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The patient with congenital heart disease in ambulatory surgery

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The number of patients with congenital heart disease (CHD) undergoing ambulatory surgery is increasing. Deciding whether a CHD patient is suitable for an ambulatory procedure is still challenging. Several factors must be considered, including the type of planned procedure, the complexity of the underlying pathology, the American Society of Anesthesiologists' Physical Status classification of the patient, and other patient-specific factors, including comorbidity, chronic complications of CHD, medication, coagulation disorders, and issues related to the presence of a pacemaker (PM) or cardioverter-defibrillator.

Numerous studies reported higher perioperative mortality and morbidity rates in surgical patients with CHD than non-CHD patients. However, most of these studies were conducted in a cohort of hospitalized patients and may not reflect the ambulatory setting. The current review aims to provide the anesthesiologist

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with an overview and practical recommendations on selecting and managing a CHD patient scheduled for an ambulatory procedure.

Introduction

Congenital heart disease (CHD) is one of the most common birth defects. An epidemiological study published in 2019 reported an overall incidence of CHD of approximately 10 in 1000 live births worldwide, with patent ductus arteriosus, atrial septal defect, and ventricular septal defect being the three most frequent anomalies [1]. Fortunately, a high percentage of patients with CHD currently survive to adulthood because of the advancements in pediatric cardiology, surgery, anesthesia, and critical care [2,3]. Therefore, the term GUCH (Grown Up Congenital Heart) has become inappropriate and has recently been replaced with ACHD (Adult Congenital Heart Disease) by the European Society of Cardiology (ESC) [4]. It is not uncommon that these patients may undergo in−or out-patient noncardiac procedures and would need to be managed by a noncardiac anesthesiologist. Management of such patients in an ambulatory setting is challenging and requires a multidisciplinary approach [5,6].

According to several studies, CHD patients (both children and adults) undergoing noncardiac surgery have a higher risk of perioperative morbidity and mortality than those without CHD [7−10]. Yet, these studies have been performed in hospitalized patients and may not necessarily mirror the current ambulatory setting in this population. Therefore, to successfully manage a patient with CHD planned for an ambulatory procedure and to avoid unscheduled hospital admission, careful patient selection and an appropriate perioperative anesthesia management plan are essential. To this end, it is fundamental for the anesthesiologist to have a good knowledge of the pathophysiology of the congenital heart defect, understand its hemodynamic consequences and/or chronic complications related to the defect, and plan an applicable anesthetic technique.

The current review aims to provide the anesthesiologist with an overview of the CHD spectrum and offers practical recommendations for managing these patients who are scheduled for an ambulatory surgery/procedure.

Patient suitability for an ambulatory procedure

Ambulatory anesthesia and surgery are not new concepts. In early 1985, the Society of Ambulatory Anesthesia was established, and, ten years later, the International Association of Ambulatory Surgery (IAAS) was created [11,12]. Since then, many patients have been managed in ambulatory settings, and ambulatory or day surgery has considerably grown in various countries [13,14]. According to the IAAS, an ambulatory (day) surgery/procedure is defined as an operation or an intervention that is carried out in a hospital or a center optimized for the management of such procedures, and the patient is discharged from the hospital on the same day of the surgery/procedure. In contrast, extended-day surgery is when the patient needs to stay one night at the hospital and be discharged the following day [14,15]. The terms “ambulatory,” “outpatient,” or “day hospital” surgery/procedure will be used interchangeably in the present review.

To accomplish a successful ambulatory procedure, both the procedure and the patient must be suitable for this setting. Healthcare providers need to understand the categories of CHD and estimate the perioperative risk to appropriately classify a CHD patient as suitable for an ambulatory procedure. According to the recent ESC and American Heart Association (AHA) guidelines, the complexity of CHD can be divided into three categories: Mild/Moderate/Severe or Simple/Moderate/Great Complexity (Table 1) [4,16]. Faraoni and colleagues have demonstrated that the severity of the cardiac defect and the comorbidities are the main predictors of perioperative 30-day mortality in children with CHD [17]. Likewise, several authors have indicated that perioperative risk estimation predominantly depends on the patient's underlying pathology [18] and the type of surgery or intervention [19,20].

Table 1
Classification of congenital heart disease according to the european society of cardiology (ESC) and the American heart Association/American college of cardiology (AHA/ACC) [4,16].

ESC	AHA/ACC
Mild	Simple
Isolated small ASD/VSD/PDA Mild pulmonary stenosis Repaired secundum ASD, SVD, and PDA with no significant residual shunt Isolated AV/MV disease (e.g., bicuspid AV, MV prolapse. Excluding cleft leaflet or parachute valve)	
Moderate	**Moderate complexity**
Aortic stenosis Anomalous pulmonary venous connection Anomalous coronary artery origin AVSD (partial/complete/primum ASD) Ebstein anomaly Coarctation of aorta Pulmonary stenosis Sinus of Valsalva fistula/aneurysm SVD Repaired ToF Moderate/large unrepaired secundum ASD Moderate/large PDA VSD with abnormality/at least moderate shunt TGA (after switch operation)	Congenital AV/MV disease (any)
Severe	**Great complexity**
Cyanotic CHD Double outlet ventricle Fontan Interrupted aortic arch Univentricular Truncus arteriosus Other abnormalities of atrioventricular connection Mitral or pulmonary atresia TGA (unrepaired) Any CHD associated with pulmonary vascular disease	TGA (all)

ASD: Atrial septal defect, AV: Aortic valve, AVSD: Atrioventricular septal defect, CHD: congenital heart disease, MV: Mitral valve, PDA: Patent ductus arteriosus, SVD: Sinus venosus defect, TGA: Transposition of great arteries, ToF: Tetralogy of Fallot, VSD: Ventricular septal defect.

A recent data analysis of over two million adult day-surgery cases showed that the ASA Physical Status classification is a simple and reliable risk stratification instrument for surgeries or interventions carried out in ambulatory surgery centers [21]. Likewise, the study concluded that patients with lower ASA scores had fewer postoperative complications or mortality than those with high ASA scores after ambulatory surgery [21]. In addition, several patient-specific factors, such as associated anomalies, the coagulation status, and risk of perioperative bleeding, the presence of a pacemaker (PM) or implantable cardioverter-defibrillator (ICD), and an increased risk of arrhythmias, need to be assessed when evaluating suitability for ambulatory surgery or interventions. Finally, each ambulatory center handling CHD patients should have the facilities to manage patients postoperatively when unplanned hospital admission is required. This should also be considered during patient screening for an ambulatory procedure.

According to various studies, the incidence of unplanned hospital admission after an ambulatory procedure varies from 0.6% to 9.7% [22–24]. Koichi Yuki and colleagues showed in a retrospective study that 2,7% of the children with CHD who underwent ambulatory procedure/surgery required unplanned hospital admission [24]. This admission rate was comparable to the one recently reported in a general population of adults after ambulatory procedure/surgery [23].

Notably, anesthesia-related factors attributed to unscheduled hospital admission were inadequate postoperative pain management, postoperative nausea and vomiting (PONV), hypoxia, headache, or fever [24].

