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TWO CHILDREN PRESENTING WITH CARDIAC HAEMANGIOMAS

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The incidence of primary cardiac tumours varies between 0.00017 and 0.027% of all live births with cardiac hemangiomas comprising only 2.8% of these. The majority are diagnosed in the neonatal period. We report on two neonates with cardiac haemangiomas, one of whom was symptomatic at presentation and the other asymptomatic.

Case 1: A 7-day-old female was admitted with a right upper lobe pneumonia requiring ventilatory support. A few hours after admission she developed a supra-ventricular tachycardia, which responded to intravenous adenosine. Echocardiography and a thoracic CT scan suggested the presence of a large complex pericardial collection. There was no enhancement of the lesion with contrast administration. A pericardial tap yielded a small amount of blood which clotted rapidly. Surgical exploration through a small sub-xiphoid incision revealed an abnormal pericardial surface. A pericardial biopsy showed features of a pericardial capillary haemangioma. On further questioning the mother revealed that the baby had been diagnosed with a pericardial tumour on fetal echo. The patient was treated with alpha-interferon with reduction in size of the mass over a period of 12 months.

Case 2: An 11-day-old female was referred from an outlying hospital with a possible diagnosis of a deep venous thrombosis of the left leg. On admission her left leg was larger than the right with extensive raised areas which had a dark blue discoloration. A small blanching lesion was noted on the left side of the chest suggestive of a capillary haemangioma of the skin. An echocardiogram revealed multiple large intracardiac masses with no apparent haemodynamic compromise. A CT angiogram showed multiple contrast enhancing lesions throughout most organs, including the heart which was consistent with extensive capillary haemangiomas. The diagnosis of a Kasabach-Merrit syndrome was made in association with the presence of a thrombocytopenia on the full blood count.

AN UNUSUAL PRESENTATION OF AN AORTOPULMONARY WINDOW IN A CHILD

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We present a 1-year-old child who was admitted with a cough and increasing shortness of breath for two weeks. She was described as previously well. On admission to Chris Hani Baragwanath Hospital she was diagnosed with a severe bronchopneumonia, but was also noted to be very pale and in cardiac failure. The cardiac failure was thought at the time to be due to the severe anaemia, the cause of which was not apparent.

Initial treatment included administration of oxygen, antibiotics and a blood transfusion. The patient deteriorated needing admission to the intensive care unit for ventilation, where she was found to be pale, and required further transfusions, high pressure ventilator settings and inotropic support. It became apparent that the cause of the anaemia was intermittent pulmonary haemorrhages with bleeding into the endotracheal tube.

Echocardiography showed normal intracardiac anatomy but the aortic arch was poorly visualized and appeared narrowed. A reconstructed CT

angiographic study revealed a large Aortopulmonary (AP) connection and a hypoplastic aortic isthmus. A repeat echocardiographic examination clearly showed the AP connection with predominantly right to left shunting associated with severe pulmonary hypertension and probable Eisenmenger's syndrome. Sildenafil to treat the pulmonary hypertension was commenced, but the patient died very shortly afterwards before a cardiac catheterisation could be performed.

Aortopulmonary connections are rare congenital heart anomalies. A high index of suspicion is required to make the diagnosis. The late presentation in a previously well child and the eventual presentation with pulmonary haemorrhages and anaemia due to the severe pulmonary hypertension makes this case most unusual.

FIVE YEARS' CLINICAL EXPERIENCE WITH THE '3F' STENTLESS AORTIC BIOPROSTHESIS

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Background: The 3F aortic bioprosthesis is a stentless biological heart valve fabricated from three equal leaflets of equine pericardium, assembled in a tubular shape, and implanted in the native aortic root, to replace the patient's diseased aortic leaflets. Clinical trials began October 2001 and being one of the first centres to implant this prosthesis in humans, we would like to present our experiences with this device.

Methods: From January till October 2002, we implanted 47 '3F' aortic bioprostheses. Effective orifice area, mean gradients, ejection fraction and clinical outcomes were evaluated at regular intervals after surgery. Furthermore, coronary blood flow before and after the implantation of the device were assessed via MRI-angiography.

Results: At 5 years, the 3F bioprostheses showed good haemodynamic performance. Mean gradients were 15.9 mmHg, mean effective orifice areas were 1.7 cm² and mean ejection fraction 61.5%. Overall mortality was 17% (N=8). Two patients required reoperation due to paravalvular leaks (4.2%). Two patients suffered a stroke (4.2%). There were no cases of endocarditis or bleeding complications. Coronary blood flow had significantly improved compared to preoperative values and reversal of preoperative systolic flow was abolished.

Conclusion: The clinical performance of the new 3F aortic bioprosthesis at 5 years is comparable to that of regular stentless aortic valves. Its unique design features make it easier and quicker to implant than conventional stentless valves, and excellent restoration of coronary blood flow is achieved.

LEAFLET RECONSTRUCTION OF INCOMPETENT BICUSPID AORTIC VALVES: MIDTERM RESULTS

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Purpose: Reoperation rates after repair of bicuspid aortic valves are higher than for mitral valve reconstruction. Satisfactory results have been reported for patch augmentation for tricuspid aortic valves. We have applied this technique for the repair of bicuspid aortic valves.

Methods: Autologous pericardium is sutured to the free edge of the prolapsing bicuspid leaflet. A large coaptation surface is created and competence of the bicuspid valve is achieved. 35 patients underwent



reconstruction of their bicuspid aortic valves by pericardial patch augmentation. Patients were followed up at regular intervals by echocardiography for 3 years.

Results: There were no perioperative deaths. 1 year postoperatively, one patient died due to endocarditis. 7 patients had aortic regurgitation grade I, and the other 28 patients had non or trivial aortic regurgitation at discharge. At 3 years postoperatively, only 4 patients had aortic regurgitation grade I. There were no cases of progression of regurgitation. Planimetric effective orifice areas ranged above 2 cm². Mean aortic gradients were 8.2 ± 4.8 mmHg and the mean height of coaptation surface was 14.7 ± 2.1 mm.

Conclusions: The pericardial patch augmentation technique increases coaptation surface, and thus provides reliable early and midterm competence of reconstructed bicuspid aortic valves.

TWO-DIMENSIONAL MEASUREMENTS OF CARDIAC SIZE AND FUNCTION IN PREMATURE INFANTS OF THE FREE STATE AND NORTHERN CAPE

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Background: Cardiac size and function are of practical importance in decision making in infants with cardiac problems. Sufficient echocardiographic analysis of South African premature infants is not available.

Objective: To determine normal 2-dimensional data in a group of South African premature infants.

Methods: This was a prospective study conducted in premature infants born in the high-risk unit of the Universitas academic hospital complex. All infants meeting the inclusion criteria had an Echocardiogram (ECHO) done and a Doppler based performance index determined. Measurements were performed according to the standards of the American Society of Echocardiography.

Results: Thirty-four patients were excluded and data of 100 echocardiograms available for analysis. The median age at ECHO was 7 (3-14) days and gestational age 28 (range: 26 - 36) weeks. Forty one percent were small for gestational age infants (SGA). The median weight was 1,36 (range: 0,78 - 2,44) kg. The shortening fraction was 32±5% and the left ventricular end diastolic dimension 13,4±2mm. Doppler derived myocardial performance index was 0,28±0.18 for the left ventricle and 0,18±0.11 for the right ventricle. We also compared the measurements with known international reference values and for most no difference were observed ($p > 0.5$) except for aortic root measurements ($p = 0.008$) in most weight categories.

Conclusion: This study gives an indication of normal values for a group of South African premature infants. In general, values are in agreement with the international reference standards except for aortic root measurements. There exists a need for larger local studies to determine reference values for children of Africa.

PREVALENCE OF OBESITY AND HYPERTENSION IN URBAN AND RURAL COMMUNITIES: A SOUTH-TO-SOUTH HEMISPHERE COMPARISON

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Background: Key risk factors have driven an epidemic of cardiovascular disease (CVD) in *developed* countries and will likely drive a future epidemic

of CVD in *developing* countries undergoing epidemiologic transition. We directly compared differences between the community prevalence of two easily measured risk factors (blood pressure [BP] and body mass index [BMI]) in rural and urban communities in Soweto, South Africa and in Australia.

Methods: We recently screened a total of 1,127 black Africans in urban Soweto (Urban-RSA) for common risk factors for CVD. Using the same methods in Australia as part of a National Blood Pressure Screening Day, we recently screened 10,649 and 3,038 participants in urban (Urban-Aus) and rural (Rural-Aus) communities (predominantly Caucasian) for the same risk factors.

Results: The median (IQR) age of participants in Urban-RSA, Urban-Aus, and Rural-Aus was 45 (IQR 34 to 55) years, 48 (36 to 59) years and 51 (39 to 62) years. The proportion of female participants in Urban-RSA, Urban-Aus, and Rural-Aus was 63%, 55% and 57%, respectively. In Urban-RSA, 23% of men and 51% of women were obese (BMI ≥30 kg/m²) compared to 25% and 26%, and 33% and 33% of men and women respectively living in Urban-Aus and Rural-Aus, respectively. Overall, 30% of men (median BP 130/80 mmHg) and 31% of women (BP 130/81) in Urban-RSA were found to be hypertensive (systolic BP ≥140 mmHg and/or diastolic BP ≥90 mmHg) compared to 41% of men (134/81 mmHg) and 25% of women (125/77 mmHg) in Urban-Aus and 49% of men (137/82 mmHg) and 34% of women (130/79 mmHg) in Rural-Aus.

Conclusions: Based on these two easily measured risk factors, there appears to be a high level of risk in all three target communities with some potentially important differences. A formal age and sex-specific comparison of these data is planned to further delineate their public health implications.

SURGERY FOR COMPLICATIONS ASSOCIATED WITH AMPLATZER DEVICE INSERTION INTO THE PATENT DUCTUS ARTERIOSUS (PDA)

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Surgical closure of the PDA has a long tradition associated with very low risk. The treatment has changed considerably in recent years. Nowadays many PDAs are closed using transcatheter techniques. Closure of the PDA has become another indication for the Amplatzer device. Severe complications associated with these techniques and devices however mandate surgical backup. The aim of this presentation is to discuss the principles of such surgery.

We report three cases of Amplatzer device complications. In the first case (8 years old) the device protruded into the main pulmonary artery. It was retrieved through a left thoracotomy on cardiopulmonary bypass (CPB) using moderate hypothermia and an aortic incision. In the second case (15 months old) the Amplatzer device protruded into the aorta. The device was retrieved in the same manner as in the first case. In the third case (2 years old) there was a residual PDA and protrusion of the device into the main pulmonary artery. Three days after insertion the device was retrieved by a transcatheter procedure in the cath lab. The right ventricle was perforated and the tricuspid valve damaged. The patient developed tamponade and arrested. An emergency median sternotomy was done to relieve the tamponade. CPB was instituted. The tricuspid valve was repaired and the right ventricle perforations were closed. In all three cases the PDA was surgically closed. The outcome in the first 2 cases was favourable, but the 3rd patient suffered some neurological damage due to the cardiac arrest.

In conclusion, Amplatzer device closure of the PDA is widely accepted but complications may be severe and may require careful surgical intervention.



ENDOVASCULAR THERAPY OF CHRONIC MESENTERIC ISCHAEMIA

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Background: Atherosclerotic disease resulting in progressive stenosis or occlusion of one or more mesenteric arteries is considered the most common cause for chronic bowel ischaemia (CMI). This condition is associated with gastrointestinal symptoms such as diarrhea, postprandial abdominal pain and significant weight loss. Historically, the treatment for CMI has been surgical revascularisation; however, surgery carries a significant procedural complication rate and mortality.

Objective: To evaluate the safety and assess role of endovascular therapy in patients (pts) with mesenteric vascular occlusive disease.

Patients and methods: Between Sept 2002 and Jan 2006, we treated 32 pts (45% male; mean age 60±12 years). Thirty patients had atheromatous stenoses and two had Takayasu arteritis. 12.5% of the lesions were chronic occlusions. We evaluated perioperative morbidity and mortality, restenosis and recurrent symptoms.

Results: Technical success was achieved in 31 pts (96.8%). The brachial access was used in 83.3% and the femoral approach in 16.6% of patients. During a follow-up of 1-83 months (mean 29 months), just one patient had recurrent pain due to restenosis of stent, that was treated with new endovascular intervention.

Conclusions: Percutaneous endovascular techniques for CMI are safe and accurate. The inherent lower procedural morbidity and mortality makes the endovascular approach the preferred revascularization treatment for these patients with chronic mesenteric ischaemia.

CLINICAL FEASIBILITY AND ANGIOGRAPHIC FOLLOW-UP OF ENDOVASCULAR STENT-GRAFT FOR TYPE B DISSECTION (STANTFORD): EXPERIENCE OF A SINGLE CENTRE IN BRAZIL

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Background: Surgery of the aorta is technically demanding and may be followed by perioperative mortality and morbidity. Endovascular repair with stent-graft placement (EVAR) for type B dissection has shown encouraging clinical results.

Objective: To show by our experience, the outcomes of patients (pts) who underwent EVAR.

Methods: A retrospective review at our institution between 2003 and December 2006 identified 85 patients with type B dissection. Patient characteristics include mean age 62±12 years, males 55.55%, diabetes 23%, hypertension 90%, multi-vessel disease 56%, renal dysfunction 14% and chronic lung disease 20%. All patients underwent computed tomography before the procedure, after EVAR and at follow-up (30 days, six and twelve months). 81 pts (95.2%) were treated by conventional stent-grafts. The procedure was done with spinal anaesthesia and intravenous sedation without mechanical ventilation.

Results: Angiographic success, defined as immediate obliteration of target lesion without major endoleak, was obtained in 98.7%. There was 1 death (laceration of iliac artery). Complications included 2 subclavian artery occlusions (asymptomatic). The angiographic follow-up was done in 91%, and there was 1 patient with endoleak, corrected with endovascular stent-graft repair.

Conclusions: Endovascular stent-graft should be considered as a safe and effective treatment for type B dissection particularly in pts with high risk of surgical mortality.

