical delay in the AV node. In most APs, the action potential relies on rapid inward
sodium currents. As a result, the stimuli travel towards the ventricles faster than usual, causing early depolarisation of the ventricular myocardium, a condition named “pre-excitation” [5]. The quick transmission of cardiac impulses combined with fast HRS may predispose to detrimental ventricular tachycardias (VTs), even ventricular fibrillation (VF) [6-8].

More than one APs may exist in the same patient, located almost anywhere in the AV groove [5]. The AP conduction is usually bidirectional, but rarely it may be only retrograde (ventriculo-atrial). In this case, the pathways are called “concealed” due to the absence of electrocardiographic signs of pre-excitation [3, 5].

In the past, the term “pre-excitation” was exclusively used for the Wolff-Parkinson-White (WPW) syndrome. Today, other pre-excitation mechanisms due to different APs have also been identified. Specifically, Mahaim Fiber and Lown-Ganong-Levine syndrome represent pre-excitation conditions which are encountered more rarely than the well known WPW.

Perioperative management of patients with pre-excitation may become quite challenging, especially if there is no time for adequate preoperative investigation and clinical optimisation – as in emergencies –, or even worse, in undiagnosed cases. It is possible that the poorly controlled or unknown underlying electrophysiological abnormality will become unmasked during anaesthesia and surgery, giving rise to potentially life-threatening arrhythmias.

In the literature, pre-excitation syndromes have been mainly approached from the view of the disease (i.e. presentation, diagnosis, treatment), while anaesthetic data are scarce. The present review aims to focus on the perioperative management of patients with WPW and two other, less known pre-excitation syndromes.

We conducted a Pubmed literature search for all types of published articles (up to February 2018) using the terms: “Preexcitation”, “Pre-excitation”, “Wolff-Parkinson-White syndrome”, “WPW syndrome” “Mahaim pre-excitation”, “Mahaim fibers”, “Lown-Ganong-Levine syndrome” and “anaesthesia” or “perioperative management” or “perioperative care” in all possible combinations. Articles in languages other than English were used, if they had a detailed English abstract containing specific and relevant information. We identified 75 suitable articles. Additionally, seven publications were found by manual searching. Since the number of randomised controlled trials (RCTs) was small, the articles we considered were mostly observational studies, retrospective studies, case series and case reports. We also used articles providing information on genetics, clinical presentation, diagnostic and therapeutic approach of the disorders. In total 119 articles were included in the present review.

The WPW syndrome

Pathophysiology and clinical features of WPW syndrome

The main pathophysiological characteristic of the syndrome is the existence of an accessory AV pathway, named “bundle of Kent”. It represents an abnormal fibro-muscular connection which ends up directly into the ventricular myocardium and has remained after incomplete AV separation [9, 10]. It may connect the left atrium and ventricle (type A), or the right atrium and ventricle (type B), and can conduct the stimuli bidirectionally [5, 10]. The prevalence of WPW syndrome is 0.1-3.1% in general population, higher in men [10]. Most cases are sporadic, while the more rare familial form is characterised by autosomal dominant inheritance and has been linked to chromosome 7q34-q36 and mutations in the gamma-2 regulatory subunit of AMP-activated protein kinase gene (PRKAG2) [11].

The ECG pattern consists of a short PQ interval (< 120 ms), a slurred upstroke at the beginning of QRS (delta wave) and a broad QRS complex (> 120 ms). This characteristic “WPW pattern” represents an exclusively ECG feature, found 10-100 times more frequently than the actual “WPW syndrome” which refers to the combination of the ECG pattern with symptomatology [9, 12]. While many individuals with a WPW-pattern will remain asymptomatic for life, patients with WPW-syndrome usually develop arrhythmias between the ages 20-40 years [10]. The commonest symptom is palpitations, while dizziness, lightheadedness, chest pain, shortness of breath and syncopal episodes may also occur. Rarely, the first manifestation of the disorder is cardiac arrest [10].

The most common arrhythmias are atioventricular reentrant tachycardias (AVRTs) [10], usually orthodromic. In orthodromic tachycardias, the stimuli travel down the AV node and return back via the AP, producing narrow QRS complexes on the ECG [9]. Less often, the conduction proceeds down the AP and retrogradely up the AV node, causing antidromic tachycardias with a wide QRS [9]. Finally, AF or atrial flutter may develop; both rhythms can be dangerous if rapid anterograde conduction occurs through an AP with a short refractory period. In this case, many atrial impulses may travel towards ventricles, causing extremely fast ventricular responses, VF and sudden cardiac death [12]. Risk factors for life-threatening arrhythmias are APs with short anterograde effective refractory period, short RR interval in pre-excited AF, inducible AVRT during electrophysiology studies (EPS), multiple APs, male gender and young age [13, 14]. Sometimes, pre-excitation may imitate acute myocard-
dial infarction, while negative delta waves can also be confused with pathological Q waves [15, 16]. Further investigation for definite diagnosis includes cardiac troponin I levels or echocardiographic assessment of myocardial wall movement [15, 16].

The initial diagnosis of WPW syndrome is based on the ECG pattern combined with symptomatology. More specific examination includes Holter monitoring and exercise or pharmacological testing with ajmaline or procainamide [12]. Finally, EPS may be required to elucidate the abnormality and identify the exact AP location and its characteristics.

Treatment, conservative or invasive, is case-specific, since both long-term antiarrhythmic therapy and AP catheter ablation are not without risks, namely drug-side effects, or heart injury, ischaemia and thromboembolism due to catheterisation [10]. The decision depends on the risk/benefit ratio, according to the type, severity and frequency of symptoms [10].

**Anaesthetic considerations in WPW syndrome**

Patients with WPW syndrome may require anaesthesia for EPS/ablation or for non-ablative procedures. In diagnostic/therapeutic interventions, anaesthetics should not interfere with cardiac electrophysiology, while in non-ablative surgery, drugs should ideally prevent – or at least not facilitate – the generation of arrhythmias associated with APs.

Anaesthesia and surgery may unmask an undiagnosed syndrome, and suspicious cases should be referred for further investigation before elective surgery. A detailed cardiac history during pre-anaesthetic evaluation is the cornerstone of not missing undiagnosed patients [12]. Apart from a 12-lead ECG, Holter monitoring may be useful, especially in intermittent WPW. Echocardiography can reveal congenital cardiac abnormalities that coexist in 7-20% of patients, such as Ebstein’s anomaly, valve lesions, cardiac hypertrophy, atrial aneurysms and septal defects [10, 11, 17]. An EPS may also be needed; the benefit of postponing an elective surgery for EPS testing should be examined. The capability of rapid anterograde AP conduction increases the risk of sudden death and is an indication for radiofrequency catheter ablation (RFCA) to precede surgery [17]. Diagnosed patients should be carefully assessed preoperatively regarding their symptoms and current treatment, and should be clinically optimised. Cardiological consultation and close cooperation are mandatory for a safe management plan.

