

Challenges of congenital heart disease in grown-up patients

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Summary

Nowadays, more than 90% of all children born with congenital heart disease (CHD) reach adult life. Although initially considered to be cured, the majority of them continue to need specialised follow-up because they require redo interventions or are at increased risk of cardiovascular complications and premature death. Arrhythmias are the most common cause of unscheduled hospital visits for grown-up CHD (GUCH) patients, accounting for one third of emergency admissions in these patients. Some GUCH patients are also at increased risk for sudden cardiac death. The principles of arrhythmia management and the prevention of sudden cardiac death in GUCH patients are similar to those used in adults with acquired heart disease, but are not evidence based. Decompensated heart failure is the other leading cause of death. Conventional medical heart-failure therapy for left ventricular dysfunction is not effective in GUCH patients at highest risk of heart failure, i.e., those with right or single ventricular failure. Careful haemodynamic assessment and structural interventions are the first step to consider in GUCH patients presenting with heart failure symptoms. Adults with moderate or complex CHD and regular follow-up in specialised GUCH centres have a survival benefit compared with patients without such follow-up. Cardiac surgery in GUCH patients should be performed by surgeons trained in treatment of CHD, i.e., surgeons also operating on paediatric patients. A structured transition programme with a defined transfer of care from the paediatric to the adult care environment is important to avoid lapses of care in today's adolescents with CHD. For GUCH patients with an intervention performed decades ago and no specific cardiac follow-up in later life, referral to a specialised GUCH centre is recommended and may save lives.

Key words: congenital heart disease, arrhythmia, stroke, infective endocarditis, heart failure, pregnancy, transition, advanced care planning, specialized care

Introduction

Cardiac defects are the most common birth defect and affect approximately 0.8% of newborns [1]. In Switzerland,

every year approximately 700 children are born with congenital heart disease (CHD). Half of these children have simple defects (e.g., a ventricular septal defect), the other half have moderately complex (e.g., coarctation of the aorta) or complex (e.g., transposition of the great arteries) defects. Many of the simple defects close spontaneously during the first years of life, but eventually 200 to 300 of the infants with CHD will require an intervention.

Nowadays, more than 90% of children with repaired CHD survive into adulthood [2]. The majority of them enter adult life with the need for a lifelong specialised follow-up. In the current era, the number of grown-up CHD (GUCH) patients has outgrown the paediatric CHD cohort: two out of three CHD patients are adults [3]. With further reductions in mortality resulting from advances in diagnostic methods, cardiac surgery, catheter interventions and electrophysiology, the prevalence of GUCH patients will continue to increase. It is estimated that currently 2.5 million adults with CHD are living in Europe, of whom 20 000 in Switzerland [4]. The following review article highlights historical milestones in CHD and the resulting challenges GUCH patients have to face during their lifetime.

Historical background

The medical success story of CHD was strongly related to pioneering work in cardiac surgery, and started with a palliation procedure. In 1945, the John Hopkins' surgeon Alfred Blalock performed a shunt operation in a 15-month-old child with cyanotic heart disease and reduced pulmonary blood flow. He sutured the left subclavian artery end-to-side on the left pulmonary artery (fig. 1), with the aim to increase pulmonary blood flow without the risk of inducing shunt-related pulmonary hypertension. This procedure, later called the Blalock-Taussig shunt, reduced CHD mortality from 23 to 17% in older cyanotic children [5]. In 1952, palliative procedures were succeeded by intracardiac surgery. James Lewis closed an atrial septal defect in a 5-year-old girl, using a special clamp technique to avoid exsanguination of the beating heart. In 1955, John W. Kirklin at the Mayo Clinic in Rochester reported the successful use of a heart-lung machine and heralded the era of open-heart surgery with use of extracorporeal circula-

