Pregnancy, rheumatic heart disease, and cardiomyopathies: more on under-recognized entities

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Pregnancy is associated with marked haemodynamic changes, such as an increase in cardiac output (among others), that may cause problems particularly in the presence of hypertension and in women with congenital or structural heart disease. Indeed, valvular heart disease, affecting either a single valve or several valves, increases the risk of pregnancy for the mother and the child and therefore requires a careful risk assessment before and during pregnancy and specialized care to minimize maternal and foetal morbidity and mortality. A timely Clinical Review article, entitled ‘Management of valvular disease in pregnancy: a global perspective’, by Karen Sliwa from the University of Cape Town in South Africa provides a guide for risk assessment and for optimal management taking into consideration the resources available in higher as well as lower to middle income countries.

Peripartum cardiomyopathy is another potentially life-threatening heart disease emerging towards the end of pregnancy or in the first post-partum months in previously healthy women. A major challenge is to distinguish peripartum discomforts such as fatigue, shortness of breath, or oedema from the pathological symptoms of peripartum cardiomyopathy. Of note, pre-eclampsia, myocarditis, or underlying genetic disease show symptoms partly similar to peripartum cardiomyopathy, which may explain why peripartum cardiomyopathy remain underdiagnosed. The second Clinical Review on this subject, entitled ‘Peripartum cardiomyopathy: current management and future perspectives’, by Denise Hilfiker-Kleiner from the Hannover Medical School in Germany focuses on novel aspects of physiological and pathophysiological changes of the maternal cardiovascular system by comparing normal conditions, hypertensive complications, genetic aspects, and infectious disease in pregnancies associated with peripartum cardiomyopathy. It also presents clinical and basic science data on the current state of knowledge on peripartum cardiomyopathy and brings them into context, thereby highlighting promising new insights in diagnostic tools and therapeutic approaches and management.

Rheumatic heart disease may become symptomatic during pregnancy or spontaneously, and was—and unfortunately also remains—the cause of untimely death for over a million women annually, particularly in lower income countries. This is outlined in the first clinical research paper of this issue entitled ‘Characteristics, complications, and gaps in evidence-based interventions in rheumatic heart disease: the Global Rheumatic Heart Disease Registry (the REMEDY study)’ by Bongani M. Mayosi from the University of Cape Town in South Africa2 accompanied by an Editorial by Jonathan Carapetis from the Menzies School of Health Research in Casuarina, Australia. The prospective registry enrolled 3343 patients with rheumatic heart disease from 12 African countries, India, and Yemen. Two-thirds had moderate to severe multivalvular disease, which in a third was complicated by congestive heart failure, in a quarter by pulmonary hypertension or atrial fibrillation, and in a small percentage by stroke, infective endocarditis, or major bleeding.

A quarter of adults had dilated ventricles and decreased left ventricular function, which was less common in children (14% and 5%). More than half of the patients were on secondary antibiotic prophylaxis or oral anticoagulants, which in the latter case were, in the majority of the patients, insufficiently dosed. Among women of childbearing age, only 3.6% were on contraceptives. The utilization of valvuloplasty and valve surgery was higher in upper to middle than lower income countries. The authors conclude that rheumatic heart disease is prevalent in Africa, India, and Yemen, with the majority of patients being young, predominantly female, and having a high prevalence of major cardiovascular complications. There is suboptimal utilization of secondary antibiotic prophylaxis, oral anticoagulation, and contraception, and in the use of percutaneous and surgical interventions, particularly in low income countries. Thus, in these areas of the world, better education of patients as successfully done in heart failure, and implementation of modern management strategies for rheumatic heart disease should be mandatory.

The second paper, ‘A study of implanted cardiac rhythm recorders in advanced cardiac AL amyloidosis’, by Julian Gillmore from the University College London Medical School in the UK, sought to characterize the nature of cardiac arrhythmias in severe cardiac AL amyloidosis using implanted cardiac rhythm recorders. Indeed, although AL amyloidosis may respond to chemotherapy, most patients with severe cardiac involvement die within a year, mostly due to tachyarrhythmias or electromechanical dissociation.
Implantable loop recorders were inserted into 20 consecutive patients with newly diagnosed severe cardiac AL amyloidosis and symptoms of syncope or pre-syncope. Weekly implantable loop recordings and additional recordings at the time of symptoms were obtained during almost a year of follow-up. In evaluable cases, death was heralded by bradyarrhythmia, usually associated with a high degree atrioventricular block, followed by pulseless electrical activity.

New York Heart Association (NYHA) class, global left ventricular strain on echocardiography, and N-terminal pro brain natriuretic peptide values differed between survivors and non-survivors. Therefore, the authors conclude that bradyarrhythmias heralded terminal cardiac decompensation in patients with severe cardiac AL amyloidosis. These results therefore would support a study of prophylactic pacemaker insertion in this patient population in the future.

Pulse pressure has been used as a prognostic indicator in a variety of cardiovascular conditions such as hypertension, coronary artery disease, and chronic heart failure. In the third manuscript, ‘Differing prognostic value of pulse pressure in patients with heart failure with reduced or preserved ejection fraction: results from the MAGGIC individual patient meta-analysis’, Colette Elizabeth Jackson and colleagues from the University of Glasgow in the UK found titin, plakophilin-2, myosin-binding protein-C 3, desmoplakin, and channelopathy-causing mutations turned out to be considerable. And 13% had three or more mutations. The authors conclude that bradyarrhythmias heralded terminal cardiac decompensation in patients with severe cardiac AL amyloidosis. These results therefore would support a study of prophylactic pacemaker insertion in this patient population in the future.

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In the fourth basic science paper ‘Atlas of the clinical genetics of human dilated cardiomyopathy’, Benjamin Meder and colleagues from the University of Heidelberg in Germany utilized next-generation sequencing to screen all dilated cardiomyopathy genes in a large cohort. In their multicentre, multinational study, the authors enrolled 639 patients with sporadic or familial cardiomyopathy and found the highest number of known cardiomyopathy mutations in dilated cardiomyopathy, hypertrophic cardiomyopathy, and channelopathy-causing mutations turned out to be considerable. Of note, >38% of patients had compound or combined mutations, and 13% had three or more mutations. The authors conclude that by comprehensively investigating the genetics of dilated cardiomyopathy in a large cohort and across a broad gene panel of the known cardiomyopathy-causing genes, a sound database of the genetic causes of dilated cardiomyopathy could be established. The high number of mutations causing the disease as well as the considerable overlap of different phenotypes with the same or a similar genetic background warrants further studies on potential post-transcriptional mechanisms involved in this phenomenon.

The editors hope that this issue of the European Heart Journal will be of interest to its readers.

References


