

complications and improvement of acute and mid-term hemodynamic results. However, possible complications mandate careful patient selection and restriction to experienced centers.

Table: Advantages and potential drawbacks of percutaneous septal ablation and surgical myectomy

Percutaneous septal ablation (PTSMA)	Surgical myectomy
<p>Advantage:</p> <ul style="list-style-type: none"> ◆ Avoidance of cardiopulmonary bypass with attendant risks - Elderly patients with concomitant non-cardiac disease ◆ Treatment of patients with isolated mid-cavitary or combined subaortic and mid-cavitary obstruction ◆ Short hospital stay ◆ Short recovery time ◆ Lower costs 	<p>Advantage:</p> <ul style="list-style-type: none"> ◆ Immediate and complete relief of resting and provoked obstruction and concomitant mitral regurgitation ◆ Documented long-term results up to 30 years ◆ Ability to treat coexistent cardiac disease - Coronary artery disease, valve disease ◆ Additional treatment of papillary muscle in extended myectomy
<p>Potential drawbacks:</p> <ul style="list-style-type: none"> ◆ Risk of damage to the left coronary artery with - Emergency bypass surgery or left main / LAD stenting ◆ Technical impossibility of reaching or identifying a target septal branch ◆ Lower success in patients with - Mitral valve leaflet and papillary muscle abnormalities - Large septal thickness (younger patients) 	<p>Potential drawbacks:</p> <ul style="list-style-type: none"> ◆ Necessity of high individual surgical experience - High surgical mortality in unexperienced centers ◆ Low risk of postoperative aortic regurgitation ◆ LV deterioration after extended myectomy during long-term follow-up, possibly due to - High incidence of left bundle branch block ◆ More invasive approach requiring extracorporeal circulation

NOTES BY SHARON BATES

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H. Seggewiss – Improve quality of life with no negative affects on life. Echo guided septal ablation has improved procedures over the years. Placement of target vessel is the key to correct & successful ablation. Keep patients in hospital up to 8 days due to increase of gradient for day 3 and reduced by day 7 and then more at 6 months and 1 year.

12:32 Restorative tailored surgical approach
M. Yacoub (London, UK)

LECTURE ABSTRACTS

LEFT VENTRICULAR ASSIST DEVICE AND DRUG THERAPY FOR THE REVERSAL OF HEART FAILURE

MAGDI YACOUB

Email: m.yacoub@imperial.ac.uk

E.J. Birks, P.D. Tansley, J. Hardy, R.S. George, C.T. Bowles, M. Burke, N.R. Banner, A. Khaghani, M.H. Yacoub; Royal Brompton and Harefield National Health Service Trust, Harefield Middlesex. United Kingdom

Background: In patients with severe heart failure, prolonged unloading of themyocardium with the use of a left ventricular assist device has been reported to lead to myocardial recovery. In small numbers of patients for varying periods of time. Increasing the frequency and durability of myocardial recovery could reduce or postpone the need for subsequent heart transplantation.

Method: We enrolled 15 patients with severe heart failure due to nonischemic cardiomyopathy and with no histologic evidence of active myocarditis. All had markedly reduced cardiac output and were receiving inotropes. The patients underwent implantation of left ventricular assist devices and were treated with lisinopril, carvedilol, spironolactone, and losartan to enhance reverse remodeling. Once regression of left ventricular enlargement had been achieved, the beta2-adrenergic-receptor agonist clenbuterol was administered to prevent myocardial atrophy.

Results: Eleven of the 15 patients had sufficient myocardial recover to undergo explanation of the left ventricular assist device a mean (\pm SD) of 320 \pm 186 days after implantation of the device. One patient died of intractable arrhythmias 24 hours after explanation: another died of carcinoma of the lung 27 months after explanation. The cumulative rate of freedom from recurrent heart failure among the surviving patients was 100% and 88.9% 1 and 4 years after explantation, respectively. The quality of life

as assessed by the Minnesota Living with Heart Failure Questionnaire score at 3 years was nearly normal. Fifty-nine months after explantation, the mean left ventricular ejection fraction was $64 \pm 12\%$, the mean left ventricular end-systolic diameter was 42.5 ± 13.2 mm, and the mean maximal oxygen uptake with exercise was $26.3 + 6.0$ ml per kilogram of body weight per minute.