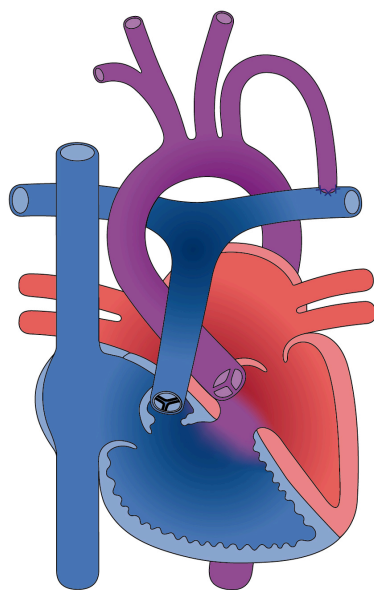
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tion. However, the reported mortality for intracardiac correction of tetralogy of Fallot, the most common cyanotic defect, was 39% with the heart-lung machine, substantially higher than the reported 17% mortality after palliation with a Blalock-Taussig shunt [6]. The policy was adopted to initially palliate complex defects by a shunt or banding procedure with the aim to gain time and let the infants and their cardiovascular structures grow, and thereby delay the repair of the cardiac defect into early childhood (secondary repair).

During the 1970s, a new intraoperative strategy was established, consisting of cooling the patient to about 20°C on the heart-lung machine, with subsequent cardiac arrest during the repair operation, followed by rewarming prior to weaning from the bypass. With this strategy, correction of almost all cardiac anomalies was progressively mastered. Further advances in surgical techniques and perioperative care allowed a stepwise abandonment of secondary repair strategies for primary repair, without the need for an initial palliation procedure. Even the most complex lesions, such as a heart with a single ventricle due to hypoplasia of one of the ventricles, can nowadays be approached (fig. 2) [7]. Contemporary mortality in the first year of life for children with non-severe CHD is now 2 to 4%, with an operative mortality of 0 to 2%. For children with severe CHD, the corresponding mortality is 8 to 13%, with an operative mortality of 4 to 6% [8]. Figure 3 depicts the remarkable increase in survival rates with CHD over the past 70 years.

Figure 1: Blalock-Taussig-Shunt. Classic Blalock-Taussig shunt for palliation of cyanotic heart disease due to reduced pulmonary blood flow. The subclavian artery is sacrificed and increases blood flow in the corresponding pulmonary artery, thereby increasing systemic oxygen saturation. Illustration taken from <http://www.chd-diagrams.com>. Creative Commons License Illustrations are licensed under Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License by the New Media Center of the University of Basel

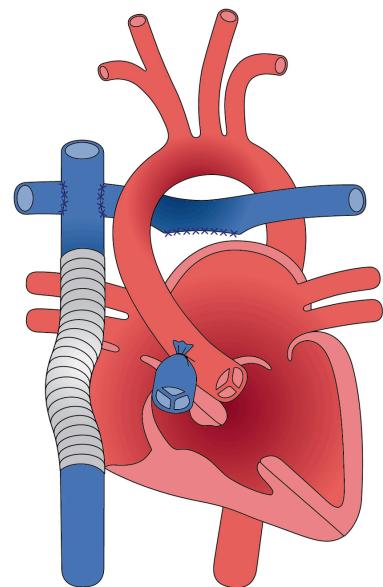


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Repaired does not mean cured

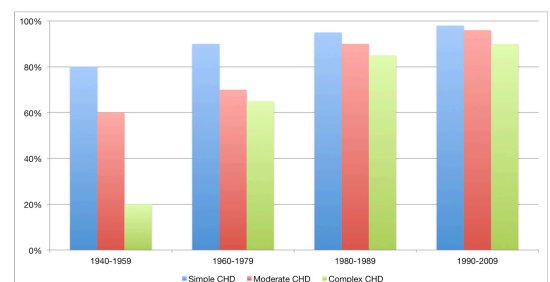
Although originally assumed to be curative, it was realised over time that most interventions to treat CHD allowed these children to grow up without severe physical limitations, but with an – initially unexpected – increased long-term morbidity and mortality, manifesting once these patients were in the midst of their adult life [10]. Figure 4 shows some of the factors contributing to the cardiovascular burden observed in GUCH patients. Table 1 lists com-

Figure 2: Fontan physiology. Modern type of Fontan procedure (total cavopulmonary anastomosis with an extracardiac conduit) for palliation of univentricular physiology, i.e. tricuspid atresia. Derouting of systemic venous return directly to the pulmonary artery avoids mixing of de-oxygenated and oxygenated blood in the functional single ventricle, and unloads the heart. The original concept was introduced by Fontan in 1971. Illustration taken from <http://www.chd-diagrams.com>. Creative Commons License Illustrations are licensed under Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License by the New Media Center of the University of Basel.