**Anaesthesia for electrophysiological investigation and ablation**

Although cardiac complications during EPS are usually not related to the anaesthetic technique [18, 19], accurate diagnostic mapping and successful ablation may be impaired by anaesthetic drugs that alter the conduction in the normal and accessory pathways [20]. Thus, agents with minimal effects on cardiac electrophysiology should be preferred [19].

Among general anaesthetics, propofol is considered appropriate for use in EPS/ablations, as it does not impede the generation of diagnostic supraventricular tachycardias (SVTs) [20]. Despite reports about bradycardia, AV block, even asystole [21], Sharpe and colleagues suggest that propofol per se has no direct effects on SA or AV node or intra-atrial conduction [22]. Moreover, propofol did not affect the refractory periods of normal and accessory pathways and did not interfere with EPS/ablation in a study with adult WPW patients [22]. Also, in children, propofol-based anaesthesia did not cause problems during EPS/RFCA [23, 24]. Two randomised studies in youngsters undergoing RFCA, showed that propofol was similarly suitable with isoflurane, both leaving unaffected the SA and AV node function [25, 26]. Nevertheless, the findings about isoflurane are not consistent; experimental research has shown that it may depress the SA node discharge, and prolong the AV and ventricular conduction [21]. Similarly, in adults undergoing surgical cryoablation, isoflurane at 1 MAC was associated with slower conduction in normal and aberrant systems [27]. Also, a retrospective study of children showed that isoflurane prolongs the atrial, ventricular and AP antegrade effective refractory period [28]. Notably, not only isoflurane [29], but also enfurane [29, 30], and halothane [29] are considered rather unsuitable agents for EPS/ablations, since they prolong the refractory period and slow the conduction in normal and accessory pathways, thus interfering with interpretation of tests and determination of ablation success. In patients receiving halothane anaesthesia, persistent delta waves and difficulty with AP localisation have also been reported [31]. Conversely, experimental studies have shown that sevoflurane exerts only moderate effects on cardiac electrophysiology [21]. Additionally, clinical data suggest that sevoflurane has a favorable profile for use in EPS/ablations [32, 33]. In adults, it did not affect the function of SA node or AV and AP conduction, and did not prevent diagnostic reciprocating tachycardias [32]. Also in children, when given after propofol, sevoflurane caused only moderate prolongation of the AP antegrade effective refractory period, without interfering with the ablative procedure [33]. On the other hand, desflurane at 1 MAC prolonged the AP effective refractory period and impaired the induction of diagnostic SVTs in paediatric patients [24]. The authors considered it unsuitable for EPS/ablative procedures [24].

Regarding opioids, animal studies have shown that morphine may exert a direct negative action on SA
and AV nodes, while fentanyl may decrease the SR, but this action is not clinically significant [21]. In WPW patients, fentanyl was found to exert no effect on AP refractory period or SA conduction [34, 35], but it may increase the cardiac vagal tone causing prolongation of the sinus node recovery time, especially when combined with propofol [35]. More pronounced effects have been reported for remifentanil: both experimental and clinical studies have shown that it may depress sinus node automaticity and delay SA and AV conduction [21, 36-38]. These properties render remifentanil less attractive for use in EPS, as it may interfere with testing and results. On the contrary, sufentanil is devoid of significant direct actions on the normal intracardiac or AP conduction [21, 29]. Also, an alfentanil-/midazolam-based anaesthesia has been suggested as suitable, because it leaves unaffected both the AV node and aberrant bundle [39].

Among adjuvants, dexmedetomidine has been studied in EPS/ablative procedures. A prospective trial in children receiving thiopental/ketamine anaesthesia showed that i.v. dexmedetomidine (1 μg/kg followed by 0.7 μg/kg/h infusion), given after successful ablation caused significant sinus and AV node depression, without affecting the atrial or ventricular refractoriness [40]. The authors consider that the drug may interfere with the generation of tachycardias and interpretation of measurements [40]. Different findings were reported by a retrospective study of children receiving propofol or sevoflurane: dexmedetomidine (0.5-1 μg/kg followed by 0.5-1 μg/kg/h infusion) given during EPS/ablation was associated with increased need for isoproterenol for SVT induction, but did not affect the electrophysiological parameters or the interventions and their success [41]. Regarding other factors, controlled ventilation in patients subjected to RFCA under GA was found to facilitate the procedure and increase the ablation success [42].

The published clinical trials on the use of anaesthetics for EPS/ablation are presented in Table 1.

**Anaesthesia for non-ablative surgical procedures**

In patients with WPW syndrome undergoing non-ablative surgery, antiarrhythmic treatment should be continued perioperatively. Additionally, sympathetic stimulation should be avoided, because it may shorten the AP refractory period and facilitate life-threatening arrhythmias [43-47]. Anaesthetics and adjuvants that increase AP refractoriness should be preferred.

Regarding volatiles, early research demonstrated that enflurane was advantageous over both halothane and isoflurane, because it increased the AP refractoriness more than the other two agents, while it was the only volatile that did not prolong the coupling interval, thus the vulnerable time for SVT generation [29]. Also, in two older case reports enflurane was described as a safe agent for WPW patients [48, 49]. Conversely, Dobkowski and colleagues suggested that enflurane may trigger arrhythmias and should not be used in WPW syndrome [30]. In a more recent report, isoflurane was preferred among other volatiles due to its property to prolong the AP refractory period and decrease the likelihood of tachycardias [50]. Similarly, most investigators consider isoflurane as a safe choice for patients with WPW [45, 51-53]. Although halothane could be preferred for its bronchodilatory properties in selected cases [52], its potential to sensitize myocardium to catecholamines is a major drawback. Sevoflurane seems less advantageous than isoflurane, since it does not prolong the AP refractory period [32]. Nevertheless, it has been used safely for the maintenance of anaesthesia in a number of cases [54-57]. Also, no adverse effects have been reported for nitrous oxide [34, 48-49, 54, 58]. Finally, desflurane has been found to prolong the refractory period of AP, without affecting the electrophysiology of sinoatrial node and normal pathways [24], but the available data are limited [59].

Among intravenous anaesthetics, propofol does not affect the AP refractory period [22]. Induction or maintenance of anaesthesia with propofol combined with fentanyl has been associated with cardiovascular stability in WPW patients [44, 50, 54, 55, 58-63]. Moreover, it was reported that delta waves disappeared after propofol administration and reappeared after its discontinuation [58]. Similarly, a decrease of QRS duration and return of PR interval into normal ranges were observed when propofol infusion (25 μg/kg/min) was added to isoflurane anaesthesia [63]. Thiopental has been used in several cases, and small (50 mg) to moderate doses (4 mg/kg) did not cause conduction problems [18, 45, 48, 51, 53, 64, 65]. Nevertheless, in the early 70’s its safety had been questioned due to observed ECG changes after anaesthesia induction; the ECG signs of acute myocardial infarction were attributed to enhanced conduction in the AP caused by thiopental [66-68]. Experimental data suggest that etomidate in high plasma concentrations may decrease the SR and prolong AV conduction [21]. Nevertheless, in common clinical doses it offers significant cardiovascular stability, quite desirable in patients with WPW, although specific data are rather limited [49]. Ketamine should better be avoided due to its sympathomimetic effects.

