

Workshop Abstracts

The genetics of cardiac arrhythmias

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The spectrum of the contribution of genetics to the genesis of cardiac arrhythmias is broad. At the simplest level, the monogenetic disorders are the result of mutations in single genes. The mutations tend to be “private”; they are specific for a particular family. They result in “clue diseases” that have provided a wealth of information on the electrophysiology of the heart and the disturbances that lead to arrhythmias. They provide models of more common diseases. Polymorphisms are variations in single genes that are a basis for variations in disease susceptibility. Diseases such as hypertension and atrial fibrillation are polygenetic disorders. Small variations in multiple genes that do not by themselves cause disease provide the heritable component of common diseases. This presentation shall focus on the monogenetic disorders that are the basis for certain cardiac arrhythmias. These are primarily the result of mutations of the Na⁺, K⁺, and Ca²⁺ channels and their regulatory genes that control the generation of the cardiac action potential. The functional disturbances produced include: slow conduction, delayed repolarization, sometimes accompanied by sensorineural deafness, and rapid repolarization.

The contributions of molecular genetics and contemporary membrane biophysics have engendered remarkable advances in these areas. I have selected illustrative examples of these syndromes for presentation. Increased awareness of these uncommon entities may lead to their early identification and treatment.

In 1992, Pedro and Joseph Brugada described an autosomal dominant syndrome of apparent right bundle branch block, recurrent syncope and sudden death from ventricular fibrillation. Although the syndrome is autosomal dominant, the phenotype is predominantly seen in males. In South East Asia, the syndrome may affect 1% of the population. Mutations in cardiac Na⁺ and Ca²⁺ channel genes have been identified as the basis of the syndrome in 20–40% of affected patients. The mutations result in slow conduction and rapid repolarization of the cardiac action

potential. The Jervell and Lange-Neilsen syndrome is an autosomal recessive disorder associated with prolongation of the QT interval on the electrocardiogram (ECG), sensorineural deafness, the stereotypic polymorphic ventricular tachycardia of torsade de pointes and sudden death. The Romano-Ward syndrome is a related disorder inherited as an autosomal dominant and associated with normal hearing. Collectively, these constitute the inherited long QT syndrome (LQTS). Mutations in K⁺ channel genes or their regulatory proteins account for the majority of LQTS cases.

The mutations result in delayed cardiac repolarization; mutation of the same channel in the inner ear account for the sensorineural deafness. An analogous disorder short QT syndrome, the result of over expression of K⁺ channels, results in rapid repolarization, shortening of the QT interval and polymorphic ventricular tachycardia.

Catechominergic polymorphic ventricular tachycardia (CPVT) is a disorder of the intracellular proteins that regulate calcium homeostasis. Catechominergic polymorphic ventricular tachycardia produces a characteristic bidirectional ventricular tachycardia. It may be inherited as an autosomal dominant or recessive disease. Characteristically, arrhythmias are precipitated by exercise. All these syndromes are electrical diseases; the heart is structurally normal and the prognosis is good if sudden death can be prevented. The implantable cardioverter-defibrillator (ICD) is the mainstay of treatment. Drugs, primarily β -blockers play a role in the treatment of specific variants of these syndromes; cardiac denervation and catheter ablation play a minor role.

A second group of disorders is associated with gross structural abnormalities of the heart. Arrhythmogenic right ventricular dysplasia/cardiomyopathy is the result of mutations in the gap junction proteins between cardiac cells. It results in fibro-fatty infiltration of the right ventricle and monomorphic ventricular tachycardia. The hypertrophic cardiomyopathies and certain of the dilated cardiomyopathies are the result of mutations in the contractile protein genes and the energy-generating mechanism in the cardiac cell. The force generating mechanisms is impaired and pathological hypertrophy or dilation occurs. Although sudden death may be prevented by ICD implantation,

patients with these structural cardiomyopathies may eventually succumb to heart failure.

