

Scientific Program (version: 9 March 2010)

April 11 (Sunday)

5:30-6:30 pm **Reception at Delzano's Restaurant (walk 1 block south of hotel)**

7:30 pm **Jonathan Braun (Dept of Pathology, UCLA School of Medicine)**

"Welcoming remarks from UCLA"

8:00 pm **Keynote Speaker: Carlo Croce (Ohio State Univ, Columbus)**

"TCL1 in B and T cell malignancies"

April 12 (Monday)

8:45 am - 10:15 am **NEUROPATHOGENESIS OF A-T**
Chairs: Peter McKinnon (St. Jude Hospital, Memphis)
Harry Vinters (UCLA, Los Angeles)

Tom Crawford

"A-T: a developmental or neurodegenerative disorder?"

Harry Vinters

"Ectopic Purkinje Cells in A-T Cerebellae"

Youngsoo Lee & Peter McKinnon (St. Jude Hospital, Memphis)

"ATM and XRCC1 collaborate during cerebellar development"

Zhao-Qi Wang et al. (Jena, Germany)

"Role of NBS1-Atm-mediated DNA damage response in Purkinje cells"

Tej Pandita et al. (Univ Texas Southwest, Dallas)

"MOF depletion results in 'AT-like' neurological phenotype in mice"

10:30 am - 12:00 pm **NEURAL STEM CELLS**
Chairs: Martin Lavin (QIMR, Brisbane, Australia)
Domenico Delia (National Tumor Inst., Milan Italy)

William Lowry (UCLA)

" Probing the potential of human pluripotent stem cells"

Paul Wong and Jeesun Kim (MDAnderson Hospital, Houston)

"Elucidation of neurodegenerative pathways and the potential for neural stem cells to treat neurological disorders"

Masatoshi Takagi et al. (Tokyo, Japan)

"ATM regulates adipocyte differentiation"

Domenico Delia et al. (Milan, Italy)

"Human ATM-deficient neural stem cells as in vitro model systems to study neurodegeneration"

12:00 pm - 1:30 pm

Hailiang Hu et al. (UCLA)

"N-myc upregulated MicroRNA-421 downregulates ATM"

Lunch

1:30 pm - 3:00 pm

IMMUNOBIOLOGY OF A-T/ATM

Chairs: Andre Nussenzweig (NIAID, Bethesda)

Chaim Roifman (Hospital for Sick Kids, Toronto)

Barry Sleckman (Washington U, St. Louis)

"Regulation of DNA end processing by ATM"

JI Loizou et al. (London, UK)

"ATMIN regulates ATM function in class switch recombination and tumour suppression"

Jeremy A. Daniel et al. (NIH, Bethesda)

"PTIP promotes chromatin changes critical for immunoglobulin switch recombination"

Shan Zha et al (Harvard Medical School, Boston)

"ATM-deficient thymic lymphoma is associated with aberrant TCR-delta rearrangement and gene amplification"

Dipanjan Chowdhury et al (Harvard Medical School, Boston)

"A PP4-phosphatase complex dephosphorylates RPA2 facilitating homologous recombination-mediated DNA repair"

3:30 pm - 5:00 pm

TUMOR FORMATION AND CANCER PREDISPOSITION IN

A-T

Chairs: Fred Alt (Harvard Medical School, Boston)

Tatjana Stankovic (Univ Birmingham, UK)

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"The role of ATM in lymphoid development and the biological behaviour of sporadic lymphoid tumors"

Oscar Fernandez-Capetillo et al. (Madrid, Spain)

"Synthetic lethality of ATR and c-myc"

Darius Filsuf et al. (NIH, Bethesda)

"Loss of ATM kinase activity leads to embryonic lethality in mice"

Elaine Willmore (Newcastle Cancer Center, UK)

"The DNA-dependent protein kinase inhibitor, NU7441, chemosensitizes ATM mutant chronic lymphocytic leukaemia cells"

5:00 pm - 6:00 pm

POSTER VIEWING

6:00 pm - 7:30 pm

Dinner

7:30 pm - 8:35 pm

STRUCTURE/FUNCTION ANALYSES OF ATM

Chairs: Patrick Concannon (Univ Virginia,

Charlottesville)
Sean Tavtigian (Univ of Utah, Salt Lake City)

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"Evaluating the consequences of missense variants"

John A. Tainer (Scripps Inst., San Diego)
"MRE11-RAD50-NBS1 functional interactions with ATM and other partners"

Laura Elnitski et al (NHGRI, Bethesda)
"Ascertainment and prediction of genomic sequence variants causing exon skipping events"

8:45 pm - 10:15 pm

LATE-BREAKING REPORTS

Chairs: Stephen Meyn (Hospital for Sick Children, Toronto)

Susan Lees-Miller (Univ Calgary, Canada)

Fred Alt (Harvard Medical School, Boston)
"Overlapping functions of ATM and XLF/Cernunnos in non-homologous end-joining"

Tanya Paull (Univ Texas, Austin)
"ATM activation by oxidative stress"

Susan Lees-Miller et al. (Univ Calgary, Canada)
"ATM-deficiency sensitizes mantle cell lymphoma cells to PARP-1 inhibitors"

10:15 pm - 11:15 pm

POSTER VIEWING (Hosted Bar)

April 13 (Tuesday)

8:45 pm - 10:15 am

RADIOBIOLOGY OF A-T AND ATM DEFICIENT MODELS

Chairs: Steve Jackson (Univ Cambridge, UK)

