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# ABSTRACTS

## SA HEART CONGRESS 2015

### **A cardioprotective and antiarrhythmic role of magnesium pre-treatment against ischaemic injury in isoprenaline-induced hypertrophic rat hearts**

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**Introduction:** The stress of cardiovascular diseases induces a pathological cardiac remodelling process that leads to hypertrophy. Cardiac hypertrophy is associated with myocardial dysfunction and arrhythmogenesis, as well as poor prognosis of ischaemic heart diseases. Magnesium (Mg<sup>2+</sup>) is used to treat several cardiovascular conditions, but its effects in pathological cardiac hypertrophy remain unclear. This study investigated the effects of Mg<sup>2+</sup> on ischaemia/reperfusion (I/R) injury in isoprenaline (ISO) induced hypertrophic hearts.

**Method:** Five groups of adult Wistar rats were treated daily for seven days with either ISO (1.25mg/kg subcutaneously), MgSO<sub>4</sub> (270mg/kg intraperitoneally), saline or combinations thereof. Hearts perfused on a constant-pressure Langendorff system were subjected to regional I/R injury, except the sham group. Haemodynamic and electrogram parameters were recorded on a PowerLab data-acquisition system. Infarct size was determined by triphenyltetrazolium chloride staining and quantified as a percentage of the area-at-risk delineated by Evans blue staining.

**Results and conclusion:** Co-treatment with Mg<sup>2+</sup> decreased I/R-induced infarct size from  $49.0 \pm 8.5\%$  to  $25.1 \pm 5.4\%$  ( $p=0.003$ ,  $n=6/\text{group}$ ) in ISO treated hearts, but not in control hearts. Mg<sup>2+</sup> also reduced episodes of I/R induced ventricular premature beats, ventricular tachycardia, ventricular fibrillation and the overall arrhythmia score in ISO treated hearts from  $13.4 \pm 2.0$  to  $3.2 \pm 1.1$  ( $p<0.05$ ,  $n=5-7/\text{group}$ ). Neither ISO, nor Mg<sup>2+</sup> had detrimental effects on the I/R-induced changes in haemodynamic parameters, including left ventricular developed pressure and time constant of relaxation ( $\tau$ ), or the coronary flow rate. Mg<sup>2+</sup> did not reverse the ISO induced increase in heart weight-to-body weight ratio. These results suggest that Mg<sup>2+</sup> prophylaxis reduces I/R-induced myocardial injury and arrhythmias in hypertrophied hearts, without altering heart weight. Thus there is a potential role for Mg<sup>2+</sup> in the management of pathological cardiac hypertrophy, but its benefits in non-diseased hearts remain unclear.

### **Improvement of diabetes-induced cardiac autonomic dysfunction and left ventricular diastolic pressure-volume response by magnesium pre-treatment in streptozotocin-induced diabetic rats**

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**Introduction:** Diabetes is a growing health problem affecting approximately 9% of the world's population. Morbidity and mortality in diabetes are mainly due to cardiovascular complications, which include autonomic dysfunction and pathological cardiac remodelling. Diabetes is associated with electrolyte abnormalities such as hypomagnesaemia, a known cause of cardiac dysfunction and remodelling. This study investigates the effects of magnesium (Mg<sup>2+</sup>) treatment on diabetes-induced cardiac complications.

**Method:** Four groups of adult Wistar rats were each treated with a single dose of, either streptozotocin (50mg/kg intraperitoneally) or vehicle, and daily for seven days with either MgSO<sub>4</sub> (270mg/kg intraperitoneally) or saline. A blood glucose level above 15mmol/L was taken as the threshold for diabetes. In-vivo tail pulse plethysmography was recorded for heart rate variability (HRV) analysis and ex-vivo Langendorff perfusion-based left ventricular (LV) pressure-volume response parameters were recorded using a PowerLab data-acquisition system.

**Results and conclusion:** Mg<sup>2+</sup> treatment improved diabetes-induced reduction in heart rate, root mean square of successive differences in NN intervals (RMSSD), high frequency (HF) power and low frequency/high frequency power (LF/HF) ratio ( $p<0.05$ ,  $n=7/\text{group}$ ), suggesting restoration of HRV. Mg<sup>2+</sup> also improved the head-up-tilt orthostatic stress response in heart rate and LF/HF ratio ( $p<0.05$ ,  $n=7/\text{group}$ ). Mg<sup>2+</sup> reversed the diabetes-induced right shift in the end-diastolic equilibrium volume intercept ( $V_0$ ) from  $49 \pm 6\mu\text{l}$  to  $25 \pm 5\mu\text{l}$  ( $p=0.014$ ,  $n=6/\text{group}$ ), without altering the LV developed pressure. Mg<sup>2+</sup> treatment did not reverse diabetes-induced hyperglycaemia. These results suggest that Mg<sup>2+</sup> pre-treatment may protect against diabetes-induced reduction in HRV and shift of LV diastolic pressure-volume response, without altering the degree of hyperglycaemia. These findings point toward a potential therapeutic role for Mg<sup>2+</sup> in diabetes-induced cardiac disease.

## Predictive value of a 12-lead ECG in an African population with pulmonary hypertension: Data from the Pan African Pulmonary hypertension Cohort (PAPUCO) study

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**Introduction:** Pulmonary Hypertension (PH) is a major global health issue. Despite the improvements in understanding PH and the development of novel therapies, the condition is still diagnosed at the advanced stage, reducing quality of life and survival. We assessed the diagnostic utility of a standard 12-lead ECG for PH in resource-limited settings.

**Methods:** Sixty-five patients diagnosed with PH from the multi-center Pan African Pulmonary hypertension Cohort (PAPUCO) study were compared to 285 heart disease-free subjects from the Heart of Soweto study. Prevalence, sensitivity, specificity, positive and negative predictive values of ECG parameters, indicative of PH and right heart strain, were calculated.

**Results:** The most common aetiology was PH due to left heart disease (46%). Compared to the control group, all abnormalities were more frequent in the PH cohort. The most prevalent (cases vs. control) ECG abnormalities were: pathologic Q-wave (47.7% vs. 6.7%,  $p < 0.05$ ), left ventricular hypertrophy (38.5% vs. 9.8%,  $p < 0.05$ ) and p-pulmonale (36.9% vs. 20.7%,  $p < 0.05$ ). No patient had a completely normal ECG, as opposed to 15% in the control group. The sensitivity of ECG criteria for right heart strain ranges between 1.5 and 47.7% while specificity ranges between 79.3 and 100%. The negative predictive value ranges between 81.5 and 88.9%. Positive predictive value was lowest at 25% for RBBB and QRS right axis deviation ( $\geq 100^\circ$ ) and highest at 100% for QRS axis  $\geq +100^\circ$  combined with R/S ratio  $\geq 1$  or R in VI  $> 7\text{mm}$ .

**Conclusion:** ECG abnormalities are common, but non-specific, among patients with PH. Furthermore, those relating to right heart strain are less frequent. ECG patterns focusing on the R and S amplitude in VI and right axis deviation had good negative predictive values for PH, but due to high prevalence of ECG abnormalities, this should be interpreted with caution. Pending further studies, other tools are needed for screening PH.

## An in vivo study of the extracellular matrix after prolonged cold ischaemic harvesting time comparing cryopreservation to decellularisation

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**Objectives:** Cryopreserved pulmonary allografts are still the golden standard for right ventricular outflow tract reconstruction, however availability is restricted and durability is limited by immunogenic response. This study was performed to investigate the early impact of prolonged cold ischaemic harvesting time and decellularisation on pulmonary allografts to improve durability.

**Methods:** Two groups of pulmonary allografts were created, both with a post-mortem cold ischaemic harvesting time of 48 hours, including cryopreserved pulmonary allografts (CPAs) (group 1; n=5) and decellularised pulmonary allografts (DPAs) (group 2; n=5). Valves of both group were explanted after 14 days of implantation in the juvenile sheep model. Integrity of the extracellular matrices of CPAs and DPAs were examined by tensile strength (TS) and Young's modulus (YM) test. Histological evaluation of the extracellular matrix included H&E, Picoserius red and transmission electron microscopy.

**Results:** Tensile strength and YM of the wall were, in group 1 ( $0.48 \pm 0.12$  MPa, and  $0.99 \pm 0.16$  MPa) and in group 2 ( $0.96 \pm 0.56$  MPa, and  $0.78 \pm 0.12$  MPa) respectively. The leaflet TS and YM were, in group 1 ( $2.79 \pm 1.00$  MPa, and  $11.47 \pm 5.05$  MPa) and group 2 ( $5.64 \pm 1.67$  MPa, and  $36.20 \pm 19.47$  MPa) respectively. Histologic examination (H&E and Picoserius Red staining) demonstrated a well-preserved extracellular matrix in both groups. Transmission electron microscopy showed partial destruction of collagen fibres in the CPAs. The DPAs, however, showed a complete intact extracellular matrix with the absence of micro-structural lesions.

**Conclusion:** This experimental study supports the potential to prolong post-mortem cold ischaemic harvesting time of pulmonary allografts. However, if cryopreservation is avoided in favour of decellularisation of pulmonary allografts, micro-lesions of the extracellular matrices can be avoided.

### **“Blackouts” and sudden death in the apparently well and young - the case of Long QT syndrome: Missed opportunities for a diagnosis and treatment**

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The long QT syndrome (LQTS) is pro-arrhythmic cardiac disorder associated with blackouts (Transient Loss of Consciousness; TLOC). It is necessary to recognise that these events are syncope and not epilepsy or another type of attack. Sometimes the syncope-causing ventricular tachycardia may degenerate into ventricular fibrillation and death. LQTS is autosomal dominantly inherited and causal-mutations (1 000+) identified in 13 genes. Through cascade screening of relatives of 26 LQTS index cases, we identified 203 living persons with the same potassium channel mutation, namely KCNQ1 A341V. Histories have been collected on all of them. We also collected information on deaths of close relatives. Many, symptomatic with a history of blackouts, were not diagnosed with LQTS. We set out to quantitate these missed diagnoses.

Of the mutation carriers 79% experienced blackouts. Only 26% was diagnosed as LQTS and appropriately treated. Epilepsy was the diagnosis in 40%. Another 34% received either laymen's explanations or medical, "vasovagal", "sick sinus syndrome", etc. A number of "near drowning" events were documented. Historic deaths before age 20, were 23. Half of these were drownings, all in able swimmers. Other examples are of a girl, aged 13, dying on a skating rink while under treatment for epilepsy and a boy, aged 5, who "choked on water".

Our experience shows gross disparities in diagnosis and consequent management in a treatable risk of sudden death. Missed opportunities for diagnoses ranged from prior to medical encounters to at, and after, the first presentation. The most common misdiagnosis was epilepsy, in the living, and drowning, in the dead. Missed and misdiagnosis may be prevented in how both lay persons and medical professionals perceive TLOC and, also, near drowning and drowning events. In the words of one of my teachers: "You only recognise what you know".

### **Infective endocarditis in central South Africa**

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**Introduction:** Infective endocarditis (IE) is an uncommon disease that carries significant morbidity and mortality despite advances in antimicrobial therapy and anaesthetic, surgical and post-operative care in the last three decades. Research regarding surgery for native infective endocarditis (IE) in South Africa is limited. The present study is a retrospective analysis of patients with IE requiring surgery in central South Africa between 2006 and 2014.

**Method:** This study is a retrospective analysis of the demographics, diagnoses, pre-disposing factors, HIV status, pathogens, intra-operative and post-operative records and clinical outcomes of patients presenting with IE in central South Africa.

**Results:** Hundred adult patients were operated for IE during this period. The average age was 43 years (14 - 78 years) with a male predominance (60%). In total, 127 valve procedures were performed of which 98 were valve replacements and 29 valve repairs. The procedures included repair or replacement of 47 aortic, 60 mitral and 17 tricuspid valves. Organisms were similar to previous studies, with 80% of cases due to Staphylococcus and Streptococcus infections. The predominant underlying cause was found to be rheumatic heart disease, with congenital causes being the second most common cause. Thirty-three percent of patients (17/52 patients) who consented to be tested were HIV-positive. The 30-day mortality rate was 8% of which 1 patient was HIV+.

**Conclusion:** IE remains an important surgical disease in the South African population. Rheumatic heart disease predominates as an important cause with the second most common cause being congenital heart disease. This prospective study also suggests that HIV does not contribute significantly to IE peri-operative mortality rates, but the prevalence of at least 17% in this cohort is higher than previously published data.

## Acute coronary syndrome in South Africans – with a specific focus on statin treatment – SA Heart SHARE registry results (2004 - 2012)

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**Introduction:** This study investigates acute coronary syndrome (ACS) in South Africans in association with risk factors and other variables, with the aim of improving the management of STEMI. Prior admission to Cath lab treatment with statins is scrutinised in relation to ACS sub-type, indication for treatment and other relevant factors.

**Method:** ACS phenotype (ASC-p) was defined by clinical subtype and prior intervention and 10 498 of 15 243 admissions to Cath labs were defined as such. Data on ethnicity were only available for 5 747 ACS-p patients and data on location of centre for 9 985 patients. Prior medication prescribed was reported upon for 6 198 patients.

**Result:** STEMI was most prevalent in subjects from African ascent (AA) (18%), Stable Angina Pectoris (SAP) in Caucasians (59%) and Non-STEMI (NS) in patients from mixed ancestry (MA) (42%). Family history of heart disease and diabetes was highest in Indian males (59% and 39%), and hypertension in AA males (72%). Statin treatment was reported in 54%, 66%, 73% and 77% in Caucasian, AA, MA and Indian patients respectively, and in 41%, 35% and 44% of STEMI, SAP and NS patients. Risk for STEMI in ACS-p patients increased by 20% per three-year period (RR 1.2; 95% CI 1.1-1.3) and was not ameliorated by statin treatment, which increased from 37% to 57% during the first and last tertile of the study. More patients were diagnosed with STEMI in public vs. private centres (15% vs. 7%) during 2010 - 2012, while more patients were treated with statins in public centres (68% vs. 49%).

**Conclusion:** STEMI cases appear to be on the increase in public centres, despite a higher percentage of patients being treated with statins. This relationship needs to be explored in greater detail, taking the duration of treatment and ancestry into consideration.

## Are sutureless aortic valves suitable for very high-risk patients suffering from active infective aortic prosthetic valve endocarditis?

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**Introduction:** Sutureless aortic valves are easy to handle and therefore we evaluated the patient benefits of these bio-prostheses in very high-risk patients with active infective aortic prosthetic valve endocarditis.

**Materials and methods:** Between April 2014 and July 2015, a total of 42 patients received a sutureless Perceval aortic valve (Sorin Biomedica Cardio Srl, Saluggia, Italy) for different indications. Four of these patients, with a median age of 63 years (range 47 - 74 years) suffered from active infective aortic prosthetic valve endocarditis, including TAVI. The median euroSCORE II of this highly selected patient cohort was 75.9 % (range 73.7 - 87.7 %). Patients were followed and measured, including operative data and early in-hospital morbidity and mortality.

