LQTS is characterized by delayed repolarization of individual action potentials and ECG, potentially resulting in QT interval prolongation and subsequent tachyarrhythmias [Roden DM et al. *J Cardiovasc Electrophysiol* 1999]. The most common LQTS genotype that is associated with sudden cardiac arrest at age <40 years is LQT1, though LQTS patients maintain a high risk for life-threatening cardiac events even after age 40. β-blockers are effective for treatment of LQTS, particularly among patients with LQT1 and LQT2 genotypes ($p<0.001$) [Moss AJ et al. *Circulation* 2000].

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a genetically transmitted cardiac ion channel disorder with an extremely high risk of SCD. If untreated, approximately 50% of individuals will present with a cardiac event that is possibly lethal by the age of 30 years. First recognized in 1975, this disorder involves mutations that lead to loss of calcium homeostasis in the heart, resulting in malignant arrhythmias.

The most common symptom is syncope, usually during periods of emotional or exertional stress. Syncope most often appears during the first or second decade of life. It is important to distinguish this from LQTS, since β-blocker therapy in individuals with this syndrome is less effective. In addition, the cardinal finding of prolongation of the QT interval that is seen in LQTS is absent in this disorder [Liu N et al. *Progress in Cardiovascular Diseases* 2008].

ARVD is a genetically determined disease of the heart muscle that is associated with arrhythmia, heart failure, and SCD. It is thought to be most common cause of SCD in the young in European countries. Unfortunately, SCD is often the first clinical manifestation of ARVD, especially among young people who are engaged in strenuous activity. While ARVD is often characterized by intramyocardial RV fatty infiltration, as seen on MRI, Dr. Stephenson cautions that reliance on this diagnostic indicator alone is not sufficient. Therefore, it is important to determine the diagnosis of ARVD, based on the composite of a number of possible findings, including ECG abnormalities (T-wave inversion in V1 to V3, RBBB, and/or epsilon wave), right ventricular structural abnormalities that are seen on imaging, abnormal myocardial biopsy, positive family history, and genetic testing. No one criterion is robust enough to adequately diagnose ARVD with confidence.

Detecting an increased risk of SCD in young patients is a clinical challenge, and often this risk is not detected until an event occurs, at which point it may be too late. However, there are various syndromes that may serve as early indicators of SCD risk. Thus, identifying these predictive markers is crucial to early detection and risk reduction. Recognizing the ECG footprints of these disorders is paramount to making a diagnosis in the asymptomatic individual. Genetic testing may also provide valuable prognostic and diagnostic data, as genetic mutations play a large role in SCD in the young.

CLARIFY and Its Relevance to the Caribbean Population

The Prospective Observational Longitudinal Registry of Patients with Stable Coronary Artery Disease (CLARIFY) study is a large (n=31,040) international trial that is aimed at evaluating outcomes and long-term prognostic determinants in outpatients with stable coronary artery disease (CAD), including heart rate (HR), that adequately represent this contemporary population worldwide. Ronald Henry, MD, Regional Coordinator of the CLARIFY study, Trinidad and Tobago, discussed the significance of the CLARIFY trial as it applies to the Caribbean.

While there is evidence that HR may be a prognostic indicator for cardiovascular (CV) outcomes, it has not yet been incorporated into CV risk assessment or therapeutic guidelines [Fox K et al. *Lancet* 2008]. Therefore, the CLARIFY registry offers a unique perspective regarding the role of HR in the setting of stable CAD, and this study may provide robust data about the disease presentation, risk management, and therapeutic strategies that are associated with HR. The prevalence of CAD has declined in developed countries, but this decline is offset by its increase in developing countries [Allender S et al. *British Heart Foundation* 2007]. Thus, CLARIFY may offer solutions to help alleviate the disease burden in regions where CAD is increasing despite contemporary drug therapies, such as the Caribbean. Additionally, Caribbean involvement in this study allows for soft entry of the region into international CV studies while adding to the learning curve for regional networking. This involvement also fosters the alignment of clinical practices with regional or international guidelines.

Data will be collected at baseline and every 6 months over the course of 5 years. CLARIFY analyses will include demographic data; risk factors and lifestyle; medical history; physical examination and vital signs, including resting HR, measured by pulse palpation and standard 12-lead ECG; current symptoms; medications; and most recent laboratory (eg, fasting blood glucose, lipid panel) and CAD assessment measurements (eg, stress testing results, echocardiography, myocardial imaging), when available. This comprehensive analysis will provide important information on the demographic and clinical

profiles of "real-world" outpatients with CAD. The HR data will potentially offer dynamic estimations of the changing patterns of HR management, disease presentation, and therapy and may help with the development of a reliable risk prediction tool that is based on HR. CLARIFY will improve knowledge about the contemporary CAD population and may help solidify the role of HR as a prognostic indicator. This study is ongoing, and results are expected in 2015.

