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REVIEW
Adult congenital heart disease

Clinical practical advice for anaesthetic management of adults with congenital heart disease undergoing non-cardiac surgery: A clinical consensus statement of the ESC Working Group on Adult Congenital Heart Disease, the European Society of Anaesthesiology and Intensive Care, the European Association of Cardiothoracic Anaesthesiology and Intensive Care and the International Society of Adult Congenital Heart Disease

Laura Dos-Subirà [1,2,3,*], Koen Rijs [4], Montserrat Ribas-Ball [5], Margarita Brida [6,7,8], Paul Clift [9], Gerhard Diller [6,10], Michael Gatzoulis [6,7], Jane Heggie [11], Dan Longrois [12], Sascha Meier [13], Raquel Prieto-Arévalo [2,14,15], Marco Ranucci [16], Steffen Rex [17], Francisco J. Ruperti [2,3,18,19,20], Ehrenfried Schindler [21], Vera Von Dossow [22], and Jolien Roos-Hesselink [3,23]

[1]Department of Cardiology, Adult Congenital Heart Disease Unit, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Passeig de la Vall d'Hebron, 119-129, Barcelona 08035, Spain; [2]Centro de Investigación Biomédica en Red de Enfermedades Cardiovasculares (CIBERCV), Instituto de Salud Carlos III (ISCIII), Madrid, Spain; [3]European Reference Network for Rare, Low-Prevalence, or Complex Diseases of the Heart (ERN GUARD-Heart), Amsterdam, The Netherlands; [4]Department of Anesthesiology, Erasmus University MC, Rotterdam, The Netherlands; [5]Department of Anesthesiology, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Barcelona, Spain; [6]Adult Congenital Heart Disease Centre, Royal Brompton & Harefield, Guy's St Thomas's NHS Foundation Trust, London, UK; [7]National Heart and Lung Institute, Imperial College, London, UK; [8]Medical Faculty, University of Rijeka, Rijeka, Croatia; [9]Department of Adult Congenital Heart Disease, Queen Elizabeth Hospital, University Hospital, Birmingham, UK; [10]Department of Cardiology III: Congenital Heart Malformations (ACHD) and Valve Diseases, University Hospital Münster, Münster, Germany; [11]Department of Anesthesia and Pain Medicine, Peter Munk Cardiac Centre, Toronto General Hospital, Toronto, Ontario, Canada; [12]Bichat-Claude Bernard and Louis Mourier Hospitals, Assistance Publique-Hôpitaux de Paris, Paris Cité University, Paris, France; [13]Department of Anesthesiology, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands; [14]Department of Cardiology, Hospital General Universitario Gregorio Marañón, Facultad de Medicina, Universidad Complutense de Madrid, Instituto de Investigación Sanitaria Gregorio Marañón, Madrid 28007, Spain; [15]Cardiovascular Diseases Department, Health Research Institute Gregorio Marañón, Madrid 28009, Spain; [16]Department of Cardiothoracic, Vascular Anesthesia and Intensive Care, IRCCS Policlinico San Donato, San Donato Milanese, Italy; [17]Department of Cardiovascular Sciences, Unit Anesthesiology and Algology, Biomedical Sciences Group, University of Leuven (KU Leuven), Leuven, Belgium; [18]Division of Cardiology, University Hospital of Basel, Basel, Switzerland; [19]Department of Cardiology, Adult Congenital Heart Disease Unit, Hospital Universitario Virgen del Rocio, Instituto de BioMedicina de Sevilla (IBIS), Sevilla, Spain; [20]Division of Cardiology, Cardiovascular Research Institute Basel, University Hospital Basel, Basel, Switzerland; [21]Department of Anesthesiology and Intensive Care Medicine, University Hospital Bonn, University of Bonn, Bonn, Germany; [22]Institute of Anaesthesiology and Pain Therapy, Heart and Diabetes Centre North Rhine Westphalia, University Hospital of Ruhr-University Bochum, Bad Oeynhausen, Germany; and [23]Department of Cardiology, Cardiovascular Institute, Thorax Center, Erasmus MC, Rotterdam, The Netherlands

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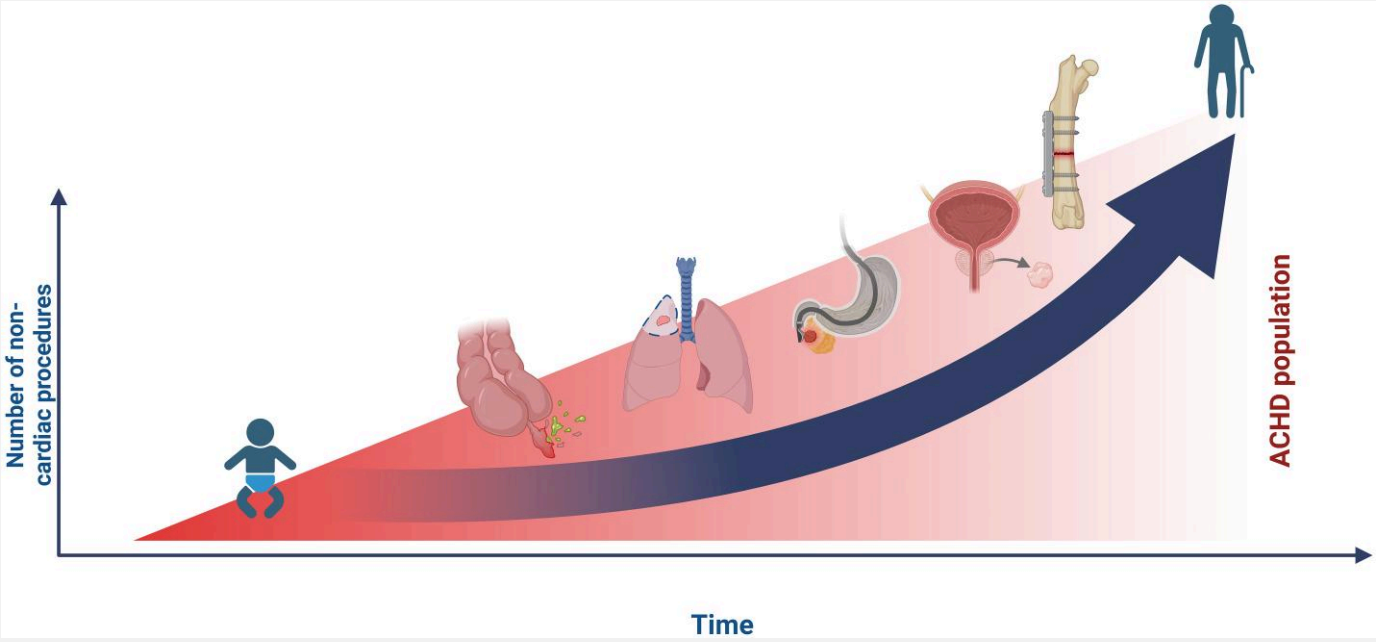
* Corresponding author. Tel: 93 489 31 52, Fax: 93 489 30 39, Email: laura.dos@vallhebron.cat

Abstract

Adult congenital heart disease (ACHD) patients often require non-cardiac surgery (NCS) and the need of such interventions is growing as the population ages. The intricacies inherent to their distinct physiology pose ACHD patients at a higher risk of perioperative complications, perioperative mortality, longer length of stay, and higher hospital charges compared with the general population. While 2020 ESC guidelines comprehensively address most aspects of ACHD care, they do not cover this specific area. Furthermore, although risk assessment for non-cardiac surgery is included in the 2022 ESC Guidelines on Cardiovascular Assessment and Management of Patients Undergoing Non-Cardiac Surgery, the unique pathophysiology of ACHD patients necessitates tailored considerations. The aim of this clinical consensus statement is to address the specific situations and needs of ACHD patients undergoing NCS from a multidisciplinary and practical point of view and to describe the organization of care for this complex group of patients. A group of experienced professionals from 4 cardiology and anaesthesiology scientific societies [the ESC Working Group on ACHD, the European Society of Anaesthesiology and Intensive Care (ESAIC), the European Association of Cardiothoracic Anaesthesiology and Intensive Care (EACTAIC), and the International Society of Adult Congenital Heart Disease (ISACHD)] have evaluated the available scientific data and reached expert consensus in areas of gaps in evidence.

