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Guidelines

Guidelines for anaesthesia of adults with congenital heart disease in non-cardiac surgery☆,☆☆

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☆ PROFESSIONAL PRACTICE GUIDELINES *of the* SOCIETE FRANÇAISE D'ANESTHESIE ET REANIMATION (SFAR) *with the participation of the* SOCIETE FRANÇAISE DE CARDIOLOGIE (SFC), *the* SOCIETE FRANÇAISE DE PEDIATRIE (SFP), *the* CLUB ANESTHESIE-REANIMATION EN OBSTETRIQUE (CARO), and the SOCIETE FRANÇAISE DE CHIRURGIE THORACIQUE ET CARDIO-VASCULAIRE (SFCTCV).

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Abbreviations: ACEI, angiotensin-converting enzyme inhibitor; ACHD, adult with congenital heart disease; AES, atrial extra-systole; AF, atrial fibrillation; AHA, American Heart Association; ALCAPA, anomalous left coronary artery from the pulmonary artery; APVC, abnormal pulmonary venous connection; AR, aortic regurgitation; ASD, atrial septal defect; AVB, atrioventricular block; AVF, arteriovenous fistula; AVS, aortic valve stenosis; AVSD, atrioventricular septal defect; BDG, Bidirectional Glenn; BTS, Blalock-Taussig shunt; CCHD, complex congenital heart disease; CHD, congenital heart disease; CVP, central venous pressure; DOA, Direct oral anticoagulant; DORV, double outlet right ventricle; EA, epidural anesthesia; ESC, European Society of Cardiology; E_tCO_2, end-tidal carbon dioxide; F_iO_2, fraction of inspired oxygen; GA, general anesthesia; HAS, Haute Autorité de Santé (France); HR, heart rate; IAS, inter-atrial septum; IVC, inferior vena cava; IVS, interventricular septum; JET, junctional ectopic tachycardia; LA, left atria; LPA, left pulmonary artery; LRA, locoregional anesthesia; LV, left ventricle; LVH, left ventricular hypertrophy; LVOTO, Left ventricular outflow tract obstruction; M3C, Complex congenital cardiac malformation network (France); MP, Main pulmonary artery; MR, mitral valve insufficiency regurgitation; MS, mitral valve stenosis; PA-IVS, pulmonary atresia with an intact ventricular septum; PA-VSD, pulmonary atresia with ventricular septal defect; P_aCO_2, Partial arterial pressure of carbon dioxide; PAH, pulmonary arterial hypertension; m/s/dAP, mean/systolic/diastolic Arterial Pressure; PDA, patent ductus arteriosus; PEEP, positive end-expiratory pressure; PFO, patent foramen ovale; PNDS, *plan national de soin* (France); PVR, pulmonary vascular resistance; PVS, pulmonary valve stenosis; RA, right atria; RPA, right pulmonary artery; RV-PA, right ventricle – pulmonary artery; RV, right ventricle; SFC, *société française de cardiologie*; SVC, superior vena cava; SVR, systemic vascular resistance; TA, truncus arteriosus; TCPC, total cavopulmonary connection; TGV, transposition of the great vessels; TOF, tetralogy of Fallot; TR, tricuspid regurgitation; VES, ventricular extra-systole; VKA, vitamin K antagonist; VSD, Ventricular septal defect; VT, ventricular tachycardia.

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ABSTRACT

Objective: To provide guidelines for the anesthetic management of adults with congenital heart disease for non-cardiac surgery.
Design: A consensus committee of 16 experts was convened. A formal conflict-of-interest (COI) policy was developed at the beginning of the process and enforced throughout. The entire guidelines process was conducted independently of any industrial funding (*i.e.,* pharmaceutical, medical devices). The authors were required to follow the rules of the Grading of Recommendations Assessment, Development, and Evaluation (GRADE®) system to guide the assessment of the quality of evidence. The potential drawbacks of making strong recommendations in the presence of low-quality evidence were emphasized.
Methods: The committee studied 10 questions within 4 fields: preoperative evaluation, intraoperative management, postoperative care, and obstetrics. Each question was formulated in a PICO (Patients Intervention Comparison Outcome) format and evidence profiles were produced. The literature review and recommendations were made according to the GRADE® methodology.
Results: The experts' synthesis work and the application of the GRADE® method resulted in 11 expert opinions. Some of the questions did not find any response in the literature. After one round of scoring, a strong agreement was reached for all recommendations.
Conclusions: There was strong agreement among experts for 10 recommendations to improve practices for the anesthetic management of adults with congenital heart disease for non-cardiac surgery.

1. Introduction

1.1. Preamble

Anaesthetizing an adult congenital heart disease (ACHD) patient can be a source of anxiety for the anaesthetist/intensivist working in a non-expert center. The complexity of the abnormalities and previous surgical repairs associated with (left, right, or overall) cardiac insufficiency and/or severe pulmonary arterial hypertension (PAH) puts these patients at high risk of perioperative decompensation. Anaesthesia is a source of hemodynamic perturbations of which the potential consequences are aggravated in the event of underlying congenital heart disease.

For an anesthetist/intensivist not used to treat this type of patient, the simplest and safest solution, if possible, would be to refer the patient to a specialized center, as is recommended in most guidelines for the management of adult congenital heart disease (ACHD) patients [1,2]. However, these guidelines provide no advice on anesthetic strategies suitable for such patients. The only published review on anesthesia management for ACHD patients having to receive non-cardiac surgery suggested multidisciplinary treatment in a specialized center [3].

The goals of the following recommendations are not only to provide practical guidance for preoperative, per-operative, and postoperative treatment of an ACHD patient who, on account of the need for emergency surgery, cannot be referred and transferred to an expert center, but also to enable professionals to recognize the situations least likely to entail complications, and that can be surgically managed exterior to centers with expertise in congenital heart disease.

We will begin by drawing up a panorama of the main types of congenital heart disease patients, who will be divided into three groups: (1) those entailing no specific problem, and for whom anesthesia will be similar to that given to any other patient, (2) those at moderate risk, and who may require special attention, and (3) those at high risk of acute perioperative decompensation [4].

1.2. Epidemiology

Congenital heart disease (CHD) is found in nearly 0.8% of live births and is one of the most frequent congenital malformations [5].

Improved CHD management, particularly in cases necessitating intervention during the neonatal period, has considerably modified these patients' prognoses and life expectancy.

Remarkable therapeutic advancements have paved the way to corrective CHD surgery survival rates neighboring 95% [6]. As a result, 98% of patients with mild congenital malformations, 90% of those with moderate cardiac malformations, and 56% of those suffering from complex cardiac malformations now survive for at least 18 years [7], and the annual growth of this population is estimated at approximately 5%. The need for anesthesia management in non-cardiac surgery in adult patients with simple or complex, corrected, or non-corrected congenital heart disease, will inevitably become more frequent.

1.3. The caregiving networks

Medical and/or surgical management in expert centers ensures higher survival rates among these patients, particularly those suffering from complex congenital heart disease (CHD) [1].

For many years in France, many patients have left the healthcare system or, due to the absence of a specialized management network, have not been referred to expert centers for adults. However, successive "rare disease" plans, especially those of 2011 and 2017, have led to the recognition of a nationwide network of complex CHD management (the French M3C network), in the framework of the "Cardiogen" branch. As of today, the M3C network encompasses 23 reference and competence centers in all French regions, including the French West Indies (Martinique) and the Indian Ocean (CHU la Réunion). It assumes a role of expertise in CHD management and the training of network actors, facilitates cooperation, and coordinates management and good practices according to the *Protocole National De Soins* (PNDS) under the

supervision of the official French health authority (HAS). Coverage throughout French territory is complemented by the pediatric congenital cardiology subsidiary (FCPC) of the *Société Française de Cardiologie* (SFC), which helps to identify pediatric congenital cardiology practices and centers, the objectives being to refer patients and to facilitate access to specialized care.

It is consequently possible at all times to contact an M3C network center, to seek advice from experts in each field on the management of ACHD patients, and to have them referred according to existing demand and/or possibilities for management. Even if France has yet to attain the "standards of care" instituted by the governments of some countries and is lacking in the accreditations and ACHD-specific qualifying training programs proposed by the European Society of Cardiology (ESC), much ground has been covered, and substantial advances include: recognition of a pediatric congenital cardiology specialty in the training of young doctors, authorization for interventional cardiology activities in a specific setting specialized in ACHD patient management. So it is that all CHD patients, as well as doctors (non-ACHD specialists), are in a position to contact an expert center.

1.4. The main types of congenital heart disease (CHD)

The diverse nature of CHD types, the different therapeutic strategies that have evolved over time, the advancements in neonatal management, and the development of interventional catheterization are parameters complicating the appraisal of cardiopathy and patient status.

So it is that the same heart disease, whether it has been corrected or not, whether with or without residual lesions, regularly monitored or not, may have evolved differently. These differences can lead to variable degrees of cardiac insufficiency that do not impose the same perioperative constraints. The learned cardiology societies have proposed a classification of heart diseases according to their degree of complexity and the patient's physiological state. However, these classifications do not directly integrate the pathophysiological specificities of the residual lesions associated with CHD that have a direct impact on anesthetic management.

We have decided to draw up a classification of the different cardiopathies in terms of the pathophysiological groups that condition anesthetic management.

1.4.1. Shunts

1.4.1.1. Left-right shunts. Left-right shunts can be atrial, ventricular, pulmonary venous, or arterial; pathophysiology depends on the response of the pulmonary vascular bed to shunt flow. In the long term, sizable left-to-right shunt can lead to irreversibly increased pulmonary vascular resistance.

When pulmonary vascular resistance exceeds systemic resistance, the shunt reverses, becoming mainly a right-to-left shunt, resulting in systemic arterial desaturation; this is known as Eisenmenger syndrome (see "right-to-left shunt") and entails a guarded long-term prognosis and reduced life expectancy [8].

