Progress in Pediatric Cardiology xxx (2012) xxx–xxx

Contents lists available at SciVerse ScienceDirect

Progress in Pediatric Cardiology

journal homepage: www.elsevier.com/locate/ppedcard

Congenital heart disease clinics — How to keep the adult patients on board

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ARTICLE INFO

Available online xxxx

Keywords:
Congenital heart disease
Long-term prognosis
Transition
Follow-up

ABSTRACT

The steadily growing number of adults with congenital heart disease has led with some delay to an increase of specialized programs dedicated to the care of these patients. Still, in many countries the prevalence of congenital heart disease in adults is not known, nor the number of adults requiring specialized care. By extrapolating the results of epidemiological studies on the population in Europe and North America, only half or even less of all adults with moderate or complex congenital heart disease is currently under follow-up in specialized centers. Lapse of appropriate medical care in adults with congenital heart disease is associated with unnecessary morbidity and an increased likelihood of the need for urgent interventions. In order to improve medical care for adults with moderate or complex congenital heart disease, the number of patients with specialized follow-up in congenital cardiac clinics has to increase. One approach is to avoid lapses of care in patients already followed by a pediatric or adult congenital heart disease center. Therefore, congenital heart disease clinics should try to identify among their patients the ones at risk for lost to follow-up, and focus some of their efforts on measures to increase the awareness for life-long medical care among these patients. Characteristics of the adult congenital cardiac patient at high risk of inappropriate medical follow-up include having few interventions in early life despite a complex cardiac defect, being born in an earlier decade, receiving no specific follow-up recommendations, and having no education about defect- or surgery-related sequelae or residual lesions. Implementation of a structured transition program, teaching patients about their heart defect and the consequences of repair or palliation in earlier life, establishing a continuity of care within the congenital cardiac clinic, optimizing organization and communication between different care-givers, and encouraging patients to play an active role regarding their personal health care, are some of the recommendations on how we should try to avoid another generation of adult congenital heart disease patients from being lost to follow-up.

1. Introduction

The prevalence of congenital heart disease at birth has not substantially changed over the past 30 to 40 years [1]. New diagnostic tools, such as the development and widespread use of echocardiography, improved the detection rate of minor defects and led to an apparent increase of congenital heart disease in infants in the 1970s and 1980s [2]. These days, a prevalence of 8 children with congenital heart disease per 1000 live births is an often quoted estimate [3]. Several population-based studies from North America and Europe have documented a decreasing mortality of congenital heart disease in children and adults in the past decades [4–6]. Currently, 9 of 10 children with congenital heart disease have the prospect to become an adult, and today adults with congenital heart disease have the prospect to live longer than previous generations of adults with cardiac defects. From 1999 to 2006, the mortality from congenital heart disease in the United States decreased 3% annually among adults 18 to 34 years of age, 3–4% among adults 35 to 49 years of age, and 4–5% among adults 50 to 64 years of age. Mortality due to congenital heart disease has a bimodal distribution: it affects primarily the infant, and the adult patient [4].

Specialized life-long care is emphasized for adults with moderate or complex congenital heart disease [7,8]. The steadily growing number of adults with congenital heart disease has led with some delay to an increase of specialized programs dedicated to the care of these patients. In Europe, the number of programs increased from 1 in 1964 to more than 70 in 2007 [9]. There are on average 1500 patients in active follow-up per center, caring for an estimated 130,000 adults with congenital heart disease throughout Europe. As in many other countries, we do not know how many adults with congenital cardiac defects actually live in Europe, or how many of them have moderate or complex congenital heart disease and qualify for specialized care.

Based on the data from Quebec for the year 2000 [10], the current prevalence of congenital heart disease is expected to exceed 4 per 1000 adults, with 1 of 10 patients having a complex or severe lesion. In 2008, the combined population of the European Union was at 500 million, 77% of them aged 19 or older. Based on these numbers,

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http://dx.doi.org/10.1016/j.ppedcard.2012.08.010

1.8 million adults with congenital heart disease are supposed to live in Europe, and 180,000 have severe or complex cardiac defects. Assuming that the proportion of adults with severe congenital heart disease is 30 to 40% [11] in specialized congenital cardiac centers, still more than half of all adults with complex congenital heart disease in Europe are not seen in specialized programs, and this percentage is probably even lower in adults with moderate lesions.