Graphical Abstract



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Keywords Adult congenital heart disease • Anaesthesia • Non-cardiac surgery • Perioperative management

Introduction

As a result of better diagnostics, improvements in surgical and interventional treatment and more optimal perioperative care, over 95% of children born with a congenital heart defect (CHD) now reach adult age. Not only are the numbers of adult congenital heart disease (ACHD) patients increasing, but their complexity has also become more severe. Survival to adulthood following Fontan palliation of all univentricular heart conditions is now expected. Despite the spectacular improvements in survival, concerns remain regarding the high incidence of morbidity and reduced life expectancy. ACHD patients often require cardiac (re)interventions due to residual lesions but also often need non-cardiac surgery (NCS) or interventions for other morbidities or, in the case of women, face gestation with possible need of caesarean section. All these procedures involve anaesthetic management which may pose challenges in the setting of CHD, particularly when unusual physiology coexists. ACHD patients are at increased risk of perioperative complications, perioperative mortality, longer length of stay, and higher hospital charges compared with the general population.[1,2] The optimal management has been scarcely investigated and rarely described, but an inadequate preoperative assessment accounts for up to 40% of the adverse events in this scenario.[3] Despite these high risks, up to 74% of NCS in ACHD patients occur at non-ACHD centers,[4] and complexity-based referral to a tertiary centre may be needed.[5] In the

European Society of Cardiology guidelines regarding the management of ACHD[6] most aspects of care have been well addressed. However, the topic of anaesthetic management in ACHD patients, both for cardiac and for non-cardiac interventions, is not discussed. On the other hand, although both the procedure and the patient's risk assessment can be evaluated following the 2022 ESC Guidelines on cardiovascular assessment and management of patients undergoing NCS,[7] the unique pathophysiology of certain CHD merit specific considerations. The aim of this clinical consensus statement is to address the specific situations and needs from a multidisciplinary point of view, to optimize anaesthetic management and describe the organization of care for this complex group of patients.

Anaesthetic basic concepts

The anaesthetic act allows the performance of invasive procedures by avoiding pain, controlling anxiety and providing muscle relaxation if needed, while maintaining physiologic balance. Most important is an adequate preoperative assessment in which the type of anaesthetic agent/technique and approach are oriented based on the type of procedure and the risk evaluation of the individual patient. It continues with the perioperative management and finalizes with the post-operative care.

An overview of the different available anaesthetic approaches[8] is described here:

1. *General anaesthesia.* The most appropriate technique for major or prolonged surgeries, for patients not able to tolerate the procedure while awake, for procedures requiring complete immobility and for those with limited airway access.

   It can be divided in three phases:

   (i) Induction: The combination of a hypnotic agent (e.g. propofol, etomidate, ketamine, midazolam) and one or more analgesics (e.g. opioid, lidocaine) is the usual practice, with or without a neuromuscular blocking agent (NMBA). Patients require airway management and mechanical ventilation for which an endotracheal tube or a supraglottic airway device, typically a laryngeal mask, are used.
   (ii) Maintenance: Combinations that include one or more hypnotics (e.g. total intravenous (IV) anaesthesia with propofol or inhalational anaesthesia with sevoflurane) combined with opioids (short-acting remifentanil), with or without a NMBA (e.g. vecuronium, rocuronium, atracurium, cisatracurium) are often employed.
   (iii) Emergence: After discontinuation of anaesthetic and adjuvant agents and tapering/reversing NMBA effects, the patient recovers consciousness, spontaneous breathing and can be safely extubated.

2. *Neuraxial anaesthesia (NA):* Spinal anaesthesia is achieved by a single injection of a local anaesthetic into the subarachnoidal space, whereas epidural anaesthesia usually requires continuous infusion in the epidural space. A CSE uses both approaches. NA produces blockade of motor and sensory nerves but also blocks sympathetic nerves and compensatory reflexes and may therefore cause hypotension (reduction of SVR, peripheral blood pooling and decrease of venous return) and bradycardia. NA is commonly used in lower abdominal and lower extremities surgery and is the procedure of choice for labour analgesia and caesarean section in most congenital heart disease patients.
3. *Peripheral locoregional anaesthesia:* Consists of a single injection or continuous infusion of a local anaesthetic (e.g. lidocaine, mepivacaine, ropivacaine) adjacent to the nerve or plexus (peripheral nerve block [PNB]). PNB is commonly used for limb surgery. Fascial plane blocks have been suggested as a complement to other anaesthetic approaches in abdominal o thoracic surgery to enhance post-operative analgesia, particularly when using long-acting drugs (bupivacaine or ropivacaine).
4. *Intravenous regional anaesthesia:* Is a rarely used alternative to PNB for short procedures, generally on the hand and forearm.

Monitored anaesthesia care: Includes administration of sedative (e.g. propofol, dexmedetomidine), anxiolytic and/or analgesic medications (remifentanil, sufentanil, alfentanil) and generally does not involve complete loss of consciousness of the patient.

Preanaesthetic assessment

The ACHD population is heterogeneous, and even patients with similar anatomy can show very different pathophysiology. An atrial septal defect, an anatomically simple CHD, requires a very different management when pulmonary hypertension coexists. Thus, no single anaesthetic technique fits every CHD lesion, or surgical procedure.[9]

Therefore, understanding past cardiac surgeries, percutaneous interventions, medical history, and current cardiac anatomy and physiology is crucial and provides critical insights not only for risk assessment but also for preoperative optimization to reduce adverse events related to the NCS.[6] To fulfil this assessment, current data on symptoms, exercise capacity, oxygen saturation, laboratory values, ECG, echocardiography and medication are needed. When available, additional data from complementary imaging techniques (i.e. computed tomography, magnetic resonance) or invasive hemodynamic assessment are helpful.

Table 1 provides a list of aspects to be considered in the setting of ACHD preoperative evaluation for NCS.

Procedural-related risk

A surgical risk classification according to type of surgery/intervention is depicted in *Table 2*. Of note, laparoscopic procedures could be considered as high-risk procedures among high-risk ACHD patients, as pneumoperitoneum and Trendelenburg position may significantly impair the haemodynamics and blood viscosity of some of these patients (see also Laparoscopy).[14,15] Gastrointestinal endoscopies performed as part of national screening programs represent a significant source of anesthetic activity in ACHD patients and are considered low-risk procedures (not included in *Table 2*).