Birth defects leading to a left-to-right shunt (Figs. 1–7):

- Total abnormal pulmonary venous connection (TAPVC)
- Atrial septal defect (ASD)
- Ventricular septal defect (VSD)
- Patent ductus arteriosus (PDA)
- Partial (≈ASD physiology) or complete (≈VSD physiology) atrioventricular septal defect (AVSD)
- Truncus arteriosus (a single vessel that comes from the heart above a large ventricular septal defect and the truncal valve, leading to indivision between the aorta, the pulmonary artery, and the coronary vessels)
- Aortopulmonary window (communication between the ascending aorta and the main pulmonary artery)

The risk of PAH entailed by shunt reversal (Eisenmenger syndrome) occurs progressively according to shunt flow, which depends on its resistance (size of the communicating orifice and pressure gradient). The pressure gradient is higher at the arterial level (between the aorta and the pulmonary artery: aortopulmonary window, truncus arteriosus) than at the ventricular level (VSD, complete AVSD) and at the atrial level (partial AVSD, ASD, APVC).

Shunts at the atrial or venous level (pre-tricuspid) are low pressure and entail the passage of excessive blood throughout the right heart, thereby provoking volume overload in the atrium and the right ventricle. While major pulmonary hypertension is rare, prolonged right atrial overload predisposes the patient to atrial arrhythmia. At the same time, right ventricular insufficiency may appear in adults at the ultimate stage of the natural history of these diseases.

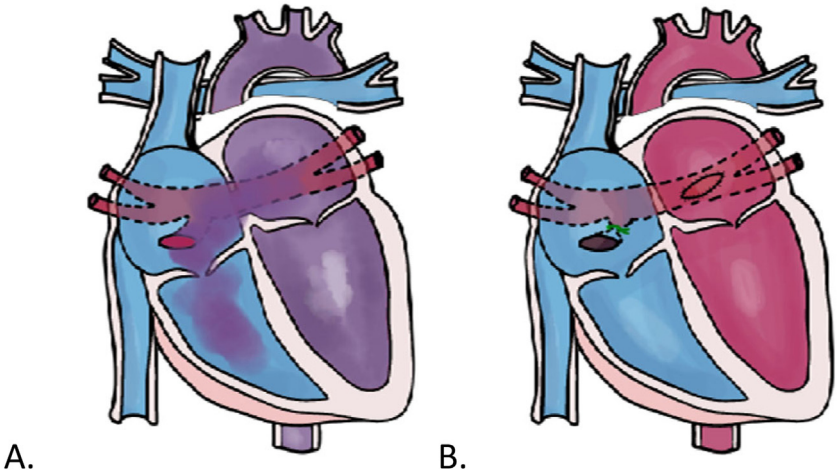


Fig. 1. Total abnormal pulmonary venous connection (TAPVC). (A) malformation: the pulmonary veins are joined at the level of a collector that empties the blood into the right atrium; (B) repair: communication with the collector is closed, and the collector is anastomosed to the left atrium.

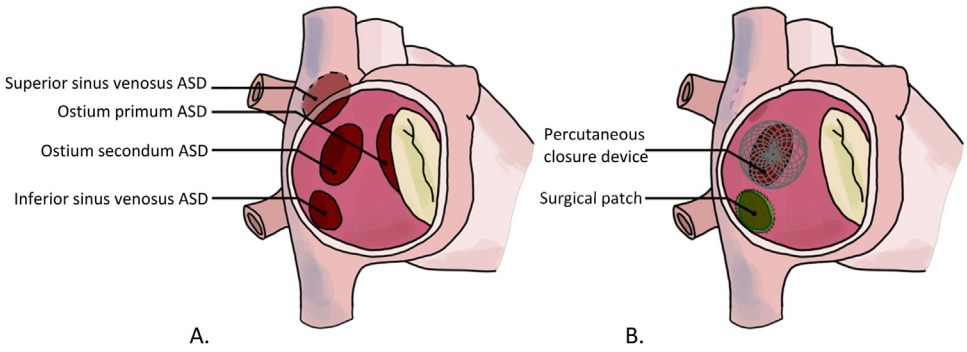


Fig. 2. Atrial septal defect (ASD). (A) Malformation: different types of ASD; B. repair: surgical patch or percutaneous closure device.

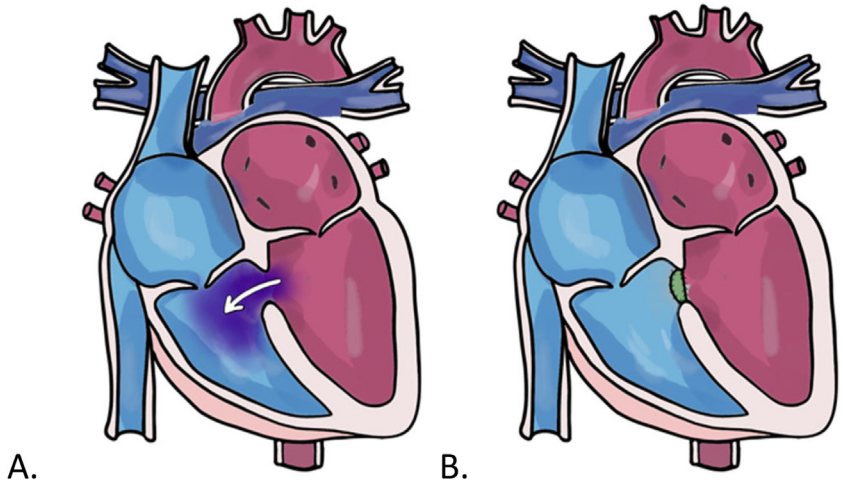


Fig. 3. Ventricular septal defect (VSD). (A) Malformation: ventricular septal defect with left-to-right shunt; B. repair: surgical patch.

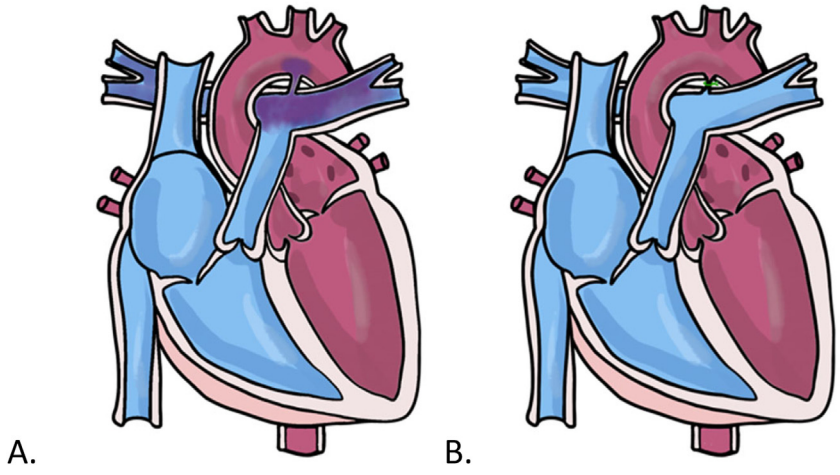


Fig. 4. Patent ductus arteriosus (PDA). (A) Malformation: communication between the aorta and the pulmonary artery by the ductus arteriosus; B. repair: surgical ligation of the ductus arteriosus (or percutaneous closure).

By provoking increased pulmonary blood flow, post-tricuspid shunts at the ventricle level or the large vessels yield left ventricular volume overload, a sign of significant flow across the shunt. With large shunts, the pulmonary vascular system is exposed to high pressure, a factor contributing to pulmonary vascular disease and Eisenmenger syndrome.

Most (surgically or percutaneously) closed shunts leave no residual lesions, insofar as the required procedures have been carried out at the early stages of the disease. That said, there exist rare situations in which PAH can continue to evolve after the closure of the shunt, and the prognosis is comparable to that associated with idiopathic PAH. In the event of CHD with the shunt

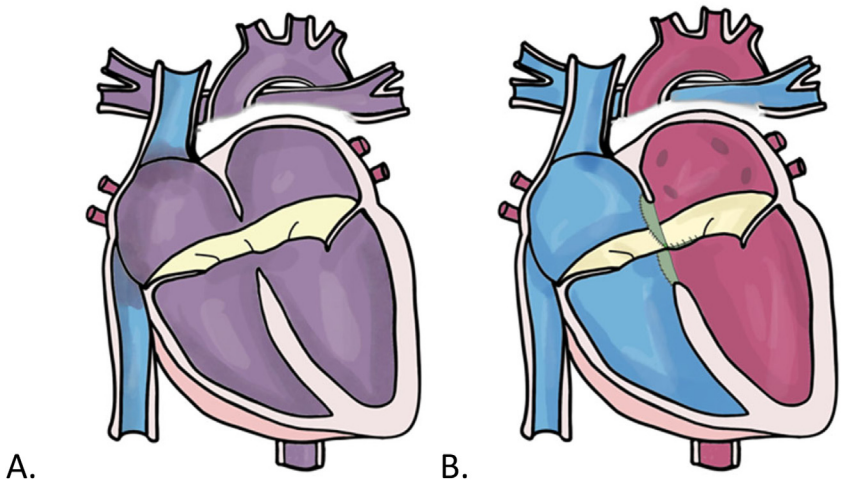


Fig. 5. Atrioventricular septal defect (AVSD). (A) Malformation: abnormalities of the atrioventricular valves and the interatrial and interventricular septa, creating a sizable shunt; (B) Repair: reconstruction of the atrioventricular valve and closure of the atrial and ventricular septal defect.