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Figure 3: Survival rates (first year) with congenital heart disease (CHD) over different decades. The most remarkable increase in survival was observed in children with complex CHD, from 20% for children born in the 1950s to 90% for children born in the past 20 years [2, 9].



mon heart defects and the expected complications in adult life.

The increased morbidity and mortality risk is reflected by a steadily growing number of re-hospitalisations within the adult CHD population, out of proportion to the pure increase in the population size over time [11]. Padrutt et al. recently presented numbers for GUCH patients followed up at the University Hospital Zurich: from 2005 to 2015, the number of adults under specialised care doubled, and the frequency of patients requiring hospital admission (e.g., for cardioversion) increased from 3.1 to 5.4% per year, a relative increase of 75% [12].

The main reasons for hospital admissions in adult CHD patients are supraventricular arrhythmias, lesion-specific surgery or percutaneous interventions, decompensated heart failure, ventricular tachycardias, the need for a pacemaker, an intracardiac defibrillator (ICD) or other elec-

trophysiology procedures, pregnancy or delivery, evaluation and treatment for pulmonary hypertension, infective endocarditis and perioperative care for noncardiac surgery [13, 14]. In addition, within the aging CHD population, acquired morbidities (e.g., coronary artery disease, renal failure, diabetes, liver dysfunction, endocrinological and haematological disorders or cancer) are becoming important outcome factors in conjunction with the underlying congenital heart defect and add to the medical burden of these patients [15]. Some of today's adult patients also face complications related to limited diagnostic possibilities in the past. Screening blood products for the presence of hepatitis C virus became routine only in 1992. Adults with congenital heart surgery who received blood transfusions before 1992 have a 5- to 20-fold increased prevalence of hepatitis C infection [16].

Specialised care saves lives

Based on experience and outcome data, specialised care by trained cardiologists of GUCH patients has been endorsed and recommended by different national and international societies within the last 20 years [4, 17, 18]. Minimal standards of care for GUCH patients in Switzerland are listed in table 2.

Mylotte et al. observed a decline in GUCH mortality in a province-wide database of Québec, Canada after the publication of the first Canadian GUCH guidelines in 1997, which emphasised the importance of specialised care for adults with moderate or complex cardiac defects [19]. In a case-control analysis, referral to a specialised GUCH centre was associated with lower risk of death (odds ratio [OR] 0.82, 95% confidence interval [CI] 0.68–0.97), independent of comorbidities, age and gender. Survival improvement was highest for patients with complex defects. Several explanations are provided: the level of training and experience of the healthcare providers in GUCH centres, a

Figure 4: Factors contributing to the cardiovascular burden over their lifetime in grown-up congenital heart disease (GUCH) patients. Prior to the repair procedure or intervention, the haemodynamic consequences of the defect and hypoxaemia in cases of cyanotic conditions pose an increased load on one or both ventricles. Peri-interventional damage and residual lesions or sequelae contribute to the cardiovascular burden after repair. Over time, aging and the emergence of acquired heart disease are an additional source of increasing cardiovascular morbidity.

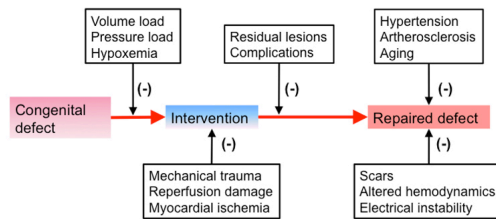


Table 1: Late complications in common congenital cardiac defects.

