Wednesday, July 9, 2014

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

Session 1: GenTAC Research

Chairs: Scott LeMaire, M.D.; Alan Braverman, M.D.

8:40  Overview of research and new tools for clinical aortic research
      Richard B. Devereux, M.D., Ph.D., Weill Cornell Medical Center, New York, NY

8:50  Vertebral Artery Tortuosity Index is a Novel Biomarker of Surgery and Aortic Dissection or Rupture in Children and Young Adults: Findings From the National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC)
      Shaine A. Morris, M.D., Baylor College of Medicine, Houston, TX

9:00  Turner syndrome/CNV analysis
      Siddharth Prakash, M.D., University of Texas Health Science Center, Houston, TX

9:10  Calcium channel blockers and aortic disease
      Hal Dietz, M.D., Johns Hopkins University, Howard Hughes Medical Institute, Baltimore, MD

9:20  Incident Aortic Dissection Among Patients with Genetically Mediated Aortic Aneurysms – Initial Results from the GenTAC Registry
      Jonathan Weinsaft, M.D. Ph.D., Weill Cornell Medical Center, New York, NY

9:30  Vascular surgery in patients with vascular Ehlers Danlos Syndrome
      Sherene Shalhub, M.D., M.P.H., University of Washington, Seattle, WA

9:40  Question and Answer/Discussion

9:55  Break

Session 2: Genetic Predisposition to Thoracic Aortic Disease and Development of the Aorta

Chairs: Cheryl Maslen, Ph.D.; Peter Byers, M.D.

10:15  Arterial wall construction: mixing, sequential induction and controlled invasion
      Daniel Greif, M.D., Yale University, New Haven, CT

10:30  Role of SCA1+ adventitial cells in vasculature
      Mark Majesky, Ph.D., University of Washington, Seattle, WA

10:45  Myocardin-like Transcriptional Coactivators and the Pathogenesis of Heritable Forms of Arterial Aneurysm
      Michael S. Parmacek, M.D., Hospital of the University of Pennsylvania, Philadelphia, PA

11:00  Novel genes and molecular pathways for familial thoracic aortic disease
      Dianna Milewicz, M.D., Ph.D., University of Texas Health Science Center, Houston, TX

11:15  Novel genes and modifiers for Thoracic Aortic Disease
      Catherine Boileau Pharm.D., Ph.D., Paris, France

11:45  Elucidation of a complex architecture for genetic modification of the TGFβ vasculopathies
      Juan F. Calderon, Ph.D., McKusick-Nathans Institute of Genetic Medicine, The Johns Hopkins University, Baltimore, MD

12:00  Bicuspid aortic valve: genome wide association study and rare variant analysis
      Simon Body, MBChB, MPH, Brigham and Women’s Hospital, Boston, MA

12:15  Modeling genetically triggered vascular diseases using human induced pluripotent stem cells
      Sanjay Sinha, Ph.D., University of Cambridge, Cambridge, UK

12:30  Question and Answer/Discussion

1:00  Lunch
Session 3: Thoracic Aortic Disease in Mouse Models

Chair: Hal Dietz, M.D., Alan Daugherty, Ph.D., Jennifer Habashi, M.D.

2:00 Mouse models: TGFBR1, TGFBR2, and FBN1 updates
Hal Dietz, M.D., Johns Hopkins University, Howard Hughes Medical Institute, Baltimore, MD

2:15 Tgfbr2 disruption in postnatal smooth muscle impairs aortic wall homeostasis
George Tellides, M.D., Ph.D., Yale University, New Haven, CT

2:30 FBLN4−/− mouse model of thoracic aortic disease
Hiromi Yanagisawa, M.D., Ph.D., UT Southwestern, Dallas, TX

2:45 Fibrillin-1 is a component of the muscle mechanosignaling complex
Francesco Ramirez, Ph.D., Mount Sinai Medical Center, New York, NY

3:00 Acta2 mutant mouse model
Jiyuan Chen, Ph.D. University of Texas Health Science Center at Houston

3:15 Angiotensin and TGF-β signaling in aortic disease
Alan Daugherty, Ph.D., D.Sc., University of Kentucky, Lexington, KY

3:30 Panel - Question and Answer/Discussion

3:45 Break

4:00 The role of ADAMTS proteins in thoracic aortic disease
Scott LeMaire, M.D., Baylor College of Medicine, Houston, TX

4:15 Pregnancy and the Marfan mouse
Jennifer Habashi, M.D., Johns Hopkins University, Baltimore, MD