**Results:** All patients needed abscess closure with pericardium. Two patients underwent left atrial appendix closure, one left ventricular thrombectomy, one coronary bypass grafting and one partial aortic arch redo was performed. Median aortic cross-clamp and cardiopulmonary bypass time was, respectively 54 minutes (range 30 - 88 min) and 85 minutes (range 52 - 133 min). The median intubation time was 13 hours (range 7 - 19 hours). Post-operative morbidity in these very high-risk patients were, reintubation due to extensive delirium in one patient, re-exploration in one patient, temporary dialysis in one patient and one patient with sternal refixation. No pacemaker implantation was needed. There was one in-hospital death (25%).

**Discussion:** Sutureless aortic valve replacement, in very high-risk patients suffering from active infection aortic prosthetic valve endocarditis, seems to be an option with limited morbidity and acceptable mortality.

## Beta-blockers in pregnancy: University of Cape Town cardiac diseases maternity cohort study

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**Background:** Data regarding the use of beta-blockers in pregnant women with structural heart disease, pertaining to fetal growth restriction and fetal outcomes, are limited. We investigated the impact of beta-blockers use in pregnancy on offspring outcome.

**Methods:** A prospective cohort study was conducted in a tertiary multidisciplinary maternal care facility in South Africa from 2010 to 2012. From 124 consecutive pregnant women with structural heart disease, 19 received beta-blockers (n= 9) in pregnancy. Poor fetal outcome was defined as: perinatal death, low birth weight (LBW), Apgar scores <7 and premature birth (<38 weeks). In addition, fetal outcomes of patients with cardiomyopathies were compared to other cardiac diseases in pregnancy.

**Results:** Fifteen percent (19/124) of patients were on beta-blockers throughout pregnancy. There was no statistical difference in the offspring birthweight of women receiving beta-blockers (BB) vs. non beta-blockers (NBB) (2 842.8kg vs. 2 853.2kg,  $p=0.92$ ). There was an increased risk of LBW babies in mothers on BB of 18.6% (RR:1.186, CI:0.38 to 2.58). No association was found between length/dose of BB used in pregnancy and LBW or poor fetal outcomes. Forty percent (8/19) of patients with cardiomyopathies were on BB during pregnancy, compared to 10% (11/104) of patients with other cardiac diseases ( $p=0.025$ ). Cardiomyopathy patients' mean birthweight on BB was less than that of patients with other cardiac diseases on BB (2 810.7kg vs. 3 024.5kg,  $p=0.55$ ). No adverse fetal events were found in the cardiomyopathy group and Apgar scores were 8 and 9.2 respectively with good outcomes.

**Conclusion:** Beta-blockers in pregnancy were found to have a minor impact on fetal birthweight, but had no impact on other fetal outcomes. This suggests that we should balance fetal risk with maternal need when considering beta-blocker use in pregnant women with structural heart disease.

## Ethnic differences in heart failure: The New Zealand Maori and Pacific Islanders

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**Introduction:** Differences in heart failure (HF) prevalence, outcome and management exist across the world. Some of these relate to economic factors and health care opportunities. Within one country however, significant differences seem to primarily relate to ethnicity. Such disparities exist in HF patients within New Zealand (NZ), a country with a predominantly European population. Between 10 - 25% of the population, depending on region, are of Maori or Pacific Island (M/PI) ethnicity.

**Method:** For approximately 10 years, data have been collected on HF admissions across NZ (NZ HF Registry). An increasing number of consecutive admissions in more recent years provide confidence that the data collected is representative of national demographics and outcomes.

**Results:** Compared to NZ European HF admissions, M/PI patients are more likely to be male and on average 17 years younger (62 vs. 79 years). They are more likely to have reduced ejection fraction and diabetes, are less likely to have coronary artery disease. These demographics are more similar to African, rather than European and American HF registries.

Due to the incidence of rheumatic heart disease amongst this ethnic population, a greater number of HF presentations are associated with valve disease. Differences are also noted in medication adherence (poorer in M/PI) but no difference in prognostic medication prescription, and re-admission at 90 days.

**Conclusions:** Registries suggest NZ Maori and Pacific Islander HF patients are more similar to African than European and American patients. NZ ethnic differences in HF aetiology and demographics at admission are seen, especially age. Once admitted, management and outcomes are similar. Focus is needed on prevention, identification and management of risks and early disease.

## Peripartum cardiomyopathy: Experiences in a low resource setting

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**Introduction:** Peripartum cardiomyopathy (PPCM) is a rare, but devastating, complication of pregnancy. Although relatively common among African women, there is limited outcome data from Africa. We sought to evaluate clinical outcome and risk factors for adverse outcomes among patients with PPCM in Zimbabwe.

**Methods:** A prospective cohort study of patients with PPCM was carried out at a tertiary hospital in Zimbabwe. Clinical assessment and echocardiograms were performed at enrolment and at 6 months. Baseline characteristics associated with the primary outcome [a composite of death, left ventricular ejection fraction (LVEF) <50%, New York Heart Association (NYHA) functional class III/IV at 6 months] were determined.

**Results:** Fifty-seven patients with a new diagnosis of PPCM were recruited. Twenty-nine patients (51.8%) had an adverse outcome. Seven patients (12.3%) died. Factors associated with adverse outcomes were lower maternal age (OR 0.89, 95% CI 0.80-0.99,  $p=0.025$ ), NYHA III (OR 10.9, 95% CI 2.69-43.9,  $p=0.001$ ), onset of symptoms after the postpartum period (OR 3.57 95% CI 1.06-12.1,  $p=0.040$ ), increased left ventricular end systolic dimension (LVDs) (OR 1.15, 95% CI 1.03-1.29,  $p=0.014$ ) and lower LVEF (OR 0.92, 95% CI 0.86-0.98,  $p=0.015$ ). Seven of the 8 patients with thrombotic complications had the primary outcome ( $p=0.052$ ). Forty-three patients had echocardiography at 6 months; 21 (48.8%) experienced complete recovery of left ventricular function defined as LVEF >50%. Overall, LVEF improved significantly (baseline  $29.7 \pm 9.9\%$  to  $44.5 \pm 14.2\%$  after 6 months,  $p<0.001$ ).

**Conclusion:** A substantial proportion of patients fully recovered left ventricular function, but adverse outcomes, including mortality were frequent. Lower maternal age, worse NYHA functional class, onset of symptoms after the postpartum period, increased LVDs and reduced LVEF at baseline were associated with adverse outcomes.

## Radiofrequency catheter ablation in infants and children: A single centre experience

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**Background:** Diagnosis and treatment of paediatric cardiac arrhythmias remain challenging. Antiarrhythmic drugs, beta-blockers, class Ic and class III drugs were the only treatment available for many years. Radiofrequency (RF) catheter ablation was introduced for adult patients in 1981 and has been applied in children since 1989. Many questions still remain regarding the efficacy and safety in paediatric patients.

**Method:** A retrospective analysis of patient records of patients, 12 years of age and younger who underwent RF catheter ablation was performed.

**Results:** From January 2011 - July 2015, a total of 51 ablations were performed in 44 patients younger than 12 years of age; mean age was 7 years (range 3 weeks - 12 years) with 11 (21.5%) aged 2 years or younger; mean weight of 16.5kg (range 3.4 - 56kg); average screening time 33.5min (SD  $\pm$  22.7) and average radiation dose of 37mGy (SD  $\pm$  436.52). Echocardiography was normal in 32 (72.7%) of patients. All cases were done under general anaesthesia. Diagnosis on invasive electrophysiological study: atrioventricular re-entry tachycardia in 26 (50.9%), atrial ectopic tachycardia in 7 (13.7%), intra-atrial re-entry tachycardia in 5 (9.8%), atrioventricular nodal re-entry tachycardia in 11 (21.6%) and ventricular tachycardia in 2 (3.9%) of cases. RF ablation was successful in 44 (86.3%) of procedures with recurrence rate of 13.7%. No significant complications were noted.

**Conclusion:** RF catheter ablation can be performed safely and effectively in even the very young infant. The recurrence and complication rates are similar to those reported in adults. Therefore, RF ablation should be the treatment of choice in paediatric patients with serious arrhythmias.

## Supracardiac total anomalous pulmonary venous drainage in an asymptomatic adult: The importance of pulmonary vein localisation in atrial septal defect assessment

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**Introduction:** Total anomalous pulmonary venous drainage (TAPVD) is a cyanotic congenital abnormality in which all 4 pulmonary veins fail to connect to the left atrium (LA), resulting in drainage of the entire pulmonary venous return to the systemic venous circulation. It is the fifth most common cyanotic congenital heart condition (incidence: 0.6 - 1.2 per 10 000 live births). (1) There are 4 anatomic variants (supracardiac, cardiac, infracardiac and mixed) that determine where oxygenated pulmonary venous blood mixes with deoxygenated blood from the systemic system. Right-to-left shunting usually occurs through an atrial septal defect (ASD). TAPVD presents almost exclusively in infancy and, if untreated, carries a 80% mortality at one year. (2) We report an asymptomatic case presenting at 18 years.

**Case:** Our patient was referred to adult cardiology at Tygerberg Hospital for evaluation of presumed rheumatic heart disease. He reported always being physically well and even playing wing for his local rugby team. He was acyanotic and cardiac examination suggested an ASD. Transthoracic echocardiography confirmed a secundum ASD with marked right heart enlargement. A large venous structure was visible anterior to the descending thoracic aorta. On transoesophageal echocardiography no pulmonary veins were visible entering the LA. Cardiac catheterisation demonstrated supracardiac TAPVD with all 4 pulmonary veins draining into a confluent chamber, posterior to the LA, which connected via a vertical vein to the innominate vein. The Qp:Qs was 4 and the pulmonary vascular resistance was 1 wood unit. Computed tomography scan confirmed the anatomy. Our patient is acyanotic due to preferential streaming of oxygenated blood across the ASD into the LA and is awaiting corrective surgery.

**Discussion:** It is rare for TAPVD to present undiagnosed in adulthood. Secundum ASD is a common echocardiographic diagnosis that should always prompt a search for associated congenital abnormalities, such as partial or total anomalous pulmonary venous drainage.

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## An unusual cause of a reversible dilated cardiomyopathy

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Hypocalcaemia is a well-known, although rare, cause of a dilated cardiomyopathy that may occur in infants and children. Causes of the hypocalcaemia may be varied, but may include rickets and prematurity.

Patient TM, who is now a 13-year-old male, presented for the first time to our hospital in biventricular failure at the age of 10 years. He had had symptoms since the age of 5 years and was examined in private practice, but no cause was found. On echocardiography, a dilated cardiomyopathy with an ejection fraction of 32% was diagnosed but with no obvious cause found. He was started on anti-failure therapy and discharged well. He then presented for the second time with a seizure when hypocalcaemia was documented. The low calcium was treated, and a full etiologic workup was done.

Normocalcaemia was established, on Vitamin D and calcium supplements, with demonstration of the LV function returning to normal over subsequent visits. However, he presented again years later to a private hospital with neurologic symptoms and a MRI Brain showed bilateral basal ganglia calcification. The significant endocrine abnormality that has been documented in this child, is that of a markedly raised parathyroid hormone (PTH), consistent with a diagnosis of pseudohypoparathyroidism.

## Congenital Rubella syndrome – not gone, not to be forgotten

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Rubella is endemic to South Africa, with clusters often occurring in autumn and spring. Although it is a preventable condition, rubella immunisation is not part of the EPI programme. It is not a notifiable condition, although congenital rubella syndrome (CRS) is.

WHO Global estimates in 2008 of the burden of rubella suggest that the number of infants born with CRS exceeded 1 100 001 which makes rubella a leading cause of preventable congenital defects, the bulk of cases occurring in the South East Asian and African regions. In the WHO African region, in 1996, it was estimated that >22 500 infants are born with CRS annually.

The exact incidence of congenital rubella syndrome in South Africa is unknown, but it is estimated that 600 - 700 cases occur annually.

Congenital rubella syndrome is a devastating condition, presenting with the classical triad of cataracts, deafness and cardiac defects.

A series of 3 patients are presented, highlighting the clinical aspects of this condition which often exacts a comparatively heavy cost to families and to strained healthcare systems, when a cheap vaccine is available.

## Renal denervation reduces office blood pressure in a sham-controlled trial after one year follow up

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**Introduction:** One large randomised sham-controlled renal denervation (RD) trial, SYMPPLICITY-HTN-3, could not demonstrate a significant reduction in office blood pressure at one year follow up. Incomplete renal denervation has been suggested as one of the reasons for its failure. We tested the hypothesis that RD would significantly reduce office blood pressure when compared to sham-controls, if performed correctly.

**Methods:** We performed bilateral four quadrant RD in 10 patients who had an office or 24 hour ambulatory blood pressure of  $\geq 160/90$  mmHg in non-diabetics or  $\geq 150/90$  mmHg in diabetics. All patients  $\geq 55$  years were on  $\geq 3$  antihypertensive agents, including a diuretic. They had to be in sinus rhythm and have echocardiographic evidence of hypertensive heart disease (enlarged left atrium with left ventricular hypertrophy). Patients with untreated thyroid disease, significant valvular heart disease and renal impairment ( $eGFR < 45$  ml/min/1.73m<sup>2</sup>) were excluded. Office blood pressure was measured electronically at least 5 minutes prior to an exercise stress test at baseline and one year after the intervention. Responders were defined as those with an SBP reduction of  $\geq 5$  mmHg at follow up. A paired T-test, with equal variance, was used to assess statistical significance and a p-value of  $< 0.05$  was considered significant.

**Results:** Age, sex, blood pressure medication and ambulatory blood pressure measurements did not differ significantly. Eleven subjects underwent sham RDN. Office systolic blood pressure was significantly reduced in the RD treated group only after one year follow up (SBP\_RD1 =  $143.2 \pm 16.39$  mmHg vs. SBP\_RD2 =  $130.7 \pm 18.2$  mmHg at one year follow up; with absolute reduction of 12.5 mmHg vs. SBP\_sham-RD1 =  $132.09 \pm 14.43$  mmHg vs. SBP\_sham-RD2 =  $140.18 \pm 16.63$  mmHg at one year follow up; with absolute increase of 8.1 mmHg;  $p=0.0361$ ). Office diastolic BP was only reduced in the intervention group: DBP\_RD1 =  $84.1 \pm 10.71$  mmHg vs. DBP\_RD2 =  $77.9 \pm 8.56$  mmHg at one year follow up; with absolute reduction of 6.2 mmHg vs. DBP\_sham-RD1 =  $76.55 \pm 6.47$  mmHg vs. DBP\_sham-RD2 =  $83 \pm 8.59$  mmHg at one year follow up.

**Conclusion:** Office blood pressure was significantly reduced by RD after one year of follow up. Larger trials are awaited to confirm these results.