Complications and long-term consequences of CHD

As mentioned earlier, patients' comorbidities are associated with a high risk of unplanned hospital admission and postoperative mortality [17,23]. Therefore, it is essential for healthcare providers managing CHD patients to have adequate knowledge of complications and late consequences of CHD. Adult patients with a history of CHD may present with one or more of the following long-term consequences of CHD: heart failure, pulmonary hypertension (PH), arrhythmias, chronic hypoxemia, and polycythemia [25,26]. Other complications, such as infective endocarditis (IE), neurological or renal abnormalities, and socio-economic factors, must also be considered [25,27,28].

Heart failure

Heart failure is one of the fundamental reasons for death in adults with CHD [29]. The pathophysiology of heart failure in CHD patients [30] is complex. It comprises, for example, long-lasting volume overload of the ventricles, pressure overload of the subpulmonary ventricle or the systemic ventricle, as in patients with a congenitally corrected transposition of the great arteries or after an atrial switch (Mustard or Senning) operation, arrhythmia, chronic hypoxemia, polycythemia, and other mechanisms [6,25,31]. Patients with CHD suffering from heart failure are, per definition, considered high-risk patients and recommended to be evaluated in advance by a multidisciplinary team for ambulatory surgery eligibility.

Chronic hypoxemia and polycythemia

Hypoxemia (reduced oxygen levels in the blood) [32] is a common feature in patients with a right-to-left shunt [33]. As a result, erythrocytosis develops to preserve oxygen content and delivery [34]. Also, patients with chronic hypoxemia and polycythemia have a higher risk of thrombosis and bleeding due to increased blood viscosity and decreased levels of vitamin K-dependent coagulation factors and Von Willebrand factor, respectively [35]. Again, these patients may not be ideally managed in an ambulatory surgery setting. An optimal perioperative strategy includes maintaining adequate pre-, per- and post-operative hydration, interrupting anticoagulant therapy only as short as possible, and aiming for early postoperative mobilization [35,36].

Pulmonary hypertension

Pulmonary hypertension (PH) is one of the essential prognostic elements in patients with CHD [4]. Of note, the definition of PH has recently changed; a patient is now considered to have PH when his invasively measured mean pulmonary artery pressure (mPAP), at rest, is above 20 mmHg [4,37]. According to the underlying pathology of CHD, PH can be subdivided into pre-capillary, post-capillary, and combined pre- and post-capillary PH (Table 2) [4,37,38]. It is essential to differentiate between these subtypes of PH related to CHD, as only the pre-capillary PH subtype may benefit from pulmonary vasodilators [25]. In general, patients with moderate to severe PH are not advisable to be managed in an ambulatory surgery setting [39].

Arrhythmias

CHD patients are more likely to experience arrhythmias because of the natural history of the underlying pathology [40]. Specifically, the etiology of arrhythmias can be attributed to congenitally abnormal anatomy/physiology, chronic hypoxemia, a surgical injury that occurred in a previous cardiac operation, or chronic dilation of the atria and/or ventricles [35,41]. Arrhythmias, particularly atrial arrhythmias, are associated with an increased risk of hospital admission and morbidity in CHD patients

Table 2
Definition, mechanisms, and pulmonary hypertension subgroups in congenital heart disease. [4,37,38].

PH subgroups	Definitions	Mechanisms and clinical settings
Pre-capillary (PAH)	•mPAP >20 mmHg •PVR ≥3 WU (240 dyn s•cm^{-5}) •PCWP ≤15 mmHg	•Systemic to pulmonary shunt lesions •Eisenmenger syndrome •Univentricular heart defect
Post-capillary	•mPAP >20 mmHg •PVR <3 WU (240 dyn s•cm^{-5}) •PCWP >15 mmHg	•Systemic ventricular failure •Systemic AV valve stenosis •Systemic AV valve regurgitation •Pulmonary vein stenosis •Cor triatriatum
Combined pre- and post-capillary	•mPAP >20 mmHg •PVR ≥3 WU (240 dyn s•cm^{-5}) •PCWP >15 mmHg	•Mechanisms listed under post-capillary PH •Mechanisms listed under post-capillary PH in combination with systemic to pulmonary shunt lesions or severe CHD

AV: atrioventricular, CHD: congenital heart disease, mPAP: mean pulmonary artery pressure, PAH: pulmonary arterial hypertension, PCWP: pulmonary capillary wedge pressure, PH: pulmonary hypertension, PVR: pulmonary vascular resistance, UVH: univentricular heart, WU: Wood units.

[42,43]. A recent multicenter study included 482 adult patients with CHD and atrial arrhythmias. It demonstrated that intra-atrial re-entry tachycardia, atrial fibrillation, and focal atrial tachycardia are the most common form of atrial arrhythmias in this population cohort [44]. Patients with CHD may also suffer from ventricular arrhythmias, particularly those with poor systemic ventricular function or those who developed myocardial fibrosis because of the previous ventriculotomy, like in patients with tetralogy of Fallot [45,46]. It is not uncommon for these patients to have a PM or an ICD that may or may not require reprogramming by the cardiologist before and after the procedure. The necessity to reprogram depends on the expected electromechanical interference during the case and the distance of the intervention site to the device. A defibrillator should be present in the operating room. The anesthesiologist should be acquainted with managing arrhythmias associated with CHD and should have full access to anti-arrhythmic and other emergency medications.

Infective endocarditis and CHD

Patients with CHD have a higher risk of infective endocarditis (IE) than the general population [47–49]. This may be attributed to numerous mechanisms like the presence of prosthetic valves and other foreign materials, stagnation of blood flow because of arrhythmia or low ventricular function, the presence of an intracardiac shunt or outflow tract obstruction resulting in blood flow disturbance with micro damage to endothelial cells, frequent re-interventions, and recurrent hospital admissions [50–53]. IE accounts for 4% of patient admissions per year in ACHD units in the United Kingdom. All patients should therefore be adequately educated about the signs and symptoms of IE [54,55].

In 2015, the ESC published guidelines concerning the indication of antibiotics to prevent IE [56]. According to these guidelines, antibiotic prophylaxis should only be administered to CDH patients with a higher risk of developing IE: those with a previous history of IE, all cyanotic CHD patients, and patients who underwent palliative correction. Of note, in CHD patients with a complete defect repair, IE prophylaxis is indicated until six months post-repair. Lifelong antibiotic prophylaxis for IE needs to be considered if a residual defect, shunt, or valve regurgitation remains [56]. For CHD patients mentioned above, antibiotic prophylaxis should be administered 30–60 min before the ambulatory procedure. The following are examples of procedures where high-risk patients need to receive antibiotic prophylaxis: dental care procedures involving manipulation of the gingival or periapical region of the teeth, perforation of the oral mucosa, procedures involving incision or biopsy of the respiratory tract mucosa, or inserting of a chest tube for abscess drainage, placement of a PM or an ICD device, an invasive gastrointestinal procedure in a patient with an infection or needs to receive antibiotics to avoid wound infection or sepsis related to this procedure, cystoscopy or other elective urinary tract manipulations in a patient with urinary tract infection or colonization, and procedures on infected skin or musculoskeletal tissue (Table 3) [56–58].

