

Original Article

The 2017 Seventh World Congress of Pediatric Cardiology & Cardiac Surgery: week in review – ambulatory pediatric cardiology*

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Abstract The Seventh World Congress of Pediatric Cardiology was held in Barcelona in July, 2017. The central philosophy of the congress was “bridging together” all major specialties in the field. This article summarises the highlights of the meeting as it relates to ambulatory paediatric cardiology. There is a now a more unified approach to children with CHD, including assessment of neuro-developmental outcomes. The new World Heart Foundation criteria for the diagnosis of rheumatic fever remain controversial.

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THE SEVENTH WORLD CONGRESS OF PAEDIATRIC cardiology was held in Barcelona in July 2017 – 5 days of great knowledge exchange, multiple talks, abstracts, and posters. The central philosophy of the congress was “bridging together” all major specialties in the field. Accordingly, the scientific programme was carefully planned to address all interests and expertise with concentration streams on paediatric cardiology, paediatric cardiac surgery, adult CHD, anaesthesia, intensive care, and nursing. The aim of the Seventh World Congress of Pediatric Cardiology & Cardiac Surgery (WCPCCS) was to bring together all professionals involved in the care of children’s heart disease and CHD of all ages, from the fetus to the aged. The congress provided a unique opportunity to meet the leaders of specialties from around the world, to learn about the latest innovations and results of procedures, and to contribute to the discussions, debates, and plenary sessions with renowned speakers.

In ambulatory cardiology a wide range of topics were covered by speakers from all over the world. In the field of preventive cardiology, cardiometabolic

syndrome, hypertension, and familial lipid disorders were discussed. Genetic permutations can disrupt energy homeostasis and alter obesity and cardiovascular disease risk. In the exponentially expanding field of epigenetics, there are strong correlations between epigenetic changes and obesity. Furthermore, the process of epigenetic alterations appears to be the most promising mechanistic explanation for the strong epidemiologic findings within the developmental origins of health and disease field. But it is important to remember that a lot of these newly discovered risk factors and mechanisms are still driven, largely in part, by the long understood drivers of cardiovascular disease like diet and activity.

Furthermore, the consequences of obesity on cardiovascular risk factor were discussed. Obesity is not a problem of only high income countries but is a world-wide one. There is no consensus on the diagnosis of metabolic syndrome in the young but obesity relates to cardiovascular risk factors and causes damage to the end organs. Lifestyle modifications can make a difference in outcome; however, these lifestyle modifications are difficult to sustain. Primary prevention of obesity at a community level is also necessary. Maternal obesity can also have epigenetic influences on the fetus. With increasing prevalence of obesity there is also an increase in the incidence and prevalence of hypertension. Hypertension is also associated with target end organ damage. The causes

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of essential hypertension is multifactorial, including altered autonomic tone, insulin resistance, and activation of renin–angiotensin–aldosterone system. As per the new guidelines the goal is to maintain systolic blood pressure less than 120 ml of mercury. The new guidelines for diagnosis and management of hypertension in children have recently been published – paediatrics 2017. As per the new guidelines the goal is to maintain systolic blood pressure <120 ml of mercury. The deleterious effects of nicotine was also discussed. Nicotine is not a benign molecule and causes major cardiovascular morbidity. E-cigarettes can also be lethal to paediatric patients. In addition, lipid disorders and familial hyperlipidaemia were discussed. Familial hypercholesterolaemia is common in the population with a prevalence of 1/250 with a high risk for cardiac events. Primary prevention can decrease morbidity and mortality. Screening is key to improving outcomes by increase percentage of treated familial hyperlipidaemia patients. Cascade screening is successful in certain populations. Ongoing evaluations of country-specific interventions are necessary.

There were several sessions that focused on exercise. Exercise was strongly encouraged in patients with CHD. It was recommended that patients with significant right to left shunts and pulmonary vascular disease, even if they have metabolic equivalents greater than 13, should be restricted from exercise. Patients with significant pulmonary hypertension should be restricted from sports at high altitude. Furthermore, changes in an athlete's heart was discussed in detail. An athlete's heart needs to be differentiated from hypertrophic cardiomyopathy. Athlete's heart is commonly seen in endurance and black male athletes. Overall, 95% of the patients with hypertrophic cardiomyopathy have electrocardiogram abnormalities like lateral T wave inversion, ST segment depression, and q waves. Relative wall thickness of greater than 0.45 suggests hypertrophic cardiomyopathy. Relative wall thickness is calculated as a sum of inter-ventricular septal thickness in diastole and posterior wall in diastole divided by left ventricular end diastolic dimension. On echocardiogram presence of asymmetric hypertrophy, impaired diastolic dysfunction, myocardial fibrosis, and non-regression of hypertrophy with de-training are all indicative of hypertrophic cardiomyopathy. An athlete will have peak VO_2 greater than 120% of predicted. All these features help separate athlete's heart from hypertrophic cardiomyopathy.

It was very encouraging to note that outcomes of CHD were being looked at all over the world. There were several presentations and abstracts from India and South Africa on the topic. The talk covered the neurodevelopment impairment seen after open heart surgery in children. All domains – motor, language,

behaviour, and learning – of neurodevelopment function can be affected. These impairments have a high prevalence but low severity and often exist in combination. There was a great emphasis on providing early intervention to these children. This topic highlighted that the focus of CHD was not limited to survival alone but quality of life of the child was equally important.