As Meijboom and Mulder recently pointed out, one can question the validity of extrapolating Canadian numbers to estimate the size of an adult congenital heart disease population in other countries [12]. Socioeconomic differences in the past, differences in access to surgery and surgical techniques, may all have had an influence on the long term outcome and hence the currently living number of adults with congenital heart disease in Europe and elsewhere. However, it is hard to imagine that differences in medical care between Europe and North America 30 to 40 years ago can sufficiently explain the gap between the expected number of adult congenital heart disease patients in Europe and the number of patients currently seen in the programs. This leaves us with questions about the amount and type of care an important part of adult congenital patients currently receive. Why are some patients not followed by specialized programs? When and why were they lost to follow-up? And how can congenital heart disease clinics keep future and present patients on board of established programs, as recommended by several guidelines?

2. Lost to follow-up

In 2009, Mackie et al. reported that out of 643 children born in 1983 in the province of Quebec, diagnosed with congenital heart disease before 6 years of age, and still alive at 22 years of age, overall 60% did not receive cardiac follow-up after the 18th birthday [13]. Between ages 6 and 12, 28% of children with congenital heart disease failed to receive continuous cardiac care. Another 19% of 643 children were lost between ages 12 and 17, and finally 94 young adults, or 15% of the initial study population, did not have specialized follow-up after age 18. More than 1 out of 5 young adults with severe congenital heart disease was not seen by a cardiologist during the 5 year period from age 18 to 22. The number of adults with complex lesion and no specialized cardiac follow-up is likely even more alarming, as the authors of this study could not distinguish between a follow-up visit at a general cardiologist's office or a follow-up visit at a specialized congenital cardiac center. Similar numbers were published by the German Heart Center in Munich in 2005 [14]. In their registry, 76% of 10,000 adults with congenital heart disease failed to return for a scheduled follow-up visit in a 5 year period. Seven out of 10 adults with a lapse of follow-up declared that they had had no medical care at all. The ones with at least occasional medical visits were usually seen by general practitioners (25%) or internists (42%).

Both studies confirm the initially raised suspicion that half or even more of all adults with moderate or complex congenital heart disease are currently not seen in specialized cardiac programs. They are either completely lost to follow-up or are seen by non-specialized cardiologists or general practitioners. These studies do not provide answers as to why these patients were lost. No access to specialized care, no intrinsic motivation, no understanding that life-long specialized follow-up may be needed despite the absence of cardiac symptoms, are all potential but also modifiable reasons. The easiest way to improve long-term follow-up in adults with congenital heart disease is to avoid a lapse of care in patients already followed by a center. In our experience, it is more challenging to bring adult congenital heart disease patients with no specialized medical follow-up back to established programs, than to keep current patients on board. Therefore, congenital cardiac centers should identify among their patients the ones at risk for being lost to follow-up, and focus some of their efforts on measures to increase the awareness for life-long adequate care among all congenital heart disease patients.

3. Predictors of lapse of care and the consequences

Reid et al. investigated the issue of going lost to follow-up among 360 young adults with complex congenital heart disease in Canada [15]. This study aimed to identify predictors of a successful transfer of care from a tertiary pediatric cardiology program to a specialized adult program within Canada. Another analysis was published in 2008 by Yeung et al., summarizing the experience of the Adult Congenital Cardiac Clinic in Denver [16]. In both studies, the percentage of adults lost to follow-up after transfer of care form the pediatric to the adult setting was high (53–63%). The numbers were similar to the results of the previously presented studies from Quebec [13] and Munich [14]. In a more recent study of 794 adolescents with congenital heart disease in Belgium, only 14% did not have an adequate transfer of care and did not receive specialist care at the time of adulthood [17]. Using a matched case–control design, Mackie et al. compared clinical and personal characteristics in 74 adults with congenital heart disease and no follow-up in the past 3 years, and in 222 controls with a similar complex defect and ongoing specialized follow-up [18]. Few interventions in early life despite a complex cardiac defect, being born in an earlier decade, no specific follow-up recommendations, and having no education about defect- or surgery-related sequelae or residual lesions, all characterize the adult congenital heart disease patient at risk of inappropriate medical follow-up (see Table 1). These characteristics apply for example to the asymptomatic adult with an atrial switch procedure for complete transposition of the great arteries or with repaired tetralogy of Fallot, or to some degree to the adult with repaired coarctation and no apparent symptoms of hypertension or aortic valve issues. The studies indicate that in the past, limited attention was paid to inform young adults with moderate or complex congenital heart disease about the needs for life-long appropriate medical care, and the Belgian example illustrates that lessons have been learned.