Most opioids are suitable for patients at risk for tachyarrhythmias, since they maintain haemodynamic stability with a mild reduction of HR [21]. Additionally, alfentanil/midazolam anaesthesia produces no significant effects on AP conduction and the refractory period [39], while the sufentanil/lorazepam combination may even cause a mild prolongation of the AP effective
**Table 1. Clinical studies on the anaesthetic management of patients with Wolff Parkinson White syndrome undergoing electrophysiological studies and/or ablation of accessory pathways**

<table>
<thead>
<tr>
<th>Author / study</th>
<th>Patients</th>
<th>Type of anaesthesia / Drugs used</th>
<th>Findings / Suggestions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Irish (1988)²⁸, retrospective</td>
<td>Children, adolescents, adults with AV APs n = 181 (197 procedures)</td>
<td>GA with various drugs: diazepam, fentanyl, scopolamine, droperidol, thiopental, halothane, enflurane, isoflurane, N₂O, pancuronium, succinylcholine, tubocurarine</td>
<td>Incidence of arrhythmias not related to anaesthetic agent; standard anaesthetic methods and drugs can be used in most cases</td>
</tr>
<tr>
<td>Joeng (2006)²⁹, retrospective</td>
<td>Children, adolescents n = 131 (47 WPW)</td>
<td>Various anaesthetic techniques: No sedation, conscious sedation, deep sedation, GA (midazolam, fentanyl, ketamine, propofol)</td>
<td>Complication incidence did not differ and was not related to anaesthetic method</td>
</tr>
<tr>
<td>Moore (2011)³⁰, retrospective</td>
<td>Children, adolescents n = 151</td>
<td>GA: propofol infusion</td>
<td>GA may interfere with risk stratification / Propofol is a good choice</td>
</tr>
<tr>
<td>Sharpe (1995)³¹, prospective</td>
<td>Adults, n = 12</td>
<td>GA: alfentanil, midazolam, vecuronium / ± propofol for maintenance</td>
<td>Propofol: no effect on AV and AP conduction / suitable for EPS / ablation</td>
</tr>
<tr>
<td>Pappone (2004)³², RCT</td>
<td>Children, n = 47 (20 received intervention)</td>
<td>GA: propofol</td>
<td>Propofol: no significant complications</td>
</tr>
<tr>
<td>Hino (2017)³³, randomized crossover study</td>
<td>Children, n = 36</td>
<td>GA with propofol or desflurane at 0.5 or 1 MAC</td>
<td>Desflurane 1 MAC: ↑ AP refractory period, no effect on normal pathways, impaired SVT induction. Desflurane: unsuitable for EPS / ablation. Propofol: suitable for EPS / ablation</td>
</tr>
<tr>
<td>Lavoie (1995)²⁵, RCT</td>
<td>Children, adolescents n = 20 (9 WPW)</td>
<td>GA: alfentanil, thiopentol, pancuronium, N₂O, propofol or isoflurane</td>
<td>Neither propofol nor isoflurane had significant effects on SA or AV node</td>
</tr>
<tr>
<td>Erb (2002)³⁴, RCT</td>
<td>Children, adolescents n = 60</td>
<td>GA: midazolam, sevoflurane, pancuronium, fentanyl, N₂O, isoflurane or propofol</td>
<td>SVT induction independent of propofol or isoflurane administration. Isoflurane and propofol equally suitable</td>
</tr>
<tr>
<td>Dobkowski (1990)³⁵, prospective*</td>
<td>Adults, n = 7</td>
<td>GA: lorazepam, morphine, sufentanil, vecuronium, isoflurane</td>
<td>Isoflurane 1 MAC: prolonged the conduction in both normal pathways and AP</td>
</tr>
<tr>
<td>Chang (1996)³⁶, retrospective</td>
<td>Infants, children, adolescents n = 51</td>
<td>Sedation (pethidine, promethazine, chlorpromazine) versus GA with isoflurane</td>
<td>Compared to sedatives, isoflurane prolonged the refractory periods in AP (antegrade), atria and ventricles. Isoflurane: interfered with EPS</td>
</tr>
<tr>
<td>Sharpe (1994)³², RCT</td>
<td>Adults, n = 21</td>
<td>GA: lorazepam, sufentanil, vecuronium, volatile (halothane or isoflurane or enflurane)</td>
<td>Lorazepam / sufentanil: no effect on AP. Volatiles: ↑ in AV and AP refractoriness (mostly enflurane)</td>
</tr>
<tr>
<td>Dobkowski (1991)³⁷, prospective*</td>
<td>Adults, n = 7</td>
<td>GA: lorazepam, morphine, sufentanil, vecuronium, enflurane</td>
<td>Enflurane: ↓ AV and AP conduction / should not be used in WPW patients</td>
</tr>
<tr>
<td>Tempe (1997)³³, retrospective</td>
<td>Adults, n = 13</td>
<td>GA: morphine, halothane, pancuronium</td>
<td>Halothane: interferes with EPS / ablation</td>
</tr>
<tr>
<td>Sharpe (1999)³³², prospective</td>
<td>Adults, n = 15</td>
<td>GA: midazolam, alfentanil, vecuronium, sevoflurane</td>
<td>Sevoflurane: no effect on AV or AP conduction / suitable for ablations</td>
</tr>
<tr>
<td>Pérez (2006)³³³, prospective</td>
<td>Children, n = 15</td>
<td>GA: fentanyl, vecuronium, propofol replaced by sevoflurane</td>
<td>Sevoflurane: moderate changes in AP properties / suitable for ablations</td>
</tr>
<tr>
<td>Fujii (2011)³³⁶, Children, n = 60 (29 completed study)</td>
<td>GA: propofol, remifentanil, vecuronium</td>
<td>Remifentanil: ↓ SN automaticity, ↓ SA conduction</td>
<td></td>
</tr>
<tr>
<td>Sharpe (1992)³³⁸, prospective</td>
<td>Adults, n = 8</td>
<td>GA: lorazepam, alfentanil, midazolam, vecuronium</td>
<td>Alfentanil / midazolam anaesthesia: no effect on AV node or AP conduction</td>
</tr>
</tbody>
</table>

**Notes:**
- * study published in the form of abstract
can be safely used in conjunction with fentanyl, alfentanil and represent more attractive choices [32, 39]. They found to exert no significant effects on AP conduction, short acting midazolam (half life: 2 h) have also been intermediate acting lorazepam (half life: 14 h) or the disadvantage, especially for outpatient cases. The nevertheless, its long duration of action (half life: 43 h) is without effects on the AP refractory period [34]. Ne-benzodiazepines, diazepam is cardiovascularly stable, anaesthesia for minor surgical procedures [46]. Among can be administered alone or as supplement to local cholinesterase, nicotinic and muscarinic receptors [73]. reversing; it seems to exert no significant actions on period in the AP [72]. Thus, it seems that the standard the anterograde and retrograde effective refractory period may become prolonged and AP conduction facilitated due to unopposed parasympathetic tone [75, 76]. Cases of WPW syndrome unmasked due to high levels of spinal blocks are not rare, especially if combined with other vagal stimulants [17, 76, 77]. Thoracic epidural and single shot spinal anaesthesia are more likely to cause cardiac sympathectomy, thus careful dosing and increased vigilance for a high blockade are required. The spinal dose of local anaesthesia should ideally produce the minimum level of sensory block required for the surgery. Addition of opioids allows a reduction in local anesthetic doses [78, 79], while selective spinal opioid analgesia may be used in some cases (i.e. labor), thus minimising haemo-dynamic fluctuations [78, 80].