Screening by genetic testing is now available commercially. Mutations at specific loci such as the pore region of ion channels may carry a particularly high risk of sudden death. The identification of a particular mutation in the proband may identify asymptomatic family members at risk for disease. They may also inform lifestyle modification. The syndromes discussed are uncommon disorders. However, there is a high premium on early identification and treatment.

Surgical treatment of atrial fibrillation

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Objective: The Cox maze procedure was introduced in 1987 for the treatment of atrial fibrillation. This study evaluated the predictors of late atrial fibrillation recurrence in 276 consecutive patients who underwent this procedure at our institution.

Methods: From 1987 through June 2003, 276 patients (79 female and 197 male patients; mean age 55 ± 11 years) underwent the Cox maze procedure. Thirty-three patients had Cox maze procedure I, 16 patients had Cox maze procedure II, and 197 patients had Cox maze procedure III. The last 30 patients underwent a modified procedure (Cox maze procedure IV) with bipolar radiofrequency ablation. There were 113 (41%) patients who had a concomitant operation, most commonly either a mitral valve procedure (19%) or coronary artery bypass grafting (20%). Data were analysed by means of univariate analysis, with preoperative and perioperative variables used as covariates. Patient follow-up was conducted by means of questionnaire, physician examination and electrocardiographic documentation. All patients had a minimum of six months of follow-up.

Results: Patient follow-up was achieved in 92.8% of cases, with a mean follow-up time of 5.8 ± 3.6 years. Risk factors for late atrial fibrillation recurrence were duration of preoperative atrial fibrillation ($p = 0.01$) and Cox maze procedure version ($p = 0.001$). There was no difference in actuarial 10-year survival between the Cox maze procedure versions.

Conclusion: The Cox maze procedure remains the gold standard for the treatment of atrial fibrillation and has excellent long-term efficacy. The most significant predictor of late recurrence was duration of preoperative atrial fibrillation, suggesting that earlier surgical intervention would further increase efficacy.

Cholesterol and cardiac disease

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Seventy-five per cent of the cholesterol used by the body is endogenously synthesized. The remaining 25% is derived from the diet. The metabolism of lipids including cholesterol is inseparable from the synthesis, interconversion and catabolism of the lipoproteins which include very low density lipoproteins (VLDL), intermediate density lipoproteins (IDL), low density lipoproteins (LDL), high density lipoproteins (HDL) and chylomicra.

Low plasma HDL or increased LDL could result in atherosclerosis. It has been established that once the LDL level exceeds a critical concentration, the process of endothelial desquamation sets in in the vessels with highest blood pressure including the coronary arteries. This is exacerbated by other synergistic factors such as hypertension, hyperglycaemia and hormonal dysfunction. The partial or total occlusion of the coronaries results in a reduced supply of nutrients and oxygen to the heart, eventually resulting in angina and myocardial infarction.

Renal ischaemia leads to increased renin production which ultimately results in increased angiotensin II. The latter elicits two systemic actions both of which cause increased blood pressure. Persistent, untreated hypertension can lead to cardiomegaly which can also result in myocardial infarction.

A patient with hypercholesterolaemia is best treated initially by lifestyle changes. If the desired levels of cholesterol are still unattainable then the introduction of drug therapy is necessary. Statins inhibit the rate-limiting step of the cholesterol biosynthetic pathway and could, therefore, be the drug of choice.

HIV and cardiovascular disease

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HIV infection results in chronic disease and, if left untreated, AIDS-related death for almost all infected persons. Over the last decade, HIV-related morbidity and mortality have decreased dramatically due to improved antiretroviral therapy (ART) access. Although ART can markedly increase the lifespan of persons living with HIV/AIDS, the life expectancy of these individuals remains shorter than that of HIV-uninfected persons, even with optimal ART treatment. The majority of these premature deaths are attributable to chronic inflammatory diseases not associated with AIDS, of which cardiovascular disease is a leading

cause. Chronic usage of certain antiretroviral drugs can result in dyslipidaemia and increased risk of death due to cardiovascular disease. However, most antiretroviral drugs are not associated with an increased risk of cardiovascular events, yet HIV-infected persons treated with these drugs still have an increased risk of premature death due to cardiovascular disease. Recent observations demonstrate that HIV-infected persons treated optimally with ART display low levels of chronic inflammation, which is likely to contribute to premature cardiovascular events. These data demonstrate that HIV infection is a cardiovascular disease risk factor that should be considered when managing HIV-infected patients.

