THE OFFICIAL PATIENT'S SOURCEBOOK *on*

PRIMARY SCLEROSING CHOLANGITIS

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Dedication

To the healthcare professionals dedicating their time and efforts to the study of primary sclerosing cholangitis.

Acknowledgements

The collective knowledge generated from academic and applied research summarized in various references has been critical in the creation of this sourcebook which is best viewed as a comprehensive compilation and collection of information prepared by various official agencies which directly or indirectly are dedicated to primary sclerosing cholangitis. All of the *Official Patient's Sourcebooks* draw from various agencies and institutions associated with the United States Department of Health and Human Services, and in particular, the Office of the Secretary of Health and Human Services (OS), the Administration for Children and Families (ACF), the Administration on Aging (AOA), the Agency for Healthcare Research and Quality (AHRQ), the Agency for Toxic Substances and Disease Registry (ATSDR), the Centers for Disease Control and Prevention (CDC), the Food and Drug Administration (FDA), the Healthcare Financing Administration (HCFA), the Health Resources and Services Administration (HRSA), the Indian Health Service (IHS), the institutions of the National Institutes of Health (NIH), the Program Support Center (PSC), and the Substance Abuse and Mental Health Services Administration (SAMHSA). In addition to these sources, information gathered from the National Library of Medicine, the United States Patent Office, the European Union, and their related organizations has been invaluable in the creation of this sourcebook. Some of the work represented was financially supported by the Research and Development Committee at INSEAD. This support is gratefully acknowledged. Finally, special thanks are owed to Tiffany LaRochelle for her excellent editorial support.

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About ICON Health Publications

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- The Official Patient's Sourcebook on Appendicitis
- The Official Patient's Sourcebook on Autoimmune Hepatitis
- The Official Patient's Sourcebook on Bacteria and Foorborne Illness
- The Official Patient's Sourcebook on Barrett's Esophagus
- The Official Patient's Sourcebook on Celiac Disease
- The Official Patient's Sourcebook on Cirrhosis of the Liver
- The Official Patient's Sourcebook on Constipation
- The Official Patient's Sourcebook on Crohn Disease
- The Official Patient's Sourcebook on Cyclic Vomiting Syndrome
- The Official Patient's Sourcebook on Diarrhea
- The Official Patient's Sourcebook on Diverticular Disease
- The Official Patient's Sourcebook on Fecal Incontinence
- The Official Patient's Sourcebook on Gallstones
- The Official Patient's Sourcebook on Gas
- The Official Patient's Sourcebook on Gastritis
- The Official Patient's Sourcebook on Gastroparesis
- The Official Patient's Sourcebook on Hemolytic Uremic Syndrome
- The Official Patient's Sourcebook on Hemorrhoids
- The Official Patient's Sourcebook on Hepatitis A
- The Official Patient's Sourcebook on Hepatitis B
- The Official Patient's Sourcebook on Hepatitis C
- The Official Patient's Sourcebook on Hiatal Hernia
- The Official Patient's Sourcebook on Hirschsprung
- The Official Patient's Sourcebook on Indigestion
- The Official Patient's Sourcebook on Inguinal Hernia
- The Official Patient's Sourcebook on Intestinal Pseudo-obstruction
- The Official Patient's Sourcebook on Irritable Bowel Syndrome
- The Official Patient's Sourcebook on Lactose Intolerance
- The Official Patient's Sourcebook on Ménétrier
- The Official Patient's Sourcebook on Pancreatitis
- The Official Patient's Sourcebook on Peptic Ulcer
- The Official Patient's Sourcebook on Porphyria
- The Official Patient's Sourcebook on Primary Biliary Cirrhosis
- The Official Patient's Sourcebook on Proctitis
- The Official Patient's Sourcebook on Rapid Gastric Emptying

- The Official Patient's Sourcebook on Short Bowel Syndrome
- The Official Patient's Sourcebook on Ulcerative Colitis
- The Official Patient's Sourcebook on Whipple Disease
- The Official Patient's Sourcebook on Wilson's Disease
- The Official Patient's Sourcebook on Zollinger-ellison Syndrome

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INTRODUCTION

Overview

Dr. C. Everett Koop, former U.S. Surgeon General, once said, "The best prescription is knowledge."[1] The Agency for Healthcare Research and Quality (AHRQ) of the National Institutes of Health (NIH) echoes this view and recommends that every patient incorporate education into the treatment process. According to the AHRQ:

> Finding out more about your condition is a good place to start. By contacting groups that support your condition, visiting your local library, and searching on the Internet, you can find good information to help guide your treatment decisions. Some information may be hard to find—especially if you don't know where to look.[2]

As the AHRQ mentions, finding the right information is not an obvious task. Though many physicians and public officials had thought that the emergence of the Internet would do much to assist patients in obtaining reliable information, in March 2001 the National Institutes of Health issued the following warning:

> The number of Web sites offering health-related resources grows every day. Many sites provide valuable information, while others may have information that is unreliable or misleading.[3]

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[1] Quotation from **http://www.drkoop.com**.
[2] The Agency for Healthcare Research and Quality (AHRQ): **http://www.ahcpr.gov/consumer/diaginfo.htm**.
[3] From the NIH, National Cancer Institute (NCI): **http://cancertrials.nci.nih.gov/beyond/evaluating.html**.

Since the late 1990s, physicians have seen a general increase in patient Internet usage rates. Patients frequently enter their doctor's offices with printed Web pages of home remedies in the guise of latest medical research. This scenario is so common that doctors often spend more time dispelling misleading information than guiding patients through sound therapies. *The Official Patient's Sourcebook on Primary Sclerosing Cholangitis* has been created for patients who have decided to make education and research an integral part of the treatment process. The pages that follow will tell you where and how to look for information covering virtually all topics related to primary sclerosing cholangitis, from the essentials to the most advanced areas of research.

The title of this book includes the word "official." This reflects the fact that the sourcebook draws from public, academic, government, and peer-reviewed research. Selected readings from various agencies are reproduced to give you some of the latest official information available to date on primary sclerosing cholangitis.

Given patients' increasing sophistication in using the Internet, abundant references to reliable Internet-based resources are provided throughout this sourcebook. Where possible, guidance is provided on how to obtain free-of-charge, primary research results as well as more detailed information via the Internet. E-book and electronic versions of this sourcebook are fully interactive with each of the Internet sites mentioned (clicking on a hyperlink automatically opens your browser to the site indicated). Hard copy users of this sourcebook can type cited Web addresses directly into their browsers to obtain access to the corresponding sites. Since we are working with ICON Health Publications, hard copy *Sourcebooks* are frequently updated and printed on demand to ensure that the information provided is current.

In addition to extensive references accessible via the Internet, every chapter presents a "Vocabulary Builder." Many health guides offer glossaries of technical or uncommon terms in an appendix. In editing this sourcebook, we have decided to place a smaller glossary within each chapter that covers terms used in that chapter. Given the technical nature of some chapters, you may need to revisit many sections. Building one's vocabulary of medical terms in such a gradual manner has been shown to improve the learning process.

We must emphasize that no sourcebook on primary sclerosing cholangitis should affirm that a specific diagnostic procedure or treatment discussed in a research study, patent, or doctoral dissertation is "correct" or your best option. This sourcebook is no exception. Each patient is unique. Deciding on

appropriate options is always up to the patient in consultation with their physician and healthcare providers.

Organization

This sourcebook is organized into three parts. Part I explores basic techniques to researching primary sclerosing cholangitis (e.g. finding guidelines on diagnosis, treatments, and prognosis), followed by a number of topics, including information on how to get in touch with organizations, associations, or other patient networks dedicated to primary sclerosing cholangitis. It also gives you sources of information that can help you find a doctor in your local area specializing in treating primary sclerosing cholangitis. Collectively, the material presented in Part I is a complete primer on basic research topics for patients with primary sclerosing cholangitis.

Part II moves on to advanced research dedicated to primary sclerosing cholangitis. Part II is intended for those willing to invest many hours of hard work and study. It is here that we direct you to the latest scientific and applied research on primary sclerosing cholangitis. When possible, contact names, links via the Internet, and summaries are provided. It is in Part II where the vocabulary process becomes important as authors publishing advanced research frequently use highly specialized language. In general, every attempt is made to recommend "free-to-use" options.

Part III provides appendices of useful background reading for all patients with primary sclerosing cholangitis or related disorders. The appendices are dedicated to more pragmatic issues faced by many patients with primary sclerosing cholangitis. Accessing materials via medical libraries may be the only option for some readers, so a guide is provided for finding local medical libraries which are open to the public. Part III, therefore, focuses on advice that goes beyond the biological and scientific issues facing patients with primary sclerosing cholangitis.

Scope

While this sourcebook covers primary sclerosing cholangitis, your doctor, research publications, and specialists may refer to your condition using a variety of terms. Therefore, you should understand that primary sclerosing cholangitis is often considered a synonym or a condition closely related to the following:

- Chronic Obliterative Cholangitis
- Fibrosing Cholangitis
- Primary Sclerosing Cholangitis
- Stenosing Cholangitis

In addition to synonyms and related conditions, physicians may refer to primary sclerosing cholangitis using certain coding systems. The International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) is the most commonly used system of classification for the world's illnesses. Your physician may use this coding system as an administrative or tracking tool. The following classification is commonly used for primary sclerosing cholangitis:[4]

- 576.1 cholangitis

For the purposes of this sourcebook, we have attempted to be as inclusive as possible, looking for official information for all of the synonyms relevant to primary sclerosing cholangitis. You may find it useful to refer to synonyms when accessing databases or interacting with healthcare professionals and medical librarians.

Moving Forward

Since the 1980s, the world has seen a proliferation of healthcare guides covering most illnesses. Some are written by patients or their family members. These generally take a layperson's approach to understanding and coping with an illness or disorder. They can be uplifting, encouraging, and highly supportive. Other guides are authored by physicians or other healthcare providers who have a more clinical outlook. Each of these two styles of guide has its purpose and can be quite useful.

As editors, we have chosen a third route. We have chosen to expose you to as many sources of official and peer-reviewed information as practical, for the purpose of educating you about basic and advanced knowledge as recognized by medical science today. You can think of this sourcebook as your personal Internet age reference librarian.

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[4] This list is based on the official version of the World Health Organization's 9th Revision, International Classification of Diseases (ICD-9). According to the National Technical Information Service, "ICD-9CM extensions, interpretations, modifications, addenda, or errata other than those approved by the U.S. Public Health Service and the Health Care Financing Administration are not to be considered official and should not be utilized. Continuous maintenance of the ICD-9-CM is the responsibility of the federal government."

Why "Internet age"? All too often, patients diagnosed with primary sclerosing cholangitis will log on to the Internet, type words into a search engine, and receive several Web site listings which are mostly irrelevant or redundant. These patients are left to wonder where the relevant information is, and how to obtain it. Since only the smallest fraction of information dealing with primary sclerosing cholangitis is even indexed in search engines, a non-systematic approach often leads to frustration and disappointment. With this sourcebook, we hope to direct you to the information you need that you would not likely find using popular Web directories. Beyond Web listings, in many cases we will reproduce brief summaries or abstracts of available reference materials. These abstracts often contain distilled information on topics of discussion.

While we focus on the more scientific aspects of primary sclerosing cholangitis, there is, of course, the emotional side to consider. Later in the sourcebook, we provide a chapter dedicated to helping you find peer groups and associations that can provide additional support beyond research produced by medical science. We hope that the choices we have made give you the most options available in moving forward. In this way, we wish you the best in your efforts to incorporate this educational approach into your treatment plan.

The Editors

PART I: THE ESSENTIALS

ABOUT PART I

Part I has been edited to give you access to what we feel are "the essentials" on primary sclerosing cholangitis. The essentials of a disease typically include the definition or description of the disease, a discussion of who it affects, the signs or symptoms associated with the disease, tests or diagnostic procedures that might be specific to the disease, and treatments for the disease. Your doctor or healthcare provider may have already explained the essentials of primary sclerosing cholangitis to you or even given you a pamphlet or brochure describing primary sclerosing cholangitis. Now you are searching for more in-depth information. As editors, we have decided, nevertheless, to include a discussion on where to find essential information that can complement what your doctor has already told you. In this section we recommend a process, not a particular Web site or reference book. The process ensures that, as you search the Web, you gain background information in such a way as to maximize your understanding.

CHAPTER 1. THE ESSENTIALS ON PRIMARY SCLEROSING CHOLANGITIS: GUIDELINES

Overview

Official agencies, as well as federally-funded institutions supported by national grants, frequently publish a variety of guidelines on primary sclerosing cholangitis. These are typically called "Fact Sheets" or "Guidelines." They can take the form of a brochure, information kit, pamphlet, or flyer. Often they are only a few pages in length. The great advantage of guidelines over other sources is that they are often written with the patient in mind. Since new guidelines on primary sclerosing cholangitis can appear at any moment and be published by a number of sources, the best approach to finding guidelines is to systematically scan the Internet-based services that post them.

The National Institutes of Health (NIH)[5]

The National Institutes of Health (NIH) is the first place to search for relatively current patient guidelines and fact sheets on primary sclerosing cholangitis. Originally founded in 1887, the NIH is one of the world's foremost medical research centers and the federal focal point for medical research in the United States. At any given time, the NIH supports some 35,000 research grants at universities, medical schools, and other research and training institutions, both nationally and internationally. The rosters of those who have conducted research or who have received NIH support over the years include the world's most illustrious scientists and physicians.

[5] Adapted from the NIH: **http://www.nih.gov/about/NIHoverview.html**.

Among them are 97 scientists who have won the Nobel Prize for achievement in medicine.

There is no guarantee that any one Institute will have a guideline on a specific disease, though the National Institutes of Health collectively publish over 600 guidelines for both common and rare diseases. The best way to access NIH guidelines is via the Internet. Although the NIH is organized into many different Institutes and Offices, the following is a list of key Web sites where you are most likely to find NIH clinical guidelines and publications dealing with primary sclerosing cholangitis and associated conditions:

- Office of the Director (OD); guidelines consolidated across agencies available at **http://www.nih.gov/health/consumer/conkey.htm**
- National Library of Medicine (NLM); extensive encyclopedia (A.D.A.M., Inc.) with guidelines available at **http://www.nlm.nih.gov/medlineplus/healthtopics.html**
- National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK); guidelines available at **http://www.niddk.nih.gov/health/health.htm**

Among these, the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) is particularly noteworthy. The NIDDK's mission is to conduct and support research on many of the most serious diseases affecting public health.[6] The Institute supports much of the clinical research on the diseases of internal medicine and related subspecialty fields as well as many basic science disciplines. The NIDDK's Division of Intramural Research encompasses the broad spectrum of metabolic diseases such as diabetes, inborn errors of metabolism, endocrine disorders, mineral metabolism, digestive diseases, nutrition, urology and renal disease, and hematology. Basic research studies include biochemistry, nutrition, pathology, histochemistry, chemistry, physical, chemical, and molecular biology, pharmacology, and toxicology. NIDDK extramural research is organized into divisions of program areas:

- Division of Diabetes, Endocrinology, and Metabolic Diseases
- Division of Digestive Diseases and Nutrition
- Division of Kidney, Urologic, and Hematologic Diseases

[6] This paragraph has been adapted from the NIDDK: **http://www.niddk.nih.gov/welcome/mission.htm**. "Adapted" signifies that a passage is reproduced exactly or slightly edited for this book.

The Division of Extramural Activities provides administrative support and overall coordination. A fifth division, the Division of Nutrition Research Coordination, coordinates government nutrition research efforts. The Institute supports basic and clinical research through investigator-initiated grants, program project and center grants, and career development and training awards. The Institute also supports research and development projects and large-scale clinical trials through contracts. The following patient guideline was recently published by the NIDDK on primary sclerosing cholangitis.

What Is Primary Sclerosing Cholangitis?[7]

In primary sclerosing cholangitis (PSC), the bile ducts inside and outside the liver become inflamed and scarred. As the scarring increases, the ducts become blocked. The ducts are important because they carry bile out of the liver. Bile is a liquid that helps break down fat in food. If the ducts are blocked, bile builds up in the liver and damages liver cells. Eventually, PSC can cause liver failure.
Researchers do not know what causes PSC. Among the theories under investigation are the possible role of bacteria, viruses, and immune system problems. PSC appears to be associated with ulcerative colitis, a type of inflammatory bowel disease.

The disease usually begins between ages 30 and 60 and is more common in men than women. PSC progresses slowly, so a person can have the disease for years before symptoms develop. The main symptoms are itching, fatigue, and jaundice, which causes yellowing of the eyes or skin. An infection in the bile ducts can cause chills and fever.

PSC is diagnosed through cholangiography, which involves injecting dye into the bile ducts and taking an x-ray. Treatment includes medication to relieve itching, antibiotics to treat infections, and vitamin supplements, as people with PSC are often deficient in vitamins A, D, and K. In some cases, surgery to open major blockages in the common bile duct is also necessary. Liver transplantation may be an option if the liver begins to fail.

[7] Adapted from the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK): **http://www.niddk.nih.gov/health/digest/summary/psc/psc.htm**.

For More Information

More information is available from:

American Liver Foundation
75 Maiden Lane, Suite 603
New York, NY 10038
Phone: 1-800-GO-LIVER (465-4837)
Email: info@liverfoundation.org
Internet: **www.liverfoundation.org**

Additional Information on Primary Sclerosing Cholangitis

The National Digestive Diseases Information Clearinghouse collects resource information on digestive diseases for the Combined Health Information Database (CHID). CHID is a database produced by health-related agencies of the Federal Government. This database provides titles, abstracts, and availability information for health information and health education resources.

To provide you with the most up-to-date resources, information specialists at the clearinghouse created an automatic search of CHID. To obtain this information you may view the results of the automatic search on Primary Sclerosing Cholangitis.

Or, if you wish to perform your own search of the database, you may access the CHID Online web site and search CHID yourself.

National Digestive Diseases Information Clearinghouse
2 Information Way
Bethesda, MD 20892-3570
Email: nddic@info.niddk.nih.gov
The National Digestive Diseases Information Clearinghouse (NDDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). NIDDK is part of the National Institutes of Health under the U.S. Department of Health and Human Services. Established in 1980, the clearinghouse provides information about digestive diseases to people with digestive disorders and to their families, health care professionals, and the public. NDDIC answers inquiries; develops and distributes publications; and works closely with professional and patient organizations and Government agencies to coordinate resources about

digestive diseases. Publications produced by the clearinghouse are carefully reviewed by both NIDDK scientists and outside experts.

More Guideline Sources

The guideline above on primary sclerosing cholangitis is only one example of the kind of material that you can find online and free of charge. The remainder of this chapter will direct you to other sources which either publish or can help you find additional guidelines on topics related to primary sclerosing cholangitis. Many of the guidelines listed below address topics that may be of particular relevance to your specific situation or of special interest to only some patients with primary sclerosing cholangitis. Due to space limitations these sources are listed in a concise manner. Do not hesitate to consult the following sources by either using the Internet hyperlink provided, or, in cases where the contact information is provided, contacting the publisher or author directly.

Topic Pages: MEDLINEplus

For patients wishing to go beyond guidelines published by specific Institutes of the NIH, the National Library of Medicine has created a vast and patient-oriented healthcare information portal called MEDLINEplus. Within this Internet-based system are "health topic pages." You can think of a health topic page as a guide to patient guides. To access this system, log on to **http://www.nlm.nih.gov/medlineplus/healthtopics.html**. From there you can either search using the alphabetical index or browse by broad topic areas.

If you do not find topics of interest when browsing health topic pages, then you can choose to use the advanced search utility of MEDLINEplus at the following: **http://www.nlm.nih.gov/medlineplus/advancedsearch.html**. This utility is similar to the NIH Search Utility, with the exception that it only includes material linked within the MEDLINEplus system (mostly patient-oriented information). It also has the disadvantage of generating unstructured results. We recommend, therefore, that you use this method only if you have a very targeted search.

The Combined Health Information Database (CHID)

CHID Online is a reference tool that maintains a database directory of thousands of journal articles and patient education guidelines on primary sclerosing cholangitis and related conditions. One of the advantages of CHID over other sources is that it offers summaries that describe the guidelines available, including contact information and pricing. CHID's general Web site is **http://chid.nih.gov/**. To search this database, go to **http://chid.nih.gov/detail/detail.html**. In particular, you can use the advanced search options to look up pamphlets, reports, brochures, and information kits. The following was recently posted in this archive:

- **Cirrhosis of the Liver**

  Source: Bethesda, MD: American Gastroenterological Association. 199x. [4 p.].

  Contact: American Gastroenterological Association (AGA). 7910 Woodmont Avenue, Seventh Floor, Bethesda, MD 20814. (800) 668-5237 or (301) 654-2055. Fax (301) 652-3890. Website: www.gastro.org. PRICE: Single copy free; bulk copies available.

  Summary: When chronic diseases cause the liver to become permanently injured and scarred, the condition is called cirrhosis. This brochure, from the American Gastroenterological Association (AGA), reviews the problem of cirrhosis. Topics include the major causes of cirrhosis, the symptoms of the condition, diagnostic methods used to confirm cirrhosis, treatment strategies, and treatment options for the complications of cirrhosis. Cirrhosis can result from direct injury to the liver cells (i.e., hepatitis), or from indirect injury via inflammation or obstruction to bile ducts (e.g., primary biliary cirrhosis, primary sclerosing cholangitis), which drain the liver cells of bile. Chronic alcoholism is the most common cause of cirrhosis in the United States. People with cirrhosis often have few symptoms at first. The two major problems that eventually cause symptoms are loss of functioning liver cells and distortion of the liver caused by scarring. Associated problems include fluid accumulation (ascites), jaundice (yellow skin), gallstones, intense itching, loss of appetite, fatigue and weakness, buildup of toxins, slowed drug processing, portal hypertension (high blood pressure in the main veins of the liver), and varices (thin walled, enlarged blood vessels). Diagnosis is confirmed from the patient's symptoms and from diagnostic tests such as CT scan, ultrasound, and biopsy. Treatment of cirrhosis is aimed to stop the development of scar tissue in the liver and prevent complications. Regardless of the cause of cirrhosis, every patient must avoid all substances, habits, and drugs that may further damage the liver,

cause complications, or speed the progression to liver failure. Liver failure refers to the end stage of liver disease and cirrhosis when the liver stops working and cannot support life. The brochure includes a list of references and a diagram of the digestive tract, with organs labeled. 3 figures. 6 references.

The National Guideline Clearinghouse™

The National Guideline Clearinghouse™ offers hundreds of evidence-based clinical practice guidelines published in the United States and other countries. You can search their site located at **http://www.guideline.gov** by using the keyword "primary sclerosing cholangitis" or synonyms.

Healthfinder™

Healthfinder™ is an additional source sponsored by the U.S. Department of Health and Human Services which offers links to hundreds of other sites that contain healthcare information. This Web site is located at **http://www.healthfinder.gov**. Again, keyword searches can be used to find guidelines. The following was recently found in this database:

- **American Family Physician (Journal)**

  Summary: The American Family Physician is the official clinical journal of the American Academy of Family Physicians.

  Source: American Academy of Family Physicians

  http://www.healthfinder.gov/scripts/recordpass.asp?RecordType=0&RecordID=1333

- **American Medical News (AMNews)**

  Summary: Published weekly, this publication covers professional, social, economic and policy issues in medicine that are of interest to physicians and their practices.

  Source: American Medical Association

  http://www.healthfinder.gov/scripts/recordpass.asp?RecordType=0&RecordID=1936

The NIH Search Utility

After browsing the references listed at the beginning of this chapter, you may want to explore the NIH Search Utility. This allows you to search for documents on over 100 selected Web sites that comprise the NIH-WEB-SPACE. Each of these servers is "crawled" and indexed on an ongoing basis. Your search will produce a list of various documents, all of which will relate in some way to primary sclerosing cholangitis. The drawbacks of this approach are that the information is not organized by theme and that the references are often a mix of information for professionals and patients. Nevertheless, a large number of the listed Web sites provide useful background information. We can only recommend this route, therefore, for relatively rare or specific disorders, or when using highly targeted searches. To use the NIH search utility, visit the following Web page: **http://search.nih.gov/index.html**.

Additional Web Sources

A number of Web sites that often link to government sites are available to the public. These can also point you in the direction of essential information. The following is a representative sample:

- AOL: **http://search.aol.com/cat.adp?id=168&layer=&from=subcats**
- drkoop.com®: **http://www.drkoop.com/conditions/ency/index.html**
- Family Village: **http://www.familyvillage.wisc.edu/specific.htm**
- Google: **http://directory.google.com/Top/Health/Conditions_and_Diseases/**
- Med Help International: **http://www.medhelp.org/HealthTopics/A.html**
- Open Directory Project: **http://dmoz.org/Health/Conditions_and_Diseases/**
- Yahoo.com: **http://dir.yahoo.com/Health/Diseases_and_Conditions/**
- WebMD®Health: **http://my.webmd.com/health_topics**

Vocabulary Builder

The material in this chapter may have contained a number of unfamiliar words. The following Vocabulary Builder introduces you to terms used in this chapter that have not been covered in the previous chapter:

Alimentary: Pertaining to food or nutritive material, or to the organs of digestion. [EU]

American Medical Association: Professional society representing the field of medicine. [NIH]

Antibiotic: A chemical substance produced by a microorganism which has the capacity, in dilute solutions, to inhibit the growth of or to kill other microorganisms. Antibiotics that are sufficiently nontoxic to the host are used as chemotherapeutic agents in the treatment of infectious diseases of man, animals and plants. [EU]

Ascites: Effusion and accumulation of serous fluid in the abdominal cavity; called also abdominal or peritoneal dropsy, hydroperitonia, and hydrops abdominis. [EU]

Bacteria: Unicellular prokaryotic microorganisms which generally possess rigid cell walls, multiply by cell division, and exhibit three principal forms: round or coccal, rodlike or bacillary, and spiral or spirochetal. [NIH]

Bile: An emulsifying agent produced in the liver and secreted into the duodenum. Its composition includes bile acids and salts, cholesterol, and electrolytes. It aids digestion of fats in the duodenum. [NIH]

Biliary: Pertaining to the bile, to the bile ducts, or to the gallbladder. [EU]

Biopsy: The removal and examination, usually microscopic, of tissue from the living body, performed to establish precise diagnosis. [EU]

Cholangiography: Roentgenography of the biliary ducts after administration or injection of a contrast medium, orally, intravenously or percutaneously. [EU]

Cholangitis: Inflammation of a bile duct. [EU]

Chronic: Persisting over a long period of time. [EU]

Cirrhosis: Liver disease characterized pathologically by loss of the normal microscopic lobular architecture, with fibrosis and nodular regeneration. The term is sometimes used to refer to chronic interstitial inflammation of any organ. [EU]

Colitis: Inflammation of the colon. [EU]

Endocrinology: A subspecialty of internal medicine concerned with the metabolism, physiology, and disorders of the endocrine system. [NIH]

Fatigue: The state of weariness following a period of exertion, mental or physical, characterized by a decreased capacity for work and reduced efficiency to respond to stimuli. [NIH]

Hematology: A subspecialty of internal medicine concerned with morphology, physiology, and pathology of the blood and blood-forming tissues. [NIH]

Hepatitis: Inflammation of the liver. [EU]

Hypertension: Persistently high arterial blood pressure. Various criteria for its threshold have been suggested, ranging from 140 mm. Hg systolic and 90 mm. Hg diastolic to as high as 200 mm. Hg systolic and 110 mm. Hg diastolic. Hypertension may have no known cause (essential or idiopathic h.) or be associated with other primary diseases (secondary h.). [EU]

Inflammation: A pathological process characterized by injury or destruction of tissues caused by a variety of cytologic and chemical reactions. It is usually manifested by typical signs of pain, heat, redness, swelling, and loss of function. [NIH]

Invasive: 1. having the quality of invasiveness. 2. involving puncture or incision of the skin or insertion of an instrument or foreign material into the body; said of diagnostic techniques. [EU]

Jaundice: A clinical manifestation of hyperbilirubinemia, consisting of deposition of bile pigments in the skin, resulting in a yellowish staining of the skin and mucous membranes. [NIH]

Molecular: Of, pertaining to, or composed of molecules : a very small mass of matter. [EU]

Proteins: Polymers of amino acids linked by peptide bonds. The specific sequence of amino acids determines the shape and function of the protein. [NIH]

Spectrum: A charted band of wavelengths of electromagnetic vibrations obtained by refraction and diffraction. By extension, a measurable range of activity, such as the range of bacteria affected by an antibiotic (antibacterial s.) or the complete range of manifestations of a disease. [EU]

Toxicology: The science concerned with the detection, chemical composition, and pharmacologic action of toxic substances or poisons and the treatment and prevention of toxic manifestations. [NIH]

Toxin: A poison; frequently used to refer specifically to a protein produced by some higher plants, certain animals, and pathogenic bacteria, which is highly toxic for other living organisms. Such substances are differentiated from the simple chemical poisons and the vegetable alkaloids by their high molecular weight and antigenicity. [EU]

Transplantation: The grafting of tissues taken from the patient's own body or from another. [EU]

Urology: A surgical specialty concerned with the study, diagnosis, and treatment of diseases of the urinary tract in both sexes and the genital tract in the male. It includes the specialty of andrology which addresses both male genital diseases and male infertility. [NIH]

Veins: The vessels carrying blood toward the heart. [NIH]

Viruses: Minute infectious agents whose genomes are composed of DNA or RNA, but not both. They are characterized by a lack of independent metabolism and the inability to replicate outside living host cells. [NIH]

CHAPTER 2. SEEKING GUIDANCE

Overview

Some patients are comforted by the knowledge that a number of organizations dedicate their resources to helping people with primary sclerosing cholangitis. These associations can become invaluable sources of information and advice. Many associations offer aftercare support, financial assistance, and other important services. Furthermore, healthcare research has shown that support groups often help people to better cope with their conditions.[8] In addition to support groups, your physician can be a valuable source of guidance and support. Therefore, finding a physician that can work with your unique situation is a very important aspect of your care.

In this chapter, we direct you to resources that can help you find patient organizations and medical specialists. We begin by describing how to find associations and peer groups that can help you better understand and cope with primary sclerosing cholangitis. The chapter ends with a discussion on how to find a doctor that is right for you.

Associations and Primary Sclerosing Cholangitis

As mentioned by the Agency for Healthcare Research and Quality, sometimes the emotional side of an illness can be as taxing as the physical side.[9] You may have fears or feel overwhelmed by your situation. Everyone has different ways of dealing with disease or physical injury. Your attitude, your expectations, and how well you cope with your condition can all

[8] Churches, synagogues, and other houses of worship might also have groups that can offer you the social support you need.
[9] This section has been adapted from **http://www.ahcpr.gov/consumer/diaginf5.htm**.

influence your well-being. This is true for both minor conditions and serious illnesses. For example, a study on female breast cancer survivors revealed that women who participated in support groups lived longer and experienced better quality of life when compared with women who did not participate. In the support group, women learned coping skills and had the opportunity to share their feelings with other women in the same situation.

In addition to associations or groups that your doctor might recommend, we suggest that you consider the following list (if there is a fee for an association, you may want to check with your insurance provider to find out if the cost will be covered):

- **American Liver Foundation**

  Address: American Liver Foundation 75 Maiden Lane, Suite 603, New York, NY 10038

  Telephone: (212) 668-1000 Toll-free: (800) 465-4837

  Fax: (973) 256-3214

  Email: webmail@liverfoundation.org

  Web Site: http://www.liverfoundation.or

  Background: The American Liver Foundation is a national voluntary not-for-profit organization dedicated to the prevention, treatment, and cure of diseases of the liver through programs of research and education. Established in 1976, the Foundation's activities include support groups, patient advocacy, support of medical research, and patient and professional education. Educational materials include brochures on Hepatitis, Cirrhosis, Biliary Atresia, liver transplantation, gallstones, and Hereditary Hemochromatosis. Fact sheets are also available on a variety of liver diseases including Alagille Syndrome, Alpha-1-Antitrypsin Deficiency, Cancer of the Liver, Fatty Liver, Gilbert Syndrome, Primary Biliary Cirrhosis, Porphyria, and others. Videotapes produced by the Foundation include 'A Healthy Liver: A Happier Life,' 'Foundations for Decision Making,' 'Hepatitis B: Patient Information,' 'Hepatitis C: A Guide for Primary Care Physicians,' and 'The Visionaries.' The Foundation also offers liver wellness and substance abuse prevention programs to elementary schools and corporations.

  Relevant area(s) of interest: Gallstones, Hepatitis C, Porphyria, Wilson's Disease

- **Canadian Liver Foundation**

  Address: Canadian Liver Foundation 365 Bloor Street, Suite 200, Toronto, Ontario, M4W 3L4, Canada

Telephone: (416) 964-4935 Toll-free: (800) 563-5483

Fax: (416) 964-0024

Email: clf@liver.ca

Web Site: http://www.liver.c

Background: The Canadian Liver Foundation (CLF) is a not-for-profit health organization committed to reducing the incidence and impact of liver disease by providing support for research and education into the causes, diagnosis, prevention and treatment of more than 100 diseases of the liver. Established in 1969, the CLF has established 30 chapters across Canada and provides information in both English and French. Some of the liver diseases discussed in brochures and medical information sheets available from CLF include gallstones, hemochromatosis, primary biliary cirrhosis, several forms of hepatitis, porphyria, fatty liver, and liver cancer. Further information is provided on liver transplantation, the effects of sodium, and management of variceal bleeding. The Foundation also produces a newsletter and maintains World Wide Web site at http://www.liver.ca.