Patient-related risk

Based on the underlying CHD and after specific risk assessment, patients can be classified into low, intermediate or high-risk subgroups[7] (*Table 3*). As the ACHD population ages, acquired cardiovascular disease (CVD) is becoming more relevant[16] and this risk should also be evaluated based on age, cardiovascular risk factors (such as smoking, hypertension, diabetes, hypercholesterolemia, and family history), established CVD, and comorbidities.[7]

Overall risk classification

The scenarios resulting from the combination of both the procedural- and patient-related risks can be divided into three risk categories with different requirements (*Figure 1*). Patients in the High-risk Scenario (high-risk patients undergoing intermediate- to high-risk procedures and intermediate-risk patients undergoing high-risk procedures) require consultation with a specialized ACHD unit and with a cardiac anaesthesiologist with ACHD expertise and should be managed in centres with a multidisciplinary ACHD program. Ideally, this tertiary referral center should maintain a network of anesthesiologists with a specific interest in ACHD, integrated across various surgical specialties (e.g. obstetrics, orthopedics), and coordinated by the cardiac anaesthesiologist with expertise in ACHD.

The preoperative advice here is reserved for time-sensitive and elective surgery. Immediate and urgent surgeries should proceed

Table 1 ACHD preoperative evaluation for non-cardiac surgery

Demographics	• Age, BMI, smoking status
Clinical examination	• Including blood pressure, pulse and SpO_2
Type of CHD	• Native cardiac lesion
Surgical/medical CHD-related history	• History of interventions/operation for palliations, reparations, and re-interventions • History heart failure, pulmonary hypertension
Current cardiac physiology	• Identifying cardiac chamber for pulmonary and systemic venous return • Defining bi-ventricular vs. univentricular physiology • Identifying the morphology of the subpulmonary and subaortic ventricle (left ventricle vs. right ventricle) • Identifying intra- and extracardiac shunts • Identifying systemic-to-pulmonary venous and arterial collaterals
Current cardiac status	• Defining (bi-)ventricular function • Identifying residual lesions (such as residual shunt, valve regurgitation, valve stenosis) • Current heart rhythm • Identifying and classifying pulmonary hypertension as defined in current guidelines[10]
Current physical status	• NYHA functional class • ASA physical status[11] • CPET if available
Arrhythmia/cardiac device status (pacemaker, CRT-system, AICD)	• History of previous arrhythmias, underlying rhythm, device function, pacemaker-dependency, battery status, battery location.
Vascular accesses	• Vascular accesses should be documented to facilitate location of the arterial or venous lines and gas exchange (be prepared for scarring or thrombosis from previous cannulation sites or limitations posed by previous shunts, arteriovenous or veno-venous fistulas)
Medication	• Assessment of prescribed and over-the-counter medication (with special attention to antiarrhythmic drugs, heart failure drugs, anticoagulants, and antiplatelet agents)
Assessing risk of bleeding	• Specially in specific situations, such as in chronic cyanotic patients, in which bleeding risk is increased due to multiple collaterals, platelet dysfunction, and alterations in the coagulation cascade[12] • Stopping/bridging anticoagulation drugs
Comorbidities	• Such as hypertension, diabetes, endocrine disorders or other organ damage (CKD, lung disease, liver disease)
Laboratory tests	• For the assessment of anaemia, impaired renal/liver function, and coagulation status • INR assessment in cyanotic patients should be performed using citrate tubes and adjusted to the patient's haematocrit
Need for endocarditis prophylaxis	• As defined in the most recent version of the guidelines[13]
Assessment of genetic syndromes	• As they may interfere with anaesthetic management (i.e. airway abnormalities, cognitive impairment, etc.)

ACHD, adult congenital heart disease; AICD, automatic implantable cardiac defibrillator; ASA, American Society of Anesthesiology; BMI, body mass index; CKD, chronic kidney disease; CPET, Cardiopulmonary exercise test; CRT, cardiac resynchronization therapy; NYHA, New York Heart Association; SpO_2, peripheral oxygen saturation.

without unnecessary delay to save life, limb, or organ function and may need to be addressed in community centres. Clear pathways should be stablished for referral to tertiary centres if required (High-risk Scenario). On the other hand, a patient's desire for timely care for NCS close to their homes with minimal disruption to their personal life must be balanced against the need for adequate optimization of the patient and careful perioperative planning by the care team. Preoperative discussions between the regional hospital and the tertiary center are essential to determine the most appropriate location for the surgery.

Finally, patient empowerment and active involvement in self-care are essential. Individuals classified as categories 2B–C and 3A–C should be made aware of the critical role of the ACHD team in the planning and management of non-cardiac surgical procedures. They should be encouraged to contact their ACHD specialist well in advance to ensure appropriate coordination and care.

General considerations

Scheduling high risk patients as the first case of the day minimizes fasting time and therefore reduces the risk of dehydration in preload dependent conditions, such as Fontan patients. It also ensures the presence of adequate personnel and consultant expertise to assist if complications occur during or immediately after surgery. Endocarditis risk assessment and prophylaxis should be performed according to current ESC Guidelines for the management of endocarditis.[13] Anaesthetic management may be needed in mentally impaired patients to tolerate procedures that would not require anaesthesia in the general population

Table 2 Procedural-related risk classification according to type of surgery or intervention

Low surgical risk (Group A)	Intermediate surgical risk (Group B)	High surgical risk (Group C)
• Breast • Dental • Endocrine: thyroid • Eye • Gynaecological: minor • Orthopaedic minor (meniscectomy) • Reconstructive • Superficial surgery • Urological minor: (transurethral resection of the prostate) • VATS minor lung resection	• Carotid asymptomatic (CEA or CAS) • Carotid symptomatic (CEA) • Endovascular aortic aneurysm repair • Head or neck surgery • Intraperitoneal: splenectomy, hiatal hernia repair, cholecystectomy • Intrathoracic: non-major • Neurological or orthopaedic: major (hip and spine surgery) • Peripheral arterial angioplasty • Renal transplants • Urological or gynaecological: major • Caesarean section	• Adrenal resection • Aortic and major vascular surgery • Carotid symptomatic (CAS) • Duodenal-pancreatic surgery • Liver resection, bile duct surgery • Oesophagectomy • Open lower limb revascularization for acute limb ischaemia or amputation • Pneumonectomy (VATS or open surgery) • Pulmonary or liver transplant • Repair of perforated bowel • Total cystectomy

Adapted from Halvorsen S. *et al.*[7]
CAS, carotid artery stenting; CEA, carotid endarterectomy; CV, cardiovascular; MI, myocardial infarction; VATS, video-assisted thoracic surgery.

Table 3 Patient-related risk stratification for non-cardiac surgery in adult CHD

Low risk (Group 1)	Patients with small, uncorrected defects, and no need for medication or any other treatment. Patients with successfully corrected CHD with no symptoms, no relevant residua, and no need for medication.
Intermediate risk (Group 2)	Patients with corrected or uncorrected conditions with residual hemodynamic abnormality (i.e. severe AV valve regurgitation) or arrhythmias, with or without medication.
High risk (Group 3)	Patients with uncorrected cyanotic heart disease, pulmonary hypertension, other complex CHD, ventricular dysfunction requiring medication, life-threatening arrhythmias and patients listed for heart transplantation.

Adapted from Halvorsen S. *et al.*[7]
AV, atrioventricular; CHD, congenital heart disease.