Electrophysiology was discussed in detail in a separate section; however, in the ambulatory section drugs causing QT prolongation were discussed. There is a general increase in the number of rare variants in arrhythmia genes, and in metabolic and mitochondrial diseases. There is an increased recognition of patients who are a slow metabolisers of CYP3, which can augment the QT prolongation with medications. Psychotropic medications can prolong QT interval with greatest risk in patients with dual therapy or other underlying conditions like anorexia nervosa, and hypothyroidism or with medications like macrolide antibiotics. There was additional discussions on premature ventricular contractions. It was important to classify these premature beats as benign or not. Those that were thought to be benign did not require any further intervention.

Pulse oximetry screening for CHD was discussed in various platforms. There were abstracts, talks, and panel discussion on the topic. Screening with pulse oximetry can help detect many, but not all, cases of critical CHD. Pulse oximetry may need to be modified and customised for the community, like the out-of-hospital deliveries. Pulse oximetry can be extended to detect babies with low saturations due to other causes like pneumonia, persistent pulmonary hypertension, and sepsis. Furthermore, the panel strongly felt that it was important to get a count of the total number of the children in the world with CHD to allocate resources and to develop infrastructure to take care of children with CHD.

General paediatric issues like immunisation as it relates to children with heart disease were discussed. Despite world-wide efforts to increase vaccinations, many still refuse or delay at least some vaccines. Every attempt should be made to vaccinate *before* heart transplant. For vulnerable groups, all caretakers and household contacts should be vaccinated. Alternative anti-platelet agents should be substituted for aspirin therapy when the varicella vaccine is administered.

Non-cardiac surgery in children with CHD was also discussed. Children with moderate or severe heart disease have a greater morbidity and mortality with non-cardiac surgery. A skilled multidisciplinary team approach is needed to provide care to children during non-cardiac surgery. Subacute endocarditis prophylaxis is indicated for high-risk groups.

Rheumatic heart disease

Rheumatic heart disease still continues to be a major cause of cardiac disease in the world. Global Burden is estimated as 15.7 million with 282,000 new cases added per year and 23,000 deaths per year – these numbers maybe underestimated. Diagnosis by echocardiogram is reliable. Prophylaxis can be provided using injectable penicillin and is thought to be effective and inexpensive. There is a high global need for increased hospitals and surgeons to provide care to these children. There continues to be a controversy whether echocardiogram should be used as a diagnostic tool to increase the detection of rheumatic heart disease. Increasing the pool of patients with rheumatic heart disease may further burden the already limited resource and capacity in low to mid income countries.

The modified Jones criteria for rheumatic fever were debated upon. There has been a revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography (<http://www.acc.org/latest-in-cardiology/ten-points-to-remember/2015/05/08/15/22/revision-of-the-jones-criteria-for-the-diagnosis-of-acute-rheumatic-fever>).

The need for diagnostic criteria was felt to be necessary to facilitate secondary prophylaxis and to avoid under- and over-diagnosis of rheumatic heart disease. It was argued that it is difficult to get accurate epidemiological data and laboratory data in low income countries, which makes it difficult to use the modified protocol. However, no single test can estimate the true probability of the disease. Studies in Africa showed that only 50% of the children with rheumatic heart disease had a positive anti streptococcus A. Exposure and without penicillin, after 20 years in pre-penicillin era, causes 90% of patients with clinical carditis as a result of recurrences. Further fine tuning of the Jones Criteria was thought to be an intellectual exercise for the developed world and has less relevance for the developing world. Lack of resources make it an overwhelming task to implement the modified Jones criteria. Availability of laboratory tests and echocardiography and non-availability of Penicillin provide additional challenges to utilise the modified Jones criteria.

Novel immunological mechanisms in rheumatic fever were discussed. Rheumatic fever and rheumatic heart disease are under the control of several genes. Specific integrins, selectins, chemokines, and cytokines mediate the inflammatory process of rheumatic fever and progression of the valve damage, leading to rheumatic heart disease. Vimentin, collagen VI, and lumican proteins are major targets of rheumatic heart

disease—valvular autoimmune process. Molecular mimicry, epitope spreading and T-cell degeneracy, and low numbers of T regulatory cells are the mechanisms that mediate rheumatic fever and rheumatic heart disease lesions.

In summary, while it is very true that echo screening enables identification of many more cases of rheumatic heart disease when compared with clinical evaluation, there are important issues that need to be considered before echo screening can be recommended as a public health strategy for rheumatic heart disease prevention. The challenges in low resource environments with rheumatic heart disease are massive. They relate to massive disease burden and dysfunctional health systems. Currently, it is difficult to deal with the burden of clinical rheumatic heart disease. Adding a massive burden of subclinical rheumatic to this will completely overwhelm fragile health systems in low- and middle-income countries. There are very good examples of success stories with rheumatic control that do not involve the use of echo screening. Implementation of penicillin prophylaxis is the single biggest challenge in rheumatic heart disease prevention. There is no one willing to manufacture good quality injectable benzathine penicillin today. Price control policies makes this unprofitable for pharmaceutical companies. Most primary health centers are unwilling to administer injectable penicillin for fear on anaphylaxis – this is justifiable because there have been a fair number of fatal events. This is believed to result from impurities and not penicillin itself. Quality control is a huge issue. The available preparations are of low water solubility and really painful. Oral penicillin is a poor substitute but this too is unavailable.

The 7th world congress was truly a thought-provoking meeting that tried to bridge the geographical, intellectual, and interdisciplinary differences so that the future of a child is no longer decided by imaginary lines on the earth and every child has an equal chance of living a full life.

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Conflicts of Interest

None.