Relevant area(s) of interest: Cirrhosis of the Liver, Gallstones, Hepatitis A, Hepatitis B, Hepatitis C, Porphyria

- **United Liver Association**

  Address: United Liver Association 11646 West Pico Boulevard, Los Angeles, CA 90064-2987

  Telephone: (310) 445-4204

  Fax: (310) 575-387

  Background: The United Liver Association is a not-for-profit organization dedicated to providing support services to individuals with liver disease, family members, and caregivers. The Association seeks to increase awareness and understanding of liver diseases in the medical and lay communities and promote ongoing medical research into the causes, management, and potential cure of these disorders. Established in 1985, the United Liver Association has a telephone hot line under the direction of liver specialists and other trained professionals; and holds group support sessions for affected individuals and families. It offers psychologist-led sessions and conducts educational seminars for patients, families, and health care professionals. The Association also gives appropriate referrals; provides limited direct assistance for out-of-area liver transplant patients and families, based on financial need; engages in patient, family, and community education; and promotes patient and

family advocacy. The United Liver Association also provides a variety of educational materials, including a regular newsletter and a brochure.

Relevant area(s) of interest: Wilson's Disease

Finding More Associations

There are a number of directories that list additional medical associations that you may find useful. While not all of these directories will provide different information than what is listed above, by consulting all of them, you will have nearly exhausted all sources for patient associations.

The National Health Information Center (NHIC)

The National Health Information Center (NHIC) offers a free referral service to help people find organizations that provide information about primary sclerosing cholangitis. For more information, see the NHIC's Web site at **http://www.health.gov/NHIC/** or contact an information specialist by calling 1-800-336-4797.

DIRLINE

A comprehensive source of information on associations is the DIRLINE database maintained by the National Library of Medicine. The database comprises some 10,000 records of organizations, research centers, and government institutes and associations which primarily focus on health and biomedicine. DIRLINE is available via the Internet at the following Web site: **http://dirline.nlm.nih.gov/**. Simply type in "primary sclerosing cholangitis" (or a synonym) or the name of a topic, and the site will list information contained in the database on all relevant organizations.

The Combined Health Information Database

Another comprehensive source of information on healthcare associations is the Combined Health Information Database. Using the "Detailed Search" option, you will need to limit your search to "Organizations" and "primary sclerosing cholangitis". Type the following hyperlink into your Web browser: **http://chid.nih.gov/detail/detail.html**. To find associations, use the

drop boxes at the bottom of the search page where "You may refine your search by." For publication date, select "All Years." Then, select your preferred language and the format option "Organization Resource Sheet." By making these selections and typing in "primary sclerosing cholangitis" (or synonyms) into the "For these words:" box, you will only receive results on organizations dealing with primary sclerosing cholangitis. You should check back periodically with this database since it is updated every 3 months.

The National Organization for Rare Disorders, Inc.

The National Organization for Rare Disorders, Inc. has prepared a Web site that provides, at no charge, lists of associations organized by specific diseases. You can access this database at the following Web site: **http://www.rarediseases.org/cgi-bin/nord/searchpage**. Select the option called "Organizational Database (ODB)" and type "primary sclerosing cholangitis" (or a synonym) in the search box.

Online Support Groups

In addition to support groups, commercial Internet service providers offer forums and chat rooms for people with different illnesses and conditions. WebMD®, for example, offers such a service at their Web site: **http://boards.webmd.com/roundtable**. These online self-help communities can help you connect with a network of people whose concerns are similar to yours. Online support groups are places where people can talk informally. If you read about a novel approach, consult with your doctor or other healthcare providers, as the treatments or discoveries you hear about may not be scientifically proven to be safe and effective.

- **PSC Support Group**
  http://www.psc-support.demon.co.uk/
- **Yahoo! Groups**
  http://groups.yahoo.com/group/psc-support/

Finding Doctors

One of the most important aspects of your treatment will be the relationship between you and your doctor or specialist. All patients with primary sclerosing cholangitis must go through the process of selecting a physician.

While this process will vary from person to person, the Agency for Healthcare Research and Quality makes a number of suggestions, including the following:[10]

- If you are in a managed care plan, check the plan's list of doctors first.
- Ask doctors or other health professionals who work with doctors, such as hospital nurses, for referrals.
- Call a hospital's doctor referral service, but keep in mind that these services usually refer you to doctors on staff at that particular hospital. The services do not have information on the quality of care that these doctors provide.
- Some local medical societies offer lists of member doctors. Again, these lists do not have information on the quality of care that these doctors provide.

Additional steps you can take to locate doctors include the following:

- Check with the associations listed earlier in this chapter.
- Information on doctors in some states is available on the Internet at **http://www.docboard.org**. This Web site is run by "Administrators in Medicine," a group of state medical board directors.
- The American Board of Medical Specialties can tell you if your doctor is board certified. "Certified" means that the doctor has completed a training program in a specialty and has passed an exam, or "board," to assess his or her knowledge, skills, and experience to provide quality patient care in that specialty. Primary care doctors may also be certified as specialists. The AMBS Web site is located at **http://www.abms.org/newsearch.asp**.[11] You can also contact the ABMS by phone at 1-866-ASK-ABMS.
- You can call the American Medical Association (AMA) at 800-665-2882 for information on training, specialties, and board certification for many licensed doctors in the United States. This information also can be found in "Physician Select" at the AMA's Web site: **http://www.ama-assn.org/aps/amahg.htm**.

If the previous sources did not meet your needs, you may want to log on to the Web site of the National Organization for Rare Disorders (NORD) at **http://www.rarediseases.org/**. NORD maintains a database of doctors with

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[10] This section is adapted from the AHRQ: **www.ahrq.gov/consumer/qntascii/qntdr.htm**.

[11] While board certification is a good measure of a doctor's knowledge, it is possible to receive quality care from doctors who are not board certified.

expertise in various rare diseases. The Metabolic Information Network (MIN), 800-945-2188, also maintains a database of physicians with expertise in various metabolic diseases.

Selecting Your Doctor[12]

When you have compiled a list of prospective doctors, call each of their offices. First, ask if the doctor accepts your health insurance plan and if he or she is taking new patients. If the doctor is not covered by your plan, ask yourself if you are prepared to pay the extra costs. The next step is to schedule a visit with your chosen physician. During the first visit you will have the opportunity to evaluate your doctor and to find out if you feel comfortable with him or her. Ask yourself, did the doctor:

- Give me a chance to ask questions about primary sclerosing cholangitis?
- Really listen to my questions?
- Answer in terms I understood?
- Show respect for me?
- Ask me questions?
- Make me feel comfortable?
- Address the health problem(s) I came with?
- Ask me my preferences about different kinds of treatments for primary sclerosing cholangitis?
- Spend enough time with me?

Trust your instincts when deciding if the doctor is right for you. But remember, it might take time for the relationship to develop. It takes more than one visit for you and your doctor to get to know each other.

[12] This section has been adapted from the AHRQ: **www.ahrq.gov/consumer/qntascii/qntdr.htm**.

Working with Your Doctor[13]

Research has shown that patients who have good relationships with their doctors tend to be more satisfied with their care and have better results. Here are some tips to help you and your doctor become partners:

- You know important things about your symptoms and your health history. Tell your doctor what you think he or she needs to know.
- It is important to tell your doctor personal information, even if it makes you feel embarrassed or uncomfortable.
- Bring a "health history" list with you (and keep it up to date).
- Always bring any medications you are currently taking with you to the appointment, or you can bring a list of your medications including dosage and frequency information. Talk about any allergies or reactions you have had to your medications.
- Tell your doctor about any natural or alternative medicines you are taking.
- Bring other medical information, such as x-ray films, test results, and medical records.
- Ask questions. If you don't, your doctor will assume that you understood everything that was said.
- Write down your questions before your visit. List the most important ones first to make sure that they are addressed.
- Consider bringing a friend with you to the appointment to help you ask questions. This person can also help you understand and/or remember the answers.
- Ask your doctor to draw pictures if you think that this would help you understand.
- Take notes. Some doctors do not mind if you bring a tape recorder to help you remember things, but always ask first.
- Let your doctor know if you need more time. If there is not time that day, perhaps you can speak to a nurse or physician assistant on staff or schedule a telephone appointment.
- Take information home. Ask for written instructions. Your doctor may also have brochures and audio and videotapes that can help you.

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[13] This section has been adapted from the AHRQ: **www.ahrq.gov/consumer/qntascii/qntdr.htm**.

- After leaving the doctor's office, take responsibility for your care. If you have questions, call. If your symptoms get worse or if you have problems with your medication, call. If you had tests and do not hear from your doctor, call for your test results. If your doctor recommended that you have certain tests, schedule an appointment to get them done. If your doctor said you should see an additional specialist, make an appointment.

By following these steps, you will enhance the relationship you will have with your physician.

Broader Health-Related Resources

In addition to the references above, the NIH has set up guidance Web sites that can help patients find healthcare professionals. These include:[14]

- Caregivers:
  http://www.nlm.nih.gov/medlineplus/caregivers.html
- Choosing a Doctor or Healthcare Service:
  http://www.nlm.nih.gov/medlineplus/choosingadoctororhealthcareservice.html
- Hospitals and Health Facilities:
  http://www.nlm.nih.gov/medlineplus/healthfacilities.html

Vocabulary Builder

The following vocabulary builder provides definitions of words used in this chapter that have not been defined in previous chapters:

Porphyria: A pathological state in man and some lower animals that is often due to genetic factors, is characterized by abnormalities of porphyrin metabolism, and results in the excretion of large quantities of porphyrins in the urine and in extreme sensitivity to light. [EU]

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[14] You can access this information at:
http://www.nlm.nih.gov/medlineplus/healthsystem.html.

CHAPTER 3. CLINICAL TRIALS AND PRIMARY SCLEROSING CHOLANGITIS

Overview

Very few medical conditions have a single treatment. The basic treatment guidelines that your physician has discussed with you, or those that you have found using the techniques discussed in Chapter 1, may provide you with all that you will require. For some patients, current treatments can be enhanced with new or innovative techniques currently under investigation. In this chapter, we will describe how clinical trials work and show you how to keep informed of trials concerning primary sclerosing cholangitis.

What Is a Clinical Trial?[15]

Clinical trials involve the participation of people in medical research. Most medical research begins with studies in test tubes and on animals. Treatments that show promise in these early studies may then be tried with people. The only sure way to find out whether a new treatment is safe, effective, and better than other treatments for primary sclerosing cholangitis is to try it on patients in a clinical trial.

[15] The discussion in this chapter has been adapted from the NIH and the NEI: **www.nei.nih.gov/netrials/ctivr.htm**.

What Kinds of Clinical Trials Are There?

Clinical trials are carried out in three phases:

- **Phase I.** Researchers first conduct Phase I trials with small numbers of patients and healthy volunteers. If the new treatment is a medication, researchers also try to determine how much of it can be given safely.
- **Phase II.** Researchers conduct Phase II trials in small numbers of patients to find out the effect of a new treatment on primary sclerosing cholangitis.
- **Phase III.** Finally, researchers conduct Phase III trials to find out how new treatments for primary sclerosing cholangitis compare with standard treatments already being used. Phase III trials also help to determine if new treatments have any side effects. These trials--which may involve hundreds, perhaps thousands, of people--can also compare new treatments with no treatment.

How Is a Clinical Trial Conducted?

Various organizations support clinical trials at medical centers, hospitals, universities, and doctors' offices across the United States. The "principal investigator" is the researcher in charge of the study at each facility participating in the clinical trial. Most clinical trial researchers are medical doctors, academic researchers, and specialists. The "clinic coordinator" knows all about how the study works and makes all the arrangements for your visits.

All doctors and researchers who take part in the study on primary sclerosing cholangitis carefully follow a detailed treatment plan called a protocol. This plan fully explains how the doctors will treat you in the study. The "protocol" ensures that all patients are treated in the same way, no matter where they receive care.

Clinical trials are controlled. This means that researchers compare the effects of the new treatment with those of the standard treatment. In some cases, when no standard treatment exists, the new treatment is compared with no treatment. Patients who receive the new treatment are in the treatment group. Patients who receive a standard treatment or no treatment are in the "control" group. In some clinical trials, patients in the treatment group get a new medication while those in the control group get a placebo. A placebo is a harmless substance, a "dummy" pill, that has no effect on primary sclerosing cholangitis. In other clinical trials, where a new surgery or device

(not a medicine) is being tested, patients in the control group may receive a "sham treatment." This treatment, like a placebo, has no effect on primary sclerosing cholangitis and does not harm patients.

Researchers assign patients "randomly" to the treatment or control group. This is like flipping a coin to decide which patients are in each group. If you choose to participate in a clinical trial, you will not know which group you will be appointed to. The chance of any patient getting the new treatment is about 50 percent. You cannot request to receive the new treatment instead of the placebo or sham treatment. Often, you will not know until the study is over whether you have been in the treatment group or the control group. This is called a "masked" study. In some trials, neither doctors nor patients know who is getting which treatment. This is called a "double masked" study. These types of trials help to ensure that the perceptions of the patients or doctors will not affect the study results.

Natural History Studies

Unlike clinical trials in which patient volunteers may receive new treatments, natural history studies provide important information to researchers on how primary sclerosing cholangitis develops over time. A natural history study follows patient volunteers to see how factors such as age, sex, race, or family history might make some people more or less at risk for primary sclerosing cholangitis. A natural history study may also tell researchers if diet, lifestyle, or occupation affects how a disease or disorder develops and progresses. Results from these studies provide information that helps answer questions such as: How fast will a disease or disorder usually progress? How bad will the condition become? Will treatment be needed?

What Is Expected of Patients in a Clinical Trial?

Not everyone can take part in a clinical trial for a specific disease or disorder. Each study enrolls patients with certain features or eligibility criteria. These criteria may include the type and stage of disease or disorder, as well as, the age and previous treatment history of the patient. You or your doctor can contact the sponsoring organization to find out more about specific clinical trials and their eligibility criteria. If you are interested in joining a clinical trial, your doctor must contact one of the trial's investigators and provide details about your diagnosis and medical history.

If you participate in a clinical trial, you may be required to have a number of medical tests. You may also need to take medications and/or undergo surgery. Depending upon the treatment and the examination procedure, you may be required to receive inpatient hospital care. Or, you may have to return to the medical facility for follow-up examinations. These exams help find out how well the treatment is working. Follow-up studies can take months or years. However, the success of the clinical trial often depends on learning what happens to patients over a long period of time. Only patients who continue to return for follow-up examinations can provide this important long-term information.

Recent Trials on Primary Sclerosing Cholangitis

The National Institutes of Health and other organizations sponsor trials on various diseases and disorders. Because funding for research goes to the medical areas that show promising research opportunities, it is not possible for the NIH or others to sponsor clinical trials for every disease and disorder at all times. The following lists recent trials dedicated to primary sclerosing cholangitis.[16] If the trial listed by the NIH is still recruiting, you may be eligible. If it is no longer recruiting or has been completed, then you can contact the sponsors to learn more about the study and, if published, the results. Further information on the trial is available at the Web site indicated. Please note that some trials may no longer be recruiting patients or are otherwise closed. Before contacting sponsors of a clinical trial, consult with your physician who can help you determine if you might benefit from participation.

- **Pilot Study of Budesonide for Patients with Primary Sclerosing Cholangitis**

  Condition(s): Cholangitis, Sclerosing; Liver Cirrhosis, Biliary

  Study Status: This study is no longer recruiting patients.

  Sponsor(s): National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK); Mayo Clinic

  Purpose - Excerpt: Objectives: I. Assess the safety and effectiveness of budesonide in patients with primary sclerosing cholangitis or primary biliary cirrhosis experiencing a suboptimal response to ursodeoxycholic acid. II. Estimate the efficacy of this therapy in these patient groups as a means of evaluating the feasibility of a long-term randomized trial.

  Phase(s): Phase I

[16] These are listed at **www.ClinicalTrials.gov**.

Study Type: Interventional

Contact(s): Keith D. Lindor 507-284-4823. Study chairs or principal investigators: Keith D. Lindor, Study Chair; Mayo Clinic

Web Site:
http://clinicaltrials.gov/ct/gui/show/NCT00004842;jsessionid=50F01501E27471F199033318F3B564D2

- **Phase II Pilot Study of Cladribine (2-Chlorodeoxyadenosine; 2-CdA) for Early Stage Primary Sclerosing Cholangitis**

  Condition(s): Cholangitis, Sclerosing

  Study Status: This study is completed.

  Sponsor(s): National Center for Research Resources (NCRR); Scripps Clinic

  Purpose - Excerpt: Objectives: I. Evaluate the effects of cladribine (2-chlorodeoxyadenosine; 2-CdA) on biochemical, radiologic, and histologic parameters in patients with early stage primary sclerosing cholangitis.

  Phase(s): Phase II

  Study Type: Interventional

  Contact(s):. Study chairs or principal investigators: Paul J. Pockros, Study Chair; Scripps Clinic

  Web Site:
  http://clinicaltrials.gov/ct/gui/show/NCT00004762;jsessionid=50F01501E27471F199033318F3B564D2

Benefits and Risks[17]

What Are the Benefits of Participating in a Clinical Trial?

If you are interested in a clinical trial, it is important to realize that your participation can bring many benefits to you and society at large:

- A new treatment could be more effective than the current treatment for primary sclerosing cholangitis. Although only half of the participants in a clinical trial receive the experimental treatment, if the new treatment is proved to be more effective and safer than the current treatment, then

[17] This section has been adapted from ClinicalTrials.gov, a service of the National Institutes of Health:
http://www.clinicaltrials.gov/ct/gui/c/a1r/info/whatis?JServSessionIdzone_ct=9jmun6f291.

those patients who did not receive the new treatment during the clinical trial may be among the first to benefit from it when the study is over.

- If the treatment is effective, then it may improve health or prevent diseases or disorders.
- Clinical trial patients receive the highest quality of medical care. Experts watch them closely during the study and may continue to follow them after the study is over.
- People who take part in trials contribute to scientific discoveries that may help other people with primary sclerosing cholangitis. In cases where certain diseases or disorders run in families, your participation may lead to better care or prevention for your family members.

The Informed Consent

Once you agree to take part in a clinical trial, you will be asked to sign an "informed consent." This document explains a clinical trial's risks and benefits, the researcher's expectations of you, and your rights as a patient.

What Are the Risks?

Clinical trials may involve risks as well as benefits. Whether or not a new treatment will work cannot be known ahead of time. There is always a chance that a new treatment may not work better than a standard treatment. There is also the possibility that it may be harmful. The treatment you receive may cause side effects that are serious enough to require medical attention.

How Is Patient Safety Protected?

Clinical trials can raise fears of the unknown. Understanding the safeguards that protect patients can ease some of these fears. Before a clinical trial begins, researchers must get approval from their hospital's Institutional Review Board (IRB), an advisory group that makes sure a clinical trial is designed to protect patient safety. During a clinical trial, doctors will closely watch you to see if the treatment is working and if you are experiencing any side effects. All the results are carefully recorded and reviewed. In many cases, experts from the Data and Safety Monitoring Committee carefully monitor each clinical trial and can recommend that a study be stopped at any

time. You will only be asked to take part in a clinical trial as a volunteer giving informed consent.

What Are a Patient's Rights in a Clinical Trial?

If you are eligible for a clinical trial, you will be given information to help you decide whether or not you want to participate. As a patient, you have the right to:

- Information on all known risks and benefits of the treatments in the study.
- Know how the researchers plan to carry out the study, for how long, and where.
- Know what is expected of you.
- Know any costs involved for you or your insurance provider.
- Know before any of your medical or personal information is shared with other researchers involved in the clinical trial.
- Talk openly with doctors and ask any questions.

After you join a clinical trial, you have the right to:

- Leave the study at any time. Participation is strictly voluntary. However, you should not enroll if you do not plan to complete the study.
- Receive any new information about the new treatment.
- Continue to ask questions and get answers.
- Maintain your privacy. Your name will not appear in any reports based on the study.
- Know whether you participated in the treatment group or the control group (once the study has been completed).

What about Costs?

In some clinical trials, the research facility pays for treatment costs and other associated expenses. You or your insurance provider may have to pay for costs that are considered standard care. These things may include inpatient hospital care, laboratory and other tests, and medical procedures. You also may need to pay for travel between your home and the clinic. You should find out about costs before committing to participation in the trial. If you

have health insurance, find out exactly what it will cover. If you don't have health insurance, or if your insurance company will not cover your costs, talk to the clinic staff about other options for covering the cost of your care.

What Questions Should You Ask before Deciding to Join a Clinical Trial?

Questions you should ask when thinking about joining a clinical trial include the following:

- What is the purpose of the clinical trial?
- What are the standard treatments for primary sclerosing cholangitis? Why do researchers think the new treatment may be better? What is likely to happen to me with or without the new treatment?
- What tests and treatments will I need? Will I need surgery? Medication? Hospitalization?
- How long will the treatment last? How often will I have to come back for follow-up exams?
- What are the treatment's possible benefits to my condition? What are the short- and long-term risks? What are the possible side effects?
- Will the treatment be uncomfortable? Will it make me feel sick? If so, for how long?
- How will my health be monitored?
- Where will I need to go for the clinical trial? How will I get there?
- How much will it cost to be in the study? What costs are covered by the study? How much will my health insurance cover?
- Will I be able to see my own doctor? Who will be in charge of my care?
- Will taking part in the study affect my daily life? Do I have time to participate?
- How do I feel about taking part in a clinical trial? Are there family members or friends who may benefit from my contributions to new medical knowledge?

Keeping Current on Clinical Trials

Various government agencies maintain databases on trials. The U.S. National Institutes of Health, through the National Library of Medicine, has

developed ClinicalTrials.gov to provide patients, family members, and physicians with current information about clinical research across the broadest number of diseases and conditions.

The site was launched in February 2000 and currently contains approximately 5,700 clinical studies in over 59,000 locations worldwide, with most studies being conducted in the United States. ClinicalTrials.gov receives about 2 million hits per month and hosts approximately 5,400 visitors daily. To access this database, simply go to their Web site (**www.clinicaltrials.gov**) and search by "primary sclerosing cholangitis" (or synonyms).

While ClinicalTrials.gov is the most comprehensive listing of NIH-supported clinical trials available, not all trials are in the database. The database is updated regularly, so clinical trials are continually being added. The following is a list of specialty databases affiliated with the National Institutes of Health that offer additional information on trials:

- For clinical studies at the Warren Grant Magnuson Clinical Center located in Bethesda, Maryland, visit their Web site: **http://clinicalstudies.info.nih.gov/**
- For clinical studies conducted at the Bayview Campus in Baltimore, Maryland, visit their Web site: **http://www.jhbmc.jhu.edu/studies/index.html**
- For trials on diseases of the digestive system and kidneys, and diabetes, visit the National Institute of Diabetes and Digestive and Kidney Diseases: **http://www.niddk.nih.gov/patient/patient.htm**

General References

The following references describe clinical trials and experimental medical research. They have been selected to ensure that they are likely to be available from your local or online bookseller or university medical library. These references are usually written for healthcare professionals, so you may consider consulting with a librarian or bookseller who might recommend a particular reference. The following includes some of the most readily available references (sorted alphabetically by title; hyperlinks provide rankings, information and reviews at Amazon.com):

- **A Guide to Patient Recruitment : Today's Best Practices & Proven Strategies** by Diana L. Anderson; Paperback - 350 pages (2001),

CenterWatch, Inc.; ISBN: 1930624115;
http://www.amazon.com/exec/obidos/ASIN/1930624115/icongroupinterna

- **A Step-By-Step Guide to Clinical Trials** by Marilyn Mulay, R.N., M.S., OCN; Spiral-bound - 143 pages Spiral edition (2001), Jones & Bartlett Pub; ISBN: 0763715697;
http://www.amazon.com/exec/obidos/ASIN/0763715697/icongroupinterna
- **The CenterWatch Directory of Drugs in Clinical Trials** by CenterWatch; Paperback - 656 pages (2000), CenterWatch, Inc.; ISBN: 0967302935;
http://www.amazon.com/exec/obidos/ASIN/0967302935/icongroupinterna
- **The Complete Guide to Informed Consent in Clinical Trials** by Terry Hartnett (Editor); Paperback - 164 pages (2000), PharmSource Information Services, Inc.; ISBN: 0970153309;
http://www.amazon.com/exec/obidos/ASIN/0970153309/icongroupinterna
- **Dictionary for Clinical Trials** by Simon Day; Paperback - 228 pages (1999), John Wiley & Sons; ISBN: 0471985961;
http://www.amazon.com/exec/obidos/ASIN/0471985961/icongroupinterna
- **Extending Medicare Reimbursement in Clinical Trials** by Institute of Medicine Staff (Editor), et al; Paperback 1st edition (2000), National Academy Press; ISBN: 0309068886;
http://www.amazon.com/exec/obidos/ASIN/0309068886/icongroupinterna
- **Handbook of Clinical Trials** by Marcus Flather (Editor); Paperback (2001), Remedica Pub Ltd; ISBN: 1901346293;
http://www.amazon.com/exec/obidos/ASIN/1901346293/icongroupinterna

Vocabulary Builder

The following vocabulary builder gives definitions of words used in this chapter that have not been defined in previous chapters:

Biochemical: Relating to biochemistry; characterized by, produced by, or involving chemical reactions in living organisms. [EU]

Budesonide: A glucocorticoid used in the management of asthma, the treatment of various skin disorders, and allergic rhinitis. [NIH]

Cladribine: An antineoplastic agent used in the treatment of lymphoproliferative diseases including hairy-cell leukemia. [NIH]

PART II: ADDITIONAL RESOURCES AND ADVANCED MATERIAL

ABOUT PART II

In Part II, we introduce you to additional resources and advanced research on primary sclerosing cholangitis. All too often, patients who conduct their own research are overwhelmed by the difficulty in finding and organizing information. The purpose of the following chapters is to provide you an organized and structured format to help you find additional information resources on primary sclerosing cholangitis. In Part II, as in Part I, our objective is not to interpret the latest advances on primary sclerosing cholangitis or render an opinion. Rather, our goal is to give you access to original research and to increase your awareness of sources you may not have already considered. In this way, you will come across the advanced materials often referred to in pamphlets, books, or other general works. Once again, some of this material is technical in nature, so consultation with a professional familiar with primary sclerosing cholangitis is suggested.

CHAPTER 4. STUDIES ON PRIMARY SCLEROSING CHOLANGITIS

Overview

Every year, academic studies are published on primary sclerosing cholangitis or related conditions. Broadly speaking, there are two types of studies. The first are peer reviewed. Generally, the content of these studies has been reviewed by scientists or physicians. Peer-reviewed studies are typically published in scientific journals and are usually available at medical libraries. The second type of studies is non-peer reviewed. These works include summary articles that do not use or report scientific results. These often appear in the popular press, newsletters, or similar periodicals.

In this chapter, we will show you how to locate peer-reviewed references and studies on primary sclerosing cholangitis. We will begin by discussing research that has been summarized and is free to view by the public via the Internet. We then show you how to generate a bibliography on primary sclerosing cholangitis and teach you how to keep current on new studies as they are published or undertaken by the scientific community.

The Combined Health Information Database

The Combined Health Information Database summarizes studies across numerous federal agencies. To limit your investigation to research studies and primary sclerosing cholangitis, you will need to use the advanced search options. First, go to **http://chid.nih.gov/index.html**. From there, select the "Detailed Search" option (or go directly to that page with the following hyperlink: **http://chid.nih.gov/detail/detail.html**). The trick in extracting studies is found in the drop boxes at the bottom of the search page where

"You may refine your search by." Select the dates and language you prefer, and the format option "Journal Article." At the top of the search form, select the number of records you would like to see (we recommend 100) and check the box to display "whole records." We recommend that you type in "primary sclerosing cholangitis" (or synonyms) into the "For these words:" box. Consider using the option "anywhere in record" to make your search as broad as possible. If you want to limit the search to only a particular field, such as the title of the journal, then select this option in the "Search in these fields" drop box. The following is a sample of what you can expect from this type of search:

- **Features of Recurrent Primary Sclerosing Cholangitis in Two Consecutive Liver Allografts After Liver Transplantation**

  Source: Journal of Clinical Gastroenterology. 32(2): 151-154. February 2001.

  Contact: Available from Lippincott Williams and Wilkins, Inc. 12107 Insurance Way, Hagerstown, MD 21740. (800) 638-3030 or (301) 714-2300.

  Summary: Primary sclerosing cholangitis (PSC) is a disease of unknown etiology (cause) that is characterized by inflammation and fibrosis (scarring) of the biliary tree, which results in stricture formation and, eventually, in liver failure. Recurrence of PSC after liver transplantation is very uncommon. The true incidence of recurrence is unknown, mainly because of the difficulty in differentiating ischemic strictures from that of recurrent disease. PSC and ischemic strictures have identical histopathologic and cholangiographic (a type of diagnostic test) features. In this article, the authors report the case of a young man who had recurrence of PSC in two allografts (transplants) and also report their experience with 32 patients who had liver transplantation for PSC. Six patients (18 percent) had evidence of non anastomotic strictures and, of these, only one patient (reported here) had unequivocal evidence of true recurrence. The strictures in the other five patients happened because of ischemia (lack of blood flow to the organ involved). The authors conclude that the recurrence of the disease in two allografts in an immunosuppressed patient (taking drugs to avoid rejection of the transplant), in the absence of ischemia, chronic rejection, or any known pathogen, raises the question of the role of an unidentified infectious agent in the cause of PSC. 2 figures. 2 tables. 14 references.

- **Primary Sclerosing Cholangitis**

  Source: Canadian Journal of Gastroenterology. 14(4): 311-315. April 2000.

Contact: Available from Pulsus Group, Inc. 2902 South Sheridan Way, Oakville, Ontario, Canada L6J 7L6. Fax (905) 829-4799. E-mail: pulsus@pulsus.com.

Summary: This article reviews the treatment of primary sclerosing cholangitis (PSC), a chronic cholestatic liver disease, characterized by fibrosing inflammation and obliteration of intra and or extrahepatic bile ducts. The disease is one of the most common cholestatic diseases in adults and is diagnosed with increasing frequency. It is very often associated with ulcerative colitis (UC). Patients with PSC have an increased incidence of bile duct carcinomas (cancer), and those with UC also have an increased incidence of colonic carcinomas. In end stage disease, liver transplantation is the treatment of choice. Immunosuppressive treatment has little effect. Ursodeoxycholic acid (UDCA), which has been shown to improve liver histology and survival in patients with primary biliary cirrhosis, has a beneficial effect in PSC, provided that patients who develop major duct stenoses (narrowing) are treated endoscopically. The aim is to treat patients as early as possible to prevent progression to the advanced stages of the disease. During treatment with UDCA, stenoses of major ducts may develop, and early endoscopic dilation is highly effective. Because UDCA treatment improves but does not cure cholestatic liver diseases, permanent treatment seems to be necessary. Such prolonged treatment with UDCA may be recommended because, until now, no side effects have been reported. In patients with end stage disease, UDCA is not effective and liver transplantation is indicated. 3 tables. 43 references.

- **Case of Sclerosing Cholangitis Managed by a Percutaneous Approach**

Source: Journal of Clinical Gastroenterology. 30(2): 205-209. March 2000.

Contact: Available from Lippincott Williams and Wilkins, Inc. 12107 Insurance Way, Hagerstown, MD 21740. (800) 638-3030 or (301) 714-2300.

Summary: This article reports on a case in which, in 1992, a 61 year old man who complained of recurrent episodes of fever and jaundice was diagnosed as having sclerosing cholangitis. In the three years that followed, the clinical picture progressively worsened; and, in 1995, the patient was hospitalized again for biliary obstruction. A liver transplantation was excluded because of concomitant severe coronary heart disease. A percutaneous transhepatic cholangiogram showed several critical strictures of the intrahepatic biliary tree and a temporary internal external biliary drainage was placed to relieve the obstruction. After 40 days, a two step percutaneous biliary balloon dilation was performed followed by topical steroid treatment through the catheter. After 45 days, the catheter was removed and steroid treatment tapered

orally. In the three years that followed, the patient was well. He experienced only about 1 to 2 episodes of ascending cholangitis per year requiring antimicrobial therapy. Laboratory analysis showed a gradual improvement in hepatic chemistry, serum bilirubin, and erythrocyte sedimentation rate (ESR). In the patient, the association of percutaneous balloon dilation and topical steroid treatment improved both the clinical and radiological picture, without significant side effects. This approach should be considered a valuable and cost effective option in primary sclerosing cholangitis (PSC), mainly for patients not eligible for liver transplantation.

- **Sclerosing Cholangitis**

  Source: Current Opinion in Gastroenterology. 14(5): 408-411. September 1998.

  Contact: Available from Lippincott Williams and Wilkins Publishers. 12107 Insurance Way, Hagerstown, MD 21740. (800) 637-3030. Fax (301) 824-7390.

  Summary: Primary sclerosing cholangitis (PSC) is associated with inflammatory bowel diseases (IBD) in most cases. There is no proven medical therapy, but endoscopic palliation may benefit some patients. However, orthotopic liver transplantation (OLT) remains the only effective treatment. This article brings readers up to date on the diagnosis and management of PSC. PSC is defined as a chronic cholestatic liver disease of unknown etiology that causes progressive obliterative fibrosis of the biliary tree, ultimately leading to death from liver failure of cholangiocarcinoma. The majority of patients have a circulating antineutrophil cystoplasmic antibody, which does not have a high enough sensitivity to be a good disease marker. Patients with ulcerative colitis (UC) and sclerosing cholangitis are at a higher risk for the development of colonic dysplasia and neoplasia, and yearly colonoscopic surveillance may be warranted. Five year survival after liver transplantation is 85 percent. However, if a cholangiocarcinoma (pancreatic cancer) is present at the time of transplantation, survival decreases markedly. The disease may recur after transplantation, but this does not appear to have any clinical implications. Posttransplant patients who have UC and who are receiving immunosuppressive therapy are also at a high risk for the development of colon cancer. 54 references.