Table 3
Recommended antibiotic prophylaxis for high-risk procedures in specific CHD patients. [56–58].

Patient selection	Procedures associated with a high risk of endocarditis infection	Types and doses of Antibiotics prophylaxis. AB should be given 30–60 min before the procedure
• Any type of cyanotic CHD • Any type of CHD repaired with a prosthetic material for up to 6 months. • Lifelong IE antibiotic prophylaxis if residual shunt or valvular regurgitation remains. • Previous history of IE.	**Dental procedures** with manipulation of the gingival or periapical region or perforation of the oral mucosa.	**First choice:** amoxicillin or ampicillin - Adult 2 g oral or IV. - Children 50 mg/kg oral or IV (max 2 g) **Alternatively:** cefazolin or ceftriaxone - Adults 1 g IV - Children 50 mg/kg IV (max 2 g) In case of an allergy to penicillin or ampicillin, one of the following antibiotics can be given Clindamycin - Adults 600 mg oral, IV or IM - Children 20 mg/kg oral, IV or IM (max. 600 mg) Cephalexin - Adults 2 g IV - Children 50 mg/kg IV (max 2 g) Azithromycin or clarithromycin - Adults 500 mg oral or IV infusion over 60 min - Children 15 mg/kg (max 500 mg) oral or IV infusion over 60 min.
	Invasive respiratory tract procedures to treat an established infection or take biopsies.	Antibiotic agents active against S.viridans, Anti-staphylococcal drug (i.e., ampicillin or cefazoline) if infection with S.aureus is known or suspected
	Invasive gastrointestinal and **genitourinary tract** procedures; AB prophylaxis is indicated to avoid wound infection or sepsis related to these procedures.	**Anti-enterococcal** drug (i.e., ampicillin, amoxicillin, vancomycin).
	Procedures on **infected skin** or **musculoskeletal tissue.**	Antibiotic agents active against **staphylococci** and **beta-hemolytic streptococci** (i.e., anti-staphylococcal penicillin or a first-generation cephalosporin)
	Placement of a PM or an ICD device.	AB regimes similar to that used for dental procedures.

AB: antibiotic, CHD: congenital heart disease, ICD: implantable cardioverter-defibrillator, IE: infective endocarditis, IM: Intramuscular, IV: intravenous, max: maximum, min: minute, PM: pacemaker.

To our knowledge, insufficient data are available concerning the incidence of IE in CHD patients following ambulatory surgery. Nevertheless, both the recommendations mentioned above, and local endocarditis prophylaxis guidelines should be strictly obeyed.

Anesthetic considerations and management

Preoperative assessment

Patients with CHD scheduled for an ambulatory procedure require an advanced preanesthetic assessment. This should include a medical and surgical history record, results and reports of recent investigations, if applicable, and a thorough physical examination. In addition, the anesthesiologist needs to assess whether the CHD patient is feasible to be managed in an ambulatory setting. Finally, an appropriate anesthesia management plan should be established and discussed with the patient (and/or their parents, in the case of minors).

Of note, several CHD patients may have other congenital anomalies associated with difficult airway management, such as trisomy 21, Beckwith-Wiedemann syndrome, Pierre Robin syndrome, and other anomalies [59–61]. Additionally, late sequels of CHD, such as heart failure, PH, chronic hypoxemia and polycythemia, arrhythmia, and protein loss enteropathy, must be identified.

Premedication with an anxiolytic drug could be of great value in reducing preoperative anxiety in CHD patients. However, these medications should be cautiously administered, particularly in patients with PH, because of the risk of carbon dioxide retention resulting from hypoventilation, as this may worsen the condition. The preoperative anesthesia assessment is presented in detail in Table 4.

Preoperative fasting

International and institutional preoperative fasting guidelines should be adhered to [62,63].

As mentioned earlier, patients with polycythemia as a consequence of long-term hypoxemia are at an increased risk of thrombotic complications if preoperative fasting is extensively prolonged. Likewise, patients with Fontan circulation depend on adequate preload levels to sustain passive flow across their pulmonary circulation [64]. In both cases, when prolonged fasting is anticipated, fluids should be given intravenously to maintain euvolemia while patients remain fasted.

Intraoperative management

Considering the underlying pathology (i.e., category of CHD) and type of surgery/procedure, key factors related to the intraoperative management of a patient with CHD are to provide adequate oxygenation and ventilation and to ensure a stable hemodynamic status through achieving an optimal balance between systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR). To our knowledge, there is no specific guidelines concerning the best anesthetic agent or anesthesia technique for CHD patients. Nevertheless, techniques that will not significantly alter hemodynamics should be

Table 4
Anesthetic assessment checklist for patients with congenital heart disease.

What are the underlying pathology and anatomical configuration of the heart and great vessels?
• Repaired/Unrepaired/Partial/Palliative correction?
• Residual shunts?
• Recent investigations?
• Potential issues with IV access?
What is the ASA-PS status of the patient?
What is the risk of surgery?
• Minor, intermediate, or High-risk surgery?
Are there any associated anomalies?
• Some syndromes with CHD also carry increased airway difficulties due to anatomical variance.
Are there any comorbidities or chronic sequels of CHD?
• Any end-organ problems?
• Ventricular function?
• Pulmonary hypertension?
Current physical status?
• Baseline saturation to air?
• Exercise capacity?
• NYHA Functional Classification?
Any history of arrhythmias?
• Implantable cardiac device present?
• Pacemaker present?
• Need for reprogramming/magnet application?
Anticoagulation issues
• Surgical bleeding risk?
• Indication for stopping/bridging of anticoagulation drugs?
Medication
• Determine which medication should be continued/taken on the day of surgery
• Any need for premedication?
• Is infective endocarditis prophylaxis indicated?
Anesthesia technique and monitoring
• GA/LRA/PNB/Sedation/LA?
• Need for invasive monitoring?

ASA PS: America Society of Anesthesiologists Physical Status, CHD: Congenital Heart Disease, GA: General anesthesia, LA: Local anesthesia, LRA: Locoregional anesthesia, NYHA: New York Heart Association Functional Classification, PNB: Peripheral nerve block.

favored. Almost any anesthetic medication can be used for CHD patients if the anesthesiologist is fully aware of its pharmacokinetic and pharmacodynamic profile and thoroughly understands the pathophysiology and the hemodynamic changes associated with CHD. Several anesthetics, such as sevoflurane and propofol, cause a dose-dependent reduction in SVR and have a slightly negative inotropic effect on the heart. Therefore, unnecessary overdosing of these agents should be avoided, particularly in patients with poor systemic ventricular function and/or PH [65–69]. More details on intraoperative anesthesia management of CHD patients are described in Fig. 1 [35,70–72].