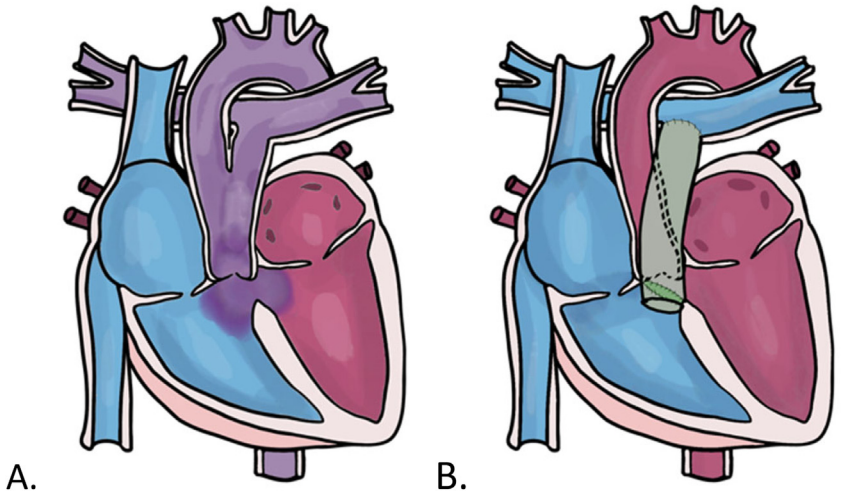


Fig. 6. Truncus arteriosus (TA) (A) Malformation: common arterial trunks between the aorta and the pulmonary artery; (B) Repair: separation of two arterial trunks by installation of patches and prostheses.

(AVSD, truncus arteriosus. . .), other lesions, many of them valvular, may appear and justify complementary cardiological evaluation at an expert center.

1.4.1.2. Right-to-left shunts. Adult CHD patients with right-to-left shunt (“cyanotic cardiopathy”) represent a highly heterogeneous population, and can be divided into two sub-groups (Table 1):

- Patients without pulmonary arterial hypertension (PAH)
- Patients with PAH secondary to a non-restrictive shunt between systemic and pulmonary circulation (Eisenmenger).

The patient with ventricular septal defect at the Eisenmenger stage (high pulmonary arterial pressure/resistance) is the most classic “cyanotic” example. Conversely, one may encounter cases of cyanotic CHD with normal pulmonary vascular resistance, such as single ventricle cardiopathy palliated by Fontan circulation (*Cf infra 1.4.4*) complicated by intrapulmonary arteriovenous fistulas. Between these two extremes, numerous variants exist.

Even though there exists a common approach addressed to all patients with cyanotic congenital heart disease, each lesion necessitates expertise to understand the underlying anatomy, the pathophysiology, and the therapeutic specificities. The risk entailed by anesthesia consists of aggravation of the right-to-left shunt and cyanosis in the event of lower SVR or higher PVR. The key question remains the presence or absence of PAH, which is closely associated with a risk of severe complications, possibly leading to death.

1.4.2. Congenital right heart disease

- Tetralogy of Fallot and pulmonary atresia with IVC

Tetralogy of Fallot (TOF) features four main characteristics: (1) right ventricular outflow tract obstruction, (2) VSD, (3) ascending aorta astride the VSD, and (4) right ventricular hypertrophy. Outflow tract obstruction is a clinically relevant lesion and can be sub-valvular, valvular, supravalvular, or present at several levels. When carried out in early infancy, repair consists of closing the INV and relieving the right ventricular outflow tract obstruction, which is in some instances with pulmonary artery plasty. A systemic-pulmonary (Blalock-Taussig) anastomosis between the brachio-cephalic artery and the pulmonary artery is sometimes performed to increase pulmonary flow and improve hematosis while awaiting weight gain allowing for definitive repair (Fig. 8).

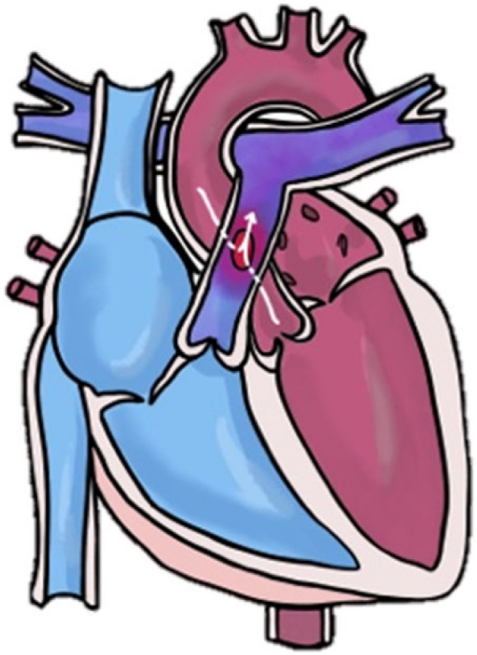


Fig. 7. Aortopulmonary window (AP Window): communication between the aorta and the pulmonary artery creating a left-to-right shunt.

Pulmonary atresia with VSD, otherwise known as pulmonary atresia with ventricular septal defect (PA-VSD) is an extreme form of the tetralogy of Fallot and refers to incomplete development of the pulmonary valve and/or the pulmonary arteries.

On principle, surgical treatment consists of closing the VSD and inserting a right ventricular pulmonary (RV-PA) duct. However, this type of repair is not possible before the pulmonary arteries have reached a certain size. Stenoses or hypoplasia of the peripheral pulmonary arteries are responsible for right ventricular hypertension and can prevent complete closure of the VSD, which serves if necessary, as a valve (right-left shunt), once continuity between the right ventricle and the pulmonary arteries has been attained.

At least one catheterization session can be organized with percutaneous pulmonary angioplasty +/− stenting to improve pulmonary blood flow toward affected pulmonary segments, thereby rehabilitating the pulmonary arterial tree.

At times, pulmonary atresia with VSD can involve side branches (collaterals) arising from the aorta and connected to the pulmonary arterial vasculature to compensate for pulmonary arterial hypoplasia. In these severe forms, surgical procedures should help to connect the systemic pulmonary collateral arteries with the central pulmonary arteries (unifocalisation) (Fig. 9).

Pulmonary outflow tract obstruction relief is generally complicated by severe residual pulmonary insufficiency. Over time in an adult, chronic severe pulmonary insufficiency entails dilation and right ventricular dysfunction yielding exercise intolerance, and pulmonary valve replacement (PVR) may be necessary, generally involving installation of a valvular prosthesis (homograft, porcine, or bovine prosthesis).

Other complications include residual pulmonary vascular obstruction, residual VSD, right ventricular systolic and diastolic dysfunction (restrictive right ventricle), left ventricular dysfunction, dilated aortic root (associated or not with aortic regurgitation/insufficiency, entailing minimal risk of dissection) and ventricular and atrial arrhythmia.

Table 1
Right-to-left shunt (cyanotic heart disease).

Right-to-left shunt without PAH	Right-to-left shunt with PAH
Pulmonary tract obstruction: Tetralogy of Fallot Double-chambered right ventricle with pulmonary stenosis Double discordance + VSD + pulmonary stenosis Pulmonary atresia with or without VSD and aortopulmonary collaterals All forms of single-ventricle heart with pulmonary outflow tract obstruction Intrapulmonary arteriovenous malformations after a cavopulmonary connection Congenital malformation without pulmonary outflow tract obstruction: Ebstein anomaly with permeable foramen or ASD	"Simple" CHD Non-restrictive VSD ASD (possible coincidence with genetic predisposition to PAH) Complex CHD without "protective" pulmonary obstacle: All forms of single-ventricle heart without pulmonary outflow tract obstruction Double discordance + VSD Truncus arteriosus Aortopulmonary connections: Patent ductus arteriosus Aortopulmonary window Large aortopulmonary collateral Potts anastomosis (between the ascending aorta and left PA) / Waterston anastomosis (between the ascending aorta and right PA)

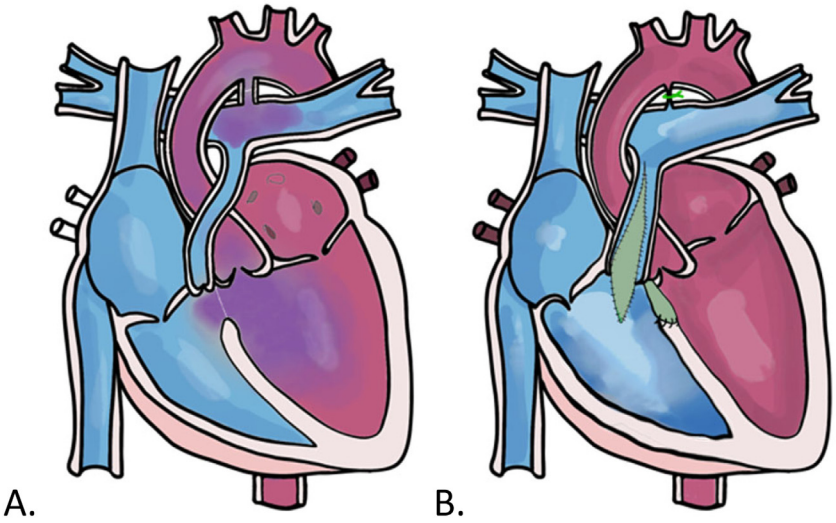


Fig. 8. Tetralogy of Fallot (TOF). (A) Malformation: association of four anomalies: VSD, pulmonary stenosis, dextroposition of the aorta, and right ventricular hypertrophy; (B) Repair: closure of the VSD by patch, enlarging of the RV-PA tube.

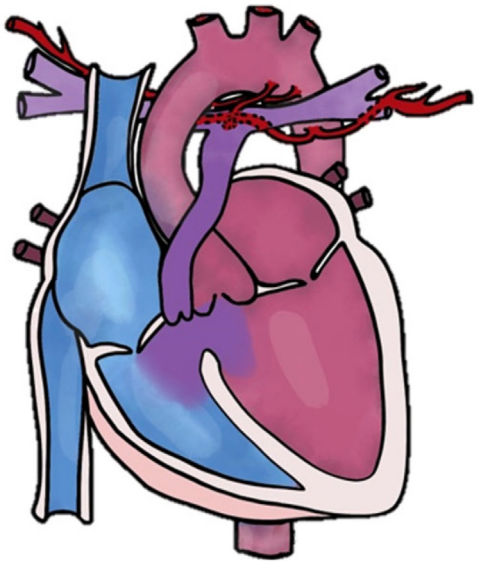


Fig. 9. Pulmonary atresia with ventricular septal defect associated with major aortopulmonary collateral arteries (MAPCAs).