DIAGNOSING PERICARDIAL EFFUSIONS

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Tuberculous pericarditis is a major health problem in developing countries like South Africa. If untreated, the mortality of tuberculous pericarditis is greater than 80%. Definitive diagnosis of tuberculous pericarditis requires isolation of the tubercle bacillus from pericardial fluid, but isolating the organism is often difficult. A diagnostic index was proposed retrospectively to improve diagnostic efficiency for tuberculous pericarditis using available tests.

Objective: To validate the proposed diagnostic index in patients presenting with large pericardial effusions to Tygerberg Hospital

Design: All large pericardial effusions have been included since 2001 as part of an ongoing study at Tygerberg Hospital. ADA, as well as routine diagnostic tests (including microscopy, culture, haematology and biochemistry), was determined on all patients. The diagnostic index consists of 5 elements, 3 being clinically orientated (fever, night sweats and weight loss) and 2 being simple blood tests looking at serum white cell count and serum globulin levels.

Results: The diagnostic index yielded a sensitivity, specificity and positive and negative predictive value of 92%, 84%, 92% and 84%, respectively. The diagnostic index proved highly effective in HIV positive patients yielding 100% sensitivity with 50% specificity. The diagnostic efficiency of the scoring index (90%) was significantly better than many other currently used diagnostic tools.

Conclusion: In a peripheral setting where specialized diagnostic facilities are unavailable the use of this non-invasive and cheap diagnostic scoring index can be used to facilitate the diagnosis of tuberculous pericarditis.

CT ANGIOGRAPHY IN DIAGNOSIS OF CORONARY ANEURYSMS IN KAWASAKI SYNDROME

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A 3 year old boy with atypical Kawasaki syndrome is presented. Giant coronary artery aneurysms were noted on echocardiography. Over time the right coronary artery size normalized whereas the left anterior descending branch of the left coronary artery remained aneurysmal. A 64-slice CT coronary angiogram was performed to outline the coronary arteries and to demonstrate the aneurysmal dilatations. The report emphasizes the use of this procedure in defining coronary artery abnormalities in children specifically in patients with Kawasaki syndrome.

THE TRANS-APICAL APPROACH FOR AORTIC VALVE IMPLANTATION

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Background: Percutaneous aortic valve implantation has been performed clinically in high risk patients with severe aortic stenosis. Retrograde valve delivery has been problematic due to concomitant peripheral vascular disease as well as the size of the delivery system. We report on a minimally invasive approach through the left ventricular apex that allows accurate antegrade placement of a catheter deliverable aortic valve.

Methods: Transapical aortic valve implantation was performed on 22 consecutive patients (83±6.4 years), using 23mm and 26mm pericardial valve (Edwards Lifesciences, Irvine, CA, USA) mounted on a stainless steel stent.



A limited anterolateral incision, in 5th intercostal space, was used to access the apex of the heart. The valve was crimped and placed into a 24 French sheath, and introduced into the left ventricle through purse string sutures. Fluoroscopy and transesophageal echo (TEE) was used to guide the catheter across the native valve and direct deployment of the stent at the level of the annulus. During deployment, the heart was unloaded with extracorporeal support (ECS), or with rapid ventricular pacing.

The average logistic Euroscore predicted risk for mortality was 26±9%.

Results: All valves were successfully deployed at the target. There were 2 in hospital deaths, 1 due to perforation of the right ventricle and 1 due to dissection of the aortic root. There were another 3 non-procedure related deaths. There were 2 cases of conversion to open surgery. In one patient valve embolisation into the aortic arch occurred, and in another patient, type A-dissection developed after balloon dilatation. In two patients, the left main stem was partially obstructed by the native valve and required stent angioplasty. On echocardiography, all valves showed good hemodynamic function with only minor paravalvular leakages.

Conclusion: Initial results of the transapical approach are encouraging.

HYBRID APPROACHES TO COMPLEX CONGENITAL CARDIAC SURGERY

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Objectives: A hybrid operation is a joint procedure involving the interventional cardiologist and the cardiac surgeon concomitantly to optimise surgical management. The aim of our study was to demonstrate the conceptual development and the feasibility of a hybrid approach to complex congenital cardiac surgery in Red Cross Children's Hospital.

Methods: Descriptive study for concomitant intervention by the cardiologist and the cardiac surgeon. 12 patients with complex congenital heart defects requiring high risk operative interventions were included in the study. The procedures were: (1) intraoperative stenting of a pulmonary artery stenosis with concomitant additional surgical procedures (n=2). (2) Balloon dilatation of Blalock-Taussig shunts to establish flow (n=1). (3) Muscular VSD (n=3). Neonatal Coarctations (n=5). Congenital Mitral Stenosis (n=1).

Results: 11 patients had successful hybrid procedures. Two patients had failed hybrid procedures due to technical problems. There were no important complications related to the temporal proximity of the interventional procedure and cardiac surgery, the latter being significantly facilitated by the former.

Conclusions: Hybrid procedures utilising both cardiological and cardio-surgical expertise can safely be used in patients with complicated congenital heart disease. These procedures may be complementary in patients with complex lesions by providing a better result in combination than either alone can offer.

GENETIC CONTRIBUTION TO RHEUMATIC FEVER: A SYSTEMATIC REVIEW AND META-ANALYSIS OF TWIN STUDIES

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Background: Rheumatic heart disease (RHD) results from repeated attacks of rheumatic fever (RF), which, in turn, is closely related to a preceding untreated pharyngitis caused by group A streptococcus. Together, RF and RHD are the leading cause of acquired heart disease in children in the world affecting 15.6 million people worldwide. A genetic component for the development of RF/RHD has been postulated, but not formally quantified in a genetic epidemiology study.

Objective: This systematic review aims to quantify the degree of genetic contribution to RF/RHD from twin studies, and to determine the extent of heterogeneity in the strength of associations between studies.

Methods: We searched all reports from original papers published in a number of databases from their earliest manuscripts to August 2007. Predefined criteria were used to identify studies examining the concordance for RF/RHD in monozygotic (MZ) versus dizygotic (DZ) twins. Two observers (RS, JV) independently evaluated the search outputs and recorded relevant data. A third reviewer (ME) verified the data extraction. The degree of genetic effect was estimated using probandwise concordance rate and odds ratio (OR). Heterogeneity between studies was calculated with the χ^2 method.

Results: Two studies on RF with a total of 71 twin pairs from three populations were included in the analysis. Probandwise concordance rates ranged from 32% to 100%, and from 5% to 67% for MZ and DZ twins in individual studies, respectively. The pooled probandwise concordance rates were 0.52 for MZ and 0.18 for DZ pairs. Random-effects meta-analysis confirmed that MZ twins were more likely to be concordant for RF than DZ twins (O.R., 8.25; 95% confidence interval 1.52; 44.75, $p=0.01$). No statistical heterogeneity was present between studies ($p=0.95$).

Conclusion: This first meta-analysis of twin studies suggests a strong genetic contribution to the development of RF. Large-scale molecular genetic studies are warranted to elucidate the genetic architecture of RF.

NO EVIDENCE FOR AN ASSOCIATION BETWEEN THE MITOCHONDRIAL VARIANT T16189C AND DILATED CARDIOMYOPATHY: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Mitochondrial (mt) point mutations are associated with maternally inherited and sporadic cases of hypertrophic and dilated cardiomyopathy (DCM).

Objective: This systematic review examined the association of mt T16189C variant with DCM, a disease characterised by dilatation and impaired contraction of the heart.

Methods: The authors searched all reports from original papers published in a number of databases up to August 2007, with no restriction on language. Predefined criteria were used to identify case-control studies examining the variant's association with DCM. Two observers (ME, GS) working independently evaluated the search outputs and two studies, supplemented by one unpublished report, met the inclusion criteria. Reviews and studies with an outcome other than DCM, other polymorphisms, or those not examining gene-association, were excluded. From each study, year of publication, origin and demographics of participants, matching procedures, diagnostic criteria, and information on variants were recorded. In the study which included subjects from differing ethnic backgrounds/geographical locations, data were extracted separately. The third reviewer served as arbitrator where necessary.

Results: Four populations conducted in European and African groups, contributed a total of 1002 subjects for analysis. Control groups comprised mainly hospital inpatients. Random-effects meta-analysis revealed a per-variant odds ratio of 1.68 (confidence interval: 0.89; 3.14 $p=0.107$) for T16189C. There was no statistically significant between-study heterogeneity for any of the comparison groups (χ^2 -based Q statistic, $p>0.1$).

Conclusion: Thus, in this first systematic review on the effect of mt T16189C variant on DCM risk, the authors found no evidence for an association between the variant and DCM in case-control studies. This variant is not likely to be a significant risk factor for DCM.



UNUSUAL CONGENITAL CORONARY ARTERY ABNORMALITY

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Congenital anomalies of the coronary vessels are rare. They may exist as:

- Anomalies of aortic origin
- Origin of one/more coronary arteries from the pulmonary arteries
- Fistulae from the coronary arteries to cardiac chambers or surrounding vessels ~ Coronary Arteriovenous and Coronary-Cameral Fistulae (C-CF)
- Idiopathic diseases of the coronary arteries
- Anomalies of the coronary sinus.

C-CF constitute 0.2-0.4% of all congenital cardiac defects. C-CF observed in 0.08-0.18% of routine coronary angiograms in adults. In primitive myocardium, wide intertrabecular spaces shrink to form the sinusoids – persistence of some of these spaces could lead to C-CF. 55-65% of fistulae arise from the RCA. About 90% of fistulae drain into the lesser circulation – RV most common followed by RA and PA. The fistulous coronary may be small or dilated and tortuous and enters the affected chamber by a single or several openings. Saccular aneurysmal dilation does occur. C-CF usually exists as isolated defects but other congenital cardiac lesions may coexist.

The large epicardial coronary arteries run on the surface of the heart, with only the terminal branches penetrating the muscle. In 50% of people, part of the epicardial vessel dips beneath the epicardial muscle so that there is a muscle bridge over the large artery - symptoms of myocardial ischaemia or infarction.

A neonate with clinical features of diastolic run-off and unusual course of the LCA will be presented. Data will include echo images, angiographic demonstration of the LCA as well as PM images of this unusual coronary artery anomaly.

This case was presented at the CARDIOLOGY IN THE YOUNG SYMPOSIUM 2007 in London (May 2007).

EARLY EXPERIENCE WITH INTRAVASCULAR STENTING FOR TREATMENT OF COARCTATION OF THE AORTA IN CHILDREN AND ADULTS

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Background: We report our initial experience with the use of balloon expandable bare and covered stents for treatment of native and recoarctation of the aorta in both children and adults.

Methods and results: Between Oct 2003 and Sept 2007, seven patients underwent percutaneous stent implantation for coarctation of the aorta. Five patients underwent a stent implantation for a native coarctation, one patient had a stent for recoarctation and one patient had a stent of an abdominal coarctation complicating a Takayasu's arteritis.

Median age of the patients was 17 years (range 6 years to 46 years). There was significant improvement ($p < 0.01$) in pre versus post stent mean coarctation dimensions (6.2 mm versus 16.6 mm), systolic gradient (mean value 39 mmHg versus 4.1 mmHg) and a mean ratio of the coarctation segment to the descending aorta (0.34 versus 0.89). Angiographic control showed good relief of the stenoses. The procedure was successful in all cases with no early complications. During a median follow up of two months (range 1 month to 48 months), hypertension has been resolved in all except one adult who is controlled with antihypertensive medication.

Conclusions: Stent implantation for native coarctation and recoarctation of the aorta in children and adults provides good immediate results and is a relatively safe and effective alternative approach for relieving the obstruction with a low complication rate.

QT PROLONGATION ≥ 460 MS PRIOR TO CORONARY ANGIOGRAPHY PREDICTS POORER LV SYSTOLIC FUNCTION AND INCREASED 6-MONTH MORTALITY

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Background: QT prolongation is associated with increased risk of sudden cardiac death in the general population. The predictive value of QT prolongation prior to diagnostic coronary angiography is currently unknown.

Methods: Utilizing the SAHA-Tygerberg Hospital cathlab registry, we prospectively recorded heart rate-corrected QT (QTc) intervals in 186 patients on the morning prior to diagnostic coronary angiography (CA). Patients were stratified into those with normal QTc-intervals (NQT) (QTc<440ms) and those with prolonged QTc intervals (LQT) (QTc \geq 460ms) and followed for 6 months. Patients with atrial fibrillation, bundle branch block or QTc between 440-459ms were excluded.

Results: Of the 125 patients that were included for analysis, patients in the LQT-subgroup (n=39) were younger than those in the NQT-subgroup (n=86) (58 \pm 13.7 years vs. 65 \pm 11.7 years, p =NS). Left ventriculography was performed in 46 patients (37% of the group). Calculated ejection fractions of the two groups differed significantly (NQT vs. LQT: 63 \pm 6% vs. 47 \pm 9%, p <0.01). Obstructive CAD was more prevalent in the NQT than in LQT group and normal angiograms were more prevalent in the LQT than in the NQT group. Patients with QTc>500ms were more likely to receive medical therapy only compared to those with QTc<440ms (80% vs. 33%; p <0.01). Patients with QTc<440ms were referred twice more commonly for CABG than patients with QTc>500ms (40% vs.20%). Ten deaths occurred in 82 patients (66% of the group) during the six month follow-up (LQT=24 and NQT=58). Six month mortality was higher in LQT compared to NQT (25% vs. 6.9%, p =0.06). All the deaths in the LQT group were sudden and occurred within 70 days post-coronary angiogram.

Conclusion: QT prolongation ≥ 460 ms immediately prior to coronary angiography predicts poorer LV systolic function and increased six month mortality.

A SIMPLIFIED INDIVIDUALISED APPROACH TO REPAIR OF COMPLETE AV CANAL

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Traditional 1- and 2-patch techniques for repair of complete AV canal defects (AVCDs) can be unnecessarily complex, and the repair can, in some patients, be considerably simplified by direct suture of either the VSD (direct suture of the common atrioventricular valve leaflets to the crest of the ventricular septum) or the ASD, or occasionally of both components.