Brain natriuretic peptide in heart failure

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The aim of this presentation is to outline the mechanisms of brain natriuretic peptide (BNP) in the management of heart failure.

Heart failure can be defined as the inability of the heart to meet the demands of the body in terms of a relative reduction in the stroke volume. Hence, determining the ejection fraction is the gold standard in evaluating heart failure.

The clinical assessment of heart failure is difficult especially in the clinical practice of primary physicians, because the signs and symptoms of heart failure are non-specific. Furthermore, the prognosis of heart failure remains poor despite immense progress in the treatment of it.

The heart defends itself by utilizations of Starling's Law as well as the release of BNP as a hormone by the ventricles, and both of these mechanisms are induced by stretch of the ventricles due to volume/pressure changes during the cardiac cycle. The interaction between the actin and myosin filaments in the myocyte forms the basis of the Starling's Law and determines the contractile strength of the heart, whilst the release of BNP causes vasodilation and natriuresis. Vasodilation reduces the afterload to enhance adequate ejection and furthermore decreases the preload, since the venous return will decline. Additionally, natriuresis decreases the blood volume leading to reduction in the venous return and end-diastolic volume (EDV).

The mechanisms of Starling's Law and the release of BNP therefore protect the heart from volume overload that could lead to heart failure when the ventricles are overstretched. The function of the heart as stated here represents one of the key biological findings of how the structure of an organ is related to its function, and in particular now why the heart is also an endocrine organ. The stretching of the heart initially leads to compensated, but overstretching induces decompensated heart failure.

Brain natriuretic peptide can therefore be assessed as a marker to determine the magnitude of heart failure, since its level will reflect the volume/pressure changes in the ventricle during the cardiac cycle. Serial measurements of BNP levels could form the basis for diagnosis, the monitoring of treatment and the prognosis of heart failure.

Research in BNP and heart failure is therefore warranted not only in improving the treatment, but also in reducing the cost of managing heart failure.

Nutritional approach to heart disease: a review of the various diets

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Background: Dietary approaches are assumed to be efficacious and cost-effective in reducing cardiovascular disease (CVD) mortality and morbidity and are included as part of risk factor management interventions.

Objectives: A scoping review conducted to assess the effects of varying nutritional strategies on preventing or reducing CVD and its risk factors.

Methods: A search for systematic reviews (SRs)/meta-analysis examining nutritional approaches to coronary heart disease (CHD) prevention and reduction was conducted. Those examining dietary patterns and not single nutrients or foods were selected. Cardiovascular disease outcomes assessed included elevated blood pressure, excess weight, elevated blood lipids and CHD. Systematic reviews and meta-analyses were appraised using the "Practical Evidence About Real Life Situations (PEARLS)" approach.

Results: A total of 13 SRs and one overview of reviews were examined. Two main dietary approaches were studied in clinical trials and cohort studies 1) Mediterranean diet and 2) Dietary Approaches to Stop Hypertension (DASH) diet. Findings from meta-analyses showed reductions in total CHD (RR = 0.79; 95% CI, 0.71, 0.88) for persons following DASH-style diet with pre-existing risk factors. Adherence to the Mediterranean diet was associated with a 9% reduction in CVD mortality (RR = 0.91; 95% CI, 0.87, 0.95). Overall, both the DASH-style and Mediterranean diets showed reductions in blood pressure, total cholesterol and low density lipoprotein-cholesterol. Other approaches such as dietary salt reduction and reduced or modified dietary fat by itself for preventing CHD were not supported by the evidence.

Conclusion: Dietary approaches may be beneficial in the reduction and prevention of CHD but are limited to two dietary patterns assessed in SRs. Other dietary pattern approaches to assess primary, secondary and tertiary CVD prevention need to be conducted.