Dose titration and gradual elevation of the block can be achieved with lumbar epidural and combined low-dose-spinal/epidural anaesthesia. These modalities offer more haemodynamic stability and should probably be preferred over single-shot spinal anaesthesia [81]. There are several reports of operations (caesarean deliveries included) performed safely under a simple epidural [64, 82-84] or a combined spinal/epidural anaesthesia [81, 85-88]. In selected cases, combined general/regional anaesthesia may be used; successful thoracotomy under propofol anaesthesia together with thoracic epidural analgesia has been reported [61]. Reduced venous return and atrial filling due to an extensive sympathetic blockade may precipitate arrhythmias. Adequate fluid loading should be con-
sidered in order to reduce – as possible – the risk and magnitude of hypotension, subsequent sympathetic activation or need for sympathomimetic drugs [88, 89]. If hypotension occurs, a pure alpha-adrenergic agonist (i.e. phenylephrine) is the vasopressor of choice [88, 90, 91]. The addition of epinephrine to local anaesthetics should be avoided, as it may shorten the AP refractory period and facilitate arrhythmias [43]. Finally, supplementation of any regional technique with adequate sedation in order to reduce anxiety and sympathetic stimulation is advisable [16].

In pregnant patients with WPW, physiological adaptation and emotional factors increase the risk of arrhythmias; estrogens, intravascular volume expansion, haemodynamic changes, pain, stress and oxytocin given during labor may all trigger SVTs, especially when an extended subarachnoid sympathetic block is established [87, 92, 93]. In this regard, a simple epidural or a combined spinal/epidural are the modalities of choice for caesarean delivery. Vaginal delivery has also been accomplished safely in WPW parturients, even in cases with severe co-morbidities, under a carefully managed labor epidural analgesia with local anaesthetic/opioid combination [83, 84]. Regarding oxytocin, it should be omitted if possible [71, 88], since it has been associated with paroxysmal SVT, even in common clinical doses [90].

Management of perioperative arrhythmias

Orthodromic AVRTs are regular narrow-complex tachycardias (constant R-R intervals, QRS < 120 ms) that should be treated as any other paroxysmal SVT with vagal maneuvers, such as carotid sinus massage, Val-salva or Valsalva-like maneuver in case of mechanical ventilation, along with 100% oxygen [94]. If these measures fail, adenosine IV boluses should be given, starting with 6 mg, followed by 12 mg and further 12 mg, if tachycardia persists. Adenosine suppresses both sinus automaticity and AV conductivity, and usually converts an SVT to normal SR [90, 95]. It should be given as a rapid push, because of its very fast elimination. Its ultra-short duration of action also renders adenosine suitable for parturients [87, 92], even though it may cause temporary bradycardia to the fetus; thus, fetal HR monitoring is suggested during administration [90].

Beta blockers or more rarely calcium channel blockers have been used to treat WPW-related SVTs [44, 51, 91-94]. Beta blockers can also be given preventively before laryngoscopy [96]. The very short acting esmolol is preferred over long acting agents [44], especially in parturients, even though the risk of causing fetal distress still exists [92].

Both ephedrine and phenylephrine have been used to treat hypotension [18, 81, 86, 90]. Moreover, phenylephrine was reported to resolve a paroxysmal SVT resistance to various other measures and drugs [91].

Antidromic AVRT presents as regular wide-complex tachycardia (QRS > 120 ms), sometimes difficult to differentiate from VT [9]. Amiodarone or propacainamide can be used to restore the rhythm in haemodynamically stable patients. If there is doubt about the type of arrhythmia, it should be treated as VT [97].

Pre-excited AF presents as irregular wide complex tachycardia, often resembling VF or Torsades de Pointes [97]. Amiodarone (300 mg i.v. in 20 min) can be given, while digoxin and verapamil are contraindicated, and also adenosine and diltiazem should be avoided, since they block the AV node while concomitantly facilitating AP conduction, thus increasing the risk of VF [90, 97]. Propacainamide and propranolol can be useful because they prolong the AP refractory period [57]. Expert cardiological consultation is required in such high risk situations.

Electrical synchronised cardioversion is indicated for SVT or AF resistant to antiarrhythmic drugs or in cases with extreme tachycardia and/or hypotension/shock, myocardial ischaemia, heart failure/pulmonary oedema or syncope [17, 59, 81, 97, 98]. Immediate defibrillation should be performed in VF or pulseless VT [97]. Antiarrhythmic drugs and a defibrillator should be prepared before anaesthesia induction [57, 81].

Sympathetic stimulation should be avoided: alleviation of anxiety, adequate intraoperative anaesthesia/analgesia, suppression of response to intubation, avoidance of anticholinergic/sympathomimetic drugs are mandatory [96]. The patient should be kept warm, normovolaemic, normocarbic and balanced regarding the acid base and electrolyte status [51]. Smooth recovery from anaesthesia, sufficient postoperative analgesia, prevention of nausea/vomiting that may cause stress and tachycardia are also important.

A concealed WPW syndrome should be considered in the differential diagnosis of paroxysmal SVTs occurring during anaesthesia. If the syndrome is suspected, cardiological consultation and postoperative ECG Holter should be requested [99]. Appearance of the electrocardiographic WPW pattern after induction of anaesthesia should not necessarily lead to surgery cancellation, especially if the patient has no history of suspicious symptoms and is haemodynamically stable [100, 101]. Nevertheless, in such cases the anaesthesiologist should be fully prepared to treat any arrhythmias that may arise [99]. It is also important to note that asymptomatic patients with intermittent WPW do not receive antiarrhythmic treatment and generally carry a low risk for tachyarrhythmias [47, 100, 102, 103]. Adequate preparation, invasive blood pressure monitoring, and a high level of vigilance are of paramount importance.
The identified publications on the anaesthetic management of patients with WPW-syndrome undergoing non-ablative surgical procedures are presented in Table 2.

