

Angioma Alliance 7th Annual

Pathobiology of CCM Scientific Workshop

Dolce Chantilly Hotel - Chantilly, France; November 16th - 18th, 2011

WEDNESDAY, NOVEMBER 16TH, 2011

3:00 Welcome & Opening Remarks

> **Amy Akers** Angioma Alliance

Sara Sukalich Chair, Angioma Alliance Board of Directors

I. SIGNALING & MOLECULAR BIOLOGY - MODERATED BY DOUG MARCHUK			
3:10	Inactivation of Dll4-Notch silenced endothelial cells	signaling in CCM3 gene mutated familial CCM and in CCM3-	
	Yuan Zhu	University of Euisberg-Essen	
3:35	-	Structure-function analysis of the STRIPAK complex identifies direct interactions between CCM3/PDCD10 and other STRIPAK members and reveals a role for STRIPAK in Golgi polarization	
	Michelle Kean	Samuel Lunenfeld Research Institute	
4:00	Coffee Break		
4:10	A GCKIII kinase mediates	A GCKIII kinase mediates CCM2-dependent death in medulloblastoma cells	
	Barbara Costa	Weizmann Institute of Science	
4:35	CCM3-dependent redistribution of Mst4 upon oxidative stress is essential for phosphorylation of Ezrin/Radixin/Moesin and protects cells from necrosis		
	Juan Zalvide	University of Santiago de Compostela	
5:00	Identification of a novel KRIT1 interactor involved in the control of actin cytoskeleton dynamics and cell resistance to oxidative stress		
	Francesco Retta	University of Torino	

5:25 **DISCUSSION OF SESSION I**

6:00 CLOSE OF DAY 1

7:00 **DINNER** – SERVED IN THE **ETOILE RESTAURANT** AT DOLCE CHANTILLY

THURSDAY, NOVEMBER 17TH, 2011

II. VASCULAR BIOLOGY & ANIMAL MODELS - MODERATED BY ELISABETH TOURNIER-LASSERVE

8:30 Loss of KRIT1 leads to increased vascular permeability and modifies inflammatory

responses in vivo

Angela Glading University of Rochester

8:55 Defective vascular integrity upon ICAP-1/Krit1 complex loss correlates with aberrant

beta 1 integrin-dependent dialog between the endothelial cell and its extracellular

matrix

Eva Faurobert INSERM, Grenoble

9:20 *CCM1* and *ICAP1* induce Notch signaling to inhibit angiogenesis. Animal models and

therapeutic implications

Andreas Fischer Heidelberg University

9:45 COFFEE BREAK

10:00 Loss of Ccm3 in neuroglia leads to cerebral cavernous malformations and vascular

pathology

Angeliki Louvi Yale School of Medicine

10:25 CCM2 Regulates Superoxide and Nitric Oxide in the Endothelium

Christopher Gibson University of Utah

10:50 Fasudil Decreases Lesion Burden in a Murine Model of Cerebral Cavernous

Malformation Disease

Dave McDonald Duke University Medical Center

11:15 Discussion of Session II

12:00 END OF DAY 2

FRIDAY, NOVEMBER 18TH, 2011

III. CLINICAL & HUMAN STUDIES - MODERATED BY ISSAM AWAD

9:00 Cavernous malformations of the human brainstem: quasi-automatic three-

dimensional-reconstruction from a stack of serial histological slides with a

computerized technique

François Chapon CHU de Caen

9:25 Mutation Profiling in Familial Cerebral Cavernous Malformations and Development of a Familial Cavernous Malformation Patient Register

Jonathan Berg University of Dundee

9:50 **Coffee Break**

10:05 Therapeutic guidelines for the management of cerebral cavernous malformations in adults: quality of the evidence

Rustam Al-Shahi Salman University of Edinburgh

10:45 Discussion of Morning Session

11:00 **Discussion of Clinical Trials**

Lead by Issam Awad – University of Chicago and Rustam Al-Shahi Salman

12:00 LUNCH - SERVED IN THE HOTEL

IV. GENETIC & CLINICAL STUDIES - MODERATED BY ISSAM AWAD

1:30 Genetic Screening for Enhancer Loci of Intracranial Hemorrhage in Zebrafish

Brant Weinstein NICHD, National Institutes of Health

1:55 Brain Vascular Malformation Consortium (BVMC): Recruitment Strategies and Preliminary Data on Clinical Characteristics and Lesion Burden in a Cohort of CCM1-CHM Patients

Helen Kim University of California, San Francisco

Beth Baca University of New Mexico

2:20 COFFEE BREAK

2:35 Quantitative Analysis of Blood-to-Brain Influx Rate Within Lesions and Nonlesional Brain Using Dynamic Contrast-Enhanced MRI in Patients with Cerebral Cavernous Malformations

Blaine Hart University of New Mexico

3:00 Neuroimaging Markers of Permeability in Patients with Cerebral Cavernous Malformations Type 1: Effects of Statins

Leslie Morrison University of New Mexico

3:25 DISCUSSION OF AFTERNOON SESSION

4:00 CLOSE OF WORKSHOP