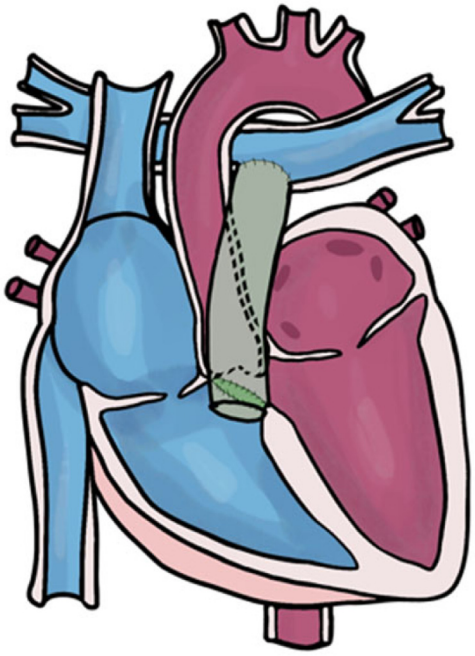


Fig. 10. Repaired persistent truncus arteriosus.

- Truncus arteriosus (TA)

Surgical repair of the TA consists of closing the VSD and implanting a tube between the right ventricle and the pulmonary artery. Tube dysfunction can lead to pulmonary leakage (regurgitation) and/or obstruction, entailing the same consequences as those following a tetralogy of Fallot operation (Fig. 10).

- Pulmonary atresia with intact ventricular septum (PA-IVS)

PA/IVS is characterized by pulmonary valve atresia, hypertrophy and right ventricular fibrosis, tricuspid valve dysplasia, and anomalies of the coronary artery. The most striking characteristic is severe hypertrophy of the right ventricle and various degrees of hypoplasia of the resulting cavity. While two-ventricle repair is considered the best option, percutaneous pulmonary valvuloplasty yields excellent results; that said, surgical pulmonary valvuloplasty or RV-PA duct enlargement is an alternative approach. A combination of pulmonary valvuloplasty or valvotomy with a bidirectional Glenn (anastomosis of the SCV to the pulmonary arteries) can be considered in cases of moderate tricuspid and right ventricular hypoplasia in which establishment of biventricular circulation remains uncertain; the procedure is known as "one and half ventricle repair". Complications include pulmonary and tricuspid regurgitation associated with fibrosis and hypertrophy of the right ventricle, which is responsible for the significant alteration of its diastolic function, with heightened right atrial and venous pressure. As a result, there exists a risk of atrial and/or ventricular arrhythmia and, in many cases, a need for replacement of the pulmonary valve (Figs. Fig. 1111 and Fig. 1212).

- Ebstein

The Ebstein anomaly is characterized by apical displacement of a tricuspid valve that is often dysplastic, with a possibly defective right ventricle. Apical displacement mainly affects the septal and posterior leaflets of the valve. The tricuspid leaflets are generally

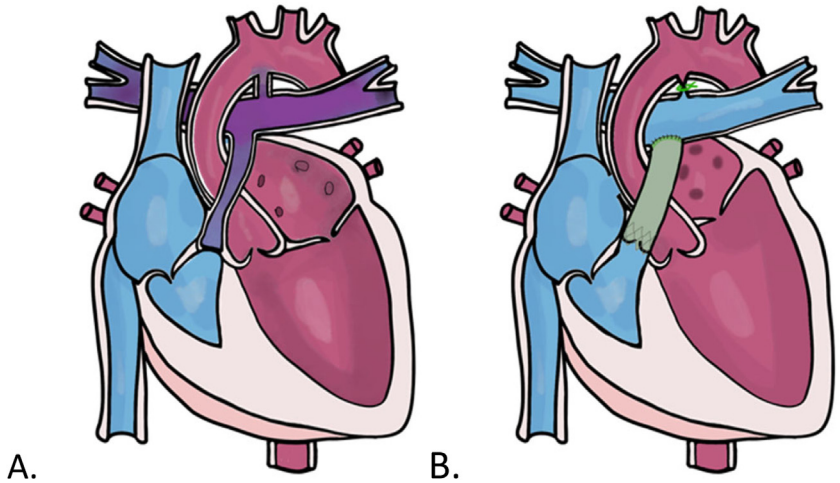


Fig. 11. Pulmonary atresia with intact ventricular septum (PA-IVS). (A) Malformation: pulmonary atresia, resulting in hypoplasia of the right ventricle, right-left shunt by ASD, and left-right shunt by PDA. (B) Repair: ligation of the PDA and enlargement of the RV-PA duct.

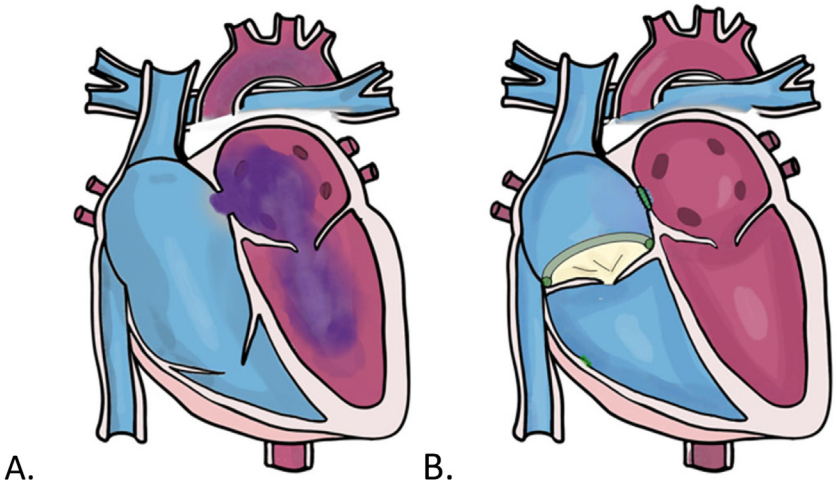


Fig. 12. Ebstein Malformation. (A) Malformation RV atrialisation; (B) Repair: tricuspid valvuloplasty and ASD closure.

small and attached to the ventricular wall (defective delamination of the leaflets). Typically, the anterior leaflet is large. The right ventricle above the valvular plane (atrialised chamber) is often thin, sometimes fibrous, and can present with diastolic and systolic dysfunction. The subvalvular right ventricle (or functional ventricle) is of determinative importance in the prognosis for this lesion. Most of the time, significant tricuspid leakage occurs. Half of the patients also present with ASD or FOP, which can lead to right-left shunt if RA diastolic pressure increases (by TR and/or RV defect). Fifteen percent of patients present with accessory bypass tracts, which are clinically manifested by the de Wolff-Parkinson-White syndrome. The main complication of the Ebstein anomaly consists of tricuspid regurgitation or right-sided cardiac insufficiency. If the latter is sufficiently severe, the implantation of a valvular prosthesis or valvular repair (if the valvular anatomy is compatible) is indicated. Scenesters associate valve surgery and bidirectional Glenn (anastomosis of the SCV to the right pulmonary artery), the objective being to unload a defective right ventricle (“one and half ventricle repair”).

1.4.3. Left-sided CHD

- Left ventricle lesions.

They encompass congenital mitral valve obstructive lesions involving the subvalvular and supravalvular apparatus: the supravalvular membrane, tri-atrial heart, or pulmonary venous stenosis. Their pathophysiology recalls that of mitral valve stenosis. While these lesions can be diagnosed during adulthood, they are often operated on during childhood when symptoms appear. Valvuloplasty/membrane resection is usually recommended as first-line treatment but with a significant risk of mitral valve redo surgery due to a high rate of regurgitation and/or residual obstruction.

Mitral valve regurgitation is another left ventricle lesion necessitating hospitalization, which is invariably due to a congenital valvular anomaly (parachute, cleft, or prolapsed mitral valve) or residual regurgitation after valvuloplasty. The pathophysiology is that of mitral valve insufficiency.

- Left ventricular outflow tract obstruction.

LVOTO includes aortic valvular (unicuspidal or bicuspid valve), subvalvular, and supravalvular stenoses; its pathophysiology is the same as that of aortic valve stenosis. In cases of valvular and subvalvular lesions, aortic insufficiency can be associated with the obstacle. The bicuspid valve is also associated with a risk of aortopathy (aneurysm of the ascending aorta, risk of aortic dissection). These lesions can be diagnosed during adulthood or in the event of obstacle recurrence following aortic valvuloplasty and subvalvular stenosis resection.

- Hypoplasia/interrupted aortic arch/aortic coarctation.

In coarctation of the aorta, narrowing/stenosis is generally situated in the proximal portion of the descending aorta, just below the left subclavian artery. The extreme form of coarctation is the interruption of the arch, and an extended form is characterized as hypoplastic. Adult patients can be divided into two groups: native (non-repaired), and repaired coarctation, which may sometimes entail residual stenosis. Some coarctations are mild and hemodynamically insignificant. The clinical gradient can be measured by comparing the highest systolic pressure of the (right) arm and the systolic pressure of the lower limbs (measured with a blood pressure cuff) (Fig. 13).

1.4.4. Single ventricle

“Single ventricle” brings together several cardiac malformations in which only one ventricle is functional; its morphology can be right, left, or intermediate.

The single ventricle “Fontan” palliation is based on the supposition that a subpulmonary ventricular pump is not required to ensure systemic venous return through the pulmonary vascular bed. Fontan palliation (the procedure is not considered a repair) encompasses at least two surgical steps:

- As a direct connection between the superior vena cava (SVC) and the right pulmonary artery (RPA), a superior cavopulmonary connection is also known as the bidirectional Glenn intervention (BDG).
- As a connection of the inferior vena cava (IVC) to the RPA, most often via an intracardiac or extra-cardiac tube, total cavopulmonary connection (TCPC) is known as the Fontan intervention. Pulmonary blood flow is ensured by the gradient between systemic venous pressure, which in most cases is moderately elevated, and pulmonary arterial pressure (Fig. 14).