Conclusions: In this single-center study, we found that sustained reversal of severe heart failure secondary to nonischemic cardiomyopathy could be achieved in selected patients with the use of a left ventricular assist device and a specific pharmacologic regimen.

N. Engl J Med 2006; 355: 1873-84

NOTES BY SHARON BATES

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M. Yacoub – Alcohol vs. surgery (the pendulum swings back to surgery) diagrams of the various flow and function believe the left ventricular and right ventricular chambers. Real pictures of the myectomy and exact reduction portion. Some re-build anterior and posterior leaflet, papillary muscle, abnormalities, accessory bands, and other issues. Some valve replacement has been written about but Dr. Yacoub feels this is unnecessary and reduces life span of patients.

Panel discussion all speakers – Drs. Ommen, Seggewiss, & Yacoub

NOTES BY SHARON BATES

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Panel - Is there are reduction of skills to provide myectomies in Germany? Dr. Seggewiss – patients are getting myectomies done at expert centers, but this is the patients choice to have surgery done at home center which can lead to less morality. In both techniques skill and training is extremely important for patients and long term value of HCM research / longevity of doctor skills.

Do you thing pediatric surgeons / backgrounds are more appropriate to skill? Yes at some but not critical advantages for knowledge of CHD but again not critical. Knowledge can be gained at adult level as long as the surgeon is not only a bi-pass surgeon.

What is the average weight of tissue removed from extended myectomy in grams? Varies based on size of septum. Make sure it is a tailored approach.

13:30 Lunch Break

NOTES BY SHARON BATES

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Spoke to Dr. Barry Maron about reviewing the ABF poster #8 to answer his statement on improving questionnaires in correlation to find more HCM patients.

15:00 **Session 5**

16:36 **LV dysfunction, dilated Cardiomyopathy and ARVC**

Chairmen: **C. Ho** (Boston, USA), **L. Tavazzi** (Pavia, I)

CAROLYN HO

Cardiovascular Division, Brigham and Women's Hospital, Boston, USA

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LUIGI TAVAZZI

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15:00 Genetic screening in the clinical setting: state-of-the-art
W.J. McKenna (London, UK)

LECTURE ABSTRACTS

GENETIC SCREENING IN THE CLINICAL SETTING: STATE-OF-THE-ART

WILLIAM J MCKENNA

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DCM and ARVC are inherited autosomal dominant cardiomyopathies. Both exhibit age-related penetrance and the majority of gene carriers show incomplete disease expression. In DCM, mutations in sarcomere, cytoskeletal and lamin A/C genes cause disease, through precise prevalence data are not available. In ARVC, mutations in cell adhesion genes (plakoglobin, plakophilin, desmoplakin, desmoglein, desmocollin) account for approximately 3-50% of disease.

Familial evaluation is recommended in both DCM and ARVC. The prevalence of familial disease is approximately 50%. In DCM, initial presentation with a major complication is unusual. Mutation analysis facilitates familial evaluation with identification of "at risk" individuals. In addition, particular mutations (e.g.

lamin A/C) are associated with conduction disease and risk of lethal ventricular arrhythmias, both of which may develop at what appears to be an early state of disease. In ARVC, diagnosis requires a number of investigations. Identification of family members who carry disease causing mutations and are at risk of disease complications is particularly important, as sudden death may be the presenting disease feature. Genotype phenotype studies are in the infancy. Left ventricular forms are associated with desmoplakin mutations, while severe forms of ARVC often are associated with compound heterozygosity/homozygosity. Cardiocutaneous syndromes have been described in recessive ARVC families with plakoglobin and desmoplakin mutations.

NOTES BY SHARON BATES

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W. McKenna - Now generally known as a genetic source / not viral studies showed where 20 – 25% have familial expression. What trips the DCM patient into CHF is a virus, pregnancy, salt intake, or alcohol – not just that a virus attacked the heart that DCM was present and slow and insidious to become symptomatic or crash.

Mutation evaluation of DCM - List of various types of genes is a huge challenge to analyze the conductors. Lamin A/C phenotype – up to 60% will test positive. Usually Rx of ace inhibitors instead of Beta blocker 5 yearly adult screening (20-40) over 40 are more likely not to get the condition. ARVC diagnosis rests on myocarditis cell death. Clinical diagnosis is very hard to make, scoring system makes it difficult to identify unless there is already familiar connection. Cell adhesion genes have also been identified to arise at helpful diagnosis. Late enhancement has helped uncover some other types of ALVC. ARVC – family history is important, pick up people early link disease progression and sudden death to exercise more than other cardiomyopathies, beta blockers is first pharma with life style counseling.

