Geriatric congenital heart disease: a new challenge in the care of adults with congenital heart disease?

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This editorial refers to 'Congenital heart disease beyond the age of 60: emergence of a new population with high resource utilization, high morbidity, and high mortality', by O. Turatel et al., on page 725 and 'Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of 'Grown-up Congenital Heart Disease' in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology', by H. Baumgartner et al., on page 686.

The major advances in paediatric heart surgery and paediatric cardiology over recent decades resulted in a markedly increasing number of patients with congenital heart disease who survive into adulthood.1,2 While 90% of patients with complex disease died during childhood when born in the 1950s, the majority already survived beyond 18 years when born in the 1980s.3,4

Thus, we are faced now with a new population of grown-ups with congenital heart disease (CHD) who need very special care.1,2

Why are adults with congenital heart disease so special?

Although the number of adults with CHD currently living in the European Union must be estimated at around 1 million patients, they nevertheless represent only a minority within the population of adults with cardiovascular disease1 considering that ~24 million may, for example, suffer from coronary artery disease as one of the major players.5 Thus, CHD is rarely seen in the non-specialized clinical practice of cardiology. In addition, the diversity of CHD and the resulting clinical problems are immense.1 Although knowledge of adult CHD (ACHD) has grown in recent years, the spectrum of disease and late complications are constantly changing due to the development of improved surgical techniques over the years. A typical example is transposition of the great arteries. In the current population of adults, those who underwent atrial switch operation are still dominating, and common late complications that need to be taken care of are arrhythmias, failure of the systemic right ventricle, and baffle obstruction or leakage.6 Adolescents and young adults have already undergone arterial switch, with a markedly differing spectrum of late complications including pulmonary stenoses, dilation of the neo-aorta with aortic regurgitation, and problems with the coronary arteries.7

Although paediatric care of CHD has become very successful and enables the vast majority of patients to survive into adulthood, most defects cannot be cured and special efforts are required to maintain the good results of paediatric cardiac medicine.1,2 So far, significant morbidity and excess mortality are an issue, particularly in moderate and complex CHD.8 Optimal care with reduction of morbidity and mortality remains a special challenge. In the current population of ACHD, the vast majority of deaths are cardiac and mostly CHD related, with only a few exceptions of milder lesions such as early corrected atrial septal defect and patent ductus arteriosus.8 The leading causes of death are heart failure, sudden death (mainly due to arrhythmia), and operative death at the time of surgical re-intervention.8 It is likely that timely detection of defect residua and sequelae with their potential of late complications followed by timely intervention and re-intervention can reduce late morbidity and mortality. However, this will require highly specialized care. A typical example is tetralogy of Fallot. A complex congenital heart defect that rarely allowed survival into adulthood has now turned into a defect with good outcome since surgical closure of the ventricular septal defect and relief of the right ventricular outflow tract obstruction have become possible. Since then, most patients have survived into adult adulthood with a good quality of life. However, excess mortality compared with the general population has been demonstrated,9 and late complications such as arrhythmias with a significant rate of sudden cardiac death and heart failure are not uncommon.8 A key player in this context appears to be residual pulmonary regurgitation that was for a long time thought to be benign but causes right heart failure and arrhythmias in the long run.1 Since the only treatment for pulmonary regurgitation in most patients remains surgical valve replacement, the timing of the intervention is still a
dilemma when considering the consequences of valve replacement during childhood and the expected number of re-operations required during adulthood. This is of particular importance as repeat re-operations contribute significantly to long-term mortality. The advent of transcatheter valve implantation may lead to paradigm shift now as it may markedly reduce the number of re-operations and thus allow earlier first intervention. However, a lot of research is still required to define the optimal treatment algorithms for this heart defect.

Besides all these surgical and medical issues directly related to the heart defect, it has become widely recognized that ACHD patients require special integrated care, taking into consideration other medical issues (pregnancy, non-cardiac surgery, etc.) as well as psycho-social issues.

Thus, there is general agreement today that ACHD care requires super-regional specialist centres, and special training programmes for the caring physicians and health professionals need to be established.

Who should take care of patients with adult congenital heart disease and where?

There is an ongoing discussion regarding who should primarily take care of this patient group—paediatric or adult cardiology—and where should that happen, in a paediatric or in an adult setting. Although cardiology as a discipline started primarily with structural heart disease—acquired valvular and congenital—adult cardiology has obviously developed in a completely different direction over the decades. Coronary artery disease, arrhythmias, and heart failure have become the main focus. The adult cardiologist has thus become more and more unfamiliar with CHD in general. The new insights gained in paediatric cardiology and the upcoming problems of patients with operations not seen in the past have led to a particular lack of knowledge of the spectrum of ACHD and its late complications that we are currently faced with. The interest in late outcome and the maintenance of the treatment success accomplished during childhood together with the impression that adult cardiology may not take proper care of these patients, as well as the trend towards decreasing numbers of children with CHD, has led in some countries to the integration of ACHD care into paediatric cardiology departments.