A retrospective review is presented of 69 consecutive patients over six years who had repair of complete AVCD with an individualised technique determined by the detailed anatomy of the lesion. In 21 patients the VSD was closed by direct suture, in 8 patients the ASD was closed by direct suture, and in three patients no patch material at all was used, both ASD and



VSD components being closed directly. An additional 9 patients had a partial direct-suture of the VSD. The remaining 28 patients had a standard 2-patch repair.

Of the patients in whom the VSD was closed by direct suture, two required a takedown of the repair and a standard 2-patch repair instead, after TOE showed an inadequate repair after bypass was weaned. One patient required repeat surgery for a residual VSD. One patient had prolonged ICU stay with poor cardiac output. There were no deaths and no other significant morbidity.

Of the patients with standard 2-patch repair, two required redo surgery for a residual defect. One required repeat surgery for endocarditis on the patch, and subsequently died.

In conclusion, this individualised simplified approach to AVCD repair is safe and effective. However, longer follow-up is required to establish the long-term durability of the repairs and to more precisely define the applicability.

SURGERY IN THE FACE OF HIV INFECTION

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There have been many reasons on the part of clinicians treating patients infected with human immunodeficiency virus (HIV) infection, to deny major surgery to these patients, but most of these have proven groundless. The introduction of highly active antiretroviral therapy (HAART) in the mid-1990s dramatically impacted the natural history of human immunodeficiency virus (HIV) infection. As a result more and more of these patients are being referred for cardiac surgery. Based on our experience with a small number of patients, on reviews of the published data, and taking cognizance of the ethics of denying surgery and the problem of scarce resources, as well as the potential risks to health personnel, a provisional protocol was developed for an approach to HIV-infected children who need cardiac surgery.

Over the three years that HAART has been available to us 17 HIV-infected children were subjected to a range of cardiothoracic surgical procedures. 12 of these patients had AIDS, while the remainder were immunocompetent at the time of surgery. There was one early (<30 days) and two late (>30 days) deaths in the follow-up period, all due to ongoing sepsis in patients with AIDS. In no case was the immunocompetence of the patient adversely affected by the procedure as measured by pre- and postoperative CD4 counts.

We recommend that:

- HIV-infected children should have the same surgery for the same indications as non-infected children, provided that:
- HAART is given for the same indications as children not requiring surgery, and
- Surgery is delayed when possible for the expected reduction of viral load and recovery of CD4 count that will be seen on HAART over time. This will also minimise risks from needle stick injuries.

CASE PRESENTATION: ACUTE MITRAL VALVE REGURGITATION DUE TO PAPILLARY MUSCLE RUPTURE. AN UNUSUAL PRESENTATION OF ANOMALOUS ORIGIN OF LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA)

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A 29-year-old previously healthy man presented with a history of increasing dyspnoea of one month duration. He was in severe biventricular failure

clinically. A pansystolic murmur consistent with mitral regurgitation was heard on cardiac auscultation.

On echocardiography there was severe prolapse of the anterior mitral valve leaflet due to papillary muscle rupture with severe mitral regurgitation. Mild anterior wall hypokinesis was also observed. An abnormal artery was noted on the parasternal short axis views and a diagnosis of anterior papillary muscle rupture due to myocardial ischaemia caused by an anomalous left coronary artery arising from a pulmonary artery (ALCAPA) was made.

This diagnosis was confirmed with coronary angiography and the patient underwent mitral valve repair and coronary vessel grafting to the LAD.

The report discusses the unusual presentation of ALCAPA in an adult, illustrating the value of transthoracic echocardiography in detecting the anomalous artery, and shows images obtained by TEE.

CLINICAL PRESENTATION OF HOLT ORAM SYNDROME

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Holt-Oram syndrome is an autosomal dominant condition characterized by preaxial radial ray abnormalities and congenital heart defects. This syndrome has been linked to mutations in the TBX5 transcription factor gene at chromosome 12q24.1. The most common cardiac lesions described are atrial septal defects (ASD), however ventricular septal defects (VSD), conduction abnormalities and more complex lesions including isomerism have been described. The limb abnormalities range from sloping shoulders to phocomelia. Thumb abnormalities are the most common and include absent, displaced, duplicated or triphalangeal thumbs. The syndrome must be differentiated from other Hand-Heart syndromes that differ both in clinical presentation and gene defect.

We present our findings in three patients with a clinical diagnosis of Holt Oram syndrome: Patient 1 with large perimembranous VSD, patent ductus arteriosus (PDA) and duplication of the left thumb, patient 2 with complete atrioventricular septal defect and bilateral triphalangeal thumbs and patient 3 with secundum ASD, right hemianomalous pulmonary venous drainage, large perimembranous VSD, PDA, a left SVC and second degree heart block with a right triphalangeal thumb.

THE EFFECT OF COMBINED CLOPIDOGREL AND ASPIRIN THERAPY FOLLOWING OFF PUMP CORONARY SURGERY

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Objective: The purpose of this study is to evaluate the effect of combined regimen of clopidogrel and aspirin on graft patency following off-pump CABG, and its association with a major adverse cardiac events and other undesirable effects. We therefore designed a prospective randomized study to evaluate the safety and efficacy of a combined clopidogrel and aspirin regimen started perioperatively on OPCAB surgery. This paper will present results from the first 20 patients (pilot study).

Methods: 20 patients who underwent standard OPCAB through median sternotomy were randomized immediately following surgery in two groups. Patients in group A (10) received 100 mg of aspirin starting preoperatively and continuing postoperatively for 3 months. Patients in group B received aspirin as patients in group A, and in addition 75 mg of clopidogrel for 3 months. Multiple perioperative parameters were statistically compared. Angiography was repeated 3 months after surgery to determine patency and quality of grafts. Fitzgibbons method of graft patency classification was utilized.

Results: There was no difference with respect to preoperative risk factors between patient groups. There was no significant difference in



average number of distal anastomosis ($p=0.572$) operation time ($p=0.686$), postoperative bleeding ($p=0.256$), ventilation time ($p=0.635$) and ICU stay ($p=0.065$). Patients in group B had shorter hospitalisation ($p=0.024$). There were no postoperative complications in both groups. 8 of 27 grafts in group A and 2 of 29 grafts in group B ($p=0,061$) were occluded at the time of control angiography.

Conclusion: Early administration of combined regimen of clopidogrel and aspirin following off-pump CABG is not associated with increased postoperative bleeding and other major complications. Despite the small number of patients in this study and small number of examined grafts, results suggest that clopidogrel increased graft patency following off pump CABG.

MECHANICAL CARDIAC SUPPORT IN END STAGE CARDIAC FAILURE: AN 8 YEAR CAPE TOWN EXPERIENCE

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Background: Although cardiac transplantation remains the gold standard in the management of end-stage cardiac failure, this procedure is limited by patient selection and organ shortage. In addition to the heart transplant program, an artificial heart program was started at the Christiaan Barnard Memorial Hospital 8 years ago.

Methods and patients: During this period 27 patients were supported using a mechanical cardiac device. Our institution uses the Berlin heart system for long-term mechanical support and eventual bridge to transplantation. For shorter term support (1-3 weeks) a centrifugal left ventricular assist device (LVAD) is used. Of the 26 patients, 23 required short-term LVAD support. The indication in this group was post cardiomy failure ($n=20$) and acute donor heart failure ($n=3$). The remaining 4 patients were supported on the Berlin heart. The indication in this latter group was cardiomyopathy with end-stage failure.

Results: Of the 4 patients supported by the Berlin Heart, 3 were successfully transplanted at 2 weeks to 7 weeks and were discharged home. Of the short-term centrifugal support ($n=23$), 20 patients were weaned of the LVAD (86.9%) and 17 were discharged home (73.9%). All 3 patients with acute donor heart failure make a full recovery on the LVAD and could be discharged home (100%).

Conclusion: Early results of the artificial heart program at the Christiaan Barnard Memorial Hospital compare favourably with reported international outcomes. Mechanical cardiac support is a limited but effective option in the management of post cardiomy failure as well as end-stage cardiac failure. Due to the progress, the program is to be extended to axial flow technology.

THE CHALLENGES OF CARDIAC SURGERY IN ZAMBIA

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1. History of CTS in Zambia: The first open heart surgery was performed in 2001 at the University Teaching Hospital in Lusaka. Before this period no significant cardiac surgery was being performed. Since the first heart operation only about 50 cardiac surgical procedures have been carried out to date. Due to the high costs of establishing a CTS center and the lack of human resource to support this service the development of cardiac surgery in Zambia has been very slow. Currently there is no full time cardiothoracic surgeon in Zambia. The procedures that have been performed to date have been done by a visiting surgeon from the Uzbekistan. Procedures that have been performed include: mitral valve replacements, valvotomies, pericardiomy and closure of PDAs.

2. Current situation of CTS in Zambia

a. Human resources:

- i. Cardiothoracic surgeon: None full time
- ii Assistant surgeon: General surgeon/vascular surgeon
- iii. Cardiac anaesthetist: One
- iv. Cardiac perfusionist: One
- v. Theatre nurses: Two
- vi. Other theatre assistants: Four

b. Infrastructure:

- i. One cardiac theatre at the UTH

c. Surgical supplies: Through local and international suppliers.

d. Intensive/critical care unit: One intensive care unit that serves all critically ill patients at UTH

e. Interventional & non-interventional cardiology support services: Currently no interventional cardiology services exist in Zambia. Non-interventional cardiology support services are present.

f. Maintenance costs: Each open heart operation may cost in excess of \$15 000 to perform locally.

g. Paediatric patient case load: There are currently over 300 patients waiting for cardiac surgery. The cases range from rheumatic heart disease to various forms of congenital cardiac malformations. A small percentage of these patients are supported by NGOs and the Government in order to travel to places like India and South Africa for operations which cost in excess of \$25 000.

h. Adult patient case load: Most cases comprise RHD.

3. Future of CTS in Zambia: Rests on setting up a development plan and ensuring sustainability as a key element of success.

4. Development plan: a development plan should focus on a multifaceted approach to establishing and maintaining the service.

LONG TERM FOLLOW UP OF THE MITROFLOW AORTIC BIOPROSTHESIS: ABOUT 299 PATIENTS

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Background and aim of the study: In older patients and those unable to tolerate oral anticoagulants, bioprosthesis provide a good alternative to mechanical prosthesis. The main disadvantage of bioprosthesis is a significant structural deterioration 15 to 20 years after implantation, which can lead to reoperation. Therefore, the international guidelines recommend using bioprosthesis in patient older than 70 years old.

The aim of the study was to evaluate long term follow-up after aortic valve replacement with Mitroflow bioprosthesis in 299 patients.

Methods: Between March 1988 and March 2006, a total of 299 Mitroflow valves were implanted in 299 patients. Their mean age was 75.5 yrs (40 to 87) and 160 were male. An isolated aortic valve replacement was performed in 168 patients and associated procedures were CABG (35%), mitral valve replacements (6.3%), mitral valvuloplasty (3.3%), or a replacement of the ascending aorta (3.7%). Valve sizes were 19 (34), 21 (117), 23 (100), 25 (43).

Results: Mean and maximum follow up was 3.2 and 17.5 years. Total follow up was 910 patient-years. Operative mortality was 7.7%. Actuarial survival at 10 years was 29.9%. Survival rate of valve-related death was 84% at 10 years. Fives cases of structural valve deterioration were found. Six patients were reoperated for valve-related problem (1 endocarditis, 1 perivalvular leak with haemolysis and 4 structural deterioration after respectively 7, 8.8, 9 and 9 yrs).



The retrospective study showed that we had a mismatch for only one patient among 299.

Conclusions: The design of the Mitroflow aortic valve bioprosthesis allowed an aortic valve replacement with no aortic enlargement and no mismatch. The long term experience shows the long term results are good and that the structural deterioration rate at 10 years is low.

EVERY ECG TELLS A STORY: ECG CHARACTERISTICS OF HEART OF SOWETO STUDY PATIENTS WITH HEART FAILURE

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Background: The 'Heart of Soweto' Study is currently examining the pattern of cardiovascular disease (CVD) in Soweto, South Africa, which comprises of townships with an estimated population of 1 to 1.5 million. The electrocardiogram (ECG) has the potential to offer a cheap and practical way to identify adults with undetected heart failure (HF).

Methods: As part of a large clinical registry of CVD patients presenting to the Baragwanath Hospital in Soweto, a 12 lead ECG were performed on 91% 'de novo' cases in 2006. These were systematically analysed using detailed Minnesota coding and correlated with a clinical presentation of HF.

Results: In 2006, a total of 756 de novo patients with HF (57% women, 88% Black African and mean 55 ± 16 years) had a 12 lead ECG. Overall, a major ECG abnormality was detected in 91% of cases; consisting of S-T segment changes, T wave changes, conduction abnormalities, bundle branch blocks and arrhythmias. A further 97% of ECGs had minor abnormalities such as axis deviation, high amplitude R waves and minor T wave changes. Left ventricular hypertrophy was evident in 15% (22% in those with a dilated cardiomyopathy) and a bundle branch block in 7.4% of ECGs (11% in those with an ischaemic cardiomyopathy). In respect to cardiac rhythm, 63% of patients were in sinus rhythm, 26% in sinus tachycardia (26%), 8% in atrial fibrillation/flutter, 2% in sinus bradycardia and 1% in complete heart block. Further analyses of ECG and echocardiographic data are planned.

Conclusion: Detailed examination of the presenting 12-lead ECGs of a large group of previously undiagnosed patients with HF in the Heart of Soweto revealed many identifiable abnormalities. In addition to reconfirming the clinical utility of this cheap and practical diagnostic tool, these data highlight its potential role in screening for heart disease in low income communities.

ECG CHARACTERISTICS OF PATIENTS IN THE HEART OF SOWETO STUDY

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Preventative Cardiology, Baker Heart Research Institute, Melbourne

Background: The 'Heart of Soweto' Study is currently examining the pattern and subsequent burden of cardiovascular disease (CVD) in Soweto, South Africa. The electrocardiogram (ECG) has the potential to offer a cheap and practical way to screen adults who have undetected heart disease.

Methods: As part of a large clinical registry of CVD patients presenting to the Baragwanath Hospital in Soweto, a 12 lead ECG were performed on 91% 'de novo' cases in 2006. These were systematically analysed using detailed Minnesota coding and correlated with clinical diagnoses.