Other Pre-Excitation Syndromes with different Accessory Pathways

Mahaim Fibers

In 1938, I. Mahaim described anatomic connections between the AV node and ventricles [104]. These “Mahaim Fibers” were initially considered to connect the AV node or the His bundle/fascicles with the right ventricle (nodo- or fasciculo-ventricular fibers) [104], but later, it was demonstrated that they usually originated in the right atrium and terminated in the right ventricular wall (atrio-ventricular) or near the right bundle branch (atrio-fascicular) [105]. Mahaim Fibers (MFs) are found in 0.5-1:10 000 of the general population, comprising about the 3% of APs [106, 107], and may coexist with other cardiovascular pathology, such as Ebstein’s anomaly [107].

The MF pathways exhibit longer conduction times compared to the bundle of Kent, while they share some of the AV properties, such as similar conduction velocity, conduction delay at high atrial rates and sensitivity to adenosine [107]. They allow only antegrade conduction and may be part of a circuit with retrograde conduction through the AV node; these antidiatomic AVRTs are characterized by a wide QRS complex and may be difficult to differentiate from ventricular arrhythmias [107, 108]. Tachycardia with a left bundle branch block pattern may be seen as the result of right ventricle pre-excitation (stimulated before LV) via MFs [106]. The ECG may have features of pre-excitation, but usually it has a normal PQ interval and minimal or no delta wave [106, 107]. An rS pattern in lead III is found in about 60% of the patients, while an additional suspicious finding is the absence of q wave in lead I [106]. The combination of such an ECG with young age and history of tachyarrhythmias strongly suggests the existence of MFs, but certain diagnosis requires an EPS.

Beta-blockers, class IA and IC antiarrhythmics are used to prevent tachycardias [107], but definite treatment with AP ablation is preferred in many cases [107, 108]. Anaesthetic data on the management of patients with MFs are limited. Conditions and drugs causing sympathetic activation and catecholamine release should be avoided; Zweifler and colleagues suggested that perioperative stress and pain were possibly the triggering factors of a wide complex tachycardia (MF pathway conduction with AV nodal reentry) in a woman with undiagnosed MFs (Table 3) [108].

Lown- Ganong-Levine syndrome

The Lown-Ganong-Levine (LGL) syndrome is another rare pre-excitation condition first described in 1952 [109]. Its ECG pattern is characterised by a short PQ interval (< 120 ms), a normal or inverted P wave, and a QRS complex of normal morphology and duration [109, 110]. The combination of the above features with paroxysmal SVTs is suggestive of LGL syndrome, which comprises the 17% of cases with short PQ interval [109]. Compared to WPW, it has shorter PJ and PQ intervals which remain constant over the years, while the QRS complex usually has a duration of 80 ms or less, without the characteristic slurring (i.e. no delta wave) [109].

The short PQ interval reflects a fast conduction of the action potential from atria to ventricles, without the normal delay in the AV node. The involved mechanisms are not quite clear: enhanced AV conduction may be caused by a congenitally hypoplastic and malfunctioning AV node, which allows atrial impulses to be rapidly conducted to the ventricles [111]. Another possible mechanism is the presence of James or Brechenmacher fibers which bypass – partially or completely – a normal AV node: James fibers connect the upper part of the AV node with its lower part or with the His bundle [112], while the Brechenmacher fibers form a route between the atrium and bundle of His, thus completely bypassing the AV node [113]. In both cases, the avoidance of normal AV delay results in short PQ intervals, while the normal stimulation of ventricles through the His-Purkinje fibers gives normal QRS complexes. The normal and the bypass tract form a circuit for reentry arrhythmias. Atrial flutter, AF and VT may develop, while the deterioration of a paroxysmal AF into VF can cause sudden cardiac death [109, 114, 115].

The syndrome is mainly diagnosed in women who develop tachyarrhythmias usually in their fourth decade of life [109, 110, 116]. Sympathetic stimulation, changes in cardiac automaticity/conduction, and pregnancy-associated physiologic adaptations may trigger or facilitate SVTs [109, 117].

As in WPW, perioperative management of patients with LGL should first aim to minimise possible triggering factors of arrhythmias. Benzodiazepines, such as midazolam or diazepam [110, 116], could relieve pre-operative anxiety, while drugs with sympathomimetic properties, such as ketamine or pancuronium, should be avoided [65]. Patients should continue their antiarrhythmic medication perioperatively, while beta blockers – especially short acting ones, such as esmolol – could be useful [116]. Episodes of SVT should be managed with vagal maneuvers and adenosine [110].