Congenital heart defect type	Possible complications / sequelae
1. Simple defects	
Isolated small or repaired ventricular septal defect	Arrhythmia, endocarditis
Small or repaired secundum atrial septal defect	Arrhythmia
Mild pulmonary stenosis	Endocarditis
Bicuspid aortic valve	Endocarditis, ascending aortic dilation, valvular dysfunction
2. Moderately complex defects	
Atrioventricular septal defects (native or repaired)	Arrhythmia, atrioventricular block, endocarditis, heart failure, pulmonary hypertension
Large unrepaired secundum atrial septal defect or sinus venosus defect	Arrhythmia, pulmonary hypertension, heart failure
Aortic coarctation (native or repaired)	Systemic hypertension, local pseudoaneurysm or re-stenosis, cerebral aneurysm
Unrepaired anomalous pulmonary venous drainage	Arrhythmia, pulmonary hypertension, heart failure
Repaired sinus venosus defect	Arrhythmia, pulmonary hypertension
Ebstein's anomaly (native or repaired)	Arrhythmia, heart failure, endocarditis
Repaired tetralogy of Fallot	Arrhythmia, heart failure, endocarditis
3. Complex defects	
Implanted conduits (e.g., RV-PA conduit)	Arrhythmia, endocarditis, heart failure
Repaired truncus arteriosus	Arrhythmia, endocarditis, heart failure, pulmonary hypertension
Transposition of the great arteries	
– After atrial switch procedure	Arrhythmia, sinus node dysfunction, heart failure, pulmonary hypertension
– After arterial switch procedure	Ostial coronary occlusion, right ventricular outflow tract obstruction, ascending aorta dilatation, heart failure,
– Congenitally corrected transposition	Arrhythmia, atrioventricular block, heart failure
Cyanotic heart condition, including Eisenmenger syndrome	Arrhythmia, heart failure, endocarditis, paradoxical embolism / stroke, pulmonary hypertension, gout, renal failure
Single ventricle after Fontan procedure	Heart failure, arrhythmia, liver disease, protein losing enteropathy, varicosis

higher likelihood of use of practice guidelines and therefore less variability in care, the maximised use of medical advances, and the availability of congenital cardiac surgeons. There is growing evidence that substantially lower mortality rates (1.8 vs 4.8%) are achieved when a congenital heart surgery team is operating on GUCH patients, i.e., a cardiac surgeon who also operates on children, and not a “general” cardiac surgeon [20]. Currently, the University Hospitals of Lausanne/Geneva, Bern and Zurich provide the optimal surgical environment to operate on GUCH patients in Switzerland. Cardiac surgery in GUCH patients is characterised by a wide anatomical-clinical spectrum, frequent multiorgan involvement, and the absence of evidence-based data [21]. In contrast to cardiac surgery for acquired heart disease, there is no validated surgical risk score for GUCH patients [22]. All these factors enforce the need for meticulous preoperative planning and preparation of surgical interventions in GUCH patients, by a multidisciplinary team with experience in CHD.

Transition and patient education

CHD patients and their families need to be aware of the importance of uninterrupted specialised care. This requires education and a structured transition and transfer process in adolescence [23]. Studies on transfer destinations have shown that without a structured transition, up to 75% of adolescents with CHD are lost to follow-up once they reached adult life [24]. This has important implications: adults with CHD and a lapse of care are more likely to be symptomatic at the time they are readmitted to a hospital (OR 2.5, 95% CI 1–6), are more likely to have new haemodynamic problems or an additional cardiac diagnosis (OR 9.6, 95% CI 4–23), and have a 3-fold (95% CI 2–7) greater likelihood of requiring urgent cardiac interventions [25]. Multiple efforts are necessary to keep patients on board [26]. Protective and risk factors for care gaps in GUCH patients are presented in table 3. Organised transition programmes providing education on the heart defect, on healthy lifestyle, and coping and self-management strategies are emphasised, [27]. A transition coordinator is recommended to facilitate the transition process,

Table 2: Minimal standards of care for adolescents and adults with congenital heart disease.

Transition	– Transition process begins in teenage years – Structured transfer from paediatric to adult care centre
Regular follow-up	– Every non-simple repaired or unrepaired defect should be presented once in a GUCH centre – Individualised but lifelong regular follow-up is mandatory for all non-simple lesions
Pregnancy	– Prepregnancy counselling for all women with CHD – High-risk pregnancy managed by a multidisciplinary team at GUCH centre
Cardiac interventions	– Expertise of a congenital cardiac surgeon or congenital invasive cardiologist mandatory
Non-cardiac surgery	– Cyanotic patients, patients with a systemic right ventricle or with single ventricle physiology need referral to specialised centre for even minor surgery
CHD = congenital heart disease; GUCH = grown-up CHD patients	

and nurses are predestined for this position. Fifteen to 60 min educational sessions led by nurses have shown to increase patient’s CHD knowledge [28, 29].