## Registries and research provide a remedy for public policy; rheumatic heart disease in Namibia

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**Background:** Although prevalence is high (30/1 000 in children 7 - 17 years), Rheumatic Heart Disease (RHD) is much neglected in Africa. Before the registry, the burden of RHD in Namibia was unknown with no data to inform public policy. We report data which informed the development of a National Programme (ASAP) for the prevention and control of RHD.

**Materials and methods:** The national registry of RHD was established at Windhoek Central Hospital in January 2010. From January 2010 - December 2014, 463 patients were enrolled in this prospective, national, hospital-based registry. Questionnaires document demographics, presentation, complications, ECG, echocardiogram and management. Baseline data is presented on the first 281 patients entered into Remedy.

**Results:** Distribution of cases reflects population density, 61% are female and 39% male. Ninety-seven (34%) are children and 83% under 40 years. Thirty-two percent were NYHA III - IV and 48% in heart failure. Nineteen percent had atrial fibrillation, 6% stroke and 13% previous surgery. Only 34% of patients were receiving secondary penicillin prophylaxis. Of patients needing anticoagulation, 39% were receiving warfarin, 73% had no INR analysis the preceding 6 months. Only 7 of 47 who had had surgery, had mitral valve repair.

**Conclusions:** The burden of disease is significant. Patients are seen late with advanced disease. Low numbers over 50 years of age (4%) reflect high mortality. Low numbers on secondary penicillin prophylaxis and poor compliance with anticoagulation protocols reflect a lack of awareness about RHD amongst health workers. Gaps identified in the organisation and delivery of care pointed to the need for a national programme for the prevention and control of RHD, established in March 2014. A national Advisory Committee for RHD was established in May 2015. This example of "research to action", emphasises the public policy benefits to be gained from clinical science.

## Why they survive? Native congenital heart disease in a grown-up African population without access to care

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**Objectives:** We describe 104 patients with congenital heart disease (CHD), older than 13 years, from an environment with no cardiac services.

**Methods:** Case notes of patients seen at Windhoek Central Hospital, between January 2009 and December 2013, were reviewed for CHD. Clinical features, ECG, chest X-ray, echocardiogram, catheterisation, operation and follow-up records were analysed. Diagnosis was established by echocardiogram. Surgery was conducted at Windhoek Central or Christiaan Barnard Memorial Hospital.

**Results:** In 104 patients, age 13 - 86 years (mean 25), 68 were female. Pathology included: VSD 30, secundum ASD 32, PDA 12, AVSD 10 (9 primum ASD), AS 4, coarctation of aorta 5. Cyanotic disease included Tetralogy of Fallot 7 (TOF), DORV with TOF 4, PS 7, Ebstein's anomaly 2 and TGA/VSD/PS 2.

Thirty-one had undergone cardiac catheterisation. Seventy-eight were deemed "operable". There were 7 interventions for: coarctation (2), PDA (2), ASD (2) and valvular PS (1). Surgery was performed on 52: VSD 15, ASD 2 16, PDA 5, primum ASD 5, TOF 6 (3 with DORV). Three with complex disease were repaired. Eleven were inoperable.

Over 5 year follow-up (mean 32 months), three patients died: 2 with inoperable secundum ASD and 1 after shunt surgery for TOF with pulmonary atresia. Fifteen patients were lost.

**Conclusions:** This cohort reflects a time with no diagnostic or curative services for CHD. A high number of patients with large, "simple" lesions (VSD, ASD and PDA), advanced pulmonary hypertension and congestive cardiac failure are still operable. There is a sub-set of patients with Tetralogy of Fallot who survive to adulthood. Their anatomy and the surgical challenge is different from that in the infant. The high number "lost" reflects system challenges in a vast, low income country.

## The use of mechanical heart valves without anticoagulation. A bridge too far?

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**Objective:** The thrombogenicity of mechanical prosthetic heart valves limit their use. Patients are subjected to lifelong warfarin use, balancing bleeding risk with thrombosis. Designing a valve that will negate the use of warfarin remains the Holy Grail in mechanical valve design research. The thrombotic properties of a re-engineered valve design (the UCT/Frater valve) were tested in the ovine model to assess its thrombogenic potential.

**Methods:** The UCT valve was modified along modern flow dynamic principles and designs. These valves were implanted in the pulmonary position during 2013 - 2014 in the juvenile ovine model (males, mean age 6 months). Sorin Carbomedics mechanical prostheses were used as control valves. The study population was divided into two groups, based on post-operative follow up (Group 1 = 6 month and Group 2 = 12 month follow-up). A total of 12 pulmonary valve replacements were performed. Each group consisted of 1 control (Carbomedics) and 5 UCT/Frater valves. No coumadin or anti-platelet therapy was used throughout the study interval.

Full blood counts, liver functions, haptoglobin, platelet activation and fragmentation were performed during follow-up and macro- and microscopic inspection upon explantation.

**Results:** Two UCT/Frater valves thrombosed secondary to infective endocarditis (IE). None of the other valves, control valves included, showed any macroscopic or microscopic thrombi. Biochemistry and hematology did not demonstrate hemolysis, activation of coagulation or platelet activity. Histology showed no thrombi on the sewing cuff, housing, poppet or struts. None of the subjects had embolic events and their lungs had no pulmonary emboli. Cardiac echocardiography confirmed normal prosthetic function in all valves, except those with IE.

**Conclusion:** The re-engineered UCT/Frater valve design, without anticoagulation and anti-platelet treatment, performs well in the ovine model with acceptable haemodynamics. This prosthesis should be tested in a more aggressive coagulation model.

## Scimitar Syndrome case series

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**Introduction:** Scimitar Syndrome is a rare congenital anomaly, a form of Partial Anomalous Pulmonary Venous Drainage. The typical anatomy comprises of the right pulmonary venous return draining into the inferior vena cava, amongst other associated abnormalities. The name is derived from the similarity in shape of the vein to a scimitar blade. There is a bi-modal presentation, with infantile types presenting early with cardiac failure, pulmonary hypertension and significant mortality.

**Cases:** We present a series of 3 cases seen at our hospital from 2004 - 2015.

All 3 cases were female, had common findings of a chest X-ray showing dextroposition, a hypoplastic right lung and, on angiography, the classical Scimitar vein draining into the inferior vena cava.

The younger patient, aged 5 months, presented with heart failure and significant pulmonary hypertension, whereas the 6-year-old and the 9-year-old were far less symptomatic. The six-year-old was the only patient who had an associated intra-cardiac defect, namely an Atrial Septal Defect. The 5-month-old and the 9-year-old had collateral arterial supply to the right lung and both were subsequently occluded during catheterisation.

The 5-month-old was lost to follow up after coiling of the collateral arterial supply. Surgical correction was successfully done in the 6-year-old and the 9-year-old is awaiting surgery.

**Discussion:** The aim of this case series is to review this unusual congenital anomaly and highlight the clinical, radiologic and management aspects.

## Cardiac transplantation vs. mechanical cardiac support: Current status in South Africa

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**Introduction:** Although donor heart transplantation is still considered to be the gold standard in the treatment of end-stage cardiac failure, technological advances are progressively becoming a more attractive option. This abstract reports on the activities and current status in long-term mechanical cardiac support in South Africa.

**Method:** Over the past 15 years, 46 patients were supported with mechanical assist devices. Nine patients were successfully treated using the HeartWare ventricular assist pump during the past 3 years as a bridge-to-transplantation. Twenty-eight patients were supported using the Jostra

Centrifugal pump. Twenty-four of these patients had post cardiectomy shock following coronary artery surgery. Four patients had donor heart failure at the time of transplantation. Nine patients received Berlin Heart BiVAD implantation. Ages ranged from 11 - 61 years. The respective indications and techniques of implantation and post-operative management are discussed.

**Results:** Seven of the nine patients (78%) receiving the HeartWare device were discharged and all 7 are back at work. Of the 28 patients who received the Jostra centrifugal pump, 24 (84%) were weaned off and 22 (79%) were discharged from hospital. Of the 9 patients on the BiVAD Berlin Heart, 7 (78%) patients were transplanted and 6 (67%) discharged from hospital. All 3 patients of the 4 requiring donor heart support were successfully weaned and discharged home.

**Conclusion:** These results compare favourably with the outcomes reported upon in international literature. Due to organ shortage in South Africa, an increase in demand for mechanical assistance can be expected in the future.

## The integrity of the coronary artery after cardiac transplantation: A review of the challenges to the transplant team in 150 cardiac recipients

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**Introduction:** Coronary artery disease is an indication for cardiac transplants in 45 - 50% of reported cases. After the transplant, transplant related coronary disease is the leading cause for late mortality. Several factors contribute to the development of transplant vasculopathy, of which tissue mismatch, CMV exposure, poor rejection control, diabetes, serum lipid profile and atherosclerosis are leading causes.

Interventions, such as percutaneous intervention (PCI), coronary surgery and transmyocardial laser revascularisation can be used to improve myocardial perfusion. Intravascular ultrasound (IVUS) can be useful in the diagnosis, quantification and management of transplant coronary artery disease.

**Method:** Hundred-and-fifty patients which receive cardiac transplants, over a period of 15 years, were reviewed. All 150 patients had annual routine coronary angiograms. Seventy-five patients had angiographical evidence of coronary pathology, post transplantation. One patient presented with angina and 23 patients with deteriorating ventricular function. Twenty-four required percutaneous coronary intervention. One patient had a transmyocardial revascularisation procedure. One patient had coronary artery bypass surgery.

**Results:** Of the 75 patients, 2 (3%) patients showed evidence of regression on medical therapy. Of the 24 patients requiring PCI 4 (32%) patients needed repeat PCI and 2 patients required repeat transplantation (3%). The 2 (3%) patients that underwent CABG and TMLR died.

**Conclusion:** As transplant vasculopathy is a leading cause of mortality in the transplant recipient, the integrity of the coronary artery forms a cornerstone of long-term management of the cardiac recipient. A team approach, as well as a sound understanding of drug interaction, is required to minimise development of transplant related vasculopathy.

## Albuminuria is related to troponin in hypertensive black women

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**Objective:** In hypertensives, troponin T, as a marker of cardiomyocyte damage, predicts the development of renal disease as reflected by micro- and macro-albuminuria. However, urinary albumin excretion also reflects general endothelial damage and relates to arterial stiffness, which in turn increases cardiac afterload and damage. We investigated the link between troponin T and albuminuria in black women, without renal dysfunction, but in which hypertensive heart disease is common.

**Design and method:** This sub-study included a cohort of 122 black women (mean age: 60.6 years) from the larger Prospective Urban and Rural Epidemiology study. ECGs, along with conventional cardiovascular assessments, were performed. Biochemical analyses included urinary albumin and creatinine, and serum troponin T. We assessed renal function with estimated creatinine clearance and glomerular filtration rate (eGFR).

**Results:** In bivariate analysis, ACR correlated positively with the Cornell-product, central systolic blood pressure, central pulse pressure, aortic pulse wave velocity and troponin T (all  $p < 0.05$ ). After adjusting for age, body surface area, hypertension status and eGFR, the results remained unchanged for troponin T ( $r = 0.30$ ;  $p = 0.010$ ) and the Cornell-product ( $r = 0.27$ ;  $p = 0.028$ ). After performing a multiple regression analysis, we observed the same trends, however with a significant interaction with hypertension status ( $F = 14.3$ ;  $p = 0.031$ ). When repeating the same model in hypertensives only, troponin T associated with ACR (Adj.  $R^2 = 0.48$ ;  $\beta = 0.55$ ;  $p = 0.006$ ). This was not the case for normotensives.

**Conclusion:** Troponin T was positively associated with albuminuria in hypertensive black women. Our result indicates that albuminuria, even in normal range, is linked to adverse cardiomyocyte damage in non-diabetic hypertensive black women, possibly via the increased arterial stiffness – cardiac overload mechanism.

## Comparison of the clinical phenotype of the common heterozygous familial hypercholesterolaemia mutations at the LDL-receptor in the Afrikaner population

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**Introduction:** It has been previously reported that South African whites have one of the highest death rates from coronary artery disease in the Western world. The reasons for this are likely multifactorial but one of the proposed reasons is the high prevalence of familial hypercholesterolaemia (FH) in this patient population. The 3 most common mutations occurring at the LDL receptor are designated Afrikaner 1 (FH1), Afrikaner 2 (FH2) and Afrikaner 3 (FH3). Little is known regarding the clinical phenotype of these patients with only one study to date with very small numbers being published.

**Methods:** A retrospective analysis from 1981 - 2015 was undertaken at the Lipid Clinic of CMJAH. All patients heterozygous for FH1, FH2 and FH3 mutations were analysed by file review.

**Results:** Four-hundred-and-ninety-two patients were analysed, 200 males and 292 females. Two-hundred-and-eighty-six patients were FH1, 183 were FH2 and 23 were FH3. The average age at diagnosis was 36 years with FH2 patients tending to present younger and FH3 patients older. The average total cholesterol at diagnosis was 8.9mmol/L for FH1, 9.5mmol/L for FH2 and 8.1mmol/L for FH3. This difference was significant ( $p=0.0038$ ) amongst the 3 groups and between FH2 and FH3 ( $p<0.01$ ). The prevalence of ischaemic heart disease was 24% in FH1, 22% in FH2 and 13% in FH3 which was not significant for difference amongst the 3 groups.

**Conclusion:** This review is the first of its kind to compare the phenotypic severity of the 3 most common Afrikaner mutations. It showed that FH3 is the mildest mutation in terms of phenotypic characteristics with a significantly lower untreated total cholesterol and prevalence of ischaemic heart disease. FH2 appears to be the worst mutation in terms of clinical severity with a higher total cholesterol and prevalence of ischaemic heart disease, although this did not achieve statistical significance.

## Comparison of the response to statin therapy in heterozygous familial hypercholesterolaemia within the Afrikaner population

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**Introduction:** The 3 most common mutations occurring at the LDL receptor are designated Afrikaner 1 (FH1), Afrikaner 2 (FH2) and Afrikaner 3 (FH3). These are responsible for 90% of the cases of familial hypercholesterolaemia (FH) within the Afrikaner population in South Africa. Little is known regarding the response to lipid-lowering therapy in these patients with only one study to date with very small numbers being published.

**Methods:** A retrospective analysis, from 1981 - 2015, was undertaken at the Lipid Clinic of CMJAH. All patients heterozygous for FH1, FH2 and FH3 mutations were analysed by lipogram assessments at baseline and after 3 months and 24 months of statin therapy (varying doses, however a mean effect was calculated within each group). Percentage reduction in total cholesterol was calculated.

**Results:** Four-hundred-and-ninety-two patients were analysed, 200 males and 292 females. Two-hundred-and-eighty-six patients were FH1, 183 were FH2 and 23 were FH3. The average total cholesterol at diagnosis was 8.9mmol/L for FH1, 9.5mmol/L for FH2 and 8.1mmol/L for FH3. The average total cholesterol (TC) for at 3 months was 7.3mmol/L (18% reduction) for FH1, 7.5mmol/L (21% reduction) for FH2 and 6.9mmol/L (15% reduction) for FH3. At 24 months the average TC was 6.43mmol/L (28% reduction) for FH1, 6.1mmol/L (36% reduction) for FH2 and 5.7mmol/L (30% reduction) for FH3. The effect of both time ( $p<0.0001$ ) and type of mutation ( $p<0.001$ ) was significant.