- **Review Article: The Management of Primary Sclerosing Cholangitis**

  Source: Alimentary Pharmacology and Therapeutics. 11(1): 33-43. February 1997.

Contact: Available from Mercury Airfreight International, Ltd. 2323 EF, Randolph Avenue, Avenel, NJ 07001. E-mail: journals.cs@blacksci.co.uk.

Summary: This article reviews the management of primary sclerosing cholangitis (PSC), a chronic cholestatic liver disease characterized by a progressive obliterating fibrosis of the intrahepatic and extrahepatic bile ducts. The pathogenesis of PSC is poorly understood but it is thought to be an immune-mediated disease. The optimal therapy that successfully improves symptoms, delays progression towards liver failure and transplantation, and prevents the onset of cholangiocarcinoma remains elusive. None of the current therapeutic agents have been shown to retard and reverse the rate of disease progression. The authors review the role of cupruretics, corticosteroids, methotrexate, anti-fibrogenic agents, and ursodeoxycholic acid in the treatment of PSC. Orthotopic liver transplantation remains the only therapeutic option for advanced PSC but the timing of transplantation remains controversial and the possibility of recurrence of the disease in the graft is increasingly recognized. The authors hope that greater insight into the pathogenetic mechanisms involved in PSC will allow therapy to be targeted more specifically at the biliary epithelium. 1 table. 65 references. (AA-M).

- **Primary Sclerosing Cholangitis: A Review of Its Clinical Features, Pathogenic Mechanisms, Natural History, and Treatment**

  Source: Advances in Gastroenterology, Hepatology and Clinical Nutrition. 2(4): 111-123. August 1997.

  Contact: Available from Lippincott-Raven Publishers. 12107 Insurance Way, Hagerstown, MD 21740. (800) 638-3030 or (301) 714-2300.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease characterized by progressive obliterative fibrosis of the biliary tree. This article reviews the clinical features, pathogenic mechanisms, natural history, and treatment of PSC. The gold standard for diagnosis is endoscopic retrograde cholangiopancreatography (ERCP), as liver biopsy is rarely diagnostic. Although the etiology of PSC is unknown, a large body of evidence supports immune-mediated mechanisms playing an important role. Genetic factors are also important, as evidenced by the close association between PSC and certain HLA haplotypes. The majority of patients with PSC also have ulcerative colitis (UC). Disease progression can be variable, but ultimately patients develop secondary biliary cirrhosis and liver failure. A significant proportion develop cholangiocarcinoma. There is no effective proven medical treatment, but the authors note that initial results from liver transplantation centers are encouraging. 5 figures. 4 tables. 128 references. (AA).

- **Diagnosing and Managing Primary Sclerosing Cholangitis**

  Source: IM. Internal Medicine. 18(10): 41-43, 47-48, 57. October 1997.

  Contact: Available from Medical Economics. 5 Paragon Drive, Montvale, NJ 07645. (800) 432-4570.

  Summary: This article offers an update on primary sclerosing cholangitis (PSC), a chronic cholestatic liver disease that most often affects young men. About 70 to 80 percent of cases are associated with inflammatory bowel disease (IBD), usually chronic ulcerative colitis (CUC). Topics include the etiology of PSC, diagnostic criteria and clinical features, the evolution of PSC into a ductopenic syndrome, the lack of treatment for PSC, managing cholestasis and portal hypertension, prognostic issues, complications specific to PSC, liver transplantation for end-stage PSC, and the search for more effective therapies for the disease. Complications of PSC include: bacterial cholangitis, gallbladder and biliary stones, dominant bile duct strictures, and bile duct carcinoma. Pruritus (itching) can be debilitating and can lead to severe excoriations and poor quality of life. Since no therapy currently achieves a complete clinical, biochemical, and histologic remission, liver transplantation continues to be an important therapeutic intervention to prolong survival and improve quality of life in patients with end-stage PSC. 2 figures. 43 references.

- **Immunogenetic Aspects of Primary Sclerosing Cholangitis: Implications for Therapeutic Strategies**

  Source: American Journal of Gastroenterology. 90(6): 893-900. June 1995.

  Summary: In this article, the authors summarize the current knowledge of immunogenetic factors in the pathogenesis of primary sclerosing cholangitis (PSC), a chronic cholestatic liver disease often associated with inflammatory bowel disease (IBD). They first outline the genetic and immunologic factors in the pathogenesis of PSC, then present support for drug therapy with ursodeoxycholic acid in the treatment of PSC. They conclude that the finding of cellular and humoral immune abnormalities, as well as the demonstrated genetic predisposition, all point to an autoimmune pathogenesis of PSC. 3 figures. 1 table. 97 references. (AA-M).

- **Endoscopic Therapy in Primary Sclerosing Cholangitis**

  Source: European Journal of Gastroenterology and Hepatology. 4(4): 284-287. 1992.

  Summary: This article discusses the role of endoscopic retrograde cholangiopancreatography (ERCP) in treating primary sclerosing

cholangitis (PSC). The authors stress the role of ERCP in avoiding more invasive transhepatic and surgical methods of improving biliary drainage. Topics include diagnostic ERCP, therapeutic ERCP, nasobiliary drainage, and endoscopic dilatation and stenting. The authors conclude that liver transplantation should be considered when continued hepatocellular impairment is seen, despite endoscopic maneuvers to provide good biliary drainage. 2 figures. 11 annotated references. (AA-M).

- **Primary Sclerosing Cholangitis: What Are the Nursing Implications?**

  Source: Gastroenterology Nursing. 14(4): 215-218. February 1992.

  Summary: Primary sclerosing cholangitis (PSC) is characterized by inflammation and fibrotic strictures of the intra-and extrahepatic bile ducts. This article explores the nursing implications of PSC. The author considers the etiology, diagnosis, and treatment for this condition. The treatment goals in PSC are to provide relief of symptoms, to relieve biliary obstruction, and to protect the liver from cirrhosis and portal hypertension. The patients need emotional support as well as information about procedures and medical therapy. 2 figures. 21 references. (AA-M).

- **Advances in Therapy for Primary Sclerosing Cholangitis**

  Source: European Journal of Gastroenterology and Hepatology. 4(4): 276-283. April 1992.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease frequently associated with inflammatory bowel disease (IBD). The pathogenesis of PSC is thought to be related to immunologic damage directed at bile duct epithelial cells. This article discusses advances in therapy for PSC. Topics include the natural history of the disease and the timing of therapy; managing complications of chronic cholestasis; managing complications of portal hypertension and liver failure; managing complications specific to PSC; and primary medical, radiologic/endoscopic, and surgical treatment for PSC. Drug agents discussed include d-penicillamine, corticosteroids, azathioprine, cyclosporine, and methotrexate. 1 table. 63 annotated references. (AA-M).

- **Natural History and Prognosis in Primary Sclerosing Cholangitis**

  Source: European Journal of Gastroenterology and Hepatology. 4(4): 272-275. April 1992.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease frequently associated with inflammatory bowel disease. This

article discusses the natural history and prognosis of PSC. Topics covered include the presentation of disease, survival statistics, outcome, occurrence of asymptomatic patients, and a multivariate analysis of prognostic factors. The authors note that the outcome for patients with PSC remains a controversial issue, although two survival studies, involving over 100 patients each, reported that the median survival was 12 years and that one-third of patients died or underwent liver transplantation. Clinical, laboratory, and histological features have been shown to be prognostically significant in multivariate analyses. 1 figure. 2 tables. 8 annotated references. (AA-M).

- **Hepatobiliary Pathology of Primary Sclerosing Cholangitis**

  Source: European Journal of Gastroenterology and Hepatology. 4(4): 266-271. April 1992.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease frequently associated with inflammatory bowel disease (IBD). In this article, the histopathological appearances of PSC are reviewed. Although there are no unique features, ductopenia, cholangiectasia, and a fibro-obliterative duct lesion are all highly characteristic, especially when present in combination. The author discusses the many non-specific features of PSC, as well as some recent developments in the investigation of the pathogenesis. The prognostic significance of staging is also discussed. 3 figures. 1 table. 27 annotated references. (AA-M).

Federally-Funded Research on Primary Sclerosing Cholangitis

The U.S. Government supports a variety of research studies relating to primary sclerosing cholangitis and associated conditions. These studies are tracked by the Office of Extramural Research at the National Institutes of Health.[18] CRISP (Computerized Retrieval of Information on Scientific Projects) is a searchable database of federally-funded biomedical research projects conducted at universities, hospitals, and other institutions. Visit the CRISP Web site at **http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket**. You can perform targeted searches by various criteria including geography, date, as

[18] Healthcare projects are funded by the National Institutes of Health (NIH), Substance Abuse and Mental Health Services (SAMHSA), Health Resources and Services Administration (HRSA), Food and Drug Administration (FDA), Centers for Disease Control and Prevention (CDCP), Agency for Healthcare Research and Quality (AHRQ), and Office of Assistant Secretary of Health (OASH).

well as topics related to primary sclerosing cholangitis and related conditions.

For most of the studies, the agencies reporting into CRISP provide summaries or abstracts. As opposed to clinical trial research using patients, many federally-funded studies use animals or simulated models to explore primary sclerosing cholangitis and related conditions. In some cases, therefore, it may be difficult to understand how some basic or fundamental research could eventually translate into medical practice. The following sample is typical of the type of information found when searching the CRISP database for primary sclerosing cholangitis:

- **Project Title: Catalase Immunity in Primary Sclerosing Cholangitis**

  Principal Investigator & Institution: Versalovic, James; ; Massachusetts General Hospital 55 Fruit St Boston, Ma 02114

  Timing: Fiscal Year 2000; Project Start 1-SEP-2000; Project End 0-JUN-2005

  Summary: (adapted from the application) Primary sclerosing cholangitis (PSC) includes features of autoimmunity. The production of autoantibodies in the majority of patients is highlighted by the prevalence of atypical pANCAs and specific anti-catalase immunoglobulins. In addition to self-reactive antibodies, autoreactive T lymphocytes likely participate in PSC as CD4- and CD8-positive T lymphocytes infiltrate affected bile ducts. In the proposed experiments, we will investigate the role of a conserved autoantigen, catalase. Catalase is highly conserved in bacteria and mammals and may induce autoimmune responses by molecular mimicry. Previous studies indicate that catalase is highly expressed in both hepatocytes and biliary epithelium. Affinity-purified autoantibodies from PSC patients will be used to map epitopes of cloned human catalase cDNA as well as selected enterohepatic Helicobacter catalases. Immunodominant epitope-containing peptides isolated by filamentous phage display will be used to examine immunopositivity in PSC patients. To investigate T cell epitopes within this candidate antigen, human and bacterial catalase peptides capable of binding MHC class I antigens have been identified by a computer-assisted HLA-A2.1 peptide binding algorithm. Candidate peptides will be evaluated for in vitro binding with IHLA-A2.1 on T2 hybridoma cells. Peptides which bind HLA-A2.1 will be used to isolate cytotoxic T lymphocyte (CTL) clones from healthy blood donors and the common bile ducts of PSC patients. Molecular mimicry will be examined in vivo by inoculation of wild type and IL-10-deficient mice with H. hepaticus catalase and subsequent analysis of anti-murine catalase immune responses. Splenocyte transfer into syngeneic animals will be

performed to address contributions of anti-catalase cell mediated immune responses to the development of sclerosing cholangitis. Finally, we propose to establish a mouse model linking IBD and sclerosing cholangitis in IL-10-deficient mice. AJ/Cr (IL-10 -/-) mice pruned by immunization with immunodominant H hepaticus catalase peptides will be infected with Helicobacter hepaticus. These mice will likely develop chronic colitis, cholangitis, anti-neutrophil and anti-catalase autoantibodies, an unopposed Thl immune response (consistent with PSC), and an inherent predisposition to fibrosis. Hepatobiliary pathology will be correlated with anti-catalase immune responses. The proposed studies will enhance our understanding of the role of molecular mimicry and autoreactive T cells in inflammatory diseases of the hepatobiliary tract. The proposed animal model will facilitate the evaluation of diagnostic and therapeutic strategies for PSC in vivo.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Mycophenolate Mofetil in Primary Sclerosing Cholangitis**

  Principal Investigator & Institution: Shiffman, Mitchell; ; Virginia Commonwealth University 901 W Franklin St Richmond, Va 23284

  Timing: Fiscal Year 2000

  Summary: An attempt to discover if Mycophenolate Mofetil (MMF) can be of use when treating primary sclerosing cholangitis, the hardening of inflamed bile ducts (PSC). PSC is a progressive liver disorder characterized by the constriction of bile ducts. This results in the obstruction of bile flow within the liver and leads to cirrhosis (scarring). The underlying causes of this disorder are not well understood. However, the constricting process is thought to be based on immune distruction of bile duct lining tissue. This hypothesis is supported by the observation that many people with PSC have other coexistent autoimmune disorders and ulcerative colitis. Several immune suppressive medications have been studied for the treatment of PSC. These include prednisone, azathioprine, and methotrexate, all of little or no benefit. Mycophenolate mofetil (MMF) has been shown to be a strong immunosuppressant for use in solid organ transplant. A total of 30 patients who meet the eligibility criteria and have PSC will be treated in this study. They will be randomly assigned to one of two groups. The first group will receive the currently accepted PSC treatment of ursodeoxycholic acid (UDCA), a dose given according to body weight. The second group will receive the same dose of UDCA plus MMF. Patients will be treated for a total of 24 months. Liver function and enzymes will be tested at baseline, 1,2, and 4 weeks. Tests will be done at

monthly intervals. After 24 months of treatment, all patients will undergo a liver biopsy to determine the results of the treatment.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Trial of 4 Treatment Regimens in PTS with Primary Sclerosing Cholangitis**

  Principal Investigator & Institution: Lee, Young-Mee; ; New England Medical Center Hospitals 750 Washington St Boston, Ma 02111

  Timing: Fiscal Year 2000

  Summary: Primary sclerosing cholangitis (PSC) is a chronic progressive liver disease that is characterized by on-going inflammation and destruction of intrahepatic and extrahepatic bile ducts. Primary sclerosing cholangitis generally affects middle aged men and is the fourth leading indication for liver transplantation in adults in the United States. In this pilot trial, the efficacy of four different treatment regimens will be prospectively studied: 1)Ursodiol alone 2) ursodol and rotating oral antibiotics 3)ursodiol and low-dose oral methotrexate, and 4) Ursodiol with methotrexate and oral antibiotics.

  Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Biliary Calmodulin Dependent Protein Kinase II Isoforms**

  Principal Investigator & Institution: Stansfield, Ann P.; Medicine; Indiana Univ-Purdue Univ at Indianapolis 355 N Lansing Indianapolis, in 46202

  Timing: Fiscal Year 2000; Project Start 1-APR-1996; Project End 1-MAR-2001

  Summary: The underlying cause of the most common inherited disease among Caucasians, cystic fibrosis, is a mutation in a cAMP-dependent Cl-channel. In addition to the respiratory problems associated with this disease, some patients also suffer from cholestasis (decreased bile flow resulting in Cirrhosis). In turn, this suggests that other cholestatic liver diseases such as primary biliary cirrhosis, primary sclerosing Cholangitis, sarcoidosis, liver transplant rejection and AIDS Cholangiopathy may also occur from or cause alterations in Cl- secretion. In biliary epithelial cells (BECs), another type of Cl- channel is regulated by Ca2+, in part, through the actions of Ca2+/calmodulin- dependent protein kinase II (CaMKII). The long-term goals of this project are to understand the mechanisms by which Ca2+ signals arising from extracellular stimuli are transduced into CaMKII activation and regulate Ca2+ dependent Cl- secretion in both normal and diseased states. In BECs, multiple isoforms of CaMKII are expressed which differ in their association domains. These domains are

involved primarily in holoenzyme formation and subcellular localization. The hypothesis of this proposal is that the CaMKII subunits primary structures will determine the holoenzymes' structural and functional characteristics and that only a subset of these holoenzymes will regulate Cl secretion. To test this hypothesis the specific aims of this grant are to: (1) characterize the activities and holoenzyme structures of CaMKII isoforms overexpressed in a biliary epithelial cell line; (2) using subcellular fractionation and immunocytochemical methods, determine the subcellular localizations of CaMKII isoforms; and (3) using [125I] efflux and patch clamp recordings, determine which CaMKII isoforms regulate Ca2+ dependent Cl- secretion. These experiments are unique since they will identify and characterize specific CaMKII isoforms involved in regulating Ca2+-dependent Cl- secretion.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: CFTR Function/ Gene Mutation in Primary Sclerosing Chola**

  Principal Investigator & Institution: Freedman, Steven D.; ; Beth Israel Deaconess Medical Center 330 Brookline Ave Boston, Ma 02215

  Timing: Fiscal Year 2000; Project Start 1-DEC-1977; Project End 0-NOV-2004

  Summary: The purpose of this study is to investigate the pathogenesis of PSC by studying the function of CFTR chloride channels as well as performing limited DNA analysis of the CFTR gene.

  Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Dysregulation of Cholangiocyte Apoptosis by Cytokines**

  Principal Investigator & Institution: Patel, Tushar C.; ; Scott and White Memorial Hospital 2401 S 31St St Temple, Tx 76508

  Timing: Fiscal Year 2000; Project Start 0-SEP-1998; Project End 1-AUG-2003

  Summary: Chronic inflammation in gastrointestinal epithelia is characterized by alterations in the cytokine milieu and often predisposes to the development of neoplasia. Primary sclerosing cholangitis, a chronic inflammatory condition affecting the biliary system, is associated with cytokine alterations and the development of cholangiocarcinoma. Dysregulation of apoptosis has recently been implicated as an important contributor to carcinogenesis. We have recently shown that the pro-inflammatory cytokine IL-6 alters the susceptibility of biliary epithelial cells to undergo apoptosis by downregulating Bax, a member of the Bcl-2 family and a dominant tumor suppressor and pro-apoptotic gene. Thus,

our HYPOTHESIS is that dysregulation of apoptosis by cytokines is an important mechanism of malignant transformation of biliary epithelia. The SPECIFIC AIMS for this proposal are: 1) To test the hypothesis that pro-inflammatory cytokines such as IL-1, IL-6 and TNFalpha regulate apoptosis by altering the expression or activity of members of the Bcl-2 family of apoptosis regulators; 2) to test the hypothesis that IL-6 alters expression of Bax by a ligand mediated activation of specific signaling pathways resulting in altered gene transcription; and 3) to test the hypothesis that dysregulation of apoptosis by cytokines enhances malignant transformation in biliary epithelial cells due to genotoxic injury. These studies will help to determine the role of cytokines in injury and transformation of biliary epithelia and expand our understanding of the mechanisms mediating malignant transformation in inflammatory states. This information has the promise of ultimately contributing to the development of specific therapies for the cholangiopathies as well as interventions to prevent neoplasia.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Gene Delivery to Bile Duct Epithelial Cells in Mice**

  Principal Investigator & Institution: Subbotin, Vladimir M.; ; Mirus Corporation 505 S Rosa Rd, #104 Madison, Wi 53711

  Timing: Fiscal Year 2001; Project Start 8-SEP-2001; Project End 1-JUL-2002

  Summary: Primary biliary cirrhosis and primary sclerosing cholangitis are irreversible liver diseases of unknown etiology with no available treatment or preventive therapy, and only liver transplantation can prolong life of patients with disease progression. The hallmark of these diseases is fibrotic accumulation around hepatic bile ducts that normally drain bile from liver to intestine. This periductular fibrosis causes bile duct compression and cholestasis. Biliary epithelial cells (BEC) line each of these bile ducts and are in direct contact with the accumulating fibrotic tissue. Our goal is to use these BEC as a target cell type for gene therapy intervention aimed at halting disease progression. The advantages of this approach in terms of gene delivery potential are: 1) BECs are always in a direct contact with compressing fibrotic tissue; and 2) BECs are readily accessible using a endoscopic procedure commonly used in the clinic. A mouse model of peribiliary fibrosis/cirrhosis induced by common bile duct ligation has been developed and will be ideal for these studies. We will use this mouse model to deliver gene complexes to BEC's via a catheter. The overall goal is to identify non-viral transfection vectors and/or methodologies that deliver genes to the BEC with high efficiency. Phase II of this research will incorporate the most efficient gene delivery complexes to identify specific genes that are effective in blocking

peribiliary cirrhosis development. PROPOSED COMMERCIAL APPLICATIONS: Primary biliary cirrhosis and primary sclerosing cholangitis are irreversible liver diseases in which liver transplantation is the only viable treatment. However, this treatment is limited by shortage of donor livers and extremely high cost. This grant is ultimately aimed at identifying alternative gene therapy treatments for these diseases. The phase I grant will determine the feasibility of delivering genes into the disease associated cells using non-viral vectors.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Liver Transplantation--Models for Patient Management**

  Principal Investigator & Institution: Dickson, E R.; Mary Lowell Leary Professor of Medicine; Mayo Clinic Rochester 200 1St St Sw Rochester, Mn 55905

  Timing: Fiscal Year 2000; Project Start 5-JAN-1986; Project End 0-NOV-2001

  Summary: (Adapted from Investigator's Abstract) Liver transplantation (OLT) is now accepted as an effective therapy for a variety of otherwise untreatable acute and chronic liver diseases. The success and broadened indications for OLT have resulted in a growing disparity between the number of potential recipients and the limited availability of donor organs. During the past decade, research has documented the efficacy of OLT and survival models have been developed for cohorts of patients with primary biliary cirrhosis and primary sclerosing cholangitis (PBC and PSC). The ultimate need for organ transplantation prior to the development of end stage liver disease in PBC and PSC patients can be predicted. A continuing challenge for OLT, as a discipline, is to more precisely define and predict its effects for individual subjects, both for the selection and timing of this important therapeutic procedure. Currently, there are three important needs and opportunities in this area as follow: 1) broaden the scope of models to include patients with chronic hepatitis C and alcoholic liver disease; 2) move from models based upon cohorts of patients to those focused on individual patients, and 3) more thoroughly investigate previously identified risk factors, such as malnutrition, in order to better understand their impact on patient outcome and health care expenditures. Health care reform will increasingly demand greater accountability for economic efficiency, maintenance of quality of medical care, and the apportionment of limited resources to those patients who will optimally benefit. This research is intended to establish models that will provide objective data to assist patients, physicians and third party carriers in making crucial medical and health policy decisions.

  Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: Multicentered Randomized Trial of High-Dose URSO in PSC**

  Principal Investigator & Institution: Lindor, Keith D.; Professor and Director; Mayo Clinic Rochester 200 1St St Sw Rochester, Mn 55905

  Timing: Fiscal Year 2001; Project Start 1-SEP-2001; Project End 1-AUG-2006

  Summary: (provided by applicant): Primary sclerosing cholangitis (PSC) is a progressive chronic cholestatic liver disease of unknown etiology that is commonly associated with chronic colitis. PSC, a common liver disease among adults, usually leads to advanced liver disease and liver failure, and as such, is an important indication for liver transplantation. Unfortunately, no effective medical therapy currently exists for PSC. The group of investigators has a longstanding track record of clinical trials in chronic cholestatic liver disease, particularly in primary sclerosing cholangitis. Recently we have generated promising results from a pilot study using high doses of ursodeoxycholic acid in the treatment of PSC. Lower doses of ursodeoxycholic acid in patients with PSC led to biochemical improvement but did not affect other clinically important endpoints in a previous study. Our pilot data is supported by data from another group showing in a small, randomized trial that high dose ursodeoxycholic acid led to biochemical, histologic, and cholangiographic improvement compared to placebo at two years. In this submission, we propose a large-scale multi-center placebo-controlled randomized trial with a minimum follow-up of four years for 150 patients with primary sclerosing cholangitis. Primary endpoints of the study will include histologic progression to cirrhosis, development of esophageal or gastric varices, need for liver transplantation, and survival. Secondary endpoints will include measurements of the effects of urosodeoxycholic acid (28-30 mg/kd/d) on liver biochemistries, histologic stage, cholangiographic features, Mayo risk score, and quality of life, using validated questionnaires. This study will be the largest ever conducted in PSC and the follow-up will be the most extensive. This will provide an invaluable resource for studying the natural history of this disease and as part of this study we will also collect serum, cells for extraction of DNA, bile, and tissue from the liver and colon as a resource for future studies. The multi-centered nature of this trial will allow recruitment of patients into this study from a diverse patient population, representative of the gender and racial distribution of this disease. Chances of successful completion of this study are enhanced by the large PSC patient population that the participating centers currently manage, recognition of these sites as referral centers for new patients with PSC, as well as a longstanding track record in clinical trials in cholestatic liver disease.

Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

- **Project Title: TCR Usage and Function in IBD**

  Principal Investigator & Institution: Saubermann, Lawrence J.; ; Boston Medical Center 1 Boston Medical Ctr Pl Boston, Ma 02118

  Timing: Fiscal Year 2000; Project Start 0-SEP-1998; Project End 9-SEP-2003

  Summary: (taken from application) T-cells are likely to play a crucial role in inflammatory bowel disease (IBD) pathogenesis. Our focus has centered on the analysis of the T-cell receptor (TCR), the specific antigen recognition element of the T-cell, as a means to implicate T-cell responses to specific antigen(s) in disease pathogenesis and to potentially identify disease relevant T-cell clones. Our previous studies on TCR usage have clearly shown that a highly specific T-cell response is present in individuals with IBD and this response is characterized by persistent T-cell expansions which are shared by genetically related individuals. This directly implicates a response by T-cells in IBD affected individuals to conventional antigen(s). The major goals of this grant proposal are to further define the nature of these T-cell expansions in IBD and investigate for potential T-cell recognized antigen(s) relevant to IBD pathogenesis. Specifically, we plan to characterize: (1) the TCR usage of the IL-2 receptor positive subset of intestinal T-cells; (2) the TCR usage of the bile-duct associated T-cells in the clinically distinct subset of individuals with primary sclerosing cholangitis, and; (3) the relationship between major histocompatibility complex (MHC) and TCR usage in families with IBD. By utilizing this information on TCR usage, specific T-cell clones of interest, which express TCRs that are a part of a public or private motif, will be isolated and utilized in long-term, molecular and cellular based strategies. These strategies, which are based on rapidly emerging technologies, will aim to define the cognate antigen(s) of these potentially disease relevant T-cells, and thus allow further insight into the etiopathogenesis of IBD.

  Website: http://commons.cit.nih.gov/crisp3/CRISP.Generate_Ticket

E-Journals: PubMed Central[19]

PubMed Central (PMC) is a digital archive of life sciences journal literature developed and managed by the National Center for Biotechnology

[19] Adapted from the National Library of Medicine: **http://www.pubmedcentral.nih.gov/about/intro.html**.

Information (NCBI) at the U.S. National Library of Medicine (NLM).[20] Access to this growing archive of e-journals is free and unrestricted.[21] To search, go to **http://www.pubmedcentral.nih.gov/index.html#search**, and type "primary sclerosing cholangitis" (or synonyms) into the search box. This search gives you access to full-text articles. The following is a sample of items found for primary sclerosing cholangitis in the PubMed Central database:

- **Identification of Helicobacter pylori and Other Helicobacter Species by PCR, Hybridization, and Partial DNA Sequencing in Human Liver Samples from Patients with Primary Sclerosing Cholangitis or Primary Biliary Cirrhosis** by Hans-Olof Nilsson, Jalal Taneera, Maria Castedal, Elisabeth Glatz, Rolf Olsson, and Torkel Wadstrom; 2000 March http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=86342&rendertype=external

The National Library of Medicine: PubMed

One of the quickest and most comprehensive ways to find academic studies in both English and other languages is to use PubMed, maintained by the National Library of Medicine. The advantage of PubMed over previously mentioned sources is that it covers a greater number of domestic and foreign references. It is also free to the public.[22] If the publisher has a Web site that offers full text of its journals, PubMed will provide links to that site, as well as to sites offering other related data. User registration, a subscription fee, or some other type of fee may be required to access the full text of articles in some journals.

To generate your own bibliography of studies dealing with primary sclerosing cholangitis, simply go to the PubMed Web site at **www.ncbi.nlm.nih.gov/pubmed**. Type "primary sclerosing cholangitis" (or

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[20] With PubMed Central, NCBI is taking the lead in preservation and maintenance of open access to electronic literature, just as NLM has done for decades with printed biomedical literature. PubMed Central aims to become a world-class library of the digital age.

[21] The value of PubMed Central, in addition to its role as an archive, lies the availability of data from diverse sources stored in a common format in a single repository. Many journals already have online publishing operations, and there is a growing tendency to publish material online only, to the exclusion of print.