(i.e. cataract surgery). Prophylactic phlebotomy to maintain haematocrit within an arbitrary predetermined level is not advised.[17]

Patients with cardiac devices—such as pacemakers or defibrillators—and those at high risk for post-operative arrhythmia should undergo thorough evaluation involving electrophysiologists with ACHD expertise. Beyond standard considerations like reprogramming pacemakers to asynchronous modes to prevent electromagnetic interference in pacemaker-dependent patients, or disabling defibrillator therapies to avoid inappropriate shocks, certain CHD introduce additional complexities. Anomalous vascular anatomy—such as occluded vessels from prior catheterizations, heterotaxy syndromes, or surgically altered circulatory pathways (e.g. Glenn or Fontan procedures)—can complicate vascular access and may limit the feasibility of transvenous pacing if required.

Preanaesthetic optimization

The unique cardiac physiology of ACHD impacts other organ systems that may need adjustment or optimization in anticipation of the surgery and the degree of dysfunction should be ascertained.

More than half of the ACHD patients will have some restrictive lung function,[18,19] having a major impact especially on patients with Fontan circulation. Inspiratory muscle training improves inspiratory muscle strength, ventilatory efficiency of exercise and resting cardiac output in young Fontan patients.[20]

Liver function may be impaired due to several factors: increased central venous pressure and consequent liver congestion, an increased incidence of hepatitis C related to previous transfusions and the use of medication, such as amiodarone.[21,22] Renal dysfunction in this population is also common and multifactorial: cyanosis, increased central venous pressure and reduced cardiac output.[23,24]

Anaemia affects up to 30% of non-cyanotic ACHD patients. The most common cause is iron deficiency, aggravated by malabsorption in the context of increased right-side pressures and congestion, renal dysfunction, and blood loss due to antiplatelet therapy/anticoagulation.[25] The risk of thrombosis is elevated in ACHD caused by blood stasis, presence of prosthetic material and frequent vascular access for invasive procedures.[26,27]

It is of upmost importance that the treating cardiologist optimizes the patient's medication prior to NCS. Maximum titration of pulmonary vasodilator drugs and heart failure medication is advisable. However, those treated with systemic vasodilators may develop and require treatment for hypotension, especially during induction. For patients under anticoagulation, thromboembolic and bleeding risks should

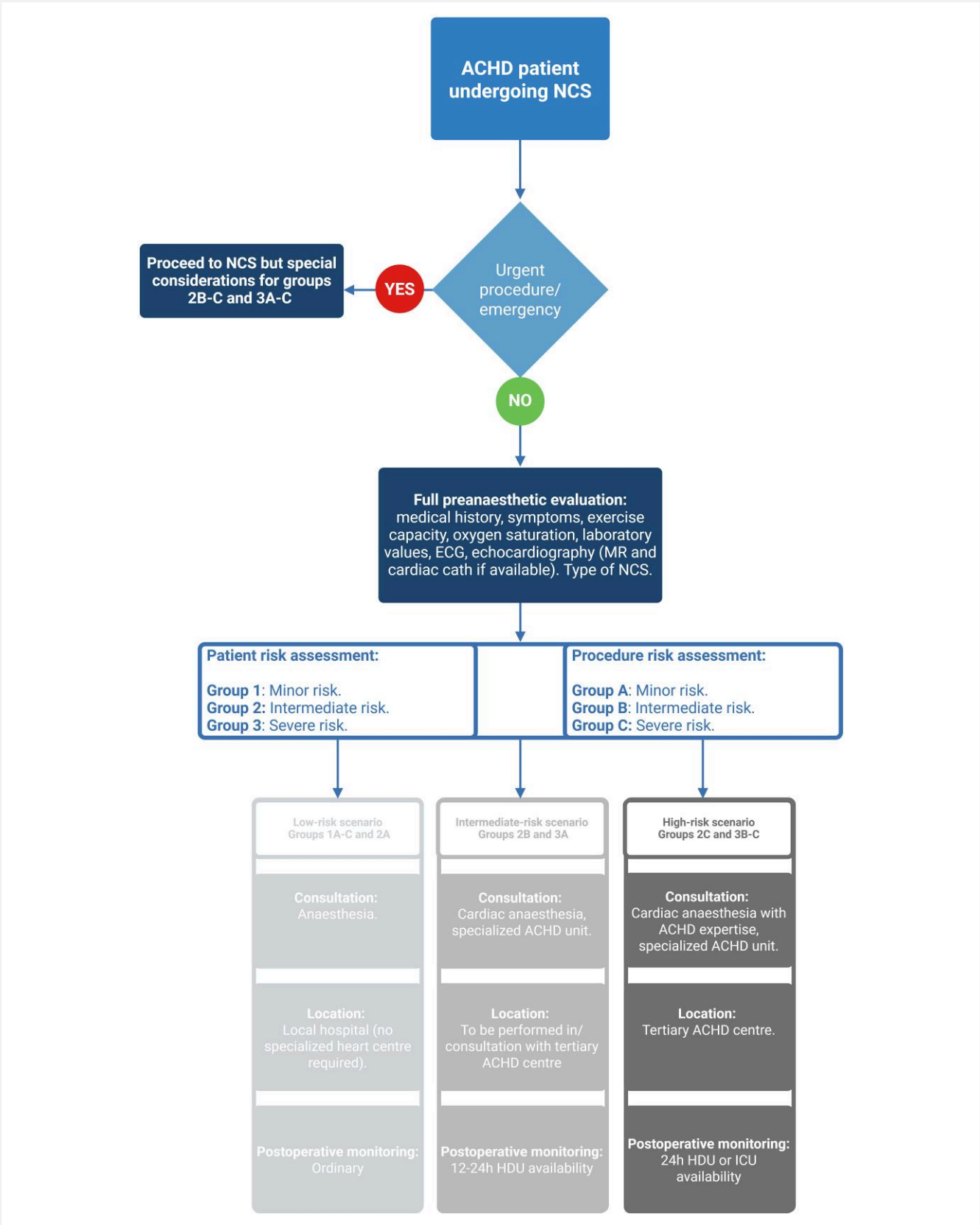

Figure 1 Decision making flowchart for ACHD patients undergoing NCS. Classification into 3 risk Scenarios combining patient-related and intervention-related risks. Created in BioRender. Dos Subirà, L. (2025), https://BioRender.com/ekg0mg1

be balanced, and a plan for treatment (dis)continuation should be made prior to NCS, if needed in multidisciplinary setting.[7]

Multimodal prehabilitation programs that include exercise, nutritional, and cognitive-behavioural interventions have been found to be beneficial in other groups of patients[28] and are promising in ACHD patients undergoing NCS although there is no evidence published to support this.

Periprocedural management

The general principles of periprocedural management are described in this section and thereafter we will discuss specific subgroups in more detail.

Choice of anaesthetic agent

Anaesthetic agents have varying effects on left and right ventricular afterload, contractility, and preload, making selection crucial based on the patient's anatomy, physiology, presence/direction of a shunt, and hemodynamic status.[29,30] *Table 4* summarizes the impact of anaesthesia on cardiovascular variables and their consequences for patients with ACHD. There is limited data on the preferable anaesthetic technique for NCS in ACHD patients[9,31] and there is no clear evidence favouring one anaesthetic agent over another in induction or maintaining anaesthesia, highlighting the need for more research in this field.

Propofol and sevoflurane belong to the most used agents. However, it should be kept in mind that propofol can cause dose-dependent hypotension due to reduction of systemic vascular resistance (SVR), bradycardia and decrease of myocardial contractility. Volatile inhalation agents cause dose-dependent reduction of SVR and tachycardia.