Hospital admission rates are 2 to 3 times higher in adult congenital heart disease patients compared to the general population, and particularly in congenital heart disease patients older than 40 years [19,20]. One in 4 adults with congenital heart disease is likely to be admitted to a hospital at least once during a five year period, and 2 out of 5 hospitalizations will be admissions via the emergency department, with arrhythmias, heart failure, coronary artery disease, pulmonary hypertension or infective endocarditis as the predominant cardiac problem [21]. We do not know how many of these emergency admissions can be prevented by a continuous specialized follow-up. We know, however, that adults with congenital heart disease and a lapse of care are more likely to be symptomatic at the time they are re-admitted to a hospital (OR 2.5, 95% CI 1–6), are more likely to have new hemodynamic problems or an additional cardiac diagnosis (OR 9.6, 95% CI 4–23), and have a 3 times (95% CI 2–7) greater likelihood of requiring urgent cardiac interventions [16]. With this in mind, timely recognition of evolving or persistent hemodynamic problems in adult congenital heart disease patients due to an appropriate medical follow-up may be life saving.

4. How to keep patients on board of existing programs

In medical and nursing science literature, some recommendations consistently emerge on how the number of lost adults with congenital heart disease may be reduced. None of these recommendations fulfills the stringent criteria of evidence-based medicine, but they have proven to be useful in building up an adult congenital heart disease clinic and are also derived from the experiences of well-established programs. These recommendations encompass mainly the transition process and address organizational issues.

4.1. Implement a transition program

The transition process should prepare adolescents to assume responsibility for their own health, and this necessarily includes medical

Table 1
Predictors of continuous care or lapse of follow-up.

Predictor variables	Favoring continuous follow-up	Favoring lost to follow-up
Very strong predictors (OR >5)		
Recent intervention	Catheterization in the past 5 years; OR 5 (95% CI 1.7–10) [18]	
Unhealthy lifestyle		Substance use: smoking, binge drinking, marijuana use or any illegal drug use; OR 5.6 (95% CI 2–14.3) [15]
Independence	Attending appointments without parents; OR 6.6 (95% CI 1.6–27.0) [15]	Living independently from parents; OR 4.1 (95% CI 1.7–10.1) in univariate analysis [16]
Strong predictors (OR >2–5)		
Number of cardiovascular interventions	≥2 interventions; OR 2.5 (95% CI 1.4–4.4) [15]	
Complexity of heart defect		No prior heart surgery; OR 3.3 (95% CI 1.9–5.8) Moderately complex lesion vs. complex lesion; OR 2.8 (95% CI 1.2–6.7) in [16] OR 1.6 (95% CI 1.0–2.6) in [17]
Follow-up recommendations	Advise to have follow-up in specialized congenital clinics (vs. no details, or general adult cardiologist); OR 3.6 (95% CI 1.7–7.7) in [15] OR 2.5 (95% CI 1.3–5) in [18]	
Medical knowledge	Awareness of the needs for dental antibiotic prophylaxis; OR 4.2 (95% CI1.5–12.0) [15]	
Weak predictors (OR 1–2)		
Age at last pediatric visit	Increasing age in 1-year intervals from age 13; OR 1.3 (95% CI 1.1–1.5) [15]	
Gender		Male gender; OR 1.6 (95% CI 1–2.6) [17]
Patient's age		Older age (per year); OR 1.1 (95% CI 1.0–1.1) in univariate analysis [16]
Family income	Higher income (per $10,000 CAD increase); OR 1.1 (95% CI 1–1.3) [18]	

education and information about the anatomical and functional aspects of the heart defect and the needs for ongoing specialized medical care throughout adulthood [8]. One important aspect of the transition process is to inform patients with moderate or complex lesions, that a surgical scar and the absence of symptoms cannot be interpreted as being "cured" and do not discharge them from further follow-up [22]. In addition, these adults need to know that only few adult cardiologists are trained in congenital heart disease, and most general cardiologists will not be very familiar with their specific cardiac problems. These 2 core messages should be communicated during the transition process to patients and parents [22].

The transition process is usually initiated by the pediatric health care team, but it is important that the adult congenital cardiologist does not avoid responsibility. Transition consists of collaboration, and it can be helpful for the pediatric cardiologists if the adult team provides them with written information (e.g. in form of a leaflet) about the adult congenital team, the localization of the adult care facilities, and the likely more "business-like" atmosphere patients may encounter after the transfer of care. An instructive example of such a leaflet designed by the University of Birmingham can be accessed online at http://adc.bmj.com/content/92/10/927/suppl/DC1 [23]. Further detailed information on how to design a transition program for adolescents with congenital heart disease can be found at the same link and in other comprehensive reviews [8,23–25]. Further recommendations based on this literature are summarized in Table 2.