1.4.5. Systemic right ventricle

Systemic right ventricle refers to a cardiac anomaly in which the morphological right ventricle serves as the systemic ventricle. The

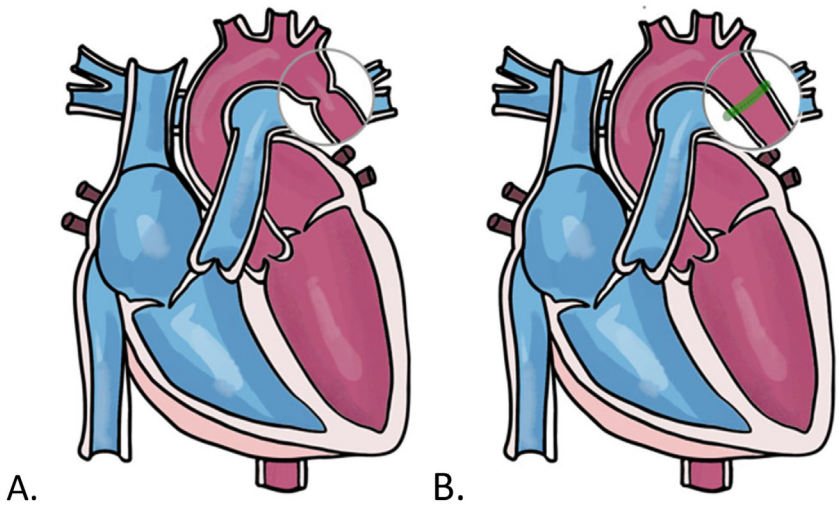


Fig. 13. Aortic coarctation. (A) Malformation; (B) Repair: enlargement of the coarcted zone.

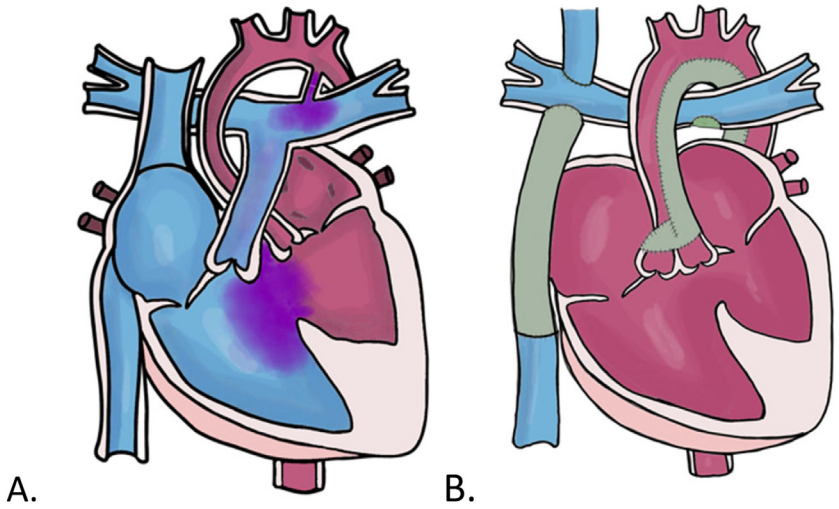


Fig. 14. (A) Single ventricle, hypoplasia of the left ventricle, IVC, and PDA. (B) Fontan circulation (total cavopulmonary connection).

morphological ventricle is determined by some anatomical characteristics. The morphologically right ventricle has a tricuspid atrioventricular valve (with attachments to the septum and apical displacement to the mitral valve) as well as numerous trabeculations.

The systemic right ventricle is found in two types of biventricular CHDs: double discordance or "corrected" transposition of the great vessels and transposition of the great vessels corrected by the switch (Senning or Mustard intervention), which essentially consists of redirecting venous blood toward the subpulmonary right ventricle and pulmonary venous blood toward the systemic right ventricle. This procedure was frequently carried out during the 1980s.

During the first decades of human life, the right ventricle can sustain high-pressure systemic circulation; during adulthood, on the other hand, in more than half of patients, the ventricular function begins to deteriorate; in these types of cardiopathy, cardiac insufficiency is the major complication. In the event of double discordance, this phenomenon can be aggravated by tricuspid valve regurgitation, which is often "Ebsteinoidally" abnormal; moreover, supraventricular arrhythmia frequently occurs following the aforementioned atrial switch.

1.4.6. Coronary artery anomalies

There are three types of coronary artery anomalies:

- Native coronary artery anomalies

ALCAPA (Anomalous Left Coronary Artery from the Pulmonary Artery) is responsible during the first months of life for myocardial ischemia justifying surgical reimplantation. This malformation is seldom diagnosed during adulthood (Fig. 15).

- Anomalous course of coronary artery

One or both coronary arteries can originate in the right sinus of Valsalva. In that case, the left coronary artery branch is situated between the aorta and the main pulmonary artery and is subjected during each systole to cyclic compression. Cases of unexplained sudden death in adolescents and young adults, particularly during intense efforts, have been ascribed to this malformation.

It is also possible that one or both coronary arteries arise from the left sinus of Valsalva, in which case the right coronary artery is situated between the aorta and the subpulmonary infundibulum; while compression may occur, sudden death seldom does.

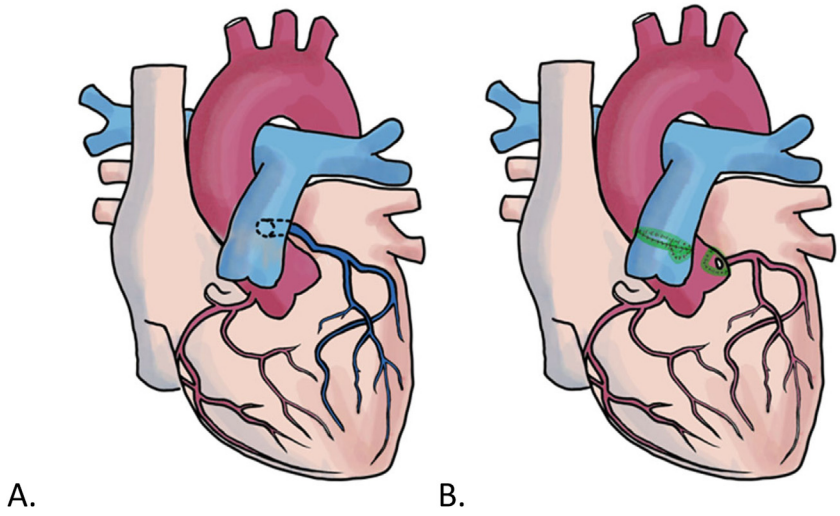


Fig. 15. ALCAPA. (A) Malformation; (B) Repair.

The term "myocardial bridge" designates the intramyocardial course of a proximal coronary artery segment, leading in some rare cases to compression and myocardial ischemia entailing a risk of sudden death.

These lesions are diagnosed either fortuitously during an imaging exam or due to anginal symptoms. An indication for surgical or percutaneous revascularization is given in the event of symptoms or documented myocardial ischemia.

Coronary lesions acquired following cardiac surgery.

Coronary restenosis is possible following:

- Transposition of the great arteries repaired by arterial switch and necessitating transfer and coronary reimplantation.
- Treatment of birth defect or abnormal coronary course.

Any suspect symptomatology or ECG aspect necessitates a coronary check-up by scanner or angiography before anesthesia.

1.5. Classification in groups according to severity levels of congenital heart disease

The CHD complexity and severity spectrum is signally wide. It ranges from a simple corrected cardiopathy, such as the closure of an intracardiac shunt that normalizes cardiac physiology and renders anesthesia management for non-cardiac surgery relatively routine, to the other extreme, one example being the single ventricle, which renders anesthesia management for a non-cardiac surgical procedure decidedly complex.

In addition to anatomic complexity, physiological complexity can be brought to bear; the same malformation can evolve in several different directions according to previous repair, presence or absence of residual lesion, alteration in myocardial function, rhythm, and/or conduction disorders.

In the spirit of the learned cardiology societies having issued guidelines [2,9,10], we are proposing three groups in ascending order of complexity regarding CHD [4,11] (Table 2):

1 *Low-risk group*: CHD patients for whom anesthetic management does not justify specific attention.
2 *Intermediate-risk group*: CHD patients for whom anesthetic management justifies specific attention.
3 *High risk of acute decompensation and perioperative death.*

To refine levels of CHD risk, it is necessary to integrate physiological status according to the 2018 guidelines of the American Heart Association [10] (Table 3).

To appraise the anesthetic risk associated with congenital heart disease (Table 4), it is necessary to associate the risk entailed by the heart disease (as defined in Table 2) with physiological status (as defined in Table 3).

In addition to the risk associated with CHD (inherent to its complexity and the patient's physiological status), the risk associated with the surgical procedure should be considered (Table 5) [12]:

In the final analysis, it is possible to determine the composite risk of the procedure considering the "patient" risk associated with the heart disease, his/her physiological status, and the risk associated with the surgery.

2. Methodology

These recommendations result from the work of a group of experts brought together by the SFAR, with the participation of the SFC, the SFP, and the SFCTCV. Each expert filled out a declaration of potential competing interests before engaging in the work of analysis. As a first step, the organizing committee determined the

Table 2
Groups at risk according to type of congenital heart disease.

Minor risk CHD	Intermediate risk CHD	Severe risk CHD
Patients with minor, 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.	Patients with corrected or uncorrected conditions with residual haemodynamic abnormality, with or without medication.	Patients with uncorrected cyanotic heart disease, pulmonary hypertension, other complex CHD, ventricular dysfunction requiring medication, and patients listed for heart transplantation.

*Palliation is the therapeutic strategy adopted when a two-ventricle anatomic repair is not possible. Example: Fontan palliation for single ventricle. Drawn from the ESC 2022 guidelines [11].

Table 3
Classification of adult congenital heart diseases according to physiological status.

