



Second Thoracic Aortic Disease Summit

Wednesday, August 1, 2012

- 8:00 Continental Breakfast
- 8:30 Opening Remarks
Eser Tolunay, Ph.D., National Heart, Lung, and Blood Institute, Bethesda, MD

Session 1: Role of Registry-Based Research

Chair: Cheryl Maslen, Ph.D., Oregon Health & Science University, Portland, OR

- 8:40 Introduction
Cheryl Maslen, Ph.D., Oregon Health & Science University, Portland, OR
- 8:55 Scope of the Problem
Richard B. Devereux, M.D., Ph.D., Weill Cornell Medical Center, New York, NY
- 9:10 The Role of IRAD in Registry-Based Research
Eric M. Isselbacher, M.D., Massachusetts General Hospital, Boston, MA
- 9:25 GenTAC II
Patrice Desvigne-Nickens, M.D., National Heart, Lung, and Blood Institute, Bethesda, MD
- 9:40 Aortic Valve Operative Outcomes in Marfan Patients: A Study Update
Joseph Coselli, M.D., Texas Heart Institute at St. Luke's Episcopal Hospital, Baylor College of Medicine, Houston TX
- 9:55 Thoracic Aortic Disease Coalition
Loren F. Hiratzka, M.D., TriHealth, Cincinnati, OH
- 10:10 Break

Session 2: Genetics and Developmental Aspects of Thoracic Aortic Disease

Chair: Reed Pyeritz, M.D., Ph.D., University of Pennsylvania, Philadelphia, PA

- 10:30 The Genetics of Thoracic Aortic Disease
Dianna Milewicz, M.D., Ph.D., University of Texas Health Science Center, Houston, TX
- 10:55 Inheritance risk and familial patterns for BAV, dilated aortas
Mark B. Lewin, M.D., University of Washington, Seattle, WA
- 11:15 Diversity of genetic mutations and phenotypes in adults with inherited aortopathies
Elizabeth McNally, M.D., Ph.D., University of Chicago, Chicago, IL
- 11:35 The Role of MicroRNAs in Aneurysm Development
Michael Fischbein, M.D., Ph.D., Stanford University, Palo Alto, CA





Contributed Abstracts

- 11:55 Bicuspid Aortic Valves and Rare Copy Number Variants Are Enriched in Early Onset Thoracic Aortic Disease
Siddharth K. Prakash, M.D., Ph.D., University of Texas Health Science Center, Houston, TX
- 12:05 Genetic Dissection of Anatomical Compartments & Embryonic Lineages in Vascular Pathology in MFS
Jason R. Cook, Mount Sinai Medical Center, New York, NY
- 12:15 Transgenic expression of mutant Acta2 R258C in mice leads to aortic disease and increased smooth muscle cell proliferation
Carlos A. Villamizar, University of Texas Health Science Center, Houston, TX
- 12:30 Question and Answer
- 12:45 Lunch Break

Session 3: Pathogenesis of Thoracic Aortic Disease and Translational Advances

Chair: Hal Dietz, M.D., Johns Hopkins University, Howard Hughes Medical Institute, Baltimore, MD

- 1:45 Animal Modeling of Aortic Maldevelopment in TGF- β vasculopathies
Mark E. Lindsay, M.D., Ph.D., Johns Hopkins University, Baltimore, MD
- 2:05 Smooth Muscle Cell Phenotype and Matrix Homeostasis in Ascending Aneurysms of Bicuspid and Trileaflet Aortic Valve Patient
Karianna Milewski R.Ph., M.D., Ph.D., University of Pennsylvania, Philadelphia, PA
- 2:25 S100 proteins as mediator of aortic smooth muscle dysfunction and aortic wall remodeling
Marion Hofmann-Bowman, M.D., Ph.D., University of Chicago, Chicago, IL
- 2:45 Matrix metallo-proteinases and aortic aneurysms in Marfan Syndrome
Cornelis Van Breemen, D.V.M., Ph.D., University of British Columbia, Vancouver, BC
- 3:05 Mechanisms of Angiotensin II Provoked Ascending Aortic Aneurysms
Alan Daugherty, Ph.D., D.Sc., University of Kentucky, Lexington, KY
- 3:25 *Break*

Contributed Abstracts

- 3:40 MMP-2 Regulates Erk1/2 Phosphorylation and Aortic Aneurysm in Marfan Syndrome
Wanfen Xiong M.D., Ph.D., University of Nebraska Medical Center, Omaha, NE
- 3:50 Carotid Intima Media Thickness Provides Evidence that Ascending Aortic Aneurysm Protects Against Systemic Atherosclerosis
John Elefteriades, M.D., Yale University School of Medicine, New Haven, CT