In cyanotic patients, induction with ketamine in combination with a gamma-aminobutyric acid (GABA)-$_A$- receptor-agonist (e.g. etomidate, midazolam, propofol) may be advantageous in avoiding increases in right-to-left shunt. Additionally, ketamine's sympathomimetic effects can be beneficial in patients with severely compromised ventricular function, obstructive lesions, or severe pulmonary hypertension.[32]

Regional anaesthesia offers an alternative, allowing for the avoidance of mechanical ventilation and providing excellent pain control. Central neuraxial anaesthesia (NA), either epidural or combined spinal-epidural (CSE), has been successfully used in virtually every malformation but requires extra precautions in certain populations (see Special settings and Special populations). Employing low-dose and incremental techniques allows to minimize hemodynamic instability.[33]

Hemodynamic management

Hemodynamic management must be tailored to the pathophysiology of the underlying malformation, particularly in patients with a shunt. In left to right shunts, factors increasing SVR (i.e. light anaesthesia, pain, sympathetic hyperstimulation, vasopressors, hypothermia, hypovolemia) and/or decreasing pulmonary vascular resistance (PVR) (i.e. hypocarbia, hyperoxia, alkalosis, pulmonary vasodilators) should be avoided. In right to left shunts, factors decreasing SVR and/or increasing PVR should be avoided,[30] as depicted in *Figure 2* and addressed in Unrepaired cyanotic congenital heart diseases and Eisenmenger physiology.

In pacemaker-dependent patients, device reprogramming to higher heart rates may be beneficial to support hemodynamics under anesthesia.

Ventilation

Ventilatory management aims to maintain adequate oxygenation and ventilation while minimizing its impact on PVR. Lung-protective ventilation strategies should be employed to minimize alveolar overinflation and reduce right ventricular afterload. Low levels of positive end-expiratory pressure may be beneficial in optimizing oxygenation and preventing atelectasis while maintaining low PVR.[34]

Monitoring

Monitoring during the procedure adheres to standard anaesthesia guidelines,[35,36] with the addition of invasive monitoring (arterial and central venous catheterization) as necessary based on the invasiveness of the procedure and type/severity of the underlying disease. Intravascular catheters also offer the opportunity for regular arterial and/or central venous blood gas analyses. This should be balanced with the risk of vascular access related complications favouring non-invasive hemodynamic measurements whenever possible. In critical patients, intra-arterial catheterization for continuous blood pressure monitoring should be performed before induction of general anaesthesia (GA) or needle placement for NA. Electroencephalogram-based depth of anaesthesia monitoring allows for anaesthetic dose titration. Transoesophageal echocardiography (TOE) monitoring may be useful during major surgeries.

Post-operative care

Post-operative care is crucial since up to 50% of adverse events occur in this phase.[3] Post-operative pain and anxiety should be treated to minimize sympathetic stimulation to avoid arrhythmias and hypertension, ensure adequate ventilation and avoid increase in PVR. A regional technique may be appropriate to control pain after certain procedures and intravenous (IV) patient-controlled analgesia (PCA) may be useful to treat moderate-intense pain. Post-operative availability of intensive care unit (ICU) or high dependency unit (HDU) facilities is required for patients with high-risk ACHD, particularly after high-risk procedures (High-risk Scenario, *Figure 1*).

Extubation is a challenging phase with high risk of hemodynamic instability and arrhythmias. Hypoxemia, hypercarbia, or acidosis can worsen pulmonary vascular resistance and compromise cardiac output, particularly in vulnerable populations such as Eisenmenger syndrome or Fontan physiology. Additionally, ACHD patients may have restrictive lung disease due to previous surgeries or thoracic abnormalities, airway anomalies, or ciliary dysmotility and are at elevated risk for extubation failure and need for reintubation. Close monitoring of high-risk patients is mandatory (see Special populations).

When feasible, early extubation strategies (EES) are an attractive option. They reduce the need for vasoactive drugs and fluid resuscitation while achieving a low complication rate and a shorter intensive care stay was found in ACHD patients undergoing cardiac surgery.[37] EES require an anaesthesia technique with short acting agents and a well-structured strategy for post-operative pain management. On the other hand, patient selection is crucial. If extubation is premature, particularly in obese patients or those with obstructive sleep apnoea, hypercarbia can induce an increase of PVR and lead to hemodynamic compromise.[38]

A structured hand-over to the admitting ward or ICU is mandatory because of the often-complex medical history. Because memories are often incorrect or incomplete it is advised to work with tools like the SBAR concept (S: 'situation', B: 'background', A: 'assessment', R: 'recommendation') for standardization of the patient handover.[39,40] SBAR helps to ensure clear and structured communication, which is particularly important in stressful or time-critical situations. When using the SBAR method, specific and relevant information should be conveyed to ensure clear and concise communication. It is not necessary to provide all available information; only key aspects should be included in each section of SBAR and should be adapted to the respective circumstances and needs (*Table 5*).[41]

Special settings

Gestation, labour, and delivery

Pregnancy induces profound physiological changes, particularly in the cardiovascular system, to accommodate the growing needs of both the mother and the fetus.[42] Note that most heart failure medications

Table 4 Impact of anesthesia on cardiovascular variables and consequences for patients with ACHD

Item	Effect	Typical peri-interventional triggers	Consequences
LV Afterload	↓ SVR	• Inhalational anaesthetics • Propofol • Neuraxial anaesthesia • Inadequately deep anaesthesia inflammation/ sepsis/anaphylaxis co-medication	• Poorly tolerated in obstructive lesions • ↑ Right-to-left shunting • ↓ Left-to-right shunting • Detrimental in patients with PH and RV dysfunction
	↑ SVR	• Endotracheal intubation without adequate analgesia • Insufficient depth of anaesthesia/analgesia • Pain/Stress	• Poorly tolerated in patients with impaired systolic function
RV Afterload	↑ PVR	• Hypercapnia • Hypoxia • Acidosis • Hypothermia • Positive pressure ventilation; coughing/pushing	• ↑ Right-to-left shunting • Can be (to some extent) beneficial to reduce left-to-right shunting • Detrimental in patients with PH and RV dysfunction
	↓ PVR	• Hypocapnia • Hyperoxia • Alkalosis	• ↑ Left-to-right shunting • Can be (to some extent) beneficial to reduce right-to-left shunting • Can improve lung flow and RV performance
Preload	Decrease	• Neuraxial anaesthesia • High dose of general anaesthetics; inflammation/sepsis/anaphylaxis • Hypovolemia (diuretics) • Haemorrhage • Fasting	• Poorly tolerated in obstructive lesions (e.g. aortic stenosis) • Hypertrophic RV or LV needs adequate preload to preserve function
	Increase	• Inadequate fluid therapy	• Poorly tolerated in patients with impaired systolic function
Heart rate	Decrease	• Inadequately high rostral spread of neuraxial anaesthesia	• Poorly tolerated in patients with impaired systolic function and/or with valvular regurgitation
	Increase	• Endotracheal intubation without adequate analgesia • Insufficient depth of anaesthesia/analgesia • Stress	• Poorly tolerated in obstructive lesions (e.g. aortic stenosis) • Dangerous increase in MVO_2 in hypertrophied RV or LV
Contractility	Decrease	• Inadequately high rostral spread of neuraxial anaesthesia • Most general anaesthetics (except ketamine)	• Poorly tolerated in general, particularly by patients with obstructive lesions
	Increase	• Stress • Pain	• Causes increase in MVO_2 → poorly tolerated by patients prone to myocardial ischemia
Positive pressure ventilation	↑ RV afterload ↓ RV preload ↓ LV afterload ↓ LV preload		• Poorly tolerated in Fontan patients and patients with RV dysfunction • Relatively well tolerated in patients with LV failure

