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Special article

2013 Update on Congenital Heart Disease, Clinical Cardiology, Heart Failure, and Heart Transplant

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ABSTRACT

This article presents the most relevant developments in 2013 in 3 key areas of cardiology: congenital heart disease, clinical cardiology, and heart failure and transplant. Within the area of congenital heart disease, we reviewed contributions related to sudden death in adult congenital heart disease, the importance of specific echocardiographic parameters in assessing the systemic right ventricle, problems in patients with repaired tetralogy of Fallot and indication for pulmonary valve replacement, and confirmation of the role of specific factors in the selection of candidates for Fontan surgery. The most recent publications in clinical cardiology include a study by a European working group on correct diagnostic work-up in cardiomyopathies, studies on the cost-effectiveness of percutaneous aortic valve implantation, a consensus document on the management of type B aortic dissection, and guidelines on aortic valve and ascending aortic disease. The most noteworthy developments in heart failure and transplantation include new American guidelines on heart failure, therapeutic advances in acute heart failure (serelaxin), the management of comorbidities such as iron deficiency, risk assessment using new biomarkers, and advances in ventricular assist devices.

Actualización 2013 en cardiopatías congénitas, cardiología clínica e insuficiencia cardiaca y trasplante

Palabras clave:
Cardiopatías congénitas
Cardiología clínica
Insuficiencia cardiaca
Trasplante cardiaco

RESUMEN

En este artículo se presentan las novedades más relevantes de 2013 en tres áreas clave de la cardiología: cardiopatías congénitas, cardiología clínica e insuficiencia cardiaca y trasplante. En cardiopatías congénitas se han revisado las aportaciones relacionadas con la muerte súbita del adulto con cardiopatía congénita, la importancia de algunos parámetros ecocardiográficos en la valoración del ventrículo derecho sistémico, los problemas del paciente con tetralogía de Fallot corregida e indicación de sustitución valvular pulmonar y la confirmación del papel que algunos factores tienen en la selección del candidato a cirugía de Fontan. Entre las novedades del área de cardiología clínica, están el documento para el correcto diagnóstico de las miocardiopatías elaborado por un grupo de trabajo europeo, estudios de coste-efectividad sobre implante percutáneo de prótesis aórticas, un consenso sobre el manejo de la disección de aorta tipo B y una guía de la enfermedad valvular aórtica y de aorta torácica ascendente. En insuficiencia cardiaca y trasplante, las novedades más importantes son las nuevas guías estadounidenses de insuficiencia cardiaca, los avances terapéuticos en la insuficiencia cardiaca aguda (serelaxina), el manejo de las comorbilidades como el déficit de hierro, la evaluación del riesgo con los nuevos biomarcadores y los avances en asistencia ventricular mecánica.

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Abbreviations

ACD: acute coronary syndrome
AMR: antibody-mediated rejection
CMR: cardiac magnetic resonance
HF: heart failure
SD: sudden death
TAVI: transcatheter aortic valve implantation

DEVELOPMENTS IN CONGENITAL HEART DISEASE

Sudden Death

Two studies have analyzed the risk factors for sudden death (SD) in adult congenital heart disease. Koyak et al[1] conducted a multicenter study using 3 large databases that included more than 25000 patients. The total mortality rate was 5%, of which 19% was due to SD. The underlying congenital heart disease in SD patients was classified as mild, moderate, and severe in 12%, 33%, and 55%, respectively. Risk factors associated with SD were supraventricular arrhythmias, ventricular dysfunction, increased QRS duration, and QT dispersion. In Spain, Gallego et al[2] analyzed the incidence of SD in 936 adults with previously repaired congenital heart disease. During follow-up, the total mortality rate was 5.3% and the incidence of SD was 2.6/1000 patient-years. The highest incidence of SD occurred in patients with transposition complexes. Independent risk factors of SD were delayed initial examination in adults and severe systemic ventricular dysfunction.