Results: In 2006, a total of 1593 newly diagnosed patients were identified with the majority being black African (85%) and women (60%). Of these, a total of 1437 (90%) had a 12 lead ECG of which 643 had a diagnosis of heart failure (45%).

The remaining 55% (n=794) were categorised as valvular disease (n=248, 17%), hypertension (n=276, 19%), coronary artery disease (n=142, 10%)

and other (n=128, 9%). Specifically relating to those with diagnoses other than heart failure; ischaemic changes were evident in 21% of ECGs and T wave changes seen in 57% of cases; 82% of the group diagnosed with CAD demonstrating T wave changes. In relation to underlying cardiac rhythm, the majority of patients were in sinus rhythm (n=553, 69%) with 17% having in sinus tachycardia (n=142) and a further 2.5% with a heart rate less than 50 bpm. A total of 60 patients had ECGs with atrial fibrillation (n=60, 7.5%). Examining axis deviation revealed that the majority had a normal axis (n=642, 81%) with an equal proportion demonstrating left and right axis deviation.

Conclusion: In this large African cohort with established CVD, significant changes on the 12-lead ECG were detected in the majority of patients. As such, it represents a potentially useful community-based screening tool to detect un-identified pathological abnormalities (e.g. myocardial ischaemia and cardiac arrhythmias).

EXCESS ADIPOSITY IS ASSOCIATED WITH LEFT VENTRICULAR DIASTOLIC DYSFUNCTION BEYOND BLOOD PRESSURE OR DIABETES MELLITUS IN A POPULATION SAMPLE WITH A HIGH PREVALENCE OF OBESITY

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Introduction: Left ventricular (LV) diastolic dysfunction, which may be caused by obesity, is an independent predictor of cardiovascular outcomes. The contribution of excess adiposity to LV diastolic dysfunction independent of blood pressure (BP) and diabetes mellitus in a population with a high prevalence of obesity has not been determined.

Methods: We studied 384 randomly selected subjects of African descent living in Soweto (minimum age of 17 years), of whom 290 had successful ambulatory BP measurements (SpaceLab model 90207). Left ventricular diastolic function (the ratio of early-to-late transmitral velocities [E/A]) and mass were determined using echocardiography (single observer with a high level of intra-observer reproducibility). Conventional BP measurements were standardized and conducted by trained observers (non-physicians).

Results: ~66% of the sample were either overweight (26%) or obese (42%), ~41% had hypertension, ~22% were receiving treatment for hypertension, ~23% had diabetes mellitus (treatment or an HbA1c >6.1%) and only ~7% were receiving treatment for diabetes mellitus. After adjustments for age, gender, conventional systolic BP, heart rate, antihypertensive treatment, left ventricular mass index, the presence of diabetes mellitus and an HbA1c >6.1% and non-independence of family members; waist circumference ($r=-0.17, p<0.002$) and body mass ($r=-0.10, p<0.05$) were independently associated with E/A. With adjustments for 24-hour ambulatory systolic BP rather than conventional systolic BP in addition to other potential confounders; waist circumference ($r=-0.14, p=0.02$) was similarly independently associated with E/A. In a multivariate stepwise regression model, waist circumference was noted to explain ~11% of the variation in E/A, whilst systolic BP explained ~30% and diabetes mellitus explained ~2% of the variation in E/A.

Conclusions: Excess adiposity is associated with LV diastolic dysfunction beyond BP and diabetes mellitus in a population sample with a high prevalence of obesity. The assessment of LV diastolic dysfunction may therefore be an important measurement when assessing cardiovascular risk in overweight and obese patients.



EVALUATION OF PATIENTS POST PERCUTANEOUS CORONARY INTERVENTION WITH MYOCARDIAL PERFUSION IMAGING

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Background: Residual ischemia due to incomplete revascularization or in-stent stenosis is complications of percutaneous coronary interventions (PCI). Stress MPI has been shown to be a successful tool to risk stratify patients post-PCI for cardiac events (death, non-fatal myocardial infarction, revascularization). Data suggest that it may be reasonable to perform screening with stress myocardial perfusion imaging even on asymptomatic patients post PCI as 45% of chest pain is not due to restenosis and 30 - 65% of in-stent stenosis is asymptomatic.

Objective: To evaluate occurrence of ischemia in patients post PCI whether symptomatic or asymptomatic with the use of MPI.

Methods: 122 patients that underwent PCI were referred for stress myocardial perfusion imaging. There findings were analyzed for presence of ischemia or infarction, whether symptomatic or asymptomatic. There were 39 females (32%) mean age 61.7±10.5 years and 83 males (68%) 56.6±9.3 years.

Results:

Finding	Symptomatic	Asymptomatic	p
	68 pts (56%)	54 pts (44%)	
Normal perfusion	14 (21%)	11 (20%)	0.93 (NS)
Infarction without ischemia	13 (19%)	13 (24%)	0.01
Ischemia with or without infarction	41 (60%)	30 (56%)	0.93 (NS)

Conclusion: There was no statistically significant difference in the incidence of ischemia between symptomatic and asymptomatic patients. The presence of ischemia with or without infarction in 56% of asymptomatic patients post PCI suggests the importance of screening these patients for incomplete revascularization or progression of disease independently of symptoms, for risk stratification post procedure. MPI proves to be a highly sensitive non-invasive technique for post-PCI screening.

VALUE OF MYOCARDIAL PERFUSION IMAGING IN PATIENTS POST CORONARY ARTERY BYPASS GRAFT SURGERY

Libhaber C D, Libhaber E, Perumal S, Jogi H, Purbhoo K, Zambakides C A, Esser J D, Sliwa K, Essop M R, Vangu M D T H W

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Background: Risk stratification for cardiac events after coronary artery bypass graft (CABG) has been reported to be accurately obtained by myocardial perfusion imaging (MPI). Multiple reports state that five years after CABG, the yearly cardiac death rate in patients with moderate to severe

ischemia during stress MPI reaches 3.1% as compared to 0.7% in patients with no evidence of ischemia. It is suggested that in patients post-CABG with uncontrolled risk factors, recurrence of symptoms or decrease effort tolerance, stress MPI should be performed.

Objective: To evaluate occurrence of ischemia in patients post CABG, whether symptomatic or asymptomatic, with the use of MPI.

Methods: We analyzed findings in 142 patients post-CABG referred for stress myocardial perfusion imaging studies, whether symptomatic or asymptomatic and analyzed for presence of ischemia. There were 32 females (23%) mean age 61.8±10.8 years and 110 males (77%) 59.9±10.5 years.

Results:

Finding	Symptomatic	Asymptomatic	p
	81 pts (57%)	61 pts (43%)	
Normal perfusion	15 (18%)	7 (11%)	0.008
Infarction without ischemia	12 (15%)	12 (20%)	NS
Ischemia with or without infarction	54 (67%)	42 (68%)	0.43 (NS)

Conclusion: Although no statistically significant difference was found in the incidence of ischemia between symptomatic and asymptomatic patients, the presence of ischemia with or without infarction in 68% of asymptomatic patients post CABG suggests the importance of screening these patients for incomplete revascularization or progression of disease, not relying on symptoms for risk stratification post surgery. MPI proves to be a highly sensitive non-invasive technique for post-CABG screening.

DOES AMBULATORY BLOOD PRESSURE PREDICT LEFT VENTRICULAR DIASTOLIC DYSFUNCTION INDEPENDENT OF CONVENTIONAL BLOOD PRESSURE IN GROUPS OF AFRICAN DESCENT?

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Introduction: In contrast to other population groups, where ambulatory blood pressure (BP) refines the ability to predict left ventricular mass (LVM) over conventional BP, we have recently been unable to confirm this in urban developing communities of African descent. However, whether ambulatory BP predicts LV diastolic dysfunction, an independent predictor of cardiovascular events, beyond conventional BP, is uncertain.

Methods: We explored whether ambulatory BP (SpaceLabs model 90207) predicts LV diastolic dysfunction beyond conventional BP in 290 randomly selected subjects of African descent living in Soweto (minimum age of 17 years). Left ventricular diastolic function (the ratio of early-to-late transmitral velocities [E/A]) and LVM were determined using echocardiography (single observer with a high level of intra-observer reproducibility). Conventional BP measurements (x5) were standardized and conducted by trained observers (non-physicians).



Results: ~40% of the sample had hypertension and only ~22% were receiving treatment for hypertension. After adjustments for age, gender, waist circumference, heart rate, antihypertensive treatment, left ventricular mass index, the presence of diabetes mellitus and an HbA1c >6.1%; both 24-hour ($r=-0.14$, $p<0.03$) and daytime ($r=-0.18$, $p<0.003$) systolic BP were independently associated with E/A. However, after adjustments for conventional systolic BP and other confounders neither 24-hour ($r=0.01$) nor daytime ($r=-0.08$) systolic BP were independently associated with E/A. Sensitivity analysis conducted in 222 untreated subjects produced essentially the same outcomes. We also confirmed that after adjustments for conventional systolic BP and other confounders, neither 24-hour ($r=0.04$) nor daytime ($r=0.04$) systolic BP were independently associated with LVM indexed to body surface area.

Conclusions: Ambulatory BP does not predict either LV diastolic dysfunction or LVM index beyond conventional BP in a group of African ancestry. These data support the notion that ambulatory BP adds no additional value to conventional BP when predicting cardiac target organ changes.

NURSE-LED HEART FAILURE CLINIC IN A RURAL SETTING A SUCCESS

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Background: Nurse-led heart failure clinics elsewhere have been shown to improve patient outcomes and reduce health costs. A semi-autonomous nurse-led cardiologist-overseen clinic in a small rural hospital was therefore set up.

Methods: A total of 107 heart failure patients admitted to the outpatient clinic were evaluated.

Evaluation of the Hawera Heart Failure Clinic (Oct 2003 - Feb 2007) included examination of hospital re-admission rates, bed-days and surveys from patients, GPs and hospital physicians. Additional comments were also encouraged on the survey forms.

Results: Reductions in hospital days were shown by comparing admissions during an equal period before and after the first heart failure first clinic. In this way patients acted as their own control.

Readmission rates at three months, six months and one year showed 229, 153 and 118 fewer hospital days respectively. As one heart failure admission costs approximately NZ\$3257 the results can also be translated into cost savings.

Patients reported improvements in their condition, quality of life and their ability to self-manage their heart failure. With one exception there was a positive response from the GPs and hospital physicians with regard to the pharmacological and non-pharmacological management of patients as well as to the inter-disciplinary team-work that occurred.

Conclusions: Results of the evaluation suggested positive trends with reduced morbidity and costs. Also important was the positive feedback from the intra-disciplinary team and patients involved. On the basis of this audit and strength of data from clinical trials a new intra-sectoral community clinic has been set up. The clinic is run autonomously by a prescribing nurse practitioner but with strong links to the cardiology department.

burden, aspiration thrombectomy is a rapidly evolving technique.

Methods: We analyzed 63 consecutive patients who underwent primary percutaneous coronary intervention (PCI) in last one year i.e. from September 2006 to Aug 2007. The aspiration catheters were used in 11 patients. Use of aspiration catheter was done in a selective group only viz. >50% luminal thrombus burden in an infarct related artery (IRA) after initial wire crossing and the artery being >2.5mm in diameter. Rio catheter (Boston Scientific) was used in 4 patients and Export catheter (Medtronic) was used in 7 patients. All these were compatible to 6F guiding catheter. In each case, 2-3 passes were done. IRA was left anterior descending (LAD) artery in 5 patients, right coronary artery (RCA) in 4 patients and left circumflex (LCX) artery in 2 patients.

Observations: Macroscopic thrombotic fragments in the aspirate were seen in only 5 patients, though the thrombus burden was significantly reduced in 10 out of 11 patients as seen by immediate check angiogram. There was no change in the thrombus appearance in 1 case. One case with LCX artery occlusion showed no residual stenosis after aspiration and did not require any further ballooning or stenting. None of the patients with angiographic reduction of thrombus burden after aspiration demonstrated slow flow/no-reflow phenomenon and there was TIMI-3 flow at the end of the procedure in all these patients. No procedure related complication was noted.

Conclusion: Aspiration thrombectomy is a safe and effective procedure during primary PCI. A selective rather than universal use of this approach could be more feasible in order to minimize the cost and time of the procedure while optimizing the outcomes.

THE POTENTIAL LONG-TERM CARDIAC IMPLICATIONS OF ANTECEDENT CRANIOCEREBRAL INJURY AND THE ROLE OF CATECHOLAMINES IN THE PRODUCTION OF CARDIAC HYPERTROPHY AND CARDIOMYOCYTE NECROSIS

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While donor hearts for cardiac transplantation have traditionally been harvested from head injured brain dead victims on the premise that these donors manifest neither macroscopic nor microscopic evidence of cardiac damage, it has been shown both experimentally and clinically that brain death is associated with severe haemodynamic deterioration. However, it has also been shown that acute severe haemodynamic derangement can be fully reversed and that this, therefore, does not constitute an impediment or contraindication to transplantation of a donor heart. In contrast to the acute cardiac impairment which occurs in the brain death period immediately preceding cardiac transplantation, however, few studies have attempted to address the issue of the long-term cardiac implications of head injury, implications which may carry profound connotations both for the integrity of the transplanted heart and the recipient's future clinical cardiovascular status.

In an attempt to examine this issue, a prospective study was undertaken of all cases of cranio-cerebral injury coming to medicolegal autopsy over a six-month period in order to determine the nature of any histopathological changes which might arise following head injury. More importantly, the issue of whether any progression and evolution of these latter changes was explored, changes which might, with chronicity, carry potential implications for the patient's future cardiovascular status.

In this study the role played by catecholamines in the induction of cardiac hypertrophy and cardiomyocyte necrosis was explored together with the signalling mechanisms involved. In addition, in view of the effect of head injuries and intracranial lesions on the myocardium, the electrocardiographic abnormalities occurring in association with these lesions and mimicking myocardial infarction and myocardial ischaemic changes was reviewed, hopefully alerting clinicians to the presence of unsuspected intracranial pathology.