- 4:00 Advanced Aortic Atherosclerosis is Associated with Increased Medial Degeneration in Sporadic Ascending Aortic Aneurysms
Paul Albini, M.D., Baylor College of Medicine, Houston, TX
- 4:10 Homing, Activity, and Fate of Bone Marrow-derived Stem Cells in Aortic Aneurysms and Dissections
Sili Zou, M.D., Texas Heart Institute at St. Luke's Episcopal Hospital, Baylor College of Medicine, Houston TX
- 4:20 AT1 receptor activation by mechanical forces on the ascending aorta induces aortic remodeling and dilatation in a mouse model following transverse aortic banding
Shao-Qing Kuang, M.S., Ph.D., University of Texas Health Science Center, Houston, TX
- 4:30 Identification of a major modifier locus for TGF β vasculopathies in mice
Juan F. Calderon, B.Sc., McKusick-Nathans Institute of Genetic Medicine, The Johns Hopkins University, Baltimore, MD
- 4:40 Question and Answer

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- 6:00 Cocktail Hour and Poster Presentations
- 7:00 Aortic Summit Dinner (*registered guests only*)

Keynote address: Amy Yasbeck

Performance: Patty Peterson



Poster Presentations

1. Role of Bmp signals in Marfan syndrome
Yan Bai, James F. Martin

2. Normal values for ascending aortic diameters with 2D echocardiography
Laurence Campens, Laurent Demulier, Mary Roman, Richard Devereux, Anne De Paepe, Julie De Backer

3. Circulating Microfibril Fragments: Biomarkers of Aortic Disease
Eric J. Carlson, L. M. Marshall, J. O'Malley J, C. K. Snyder, Noe L. Charbonneau, Susan J. Hayflick, Joseph S. Coselli, Scott A. LeMaire, Lynn Y. Sakai

4. Cardiomyopathy is a primary cardiovascular manifestation in Marfan syndrome
Jason R Cook, Luca Carta, Ludovic Benard, Elie R. Chemaly, Emily Wilson

5. Preventing Paraplegia after Extensive Distal Thoracic Aortic Replacement
Gabriele Di Luozzo, Sarah Geisbuesch, Hung-Mo Lin, Moritz Bischoff, Randall B. Griep

6. Frequency of Reoperations in Patients with Marfan Syndrome
Sarah Geisbuesch, Deborah Schray, Moritz S. Bischoff, Hung-Mo Lin, Gabriele Di Luozzo, Randall B. Griep

7. Transforming Growth Factor Beta signaling in Thoracic Aortic Aneurysms
David Howell, Richard Metz, Natasha Popovic, Emily Wilson

8. Possible Mechanical Role of GAGs in Thoracic Aortic Dissections
Jay Humphrey, S. Roccabianca, G. Ateshian

9. A rare variant in MYH11, R247C, acts as a genetic modifier to cause aortic dilatation in mice haploinsufficient for Acta2
Callie Kwartler, Callie S. Kwartler, Shao-Qing Kuang, Siddharth Prakash, Katerina L. Byanova, John Pham, Jian Huang, Kristine Kamm, James Stull, H. Lee Sweeney, Dianna M. Milewicz
10. AKT2-Deficient Mice Are Susceptible to Aortic Aneurysms and Dissections
Scott A. LeMaire, Lin Zhang, Mary Nguyen, Pingping Ren, Darrell Wu, Joseph S. Coselli, Ying H. Shen

11. A microRNA profile comparison between thoracic aortic dissection and normal thoracic aorta indicates the potential role of microRNAs in contributing to thoracic aortic dissection pathogenesis
Mingfang Liao, Sili Zou, Mingfang Liao, Sili Zou, Jianfeng Weng, Lewei Hou, Lin Yang, Zhiqing Zhao, Junmin Bao, Zaiping Jing
12. Dissecting the Role of the TGF- β Pathway in Genetically Mediated Vascular Disorders
Rachel Morissette, Florian Schoenhoff, Jennifer L. Fiori, Benjamin F. Griswold, Leslie J. Sloper, Hal Dietz, Jennifer E. Van Eyk, Nazli B. McDonnell
13. ADAMTS-1 and ADAMTS-4 are Elevated in Thoracic Aortic Aneurysms and Dissections
Pingping Ren, Pingping Ren, Lin Zhang, Gaiping Xu, Laura C. Palmero, Joseph S. Coselli, Ying H. Shen, Scott A, LeMaire
14. ADAMTS-4 Deficiency Attenuates Aortic Aneurysm and Dissection Formation in Mice
Pingping Ren, Pingping Ren, Sili Zou, Mary T. Nguyen, Joseph S. Coselli, Ying H. Shen, Scott A. LeMaire



