REVIEW

Key issues of daily life in adults with congenital heart disease

Questions de la vie quotidienne des patients adultes ayant une cardiopathie congénitale

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Summary Increasing survival rates of patients with congenital heart disease have resulted in a new and growing patient population of adults with operated congenital heart disease. Medical professionals face the specific medical needs of these patients but must also deal with their daily life issues. Adult patients with congenital heart disease report difficulties in several areas of daily life, such as sport, employment, insurability and travel or driving. Moreover, they must have a healthy lifestyle to prevent cardiovascular complications. All these issues can be addressed in a specific educational program. In this review, we discuss the different daily life issues of adults with congenital heart disease and the preventive measures that can be proposed to improve their quality of life.
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Abbreviations: ACHD, adult congenital heart disease; CAD, coronary artery disease; CHD, congenital heart disease; TQTH, Reconnaissance Qualité Travailleur Handicapé; VO2, oxygen uptake.
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Introduction

The improvement in surgical techniques over the last decades has led to a dramatic increase in survival rates of patients with congenital heart disease (CHD). This success has resulted in the emergence of a growing population of adults with operated CHD. Medical professionals are currently managing the specific medical needs of these patients but have to deal concomitantly with their daily life issues. A minority of adults with mild CHD but a majority of those with complex CHD report difficulties in their daily life [1], with an impact on quality of life [2,3]. It is of note that a high proportion of these patients have an inadequate level of knowledge about their disease and their potential in a variety of areas [4,5]. Patients with complex CHD frequently felt that the direct limitation to their participation in sport, employment or education was their heart disease. Knowledge about reproduction issues, 'family planning' and risk related to pregnancy was found to be poor among women with CHD [6]. An additional difficulty is access of these patients to life insurance and loan applications [7]. Together with these issues, adult patients with CHD should have preventive attitudes in supplementary domains to prevent common cardiovascular complications and endocarditis. Acquiring autonomy to tackle all these aspects of daily life with a CHD is a complex process for adolescents and young adults. The development of a specific transition/transfer program with counseling and educational content related to these issues is crucial for this population. Hopefully, developing these programs should lead to a reduction in complications and an improvement in quality of life.

In this review, we will discuss the key daily life issues for patients with adult congenital heart disease (ACHD). These issues are sport and more general physical activities, employment, family planning, risky behaviors, traveling and driving. We will also analyze the proposals that have been made in the literature for managing these difficulties.

Physical activity

Exercise capacity is a major issue in ACHD that has a significant impact on daily activity and quality of life. Additionally, physical activity has a direct link with cardiac status and a variety of risks, such as arrhythmias or syncope. A recent review by Kempny et al. [8] aggregated the available data on exercise capacity in ACHD comparing cardiopulmonary exercise test results in ACHD in their institution with the data published by other centres. This study confirmed that exercise capacity differs significantly across the spectrum of ACHD. The practical clinical consequence of these findings is that improving exercise capacity through aerobic training is probably worthwhile in virtually all ACHD patients. Kempny et al. [8] proposed different occupations according to peak oxygen uptake (VO2) results derived from their study. These findings could help greatly in choosing/prescribing recreational activities when counseling a patient with ACHD.

Today, it is widely recognized that physical activity has long-term beneficial effects not only on quality of life but also on long-term morbidity and overall mortality [9,10]. As an example, exercise training in chronic heart failure reduces heart failure-related events, including mortality and hospital admission for worsening of heart failure [10,11]. Data remain scarce on the feasibility and efficacy of exercise training programs in ACHD patients [12]. Still, all these programs were safe and they all demonstrated an increase in exercise capacity or an improvement in quality of life [13–17] (Table 1), despite the controversial link between quality of life and exercise capacity. Indeed, self-estimated physical functioning, evaluated using quality of life scales, poorly predicts actual exercise capacity [18]. It is worthwhile noting that even patients with pulmonary hypertension, who have the poorest VO2, experienced improvement in exercise endurance, symptomatic status and quality of life after carefully designed exercise training [19,20]. The limited compliance of the patients was the main limitation in these studies that described physical training. Indeed, Swan et al. showed that only one third of ACHD patients regularly practiced at least moderate exercise and one third had no physical activity despite their mean age being 26 years [21]. To be efficient on outcomes, these programs have to be developed over a long period of time. To be sustainable, the programs need patient reinforcement and are therefore necessarily time consuming. Patient education on the short- and long-term benefits of exercise must be part of ACHD management. Recommendations regarding physical training, exercise and sport...
should begin in early adolescence (Fig. 1). It is important that the patient and their family are informed that exercise restriction is very rarely indicated. Obviously, safety is a major concern when prescribing exercise and athletic activities for young adults with cardiac disease, especially those with hypertrophic cardiomyopathy, coronary artery anomalies, Marfan syndrome and aortic valve disease, which have all been associated with sudden cardiac death. For major ACHD groups, recommendations are provided by the American Heart Association [22,23]. Certainly, these guidelines