15:48 Update on ARVC: morphological spectrum and outcome
C. Basso (Padua, I)

LECTURE ABSTRACTS

UPDATE ON ARVC: MORPHOLOGICAL SPECTRUM AND OUTCOME

CRISTINA BASSO

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Arrhythmogenic right ventricular Cardiomyopathy (ARVC) is a myocardial disease mostly affecting the right ventricle (RV) and characterized by ventricular electrical instability. ARVC is nowadays defined as a disease of cell junctions, since both in the recessive and dominant forms pathogenic mutations have been identified in genes encoding for intercellular junction proteins.

The replacement of the RV myocardium by fibro-fatty tissue is progressive, starts from the epicardium or mid-myocardium and extends downward to become transmural, thus accounting for wall thinning and the development of aneurysms, the gross hallmark of ARVC. Left ventricular involvement, usually confined to be subepicardial layers the postero-lateral free wall, is present in more than half of the hearts studied at post-mortem or after cardiac transplantation. Three main distinct clinical-pathologic patterns are nowadays recognized: 1) the classical RV phenotype, with isolated RV disease or minimal left ventricular involvement in association with significant RV involvement; 2) the left dominant phenotype, with early and prominent left ventricular manifestations and relatively mild RV disease; and 3) the biventricular phenotype, characterized by equal involvement of both ventricles.

Histological examination reveals islands of surviving myocytes interspersed with fibrous and fatty tissue, ongoing myocyte death (either apoptosis or necrosis) and frequent inflammatory infiltrates. It is still unknown whether this is a reactive phenomenon to cell death or the consequence of infective or immune mechanisms. Ultrastructural investigation in gene-positive ARVC patients revealed intercalated disk remodeling with desmosomal abnormalities and intercellular gap widening. Impaired mechanical coupling may account for impaired electrical coupling by gap junction remodeling. Rather than being a continuous process, disease progression may occur during periodic “burst” which may be characterized by life-threatening arrhythmic storms. Environmental factors such as exercise or inflammation may worsen impaired cell-to-cell adhesion and trigger disease progression. Cardiac arrest due to ventricular fibrillation may occur any time during the disease course, and the occurrence of sudden death varies from 0.1 to 3% per year in adults with diagnosed ARVC, but may be higher in the young, when the disease is still undiagnosed and sudden death can be the first manifestation. More exceptional presentations are isolated RV and / or biventricular heart failure as to mimic dilated Cardiomyopathy and to require cardiac transplantation.

NOTES BY SHARON BATES

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C. Basso – Fibro / fatty replacement & arterial wall (right ventricular) free wall some misdiagnosis of

chronic myocarditis as there have been studies showing inflammation / secondary to the fibro / fatty replacement. Get the “Guidelines for Autopsy” from the author for PHW.

16:12 Clinical management of ARVC

B. Bauce (Padua, I)

LECTURE ABSTRACTS

CLINICAL MANAGEMENT OF ARVC

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Arrhythmogenic right ventricular cardiomyopathy is a myocardial disease of genetic origin. In the last few years several genes linked to the disease have been identified, the majority of whom encoding for intercellular junction proteins. Results of genotype – phenotype studies in ARVC families should help to establish appropriate clinical management in mutations carriers.

