Conference report - The Cardiac Patient from Birth to Adulthood

THE MEETING
International experts gathered in Sweden in late February to discuss the art of adult congenital heart disease and the transition from pediatric to adult cardiology. Held at the Swedish Society of Medicine in Stockholm, it was jointly organised by Acta Paediatrica, Journal of Internal Medicine and The Swedish Society of Medicine. The aim of the symposium was to present the latest state-of-the-art developments in areas such as adult congenital heart disease, the early development of atherosclerosis, arrhythmia and the cardiovascular outcomes of prematurity. There was a special focus on the transition from paediatrics to adult cardiology and these were illustrated by clinical case presentations. The seminar also discussed how to prevent cardiovascular diseases in the womb and during early childhood.

THE TOPIC
The causes of congenital cardiac diseases often seem to be unknown, but genetic factors are thought to be important and may affect the phenotype in the embryo. Acquired diseases, such as coronary heart disease, hypertension and stroke, may also originate during fetal and early postnatal life, due to environmental factors that cause metabolic and developmental programming. These go on to affect blood pressure and metabolic homeostasis. That is why the prevention and treatment of cardiovascular diseases hypothetically must be initiated as early as possible, even before conception.

Surgical treatment of congenital heart disease (CHD) in small children has resulted in a remarkable increase in the survival rate and treating the fetus may further improve outcomes in the near future. Up to the end of the 20th century, many of these children used to die before they reached adulthood. Although most adults with CHD seem to achieve a relatively good quality of life, there are many residual problems. In many cases, the long-term prognosis is still unknown and this is a new challenge for physicians working in adult medicine.

THE SESSIONS
Adult congenital heart disease: past, present and future

Advances in diagnosis and management mean that more than 90% of children with CHD survive well into adulthood. Michael A Gatzoulis, Royal Brompton Hospital and National Heart and Lung Institute, UK, attributed this to remarkable advances in paediatric cardiology, cardiac surgery and catheter interventions. However, they stated that many patients face residual and progressive haemodynamic lesions, exercise intolerance, arrhythmias, heart failure and premature death. Clinicians urgently need to understand late pathophysiology better and provide evidence about drug therapy, devices and transplantation.

The changing epidemiology in congenital heart disease

Ariane Marelli, McGill University Health Center, Canada, discussed the impact of demographic shifts in healthcare delivery and research directions. CHD is a lifespan condition that requires special consideration with regard to how care is delivered and in terms of the knowledge gaps that need to be addressed. The learning points presented centred around the importance of aligning paediatric and adult cardiology to tackle the challenges that lie ahead in the next decade.

Atherosclerosis in patients with congenital heart disease

Coronary heart disease is the single most common cause of death before the age of 55 in Europe and is rapidly increasing in countries like India, where it was previously rare. Eero Jokinen, University of Helsinki, Finland, pointed out that children and adolescents with acyanotic CHD have almost the same risk profile as their healthy peers, but patients with cyanotic heart defects are protected against atherosclerosis. The risk possibly returns to the average level when the defect becomes acyanotic after surgery.

Arrhythmia interventions in complex congenital heart disease

Sabine Ernst, Royal Brompton Hospital, UK focused on state-of-the-art ablation techniques for both atrial and ventricular arrhythmias in patients with CHD. Catheter ablation is now one of the pillars of managing sustained tachycardia in patients with CHD. Recent improvements in mapping and ablation technology have improved ablation outcomes substantially in this complex patient group. The success of atrial or ventricular tachycardia ablation in CHD is influenced by the
underlying cardiac anatomy and surgical repairs, along with the current haemodynamic sequelae of the anatomy and repairs.

**Single ventricle physiology**

Marc Gewillig, Catholic University of Leuven, Belgium, described various aspects of the Fontan operation, which still after nearly five decades is the final palliation for patients with univentricular hearts. Although thousands of operations have been carried out, mortality and morbidity remain high. Standard paradigms of management have been religiously pursued, with minimal effects on outcomes. The major determinants of outcome are: exercise intolerance, acute and chronic heart failure, cyanosis, arrhythmias, thromboembolism, protein losing enteropathy, plastic bronchitis, liver disease, renal dysfunction and premature death.