4:30 Structural alterations in the aorta due in mouse models of aortic disease
Elaine Davis, Ph.D., McGill University, Montreal, Canada,
Poster Presentations

1. **Pregnancy and the Risk of Aortic Dissection: a Case-Crossover Analysis**
   Hooman Kamel, Mary J. Roman, Richard B. Devereux

2. **Twins with Progressive Thoracic Aortic Aneurysm, Recurrent Dissection and ACTA2 Mutation**
   Robert B. Hinton, Amy Shikany, Benjamin J. Landis, Jeanne F. James, Stephanie M. Ware

3. **Open Repair of Descending Thoracic or Thoracoabdominal Aortic Aneurysms in Patients With Chronic DeBakey Type I Dissection: Early and Long-term Outcomes.**
   Michael S. Hughes, Joseph S. Coselli, Susan Y. Green, Samantha Zarda, Courtney Nalty, Matt D. Price, Ourania Preventza, Kim I. de la Cruz, Scott A. LeMaire

4. **NLRP3 Inflammasome Degrades Contractile Proteins Leading to Thoracic Aortic Biomechanical Failure in a Murine Model**
   Yanqiu Zheng, Darrell Wu, Michael Hughes, Lin Zhang, Joseph S. Coselli, Ying H. Shen, Scott A. LeMaire

5. **Changes in Gender-Related Differences in Acute Aortic Dissection over Time**
   Stephen Philip, Linda Pape, Toru Suzuki, Lori Conklin, James Froehlich, Rossella Fattori, Eva Kline-Rogers, Justin Jabara, Guillaume Jondeau, Hasan Siddiqi, Eduardo Bossone, Daniel Montgomery, Eric Isselbacher, Christoph A. Nienaber

6. **Decreased TGFβ activity is associated with formation of thoracic aortic aneurysm in patients with bicuspid aortic valve**

7. **Role for Notch1 Signaling in Ascending Aortic Aneurysms**
   Sara N. Koenig, Patrick Rowland, Nianyuan Huang, Vidu Garg

8. **New Genes in Old Families with Heritable Thoracic Aortic Disease**
   Julie De Backer, L. Campens, B. Callewaert, L. Muino Mosquera, S. De Nobele, A. De Paepe, P Coucke

9. **Mutation detection rate and -characteristics in TAA related disorders: results from NGS panel testing J**
   Julie De Backer, L. Campens, L. Muiño Mosquera, B. Callewaert, M. Baetens, M. Renard A. De Paepe, P. Coucke

10. **Comprehensive Genetic Screening for Thoracic Aortic Aneurysm via Whole Exome Sequencing**

11. **Advanced glycation end products and their soluble receptors: A novel risk factor in the pathogenesis of aortic aneurysm**
    Bulat A. Ziganshin, Kailash Prasad, Abdullah Sarkar, Courtney Mayerick, Mohammad A. Zafar, Ahmed Shoker, Hamdi El Moselhi, Maryann Tranquilli, John A. Elefteriades

12. **Comparison of 3-Dimensional Echocardiography and Cardiac Magnetic Resonance Imaging for the Assessment of Aortic Dilation**
    Justin Uhl, Luciana Young, Jennifer Acevedo, Elizabeth Cappella, Karen Rychlik, Grace Choi

13. **Captopril Treatment Worsens Aortic Remodeling Associated with Aortic Constriction and Aneurysm Formation in Acta2/-/- Mice**
    Andrew Peters, Jiyuan Chen, Siddharth Prakash, Shao-Qing Kuang, Corey Reynolds, Dianna Milewicz
14. Integrin interaction with fibrillin-1 triggers an integrated response in microRNA expression
   Dieter Reinhardt, Karina Zeyer, Heena Kumra, Amani Hassan

15. SM22 knockout mouse model: the roles of cytoskeletal remodeling-induced oxidative stress and inflammation in aneurysm formation
   Li Li, Xiaohua Dai, Neeraja Priyanka Annam, Gannon Curtis, Pamela C Udeh-Ubaka

   Michael Silberbach, Cheryl Maslen, Dianna M. Milewicz

17. Premature Aortic Smooth Muscle Cell Differentiation Contributes to Matrix Dysregulation in Marfan Syndrome
   Matthew Dale, Wanfen Xiong, Trevor Meisinger, Jeffrey Carson, B. Timothy Baxter

18. Echocardiography Underestimates Aortic Size in Children with Bicuspid Aortic Valve
   Glen Iannucci, Timothy Slesnick, James Parks, William Mahle, Denver Sallee,
Thursday, July 10, 2014