In our experience the detection of a genetic mutation in a proband or family member may lead to different management problems depending on circumstances as described below: 1) presence of an ARVC genetic mutation in an affected proband. The proband usually presents an overt form of the disease and genetic result leads only to a confirmation of the disease diagnosis but it does not modify the clinical management that is related to the extent of the disease and degree of electrical instability; 2) detection of a genetic mutation in a subject, usually a family member, with no clinical signs of disease. This situation can be due to the young age of the subject who has not yet developed the disease or to the fact that the mutation is characterized by a low expression. There are no guidelines for management of these healthy carriers of the disease. Since a myocardial stretching may facilitate the onset and progression of the disease, in these kind of subjects we allow only a limited physical activity without isotonic efforts, whereas a competitive physical activity is always forbidden; 3) identification of a genetic mutation in a subject with minor signs of the disease. In this case we can assume that a mild anatomic abnormality is present. This patient may be asymptomatic and in this case we allow only a mild physical activity with a systematic clinical evaluation. The detection of ventricular arrhythmias has lead the physician to start an antiarrhythmic therapy and to suggest the avoidance of physical activity; 4) identification of a genetic mutation in family members with an overt form of the disease (previously unknown); in this cases there is a strong correlation between presence of a gene mutation and disease expression, thus the clinical management will be the same as that of the proband. In subjects classified as affected, physical activity must be prohibited, both in competitive as well as non competitive sports. In conclusion, advances in the genetics of ARVC has not helped the clinical management of the affected patients, as therapeutic decisions relate to clinical features and degree of electrical instability. At this time, we are not able to modify the disease progression with certainty. However, genetic analysis of family members allows the early detection of subjects risk of life-threatening ventricular arrhythmias and sudden death. In these subjects it is important to erase those factors that theoretically favor the onset to the clinical disease. We do not have a clear knowledge on pejorative factors, even if we hypothesize that myocardial cells stretching due to strenuous physical activity can favor the onset of the pathologic process. In these subjects physical activity has to be limited or prohibited; moreover, they must be followed systematically with frequent non invasive clinical evaluations aim of promptly identifying the onset of the arrhythmias and to prevent serious life threatening ventricular arrhythmia by antiarrhythmic therapy.

NOTES BY SHARON BATES

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B. Bauce – Family history, physical exam (12-lead, signal ECG, 24 hour monitors) echo, etc. Risk stratification must be done. Familiar disease, must analyze the family. Genetic tests must be done on all mutations due to compound genotype – phenotype. Not so rare, it causes more risk. Genetics just confirm diagnosis. But for genetics in family, no clinical signs of disease and reduces risk of sudden death, concern on limiting strenuous physical activity which progresses the disease over time. Also allows the good news to be given to family members genetically negative. Use of electrical instability to determine extent and management of disease.

16:36 Coffee Break

NOTES BY SHARON BATES

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Spoke to Dr. Basso, and asked fro the study on “Autopsy” to share with pathologists and parents in the US. Also, Dr. Cecchi invited me to attend the faculty dinner at the remainder of today’s sessions.

17:00 **Session 6**

18:12 **Restrictive cardiomyopathies and children**

Chairmen: **E. Arbustini** (Pavia, I), **M. Yacoub** (London, UK)

ELOSISA ARBUSTINI

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MAGDI YACOUB

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17:00 Cardiomyopathies in children including LV non compaction
J.A. Towbin (Houston, USA)

LECTURE ABSTRACTS

EPIDEMIOLOGY AND CAUSE-SPECIFIC OUTCOME OF HYPERTROPHIC CARDIOMYOPATHY IN CHILDREN: FINDINGS FROM THE PEDIATRIC CARDIOMYOPATHY REGISTRY

JEFFREY A. TOWBIN

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*S.D. Colan, S.E. Lipshultz, A.M. Lowe, L.A. Sleeper, J. Messere, G.F. Cox, P.R. Lurie, E.J. Orav, J.A. Towbin
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Background: Current information on the epidemiology and outcomes of Hypertrophic Cardiomyopathy (HCM) in children is limited by disease diversity and small case series.

Methods and results: The Pediatric Cardiomyopathy Registry has collected prospective and retrospective data on children diagnosed with HCM since 1990. We identified the various causes of HCM in childhood and determined the relationship between outcome, cause, and age at presentation. Of 855 patients < 18 years of age with HCM, 8.7% (n = 74) had inborn errors of metabolism, 9.0% (n = 77) had malformation syndromes, 7.5% (n = 64) had neuromuscular disorders, and 74.2% (n = 634) had idiopathic HCM. Children with HCM associated with inborn errors of metabolism and malformation syndromes have significantly worse survival than the other 2 groups. Patients with idiopathic HCM diagnosed before 1 year of age (n = 227) had worse survival from the time of diagnosis than those diagnosed after 1 year of age (n = 407). Patients with idiopathic HCM who survived to at least 1 year of age, however, had an annual mortality rate of 1% that was similar regardless of whether they were diagnosed before or after 1 year of age. **Conclusion:** In children, HCM is a diverse disorder with outcomes that depend largely on cause and age. Patients presenting before 1 year of age have the broadest spectrum of causes and the poorest outcome. In those children with idiopathic HCM who survive beyond age 1, however, survival is independent of age at diagnosis, with an annual mortality rate (1%) that is much lower than previously reported in children and is not different from has been found in population-based studies in adults.