As shown in Table 3, anaesthetic data are rather limited; thiopental and propofol have been used without
<table>
<thead>
<tr>
<th>First author (year)</th>
<th>Syndrome</th>
<th>No of pts/ age</th>
<th>Surgery/ procedure</th>
<th>Anaesthesia</th>
<th>Complications &amp; Management (*)</th>
<th>Outcome / Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lustik (1999)</td>
<td>WPW†</td>
<td>1F/34y</td>
<td>Uterus dilation / evacuation (17w)</td>
<td>RA SA (no further details)</td>
<td>Reported palpitations / chest pain</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Nago (2009)</td>
<td>WPW</td>
<td>1F/17y</td>
<td>Thyroidectomy</td>
<td>GA IN: Midazolam, fentanyl, propofol, pancuronium MNT: halothane</td>
<td>SVT after pancuronium / tracheal intubation *Adenosine, esmolol</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Richmond (1988)</td>
<td>WPW</td>
<td>1M/36w‡, 40w‡</td>
<td>1) IPPV for pneumonia 2) Pyelotomy</td>
<td>1) Pancuronium for IPPV 2) GA with thioental, vecuronium, isoflurane, N₂O *Wound infiltration: bupivacaine</td>
<td>1) Prolonged SVT episode (289 bpm) *Sync cardioversion 2) None</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Schmitz (1997)</td>
<td>WPW</td>
<td>1F/26y</td>
<td>Teeth surgical removal</td>
<td>•Premed: midazolam / Sedation: fentanyl, midazolam, N₂O *Local anaesthesia (bupivacaine)</td>
<td>None / Sedation associated with ECG normalization</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Wakita (2007)</td>
<td>Intermittent WPW†</td>
<td>1F/49y</td>
<td>Tooth extraction</td>
<td>IV sedation: propofol Local anaesthesia: lidocaine (± epinephrine)</td>
<td>Repeated appearance of δ-waves</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Okada (1990)</td>
<td>WPW</td>
<td>1M/29y</td>
<td>Maxillary cyst operation</td>
<td>GA (premed: atropine, hydroxyzine, pathetilorfan) IN: thioental, succiny1choline MNT: enflurane, N₂O</td>
<td>Postop retrosternal discomfort + ECG changes: Glycopyrrolate suspected</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Janes (1989)</td>
<td>WPW</td>
<td>1F/35y</td>
<td>Laparoscopic sterilisation</td>
<td>GA (premed: temazepam) IN: etomidate, fentanyl, atracurium MNT: enflurane, N₂O RV: glycopyrrolate/neostigmine</td>
<td>*Vigilance for level of neuraxial block</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Sinha (2010)</td>
<td>WPW/ Ebstein’s anomaly/ MVS</td>
<td>1F/23y</td>
<td>Danielson’s repair &amp; MVR</td>
<td>GA (premed: dazepam, morphine) IN: propofol, fentanyl, midazolam, vecuronium MNT: isoflurane</td>
<td>*SVTs *Adenosine, amiodarone, IV fluids Pt discharged with persistent pre-excitation on ECG</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Goldhill (1988)</td>
<td>WPW</td>
<td>1M/46w‡</td>
<td>3 surgeries for VPS and hydroceles / 1 CT</td>
<td>GA (± premed with atropine) Thioental, atracurium, isoflurane, N₂O</td>
<td>*Vagal stimulation, propranolol or verapamil</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Laloyaux (1998)</td>
<td>WPW/ Cantrell’s pentalogy</td>
<td>1M/42w‡ &amp; 48w‡</td>
<td>1) Inguinal hernia repair 2) Blalock-Taussig shunt</td>
<td>1) GA: halothane / RA: Caudal with mepivacaine, bupivacaine 2) GA: sufentanil, pancuronium, lidocaine, diazepam</td>
<td>*Vagal preop / transient tachycardia intraop *no extra drugs (already on digoxin, amiodarone)</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>Kumar (1986)</td>
<td>WPW</td>
<td>1M/30y</td>
<td>Lumbar laminectomy</td>
<td>GA (premed: papaveretum, hyoscine) IN: thioental, succiny1choline MNT: isoflurane, N₂O, fentanyl, vecuronium</td>
<td>None</td>
<td>Uneventful course</td>
</tr>
<tr>
<td>Kadoya (1999)</td>
<td>Intermittent WPW</td>
<td>1M/67y</td>
<td>Laryngeal microsurgery</td>
<td>GA IN: propofol, fentanyl, vecuronium MNT: sevoflurane, N₂O RV: Neostigmine without atropine</td>
<td>AF with narrow QRS after sevoflurane/N₂O discontinuation</td>
<td>Exubation \rightarrow ICU transfer \rightarrow recovery uncomplicated \rightarrow Avoidance of anti-cholinesterases</td>
</tr>
<tr>
<td>Author</td>
<td>Year</td>
<td>Procedure</td>
<td>Diagnosis</td>
<td>Details</td>
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<tr>
<td>Şahin</td>
<td>2015</td>
<td>WPW</td>
<td>1M/51y</td>
<td>Laparoscopic cholecystectomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nakamura</td>
<td>2009</td>
<td>WPW/ operation</td>
<td>1M/34y</td>
<td>Thoracoscopic sigmoid colectomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sengul</td>
<td>2016</td>
<td>WPW</td>
<td>1F/23y</td>
<td>Craniotomy &amp; tonsillectomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Seki</td>
<td>1999</td>
<td>WPW</td>
<td>1F/29y</td>
<td>Video-assisted thoracic surgery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sato</td>
<td>2014</td>
<td>WPW</td>
<td>1M/59y</td>
<td>Postop ECG: WPW pattern</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yamaguchi</td>
<td>1998</td>
<td>WPW</td>
<td>1M/62y</td>
<td>Hypercarbia did not cause tachyarrhythmias</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kajikawa</td>
<td>2001</td>
<td>WPW</td>
<td>1M/57y</td>
<td>Minimal invasive direct CAB</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Takayama</td>
<td>2000</td>
<td>WPW</td>
<td>1M/55y</td>
<td>Minimally invasive direct CAB</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gupta</td>
<td>2013</td>
<td>WPW</td>
<td>1F/30y</td>
<td>No complications associated with GA / EA</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Klepper</td>
<td>1981</td>
<td>WPW</td>
<td>1F/28y, pregnant</td>
<td>No complications associated with GA / EA</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sadowski</td>
<td>1979</td>
<td>WPW</td>
<td>3½y-64y</td>
<td>Bilateral adrenalectomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hannington-Kift</td>
<td>1968</td>
<td>WPW</td>
<td>1F/16y</td>
<td>Surgical pack change</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Campkin | 1969 | WPW | 1M/34y | Clinical course
| Van der Starre | 1978 | WPW | 1M/22y | Knee arthroscopy |
| Suppan | 1979 | WPW | 1F/45y | Laparoscopic ligation of the Fallopian tubes |
| Şahin | 2015 | WPW | 1M/51y | Laparoscopic cholecystectomy |
| Nakamura | 2009 | WPW/ operation | 1M/34y | Thoracoscopic sigmoid colectomy |
| Sengul | 2016 | WPW | 1F/23y | Craniotomy & tonsillectomy |
| Seki | 1999 | WPW | 1F/29y | Video-assisted thoracic surgery |
| Sato | 2014 | WPW | 1M/59y | Postop ECG: WPW pattern |
| Yamaguchi | 1998 | WPW | 1M/62y | Hypercarbia did not cause tachyarrhythmias |
| Kajikawa | 2001 | WPW | 1M/57y | Minimal invasive direct CAB |
| Takayama | 2000 | WPW | 1M/55y | Minimally invasive direct CAB |
| Gupta | 2013 | WPW | 1F/30y | No complications associated with GA / EA |
| Klepper | 1981 | WPW | 1F/28y, pregnant | No complications associated with GA / EA |
| Sadowski | 1979 | WPW | 3½y-64y | Bilateral adrenalectomy |
| Hannington-Kift | 1968 | WPW | 1F/16y | Surgical pack change |
| Campkin | 1969 | WPW | 1M/34y | Clinical course
| Van der Starre | 1978 | WPW | 1M/22y | Knee arthroscopy |
| Suppan | 1979 | WPW | 1F/45y | Laparoscopic ligation of the Fallopian tubes |

### Notes

- **WPW**: Wolff-Parkinson-White syndrome
- **GA**: General anesthesia
- **EA**: Epidural anesthesia