ACHD, adult congenital heart disease; LV, left ventricular; MVO_2, myocardial oxygen consumption; PVR, pulmonary vascular resistance; RV, right ventricular; SVR, systemic vascular resistance.

are contraindicated in pregnancy, exposing the pregnant women to an additional risk.[43] This places unique challenges on the anaesthetic management of ACHD patients. The modified World Health Organization (mWHO) classification provides a valuable framework for assessing these risks, distinguishing between no, small, increased, and extremely increased risks of maternal mortality or severe morbidity.[44] The anaesthetic management of ACHD patients during gestation, particularly those at higher risk (mWHO III and IV) requires a multidisciplinary approach and a meticulous perioperative planning to be discussed and prepared well in advance (mode of delivery, need for ICU stay, anaesthetic details, length of stay after delivery). A detailed delivery plan must be available at the beginning of the third trimester and patients with

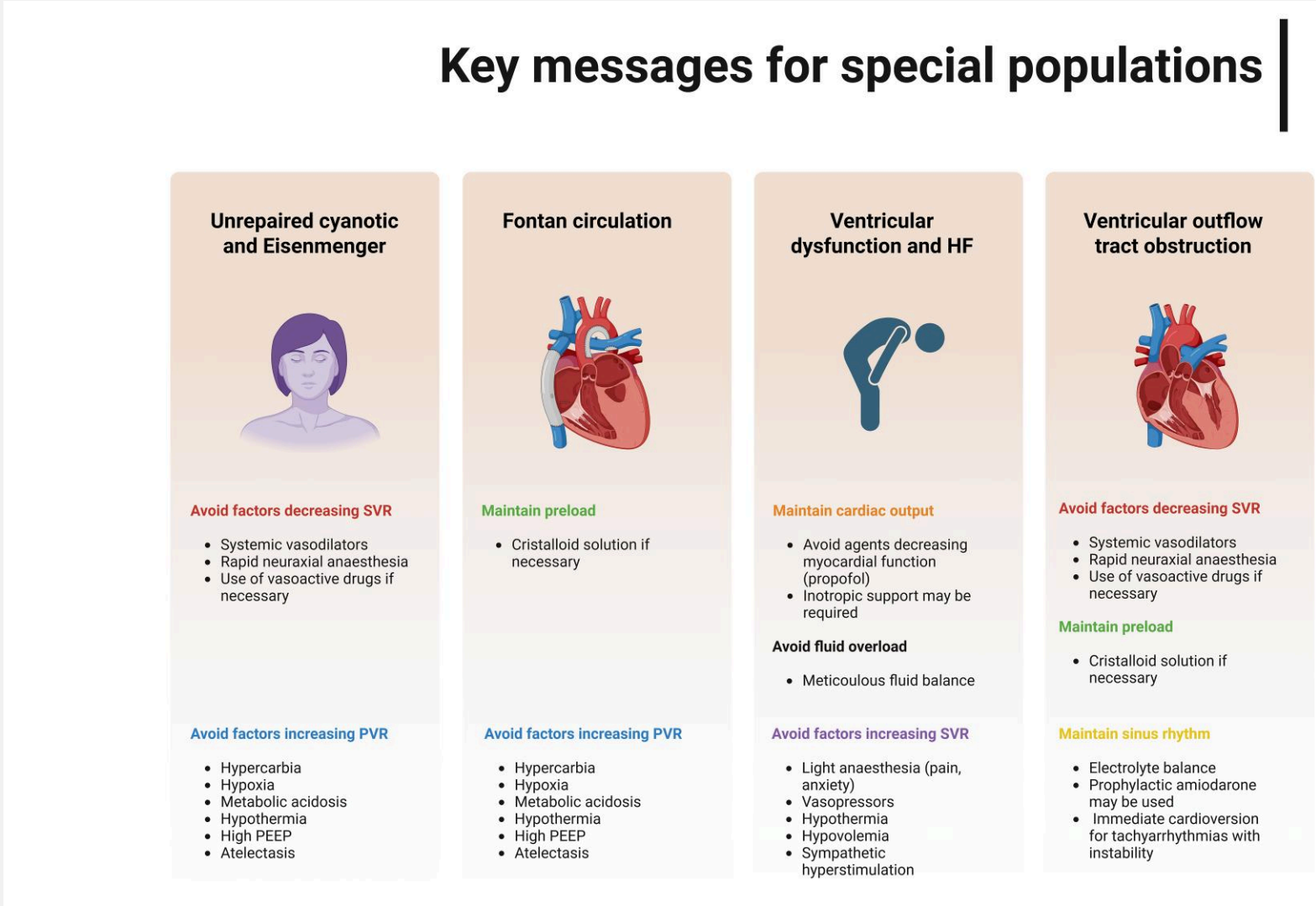

Figure 2 Key messages for special populations. Created in BioRender. Dos Subirà, L. (2025), https://BioRender.com/ekg0mg1

Table 5 Example for a structured handover protocol from the operation room to the intensive care unit or recovery room following the SBAR principle

Situation	Patient ID
	Main diagnosis
	Operation/reason for ICU admission
	Type of anaesthesia
	ASA classification[11]
	Airway management
Background	Allergies/caution
	Preoperative diagnostics
	Preoperative drug therapy
	Any valuables taken into safekeeping
Assessment	IV access
	Circulation
	Mechanical ventilation parameters
	POC Lab
	Medication/antibiotics/catecholamines
	Coagulation
	Blood products
	Fluid balance
	Intraoperative events
	Drains
Recommendation	From surgeon
	From anaesthesiologist
	Any other information for post-operative period

ASA, American Society of Anesthesiologists; iv, intravenous; POC, point of care; SBAR, Situation, Background, Assessment, Recommendation.

mWHO III and IV risk should be taken care of in an expert center for pregnancy and cardiac disease.[45]

Vaginal delivery is safe for most ACHD women and is the preferred mode of delivery,[46] caesarean section being primarily indicated for obstetric reasons. CHD where a caesarean section is advised are aggressive aortic disease (severe dilatation/dissection of the aorta), severe and intractable heart failure, when the mother is still using oral anticoagulation (to reduce the risk of foetal intracranial haemorrhage) or in severe forms of pulmonary hypertension (e.g. Eisenmenger patients).[44]

Neuraxial anaesthesia is the technique of choice for most patients. Absolute contraindications for NA include refusal of the patient, anticoagulation/bleeding diathesis, local anesthetic allergy, and infections at the insertion site. In afterload dependent heart disease, like aortic stenosis or hypertrophic cardiomyopathy, it is crucial to use vasopressors to maintain adequate coronary perfusion pressure. To lower the risk of hypotension, incremental techniques such as epidural anaesthesia and CSE are often used because they facilitate slow titration. Slow titration of NA is also crucial in Eisenmenger syndrome, as a decrease in SVR can lead to increased right-to-left shunting and subsequent cyanotic crisis. In women with pulmonary hypertension, it is important to minimize PVR by avoiding hypoxemia, hypercarbia, hypothermia, pain and sympathetic stimulation.

It is also important to consider the timing of NA techniques, because many women with cardiac disease are prescribed anti-coagulant therapy. Treatment with anti-vitamin K (VKA) should be switched to low molecular weight heparin (LMWH) or unfractionated heparin (UFH) after week 36 or if delivery is expected within a week. For patients using VKAs who present in labour or need a caesarean section, an international normalized ratio (INR) ≤ 1.4 is advised before performing a neuraxial block. For those on UFH, a minimum of 4 h without UFH, with a normal activated partial thromboplastin time (aPTT) and platelet count, is required before a neuraxial block. For parturient on therapeutic LMWH, a 24-h interval should pass after the last dose before performing a neuraxial block.