Postoperative management and hospital discharge

The main goal of the recovery process is to return the patient to their preoperative physiological state. In this respect, patients with CHD do not differ from other patients undergoing ambulatory surgery. Many ambulatory centers use the modified Aldrete Scoring System, which determines patient readiness for Post-Anesthesia Care Unit (PACU) discharge [73,74]. This score is based on vital parameters such as respiration, blood pressure, motoric activity, consciousness, and oxygen saturation. Each parameter is scored between 0 and 2. When patients score $\geq$9, they are permitted to leave the PACU and be admitted to a Step-Down Unit (SDU), an intermediate unit with a specific level of care between PACU and hospital discharge [2]. Of note, for cyanotic CHD patients with preoperative oxygen (O_2) saturation <90%, the discharge criteria must be modified according to the patient's preoperative O_2 saturation levels.

In the SDU, a written protocol with specific discharge criteria is advisable [73,74]. Pain and PONV are not included in the modified Aldrete scoring system. Therefore, these complications should be strictly managed before patient discharge.

At home, a dedicated nurse should call patients (or their parents) to inquire about pain and postoperative complications [75–77]. In case of difficulties or not fulfilling the discharge criteria, the patient should be admitted to the hospital.

Furthermore, there are still some specific considerations that one must consider in patients with CHD. For example, patients with a conduit, shunt, or prosthetic valve are at risk of a thrombotic event after cessation of anticoagulant drugs. Hence, these patients must restart their anticoagulant as soon as possible while considering the surgical bleeding risk. In addition, if a PM or ICD is present, device testing after surgery is needed if its original settings have been changed.

Locoregional anesthesia for CHD

Many procedures in the ambulatory surgery department can be done under locoregional anesthesia, sometimes combined with periprocedural sedation or even general anesthesia. Numerous centers also offer regional anesthesia to the pediatric population (e.g., caudal block). The use of locoregional anesthesia can provide adequate pain relief both intra- and post-operatively at home. The feasibility of these techniques in children with CHD is generally the same as in the normal pediatric population [78]. The same holds true for adult patients with CHD.

Patients with CHD will often be treated with one or more anticoagulants. This may preclude the use of central nervous blocks such as spinal, epidural, or caudal anesthesia. In contrast, peripheral nerve blocks are usually not contra-indicated, as stated in most international regional anesthesia guidelines [79,80]. Some studies suggest that CHD patients are at increased risk of acquired coagulation disorders, so this should be considered during preoperative assessment [81].

Using regional anesthesia as the sole anesthetic technique, airway manipulation with positive pressure ventilation and positive end-expiratory pressure (which would increase PVR) can be avoided altogether. Adequate pain relief will also keep sympathetic stimulation down.

On the other hand, SVR might drop with the use of neuraxial techniques (more in older children and adults) and could reverse a left-to-right shunt over nonrestrictive defects into a right-to-left shunt. These side effects can be mitigated by using low doses of a vasopressor agent, maintenance of a euvolemic state, using incremental techniques such as (continuous spinal-) epidural anesthesia, and low doses of local anesthetics [82].

Likewise, adding epinephrine five micrograms per milliliter to the local anesthetic solution for a caudal block can increase cardiac output and decrease mean arterial pressure through the beta effects

O_2: Oxygen, PEEP: positive end expiratory pressure, PVR: pulmonary vascular resistance, SVR: systemic vascular resistance; VR, vascular resistance.
* Considered as high-risk patient and are uncommon to be managed in day surgery.
** Should be administered with caution. Only use of incremental and low dose neuraxial anesthesia (Epidural or combined spinal and epidural (CSE)).

Fig. 1. Intraoperative anesthesia management of congenital heart disease patients [35,70–72].

Table 5
Anesthesia technique and its related hemodynamic changes in congenital heart disease patients.

	Neuraxial anesthesia	General anesthesia
Preload	↓↓	↔
Afterload	↓↓	↓
Contractility	↔	↓
Heart rate	↓ or ↔	↔ or ↑
Advantages	• Spontaneous breathing • Excellent analgesia. • Less incidence of PONV.	• Control and monitoring of gas exchange (FiO_2 and $etCO_2$).
Disadvantages	• Contraindicated in patients with active anticoagulation • Can cause significant systemic hypotension and must, therefore, be used with caution in patients with obstructive valve lesions or patients with Right-to-Left shunt	Risk of: • Failed intubation • Sympathetic stimulation due to direct laryngoscopy and intubation procedure • Need for positive pressure ventilation with associated decrease in venous return and increased PVR • Higher incidence of PONV • Most inhalation and intravenous anesthetics (ketamine to a lesser extent) can significantly affect loading conditions and cardiac inotropy and should, therefore, carefully be administered in patients with moderate and severe CHD

↔: no change, ↓: slight decrease, ↓↓: considerable decrease, CHD: congenital heart disease, etCO2: End-tidal carbon dioxide, FiO2: fraction of inspired oxygen, PONV: postoperative nausea and vomiting, PVR: pulmonary vascular resistance, RV: right ventricle.

of adrenaline after epidural absorption [83]. Specific advantages and disadvantages of neuraxial versus general anesthesia are displayed in Table 5.

Although patients with CHD are known to be at increased risk for arrhythmias, to our knowledge, no studies have indicated an increased risk of Local Anesthetic Systemic Toxicity. However, as with all forms of regional anesthesia, it is advised to adhere to local protocols or international guidelines for maximum doses of local anesthetics [84,85].

Procedural sedation for CHD

Monitored anesthesia care (MAC) is rapidly gaining popularity for some ambulatory and outpatient procedures. Shorter acting sedative agents offer an attractive alternative to general anesthesia for shorter diagnostic or therapeutic procedures. Patient-specific concerns for children and/or adults with CHD are mostly related to the depth of anesthesia and its repercussions on pulmonary and systemic vascular resistance. Many anesthetic agents are available for procedural sedation, each with its own pharmacokinetic and pharmacodynamic profile and advantages or disadvantages (Fig. 2).

One of the most widely used techniques in the ambulatory setting is propofol sedation because of its suitable pharmacological profile and few side effects (quick onset and offset, reliable effect, less PONV). Although not contraindicated, the use of propofol in the CHD group warrants cautions, particularly in those patients with an intracardiac shunt, as it can drop SVR and worsen a right-to-left shunt [86,87].

Midazolam is widely used for procedural sedation in pediatric and adult populations. A significant advantage is the availability of an oral form, which facilitates administration in pediatric patients. Nevertheless, there are a few shortcomings to using midazolam: an unpredictable duration of its effect (especially with repeated doses), the potential for paradoxical reactions [88], and depression of ventilatory drive with an increase of PVR [89]. The latter may critically impact the patient's perioperative hemodynamics and outcome.

Dexmedetomidine is a comparatively new product, but it has been extensively investigated for its use in procedural sedation for CHD patients [90–92]. Effects on airway and breathing are minimal, and supplemental oxygen is rarely needed. It can be given intranasally, which generally suffices for

↑: increased, ↓: decreased, *FiO_2: Fraction of inspired oxygen, IM: intramuscular, PONV: postoperative nausea and vomiting, POD: postoperative delirium, POCD: postoperative cognitive dysfunction, PVR: pulmonary vascular resistance, R-L: right to left, SpO_2: oxygen saturation, SVR: systemic vascular resistance.*

Fig. 2. Pharmacological and non-pharmacological options and considerations for monitored anesthetic care in the congenital heart disease patient.

nonpainful procedures such as transthoracic echocardiography or magnetic resonance imaging scans [93,94]. With a more painful procedure such as cardiac catheterization, a second sedative or analgesic is often associated [95].

