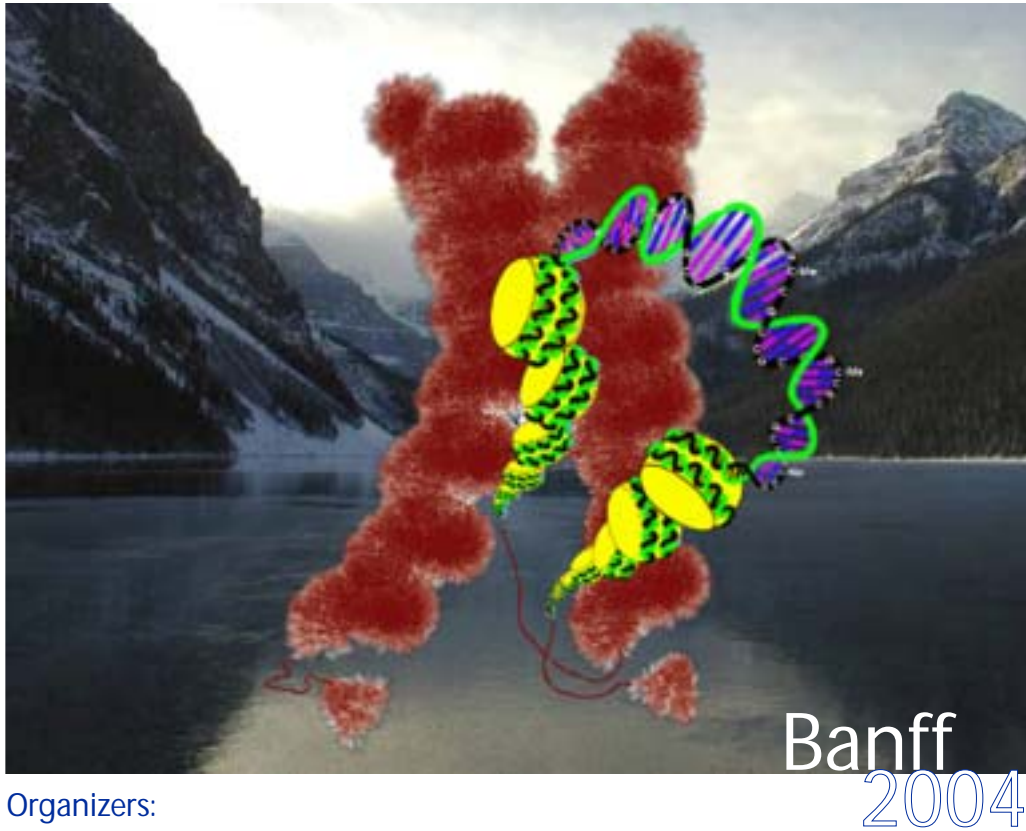


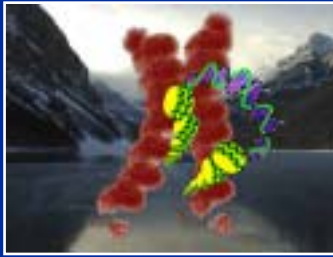
4th International Conference on
Unstable Microsatellites and Human Disease



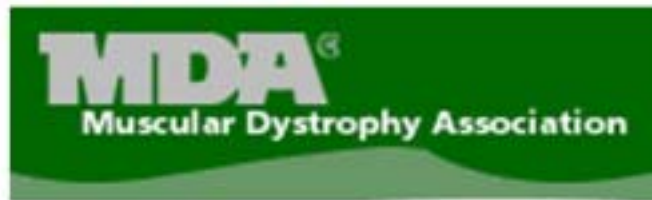
Organizers:
Christopher E. Pearson, Ph.D
Robert G. Korneluk, Ph.D

February 28 to March 4, 2004
The Banff Centre, Banff, Alberta, Canada

Website: www.microsatellites.ca



4th International Conference on
Unstable Microsatellites and Human Disease
SPONSORS



THE 4TH INTERNATIONAL CONFERENCE ON
UNSTABLE MICROSATELLITES AND HUMAN DISEASE

PROGRAM SCHEDULE

(All Speaker and Workshop Presentations will be held in Max Bell Auditorium)
(All meals served in Main Dining Room)