The postpartum period poses high risk for maternal cardiac complications, with haemorrhage being twice as common in women with heart disease. Prophylactic uterotonics reduce this risk. Oxytocin should be used with caution due to its negative effect on SVR. A bolus dose of 2 IU over 10 min, followed by 12 mU/min for a total of 4 h is advised.[47] Carbetocin, a heat-stable alternative to oxytocin, is advised in situations where oxytocin is unavailable, or its quality cannot be guaranteed.

Carboprost and methylergonovine should be used with extreme caution due to their potential side-effects, including severe hypertension and arterial vasospasm causing myocardial ischaemia. Of note, nitroprusside and nitroglycerin should be avoided immediately after delivery for their myometrial relaxant effect that may lead to uterine atony and postpartum haemorrhage.

Urgent surgery

Urgent surgery is a challenging situation for the ACHD population, particularly for high-risk patients.

Perioperative period

Some warning signs should be sought in all patients in the perioperative period: deterioration in systemic pressure or oxygen saturation from baseline (especially in cyanotic patients) or new onset arrhythmias or difficult rate control in a previously stabilized atrial arrhythmia. Hypovolemia and tachycardia require aggressive treatment in patients with preexisting severe outflow tract obstruction, unrestrictive shunts, Fontan circulation or cyanosis.

Intraoperative care

Despite the urgency, baseline vital signs should be recorded before induction. For high-risk patients, redo or high-risk procedures, invasive intraarterial and/or central venous monitoring is advised, especially in those more vulnerable to reduction in preload. Regarding venous access in these cases, large-bore IV catheters are required. New techniques of non-invasive cardiac output monitoring and brain oxygen regional saturation to identify global hypoperfusion related to a reduction in cardiac output are advisable, as well as TOE for major surgery. It should be noted that brain oxygen regional saturation monitoring has not been validated in patients with high haematocrit. External pads should also be available.

Anaesthetic technique

The choice of anaesthetic technique will depend on the duration of the surgery and the expected hemodynamic consequences according to the underlying cardiac condition and cardiac reserve. Peripheral nerve blocks (PNBs) and opioid minimization are advised. For patients in whom a decrease in SVR is not a concern, NA is advised.

Laparoscopy

This surgical technique is advisable due to its better post-operative recovery and is widely used in ACHD patients.[48] However, table position and the pneumoperitoneum caused with intraabdominal carbon

Table 6 Advised management of patients in the high-risk scenario

Preoperative
Prioritize non-invasive options if possible.
Planned procedures should be undertaken in centres with ACHD expertise.
Multidisciplinary team involved and case discussion in a timely fashion.
Anaesthesiologist with expertise in ACHD in charge.
Regional anesthesia preferred if possible.
Optimization of medications and underlying conditions (e.g. heart failure, anaemia, arrhythmia).
Ceilings of care to be discussed.
Complete medical and surgical history required. If no echocardiogram results available, perform echocardiogram before anaesthetic act.
Intraoperative
External pacing/cardioversion/defibrillator pads prior to induction (increased risk of arrhythmias and poorer tolerance).
Arterial line placed prior to induction in intermediate and high-risk procedures.
Avoid fluctuations of PVR or SVR (prophylactic use of vasopressors for intermediate and high-risk procedures).
Careful fluid management
Slow titration of NA anaesthesia.
ECMO support available as rescue for intermediate and high-risk procedures.
Post-operative
Availability of intensive care facilities after intermediate and high-risk procedures.

ACHD, adult congenital heart disease; ECMO, extracorporeal membrane oxygenation; NA, neuraxial anaesthesia; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.

dioxide (CO_2) insufflation with its consequent increase in intraabdominal pressure, may compromise venous return. On the other hand, hypercarbia caused by CO_2 absorption and atelectasis due to lung compression, may increase PVR. These circumstances may be deleterious for patients with pulmonary hypertension, right to left shunts and preload dependent conditions (i.e. Fontan circulation). In these scenarios, insufflation onset must be gradual and up 8–12 mm Hg, with close monitoring during onset and release and strict CO_2 partial pressure control. It is mandatory to discuss potential effects of pneumoperitoneum with the surgeon before the operation and to maintain close communication intraoperatively. Lower limb compression is advised. Analgesia should be based on PNB. Protective positive ventilation is mandatory and should always take into account the possible interaction with the underlying cardiac physiology.

Special populations

Intermediate- and severe-risk ACHD patients are more prone to complications during NCS, particularly when undergoing major procedures and they require an exquisite anaesthetic management. *Table 6* depicts a checklist of considerations for the management of patients at the High-risk Scenario. *Figure 2* highlights the key messages for these special populations when undergoing NCS.

Unrepaired cyanotic congenital heart diseases and Eisenmenger physiology

Patients with Eisenmenger physiology represent the highest risk CHD cohort, having both chronic cyanosis and pulmonary vascular disease.

Preoperative evaluation

When assessing preoperative risk stratification, the following are recognized predictors of peri-interventional morbidity and mortality: cyanosis with oxygen saturation (SpO_2) <85%, impaired ventricular function (ejection fraction < 30%), atrial fibrillation, pulmonary arterial hypertension, New York Heart Association (NYHA) stage ≥ 2, poor pre-intervention CHD appraisal. Not surprisingly, emergencies are associated with excess mortality.

Perioperative management

Air filters are advised for cyanotic patients and unrestricted shunts to prevent paradoxical air embolism, but with some precautions. Air filters need to be avoided for high-density products infusion (i.e. blood products or propofol) because they get easily clogged. In these cases, extreme care to avoid air bubbles in the line is required. Dilution of some high-density fluids (i.e. propofol) may be an option to keep air filters in place. On the other hand, in-line IV filters back-siphon when lowered below the patient's heart level and have a blousing effect when raised above it. This may induce variations in the infusion rate that may be particularly concerning for vasoactive drugs.

Most anaesthesia complications relate to hypotension and/or hypoxemia. Therefore, the anaesthesia care should focus on avoiding stimuli that could cause sudden increases in PVR and subsequent pulmonary hypertension crises such as hypoxia, hypercarbia, hypothermia, or acidosis while maintaining stable SVR. Thus, the prophylactic use of vasopressor and/or inotrope infusions (low dose to avoid undesirable tachycardia or induction of tachyarrhythmias) may be advisable starting during anaesthetic induction.[49] Vasopressin is preferred as it increases SVR without increasing PVR.[50] Parenteral pulmonary vasodilators (e.g. inhaled nitric oxide, inhaled iloprost, IV epoprostentol, IV sildenafil)

should be available. Pulmonary hypertension/cyanotic crisis should be promptly detected and aggressively treated with vasopressors, inotropes and inhaled pulmonary vasodilators.[51]

Post-operative care

This is a very high-risk period, requiring meticulous attention for hemodynamic changes. ICU or HDU admission is mandatory for intermediate and high-risk procedures since adverse events can occur up to 72 h after an apparently uneventful procedure. For most cyanotic patients, haemoglobin thresholds for erythrocyte transfusion are lower for cyanotic patients (should be 12–15 mg/dL, rather than 7–8 mg/dL).