**Conclusion:** This study is the largest of its kind to compare the response to statin therapy amongst the three most common Afrikaner mutations. It showed that FH2 patients, although starting at a higher baseline TC, respond significantly better to statin therapy than FH1 and FH3 patients do.

## One year survival of patients with symptomatic dilated cardiomyopathy compared to peripartum cardiomyopathy

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**Background:** Little data are available from sub-Saharan Africa on the clinical characteristics and prognosis of symptomatic DCM.

**Objective:** The purpose of this study was to compare the 1 year outcomes of patients with symptomatic DCM compared to those with symptomatic PPCM, in a contemporaneous African setting with availability of evidence-based therapies.

**Methods:** One-hundred-and-forty-six patients were included in the study, comprising 92 DCM patients (51 female, mean age  $36.6 \pm 9.4$  years) and 54 PPCM patients (mean age  $38.4 \pm 9.8$  years) that were followed up serially between 1 January 2014 and 31 December 2014, in a dedicated specialist cardiomyopathy clinic at Groote Schuur Hospital, Cape Town, South Africa. Clinical characteristics, medical therapy, cardiovascular events and mortality were compared between the 2 groups.

**Results:** There were no significant differences between the baseline clinical features of patients with DCM and PPCM (besides gender). A high proportion of patients were treated with evidence-based therapies, including angiotensin converting enzyme inhibitors/angiotensin receptor blockers, beta-blockers, aldosterone inhibitors and diuretics and there were no significant differences in the management of DCM and PPCM patients. Patients with PPCM had a modest improvement in systolic function ( $p=0.026$ ) while there was no significant change in the LV ejection fraction (LVEF) of DCM patients ( $p=0.844$ ) over the follow-up period of 1 year. The 1 year case fatality rate of symptomatic DCM was 8.7% vs. 7.4% for symptomatic PPCM ( $p=0.783$ ). In this study, there was no association between LVEF and mortality ( $p=0.902$ ). Importantly, these data do not reflect the early outcome of PPCM and DCM, often associated with higher mortality.

**Conclusions:** The case fatality rate remains high in our setting despite the full application of evidence-based therapies. Our data emphasise the importance of individualised follow-up care and long-term continuity of care of patients with cardiomyopathy.

## Genetic contribution to the risk for metabolic syndrome

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**Introduction:** Metabolic syndrome (MS) is associated with lifestyle, environmental and genetic factors, with obesity and insulin resistance (IR) as driving factors for its progression.

**Aims:** To establish the genetic profiles of subjects in the Indian community of Phoenix and to determine genetic patterns associated with MS.

**Methodology:** This study investigated 1 000 subjects from the Phoenix Lifestyle Project. MS was diagnosed according to NCEP ATPIII, IDF and harmonised definitions and IR (HOMA-IR values  $>2.6$ ). The apolipoprotein A5 Q139X, lipoprotein lipase Hinf I, human paraoxonase 1 192Arg/Gln, cholesteryl ester transfer protein TaqIB, adiponectin 45T  $>G$  and leptin 25CAG were genotyped by real time PCR.

**Results:** The prevalence of MS was high (44.3%: NCEP ATPIII, 51.6%: IDF, 49.0%: harmonised). More females had MS than males [harmonised (51.0% vs. 42.8%)]. IR was high in MS subjects (harmonised 76.3%), with more females being IR than males [harmonised (78.3% vs. 69.2%)]. Genotypes/alleles of the polymorphisms in MS subjects varied, with no significance detected. Males with adiponectin TG genotype and human paraoxonase 1 AA genotypes were more inclined to have reduced HDL-C (harmonised  $p=0.001$ ) and higher systolic BP (harmonised  $p=0.02$ ) respectively. IR males (HOMA IR values  $>2.6$ ) with the lipoprotein lipase HinfI (GG genotype) and human paraoxonase 1 192Arg/Gln (AA genotype) were more likely to have higher systolic and diastolic BP (harmonised  $p<0.05$ ).

**Conclusion:** A high number of Indians living in Phoenix have MS. No association between the polymorphisms studied and the risk for MS was observed, except for an association with low HDL and hypertension (in males). The genetic risk for MS may lie in its components, rather than in MS as an entity.

## Depression symptoms and ocular perfusion pressure in black Africans - the eyes and not the carotid tell it all: The SABPA study

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**Introduction:** High pulse pressure exerts shear stress on vessel walls and may impair carotid and retinal perfusion, contributing to arterial stiffness and remodelling. Depression symptoms have been associated with poor prognosis of coronary artery disease and cardiac wall remodelling. We therefore aimed to assess the link between depressive symptoms and reduced perfusion to the carotid and the retina.

**Method:** A sub-Saharan African bi-ethnic gender cohort (Africans/Men  $\pm$  48%), aged  $48.6 \pm 9$  years, was included ( $n=359$ ). Cardiometabolic risk markers were obtained under fasting well-controlled conditions using standardised protocols. Depressive symptoms scores were calculated using DSMIV criteria. Ambulatory BP, ECG and ultrasound B-mode carotid far wall measures were obtained. DBP – Intra-ocular pressure defined ocular perfusion. Cardiometabolic retinopathy conditions were evaluated whilst static and dynamic retinal vascular calibre and responses were quantified from digital photographs using standardised protocols.

**Result:** Africans revealed more depressive symptoms, silent ischaemia, higher 24 hour pulse pressure, retinal remodelling (arteriolar narrowing) and augmented venular dilation during flickering light, when compared to Caucasians. Overall, depressive symptoms were associated with retinal arteriolar

narrowing, but not carotid remodelling in the bi-ethnic cohort. Depressive symptoms and ocular perfusion pressure were associated with a wider venular calibre ( $p<0.01$ ), a stroke risk marker; in Africans only. Ocular perfusion pressure was inversely associated with arteriolar calibre and dilation ( $p<0.01$ ) in African men.

**Conclusion:** More depressive symptoms may trigger changes in ocular haemodynamics of the Africans. Reduced perfusion in the eye might act as a trigger to increase blood pressure as compensatory mechanism to alleviate reduced supply. The increased risk for stroke in the Africans underscores the need for screening of diastolic ocular perfusion pressure and presence of depression symptoms to detect early onset of vascular pathology and potential subcortical ischaemia. It may improve insight on the retinal-heart circulation and future stroke.

### Infective endocarditis due to intravenous Nyaope use: The beginning of an epidemic?

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**Introduction:** Infective endocarditis (IE), secondary to intravenous drug abuse (IVDA), is uncommon in an African population. We recently described 3 cases of IE secondary to IV Nyaope (variable mixture of heroin, cannabis, antiretrovirals, metamphetamines) use. Since then we have encountered an additional 21 cases.

**Methods:** Patients presenting with IE, due to IVDA, at Chris Hani Baragwanath Academic Hospital were included in this prospective case series. All underwent echocardiography and additional investigations at the discretion of the treating physician.

**Results:** Mean age was 24.8 years (23 males). The majority presented with dyspnea (58%), symptoms of withdrawal (29%), peripheral suppurative infection (25%) and right ventricular (RV) failure (8%). Most were HIV reactive (87%) and not on ARVs. Three patients had a CD4 count  $\leq 200$  cells/ $\mu$ l. All were anaemic with raised infective markers. Renal failure requiring dialysis was present in two patients. Four patients had evidence of prior Hepatitis B infection and Hepatitis C antibody were detected in 42%. Staphylococcus aureus was cultured in 58% and Pseudomonas aeruginosa in 1 patient. Polymicrobial infection was found in 4 patients. Sterile blood cultures were present in 6 patients. Tricuspid valve endocarditis was present in the majority. Concomitant involvement of mitral valve and pulmonary valve was present in the minority. Only one patient had isolated aortic valve endocarditis. The mean vegetation size was 12.5mm and 9 patients had pulmonary hypertension and RV dysfunction. Septic pulmonary emboli were documented in 11 patients. All were treated with antimicrobials and only 3 patients underwent surgery. One patient died within 24 hours of admission secondary to respiratory failure.

**Conclusion:** IE secondary to IVDA is on the rise among young black South Africans and is responsible for morbidity and mortality in this group. A high degree of suspicion must be maintained in order to make an early diagnosis and initiate treatment.

### Left atrial volume and strain parameters using echocardiography in a normal sub-Saharan African population

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**Introduction:** Left atrial (LA) volume and function are important predictors of cardiovascular morbidity and mortality. No normative data are available for black Africans. Furthermore, patterns of left ventricular (LV) remodelling differ between races. We therefore sought to establish reference values for LA volume and strain ( $\epsilon$ ) in a normal black African population. We thus sought to: (1) establish normative values for LA volumetric and  $\epsilon$  parameters and (2) determine the effect of age related changes in LV diastolic function on measures of LA function.

**Methods:** The study comprised 120 normal individuals aged between 18 - 70 years (50% males). LA volumes and  $\epsilon$  were measured with biplane Simpson's method and Philips Qlab 9 speckle tracking software. LV diastolic function was assessed using the  $e/e'$  ratio.

**Results:** The mean age was  $38.7 \pm 12.8$  years. Overall maximum LA volume indexed to BSA (LAVi) was  $19.7 \pm 5.9$ ml/m<sup>2</sup> and minimum LAVi was  $7.7 \pm 3.2$ ml/m<sup>2</sup>. Peak global longitudinal strain in the reservoir phase ( $\epsilon_R$ ) was  $39.0 \pm 8.3\%$  and the peak strain in the contractile phase was  $-2.7 \pm 2.5\%$ . LA pump function increased with age ( $r=0.2$ ,  $p=0.02$ ), as the conduit function decreased with age ( $r=-0.3$ ,  $p<0.001$ ).  $\epsilon_R$  declined with age ( $p<0.001$ ). More advanced age was associated with decrease in diastolic function ( $r=0.4$ ,  $p<0.001$ ).

A positive correlation between  $e/e'$  lateral and LA conduit and booster pump parameters ( $r=0.3$ ,  $p<0.05$ ) was noted.  $\epsilon_R$  correlated negatively with diastolic function ( $r=-0.3$ ,  $p<0.002$ ). On multivariate regression analysis male gender and  $e/e'$  lateral were predictors of maximum LAVi ( $p<0.05$ ). Age and  $e/e'$  medial remained determinants of  $\epsilon_R$  ( $p<0.05$ ).

**Conclusion:** LA pump and conduit functions show a compensatory increase with normal ageing as diastolic function worsens. This occurs before alteration in maximum LAVi. Additionally,  $\epsilon_R$  may be a more sensitive marker for assessing LA function than maximum LAVi.

## Right ventricular strain using 2D Speckle tracking echocardiography in a normal African population

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**Introduction:** Right ventricular (RV) function is an important prognostic determinant in various cardiovascular diseases. RV strain ( $\epsilon$ ) is a relatively new technique for assessing subclinical RV dysfunction. There is a paucity of data regarding RV  $\epsilon$  in a black population. We thus sought to establish: (1) Normative RV  $\epsilon$  data in a black population; (2) determinants of RV  $\epsilon$ ; and (3) correlation of RV  $\epsilon$  with age and RV tissue Doppler parameters.

**Methods:** A prospective cross-sectional study, comprising 109 normal black African subjects was conducted at CHBAH from 2014 - 2015. The subjects were categorised into four age groups: 18 - 29, 30 - 39, 40 - 49 and >50 years. Peak RV free wall  $\epsilon$  and tissue Doppler parameters were measured on echocardiography (Phillips IE33 and QLAB 9 speckle tracking software).

**Results:** The mean age was  $36.3 \pm 13.2$  years (49% females). The RV  $E'$  decreased with increasing age on sub-group analysis ( $p=0.02$ ). There was a negative correlation between RV  $E'$  and age ( $r=-0.39$ ,  $p<0.001$ ). RV  $\epsilon$  was  $-19.1 \pm 3.5\%$ . There was no difference in  $\epsilon$  in between the age categories ( $p=0.059$ ). As the RV  $\epsilon$  increased (more negative), the RV systolic function ( $S'$ ) and early RV filling ( $E'$ ) increased ( $r=-0.21$ ,  $p=0.02$ ;  $r=-0.26$ ,  $p=0.005$ ). On multivariate linear regression analysis, age and RV  $E'$  emerged as the most important predictors of strain after adjusting for gender, RV  $S'$  and systolic blood pressure ( $r=0.4$ ,  $p<0.001$ ).

**Conclusion:** We have provided normative data for RV  $\epsilon$  in a normal African population. Age and RV  $E'$  were important determinants of peak systolic RV free wall  $\epsilon$ . This study provides a framework for future risk assessment and prognostication in varied cardiovascular diseases.

## The changing spectrum of chronic rheumatic mitral regurgitation in Soweto, South Africa

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**Introduction:** Previous studies have documented the high prevalence of rheumatic fever and rheumatic heart disease in the population of Soweto and have provided a detailed description of the echocardiographic findings in these patients. We sought to determine whether the clinical and echocardiographic characteristics of rheumatic mitral regurgitation (MR) had changed in a more contemporary population.

**Methods:** This prospective cross-sectional study was conducted at Chris Hani Baragwanath Academic Hospital (CHBAH) between January 2014 and October 2014. Eighty-four patients with moderate, or severe, chronic rheumatic MR who met the selection criteria were included in the study.

**Results:** The study comprised 84% females with a mean age of  $44 \pm 15.3$  years. Acute rheumatic fever (ARF) was documented in only 1 patient. Hypertension and HIV were present in 52% and 26% respectively. Echocardiography showed leaflet thickening and calcification, restricted motion and subvalvular disease in 41%, 25% and 34%, respectively. Carpentier IIIa leaflet dysfunction occurred in 80%. Leaflet prolapse was seen in only 20%. Patients older than 30 years more commonly had hypertension (69% vs. 9%,  $p<0.01$ ) and HIV (32% vs. 9%,  $p=0.03$ ). These findings are in marked contrast to previous literature, in which patients were younger (mean 19 years), commonly presented with rheumatic carditis, and had no comorbidities. Leaflets were pliable, with 84% having isolated leaflet prolapse, and no commissural fusion. Elongated (92%) and ruptured (25%) chordae predominated.

**Conclusion:** Contemporary patients with rheumatic MR are older with greater comorbidities. Acute rheumatic fever, in our setting, has become rare. Echocardiographic features of advanced valvular and subvalvular disease is common with very little isolated annular dilatation or leaflet prolapse. These findings may have significant implications for the management of rheumatic disease in the modern era and serve to inform strategies for future management of this disease.

## Utility of cardiac MRI in a case of sub-aortic aneurysm

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**Introduction:** Sub-aortic (SA) aneurysms are a rare entity of variable aetiology. Most are congenital and a result of a defect between the ventricular wall and valvular annuli. We report on the first case of SA aneurysm imaged using cardiac MRI.