Circulation 2007; 115: 773-781

LECTURE ABSTRACTS

CLINICAL CHARACTERIZATION OF LEFT VENTRICULAR NONCOMPACTION IN CHILDREN: A RELATIVELY COMMON FORM OF CARDIOMYOPATHY

JEFFREY A. TOWBIN

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S.Clunie, S. Fernbach, N.E. Bowles, J.A. Towbin
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Background: Left ventricular noncompaction (LVNC) is a reportedly uncommon genetic disorder of endocardial morphogenesis with a reportedly high mortality rate. The purpose of this study was to identify the clinical characteristics of children with LVNC. **Methods and results:** We retrospectively reviewed 36 children with LVNC evaluated at Texas children's Hospital (TCH) from January 1997 to December 2002. Five children had associated cardiac lesions. There were 16 girls and 20 boys. The median age at presentation was 90 days (range, 1 day to 17 years). The median duration of follow-up was 3.2 years (range, 0.5 to 12 years). Twenty-seven patients (75%) had ECG abnormalities, most commonly biventricular hypertrophy (10 patients, 28%). Both ventricles were involved in 8 patients (22%) and only the left ventricle in 28 patients (78%). Left ventricular systolic function was depressed in 30 patients (83%), with a median ejection fraction of 30% (range, 15% to 66%) at diagnosis. Nine patients presenting in the first year of life with depressed left ventricular contractility had a transient recovery of function: however, ejection fraction deteriorated later in life, at a median interval of 6.3 years (range, 3 to 12 years). Two patients had an "undulating" phenotype from dilated to hypertrophic cardiomyopathy. Two patients (6%) were identified with an underlying G4.5 gene mutation. Five patients (14%) died during the study. **Conclusion:** LVNC does not have an invariably fatal course when diagnosed in the neonatal period. A significant number of patients have transient recovery of function followed by later deterioration, which

may account for many patients presenting as adults, some manifesting an “undulating” phenotype.

Circulation 2003; 108: 2672-2678

NOTES BY SHARON BATES

J. Towbin – Heart failure in young. All forms of Cardiomyopathy except ARVC (usually in adolescence). X-linked Dilated Cardiomyopathy passed to boys from mom. Dystrophic – mutations in 5-Dys resulting in early onset. Final common pathways.

10N channels, sarcomere, sarcolemma, sarcomere link
(--- Cascade pathways ---)

HCM

DCM

Ventricular Arrhythmias

Dystrophic is a key player to membrane of cardiac

Muscular dystrophies (form) also are related to genetic (DNA) mutation of muscular skeletal issues. Protein / Protein disorders that lead down the path of presented disease. Fatigue without presentation of cardiac issues are still related to muscular disease. Neuro muscular doctors need to be included in the care of these kids with DCM genes.

Pediatric HCM

The sarcomere, the youngest of kids have metabolic relation which has the worst outcome. Other forms of HCM in “childhood” Same list as DCM. Enzymes are necessary to introduce to prevent death at early life. LV non-compaction can be misleading as HCM, Hypoplastic Left Heart, usually leads to death or heart transplant. Can also change to dilated cardiomyopathy over time.

Childhood heart failure occurs wide spectrum of etiologies. Look at muscle disease commonly is associated with cardiac dysfunction in children.

* talk to Dr. Towbin about hyper-extension of finger & knee & ankles. Other in PHW same issues & massive nose bleeds.

17:24 Differential diagnosis and management of cardiac amyloidoses

C. Rapezzi (Bologna, I)

LECTURE ABSTRACTS

DIFFERENTIAL DIAGNOSIS AND MANAGEMENT OF CARDIAC AMYLOIDOSES

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Most studies of amyloidic cardiomyopathy consider as a single entity the three main systemic cardiac amyloidoses: 1) acquired monoclonal immunoglobulin light-chain amyloidosis (AL), 2) hereditary, transthyretin-related amyloidosis (ATTR), 3) systemic “senile” amyloidosis (SSA).