15. Reoperative repair of multiple thoracic aneurysms following progressive aneurysmal degeneration in a patient with incompletely treated temporal arteritis
Mark J. Russo, Karin Dill, Atman Shah, Darwin Eton
16. In Search of a New Therapeutic Target for the Treatment of GentAC (ii): Impact of the Segmental Development of the Aortic Tree
Hisham M.F. Sherif
17. In Search of a New Therapeutic Target for the Treatment of Genetically Triggered Thoracic Aortic Aneurysms: Insights from Human and Animal Lathyrism
Hisham M.F. Sherif
18. Aortic Wall Thickness: An Independent Risk Factor for Aortic Dissection in Connective Tissue Disorders?
Hadas Shiran, Justin Odegaard, Gerald Berry, D. Craig Miller, Michael Fischbein, David Liang
19. Amplification of the NLRP3 Inflammasome Cascade in Thoracic Aortic Aneurysms and Dissections: A Potential Mechanism for Matrix Metalloproteinase-9 Activation
Darrell Wu, Darrell Wu, Gaiping Xu, Wanmu Xie, Pingping Ren, Lin Zhang, Laura Palmero, Joseph S. Coselli, Ying H. Shen, Scott A. LeMaire
20. Notch Signaling in Thoracic Aortic Aneurysms and Dissections
Sili Zou, Pingping Ren, Mary Nguyen, Joseph S. Coselli, Ying H. Shen, Scott A. LeMaire





Thursday, August 2, 2012

7:30 AM Continental Breakfast

Session 4: Natural History, Clinical History, and Medical Management

Nazli McDonnell, M.D., Ph.D. National Institute of Aging

- 8:00 The Imperative for Investing in Understanding Natural and Clinical History
Reed Pyeritz, M.D., Ph.D., Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA
- 8:20 New insights into the pathogenesis and treatment of Marfan syndrome and related disorders
Hal Dietz, M.D., Johns Hopkins University, Howard Hughes Medical Institute, Baltimore, MD
- 8:40 Pregnancy in Women with Aortopathy
Alan Braverman, M.D., Washington University School of Medicine in St. Louis
- 9:00 Aorta aneurysm in Turner Syndrome: Who to treat, when to worry
Michael Silberbach, M.D., Oregon Health & Science University, Portland, OR
- 9:20 Death is a rumor to the young: BAV outcomes in the community-The Mayo Clinic experience
Hector Michelena, M.D., Mayo Clinic, Rochester, MN

Contributed Abstracts

- 9:40 Rationale and proposed design of a prospective, collaborative meta-analysis of angiotensin receptor antagonists in Marfan syndrome
Alex Pitcher, M.A., BMBCh, MRCP, University of Oxford, Oxford, UK
- 10:00 Panel Discussion
- 10:30 Break

Session 5 Imaging, Interventional Management and Outcomes

Chairs: Federico Asch, M.D., FASE, Washington Hospital Center, Washington, DC

William Ravekes, M.D., Johns Hopkins University, Baltimore, MD

- 10:45 Report on the ASE and EAE Guidelines for Multimodality Imaging for Diseases of the Aorta in Adults
Steven Goldstein, M.D., Washington Hospital Center, Washington, DC
- 11:05 Complex blood flow disturbance and wall shear stress in Marfan syndrome: insights from 4D flow CMR.
Alex Pitcher, M.A., BMBCh, MRCP, University of Oxford, Oxford, UK
- 11:25 Controversies in imaging the aorta: Should children images be handled different from adults?



Ronald Lacro, M.D., Harvard University, Boston, MA

11:45 LUNCH

12:45 Aortic Surgery in Patients with a Bicuspid Aortic Valve

Joseph Bavaria, M.D., University of Pennsylvania Medical School, Philadelphia, PA

1:00 Valve-sparing aortic root replacement in MFS and other (non-bicuspid) patients

D. Craig Miller, M.D., Stanford University, Palo Alto, CA

1:15 Aortic Surgery in Childhood

Duke Cameron, M.D., Johns Hopkins University, Baltimore, MD

1:30 Stenting and dissection

G. Chad Hughes, M.D., Duke University, Durham, NC

1:45 Indications and Contraindications of TEVAR for Genetically Triggered Aortic Aneurysmal Disease

Thomas G. Gleason, M.D., University of Pittsburgh, Pittsburgh, PA

Contributed Abstracts

2:00 A measured and conserving approach to the Marfan aortic root by use of a customised external support

Tom Treasure, M.D., M.S., FRCS, FRCP, University College London

2:10 Total aortic exclusion during thoracoabdominal aortic aneurysm repair using branched surgical grafts to eliminate all at-risk aorta

Brandon W. Propper, M.D., Johns Hopkins University, Baltimore, MD

2:20 Panel Discussion

Closing Remarks

3:15 Future Directions

Peter Byers, M.D., University of Washington, Seattle, WA

3:45 Adjourn