[22] PubMed was developed by the National Center for Biotechnology Information (NCBI) at the National Library of Medicine (NLM) at the National Institutes of Health (NIH). The PubMed database was developed in conjunction with publishers of biomedical literature as a search tool for accessing literature citations and linking to full-text journal articles at Web sites of participating publishers. Publishers that participate in PubMed supply NLM with their citations electronically prior to or at the time of publication.

synonyms) into the search box, and click "Go." The following is the type of output you can expect from PubMed for "primary sclerosing cholangitis" (hyperlinks lead to article summaries):

Vocabulary Builder

Antibody: An immunoglobulin molecule that has a specific amino acid sequence by virtue of which it interacts only with the antigen that induced its synthesis in cells of the lymphoid series (especially plasma cells), or with antigen closely related to it. Antibodies are classified according to their ode of action as agglutinins, bacteriolysins, haemolysins, opsonins, precipitins, etc. [EU]

Antigens: Substances that cause an immune response in the body. The body "sees" the antigens as harmful or foreign. To fight them, the body produces antibodies, which attack and try to eliminate the antigens. [NIH]

Antimicrobial: Killing microorganisms, or suppressing their multiplication or growth. [EU]

Asymptomatic: No symptoms; no clear sign of disease present. [NIH]

Atypical: Irregular; not conformable to the type; in microbiology, applied specifically to strains of unusual type. [EU]

Autoimmunity: Process whereby the immune system reacts against the body's own tissues. Autoimmunity may produce or be caused by autoimmune diseases. [NIH]

Bilirubin: A bile pigment that is a degradation product of HEME. [NIH]

Calmodulin: A heat-stable, low-molecular-weight activator protein found mainly in the brain and heart. The binding of calcium ions to this protein allows this protein to bind to cyclic nucleotide phosphodiesterases and to adenyl cyclase with subsequent activation. Thereby this protein modulates cyclic AMP and cyclic GMP levels. [NIH]

Carcinoma: A malignant new growth made up of epithelial cells tending to infiltrate the surrounding tissues and give rise to metastases. [EU]

Catalase: An oxidoreductase that catalyzes the conversion of hydrogen peroxide to water and oxygen. It is present in many animal cells. A deficiency of this enzyme results in acatalasia. EC 1.11.1.6. [NIH]

Catheter: A tubular, flexible, surgical instrument for withdrawing fluids from (or introducing fluids into) a cavity of the body, especially one for introduction into the bladder through the urethra for the withdraw of urine. [EU]

Cholestasis: Impairment of biliary flow at any level from the hepatocyte to

Vater's ampulla. [NIH]

Colonoscopy: Endoscopic examination, therapy or surgery of the luminal surface of the colon. [NIH]

Colorectal: Pertaining to or affecting the colon and rectum. [EU]

Concomitant: Accompanying; accessory; joined with another. [EU]

Constriction: The act of constricting. [NIH]

Coronary: Encircling in the manner of a crown; a term applied to vessels; nerves, ligaments, etc. The term usually denotes the arteries that supply the heart muscle and, by extension, a pathologic involvement of them. [EU]

Cytokines: Non-antibody proteins secreted by inflammatory leukocytes and some non-leukocytic cells, that act as intercellular mediators. They differ from classical hormones in that they are produced by a number of tissue or cell types rather than by specialized glands. They generally act locally in a paracrine or autocrine rather than endocrine manner. [NIH]

Cytotoxic: Pertaining to or exhibiting cytotoxicity. [EU]

Dilatation: The condition, as of an orifice or tubular structure, of being dilated or stretched beyond the normal dimensions. [EU]

Dysplasia: Abnormality of development; in pathology, alteration in size, shape, and organization of adult cells. [EU]

Enterohepatic: Of or involving the intestine and liver. [EU]

Enzyme: A protein molecule that catalyses chemical reactions of other substances without itself being destroyed or altered upon completion of the reactions. Enzymes are classified according to the recommendations of the Nomenclature Committee of the International Union of Biochemistry. Each enzyme is assigned a recommended name and an Enzyme Commission (EC) number. They are divided into six main groups; oxidoreductases, transferases, hydrolases, lyases, isomerases, and ligases. [EU]

Epithelium: The covering of internal and external surfaces of the body, including the lining of vessels and other small cavities. It consists of cells joined by small amounts of cementing substances. Epithelium is classified into types on the basis of the number of layers deep and the shape of the superficial cells. [EU]

Epitopes: Sites on an antigen that interact with specific antibodies. [NIH]

Extracellular: Outside a cell or cells. [EU]

Extraction: The process or act of pulling or drawing out. [EU]

Fibrosis: The formation of fibrous tissue; fibroid or fibrous degeneration [EU]

Gastrointestinal: Pertaining to or communicating with the stomach and intestine, as a gastrointestinal fistula. [EU]

Haplotypes: The genetic constitution of individuals with respect to one member of a pair of allelic genes, or sets of genes that are closely linked and tend to be inherited together such as those of the major histocompatibility complex. [NIH]

Helicobacter: A genus of gram-negative, spiral-shaped bacteria that is pathogenic and has been isolated from the intestinal tract of mammals, including humans. [NIH]

Hepatobiliary: Pertaining to the liver and the bile or the biliary ducts. [EU]

Hepatocellular: Pertaining to or affecting liver cells. [EU]

Hepatocytes: The main structural component of the liver. They are specialized epithelial cells that are organized into interconnected plates called lobules. [NIH]

Histocompatibility: The degree of antigenic similarity between the tissues of different individuals, which determines the acceptance or rejection of allografts. [NIH]

Humoral: Of, relating to, proceeding from, or involving a bodily humour - now often used of endocrine factors as opposed to neural or somatic. [EU]

Hybridization: The genetic process of crossbreeding to produce a hybrid. Hybrid nucleic acids can be formed by nucleic acid hybridization of DNA and RNA molecules. Protein Hybridization allows for hybrid proteins to be formed from polypeptide chains. [NIH]

Immunity: The condition of being immune; the protection against infectious disease conferred either by the immune response generated by immunization or previous infection or by other nonimmunologic factors (innate i.). [EU]

Immunization: The induction of immunity. [EU]

Immunosuppressant: An agent capable of suppressing immune responses. [EU]

Ischemia: Deficiency of blood in a part, due to functional constriction or actual obstruction of a blood vessel. [EU]

Lesion: Any pathological or traumatic discontinuity of tissue or loss of function of a part. [EU]

Ligation: Application of a ligature to tie a vessel or strangulate a part. [NIH]

Localization: 1. the determination of the site or place of any process or lesion. 2. restriction to a circumscribed or limited area. 3. prelocalization. [EU]

Malignant: Tending to become progressively worse and to result in death. Having the properties of anaplasia, invasion, and metastasis; said of tumours. [EU]

Methotrexate: An antineoplastic antimetabolite with immunosuppressant

properties. It is an inhibitor of dihydrofolate reductase and prevents the formation of tetrahydrofolate, necessary for synthesis of thymidylate, an essential component of DNA. [NIH]

Pathogen: Any disease-producing microorganism. [EU]

Penicillamine: 3-Mercapto-D-valine. The most characteristic degradation product of the penicillin antibiotics. It is used as an antirheumatic and as a chelating agent in Wilson's disease. [NIH]

Percutaneous: Performed through the skin, as injection of radiopacque material in radiological examination, or the removal of tissue for biopsy accomplished by a needle. [EU]

Predisposition: A latent susceptibility to disease which may be activated under certain conditions, as by stress. [EU]

Prednisone: A synthetic anti-inflammatory glucocorticoid derived from cortisone. It is biologically inert and converted to prednisolone in the liver. [NIH]

Prevalence: The number of people in a given group or population who are reported to have a disease. [NIH]

Progressive: Advancing; going forward; going from bad to worse; increasing in scope or severity. [EU]

Pruritus: Itching skin; may be a symptom of diabetes. [NIH]

Receptor: 1. a molecular structure within a cell or on the surface characterized by (1) selective binding of a specific substance and (2) a specific physiologic effect that accompanies the binding, e.g., cell-surface receptors for peptide hormones, neurotransmitters, antigens, complement fragments, and immunoglobulins and cytoplasmic receptors for steroid hormones. 2. a sensory nerve terminal that responds to stimuli of various kinds. [EU]

Recurrence: The return of a sign, symptom, or disease after a remission. [NIH]

Remission: A diminution or abatement of the symptoms of a disease; also the period during which such diminution occurs. [EU]

Respiratory: Pertaining to respiration. [EU]

Retrograde: 1. moving backward or against the usual direction of flow. 2. degenerating, deteriorating, or catabolic. [EU]

Sarcoidosis: An idiopathic systemic inflammatory granulomatous disorder comprised of epithelioid and multinucleated giant cells with little necrosis. It usually invades the lungs with fibrosis and may also involve lymph nodes, skin, liver, spleen, eyes, phalangeal bones, and parotid glands. [NIH]

Secretion: 1. the process of elaborating a specific product as a result of the activity of a gland; this activity may range from separating a specific

substance of the blood to the elaboration of a new chemical substance. 2. any substance produced by secretion. [EU]

Sedimentation: The act of causing the deposit of sediment, especially by the use of a centrifugal machine. [EU]

Serum: The clear portion of any body fluid; the clear fluid moistening serous membranes. 2. blood serum; the clear liquid that separates from blood on clotting. 3. immune serum; blood serum from an immunized animal used for passive immunization; an antiserum; antitoxin, or antivenin. [EU]

Species: A taxonomic category subordinate to a genus (or subgenus) and superior to a subspecies or variety, composed of individuals possessing common characters distinguishing them from other categories of individuals of the same taxonomic level. In taxonomic nomenclature, species are designated by the genus name followed by a Latin or Latinized adjective or noun. [EU]

Topical: Pertaining to a particular surface area, as a topical anti-infective applied to a certain area of the skin and affecting only the area to which it is applied. [EU]

Viral: Pertaining to, caused by, or of the nature of virus. [EU]

CHAPTER 5. PATENTS ON PRIMARY SCLEROSING CHOLANGITIS

Overview

You can learn about innovations relating to primary sclerosing cholangitis by reading recent patents and patent applications. Patents can be physical innovations (e.g. chemicals, pharmaceuticals, medical equipment) or processes (e.g. treatments or diagnostic procedures). The United States Patent and Trademark Office defines a patent as a grant of a property right to the inventor, issued by the Patent and Trademark Office.[23] Patents, therefore, are intellectual property. For the United States, the term of a new patent is 20 years from the date when the patent application was filed. If the inventor wishes to receive economic benefits, it is likely that the invention will become commercially available to patients with primary sclerosing cholangitis within 20 years of the initial filing. It is important to understand, therefore, that an inventor's patent does not indicate that a product or service is or will be commercially available to patients with primary sclerosing cholangitis. The patent implies only that the inventor has "the right to exclude others from making, using, offering for sale, or selling" the invention in the United States. While this relates to U.S. patents, similar rules govern foreign patents.

In this chapter, we show you how to locate information on patents and their inventors. If you find a patent that is particularly interesting to you, contact the inventor or the assignee for further information.

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[23] Adapted from The U. S. Patent and Trademark Office: **http://www.uspto.gov/web/offices/pac/doc/general/whatis.htm**.

Patents on Primary Sclerosing Cholangitis

By performing a patent search focusing on primary sclerosing cholangitis, you can obtain information such as the title of the invention, the names of the inventor(s), the assignee(s) or the company that owns or controls the patent, a short abstract that summarizes the patent, and a few excerpts from the description of the patent. The abstract of a patent tends to be more technical in nature, while the description is often written for the public. Full patent descriptions contain much more information than is presented here (e.g. claims, references, figures, diagrams, etc.). We will tell you how to obtain this information later in the chapter. The following is an example of the type of information that you can expect to obtain from a patent search on primary sclerosing cholangitis:

- **Methods for selectively detecting perinuclear anti-neutrophil cytoplasmic antibody of ulcerative colitis or primary sclerosing cholangitis**

  Inventor(s): Targan; Stephan R. (Los Angeles, CA), Vidrich; Alda (Pacific Palisades, CA)

  Assignee(s): Cedars-Sinai Medical Center (Los Angeles, CA)

  Patent Number: 5,750,355

  Date filed: October 7, 1994

  Abstract: The invention is directed to methods and kits for detecting and measuring the presence or absence of perinuclear anti-neutrophil cytoplasmic autoantibody of ulcerative colitis or primary sclerosing cholangitis. The methods and kits of the present invention provide safe and reliable means for diagnosing ulcerative colitis and primary sclerosing cholangitis. The antigens reactive with perinuclear anti-neutrophil cytoplasmic autoantibody of ulcerative colitis and primary sclerosing cholangitis are also provided.

  Excerpt(s): The invention relates to methods of detecting and measuring the presence or absence of perinuclear anti-neutrophil cytoplasmic autoantibodies of ulcerative colitis or primary sclerosing cholangitis. More specifically, the methods of the present invention employ DNase treatment of neutrophils in assays such as ELISA and immunofluorescence to elicit the loss of a positive control value when the autoantibody is present. ... The present invention provides methods of detecting and measuring the presence or absence of perinuclear anti-neutrophil cytoplasmic autoantibodies (p-ANCA) of ulcerative colitis (UC) or primary sclerosing cholangitis (PSC) in a sample. More specifically, the presence of p-ANCA of UC or PSC is detected by

assaying for the loss of a positive value (i.e., loss of a detectable marker as compared to a control) upon treatment of neutrophils with DNase. ... The present invention provides methods and kits for detecting the presence of perinuclear anti-neutrophil cytoplasmic autoantibody (p-ANCA) for ulcerative colitis (UC) or primary sclerosing cholangitis (PSC) in a sample. Inventive methods involve assaying for the loss of a positive value (as compared to a control) upon treatment of neutrophils with DNAase.

Web site: http://www.delphion.com/details?pn=US05750355__

Patent Applications on Primary Sclerosing Cholangitis

As of December 2000, U.S. patent applications are open to public viewing.[24] Applications are patent requests which have yet to be granted (the process to achieve a patent can take several years).

Keeping Current

In order to stay informed about patents and patent applications dealing with primary sclerosing cholangitis, you can access the U.S. Patent Office archive via the Internet at no cost to you. This archive is available at the following Web address: **http://www.uspto.gov/main/patents.htm**. Under "Services," click on "Search Patents." You will see two broad options: (1) Patent Grants, and (2) Patent Applications. To see a list of granted patents, perform the following steps: Under "Patent Grants," click "Quick Search." Then, type "primary sclerosing cholangitis" (or synonyms) into the "Term 1" box. After clicking on the search button, scroll down to see the various patents which have been granted to date on primary sclerosing cholangitis. You can also use this procedure to view pending patent applications concerning primary sclerosing cholangitis. Simply go back to the following Web address: **http://www.uspto.gov/main/patents.htm**. Under "Services," click on "Search Patents." Select "Quick Search" under "Patent Applications." Then proceed with the steps listed above.

Vocabulary Builder

Assay: Determination of the amount of a particular constituent of a mixture,

[24] This has been a common practice outside the United States prior to December 2000.

or of the biological or pharmacological potency of a drug. [EU]

Neutrophils: Granular leukocytes having a nucleus with three to five lobes connected by slender threads of chromatin, and cytoplasm containing fine inconspicuous granules and stainable by neutral dyes. [NIH]

CHAPTER 6. BOOKS ON PRIMARY SCLEROSING CHOLANGITIS

Overview

This chapter provides bibliographic book references relating to primary sclerosing cholangitis. You have many options to locate books on primary sclerosing cholangitis. The simplest method is to go to your local bookseller and inquire about titles that they have in stock or can special order for you. Some patients, however, feel uncomfortable approaching their local booksellers and prefer online sources (e.g. **www.amazon.com** and **www.bn.com**). In addition to online booksellers, excellent sources for book titles on primary sclerosing cholangitis include the Combined Health Information Database and the National Library of Medicine. Once you have found a title that interests you, visit your local public or medical library to see if it is available for loan.

Book Summaries: Federal Agencies

The Combined Health Information Database collects various book abstracts from a variety of healthcare institutions and federal agencies. To access these summaries, go directly to the following hyperlink: **http://chid.nih.gov/detail/detail.html**. You will need to use the "Detailed Search" option. To find book summaries, use the drop boxes at the bottom of the search page where "You may refine your search by." Select the dates and language you prefer. For the format option, select "Monograph/Book." Now type "primary sclerosing cholangitis" (or synonyms) into the "For these words:" box. You will only receive results on books. You should check back periodically with this database which is updated every 3 months. The

following is a typical result when searching for books on primary sclerosing cholangitis:

- **Liver Disorders Sourcebook**

  Source: Detroit, MI: Omnigraphics. 2000. 591 p.

  Contact: Available from Omnigraphics, Inc. 615 Griswold, Detroit, MI 48226. (800) 234-1340. Fax (800) 875-1340. PRICE: $78.00 plus shipping and handling. ISBN: 0780802403.

  Summary: This Sourcebook provides basic health care information about liver functions, guidelines for liver health, and tests that assess liver distress. The book also presents the symptoms, treatments, and preventive measures available for liver cancer; hepatitis A, B, C, D and E; genetically based liver diseases; and other liver diseases. The liver transplantation process is explained. Specific topics include strategies for protecting the liver, risk factors, common laboratory tests in liver disease, liver biopsy, cancer tumor markers, cirrhosis (scarring of the liver), infectious agents and parasites, pregnancy and the liver, jaundice in the healthy newborn, the liver's response to drugs, alcohol and the liver, acetaminophen, herbs and alternative medicine, galactosemia, Gaucher disease, hereditary hemochromatosis, Niemann-Pick disease, Wilson's disease, biliary atresia, cystic disease of the liver, fatty liver, gallstones, primary biliary cirrhosis, primary sclerosing cholangitis, organ donation, and the bioartificial liver. A glossary, a directory of organizations and support groups with up to date contact information (including websites and email addresses), a listing of transplant centers, and a subject index conclude the volume.

- **Gallbladder and Biliary Tract Diseases**

  Source: New York, NY: Marcel Dekker, Inc. 2000. 928 p.

  Contact: Available from Marcel Dekker, Inc. Cimarron Road, P.O. Box 5005, Monticello, NY 12701. (800) 228-1160 or (845) 796-1919. Fax (845) 796-1772. E-mail: custserv@dekker.com. International E-mail: intlcustserv@dekker.com. Website: www.dekker.com. PRICE: $250.00 plus shipping and handling. ISBN: 0824703111.

  Summary: The gallbladder and biliary tract are the 'orphan' organs of the digestive system, falling between the realms of the solid organ liver specialist and the hollow organ intestinal expert. This comprehensive text covers the gallbladder and biliary tract disease, noting that the management of gallbladder and biliary disease is truly multidisciplinary, involving gastroenterologists, surgeons, endoscopists, and radiologists. The text attempts to translate advances in basic science into clinically

relevant treatment and to bridge the gap between clinical disciplines. Parts I and II focus on important physiological and pathophysiological principles, with a special emphasis on gallstones. In Parts III to V, the authors focus on clinical disorders of the gallbladder and biliary tree, with input on management from surgeons, endoscopists, and radiologists. New imaging techniques, such as magnetic resonance cholangiography and endoscopic ultrasound, are discussed from both the radiologist's and endoscopist's perspective, and their role in disease management is defined. The 37 chapters cover the neurobiology of the gallbladder, gallbladder mucosal function, gallbladder smooth muscle function and dysfunction, canalicular lipid secretion, bile ductal secretion and its regulation, the pathogenesis of gallstones, pigment gallstones, cholesterol crystallization in bile, normal gallbladder motor functions, gallbladder motility and gallstones, the role of intestinal transit, prevention of gallstones, the gallbladder and biliary tree in cystic fibrosis, the silent gallstone, biliary crystals and sludge, biliary colic and acute cholecystitis (gallbladder infection), laparoscopic cholecystectomy (removal of the gallbladder), nonsurgical therapy of gallstones, biliary lithotripsy, topical contact dissolution of gallbladder stones, common bile duct stones, acalculous cholecystitis, gallbladder cancer, primary sclerosing cholangitis, vanishing bile duct syndrome, cholangiocarcinoma (bile duct cancer), ampullary tumors, infections of the bile ducts, and bile duct injuries. Each chapter includes extensive references and the text concludes with a detailed subject index.

- **Clinical Practice of Gastroenterology. Volume Two**

  Source: Philadelphia, PA: Current Medicine. 1999. 861 p.

  Contact: Available from W.B. Saunders Company. Order Fulfillment, 6277 Sea Harbor Drive, Orlando, FL 32887. (800) 545-2522. Fax (800) 874-6418 or (407) 352-3445. Website: www.wbsaunders.com. PRICE: $235.00 plus shipping and handling. ISBN: 0443065209 (two volume set); 0443065217 (volume 1); 0443065225 (volume 2).

  Summary: This lengthy textbook brings practitioners up to date on the complexities of gastroenterology practice, focusing on the essentials of patient care. This second volume includes 113 chapters in five sections: liver, gallbladder and biliary tract, pancreas, pediatric gastroenterology, and special topics. Specific topics include hepatic (liver) structure and function, jaundice, viral hepatitis, alcoholic liver injury, liver tumors, parasitic diseases of the liver, Wilson's disease, hemochromatosis, the pregnancy patient with liver disease, portal hypertension, hepatic encephalopathy, fulminant hepatic failure, liver transplantation, the anatomy of the gallbladder and biliary tract, gallstones, laparoscopic

cholecystectomy (gallbladder removal), cholecystitis (gallbladder infection), primary sclerosing cholangitis, biliary obstruction, pancreatic anatomy and physiology, acute pancreatitis, pancreatic fistulas and ascites (fluid accumulation), chronic pancreatitis, cancer of the pancreas, endoscopic retrograde cholangiopancreatography, esophageal atresia, gastroesophageal reflux in infants and children, achalasia and esophageal motility disorders, caustic and foreign body ingestion, vomiting, chronic abdominal pain, gastritis and peptic ulcer disease in children, malabsorption syndromes in children, inflammatory bowel disease in children and adolescents, acute appendicitis, cystic fibrosis, constipation and fecal soiling (incontinence), hepatitis in children, liver transplantation in children, failure to thrive, pediatric AIDS, the gastrointestinal manifestations of AIDS, the evaluation and management of acute upper gastrointestinal bleeding, principles of endoscopy, eating disorders, nutritional assessment, enteral and parenteral nutrition, gastrointestinal diseases in the elderly and in pregnancy, nosocomial infections, and the psychosocial aspects of gastroenterology (doctor patient interactions). The chapters include figures, algorithms, charts, graphs, radiographs, endoscopic pictures, intraoperative photographs, photomicrographs, tables, and extensive references. The volume concludes with a detailed subject index and a section of color plates.

- **Evidence Based Gastroenterology and Hepatology**

  Source: London, UK: BMJ Publishing Group. 1999. 557 p.

  Contact: Available from BMJ Publishing Group. BMA Books, BMA House, Tavistock Square, London WCIH 9JR. Fax 44 (0)20 7383 6402. E-mail: orders@bmjbooks.com. Website: www.bmjbooks.com. PRICE: Contact publisher for price. ISBN: 0727911821.

  Summary: This book emphasizes the approaches of evidence based medicine in gastroenterology (the study of the gastrointestinal tract and gastrointestinal diseases) and hepatology (the study of the liver and liver diseases). The authors use clinical epidemiology to present the strongest and most current evidence for interventions for the major diseases of the gastrointestinal tract and liver. Thirty chapters are included: an introduction to evidence based gastroenterology and hepatology; gastroesophageal reflux disease (GERD); ulcer disease and Helicobacter pylori; ulcer disease and nonsteroidal antiinflammatory drugs; treatment options for non-variceal gastrointestinal hemorrhage; the diagnosis and treatment of functional dyspepsia (indigestion); the diagnosis, treatment, and prognosis of celiac disease (gluten intolerance); the treatment of Crohn's disease; the diagnosis, prognosis, and treatment of ulcerative colitis (UC); pouchitis after restorative proctocolectomy; metabolic bone

disease in gastrointestinal disorders; colorectal cancer in UC and the role of surveillance; population based screening and surveillance for colorectal cancer; irritable bowel syndrome (IBS); the surgical treatment of gallstone disease; the prognosis and treatment of acute pancreatitis; hepatitis C; hepatitis B; the screening and treatment of alcoholic liver disease; hemochromatosis and Wilson disease; primary biliary cirrhosis (PBC); autoimmune hepatitis; primary sclerosing cholangitis (PSC); the prevention and treatment of portal hypertensive bleeding; ascites, hepatorenal syndrome, and spontaneous bacterial peritonitis; hepatic encephalopathy; hepatocellular carcinoma; fulminant hepatic failure; the prevention and treatment of rejection after liver transplantation; and the prevention and treatment of infection after liver transplantation. Each chapter features the grading of recommendations and levels of evidence used by the authors to note the research basis on which their clinical guidelines are formed. Chapters conclude with extensive reference lists; the text concludes with a subject index. A glossary of acronyms is also provided.

- **Autoimmune Liver Diseases. 2nd ed**

  Source: New York, NY: Elsevier Science, Inc. 1998. 656 p.

  Contact: Available from Elsevier Science, Inc. P.O. Box 945, Madison Square Station, New York, NY 10160-0757. (888) 437-4636 or (212) 633-3730. Fax (212) 633-3680. E-mail: usinfo-f@elsevier.com. PRICE: $284.50. ISBN: 0444828036.

  Summary: This book is devoted to summarizing the current knowledge of autoimmune liver diseases, focusing on potential pathogenic mechanisms and potential therapies that may prove to be of benefit in treating these diseases. Thirty-four chapters, each written by experts in the field, cover topics including: the concept of autoimmunity and autoimmune disease; the pathogenesis of autoimmune hepatitis; the pathogenesis of primary biliary cirrhosis; immunogenetic studies; the histopathology of autoimmune hepatitis, primary biliary cirrhosis, and primary sclerosing cholangitis; current medical therapies of primary biliary cirrhosis and primary sclerosing cholangitis; drug-induced autoimmune liver disease; and autoimmune manifestations of alcoholic liver disease. Each chapter includes extensive references and a detailed subject index concludes the volume.

- **Handbook of Liver Disease**

  Source: Philadelphia, PA: Churchill-Livingstone. 1998. 534 p.

Contact: Available from W.B. Saunders Company. Book Order Fulfillment Department, 6277 Sea Harbor Drive, Orlando, FL 32887-4430. (800) 545-2522. Fax (800) 874-6418. E-mail: wbsbcs@harcourtbrace.com. PRICE: $73.00 plus shipping and handling. ISBN: 0443055203.

Summary: This comprehensive handbook in outline format offers easy access to information on the full range of liver disorders, and covers symptoms, signs, differential diagnoses, and treatments. A total of 34 chapters cover the following topics: assessment of liver function and diagnostic studies, acute liver failure, chronic viral hepatitis, acute viral hepatitis, autoimmune hepatitis, alcoholic liver disease, fatty liver and nonalcoholic steatohepatitis, drug induced and toxic liver disease, cirrhosis and portal hypertension, portal hypertension and gastrointestinal bleeding, ascites and spontaneous bacterial peritonitis, hepatorenal syndrome, hepatic encephalopathy, primary biliary cirrhosis, primary sclerosing cholangitis, hemochromatosis, Wilson's disease and related disorders, alpha 1 antitrypsin deficiency and other metabolic liver diseases, Budd Chiari syndrome and other vascular disorders, the liver in heart failure, the liver in pregnancy, the liver in systemic disease, pediatric liver disease, liver disease in the elderly, HIV and the liver, granulomatous liver disease, hepatic tumors, hepatic abscesses and cysts, other infections involving the liver, surgery in the patient with liver disease and postoperative jaundice, liver transplantation, cholelithiasis and cholecystitis, diseases of the bile ducts, and tumors of the biliary tract. The book features lists that summarize key information and numerous figures and tables on topics such as acetaminophen toxicity, classifications of chronic hepatitis, and indications for liver transplantation. Each chapter was written by an acknowledged expert in the field and includes references for additional study. A subject index concludes the volume.

- **Chronic Hepatitis**

Source: Disease-a-Month, Masters in Medicine. 39(2): 53-126. February 1993.

Contact: Available from Mosby-Year Book, Inc. 11830 Westline Industrial Drive, St. Louis, MO 63146-3318. (800) 325-4177. Fax (800) 535-9935. PRICE: $15 (as of 1995); bulk prices available.

Summary: This monograph presents an overview of chronic hepatitis. Topics covered include a classification of chronic hepatitis based on histologic manifestations; historical perspectives; differential diagnosis; disorders that may simulate chronic hepatitis, including primary biliary cirrhosis, primary sclerosing cholangitis, alcohol induced liver disease, nonalcoholic steatohepatitis, and alpha1 antitrypsin deficiency; chronic

hepatitis B and C; idiopathic autoimmune chronic active hepatitis; chronic active hepatitis from therapeutic drugs; and Wilson's disease (hepatolenticular degeneration). The author focuses on evaluation, management, and treatment. He concludes that advances in diagnosis, increased understanding of the courses of the major disorders, and availability of effective treatment has heightened interest in chronic hepatitis. 7 figures. 8 tables. 223 references.

Book Summaries: Online Booksellers

Commercial Internet-based booksellers, such as Amazon.com and Barnes & Noble.com, offer summaries which have been supplied by each title's publisher. Some summaries also include customer reviews. Your local bookseller may have access to in-house and commercial databases that index all published books (e.g. Books in Print®). The following have been recently listed with online booksellers as relating to primary sclerosing cholangitis (sorted alphabetically by title; follow the hyperlink to view more details at Amazon.com):

- **Primary Sclerosing Cholangitis** by M.P. Manns (Editor), R.W. Chapman (Editor), A. Stiehl (Editor), R. Wiesner; http://www.amazon.com/exec/obidos/ASIN/b>Publishe/icongroupinterna

The National Library of Medicine Book Index

The National Library of Medicine at the National Institutes of Health has a massive database of books published on healthcare and biomedicine. Go to the following Internet site, **http://locatorplus.gov/**, and then select "Search LOCATORplus." Once you are in the search area, simply type "primary sclerosing cholangitis" (or synonyms) into the search box, and select "books only." From there, results can be sorted by publication date, author, or relevance. The following was recently catalogued by the National Library of Medicine:[25]

[25] In addition to LOCATORPlus, in collaboration with authors and publishers, the National Center for Biotechnology Information (NCBI) is adapting biomedical books for the Web. The books may be accessed in two ways: (1) by searching directly using any search term or phrase (in the same way as the bibliographic database PubMed), or (2) by following the links to PubMed abstracts. Each PubMed abstract has a "Books" button that displays a facsimile of the abstract in which some phrases are hypertext links. These phrases are also found in the books available at NCBI. Click on hyperlinked results in the list of books in which the phrase is found. Currently, the majority of the links are between the books and

- **Autoimmune liver disease: its recent advances: proceedings of the International Symposium of Digestive Diseases Week held in Hiroshima on 29-30, October 1999.** Author: editors, Mikio Nishioka, Seishiro Watanabe, and Keiji Arima; Year: 2000; Amsterdam; New York: Elsevier, 2000; ISBN: 044450527X (alk. paper) http://www.amazon.com/exec/obidos/ASIN/044450527X/icongroupinterna
- **Effect of a sclerosing agent on the process of odontogenesis.** Author: Mehlisch, Donald Robert; Year: 1970; [Minneapolis] 1970
- **Manual of sclerotherapy.** Author: Neil S. Sadick; Year: 2000; Philadelphia: Lippincott Williams & Wilkins, c2000; ISBN: 0397517424 http://www.amazon.com/exec/obidos/ASIN/0397517424/icongroupinterna

Chapters on Primary Sclerosing Cholangitis

Frequently, primary sclerosing cholangitis will be discussed within a book, perhaps within a specific chapter. In order to find chapters that are specifically dealing with primary sclerosing cholangitis, an excellent source of abstracts is the Combined Health Information Database. You will need to limit your search to book chapters and primary sclerosing cholangitis using the "Detailed Search" option. Go directly to the following hyperlink: **http://chid.nih.gov/detail/detail.html**. To find book chapters, use the drop boxes at the bottom of the search page where "You may refine your search by." Select the dates and language you prefer, and the format option "Book Chapter." By making these selections and typing in "primary sclerosing cholangitis" (or synonyms) into the "For these words:" box, you will only receive results on chapters in books. The following is a typical result when searching for book chapters on primary sclerosing cholangitis:

- **Stricture Management in Primary Sclerosing Cholangitis**

  Source: in Bayless, T.M. and Hanauer, S.B. Advanced Therapy of Inflammatory Bowel Disease. Hamilton, Ontario: B.C. Decker Inc. 2001. p. 303-304.

  Contact: Available from B.C. Decker Inc. 20 Hughson Street South, P.O. Box 620, L.C.D. 1 Hamilton, Ontario L8N 3K7. (905) 522-7017 or (800) 568-

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PubMed. In the future, more links will be created between the books and other types of information, such as gene and protein sequences and macromolecular structures. See **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Books.**

7281. Fax (905) 522-7839. Email: info@bcdecker.com. Website: www.bcdecker.com. PRICE: $129.00 plus shipping and handling. ISBN: 1550091220.

Summary: This chapter on stricture management in primary sclerosing cholangitis is from the second edition of a book devoted to the details of medical, surgical, and supportive management of patients with Crohn's disease (CD) and Ulcerative Colitis (UC), together known as inflammatory bowel disease (IBD). Sclerosing cholangitis is a cholestatic liver disorder characterized by diffuse strictures (scar tissue causing a narrowing) of the bile ducts. The disorder may be primary (idiopathic) or secondary due to structural abnormalities of the bile ducts. Primary sclerosing cholangitis (PSC) is clinically indistinguishable from disorders that cause secondary sclerosing cholangitis. There is a strong association of PSC and IBD, particularly ulcerative colitis. Approximately 4 percent of patients with IBD will either have or develop PSC. The mainstay of treatment of PSC is liver transplantation with survival rates of greater than 80 percent at 5 years posttransplant. The author cautions that the use of other palliative treatment options such as endoscopic, nontransplant surgical or radiologic interventions should be evaluated in the context of the effectiveness of liver transplantation. The concern of underlying cholangiocarcionoma (biliary tract cancer) should be a priority when evaluating and treating these strictures. 11 references.

- **Pancreatitis in Inflammatory Bowel Disease**

  Source: in Bayless, T.M. and Hanauer, S.B. Advanced Therapy of Inflammatory Bowel Disease. Hamilton, Ontario: B.C. Decker Inc. 2001. p. 329-332.

  Contact: Available from B.C. Decker Inc. 20 Hughson Street South, P.O. Box 620, L.C.D. 1 Hamilton, Ontario L8N 3K7. (905) 522-7017 or (800) 568-7281. Fax (905) 522-7839. Email: info@bcdecker.com. Website: www.bcdecker.com. PRICE: $129.00 plus shipping and handling. ISBN: 1550091220.

  Summary: This chapter on pancreatitis is from the second edition of a book devoted to the details of medical, surgical, and supportive management of patients with Crohn's disease (CD) and Ulcerative Colitis (UC), together known as inflammatory bowel disease (IBD). There is a higher incidence and prevalence of pancreatitis in patients with inflammatory bowel disease (IBD) than in the general population. The pancreatitis can be acute or chronic, or subclinical or overt, and has many causes. The most common cause is medications used to treat IBD, especially azathioprine and 6 mercaptopurine. Other causes of pancreatitis include duodenal involvement from Crohn's disease (CD),

gallstones (cholelithiasis), and primary sclerosing cholangitis (PSC). Pancreatitis also can be caused by high serum concentrations of triglycerides during total parenteral nutritional (TPN) therapy for CD, and may also be a primary extra-intestinal manifestation of IBD. Treatment is different for each cause. For drug-induced pancreatitis, discontinuation of the drug should improvethe pancreatitis. For TPN-induced pancreatitis, oral medium-chain triglycerides should be substituted for the lipid emulsion. For pancreatitis that has developed from gallstones, the usual treatment is laparoscopic cholecystectomy (removal of the gallbladder). Idiopathic (of unknown cause) pancreatitis is often successfully treated by treating the underlying IBD. 1 table. 10 references.

- **Liver Transplantation for Primary Sclerosing Cholangitis**

  Source: in Bayless, T.M. and Hanauer, S.B. Advanced Therapy of Inflammatory Bowel Disease. Hamilton, Ontario: B.C. Decker Inc. 2001. p. 305-310.

  Contact: Available from B.C. Decker Inc. 20 Hughson Street South, P.O. Box 620, L.C.D. 1 Hamilton, Ontario L8N 3K7. (905) 522-7017 or (800) 568-7281. Fax (905) 522-7839. Email: info@bcdecker.com. Website: www.bcdecker.com. PRICE: $129.00 plus shipping and handling. ISBN: 1550091220.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic, usually progressive, liver disease characterized by intrahepatic or extrahepatic (inside or outside the liver, respectively) duct stricturing (narrowing) and dilatation ('beaded' appearance). This chapter on liver transplantation for PSC is from the second edition of a book devoted to the details of medical, surgical, and supportive management of patients with Crohn's disease (CD) and ulcerative colitis (UC), together known as inflammatory bowel disease (IBD). About 70 percent of the patients with PSC have IBD, with UC being more common than CD (by a ratio of 9 to 1). Dominant, symptomatic strictures can be managed endoscopically. Reconstructive biliary surgery has no role in the management of PSC. Liver transplantation is the treatment of choice. Five-year survival after liver transplantation is approximately 80 to 90 percent. Actuarial patient survival after liver transplantation is significantly better than the survival rate of patients treated with nontransplantation biliary surgery or the predicted survival from prognostic models. Ten to 15 percent of patients with PSC develop cholangiocarcinoma (biliary tract cancer), which is difficult to diagnose and rarely curable; however, in patients with small incidental cholangiocarcinomas, recognized only at liver exploration (in the absence of lymph node spread), the prognosis is more encouraging. It

is recommended that patients with PSC are transplanted earlier than patients with other liver diseases. The high risk and poor prognosis for cholangiocarcinoma in longstanding PSC are an argument in favor of performing liver transplantation in an earlier phase of the disease. Colonoscopic surveillance for dysplasia (abnormal tissue) is essential. 2 figures. 1 table. 21 references.