**Case:** A 33-year-old female, with HIV and on HAART, presented with syncope and dyspnea. Clinical examination was compatible with moderate to severe aortic regurgitation (AR) confirmed by transthoracic and transesophageal echocardiograms. An eccentric jet precluded accurate assessment of the AR severity on echocardiography. Cardiac MRI was done to elucidate the aetiology, severity and mechanism of regurgitation. The anatomic relationships of the aneurysm were clearly defined. It confirmed the presence of a SA aneurysm below the left coronary cusp and its retraction resulting in an eccentric AR jet. An assessment of moderate AR, based on regurgitant volume, was made. Furthermore, compression of the left atrium, pulmonary artery and RVOT was noted. No coronary artery impingement was noted.

**Conclusion:** CMR is a useful, non-invasive technique for comprehensive assessment of SA aneurysm.

## Focal atrial tachycardia: A retrospective review of 11 cases at Groote Schuur Hospital, an experience from January 2013 - July 2015

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**Background:** Focal atrial tachycardia (AT) is an uncommon cause of a supraventricular tachycardia (SVT). There is a paucity of information of AT in the South African population. The aim of this study was to describe the incidence, clinical, ECG and electrophysiological characteristics of patients with AT.

**Methods:** A retrospective study was performed of consecutive patients with AT who underwent electrophysiological studies (EPS) with 3D mapping and radio frequency ablation (RFA) between January 2013 and July 2015 at Groote Schuur and UCT Private Academic Hospitals.

**Results:** A total of 262 patients with supraventricular tachycardia underwent an EPS. Eleven (3%) patients had a diagnosis of AT. The mean age of patients was 43 years of which 7 patients were female (64%). All patients presented with palpitations (100%) with a mean symptom duration of 8 months. Six (64%) patients had underlying left ventricular systolic dysfunction at presentation. Seven (100%) patients with left ventricular systolic dysfunction had incessant AT. Six (64%) patients had a long RP (short PR) tachycardia on ECG. The mean atrial rate of the AT was 162bpm (range: 120 - 210bpm). At 6 months post EPS and RFA, 8/9 (89%) had no palpitations and no recurrence of AT. There was a significant recovery in left ventricular systolic function in 7 (100%) of the patients (tachycardia induced cardiomyopathy) at 6 months post ablation.

**Conclusion:** AT is an uncommon cause of a SVT with a female preponderance. AT can cause both a long PR and short PR tachycardia. AT results in a tachycardia induced cardiomyopathy in a significant proportion of patients. EPS and RFA has a high success rate in treating AT.

## The proportion of patients in the Uganda Rheumatic Heart Disease Registry with advanced disease requiring urgent surgical interventions

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**Introduction:** Since the establishment of the Uganda Rheumatic Heart Registry, over 900 patients have been enrolled. We sought to stratify the patients in the registry, according to disease severity and optimal management strategy, with a view to categorise those who need invasive intervention.

**Methods:** We reviewed data of 618 patients who had enrolled in the Registry between March 2010 and February 2013. Of these, 67 patients who had died were excluded leaving 551 patients who were recruited. The optimum management strategy was determined according to the 2007 European Society of Cardiology Guidelines on the management of valvular heart disease.

**Results:** Out of the 551 patients' records evaluated, 398 (72.3%) require invasive intervention, with 332 (60.3%) patients requiring surgery and 66 (12.0%) requiring percutaneous mitral commissurotomy (PMC). This leaves only 27.7% of patients who require only medical management. Currently, the majority of the patients (498, 90.4%) in the registry are on medical treatment. Of the 60.3% requiring surgical intervention, only 8.0% (44 patients) underwent valvular surgery and 5 (1.0%) patients of the 66 (12.0%) underwent PMC successfully.

**Conclusion:** There is a high proportion of patients with advanced severe RHD who require invasive treatment, yet they cannot access this therapy due to the high cost.

## Giant right atrium due to congenital dysplastic tricuspid valve: A report on 2 cases

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**Introduction:** Congenital dysplastic tricuspid valve (TV) is a rare anomaly associated with tricuspid valve insufficiency that usually presents with right heart failure in the neonatal period.

**Case reports:** We report on 2 cases. The first patient is a 14-year-old male who presented for the first time with right heart failure and atrial fibrillation. The 2D echocardiogram revealed a giant right atrium with spontaneous contrast, a dysplastic TV and hypoplastic right ventricle (RV). Right atrial (RA) tumours were excluded. The second patient is a 11-year-old male who presented at age 6 years with right heart failure. He had recurrent pericardial effusions that required pericardiocentesis a pericardial window. He suffered a cerebral vascular accident post cardiac tamponade. The 2D echocardiogram showed a giant right atrium with spontaneous contrast, dysplastic TV and hypoplastic RV.

**Discussion:** Dysplastic TV encompasses a morphologic spectrum of congenital malformation of the Tricuspid valve leaflets, chordae tendinae and papillary muscles that include either focal or diffuse thickening of valve leaflets, underdevelopment of chordae tendinae and papillary muscles, incomplete separation of valve components from the RV wall or focal agenesis of valvular tissue. In some cases annular dilatation is more significant than the dysplastic changes. Annular attachment of leaflets is normal without apical displacement.

There is functional impairment of the RV with severe TR that retards prograde flow through the right heart, leading to functional or anatomical RVOTO and RA dilatation. Differential diagnoses include: Ebstein anomaly, Uhl's anomaly, unguarded TV orifice and endomyocardial fibrosis of the RV. Timing of presentation depends on the degree.



## Multiple large vessel aneurysms with airway obstruction: A case of Loey-Dietz Syndrome

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**Introduction:** Loey-Dietz syndrome is a rare inherited connective tissue disorder (CTD) that presents with multiple aneurysmal dilatations of the aorta and its branches.

**Case report:** A 6-month-old girl presented with a lower respiratory tract infection in severe respiratory distress. Clinically she had subtle dysmorphism and a pulsatile mass in the right axilla. A chest X-ray revealed a large radio-opaque lesion occupying most of the superior mediastinum and left hemithorax. A 2D echocardiogram revealed a large aneurysm of the transverse aorta with a moderate patent ductus arteriosus, massively dilated pulmonary arteries and no pulmonary hypertension. The CT angiography revealed multiple aneurysmal dilatations of the aorta, common carotid arteries, subclavian and axillary arteries. Genetic analysis revealed a heterozygous c.1460G>A (p.ARG487Gln) TGFBR1 mutation which confirmed the diagnosis of Loey-Dietz syndrome. Aortic dilatation resulted in severe airway compression manifesting clinically with respiratory distress. As there was no cardiothoracic intervention available to treat such severe aortic dilatation and airway compression, supportive management was employed. The child demised a few months later, most likely from a ruptured aneurysm.

**Discussion:** Loey-Dietz syndrome is a rare autosomal dominant inherited CTD. Its exact inheritance is not known. It is caused by genetic mutations in the transforming growth factor beta receptor (TGFBR) 1 and 2 and TGFBR2. It should be considered in patients that present with thoraco-abdominal aneurysms. The vascular abnormalities include: aortic root dilatation and dissection in addition to aneurysms and tortuosity throughout the arterial tree. It can also present with skeletal, skin and craniofacial abnormalities. Differential diagnoses include: Ehler-Danlos Syndrome, atypical Marfan Syndrome and familial thoracic aortic aneurysm. Management should be individualised to the needs of the patient.

## Catheterisation induced Takotsubo cardiomyopathy

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Takotsubo cardiomyopathy, or apical ballooning syndrome, was first described in 1990 in the Japanese population. Increasing reports of the condition have led to it being incorporated in 2006 into the American Heart Association classification of primary acquired cardiomyopathies. Interestingly, even though Takotsubo cardiomyopathy has classically been described in the literature as primarily an adult disease, it has now been identified in neonates as young as 2 weeks old.

Miss KM is a 21-year-old female who was diagnosed with coarctation of the aorta, ventricular septal defect and hypothyroidism at the age of 1 year. She has been for a subclavian flap, pulmonary artery banding, VSD closure and pulmonary artery debanding. Twenty years later it is noted that she has increasing gradient across the coarctation repair site necessitating catheterisation for percutaneous stent insertion. She developed signs of cardiac failure with pulmonary oedema and features of a right hemiplegia during stent insertion. Angiography of the descending aorta showed no obvious problems with the stent. ECG showed new onset ST changes and echocardiography revealed a 'ballooned out', myopathic apical segment of the left ventricle with hypercontractile basal segment, as described in Takotsubo cardiomyopathy.

The recognition of this disease in the paediatric populations necessitates a closer look at what is known about this cardiomyopathy in children, as well as the management and predisposing factors, so that it may be correctly recognised and managed.

## Coronary cameral fistulas

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Coronary cameral fistula is a rare congenital abnormality of the coronary arteries accounting for 0.2 - 0.4% of congenital cardiac abnormalities. In the adult population it is found in 0.08 - 0.3% of patients undergoing angiography for other reasons. It has a wide spectrum of presentation and severity. This is a presentation of 3 cases of coronary cameral fistula, in an 8-day-old neonate, a 4-year-old child and a 30-year-old adult.

Case 1 is a 8-day-old neonate who presented with severe metabolic acidosis, renal failure and signs of right heart failure, together with a continuous murmur and displaced apex. ECG showed ST-depression and t-wave inversion in V4 and V5. Echocardiography revealed a massive left coronary cameral fistula draining into the apex of the right ventricle.

Case 2 is a 4-year-old boy who presented with gastroenteritis and the incidental finding of a continuous murmur with cardiomegaly on chest X-ray. He remained asymptomatic. There were no ischaemic changes on ECG. Angiography revealed an aneurysmal coronary cameral fistula of the right coronary artery.

Case 3 is that of a 30-year-old female presenting for the first time with atypical chest pain and ventricular bigemy together with a continuous murmur. Angiography revealed a massively dilated anomalous coronary arising off the left coronary cusp and draining into the right atrium. A normal left anterior descending artery and circumflex artery seem to arise from the aneurysm.

The management of these patients is always a topic for discussion as to whether the surgical or percutaneous route is the more favourable one, and the implications thereof with regards to thrombosis and embolism, and whether any management is necessary at all if the patient remains asymptomatic.

## Holt-Oram syndrome

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**Introduction:** Hand-heart syndromes are genetically acquired disorders that compose of congenital cardiac and limb deformities. We present a case of Holt-Oram Syndrome (HOS) with a family history that spans 3 generations.

**Case presentation:** A 25-year-old female, presented in preterm labour at 20 weeks. The foetus was born without thumbs in both hands and demised at 3 weeks. On enquiry the patient reports a 10 year history of dizziness, palpitations and exertional dyspnoea. General examination revealed bifid fingerised right thumb, absent left thumb, clinodactyly, bilateral hypoplastic thenar eminences, narrow sloping shoulders and pectus excavatum. On cardiac examination, there were features suggestive of an atrial septal defect (ASD) which was confirmed on transthoracic echocardiogram. An ECG showed Atrial Flutter. Family history revealed that the patient's mother was born with abnormal thumbs. She had 3 children with different partners. The index patient is the eldest child and, at birth, was found to have an absent thumb on the left hand. The second child died at seven months, after surgery to correct a congenital cardiac defect. The third child underwent a ventricular septal defect (VSD) closure at the age of one and also has skeletal abnormalities.

**Discussion:** HOS is a rare genetic condition. It has an autosomal-dominant mode of genetic transmission. The responsible gene has been mapped to 12q24.1 which encodes human transcription factor TBX5. TBX5 provides instructions for making a protein that plays a role in the development of the heart and upper limbs. It is the mutations in this gene that lead to a wide range of phenotypes. Abnormal carpal bones are present in all cases. There is usually a personal or family history of congenital heart disease (ASD or VSD). ECGs may reveal conduction abnormalities. Management will depend on the underlying cardiac and skeletal defect. Genetic counselling should be offered to all these patients.

## Unravelling right ventricular dysfunction in isolated left ventricular non-compaction

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**Introduction:** The prevalence of right ventricular (RV) dysfunction varies amongst patients with isolated left ventricular non-compaction (ILVNC) and has not been systematically studied in a sub-Saharan population. We aimed to (1) determine the prevalence of RV dysfunction in a sub-Saharan population of ILVNC patients; (2) compare the clinical and echocardiographic characteristics of ILVNC patients with RV dysfunction to those without RV dysfunction.

**Methods:** Forty-nine patients with ILVNC, with no evidence of RV non-compaction, were enrolled in this cross-sectional study at Chris Hani Baragwanath Hospital, and assessed for RV dysfunction using echocardiography. RV systolic function was evaluated using the tricuspid annular peak systolic velocity (S'). Patients with an RV S' < 10cm/s were classified as having RV systolic dysfunction.

**Results:** The mean age was  $41.9 \pm 12.0$  years (43% male). The left ventricular end diastolic diameter and ejection fraction (EF) were  $58.3 \pm 10.3$ mm and  $28.2 \pm 1.5\%$ , respectively. The pulmonary artery systolic pressure (PASP) was  $41.8 \pm 19.8$ mmHg with 39% having a PASP > 35mmHg. Moderate or severe tricuspid regurgitation was found in 35% of cases. RV dysfunction was noted in 49% of patients. No differences were noted with regard to NYHA class, LVEF, LV diastolic parameters, parameters of LV remodelling, the presence of moderate/severe mitral regurgitation or pulmonary hypertension between patients with ILVNC, with or without RV dysfunction.

**Conclusion:** Almost 50% of patients with ILVNC have RV dysfunction. RV dysfunction does not appear to be related to the severity of left ventricular dysfunction and must therefore reflect an intrinsic abnormality of RV contractile function.

## Should immunologist be part of the heart team? A glimpse into the management of rare (likely, under reported) cause of acute coronary syndrome

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**Introduction:** Kounis syndrome is characterized by a group of symptoms that manifest as unstable vasospastic or non-vasospastic angina secondary to a hypersensitivity reaction. It was first described by Nicholas Kounis and Zavras in 1991 as a concurrence of an allergic reaction with anaphylactoid or anaphylaxis and coronary artery spasm, or even myocardial infarction. In addition to coronary arterial involvement, the entity "Kounis syndrome" today encompasses other arterial systems with similar physiology, such as mesenteric and cerebral circulation, resulting in ischaemia/infarction of the vital organs supplied. Incidence of this condition is hard to delineate due to the number of potential instigating factors and its relatively infrequent documentation in literature. It remains unclear whether the reported lower rates of this condition are the direct result of remissness.