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Training programs evaluated in patients with congenital heart disease.</th>
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<tr>
<td><strong>Population</strong></td>
<td><strong>Training</strong></td>
</tr>
<tr>
<td>Fifty ACHD patients</td>
<td>Walk 5/7 days for 10 weeks</td>
</tr>
<tr>
<td>Nine adults with ToF</td>
<td>Individual program three times/week for 3 months</td>
</tr>
<tr>
<td>Eighteen children with CHD</td>
<td>12 weeks</td>
</tr>
<tr>
<td>Thirty adults with PAH</td>
<td>Daily structured exercise for 15 weeks</td>
</tr>
</tbody>
</table>

ACHD: adult congenital heart disease; CHD: congenital heart disease; CPET: cardiopulmonary exercise test; PAH: pulmonary artery hypertension; QoL: quality of life; ToF: tetralogy of Fallot; VO₂: oxygen uptake.

**Figure 1.** Classification of sport from Sable et al. [68], based on American Heart Association recommendations [23], with kind permission from Wolters Kluwer Health, 2012.
should be adapted to individual status. To guide the exercise prescription and the choice of recreational activities, a cardiopulmonary exercise test may provide valuable information on the level/type of activity that is appropriate for each patient. Self-monitoring of physical activity through diaries and the use of simple devices such as accelerometers [13] may also enhance awareness and motivation.

Education, employment and career

A recent multicentre study [24] compared a large sample of adult patients with various types of CHD with a reference group, with regard to sociodemographic characteristics and lifestyle. The study showed that young ACHD patients (aged <40 years) had poorer outcomes in educational attainment, employment and relationships compared with the reference group. This difference was found not only for moderate-to-severe defects but also for mild defects. These poorer outcomes were more pronounced in men, as in former studies [25–28]. Actually, there could be discrimination against job applicants when they declare they have a CHD. Certainly, complex CHDs have become more prevalent than in the past — when patients did not survive until adulthood — and are more often associated with disability; their impact on employment might be consequentially more visible. While Kamphuis et al. [25] and Ternestedt et al. [29] found no differences in employment status for ACHD patients compared with a reference group, the latter study did indeed find a clear difference between mild and severe defects, with better employment status for those with mild defects. Another cause for these differences in employment might be educational attainment, which Van Rijen et al. [28] reported to be lower in young ACHD patients compared with a reference group. Absence from school because of illness, need for hospitalization, recovery from invasive procedures and learning disabilities/neurocognitive defects could explain these poorer outcomes in patients with CHD [30,31].

In 2005, Crossland et al. [32] reported that 33% of ACHD patients were unemployed (16% in matched controls) and that 25% had been unemployed for more than 1 year (3% in matched controls). In this study, only one fifth of ACHD patients received counseling regarding their career, while this counseling was more frequent in controls (31%). In 42% of ACHD patients, this counseling aimed to exclude some inappropriate occupations. Receiving career counseling was significantly associated with increased employment in the ACHD population, with 73% of ACHD patients given advice being employed compared with 46% of those not given advice. This pattern was not seen in controls. ACHD patients are less likely to receive useful advice regarding potential careers and find the advice given less helpful than controls, although receiving suitable advice can be considered efficient in improving the proportion of ACHD patients who get a job. Many patients were advised not to apply for specific jobs. This advice was often given by medical staff not trained in this field or by career advisors not trained to give advice based on the medical condition of the patient.