- **Primary Sclerosing Cholangitis**

  Source: in Friedman, L.S. and Keeffe, E.B., eds. Handbook of Liver Disease. Philadelphia, PA: Churchill-Livingstone. 1998. p. 215-225.

  Contact: Available from W.B. Saunders Company. Book Order Fulfillment Department, 6277 Sea Harbor Drive, Orlando, FL 32887-4430. (800) 545-2522. Fax (800) 874-6418. E-mail: wbsbcs@harcourtbrace.com. PRICE: $73.00 plus shipping and handling. ISBN: 0443055203.

  Summary: Primary sclerosing cholangitis (PSC) is a chronic cholestatic disease that occurs most commonly in middle aged males and is frequently found in association with inflammatory bowel disease (IBD), most commonly ulcerative colitis (UC). This chapter on PSC is from a comprehensive handbook in outline format that offers easy access to information on the full range of liver disorders and covers symptoms, signs, differential diagnoses, and treatments. The diagnosis of PSC is based on clinical, biochemical, and most important, cholangiography findings. The exclusion of identifiable causes of secondary sclerosing cholangitis is important. Diagnosis is often made on the basis of a cholestatic biochemical profile found in a patient with long standing UC. The gradual onset of progressive fatigue and pruritus followed by jaundice represents the most frequent symptom complex leading to the diagnosis. The etiology of PSC remains unknown, but evolving evidence points to autoimmune mechanisms. PSC is usually slowly progressive, leading to significant complications, some of which are specific to the syndrome, such as formation of dominant biliary strictures, choledocholithiasis (common bile duct stones), and cholangiocarcinoma. Several models that can be used to estimate survival for the individual patient have been formulated. None of the medical approaches (drug therapies) used to date have been shown to have a major impact on prolonging survival or preventing complications. Surgical therapy includes biliary tract reconstruction and proctocolectomy for UC. Neither of these has been shown to halt the progression of PSC, however. The results of liver transplantation have been quite good, with 5 year survival rates of 75 to 80 percent. Recurrence of PSC after liver transplantation has been described, but appears to be infrequent and often clinically insignificant. 1 figure. 4 tables. 6 references.

- **Hepatobiliary Complications of Ulcerative Colitis and Crohn's Disease**

  Source: in Snape, W.J., ed. Consultations in Gastroenterology. Philadelphia, PA: W.B. Saunders Company. 1996. p. 741-749.

  Contact: Available from W.B. Saunders Company. Order Fulfillment, 6277 Sea Harbor Drive, Orlando, FL 32887. (800) 545-2522. Fax (800) 874-6418 or (407) 352-3445. PRICE: $125.00. ISBN: 0721646700.

  Summary: This chapter, from a gastroenterology yearbook, covers the hepatobiliary complications of ulcerative colitis (UC) and Crohn's disease. Although hepatobiliary abnormalities occur frequently in patients with inflammatory bowel disease (IBD), there are only three associated conditions of major clinical importance: primary sclerosing cholangitis (PSC), autoimmune chronic active hepatitis, and cholelithiasis (gallstones). The authors comment briefly on the latter two diseases, but concentrate primarily on PSC. PSC is a chronic, progressive, idiopathic, cholestatic liver disease that principally affects young men and is characterized by diffuse inflammation and fibrosis of the entire biliary tree. The natural history of PSC usually is one of slow progression with eventual development of cirrhosis, portal hypertension with its accompanying complications, and death from liver failure unless liver transplantation is performed. The authors discuss the etiology, clinical features, diagnosis, natural history and course, relationship to IBD, differential diagnosis, treatment, and complications of PSC. The authors note that patients with PSC and UC are doubly at risk for malignancies of the colon and biliary system. Medical therapies that may beneficially affect both PSC and UC are being assessed, and liver transplantation is life saving for patients with advanced PSC. 2 figures. 3 tables. 15 references. (AA-M).

General Home References

In addition to references for primary sclerosing cholangitis, you may want a general home medical guide that spans all aspects of home healthcare. The following list is a recent sample of such guides (sorted alphabetically by title; hyperlinks provide rankings, information, and reviews at Amazon.com):

- **The Digestive System (21st Century Health and Wellness)** by Regina Avraham; Library Binding (February 2000), Chelsea House Publishing (Library); ISBN: 0791055264; **http://www.amazon.com/exec/obidos/ASIN/0791055264/icongroupinterna**
- **American College of Physicians Complete Home Medical Guide (with Interactive Human Anatomy CD-ROM)** by David R. Goldmann (Editor),

American College of Physicians; Hardcover - 1104 pages, Book & CD-Rom edition (1999), DK Publishing; ISBN: 0789444127; **http://www.amazon.com/exec/obidos/ASIN/0789444127/icongroupinterna**

- **The American Medical Association Guide to Home Caregiving** by the American Medical Association (Editor); Paperback - 256 pages 1 edition (2001), John Wiley & Sons; ISBN: 0471414093; **http://www.amazon.com/exec/obidos/ASIN/0471414093/icongroupinterna**
- **Anatomica : The Complete Home Medical Reference** by Peter Forrestal (Editor); Hardcover (2000), Book Sales; ISBN: 1740480309; **http://www.amazon.com/exec/obidos/ASIN/1740480309/icongroupinterna**
- **The HarperCollins Illustrated Medical Dictionary : The Complete Home Medical Dictionary** by Ida G. Dox, et al; Paperback - 656 pages 4th edition (2001), Harper Resource; ISBN: 0062736469; **http://www.amazon.com/exec/obidos/ASIN/0062736469/icongroupinterna**
- **Mayo Clinic Guide to Self-Care: Answers for Everyday Health Problems** by Philip Hagen, M.D. (Editor), et al; Paperback - 279 pages, 2nd edition (December 15, 1999), Kensington Publishing Corp.; ISBN: 0962786578; **http://www.amazon.com/exec/obidos/ASIN/0962786578/icongroupinterna**
- **The Merck Manual of Medical Information : Home Edition (Merck Manual of Medical Information Home Edition (Trade Paper)** by Robert Berkow (Editor), Mark H. Beers, M.D. (Editor); Paperback - 1536 pages (2000), Pocket Books; ISBN: 0671027263; **http://www.amazon.com/exec/obidos/ASIN/0671027263/icongroupinterna**

Vocabulary Builder

Acetaminophen: Analgesic antipyretic derivative of acetanilide. It has weak anti-inflammatory properties and is used as a common analgesic, but may cause liver, blood cell, and kidney damage. [NIH]

Anaemia: A reduction below normal in the number of erythrocytes per cu. mm., in the quantity of haemoglobin, or in the volume of packed red cells per 100 ml. of blood which occurs when the equilibrium between blood loss (through bleeding or destruction) and blood production is disturbed. [EU]

Appendicitis: Acute inflammation of the vermiform appendix. [NIH]

Caustic: An escharotic or corrosive agent. Called also cauterant. [EU]

Cholecystectomy: Surgical removal of the gallbladder. [NIH]

Cholecystitis: Inflammation of the gallbladder. [EU]

Cholelithiasis: The presence or formation of gallstones. [EU]

Cholesterol: The principal sterol of all higher animals, distributed in body tissues, especially the brain and spinal cord, and in animal fats and oils. [NIH]

Colic: Paroxysms of pain. This condition usually occurs in the abdominal region but may occur in other body regions as well. [NIH]

Constipation: Infrequent or difficult evacuation of the faeces. [EU]

Crystallization: The formation of crystals; conversion to a crystalline form. [EU]

Dyspepsia: Impairment of the power of function of digestion; usually applied to epigastric discomfort following meals. [EU]

Encephalopathy: Any degenerative disease of the brain. [EU]

Endoscopy: Visual inspection of any cavity of the body by means of an endoscope. [EU]

Epidemic: Occurring suddenly in numbers clearly in excess of normal expectancy; said especially of infectious diseases but applied also to any disease, injury, or other health-related event occurring in such outbreaks. [EU]

Fistula: An abnormal passage or communication, usually between two internal organs, or leading from an internal organ to the surface of the body; frequently designated according to the organs or parts with which it communicates, as anovaginal, brochocutaneous, hepatopleural, pulmonoperitoneal, rectovaginal, urethrovaginal, and the like. Such passages are frequently created experimentally for the purpose of obtaining body secretions for physiologic study. [EU]

Gastritis: Inflammation of the stomach. [EU]

Gluten: The protein of wheat and other grains which gives to the dough its tough elastic character. [EU]

Hemorrhage: Bleeding or escape of blood from a vessel. [NIH]

Hemorrhoids: Varicosities of the hemorrhoidal venous plexuses. [NIH]

Hernia: (he protrusion of a loop or knuckle of an organ or tissue through an abnormal opening. [EU]

Idiopathic: Of the nature of an idiopathy; self-originated; of unknown causation. [EU]

Incidental: 1. small and relatively unimportant, minor; 2. accompanying, but not a major part of something; 3. (to something) liable to occur because of something or in connection with something (said of risks, responsibilities, ...) [EU]

Incontinence: Inability to control excretory functions, as defecation (faecal i.) or urination (urinary i.). [EU]

Ingestion: The act of taking food, medicines, etc., into the body, by mouth. [EU]

Lipid: Any of a heterogeneous group of flats and fatlike substances characterized by being water-insoluble and being extractable by nonpolar (or fat) solvents such as alcohol, ether, chloroform, benzene, etc. All contain as a major constituent aliphatic hydrocarbons. The lipids, which are easily stored in the body, serve as a source of fuel, are an important constituent of cell structure, and serve other biological functions. Lipids may be considered to include fatty acids, neutral fats, waxes, and steroids. Compound lipids comprise the glycolipids, lipoproteins, and phospholipids. [EU]

Lipodystrophy: 1. any disturbance of fat metabolism. 2. a group of conditions due to defective metabolism of fat, resulting in the absence of subcutaneous fat, which may be congenital or acquired and partial or total. Called also lipoatrophy and lipodystrophia. [EU]

Lithotripsy: The destruction of a calculus of the kidney, ureter, bladder, or gallbladder by physical forces, including crushing with a lithotriptor through a catheter. Focused percutaneous ultrasound and focused hydraulic shock waves may be used without surgery. Lithotripsy does not include the dissolving of stones by acids or litholysis. Lithotripsy by laser is lithotripsy, laser. [NIH]

Malabsorption: Impaired intestinal absorption of nutrients. [EU]

Motility: The ability to move spontaneously. [EU]

Nosocomial: Pertaining to or originating in the hospital, said of an infection not present or incubating prior to admittance to the hospital, but generally occurring 72 hours after admittance; the term is usually used to refer to patient disease, but hospital personnel may also acquire nosocomial infection. [EU]

Palliative: 1. affording relief, but not cure. 2. an alleviating medicine. [EU]

Pancreas: An organ behind the lower part of the stomach that is about the size of a hand. It makes insulin so that the body can use glucose (sugar) for energy. It also makes enzymes that help the body digest food. Spread all over the pancreas are areas called the islets of Langerhans. The cells in these areas each have a special purpose. The alpha cells make glucagon, which raises the level of glucose in the blood; the beta cells make insulin; the delta cells make somatostatin. There are also the PP cells and the D1 cells, about which little is known. [NIH]

Pancreatitis: Inflammation (pain, tenderness) of the pancreas; it can make the pancreas stop working. It is caused by drinking too much alcohol, by disease in the gallbladder, or by a virus. [NIH]

Parasitic: Pertaining to, of the nature of, or caused by a parasite. [EU]

Parenteral: Not through the alimentary canal but rather by injection through some other route, as subcutaneous, intramuscular, intraorbital, intracapsular,

intraspinal, intrasternal, intravenous, etc. [EU]

Peptic: Pertaining to pepsin or to digestion; related to the action of gastric juices. [EU]

Perinatal: Pertaining to or occurring in the period shortly before and after birth; variously defined as beginning with completion of the twentieth to twenty-eighth week of gestation and ending 7 to 28 days after birth. [EU]

Peritonitis: Inflammation of the peritoneum; a condition marked by exudations in the peritoneum of serum, fibrin, cells, and pus. It is attended by abdominal pain and tenderness, constipation, vomiting, and moderate fever. [EU]

Postoperative: Occurring after a surgical operation. [EU]

Reflux: A backward or return flow. [EU]

Sclerotherapy: Treatment of varicose veins, hemorrhoids, gastric and esophageal varices, and peptic ulcer hemorrhage by injection or infusion of chemical agents which cause localized thrombosis and eventual fibrosis and obliteration of the vessels. [NIH]

Subclinical: Without clinical manifestations; said of the early stage(s) of an infection or other disease or abnormality before symptoms and signs become apparent or detectable by clinical examination or laboratory tests, or of a very mild form of an infection or other disease or abnormality. [EU]

Systemic: Pertaining to or affecting the body as a whole. [EU]

Toxicity: The quality of being poisonous, especially the degree of virulence of a toxic microbe or of a poison. [EU]

Vascular: Pertaining to blood vessels or indicative of a copious blood supply. [EU]

CHAPTER 7. MULTIMEDIA ON PRIMARY SCLEROSING CHOLANGITIS

Overview

Information on primary sclerosing cholangitis can come in a variety of formats. Among multimedia sources, video productions, slides, audiotapes, and computer databases are often available. In this chapter, we show you how to keep current on multimedia sources of information on primary sclerosing cholangitis. We start with sources that have been summarized by federal agencies, and then show you how to find bibliographic information catalogued by the National Library of Medicine. If you see an interesting item, visit your local medical library to check on the availability of the title.

Bibliography: Multimedia on Primary Sclerosing Cholangitis

The National Library of Medicine is a rich source of information on healthcare-related multimedia productions including slides, computer software, and databases. To access the multimedia database, go to the following Web site: **http://locatorplus.gov/**. Select "Search LOCATORplus." Once in the search area, simply type in primary sclerosing cholangitis (or synonyms). Then, in the option box provided below the search box, select "Audiovisuals and Computer Files." From there, you can choose to sort results by publication date, author, or relevance. The following multimedia has been indexed on primary sclerosing cholangitis. For more information, follow the hyperlink indicated:

- **Cholangioscopic differentiation of various bile duct lesions.** Source: Dong Wan Seo ... [et al.]; Year: 1999; Format: Videorecording; Timonium, MD: Milner-Fenwick [distributor], [1999?]

- **Surgical treatment of sclerosing cholangitis.** Source: author, Henry A. Pitt; produced by Davis & Geck, Medical Device Division; Year: 1987; Format: Videorecording; Danbury, Conn.: American Cyanamid, c1987

Vocabulary Builder

Orthopaedic: Pertaining to the correction of deformities of the musculoskeletal system; pertaining to orthopaedics. [EU]

Prosthesis: A man-made substitute for a missing body part such as an arm or a leg; also an implant such as for the hip. [NIH]

CHAPTER 8. PHYSICIAN GUIDELINES AND DATABASES

Overview

Doctors and medical researchers rely on a number of information sources to help patients with their conditions. Many will subscribe to journals or newsletters published by their professional associations or refer to specialized textbooks or clinical guides published for the medical profession. In this chapter, we focus on databases and Internet-based guidelines created or written for this professional audience.

NIH Guidelines

For the more common diseases, The National Institutes of Health publish guidelines that are frequently consulted by physicians. Publications are typically written by one or more of the various NIH Institutes. For physician guidelines, commonly referred to as "clinical" or "professional" guidelines, you can visit the following Institutes:

- Office of the Director (OD); guidelines consolidated across agencies available at **http://www.nih.gov/health/consumer/conkey.htm**
- National Institute of General Medical Sciences (NIGMS); fact sheets available at **http://www.nigms.nih.gov/news/facts/**
- National Library of Medicine (NLM); extensive encyclopedia (A.D.A.M., Inc.) with guidelines: **http://www.nlm.nih.gov/medlineplus/healthtopics.html**
- National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK); guidelines available at **http://www.niddk.nih.gov/health/health.htm**

NIH Databases

In addition to the various Institutes of Health that publish professional guidelines, the NIH has designed a number of databases for professionals.[26] Physician-oriented resources provide a wide variety of information related to the biomedical and health sciences, both past and present. The format of these resources varies. Searchable databases, bibliographic citations, full text articles (when available), archival collections, and images are all available. The following are referenced by the National Library of Medicine:[27]

- **Bioethics:** Access to published literature on the ethical, legal and public policy issues surrounding healthcare and biomedical research. This information is provided in conjunction with the Kennedy Institute of Ethics located at Georgetown University, Washington, D.C.: **http://www.nlm.nih.gov/databases/databases_bioethics.html**
- **HIV/AIDS Resources:** Describes various links and databases dedicated to HIV/AIDS research: **http://www.nlm.nih.gov/pubs/factsheets/aidsinfs.html**
- **NLM Online Exhibitions:** Describes "Exhibitions in the History of Medicine": **http://www.nlm.nih.gov/exhibition/exhibition.html**. Additional resources for historical scholarship in medicine: **http://www.nlm.nih.gov/hmd/hmd.html**
- **Biotechnology Information:** Access to public databases. The National Center for Biotechnology Information conducts research in computational biology, develops software tools for analyzing genome data, and disseminates biomedical information for the better understanding of molecular processes affecting human health and disease: **http://www.ncbi.nlm.nih.gov/**
- **Population Information:** The National Library of Medicine provides access to worldwide coverage of population, family planning, and related health issues, including family planning technology and programs, fertility, and population law and policy: **http://www.nlm.nih.gov/databases/databases_population.html**
- **Cancer Information:** Access to caner-oriented databases: **http://www.nlm.nih.gov/databases/databases_cancer.html**

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[26] Remember, for the general public, the National Library of Medicine recommends the databases referenced in MEDLINE*plus* (**http://medlineplus.gov/** or **http://www.nlm.nih.gov/medlineplus/databases.html**).

[27] See **http://www.nlm.nih.gov/databases/databases.html**.

- **Profiles in Science:** Offering the archival collections of prominent twentieth-century biomedical scientists to the public through modern digital technology: **http://www.profiles.nlm.nih.gov/**
- **Chemical Information:** Provides links to various chemical databases and references: **http://sis.nlm.nih.gov/Chem/ChemMain.html**
- **Clinical Alerts:** Reports the release of findings from the NIH-funded clinical trials where such release could significantly affect morbidity and mortality: **http://www.nlm.nih.gov/databases/alerts/clinical_alerts.html**
- **Space Life Sciences:** Provides links and information to space-based research (including NASA): **http://www.nlm.nih.gov/databases/databases_space.html**
- **MEDLINE:** Bibliographic database covering the fields of medicine, nursing, dentistry, veterinary medicine, the healthcare system, and the pre-clinical sciences: **http://www.nlm.nih.gov/databases/databases_medline.html**
- **Toxicology and Environmental Health Information (TOXNET):** Databases covering toxicology and environmental health: **http://sis.nlm.nih.gov/Tox/ToxMain.html**
- **Visible Human Interface:** Anatomically detailed, three-dimensional representations of normal male and female human bodies: **http://www.nlm.nih.gov/research/visible/visible_human.html**

While all of the above references may be of interest to physicians who study and treat primary sclerosing cholangitis, the following are particularly noteworthy.

The Combined Health Information Database

A comprehensive source of information on clinical guidelines written for professionals is the Combined Health Information Database. You will need to limit your search to "Brochure/Pamphlet," "Fact Sheet," or "Information Package" and primary sclerosing cholangitis using the "Detailed Search" option. Go directly to the following hyperlink: **http://chid.nih.gov/detail/detail.html**. To find associations, use the drop boxes at the bottom of the search page where "You may refine your search by." For the publication date, select "All Years," select your preferred language, and the format option "Fact Sheet." By making these selections and typing "primary sclerosing cholangitis" (or synonyms) into the "For

these words:" box above, you will only receive results on fact sheets dealing with primary sclerosing cholangitis. The following is a sample result:

- **Terminology of Chronic Hepatitis: International Working Party Report**

  Source: American Journal of Gastroenterology. 90(2): 181-189. February 1995.

  Summary: In this article, the authors present the proceedings of two panels on chronic hepatitis of the World Congresses of Gastroenterology (WCPG) working party. The authors hope that the suggested terms, definitions, and codes can: be used by clinicians and laboratory physicians as they jointly care for patients; keep the language in hepatology contemporary; and help to create data that lend themselves to improved computerized coding and retrieval in the interest of patient service and research. After a discussion of background and objectives, the authors present definitions, synonyms, and clinical, laboratory, and histological findings for the following conditions: autoimmune hepatitis; chronic hepatitis B; chronic hepatitis D; chronic hepatitis C; chronic drug hepatitis; primary biliary cirrhosis; primary sclerosing cholangitis; Wilson's disease of the liver; and alpha-1-antitrypsin deficiency. One brief section discusses the grading and staging of chronic hepatitis. 2 tables. 28 references.

The NLM Gateway[28]

The NLM (National Library of Medicine) Gateway is a Web-based system that lets users search simultaneously in multiple retrieval systems at the U.S. National Library of Medicine (NLM). It allows users of NLM services to initiate searches from one Web interface, providing "one-stop searching" for many of NLM's information resources or databases.[29] One target audience for the Gateway is the Internet user who is new to NLM's online resources and does not know what information is available or how best to search for it. This audience may include physicians and other healthcare providers, researchers, librarians, students, and, increasingly, patients, their families, and the public.[30] To use the NLM Gateway, simply go to the search site at

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[28] Adapted from NLM: **http://gateway.nlm.nih.gov/gw/Cmd?Overview.x**.

[29] The NLM Gateway is currently being developed by the Lister Hill National Center for Biomedical Communications (LHNCBC) at the National Library of Medicine (NLM) of the National Institutes of Health (NIH).

[30] Other users may find the Gateway useful for an overall search of NLM's information resources. Some searchers may locate what they need immediately, while others will utilize the Gateway as an adjunct tool to other NLM search services such as PubMed® and MEDLINEplus®. The Gateway connects users with multiple NLM retrieval systems while

http://gateway.nlm.nih.gov/gw/Cmd. Type "primary sclerosing cholangitis" (or synonyms) into the search box and click "Search." The results will be presented in a tabular form, indicating the number of references in each database category.

Results Summary

Category	Items Found
Journal Articles	343670
Books / Periodicals / Audio Visual	2561
Consumer Health	292
Meeting Abstracts	3093
Other Collections	100
Total	349716

HSTAT[31]

HSTAT is a free, Web-based resource that provides access to full-text documents used in healthcare decision-making.[32] HSTAT's audience includes healthcare providers, health service researchers, policy makers, insurance companies, consumers, and the information professionals who serve these groups. HSTAT provides access to a wide variety of publications, including clinical practice guidelines, quick-reference guides for clinicians, consumer health brochures, evidence reports and technology assessments from the Agency for Healthcare Research and Quality (AHRQ), as well as AHRQ's Put Prevention Into Practice.[33] Simply search by "primary sclerosing

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also providing a search interface for its own collections. These collections include various types of information that do not logically belong in PubMed, LOCATORplus, or other established NLM retrieval systems (e.g., meeting announcements and pre-1966 journal citations). The Gateway will provide access to the information found in an increasing number of NLM retrieval systems in several phases.

[31] Adapted from HSTAT: **http://www.nlm.nih.gov/pubs/factsheets/hstat.html**.

[32] The HSTAT URL is **http://hstat.nlm.nih.gov/**.

[33] Other important documents in HSTAT include: the National Institutes of Health (NIH) Consensus Conference Reports and Technology Assessment Reports; the HIV/AIDS Treatment Information Service (ATIS) resource documents; the Substance Abuse and Mental Health Services Administration's Center for Substance Abuse Treatment (SAMHSA/CSAT) Treatment Improvement Protocols (TIP) and Center for Substance Abuse Prevention (SAMHSA/CSAP) Prevention Enhancement Protocols System (PEPS); the Public Health Service (PHS) Preventive Services Task Force's *Guide to Clinical Preventive Services*; the independent, nonfederal Task Force on Community Services *Guide to Community Preventive Services*; and the Health Technology Advisory Committee (HTAC) of the Minnesota Health Care Commission (MHCC) health technology evaluations.

cholangitis" (or synonyms) at the following Web site: **http://text.nlm.nih.gov**.

Coffee Break: Tutorials for Biologists[34]

Some patients may wish to have access to a general healthcare site that takes a scientific view of the news and covers recent breakthroughs in biology that may one day assist physicians in developing treatments. To this end, we recommend "Coffee Break," a collection of short reports on recent biological discoveries. Each report incorporates interactive tutorials that demonstrate how bioinformatics tools are used as a part of the research process. Currently, all Coffee Breaks are written by NCBI staff.[35] Each report is about 400 words and is usually based on a discovery reported in one or more articles from recently published, peer-reviewed literature.[36] This site has new articles every few weeks, so it can be considered an online magazine of sorts, and intended for general background information. You can access the Coffee Break Web site at the following hyperlink: **http://www.ncbi.nlm.nih.gov/Coffeebreak/**.

Other Commercial Databases

In addition to resources maintained by official agencies, other databases exist that are commercial ventures addressing medical professionals. Here are a few examples that may interest you:

- **CliniWeb International:** Index and table of contents to selected clinical information on the Internet; see **http://www.ohsu.edu/cliniweb/**.
- **Image Engine:** Multimedia electronic medical record system that integrates a wide range of digitized clinical images with textual data stored in the University of Pittsburgh Medical Center's MARS electronic medical record system; see the following Web site: **http://www.cml.upmc.edu/cml/imageengine/imageEngine.html**.

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[34] Adapted from **http://www.ncbi.nlm.nih.gov/Coffeebreak/Archive/FAQ.html**.

[35] The figure that accompanies each article is frequently supplied by an expert external to NCBI, in which case the source of the figure is cited. The result is an interactive tutorial that tells a biological story.

[36] After a brief introduction that sets the work described into a broader context, the report focuses on how a molecular understanding can provide explanations of observed biology and lead to therapies for diseases. Each vignette is accompanied by a figure and hypertext links that lead to a series of pages that interactively show how NCBI tools and resources are used in the research process.

- **Medical World Search:** Searches full text from thousands of selected medical sites on the Internet; see **http://www.mwsearch.com/**.
- **MedWeaver:** Prototype system that allows users to search differential diagnoses for any list of signs and symptoms, to search medical literature, and to explore relevant Web sites; see **http://www.med.virginia.edu/~wmd4n/medweaver.html**.
- **Metaphrase:** Middleware component intended for use by both caregivers and medical records personnel. It converts the informal language generally used by caregivers into terms from formal, controlled vocabularies; see the following Web site: **http://www.lexical.com/Metaphrase.html**.

The Genome Project and Primary Sclerosing Cholangitis

With all the discussion in the press about the Human Genome Project, it is only natural that physicians, researchers, and patients want to know about how human genes relate to primary sclerosing cholangitis. In the following section, we will discuss databases and references used by physicians and scientists who work in this area.

Online Mendelian Inheritance in Man (OMIM)

The Online Mendelian Inheritance in Man (OMIM) database is a catalog of human genes and genetic disorders authored and edited by Dr. Victor A. McKusick and his colleagues at Johns Hopkins and elsewhere. OMIM was developed for the World Wide Web by the National Center for Biotechnology Information (NCBI).[37] The database contains textual information, pictures, and reference information. It also contains copious links to NCBI's Entrez database of MEDLINE articles and sequence information.

To search the database, go to **http://www.ncbi.nlm.nih.gov/Omim/searchomim.html**. Type "primary sclerosing cholangitis" (or synonyms) in the search box, and click "Submit Search." If too many results appear, you can narrow the search by adding

[37] Adapted from **http://www.ncbi.nlm.nih.gov/**. Established in 1988 as a national resource for molecular biology information, NCBI creates public databases, conducts research in computational biology, develops software tools for analyzing genome data, and disseminates biomedical information--all for the better understanding of molecular processes affecting human health and disease.

the word "clinical." Each report will have additional links to related research and databases. By following these links, especially the link titled "Database Links," you will be exposed to numerous specialized databases that are largely used by the scientific community. These databases are overly technical and seldom used by the general public, but offer an abundance of information. The following is an example of the results you can obtain from the OMIM for primary sclerosing cholangitis:

- **Immune Deficiency Disease**
  Web site: http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispmim?242850
- **Inflammatory Bowel Disease 1; Ibd1**
  Web site: http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispmim?266600
- **Nephropathy, Progressive Tubulointerstitial, with Cholestatic Liver Disease**
  Web site: http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispmim?602114
- **Pancreatitis, Sclerosing Cholangitis, and Sicca Complex**
  Web site: http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispmim?260480

Genes and Disease (NCBI - Map)

The Genes and Disease database is produced by the National Center for Biotechnology Information of the National Library of Medicine at the National Institutes of Health. This Web site categorizes each disorder by the system of the body associated with it. Go to **http://www.ncbi.nlm.nih.gov/disease/**, and browse the system pages to have a full view of important conditions linked to human genes. Since this site is regularly updated, you may wish to re-visit it from time to time. The following systems and associated disorders are addressed:

- **Immune System:** Fights invaders.
  Examples: Asthma, autoimmune polyglandular syndrome, Crohn's disease, DiGeorge syndrome, familial Mediterranean fever, immunodeficiency with Hyper-IgM, severe combined immunodeficiency.
  Web site: **http://www.ncbi.nlm.nih.gov/disease/Immune.html**
- **Metabolism:** Food and energy.
  Examples: Adreno-leukodystrophy, Atherosclerosis, Best disease, Gaucher disease, Glucose galactose malabsorption, Gyrate atrophy,

Juvenile onset diabetes, Obesity, Paroxysmal nocturnal hemoglobinuria, Phenylketonuria, Refsum disease, Tangier disease, Tay-Sachs disease.
Web site: **http://www.ncbi.nlm.nih.gov/disease/Metabolism.html**

- **Muscle and Bone:** Movement and growth.
  Examples: Duchenne muscular dystrophy, Ellis-van Creveld syndrome, Marfan syndrome, myotonic dystrophy, spinal muscular atrophy.
  Web site: **http://www.ncbi.nlm.nih.gov/disease/Muscle.html**
- **Signals:** Cellular messages.
  Examples: Ataxia telangiectasia, Baldness, Cockayne syndrome, Glaucoma, SRY: sex determination, Tuberous sclerosis, Waardenburg syndrome, Werner syndrome.
  Web site: **http://www.ncbi.nlm.nih.gov/disease/Signals.html**
- **Transporters:** Pumps and channels.
  Examples: Cystic Fibrosis, deafness, diastrophic dysplasia, Hemophilia A, long-QT syndrome, Menkes syndrome, Pendred syndrome, polycystic kidney disease, sickle cell anemia, Wilson's disease, Zellweger syndrome.
  Web site: **http://www.ncbi.nlm.nih.gov/disease/Transporters.html**

Entrez

Entrez is a search and retrieval system that integrates several linked databases at the National Center for Biotechnology Information (NCBI). These databases include nucleotide sequences, protein sequences, macromolecular structures, whole genomes, and MEDLINE through PubMed. Entrez provides access to the following databases:

- **PubMed:** Biomedical literature (PubMed),
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=PubMed**
- **Nucleotide Sequence Database (Genbank):**
  Web site:
  http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Nucleotide
- **Protein Sequence Database:**
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Protein**
- **Structure:** Three-dimensional macromolecular structures,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Structure**
- **Genome:** Complete genome assemblies,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Genome**
- **PopSet:** Population study data sets,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Popset**

- **OMIM:** Online Mendelian Inheritance in Man,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=OMIM**
- **Taxonomy:** Organisms in GenBank,
  Web site:
  http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=Taxonomy
- **Books:** Online books,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=books**
- **ProbeSet:** Gene Expression Omnibus (GEO),
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=geo**
- **3D Domains:** Domains from Entrez Structure,
  Web site: **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=geo**
- **NCBI's Protein Sequence Information Survey Results:**
  Web site: **http://www.ncbi.nlm.nih.gov/About/proteinsurvey/**

To access the Entrez system at the National Center for Biotechnology Information, go to **http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?CMD=search&DB=genome**, and then select the database that you would like to search. The databases available are listed in the drop box next to "Search." In the box next to "for," enter "primary sclerosing cholangitis" (or synonyms) and click "Go."

Jablonski's Multiple Congenital Anomaly/Mental Retardation (MCA/MR) Syndromes Database[38]

This online resource can be quite useful. It has been developed to facilitate the identification and differentiation of syndromic entities. Special attention is given to the type of information that is usually limited or completely omitted in existing reference sources due to space limitations of the printed form.

At the following Web site you can also search across syndromes using an alphabetical index: **http://www.nlm.nih.gov/mesh/jablonski/syndrome_toc/toc_a.html.** You can search by keywords at this Web site: **http://www.nlm.nih.gov/mesh/jablonski/syndrome_db.html**.

[38] Adapted from the National Library of Medicine:
http://www.nlm.nih.gov/mesh/jablonski/about_syndrome.html.

The Genome Database[39]

Established at Johns Hopkins University in Baltimore, Maryland in 1990, the Genome Database (GDB) is the official central repository for genomic mapping data resulting from the Human Genome Initiative. In the spring of 1999, the Bioinformatics Supercomputing Centre (BiSC) at the Hospital for Sick Children in Toronto, Ontario assumed the management of GDB. The Human Genome Initiative is a worldwide research effort focusing on structural analysis of human DNA to determine the location and sequence of the estimated 100,000 human genes. In support of this project, GDB stores and curates data generated by researchers worldwide who are engaged in the mapping effort of the Human Genome Project (HGP). GDB's mission is to provide scientists with an encyclopedia of the human genome which is continually revised and updated to reflect the current state of scientific knowledge. Although GDB has historically focused on gene mapping, its focus will broaden as the Genome Project moves from mapping to sequence, and finally, to functional analysis.