**Case:** We report a case of a 39-year-old man, HIV negative, with no coronary risk factors nor family history of premature coronary artery disease who developed, following administration of ciprofloxacin for dysuria, severe sudden onset of retrosternal chest pain, radiating to both arm, with associated autonomic symptoms, in addition to the severe generalised pruritic erythematous rash and swelling of the hands, feet and oedema. An electrocardiogram (ECG) showed inferior ST-Segment elevation with posterolateral extension. A diagnosis of acute coronary syndrome: Inferior STEMI with posterolateral extension was made and Tenecteplase, a thrombolytic therapy was administered, no resolution of symptoms, nor of ST-Segment on serial ECG's. He was subsequently referred to our unit (PCI-capable center) for rescue PCI. On arrival in our institution and after meticulous history taking, allergic myocardial infarction... Kounis syndrome, was entertained, a blood specimen for tryptase and IgE, among other investigation was sent to the laboratory. Prednisone, diphenhydramine, ranitidine and amlodipine were administered, symptoms subsided, ST-Segment elevation resolved. Cardiac catheterisation and coronary angiogram done within 6 hours after admission as a pharmaco-invasive strategy, showed unobstructed coronaries. Allergy physicians were consulted for further management.

**Conclusions:** There is paucity of data as to the incidence and prevalence of Kounis syndrome in this country (South Africa), rare as it is, it remains not infrequent. It is important to be aware of its existence. Treatment guidelines for the patients with Kounis syndrome have not been established, and most of the information about the treatment of this syndrome comes from individual case reports or case series. Kounis syndrome should be considered in young, healthy patients with no atherosclerotic risk factors when they develop acute coronary syndrome (especially inferior myocardial infarction) after administration of potentially allergic agent. These patients need treatment with steroids, antihistamines, fluid resuscitation, possibly epinephrine, oxygen, and anti-thrombotics before transfer to the cardiac catheterisation laboratory. An allergy work-up should include the assessment of other allergies to food, insect stings and other environmental agents. Skin tests and food challenges may be useful in identifying the culprit agent. The intricate management of patients with Kounis syndrome, warrants allergy physicians (Immunologists) to be remote members of the "Heart Team".

## Efficacy and safety of liraglutide vs. sulfonylurea, both in combination with metformin during Ramadan in subjects with Type 2 diabetes (LIRA-Ramadan): A randomised trial

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**Introduction:** Subjects with type 2 diabetes (T2D) who fast during Ramadan have a 5 and 7.5 fold increased risk of severe hyper- and hypoglycemia, respectively. The effect of liraglutide (lira) vs. sulfonylurea (SU), both + metformin (Met), on glycaemic control in subjects with T2D who fasted during Ramadan was examined.

**Methods:** In this up to 33 week, open-label trial, adults (HbA1c 7 - 10%; BMI >20 kg/m<sup>2</sup>; stable SU + Met; intent to fast during Ramadan) were randomised to either switch to once daily lira 1.8mg (n=172) or continue pretrial SU (n=171), both + Met. After 3 week dose escalation, a 6 - 9 week maintenance period preceded Ramadan. The primary endpoint was a change in fructosamine (FA), from start to end of Ramadan (lira n=151; SU n=165).

**Results:** During Ramadan, despite lower mean FA and HbA1c at start of Ramadan in the lira arm, a similar reduction in FA with lira and SU was seen (mean reduction 12.8umol/L and 16.4umol/L respectively). Confirmed hypoglycemic episodes appeared to be lower with lira and fewer subjects withdrew during Ramadan (lira 3, SU 11). During Ramadan, lira was associated with greater weight loss [1.4kg compared to SU (0.89kg)]. AE frequencies appeared similar: lira 23.7%; SU 20.9%. GI AEs were more common for lira (10.5%; SU 3.7%). A low incidence of SAEs was observed (lira 1.3%; SU 0%).

**Conclusion:** During Ramadan, lira showed similar improvements in glycaemic control based on FA and HbA1c levels compared to SU with a similar number of AEs, apparently fewer confirmed hypoglycemic episodes and better weight control.

## Cardiovascular magnetic resonance characterisation of myocardial involvement in tuberculous pericarditis with, and without, HIV co-infection

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**Background:** Little is known about myocardial involvement in tuberculous pericarditis (TBP). Cardiovascular magnetic resonance (CMR) can non-invasively assess cardiac function, myocardial oedema, inflammation and fibrosis.

**Objective:** The purpose of this study was to assess cardiac and pericardial structure and function in patients with TBP, with and without HIV co-infection, and to assess the relationship of LV function with other imaging biomarkers.

**Methods:** Seventy-two patients with TBP [37 male (51.3%), mean age  $40 \pm 14.3$ ] were included in the study. Of these, 35 were HIV-infected [17 male (48.6%), mean age  $34 \pm 8$ ] and 37 were HIV-uninfected [20 male (54.1%), mean age  $51 \pm 16$ ]. Assessments included clinical examination, ECG, echocardiography, serum and pericardial biomarkers and CMR (biventricular volumes and function, oedema and late gadolinium enhancement - LGE).

**Results:** HIV-infected TBP patients were younger ( $p < 0.001$ ), had lower serum haemoglobin ( $p < 0.001$ ) and were more likely to have NYHA class III and IV symptoms ( $p < 0.001$ ). There were no differences on ECG and echocardiography between HIV-infected and uninfected TBP patients. There were also no differences in global systolic function between HIV-infected and uninfected TBP patients. Focal fibrosis on LGE was found more commonly in those with HIV infection ( $p < 0.001$ ). Pericardial effusions were frequent ( $> 50\%$ ) in both groups of TBP patients. Determinants of LV ejection fraction in TBP included heart rate, LV size, E/A ratio, pericardial LGE and pericardial thickness (all  $p < 0.01$ ).

**Conclusions:** HIV co-infection is associated with increased focal myocardial fibrosis in TBP patients, suggesting increased myocardial inflammation in those with HIV co-infection. In the future, it will be important to assess the prognostic significance of these findings.

## Risk factors associated with acute coronary syndromes in South Africa

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**Aims:** To examine the association between traditional risk factors and acute coronary syndrome (ACS) in the South Asian Indian population in KwaZulu-Natal, South Africa.

**Methods and results:** The study population comprised 4 418 patients with a mean age of  $54.6 \pm 10.9$  years, of whom 67% were males. The majority presented with STEMI (75%), 16% had NSTEMI and 9% unstable angina. Visceral obesity (82%, mean waist circumference  $101.43 \pm 10.34$ cm) was the most commonly observed risk factor, while 78% had hypercholesterolaemia (mean  $5.97 \pm 1.11$  mmol/L) and 74% had a family history of vascular disease. More males compared to females were smokers ( $p < 0.0001$ ), while females were more likely to have visceral obesity, diabetes, hypertension, increased BMI and low HDL cholesterol levels ( $p < 0.0001$ ). Young patients ( $\leq 45$  years,  $n=968$ ) had a higher incidence of family history of vascular disease (83%,  $p=0.019$ ), smoking (79%,  $p < 0.0001$ ) and hypertriglyceridaemia (62%,  $p=0.014$ ) compared to middle (46 - 65 years,  $n=2 708$ ) or old age ( $> 65$  years,  $n=742$ ) groups, whilst older patients were more likely to have diabetes (59%,  $p=0.001$ ) and hypertension (68%,  $p < 0.0001$ ).

**Conclusion:** Asian Indians in South African have multiple risk factors for ACS, possibly contributing to the increased incidence of coronary heart disease at a young age. This study further confirms that a family history of vascular disease is strongly associated with the presence of ACS, and should be incorporated in future risk factor analyses.

## Admission, 24 hours and discharge troponin T amongst acute myocardial infarction patients: Differing by prognostic contribution

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**Aims:** We examined the prognostic performance of measurements of cTnT concentrations at admission, compared to discharge, in predicting major cardiovascular events during hospital admission and at six months follow-up.

**Methods and results:** The study population comprised 1 351 patients with AMI and a mean age of  $57.5 \pm 11.4$  years, of whom 66% were males. Cardiac TnT was measured on admission, 24 hours, and at discharge using the Elecsys 2010 (Roche Diagnostics). No significant difference was found in patients who were cTnT negative at admission [ $n=345$  (26%)] compared to the cTnT positive group [ $n=1 006$  (74%)], with respect to baseline characteristics, infarct pattern, biochemical data and major cardiac events. In 475 patients (35%), serum cTnT levels were found to be higher on discharge from the CCU compared to admission/24 hour levels. A significantly greater proportion of patients had hypertension (63% vs. 50%,  $p < 0.001$ ), higher systolic blood pressures (133, IQR 115 - 154 vs. 127, IQR 111 - 147,  $p < 0.001$ ), history of previous AMI (17% vs. 9%,  $p < 0.001$ ) and previous angina (17% vs. 9%,  $p = 0.001$ ) if the discharge cTnT levels exceeded the admission/24 hour levels. A total of 120 deaths occurred during the study period with a significantly greater number of deaths recorded in patients whose discharge cTnT levels were higher than the admission/24 hour values [54 (11%) vs.

66 (8%);  $p=0.02$ , respectively]. Multivariable analysis using logistic regression showed that cardiogenic shock [OR 5.92 {95% CI 2.86 - 12.28};  $p<0.001$ ], cardiac failure [OR 4.80 {95% CI 2.61 - 8.82};  $p<0.001$ ], cerebrovascular accident [OR 3.95 {95% CI 1.48 - 10.58};  $p=0.01$ ], complete heart block [OR 3.50 {95% CI 1.22 - 10.09};  $p=0.02$ ], increasing age [OR 1.04 {95% CI 1.02 - 1.01};  $p<0.001$ ] and a greater discharge cTnT value [OR 1.61 (95% CI 1.01 - 2.56);  $p=0.04$ ] conferred a significantly higher odds of mortality.

**Conclusions:** This study shows that, in addition to cardiogenic shock, cardiac failure, cerebrovascular accident, complete heart block and increasing age, higher cTnT level at discharge is an important independent predictor of mortality in patients with AMI, and could further improve the prognostic accuracy of admission values of cTnT, based on relevant patents.

## Lens changes and radiation safety practices amongst paediatric cardiologists in South Africa: Preliminary results

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**Introduction:** The eyes are one of the most radiosensitive organs. The effects of radiation on the eyes ranges from burns, keratinisation, macula changes and cataracts. The posterior sub-capsular lens is the most common site for cataracts and are 3.9 times more likely in interventional cardiologists. Radiation occupational workers are exposed to increased radiation risk in the absence of adequate radiation safety practices and with inadequate utilisation of personal protective equipment. Cardiologists are of the most at risk occupational categories for radiation exposure. This study was done to assess the understanding of paediatric cardiologists and their ability to mitigate these risks, and to determine if lens changes are seen in this occupational group.

**Methods:** This was a cross sectional study. Data were collected at a Paediatric Cardiology workshop (Red Cross Children's Hospital). Participants completed a survey and screened their eyes for lens changes. Data were analysed descriptively. Ethics clearance (ECUFS 44/2015) was obtained from the UFS.

**Results:** The response rate was 60% (24). Three participants were excluded because they did not meet the inclusion criteria, 38% (8) were female. The average age was 43, 19% (4) had myopia whilst 24% (5) reported chronic eye infections. Of the participants 33% (7) reported having previously been screened for cataracts with no eye changes. The average number of years worked was 9.6. The average number of diagnostic procedures per week were 4 and the interventional procedures were 2.5 per week. Everyone used a lead apron all the time and 80% (17) used a thyroid shield all the time whilst 95% (20) never used lead glasses. Of the participants 33% (7) had cataract changes and 14% (3) had other eye changes.

**Conclusion:** Preliminary results showed eye changes in 33% of participants. There was low compliance with the PPE utilisation. There needs to be greater vigilance and increased radiation safety compliance to mitigate for the risks.

## Children admitted to a paediatric intensive care unit (PICU) with Fulminant Dilated Cardiomyopathy (DCMO) or Myocarditis in a tertiary academic hospital

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**Introduction:** In South Africa curative treatment for children with DCMO/myocarditis is limited. During exacerbation these children need frequent readmission for supportive treatment.

**Method:** A retrospective folder review describing patient characteristics, PICU treatment and outcome of children with fulminant DCMO/myocarditis admitted to Red Cross PICU between January 2010 and July 2015.

**Results:** Ninety-five children (36 males, 59 female), median age of 27.8 months were identified. All presented in Ross stage 4 cardiac failure (77% in cardiogenic shock), with an overall median lactate of 6.5mmol/l on admission. On presentation the left ventricular ejection fraction was  $<30\%$  in 79% and 9 developed intra-cardiac clots. Aetiology was presumed viral myocarditis in 87% and 13% idiopathic. Adenovirus PCR was positive in 28, Parvovirus in 19 with multiple positive viral studies in 32.

The median number of ICU admissions per patient was 1.5 (range 1 - 5) and length of ICU stay was 14.9 days (1 - 69). Fifty-five percent required ventilation for median of 8.1 days. Hundred percent required inotropic support for a median of 8.2 days. Eighty-two present were treated with Milrinone, 78% on Dobutamine and 33% on Adrenaline infusions. The median maximum inotrope score was 21.9.

Complications during ICU stay included: kidney injury in 68% with two patients needing dialysis, liver derangement in 43%, neurological events in 25% and 34% suffering a cardiac arrest episode. Thirty-three percent had arrhythmias of which 27% needed electrical cardioversion and 57% drug treatment during PICU stay.

Sixty-three (66%) children survived to ICU discharge. The overall survival was 47%. Of the survivors, the median number of ward readmissions was 3.7 (range 1 - 19). Total median length of ward stay was 23.2 days (1 - 138).

Of the survivors, only 22 were seen at cardiac outpatients during 2015, the rest are presumed lost to follow up.

**Conclusion:** In our setting DCMO/myocarditis is associated with significant duration of hospital stay, morbidity and mortality.

## Paediatric Cardiac Critical Care (PCCC) in a South African tertiary academic hospital

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**Introduction:** Knowledge is limited on PCCC in South Africa, apart from cardiac surgery reports. This study describes the patient profiles of PCCC admissions in a South African paediatric intensive care unit (PICU).

**Method:** A prospective review describing consecutive patients admitted to Red Cross PICU with a cardiac diagnosis between January 2015 and June 2015.

**Results:** Two-hundred-and-twelve PCCC patients needed 234 PICU admissions (95 males, 117 females) at median age of 33.2 months (range 1 day - 199 months). One-hundred-and-thirty-seven admissions followed cardiac surgery, 7 elective general surgery and 90 were medical emergencies. Sixty-four children had a genetic diagnosis.

Indications for emergency admission included: need for respiratory support (26%), cardiogenic shock (21%), severe cyanosis (20%), decompensated cardiac failure (17%), post cardiac arrest (14%) and life threatening arrhythmias (2%).

The median durations of PICU stay, ventilation and inotropic infusion were 10.5, 4.8 and 4.4 days in the emergency group vs. 3.5, 1.6 and 2.3 days in the cardiac surgery group respectively. The median maximum inotropic score was 11 in both groups.

The first diagnosis of a cardiac problem was made on this admission in 33 patients and 54 theatre procedures (24 cardiac surgical) were undertaken from PICU. Twenty-two patients needed 32 PICU re-admissions and 13 underwent redo cardiac operations during this study period.

Complications during PICU stay for the emergency vs. the cardiac surgery group included kidney injury in 26 vs. 11, neurological events in 19 vs. 4, pulmonary hypertension requiring inhaled nitric oxide in 7 vs. 10, arrhythmias requiring intervention in 12 vs. 31 and cardiac arrest episodes in 14 vs. 4 patients.