Physiological stage
A
NYHA FC I symptoms No hemodynamic or anatomic sequelae No arrhythmias Normal exercise capacity Normal renal/hepatic/pulmonary function
B
NYHA FC II symptoms Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction) Mild valvular disease Trivial or small shunt (not hemodynamically significant) Arrhythmia not requiring treatment Abnormal objective cardiac limitation to exercise
C
NYHA FC III symptoms Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both) Moderate aortic enlargement Venous or arterial stenosis Mild or moderate hypoxemia/cyanosis Hemodynamically significant shunt Arrhythmias controlled with treatment Pulmonary hypertension (less than severe) End-organ dysfunction responsive to therapy
D
NYHA FC IV symptoms Severe aortic enlargement Arrhythmias refractory to treatment Severe hypoxemia (almost always associated with cyanosis) Severe pulmonary hypertension Eisenmenger syndrome Refractory end-organ dysfunction

From the AHA 2018 [10].

objectives, the methodology, and the fields of application, as well as the questions to be addressed because of drawing up the recommendations. These elements were subsequently modified and validated by the experts.

To the greatest possible extent, the questions were formulated following the PICO (Population – Intervention – Comparison – Outcome) format.

The recommendation fields

For the present recommendations, the experts unanimously decided on the four following fields:

FIELD 1 – Preoperative risk
FIELD 2 – Peroperative management
FIELD 3 – Postoperative management
FIELD 4 – Obstetric management

The importance of peroperative management chronology and a certain number of obstetric specificities were considered when choosing the fields.

Extensive bibliographic search covering the period from January 2002 to 30 June 2022 was carried out using the MEDLINE and COCHRANE databases by at least two experts for each field of application according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) methodology for systematic reviews.

Were included in the analysis: meta-analyses, randomized controlled trials, non-randomized prospective trials, retrospective cohorts, case series, and case reports; conducted among patients and caregivers or in their environment; dealing with anesthesia in adult congenital heart disease patients; published in French or English.

Analysis of the literature was then conducted following the GRADE® (Grade of Recommendation Assessment, Development, and Evaluation) method. The judgment criteria were preliminarily determined as follows:

- Primary endpoint: mortality (importance 9);
- Secondary endpoints: cardiovascular complications (importance 8), neurological complications (importance 8), infectious complications (importance 7), duration of stay in critical care (importance 7), duration of hospital stay (importance 6),

Table 4
Risk and pathophysiological classification of congenital heart disease.

Pathophysiological category		Physiological status= (*Cf.* Table 3)	Risk associated with CHD (*Cf.* Table 2)	Common risks
Shunts				
Left-to-right shunts	Pre-tricuspid: RVPA Large ASD (>10 mm) Partial AVSD	A B	Low	Severe RV dysfunction, TR > moderate PAHT
		C	Intermediate	Supraventricular arrythmia ++, conduction disorder + /–
		D	High	
	Post-tricuspid: VSD PDA Complete AVSD	A	Low	Severe LV dysfunction (chronic overload) PAHT
		B C	Intermediate	
		D	High	
Right-to-left shunts	With PAHT: Large VSD, CA large ASD AVSD TA PA-VSD	D	High	Cardiac insufficiency Polyglobulia/thrombopenia/ thrombopathy Hemoptysis Paradoxical embolism
	Without PAHT: VSD + Pulmonary stenosis Ebstein ASD Single ventricle Fontan	C	Intermediate	Endocarditis
		D	High	
Right heart CHD				
	Repaired right heart: Fallot PA-VSD APSI PA-IVS DORV Ross arterial switch	A	Low	Pulmonic regurgitation > mild Narrow pulmonary stenosis (>64 mmHg) RV dysfunction, TR Arrhythmia Aortopathy/ AR LV dysfunction Endocarditis (prosthesis of the RV-PA tube)
		B C	Intermediate	
		D	High	
	Tricuspid: Ebstein	A	Low	TO > moderate RV dysfunction ASD (Cf. CC cyanosis) Pre-excitation/ SVT
		B C	Intermediate	
		D	High	
Left heart CHD: Congenital MS Congenital MR Congenital ASD Aortic coarctation		A	Low	MR: HTAP, SVT MR: LV dysfunction, SVT AI: LV dysfunction, TV, aortopathy AoC: AHT, brain aneurysm, aortopathy, LV dysfunction
		B C	Intermediate	
		D	High	
Systemic RV: atrial switch (D-TGV) double discordance (L-TGV)		A	Low	Severe RV dysfunction Moderate to severe tricuspid regurgitation
		B C	Intermediate	
		D	High	
Single ventricle-Fontan		C	Intermediate	Single ventricle dysfunction, valve leakage AV > moderate Aortic stenosis SVT: conduction disturbance Fistula/ collateral (Cf. CC cyanosis) Cirrhosis, exudative enteropathy, plastic bronchitis, renal insufficiency
		D	High	
Coronary anomaly		A	Low	Ischemia Ventricular dysfunction Ventricular arrhythmia Ischemic MVI (rare)
		B C	Intermediate	
		D	High	

Table 5
Classification of mortality risk according to surgical procedure.

Low-risk: <1%	Intermediate-risk: 1–5%	High-risk: >5%
Superficial surgery Breast surgery Dental surgery Thyroid surgery Ophthalmology Reconstructive surgery Asymptomatic carotid Minor gynaecology Minor orthopaedics (ex: meniscectomy) Minor urology (TURP)	Intraperitoneal: Splenectomy, hiatal hernia, cholecystectomy Symptomatic carotid Peripheral angioplasty endovascular Aneurysm repair Head and neck Neurosurgery or Major orthopaedic (hip, spine) Major urologic or gynaecological Renal transplantation Non-major thoracic surgery Obstetrics	Major aortic or vascular surgery (revascularisation PAOD amputation) Duodeno-pancreatic surgery liver resection, bile duct surgery Oesophagectomy Repair of perforative bowel Adrenal resection Total cystectomy Pneumonectomy Transplantation (liver or lung)

Estimated surgical risk approximates the risk of death at 30 days and of myocardial infarction without considering the type of surgical procedure or considering patient comorbidities. Adapted from the ESC/ESA 2014 guidelines [12].

respiratory complications (importance 6), renal insufficiency (importance 6), hemorrhagic complications (importance 6).

Given the very small number of studies corresponding with the necessary power to the primary endpoint (*i.e.,* mortality), it was preliminarily decided to adopt the professional practice recommendations (PPR) rather than the formalized expert recommendations (FER) format. That said, the GRADE® methodology was applied to analyze the literature and draft tables summarising the data in the literature. A level of evidence was defined for each bibliographic reference according to the type of study. It was reassessed considering the methodological quality of the study, the coherence of the results between the different studies, the direct or indirect nature of the evidence, the analysis of cost, and the extent of the benefit. The recommendations were then drafted, using the PPR terminology of the SFAR: ''The experts suggest doing'' or ''The experts suggest not to do''. The proposed recommendations were presented and discussed, one by one. The goal was not necessarily to have the experts agree on all the proposals, but rather to identify points of concordance instead of points of divergence or indecision.

Each expert evaluated each recommendation and separately rated it on a scale ranging from 1 (complete disagreement) to 9 (complete agreement). The overall rating was validated by the experts according to the GRADE® grid methodology. To validate a recommendation, at least 70% of the experts had to express a converging opinion, while fewer than 20% expressed a diverging opinion. If one or more of the recommendations could not be thereby validated, it was reformulated and rated anew, the objective being to reach a consensus.

3. The results

3.1. Recommendation fields and questions

During the first meeting to organize these guidelines, the experts consensually decided to address 10 questions distributed in four fields. The following questions were selected for the collection and analysis of the literature:

FIELD 1 – Preoperative evaluation

- In adults with congenital heart disease, does preoperative assessment help to reduce the risk of perioperative morbimortality in non-cardiac surgery?
- In adults with congenital heart disease, does management in an expert center for congenital heart disease help to reduce perioperative complications in non-cardiac surgery?

Field 2 – Peroperative management

- In adults with congenital heart disease, does locoregional anesthesia in non-cardiac surgery help to reduce perioperative mortality in comparison with general anesthesia?
- In adults with congenital heart disease undergoing non-cardiac surgery, does the anesthetic strategy (equipment, specific monitoring) help to reduce the risks of perioperative morbimortality?

Field 3 – Postoperative management

- In adults with congenital heart disease undergoing non-cardiac surgery, does postoperative surveillance in critical care help to reduce postoperative complications?
- Following non-cardiac surgery in adults with congenital heart disease, does advice by adult congenital heart disease specialists help to reduce risks of morbimortality?

Field 4 – Obstetric management

- In adults with congenital heart disease during the peripartum period, does prepartum cardiopathy assessment help to reduce peripartum mortality?
- In pregnant chronic heart disease patients, does management in an expert center help to reduce complications during the peripartum stage?
- In chronic heart disease patients during the peripartum stage, does the anesthetic strategy help to reduce the risks of peripartum morbimortality?
- In adults with congenital heart disease, does postpartum surveillance in critical care help to reduce postpartum complications?

3.2. Synthesis of the results

The experts' efforts at synthesis and application of the GRADE® method led to the formalization of 11 recommendations, all of which were submitted to the group of experts for rating according to the GRADE® Grid method. After two rounds of rating, a strong agreement was reached for 100% of the recommendations.

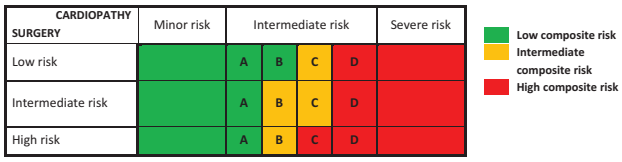
The SFAR urges all anesthesia-intensive care practitioners to comply with these PPRs to optimize the quality of patient care. When applying these recommendations, however, each practitioner is called upon to exercise his own judgment, taking into full account his area of expertise and the specificities of his establishment, to decide on the means of intervention best suited to the patient of whom he is in charge.

FIELD 1: Preoperative risk

Question: In ACHD, does preoperative assessment help to reduce the risk of perioperative morbimortality in non-cardiac surgery?