To access the GDB, simply go to the following hyperlink: **http://www.gdb.org/**. Search "All Biological Data" by "Keyword." Type "primary sclerosing cholangitis" (or synonyms) into the search box, and review the results. If more than one word is used in the search box, then separate each one with the word "and" or "or" (using "or" might be useful when using synonyms). This database is extremely technical as it was created for specialists. The articles are the results which are the most accessible to non-professionals and often listed under the heading "Citations." The contact names are also accessible to non-professionals.

Specialized References

The following books are specialized references written for professionals interested in primary sclerosing cholangitis (sorted alphabetically by title, hyperlinks provide rankings, information, and reviews at Amazon.com):

- **Blackwell's Primary Care Essentials: Gastointestinal Disease** by David W. Hay; Paperback, 1st edition (December 15, 2001), Blackwell Science Inc; ISBN: 0632045035; **http://www.amazon.com/exec/obidos/ASIN/0632045035/icongroupinterna**
- **Gastrointestinal Problems** by Martin S. Lipsky, M.D. (Editor), Richard Sadovsky, M.D. (Editor); Paperback - 194 pages, 1st edition (August 15,

[39] Adapted from the Genome Database: **http://gdbwww.gdb.org/gdb/aboutGDB.html#mission**.

2000), Lippincott, Williams & Wilkins Publishers; ISBN: 0781720540; **http://www.amazon.com/exec/obidos/ASIN/0781720540/icongroupinterna**

- **Rome II: The Functional Gastrointestinal Disorders** by Douglas A. Drossman (Editor); Paperback - 800 pages, 2nd edition (March 1, 2000), Degnon Associates Inc.; ISBN: 0965683729; **http://www.amazon.com/exec/obidos/ASIN/0965683729/icongroupinterna**

Vocabulary Builder

Nephropathy: Disease of the kidneys. [EU]

CHAPTER 9. DISSERTATIONS ON PRIMARY SCLEROSING CHOLANGITIS

Overview

University researchers are active in studying almost all known diseases. The result of research is often published in the form of Doctoral or Master's dissertations. You should understand, therefore, that applied diagnostic procedures and/or therapies can take many years to develop after the thesis that proposed the new technique or approach was written.

In this chapter, we will give you a bibliography on recent dissertations relating to primary sclerosing cholangitis. You can read about these in more detail using the Internet or your local medical library. We will also provide you with information on how to use the Internet to stay current on dissertations.

Dissertations on Primary Sclerosing Cholangitis

ProQuest Digital Dissertations is the largest archive of academic dissertations available. From this archive, we have compiled the following list covering dissertations devoted to primary sclerosing cholangitis. You will see that the information provided includes the dissertation's title, its author, and the author's institution. To read more about the following, simply use the Internet address indicated. The following covers recent dissertations dealing with primary sclerosing cholangitis:

- **Imaging of Biliary Carcinoma, Fistula and Primary Sclerosing Cholangitis and Percutaneous Metallic Stenting in Malignant Biliary**

Obstruction by Oikarinen, Helja; Phd from Oulun Yliopisto (finland), 2001, 127 pages
http://wwwlib.umi.com/dissertations/fullcit/f475937

Keeping Current

As previously mentioned, an effective way to stay current on dissertations dedicated to primary sclerosing cholangitis is to use the database called *ProQuest Digital Dissertations* via the Internet, located at the following Web address: **http://wwwlib.umi.com/dissertations.** The site allows you to freely access the last two years of citations and abstracts. Ask your medical librarian if the library has full and unlimited access to this database. From the library, you should be able to do more complete searches than with the limited 2-year access available to the general public.

PART III. APPENDICES

ABOUT PART III

Part III is a collection of appendices on general medical topics which may be of interest to patients with primary sclerosing cholangitis and related conditions.

APPENDIX A. RESEARCHING YOUR MEDICATIONS

Overview

There are a number of sources available on new or existing medications which could be prescribed to patients with primary sclerosing cholangitis. While a number of hard copy or CD-Rom resources are available to patients and physicians for research purposes, a more flexible method is to use Internet-based databases. In this chapter, we will begin with a general overview of medications. We will then proceed to outline official recommendations on how you should view your medications. You may also want to research medications that you are currently taking for other conditions as they may interact with medications for primary sclerosing cholangitis. Research can give you information on the side effects, interactions, and limitations of prescription drugs used in the treatment of primary sclerosing cholangitis. Broadly speaking, there are two sources of information on approved medications: public sources and private sources. We will emphasize free-to-use public sources.

Your Medications: The Basics[40]

The Agency for Health Care Research and Quality has published extremely useful guidelines on how you can best participate in the medication aspects of primary sclerosing cholangitis. Taking medicines is not always as simple as swallowing a pill. It can involve many steps and decisions each day. The AHCRQ recommends that patients with primary sclerosing cholangitis take part in treatment decisions. Do not be afraid to ask questions and talk about your concerns. By taking a moment to ask questions early, you may avoid problems later. Here are some points to cover each time a new medicine is prescribed**:**

- Ask about all parts of your treatment, including diet changes, exercise, and medicines.
- Ask about the risks and benefits of each medicine or other treatment you might receive.
- Ask how often you or your doctor will check for side effects from a given medication.

Do not hesitate to ask what is important to you about your medicines. You may want a medicine with the fewest side effects, or the fewest doses to take each day. You may care most about cost, or how the medicine might affect how you live or work. Or, you may want the medicine your doctor believes will work the best. Telling your doctor will help him or her select the best treatment for you.

Do not be afraid to "bother" your doctor with your concerns and questions about medications for primary sclerosing cholangitis. You can also talk to a nurse or a pharmacist. They can help you better understand your treatment plan. Feel free to bring a friend or family member with you when you visit your doctor. Talking over your options with someone you trust can help you make better choices, especially if you are not feeling well. Specifically, ask your doctor the following:

- The name of the medicine and what it is supposed to do.
- How and when to take the medicine, how much to take, and for how long.
- What food, drinks, other medicines, or activities you should avoid while taking the medicine.
- What side effects the medicine may have, and what to do if they occur.

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[40] This section is adapted from AHCRQ: **http://www.ahcpr.gov/consumer/ncpiebro.htm**.

- If you can get a refill, and how often.
- About any terms or directions you do not understand.
- What to do if you miss a dose.
- If there is written information you can take home (most pharmacies have information sheets on your prescription medicines; some even offer large-print or Spanish versions).

Do not forget to tell your doctor about all the medicines you are currently taking (not just those for primary sclerosing cholangitis). This includes prescription medicines and the medicines that you buy over the counter. Then your doctor can avoid giving you a new medicine that may not work well with the medications you take now. When talking to your doctor, you may wish to prepare a list of medicines you currently take, the reason you take them, and how you take them. Be sure to include the following information for each:

- Name of medicine
- Reason taken
- Dosage
- Time(s) of day

Also include any over-the-counter medicines, such as:

- Laxatives
- Diet pills
- Vitamins
- Cold medicine
- Aspirin or other pain, headache, or fever medicine
- Cough medicine
- Allergy relief medicine
- Antacids
- Sleeping pills
- Others (include names)

Learning More about Your Medications

Because of historical investments by various organizations and the emergence of the Internet, it has become rather simple to learn about the medications your doctor has recommended for primary sclerosing cholangitis. One such source is the United States Pharmacopeia. In 1820, eleven physicians met in Washington, D.C. to establish the first compendium of standard drugs for the United States. They called this compendium the "U.S. Pharmacopeia (USP)." Today, the USP is a non-profit organization consisting of 800 volunteer scientists, eleven elected officials, and 400 representatives of state associations and colleges of medicine and pharmacy. The USP is located in Rockville, Maryland, and its home page is located at **www.usp.org**. The USP currently provides standards for over 3,700 medications. The resulting USP DI® Advice for the Patient® can be accessed through the National Library of Medicine of the National Institutes of Health. The database is partially derived from lists of federally approved medications in the Food and Drug Administration's (FDA) Drug Approvals database.[41]

While the FDA database is rather large and difficult to navigate, the Phamacopeia is both user-friendly and free to use. It covers more than 9,000 prescription and over-the-counter medications. To access this database, simply type the following hyperlink into your Web browser: **http://www.nlm.nih.gov/medlineplus/druginformation.html**. To view examples of a given medication (brand names, category, description, preparation, proper use, precautions, side effects, etc.), simply follow the hyperlinks indicated within the United States Pharmacopoeia (USP). It is important to read the disclaimer by the USP (**http://www.nlm.nih.gov/medlineplus/drugdisclaimer.html**) before using the information provided.

Commercial Databases

In addition to the medications listed in the USP above, a number of commercial sites are available by subscription to physicians and their institutions. You may be able to access these sources from your local medical library or your doctor's office.

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[41] Though cumbersome, the FDA database can be freely browsed at the following site: **www.fda.gov/cder/da/da.htm**.

Reuters Health Drug Database

The Reuters Health Drug Database can be searched by keyword at the hyperlink: **http://www.reutershealth.com/frame2/drug.html**.

Mosby's GenRx

Mosby's GenRx database (also available on CD-Rom and book format) covers 45,000 drug products including generics and international brands. It provides prescribing information, drug interactions, and patient information. Information can be obtained at the following hyperlink: **http://www.genrx.com/Mosby/PhyGenRx/group.html**.

Physicians Desk Reference

The Physicians Desk Reference database (also available in CD-Rom and book format) is a full-text drug database. The database is searchable by brand name, generic name or by indication. It features multiple drug interactions reports. Information can be obtained at the following hyperlink: **http://physician.pdr.net/physician/templates/en/acl/psuser_t.htm**.

Other Web Sites

A number of additional Web sites discuss drug information. As an example, you may like to look at **www.drugs.com** which reproduces the information in the Pharmacopeia as well as commercial information. You may also want to consider the Web site of the Medical Letter, Inc. which allows users to download articles on various drugs and therapeutics for a nominal fee: **http://www.medletter.com/**.

Contraindications and Interactions (Hidden Dangers)

Some of the medications mentioned in the previous discussions can be problematic for patients with primary sclerosing cholangitis--not because they are used in the treatment process, but because of contraindications, or side effects. Medications with contraindications are those that could react with drugs used to treat primary sclerosing cholangitis or potentially create deleterious side effects in patients with primary sclerosing cholangitis. You should ask your physician about any contraindications, especially as these

might apply to other medications that you may be taking for common ailments.

Drug-drug interactions occur when two or more drugs react with each other. This drug-drug interaction may cause you to experience an unexpected side effect. Drug interactions may make your medications less effective, cause unexpected side effects, or increase the action of a particular drug. Some drug interactions can even be harmful to you.

Be sure to read the label every time you use a nonprescription or prescription drug, and take the time to learn about drug interactions. These precautions may be critical to your health. You can reduce the risk of potentially harmful drug interactions and side effects with a little bit of knowledge and common sense.

Drug labels contain important information about ingredients, uses, warnings, and directions which you should take the time to read and understand. Labels also include warnings about possible drug interactions. Further, drug labels may change as new information becomes available. This is why it's especially important to read the label every time you use a medication. When your doctor prescribes a new drug, discuss all over-the-counter and prescription medications, dietary supplements, vitamins, botanicals, minerals and herbals you take as well as the foods you eat. Ask your pharmacist for the package insert for each prescription drug you take. The package insert provides more information about potential drug interactions.

A Final Warning

At some point, you may hear of alternative medications from friends, relatives, or in the news media. Advertisements may suggest that certain alternative drugs can produce positive results for patients with primary sclerosing cholangitis. Exercise caution--some of these drugs may have fraudulent claims, and others may actually hurt you. The Food and Drug Administration (FDA) is the official U.S. agency charged with discovering which medications are likely to improve the health of patients with primary sclerosing cholangitis. The FDA warns patients to watch out for[42]:

- Secret formulas (real scientists share what they know)

[42] This section has been adapted from **http://www.fda.gov/opacom/lowlit/medfraud.html**.

- Amazing breakthroughs or miracle cures (real breakthroughs don't happen very often; when they do, real scientists do not call them amazing or miracles)
- Quick, painless, or guaranteed cures
- If it sounds too good to be true, it probably isn't true.

If you have any questions about any kind of medical treatment, the FDA may have an office near you. Look for their number in the blue pages of the phone book. You can also contact the FDA through its toll-free number, 1-888-INFO-FDA (1-888-463-6332), or on the World Wide Web at **www.fda.gov**.

General References

In addition to the resources provided earlier in this chapter, the following general references describe medications (sorted alphabetically by title; hyperlinks provide rankings, information and reviews at Amazon.com):

- **Drug Development: Molecular Targets for Gi Diseases** by Timothy S. Gaginella (Editor), Antonio Guglietta (Editor); Hardcover - 288 pages (December 1999), Humana Press; ISBN: 0896035891; **http://www.amazon.com/exec/obidos/ASIN/0896035891/icongroupinterna**
- **Drug Therapy for Gastrointestinal and Liver Diseases** by Michael J.G. Farthing, M.D. (Editor), Anne B. Ballinger (Editor); Hardcover - 346 pages, 1st edition (August 15, 2001), Martin Dunitz Ltd.; ISBN: 1853177334; **http://www.amazon.com/exec/obidos/ASIN/1853177334/icongroupinterna**
- **Immunopharmacology of the Gastrointestinal System (Handbook of Immunopharmacology)** by John L. Wallace (Editor); Hardcover (October 1997), Academic Press; ISBN: 0127328602; **http://www.amazon.com/exec/obidos/ASIN/0127328602/icongroupinterna**
- **A Pharmacologic Approach to Gastrointestinal Disorders** by James H. Lewis, M.D. (Editor); Hardcover – (February 1994), Lippincott, Williams & Wilkins; ISBN: 0683049704; **http://www.amazon.com/exec/obidos/ASIN/0683049704/icongroupinterna**

Vocabulary Builder

The following vocabulary builder gives definitions of words used in this chapter that have not been defined in previous chapters:

Pharmacist: A person trained to prepare and distribute medicines and to give information about them. [NIH]

Pharmacologic: Pertaining to pharmacology or to the properties and reactions of drugs. [EU]

APPENDIX B. RESEARCHING ALTERNATIVE MEDICINE

Overview

Complementary and alternative medicine (CAM) is one of the most contentious aspects of modern medical practice. You may have heard of these treatments on the radio or on television. Maybe you have seen articles written about these treatments in magazines, newspapers, or books. Perhaps your friends or doctor have mentioned alternatives.

In this chapter, we will begin by giving you a broad perspective on complementary and alternative therapies. Next, we will introduce you to official information sources on CAM relating to primary sclerosing cholangitis. Finally, at the conclusion of this chapter, we will provide a list of readings on primary sclerosing cholangitis from various authors. We will begin, however, with the National Center for Complementary and Alternative Medicine's (NCCAM) overview of complementary and alternative medicine.

What Is CAM?[43]

Complementary and alternative medicine (CAM) covers a broad range of healing philosophies, approaches, and therapies. Generally, it is defined as those treatments and healthcare practices which are not taught in medical schools, used in hospitals, or reimbursed by medical insurance companies. Many CAM therapies are termed "holistic," which generally means that the healthcare practitioner considers the whole person, including physical, mental, emotional, and spiritual health. Some of these therapies are also

[43] Adapted from the NCCAM: **http://nccam.nih.gov/nccam/fcp/faq/index.html#what-is**.

known as "preventive," which means that the practitioner educates and treats the person to prevent health problems from arising, rather than treating symptoms after problems have occurred.

People use CAM treatments and therapies in a variety of ways. Therapies are used alone (often referred to as alternative), in combination with other alternative therapies, or in addition to conventional treatment (sometimes referred to as complementary). Complementary and alternative medicine, or "integrative medicine," includes a broad range of healing philosophies, approaches, and therapies. Some approaches are consistent with physiological principles of Western medicine, while others constitute healing systems with non-Western origins. While some therapies are far outside the realm of accepted Western medical theory and practice, others are becoming established in mainstream medicine.

Complementary and alternative therapies are used in an effort to prevent illness, reduce stress, prevent or reduce side effects and symptoms, or control or cure disease. Some commonly used methods of complementary or alternative therapy include mind/body control interventions such as visualization and relaxation, manual healing including acupressure and massage, homeopathy, vitamins or herbal products, and acupuncture.

What Are the Domains of Alternative Medicine?[44]

The list of CAM practices changes continually. The reason being is that these new practices and therapies are often proved to be safe and effective, and therefore become generally accepted as "mainstream" healthcare practices. Today, CAM practices may be grouped within five major domains: (1) alternative medical systems, (2) mind-body interventions, (3) biologically-based treatments, (4) manipulative and body-based methods, and (5) energy therapies. The individual systems and treatments comprising these categories are too numerous to list in this sourcebook. Thus, only limited examples are provided within each.

Alternative Medical Systems

Alternative medical systems involve complete systems of theory and practice that have evolved independent of, and often prior to, conventional biomedical approaches. Many are traditional systems of medicine that are

[44] Adapted from the NCCAM: **http://nccam.nih.gov/nccam/fcp/classify/index.html**.

practiced by individual cultures throughout the world, including a number of venerable Asian approaches.

Traditional oriental medicine emphasizes the balance or disturbances of qi (pronounced chi) or vital energy in health and disease, respectively. Traditional oriental medicine consists of a group of techniques and methods including acupuncture, herbal medicine, oriental massage, and qi gong (a form of energy therapy). Acupuncture involves stimulating specific anatomic points in the body for therapeutic purposes, usually by puncturing the skin with a thin needle.

Ayurveda is India's traditional system of medicine. Ayurvedic medicine (meaning "science of life") is a comprehensive system of medicine that places equal emphasis on body, mind, and spirit. Ayurveda strives to restore the innate harmony of the individual. Some of the primary Ayurvedic treatments include diet, exercise, meditation, herbs, massage, exposure to sunlight, and controlled breathing.

Other traditional healing systems have been developed by the world's indigenous populations. These populations include Native American, Aboriginal, African, Middle Eastern, Tibetan, and Central and South American cultures. Homeopathy and naturopathy are also examples of complete alternative medicine systems.

Homeopathic medicine is an unconventional Western system that is based on the principle that "like cures like," i.e., that the same substance that in large doses produces the symptoms of an illness, in very minute doses cures it. Homeopathic health practitioners believe that the more dilute the remedy, the greater its potency. Therefore, they use small doses of specially prepared plant extracts and minerals to stimulate the body's defense mechanisms and healing processes in order to treat illness.

Naturopathic medicine is based on the theory that disease is a manifestation of alterations in the processes by which the body naturally heals itself and emphasizes health restoration rather than disease treatment. Naturopathic physicians employ an array of healing practices, including the following: diet and clinical nutrition, homeopathy, acupuncture, herbal medicine, hydrotherapy (the use of water in a range of temperatures and methods of applications), spinal and soft-tissue manipulation, physical therapies (such as those involving electrical currents, ultrasound, and light), therapeutic counseling, and pharmacology.

Mind-Body Interventions

Mind-body interventions employ a variety of techniques designed to facilitate the mind's capacity to affect bodily function and symptoms. Only a select group of mind-body interventions having well-documented theoretical foundations are considered CAM. For example, patient education and cognitive-behavioral approaches are now considered "mainstream." On the other hand, complementary and alternative medicine includes meditation, certain uses of hypnosis, dance, music, and art therapy, as well as prayer and mental healing.

Biological-Based Therapies

This category of CAM includes natural and biological-based practices, interventions, and products, many of which overlap with conventional medicine's use of dietary supplements. This category includes herbal, special dietary, orthomolecular, and individual biological therapies.

Herbal therapy employs an individual herb or a mixture of herbs for healing purposes. An herb is a plant or plant part that produces and contains chemical substances that act upon the body. Special diet therapies, such as those proposed by Drs. Atkins, Ornish, Pritikin, and Weil, are believed to prevent and/or control illness as well as promote health. Orthomolecular therapies aim to treat disease with varying concentrations of chemicals such as magnesium, melatonin, and mega-doses of vitamins. Biological therapies include, for example, the use of laetrile and shark cartilage to treat cancer and the use of bee pollen to treat autoimmune and inflammatory diseases.

Manipulative and Body-Based Methods

This category includes methods that are based on manipulation and/or movement of the body. For example, chiropractors focus on the relationship between structure and function, primarily pertaining to the spine, and how that relationship affects the preservation and restoration of health. Chiropractors use manipulative therapy as an integral treatment tool.

In contrast, osteopaths place particular emphasis on the musculoskeletal system and practice osteopathic manipulation. Osteopaths believe that all of the body's systems work together and that disturbances in one system may have an impact upon function elsewhere in the body. Massage therapists manipulate the soft tissues of the body to normalize those tissues.

Energy Therapies

Energy therapies focus on energy fields originating within the body (biofields) or those from other sources (electromagnetic fields). Biofield therapies are intended to affect energy fields (the existence of which is not yet experimentally proven) that surround and penetrate the human body. Some forms of energy therapy manipulate biofields by applying pressure and/or manipulating the body by placing the hands in or through these fields. Examples include Qi gong, Reiki and Therapeutic Touch.

Qi gong is a component of traditional oriental medicine that combines movement, meditation, and regulation of breathing to enhance the flow of vital energy (qi) in the body, improve blood circulation, and enhance immune function. Reiki, the Japanese word representing Universal Life Energy, is based on the belief that, by channeling spiritual energy through the practitioner, the spirit is healed and, in turn, heals the physical body. Therapeutic Touch is derived from the ancient technique of "laying-on of hands." It is based on the premises that the therapist's healing force affects the patient's recovery and that healing is promoted when the body's energies are in balance. By passing their hands over the patient, these healers identify energy imbalances.

Bioelectromagnetic-based therapies involve the unconventional use of electromagnetic fields to treat illnesses or manage pain. These therapies are often used to treat asthma, cancer, and migraine headaches. Types of electromagnetic fields which are manipulated in these therapies include pulsed fields, magnetic fields, and alternating current or direct current fields.

Can Alternatives Affect My Treatment?

A critical issue in pursuing complementary alternatives mentioned thus far is the risk that these might have undesirable interactions with your medical treatment. It becomes all the more important to speak with your doctor who can offer advice on the use of alternatives. Official sources confirm this view. Though written for women, we find that the National Women's Health Information Center's advice on pursuing alternative medicine is appropriate for patients of both genders and all ages.[45]

[45] Adapted from **http://www.4woman.gov/faq/alternative.htm**.

Is It Okay to Want Both Traditional and Alternative or Complementary Medicine?

Should you wish to explore non-traditional types of treatment, be sure to discuss all issues concerning treatments and therapies with your healthcare provider, whether a physician or practitioner of complementary and alternative medicine. Competent healthcare management requires knowledge of both conventional and alternative therapies you are taking for the practitioner to have a complete picture of your treatment plan.

The decision to use complementary and alternative treatments is an important one. Consider before selecting an alternative therapy, the safety and effectiveness of the therapy or treatment, the expertise and qualifications of the healthcare practitioner, and the quality of delivery. These topics should be considered when selecting any practitioner or therapy.

Finding CAM References on Primary Sclerosing Cholangitis

Having read the previous discussion, you may be wondering which complementary or alternative treatments might be appropriate for primary sclerosing cholangitis. For the remainder of this chapter, we will direct you to a number of official sources which can assist you in researching studies and publications. Some of these articles are rather technical, so some patience may be required.

National Center for Complementary and Alternative Medicine

The National Center for Complementary and Alternative Medicine (NCCAM) of the National Institutes of Health (**http://nccam.nih.gov**) has created a link to the National Library of Medicine's databases to allow patients to search for articles that specifically relate to primary sclerosing cholangitis and complementary medicine. To search the database, go to the following Web site: **www.nlm.nih.gov/nccam/camonpubmed.html**. Select "CAM on PubMed." Enter "primary sclerosing cholangitis" (or synonyms) into the search box. Click "Go." The following references provide information on particular aspects of complementary and alternative medicine (CAM) that are related to primary sclerosing cholangitis:

- **Detection of anti-liver cell membrane antibody using a human hepatocellular carcinoma cell line.**
  Author(s): Lobo-Yeo A, McSorley C, McFarlane BM, Mieli-Vergani G, Mowat AP, Vergani D.

Source: Hepatology (Baltimore, Md.). 1989 February; 9(2): 210-4.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=2536348&dopt=Abstract

- **Effect of dietary fiber on serum bile acids in patients with chronic cholestatic liver disease under ursodeoxycholic acid therapy.**
  Author(s): Sauter G, Beuers U, Paumgartner G.
  Source: Digestion. 1995; 56(6): 523-7.
  http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8536824&dopt=Abstract

- **Langerhans' cell histiocytosis presenting with hepatic dysfunction.**
  Author(s): Squires RH Jr, Weinberg AG, Zwiener RJ, Winick N.
  Source: Journal of Pediatric Gastroenterology and Nutrition. 1993 February; 16(2): 190-3. No Abstract Available.
  http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8450389&dopt=Abstract

- **The pathobiology of biliary epithelia.**
  Author(s): Alpini G, McGill JM, Larusso NF.
  Source: Hepatology (Baltimore, Md.). 2002 May; 35(5): 1256-68. Review.
  http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11981776&dopt=Abstract

Additional Web Resources

A number of additional Web sites offer encyclopedic information covering CAM and related topics. The following is a representative sample:

- Alternative Medicine Foundation, Inc.: **http://www.herbmed.org/**
- AOL: **http://search.aol.com/cat.adp?id=169&layer=&from=subcats**
- Chinese Medicine: **http://www.newcenturynutrition.com/**
- drkoop.com®: **http://www.drkoop.com/InteractiveMedicine/IndexC.html**
- Family Village: **http://www.familyvillage.wisc.edu/med_altn.htm**
- Google: **http://directory.google.com/Top/Health/Alternative/**
- Healthnotes: **http://www.thedacare.org/healthnotes/**
- Open Directory Project: **http://dmoz.org/Health/Alternative/**

- TPN.com: **http://www.tnp.com/**
- Yahoo.com: **http://dir.yahoo.com/Health/Alternative_Medicine/**
- WebMD®Health: **http://my.webmd.com/drugs_and_herbs**
- WellNet: **http://www.wellnet.ca/herbsa-c.htm**
- WholeHealthMD.com: **http://www.wholehealthmd.com/reflib/0,1529,,00.html**

General References

A good place to find general background information on CAM is the National Library of Medicine. It has prepared within the MEDLINEplus system an information topic page dedicated to complementary and alternative medicine. To access this page, go to the MEDLINEplus site at: **www.nlm.nih.gov/medlineplus/alternativemedicine.html.** This Web site provides a general overview of various topics and can lead to a number of general sources. The following additional references describe, in broad terms, alternative and complementary medicine (sorted alphabetically by title; hyperlinks provide rankings, information, and reviews at Amazon.com):

- **Gastrointestinal Disorders and Nutrition** by Tonia Reinhard; Paperback - 192 pages (January 24, 2002), McGraw-Hill Professional Publishing; ISBN: 0737303611; **http://www.amazon.com/exec/obidos/ASIN/0737303611/icongroupinterna**
- **Healthy Digestion the Natural Way: Preventing and Healing Heartburn, Constipation, Gas, Diarrhea, Inflammatory Bowel and Gallbladder Diseases, Ulcers, Irritable Bowel Syndrome, and More** by D. Lindsey Berkson, et al; Paperback - 256 pages, 1st edition (February 2000), John Wiley & Sons; ISBN: 0471349623; **http://www.amazon.com/exec/obidos/ASIN/0471349623/icongroupinterna**
- **No More Heartburn: Stop the Pain in 30 Days--Naturally!: The Safe, Effective Way to Prevent and Heal Chronic Gastrointestinal Disorders** by Sherry A. Rogers, M.D.; Paperback - 320 pages (February 2000), Kensington Publishing Corp.; ISBN: 1575665107; **http://www.amazon.com/exec/obidos/ASIN/1575665107/icongroupinterna**

For additional information on complementary and alternative medicine, ask your doctor or write to:

National Institutes of Health
National Center for Complementary and Alternative Medicine Clearinghouse
P. O. Box 8218
Silver Spring, MD 20907-8218

Vocabulary Builder

The following vocabulary builder gives definitions of words used in this chapter that have not been defined in previous chapters:

Diarrhea: Passage of excessively liquid or excessively frequent stools. [NIH]

Heartburn: Substernal pain or burning sensation, usually associated with regurgitation of gastric juice into the esophagus. [NIH]

Histiocytosis: General term for the abnormal appearance of histiocytes in the blood. Based on the pathological features of the cells involved rather than on clinical findings, the histiocytic diseases are subdivided into three groups: histiocytosis, langerhans cell; histiocytosis, non-langerhans cell; and histiocytic disorders, malignant. [NIH]

Membrane: A thin layer of tissue which covers a surface, lines a cavity or divides a space or organ. [EU]

APPENDIX C. RESEARCHING NUTRITION

Overview

Since the time of Hippocrates, doctors have understood the importance of diet and nutrition to patients' health and well-being. Since then, they have accumulated an impressive archive of studies and knowledge dedicated to this subject. Based on their experience, doctors and healthcare providers may recommend particular dietary supplements to patients with primary sclerosing cholangitis. Any dietary recommendation is based on a patient's age, body mass, gender, lifestyle, eating habits, food preferences, and health condition. It is therefore likely that different patients with primary sclerosing cholangitis may be given different recommendations. Some recommendations may be directly related to primary sclerosing cholangitis, while others may be more related to the patient's general health. These recommendations, themselves, may differ from what official sources recommend for the average person.

In this chapter we will begin by briefly reviewing the essentials of diet and nutrition that will broadly frame more detailed discussions of primary sclerosing cholangitis. We will then show you how to find studies dedicated specifically to nutrition and primary sclerosing cholangitis.

Food and Nutrition: General Principles

What Are Essential Foods?

Food is generally viewed by official sources as consisting of six basic elements: (1) fluids, (2) carbohydrates, (3) protein, (4) fats, (5) vitamins, and

(6) minerals. Consuming a combination of these elements is considered to be a healthy diet:

- **Fluids** are essential to human life as 80-percent of the body is composed of water. Water is lost via urination, sweating, diarrhea, vomiting, diuretics (drugs that increase urination), caffeine, and physical exertion.
- **Carbohydrates** are the main source for human energy (thermoregulation) and the bulk of typical diets. They are mostly classified as being either simple or complex. Simple carbohydrates include sugars which are often consumed in the form of cookies, candies, or cakes. Complex carbohydrates consist of starches and dietary fibers. Starches are consumed in the form of pastas, breads, potatoes, rice, and other foods. Soluble fibers can be eaten in the form of certain vegetables, fruits, oats, and legumes. Insoluble fibers include brown rice, whole grains, certain fruits, wheat bran and legumes.
- **Proteins** are eaten to build and repair human tissues. Some foods that are high in protein are also high in fat and calories. Food sources for protein include nuts, meat, fish, cheese, and other dairy products.
- **Fats** are consumed for both energy and the absorption of certain vitamins. There are many types of fats, with many general publications recommending the intake of unsaturated fats or those low in cholesterol.

Vitamins and minerals are fundamental to human health, growth, and, in some cases, disease prevention. Most are consumed in your diet (exceptions being vitamins K and D which are produced by intestinal bacteria and sunlight on the skin, respectively). Each vitamin and mineral plays a different role in health. The following outlines essential vitamins:

- **Vitamin A** is important to the health of your eyes, hair, bones, and skin; sources of vitamin A include foods such as eggs, carrots, and cantaloupe.
- **Vitamin B^1**, also known as thiamine, is important for your nervous system and energy production; food sources for thiamine include meat, peas, fortified cereals, bread, and whole grains.
- **Vitamin B^2**, also known as riboflavin, is important for your nervous system and muscles, but is also involved in the release of proteins from nutrients; food sources for riboflavin include dairy products, leafy vegetables, meat, and eggs.
- **Vitamin B^3**, also known as niacin, is important for healthy skin and helps the body use energy; food sources for niacin include peas, peanuts, fish, and whole grains

- **Vitamin B^6**, also known as pyridoxine, is important for the regulation of cells in the nervous system and is vital for blood formation; food sources for pyridoxine include bananas, whole grains, meat, and fish.
- **Vitamin B^{12}** is vital for a healthy nervous system and for the growth of red blood cells in bone marrow; food sources for vitamin B12 include yeast, milk, fish, eggs, and meat.
- **Vitamin C** allows the body's immune system to fight various diseases, strengthens body tissue, and improves the body's use of iron; food sources for vitamin C include a wide variety of fruits and vegetables.
- **Vitamin D** helps the body absorb calcium which strengthens bones and teeth; food sources for vitamin D include oily fish and dairy products.
- **Vitamin E** can help protect certain organs and tissues from various degenerative diseases; food sources for vitamin E include margarine, vegetables, eggs, and fish.
- **Vitamin K** is essential for bone formation and blood clotting; common food sources for vitamin K include leafy green vegetables.
- **Folic Acid** maintains healthy cells and blood and, when taken by a pregnant woman, can prevent her fetus from developing neural tube defects; food sources for folic acid include nuts, fortified breads, leafy green vegetables, and whole grains.

It should be noted that one can overdose on certain vitamins which become toxic if consumed in excess (e.g. vitamin A, D, E and K).