Saturday, February 28

- 5:30 p.m. Buffet Dinner
- 6:50 p.m. **Opening Comments**
Christopher E. Pearson & Robert G. Korneluk
- Keynote Addresses - Replication & Repair**
Chair: Christopher E. Pearson
- 7:00-8:00 p.m. **Melvin DePamphilis**
DNA Replication in Eukaryotic Cells
- 8:00-9:00 p.m. **Philip C. Hanawalt**
Subpathways of Nucleotide Excision Repair and their Regulation: Roles in Human Genetic Disease
- 9:00-11:00 p.m. Reception

Sunday, February 29

- 7:00-8:00 a.m. Breakfast Buffet
- 8:00-4:00 p.m. **Poster Set-Up** (*Max Bell 252*)
(all posters will be up through to Wednesday afternoon)
- Instability in Humans and Mice I**
Chair: Genevieve Gourdon
- 8:00-8:25 a.m. **Norman Arnheim**
What Cell Compartments of the Human Germline Exhibit Trinucleotide Repeat Expansion Mutations in Huntington Disease Patients?
- 8:25-8:50 a.m. **Genevieve Gourdon**
How and When? About CTG Repeat Instability in Transgenic Mice
- 8:50-9:15 a.m. **Cynthia T. McMurray**
The Mechanism by Which Msh2 Causes Expansion During Transmission and Embryonic Development
- 9:15-9:45 a.m. Coffee Break

Instability in Humans and Mice II

Chair: Be Wieringa

- 9:45-10:10 a.m. **Be Wieringa**
Cellular and Animal Models For DMI: Causes and Consequences of Somatic Variation in (CTG)_n Repeat- and DMPK Gene-dose
- 10:10-10:35 a.m. **Darren G. Monckton**
Can the Repeat Expansion Disorders be Cured?
- 10:35-11:00 a.m. **Anne Messer**
Antibody Therapeutics for Huntington's Disease
- 11:00-4:00 a.m. Free Time (Note: Ski shuttles leave hotel at 11:30 a.m.)
- 4:00-4:30 p.m. Refreshments (*Max Bell 252*)
- 4:15-6:15 p.m. **Poster Session I** (Even Numbers at *Max Bell 252*)
- 6:15-7:30 p.m. Buffet Dinner

Cis - and Trans-Factors of Instability

Chair: Albert LaSpada

- 8:00-8:25 p.m. **Yeon-Soo Seo**
Okazaki Fragment Processing and Trinucleotide Repeat Stability in S. cerevisiae; A Critical Role of the N-Terminal Domain of DNA2 in Maintenance of Trinucleotide Repeats
- 8:25-8:50 p.m. **Robert Lahue**
A New Protein Modulator of Triplet Repeat Instability in Yeast
- 8:50-9:15 p.m. **Albert LaSpada**
Molecular Determinants of CAG/CTG Repeat Instability: Identification of Cis-Acting Elements and Trans-Acting Factors
- 9:15-9:40 p.m. **Dmitry Gordenin**
Genome Instability Associated with Defects in DNA Polymerase Proofreading, Mismatch Repair and Okazaki Maturation

Monday, March 1

- 7:00-8:00 a.m. Breakfast Buffet
- Mechanisms of Instability**
Chair: Christopher E. Pearson
- 8:00-8:25 a.m. **Sergei Mirkin**
Replication and Stability of Friedreich's Ataxia (GAA)_n Repeats
- 8:25-8:50 a.m. **Peter M. Lansdorp**
Loss of G-Rich DNA with Replication
- 8:50-9:15 a.m. **Christopher E. Pearson**
A Human Cell-Free System to Process Slipped-Strand DNAs
- 9:15-9:45 a.m. Coffee Break

Instability and Chromosomes

Chair: Jack D. Griffith

- 9:45-10:10 a.m. **Jack D. Griffith**
Telomere Replication and Maintenance: Parallels with the Unstable Disease-Related Nucleotide Repeats
- 10:10-10:35 a.m. **Aaron Bensimon**
A New Paradigm for Ribosomal RNA Gene Arrays Incorporating Palindromic Pseudogenes
- 10:35-11:00 a.m. **Richard Festenstein**
Overcoming and Inducing Epigenetic Silencing in Mammals
- 11:00-4:00 p.m. Free Time (Note: Ski shuttles leave hotel at 11:30 a.m.)
- 4:00-4:30 p.m. Refreshments
- 4:30-6:15 p.m. **Workshop I** (Eight 10-min. talks)
- 6:15 -7:30 p.m. Buffet Dinner