Rheumatic heart disease – a public health approach

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Rheumatic heart disease (RHD) is the most common cardiovascular disease in children and young adults and affects about 20 million persons worldwide. It is a huge public health burden in third world countries. Rheumatic heart disease results from rheumatic fever (RF) which follows throat infections with Group A streptococcus in some persons. The heart valves are damaged by RF and become incompetent or stenotic. It is a disease of the poor in developing and middle-income countries and of indigenous people in some wealthy nations.

There are 252 000 new cases and 233 000–468 000 deaths annually from the disease. The complications of RHD are very costly to treat and reduce the lifespan and potential of large numbers of young people. Although preventable by standard public health approaches which include surveillance, identification of risk and protective factors and the development, implementation and evaluation of interventions, RF and RHD have been neglected by governments, the medical community, the media and funding agencies.

The prevalence of RHD has been significantly underestimated because of inadequate data. Resources for research and training of health personnel have been lacking and patients have had limited access to healthcare. Screening for RHD by echocardiography in recent years has indicated that the burden of the disease has been significantly underestimated in the past. Longitudinal study of subclinical RHD cases identified by echocardiography should reveal the natural history, appropriate management, especially as regards secondary prophylaxis, and the feasibility and cost-effectiveness of widespread screening by this method.

Primary prevention of hypertension

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Prevalence of overweight/obesity and co-morbidities including hypertension are high and rising, and 80% of global burden of chronic disease resides in low and low-middle income countries. These are the very countries which do not have resources to employ pharmacologic treatment as their primary strategy. Prevention is globally recognized as a preferred pathway to disease reduction but secondary prevention through lifestyle changes have proven difficult to impossible to institute at population level. Primary prevention is still being conceived and thus presents an attractive opportunity for researchers. Devel-

opmental entrainment of functional equilibrium set points and adaptive capacities contribute substantially to risk of obesity and its co-morbidities. One high-value question concerns whether we can identify molecular markers of functional capability and capacity, and use such markers to accelerate the shift in offspring biology toward disease resistant phenotypes.

Metabolic syndrome in Jamaica and the implications for heart disease

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The metabolic syndrome as defined by Grundy *et al* is “...a constellation of interrelated risk factors of metabolic origin that appear to directly promote the development of atherosclerotic cardiovascular disease”. The syndrome emerged as an important clinical entity over the last two decades following Gerald Reaven’s description of a clustering of risk factors for coronary artery disease in 1988. Current diagnostic criteria for metabolic syndrome include five components, namely: central obesity, glucose intolerance, elevated triglycerides, low levels of high density lipoprotein cholesterol (HDL), and hypertension. Several organizations have proposed diagnostic criteria for the metabolic syndrome resulting in varying estimates based on the criteria used in specific studies. In order to harmonize these diagnostic criteria, the International Diabetes Federation, American Heart Association, National Heart, Lung, and Blood Institute along with other international groups proposed unifying diagnostic criteria with components as listed below:

- * Elevated waist circumference (using ethnic specific cut points): men ≥ 94 cm, women ≥ 80 cm for Europeans and blacks; men 102 cm, women 88 cm for North America; men 90 cm, women 80 cm for Asians
- * Elevated triglycerides: ≥ 1.7 mmol/L
- * Reduced high density lipoprotein (HDL) cholesterol: men < 1.0 mmol/L, women < 1.3 mmol/L
- * Elevated blood pressure (BP): systolic BP ≥ 130 mmHg or diastolic BP ≥ 85 mmHg
- * Elevated fasting glucose: ≥ 5.6 mmol/L

Persons are classified as having the metabolic syndrome if they satisfy any three of the five components.

Worldwide, most prevalence estimates for the metabolic syndrome range from 8%–24% among men and from 7%–46% among women. In Jamaica, the prevalence of the metabolic syndrome in the Spanish Town Cohort was 27.6% among women, 10.6% among men, and 21.1% for men and women combined. Prevalence of the metabolic syndrome varied with socio-economic status, with signifi-

cantly increased odds among men with higher income and education. Among Jamaican youth, the prevalence of the metabolic syndrome was relatively low, with an estimated prevalence of 1.2% among participants of the 1986 Jamaica Birth Cohort Study. However, over 50% of participants in that study had at least one component of the metabolic syndrome.