Common cardiovascular problems in GUCH patients

The need for lifelong medical follow-up is related to the common cardiovascular complications seen in in GUCH patients.

Arrhythmias and stroke

More than half of all GUCH patients with complex CHD will develop arrhythmias by the age 65 years [30]. Arrhythmias are the most common cause of unscheduled hospital visits in adults with CHD and account for one third of all emergency admissions in this population [31]. All types of cardiac arrhythmias are prevalent, ranging from clinically occult arrhythmias, such as silent atrial fibrillation, to potentially fatal ventricular tachycardia. Table 4 illustrates the frequency of arrhythmia disturbances in different congenital defects. Residual anatomic or haemodynamic lesions and surgical scars from atrio- or ventriculotomies form the substrate for arrhythmias or dyssynchrony [32]. GUCH patients are routinely advised to consult their primary physician in the event of persistent palpitations or a sudden decrease in exercise capacity, in order to rule out arrhythmias with a routine 12-lead ECG or Holter ECG.

Table 3: Predictors of care gaps in congenital heart disease patients.

Protective factors	Risk factors
– Higher family income – ≥ 1 comorbid condition – Having a written referral to a specialist GUCH centre – Keeping first and second GUCH appointment – Patient’s belief that follow-up should be performed at specialised clinic – No substance use – Compliance with antibiotic prophylaxis – Greater independence in attending appointments	– Living independently – Male gender – Lower family income – Travel distance to GUCH care – Fewer outpatient visits in paediatric care – The last visit taking place outside a university hospital – Childhood hospitalisations – History of at least one missed appointment – Mild disease severity – No previous heart surgery
GUCH = grown-up congenital heart disease patients	

Table 4: Frequency of rhythm problems in GUCH patients.

Cardiac defect	SVT	VT	SND	AVB	DYS
Ventricular septal defect					
Secundum type atrial septal defect					
Primum type atrial septal defect					
Aortic coarctation					
Ebstein’s anomaly					
Tetralogy of Fallot					
TGA with an atrial switch procedure					
Single ventricle after Fontan palliation					

AVB = atrioventricular block; DYS = intraventricular dyssynchrony; SND = sinus node dysfunction; SVT = supraventricular tachycardia; TGA = transposition of the great arteries; VT = ventricular tachycardia. The shaded pattern ranges from minimal (no shading) to mild (light grey), moderate (medium grey), and high (dark grey) risk. Adapted from Khairy et al. [32].

A particular challenge in CHD patients with supraventricular arrhythmias is the decision when to start oral anticoagulation. Atrial fibrillation and intra-atrial re-entry tachycardia are the most common arrhythmias, and are associated with thrombosis and embolic events [33]. Among GUCH patients, the stroke incidence is 9 to 12 times higher than in the general population, particularly in younger adults (below age 55 years). Important predictors of ischaemic stroke, besides the absence of sinus rhythm, are the severity of the congenital defect, active infective endocarditis, the presence of endovenous pacemaker leads, heart failure, diabetes mellitus, and a recent history of myocardial infarction [33, 34]. The CHA₂DS₂-VASc score has proven benefits in identifying patients with nonvalvular atrial fibrillation at increased risk for stroke, but has a limited ability in GUCH patients [35]. Anticoagulation is recommended by experts in all GUCH patients with atrial fibrillation or intra-atrial re-entrant tachycardia and a history of previous intracardiac repair, cyanosis, after a Fontan palliation or in the presence of a systemic right ventricle, independent of the CHA₂DS₂ VASc score [36].

Sudden cardiac death accounts for 15 to 25% of fatalities in GUCH patients [12, 37]. The incidence of sudden cardiac death is in the range of 1 per 1000 patient-years, and similar to the incidence of sustained ventricular tachycardia in these adults [38]. Intracardiac defibrillators (ICDs) are indicated in CHD adults for secondary prevention after resuscitation from sudden cardiac death and in those with spontaneous sustained ventricular tachycardia, after a careful workup has failed to identify a clear reversible cause. Selecting candidates for primary prevention is challenging. In general, ICDs are recommended in patients with a systemic left ventricular ejection fraction $\leq 35\%$, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms [38]. They may also be considered in GUCH patients with a systemic right ventricular ejection fraction $\leq 35\%$ and NYHA class III or IV, if additional risk factors are present, such as a history of syncope, or QRS duration >140 ms [39].