1100

SELECTIVE USE OF ASPIRATION CATHETER FOR THROMBUS REDUCTION DURING PRIMARY ANGIOPLASTY

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Background: Acute ST elevation myocardial infarction (STEMI) is predominantly caused by plaque rupture and sudden thrombotic occlusion of a coronary artery. Among various strategies to mitigate the thrombus



Statistical analysis indicated a statistically significant relationship between antecedent cranio-cerebral injury and subsequent myocardial fibroses at a level of significance appropriate to exploratory research. Hence, it is suggested that all long-term survivors of head injury and, in particular, young survivors undergo routine follow-up, surveillance and monitoring of their clinical cardiovascular status in order to detect this potential complication which carries with it the risk of impairment of myocardial contractile function and, therefore, possible myocardial insufficiency. With regard to cardiac transplant recipients in whom donor hearts have been harvested from head injured brain dead victims, it is similarly suggested that they be monitored for the possible advent of cardiac decompensation on the basis of this latter complication in addition to standard monitoring for signs of transplant rejection.

TROPONIN T AND B-MYOSIN MUTATIONS HAVE DISTINCT CARDIAC FUNCTIONAL EFFECTS IN HYPERTROPHIC CARDIOMYOPATHY PATIENTS WITHOUT HYPERTROPHY

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The validity of genotype:phenotype correlation studies in human hypertrophic cardiomyopathy (HCM) has recently been questioned, yet animal models and *in vitro* studies suggest distinct effects for different mutations. The aims of this study were to investigate whether distinct hypertrophic cardiomyopathy (HCM) have different consequences on cardiac structure and function in the absence of the confounding effects of hypertrophy.

Individuals aged 20-65 years belonging to 21 R92W_{TNNT2}, R403W_{MYH7}, or A797T_{MYH7} mutation-bearing families were investigated with 2D, M-mode and Doppler echocardiography. Cardiac structural and functional parameters were compared between prehypertrophic mutation-carriers and their non-carrier family members, with concomitant adjustment for appropriate covariates. Findings were evaluated against existing animal and *in vitro* functional data.

The distinct functional effect of the R92W_{TNNT2} mutation was a relative increase in systolic functional parameters, that of the A797T_{MYH7} mutation was reduced diastolic function, while the R403W_{MYH7} mutation reduced both systolic and diastolic function. The observed early effects of the R92W_{TNNT2} mutation mechanistically fit with prolonged force-transients precipitated by increased Ca²⁺-sensitivity of the thin filament, and that of the MYH7 mutations with local ATP depletion.

Evaluation of the impact of the mutations on cardiac structure and function in prehypertrophic mutation-carriers, relative to the base-line norm provided by their non-carrier family-members, best recapitulated existing animal and *in vitro* functional data, while inclusion of mutation-carriers with hypertrophy obscured such findings. The results highlight the possibility that timely treatment aimed at ameliorating Ca²⁺-sensitivity for R92W_{TNNT2} carriers, and energy depletion for MYH7 mutation carriers, may offer a plausible approach for preventing progression from a preclinical into a decompensated state.

RISK FACTOR PROFILES IN SOUTH AFRICAN PATIENTS WITH HYPERTROPHIC CARDIOMYOPATHY CAUSED BY DISTINCT FOUNDER MUTATIONS

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Implantable cardioverter defibrillators are increasingly used to prevent sudden cardiac death (SCD) in hypertrophic cardiomyopathy (HCM)

patients deemed to be at high risk. Risk stratification assesses a number of factors, including a family history of multiple SCDs, unexplained syncope, non-sustained ventricular tachycardia on Holter ECG, maximal left ventricular wall thickness (mLVWT) ≥ 30 mm and abnormal blood pressure response during exercise testing. Early genetic studies also suggested that correlations exist between genetic mutation and survival. The aim of this study was to compare risk factor profiles between three South African HCM founder mutation groups, to assess whether genotype correlated with risk profile.

Twenty-one South African families in which one of three HCM founder mutations segregated participated in this study. Thirty R92W_{TNNT2}, 24 R403W_{MYH7}, and 27 A797T_{MYH7} HCM mutation-bearing individuals as well as 67 of their non-carrier relatives were investigated with 2D and M-mode echocardiography and exercised under a modified Bruce protocol. Patient and family histories were obtained.

Significantly more R92W_{TNNT2} individuals failed to increase systolic blood pressure by more than 20 mmHg than did individuals from either the control or other mutation groups ($p=0.015$). R92W_{TNNT2} individuals also demonstrated more syncope ($p=0.007$) than did the control or other mutation groups. On the other hand, significantly more A797T_{MYH7} individuals demonstrated overt hypertrophy (mLVWT ≥ 30 mm). Most SCDs occurred in R92W_{TNNT2} families, while A797T_{MYH7} families suffered more SCDs than R403W_{MYH7} families. These deaths most often occurred during periods of enhanced emotional or exertional stress, predominantly affected males.

Risk factor profiles differed according to causal mutation in South African HCM families, with R92W_{TNNT2}-positive individuals demonstrating the highest number of risk factors and R403W_{MYH7} the lowest. Mechanisms underlying SCD for the different mutations are not clear, but may involve alterations in calcium-handling and/or alterations in vagal tone.

SUBMITRAL LEFT VENTRICULAR ANEURYSMS IN A PAEDIATRIC POPULATION

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Submitral left ventricular aneurysms are a widely recognized but poorly known entity that has been described mainly in Sub Saharan Africa. Submitral aneurysms occur in relation to the annulus of the mitral valve around the posteromedial commissural line. Along with subaortic aneurysms they have been grouped as subvalvar aneurysms. Patients with submitral aneurysms may be asymptomatic or clinical symptoms arise as a result of valvar regurgitation or occasionally from compression of cardiac structures. The aetiology of subvalvar aneurysms is mostly thought to be a congenital weakness of the fibrous annulus of the valve. Coexistent pathology such as rheumatic carditis or tuberculosis has been described in association with this entity. Diagnosis can be made by echocardiography.

This retrospective review describes the clinical, anatomical and histological features of submitral aneurysms seen in the paediatric population at the Johannesburg and CH Baragwanath hospitals between 2001 and 2007.

There were 6 patients with submitral aneurysms, 3 male and 3 female. The age at clinical presentation varied from 13 months to 15 years. All but one of the patients presented with mitral regurgitation and congestive cardiac failure. The diagnosis was made in all instances on transthoracic echocardiography and supplemented with transoesophageal echocardiography or angiograms selectively. Five of the six patients underwent surgery and in 4 of these patients the aneurysm was related to the posterior mitral leaflet and in one patient the aneurysm was related to the anterior mitral leaflet. Histological diagnosis revealed features of rheumatic carditis in 2 patients, features of tuberculosis in one patient and nonspecific features in 2 patients.



The outcome in all patients who underwent surgery was favourable. The sixth patient died soon after the initial diagnosis.

Submitral aneurysms are not uncommon. The cause is considered to be congenital with acquired factors confounding. With surgical treatment outcome is usually favourable.

RING CHROMOSOME 4 IN A DYSMORPHIC INFANT WITH CONGENITAL HEART DEFECTS AND EARLY ONSET EISENMENGER SYNDROME

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Ring chromosome formation is a rare structural karyotypic defect which is thought to be the result of deletion of genetic material on the terminal portion of the long and short arm of a chromosome.

Ring chromosome 4 cases are rarely observed and only 16 have been documented in the literature. There is a variable phenotypic expression which includes features of both 4p and 4q deletion. The phenotype consists of severe mental and motor retardation, microcephaly, cleft palate and lip, growth retardation, facial dysmorphism, upper extremity malformations, congenital heart defects mostly ASD, VSD.

We present a descriptive case of a 1 year 5 month, female infant, born normally at 36 weeks gestation to non-consanguineous parents. Her birth weight was 2,16 kg. She did not cry well at birth and was immediately admitted to the neonatal ward for oxygen therapy for respiratory distress.

On examination at 3 weeks of age, she weighed 1,74 kg; length 45 cm; HC 28.5 cm (all below 3rd centile). She had microcephaly, micrognathia, facial dysmorphism, abnormal hands and hemi-vertebra at T10. Cardiac evaluation revealed a large apical muscular VSD and a large ASD secundum.

Chromosome analysis revealed a 46,XX,r(4),(p?q?) karyotype indicating that one of her number 4 chromosomes has a ring configuration. A FISH analysis was done to determine whether the Wolf-Hirschhorn critical region on 4p was deleted. The results showed that the 4p16,3 Wolf-Hirschhorn region was intact and therefore the deletion had occurred distal to this region.

She is being treated conservatively with regular follow ups at cardiac and genetic clinic. Currently she is 1 year and 5 months old weighing only 4.3 kg and has developed signs of severe rapidly progressive pulmonary hypertension and early onset Eisenmenger Syndrome.

ACUTE RHEUMATIC FEVER SURVEILLANCE IN SOUTH AFRICA: AN EVALUATION TOOL BASED ON THE CDC GUIDELINES FOR EVALUATING PUBLIC HEALTH SURVEILLANCE SYSTEMS

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Background: The primary objectives of disease surveillance are to monitor and control disease trends. In South Africa (SA), the system is based on law, coupled with guidelines by the department of health. An earlier study revealed a breakdown in the reporting of acute rheumatic fever (ARF) at local, provincial and national levels of the SA Health System. The CDC has published a generic set of guidelines for evaluating surveillance systems that can be adapted to different environments. We present an evaluation tool, based on these guidelines with contextual adaptations, for conducting an evaluation of the ARF surveillance system within the Cape Metro region of SA.

Methods: Five steps are incorporated; some of those outlined by the CDC guidelines will be forgone as they will not be directly applicable. The first step will be engagement of the relevant stakeholders to ensure that the

evaluation of the ARF notification system addresses appropriate questions and that it assesses the pertinent attributes of the system. Step two will be to describe the system as regards purpose, flow of information, and the resources required in maintaining the system. The third step involves a more focused structuring of the evaluation within our developing country context. Step four focuses on evidence for performance in adhering to the national ARF guidelines. This will involve engaging the daily users of the system. Thereafter, drawing conclusions and justified recommendations, to be communicated to the relevant stakeholders by way of a written or oral presentations, constitute steps five and six, respectively.

Result and conclusion: The CDC's guidelines, together with our modifications, appear to provide a practical framework on which to base a systematic yet tailored evaluation of the ARF surveillance system in South Africa. The CDC's guidelines are largely comprehensive and amendable to the contextual issues of ARF surveillance in SA.

THREE SIBLINGS WITH IDENTICAL CYANOTIC CONGENITAL HEART LESION

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We report on a family with three siblings (two brothers and one sister) with identical cyanotic congenital heart lesion. The heart lesion consists of pulmonary atresia with ventricular septal defect, absent native pulmonary artery, right sided aortic arch, major aortopulmonary collaterals and retro aortic innominate vein. We diagnosed the proband during neonatal period and subsequently discovered that he had a fourteen year old brother attending our paediatric cardiology clinic and a sister who demised at the age of four months with identical lesion.

The recurrence in this family supports the importance of the genetic basis of congenital heart defects. Molecular genetics has provided much awaited insight into genetic basis of many congenital heart defects but still proves to be extremely expensive for our setting.

THE IMPACT OF URBANISATION ON BLACK PATIENTS PRESENTING WITH CORONARY ARTERY DISEASES (CAD) AT BARAGWANATH HOSPITAL CARDIAC CLINIC, SOWETO

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Introduction: Coronary artery disease (CAD) is still rare in Africa. With epidemiologic transition to increasing risk behaviours linked to urbanization and westernization, this is expected to change. As part of the larger Heart of Soweto Study we examined the volume and characteristics of CAD in patients presenting to the Cardiology Unit at the Baragwanath Hospital in Soweto.

Methods: Baragwanath Hospital in Soweto, South Africa, provides health care to a population of 1-1.5 million mainly black Africans. We registered demographic, clinical and relevant biochemical and imaging data from all new patients with CVD attending the hospital's Cardiology Unit in 2006: this includes those referred by Soweto's primary care clinics and other smaller hospitals.

Results: We identified 4162 cases of CVD (1593 newly diagnosed) in 2006. Black Africans (85%) and women predominated (60%). Among the 1593 newly diagnosed cases, there were 165 cases of CAD (10.4%). Mean age was 56.7 ± 12.4 years, 59% were male and 47% were black African. Compared to the rest of the study cohort, women (OR 0.37, 95% CI 0.31- 0.44: $p < 0.0001$)



and black Africans (OR 0.10, 95% CI 0.07 to 0.14: $p < 0.0001$) were far less likely to be diagnosed with CAD compared to men and other racial groups, respectively. Overall, 51% had a smoking history, 37% were obese, and 36% had a positive family history or were dyslipidaemic and 21% diabetes (71% with multiple risk factors).

Conclusion: These preliminary data supports our hypothesis that epidemiological transition is still at an earlier stage but this is expected to change with increase in urbanization and westernization seen in black patients presenting with CAD to the Baragwanath Hospital in Soweto South Africa.

LEFT VENTRICULAR MASS REGRESSION, BLOOD PRESSURE CONTROL AND NT-PRO BNP CONCENTRATIONS IN BLACK AFRICAN PATIENTS WITH MODERATE TO SEVERE HYPERTENSION AFTER 1- 4 YEARS FOLLOW-UP

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Background: Hypertension is a significant cause of heart failure and concentric left ventricular hypertrophy (LVH). Left ventricular mass (LVM) and LVH prevalence can be reduced through antihypertensive therapy. Plasma brain natriuretic peptide (BNP) levels are associated with increased cardiovascular mortality, independent of age, functional class and left ventricular function. We evaluated whether blood pressure (BP) reduction with standard antihypertensive therapy is associated with a reduction in LVM and NT-pro BNP levels respectively.

Method: 25 of a total of 118 Black African patients with moderate to severe hypertension on standard antihypertensive therapy, previously studied were followed up after 1-4 years with repeat ambulatory blood pressure monitoring, echocardiography and NT-pro BNP measurements. Of the 25 patients followed up, 56% were assessed after 3-4 years and 44% after 1-2 years respectively.