These data suggest that large proportions of the Jamaican population may be at relatively high risk for cardiovascular disease and diabetes. More public health measures to ameliorate these risks should be implemented.

Coronary angiogram and angioplasty at the University Hospital of the West Indies

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While coronary angiography has been performed at the University Hospital of the West Indies (UHWI) for more than 30 years, regular angioplasty and stenting started in 2002 with the return of two former graduates each from the United States of America (USA) and Canada. Over the past 10 years, the number of angioplasty and stenting cases have more than tripled and the complexity of the cases have increased. There has been the growth of the programme to involve the deployment of drug eluting stents, performing regular emergency primary percutaneous coronary intervention (PCI) along with rescue and facilitated interventions and increased number of cases using the radial approach. The programme was also innovative, as Jamaica was the first country in the western hemisphere to use the TREK® balloon and the programme was the first in the island to perform renal angioplasty in 2008, deploy the 'StarClose' vascular closure device, deploy coated stents, use filter wires, and to introduce intravascular ultrasound and recently rotoblation technology. With these advances, we have managed to keep our complication rates within international standards. In 2012, for the first time, over 500 cases were performed. The programme was also the nidus for the development of a cardiac nursing programme in 2011.

What the physician needs to know about pacemakers

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With improved access to medical devices such as pacemaker generators, increasing numbers of Jamaican patients

have received implanted pacemaker generators. Physicians should be familiar with the indications for pacemaker insertion and the management of patients with implanted pacemakers. This presentation will seek to address the important information that physicians should know.

Out-of-hospital cardiac arrest in adults

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In the United States of America (USA), 300 000 adult out-of-hospital cardiac arrest (OHCA) events occur each year, and the majority (70–85%) is due to a cardiac cause. Once arrest has occurred, the outcome is impacted by the timeliness of cardiopulmonary resuscitation (CPR) and where appropriate, defibrillation. Factors associated with delays in accessing emergency medical services for cardiac emergencies include the presence of a relative, lower socio-economic status, underestimation of symptom severity and age in females > 65 years. Survival is highest for emergency medical services (EMS) witnessed cardiac arrest. The survival from OHCA where the initial rhythm is ventricular fibrillation is higher than for other rhythms. Patients who have return of spontaneous circulation (ROSC) after the first shock have a higher survival rate than those who require three or more shocks. Defibrillations are most effective when they are administered within the first six minutes of arrest. The use of advanced cardiac life support (ACLS) drugs has improved the ROSC but not the survival to hospital discharge.

Given the limited EMS resources in Jamaica, the initial focus should be on providing timely effective CPR and defibrillation. Ideally, this should be commenced by trained laypersons and continued by EMS personnel. Automated external defibrillators should be placed strategically in public spaces.

Cardiac rehabilitation: high value, low usage

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Cardiovascular disease (CVD) remains one of the leading causes of death globally and locally. Cardiac rehabilitation (CR) involves making lifestyle changes, such as regular participation in exercise, to attenuate or reverse the progression of CVD, thereby decreasing the cost of care. Persons with stable heart disease and who have no contraindications to exercise training are generally eligible to participate in cardiac rehabilitation programmes. In spite of this,

most persons with CVD do not attend cardiac rehabilitation; further, studies have shown that up to 50% of eligible patients who are referred do not attend. In view of the growing burden of CVD in Jamaica and the incontrovertible evidence for the effectiveness of cardiac rehabilitation in reducing that burden, healthcare practitioners and policy-makers should institute systems which will facilitate increased patient participation in cardiac rehabilitation. This presentation will examine the reasons for low patient participation in cardiac rehabilitation and suggest feasible changes in the healthcare system which may facilitate increased patient participation.