Our aim was to assess the diagnostic and clinical profiles of the three main types of systemic cardiac amyloidosis.

We conducted a longitudinal study of 115 cardiac amyloidosis patients with clear-cut etiological diagnosis (AL, n= 64; ATTR, n = 37; SSA, n = 14) seen at our institutional network for diagnosis/treatment of systemic amyloidosis since 1994 (minimum follow-up, 6 months). In addition to diagnostic ECG and echocardiographic findings, routine hemodynamic data were available for most patients (n = 71).

Average age at diagnosis was higher in AL than in ATTR; all SSA patients were elderly men. At diagnosis, mean LV wall thickness was higher in SSA but not in AL or ATTR. Hemodynamic measures (right and left filling pressures and cardiac index) differed remarkably between the three etiological types, with the highest frequencies of abnormal values always being recorded for AL. Low QRS voltage and low voltage/mass ratio were much less frequent findings in ATTR than in AL.

ATTR and SSA patients had better outcomes than AL patients in terms of both overall survival and freedom from major cardiac events. At Cox proportional hazards analysis, ATTR was a strongly favorable predictor survival, and SSA predicted freedom from major cardiac events.

Conclusion: AL, ATTR, and SSA should be considered three different cardiac diseases, probably characterized by different pathophysiological substrates and courses.

NOTES BY SHARON BATES

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C. Rapezzi – Amyloidosis grouped into HCM but there are various misformed (thick) sections of the heart in all areas (LV, RV, ART). Carpal tunnel disorder is present in 30% of the patients with amyloidosis. Frequency of low voltage is high in amyloidosis. Different uptake of radiology tracers for AL & ATTR mutations. There are different treatment plans to add to longevity of patients.

17:48 Tropical neglected cardiomyopathies
A.O.H. Mocumbi (London, UK)

LECTURE ABSTRACTS

TROPICAL NEGLECTED CARDIOMYOPATHIES

ANA OLGA H. MOCUMBI

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The practice, science and art of cardiology and cardiac surgery have witnessed unprecedented progress during the last 50 years. This resulted in considerable reduction in mortality and morbidity from cardiovascular disease, and contributed to enhance overall life expectancy and quality of life, benefits that have been highly selective, since excluding the majority of the world population who lives in developing countries.

It is widely accepted that cardiomyopathies constitute the major challenge of cardiovascular medicine in the developing world. Its burden is largely underestimated considering that to the usual forms with worldwide distribution, one must add a subset of conditions endemic in these areas, namely Chagas disease, endomyocardial fibrosis, peripartum cardiomyopathy and nutritional cardiomyopathies. Despite affecting large numbers of people, because they affect poor populations with little human and infrastructural capacity for research, little progress that been done in understanding the mechanisms of the diseases and, therefore, in identifying new therapeutic targets. Endomyocardial fibrosis is the commonest form of restrictive cardiomyopathy affecting children and young adults from poor areas developing countries. In the endemic areas of Africa, EMF is the second cause of admission for acquired cardiovascular disease in these age groups, after rheumatic heart disease, accounting for up to 20% of all cases of heart failure. The disease has an unclear etiology, the pathogenesis is unknown, carries a poor prognosis and has no specific treatment.

The availability of portable echocardiography allows for large-scale epidemiological studies on incidence, prevalence and determinants of endomyocardial fibrosis, A translational research project involving the Imperial College London, Chain of Hope-UK and the Maputo Heart Institute-Mozambique is currently being done in an endemic area of Mozambique, combining community studies with cellular and molecular research to define the basic mechanisms involved. In addition, new surgical procedures based on specific, pathophysiology present are being developed and applied, with attempts at optimizing the timing of surgery. The results of this research programme are presented and discussed.

NOTES BY SHARON BATES

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A.O.H. Mocumbi –16.7 million people die from CVD worldwide in 2002 but 80% lived in developing countries / tropical regions potentially. Endomyocardial Fibrosis (EMF) affects mainly children in poor countries. EMF tissues leads to restrictive physiology with AV valve dysfunction. Potential echocardiogram research center in rural areas of highest area of reported cases. 1200 people screened to date. Many clinical of condition were determined for mass screenings. Large distortions of heart so regular Cardiomyopathy calculations do not apply. When a person is found fibrosis tissues that separate cardium. No clinical treatment but surgery has allowed for some good survival rates. 26 patients, 24 survivals and 2 deaths. Created educational numbers in commonality. Research centers in country, mapped homes, areas with 1200 patients. Early research (2 years) with non-recurrence of disease. Good presentation, research & more!