### Details

- **Premed**: Alprazolam, ranitidine
- **MNT**: Isoflurane, propofol
- **IN**: Fentanyl, propofol
- **RV**: Atropine/neostigmine
- **MNT**: Propofol, fentanyl
- **IN & MNT**: Propofol, fentanyl
- **Premed**: Atropine, hydroxyzine
- **GA**: Postop recovery after propofol discontinuation
- **BP elevation**: ECG changes resolved
- **EA**: Ablative ssisted surgery of the trachea
- **ECG**: Electrocardiogram
- **AP ablation**: Ablation of an accessory pathway
- **SVT**: Supraventricular tachycardia
- **mifentanil**: Mifentanil
- **Sugammadex**: Sugammadex
- **MNT**: Methylprednisolone
- **IN**: Indomethacin
- **Premed**: Risk reduction
- **GA**: General anesthesia
- **tubes**: Fallopian tubes
- **Laparoscopic**: Laparoscopic surgery
- **Thoracic**: Thoracic surgery
- **Ligation**: Ligation of the Fallopian tubes
- **Knee**: Knee arthroscopy
<table>
<thead>
<tr>
<th>Author</th>
<th>WPW Type</th>
<th>Age/Gender</th>
<th>Details</th>
<th>1)</th>
<th>2)</th>
<th>3)</th>
<th>4)</th>
<th>5)</th>
<th>6)</th>
<th>7)</th>
<th>8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rahul et al.</td>
<td>WPW</td>
<td>1F/42y</td>
<td>None</td>
<td>1) RA: CSE</td>
<td>SA: hyperbaric bupivacaine 0.5% + fentanyl</td>
<td>EA: plain bupivacaine 0.375% infusion + fentanyl bolus</td>
<td>T-wave inversion in leads I-II-III without hemodynamic instability</td>
<td>No treatment</td>
<td>•Uneventful recovery</td>
<td>•ECG normalised (at 24 h postop)</td>
<td></td>
</tr>
<tr>
<td>Shiroyama et al.</td>
<td>WPW†</td>
<td>1 pt / no further details</td>
<td>No further details</td>
<td>RA Spinal, upper sensory level: C6</td>
<td>WPW pattern on ECG</td>
<td>•ECG normalized in 3d</td>
<td>•High spinal block may cause ↓ AV &amp; ↑ AP conduction and unmask intermittent WPW</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lubarsky et al.</td>
<td>WPW†</td>
<td>n = 2 1M/14y</td>
<td>1) Transurethral resection of prostate</td>
<td>1) RA Premed: pethidine, pentobarbital, cefazolin SA with tetracaine (i.v.: diazepam, ephedrine, phenylpropranolol as needed)</td>
<td>2) GA Diazepam, isoflurane, N₂O</td>
<td>Postoperative wide QRS complexes (T6 block, low Na⁺, hypothermia, nausea)</td>
<td>•furosemide/warm saline</td>
<td>•Monitor vital signs</td>
<td>•Unilateral course</td>
<td>•Intrathecal opioids for labour analgesia do not induce sympathetic block: useful in WPW</td>
<td></td>
</tr>
<tr>
<td>Devisetti et al.</td>
<td>WPW</td>
<td>1F/20y</td>
<td>Evacuation of molar pregnancy</td>
<td>RA: SA with hyperbaric bupivacaine 0.5% + fentanyl</td>
<td>Level of sensory block: T8</td>
<td>•Other drugs: midazolam, fentanyl, oxycotin</td>
<td>Transient sinus tachycardia (130 bpm) during delivery</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•CSE useful and safe</td>
<td></td>
</tr>
<tr>
<td>Namshikar et al.</td>
<td>WPW</td>
<td>n = 2 IF/28y</td>
<td>Elective CDs</td>
<td>CSE SA: hyperbaric bupivacaine 0.5% (1.7-2 mL)</td>
<td>Postoperative epidural bupivacaine 0.1-0.125% ± fentanyl 50 μg</td>
<td>Other drugs: diclofenac, fentanyl, morphine</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•CSE useful and safe</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ruiz-Castro et al.</td>
<td>WPW/MV prolapse</td>
<td>1F/22y</td>
<td>CD</td>
<td>RA: EA</td>
<td>↓ BP 45 min after block</td>
<td>*Methoxamine</td>
<td>No further incidents</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge on 2nd postnatal day</td>
<td></td>
</tr>
<tr>
<td>Robinson et al.</td>
<td>WPW/FHPP</td>
<td>1F/29y</td>
<td>Forces assisted</td>
<td>RA: EA with bupivacaine/epinephrine, fentanyl, morphine</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
<td></td>
</tr>
<tr>
<td>Misra et al.</td>
<td>WPW</td>
<td>IF/25y</td>
<td>Forces assisted</td>
<td>VA: bupivacaine, fentanyl, chloroprocaine, lidocaine</td>
<td>Mild ↓ BP after delivery</td>
<td>*Phentylephrine</td>
<td>•Unilateral course</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
<td></td>
</tr>
<tr>
<td>Tachikawa et al.</td>
<td>WPW/EB</td>
<td>Not available</td>
<td>Ankle joint fracture repair</td>
<td>RA: CSE (no further details)</td>
<td>None</td>
<td>•Unilateral course</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sahu et al.</td>
<td>WPW</td>
<td>IF/45y</td>
<td>Abdominal hysterectomy</td>
<td>RA: CSE with bupivacaine, fentanyl</td>
<td>Postop PCEA</td>
<td>↓ BP after initial epidural bolus</td>
<td>*Phentylephrine</td>
<td>Postop ICU for 24 h monitoring</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
</tr>
<tr>
<td>Palaria et al.</td>
<td>WPW</td>
<td>IF/30y</td>
<td>Emergency CD</td>
<td>RA (CSE)</td>
<td>•SA with hyperbaric bupivacaine</td>
<td>•Postop EA: bupivacaine</td>
<td>•Other drugs: oxycotin</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Discharge after 48 h</td>
<td></td>
</tr>
<tr>
<td>Kaur et al.</td>
<td>WPW</td>
<td>IF/30y</td>
<td>CD</td>
<td>RA (CSE)</td>
<td>•SA: hyperbaric bupivacaine 0.5% (1.8 mL) - block up to T8</td>
<td>•EA: bupivacaine 0.5% - level T5</td>
<td>•Postop EA: bupivacaine 0.125%</td>
<td>None</td>
<td>•Oxycotin was withheld</td>
<td>•Prepare defibrillator / antiarrhythmics</td>
<td></td>
</tr>
<tr>
<td>Kabude et al.</td>
<td>WPW</td>
<td>IF/48y</td>
<td>Abdominal hysterectomy</td>
<td>RA: EA with bupivacaine, lidocaine</td>
<td>•Other drugs: alprazolam, ranitidine, ondansetron, midazolam</td>
<td>None</td>
<td>•Unilateral course</td>
<td>•Intrathecal for haemodynamic stability</td>
<td>•Prepare defibrillator / antiarrhythmics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Author(s)</td>
<td>WPW ±</td>
<td>Age</td>
<td>Procedure</td>
<td>Description</td>
<td>Anesthesia/Agents</td>
<td>Intervention</td>
<td></td>
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<tr>
<td>Shora et al.</td>
<td>WPW</td>
<td>1F/27y</td>
<td>Emergency CD</td>
<td>RA: SA with hyperbaric bupivacaine, fentanyl</td>
<td>SVT (220 bpm) with ↓ BP</td>
<td>Pt transferred to ICU / EPS / RFCA in 2 m → neonate died due to unrelated reason</td>
<td></td>
<td></td>
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<tr>
<td>Jacobson</td>
<td>WPW</td>
<td>1M/17y</td>
<td>Eye surgery</td>
<td>GA (premed: diazepam)</td>
<td>2 SVT episodes with ↓ BP (at induction and postop)</td>
<td>Unventful course</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Van Zijl et al.</td>
<td>WP+W</td>
<td>1F/27y</td>
<td>Surgery</td>
<td>GA: ETomidate, succinylcholine</td>
<td>SVT (220 bpm) with ↓ BP</td>
<td>Unventful course</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Okamoto</td>
<td>WPW</td>
<td>1F/36y</td>
<td>CD</td>
<td>RA: EA with mepivacaine</td>
<td>4 SVT episodes</td>
<td>Verapamil, effective</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Bronheim</td>
<td>WPW</td>
<td>1M/51y</td>
<td>Surgical ablation</td>
<td>Propranolol before anaesthesia</td>
<td>Paroxysmal SVT after pericardial manipulation</td>
<td>Further procedure uneventful / Adenosine is effective in WPW syndrome</td>
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<tr>
<td>Jones et al.</td>
<td>WPW</td>
<td>n = 3 (1WPW)</td>
<td>(IWPW)</td>
<td>Cholecystectomy</td>
<td>Propranolol before anaesthesia</td>
<td>None</td>
<td></td>
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<tr>
<td>Wheeler et al.</td>
<td>WPW</td>
<td>1M/72y</td>
<td>CABG</td>
<td>GA</td>
<td>Postop wide / narrow QRS tachy</td>
<td>Unventful course</td>
<td></td>
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<tr>
<td>Nishikawa et al.</td>
<td>WPW</td>
<td>1M/50y</td>
<td>Transurethral resection of bladder tumour</td>
<td>RA: Premed: secobarbitone ▪ SA: tetracaine ▪ Obturator nerve block with lidocaine</td>
<td>Electrical stimulation for block triggered paroxysmal tachycardia</td>
<td>Uncomplicated course / Hospitalized for 16 days / Tachy episodes after discharge</td>
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<tr>
<td>Chhabra et al.</td>
<td>WPW</td>
<td>1M/15</td>
<td>Modified radical mastoidectomy</td>
<td>GA: IN: bupivacaine, fentanyl, alcuronium MNT: enflurane, N₂O RV: glycopyrrolate / neostigmine</td>
<td>WPW pattern, but haemodynamically stable ▪ Switch to enflurane, lidocaine i.v. (no effect)</td>
<td>Surgery continued and completed uneventfully / Normal SR after extubation / discharge after cardiac evaluation</td>
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<tr>
<td>Esenther et al.</td>
<td>Intermittent</td>
<td>1M/4y</td>
<td>Elective bronchoscopy</td>
<td>GA: Sevoflurane, N₂O</td>
<td>intermittent tachyarrhythmia haemodynamically stable ▪ None</td>
<td>Unventful course</td>
<td></td>
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<tr>
<td>Braun et al.</td>
<td>Intermittent</td>
<td>1F/30y</td>
<td>CD</td>
<td>GA: (RA not preferred because pt was receiving heparin SC)</td>
<td>intermittent tachyarrhythmia haemodynamically stable ▪ None</td>
<td>Unventful course (Main focus of paper is on the management of post-Fontan parturients)</td>
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</tbody>
</table>