For most adolescents, leaving pediatric care is a logical step [26]. They recognize and accept "cultural gaps" between pediatric and adult services. Nevertheless, adolescents and their parents call for a better organization of the adult congenital service, and for more communication between the different health-care providers [26]. Joint medical consultations or transition clinics are probably the most evident form of optimized communication and alignment between a pediatric and adult service. Unfortunately, the limited resources often preclude such a close and resource intensive collaboration. Regular consultations in the form of transition meetings between pediatric and the adult congenital cardiology staff about patients to be transferred are another way to improve the transition process.

Table 2
Recommendations to improve patients' adherence.

Implement a transition program
– Design a structured program in collaboration with the pediatric care team
– Prepare patient and parents for differences between pediatric and adult follow-up
– Encourage young adults to independent behavior and to assume personal responsibility
– Inform patients and parents about the need for life-long cardiac follow-up
– Inform patients and parents about the need for specialized care
– Referral notes from the pediatric to the adult program should include specific recommendations about the transfer of care, and about the cardiologist in charge during transfer period
– Organize regular consultations between pediatric and adult congenital cardiology about patients to be transferred
– Provide feedback on transferred patients to the pediatric cardiology unit
– Consider using the same medical records, hospital information system, and database as your pediatric colleagues
Organizational issues
– Keep track of your patients or of patients in the transition process by using a database
– Discuss the necessity and timing of further follow-up visits at the end of every appointment
– Send repeatedly outpatient visit reminders to patients not showing up for a scheduled visit
– Send copies of your medical reports to the patient
– Establish continuity of care
– Make your service easy to get
– Encourage patients to become a member of patients' organization
– Establish a close collaboration with referring family physicians, cardiologists, other hospitals and in-hospital services

4.2. Organizational issues

In our experience, it is helpful to have a regularly updated database for keeping track of adults with congenital heart disease and adolescents in the transition process. This database can also be used to organize outpatient clinics. Even if we encourage patients to play an active role regarding their health-care issues and to take over responsibility, it is not advisable to arrange follow-up appointments only by referral or by the patient's initiative. Discussing with the adult congenital heart disease patient at the end of a visit the need for further follow-up and fixing the date of the next appointment in advance have been proven to work well. If some patients fail to present for a scheduled visit, we re-contact them up to 3 times. If a patient believes that no further follow-up is necessary despite being told otherwise, we try to inform them about our rationale for recommending further ongoing medical follow-up. In case they strictly want to avoid the setting of a tertiary university hospital, we inform them about other congenital cardiac clinics or advise them to see a local cardiologist. All patients receive a copy of our medical report. This keeps the patients informed about ongoing issues (even if they do not understand all of the medical terminology), and the statement about the next follow-up appointment recalls the need for ongoing medical care. In addition, the patients' feedback and questions regarding the medical report tell us what they do or do not understand regarding their heart defect. Only a minority of patients wish not to be confronted with their medical records or do not bother about it.

Some patients perceive the adult congenital cardiac clinic as anonymous, formal facilities, in contrast to the familiar and relaxed atmosphere in the pediatric clinic. Some patients are unhappy about being confronted with several new faces during a single follow-up visit in the adult congenital clinic. They are understandably reluctant to retell their medical history several times a day. As many adult congenital cardiac clinics are part of a larger adult cardiology department with its different services, it is often difficult to have one and the same physician doing the physical exam, the echocardiography, the exercise test and the pacemaker interrogation. It has however proven to be useful to establish a continuity of care with respect to the congenital cardiologists in charge of the patient. To discuss ongoing issues with the same cardiologist at any visit is helpful in building up a trusted patient–doctor-relationship. As a part of this relationship, patients should also have the possibility to directly contact their cardiologist by phone or e-mail. A webpage presenting the team and the clinic and providing further information about the conduct of follow-up visits, the use of educational material and informative leaflets during the visits (as for example the adult congenital heart disease passport) complement the personal contacts (Fig. 1).

All adult congenital heart disease patients should be informed about local or national patients' organization and be encouraged to become active members. As a member of a patient organization, they can share their experiences with others, play an active role and take care into their own hands.