Yosef Shiloh (Tel Aviv Univ, Israel)

John Petrini (Mem Sloan Kettering Cancer Center, New York)

"The Mre11 complex and checkpoint responses to DNA damage"

Vincenzo Costanzo and Claudia Cosentino (London, UK)

"ATM-dependent regulation of pathways promoting cell survival and DSB repair"

Hava Segal-Raz et al. (Tel Aviv Univ, Israel)
"ATM-mediated phosphorylation of polynucleotide kinase: a direct link between ATM and DNA repair"

Junya Kobayashi et al. (Kyoto, Japan)
"The role of novel H2AX-binding factor, nucleolin in ATM-dependent DNA damage response"

10:30 am - 12:00 pm

I. UPSTREAM EVENTS OF DSB SIGNALING AND REPAIR

**Chairs: Tanya Paull (U Texas, Austin)
Shuki Mizutani (TMDU, Tokyo, Japan)**

Jiri Lukas (Danish Cancer Center, Copenhagen, Denmark)

"Dynamic organization of DNA damage response by phosphorylation and ubiquitin signaling"

Brendan D. Price et al. (Harvard Medical School, Boston)

"How chromatin links MRN and Tip60 to the activation of ATM"

Roger A. Greenberg (Univ Pennsylvania Med Sch, Philadelphia)

"An ATM-dependent transcriptional silencing program is transmitted through chromatin modification in cis to DNA double strand breaks"

Sachin Katyal et al. (St. Jude Hospital, Memphis)

"Linking ATM to DNA single-strand break repair"

12:00 pm - 1:30 pm

Lunch

1:30 pm - 3:00 pm

II. UPSTREAM EVENTS OF DSB SIGNALING AND REPAIR

**Chairs: Jiri Lukas (DCI, Copenhagen, Denmark)
Junjie Chen (M.D. Anderson, Houston)**

Steve Jackson (Univ. Cambridge, UK)

"Responses to DNA damage in the context of chromatin"

Jean Gautier et al. (Columbia Univ, New York)

"Cell cycle regulation of DNA double-strand break resection"

Steve Meyn and Peter Bradshaw (Toronto, Canada)

"Human ATM protein displays distinct spatial and temporal dynamics at sites of induced DNA damage"

Zhongsheng You et al. (Washington Univ, St. Louis)

"CTIP, a tip for integrating DNA damage checkpoint and cell cycle control with DNA repair"

Samuel Bunting et al. (NIH, Bethesda)

"53BP1 suppresses homologous recombination in BRCA1-deficient cells by inhibiting ATM-mediated resection at DNA breaks"

3:30 pm - 5:00 pm

AT-RELATED SYNDROMES (radiosensitive and neurodegenerative)

Chairs: Penny Jeggo (Univ Sussex, UK)

Thilo Dork (Hannover Medical School, Germany)

Martin F. Lavin et al. (Brisbane, Australia)

"Ataxia Oculomotor Apraxia Type 2 (AOA2): a model system to investigate neurodegeneration"

Penny Jeggo (Sussex, UK)

"DNA double strand break repair in A-T and its related disorders, ATLD, NBS, and Riddle Syndrome; the link to heterochromatin"

Shareef Nahas et al. (UCLA)

"Is the colony survival assay the only "gold standard" for radiosensitivity?"

Benjamin P.C. Chen et al. (Univ Texas Southwestern, Dallas)

"Severe anemia and hematopoietic stem cell failure in DNA-PKCS mutant mice associated with deficiencies in multiple DNA repair pathways"

Dianne Watters et al. (Brisbane, Australia)

"Interaction between ATM and the tuberous sclerosis complex"

5:00 - 6:00 pm

Special Honorary Lecture: Richard A. Gatti

"New horizons in A-T pathogenesis and treatment"

6:00 - 7:00 pm

POSTER VIEWING (Hosted Bar)

8:00 pm

Banquet/Entertainment

April 14 (Wednesday)

8:00 am - 8:30 am

Organizing Committee meeting – selection of next venue

8:45 am - 10:15 am

PROMISING THERAPEUTIC APPROACHES FOR A-T
Chairs: Gerry Berry (Harvard, Boston)
Susan Perlman (UCLA, Los Angeles)

Gerard S. Shadel (Yale Medical School, New Haven)

"Mitochondrial dysfunction and signaling in A-T: a new therapeutic target?"

Liutao Du et al. (UCLA)

"Arginine-rich cell penetrating peptide dramatically enhances AMO-mediated correction of aberrant splicing in ATM transcripts"

Claudio Pignata et al. (Naples, Italy)

"The effects of betamethasone therapy on neurological functions in A-T patients"

Andreea Nissenkorn et al. (Tel Aviv, Israel)

"Amantadine sulfate for treatment of movement disorder in A-T"

Stefan Zielen et al. (Frankfurt, Germany)

"Interstitial lung disease in A-T"

10:30 am - 12:00 pm

WRAP UP DISCUSSION

Danielo Tagle (NINDS, Bethesda)

"NIH funding opportunities for A-T Research"

**Panel Discussion: Lavin, Lowry, McBride, McKinnon,
Meyn, Shiloh, Whitehouse**
"Future directions"

Focus Topics (i.e., timely issues) will be developed by the Co-Chairs. These will be used to target discussions. They will be introduced by one of the Chairs at the beginning of each session.

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