Screening and Outcomes in Biliary Atresia

Sponsored by NIDDK, HRSA, ORD

September 11-12, 2006

Aims: To assess current knowledge of the pathogenesis, clinical presentation, natural history, surgery and outcomes of biliary atresia in the United States and elsewhere, focusing on the role of newborn screening and means of early detection of biliary atresia and the evidence for and against the effectiveness and the medical outcomes of methods of screening. This meeting also will review the current understanding of the pathogenesis of biliary atresia and possibility of new approaches to early detection using a better understanding of its pathogenesis and clinical course.

Organizers: Ronald Sokol (University of Colorado), Ross Shepherd (Washington University), Riccardo Superina (Children's Hospital, Chicago), Patricia Robuck (NIDDK), Jay Hoofnagle (NIDDK), Stephen Groft (Office of Rare Diseases), Marie Mann (Health Research Services Administration), Diane Sumner (Children's Liver Association for Support Services), and Ellis Green (American Liver Foundation)

Day One: Monday, September 11, 2006

| 8:00 – 8:15 a.m. | Welcome and Introduction | Ronald Sokol (University of Colorado) |
|--------------------|---|---|
| Session 1: | Clinical Course of Biliary Atresia | |
| 8:15 – 8:35 a.m. | Neonatal Jaundice | Jeff Maisels (William Beaumont Hospital, Michigan) |
| 8:35 – 8:55 a.m. | Epidemiology and Clinical Features of Biliary Atresia | Barbara Haber (Children's Hospital of Philadelphia) |
| 8:55 – 9:15 a.m. | Diagnosis of Biliary Atresia | Peter Whitington (Children's Memoria Hospital, Chicago) |
| 9:15 – 9:35 a.m. | Pathology of Biliary Atresia | Pierre Russo (Children's Hospital of Philadelphia) |
| 9:35 – 10:00 a.m. | Discussion | |
| 10:00 – 10:20 a.m. | Break | |

Session 4:

| Session 2: | Hepatoporto-enterostomy and Outcomes for Biliary Atresia | | |
|-------------------------|--|--|--|
| 10:20 – 10:40 a.m. | The Kasai Procedure | Riccardo Superina (Children's Memorial Hospital, Chicago) | |
| 10:40 – 11:00 a.m. | Outcomes of Biliary Atresia in the United States | Benjamin Shneider (Mt. Sinai Medical Center, NY) | |
| 11:00 – 11:20 a.m. | Outcomes of Biliary Atresia in Canada | Richard Schreiber (University of British Columbia) | |
| 11:20 – 11:40 a.m. | Outcomes of Biliary Atresia in the United Kingdom | Mark Davenport (Kings College Hospital, London) | |
| 11:40 a.m. – 12:00 noon | Outcomes of Biliary Atresia in France | Christophe Chardot (Cantonal University Hospital of Geneva, Switzerland) | |
| 12:00 noon – 12:30 p.m. | Discussion | | |
| 12:30 – 1:30 p.m. | Lunch | | |
| Day One: Afternoon | | | |
| Session 3: | Biliary Atresia: New Insights Into Etiology | | |
| 1:30 – 1:50 p.m. | Developmental Biology of the Biliary Tract | Michael Pack (University of Pennsylvania) | |
| 1:50 – 2:10 p.m. | Viral and Immunological Pathogenesis | Cara Mack (University of Colorado) | |
| 2:10 – 2:30 p.m. | Genetics of Biliary Atresia | David Perlmutter (University of Pittsburgh) | |
| 2:30 – 2:50 p.m. | Proteomics and Biomarkers | Robert Heuckeroth (Washington University, St. Louis) | |
| 2:50 – 3:10 p.m. | Discussion | | |
| 3:10 – 3:40 p.m. | Break | | |

September 11, 2006 2

Towards Improving Outcomes of Biliary Atresia

| 3:40 – 4:00 p.m. | Current Status of Newborn Screening | Brad Therrell (University of Texas Health Sciences Center, San Antonio) |
|------------------|--|---|
| 4:00 – 4:20 p.m. | Japanese Experience With Corticosteroids and Other Therapies After Hepatoportoenterosotomy | Masaki Nio (Miyagi Children's Hospital Sendai, Japan) |
| 4:20 – 4:40 p.m. | Nutritional Aspects of Biliary Atresia | Ross Shepherd (Washington University) |
| 4:40 – 5:00 p.m. | Neurodevelopmental Outcomes in Biliary Atresia | Estella Alonso (Children's Memorial Hospital, Chicago) |
| 5:00 – 5:30 p.m. | Discussion | |
| 5:30 – 8:00 p.m. | Reception and Posters | |

Day Two: Tuesday, September 12, 2006

10:45 a.m. – 12:00 noon

| Day 1 wo: 1 desday, September 12, 2000 | | | |
|--|---|---|--|
| Session 5: | Early Diagnosis and Screening for Biliary Atresia | | |
| 8:30 – 8:50 a.m. | Obstacles To Making an Early Diagnosis | Ronald Sokol (University of Colorado) | |
| 8:50 – 9:10 a.m. | Use of Serum and Urinary Bile Acids for Neonatal Screening for Biliary Atresia | James Heubi (University of Cincinnati) | |
| 9:10 – 9:30 a.m. | Serum Bilirubin as a Means of Screening for Biliary Atresia | Deirdre Kelly (Birmingham Children's Hospital, United Kingdom) | |
| 9:30 – 10:15 a.m. | Stool Color Cards for Screening for Biliary Atresia | Akira Matsui (Jichi University, Japan) Mei-Hwei Chang (National Taiwan University Hospital) Margarita Ramonet (Hospital Nacional Alejandro Posadas, Argentina) | |
| 10:15 – 10:45 a.m. | Discussion/Break | | |
| Session 6: | Research Agenda for the Future | | |

September 11-12, 2006 3

Organizers

Research Agenda Summary