8:00 Continental Breakfast

**Session 4: Losartan Trials, Cohort Studies, and Biomarkers**
*Chairs: Reed Pyeritz, M.D., Guillaume Jondeau, M.D.*

- **8:30** Timeline for US trial outcome and ancillary studies  
  *Ronald Lacro, M.D., Harvard University, Boston, MA*
- **8:45** Update on meta-analysis of the losartan trials world wide  
  *Alex Pitcher, M.A., BMBCh, MRCP, University of Oxford, Oxford, UK*
- **9:00** Overview of mouse drug trials – what are they telling us?  
  *Francesco Ramirez, Ph.D., Mount Sinai Medical Center, New York, NY*
- **9:15** Aortic Disease Outcomes Associated with ACTA2 mutations  
  *Ellen Regalado, M.S., C.G.C., University of Texas Health Science Center, Houston, TX*
- **9:30** IRAD update  
  *Kim Eagle, M.D., University of Michigan, Ann Arbor, MI*
- **9:45** Panel - Question and Answer/Discussion

- **10:15** Biomarkers for aortic disease  
  *Lynn Sakai, Ph.D., Shriners Research Center, Portland, OR*
- **10:30** Preliminary Biomarkers for identification of human ascending thoracic aortic aneurysm  
  *James D. McCully, Ph.D., Harvard University, Cambridge, MA*

**Contributed Abstracts**

- **10:45** Aortic Wall Delayed Gadolinium Enhancement in Thoracic Aortic Aneurysm Patients  
  *Hadas Shiran, M.D., Stanford University, Palo Alto, CA*
- **10:55** Aortic size indices: A more comprehensive evaluation of aortic risk  
  *Arturo Evangelista, M.D., Hospital General Universitari Vall d’Hebron, Barcelona, Spain*
- **11:05** Pediatric nonsyndromic thoracic aortic aneurysm: opportunities for early clinical stratification and therapy?  
  *Benjamin J. Landis, M.D., Cincinnati Children's Hospital Medical Center, Cincinnati, OH*
- **11:15** Prevalence of aortopathy in patients with hereditary hemorrhagic telangiectasia  
  *Christina Rigelsky, M.S., Cleveland Clinic, Cleveland, OH*
- **11:25** Panel - Question and Answer/Discussion
- **11:45** Lunch

**Session 5 Imaging and Surgical Outcomes**
*Chairs: Craig Miller, M.D. and Federico Asch, M.D.*

- **1:00** Evolving Guidelines: Multimodality imaging of the thoracic aorta  
  *Steven Goldstein, M.D., Washington Hospital Center, Washington, DC*
- **1:15** Novel MRI for aortic physiology  
  *Rachel Clough, M.D., Ph.D., King’s College, London, UK*
- **1:30** Aortic Size & Dissection – What Do We Really Know?  
  *Richard B. Devereux, M.D., Ph.D., Weill Cornell Medical Center, New York, NY*
- **1:45** Imaging of Aortic Disease with bicuspid aortic valve  
  *Arturo Evangelista, M.D., Hospital General Universitari Vall d’Hebron, Barcelona, Spain*
- **2:00** Update from the Aortic Valve Operative Outcomes in Marfan Patients Study  
  *Scott LeMaire, M.D., Baylor College of Medicine, Houston, TX*
- **2:15** Indications and Contraindications of TEVAR for Genetically Triggered Aortic Aneurysmal Disease  
  *Thomas G. Gleason, M.D., University of Pittsburgh, Pittsburgh, PA*
Tirone David V-SARR for Patients with Bicuspid Aortic Valves. Corollary: Should all aortic root surgery be performed only in established centers of excellence?
D. Craig Miller, M.D., Stanford University, Palo Alto, CA

Aortic replacement in Bicuspid Aortic Valve: How big? How much?
Thor Sundt, III, M.D., Harvard University, Boston, MA

Emergency stent-grafting (TEVAR) for complicated acute type B dissections in genetically triggered aortic disease
Karianna Milewski, M.D., Ph.D., University of Pennsylvania Medical School, Philadelphia, PA

TEVAR vs. Open surgery for chronic Type B
Eric Roselli, M.D., Cleveland Clinic, Cleveland, OH

Panel - Question and Answer/Discussion

Closing Remarks:
Overview and Future Directions for GenTAC
Kim Eagle, M.D., University of Michigan, Ann Arbor, MI,
Patrice Desvigne-Nickens, M.D., National Heart, Lung, and Blood Institute, Bethesda, MD

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