Fontan circulation

The Fontan circulation is a univentricular circulation and is dependent on systemic venous hypertension and a low PVR to ensure preload to the single ventricle and maintain cardiac output. In the absence of a sub-pulmonary ventricle, pre-load recruitment is minimal and cardiac reserve is limited, with a limited range of cardiac output.

Pre-anaesthetic assessment

An older atriopulmonary connection may pose a greater risk than a more recent total cavopulmonary connection. History of oesophageal varices should be checked before considering intraoperative TOE monitoring.

A laparoscopic approach may be feasible but requires careful planning and management (see Special setting and Laparoscopy) with a low threshold to switch to an open approach if not tolerated or the access proves challenging.

Periprocedural management

It is advisable to assume a resting right to left shunt and therefore at risk of paradoxical embolus and insert air filters in the venous accesses whenever possible and ensure IV pre-hydration. Induction agents can take a while to take effect due to slow circulation.[52] Because positive-pressure ventilation increases intrathoracic pressure and reduces pulmonary blood flow, it is best to lower airway pressure as much as possible. However, hypoventilation and atelectasis could lead to hypercarbia, hypoxemia, and elevated PVR. Therefore, to maximize pulmonary blood flow, appropriate tidal volume, respiratory rate, and PEEP are required to avoid hypercarbia and atelectasis while maintaining low airway pressure.[53] Allow spontaneous ventilation if possible and favour EES. Extubation is accompanied by an increase of the cardiac output and a drop of the pulmonary mean pressure in Fontan patients.[54] If mechanically ventilated, prioritize spontaneous breathing with pressure support minimizing PEEP and have nitric oxide available. Central venous and Swan-Ganz catheterization should be avoided in these patients due to the associated thromboembolic risks and transthoracic echo or TOE should be favored for monitoring.

Post-operative care

Use EES whenever possible. Ensure early mobilization and enhanced recovery. Early post-operative chest physiotherapy is advised. It is important to have a low threshold for transfusion and clotting products while being aware that transfusion related overload (pulmonary oedema or acute lung injury) is potentially life threatening. Fluid management maintaining adequate filling continues to be crucial in the post-operative period.

Severe systemic ventricular dysfunction and heart failure

Heart failure is more common in patients with complex CHD compared with simple lesions.[55] ACHD who have severe systemic ventricular dysfunction and heart failure present unique challenges when undergoing anaesthesia, both during induction and maintenance. Anaesthetic management must prioritize hemodynamic stability, as these patients often have impaired cardiac output and are sensitive to fluctuations in blood pressure and volume status.

Preoperative assessment

Optimization of cardiac and other medications pre-surgery is essential to have cardiac stability. Echocardiogram, exercise capacity, and natriuretic peptides, either brain natriuretic peptide (BNP) or N-terminal pro-B-type natriuretic peptide (NTproBNP), strongly correlate with the risk of cardiovascular events in patients with heart failure and should be assessed.[9,56] Furthermore, it is important to optimally treat any factor that may compromise ventricular function and complicate anaesthesia beforehand, such as anaemia, brady- or tachycardia, pulmonary hypertension, profound cyanosis, etc.

Periprocedural management

It is important to choose appropriate anaesthetic agents that maintain cardiovascular stability, such as etomidate or ketamine, and avoid the agents that may depress myocardial function or cause vasodilation, such as propofol.[49] Intraoperative monitoring and management often necessitate invasive arterial line for real-time assessment, meticulous fluid balance to prevent volume overload, maintain preload, and avoid excessive afterload. Cautious administration of crystalloids and blood products is needed, since these patients are at a high risk of decompensation and volume overload. Inotropic support may be needed to sustain cardiac output and increase contractility. The use of advanced hemodynamic support techniques may be necessary to optimize outcomes and reduce the risk of complications during and after surgery.[57,58] In contemporary management, preoperative and post-operative TOE in the operative theatre for cardiac operations is the norm to assess early operative success and provide insight into ventricular function and volume status.

Post-operative care

Close monitoring in an intensive care setting is required. Early identification and management of arrhythmias or heart failure exacerbations are critical. Afterload reduction in the post-operative period can also decrease filling pressures and improve ventricular function, especially in the presence of any systemic AV valve regurgitation. Pain management should be optimized to prevent sympathetic stimulation.

Severe outflow tract obstructive lesions

The key aspects of anaesthesia in patients with outflow tract obstruction (OTO), such as aortic or pulmonary valve stenosis or hypertrophic obstructive cardiomyopathy, is avoiding a decrease in SVR and maintaining preload and sinus rhythm. Patients with severe OTO have a relatively fixed stroke volume (SV) with a limited ability to increase on demand. Therefore, a drop in SVR will be poorly compensated and will cause hypotension. On the other hand, the ventricular hypertrophy, that develops as the OTO worsens, will compromise the filling properties of the ventricle that will depend on an adequate preload and the atrial contraction. Therefore, the main concerns include:

Maintenance of systemic vascular resistance

Hypotension may be particularly dangerous in systemic ventricular OTO due the resulting coronary hypoperfusion and subsequent myocardial ischemia of an already hypertrophied ventricle. The use of vasoactive drugs may be necessary and bolus dosing should be prepared in advance. NA should be avoided or used with extreme caution (preferably CSE with very slow up-titration of epidural anaesthesia).

Maintenance of preload

The use of crystalloid solution may be necessary to assure an adequate preload during major surgery with bleeding or large fluid shifts. However, fluid reposition should be carefully managed in patients with severe ventricular hypertrophy (since filling pressures may increase abruptly) and in patients with reduced ventricular function.

Maintenance of sinus rhythm

Arrhythmias are common in the perioperative setting, with ≈50% of adults with CHD having some rhythm disturbance during cardiac surgery.[59] In non-cardiac surgeries, factors such as electrolyte imbalances, sympathetic activation, and inflammation play a role in arrhythmia development, which is more frequent during thoracic surgery.[60] In case of supraventricular tachyarrhythmia resulting in hemodynamic instability, immediate cardioversion may be needed and even a prophylactic use of amiodarone may be an option for highest risk patients.[7,61]

Conclusions

The number and complexity of ACHD patients exposed to anaesthetic procedures is increasing. Intermediate- and severe-risk ACHD patients should be managed in centres with anaesthetic expertise in CHD or in close consultation with an ACHD program. There is a growing need for anaesthesiologists to be aware of the intricacies inherent to the individual ACHD lesions and their distinct physiology whether repaired, palliated or native.

Author contributions

Laura Dos Subira (Writing—review & editing [equal]), Ehrenfried Schindler (Writing—review & editing [equal]), Francisco J. Ruperti (Writing—review & editing [equal]), Steffen Rex (Writing—review & editing [equal]), Marco Ranucci (Writing—review & editing [equal]), Raquel Prieto-Arévalo (Writing—review & editing [equal]), Sascha Meier (Writing—review & editing [equal]), Vera Von Dossow (Writing—review & editing [equal]), Dan Longrois (Writing—review & editing [equal]), Michael Gatzoulis (Writing—review & editing [equal]), Gerhard Diller (Writing—review & editing [equal]), Paul Clift (Writing—review & editing [equal]), Margarita Brida (Writing—review & editing [equal]), Montserrat Ribas-Ball (Writing—review & editing [equal]), Koen Rijs (Writing—review & editing [equal]), Jane Heggie (Writing—review & editing [equal]), and Jolien Roos-Hesselink (Writing—review & editing [equal]).

Declarations

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No data were generated or analysed for or in support of this paper.

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