Dexmedetomidine has minimal s ide effects like bradycardia and hypotension [96], which can be well tolerated by CHD patients if used in moderate doses. While systolic arterial blood pressure tends to increase during the loading dose, pulmonary artery pressure does not [97].

Ketamine is an old drug that can be given intramuscularly if needed. It is well-known for its hemodynamic stability and reliable maintenance of airway and ventilation. Because of frequent visual hallucinations, a second drug like midazolam or dexmedetomidine is usually associated.

Hypoventilation and apnea resulting from more profound levels of sedation or excessive use of opiates should be avoided in patients with CHD because they increase the partial pressure of carbon dioxide levels and worsen PH [98]. Likewise, with more extended periods of apnea, hypoxia will contribute as well.

During procedural sedation, supplemental oxygen is typically given through a nasal cannula or a face mask. In CHD patients, a high fraction of inspired oxygen (FiO_2) can increase pulmonary blood flow in patients with left-to-right shunt and consequently cause systemic hypotension. It is therefore recommended to titrate FiO_2 during sedation in order to maintain the patient's baseline oxygen saturation levels [99]. Hypothermia should also be avoided for its negative effect on PVR and myocardial contractility, especially in prolonged cases [100].

Finally, it is worth mentioning that there is increasing experience with nonpharmacological options for perioperative sedation, such as virtual reality, audiovisual distraction, or hypnosis. Given their limited hemodynamic impact, they may pose good alternatives for MAC in centers experienced in these techniques. However, revising the advantages and limitations of these techniques is beyond the scope of the present review.

Summary

Because of the improvements in pediatric cardiology, cardiac surgery, anesthesia, and critical care medicine, a considerable number of children with CHD currently survive to adulthood and might therefore present for ambulatory noncardiac surgery/procedure later in life. However, developing an optimal anesthesia technique based on a thorough knowledge of the pathophysiology of CHD and its chronic consequences and complications makes it feasible and safe to manage these patients for ambulatory surgery. Therefore, an important role for anesthesiologists is correctly identifying CHD patients who are suitable to undergo these interventions in an outpatient setting.

Practice Points

- General anesthesia (GA) is feasible in CHD patients undergoing ambulatory procedures if the attending anesthesiologist is aware of the underlying pathology and the adverse effects of anesthetic drugs and their impact on PVR and SVR and avoids unnecessary overdosing.
- Patients' suitability for ambulatory procedures remains challenging and needs to be investigated further.
- Locoregional anesthesia is an attractive alternative for GA as it avoids the need for positive pressure ventilation and decreases sympathetic stimulation. However, the use of anticoagulants should always be checked.
- In CHD patients with a left-to-right or right-to-left shunt, providing adequate oxygenation and ventilation and ensuring a stable hemodynamic status through adequately balancing SVR and PVR, are essential. Maintain euvolemia and use vasopressor agents if needed.
- Sedation should be used cautiously as hypoventilation and apnea will affect gas exchange and PVR. Short-acting drugs with minimal effect on breathing and SVR are preferred. Be aware of excessive oxygen supply as it can affect PVR.
- Whatever anesthetic technique is used in these patients; complete ASA monitoring is always indicated.
- Ambulatory centers handling CHD patients should have the capability and a firm postoperative plan to manage CHD patients when unplanned hospital admission is required.

Research agenda

- Future studies should focus on predictive factors related to prolonged or unplanned hospital admission in CHD patients following ambulatory surgery.
- Patients' selection for ambulatory surgery is still challenging and needs further exploration.

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Declaration of competing interest

The authors declare no conflicts of interest.

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References

[1] Liu Y, Chen S, Zühlke L, et al. Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies. Int J Epidemiol 2019;48:455–63. https://doi.org/10.1093/ije/dyz009.
[2] Mulder BJM. Epidemiology of adult congenital heart disease: demographic variations worldwide. Neth Heart J 2012; 20:505–8. https://doi.org/10.1007/s12471-012-0335-1.
[3] Dray EM, Marelli AJ. Adult congenital heart disease: scope of the problem. Cardiol Clin 2015;33:503–12. https://doi.org/10.1016/j.ccl.2015.07.001. vii.
*[4] Baumgartner H, Backer JD, Babu-Narayan SV, et al. ESC Guidelines for the management of adult congenital heart disease. Eur Heart J 2020;42:563–645. https://doi.org/10.1093/eurheartj/ehaa554.
*[5] Hamilton R, Odegard KC, Yuki K. How should we care for patients with congenital heart diseases undergoing surgical procedures in ambulatory settings? Transl Perioper Pain Med 2022;9:416–20.
[6] Dearani JA, Connolly HM, Martinez R, et al. Caring for adults with congenital cardiac disease: successes and challenges for 2007 and beyond. Cardiol Young 2007;17:87–96. https://doi.org/10.1017/s1047951107001199.
[7] Flick RP, Sprung J, Harrison TE, et al. Perioperative cardiac arrests in children between 1988 and 2005 at a tertiary referral center. Anesthesiology 2007;106:226–37. https://doi.org/10.1097/00000542-200702000-00009.
[8] Baum VC, Barton DM, Gutgesell HP. Influence of congenital heart disease on mortality after noncardiac surgery in hospitalized children. Pediatrics 2000;105:332–5. https://doi.org/10.1542/peds.105.2.332.
[9] van der Griend BF, Lister NA, McKenzie IM, et al. Postoperative mortality in children after 101,885 anesthetics at a tertiary pediatric hospital. Anesth Analgesia 2011;112:1440–7. https://doi.org/10.1213/ane.0b013e318213be52.
[10] Maxwell BG, Wong JK, Kin C, et al. Perioperative outcomes of major noncardiac surgery in adults with congenital heart disease. Anesthesiology 2013;119:762–9. https://doi.org/10.1097/aln.0b013e3182a56de3.
[11] Lathouwer CD, Poullier JP. How much ambulatory surgery in the World in 1996–1997 and trends? Ambul Surg 2000;8: 191–210. https://doi.org/10.1016/s0966-6532(00)00065-2.
[12] Jarrett PEM. The international association for ambulatory surgery (IAAS). Ambul Surg 2003;10:113. https://doi.org/10.1016/j.ambsur.2003.10.001.
[13] Leroy R, Camberlin C, Lefèvre M, et al. Variability in elective day-surgery rates between Belgian hospitals - analyses of administrative data explained by surgical experts. Int J Surg 2017;45:118–24. https://doi.org/10.1016/j.ijsu.2017.07.075.
[14] Surgery Ia for A. Day surgery development and practice. 1st ed. Porto, Portugal: Classica Artes Graficas; 2006.
[15] Quemby DJ, Stocker ME. Day surgery development and practice: key factors for a successful pathway. Continuing Educ Anaesth Crit Care Pain 2014;14:256–61. https://doi.org/10.1093/bjaceaccp/mkt066.
*[16] Stout KK, Daniels CJ, Aboulhosn JA, et al. AHA/ACC guideline for the management of adults with congenital heart disease. Circulation 2018;139:1. https://doi.org/10.1161/cir.0000000000000603.
*[17] Faraoni D, Zou X, DiNardo JA, et al. Integration of the intrinsic surgical risk with patient comorbidities and severity of congenital cardiac disease does not improve risk stratification in children undergoing noncardiac surgery. Anesth Analg 2020;131:1083–9. https://doi.org/10.1213/ane.0000000000004906.
*[18] Gerardin JF, Earing MG. Preoperative evaluation of adult congenital heart disease patients for non-cardiac surgery. Curr Cardiol Rep 2018;20:76. https://doi.org/10.1007/s11886-018-1016-5.
[19] Guarracino F, Baldassarri R, Priebe HJ. Revised ESC/ESA Guidelines on non-cardiac surgery: cardiovascular assessment and management. Implications for preoperative clinical evaluation. Minerva Anestesiol 2014;81:226–33.