PCCC mortality was 5.5% vs. predicted mortality (PIM2) of 10%. Twelve of the emergency patients died in PICU vs. 1 elective cardiac surgery patient.

**Conclusion:** In this centre 18% of PICU admissions are PCCC admissions. Compared to elective admissions, emergency PCCC admissions have a higher morbidity and mortality.

## One event leads to another: First description of Danon's disease in South Africa

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**Introduction:** The first case in South Africa of Danon's disease, a X-linked dominantly inherited disease with cardiomyopathy, myopathy, mild mental retardation, liver function abnormalities in males and late onset cardiomyopathy in females, is described. It is caused by a deficiency of lysosome associated membrane protein 2 (LAMP-2).

**Case report and discussion:** Liver function abnormalities, prior to initiating isotretinoin for acne in a 16-year-old boy, prompted referral. The combination found of hypertrophic cardiomyopathy and ECG evidence of pre-excitation has a limited differential diagnosis, namely, Danon's, Fabry's and Pompe's disease, mutations in PRKAG2, some mitochondrial disorders and tuberous sclerosis. If you add mild learning disabilities and proximal muscle weakness and the death of his mother at age 40 with a dilated cardiomyopathy (DCM), in spite of cardiac resynchronisation therapy, Danon was most likely. Two siblings were unaffected.

The probands underwent skeletal muscle biopsy, endomyocardial biopsy and analysis and screening for LAMP-2 for disease-causing mutations using DNA sequencing. Autophagic vacuoles with sarcolemma features (AVSFs), the latter setting it apart from acid maltase deficiency, were seen within lysosomes. Furthermore, Immunohistochemical analysis showed a complete absence of LAMP-2 protein when compared with a control. DNA sequencing of LAMP-2 identified a c.973insC frameshift mutation, affecting all amino acids downstream not present in healthy ethnically matched controls. The poor prognosis of Danon's was exemplified in our patient when he died, age 18, with refractory heart failure after initiation of transplant planning and despite accessory pathway ablation with ICD insertion.

Recognising a unique combination of clinical symptoms, signs and events lead to the first South African diagnosis of Danon's disease. This is supported by histology, histochemistry and a highly plausible mutation.

## Symptoms and signs of heart failure at admission and discharge and outcomes in the sub-Saharan acute heart failure (THESUS-HF) registry

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**Introduction:** Symptoms and signs of heart failure (HF) are the leading reasons for admission to hospital for acute HF (AHF) and are used routinely throughout admission in clinical practice to assess the severity of disease and response to therapy.

**Methods:** The data were collected in the THESUS-HF study, a prospective, multi-center, observational survey of AHF from 9 countries in sub-Saharan Africa. Patients older than 12 years diagnosed with AHF, based on symptoms and signs, were enrolled. Signs and symptoms of HF were assessed at entry and on days 1, 2 and 7 (or discharge if earlier) and included oxygen saturation; degree of oedema and rales; body weight and level of orthopnea. Changes in dyspnea and well-being relative to admission were also assessed on days 1, 2 and 7 (or discharge, if earlier). Patient determined dyspnea, as well as general well-being, physician determined symptoms and signs of HF as well as vital sign measurements improved throughout the admission.

**Results:** After multivariable adjustment, changes in oxygen saturation and general well-being to day 7, or discharge, predicted death or HF hospitalisation through day 60, while baseline orthopnea, oedema, rales and respiratory rate and changes to day 7, or discharge, in respiratory rate, oxygen saturation and general well-being were predictive of death through day 180.

**Conclusion:** In AHF patients in sub-Saharan Africa, symptoms and signs of HF improve throughout admission and repeat assessments of oxygen saturation and general well-being are a valuable tool in predicting outcome.

## Valvular atrial fibrillation - a common form of AF among acute heart failure patients in sub-Saharan Africa: Insights from THESUS-HF

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**Background:** Rheumatic heart disease (RHD) is one of the leading non-communicable diseases in low- and middle-income countries and a common cause of heart failure in sub-Saharan Africa. Patients with RHD also suffer from complications related to atrial fibrillation (AF), infective endocarditis and the attendant consequences of thromboembolism. Little is known about the burden of Valvular AF among patients with acute heart failure in sub-Saharan Africa.

**Methods:** THESUS-HF was a prospective, multi-center, observational survey of AHF from 9 countries in sub-Saharan Africa. Patients older than 12 years, diagnosed with AHF based on symptoms and signs, were enrolled. ECGs were analysed for conduction or rhythm disturbances, evidence of myocardial ischaemia/infarction or hypertrophy. The information obtained was entered into the database registry, together with other clinical data.

**Results:** Information on the presence, or absence, of AF was available for 998 (99%) of the 1 006 patients. AF was present in 184 (18.4%) of the 998 patients. Valvular heart disease was responsible for AF in 48% of the patients with AF, second only to hypertensive heart disease with 51%. Compared to the patients without AF, the patients with AF are older (mean age 56.8 vs. 51.3 years) and more likely to be females (58.2% vs. 49.2%). The presence of AF was not associated with all cause death or readmission through 60 days, or all cause death through 180 days in the cox regression models.

**Conclusions:** In sub-Saharan African patients admitted for acute HF, AF is common as it is present in almost one fifth of patients, largely of non-ischaemic aetiology. Valvular AF causes significant morbidity. This underscores the need for proper anticoagulation to prevent thromboembolism as well as the need for research on the role of the new oral anticoagulants (NOACs) in valvular AF.

### How effective is de-endothelialisation of hearts by different concentration of deoxycholic acid in the rat model?

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**Aim:** Cardiac transplantation is a well-known treatment for the end-stage of heart failure, however cardiac allograft vasculopathy (CAV) is a serious complication, which might lead to graft failure. This study is performed to establish a model to investigate the impact of endothelial cell function on CAV.

**Material and methods:** Fourteen male Sprague Dawley rat hearts were excised and perfused with cardioplegia before attached to an Easy Cell Extraction System. De-endothelialisation of the hearts was performed by either deoxycholic acid sodium salt (DOA) perfusion at different concentrations (group 1: 1%, n=5; group 2: 0.1%, n=4; group 3: 0.01% n=5). All groups were treated for 5 minutes, extensively rinsed and perfused with Tyrode solution at the Langendorff system to study contractility. After finalising functional testing, the specimens were fixed and embedded. Hematoxylin and Eosin staining (H&E) and von Willebrand factor (vWF) staining with 3'-3-Diaminobenzidin (DAB) was used for quantified visualisation of endothelial cells persistence within the coronary system at the different areas of arteries, veins and capillaries after de-endothelialisation.

**Results:** De-endothelialisation performed with DOA, independent on the concentration of groups 1, 2 and 3, resulted in the absence of heart contractility. Group 1 showed a quantitative reduction of endothelial cells by  $66.9 \pm 3.7\%$ , in group 2:  $47.7 \pm 4.5\%$  and in group 3:  $39.3 \pm 3.6\%$ . There was a statistical significant difference between the reduction of endothelial cells in group 1 vs. group 2 ( $p \leq 0.05$ ) and group 1 vs. group 3 ( $p \leq 0.05$ ). Histological examination showed a similar pattern within the different areas of the coronary system.

**Conclusion:** Further studies are needed to optimise the concentration of de-endothelialised agents to produce an endothelial cell free heart, without losing contractility.

### Reprogramming of bone marrow stem cells into venous endothelial cells

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**Objective:** This study was initiated to evaluate reprogrammed bone marrow stem cells (BMSCs) into functional venous endothelial cells (VECs).

**Material and methods:** Pelvic bone marrow aspiration was performed in six mini-pigs to harvest BMSC using a filter with a 100µm cell strainer to determine bone remains and tissue fragments. Bone marrow-derived mononuclear cells were isolated by sucrose gradient centrifugation, adding endothelial growth factor (16µg/ml) into the culture medium to transform BMSCs into VECs. BMSCs transformation and characterisation was examined by immunofluorescence staining for CD 31 (PECAM) and von Willebrandt factor (vWF) staining as well as the expression of endothelial nitric oxide synthase (eNOS). Further NO release was examined by spectrophotometric investigation. To investigate whether transformed BMSCs could form angiogenic tubes in a 3D culture, angiogenesis assay with matrigel was achieved.

**Results:** Reprogrammed BMSCs exhibited a typical cobblestone-like endothelial cell phenotype. Immunofluorescence staining, including PECAM and vWF, showed that these differentiated BMSCs were similar to mature VECs. Furthermore, eNOS expression, by immunofluorescence staining and western blot, showed no differences between BMSCs and venous and arterial ECs. Spectrophotometric examination showed comparable NO release for reprogrammed BMSCs ( $550.0 \pm 115.0 \mu\text{mol/ml}$ ) and VEC ( $507.5 \pm 92.5 \mu\text{mol/ml}$ ) under 1mmol/l ATP stimulation and at control conditions (BMSCs:  $72.5 \pm 47.5 \mu\text{mol/ml}$  vs. VEC:  $55.0 \pm 3.8 \mu\text{mol/ml}$ ). Angiogenesis assay with matrigel indicated the tube formation of reprogrammed BMSC in vitro after 18h (grading 3.25) as seen in ECs. BMSCs cultured without endothelial growth factor; however were not able to develop tubes.

**Conclusion:** This study showed that reprogrammed BMSCs can successfully be transformed into functional venous endothelial cells.



## Saving lost lives: The Namibian Children's Heart Project

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**Introduction:** Prior to 2009, there were no cardiac services in Namibia. The aim of this project is to provide treatment for children with heart disease. Between September 2008 and December 2014, 148 patients seen by a single cardiologist in the Paediatric and Congenital Heart clinic at Windhoek Central Hospital, were referred for intervention or surgery at Christiaan Barnard Memorial Hospital.

**Objectives:** The primary aim of this study is to audit the results of this project. We describe clinical features, diagnosis, intervention or surgery, outcomes, complications and follow-up over 6 years and therefore, the medium term impact of this project.

**Methods:** This is a retrospective case series. Two data sources identified patients. Firstly, records at Windhoek Central Hospital and secondly, hospital admission records at Christiaan Barnard Memorial Hospital. Case notes were reviewed for echocardiogram, cardiac catheterisation, treatment, follow-up and outcome.

**Results:** Of 272 identified as needing surgery or intervention 148 patients, aged between 3 days and 23 years, were referred to Cape Town. Forty-nine had diagnostic and 13 interventional catheterisations. Four patients were inoperable, either through complexity or; irreversible pulmonary hypertension. Cardiac surgery was performed in 112, of which 16 were palliative procedures. Complex cardiac lesions, co-morbidities and late presentation contribute to post-operative morbidity. There were 5 early deaths (mortality rate 4.4%). Twenty-five have been lost to follow-up.

**Conclusion:** There is a heavy burden of congenital heart disease in Namibia, a low middle income country without capacity to operate on babies, small children and complex congenital heart disease. Opinion differs on whether countries with small populations should have independent units for paediatric cardiac surgery or; should refer to larger regional centers. Notwithstanding, this public-private partnership reports a large cohort of patients with comparatively good clinical outcome.

## Myocardial perfusion imaging using single photon emission computed tomography (SPECT) to differentiate ischaemic left ventricular dysfunction from dilated cardiomyopathy

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**Background:** Although dilated cardiomyopathy (DCMO) remains the most common cause for heart failure (HF) in indigent patients at this hospital, the incidence of ischaemic heart disease has risen significantly. Differentiation of DCMO from ischaemic left ventricular dysfunction (ILVD) in patients presenting with HF is crucial and has important therapeutic implications, but may be difficult. We evaluated the utility of Technetium 99m Sestamibi SPECT myocardial perfusion imaging (MPI) using coronary angiography (CA) as the gold standard.

**Methods:** Fifty-two patients with HF with reduced ejection fraction (<40%) who had both MPI and CA were identified retrospectively between January 2005 and December 2012. MPI scans were analysed for wall motion abnormality (WMA), reversible perfusion defects (RPD) and fixed perfusion defects (FPD) and were read in a blinded manner.

**Results:** Thirty-three patients (63%) had ILVD and 19 (37%) had DCMO, as defined by CA. MPI had a sensitivity of 100% and specificity 53% with a positive predictive value of 0.79. The most useful parameter on MIBI was the number of FPD. RPD and WMA were not able to reliably distinguish the 2 groups.

**Conclusion:** The high sensitivity of MPI makes it a useful non-invasive screening test to differentiate DCMO from ILVD in patients presenting with HF and low ejection fraction, but the low specificity means that the diagnosis requires confirmation with CA. The most useful parameter to make this distinction on MPI is the number and size of FPD.

## Right ventricular involvement in cardiac sarcoidosis demonstrated with cardiac magnetic resonance

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**Aims:** Cardiac involvement in sarcoidosis is reported in up to 30% of patients. Left ventricular involvement demonstrated by contrast-enhanced cardiac magnetic resonance has been well validated. We sought to determine the prevalence and distribution of right ventricular late gadolinium enhancement in patients diagnosed with pulmonary sarcoidosis.

**Methods and results:** We prospectively evaluated 86 patients diagnosed with pulmonary sarcoidosis with contrast-enhanced cardiac magnetic resonance for right ventricular involvement. Pulmonary artery pressures were non-invasively evaluated with Doppler echocardiography. Patient characteristics were compared between the groups with and without right ventricular involvement, and right ventricular enhancement was correlated with pulmonary hypertension, ventricular mass, volume and systolic function. Left ventricular late gadolinium enhancement was demonstrated in 28 patients (33%). Twelve patients (14%) had right ventricular late gadolinium enhancement, with sole right ventricular enhancement in only 2 patients. The pattern of right ventricular consisted of free wall enhancement in 6 patients, ventricular insertion point enhancement in 6 patients and enhancement of the right side of the interventricular septum in 8 patients. Pulmonary arterial hypertension correlated with the presence of insertion point enhancement ( $p < 0.001$ ). Right ventricular enhancement correlated with systolic right ventricular dysfunction ( $p < 0.001$ ), right ventricular hypertrophy ( $p = 0.001$ ) and right ventricular dilation ( $p < 0.001$ ).

**Conclusion:** Right ventricular enhancement was present in 14% of patients diagnosed with pulmonary sarcoidosis and 36% of patients with left ventricular enhancement. The presence of right ventricular enhancement correlated with pulmonary arterial hypertension, right ventricular systolic dysfunction, hypertrophy and dilation.

### Obese patients with hypertensive heart disease have faster atrio-ventricular conduction than non-obese controls

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**Introduction:** Atrial fibrillation (AF), the most common cardiac arrhythmia, associates with increased morbidity and mortality. Left atrial remodelling, an important AF-substrate, typically associates with longer PR-intervals. We hypothesised that the increased sympathetic tone seen amongst obese hypertensive patients would shorten the PR-interval and accelerate AV conduction.