18:12 **Session 7**

19:00 **Round Table**

How to implement education and translation research in cardiomyopathies

E. Arbustini (Pavia, I), **B.J. Maron** (Minneapolis, USA)

W.J. McKenna (London, UK), **J.A. Towbin** (Houston, USA)

M. Yacoub (London, UK)

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LECTURE ABSTRACTS

IMPLEMENTING EDUCATION AND TRANSLATIONAL RESEARCH IN CARDIOMYOPATHIES: THE PERSPECTIVE

GIAN FRANCO GENSINI

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The term “cardiomyopathies” identifies a large and articulated group of disturbances affecting the myocardium. These disorders often result in heart dysfunction and consequently heart failure. Cardiomyopathies are the major determinant of hospitalization in people older than 65 years of age in the USA. Therefore, involving huge loss of quantity and quality of life and relevant health expenditure. Ischemic cardiomyopathy accounts for approximately 50% of the affected patients, even if, according to recent clinical tests, the prevalence of potentially reversible nonischemic cardiomyopathy may range from 25% to 45%. The traditional classification of cardiomyopathies into dilated, hypertrophic and restrictive patterns implies classical subdivisions according to morphologic criteria. However, given recent relevant and quick progress in the genetic evaluation of cardiomyopathies, a re-definition of their taxonomy is mandatory to effectively implement systemic education and translational research in this area. Such educational and research effort will be illustrated in this oral presentation.

NOTES BY SHARON BATES

Email: sharon@anthonybates.org

G. Gensini - The Italian perspective appropriate algorithms to follow cardiomyopathies in Italy (Tuscan region).

B. Maron – Within the specific center / institution to call it the name of disease, generate data, have a structured center & organizations to be of that sub-specialty center. Collaborative efforts come from partnerships, with commitments focus & support.

ESC – Get European group to support such a center by building the groups.

W. McKennon – Agreement to have centers, standardization of Cardiomyopathies for bigger world collaborations. Funding has always been an issues, but all levels of research need to be buildt to bringing in research / education for future.

J. Towbin – Added group across the years with all subgroups of Neurology, etc. Group includes Pediatric Cardiomyopathy specimum respository for future research options. Funding sources are available broad based thinking and collaboration is the way we go!

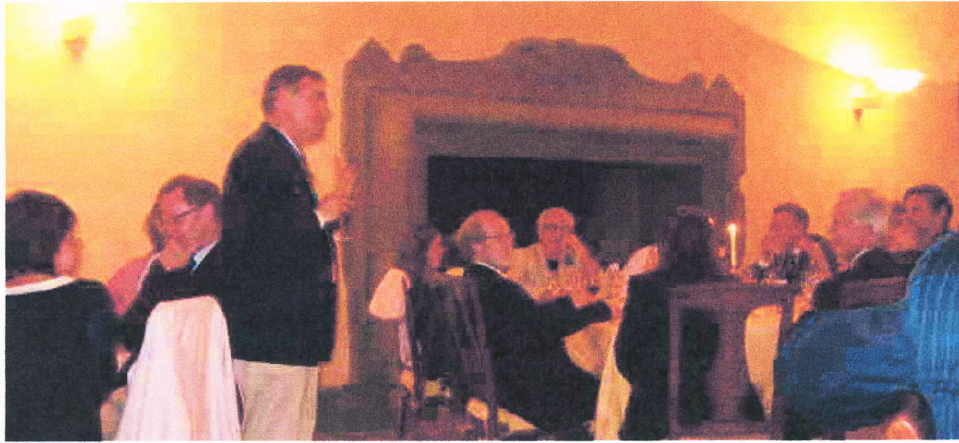
E. Aurbustini – Medical school & specialty school students are lucky to have 1 hour in cardiomyopathy training and barely have a genetic subject. More changes must occur in education of our future doctors.

