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Selection of the Best in 2016 in Congenital Heart Diseases



Selección de lo mejor del año 2016 en cardiopatías congénitas

To the Editor,

Last year saw a continuation of the trend to publish more articles on congenital heart diseases (CHD), a trend characterized by intensified interest in the epidemiological importance being acquired by this population.

One of the more notable studies on the consequences of modern interventions in CHD was the Finnish National Registry of late causes of death among patients younger than 15 years who underwent surgery between 1953 and 2009 (10 964 patients, 98% follow-up).¹ This study compared causes of death with those in the general population, subdividing the results into 2 periods: 1953 to 1989 and 1990 to 2009. Total survival was higher in the second period, and the main cause of CHD-related death was heart failure (HF); however, the rate of HF-death declined significantly over the long-term among patients who underwent surgical correction of ventricular septal defects and transposition of the great arteries. An especially notable finding was the near complete absence of sudden death in the second period among patients with tetralogy of Fallot and transposition of the great arteries. Also of interest is the higher death rate from neurological diseases and infections among CHD patients and a higher incidence of neoplasms, especially in the second period.¹

Although HF is the main cause of death among CHD patients, the indications and optimal timing for transplantation are less well defined than for other heart conditions. Patients with CHD are less likely than other heart disease patients to be fitted with a defibrillator or ventricular assist device or to be included on high-priority wait-lists, and their wait-list mortality is higher. These specific concerns prompted the American Heart Association to issue a scientific statement reviewing the particular characteristics of CHD patients that can affect transplant surgery, such as complex anatomies frequently requiring additional surgery, HLA antibody sensitization, and difficult vascular access.² The review also examines evidence on the effectiveness of ventricular assist devices in the treatment of CHD and proposes therapeutic strategies to improve transplant outcome, including specific changes to the criteria used to assign urgency status to CHD patients on transplant wait-lists.²

Studies examining sudden death among CHD patients included a meta-analysis of the use of implantable cardioverter-defibrillators in a total of 2162 individuals followed up over 3.6 ± 0.9 years.³ In this population, 1 or more appropriate shocks were recorded in 22% of patients in primary prevention and 35% of those in secondary prevention; inappropriate shocks were recorded in 25% of patients, and other defibrillator-related complications in 26%.³ These findings point to the need for continuing improvements in risk stratification and implantation programs.

There is also increasing awareness of the influence of CHD on psychosocial factors, reproductive function, and noncardiac conditions. A study published last year sought to characterize the effects of CHD on brain function, describing how neurodevelopment before birth and during infancy is influenced by a close interaction among genetic and epigenetic factors, direct disease consequences, such as severe cyanosis, and even therapeutic interventions; moreover, the cumulative burden of CHD continues into adulthood, when disease progression and the appearance of HF, arrhythmias, and comorbidities contribute to brain damage in the form of neurovascular disease.⁴ The article also suggests interesting future directions for translational research to improve prognosis and quality of life in this population. Several studies have explored the multiorgan consequences of univentricular circulation, examining post-Fontan-procedure hemodynamics and the prevalence of liver fibrosis; however, these studies have not yet been translated into a specific therapeutic program. It is nonetheless worth highlighting a small, single-center study that presented a new therapeutic option for plastic bronchitis, a serious complication after single-ventricle palliation; the authors used a percutaneous embolization technique to reduce lymphatic flow to the pulmonary parenchyma, reporting significant symptomatic improvement in 15 of the 17 patients treated.⁵ Plastic bronchitis is associated with high morbidity and mortality, and this new treatment could therefore represent an important advance if confirmed in a larger population over a longer follow-up.

Cardiac magnetic resonance is undoubtedly one of the most important imaging tools for diagnosis, risk stratification, and treatment planning. A recent review summarized advances applied to CHD such as 3 D flow imaging, which helps to elucidate underlying pathophysiology, and tissue characterization techniques such as T₁ mapping, which can detect clinical features of tetralogy of Fallot and systemic right ventricle.⁶ The article also emphasizes the need to carefully manage gadolinium contrast agent administration to patients who will require repeat examinations throughout life from an early age.

On a final note, the past year saw a continued dearth of randomized studies in large CHD cohorts; moreover, a high proportion of published articles reported retrospective single-center studies, demonstrating the enormous potential for future research in this area.

