

# The 3<sup>rd</sup> International Conference

## Unstable Microsatellites and Human Disease



**Noordwijkerhout  
The Netherlands**

**April 21th – April 25th 2001.**

Organising committee:  
**David Nelson (Houston)**  
**Ben Oostra (Rotterdam)**  
**Be Wieringa (Nijmegen)**

**The meeting place:  
Golden Tulip Conference Hotel Leeuwenhorst  
Noordwijkerhout,  
The Netherlands.**

The 3rd International Conference on

# "Unstable Microsatellites & Human Disease"

April 21 – 25 2001

Golden Tulip Conference Hotel Leeuwenhorst



Noordwijkerhout  
The Netherlands



## **Organisers:**

David Nelson (Houston)  
Ben Oostra (Rotterdam)  
Be Wieringa (Nijmegen)

## **Sponsored by:**

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# Program "Unstable Microsatellites and Human Disease" meeting Noordwijkerhout 2001

**Saturday April 21**

**Key note address:**

**Chairman: B.Wieringa**

**20.00      Jeffreys:**      Human minisatellites, repeat instability and meiotic recombination

**Get together**

**Sunday April 22**

**Morning:      *Session IA: Instability: Cis and trans acting (nuclear) factors***

**Chairman: B.A.Oostra**

**8.45      Lilley**      DNA structure and its recognition by proteins  
**9.20      Wells**      Sticky DNA, Transcription Inhibition, and Friedreich's Ataxia  
**9.55      Lindahl**      Exonucleolytic processing of unpaired DNA termini in mammalian cells

**10.30**      *Coffee break*

**11.00      Fry**      Protein Interaction with the (CGG) Trinucleotide Repeat: Potential Bearing on Repeat Expansion

**11.40      Resnick**      Yeast as an in vivo test tube to study repeat-associated instability and disease

**12.30**      *Lunch*

**Afternoon:      *Session IB: Instability: Cis and trans acting (nuclear) factors***

**Chairman: M.Resnick**

**14.00      Fishel**      Mismatch Repair and Microsatellite Instability: the mechanics of a molecular switch

**14.35      Te Riele**      Specific and redundant functions of mismatch repair proteins in mutation avoidance and suppression of cancer

**15.10 – 15.30**      *Coffee / tea break*

**15.30 – 17.30**      **Oral presentations (selected on the basis of abstracts)**

**Evening:**

**18.00**      *Dinner*

**20.00 – 21.30**      **Poster session**

## Monday April 23

- Morning: Session IC: Instability: Cis and trans acting (cellular) factors**  
**Chairman: D.Lilley**
- 8.45 McMurray Trinucleotide expansion in haploid germ cells of transgenic animals by gap repair.
- 9.20 Moncton Cis and trans-acting genetic modifiers of triplet repeat stability
- 9.55 Gourdon CTG repeat instability in its human DM environment :  
modelisation in transgenic mice

10.30 – 11.00 *coffee break*

- 11.00 Zakian Effects of trinucleotide repeats on replication fork progression and chromosome breakage in *Saccharomyces*
- 11.40 Shelbourne Mutation length and cell vulnerability in Huntington's disease

12.30 *Lunch*

**Afternoon: Social program: Tulip fields**

14.00 **Departure to Tulip fields**

18.00 *Diner*

**Evening: Session II: RNA-related effects**  
**Chairman: G.Gourdon**

- 20.00 Warren Studies of FMRP/RNA interaction
- 20.35 Mahadevan Myotonic Dystrophy: A New Twist on an Old Tail
- 21.10 Swanson RNA Dominance in Myotonic Dystrophy: Role for Human Muscleblind Proteins

## Tuesday April 24

**Morning: Session IIIA: Molecular pathology**  
**Chairman: S.Warren**

- 8.45 Orr The Molecular Basis of SCA1
- 9.20 Wieringa. DMPK gene-instability and -function(s) revisited.
- 9.55 Pandolfo Causes and consequences of frataxin deficiency

10.30 – 11.00 *Coffee break*

- 11.00 Tsuji Molecular pathogenesis of dentatorubral-pallidoluysian atrophy
- 11.40 Oostra The FMR1 gene: repeat instability and function.

12.30 *Lunch*

**Afternoon:**

**14.00 - 16.00 Oral presentation of selected abstracts**

*16.00 - 17.30 Coffee break and Posters*

**Evening:**

*18.00 Diner: Indonesian Rice table*

**Evening: Session IIIB: Molecular pathology  
Chairman: H.Orr**

**20.30 Nelson** FMR1 CGG repeat instability in YAC transgenic mice  
**21.10 MacDonald** Molecular genetics in the mouse to uncover the  
biochemical polyglutamine-expansion trigger in Huntington's  
disease

**Wednesday April 25**

**Morning: Session IV: Past, present and future: Disease, diagnosis and therapeutic  
promises  
Chairman: D.Nelson**

**8.45 Messer** Engineering antibodies for anti-Huntington's Disease therapies  
**9.20 Thornton** Muscle disease in human myotonic dystrophy and a transgenic  
model

*10.00 – 10.30 Coffee break*

**10.30 Richards** Causes and Consequences of Chromosomal Fragile Sites  
**11.10 Mandel** **To be announced**

*12.00 Lunch*