

**7th INTERNATIONAL SYMPOSIUM ON VHL & HEREDITARY
KIDNEY CANCERS
October 26 – 28, 2006**

Four Points Sheraton, London, ON

PROGRAM

October 26: Mechanisms of VHL-dependent tumorigenesis

8:30 Welcoming Remarks
Dr. Stephen Pautler and Dr. Jane Green

Session 1: Mechanisms of VHL-dependent tumorigenesis

8:40 Opening remarks on basics of E3 ubiquitin ligases in cancer
Session Chair: Stephen Lee

8:45 Plenary Seminar, Arnim Pause, Molecular Oncology McGill University, Montreal
Characterization of a von Hippel Lindau Pathway Involved in
Extracellular Matrix Remodeling, Cell Invasion, and Angiogenesis

9:30 Hiroshi Kanno Department of Neurosurgery, Yokohama City University School
of Medicine, Yokohama, Japan
Role of VHL protein in neuronal differentiation of multipotent stem cells and its
application for neuronal regeneration

9.50 Break

10:05 Rachel Giles, University Medical Center, Utrecht, The Netherlands
Zebrafish knockouts for the von Hippel-Lindau (VHL) Tumor Suppressor Gene
Display Key Aspects of the Human Disease Condition.

10:25 Question and Answer Session

10:35 Eamon Maher, Department of Medical and Molecular Genetics, University of
Birmingham School of Medicine, Institute of Biomedical Research, Birmingham,
England
VHL Target Gene Studies

10:55 Jose Claudio Rocha, Head of the national tissue bank, Rio de Janeiro, Brazil
Tissue Bank and VHL

11:15 Question and Answer Session

11:25 Lunch and networking

Session 2: HIF-dependent and -independent tumor suppressor functions of VHL

- 12:30 Opening remarks on HIF-dependent and -independent tumour suppressor functions of VHL
Session chair Arnim Pause
- 12:35 Plenary Seminar, Dr. Patrick H. Maxwell, Professor of Nephrology, Imperial College, London, England
Activation of HIF in humans with VHL mutations
- 13:35 Plenary Seminar, Stephen Lee, Molecular Medicine University of Ottawa, Canada
HIF-activated Oncogenic Pathways in VHL Tumor
- 14:20 Eijiro Nakamura, Department of Urology, Kyoto University, Graduate School of Medicine, Japan
Hypoxia Inducible Factor-Independent Function of pVHL and its Roles on the Development of Familial Pheochromocytoma
- 14:40 Question and Answer Session
- 14:50 Break
- 15:05 Dr. Jodi Maranchie, Univ. of Pittsburgh Medical Center, Pittsburgh, PA, USA
Silencing of the NOX4 NADP(H) Oxidase Abrogates the tumorigenic phenotype of VHL-deficient renal cancer cells.
- 15:25 Dorus A. Mans, Medical Oncology, University Medical Center Utrecht, The Netherlands
The von Hippel-Lindau tumor suppressor regulates primary cilia through kinesin-2 interaction
- 15:45 Mallory Lutz, Department of Microbiology and Immunology, Marion Bessin Liver Research Center and Albert Einstein Comprehensive Cancer Center, Albert Einstein College of Medicine, New York, USA
VHL syndrome renal cystic disease explained by absence of primary cilium due to dysfunctional VHL
- 16:05 Xiaoging Wu, Interdisciplinary Centre for Clinical Research, University of Erlangen-Nuremberg, Erlangen, Germany.
Hypoxic repression of the antiapoptotic protein survivin is dependent on HIF-1 and VHL in primary renal tubular and renal carcinoma cells
- 16:25 Question and Answer Session
- 16:35 Closing remarks for day
Dr. Stephen Pautler and Jane Green

18:00 Dinner

19:00 Keynote Seminar

Gladys Glenn, MD, PhD, National Cancer Institute, Rockville, MD, USA
Hereditary Kidney Cancer Syndromes beyond VHL

October 27: Nature of VHL Disease and Advances in Treatment Protocols

Session 3: Advances in diagnosis and treatment protocols

8:30 Opening remarks in diagnosis and advances in treatment protocols
Session chair, Michael Jewett

8:35 Plenary Seminar

Dr. Emily Chew, NEI, NIH, Bethesda, MD, USA
Translational Research: From the Bench to the Bedside

9:20 Alan Franklin, RFE Pharma Framingham, MA, USA
CAI is a Candidate Molecule to Treat Recalcitrant Retinal Angiomata

9:40 Marie Luise Bisgaard, Copenhagen University, Copenhagen, Denmark
Significance of isolated hemangioblastoma in central nervous system (CNS)

10:00 Sven Gläsker, NINDS, NIH and University of Freiburg, Freiburg, Germany
Hemangioblastomas originate from embryonic hemangioblast progenitor cell

10:20 Question and Answer Session

10:30 Break

10:45 Dr. Hartmut Neumann, University of Freiburg, Freiburg, Germany
Clinical Characteristics of Pheochromocytoma-associated Syndromes

11:05 Eamon Maher, Department of Medical and Molecular Genetics, University of Birmingham School of Medicine, Institute of Biomedical Research, Birmingham, England
Genotype-phenotype studies in VHL Disease

11:25 Mercedes Robledo, Spanish National Cancer Centre, Madrid, Spain
Pheochromocytoma and paraganglioma susceptibility: The five main players. A study protocol based on a Spanish series of patients