5. Conclusions

Appropriate medical care for adults with moderate or complex congenital heart disease includes follow-up in specialized congenital cardiac clinics. Implementation of a structured transition program, teaching patients about their heart defect and the consequences of repair or palliation in earlier life, establishing a continuity of care within the adult congenital cardiac clinic, optimizing organization and communication between different care-givers, and encouraging patients to play an active role regarding their personal health care, are valuable recommendations on how to avoid another generation of adult congenital heart disease patients from being lost to follow-up.

Acknowledgment

Many of the thoughts presented in this manuscript reflect my experience as a fellow at the Toronto Congenital Cardiac Center for Adults with Dr. Webb as its director at that time, and his outstanding commitment to adult congenital heart disease.

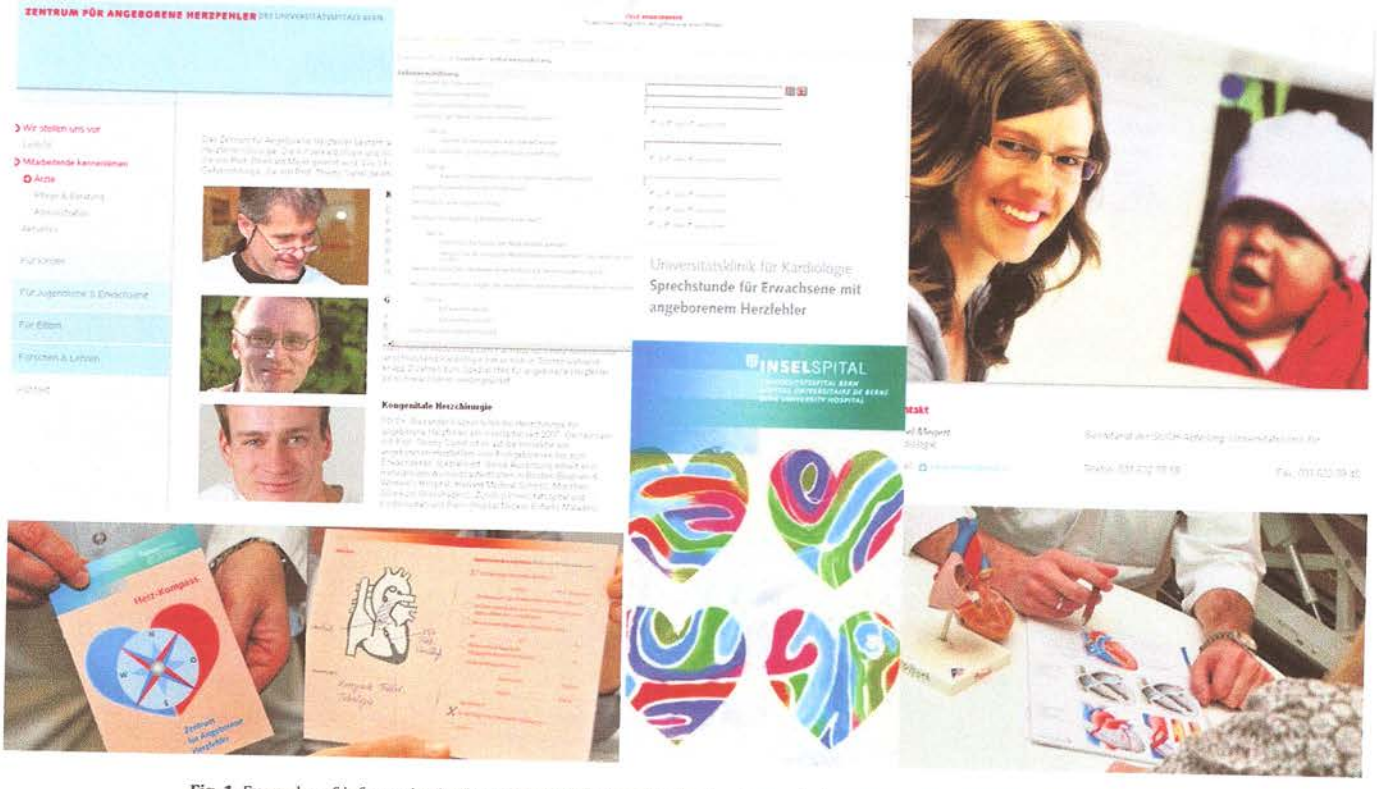

Fig. 1. Examples of informative leaflets and booklets for patient education, and of our center specific website (www.guch.ch).

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