Heart failure

Beside sudden cardiac death, heart failure is the other leading cause of death in GUCH patients. Together, they account for nearly half of all late mortality in mixed cohorts of children and adults [40, 41]. Mechanisms for heart failure in CHD are often lesion specific and may involve different pathophysiological pathways (table 5). Recognition of heart failure symptoms can be delayed in GUCH patients because many have lived their entire life with the notion of a limited exercise capacity, do not perceive changes over time and underreport symptoms [42].

The Dutch CONCOR (CONgenital CORvitia Dutch national Registry) database reported an incidence of heart failure admission in GUCH patients of 1.2 per 1000 patient-years, which was 10 times higher than the incidence in the general population of adults of the same age [43]. GUCH patients admitted for heart failure had a 5-fold higher mortality than CHD patients without heart failure history, and the 1- and 3-year mortality after first heart failure admission were 24% and 35%, respectively. GUCH patients at high risk for heart failure are the ones with cardiac defects affecting the right ventricle, i.e. corrected tetralogy of Fallot, transposition of the great arteries with the right

ventricle in systemic position, Ebstein's anomaly, or with a single ventricle physiology.

Whereas medical heart failure treatment with beta-blockers, angiotensin converting-enzyme inhibitors or angiotensin receptor blockers, and aldosterone antagonists have proven benefits in left ventricular heart failure, the role of these agents has not been established in GUCH patients with right or single ventricular dysfunction [44]. Whereas neurohormonal modulation is the cornerstone of heart failure treatment in classic left heart failure patients and invasive haemodynamic assessment is reserved for patients with refractory heart failure, the reverse is true for most CHD diagnoses: careful haemodynamic assessment and structural interventions are the first step to consider in GUCH patients presenting with cardiac dysfunction [45]. Cardiac resynchronisation therapy has been used in a few CHD patients, but data from small studies in heterogeneous CHD populations could not clearly determine which GUCH patients will benefit, irrespective of anatomical difficulties for implantation [42]. Currently, the only established therapy for end-stage heart failure in CHD patients without pulmonary hypertension is heart transplantation [46]. Although early post-transplant mortality is increased in CHD patients, the 10-year survival is similar to non-CHD transplant recipients [47]. Ventricular assist devices can be used as a bridge to transplant [48]. Given the ubiquitous shortage of donor organs and the number of GUCH patients at risk, further medical options are currently being investigated in adults with a systemic right ventricle [49].

Infective endocarditis

CHD patients are at increased lifetime risk of infective endocarditis [50]. The risk varies with the underlying defect, but on average 1 in 1000 CHD patients suffers an episode of infective endocarditis per year [51]. This incidence is 20 to 30 times higher than the incidence observed in the general population (3–5 cases per 100 000 patients per year). In contemporary GUCH cohorts, the acute mortality of infective endocarditis is still 16%, and 1-year mortality is 19% [52]. Adults with repaired or palliated pulmonary atresia and adults with corrected congenital transposition of the great arteries have the highest incidence of infective endocarditis with 5.8 (95% CI 1.8–9.8) and 2.3 (95% CI 0.5–4.1) cases per 1000 patient-years, fol-

Table 5: Pathophysiology of heart failure in grown-up congenital heart disease patients.

Mechanism	Example
Volume overload	Long-sustained regurgitation of native or repaired/replaced valves, shunt lesions (residual shunt, late diagnosis)
Pressure overload	Valve or conduit stenosis, coarctation, peripheral pulmonary artery stenosis, systemic or pulmonary hypertension
Ventricular anatomy	Systemic right ventricle, functionally single ventricle
Myocardial injury	Scars, cyanotic heart disease, limited myocardial protection during bypass, myocardial fibrosis
Coronary artery supply	Coronary artery anomalies, coronary artery disease
Myocardial architecture	Ventricular non-compaction
Ventricular dyssynchrony	Complete bundle-branch block after surgery, intractable arrhythmias