*[20] Kristensen SD, Knuuti J, Saraste A, et al. ESC/ESA Guidelines on non-cardiac surgery: cardiovascular assessment and managementThe Joint Task Force on non-cardiac surgery: cardiovascular assessment and management of the European Society of Cardiology (ESC) and the European Society of Anaesthesiology (ESA) 2014. 2014.
[21] Foley C, Kendall MC, Apruzzese P, et al. American Society of Anesthesiologists Physical Status Classification as a reliable predictor of postoperative medical complications and mortality following ambulatory surgery: an analysis of 2,089,830 ACS-NSQIP outpatient cases. Bmc Surg 2021;21:253. https://doi.org/10.1186/s12893-021-01256-6.
[22] Twersky R, Abiona M, Thorne A, et al. Admissions following ambulatory surgery: outcome in seven urban hospitals. Ambul Surg 1995;3:141–6. https://doi.org/10.1016/0966-6532(95)00020-5.
[23] Whippey A, Kostandoff G, Paul J, et al. Predictors of unanticipated admission following ambulatory surgery: a retrospective case-control study. Can J Anesth J Can D'anesthésie 2013;60:675–83. https://doi.org/10.1007/s12630-013-9935-5.
*[24] Yuki K, Koutsogiannaki S, Lee S, et al. Unanticipated hospital admission in pediatric patients with congenital heart disease undergoing ambulatory noncardiac surgical procedures. Pediatr Anesth 2018;28:607–11. https://doi.org/10.1111/pan.13388.
[25] Ministeri M, Alonso-Gonzalez R, Swan L, et al. Common long-term complications of adult congenital heart disease: avoid falling in a H.E.A.P. Expert Rev Cardiovasc Ther 2016;14:445–62. https://doi.org/10.1586/14779072.2016.1133294.
[26] Fitzsimmons S, Salmon A. Congenital heart disease in adults. Medicine 2014;42:656–9. https://doi.org/10.1016/j.mpmed.2014.08.006.
[27] Johnson WD, Dawes R, Walker J, et al. Congenital heart disease in adults. Am J Surg 1966;111:830–3. https://doi.org/10.1016/0002-9610(66)90182-6.
[28] Findlow D, Doyle E. Congenital heart disease in adults. Bja Br J Anaesth 1997;78:416–30. https://doi.org/10.1093/bja/78.4.416.
[29] Kecskemeti D, Szegedi M, Temesvari A, et al. Cause of death in adult patients with congenital heart disease: experiences of a tertiary centre. Eur Heart J 2021;42. https://doi.org/10.1093/eurheartj/ehab724.1889.
[30] Verheugt CL, Uiterwaal CSPM, Velde van der ET, et al. Mortality in adult congenital heart disease. Eur Heart J 2010;31:1220–9. https://doi.org/10.1093/eurheartj/ehq032.
[31] Smit-Fun VM, Buhre WF. Heart failure in adult patients with congenital heart disease. Anesthesiol Clin 2019;37:751–68. https://doi.org/10.1016/j.anclin.2019.08.005.
[32] Sarkar M, Niranjan N, Banyal P. Mechanisms of hypoxemia. Lung India Official Organ Indian Chest Soc 2017;34:47–60. https://doi.org/10.4103/0970-2113.197116.
[33] Zabala LM, Guzzetta NA. Cyanotic congenital heart disease (CCHD): focus on hypoxemia, secondary erythrocytosis, and coagulation alterations. Pediatr Anesth 2015;25:981–9. https://doi.org/10.1111/pan.12705.
[34] Rose SS, Shah AA, Hoover DR, et al. Cyanotic congenital heart disease (CCHD) with symptomatic erythrocytosis. J Gen Intern Med 2007;22:1775–7. https://doi.org/10.1007/s11606-007-0356-4.
[35] Cannesson M, Earing MG, Collange V, et al. Anesthesia for noncardiac surgery in adults with congenital heart disease. Anesthesiology 2009;111:432–40. https://doi.org/10.1097/aln.0b013e3181ae51a6.
*[36] Menghraj SJ. Anaesthetic considerations in children with congenital heart disease undergoing non-cardiac surgery. Indian J Anaesth 2012;56:491–5. https://doi.org/10.4103/0019-5049.103969.
[37] Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J 2019;53:1801913. https://doi.org/10.1183/13993003.01913-2018.
[38] Simonneau G, Hoeper MM. The revised definition of pulmonary hypertension: exploring the impact on patient management. Eur Heart J Suppl 2019;21. https://doi.org/10.1093/eurheartj/suz211. K4–8.
[39] Gille J, Seyfarth H-J, Gerlach S, et al. Perioperative anesthesiological management of patients with pulmonary hypertension. Anesthesiol Res Pract 2012;2012:356982. https://doi.org/10.1155/2012/356982.
[40] Khairy P, Hare GFV, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease developed in partnership between the pediatric and congenital electrophysiology society (PACES) and the heart rhythm society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of cardiology (ACC), the American heart association (AHA), the European heart rhythm association (EHRA), the Canadian heart rhythm society (CHRS), and the international society for adult congenital heart disease (ISACHD). Heart Rhythm 2014;11:e102–65. https://doi.org/10.1016/j.hrthm.2014.05.009.
[41] Yumul R, Emdadi A, Moradi N. Anesthesia for noncardiac surgery in children with congenital heart disease. Semin CardioThorac Vasc Anesth 2003;7:153–65. https://doi.org/10.1177/108925320300700204.
[42] Verheugt CL, Uiterwaal CSPM, Velde van der ET, et al. The emerging burden of hospital admissions of adults with congenital heart disease. Heart 2010;96:872. https://doi.org/10.1136/hrt.2009.185595.
[43] Bouchardy J, Therrien J, Pilote L, et al. Atrial arrhythmias in adults with congenital heart disease. Circulation 2009;120:1679–86. https://doi.org/10.1161/circulationaha.109.866319.
[44] Labombarda F, Hamilton R, Shohoudi A, et al. Increasing prevalence of atrial fibrillation and permanent atrial arrhythmias in congenital heart disease. J Am Coll Cardiol 2017;70:857–65. https://doi.org/10.1016/j.jacc.2017.06.034.
[45] Khairy P. Ventricular arrhythmias and sudden cardiac death in adults with congenital heart disease. Heart 2016;102:1703. https://doi.org/10.1136/heartjnl-2015-309069.
[46] Naidu P, Grigg L, Zentner D. Mortality in adults with congenital heart disease. Int J Cardiol 2017;245:125–30. https://doi.org/10.1016/j.ijcard.2017.05.132.
[47] Niwa K, Nakazawa M, Tateno S, et al. Infective endocarditis in congenital heart disease: Japanese national collaboration study. Heart 2005;91:795. https://doi.org/10.1136/hrt.2004.043323.
[48] Thornhill MH, Jones S, Prendergast B, et al. Quantifying infective endocarditis risk in patients with predisposing cardiac conditions. Eur Heart J 2017;39:586–95. https://doi.org/10.1093/eurheartj/ehx655.
[49] Kuijpers JM, Koolbergen DR, Groenink M, et al. Incidence, risk factors, and predictors of infective endocarditis in adult congenital heart disease: focus on the use of prosthetic material. Eur Heart J 2016;38:2048–56. https://doi.org/10.1093/eurheartj/ehw591.