Experts: Pascal AMEDRO, Catherine KOFFEL, Magalie LADOUCEUR, Nadir TAFER, Diane ZLOTNIK

R1.1 – The experts suggest the use of the following composite score, of which the construction is explained in introduction §1.2, to assess perioperative risk in adult heart disease patients in non-cardiac surgery.



A, B, C, D: physiological status (Cf Table 3) **A procedure using peripheral LRA is considered as having a low composite score, whatever the heart disease status.*

Expert opinion (Strong agreement)

Argumentation: No randomised prospective study has evaluated the effect of preoperative assessment in non-cardiac surgery of adult CHD patients. The existing data in the literature are essentially issued from a retrospective analysis of patient series/registers. Several studies have reported significantly higher perioperative mortality, ranging from 2% to 7%, in adult CHD patients [13–17]. Indeed, congenital heart disease has been reported as an independent risk factor for perioperative mortality [13,15]. Several studies have highlighted the importance of preoperative heart disease assessment [1,18–20]. In a retrospective study involving a low-population cohort, the absence of preoperative CHD identification and assessment seemed related to 50% of reported deaths [18]. In these studies, the identified risk factors for patient mortality were: type of heart disease single-ventricle palliation cyanosis with basal SpO_2 <85% altered cardiac function (LVEF < 30%) atrial fibrillation, pulmonary arterial hypertension NYHA stage ≥2. The predictive factors for perioperative morbidity were [13–20]: Resting arterial saturation <90% Altered myocardial function (LVEF < 30%) NT-proBNP assay ≥33.3 pmol/L Poor preoperative CHD appraisal-Concerning the evaluation of surgical risk, an emergency condition is a crucial factor. In a retrospective analysis of a North American cohort consisting of 10,004 CHD patients paired with 35581 non-CHD adult patients having undergone non-cardiac surgery, in a multivariate analysis, Maxwell et al. found emergency conditions to be associated with excess mortality (OR 2.13; CI95% [1.99–2.28]) [13]. Glance *et al.* divided a range of surgical procedures into three groups: low risk, intermediate risk, and high risk (Table 4) [11,21].Regarding ACHD patients, there presently exists no validated score estimating the risk of preoperative morbimortality in non-cardiac surgery. In light of this argumentation, the authors have proposed a three-category composite score associating CHD based on the classifications found in the 2018 AHA guidelines [10], the risk associated with a patient's physiological state, and the surgical risk represented by the procedure (cf. Table 6).

Question: In ACHD patients undergoing non-cardiac surgery, does management in an expert center for congenital heart disease help to reduce perioperative complications?

Experts: Stéphane LE BEL, Bertrand LEOBON

R1.2 – The experts suggest that management of ACHD with a high or intermediate score should take place in an expert center to reduce the occurrence of perioperative complications.

Expert opinion (Strong agreement)

Argumentation: Several indirect arguments favor the management of these patients in an expert center. Karamlou et al. studied the combined impact on perioperative mortality, following cardiac surgery for CHD, of a surgeon and a specialised center [22]. In this work, they compared mortality in patients operated by cardiac surgeons specialised in CHD, in a hospital likewise specialised in CHD, and in a hospital not specialized in CHD. They highlighted pronouncedly higher mortality in the ''non-specialized hospital'' (OR 9.07; CI 95% [2.99–27.56]; $p < 0.001$). These results seem to underline the importance of CHD competence throughout the medical and surgical staff in this type of complex management. The context of non-cardiac surgery was not considered. In a study of the Quebec Congenital Heart Disease Database, Mylotte et al. observed a significant increase between 1990 and 2005 in the number of patients followed in specialised centers (RR + 7.4%; CI95%, [6.6 %–8.2 %] $p < 0.0001$) [1]. This was partially due to the publication in 1998 of nationwide recommendations on the monitoring of ACHD patients in Canada. The enlarged proportion of patients managed in a specialised center was associated with reduced mortality (RR −5.0%; CI95% [−10.8% to −0.8%] $p = 0.04$), most particularly in the group of ''high-risk'' CHD patients. The specific context of non-cardiac surgery was not examined in this study. A survey of 168 anesthesists conducted by Maxwell et al. on their knowledge of anaesthesia in ACHD patients highlighted a lack of confidence in their ability to manage ACHD patients among those not specialised in pediatrics or cardiology [23]. Even though the results of these different studies essentially provide indirect arguments, they all converge when emphasising the importance of management in an expert CHD center of patients at intermediate to high risk, the common objective is to reduce perioperative complications.

FIELD 2: Peroperative management

Question: In ACHD patients, does locoregional anesthesia in non-cardiac surgery help to reduce perioperative mortality in comparison with general anesthesia?

Experts: Stéphane LE BEL, Xavier ALACOQUE

Table 6
Composite risk according to type of congenital heart disease, physiological status, and risk associated with the operation. Following a proposal issued by the expert group.

CARDIOPATHY / SURGERY	Minor risk	Intermediate risk				Severe risk
Low risk		A	B	C	D	
Intermediate risk		A	B	C	D	
High risk		A	B	C	D	

Low composite risk
Intermediate composite risk
High composite risk

A, B, C, D: physiological status (Cf Table 3).
*A procedure using peripheral locoregional anesthesia is considered as having a low composite score, whatever the heart disease status.

R2.1.1 – In ACHD patients in non-cardiac surgery, the experts suggest locoregional rather than general anaesthesia whenever possible, the objective being to reduce perioperative morbimortality.

R2.1.2 – When choosing a neuraxial locoregional anaesthesia technique, the experts suggest the use of titrated or continuous neuraxial anaesthesia rather than the application of a non-titrated technique, the objective being to reduce perioperative mortality in ACHD.

Expert opinion (Strong agreement)

Argumentation: In univariate analysis, a retrospective single-center study on non-cardiac surgery conducted by Egbe et al. and involving patients with Fontan circulation highlighted a link between intermediate or deep sedation and occurrence of perioperative or postoperative complications (HR 2.01; CI95% [1.22–3.62]; $p = 0.02$). After pairing a cohort of ACHD patients with biventricular physiology and a cohort of patients without cardiopathy, moderate or deep sedation remained associated in the Fontan group with increased incidence of perioperative complications, but the difference between the two groups was not significant (HR 1.98; CI95% [0.92–2.86]; $p = 0.06$) [17]. Published in 2002, a systematic review by Martin et al. dealt with perioperative mortality in non-cardiac surgery among Eisenmenger syndrome patients, and even though the authors highlighted greater mortality among patients having received generally as opposed to neuraxial anaesthesia (18 *vs.* 5%), the difference was not significant [24]. Moreover, no study has demonstrated increased risk due to the administration of local anaesthetics during locoregional anaesthesia in CHD patients [25]. Above and beyond the limited literature, what should probably be privileged is an anaesthesia strategy aimed at reducing hemodynamic and ventilatory variations, which are of crucial importance in CHD management. Peripheral LRA rather than GA, and titrated or continuous neuraxial LRA rather than non-titrated LRA, are probably to be prioritised.

Question: In ACDH patients undergoing non-cardiac surgery, does an anesthetic strategy (equipment, specific monitoring) help to reduce the risks of perioperative mortality?

Experts: Catherine KOFFEL, Xavier ALACOQUE, Loïc MACE, Mirela BOJAN, Bernard CHOLLEY

R2.2 – The experts suggest that the anaesthesia strategy to adapted to the specific risk of each heart disease, the objective being to reduce perioperative morbimortality:

- **Standard monitoring for procedures with low composite risk;**
- **Continuous and invasive monitoring of arterial pressure and regular measurement of $PaO_2$and $PaCO_2$for procedures entailing intermediate or high risk;**
- **Addition of continuous central venous pressure in the event of Fontan circulation (cf.** Table 6 **and fact sheet #5).**

Expert opinion (Strong agreement)

Argumentation: Large-scale retrospective series have not revealed excess perioperative mortality in CHD patients at low risk as compared to a control population [13,15]. While routine perioperative monitoring recommended by the relevant learned societies remains applicable, there are limits and traps peculiar to the CHD population. Special attention must be accorded to the management of venous access (thromboembolic risk) and to the technical difficulties that may be encountered during implantation of a central venous or invasive arterial approach. [3,26]. Most perioperative complications in ACHD patients not undergoing cardiac surgery occur following periprocedural anaesthesia; they consist mainly in hypotension and hypoxemia, which in CHD cases are integrally linked. It is consequently primordial to closely monitor these parameters in patients at intermediate to high composite risk [17,18]. The study by Maxwell et *al.* of files from the American national register on medical complaints concerning non-cardiac anaesthesia in ACHD patients showed that perioperative monitoring (or its absence) was criticized in close to 10% of the files, often because an arterial catheter had not been inserted [18].

FIELD 3: Postoperative management

Question: In ACHD patients undergoing non-cardiac surgery, does postoperative surveillance in critical care help to reduce postoperative complications?

Experts: Diane ZLOTNIK, Mirela BOJAN

R3.1 – The experts suggest that ACHD patients with an intermediate or high composite score be systematically postoperatively managed in a critical care unit for non-cardiac surgery, the objective being to reduce the risks of postoperative morbimortality.

Expert opinion (Strong agreement)

Argumentation: The data in the complaint registry published by Maxwell et *al.* demonstrate that in the context of non-cardiac surgery, 60% of the complications presented by ACHD occur during the postoperative period [18]. Moreover, the literature underlines the need to ensure follow-up in intensive care and close monitoring of the invasive arterial blood pressure of ''Fontan'' patients with preoperative LVEF $< 30\%$ [14] or preoperative cyanosis [18], the main risk factors for postoperative complications.

Question: Following non-cardiac surgery in ACHD patients, does consulting with congenital heart disease specialists help to reduce risks of morbimortality?