Like vitamins, minerals are chemicals that are required by the body to remain in good health. Because the human body does not manufacture these chemicals internally, we obtain them from food and other dietary sources. The more important minerals include:

- **Calcium** is needed for healthy bones, teeth, and muscles, but also helps the nervous system function; food sources for calcium include dry beans, peas, eggs, and dairy products.
- **Chromium** is helpful in regulating sugar levels in blood; food sources for chromium include egg yolks, raw sugar, cheese, nuts, beets, whole grains, and meat.
- **Fluoride** is used by the body to help prevent tooth decay and to reinforce bone strength; sources of fluoride include drinking water and certain brands of toothpaste.

- **Iodine** helps regulate the body's use of energy by synthesizing into the hormone thyroxine; food sources include leafy green vegetables, nuts, egg yolks, and red meat.
- **Iron** helps maintain muscles and the formation of red blood cells and certain proteins; food sources for iron include meat, dairy products, eggs, and leafy green vegetables.
- **Magnesium** is important for the production of DNA, as well as for healthy teeth, bones, muscles, and nerves; food sources for magnesium include dried fruit, dark green vegetables, nuts, and seafood.
- **Phosphorous** is used by the body to work with calcium to form bones and teeth; food sources for phosphorous include eggs, meat, cereals, and dairy products.
- **Selenium** primarily helps maintain normal heart and liver functions; food sources for selenium include wholegrain cereals, fish, meat, and dairy products.
- **Zinc** helps wounds heal, the formation of sperm, and encourage rapid growth and energy; food sources include dried beans, shellfish, eggs, and nuts.

The United States government periodically publishes recommended diets and consumption levels of the various elements of food. Again, your doctor may encourage deviations from the average official recommendation based on your specific condition. To learn more about basic dietary guidelines, visit the Web site: **http://www.health.gov/dietaryguidelines/**. Based on these guidelines, many foods are required to list the nutrition levels on the food's packaging. Labeling Requirements are listed at the following site maintained by the Food and Drug Administration: **http://www.cfsan.fda.gov/~dms/lab-cons.html**. When interpreting these requirements, the government recommends that consumers become familiar with the following abbreviations before reading FDA literature:[46]

- **DVs (Daily Values):** A new dietary reference term that will appear on the food label. It is made up of two sets of references, DRVs and RDIs.
- **DRVs (Daily Reference Values):** A set of dietary references that applies to fat, saturated fat, cholesterol, carbohydrate, protein, fiber, sodium, and potassium.
- **RDIs (Reference Daily Intakes):** A set of dietary references based on the Recommended Dietary Allowances for essential vitamins and minerals

[46] Adapted from the FDA: **http://www.fda.gov/fdac/special/foodlabel/dvs.html**.

and, in selected groups, protein. The name "RDI" replaces the term "U.S. RDA."

- **RDAs (Recommended Dietary Allowances):** A set of estimated nutrient allowances established by the National Academy of Sciences. It is updated periodically to reflect current scientific knowledge.

What Are Dietary Supplements?[47]

Dietary supplements are widely available through many commercial sources, including health food stores, grocery stores, pharmacies, and by mail. Dietary supplements are provided in many forms including tablets, capsules, powders, gel-tabs, extracts, and liquids. Historically in the United States, the most prevalent type of dietary supplement was a multivitamin/mineral tablet or capsule that was available in pharmacies, either by prescription or "over the counter." Supplements containing strictly herbal preparations were less widely available. Currently in the United States, a wide array of supplement products are available, including vitamin, mineral, other nutrients, and botanical supplements as well as ingredients and extracts of animal and plant origin.

The Office of Dietary Supplements (ODS) of the National Institutes of Health is the official agency of the United States which has the expressed goal of acquiring "new knowledge to help prevent, detect, diagnose, and treat disease and disability, from the rarest genetic disorder to the common cold."[48] According to the ODS, dietary supplements can have an important impact on the prevention and management of disease and on the maintenance of health.[49] The ODS notes that considerable research on the effects of dietary supplements has been conducted in Asia and Europe where the use of plant products, in particular, has a long tradition. However, the overwhelming majority of supplements have not been studied scientifically.

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[47] This discussion has been adapted from the NIH: **http://ods.od.nih.gov/whatare/whatare.html**.

[48] Contact: The Office of Dietary Supplements, National Institutes of Health, Building 31, Room 1B29, 31 Center Drive, MSC 2086, Bethesda, Maryland 20892-2086, Tel: (301) 435-2920, Fax: (301) 480-1845, E-mail: **ods@nih.gov**.

[49] Adapted from **http://ods.od.nih.gov/about/about.html**. The Dietary Supplement Health and Education Act defines dietary supplements as "a product (other than tobacco) intended to supplement the diet that bears or contains one or more of the following dietary ingredients: a vitamin, mineral, amino acid, herb or other botanical; or a dietary substance for use to supplement the diet by increasing the total dietary intake; or a concentrate, metabolite, constituent, extract, or combination of any ingredient described above; and intended for ingestion in the form of a capsule, powder, softgel, or gelcap, and not represented as a conventional food or as a sole item of a meal or the diet."

To explore the role of dietary supplements in the improvement of health care, the ODS plans, organizes, and supports conferences, workshops, and symposia on scientific topics related to dietary supplements. The ODS often works in conjunction with other NIH Institutes and Centers, other government agencies, professional organizations, and public advocacy groups.

To learn more about official information on dietary supplements, visit the ODS site at **http://ods.od.nih.gov/whatare/whatare.html**. Or contact:

The Office of Dietary Supplements
National Institutes of Health
Building 31, Room 1B29
31 Center Drive, MSC 2086
Bethesda, Maryland 20892-2086
Tel: (301) 435-2920
Fax: (301) 480-1845
E-mail: ods@nih.gov

Finding Studies on Primary Sclerosing Cholangitis

The NIH maintains an office dedicated to patient nutrition and diet. The National Institutes of Health's Office of Dietary Supplements (ODS) offers a searchable bibliographic database called the IBIDS (International Bibliographic Information on Dietary Supplements). The IBIDS contains over 460,000 scientific citations and summaries about dietary supplements and nutrition as well as references to published international, scientific literature on dietary supplements such as vitamins, minerals, and botanicals.[50] IBIDS is available to the public free of charge through the ODS Internet page: **http://ods.od.nih.gov/databases/ibids.html**.

After entering the search area, you have three choices: (1) IBIDS Consumer Database, (2) Full IBIDS Database, or (3) Peer Reviewed Citations Only. We recommend that you start with the Consumer Database. While you may not find references for the topics that are of most interest to you, check back periodically as this database is frequently updated. More studies can be found by searching the Full IBIDS Database. Healthcare professionals and

[50] Adapted from **http://ods.od.nih.gov**. IBIDS is produced by the Office of Dietary Supplements (ODS) at the National Institutes of Health to assist the public, healthcare providers, educators, and researchers in locating credible, scientific information on dietary supplements. IBIDS was developed and will be maintained through an interagency partnership with the Food and Nutrition Information Center of the National Agricultural Library, U.S. Department of Agriculture.

researchers generally use the third option, which lists peer-reviewed citations. In all cases, we suggest that you take advantage of the "Advanced Search" option that allows you to retrieve up to 100 fully explained references in a comprehensive format. Type "primary sclerosing cholangitis" (or synonyms) into the search box. To narrow the search, you can also select the "Title" field.

The following information is typical of that found when using the "Full IBIDS Database" when searching using "primary sclerosing cholangitis" (or a synonym):

- **A 3-year prospective study on serum tumor markers used for detecting cholangiocarcinoma in patients with primary sclerosing cholangitis.**
  Author(s): Department of Gastroenterology and Hepatology, Karolinska Hospital, Stockholm, Sweden.
  Source: Hultcrantz, R Olsson, R Danielsson, A Jarnerot, G Loof, L Ryden, B O Wahren, B Broome, U J-Hepatol. 1999 April; 30(4): 669-73 0168-8278
- **A controlled trial of calcitonin therapy for the prevention of post-liver transplantation atraumatic fractures in patients with primary biliary cirrhosis and primary sclerosing cholangitis.**
  Author(s): Division of Gastroenterology, Mayo Medical Center, Rochester, MN 55905, USA. jhay@mayo.edu
  Source: Hay, J E Malinchoc, M Dickson, E R J-Hepatol. 2001 February; 34(2): 292-8 0168-8278
- **A pilot study of pentoxifylline for the treatment of primary sclerosing cholangitis.**
  Author(s): Division of Gastroenterology, Mayo Clinic, Rochester, Minnesota 55905, USA.
  Source: Bharucha, A E Jorgensen, R Lichtman, S N LaRusso, N F Lindor, K D Am-J-Gastroenterol. 2000 September; 95(9): 2338-42 0002-9270
- **Adult celiac disease and primary sclerosing cholangitis: two case reports.**
  Author(s): Cattedra di Semeiotica e Metodologia Medica, Universita di Modena, Italy.
  Source: Venturini, I Cosenza, R Miglioli, L Borghi, A Bagni, A Gandolfo, M Modonesi, G Zeneroli, M L Hepatogastroenterology. 1998 Nov-December; 45(24): 2344-7 0172-6390
- **An atypical presentation for primary sclerosing cholangitis.**
  Author(s): Hepatology Section, Medical College of Virginia, Virginia Commonwealth University, Richmond 23298, USA.
  Source: Luketic, V A Gomez, D A Sanyal, A J Shiffman, M L Dig-Dis-Sci. 1997 October; 42(10): 2009-16 0163-2116

- **Antineutrophil cytoplasmic antibodies (ANCA) directed against cathepsin G in ulcerative colitis, Crohn's disease and primary sclerosing cholangitis.**
  Author(s): Department of Nephrology, Hopital Necker, Paris, France.
  Source: Halbwachs Mecarelli, L Nusbaum, P Noel, L H Reumaux, D Erlinger, S Grunfeld, J P Lesavre, P Clin-Exp-Immunol. 1992 October; 90(1): 79-84 0009-9104
- **Biliary lactoferrin concentrations are increased in active inflammatory bowel disease: a factor in the pathogenesis of primary sclerosing cholangitis?**
  Author(s): Gastroenterology Unit, Division of Medicine, UMDS, 5th Floor, Thomas Guy House, Guy's Hospital, London SE1 9RT, U.K.
  Source: Pereira, S P Rhodes, J M Campbell, B J KuMarch, D Bain, I M Murphy, G M Dowling, R H Clin-Sci-(Colch). 1998 November; 95(5): 637-44 0143-5221
- **Celiac disease, inflammatory colitis, and primary sclerosing cholangitis in a girl with Turner's syndrome.**
  Author(s): Department of Paediatrics, Enfants-Malades Hospital, Paris, France.
  Source: Lacaille, F Canioni, D Bernard, O Fabre, M Brousse, N Schmitz, J J-Pediatr-Gastroenterol-Nutr. 1995 November; 21(4): 463-7 0277-2116
- **Cholangiocarcinoma complicating primary sclerosing cholangitis.**
  Author(s): Department of General Surgery, Mayo Clinic, Rochester, MN 55905.
  Source: Rosen, C B Nagorney, D M Wiesner, R H Coffey, R J LaRusso, N F Ann-Surg. 1991 January; 213(1): 21-5 0003-4932
- **Colchicine treatment of primary sclerosing cholangitis.**
  Author(s): Medical Clinics, Sahlgren's Hospital, Goteborg, Sweden.
  Source: Olsson, R Broome, U Danielsson, A Hagerstrand, I Jarnerot, G Loof, L Prytz, H Ryden, B O Wallerstedt, S Gastroenterology. 1995 April; 108(4): 1199-203 0016-5085
- **Differences in colonic disease activity in patients with ulcerative colitis with and without primary sclerosing cholangitis: a case control study.**
  Author(s): Department of Gastroenterology and Hepatology, Huddinge University Hospital, Sweden.
  Source: Lundqvist, K Broome, U Dis-Colon-Rectum. 1997 April; 40(4): 451-6 0012-3706
- **Medical approaches to primary sclerosing cholangitis.**
  Author(s): Department of Medicine, New England Medical Center Hospitals, Boston, MA 02111.

Source: Kaplan, M M Semin-Liver-Dis. 1991 February; 11(1): 56-63 0272-8087

- **No beneficial effects of transdermal nicotine in patients with primary sclerosing cholangitis: results of a randomized double-blind placebo-controlled cross-over study.**
  Author(s): Department of Hepatology and Gastroenterology, University Hospital, Rotterdam, The Netherlands.
  Source: Vleggaar, F P van Buuren, H R van Berge Henegouwen, G P Hop, W C van Erpecum, K J Eur-J-Gastroenterol-Hepatol. 2001 February; 13(2): 171-5 0954-691X

- **Oral nicotine in treatment of primary sclerosing cholangitis: a pilot study.**
  Author(s): Division of Gastroenterology and Hepatology, Mayo Clinic and Mayo Foundation, Rochester, Minnesota 55905, USA.
  Source: Angulo, P Bharucha, A E Jorgensen, R A DeSotel, C K Sandborn, W J Larusso, N F Lindor, K D Dig-Dis-Sci. 1999 March; 44(3): 602-7 0163-2116

- **Post-infantile giant cell hepatitis in patients with primary sclerosing cholangitis and autoimmune hepatitis.**
  Author(s): Department of Medicine, Johannes-Gutenberg-University, Mainz, Germany.
  Source: Protzer, U Dienes, H P Bianchi, L Lohse, A W Helmreich Becker, I Gerken, G Meyer zum Buschenfelde, K H Liver. 1996 August; 16(4): 274-82 0106-9543

- **Prevalence and clinical significance of anti-lactoferrin autoantibodies in inflammatory bowel diseases and primary sclerosing cholangitis.**
  Author(s): Department of Internal Medicine, University Hospital, Groningen, The Netherlands.
  Source: Roozendaal, C Horst, G Pogany, K van Milligen de Wit, A W Kleibeuker, J H Haagsma, E B Limburg, P C Kallenberg, C G Adv-Exp-Med-Biol. 1998; 443313-9 0065-2598

- **Primary sclerosing cholangitis and celiac disease. A novel association.**
  Author(s): Mayo Clinic, Rochester, Minnesota.
  Source: Hay, J E Wiesner, R H Shorter, R G LaRusso, N F Baldus, W P Ann-Intern-Med. 1988 November 1; 109(9): 713-7 0003-4819

- **Primary sclerosing cholangitis and multiple autoimmune disorders in a patient with Down syndrome.**
  Author(s): Department of Pediatric Gastroenterology, Hahnemann University Hospital, Philadelphia, Pennsylvania, USA.
  Source: Mehta, D I Hill, I D Singer Granick, C Balloch, Z Blecker, U Clin-Pediatr-(Phila). 1995 September; 34(9): 502-5 0009-9228

- **Primary sclerosing cholangitis in India.**
  Author(s): Department of Gastroenterology, All India Institute of Medical Sciences, New Delhi.
  Source: Acharya, S K Vashisht, S Tandon, R K Gastroenterol-Jpn. 1989 February; 24(1): 75-9 0435-1339
- **Primary sclerosing cholangitis with celiac sprue: two cases.**
  Author(s): Department of Infectious Diseases, Ospedali Riuniti, Bergamo, Italy.
  Source: Fracassetti, O Delvecchio, G Tambini, R Lorenzi, N Gavazzeni, G J-Clin-Gastroenterol. 1996 January; 22(1): 71-2 0192-0790

Federal Resources on Nutrition

In addition to the IBIDS, the United States Department of Health and Human Services (HHS) and the United States Department of Agriculture (USDA) provide many sources of information on general nutrition and health. Recommended resources include:

- healthfinder®, HHS's gateway to health information, including diet and nutrition:
  http://www.healthfinder.gov/scripts/SearchContext.asp?topic=238&page=0
- The United States Department of Agriculture's Web site dedicated to nutrition information: **www.nutrition.gov**
- The Food and Drug Administration's Web site for federal food safety information: **www.foodsafety.gov**
- The National Action Plan on Overweight and Obesity sponsored by the United States Surgeon General: **http://www.surgeongeneral.gov/topics/obesity/**
- The Center for Food Safety and Applied Nutrition has an Internet site sponsored by the Food and Drug Administration and the Department of Health and Human Services: **http://vm.cfsan.fda.gov/**
- Center for Nutrition Policy and Promotion sponsored by the United States Department of Agriculture: **http://www.usda.gov/cnpp/**
- Food and Nutrition Information Center, National Agricultural Library sponsored by the United States Department of Agriculture: **http://www.nal.usda.gov/fnic/**
- Food and Nutrition Service sponsored by the United States Department of Agriculture: **http://www.fns.usda.gov/fns/**

Additional Web Resources

A number of additional Web sites offer encyclopedic information covering food and nutrition. The following is a representative sample:

- AOL: **http://search.aol.com/cat.adp?id=174&layer=&from=subcats**
- Family Village: **http://www.familyvillage.wisc.edu/med_nutrition.html**
- Google: **http://directory.google.com/Top/Health/Nutrition/**
- Healthnotes: **http://www.thedacare.org/healthnotes/**
- Open Directory Project: **http://dmoz.org/Health/Nutrition/**
- Yahoo.com: **http://dir.yahoo.com/Health/Nutrition/**
- WebMD®Health: **http://my.webmd.com/nutrition**
- WholeHealthMD.com: **http://www.wholehealthmd.com/reflib/0,1529,,00.html**

Vocabulary Builder

The following vocabulary builder defines words used in the references in this chapter that have not been defined in previous chapters:

Calcitonin: A peptide hormone that lowers calcium concentration in the blood. In humans, it is released by thyroid cells and acts to decrease the formation and absorptive activity of osteoclasts. Its role in regulating plasma calcium is much greater in children and in certain diseases than in normal adults. [NIH]

Capsules: Hard or soft soluble containers used for the oral administration of medicine. [NIH]

Carbohydrate: An aldehyde or ketone derivative of a polyhydric alcohol, particularly of the pentahydric and hexahydric alcohols. They are so named because the hydrogen and oxygen are usually in the proportion to form water, (CH2O)n. The most important carbohydrates are the starches, sugars, celluloses, and gums. They are classified into mono-, di-, tri-, poly- and heterosaccharides. [EU]

Degenerative: Undergoing degeneration : tending to degenerate; having the character of or involving degeneration; causing or tending to cause degeneration. [EU]

Fats: One of the three main classes of foods and a source of energy in the body. Fats help the body use some vitamins and keep the skin healthy. They

also serve as energy stores for the body. In food, there are two types of fats: saturated and unsaturated. [NIH]

Infantile: Pertaining to an infant or to infancy. [EU]

Iodine: A nonmetallic element of the halogen group that is represented by the atomic symbol I, atomic number 53, and atomic weight of 126.90. It is a nutritionally essential element, especially important in thyroid hormone synthesis. In solution, it has anti-infective properties and is used topically. [NIH]

Nephrology: A subspecialty of internal medicine concerned with the anatomy, physiology, and pathology of the kidney. [NIH]

Neural: 1. pertaining to a nerve or to the nerves. 2. situated in the region of the spinal axis, as the neutral arch. [EU]

Niacin: Water-soluble vitamin of the B complex occurring in various animal and plant tissues. Required by the body for the formation of coenzymes NAD and NADP. Has pellagra-curative, vasodilating, and antilipemic properties. [NIH]

Nicotine: Nicotine is highly toxic alkaloid. It is the prototypical agonist at nicotinic cholinergic receptors where it dramatically stimulates neurons and ultimately blocks synaptic transmission. Nicotine is also important medically because of its presence in tobacco smoke. [NIH]

Overdose: 1. to administer an excessive dose. 2. an excessive dose. [EU]

Paediatric: Of or relating to the care and medical treatment of children; belonging to or concerned with paediatrics. [EU]

Pentoxifylline: A methylxanthine derivative that inhibits phosphodiesterase and affects blood rheology. It improves blood flow by increasing erythrocyte and leukocyte flexibility. It also inhibits platelet aggregation. Pentoxifylline modulates immunologic activity by stimulating cytokine production. [NIH]

Potassium: An element that is in the alkali group of metals. It has an atomic symbol K, atomic number 19, and atomic weight 39.10. It is the chief cation in the intracellular fluid of muscle and other cells. Potassium ion is a strong electrolyte and it plays a significant role in the regulation of fluid volume and maintenance of the water-electrolyte balance. [NIH]

Riboflavin: Nutritional factor found in milk, eggs, malted barley, liver, kidney, heart, and leafy vegetables. The richest natural source is yeast. It occurs in the free form only in the retina of the eye, in whey, and in urine; its principal forms in tissues and cells are as FMN and FAD. [NIH]

Selenium: An element with the atomic symbol Se, atomic number 34, and atomic weight 78.96. It is an essential micronutrient for mammals and other animals but is toxic in large amounts. Selenium protects intracellular structures against oxidative damage. It is an essential component of

glutathione peroxidase. [NIH]

Thermoregulation: Heat regulation. [EU]

Thyroxine: An amino acid of the thyroid gland which exerts a stimulating effect on thyroid metabolism. [NIH]

Transdermal: Entering through the dermis, or skin, as in administration of a drug applied to the skin in ointment or patch form. [EU]

APPENDIX D. FINDING MEDICAL LIBRARIES

Overview

At a medical library you can find medical texts and reference books, consumer health publications, specialty newspapers and magazines, as well as medical journals. In this Appendix, we show you how to quickly find a medical library in your area.

Preparation

Before going to the library, highlight the references mentioned in this sourcebook that you find interesting. Focus on those items that are not available via the Internet, and ask the reference librarian for help with your search. He or she may know of additional resources that could be helpful to you. Most importantly, your local public library and medical libraries have Interlibrary Loan programs with the National Library of Medicine (NLM), one of the largest medical collections in the world. According to the NLM, most of the literature in the general and historical collections of the National Library of Medicine is available on interlibrary loan to any library. NLM's interlibrary loan services are only available to libraries. If you would like to access NLM medical literature, then visit a library in your area that can request the publications for you.[51]

[51] Adapted from the NLM: **http://www.nlm.nih.gov/psd/cas/interlibrary.html**.

Finding a Local Medical Library

The quickest method to locate medical libraries is to use the Internet-based directory published by the National Network of Libraries of Medicine (NN/LM). This network includes 4626 members and affiliates that provide many services to librarians, health professionals, and the public. To find a library in your area, simply visit **http://nnlm.gov/members/adv.html** or call 1-800-338-7657.

Medical Libraries Open to the Public

In addition to the NN/LM, the National Library of Medicine (NLM) lists a number of libraries that are generally open to the public and have reference facilities. The following is the NLM's list plus hyperlinks to each library Web site. These Web pages can provide information on hours of operation and other restrictions. The list below is a small sample of libraries recommended by the National Library of Medicine (sorted alphabetically by name of the U.S. state or Canadian province where the library is located):[52]

- **Alabama:** Health InfoNet of Jefferson County (Jefferson County Library Cooperative, Lister Hill Library of the Health Sciences), **http://www.uab.edu/infonet/**
- **Alabama:** Richard M. Scrushy Library (American Sports Medicine Institute), **http://www.asmi.org/LIBRARY.HTM**
- **Arizona:** Samaritan Regional Medical Center: The Learning Center (Samaritan Health System, Phoenix, Arizona), **http://www.samaritan.edu/library/bannerlibs.htm**
- **California:** Kris Kelly Health Information Center (St. Joseph Health System), **http://www.humboldt1.com/~kkhic/index.html**
- **California:** Community Health Library of Los Gatos (Community Health Library of Los Gatos), **http://www.healthlib.org/orgresources.html**
- **California:** Consumer Health Program and Services (CHIPS) (County of Los Angeles Public Library, Los Angeles County Harbor-UCLA Medical Center Library) - Carson, CA, **http://www.colapublib.org/services/chips.html**
- **California:** Gateway Health Library (Sutter Gould Medical Foundation)
- **California:** Health Library (Stanford University Medical Center), **http://www-med.stanford.edu/healthlibrary/**

[52] Abstracted from **http://www.nlm.nih.gov/medlineplus/libraries.html**.

- **California:** Patient Education Resource Center - Health Information and Resources (University of California, San Francisco), **http://sfghdean.ucsf.edu/barnett/PERC/default.asp**
- **California:** Redwood Health Library (Petaluma Health Care District), **http://www.phcd.org/rdwdlib.html**
- **California:** San José PlaneTree Health Library, **http://planetreesanjose.org/**
- **California:** Sutter Resource Library (Sutter Hospitals Foundation), **http://go.sutterhealth.org/comm/resc-library/sac-resources.html**
- **California:** University of California, Davis. Health Sciences Libraries
- **California:** ValleyCare Health Library & Ryan Comer Cancer Resource Center (ValleyCare Health System), **http://www.valleycare.com/library.html**
- **California:** Washington Community Health Resource Library (Washington Community Health Resource Library), **http://www.healthlibrary.org/**
- **Colorado:** William V. Gervasini Memorial Library (Exempla Healthcare), **http://www.exempla.org/conslib.htm**
- **Connecticut:** Hartford Hospital Health Science Libraries (Hartford Hospital), **http://www.harthosp.org/library/**
- **Connecticut:** Healthnet: Connecticut Consumer Health Information Center (University of Connecticut Health Center, Lyman Maynard Stowe Library), **http://library.uchc.edu/departm/hnet/**
- **Connecticut:** Waterbury Hospital Health Center Library (Waterbury Hospital), **http://www.waterburyhospital.com/library/consumer.shtml**
- **Delaware:** Consumer Health Library (Christiana Care Health System, Eugene du Pont Preventive Medicine & Rehabilitation Institute), **http://www.christianacare.org/health_guide/health_guide_pmri_health_info.cfm**
- **Delaware:** Lewis B. Flinn Library (Delaware Academy of Medicine), **http://www.delamed.org/chls.html**
- **Georgia:** Family Resource Library (Medical College of Georgia), **http://cmc.mcg.edu/kids_families/fam_resources/fam_res_lib/frl.htm**
- **Georgia:** Health Resource Center (Medical Center of Central Georgia), **http://www.mccg.org/hrc/hrchome.asp**
- **Hawaii:** Hawaii Medical Library: Consumer Health Information Service (Hawaii Medical Library), **http://hml.org/CHIS/**

- **Idaho:** DeArmond Consumer Health Library (Kootenai Medical Center), **http://www.nicon.org/DeArmond/index.htm**
- **Illinois:** Health Learning Center of Northwestern Memorial Hospital (Northwestern Memorial Hospital, Health Learning Center), **http://www.nmh.org/health_info/hlc.html**
- **Illinois:** Medical Library (OSF Saint Francis Medical Center), **http://www.osfsaintfrancis.org/general/library/**
- **Kentucky:** Medical Library - Services for Patients, Families, Students & the Public (Central Baptist Hospital), **http://www.centralbap.com/education/community/library.htm**
- **Kentucky:** University of Kentucky - Health Information Library (University of Kentucky, Chandler Medical Center, Health Information Library), **http://www.mc.uky.edu/PatientEd/**
- **Louisiana:** Alton Ochsner Medical Foundation Library (Alton Ochsner Medical Foundation), **http://www.ochsner.org/library/**
- **Louisiana:** Louisiana State University Health Sciences Center Medical Library-Shreveport, **http://lib-sh.lsuhsc.edu/**
- **Maine:** Franklin Memorial Hospital Medical Library (Franklin Memorial Hospital), **http://www.fchn.org/fmh/lib.htm**
- **Maine:** Gerrish-True Health Sciences Library (Central Maine Medical Center), **http://www.cmmc.org/library/library.html**
- **Maine:** Hadley Parrot Health Science Library (Eastern Maine Healthcare), **http://www.emh.org/hll/hpl/guide.htm**
- **Maine:** Maine Medical Center Library (Maine Medical Center), **http://www.mmc.org/library/**
- **Maine:** Parkview Hospital, **http://www.parkviewhospital.org/communit.htm#Library**
- **Maine:** Southern Maine Medical Center Health Sciences Library (Southern Maine Medical Center), **http://www.smmc.org/services/service.php3?choice=10**
- **Maine:** Stephens Memorial Hospital Health Information Library (Western Maine Health), **http://www.wmhcc.com/hil_frame.html**
- **Manitoba, Canada:** Consumer & Patient Health Information Service (University of Manitoba Libraries), **http://www.umanitoba.ca/libraries/units/health/reference/chis.html**
- **Manitoba, Canada:** J.W. Crane Memorial Library (Deer Lodge Centre), **http://www.deerlodge.mb.ca/library/libraryservices.shtml**

- **Maryland:** Health Information Center at the Wheaton Regional Library (Montgomery County, Md., Dept. of Public Libraries, Wheaton Regional Library), **http://www.mont.lib.md.us/healthinfo/hic.asp**
- **Massachusetts:** Baystate Medical Center Library (Baystate Health System), **http://www.baystatehealth.com/1024/**
- **Massachusetts:** Boston University Medical Center Alumni Medical Library (Boston University Medical Center), **http://med-libwww.bu.edu/library/lib.html**
- **Massachusetts:** Lowell General Hospital Health Sciences Library (Lowell General Hospital), **http://www.lowellgeneral.org/library/HomePageLinks/WWW.htm**
- **Massachusetts:** Paul E. Woodard Health Sciences Library (New England Baptist Hospital), **http://www.nebh.org/health_lib.asp**
- **Massachusetts:** St. Luke's Hospital Health Sciences Library (St. Luke's Hospital), **http://www.southcoast.org/library/**
- **Massachusetts:** Treadwell Library Consumer Health Reference Center (Massachusetts General Hospital), **http://www.mgh.harvard.edu/library/chrcindex.html**
- **Massachusetts:** UMass HealthNet (University of Massachusetts Medical School), **http://healthnet.umassmed.edu/**
- **Michigan:** Botsford General Hospital Library - Consumer Health (Botsford General Hospital, Library & Internet Services), **http://www.botsfordlibrary.org/consumer.htm**
- **Michigan:** Helen DeRoy Medical Library (Providence Hospital and Medical Centers), **http://www.providence-hospital.org/library/**
- **Michigan:** Marquette General Hospital - Consumer Health Library (Marquette General Hospital, Health Information Center), **http://www.mgh.org/center.html**
- **Michigan:** Patient Education Resouce Center - University of Michigan Cancer Center (University of Michigan Comprehensive Cancer Center), **http://www.cancer.med.umich.edu/learn/leares.htm**
- **Michigan:** Sladen Library & Center for Health Information Resources - Consumer Health Information, **http://www.sladen.hfhs.org/library/consumer/index.html**
- **Montana:** Center for Health Information (St. Patrick Hospital and Health Sciences Center), **http://www.saintpatrick.org/chi/librarydetail.php3?ID=41**

- **National:** Consumer Health Library Directory (Medical Library Association, Consumer and Patient Health Information Section), **http://caphis.mlanet.org/directory/index.html**
- **National:** National Network of Libraries of Medicine (National Library of Medicine) - provides library services for health professionals in the United States who do not have access to a medical library, **http://nnlm.gov/**
- **National:** NN/LM List of Libraries Serving the Public (National Network of Libraries of Medicine), **http://nnlm.gov/members/**
- **Nevada:** Health Science Library, West Charleston Library (Las Vegas Clark County Library District), **http://www.lvccld.org/special_collections/medical/index.htm**
- **New Hampshire:** Dartmouth Biomedical Libraries (Dartmouth College Library), **http://www.dartmouth.edu/~biomed/resources.htmld/conshealth.htmld/**
- **New Jersey:** Consumer Health Library (Rahway Hospital), **http://www.rahwayhospital.com/library.htm**
- **New Jersey:** Dr. Walter Phillips Health Sciences Library (Englewood Hospital and Medical Center), **http://www.englewoodhospital.com/links/index.htm**
- **New Jersey:** Meland Foundation (Englewood Hospital and Medical Center), **http://www.geocities.com/ResearchTriangle/9360/**
- **New York:** Choices in Health Information (New York Public Library) - NLM Consumer Pilot Project participant, **http://www.nypl.org/branch/health/links.html**
- **New York:** Health Information Center (Upstate Medical University, State University of New York), **http://www.upstate.edu/library/hic/**
- **New York:** Health Sciences Library (Long Island Jewish Medical Center), **http://www.lij.edu/library/library.html**
- **New York:** ViaHealth Medical Library (Rochester General Hospital), **http://www.nyam.org/library/**
- **Ohio:** Consumer Health Library (Akron General Medical Center, Medical & Consumer Health Library), **http://www.akrongeneral.org/hwlibrary.htm**
- **Oklahoma:** Saint Francis Health System Patient/Family Resource Center (Saint Francis Health System), **http://www.sfh-tulsa.com/patientfamilycenter/default.asp**

- **Oregon:** Planetree Health Resource Center (Mid-Columbia Medical Center), **http://www.mcmc.net/phrc/**
- **Pennsylvania:** Community Health Information Library (Milton S. Hershey Medical Center), **http://www.hmc.psu.edu/commhealth/**
- **Pennsylvania:** Community Health Resource Library (Geisinger Medical Center), **http://www.geisinger.edu/education/commlib.shtml**
- **Pennsylvania:** HealthInfo Library (Moses Taylor Hospital), **http://www.mth.org/healthwellness.html**
- **Pennsylvania:** Hopwood Library (University of Pittsburgh, Health Sciences Library System), **http://www.hsls.pitt.edu/chi/hhrcinfo.html**
- **Pennsylvania:** Koop Community Health Information Center (College of Physicians of Philadelphia), **http://www.collphyphil.org/kooppg1.shtml**
- **Pennsylvania:** Learning Resources Center - Medical Library (Susquehanna Health System), **http://www.shscares.org/services/lrc/index.asp**
- **Pennsylvania:** Medical Library (UPMC Health System), **http://www.upmc.edu/passavant/library.htm**
- **Quebec, Canada:** Medical Library (Montreal General Hospital), **http://ww2.mcgill.ca/mghlib/**
- **South Dakota:** Rapid City Regional Hospital - Health Information Center (Rapid City Regional Hospital, Health Information Center), **http://www.rcrh.org/education/LibraryResourcesConsumers.htm**
- **Texas:** Houston HealthWays (Houston Academy of Medicine-Texas Medical Center Library), **http://hhw.library.tmc.edu/**
- **Texas:** Matustik Family Resource Center (Cook Children's Health Care System), **http://www.cookchildrens.com/Matustik_Library.html**
- **Washington:** Community Health Library (Kittitas Valley Community Hospital), **http://www.kvch.com/**
- **Washington:** Southwest Washington Medical Center Library (Southwest Washington Medical Center), **http://www.swmedctr.com/Home/**

APPENDIX E. YOUR RIGHTS AND INSURANCE

Overview

Any patient with primary sclerosing cholangitis faces a series of issues related more to the healthcare industry than to the medical condition itself. This appendix covers two important topics in this regard: your rights and responsibilities as a patient, and how to get the most out of your medical insurance plan.