[50] Mandalenakis Z, Rosengren A, Skoglund K, et al. Survivorship in children and young adults with congenital heart disease in Sweden. Jama Intern Med 2016;177:224–30. https://doi.org/10.1001/jamainternmed.2016.7765.
[51] Knirsch W, Nadal D. Infective endocarditis in congenital heart disease. Eur J Pediatr 2011;170:1111. https://doi.org/10.1007/s00431-011-1520-8.
[52] Dodo H, Child JS. Infective endocarditis in congenital heart disease. Cardiol Clin 1996;14:383–92. https://doi.org/10.1016/s0733-8651(05)70291-5.
[53] Snygg-Martin U, Giang KW, Dellborg M, et al. Cumulative incidence of infective endocarditis in patients with congenital heart disease: a nationwide, case-control study over nine decades. Clin Infect Dis 2021;73:ciab478. https://doi.org/10.1093/cid/ciab478.
*[54] Cancer E by the ES of CM and ID (ESCMID) and by the IS of C (ISC) for I and, Members AF, Habib G, et al. Guidelines on the prevention, diagnosis, and treatment of infective endocarditis (new version 2009)The task force on the prevention, diagnosis, and treatment of infective endocarditis of the European society of cardiology (ESC). Eur Heart J 2009;30:2369–413. https://doi.org/10.1093/eurheartj/ehp285.
[55] Li W, Somerville J. Infective endocarditis in the grown-up congenital heart (GUCH) population. Eur Heart J 1998;19:166–73. https://doi.org/10.1053/euhj.1997.0821.
[56] Habib G, Lancellotti P, Antunes MJ, et al. ESC guidelines for the management of infective endocarditisThe task force for the management of infective endocarditis of the European society of cardiology (ESC)endorsed by: European association for cardio-thoracic surgery (EACTS), the European association of nuclear medicine (EANM). Eur Heart J 2015;36:3075–128. https://doi.org/10.1093/eurheartj/ehv319.
[57] Allen U. Infective endocarditis: updated guidelines. Can J Infect Dis Med Microbiol 2010;21:74–7. https://doi.org/10.1155/2010/760276.
[58] Hafner S, Albittar M, Abdel-Kahaar E, et al. Antibiotic prophylaxis of infective endocarditis in oral and maxillofacial surgery: incomplete implementation of guidelines in everyday clinical practice. Int J Oral Maxillofac Surg 2020;49:522–8. https://doi.org/10.1016/j.ijom.2019.09.007.
[59] Greenwood RD, Sommer A, Rosenthal A, et al. Cardiovascular abnormalities in the beckwith-wiedemann syndrome. Am J Dis Child 1977;131:293–4. https://doi.org/10.1001/archpedi.1977.02120160047007.
[60] Pearl W. Congenital heart disease in the Pierre Robin syndrome. Pediatr Cardiol 1982;2:307–9. https://doi.org/10.1007/bf02426978.
[61] Delany DR, Gaydos SS, Romeo DA, et al. Down syndrome and congenital heart disease: perioperative planning and management. J Congenit Cardiol 2021;5:7. https://doi.org/10.1186/s40949-021-00061-3.
[62] Practice guidelines for preoperative fasting and the use of pharmacologic agents to reduce the risk of pulmonary aspiration. Anesthesiology 2017;126:376–93. https://doi.org/10.1097/aln.0000000000001452.
[63] Frykholm P, Disma N, Andersson H, et al. Pre-operative fasting in children: a guideline from the European society of anaesthesiology and intensive care. Eur J Anaesthesiol 2022;39:4–25. https://doi.org/10.1097/eja.0000000000001599.
[64] Gewillig M, Brown SC, Eyskens B, et al. The Fontan circulation: who controls cardiac output? Interact Cardiov Th 2010;10:428–33. https://doi.org/10.1510/icvts.2009.218594.
[65] Bilotta F, Fiorani L, Rosa IL, et al. Cardiovascular effects of intravenous propofol administered at two infusion rates: a transthoracic echocardiographic study. Anaesthesia 2001;56:266–71. https://doi.org/10.1046/j.1365-2044.2001.01717-5.x.
[66] Castor G, Niedermark I, Helms J, et al. Haemodynamic effects of thiopentone, midazolam, etomidate and propofol during induction of anaesthesia. Drug Invest 1991;3:188–94. https://doi.org/10.1007/bf03259563.
[67] Liu H, Kalarickal PL, Tong Y, et al. Pulmonary Hypertension 2013. https://doi.org/10.5772/56056.
[68] Torri G. Inhalation anesthetics: a review. Minerva Anestesiol 2010;76:215–28.
[69] Tanaka S, Tsuchida H, Nakabayashi K, et al. The effects of sevoflurane, isoflurane, halothane, and enflurane on hemodynamic responses during an inhaled induction of anesthesia via a mask in humans. Anesth Analg 1996;82:821–6. https://doi.org/10.1097/00000539-199604000-00025.
[70] Andropoulos DB, Ogletree ML. Anesthesia for congenital heart disease, vols. 30–48; 2008. https://doi.org/10.1002/9780470754986.ch3.
[71] Fineman JR, Wong J, Soifer SJ. Hyperoxia and alkalosis produce pulmonary vasodilation independent of endothelium-derived nitric oxide in newborn lambs. Pediatr Res 1993;33:341–6. https://doi.org/10.1203/00006450-199304000-00007.
[72] Rothe CF, Maass-Moreno R, Flanagan AD. Effects of hypercapnia and hypoxia on the cardiovascular system: vascular capacitance and aortic chemoreceptors. Am J Physiol-Heart C 1990;259:H932–9. https://doi.org/10.1152/ajpheart.1990.259.3.h932.
[73] Kallar SK, Chung F. Practical application of postanesthetic discharge scoring system—PADS. Anesthesiology 1992;77:A12. https://doi.org/10.1097/00000542-199209001-00012.
[74] Chung F. Recovery pattern and home-readiness after ambulatory surgery. Anesth Analg 1995;80:896–902. https://doi.org/10.1097/00000539-199505000-00008.
[75] Zhu Y, Yang S, Zhang R, et al. Using clinical-based discharge criteria to discharge patients after ophthalmic ambulatory surgery under general anesthesia: an observational study. J Perianesth Nurs 2020;35:586–91. https://doi.org/10.1016/j.jopan.2020.04.012. e1.
[76] Marley RA, Swanson J. Patient care after discharge from the ambulatory surgical center. J Perianesth Nurs 2001;16:399–419. https://doi.org/10.1053/jpan.2001.28891.
[77] Veyckemans F, Momeni M. The patient with a history of congenital heart disease who is to undergo ambulatory surgery. Curr Opin Anaesthesiol 2013;26:685–91. https://doi.org/10.1097/aco.0000000000000012.
[78] Lönnqvist P-A. Blocks for pain management in children undergoing ambulatory surgery. Curr Opin Anaesthesiol 2011;24:627–32. https://doi.org/10.1097/aco.0b013e32834a276d.
[79] Horlocker TT, Vandermeuelen E, Kopp SL, et al. Regional anesthesia in the patient receiving antithrombotic or thrombolytic therapy. Region Anesth Pain M 2018;43:263–309. https://doi.org/10.1097/aap.0000000000000763.