Table 3. Publications on perioperative management, complications and outcome of patients with rare pre-excitation syndromes: Mahaim fiber and Lown-Ganong-Levine syndrome

<table>
<thead>
<tr>
<th>First author (publication year)</th>
<th>Syndrome - clinical description</th>
<th>No of pts - Age</th>
<th>Type of surgery</th>
<th>Type of anaesthesia / Drugs used</th>
<th>Complications / Management</th>
<th>Outcome / Points of interest</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sadowski (1979)</td>
<td>In total: n = 13</td>
<td>LGL: n = 2</td>
<td>1 case LGL†</td>
<td>1) bundle division 2) 2 other operations</td>
<td>1) GA: enflurane, N₂O, d-tubocurarine 2) GA: morphine, N₂O, pancuronium or d-tubocurarine</td>
<td>1) None</td>
</tr>
<tr>
<td>Zweifler (2011)</td>
<td>Mahaim fiber†</td>
<td>1F/ 38y</td>
<td>Spinal surgery</td>
<td>Plan for GA Premed in OR: midazolam, fentanyl</td>
<td>After premed: † HR (180 bpm) - wide QRS - varying morphology *Lidocaine: SR restoration</td>
<td>•12-lead ECG: T-wave inversion&lt;br&gt;•Surgery postponed / EPS &amp; AP ablation / surgery: 2w later</td>
</tr>
<tr>
<td>Sharma (2011)</td>
<td>LGL</td>
<td>1F/ 42y</td>
<td>Cholecystectomy</td>
<td>GA &amp; EA (lumbar) for postop analgesia&lt;br&gt;IN: midazolam, propofol, fentanyl, vecuronium&lt;br&gt;MNT: TIVA with propofol&lt;br&gt;RV: glycopyrrolate/neostigmine</td>
<td>1 episode of SVT&lt;br&gt;*Carotid sinus massage</td>
<td>•Uneventful course&lt;br&gt;•TIVA with propofol &amp; adequate postop analgesia may be useful</td>
</tr>
<tr>
<td>Jones (1984)</td>
<td>In total: n = 3</td>
<td>LGL: n = 1</td>
<td>1F/ 36y</td>
<td>Mastectomy</td>
<td>GA / premed: diazepam, continuation of medication (verapamil) + propranolol&lt;br&gt;IN: thiopental, fentanyl, alcuronium&lt;br&gt;MNT: enflurane, N₂O&lt;br&gt;RV: glycopyrrolate/neostigmine</td>
<td>None</td>
</tr>
</tbody>
</table>


No of pts: number of patients, †: disease undiagnosed, *: management of complication


no of pts: number of patients, †: disease undiagnosed, *: management of complication

and populations may account for some contradictory findings, although we consider that safe conclusions could be drawn for most anaesthetics used in WPW. For the more rare pre-excitation conditions, data were rather limited, since only a small number of case reports were identified. Nonetheless, we think that the present review may contribute to a better understanding of pre-excitation syndromes and provide useful information for their perioperative management.

Conclusion

Anaesthesia and perioperative care of patients with pre-excitation syndromes may be difficult, especially if they are undiagnosed, under-treated or if there is no adequate time for clinical optimisation, as in emergencies. These patients are at a high risk of developing life-threatening arrhythmias perioperatively. Close cooperation with a cardiologist is mandatory,
while increased vigilance and postoperative cardio-vascular monitoring will allow prompt therapeutic intervention in cases of arrhythmias.

Conflict of interest
Nothing to declare

References
5. Öhnell RF. Pre-excitation, cardiac abnormality, pathophysiologial, patho-anatomical and clinical studies of excitatory spread phenomenon bearing upon the problem of the WPW (Wolff, Parkinson, and White) electrocardiogram and paroxysmal tachycardia. Acta Med Scand 1944; 152: 1-167
27. Dobkowski WB, Murkin JM, Sharpe MD, Sharmar0 AD, Yee R, Guiraudon G. The effect of isoflurane (1 MAC) on the normal


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