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Selection of the Best of 2016 in Clinical Arrhythmology



Selección de lo mejor del año 2016 en aritmología clínica

To the Editor,

Clinical arrhythmology is an essential field for both electrophysiologists and cardiologists: in the case of the former to prevent the specialist from becoming a technician and in the case of the latter to fully understand the specialty. The clinical guidelines reaffirm this idea while also recommending an increasingly widespread interventional approach throughout the range of arrhythmias. This concept is clearly illustrated in the interaction between arrhythmia-cardiomyopathy and ventricular dysfunction. Between 10% and 50% of patients with heart failure have atrial fibrillation (AF), and the potential worsening of ventricular function due to inappropriate rate control is recognized. In patients with ventricular extrasystole (VE), who are often referred for electrophysiological assessment, several studies have shown an incidence of arrhythmia-induced cardiomyopathy of between 9% and 34%. Although knowledge of their pathophysiology is still incomplete, a fundamental factor is now recognized to be high total extrasystole burden, defined as more than 10 000 to 25 000 VE per day (10%-24% of the total number of complexes), as well as certain clinical characteristics (male sex, high body mass index), electrocardiographic characteristics (QRS width in VE > 153 ms), and anatomic features (VE origin other than ventricular outflow tract).¹ Although the incidence and prevalence of arrhythmia-induced cardiomyopathy is not known, early detection should be a priority, given the excellent response to treatment, which is generally interventional. The correlation between chronic consumption of caffeine-containing products and the degree of atrial and ventricular ectopy has still not been established.

The increasing prevalence of AF is a health challenge of the utmost importance. The 3 basic pillars of treatment include anticoagulation, rhythm control, and rate control.² Recent evidence reflects the benefit of reducing or modifying risk factors, for example, substantially decreasing the arrhythmic load of AF by weight loss sustained in the long term. Along these lines, the CARDIO-FIT study analyzed the impact of cardiorespiratory fitness on arrhythmia recurrence in obese individuals with AF.³ The improvement in cardiorespiratory fitness achieved through a specific training program reduced recurrences, and this benefit was in addition to that obtained through weight loss: an increase of 1 metabolic equivalent corresponded to a 9% decrease in recurrences. Furthermore, several studies have confirmed the

dose-dependent relationship between physical exercise and AF, as well as the additive effect of certain risk factors, and it was observed that more than 2000 hours of high-intensity endurance training during the course of a lifetime, tall stature (> 179 cm), abdominal obesity (> 102 cm in men and 88 cm in women), and sleep apnea syndrome were associated with AF.⁴ The role of intensive physical endurance training may also be proarrhythmic for the ventricle, and cause anatomical functional changes in the right ventricle in predisposed individuals. These changes can be detected by imaging techniques, particularly after exercise, and are associated with potentially fatal ventricular arrhythmias.

Oral anticoagulation in AF is becoming increasingly widespread, although this therapy is still underused in elderly patients and other subgroups with greater bleeding risk. In many cases, it is worth considering alternatives such as percutaneous closure of the left atrial appendage. In addition, the first specific reversal agent for dabigatran is now on the market (idarucizumab [Praxbind]). We do not know what the clinical impact of this availability will be.

Clinical history and the electrocardiogram (ECG) are the main source of information for stratification of arrhythmic risk: seek and you shall find. In patients with Brugada syndrome and no history of cardiac arrest, an S-wave ≥ 0.1 mV or duration ≥ 40 ms in lead I has been described as a marker of risk of sudden cardiac death.⁵ For this syndrome, the usefulness of quinidine in reducing malignant ventricular arrhythmias has also been confirmed. The term *early repolarization* has been used for more than 50 years now, but only in the last 10 has it been associated with sudden cardiac death. Finally, in 2015, a consensus was reached on its definition, thereby allowing appropriate characterization of this finding in the ECG for clinical investigation. Furthermore, progress in genetics is and will be important in the field of arrhythmology, and different studies continue to provide support for the clinical benefit of specific gene therapy. Genetic susceptibility determines certain aspects of the pathophysiology of many cardiac arrhythmias, and it is foreseen that the importance of genetic study will increase as the emphasis in rhythm disorders shifts to prevention.⁶

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