In This Issue:

This issue of the PSC Partners Seeking a Cure Newsletter is devoted to a brief description of the Primary Sclerosing Cholangitis Research Workshop, held at the National Institutes of Health, Lister Hill Auditorium, Bethesda, Maryland, from September 19-20, 2005. The workshop was attended by many of the leading PSC researchers from around the world. It was sponsored by the NIH, NIDDK, Office of Rare Diseases, and the Morgan Foundation (http://www.pscfoundation.org/).

All of the presentations and discussions were videotaped, and the video-casts are available for viewing at the following URLs:

Primary Sclerosing Cholangitis Research Workshop (Day 1)
http://videocast.nih.gov/ram/niddk091905.ram
Time: 6h:35min
Format: RealVideo

Primary Sclerosing Cholangitis Research Workshop (Day 2)
http://videocast.nih.gov/ram/niddk092005.ram
Time: 5h:42min
Format: RealVideo

There is really no substitute for viewing and listening to these 12 hours of informative presentations and discussions! You will need to install RealPlayer software on your computer in order to view these video-casts. The Free RealPlayer software is available from:
http://videocast.nih.gov/

You might wish to print off the agenda for the two days of talks to help guide you during the video-casts. The agenda is available from:
https://www.niddk.nih.gov/fund/other/primarysclerosing/

The organizing committee plans to publish an outline of the conference in a future issue of the journal “Hepatology”.

Again, congratulations to the organizing committee, the NIH/NIDDK, Office of Rare Diseases and the Morgan Foundation for putting on this important event. It represents a significant milestone in PSC research, and a major advance in the coordination of research efforts in this chronic liver disease.

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bicarbonate-rich bile, and 3) induction of detoxification enzymes and alternative pumps for bile acids.

A second personal highlight was the talk by Dr. David Adams on T cell immunity and PSC, and his recent work on long-lived mucosal T cell migration between gut and liver. The migration of these cells may be responsible for sustaining PSC. His discoveries in this area, if corroborated by others, may lead to some novel approaches to PSC treatment.

A third highlight was meeting Dr. Chris Bowlus and Dr. Chris Aoki from U.C. Davis, and hearing more about their work on PSC genetics, and the p-ANCA antibodies associated with PSC. Dr. Bowlus is particularly interested in contacting twins with PSC (one or both of whom are affected) to assist in identifying genetic factors. Please visit www.psc-literature.org for contact information for PSC genetic studies underway at both U.C. Davis and Mayo Clinic.

A fourth highlight was meeting Dr. Steven Freedman and his colleague, Dr. Harpreet Pall, and learning more about their recently initiated trial on DHA in PSC. DHA (docosahexaenoic acid; an omega-3 fatty acid found in fish oil) has shown benefit in cystic fibrosis patients, and in a mouse model of sclerosing cholangitis: the cystic fibrosis transmembrane conductance regulator (CFTR) deficient mouse. When given colitis, these CFTR-deficient mice develop sclerosing cholangitis, and are significantly protected from bile duct injury by oral DHA supplementation.

A fifth highlight was the announcement, by Dr. Dennis Black, concerning the initiation of a PSC registry which will greatly facilitate future collaborative research. This registry will consist of a databank of PSC patient history and DNA to facilitate studies on the genetics of PSC. The Morgan Foundation is taking the lead on this important initiative.

Dr. Roger Chapman discussed pathogenic mechanisms in PSC and concluded that it is probable that multiple interacting genetic and environmental factors contribute to the development of PSC, but the question of whether it is a true autoimmune disease remains unanswered. The male predominance and the association with lack of smoking remain unexplained.

Dr. Peter Donaldson gave an update on PSC genetics. He explained the established importance of the major histocompatibility complex (MHC) genes in determining the risk of developing PSC, and then discussed additional genes as candidate susceptibility genes, including the bile transporters (BSEP and MDR3), CFTR, and the CARD genes (CARD4 and CARD15) implicated in inflammatory bowel diseases.

There was much discussion about whether or not pediatric PSC is a different disease from adult PSC. Both Dr. Eve Roberts and Dr. Giorgina Mieli-Vergani noted that the presentation in children often looks much more like autoimmune hepatitis. Dr. Giorgina Mieli-Vergani suggested that scleros-