Session 3 continued

- 11:45 Graeme Eisenhofer, Clinical Neurocardiology Section, National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, MD, USA
Distinct phenotypes of pheochromocytomas in von Hippel-Lindau compared to other hereditary and sporadic forms of the tumor
Pheochromocytoma: Advances in diagnosis with a consensus in the making
- 12:05 Question and Answer Session
- 12:15 Lunch
- 13:15 Opening remarks on advances in diagnosis and treatment protocols
Session chair, Micheil Innes
- 13:20 Plenary Seminar
Dr. Michael Jewett, University Health Network, Toronto, ON, CANADA
Management of Small Renal Masses
- 14:05 Surena F. Matin, Department of Urology, Univ. of Texas M.D. Anderson Cancer Center, Houston TX USA
How often is minimally invasive therapy possible for patients with VHL? Results from a VHL Clinical Care Center
- 14:25 Hiroshi Kanno Department of Neurosurgery, Yokohama City University School of Medicine, Yokohama, Japan
Management of cerebellar and spinal hemangioblastomas using direct surgery or stereotaxic radiosurgery
- 14:45 Question and Answer Session
- 14:55 Break
- 15:10 Surena F. Matin, Department of Urology, Univ. of Texas M.D. Anderson Cancer Center, Houston TX USA
Targeting von Hippel-Lindau Syndrome: A Single Arm Phase II Trial of Sunitinib Malate in Patients Undergoing Surveillance of VHL-Related Lesions
- 15:30 Sven Gläsker, Surgical Neurology Branch, NINDS, NIH, Bethesda, MD, USA
Developmental effects of VHL deficiency: involvement of Wolffian duct derivatives

Session 3 continued

15:50 Charline Ladroue, Génétique Oncologique EPHE, Institut de cancérologie Gustave Roussy Villejuif et Faculté de Médecine Paris-Sud, France
A family with a novel complex *VHL* germline RXQ/R200W mutation do not present the Chuvash polycythemia phenotype

16:10 Question and Answer Session

16:20 Consensus session. Suggested changes to handbook.

16:50 Closing remarks for day
Stephen Pautler and Jane Green

17:20 Reception and poster session (Room across hall)

C.R.M Lammens, Department of Psychosocial Research and Epidemiology, The Netherlands Cancer Institute, Amsterdam the Netherlands.

Psychosocial aspects of genetic testing in families at high risk of multiple tumors at various sites and ages

Alberto Cascón, Centro Nacional de Investigaciones Oncológicas, Madrid, Spain
Loss of the actin regulator HSPC300 results in renal cell carcinoma protection in von Hippel-Lindau patients

Betty Gardie, Génétique Oncologique EPHE, Institut de cancérologie Gustave Roussy Villejuif et Faculté de Médecine Paris-Sud, France
FH germline mutations in French families affected with hereditary leiomyomatosis renal-cell cancer (HLRCC) or sporadic papillary RCC

Joyce Graff, Executive Director, VHL Family Alliance, Boston, USA, Stéphane Richard Paris, France, Harrmut Neumann Freiburg, Germany and Russell Lonser NIH, Bethesda, USA
Expanding diagnosis of von Hippel-Lindau disease

David Philippe, Service de Neurochirurgie, Le Kremlin Bicêtre, France
Microsurgical resection of spinal cord hemangioblastomas: a report of 41 cases

Xiaoping Wu on behalf of Karl Knaup, Interdisciplinary Centre for Clinical Research and Department of Nephrology & Hypertension, University of Erlangen-Nuremberg, Erlangen, Germany
Differential effects of mTOR inhibition on the HIF pathway and renal cell carcinoma cells

Session 3 continued

- 18:30 Guest Speaker: Fred Sexton, GP, Oncologist and Author, London Regional Cancer Program, U.K. Docs without Socks
- 19:00 Depart for hockey game

October 28: Living with VHL

Session 4: Managing VHL in Your Life

- 8:30 Welcoming Remarks
Stephen Pautler and Jane Green
- 8:35 Opening remarks on living with VHL
Session chair, Jane Green
- 8:40 Plenary Seminar
Michiel Innes, University of Calgary, Calgary, AB
Genetics and VHL
- 9:25 Jane Green, Memorial University of Newfoundland, St. John's, NFLB, Canada
DNA testing: When, why, and how
- 9:55 Marie Luise Bisgaard, Copenhagen University, Copenhagen, DK
Coordinated Care, Management of VHL in Denmark
- 10:25 Break
- 10:40 Taro Shuin, Department of Urology, Kochi University School of Medicine, Kochi
Japan
Health-related QOL in patients with von Hippel-Lindau disease is highly
deteriorated repetitive surgical treatments for VHL-related tumors in the CNS or
visceral organ.
- 11:10 Giuseppe Opocher, Department of Medical and Surgical Sciences, University of
Padova, Italy
The von Hippel-Lindau disease natural history: an 8-year follow up
- 11:40 Gillian Crawford, Wessex Clinical Genetics Service, Southampton, UK
Living with VHL

Session 4 continued

- 12:10 Open Forum Discussion with Patients, Families, Researchers and Clinicians
Facilitated by Stephen Pautler and Jane Green
- 12:40 Closing remarks
Stephen Pautler and Jane Green
- 12:45 Lunch
- 13:30 VHL Leadership Session
Leaders who are present from any country are invited, and anyone who would like to be a leader may also attend
- 15:00 Adjourn