[80] Kietaibl S, Ferrandis R, Godier A, et al. Regional anaesthesia in patients on antithrombotic drugs: joint ESAIC/ESRA guidelines. Eur J Anaesthesiol 2022;39:100–32. https://doi.org/10.1097/eja.0000000000001600.
[81] Waldow HC, Westhoff-Bleck M, Widera C, et al. Acquired von Willebrand syndrome in adult patients with congenital heart disease. Int J Cardiol 2014;176:739–45. https://doi.org/10.1016/j.ijcard.2014.07.104.
[82] Rex S, Devroe S. Cardiac disease in pregnancy. Best Pract Res Clin Anaesthesiol 2022;36:191–208. https://doi.org/10.1016/j.bpa.2022.02.005.
[83] Raux O, Rochette A, Morau E, et al. The effects of spread of block and adrenaline on cardiac output after epidural anesthesia in young children: a randomized, double-blind, prospective study. Anesth Analg 2004;98:948–55. https://doi.org/10.1213/01.ane.0000108133.63310.af.
[84] Neal JM, Woodward CM, Harrison TK. The American society of regional anesthesia and pain medicine checklist for managing local anesthetic systemic toxicity. Region Anesth Pain M 2018;43:150–3. https://doi.org/10.1097/aap.0000000000000726.
[85] Suresh S, Ecoffey C, Bosenberg A, et al. The European society of regional anaesthesia and pain therapy/American society of regional anesthesia and pain medicine recommendations on local anesthetics and adjuvants dosage in pediatric regional anesthesia. Region Anesth Pain M 2018;43:211–6. https://doi.org/10.1097/aap.0000000000000702.
[86] Williams GD, Jones TK, Hanson KA, et al. The hemodynamic effects of propofol in children with congenital heart disease. Anesth Analg 1999;89:1411. https://doi.org/10.1213/00000539-199912000-00016.
[87] Öklü E, Bulutcu FS, Yalçın Y, et al. Which anesthetic agent alters the hemodynamic status during pediatric catheterization? comparison of propofol versus ketamine. J Cardiothorac Vasc Anesth 2003;17:686–90. https://doi.org/10.1053/j.jvca.2003.09.009.
[88] Mancuso CE, Tanzi MG, Gabay M. Paradoxical reactions to benzodiazepines: literature review and treatment options. Pharmacotherapy 2004;24:1177–85. https://doi.org/10.1592/phco.24.13.1177.38089.
[89] Forster A, Gardaz J-P, Suter PM, et al. Respiratory depression by midazolam and diazepam. Anesthesiology 1980;53:494–7. https://doi.org/10.1097/00000542-198012000-00010.
[90] Tobias JD, Gupta P, Naguib A, et al. Dexmedetomidine: applications for the pediatric patient with congenital heart disease. Pediatr Cardiol 2011;32:1075–87. https://doi.org/10.1007/s00246-011-0092-8.
[91] Mallory MD, Travers C, Cravero JP, et al. Pediatric sedation/anesthesia for MRI: results from the pediatric sedation research consortium. J Magn Reson Imag 2022. https://doi.org/10.1002/jmri.28392.
[92] Vladinov G, Fermin L, Longini R, et al. Choosing the anesthetic and sedative drugs for supraventricular tachycardia ablations: a focused review. Pacing Clin Electrophysiol 2018;41:1555–63. https://doi.org/10.1111/pace.13511.
[93] Cozzi G, Norbedo S, Barbi E. Intranasal dexmedetomidine for procedural sedation in children, a suitable alternative to chloral hydrate. Pediatr Drugs 2017;19:107–11. https://doi.org/10.1007/s40272-017-0217-5.
*[94] Li BL, Ni J, Huang JX, et al. Intranasal dexmedetomidine for sedation in children undergoing transthoracic echocardiography study—a prospective observational study. Pediatr Anesth 2015;25:891–6. https://doi.org/10.1111/pan.12687.
[95] Tosun Z, Akin A, Guler G, et al. Dexmedetomidine-Ketamine and propofol-ketamine combinations for anesthesia in spontaneously breathing pediatric patients undergoing cardiac catheterization. J Cardiothorac Vasc Anesth 2006;20:515–9. https://doi.org/10.1053/j.jvca.2005.07.018.
[96] Gertler R, Brown HC, Mitchell DH, et al. Dexmedetomidine: a novel sedative-analgesic agent. Bayl Univ Med Cent Proc 2017;14:13–21. https://doi.org/10.1080/08998280.2001.11927725.
[97] Friesen RH, Nichols CS, Twite MD, et al. The hemodynamic response to dexmedetomidine loading dose in children with and without pulmonary hypertension. Anesth Analg 2013;117:953–9. https://doi.org/10.1213/ane.0b013e3182a15aa6.
[98] Manfredi F, Sieker HO. The effect of carbon dioxide on the pulmonary circulation. J Clin Invest 1960;39:295–301. https://doi.org/10.1172/jci104040.
[99] Andrews JS, Hashmi NK. Anesthetic management in adults with congenital heart disease. Curr Cardiol Rep 2022;24:235–46. https://doi.org/10.1007/s11886-022-01639-y.
[100] Coté CJ, Wilson S, Pediatrics AAO, et al. Guidelines for monitoring and management of pediatric patients before, during, and after sedation for diagnostic and therapeutic procedures. Pediatrics 2019;143:e20191000. https://doi.org/10.1542/peds.2019-1000.