Results: Of the 25 patients reviewed, (age 58 ± 10 years, males 12), 7 (68%) had a controlled BP of less than 140/90 mm Hg after treatment with a mean BP of $(131 \pm 13 / 80 \pm 8$ mmHg). 20% of patients were in NYHA FC I, 72% were in FC II and 8% were in FC III. No LVMI reduction was observed from baseline to follow up respectively (131 ± 53 mg/m² and 134 ± 48 mg/m²). NT-pro BNP levels were significantly reduced (from 708 ± 2040 ng/l to 229 ± 376 ng/l), with a $p < 0.0001$.

Conclusion: The interim analysis showed that in spite of successful antihypertensive therapy, no significant reduction in LVM was observed. However there was a significant reduction in NT-pro BNP levels.

ANOMALOUS ORIGIN OF THE LEFT PULMONARY ARTERY FROM THE ASCENDING AORTA IN TWO CHILDREN WITH PULMONARY ATRESIA, VSD, LEFT AORTIC ARCH AND RIGHT SIDED MAJOR AORTO-PULMONARY COLLATERAL ARTERIES

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We report two rare cases with an identical congenital heart lesion viz. anomalous origin of the left pulmonary artery (AOLPA) from the ascending

aorta associated with pulmonary atresia, a ventricular septal defect and a left aortic arch. The cases are unusual because AOLPA is reportedly more commonly associated with a right aortic arch and it is more usual for the right pulmonary artery to originate anomalously from the ascending aorta. The pulmonary blood supply to the right lung in both patients was absent and provided instead by Major Aorto-Pulmonary Collateral Arteries which were stenosed at multiple levels. The AOLPA in both patients originated from the postero-lateral aspect of the ascending aorta just distal to the sinotubular junction. Only one patient showed the more common association of an unusual aortic arch branching pattern in the form of an anomalous right subclavian artery. Neither patient was in heart failure and the chest X-ray in both revealed differential pulmonary perfusion with prominent vascularity of the left lung. As expected cardiac catheterization showed systemic pressures within the anomalous left pulmonary artery. Karyotyping revealed normal chromosomes and Fluorescent In-Situ Hybridization done in one patient was negative for chromosome 22q11.2 microdeletion. Both patients have been managed conservatively.

TIME-COURSE CHANGES IN BRAIN NATRIURETIC PEPTIDE IN SEVERE MITRAL REGURGITATION

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Aim: Brain natriuretic peptide (BNP) rises in response to ventricular muscle stretch and levels have been used to detect early heart failure. We hypothesized that BNP would be activated in patients with regurgitant valvular heart disease with concomitant ventricular dilatation. We therefore studied the pattern of changes in BNP in patients with chronic severe rheumatic mitral regurgitation who were undergoing mitral valve replacement.

Method: Twenty-seven consecutive patients undergoing surgery in the last eight months were prospectively evaluated. Simultaneous quantification of the severity of mitral regurgitation (MR), left ventricular (LV) end systolic volume (ESV), left atrial (LA) volume and Doppler filling ratios (mitral (E)/annulus (Ea)) were performed preoperatively, at 1-week and at the six-week follow-up visit. Twenty-eight controls subjects with severe MR were selected from the hospital clinic where they were being followed up.

Results: There were seven males and twenty females with a mean age of 20 years. BNP levels were markedly elevated preoperatively (262 pg/mol) in all patients selected for surgery compared to patients with similar degrees of MR (57 pg/mol) who were being followed up at the clinic ($p = 0.0001$). EchoDoppler parameters were similar in all patients preoperatively except LA size (76 vs 66 mm $p < 0.01$) and volume (309 vs 173 ml $p < 0.004$) which were elevated in the surgical group. BNP levels increased further immediately (at one week) post surgery (497 pg/mol) and subsided at the six week follow-up visit (71 pg/mol). These changes were accompanied by significant reduction in LA (205 to 175 ml $p = 0.042$) and LV chamber dimensions (ESV 83 to 64 ml $p = 0.003$) with an increase in the ejection fraction from 42% at one week to 52% at six weeks. Myocardial tissue Doppler showed an increase in the systolic wave (0.06 to 0.07 $p = 0.019$). Four patients had abnormally elevated BNP levels (> 31 pg/mol) at the 6-week follow-up visit. These were accompanied by increased E/Ea ratio (> 10) on tissue Doppler and impaired LV contractility (ejection fraction $< 40\%$). One patient demised postoperatively at 1 week follow-up. The BNP level recorded was 2065 pg/mol.

Conclusion: Elevated BNP correctly identified patients selected for surgery. BNP levels subsided at the six weeks follow-up visit after surgery, but remained elevated in those with persistent LV dysfunction. Estimation of BNP could assist in the timing of surgery in patients with regurgitant valvular heart disease prior to the onset of irreversible LV dysfunction.



FOOD CHOICES AND THEIR NUTRITIONAL VALUE IN BLACK AFRICAN PATIENTS WITH HEART FAILURE: THE HEART OF SOWETO STUDY

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Background: Heart failure (HF), an emerging health problem in one of Africa's largest urban concentrations of black Africans, requires targeted prevention and management strategies. However, there are no data on the dietary patterns of black African patients with HF to determine the potential of dietary interventions.

Methods: Fifty consecutive black African patients with HF attending the Baragwanath Hospital in Soweto were surveyed about their food choices and dietary intake via the validated quantitative food frequency questionnaires (QFFQ). Data were translated into nutrient data using Dietary Manager based on South African food composition tables and compared to recommended values.

Results: Overall, 27 (54%) of patients were female with a mean age of 49.9 ± 16.4 (range 23 to 79) years. In women, notable food choices likely to negatively impact on heart health included added sugar (75% of women: mean daily intake 15 ± 9.6g), sweet drinks (51%: 310 ± 356ml) and salted snacks (61%: 15 ± 22g). The same food choices in men were 94% (15 ± 11g added sugar), 65% (438 ± 567ml sweet drink) and 74% (16 ± 18g salted snacks). Relative to age-specific recommendations, on average women aged < 35 years and ≥ 35 years derived 74% and 66% of their calcium, 78% and 46% of vitamin C and 99% and 58% of vitamin E daily requirements. The equivalent figures for men (all aged ≥ 35 years) were 115%, 70% and 71%, respectively. Most significantly, in this HF group, the overall mean daily intake of sodium was 1470 ± 1280% (range 238 to 5523%) greater than recommended consumption levels for a healthy population. We are currently calculating the cost of healthy versus unhealthy choices for HF patients based on these data.

Conclusions: These data highlight a significant potential to improve the health status of black African patients with HF by recommending and supporting healthier food choices; particularly in respect to excess salt and sweet drink consumption levels.

CASE REPORT: IS THIS THE ELUSIVE CONGENITAL CORONARY ARTERY ANEURYSM?

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Medunsa

A report on an 8 year old boy, who was first seen aged 6 months with a suspicion of a VSD. Patient was found to have a systolic murmur and was seen after a further year, with an echocardiographic evaluation of a "funny heart". He presented again aged 5 years, with a history of chest pain on exertion and echocardiography suggesting a VSD. At 8 years he then presented with the same history and his ECG suggested ischaemic disease, his echocardiogram showed a cystic lesion, lying over the RVOT compressing it and an abnormal looking LV cavity and reduced ejection fraction. Angiography further showed compression of the RVOT, large left main and pooling of contrast just distal to the left main. Surgery found a large aneurysm over the RV, communication with the LAD and a large left main. It seemed to be a developmental aneurysm, related to the RV sinusoids, which got aneurysmal and caused ischaemia. Patient well after surgery, with improved effort tolerance and pain free.

PATIENT COMPLIANCE AND KNOWLEDGE ON HEART FAILURE IN SOUTH AFRICA

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Aims: Despite its clinical importance there is a paucity of data on treatment adherence in patients with heart failure (HF) in the African context. We examined the pattern of treatment adherence in a large cohort of HF patients from Soweto, South Africa.

Material and methods: The "Patient compliance and knowledge on Heart Failure Survey" was initially developed, validated and then applied to 200 consecutive HF patients with a left ventricular ejection fraction ≤ 45% attending the Cardiology Clinic of the Chris Hani Baragwanath Hospital in Soweto. A pill count of prescribed HF treatment was also conducted in 82 patients (41%) at 1 month post interview.

Results: The mean age of participating patients was 56 ± 14 years and 45.5% were female. Standard treatment included ACE inhibition (74%), beta-blockers (84%), loop-diuretic (93%) and spironolactone (64%), with a mean of 6 ± 2 medications and 9 ± 3 doses prescribed. Overall, 81% of patients adhered to their follow-up appointment schedule. In respect to fluid management and dietary advice 56% of patients adhered to the recommended fluid intake of < 2 L/day while only 13% reported eating 5 servings of fruit every day. As part of monitoring their HF status, only 2.5% of patients weighed themselves daily. In respect to maximising their overall health status, 38% reported to be physically active for > 20 to 30 minutes/day while 16% persisted in smoking tobacco. Alternatively, only 2% of patients had excessive alcohol intake. Overall, patient knowledge concerning their prescribed medication was poor: 56% could not name the effect or a side effect of their medication. An average score of 69% was achieved on 10 questions concerning HF management. Pill counts revealed that 71% patients were clearly compliant and 22% non-compliant with their prescribed HF regimen.

Conclusion: As in many other regions of the world, non-adherence to complex HF treatment is a substantive problem in Soweto, South Africa. These data confirm the need for a dedicated HF program to optimise HF management and outcomes.

EARLY EXPERIENCE OF TRANS-CATHETER MAZE (TCM) PROCEDURE FOR PATIENTS WITH ATRIAL FIBRILLATION IN SOUTH AFRICA

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Introduction: The main purpose of this presentation is to share the learning experience from performing pulmonary vein ablations. The author will share from this experience what he believes to be high risk patients and anatomy. Following current guidelines, catheter ablation for atrial fibrillation (AF) is a rapidly growing therapeutic modality in RSA.

Methods and results: A 3D model of the left atrium (LA) was created using the NavX Ensite system¹ and reconstructed 3D Cardiac Tomography. TCM procedure was performed in the LA (circumferential PV, roof, posterior, anterior, LA isthmus and coronary sinus lines) as described by Pappone *et al.*² Success of PV disconnection was confirmed using peak-to-peak voltage mapping. Only patients with failed drug therapy were considered for treatment. 81 patients were treated: 27 permanent; 15 persistent; 39 paroxysmal. Mean age was 61 years (30-81; M: 59, F: 22). Mean procedure time was 167.6 minutes and fluoroscopy 67.5 minutes.



All complications	N
Phrenic nerve palsy	1
Pericardial effusion requiring pericardiocentesis	8
Pericardial effusion requiring thoracotomy	1
Stroke	1
Late onset atrial flutter	13

Sinus rhythm, at least 9 months post procedure, was considered a success. Success rate was 59% in the permanent, 73% in the persistent and 86% in the paroxysmal group. Of the 20 patients not in sinus rhythm, 4 were significantly less symptomatic.

Conclusions: TCM using 3D mapping technology is an effective treatment for AF and success rate should increase with experience and improved NavX Ensite technology. Cost effectiveness remains to be assessed in the RSA.

1. St Jude Medical, St Paul, MN, USA.
2. Pappone C, Manguso F, Vicedomini G, *et al.* Prevention of iatrogenic atrial tachycardia after ablation of atrial fibrillation: A prospective randomized study comparing circumferential pulmonary vein ablation with a modified approach. *Circulation* 2004; 110: 3036-3042.

SURGICAL MANAGEMENT FOR ATRIAL FIBRILLATION: A NOVEL METHOD USING IRRIGATED MONOPOLAR ELECTROCAUTERY ABLATION

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30 to 84% of patients undergoing mitral valve surgery present with associated permanent atrial fibrillation (AF). After mitral surgery up to 20% of these patients will revert to normal sinus rhythm (NSR) but AF will persist in the majority. AF increases the risk of stroke, other associated complications, and patients require more frequent physician visits and hospitalisations.

The "cut and sew" Cox - Maze procedure remains the gold standard for restoring NSR in patients with permanent AF. Due to its complexity, and bleeding complications, alternative methods have been used to create the ablation lines - in particular radiofrequency ablation.

The cost of the commercially available probes has limited the use of surgical radiofrequency ablation at Groote Schuur Hospital. The use of *non-irrigated* electrocautery ablation has been reported previously. Criticism of this technique includes the tissue charring created and difficulty in achieving transmural atrial lesions. This prompted us to develop a novel *irrigated* monopolar electrocautery ablation to restore NSR in patients with permanent AF.

During the 2½ year period from September 2004 to April 2007, 40 cardiac surgery patients also had a modified Maze procedure. 16 were males and 24 females with a mean age of 47.15 years. All procedures were performed by a single surgeon. 38 had mitral surgery (either isolated or in combination with other procedures); one patient had an isolated ASD repair and one an isolated aortic valve replacement.

Early mortality was 7.5% - none related to the Maze procedure. Mean hospital stay was 10.1 days, with sinus-conversion rate of 70% upon discharge.

Follow up was 97.5% complete and ranged from 1 to 30 months (mean 14 months). There were three late valve related mortalities. No late electrical cardioversion was performed and 69% of patients (n=25/36) remained in sinus rhythm at last follow up.

TWO UNUSUAL CASES OF PURULENT PERICARDITIS

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Although rare in developed countries, purulent pericarditis is still seen in developing countries, and is usually caused by *Staphylococcus aureus* or *Streptococcus pneumoniae*.

We report two cases of unusual causes of bacterial pericarditis identified during the past twelve months at the Red Cross and Tygerberg Children's Hospitals, Cape Town: the first caused by *Streptococcus milleri* in a 2-year-old boy, and a second of *Escherichia coli* pericarditis in a 4-year-old girl associated with appendicitis.

IS MITOCHONRIAL VARIANT T16189C ASSOCIATED WITH AN INCREASED RISK OF DILATED CARDIOMYOPATHY IN HIV POSITIVE PATIENTS?