A variety of solutions have been proposed to improve the employment status of ACHD patients; the most straightforward seems to be helpful, positive, medically appropriate advice concerning potential careers. Guidelines based on CHD diagnosis are far from being perfect. This is particularly true for those lesions considered to be significant, as there is a wide variation in cardiac status within each headline diagnosis [33]. Kempny et al. [8] compared the required peak VO2 values for a professional activity with the distribution of oxygen uptake expected for a given CHD condition. These results could assist clinicians in deciding how likely it is that young patients with CHD may be able to cope with the physical demands of a job as a career. Individualized discussion with the patient, involving both the cardiologist and the career advisor, preferably at a single clinic, seems to be more appropriate. Counselling the patient and their family on this delicate issue during the patient’s early adolescence facilitates appropriate career planning and overcomes unrealistic expectations.

In the study by Crossland et al. [32], more than half of patients with complex ACHD experienced problems in their careers. Physical problems were frequently reported as a specific reason for quitting a job. The most important specific adaptations mentioned concerned working time and workload. Career counseling and use of job adjustment, taking into account physical abilities, are important in the ACHD population to prevent unemployment. In France, the ‘Labour Code’ recognizes the quality of disabled worker (reconnaissance qualité travailleur handicapé [RQTH]). When a patient applies, there is a hearing at the maison départementale des personnes handicapées. This procedure can be followed by an ACHD patient who is seeking a job or by an employed patient who requires an RQTH or an RQTH renewal. This labelling can be useful in ACHD patients by improving access to work or keeping them in their job.

Family planning

Sexual activity is an important component of quality of life for men and women with cardiovascular disease and their partners. The American Heart Association provides guidelines [34] and recommends that physicians and other healthcare professionals have dialogue with patients on this topic. Reported deaths or strokes during sexual activity in the ACHD population are rare. In a study by Vigu et al. [35], 9% of women with ACHD reported symptoms during sexual activity, which included dyspnoea, perceived arrhythmia, increased fatigue or syncope. Symptoms were more common in patients with severe lesions, worse functional status or cyanosis. In another survey of men with ACHD, 9% reported dyspnoea or subjective arrhythmias and 5% reported chest pain during sexual activity. Again, symptoms were more common in patients with greater functional impairment (New York Heart Association class III) [36]. According to the American Heart Association guidelines, sexual activity is safe in most patients with ACHD [34]. Patients in whom the safety of sexual activity is less certain or unclear are those with significant pulmonary hypertension, cyanotic heart disease, severe left-sided heart outflow obstruction, uncontrolled arrhythmias and anomalous coronary artery passing between the pulmonary artery and aorta [34].

Contraception and pregnancy have now become important issues in this population. Both can, however, be
associated with increased risks in women with ACHD [37,38]. Other issues, such as adverse foetal outcomes and recurrence of CHD in offspring, must be addressed. Current guidelines recommend proactive counseling about contraception and pregnancy [39]. Of note, information regarding the risks of contraception and pregnancy is currently ignored by women with ACHD [6] Overall, sexual activity in adolescents with CHD and ACHD is lower than in matched controls [40] but women are more sexually active than men. This also emphasizes the importance of starting an education program on birth control and pregnancy-associated risks very early in adolescence in female patients with CHD. Poor family functioning seems to promote risky sexual behavior [40]. In education programs on birth control, these risks must be detected in adolescents. Specialist cardiologists or gynecologists must insist on their prevention and check patient knowledge on this topic.

A high proportion of women with ACHD have limited knowledge of the appropriate contraceptive method they should use and the risk of pregnancy to their own health [5,35]. In these studies [6,35], 43–49% of the women had not been counseled about contraception, 20% used contraceptive methods that were contraindicated for their specific cardiac condition and 28% in the group with high pregnancy-associated risks were not using contraception despite having a sexual relationship [35]. For the group of women considered to be at intermediate or high risk of complications during pregnancy, 37–48% did not recall receiving this information. Further, only half of women with contraindications to pregnancy recalled having received this information [6]. Accurate information on contraception and the implications of CHD on pregnancy should be given through an education program starting during adolescence (Table 2). There are several models to address this issue, including a collaborative approach between ACHD and contraception clinics, or the addition of contraception and pregnancy counseling by advanced practice nurses within ACHD clinics.