lowed by adults with a single ventricle, a bicuspid aortic valve, and a ventricular septal defect. The risk is very low in adults with a pulmonary stenosis or an atrial septal defect, and almost non-existent in adults with a repaired or still patent arterial duct. Besides the underlying cardiac anatomy, the presence of prosthetic material also confers a high risk of infective endocarditis. Among 14 000 GUCH patients, biological or mechanical valves were independently associated with infective endocarditis, in the short and long term after implantation (0–6 months: hazard ratio [HR] 17.29, 95% CI 7.34–40.70; 6–12 months: HR 15.91, 95% CI 6.76–37.45; after 12 months: HR 5.26, 95% CI 3.52–7.86), whereas valve repair carried only an increased risk in the first 6 months following valve procedure (HR 3.34, 95% CI 1.33–8.41), but not thereafter [52].

Interestingly, infective endocarditis occurs less frequently in women (odds ratio [OR] 0.5, 95% CI 0.4–0.7), independent of the underlying defect or other risk factors [53]. Evidence supports a relationship between dental hygiene and the risk of developing endocarditis-associated bacteraemia after daily events such as tooth-brushing [54]. A large Swiss population study has shown that men are less likely to practice good dental hygiene than women [55]. It was shown that female gender and better disease knowledge are strongly related to good dental hygiene in GUCH patients [56], reinforcing the concept that patients' education plays an important role in endocarditis prevention, as well as dental care. According to the current guidelines, optimal oral health is considered more important for infective endocarditis prevention than prophylactic antibiotics for dental procedures [57]. We discourage the empiric use of antibiotic treatments for suspected bacterial infections without evident focus, unless blood-cultures have been obtained prior to starting antibiotic therapy.

GUCH and pregnancy

Many women with CHD are in childbearing age and consider pregnancy [58]. From a cardiac point of view, pregnancy is a prolonged proarrhythmic and prothrombotic state with increased cardiac output and decreased peripheral resistance. The majority of women with CHD, normal biventricular function and no severe outflow tract obstruction tolerate pregnancy well. However, an impaired contractile function (e.g., a systemic right ventricle), a fixed obstruction (e.g., aortic coarctation, mitral stenosis, severe pulmonary hypertension), mechanical valves, aortic dilatation and residual cyanotic heart disease are conditions with an increased risk for maternal cardiovascular complications during pregnancy and delivery [59]. Extremely high risks of maternal mortality or morbidity are encountered in women with pulmonary arterial hypertension of any cause, severe systemic ventricular dysfunction with an ejection fraction <30% or in functional NYHA class III or IV, severe symptomatic mitral or aortic stenosis, native severe coarctation, Marfan syndrome with an ascending aortic diameter >45 mm or a bicuspid aortic valve with an ascending aortic diameter >50 mm, and in women with a history of peripartum cardiomyopathy and residual impairment of left ventricular function. If pregnancy occurs in these situations, termination should be discussed. A multidisciplinary team of cardiologists, obstetricians and anaesthesiologists should monitor CHD patients before and during

pregnancy, delivery and the postpartum period. After delivery, cardiac output may take up to 6 months to return to prepregnancy levels. Individual preconception counselling of women with CHD concerning contraception, pregnancy risks and inheritance is prerequisite to an informed decision.

End-of-life discussions and advanced care planning

Death is not predictable in most CHD patients, and heart failure symptoms may manifest late in the disease trajectory, making early end-of-life discussions indispensable. This includes the completion of advanced directives, palliative care and resuscitation options, and identifying an attorney [60]. As in many other disciplines, advanced care planning (ACP) discussions are rarely held with GUCH patients. In a Canadian GUCH centre, out of 200 patients only 2 (1%) recalled having discussed ACP, whereas 24 of 48 (50%) healthcare providers indicated that they discussed life expectancy, ACP and resuscitation preferences regularly with their patients [61]. Patients' preferences for ACP did not depend on the CHD complexity, but healthcare providers stated that they discussed ACP more often with patients with complex lesions. Similar results were found in a recent survey of an American GUCH centre [62]. Patients favour ACP discussions prior to a life-threatening illness, but the physicians' focus on life, rather on death, hindered a timely initiation [63]. Adult CHD poses many challenges, not only to patients, but also to their treating physicians.

Disclosure statement

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