Experts: Caroline OVAERT, Pascal AMEDRO

R3.2 – The experts suggest taking the advice of a congenital heart disease specialist following non-cardiac surgery to reduce morbimortality:

- **For patients at intermediate or high composite risk (Cf.** Table 6**);**
- **In the event of a complication, whatever the degree of severity;**
- **In the event of emergency surgery without preliminary specialised cardiological advice.**

Expert opinion (Strong agreement)

Argumentation: There exists no study in the literature specifically dealing with this question, and no relevant recommendations have been formulated. Issued in 2022, the most recent European Society of Cardiology (ESC) guidelines do not take up modalities of postoperative surveillance of CHD patients having undergone a non-cardiac surgical procedure [11]. The AHA 2018 guidelines [10] insist on the fact that even low-risk operations can entail higher risks for ACHD patients, in whom major postoperative physiological variations can occur (water balance, vasoplegia, hypoxemia, etc.). The guidelines stipulate that ACHD patients presenting with an altered physiological state (physiological status B, C, or D; Cf. Table 3) should receive programmed surgery in a hospital containing an M3C expert center. A list of ''problems to consider'' also appears in these recommendations. In the event of an emer-

gency or surgery exterior to a CHD unit, these recommendations underline the need to be able to ''communicate'' with a specialised CHD team. Given that postoperative mortality is associated with the management of complications [12], specialised cardiological advice can no doubt help to optimise postoperative management, especially when the patient is hospitalised in a non-specialised center.

Field 4: Obstetric management

Question: In ACHD patients during the peripartum period, does prepartum cardiopathy assessment help to reduce peripartum morbimortality?

Experts: Magalie LADOUCEUR, Marie BRUYERE, Estelle MORAU

R4.1 – The experts suggest that at the outset of pregnancy, all pregnant CHD patients undergo congenital cardiopathy evaluation in an expert center concerning their physiological state and possible heart disease complications, the objective being to reduce peripartum morbimortality.

Expert opinion (Strong agreement)

Argumentation: There exist no studies highlighting a direct link between prepartum CHD evaluation and peripartum maternal morbimortality. The risk of maternal morbidity or mortality essentially depends on the type of congenital cardiopathy, severity of residual lesions, and ventricular function [27,28]. Mortality in the 2019 edition of the International Registry of Pregnancy And Cardiac Disease (ROPAC) was 0.2% in the congenital cardiac group, while cardiac insufficiency was found to occur in 13% of patients with severe congenital cardiopathy and 5% of patients with mild to moderate congenital cardiopathy [29]. The determinants of maternal complication were cardiac insufficiency before pregnancy or NYHA score > II, systemic ventricular ejection fraction <40%, modified WHO class 4, and anticoagulant utilisation. Several scores have been developed to assess maternal risk in cardiopathy patients, and some have shown a significant association between maternal risk of cardiovascular complications during the peripartum stage (CARPREG [30], CARPREG II [31], the ZAHARA risk score [32], and the modified WHO score proposed by the 2018 European guidelines [33]). These scores integrate several factors in cardiovascular evaluation: anterior cardiac events or arrhythmia episodes, cardiovascular treatment before pregnancy, NYHA class > II or cyanosis, a high-risk valvular disease or left ventricular outflow tract obstruction, systemic ventricular dysfunction, mechanical valve, high-risk aortopathy, pulmonary hypertension, and coronaropathy. Moreover, the CARPREG II score integrates belated management (>20 weeks of amenorrhea) as a predictive factor, suggesting a need for cardiopathy assessment during the early stages of pregnancy. Even though it remains somewhat imprecise, the modified WHO classification [34,35] seems most apt to predict cardiovascular risk in pregnant women with congenital cardiopathy [34–36], and it can serve as a baseline in preoperative assessment of these patients [33]. The type (*i.e.*, consultation, echocardiography, stress test, catheterization) and frequency of cardiovascular assessment will be indicated by a cardiologist specialised in CHD and based on anatomy, physiology, estimated maternal risk, and specific management of congenital cardiopathy [37–42].

Question: In pregnant chronic heart disease patients, does management in an expert center help to reduce complications during the peripartum stage?

Experts: Marie BRUYERE, Estelle MORAU, Magalie LADOUCEUR

R4.2 – The experts suggest that pregnant chronic heart disease patients with an intermediate or high composite score be managed during the peripartum stage in an expert center to the incidence of peripartum complications.

Expert opinion (Strong agreement)

Argumentation: Several elements in the organisation of care seem to decrease the morbidity of high-risk pregnancies: a center with many childbirths, the performance of more than 200 Caesarean sections a year, a university hospital, high caregiver/patient ratio [43,44]. Even though severe maternal morbidity is particularly high in centers managing many high-risk pregnancies, association with a high number of childbirths reduces the risk [45,46]. There presently exist no French guidelines on the organisation of maternity wards according to the level of maternal risk. The North American recommendations organise birthing centers regionally according to the level (from 1 to 4) of maternal care. Material and human resources as well as team expertise are indicated for each level of care [10]. Studies on the monitoring of pregnant women with congenital heart disease are retrospective and cover long periods [44,46,47]; all of them agree on the benefits afforded by an expert center, which is defined by the presence of a multidisciplinary team composed, at the very least, of a cardiologist, an obstetrician and an anaesthetist with expertise in the management of pregnant women with congenital cardiopathy [47–49]. The 2018 European guidelines insist on the importance of a ''Pregnancy Heart Team'' from the pre-conceptional stage to 6 months postpartum (strong recommendation, low level of evidence). As regards PAHT patients, the enactment of a personalised peripartum ''multidisciplinary management plan'' led to drastically reduced postpartum mortality [50]. It preliminarily determined factors of vigilance and anaesthetic administration (type of monitoring, type of anaesthesia, choice of vasopressor drugs...) for the day of delivery. This type of organisation of care has been highlighted as a means of achieving favourable outcomes in very high-risk patients [51]. Even though these different arguments are based on retrospective studies, expert opinions converge in their recommendations to refer the patient, starting at the pre-conceptional phase, to a multidisciplinary center with expertise in congenital cardiopathy to optimally prepare and anticipate complications during the peripartum phase.

Question: In ACHD patients during the peripartum stage, does the anesthetic strategy help to reduce the risks of peripartum morbimortality?

Experts: Estelle MORAU, Marie BRUYERE, Magalie LADOUCEUR

R4.3 - The experts suggest that pregnant ACHD patients should be administered titrated locoregional anaesthesia rather than non-titrated or general anaesthesia, the objective being to reduce peripartum morbimortality.

Expert opinion (Strong agreement)

Argumentation: As regards to anaesthesia management, the ROPAC cohort does not provide applicable data; moreover, there exists no meta-analysis or randomised study comparing anaesthesia protocols in this specific patient population. In a retrospective cohort of congenital cardiopathy patients giving birth by Caesarean section, Tsukinaga et *al.* found no difference in the occurrence of cardiovascular events according to whether the parturient population received locoregional or general anaesthesia [52]. In a more targeted population of PAH patients, LRA seems to be a protective factor regarding postpartum cardiovascular events [50,53] and maternal hemodynamic parameters [54]. However, details on the LRA protocols applied are seldom provided. Even though there exists no formal proof in the literature concerning the efficacy of titrated LRA, physiology and the known hemodynamic effects of neuraxial LRA and general anaesthesia have induced experts to recommend it given ensure improved hemodynamic stabil-

ity and reducing peripartum morbimortality in a parturient population suffering from CHD.

Question: In ACHD patients, does postpartum surveillance in critical care help to reduce postpartum complications?

Experts: Experts: Estelle MORAU, Marie BRUYERE, Magalie LADOUCEUR

R4.4 — The experts suggest systematic surveillance in a critical care unit during the postpartum stage of ACHD patients with an intermediate or high composite score, the objective being to reduce postpartum mortality.

Expert opinion (Strong agreement)

Argumentation: The immediate postpartum phase is a period entailing hemodynamic risk, marked by frequent variations in blood volume: positive due to relief from the vena cava compression syndrome and blood redistribution following childbirth; negative in the event of excessive blood loss. In ACHD, the peripartum period puts patients at a relatively high risk of decompensation. The two most widely reported decompensations in the ROPAC cohort are heart failure (5–13% of cases) and arrhythmia (from 1 to 3%). In patients at high cardiovascular risk (mWHO IV suffering from PAHT, severe systemic ventricular dysfunction, or severe aortic or mitral valvopathy [33]), heart failure occurred in 33% of cases. In this cohort, the exact moment of decompensation was not reported. In a retrospective cohort of 197 pregnancies between 2011 and 2020, Ornaghi *et al.* observed that cardiac decompensation occurred during the postpartum period in 65% of cases [55]. In a more targeted population of 227 pulmonary arterial hypertension patients from 2008 to 2018, Low *et al.* reported the most elevated risk of mortality as occurring between day 0 and day 4 postpartum [56]. There is no meta-analysis or randomised study in the literature comparing the evolution of CHD patients according to whether they were or were not systematically hospitalised in critical care after giving birth. That said, the elements of clinical and paraclinical surveillance expected by these patients (heart rate, respiratory rate, blood pressure, diuresis, cardiopulmonary auscultation, ECG tracing, fatigue, dyspnea, oedema, cardiac enzyme assay, monitoring of specific therapeutic measures. . .) necessitate adequate equipment and trained and available staff. The present-day organisation of French maternity wards and patient/caregiver ratios seldom allow for close specialized monitoring. Referral to critical care is probably beneficial to patients with a high or intermediate composite score. (*Cf.* Table 6).

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Conflicts of interests of the SFAR experts during the five years preceding the date of validation by the SFAR board of directors

Nadir Tafer: No competing interest that could compromise his independence regarding the present guidelines.

Elise Langouet: No competing interest that could compromise his independence regarding the current guidelines.

Xavier Alacoque: No competing interests that could compromise his independence regarding the present guidelines.

Pascal Amedro: No competing interest that could compromise his independence regarding the present guidelines.

Miréla Bojan: No competing interest that could compromise his independence regarding the present guidelines.

Marie Bruyère: No competing interest that could compromise his independence regarding the present guidelines.

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Hugues de Courson: No competing interest that could compromise his independence regarding the present guidelines.

Marc-Olivier Fischer: No competing interest that could compromise his independence regarding the present guidelines.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.accpm.2025.101540.

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