**Insurability**

We recently showed that patients with intermediate ACHD prognosis have the same refusal rate for insurance at standard rates than those with 'complex' ACHD [41]. Others studies observed that ACHD patients were significantly more likely to have difficulty in obtaining life insurance or a mortgage than controls, regardless of the severity of the ACHD [7]. However, prognosis varies within individual lesions, particularly in 'intermediate' ACHDs, such as tetralogy of Fallot. Recent advances in the treatment of ACHD have improved the prognosis of this group, which tends to be similar to that of control populations [42]. Therefore, it is exceedingly important that physicians emphasize in their letters for insurance purposes the presence of positive diagnostic factors and the absence of negative prognostic factors. Decisions by loan companies are often made without contacting the supervising cardiologist and the premium is probably based on a diagnosis rather than details of the hemodynamic status of an individual patient [41]. This attitude will particularly penalize patients with 'intermediate-risk’ CHD. Patients declined by one company or those offered insurance at high premiums should seek another company. Another possibility is to seek advice from a CHD patient association, which may be able to provide advice about insurance companies with a track record of providing coverage. In our own study, age did not affect insurability [41]. The "S’Assurer et Emprunter avec un Risque Aggravé de Santé” convention aims to expand access to loans for people who have or have had a severe disease. Despite this rule, patients with health risks still encounter difficulties in insurability. This is particularly true for disability that is not adequately covered.

**Cardiovascular risk factors and cardiac risky behaviors**

**Cardiovascular risk**

The prevalence of significant coronary artery disease (CAD) in the adult CHD cohort is estimated at between 7.6% and 9.2% [43,44], which is similar to that in patients of the same age without CHD. While the overall prevalence is similar, the relative risk of developing CAD is linked to the type of CHD. Indeed, systemic arterial hypertension is a strong predictor of CAD in ACHD [44] and is more frequently associated with aortic coarctation [45]. Conversely, cyanotic ACHDs have a lower risk of atherosclerosis [46-48]. Factors possibly contributing to the lower incidence of CAD in cyanotic ACHD are low cholesterol concentrations, increased bioavailability of nitric oxide, hyperbilirubinaemia and low platelet count [43]. In transposition of the great arteries after arterial switch procedure, proximal eccentric intimal thickening [49] and reduced coronary flow reserve [50] have been observed. These observations suggest that early atherosclerosis may develop in the transferred coronary arteries. Whether this will lead to late coronary events is uncertain, as the patients who underwent this surgical repair are aged <30 years. Recent studies also showed an increased carotid intima-media thickness [51,52] and significantly higher triglyceride and lower high-density lipoprotein concentrations in obese patients who had the arterial switch operation [53]. These factors may confer additional risk for future cardiovascular events in patients who underwent coronary artery reimplantation in infancy and who then may require more intensive education on 'classical' cardiovascular prevention.

A few studies have reported outcomes on classical cardiovascular risk factors in the ACHD population. Smoking prevalence varies according to the study: from 18%, which is similar to the English national average of 22% [43], to 10.9% in the Netherlands [24], which is significantly lower than the reference value of 16.4%. This last study also showed that ACHD patients were participating in sport more frequently and were less overweight than their age-matched peers; curiously, ACHD patients presented more often with diabetes [24,53]. Higher serum glucose concentrations were also found in CHD patients compared with in non-congenital patients [54].

The importance of controlling cardiovascular risk factors in ACHD has been emphasized by a recent epidemiological study demonstrating that in the current era, the main cause of death in non-cyanotic ACHD patients is myocardial
infarction [55]. Overall, primary prevention of CAD and screening for traditional cardiovascular risk factors in ACHD must be performed as they are routinely in the non-CHD population. A Canadian study demonstrated that patients with only elementary education were less able to recall important cardiovascular risk factors [56]. Education of ACHD patients is crucial in their management. Thus, CHD patients with elementary education might need a more intensive program of education concerning cardiac prevention.

### Mental health and risky behaviors

With regard to the emotional and social functioning (leisure-time activities), the sample showed favorable results [28], with better scores than in a reference group for hostility, self-esteem and neuroticism. Another study showed an increased level of psychological stress [57] in these patients and an increase in depression or anxiety in adult [58] and adolescent [59] populations with CHD. These findings highlight the need for clinical assessment and treatment by mental health professionals, if needed.