**Physical capacity in adults with congenital heart disease**

Gerhard-Paul Diller, University Hospital Munster, Germany, stated that exercise capacity is reduced in adults with CHD. Studies have shown associations between impaired objective exercise capacity and poor outcome in the mid-term and for cardiac procedures. International recommendations encourage regular exercise, but the benefits are not clear. However, epidemiology reports suggest that only a minority of patients die during exercise and the risk is relatively low.

**Quality of life in adult patients with congenital heart disease**

Philip Moons, University Hospital Leuven, Belgium, maintained that most published studies on CHD and quality of life demonstrated substantial methodological and conceptual limitations and geographic differences. Quality of life studies that were based on sound conceptual grounds could enable international comparisons. Three key learning points were identified. First, different conceptual approaches should be taken into account when interpreting findings. Second, adults with CHD have a good overall quality of life that can be even better then healthy individuals with regard to satisfaction with life. Third, country-specific factors can predict quality of life above and beyond patient-related factors.

**Persistent foramen ovale in cryptogenic stroke**
A patent foramen ovale has since many years been associated with cryptogenic strokes but previous intervention trials have been negative. Lars Søndergaard, Rigshospitalet Copenhagen, Denmark, said that the link between ischaemic stroke and paradoxical embolism, via patent foramen ovale, has recently been extensively studied. Three international randomised clinical trials have now demonstrated the therapeutic benefit of adding percutaneous patent foramen ovale closure to anti-platelet therapy to reduce the risk of recurrent ischaemic stroke in populations with previous cryptogenic strokes. Although the risk of recurrence is fairly low, it can be further reduced by two thirds by closing a patent foramen ovale. Guidelines should be updated to specify when a patent foramen ovale should be closed in such patients.

Evolutionary aspects on the origin of cardiac disease

Johan Frostegård, Karolinska Institutet, Sweden, said that his team had been studying how oxidized phospholipids, especially phosphorylcholine, played a role in immune activation induced by oxidized low density protein. Evolutionary factors could play both a direct and indirect role in atherosclerosis. These included low levels of anti-phosphorylcholine, due to a lack of exposure to microorganisms that have co-evolved with humans for millions of years. This leads to an immune deficient state, which predisposes an individual to atherosclerosis.

The inheritance of cardiovascular disease risk

Mark Hanson, Institute of Developmental Sciences and NIHR Southampton Biomedical Research Centre, UK, emphasised that interventions to prevent cardiovascular disease need to start before conception. These should promote healthy behaviours in prospective parents, as well as during pregnancy and postpartum. The risk components of cardiovascular disease are transmitted across generations by socio-economic and biological factors and epigenetic mechanisms can also be involved. Inheriting cardiovascular disease risks, and the role that social inequalities play in those risks, raises wider questions about responsibility for the health of future generations at societal and individual levels.

Early life origins of adult cardiovascular disease

Atul Singhal, UCL Great Ormond Street Institute of Child Health, UK, discussed
the idea that there is a critical window for nutrition that may permanently affect, or programme, long-term health. This theory was strongly supported by the benefits of breastfeeding on the risks of obesity and cardiovascular disease. It was also supported by early experimental randomised studies of preterm infants randomly assigned to human milk and some systematic reviews. The need for experimental studies in nutrition and the implications of nutritional programming for nutritional, clinical and public health practice was emphasised.

Preterm birth and cardiovascular risk

It is important to understand the potential role that preterm birth plays in adverse cardiovascular health in adult life, according to Mikael Norman, Karolinska Institutet, Sweden. Follow-up studies of children and young adults born preterm suggest some alterations in the structure and function of the cardiovascular system, elevated blood pressure and glucose intolerance. Whether or not these changes in physiology reflect early adaptations after preterm birth or genetic traits needs to be established. Other unresolved questions include if, and how, preterm birth results in a significant risk increase for cardiovascular disease late in life.

ABBREVIATION

CHD, Congenital heart disease