Systemic Right Ventricle

The assessment of ventricular function in the systemic right ventricle is particularly challenging. Cardiac magnetic resonance (CMR) imaging has become the gold standard, but is of limited use in patients with pacemakers or implantable cardioverter-defibrillators. A group from the University of Bern[3] compared echocardiography-derived parameters with CMR-derived systemic right ventricle ejection fraction in adults with transposition of transposition the great vessels who had undergone the Mustard/Senning procedure. Routine nongeometric echocardiographic parameters (lateral tricuspid annular plane systolic excursion, lateral right ventricle systolic motion velocities assessed by tissue Doppler, total ejection isovolume index) were weakly correlated with CMR-derived systemic right ventricle ejection fraction. The dp/dt measured across the tricuspid regurgitant jet and fractional area change were significantly correlated with the CMR-derived systemic right ventricle ejection fraction. A fractional area change $< 33\%$ (area under the receiver operating characteristic curve = 0.73) identified an systemic right ventricle ejection fraction $< 50\%$ with high sensitivity and specificity. Furthermore, a study conducted at the Royal Brompton Hospital[4] demonstrated that heart deformation or systolic right ventricle longitudinal strain is significantly reduced in patients with systemic right ventricle, is related to subpulmonary ventricular function, and predicts adverse clinical outcomes in adults following physiological repair of transposition the great vessels (atrial switch).

Van der Bom et al[5] conducted a multicenter, randomized, double-blind, placebo-controlled pilot trial of valsartan on systemic right ventricle function, functional capacity, B-type natriuretic peptide concentrations, and quality of life of patients with atrial switch or congenitally corrected transposition the great vessels. Although no significant treatment effects were found, possibly due to the small sample size, the most positive aspects were: *a)* the absence of adverse effects; *b)* a smaller increase in ventricular volume and mass in the treatment group, and *c)* less deterioration of ventricular function in symptomatic patients treated with valsartan. This pilot trial opens the door to other larger trials to explore the long-term effect of drugs routinely used in adults with ventricular dysfunction in this specific group of patients.[6]

Tetralogy of Fallot

Tetralogy of Fallot has the highest postoperative survival rate among cyanotic congenital heart disease and is the most studied congenital heart disease in the adult population. The greatest uncertainty associated with this defect is the long-term risk of SD. To date, more than 30 risk factors have been proposed, of which age at repair, QRS duration, right ventricle enlargement, and left ventricular dysfunction are most predictive.[7] Diller et al[8] included in this long list echocardiographic parameters that measure longitudinal left venticular function, particularly mitral annular plane systolic excursion and longitudinal strain. Unfortunately, the plethora of proposed risk factors indicates the difficulty of finding simple methods that have high predictive value.

Hickey et al[9] analyzed the functional health status of adults with Tetralogy of Fallot in a large patient population. Although long-term survival was excellent, almost 50% of patients had cardiorespiratory symptomatology, and physical functioning decreased with advancing age. A similar study found that functional health status was significantly worse in adults surviving Tetralogy of Fallot repair than that of their age-matched healthy siblings.[10] Chronic pulmonary disease is the main hemodynamic problem associated with decrements in functional health status. Thus, the optimal timing of pulmonary valve replacement remains an unresolved issue.[11] The use of CMR has become the main focus in the debate. If the goal is to normalize right ventricle volumes after valve replacement, surgery is recommended[12] before the end-diastolic volume index is > 160 mL/m^2 or end-systolic volume index is > 80 mL/m^2. However, if the target is midterm survival or the incidence of arrhythmias or SD, these criteria have no demonstrated predictive value.[13] Early indications based on ventricular volume should be weighed against the great number of reoperations for deterioration of prosthetic valve function. Moreover, other authors[14] have not found a ceiling beyond which right ventricle end-diastolic and end-systolic volumes did not "normalize".

It has been suggested that in both Tetralogy of Fallot and transposition of the great vessels there is an aortic pathology involving histological changes that are similar to those found in Marfan disease. This aortic disease can manifest in these patients as increased aortic root diameter, abnormal wall distensibility, and progressive aortic regurgitation.[15] However, in a multicenter study[16] of 474 adults with Tetralogy of Fallot, although nearly a third of the patients had an aortic root diameter ≥ 40 mm, the prevalence of an observed-to-expected aortic root dimension ratio > 1.5 was only 6.6%, and the prevalence of moderate or severe aortic regurgitation was only 3.5%.

Fontan Circulation

Patients born with a single functional ventricle can be treated using palliative techniques such as total cavopulmonary connection with intracardiac tunnel or extracardiac conduit. These procedures are the result of successive modifications of the original Fontan procedure or atriopulmonary connection, and attempt to prevent overpressure, progressive right atrial dilatation, and its associated complications: energy loss and antegrade flow,