Instability, Apoptosis & Therapy

Chair: Robert G. Korneluk

- 8:00-8:25 p.m. **Charles Thornton**
Molecular Pathogenesis of Myotonic Dystrophy in Brain
- 8:25-8:50 p.m. **Harry T. Orr**
Repair in Conditional SCA1 Mice
- 8:50-9:15 p.m. **Bronwen Connor**
Increased Progenitor Cell Proliferation and Striatal Neurogenesis in Huntington's Disease
- 9:15 -9:40 p.m. **Jack Puymirat**
Potential Therapeutic Tools for Myotonic Dystrophy
- 9:40-10:05 p.m. **Robert G. Korneluk**
Trinucleotide Repeats, Apoptosis & Therapy

Tuesday, March 2

- 7:00-8:00 a.m. Breakfast Buffet

Large Repeat Units & HDL2

Chair: Tetsuo Ashizawa

- 8:00-8:25 a.m. **Tetsuo Ashizawa**
Somatic and Germ-line Instability of the ATTCT Repeat in Spinocerebellar Ataxia Type 10
- 8:25-8:50 a.m. **Rune Frants**
Facioscapulohumeral Muscular Dystrophy; Macrosatellite Repeat Instability
- 8:50-9:15 a.m. **Russell Margolis**
Huntington's Disease-like 2: An Update
- 9:15 -9:45 a.m. Coffee Break

Instability, Transcription & Toxic-RNAs

Chair: Guy Rouleau

- 9:45-10:10 a.m. **Guy Rouleau**
Role of Transcriptional Slippage of Long CAG Tracts in Neurological Disease
- 10:10-10:35 a.m. **Maurice Swanson**
A Loss-of-Muscleblind mouse model for Myotonic Dystrophy
- 10:35-11:00 a.m. **Laura Ranum**
RNA Pathogenesis in Myotonic Dystrophy Type 2 and Spinocerebellar Ataxia Type 8.
- 11:00-4:00 p.m. Free Time (Note: Ski shuttles leave hotel at 11:30 a.m.)
- 4:00-4:30 p.m. Refreshments
- 4:15-6:15 p.m. **Poster Session II** (Odd Numbers at *Max Bell 252*)
- 6:15-7:30 p.m. Buffet Dinner

Instability and Fragile Sites

Chair: David L. Nelson

- 8:00-8:25 p.m. **Pietro Chiurazzi**
Epigenetic Changes After Pharmacological Reactivation of the FMR1 Gene, Silenced in the Fragile X Syndrome
- 8:25-8:50 p.m. **Robert Richards**
DNA Instability at the FRA16D Locus and its Functional Contribution to Cancer
- 8:50 -9:15 p.m. **Peng Jin /Stephen Warren**
Fragile X & Fragile X Tremor Ataxia Syndromes: RNA-Mediated Diseases
- 9:15-9:40 p.m. **David Nelson**
Animal Models for Mutations in FMR1 and Paralogues
- 9:40-10:30 p.m. **Workshop II** (Five 10-minute talks)

Wednesday, March 3

- 7:00-8:00 a.m. Breakfast Buffet
- FRAXA & FRAXA-Associated Tremor/Ataxia Syndrome**
Chair: Ben Oostra
- 8:00-8:25 a.m. **Paul J. Hagerman**
Fragile X-Associated Tremor/Ataxia Syndrome (FXTAS): An Aging Face of the FMR1 Gene
- 8:25-8:50 a.m. **Ben Oostra**
The FMR1 Gene: Demethylation of the Hypermethylated FMR1 Promoter and the Lack of FMR1 Protein Causing Enhanced LTD at Enlarged Purkinje Cell Spines Resulting in Motor Learning Deficits
- 8:50-9:15 a.m. **Hirokazu Furuya**
Some Flavonoids Prevent Cytotoxicity and CIS-Effect of Expanded CTG Repeats in Stable Transformant of PC12 Neuronal Cell

- 9:45 -11:30 a.m. **Workshop III** (Eight 10-minute talks)
- 11:30-11:50 a.m. Wrap-Up
- 12:00 -6:30 p.m. Tour of Banff or Free Time (Tour Buses Depart at 12:30)
- 6:30 p.m. Cocktails
- 7:30 -11:00 p.m. Closing Banquet and Dance

Thursday, March 4

- 7:00- 9:00 a.m. Breakfast Buffet
- Departure