19:30 Faculty Dinner – Castello Vicchiomaggio, Greve in Chianti
24:00

NOTES BY SHARON BATES

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The invitation to the faculty dinner proved to be a great opportunity to share the ABF perspective with the doctors and researchers on the front lines. Connecting with Dr. Ana Mocumbi lead to an invitation to come to Mozambique and witness the screening effort taking place in Africa. This lead to a discussion with the doctors from Egypt. Their invitation followed to assist in building a cardiac screening program for the young people of Egypt. All of which created new friends in new corners of the world.



Toast to all the participants and leadership that made this conference a success!



Dr. Franco Cecchi accepting the accolades for all the leaders of the great conference!

SATURDAY, MAY 24, 2008

8:30 **Session 8**

10:00 **Sudden death prevention: ICD patient selection and risk/benefit ratio**

Chairmen: **L. Padeletti** (Florence, I), **A. Raviele** (Venice, I)

LUIGI PADELETTI

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ANTONIO RAVIELE

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8:30 In HCM

P. Spirito (Genoa, I)

LECTURE ABSTRACTS

IN HYPERTROPHIC CARDIOMYOPATHY

PAOLO SPIRITO

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The implantable cardioverter-defibrillator (ICD) has proven to be particularly effective in preventing sudden cardiac death in patients with hypertrophic cardiomyopathy (HCM) and has altered the natural history of this disease. Indeed, the ICD reliably aborts life-threatening ventricular tachyarrhythmias in HCM patients judged to be at high risk for sudden death. An important proportion of appropriate ICD interventions occur in patients prophylactically implanted for only a single risk factor, underscoring that multiple markers of high-risk status are not always necessary to justify consideration for primary

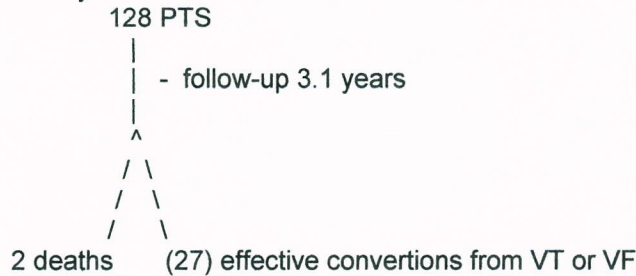
prevention defibrillator therapy. The current risk profile strategy has proved to be a useful guide to the selection of HCM candidates to prophylactic ICDs, but further investigation is required to achieve a more precise risk stratification algorithm. The available experience suggest that high risk HCM patients protected from sudden cardiac death by the ICD could potentially survive many decades of productive life, and even achieve normal or near-normal life expectancy, if not encumbered by other major HCM-related disease complications.

NOTES BY SHARON BATES

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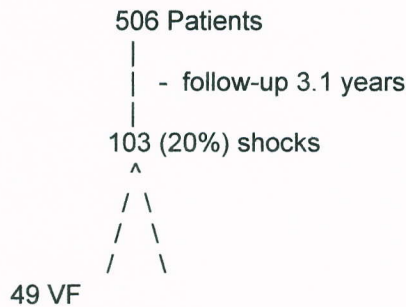
P. Spirito – Sudden Death with HCM occurs more often than 15 – 20 years ago. No evidence that beta-blockers may reduce risk of sudden death.

Efficacy of ICD



2 or more cardiac arrest (VT)

Familial sudden death, massive > 3.0 cm LVH, unexplained history of syncope and abnormal BP



Appropriate cardioversions rate is 4% per year

Only 30% had 1 Risk factor

Risk stratification & ICD decision making in HCM

- Current guide is good but having one risk factor is enough to determine need for ICD

Need to select patients with high risk of Sudden Death

No Prognostic weight in risk factors

Single risk factors will justify strong consideration for ICD

- family with history of ≥ 2 sudden deaths in young relatives
- Extreme LVH (> 3 cm) in adolescents & young patients
- Rep. & prolonged NSVT
- Unexplained syncope

ICD decision making in HCM continues to rely on the judgement of the individual physician but also the contribution of the patients. Discuss with patients ICD benefits and patients potential risk of ICD implantation.

8:45 In DCM & ARVC
G. Bonani (Bologna, I)

LECTURE ABSTRACTS

IN DILATED NON-ISCHEMIC CARDIOMYOPATHY AND ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

GIUSEPPE BORIANI

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Dilated non ischemic Cardiomyopathy