**Methods:** As part of a prospective epidemiological study to assess AF-risk, we screened >3 000 patients who visited a private cardiology practice for echocardiography, confirmed hypertensive heart disease (HTHD) over a period of 6 years (2009 - 2015). Patients with AF, permanent pacemakers, significant valvular heart disease, chronic renal impairment and untreated thyroid disease were excluded. Heart rate and the PR-interval were calculated from standard 12-lead resting ECGs. The HTHD-cohort was dichotomised into obese (BMI  $\geq 30 \text{ kg/m}^2$ ) and non-obese (BMI  $< 30 \text{ kg/m}^2$ ) subgroups. Heart rate and PR interval measurements were performed on beta-blocker (BB) naïve patients only.

**Results:** HTHD-prevalence was estimated at 500/3 000 (17%) and 246 of 500 (49%) included patients which were BB-naïve. When compared to non-obese controls (n=89), obese patients (n=157) had faster resting heart rates ( $73.06 \pm 11.85 \text{ bpm}$  vs.  $70.97 \pm 12.86 \text{ bpm}$ ) and higher office blood pressure ( $123.26/75.7 \text{ mmHg}$  vs.  $118.92/73.11 \text{ mmHg}$ ), suggesting increased sympathetic tone. They also had heavier left ventricles ( $254.06 \pm 69.92 \text{ grams}$  vs.  $240.33 \pm 52.25 \text{ grams}$ ) and larger left atria ( $41.92 \pm 4.67 \text{ mm}$  vs.  $39.37 \pm 4.58 \text{ mm}$ ,  $p < 0.0001$ ). Obese patients had shorter PR-intervals than non-obese controls ( $166.19 \pm 27 \text{ ms}$  vs.  $172.09 \pm 28.9 \text{ ms}$ ).

**Conclusion:** Despite having significantly larger left atria and heavier left ventricles, the PR-interval of obese subjects were not significantly longer when compared to non-obese subjects. This association may predispose obese subjects to faster AF episodes, compared to non-obese subjects in whom the PR-interval lengthens as atrial size increases.

### Large cardiac rhabdomyomas successfully treated with everolimus in a neonate

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**Introduction:** Congenital cardiac rhabdomyomas are rare and often benign. When there is no haemodynamic compromise, management is often conservative and the rhabdomyomas tend to regress spontaneously. When there is significant chamber blood flow obstruction, or associated arrhythmias, the management is often surgical. We report on a neonate with a large rhabdomyomas and severe left ventricular outflow tract obstruction, successfully treated medically with everolimus.

**Case Report:** A term baby was delivered by caesarean section. He was antenatally diagnosed to have a cardiac tumour. On examination he weighed 3.1 kg and was noted to have mild respiratory distress requiring administration of oxygen by nasal cannula. He had saturations of 95%, heart rate of 180/min and BP of 45/26 (mean=33 mmHg). He had decreased peripheral perfusion. There was no cardiomegaly. A 2/6 ejection systolic murmur was audible in the second right inter-coastal space, radiating to the neck. There were no signs of overt congestive cardiac failure.

Echocardiography revealed multiple large intra-cardiac masses, predominantly in the left ventricular cavity. One mass obstructed the LV outflow tract,

creating severe aortic valve stenosis with very little flow across the AV. Due to the severe LV outflow tract obstruction and resultant poor perfusion he was started on a prostaglandin infusion. His BP and peripheral perfusion markedly improved. The respiratory distress settled and he could be weaned off oxygen.

Due to the high surgical risk of a debulking procedure in a 3kg neonate through the small aortic valve annulus, a conservative approach was selected to maintain the baby on prostaglandin infusion and a trial of medical treatment with everolimus, an inhibitor of mammalian target of rapamycin (mTOR). The cardiac masses were noted to gradually become smaller and the LVOT obstruction to impressively improve. The prostaglandin infusion could be stopped after 1 week and the everolimus stopped after 8 weeks. The LV masses had markedly reduced. There was no rebound growth of the masses after the everolimus was discontinued.

**Discussion:** The conventional management for symptomatic cardiac rhabdomyomas is surgical. This has inherent risk for morbidity and mortality in a young neonate with large rhabdomyomas. We report successful treatment of large, haemodynamically significant cardiac rhabdomyomas using everolimus, a mTOR inhibitor. It has been reported to be effective against subependymal giant cell astrocytoma (SEGA) in tuberous sclerosis, renal cell carcinoma and neuroendocrine tumours. There have been case reports of the successful use of everolimus in the treatment of inoperable cardiac rhabdomyomas, which inspired its use in our patient.

## Clinical, angiographic and procedural predictors of peri-procedural myocardial infarction during percutaneous coronary intervention in Johannesburg

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**Introduction:** Percutaneous coronary intervention (PCI) has been established as an effective therapy for significant coronary artery disease (CAD). Despite medical and technological advances in coronary intervention, peri-procedural myocardial infarction (PMI) remains the most common complication of PCI. We prospectively investigated the frequency, predictors and clinical relevance of PMI in a tertiary academic teaching center in Johannesburg.

**Methods:** We prospectively enrolled 153 adult patients, undergoing PCI at the Charlotte Maxeke Johannesburg Academic Hospital from 1 February 2014 - 31 October 2014. Peri-procedural creatinine kinase-MB and hs-Troponin I were routinely measured before PCI and at 16 - 24 hours post-procedure. The Third Universal definition of myocardial infarction was used to define a PMI event. Data were prospectively collected and retrospectively analysed by an independent research team.

**Results:** One-hundred-and-fifty-two participants met the inclusion criteria and were analysed for peri-procedural myocardial infarction. Of these participants 70.4% were male. The participant mean age was 58.8 ± 10.9 years old and 42.1% of participants were white. Of the cases enrolled 80.3% presented with an acute coronary syndrome. Sixteen (10.5%) participants fulfilled the criteria for peri-procedural myocardial infarction. Side Branch Pinching with preserved TIMI III flow was noted in 62.5% of PMI cases. Duration of procedure ( $p=0.007$ ), right coronary artery intervention ( $p=0.042$ ) and total stent length ( $p=0.0496$ ) were independently associated with PMI. There was a borderline statistically significant association between patients with peri-procedural myocardial infarction and in-patient mortality ( $p=0.055$ ).

**Conclusions:** Peri-procedural myocardial infarction occurred in 10.5% of the cases undergoing PCI. Side-branch pinching was the most common risk factor. Larger studies are required in our demographic region to further define relevant predictors and outcomes of peri-procedural myocardial infarction.

## Intracoronary thrombolytic in STEMI

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**Introduction:** Patients in South Africa often present late with ST-segment myocardial infarction (STEMI). The culprit artery usually has a high thrombus burden and attempts to reperfuse the vulnerable myocardium have not been optimal with conventional methods such as thrombus aspiration.

**Objectives:** To assess safety and efficacy of an intra-coronary thrombolytic regimen in younger patients presenting late with STEMI.

**Methods:** Over a 2 year period, 8 patients with late (>24 hours) presenting STEMI and a high thrombus burden were treated with an intra-coronary thrombolytic.

**Results:** All 8 patients were male (4 patients were black and 4 were white). The average age was 42 years old (range 25 - 60). Two patients were given Streptokinase prior to repeat intra-coronary thrombolytic (Actilyse). There were no prior contra-indications for thrombolytic use. Risk factors included smoking and HIV (3 patients), hypertension (1 patient), family history (1 patient) and IV drug use (1 patient). No patients were diabetic and none were known to have dyslipidaemia or a previous myocardial infarction.

Coronary angiography was performed via the femoral artery. Four patients presented with anterior STEMI and 4 with inferior STEMI. Actilyse (average 38mg) was the thrombolytic administered with routine anti-coagulation, which consisted of aspirin, clopidogrel and intravenous heparin. GPIIb/IIIa inhibitors were not used.

The thrombus grade prior to lytic averaged 4.6 and post lytic 0.9. Five patients were treated with medical therapy (i.e. no stent was required) and 3 patients had a drug eluting stent inserted. One patient had a reperfusion arrhythmia. There were no bleeding complications and no deaths.

**Conclusion:** Intra-coronary thrombolytic appears to be a safe option with a good clinical outcome in young patients who present late (>24 hours) with STEMI and a high thrombus burden.

### Evaluation of the SUNHEART cardiology outreach programme

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**Introduction:** The demand for advanced cardiac care and specialised interventions is increasing. By decentralising cardiac care and, using a hub-and-spoke model, the SUNHEART Outreach Programme of cardiovascular care aims to improve access to advanced cardiac care in the Western Cape. Tygerberg Hospital is the central hub with the first spoke being Paarl Hospital.

**Objective:** To determine the value of the SUNHEART Outreach Programme to the public health care system.

**Methods:** An audit of patients accessing the Outreach Programme was performed for the period May 2013 - May 2014 and compared to a historical cohort of patients from October 2012 - April 2013. Access to advanced cardiac care was measured by time to initial evaluation, definitive diagnosis or intervention and patient compliance with appointments. The value to the health care system was assessed by doing a cost analysis of transport of patients and health care workers, as well as compliance with appointments. The diagnostic accuracy and adherence to guidelines by the local health care providers was assessed. We documented the spectrum of disease requiring advanced cardiac care to help guide future interventions.

**Results:** Data of 185 patients were included. Sixty-four patients were referred to tertiary care from October 2012 - April 2013 and 121 patients were referred to the outreach facility from May 2013 - May 2014. There was a significant reduction in waiting time with the median days to appointment of the historical cohort being 85 days compared to 18 days in the Outreach Programme cohort ( $p < 0.01$ ). Patient compliance with appointments was significantly superior in the Outreach Programme cohort (90% vs. 56%;  $p < 0.01$ ). Valvular (36.5%) and ischaemic heart disease (35.5%) were the major pathologies. Transport costs per patient treated was significantly reduced in the outreach programme cohort (R118.09 vs. R308.77).

**Conclusion:** Decentralisation of services, in the form of an Outreach Programme with a central hub, improves access to advanced cardiac care by decreasing waiting time, improving compliance with appointments and decreasing travel costs.

### Tri-leaflet mitral valves - when lighting strikes thrice

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A tri-leaflet mitral valve is a novel echocardiographic finding that has only been described in 2 case reports. We report on 3 patients recently found to have tri-leaflet mitral valves in the setting of atrioventricular concordance.

The first patient is a 22-year-old male referred to our congenital heart disease clinic for follow-up of a restrictive VSD. Echocardiography revealed a quadricuspid aortic valve and an unusual appearance of the mitral valve. There were 3, evenly spaced commissures with central coaptation, present as well as 3 papillary muscles – a medial, a small or rudimentary antero-lateral and a posteriorly positioned papillary muscle. Despite these morphological abnormalities, only trace regurgitation was detected. 3D echocardiography allowed for a more detailed morphological and functional assessment.

The second patient is a 49-year-old male evaluated for chronic severe mitral regurgitation. Echocardiography confirmed a dilated left ventricle with severe mitral regurgitation with a tri-leaflet mitral valve, also with 3 (medial, lateral and posterior) papillary muscles present. No other structural abnormalities were present.

The third patient is a 50-year-old male evaluated for chronic severe mitral regurgitation. Initially this was assessed as being due to prolapse, but more careful scrutiny confirmed a dilated left ventricle with severe mitral regurgitation with a tri-leaflet mitral valve with 3 papillary muscles present: a rudimentary basal medial PM, an anterolateral PM and a more apical posterior PM. 3D echocardiography clearly shows the 3 commissures.

The mitral valves of all 3 patients mimic the normal tricuspid valve morphology with 3 leaflets, 3 commissures, 3 papillary muscles (of which one is small/rudimentary) and cords from each papillary muscle (including from the rudimentary papillary muscle) to 2 separate leaflets.

The tri-leaflet mitral valve is a rare congenital abnormality that is increasingly being recognised with advances in imaging techniques.

## Critical review of paediatric and congenital cardiac transplantation at Christiaan Barnard Memorial Hospital since 1995

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**Introduction:** Cardiac transplantation commenced in 1995.

**Methods:** Referrals for transplant evaluation for end-stage cardiac failure in patients <20 years of age, and with congenital defects not suitable for conventional repair regardless of age, were analysed.

A comparison was made between two 10 year periods (A: 1995 - 2004 and B: 2005 - 2014) as well as between the first (C: 1995 - 1999) and last 5 year period (D: 2010 - 2014), of: (a) transplants done, and (b) deaths on the waiting list.

**Results:** Fifty-nine patients were referred for transplant evaluation (A=30; B=29). Of those, 48 (A=24; B=24) were wait-listed.

Transplants were done in 25 patients (A=16/24; B= 10/24), a reduction of 36% comparing the 2 periods. Comparing the first and last 5 year periods (C and D), the reduction was more pronounced with 12/13 transplanted in C, and only 2/10 in D, a reduction from 92.3 to 20%.

SA annual numbers of thoracic transplants were compared to the trend observed in this study. Period A, when compared to B, showed a 34.4% reduction in numbers and for period C to D it decreased by 38%.

Deaths on the waiting list occurred in 22/48 patients (A=7/24; B=15/24), more than doubling from 29 - 62%. Whilst 1 of 13 patients suitable for transplantation died while waiting during the first 5 years (C), the mortality was 8 out of 10 (80%) during the last 5 years (D).

Of those, 17 patients died within 3 months awaiting organs (A=6; B=11) and 5 died after >3 months (A=1; B=4).

**Conclusions:** (1) Numbers of paediatric and congenital cardiac transplants decreased more than comparative SA statistics. (2) Mortality of patients on the waiting list increased markedly over time with most deaths (77.2%) within the first 3 months on the waiting list.

## SHARE-TAVI registry

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**Background:** The SHARE-TAVI registry was set up to gather data on all the TAVI implants in South Africa with the primary aim of improving patient care in the long term. Data capture started in November 2014 and this presentation aims to provide the first feedback of the data. An outline of the capture system will be discussed.

**Results:** A total of 76 patients were entered into the system from 14 different teams, of which 8 are actively capturing data. Data for implants in non-aortic position was not collected and it is important to note that this data does not include all cases screened for TAVI. Of the 76 cases entered, 68 were approved by the Heart team for TAVIs, and 45 received implants. Reasons for not proceeding to implant included funding refusal (2), death (3), patient declined procedure (1) and anatomy not suitable or valve severity moderate (2). Risk profile: average age was 80 years and mean EuroSCORE (Log.) was 19.12, mean EuroSCORE 2 was 7.684 and mean STS score was 7.408. Of the 62% of patients who had frank contra-indications to conventional surgery, other risks included frailty (n=29), porcelain aorta (n=8), patent LIMA graft (n=10) and hostile thorax in 5 patients.

Procedural success was 93.3%, the remainder experienced either device/delivery malfunction (n=1) or residual aortic regurgitation (n=2) and complications are discussed. Access route was TF (50%), TA in 5.4% and not recorded in the remainder. Hospital stay was a mean stay in ICU of 2.28 days, high care 1.23 days and ward stay of 3.09 days. Follow up 30 day data is complete for 42% and 30-day mortality was 4.4% in patients undergoing TAVI.