The Management of Traumatic Aortic Transection

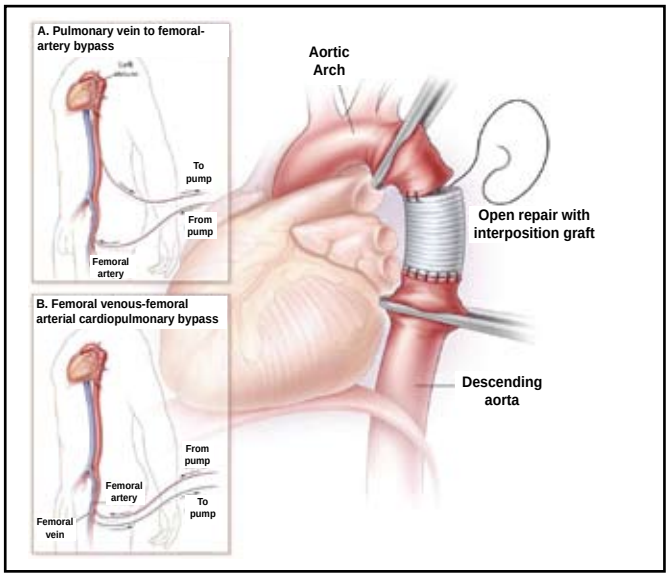
Traumatic aortic transection (TAT) is a rare but often fatal condition, with ~70% of these events caused by blunt motor vehicle trauma (it is the second most common cause of motor vehicle accident mortality [MVA]) [Michetti et al. *J Trauma* 2006]. While the majority of patients dies at the scene of the accident, the prognosis for those who initially survive remains dire, with 90% of deaths occurring within 4 months post-aortic transection. Joseph L. Blidgen, MD, MBBS, University Hospital of the West Indies (UHWI), Kingston, Jamaica, discussed management strategies for TAT, related to 5 cases that were seen at his institution between April 2006 and February 2010.

Diagnostic imaging modalities, such as chest x-ray, angiography, and computed tomography (CT) angiography, which is the current gold standard, are the first step in identifying TAT, particularly among patients who have been involved in a MVA. Further management of TAT may involve adjunctive medical therapy (ie, antihypertensive or β-blocker treatment), open repair (ie, clamp and sew or shunt bypasses), endovascular stenting, or a combination of these methods. Dr. Blidgen cautioned that life-threatening injuries (ie, severe head and abdominal injuries) should be given priority over TAT when present.

Dr. Blidgen and colleagues followed 5 cases of TAT that resulted from MVAs at UWHI. All 5 patients were unrestrained during the MVA, 3 were back seat passengers and 2 were front seat passengers (age range was 15 to 29 years). One patient was diagnosed intraoperatively and referred 4 hours post-MVA once control of intraabdominal bleeding was achieved, 3 were referred 12 hours post-MVA, and 1 was referred 2 weeks post-MVA. Excluding the 15-year-old female who was diagnosed intraoperatively, TAT was diagnosed using chest x-ray and contrast CT scan.

Three patients underwent surgical repair with cardiopulmonary bypass, 1 had left thoracotomy and clamp and sew technique without cardiopulmonary bypass, and 1 patient refused surgical intervention (Figure 1). Of the 4 patients who underwent surgical intervention, 2 had interposition graft placement and 2 had direct repair. One patient had subsequent paraplegia, but there were no reported deaths.

Figure 1. Repair Techniques.



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Endovascular repair is another option for TAT management. It is less invasive than open repair; avoids thoracotomy, single lung ventilation, systemic heparinization, and other potential complications; and can be performed using localized anesthesia. However, it is a relatively new technique; so the long-term prognosis that is associated with this procedure is unclear. Additionally, it may introduce ischemia in other regions, such as the arm or vertebral artery. While this may be a more viable option in the future, these concerns, as well as cost constraints, make endovascular repair less practical at present. Dr. Blidgen concluded that the standard open surgical approach to TAT remains the preferred strategy for the treatment of this complex condition.

Cardiac Surgery and the Appropriate Use of Blood Products in Trinidad and Tobago

The frequency of open heart surgery procedures that are performed in Trinidad and Tobago has increased since procedures began being performed at the Eric Williams Medical Science Complex in 1993, and by 2006, over 200 cases per year were being performed nationally.