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Background: Mitochondrial variant T16189C has been reported to be associated with an increased risk of dilated cardiomyopathy. It generates an uninterrupted homopolymeric C-tract (approximately 10 cytosines), which causes heteroplasmic length variation of the mtDNA, possibly as a result of replication slippage. We hypothesize that this variant predisposes to HIV associated cardiomyopathy.

Objective: To determine the frequency of the T16189C variant in HIV associated cardiomyopathy (HIVAC) patients.

Method: A detailed four year case-control study (2002- 2006) was designed and three South African black cohorts recruited: 30 HIV positive black patients with dilated cardiomyopathy, 38 HIV positive black controls without dilated cardiomyopathy and 117 HIV negative controls without dilated cardiomyopathy (background population). Where possible, cases and controls were matched for age and gender. All 185 DNA samples were sequenced in the forward and reverse directions on an ABI3100 sequencer.

Results: The T16189C variant was detected at a frequency of 70% (21/30) in the HIVACs, 66% in the HIV positive controls (25/38) and 76% in the South African background population (89/117). There was no significant difference in the frequency distribution of the variant between the three groups ($p=0.43$). The presence of a substitution, other than the T16189C variant, appeared to stabilize the homopolymeric C-tract. Numerous other variants were also found in this hypervariable region. Base substitutions were found to occur more at positions C16187 and T16189 compared with the other bases. Also observed was that more changes were prevalent in the HIV positive groups (HIVAC and HIV positive control group) compared to the background population (without HIV).

Conclusion: This study found that the T16189C variant is not a risk factor for HIV associated cardiomyopathy.

THE OCCLUSION OF RESIDUAL FORWARD FLOW IN THE SUBPULMONARY OUTFLOW TRACT IN PATIENTS UNDERGOING SINGLE VENTRICLE PALLIATION

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Six patients (4 M, 2 F age 0.5 - 15 yrs) underwent cardiac catheterisation in an attempt to decrease forward flow into the pulmonary arteries and decrease PA pressure. Three patients had tricuspid atresia; the other three patients had two-ventricle hearts in which a conventional repair was not possible.

Three patients were post final palliation (TCPC or Kawashima) and three were post recent Glenn shunt. In all patients, the procedure was successful. Various devices - plugs, duct occluders and septal defect devices were used to close the outflow tract. The procedures tended to be lengthy requiring combined femoral and jugular vein approaches.

Closure of the outflow tract resulted in improvement in the clinical condition of the patient while avoiding surgery in patients with repeated thoracotomies.



LIMITATIONS IN PREOPERATIVE ASSESSMENT OF PULMONARY HYPERTENSION

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Background: Advanced pulmonary hypertension is a well-recognised feature of untreated congenital cardiac disease. We previously presented our operative results in patients with significant pulmonary hypertension and demonstrated satisfactory results.

Objective: To determine the outcomes of patients with significant pulmonary hypertension undergoing surgery and evaluate the predictive value of present pre-operative assessment techniques.

Methods: This was a descriptive, retrospective analysis of all patients who had a VSD or AVSD repair from 2001 to 2006 at the Universitas Academic Hospital complex. Patient records were used to obtain all relevant demographic, catheterisation, echocardiographic and surgical data. Patients were divided into 3 groups according to calculated pulmonary resistance (Rp): group A = $R_p \leq 3,5U$, group B = $R_p 3,6 - 6U$ and group C = $R_p > 6U$.

Results: A total of 173 patients were available for analysis after exclusion of complex lesions and incomplete data. A total of 125 patients had surgery and 48 were either followed up or considered to be inoperable. In the surgical group there were 66 patients in Group A, 38 in group B and 21 in group C. There were 4 perioperative deaths in group A, 2 in group B and 2 in group C. Patients were followed-up echocardiographically an average of 3,6 years post-operatively and revealed a reduction in pulmonary artery pressure of 20% (VSD) and 22% (AVSD) in group A, 35% (VSD) and 42% (AVSD) in group B and 60% (VSD) and 37% (AVSD) in group C.

Conclusion: A disturbing number of children have significant pulmonary hypertension when they present to our unit, indicating the urgent need for proper and effective referral of children with congenital heart disease. Patients with severe pulmonary hypertension (pulmonary resistances of more than 6U) who retain pulmonary vascular reactivity seems to be operable and improve over medium term follow-up. The limitations of conventional pre-operative assessment techniques, the need for more definitive evaluation techniques and the interpretation of results need to be critically re-assessed to prevent denial of surgery to worthy candidates.

A CRITICAL ROLE FOR STAT-3 IN ISCHAEMIC POSTCONDITIONING

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Aim: The recently discovered phenomenon of ischaemic postconditioning (IPostC) is a promising therapy for acute myocardial infarction because such therapy can be administered immediately upon revascularisation. However, IPostC mechanisms remain unclear. We hypothesised that IPostC may protect via activation of a prosurvival signal nuclear-regulatory peptide STAT-3 (Signal Transducer and Activator of transcription-3).

Methods: Control isolated rat hearts underwent 30 min global ischemia and 30 min reperfusion (CTL). Postconditioned hearts underwent 4 cycles of 10 sec ischemia-reperfusion initiated at reperfusion onset (IPostC). In a third group, hearts were postconditioned in conjunction with AG490 (100nM), a STAT-3 inhibitor (IPostC+AG). Additional rats underwent similar protocols involving 30 min regional ischaemia and 120 min reperfusion to measure infarct size. Western blot analyses done 15 min after reperfusion were used to determine STAT-3 phosphorylation following IPostC.

Results: Control hearts had a rate pressure product (RPP: % of baseline value) of $19 \pm 4\%$. IPostC improved RPP to $45 \pm 6\%$ ($p < 0.05$ vs CTL). Perfusion of AG490 during the first 15 min of reperfusion abolished the IPostC

cardioprotective effect (RPP for IPostC+AG: $17 \pm 4\%$, $p < 0.05$ vs IPostC). Similarly, IPostC improved infarct size from $33 \pm 4\%$ (CTL) to $11 \pm 1\%$ ($p < 0.01$) and perfusion of AG490 during IPostC abolished the cardioprotective effect (IPostC+AG: $25 \pm 5\%$). Western blot analysis revealed IPostC to decrease phosphorylation of cytosolic STAT-3 (pSTAT-3) by 68% (IPostC: 0.5 ± 0.1 arbitrary units vs 1.6 ± 0.3 for control group, $p < 0.01$), suggesting translocation and activation of STAT-3 in the nucleus after IPostC.

Conclusion: Our results are first to suggest that phosphorylation of STAT-3 is essential for IPostC cardioprotection.

ANOMALOUS ORIGIN OF THE LEFT SUBCLAVIAN ARTERY FROM THE MAIN PULMONARY ARTERY ASSOCIATED WITH A LARGE PATENT DUCTUS ARTERIOSUS

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Background: Anomalous origin of a subclavian artery from the pulmonary artery (also termed isolated subclavian artery) is a rare congenital anomaly described in only a few case reports. The isolated subclavian artery fills in a retrograde fashion as blood flows around the circle of Willis and down the vertebral artery, which produces a congenital subclavian steal phenomenon.

We report on a 9 year old boy with a large hypertensive patent ductus arteriosus (PDA), a right sided aortic arch and an isolated left subclavian artery, arising from the main pulmonary artery (MPA).

Case report: A 9-year-old boy was referred to our centre for evaluation of a PDA. He had been evaluated at the referring hospital for developmental delay, left hemiparesis and mental retardation. A cardiac murmur was heard incidentally.

On examination he weighed 16 kg and was small for his age. He had minor dysmorphic features of hypertelorism, low set ears, microcephaly and frontal bossing. He was cyanosed on the left arm with saturations of 85% with early digital clubbing. He was pink centrally and in all other limbs with saturations of 95% in the right arm and both feet. His pulse was 100/min and the BP was 100/40 mmHg. There was cardiomegaly and a left precordial bulge. The second heart sound was loud. A 3/6 ejection systolic murmur was audible in the left infraclavicular area.

Chest radiography revealed cardiomegaly with plethoric lung fields and a right sided aortic arch. Electrocardiography showed a heart rate of 100/min. The QRS axis was -60 degrees and there was evidence of left ventricular hypertrophy. Echocardiography demonstrated a large PDA measuring 10 mm, with left to right shunting. The pulmonary arteries were dilated. There was no VSD or ASD. Cardiac catheterisation and angiography confirmed a large PDA with severe, but reversible pulmonary hypertension. In addition the left subclavian artery was seen arising from the MPA.

Conclusion: We describe this unusual case of an isolated left subclavian artery, arising from the MPA, associated with a large PDA and we give a general overview on this condition.

PROFILE AND HIGH PREVALENCE OF CARDIOVASCULAR RISK FACTORS IN AN URBAN BLACK AFRICAN POPULATION: UPDATE ON HEART AWARENESS DAYS IN SOWETO

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Background: While cardiovascular disease (CVD) in Africa is mostly non-ischaemic in origin, the incidence of coronary heart disease in sub-Saharan



Africa is on the increase. As a subset to the much larger 'Heart of Soweto' study, this study set out to raise awareness on CVD risk factors; and establish a baseline profile of CVD risk in the population of Soweto, South Africa.

Methods: All persons presenting to a fixed monitoring stand in Soweto, the largest black residential area in South Africa. Subjects were briefly interviewed on their demographic status, prior history of documented CVD, diabetes, or a family history thereof; and on their own behavioural risk factor history or tobacco and alcohol use. Blood pressure, waist circumference, height and weight (used to measure BMI) were measured, and blood tested for random blood glucose and cholesterol.

Results: A total of 1 691 participants were screened. 80% of the population had at least one major risk factor for CVD, 70% having high BMI and 43% being severely overweight. One third of screened persons had at least one component of blood pressure raised, while 13% had raised blood cholesterol levels. Being overweight was significantly associated with elevated blood pressure, and raised serum glucose and cholesterol levels.

Conclusions: The study findings strongly suggest a high prevalence of risk factors for CVD in this urban black African population. There is a need for proper sampling to better depict the risk factor profile.

THE INCIDENCE OF ACUTE RHEUMATIC FEVER IN THE WORLD: A SYSTEMATIC REVIEW OF POPULATION-BASED STUDIES

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Background: Acute rheumatic fever is a multi-organ disease resulting from an autoimmune response of the body to infection with Lancefield Group-A β haemolytic streptococci (GAS). Overall, industrialized countries have experienced a declining incidence of acute rheumatic fever over the past 100 years. However, despite evidence of the effectiveness of antibiotic treatment of GAS pharyngitis in reducing the incidence of ARF, developing countries continue to experience a high burden of the disease and its chronic sequel, rheumatic heart disease.

Aim: To summarise data from population-based studies on the magnitude of, and temporal trends in, the incidence of acute rheumatic fever.

Method: We conducted a comprehensive search of MEDLINE, EMBASE, and other health-related databases identifying all published prospective population-based studies of the incidence of acute rheumatic fever that fulfilled pre-specified inclusion and exclusion criteria. We critically reviewed each study assessing both mean incidence rate of first attack of acute rheumatic fever per year (calculated over the entire study period for each study), and annum specific incidence rate (for those studies documenting incident cases specific to each year of study).

Results: Our review included 10 eligible studies conducted in 10 different countries (none in Africa) The overall mean incidence rate of first attack of acute rheumatic fever per year for each study ranged from 5 to 51 per 100 000 population (mean 19 per 100 000; 95% CI 9 - 30 per 100 000). A low incidence of acute rheumatic fever of ≤ 10 per 100 000 per year was found in North and South America, and in Northern Europe. There was a high incidence of >10 per 100 000 in Eastern Europe, Middle East, Asia, and Australasia. Annum-specific incidence rates were higher in the Middle East than in other regions. There was a fall in the incidence of acute rheumatic fever over time in all countries with longitudinal data [Figure 1].

Conclusions: There has been a modest decline in the incidence of acute rheumatic fever over time globally, however, the disease still occurs relatively frequently in Eastern Europe, the Middle East, and Australasia. There are no population-based incidence studies of acute rheumatic fever in Africa, a continent that bears the highest number of cases of rheumatic heart disease in the world.

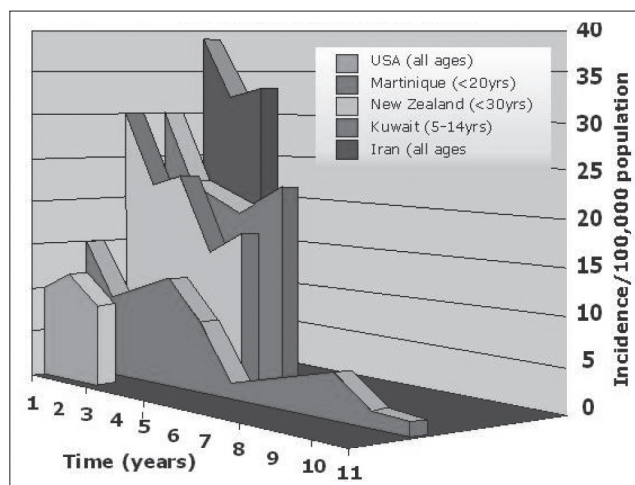


Fig. 1. Trend in incidence of first attack acute rheumatic fever over time.

A RETROSPECTIVE ANALYSIS OF CONTRAST INDUCED NEPHROPATHY (CIN) IN 501 CONSECUTIVE PATIENTS UNDERGOING CORONARY ANGIOGRAMS DONE IN A SECONDARY CENTRE

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CIN is a recognised complication of coronary angiography known to carry an adverse prognosis. 501 sequential patients undergoing Coronary angiography were analysed (2.5 years to six weeks post procedure). Age range <40 to mid-80s. Iodixanol (Visipaque) was used for all procedures, and a simple fluid regime protocol is used depending on diabetic/pre-procedure renal function (full details will be presented), etc. Follow-up investigations were done on Day 6 post procedure. Complete data were available for 485 of the patients. The renal function of the other patients was checked subsequently and was found to be normal. Two patients were dead at the time of analysis (mortality 0.4%). None died from CIN (or of subsequent cardiac surgery or PCI). One died the night after the procedure of AMI, not felt by the pathologist to be related to the procedure. One patient with significant co-morbidities was readmitted 4 days later with pneumonia and subsequently developed multi-organ failure and died.