It is obvious, however, that neither departments of cardiology nor departments of paediatric cardiology are most suitable for ACHD care per se. The knowledge gap in CHD—clinically, in imaging, and in intervention—may be the problem of the average adult department. On the other hand, paediatric cardiology does not cover the needs of adults with CHD per se as well. Not only do long-term problems of the heart defects encountered during adult life differ from those seen in children, and particular patient groups such as those with transposition of the great arteries with atrial switch or Eisenmenger patients are no longer or only rarely seen in the paediatric population, but adult patients just require adult medicine. Paediatricians are not familiar with pregnancy pre-evaluation and care, with adult cardiovascular disease such as coronary artery disease, systemic hypertension, etc., and with adult co-morbidities. Therefore, it has been generally accepted that ACHD cardiology is a new specialty in addition to both adult and paediatric cardiology. Trainees may come from either training track but need additional specific training in this new specialty as well as additional knowledge in paediatric cardiology for adult cardiologists and in adult cardiology and adult medicine in general for paediatric cardiologists.

Geriatric congenital heart disease: a new chapter in adult congenital heart disease care?

Adult congenital heart disease cardiology has so far mainly focused on young and middle-aged adults. However, the ACHD population is obviously ageing, the admission of patients older than 65 years has markedly increased, and the mortality rates in CHD have shifted away from the young towards adults of increasing age. However, little is still known about the prevalence, disease burden, and determinants of mortality in geriatric adults with CHD. Alfilalo et al. were the first to address this issue. Although theirs is a population-based study, it has major limitations due to its specific methodology. Using the administrative database of the Quebec healthcare system between 1983 and 2005, the authors had to rely on International Classification of Diseases, 9th Revision (ICD-9) codes and to work with the rather superficial data provided by such a source. Nevertheless, they set the stage for the new chapter of ‘geriatric CHD’. They reported a prevalence of ACHD in adults ≥ 65 years of 3.7 per 1000 general population. Independent predictors of mortality in this study surprisingly were dementia, gastrointestinal bleed, and chronic kidney disease, but not CHD severity and directly CHD-related complications. However, 60% of patients had shunt lesions, 37% valvular disease, and only 3% other, very rarely more complex, disease.

The study by Tutarel et al. is now the first to give more detailed insight into the prevalence, morbidity, and mortality of CHD beyond the age of 60 years. In a very large study population of 7315 ACHD patients of a tertiary centre, the prevalence of ACHD beyond 60 years increased 10-fold from 0.5% in 2000 to 5.1% in 2012. This is consistent with unpublished data from the German Competence Network of Congenital Heart Disease currently recording 5.2% of adult patients with CHD of 60 years and older. Figure 1 demonstrates the current estimate of patients with CHD in the European Union. In 2008, the number of adults with CHD exceed for the first time that of children, and the portion of adults will be constantly increasing. As the population is ageing, the percentage of ‘geriatric CHD’ will also increase and may be expected to reach 11% of the ACHD population in 2013. The paper not only demonstrates the exponential increase in elderly ACHD patients but also that—compared with younger ACHD patients—elderly ACHD patients are afflicted by substantially higher morbidity, frequently requiring hospital admissions or interventions, and thus utilize healthcare resources disproportionately. In contrast to Alfilalo’s study, one-third of patients had moderate to complex CHD, and CHD severity indeed had an important impact on morbidity and mortality. Nevertheless, coronary artery disease, symptoms of heart failure, and moderate...
to severe reduction in systemic ventricular function were the inde-
pendent predictors of mortality by multivariate analysis. Although
the spectrum of congenital lesions as well as the causes of death
may change over time, with more patients having complex CHD
reaching this age, the results emphasize the importance of additional
acquired co-morbidities such as coronary artery disease and others
for ACHD outcome. This may have important implications on
healthcare organization for ACHD.

In summary, the number of patients with CHD over 60 years
of age is constantly growing. Acquired cardiac disease such as
coronary artery disease becomes increasingly important for the
outcome, as do additional co-morbidities. This highlights the
need for integrated care in adult medicine in providing not only
adult cardiology specialties but also the entire specialties of
adult medicine. Although the actual spectrum of survival limiting
disease may change over the years with more patients suffering
from complex CHD, the study of Tutarel et al. provides the
following important messages.

- The field of ACHD is growing and the need to establish the subspe-
cialty ACHD cardiology is once more confirmed.
- ACHD patients are reaching an older age, and consideration of additional acquired cardiovascular disease and co-morbidities
deserves more attention.
- Prevention of acquired cardiovascular disease in ACHD patients
requires particular attention and should be started early.

- ACHD care requires profound knowledge not only of CHD in
general and the specific problems of adults with CHD, but also
of acquired adult cardiovascular disease and general adult medi-
cine.
- ACHD divisions should be located in settings where not only the
entire spectrum of adult and paediatric cardiology as well as
cardiac surgery are provided, but where all specialties of adult
medicine are present.

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