Selection of the Best of 2017 in Congenital Heart Disease

Selección de lo mejor del año 2017 en cardiopatías congénitas

To the Editor,

This year once again saw numerous scientific publications in the field of congenital heart disease (CHD). Much of the work stresses that, despite prognostic improvements, this population still faces significant morbidity and mortality.

Oliver et al.[1] presented a study in a cohort of 3311 patients with CHD and investigate different risk factors for excess mortality. In this study, the annual mortality rate was 0.89% and the mean survival was 75.1 (95% confidence interval [95%CI], 73-77) years with a standardized mortality rate of 2.64 (95%CI, 2.3-3.0, $P < .001$). In multivariable analysis, 11 predictors of death from any cause were identified and patients with 1 or more risk factors had a significantly higher standardized mortality (5.22, 95%CI, 4.5-6.0; $P < .001$) than those without risk factors (1.14, 95%CI, 0.9-1.5, $P = .19$).

Another noteworthy work is that of Hjortshøj et al.,[2] which described the current causes of death in patients with Eisenmenger syndrome. To do this, they followed 1546 patients from 13 countries between 1977 and 2015. During this period, 558 patients died (2.8% of patients per year), with heart failure (HF) the leading cause of death, followed by infectious etiology, sudden death, thromboembolism, hemorrhage, and periprocedural death. The authors analyzed the causes of death in 2 periods and observed a significant increase in mortality due to HF, as well as a decrease in mortality related to thromboembolism and procedures themselves. Overall, survival improved, with the average age of the deceased increasing from 36.9 ± 18.8 to 45.2 ± 16.2 years. In conclusion, patients died later and for chronic causes rather than acute events.

Crucially, HF is the end point of multiple CHDs, complicating its management, given the limited evidence on medical treatment, the difficulty of receiving a heart transplantation, and the lower probability of receiving ventricular support. This year, data were published on the use of mechanical ventricular assist devices in patients with CHD included in the INTERMACS registry,[3] which includes more than 16 000 patients with long-term mechanical ventricular assist devices implanted in the United States; only 126 of the patients had been diagnosed with CHD, less than 1%. For the analysis, patients were divided into those with biventricular circulation and systemic morphologic left ventricle (n = 63), biventricular circulation and systemic morphologic right ventricle (n = 45), and univentricular circulation (n = 17). Compared with other etiologies, patients with CHD were younger and were more likely to require biventricular assistance (21% vs 7%) and, in most cases, assistance was used as a bridge to transplantation and not as destination therapy. The most interesting aspect of this work is that, although overall mortality was higher in patients with CHD, patients with CHD and biventricular circulation with a left ventricular assist device had the same mortality as patients without CHD, regardless of the systemic ventricular morphology. Among patients with biventricular assistance, mortality was higher in those with CHD, and the need for biventricular assistance was the only independent predictor of mortality in this population (hazard ratio [HR] = 4.4, 95%CI, 1.8-11.1).

Another of last year's hot topics in CHD was liver failure due to the Fontan circulation. Among the many relevant publications, the most interesting is possibly the review by Hilscher et al.,[4] which details the clinical, analytical, imaging, and anatomopathological findings in the study of liver cirrhosis and its applicability to patients with the Fontan circulation. The authors recommend an annual clinical follow-up during the first 10 years after completion of the Fontan circulation, a complete analytical study including calculation of liver cirrhosis risk scores (the aspartate aminotransferase to platelet ratio index [APRI] and Fibrosis 4 [FIB-4]) every 2 or 3 years, and a liver ultrasound at 5 years. However, after 10 years, a complete annual assessment is recommended that includes a clinical and analytical evaluation and abdominal ultrasound. The presence of unexplained thrombocytopenia or ascites on ultrasound would be an indication for liver magnetic resonance imaging with elastography or a liver ultrasound with elastography to determine the degree of liver fibrosis. In the case of established cirrhosis, they propose liver ultrasound and a biannual alpha-fetoprotein determination to screen for hepatocellular carcinoma.

One of the diseases whose future has changed drastically is hypoplastic left heart syndrome (HLHS). Latus et al.[5] analyzed patient outcomes according to palliation technique–Norwood surgery or the hybrid procedure–using magnetic resonance parameters after the second stage, prior to the Fontan procedure, and found no significant differences in right ventricular ejection fraction, preserved in both groups, or in cardiac output; however, strain values, intraventricular synchrony parameters, and pulmonary branch development were better after Norwood. The need for reinterventions also favored the post-Norwood group.

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Selection of the Best of 2017 in Catheter Ablation

Selección de lo mejor del año 2017 en ablación con catéter

To the Editor,

The last year saw the publication of various noteworthy studies in the field of percutaneous catheter ablation aimed at reducing procedure-related complications by refining the perioperative approach, optimizing patient selection, demonstrating the viability and benefit of the invasive approach for unconventional substrates, and improving arrhythmia recurrence-free survival outcomes.