Drugs use is significantly lower in the ACHD population [19,60]. Body art in the form of tattoos and piercings has become increasingly popular among children and teenagers and is nowadays more socially acceptable despite the known infection risk. It is estimated that millions of people have been pierced or tattooed [61]. These cosmetic procedures are considered to be safe with a low risk of serious infection. Serious infections such as endocarditis in CHD were reported in a few studies [62,63] in which 17.7—43.0% of patients with CHD pierced their ears and 3% received a tattoo. Many patients (approximately 25%) experienced local skin infections after ear piercing [62] and only one out of 87 developed endocarditis. In parallel, Shebani et al. [63] reported 28 cases of endocarditis after piercing declared by cardiologist consultants. Although endocarditis after body art is rare, education concerning infectious risk in CHD is required. In our own practice, we strongly discourage all forms of body art except for ear piercing if there is careful monitoring after the procedure. Better knowledge and education about the link between body art and endocarditis is required in order to provide guidelines for doctors and patients.

### Traveling and driving

#### Traveling

Physicians are frequently asked by patients with chronic heart disease whether they can safely travel. Questions usually relate to commercial aircraft travel or visiting locations at higher altitude. Patients with cyanotic CHD, like healthy people, experience a fall in arterial oxygen saturation of about 8% from baseline but tolerate this well without supplemental oxygen; they maintain adequate tissue oxygen delivery due to the chronic rightward shift in their oxyhaemoglobin dissociation curve and to secondary erythrocytosis [64,65].

Deep venous thrombosis occurs in up to 10% of long-haul (>8 hours) airline travelers [66]. Additional preventive measures may be considered for individuals judged at increased risk of thrombosis, such as patients with cyanosis. Preventive measures are properly fitted below-knee stockings and heparin.

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**Table 2 Recommendations for contraception in females with congenital heart disease.**

<table>
<thead>
<tr>
<th>Defect/residua</th>
<th>COCs</th>
<th>Mini Pill</th>
<th>Norplant</th>
<th>Depo-Provera</th>
<th>IUD</th>
<th>Barrier</th>
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<tbody>
<tr>
<td><strong>Surgically-repaired defects</strong></td>
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<tr>
<td>No residua: ASD/VSD/PDA</td>
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<td>+</td>
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<tr>
<td>Residual shunt and/or obstruction</td>
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<td>+</td>
<td>+</td>
<td>+</td>
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<td>+</td>
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<tr>
<td>Prosthetic valves, conduits, baffles</td>
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<td>+</td>
<td>+</td>
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<td>+</td>
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<tr>
<td>Residual pulmonary and/or systemic</td>
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<td>+</td>
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<td>+</td>
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<tr>
<td>hypertension</td>
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<td><strong>Unrepaired defects, postoperative residua</strong></td>
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<tr>
<td>Small VSD</td>
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<tr>
<td>Mild-to-moderate residual shunts (ASD, VSD, PDA)</td>
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<td>+</td>
<td>+</td>
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<td>+</td>
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<tr>
<td>Residual systemic or pulmonary hypertension (coarctation of aorta)</td>
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<td>+</td>
<td>+</td>
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<td>+</td>
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<tr>
<td>Complex cyanotic defects (TA, SV, TR)</td>
<td>—</td>
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Driving

Driving a motorized vehicle is an inherently dangerous activity with associated significant mortality and morbidity. Societies have determined that driving privileges should be restricted in people who are likely to place themselves and others at unacceptable risk. Those who drive large trucks, large passenger-carrying vehicles and smaller passenger-carrying vehicles, such as taxis, must conform to stricter standards than those who drive a small personal vehicle. The medical communities have published guidelines for patients with arrhythmia that may affect consciousness [67]. These recommendations cover issues most likely to be seen in the adult with CHD. The overriding concern is that a medical problem may increase the individual’s risk when driving due to a sudden loss of consciousness or significant alteration of mental awareness.

Conclusion

ACHD patients are concerned by many daily life issues. Management of these daily life issues aims to improve quality of life and probably survival. Although some of these problems are more actively handled by ACHD physicians, the increasing prevalence of ACHD patients will raise new questions in the very near future. Sharing of knowledge on these topics between the adult congenital cardiologist, the patient and their environment is of utmost importance, as ignorance of some of these risks may have severe medical consequences. Developing education programs dedicated to these daily life issues is definitively a path to follow. To be efficient, these types of programs should start early in adolescence and be followed continuously through reinforcement and personalized sessions in adulthood. Interdisciplinarity is also crucial in this field and the educational team should probably include specialist nurses, psychologists, gynecologists, sports doctors, educational counselors and teachers. Hopefully, adding these concerns to the current management of ACHD and providing adequate counseling should prevent some serious complications and potentially improve quality of life for this growing population.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


Lifestyle in adults with congenital heart disease