There was a 2/485 (0.41%) incidence of CIN using the a stringent definition ($>25\%$ decrease in GFR, Creatinine increase >0.044 mmol/l). No patients required dialysis post procedure. The mean change in GFR was -1.08 ml/min. A non-significant greater change was noted in Maori patients. There were no correlations observed between age, race, pre-angiography creatinine clearance/function, diabetes, contrast volume and deterioration in renal function as assessed by creatinine clearance or serum creatinine.

Conclusions: A simple fluid regime is safe and effective with no evidence of CIN in a large sample of patients using this contrast agent.

AN UNUSUAL CASE OF PERICARDIAL EFFUSION

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Case report:

- A 2-year-old male presented with shortness of breath, a large pericardial effusion with tamponade, extensive bronchopneumonic changes and a hepato-splenomegaly.
- Over a period of 6 weeks, he underwent several procedures:
 1. Percutaneous and surgical pericardial drainage



2. A right limited thoracotomy for a right pleural effusion + pericardial window
 3. Pericardiectomy + partial thymectomy.
- Turbid fluid from the pericardial and pleural cavity was high in protein, LDH and triglycerides.
 - Despite surgical and medical interventions, the chylous drainage continued.
 - Chyloptysis was present in the tracheal aspirate. He demised about 10 weeks post admission from respiratory failure.
 - The final diagnosis was made at post-mortem. Lymphangiomatosis was present in the lungs, thymus, spleen and the hilum of thoracic lymph nodes.

Discussion:

- Lymphangiomatosis is a congenital anomaly of the lymphatic system.
- Although histologically a benign condition, it has the propensity to infiltrate bone and soft tissue resulting in life-threatening complications.

OUTCOME FOR PALLIATIVE SURGERY IN CONGENITAL HEART DISEASE

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Introduction:

- Palliative procedures constitute between 11-12% of paediatric congenital heart surgery cases performed at Johannesburg Hospital.

Objectives:

- To ascertain the age related and procedure related hospital mortality rates.
- To evaluate the follow-up with respect to out-patients review and further procedures.

Methods:

- A retrospective study conducted over a 10 years period between 1995 and 2004.
- The procedures were classified in 3 groups: 1 – shunts, 2 – pulmonary artery banding, 3 – miscellaneous procedures.
- Each procedural group was further subdivided according to pathology.
- Each group was subdivided into the following age categories: neonatal, infants, older children.

Results:

- A total of 158 patients was analyzed.

EARLY RESULTS OF BIVENTRICULAR REPAIR OF HYPOPLASTIC LEFT HEART COMPLEX OR SEVERE AORTIC ARCH HYPOPLASIA WITH VENTRICULAR SEPTAL DEFECT

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Background: A selected group of patients with hypoplastic heart complex (HLHC) and those with severe aortic hypoplasia and VSD (Aohy/VSD) may be candidates for biventricular repair instead of univentricular repair and ultimately a Fontan circulation.

Methods: Since July 2004 4 patients with HLHC and 5 with Aohy/VSD underwent biventricular repair. Z-value for Mitral valve and Aortic Valve ranged from -3 and -4.5 for HLHC whereas 4 patients with Aohy/VSD had aortic annular diameters 3mm or smaller. Repair for HLHC consisted of patch aortoplasty of the aortic arch and ascending aorta with ASD fenestration. Two patients underwent additionally unroofing of the coronary sinus. Repair for Aohy/VSD consisted of a Norwood-Rastelli procedure as a single stage approach in 5 patients.

Results: Hospital survival was 100%. Postoperatively no ECMO or assist device was needed.

At follow-up up to 27 months no late mortality occurred. Reoperation has been necessary in 4 patients. In HLHC patients reasons for reoperation were supra-avalvular mitral ring stenosis, supra-avalvular aortic stenosis, recoarctation and in one patient with Aohy/VSD systemic venous stenosis. Clinical condition of all patients is good except for one.

Conclusions: In this difficult subset of patients successful biventricular repair was achieved. Short term follow-up supports our policy to preserve biventricular physiology when possible.

ALDOSTERONE-TO-RENIN RATIO IS AN INDEPENDENT PREDICTOR OF BLOOD PRESSURE IN A GENERAL POPULATION OF SUBJECTS OF AFRICAN ANCESTRY

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As hypertension in subjects of African descent is associated with a low-renin state and an impaired blood pressure (BP) response to antihypertensive agents that target the renin-angiotensin system (RAS), the role of the RAS in hypertension in this ethnic group is in question. As part of the African Program on Genes in Hypertension, we evaluated the relationship between RAS activity and both conventional and ambulatory BP in a randomly selected adult (>16years) population of African ancestry. Of 671 subjects recruited, 265 had plasma renin and aldosterone concentrations determined after lying for 15 minutes, as well as 24-hour urine (complying to pre-specified quality control criteria) electrolyte excretion values, and 222 also had acceptable ambulatory BP measurements. Plasma renin was negatively related to conventional (partial $r = -0.26, p < 0.0001$), and 24-hour (partial $r = -0.14, p < 0.05$) diastolic BP after adjustments for age, sex, antihypertensive treatment, total cholesterol concentrations, waist circumference and urinary Na^+/K^+ ratios. On sensitivity analysis conducted in untreated subjects, plasma renin was related to conventional ($p < 0.02$), but not 24-hour diastolic BP after adjustments for confounders. Modest relationships between plasma aldosterone and systolic BP were noted after adjustments ($p < 0.05$). However, the aldosterone-to-renin ratio was positively associated with both conventional (partial $r = 0.32, p < 0.0001$) and 24-hour (partial $r = 0.19, p < 0.01$) systolic BP after adjustments for confounders, data confirmed in untreated subjects. Moreover, the aldosterone-to-renin ratio was independently and positively associated with both conventional (partial $r = 0.25, p < 0.0001$) and 24-hour (partial $r = 0.16, p < 0.01$) systolic BP after adjustments for confounders as well as plasma renin and aldosterone, data confirmed in untreated subjects. The aldosterone-to-renin ratio was second only to age in explaining the % variability in both conventional and 24-hour systolic BP. In conclusion, these are the first substantial data indicating that RAS activation downstream from renin is a critical determinant of BP in a population of African ancestry.

A DECREASE IN HEART PERIOD VARIABILITY AS AN INDICATOR OF ISCHAEMIC HEART DISEASE

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Introduction: Autonomic dysfunction is characteristic of chronic heart failure. Change in heart period variability (HPV) is an indicator of autonomic dysfunction. This study focused on HPV and arterial blood pressure (ABP) as indications of myocardial ischaemia during an electrocardiographic (ECG) stress test.

Methods: ABP and heart rate (HR) were recorded on three groups of subjects: a healthy group, a group on anti-hypertensive medication, and an ischemic group on anti-hypertensive medication. The measurements were



done before, during and after a stress ECG. The HPV was analysed with respect to the total power of the frequency components.

Results: Both groups on anti-hypertensive medication showed a decrease in HPV when compared to the healthy group. Comparing the groups on anti-hypertensive medication, there was a decrease in HPV in the group diagnosed with IHD. ABP increased with exercise in both groups on anti-hypertensive medication but not in the healthy group.

Conclusion: Patients with IHD could further be monitored using HPV as an indicator of IHD.

VALVE THROMBOSIS – THE INFLUENCE OF VALVE DESIGN AND THE ROLE OF ANTICOAGULANTS

M A Williams

Introduction: Valve thrombosis is a catastrophic and often fatal complication of mechanical valve replacement. This complication is especially common in poorly anticoagulated communities. This study was undertaken to assess the role of anticoagulation and the influence of valve design in six different prostheses in a poorly anticoagulated population.

Methods: Data relating to the performance of six valves was studied. Information gathered focused on the incidence of valve thrombosis and the mechanism of thrombosis in the different designs.

Results: The incidence of valve thrombosis was higher than that recorded in well anticoagulated patients.

Valve	Number of patients	Clotted valves	Anticoagulation
Lillehei-Kaster	189	18	poor
Bjork-Shiley	224	24	poor
St Jude	2000	41	30% anticoagulated
Medtronic-Hall	122	9	40% anticoagulated
Carbo-medics	82	14	40% anticoagulated
On-X	710	3	50% anticoagulated

Discussion: Most of our patients come from the third world part of the population of South Africa. Poverty, ignorance and geographic dispersion make the control of anticoagulation very difficult. In this group of patients one would expect a higher incidence of valve related complication especially thrombosis and thromboembolism.

Poor anticoagulation coverage unmasks the true thrombogenic potential of the different valves, highlighting design defects. On examination of the explanted valves it was apparent that the mechanism of thrombosis was different - turbulence, stasis and pannus ingrowth playing different roles.

Conclusion: Our experience with the On-X valve has been rewarding with a significantly lower incidence of valve thrombosis. In our view the low incidence of thrombotic complications is related to innovative design features. It is currently our valve of choice in patients where anticoagulation is uncertain.

CLINICAL, RISK FACTOR AND ANGIOGRAPHIC PROFILE OF YOUNG PATIENTS WITH ACUTE MYOCARDIAL INFARCTION – APOLLO EXPERIENCE

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Objective: The objective of this study was to evaluate clinical, risk factor and

angiographic profile of young patients (<40 years) with acute myocardial infarction (AMI), as they have a different profile and prognosis to older patients.

Methods: We included 30 patients (<40 years) with AMI admitted to our Hospital, who were also evaluated for risk factors such as a positive family history of premature coronary artery disease (CAD), smoking, hypertension, diabetes, dyslipidaemia, hyperhomocysteinemia and elevated lipoprotein(a) levels, and who also underwent coronary angiography.

Results: Of the 30 patients (2 females) included in this study, the youngest was a 21 year old male patient. The mean age of patients was 33.75±3.4 years. Six (40%) had a positive family history of premature CAD, 16 (53%) were smokers, 3 (10%) had systemic hypertension, 1 (3.3%) had diabetes, 22 (73%) had dyslipidemia (15 had high low density lipoprotein (LDL) cholesterol, 14 had low high density lipoprotein (HDL) cholesterol, 11 had hypertriglyceridemia). Hyperhomocysteinemia was seen in 20 (66%) patients (mild (>15-30 µmol/l) in 8 patients, moderate (>30 µmol/l) in 12 patients). Elevated lipoprotein(a) levels (>30mg/dl) were seen in 11 (36.6%) patients. 24 patients (80%) had anterior wall myocardial infarction (MI), 5 (16.6%) had inferior wall MI and 1 (3.3%) had posterolateral MI. Results of coronary angiography showed that 20 patients (66%) had single vessel disease (LAD 17 patients, RCA 3 patients). Of these, 12 (60%) patients had a recanalized LAD, 4 (13.3%) patients had double-vessel disease, and 2 (6.6%) patients had triple-vessel disease.

Conclusions: Dyslipidemia, smoking and positive family history of premature CAD were the major conventional risk factors in these young patients with AMI. Diabetes and hypertension were less common than the older patients. Also, non-conventional risk factors like hyperhomocysteinemia and elevated lipoprotein(a) levels were seen in this younger subset of patients with AMI. Anterior wall (80%) was the commonest site affected by MI. Single vessel disease (66%) was common and LAD (85%) was the most common culprit vessel, followed by RCA. Multi-vessel disease was more commonly seen in the relatively older age group patients, 36 to 40 years of age. The majority of patients (80%) had fair left ventricular function. This subset of young patients with AMI has a good prognosis overall, compared with older patients.

ALCAPA: FROM CRITICAL TO CURED

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an uncommon congenital heart defect occurring in 1:300 000 live births. It has the unique distinction of a mortality rate of 90% when undiagnosed in infancy yet if diagnosed and repaired timeously, yields excellent survival and functional outcome results.

Although known since the 1800's, many challenges remain in the management of patients with this condition, not least the early recognition of the anomaly. Currently there are various diagnostic modalities available to the cardiologist. Peri-operative strategies and surgical techniques have evolved over time. Children represent a particular challenge as they can present critically ill with profound LV dysfunction, even requiring mechanical support as part of their management.

To assess our diagnostic accuracy, medical and surgical management of these patients as well as reviewing the long-term results after repair, a retrospective review of the 23 patients diagnosed with ALCAPA since 1997 was undertaken.



The average length of time until definitive diagnosis ranged from 11.5 years to immediately at the initial echocardiogram. Only 13/23 patients were catheterised pre-operatively. Of the 18 patients who eventually had surgery, half had a direct re-implantation of the coronary artery and half a Takeuchi repair. Five patients died – 4 early and one late death. Follow-up confirmed excellent outcomes with 90% of patients having normal ejection fractions.

Despite a worse initial presentation in children, timeous ALCAPA repair offers the best potential for full LV functional recovery.

OF LICE AND MEN: A CASE OF BARTONELLA QUINTANA

Liesl Zühlke, Rik De Decker, John Hewitson, Neil Seller, Andrew Whitelaw, John Lawrenson

Bartonella quintana, a pathogen that is restricted to human hosts and louse vectors, has been known as the causative agent for Trench fever since 1915. Now reemerging as an agent of disease, it is implicated in a variety of conditions amongst homeless and alcoholic populations, including septicaemia and endocarditis. Endocarditis is often blood culture negative

and confirmation of the diagnosis requires the adjuvant use of serologic testing and more recently molecular diagnostics.

We present an unusual case of a 12-year-old girl, presenting for routine closure of a PDA. On examination, she had a firm splenomegaly and was pale with an haemoglobin of 5g/dl. Echocardiogram showed extensive vegetations in the PDA with extension into the MPA. Blood cultures remained negative after 21 days and serology (ACW1) did not reveal a causative agent. We elected to perform an endoarterectomy and closure of the PDA. Parts of the vegetation excised as surgery were sent for further testing. DNA was extracted from the vegetations and a portion of the 16SrRNA gene was amplified by PCR. The amplification products were sequenced and analysis of the sequence results showed 99% homology with published sequences of *Bartonella quintana*.

This case highlights the use of broad-range bacterial PCR and DNA sequencing in elucidating causes of culture negative endocarditis. This is also, as far as we can ascertain, the first case of PDA endarteritis caused by *Bartonella quintana*.