Preventing re-admission for heart failure: the role of nurse-led heart failure clinics

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Objective: To provide support and monitoring services through a nurse-led heart-failure clinic at the University Hospital of the West Indies (UHWI) and to prevent/reduce re-admissions for patients with heart failure at the UHWI.

Methods: Specially trained cardiac nurses will meet with in-hospital patients with a diagnosis of heart failure. An initial nursing assessment that includes the patient's history, a complete physical examination, the learning needs of the patient, and level of available psycho-social support will be made. Just prior to discharge, the nurse visits and arranges the appointment for the nurse-led clinic. This first visit must occur within one week after discharge and will yield an opportunity for individualized, intensive post-discharge care. Care activities in the clinic will include specifically tailored education at the patients' level, medication review, post discharge laboratory work, medication titration and identification of high risk patients. Subsequent follow-up will be undertaken at fixed intervals or may be scheduled as needed. A home visit will give additional insight into the psycho-social environment of the patient and identify gaps in care. While the clinic will be led by nurses, a multi-disciplinary healthcare team approach will be integral to the successful functioning of the clinic.

Results: Early identification of, and prompt intervention for non-adherence to medication and dietary restrictions, and improvement of the patient's health literacy will reduce the re-admission rates for patients with heart-failure.

Conclusions: Nurse-led heart failure clinics have the potential to prevent and/or reduce re-admissions, and have favourable effects on quality of life, survival results and care costs both to the institution and patients involved.

A mother's experience of heart surgery in her child and the struggles

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Treating children with heart disease in developing countries is one of the greatest challenges. Many children born in developing countries with congenital heart disease will never see their first birthday. The resources required to treat and cure congenital heart disease are not easily accessible. Skilled medical teams (doctors, surgical and intensive care nurses, and other support staff), medicines, equipment and facilities are scarce or non-existent. Our possible options for ensuring that Jamaican children receive the heart surgery they need are:

- * Referring patients to developed countries
- * Having overseas surgical teams travel to Jamaica to perform heart operations
- * Developing a specialized centre for heart surgery

A mother shares her experience of how her infant with a congenital heart defect was able to receive surgery by an overseas visiting team working alongside local doctors and nurses.

Cardiac surgery in Jamaica: reflections on the programmes at the University Hospital of the West Indies and Bustamante Hospital for Children

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Aim and Objectives: This was an observational, and both retro- and prospective study. The main aim of which was to determine 1) the number and types of cardiac cases performed over the past 50 years at both the University Hospital of the West Indies (UHWI) and Bustamante Hospital for Children (BHC), 2) past and present factors that hinders the growth of cardiac surgery and to make collective team-based recommendations regarding the future of cardiac surgery.

Methods: The following information was retrieved from the cardiac databank files: 1) number and type of cases, 2) gender, 3) cardiac risk score and 4) outcome.

Results: Over 3000 patients have benefited from cardiac surgery over the past 50 years. The number of cases per year has steadily increased due to increased availability of intensive care unit (ICU) facilities, including ICU-trained nurses, beds and other resources. The assistance of several overseas cardiac surgical teams has also served to decrease the backlog of patients waiting for cardiac surgery, at both hospitals. The number of paediatric surgery at UHWI, done mainly to correct congenital heart disease, has decreased over the past five years, coinciding with the

employment of a cardiac surgeon at the BHC. In the first thirty years, cardiac cases were mainly correction of congenital heart disease and valve replacement or repair, the latter due mainly to rheumatic heart disease. The next twenty years saw a decrease in valve surgery and a persistent increase in coronary artery bypass surgery (CABG) for coronary artery disease (CAD) and aortic surgeries such as aneurysm and dissections. This latter type of surgery has also increased steadily among females. Patients who are

diabetic, hypertensive, have renal impairment and/or of Indian descent tend to have a severe form of CAD. Factors which are needed to sustain the cardiac service and to maintain high quality care are: i) reliable supplies of drugs, equipment and other resources needed for the service, ii) retention of trained ICU nurses, iii) development of a dedicated cardiac ICU and iv) increased numbers of cardiac surgeons and perfusionists at both hospitals and support services such as renal, cardiology and haematology.