Regarding the perioperative management of atrial fibrillation (AF) ablation, the evidence supports the strategy of not interrupting anticoagulant treatment with vitamin K antagonists during the procedures. In addition, more and more patients presenting to electrophysiology laboratories are under treatment with novel oral anticoagulants (NOACs). Several studies have been launched to determine the safety of the uninterrupted approach for these patients. After the VENTURE-AF study,[1] which showed the plausibility of ablation in patients under uninterrupted treatment with rivaroxaban, the results have been published of the randomized, multicenter trial RECIRCUIT,[2] a study that assessed the safety of uninterrupted dabigatran versus warfarin during ablation procedures. With a primary end point of the rate of major bleeding events 2 months after ablation, dabigatran was clearly superior to warfarin (1.6% vs 6.9%, $P < .001$). There was only one thromboembolic event (secondary end point) in the warfarin group. The rates of minor bleeding were similar in the 2 groups. These data support the use of dabigatran for uninterrupted treatment during ablation procedures.

According to data from the European ablation registry, the profile of patients undergoing AF ablation procedures in our setting is still that of a relatively young patient (average age, 59 years) with little comorbidity and normal ventricular function (IQR 55-65). Because very few of these patients have advanced heart disease (<1%), the results of the CASTLE-AF multicenter randomized clinical trial,[3] recently presented at the European Congress of Cardiology 2017, are surprising. This study compares the efficacy of the AF ablation procedure with that of standard treatment during the follow-up of 397 patients with severe heart failure (left ventricular ejection fraction < 35%, New York Heart Association functional class > II) who have an implantable automatic cardioverter-defibrillator (ICD) capable of monitoring the arrhythmia burden. The novelty of this study is that the primary end point is death from any cause and hospitalization for HF. The primary end point occurred in 28.5% of the ablation group and in 44.6% of the control group after a mean follow-up of 37.8 months (relative risk reduction = 38%; hazard ratio [HR] = 0.62, 95% confidence interval [95%CI], 0.43-0.87). This study shows for the first time that AF ablation not only improves patients' symptoms, but also reduces morbidity and mortality in the mid-to-long term. Although they are striking results, because the patients involved were highly selected and do not reflect the typical patient profile, the results must be interpreted with caution.

Characterization of the arrhythmic substrate of patients with ventricular tachycardia (VT) and underlying heart disease allows successful ablation without the need for tachycardia mapping and reduces the rate of sustained VT recurrences versus clinical VT ablation alone during follow-up in patients with well-tolerated arrhythmia. Advances in imaging techniques, particularly delayed enhancement magnetic resonance imaging (DE-MRI) characterization of the substrate, have helped to improve ablation outcomes. The study by Andreu et al.[4] showed a reduction in the short- and long-term rates of VT recurrence with a substrate ablation strategy guided by the fibrosis detected on 3-T MRI, which identifies the regions of heterogeneity of the fibrotic scar contributing to the formation of the reentry circuits. The strategy supported by DE-MRI had an 18.5% rate of VT recurrence versus 43.8% for the control group, comprising those who could not undergo MR or whose MRI was not of good quality (Figure). In multivariable analysis, the guided ablation strategy was an independent predictor of recurrence (HR = 0.48; 95%CI, 0.24-0.96).

In addition, a multicenter and randomized study[5] showed no benefit from substrate ablation in patients with an ICD and unstable VT; time until first recurrence of VT/VF was used as the primary end point. Recurrence was observed in 51% of the patients with ablation and ICD versus 48.6% of the patients with ICD alone in a mean follow-up of 2.2 years. Ablation was still associated with a reduction in the total number of VF/VT episodes with or without therapy.

A relevant novelty is radiofrequency ablation in Brugada syndrome. There were few therapeutic options for patients with Brugada syndrome and ventricular arrhythmias. Ablation in this context, although previously reported, was limited to a small series. The recent study by Pappone et al.[6] included 135 symptomatic ICD carriers with spontaneous or inducible VT in the electrophysiological study. For better epicardial characterization of the substrate, the mapping was performed with ajmaline infusion. The objective was to identify and eliminate all delayed and fragmented potentials in the right ventricular epicardium in order to normalize the electrocardiographic pattern and ensure that no arrhythmias were induced in the electrophysiological study. After a mean follow-up of 10 months, electrocardiographic normalization and the absence of spontaneous or inducible ventricular arrhythmias persisted in 98.5% of the patients.

In summary, research in the field of percutaneous ablation during 2017 has highlighted the safety of uninterrupted intraprocedural treatment with NOACs, an improvement in mortality and hospitalizations for patients with HF and AF who undergo ablation, and the importance of the integration of imaging techniques with navigation systems to reduce the ventricular arrhythmia burden. The good results with the substrate approach in patients with ventricular arrhythmias and Brugada syndrome open the door to an expansion of the therapeutic options in complex cases and, if these results are maintained during long-term follow-up, they may