Your Rights as a Patient

The President's Advisory Commission on Consumer Protection and Quality in the Healthcare Industry has created the following summary of your rights as a patient.[53]

Information Disclosure

Consumers have the right to receive accurate, easily understood information. Some consumers require assistance in making informed decisions about health plans, health professionals, and healthcare facilities. Such information includes:

- ***Health plans.*** Covered benefits, cost-sharing, and procedures for resolving complaints, licensure, certification, and accreditation status, comparable measures of quality and consumer satisfaction, provider

[53]Adapted from Consumer Bill of Rights and Responsibilities: **http://www.hcqualitycommission.gov/press/cbor.html#head1**.

network composition, the procedures that govern access to specialists and emergency services, and care management information.

- ***Health professionals.*** Education, board certification, and recertification, years of practice, experience performing certain procedures, and comparable measures of quality and consumer satisfaction.
- ***Healthcare facilities.*** Experience in performing certain procedures and services, accreditation status, comparable measures of quality, worker, and consumer satisfaction, and procedures for resolving complaints.
- ***Consumer assistance programs.*** Programs must be carefully structured to promote consumer confidence and to work cooperatively with health plans, providers, payers, and regulators. Desirable characteristics of such programs are sponsorship that ensures accountability to the interests of consumers and stable, adequate funding.

Choice of Providers and Plans

Consumers have the right to a choice of healthcare providers that is sufficient to ensure access to appropriate high-quality healthcare. To ensure such choice, the Commission recommends the following:

- ***Provider network adequacy.*** All health plan networks should provide access to sufficient numbers and types of providers to assure that all covered services will be accessible without unreasonable delay -- including access to emergency services 24 hours a day and 7 days a week. If a health plan has an insufficient number or type of providers to provide a covered benefit with the appropriate degree of specialization, the plan should ensure that the consumer obtains the benefit outside the network at no greater cost than if the benefit were obtained from participating providers.
- ***Women's health services.*** Women should be able to choose a qualified provider offered by a plan -- such as gynecologists, certified nurse midwives, and other qualified healthcare providers -- for the provision of covered care necessary to provide routine and preventative women's healthcare services.
- ***Access to specialists.*** Consumers with complex or serious medical conditions who require frequent specialty care should have direct access to a qualified specialist of their choice within a plan's network of providers. Authorizations, when required, should be for an adequate number of direct access visits under an approved treatment plan.

- ***Transitional care.*** Consumers who are undergoing a course of treatment for a chronic or disabling condition (or who are in the second or third trimester of a pregnancy) at the time they involuntarily change health plans or at a time when a provider is terminated by a plan for other than cause should be able to continue seeing their current specialty providers for up to 90 days (or through completion of postpartum care) to allow for transition of care.
- ***Choice of health plans.*** Public and private group purchasers should, wherever feasible, offer consumers a choice of high-quality health insurance plans.

Access to Emergency Services

Consumers have the right to access emergency healthcare services when and where the need arises. Health plans should provide payment when a consumer presents to an emergency department with acute symptoms of sufficient severity--including severe pain--such that a "prudent layperson" could reasonably expect the absence of medical attention to result in placing that consumer's health in serious jeopardy, serious impairment to bodily functions, or serious dysfunction of any bodily organ or part.

Participation in Treatment Decisions

Consumers have the right and responsibility to fully participate in all decisions related to their healthcare. Consumers who are unable to fully participate in treatment decisions have the right to be represented by parents, guardians, family members, or other conservators. Physicians and other health professionals should:

- Provide patients with sufficient information and opportunity to decide among treatment options consistent with the informed consent process.
- Discuss all treatment options with a patient in a culturally competent manner, including the option of no treatment at all.
- Ensure that persons with disabilities have effective communications with members of the health system in making such decisions.
- Discuss all current treatments a consumer may be undergoing.
- Discuss all risks, benefits, and consequences to treatment or nontreatment.

- Give patients the opportunity to refuse treatment and to express preferences about future treatment decisions.
- Discuss the use of advance directives -- both living wills and durable powers of attorney for healthcare -- with patients and their designated family members.
- Abide by the decisions made by their patients and/or their designated representatives consistent with the informed consent process.

Health plans, health providers, and healthcare facilities should:

- Disclose to consumers factors -- such as methods of compensation, ownership of or interest in healthcare facilities, or matters of conscience -- that could influence advice or treatment decisions.
- Assure that provider contracts do not contain any so-called "gag clauses" or other contractual mechanisms that restrict healthcare providers' ability to communicate with and advise patients about medically necessary treatment options.
- Be prohibited from penalizing or seeking retribution against healthcare professionals or other health workers for advocating on behalf of their patients.

Respect and Nondiscrimination

Consumers have the right to considerate, respectful care from all members of the healthcare industry at all times and under all circumstances. An environment of mutual respect is essential to maintain a quality healthcare system. To assure that right, the Commission recommends the following:

- Consumers must not be discriminated against in the delivery of healthcare services consistent with the benefits covered in their policy, or as required by law, based on race, ethnicity, national origin, religion, sex, age, mental or physical disability, sexual orientation, genetic information, or source of payment.
- Consumers eligible for coverage under the terms and conditions of a health plan or program, or as required by law, must not be discriminated against in marketing and enrollment practices based on race, ethnicity, national origin, religion, sex, age, mental or physical disability, sexual orientation, genetic information, or source of payment.

Confidentiality of Health Information

Consumers have the right to communicate with healthcare providers in confidence and to have the confidentiality of their individually identifiable healthcare information protected. Consumers also have the right to review and copy their own medical records and request amendments to their records.

Complaints and Appeals

Consumers have the right to a fair and efficient process for resolving differences with their health plans, healthcare providers, and the institutions that serve them, including a rigorous system of internal review and an independent system of external review. A free copy of the Patient's Bill of Rights is available from the American Hospital Association.[54]

Patient Responsibilities

Treatment is a two-way street between you and your healthcare providers. To underscore the importance of finance in modern healthcare as well as your responsibility for the financial aspects of your care, the President's Advisory Commission on Consumer Protection and Quality in the Healthcare Industry has proposed that patients understand the following "Consumer Responsibilities."[55] In a healthcare system that protects consumers' rights, it is reasonable to expect and encourage consumers to assume certain responsibilities. Greater individual involvement by the consumer in his or her care increases the likelihood of achieving the best outcome and helps support a quality-oriented, cost-conscious environment. Such responsibilities include:

- Take responsibility for maximizing healthy habits such as exercising, not smoking, and eating a healthy diet.
- Work collaboratively with healthcare providers in developing and carrying out agreed-upon treatment plans.
- Disclose relevant information and clearly communicate wants and needs.

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[54] To order your free copy of the Patient's Bill of Rights, telephone 312-422-3000 or visit the American Hospital Association's Web site: **http://www.aha.org**. Click on "Resource Center," go to "Search" at bottom of page, and then type in "Patient's Bill of Rights." The Patient's Bill of Rights is also available from Fax on Demand, at 312-422-2020, document number 471124.

[55] Adapted from **http://www.hcqualitycommission.gov/press/cbor.html#head1**.

- Use your health insurance plan's internal complaint and appeal processes to address your concerns.
- Avoid knowingly spreading disease.
- Recognize the reality of risks, the limits of the medical science, and the human fallibility of the healthcare professional.
- Be aware of a healthcare provider's obligation to be reasonably efficient and equitable in providing care to other patients and the community.
- Become knowledgeable about your health plan's coverage and options (when available) including all covered benefits, limitations, and exclusions, rules regarding use of network providers, coverage and referral rules, appropriate processes to secure additional information, and the process to appeal coverage decisions.
- Show respect for other patients and health workers.
- Make a good-faith effort to meet financial obligations.
- Abide by administrative and operational procedures of health plans, healthcare providers, and Government health benefit programs.

Choosing an Insurance Plan

There are a number of official government agencies that help consumers understand their healthcare insurance choices.[56] The U.S. Department of Labor, in particular, recommends ten ways to make your health benefits choices work best for you.[57]

1. Your options are important. There are many different types of health benefit plans. Find out which one your employer offers, then check out the plan, or plans, offered. Your employer's human resource office, the health plan administrator, or your union can provide information to help you match your needs and preferences with the available plans. The more information you have, the better your healthcare decisions will be.

2. Reviewing the benefits available. Do the plans offered cover preventive care, well-baby care, vision or dental care? Are there deductibles? Answers to these questions can help determine the out-of-pocket expenses you may

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[56] More information about quality across programs is provided at the following AHRQ Web site:
http://www.ahrq.gov/consumer/qntascii/qnthplan.htm.

[57] Adapted from the Department of Labor:
http://www.dol.gov/dol/pwba/public/pubs/health/top10-text.html.

face. Matching your needs and those of your family members will result in the best possible benefits. Cheapest may not always be best. Your goal is high quality health benefits.

3. Look for quality. The quality of healthcare services varies, but quality can be measured. You should consider the quality of healthcare in deciding among the healthcare plans or options available to you. Not all health plans, doctors, hospitals and other providers give the highest quality care. Fortunately, there is quality information you can use right now to help you compare your healthcare choices. Find out how you can measure quality. Consult the U.S. Department of Health and Human Services publication "Your Guide to Choosing Quality Health Care" on the Internet at **www.ahcpr.gov/consumer**.

4. Your plan's summary plan description (SPD) provides a wealth of information. Your health plan administrator can provide you with a copy of your plan's SPD. It outlines your benefits and your legal rights under the Employee Retirement Income Security Act (ERISA), the federal law that protects your health benefits. It should contain information about the coverage of dependents, what services will require a co-pay, and the circumstances under which your employer can change or terminate a health benefits plan. Save the SPD and all other health plan brochures and documents, along with memos or correspondence from your employer relating to health benefits.

5. Assess your benefit coverage as your family status changes. Marriage, divorce, childbirth or adoption, and the death of a spouse are all life events that may signal a need to change your health benefits. You, your spouse and dependent children may be eligible for a special enrollment period under provisions of the Health Insurance Portability and Accountability Act (HIPAA). Even without life-changing events, the information provided by your employer should tell you how you can change benefits or switch plans, if more than one plan is offered. If your spouse's employer also offers a health benefits package, consider coordinating both plans for maximum coverage.

6. Changing jobs and other life events can affect your health benefits. Under the Consolidated Omnibus Budget Reconciliation Act (COBRA), you, your covered spouse, and your dependent children may be eligible to purchase extended health coverage under your employer's plan if you lose your job, change employers, get divorced, or upon occurrence of certain other events. Coverage can range from 18 to 36 months depending on your situation. COBRA applies to most employers with 20 or more workers and

requires your plan to notify you of your rights. Most plans require eligible individuals to make their COBRA election within 60 days of the plan's notice. Be sure to follow up with your plan sponsor if you don't receive notice, and make sure you respond within the allotted time.

7. HIPAA can also help if you are changing jobs, particularly if you have a medical condition. HIPAA generally limits pre-existing condition exclusions to a maximum of 12 months (18 months for late enrollees). HIPAA also requires this maximum period to be reduced by the length of time you had prior "creditable coverage." You should receive a certificate documenting your prior creditable coverage from your old plan when coverage ends.

8. Plan for retirement. Before you retire, find out what health benefits, if any, extend to you and your spouse during your retirement years. Consult with your employer's human resources office, your union, the plan administrator, and check your SPD. Make sure there is no conflicting information among these sources about the benefits you will receive or the circumstances under which they can change or be eliminated. With this information in hand, you can make other important choices, like finding out if you are eligible for Medicare and Medigap insurance coverage.

9. Know how to file an appeal if your health benefits claim is denied. Understand how your plan handles grievances and where to make appeals of the plan's decisions. Keep records and copies of correspondence. Check your health benefits package and your SPD to determine who is responsible for handling problems with benefit claims. Contact PWBA for customer service assistance if you are unable to obtain a response to your complaint.

10. You can take steps to improve the quality of the healthcare and the health benefits you receive. Look for and use things like Quality Reports and Accreditation Reports whenever you can. Quality reports may contain consumer ratings -- how satisfied consumers are with the doctors in their plan, for instance-- and clinical performance measures -- how well a healthcare organization prevents and treats illness. Accreditation reports provide information on how accredited organizations meet national standards, and often include clinical performance measures. Look for these quality measures whenever possible. Consult "Your Guide to Choosing Quality Health Care" on the Internet at **www.ahcpr.gov/consumer**.

Medicare and Medicaid

Illness strikes both rich and poor families. For low-income families, Medicaid is available to defer the costs of treatment. The Health Care Financing Administration (HCFA) administers Medicare, the nation's largest health insurance program, which covers 39 million Americans. In the following pages, you will learn the basics about Medicare insurance as well as useful contact information on how to find more in-depth information about Medicaid.[58]

Who is Eligible for Medicare?

Generally, you are eligible for Medicare if you or your spouse worked for at least 10 years in Medicare-covered employment and you are 65 years old and a citizen or permanent resident of the United States. You might also qualify for coverage if you are under age 65 but have a disability or End-Stage Renal disease (permanent kidney failure requiring dialysis or transplant). Here are some simple guidelines:

You can get Part A at age 65 without having to pay premiums if:

- You are already receiving retirement benefits from Social Security or the Railroad Retirement Board.
- You are eligible to receive Social Security or Railroad benefits but have not yet filed for them.
- You or your spouse had Medicare-covered government employment.

If you are under 65, you can get Part A without having to pay premiums if:

- You have received Social Security or Railroad Retirement Board disability benefit for 24 months.
- You are a kidney dialysis or kidney transplant patient.

Medicare has two parts:

- Part A (Hospital Insurance). Most people do not have to pay for Part A.
- Part B (Medical Insurance). Most people pay monthly for Part B.

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[58] This section has been adapted from the Official U.S. Site for Medicare Information: **http://www.medicare.gov/Basics/Overview.asp**.

Part A (Hospital Insurance)

Helps Pay For: Inpatient hospital care, care in critical access hospitals (small facilities that give limited outpatient and inpatient services to people in rural areas) and skilled nursing facilities, hospice care, and some home healthcare.

Cost: Most people get Part A automatically when they turn age 65. You do not have to pay a monthly payment called a premium for Part A because you or a spouse paid Medicare taxes while you were working.

If you (or your spouse) did not pay Medicare taxes while you were working and you are age 65 or older, you still may be able to buy Part A. If you are not sure you have Part A, look on your red, white, and blue Medicare card. It will show "Hospital Part A" on the lower left corner of the card. You can also call the Social Security Administration toll free at 1-800-772-1213 or call your local Social Security office for more information about buying Part A. If you get benefits from the Railroad Retirement Board, call your local RRB office or 1-800-808-0772. For more information, call your Fiscal Intermediary about Part A bills and services. The phone number for the Fiscal Intermediary office in your area can be obtained from the following Web site: **http://www.medicare.gov/Contacts/home.asp**.

Part B (Medical Insurance)

Helps Pay For: Doctors, services, outpatient hospital care, and some other medical services that Part A does not cover, such as the services of physical and occupational therapists, and some home healthcare. Part B helps pay for covered services and supplies when they are medically necessary.

Cost: As of 2001, you pay the Medicare Part B premium of $50.00 per month. In some cases this amount may be higher if you did not choose Part B when you first became eligible at age 65. The cost of Part B may go up 10% for each 12-month period that you were eligible for Part B but declined coverage, except in special cases. You will have to pay the extra 10% cost for the rest of your life.

Enrolling in Part B is your choice. You can sign up for Part B anytime during a 7-month period that begins 3 months before you turn 65. Visit your local Social Security office, or call the Social Security Administration at 1-800-772-1213 to sign up. If you choose to enroll in Part B, the premium is usually taken out of your monthly Social Security, Railroad Retirement, or Civil Service Retirement payment. If you do not receive any of the above

payments, Medicare sends you a bill for your part B premium every 3 months. You should receive your Medicare premium bill in the mail by the 10th of the month. If you do not, call the Social Security Administration at 1-800-772-1213, or your local Social Security office. If you get benefits from the Railroad Retirement Board, call your local RRB office or 1-800-808-0772. For more information, call your Medicare carrier about bills and services. The phone number for the Medicare carrier in your area can be found at the following Web site: **http://www.medicare.gov/Contacts/home.asp**. You may have choices in how you get your healthcare including the Original Medicare Plan, Medicare Managed Care Plans (like HMOs), and Medicare Private Fee-for-Service Plans.

Medicaid

Medicaid is a joint federal and state program that helps pay medical costs for some people with low incomes and limited resources. Medicaid programs vary from state to state. People on Medicaid may also get coverage for nursing home care and outpatient prescription drugs which are not covered by Medicare. You can find more information about Medicaid on the HCFA.gov Web site at **http://www.hcfa.gov/medicaid/medicaid.htm**.

States also have programs that pay some or all of Medicare's premiums and may also pay Medicare deductibles and coinsurance for certain people who have Medicare and a low income. To qualify, you must have:

- Part A (Hospital Insurance),
- Assets, such as bank accounts, stocks, and bonds that are not more than $4,000 for a single person, or $6,000 for a couple, and
- A monthly income that is below certain limits.

For more information on these programs, look at the Medicare Savings Programs brochure, **http://www.medicare.gov/Library/PDFNavigation/PDFInterim.asp?Language=English&Type=Pub&PubID=10126**. There are also Prescription Drug Assistance Programs available. Find information on these programs which offer discounts or free medications to individuals in need at **http://www.medicare.gov/Prescription/Home.asp**.

NORD's Medication Assistance Programs

Finally, the National Organization for Rare Disorders, Inc. (NORD) administers medication programs sponsored by humanitarian-minded pharmaceutical and biotechnology companies to help uninsured or under-insured individuals secure life-saving or life-sustaining drugs.[59] NORD programs ensure that certain vital drugs are available "to those individuals whose income is too high to qualify for Medicaid but too low to pay for their prescribed medications." The program has standards for fairness, equity, and unbiased eligibility. It currently covers some 14 programs for nine pharmaceutical companies. NORD also offers early access programs for investigational new drugs (IND) under the approved "Treatment INDs" programs of the Food and Drug Administration (FDA). In these programs, a limited number of individuals can receive investigational drugs that have yet to be approved by the FDA. These programs are generally designed for rare diseases or disorders. For more information, visit **www.rarediseases.org**.

Additional Resources

In addition to the references already listed in this chapter, you may need more information on health insurance, hospitals, or the healthcare system in general. The NIH has set up an excellent guidance Web site that addresses these and other issues. Topics include:[60]

- Health Insurance:
  http://www.nlm.nih.gov/medlineplus/healthinsurance.html
- Health Statistics:
  http://www.nlm.nih.gov/medlineplus/healthstatistics.html
- HMO and Managed Care:
  http://www.nlm.nih.gov/medlineplus/managedcare.html
- Hospice Care: **http://www.nlm.nih.gov/medlineplus/hospicecare.html**
- Medicaid: **http://www.nlm.nih.gov/medlineplus/medicaid.html**
- Medicare: **http://www.nlm.nih.gov/medlineplus/medicare.html**
- Nursing Homes and Long-term Care:
  http://www.nlm.nih.gov/medlineplus/nursinghomes.html

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[59] Adapted from NORD: **http://www.rarediseases.org/cgi-bin/nord/progserv#patient?id=rPIzL9oD&mv_pc=30**.

[60] You can access this information at:
http://www.nlm.nih.gov/medlineplus/healthsystem.html.

- Patient's Rights, Confidentiality, Informed Consent, Ombudsman Programs, Privacy and Patient Issues:
  http://www.nlm.nih.gov/medlineplus/patientissues.html
- Veteran's Health, Persian Gulf War, Gulf War Syndrome, Agent Orange:
  http://www.nlm.nih.gov/medlineplus/veteranshealth.html

ONLINE GLOSSARIES

The Internet provides access to a number of free-to-use medical dictionaries and glossaries. The National Library of Medicine has compiled the following list of online dictionaries:

- ADAM Medical Encyclopedia (A.D.A.M., Inc.), comprehensive medical reference: **http://www.nlm.nih.gov/medlineplus/encyclopedia.html**
- MedicineNet.com Medical Dictionary (MedicineNet, Inc.): **http://www.medterms.com/Script/Main/hp.asp**
- Merriam-Webster Medical Dictionary (Inteli-Health, Inc.): **http://www.intelihealth.com/IH/**
- Multilingual Glossary of Technical and Popular Medical Terms in Eight European Languages (European Commission) - Danish, Dutch, English, French, German, Italian, Portuguese, and Spanish: **http://allserv.rug.ac.be/~rvdstich/eugloss/welcome.html**
- On-line Medical Dictionary (CancerWEB): **http://www.graylab.ac.uk/omd/**
- Technology Glossary (National Library of Medicine) - Health Care Technology: **http://www.nlm.nih.gov/nichsr/ta101/ta10108.htm**
- Terms and Definitions (Office of Rare Diseases): **http://rarediseases.info.nih.gov/ord/glossary_a-e.html**

Beyond these, MEDLINEplus contains a very user-friendly encyclopedia covering every aspect of medicine (licensed from A.D.A.M., Inc.). The ADAM Medical Encyclopedia Web site address is **http://www.nlm.nih.gov/medlineplus/encyclopedia.html**. ADAM is also available on commercial Web sites such as drkoop.com (**http://www.drkoop.com/**) and Web MD (**http://my.webmd.com/adam/asset/adam_disease_articles/a_to_z/a)**. Topics of interest can be researched by using keywords before continuing elsewhere, as these basic definitions and concepts will be useful in more advanced areas of research. You may choose to print various pages specifically relating to primary sclerosing cholangitis and keep them on file.

Online Dictionary Directories

The following are additional online directories compiled by the National Library of Medicine, including a number of specialized medical dictionaries and glossaries:

- Medical Dictionaries: Medical & Biological (World Health Organization): **http://www.who.int/hlt/virtuallibrary/English/diction.htm#Medical**
- MEL-Michigan Electronic Library List of Online Health and Medical Dictionaries (Michigan Electronic Library): **http://mel.lib.mi.us/health/health-dictionaries.html**
- Patient Education: Glossaries (DMOZ Open Directory Project): **http://dmoz.org/Health/Education/Patient_Education/Glossaries/**
- Web of Online Dictionaries (Bucknell University): **http://www.yourdictionary.com/diction5.html#medicine**

PRIMARY SCLEROSING CHOLANGITIS GLOSSARY

The following is a complete glossary of terms used in this sourcebook. The definitions are derived from official public sources including the National Institutes of Health [NIH] and the European Union [EU]. After this glossary, we list a number of additional hardbound and electronic glossaries and dictionaries that you may wish to consult.

Acetaminophen: Analgesic antipyretic derivative of acetanilide. It has weak anti-inflammatory properties and is used as a common analgesic, but may cause liver, blood cell, and kidney damage. [NIH]

Alimentary: Pertaining to food or nutritive material, or to the organs of digestion. [EU]

American Medical Association: Professional society representing the field of medicine. [NIH]

Anaemia: A reduction below normal in the number of erythrocytes per cu. mm., in the quantity of haemoglobin, or in the volume of packed red cells per 100 ml. of blood which occurs when the equilibrium between blood loss (through bleeding or destruction) and blood production is disturbed. [EU]

Antibiotic: A chemical substance produced by a microorganism which has the capacity, in dilute solutions, to inhibit the growth of or to kill other microorganisms. Antibiotics that are sufficiently nontoxic to the host are used as chemotherapeutic agents in the treatment of infectious diseases of man, animals and plants. [EU]

Antibody: An immunoglobulin molecule that has a specific amino acid sequence by virtue of which it interacts only with the antigen that induced its synthesis in cells of the lymphoid series (especially plasma cells), or with antigen closely related to it. Antibodies are classified according to their ode of action as agglutinins, bacteriolysins, haemolysins, opsonins, precipitins, etc. [EU]

Antigens: Substances that cause an immune response in the body. The body "sees" the antigens as harmful or foreign. To fight them, the body produces antibodies, which attack and try to eliminate the antigens. [NIH]

Antimicrobial: Killing microorganisms, or suppressing their multiplication or growth. [EU]

Appendicitis: Acute inflammation of the vermiform appendix. [NIH]

Ascites: Effusion and accumulation of serous fluid in the abdominal cavity;

called also abdominal or peritoneal dropsy, hydroperitonia, and hydrops abdominis. [EU]

Assay: Determination of the amount of a particular constituent of a mixture, or of the biological or pharmacological potency of a drug. [EU]

Asymptomatic: No symptoms; no clear sign of disease present. [NIH]

Atypical: Irregular; not conformable to the type; in microbiology, applied specifically to strains of unusual type. [EU]

Autoimmunity: Process whereby the immune system reacts against the body's own tissues. Autoimmunity may produce or be caused by autoimmune diseases. [NIH]

Bacteria: Unicellular prokaryotic microorganisms which generally possess rigid cell walls, multiply by cell division, and exhibit three principal forms: round or coccal, rodlike or bacillary, and spiral or spirochetal. [NIH]

Bile: An emulsifying agent produced in the liver and secreted into the duodenum. Its composition includes bile acids and salts, cholesterol, and electrolytes. It aids digestion of fats in the duodenum. [NIH]

Biliary: Pertaining to the bile, to the bile ducts, or to the gallbladder. [EU]

Bilirubin: A bile pigment that is a degradation product of HEME. [NIH]

Biochemical: Relating to biochemistry; characterized by, produced by, or involving chemical reactions in living organisms. [EU]

Biopsy: The removal and examination, usually microscopic, of tissue from the living body, performed to establish precise diagnosis. [EU]

Budesonide: A glucocorticoid used in the management of asthma, the treatment of various skin disorders, and allergic rhinitis. [NIH]

Calcitonin: A peptide hormone that lowers calcium concentration in the blood. In humans, it is released by thyroid cells and acts to decrease the formation and absorptive activity of osteoclasts. Its role in regulating plasma calcium is much greater in children and in certain diseases than in normal adults. [NIH]

Calmodulin: A heat-stable, low-molecular-weight activator protein found mainly in the brain and heart. The binding of calcium ions to this protein allows this protein to bind to cyclic nucleotide phosphodiesterases and to adenyl cyclase with subsequent activation. Thereby this protein modulates cyclic AMP and cyclic GMP levels. [NIH]

Capsules: Hard or soft soluble containers used for the oral administration of medicine. [NIH]

Carbohydrate: An aldehyde or ketone derivative of a polyhydric alcohol, particularly of the pentahydric and hexahydric alcohols. They are so named because the hydrogen and oxygen are usually in the proportion to form

water, (CH2O)n. The most important carbohydrates are the starches, sugars, celluloses, and gums. They are classified into mono-, di-, tri-, poly- and heterosaccharides. [EU]

Carcinoma: A malignant new growth made up of epithelial cells tending to infiltrate the surrounding tissues and give rise to metastases. [EU]

Catalase: An oxidoreductase that catalyzes the conversion of hydrogen peroxide to water and oxygen. It is present in many animal cells. A deficiency of this enzyme results in ACATALASIA. EC 1.11.1.6. [NIH]

Catheter: A tubular, flexible, surgical instrument for withdrawing fluids from (or introducing fluids into) a cavity of the body, especially one for introduction into the bladder through the urethra for the withdraw of urine. [EU]

Caustic: An escharotic or corrosive agent. Called also cauterant. [EU]

Cholangiography: Roentgenography of the biliary ducts after administration or injection of a contrast medium, orally, intravenously or percutaneously. [EU]

Cholangitis: Inflammation of a bile duct. [EU]

Cholecystectomy: Surgical removal of the gallbladder. [NIH]

Cholecystitis: Inflammation of the gallbladder. [EU]

Cholelithiasis: The presence or formation of gallstones. [EU]

Cholestasis: Impairment of biliary flow at any level from the hepatocyte to Vater's ampulla. [NIH]

Cholesterol: The principal sterol of all higher animals, distributed in body tissues, especially the brain and spinal cord, and in animal fats and oils. [NIH]

Chronic: Persisting over a long period of time. [EU]

Cirrhosis: Liver disease characterized pathologically by loss of the normal microscopic lobular architecture, with fibrosis and nodular regeneration. The term is sometimes used to refer to chronic interstitial inflammation of any organ. [EU]

Cladribine: An antineoplastic agent used in the treatment of lymphoproliferative diseases including hairy-cell leukemia. [NIH]

Colic: Paroxysms of pain. This condition usually occurs in the abdominal region but may occur in other body regions as well. [NIH]

Colitis: Inflammation of the colon. [EU]

Colonoscopy: Endoscopic examination, therapy or surgery of the luminal surface of the colon. [NIH]

Colorectal: Pertaining to or affecting the colon and rectum. [EU]

Concomitant: Accompanying; accessory; joined with another. [EU]

Constipation: Infrequent or difficult evacuation of the faeces. [EU]

Constriction: The act of constricting. [NIH]

Coronary: Encircling in the manner of a crown; a term applied to vessels; nerves, ligaments, etc. The term usually denotes the arteries that supply the heart muscle and, by extension, a pathologic involvement of them. [EU]

Crystallization: The formation of crystals; conversion to a crystalline form. [EU]

Cytokines: Non-antibody proteins secreted by inflammatory leukocytes and some non-leukocytic cells, that act as intercellular mediators. They differ from classical hormones in that they are produced by a number of tissue or cell types rather than by specialized glands. They generally act locally in a paracrine or autocrine rather than endocrine manner. [NIH]

Cytotoxic: Pertaining to or exhibiting cytotoxicity. [EU]

Degenerative: Undergoing degeneration : tending to degenerate; having the character of or involving degeneration; causing or tending to cause degeneration. [EU]

Diarrhea: Passage of excessively liquid or excessively frequent stools. [NIH]

Dilatation: The condition, as of an orifice or tubular structure, of being dilated or stretched beyond the normal dimensions. [EU]

Dyspepsia: Impairment of the power of function of digestion; usually applied to epigastric discomfort following meals. [EU]

Dysplasia: Abnormality of development; in pathology, alteration in size, shape, and organization of adult cells. [EU]

Encephalopathy: Any degenerative disease of the brain. [EU]

Endocrinology: A subspecialty of internal medicine concerned with the metabolism, physiology, and disorders of the endocrine system. [NIH]

Endoscopy: Visual inspection of any cavity of the body by means of an endoscope. [EU]

Enterohepatic: Of or involving the intestine and liver. [EU]

Enzyme: A protein molecule that catalyses chemical reactions of other substances without itself being destroyed or altered upon completion of the reactions. Enzymes are classified according to the recommendations of the Nomenclature Committee of the International Union of Biochemistry. Each enzyme is assigned a recommended name and an Enzyme Commission (EC) number. They are divided into six main groups; oxidoreductases, transferases, hydrolases, lyases, isomerases, and ligases. [EU]

Epidemic: Occurring suddenly in numbers clearly in excess of normal expectancy; said especially of infectious diseases but applied also to any disease, injury, or other health-related event occurring in such outbreaks. [EU]

Epithelium: The covering of internal and external surfaces of the body, including the lining of vessels and other small cavities. It consists of cells joined by small amounts of cementing substances. Epithelium is classified into types on the basis of the number of layers deep and the shape of the superficial cells. [EU]

Epitopes: Sites on an antigen that interact with specific antibodies. [NIH]

Extracellular: Outside a cell or cells. [EU]

Extraction: The process or act of pulling or drawing out. [EU]

Fatigue: The state of weariness following a period of exertion, mental or physical, characterized by a decreased capacity for work and reduced efficiency to respond to stimuli. [NIH]

Fats: One of the three main classes of foods and a source of energy in the body. Fats help the body use some vitamins and keep the skin healthy. They also serve as energy stores for the body. In food, there are two types of fats: saturated and unsaturated. [NIH]

Fibrosis: The formation of fibrous tissue; fibroid or fibrous degeneration [EU]

Fistula: An abnormal passage or communication, usually between two internal organs, or leading from an internal organ to the surface of the body; frequently designated according to the organs or parts with which it communicates, as anovaginal, brochocutaneous, hepatopleural, pulmonoperitoneal, rectovaginal, urethrovaginal, and the like. Such passages are frequently created experimentally for the purpose of obtaining body secretions for physiologic study. [EU]

Gastritis: Inflammation of the stomach. [EU]

Gastrointestinal: Pertaining to or communicating with the stomach and intestine, as a gastrointestinal fistula. [EU]

Gluten: The protein of wheat and other grains which gives to the dough its tough elastic character. [EU]

Haplotypes: The genetic constitution of individuals with respect to one member of a pair of allelic genes, or sets of genes that are closely linked and tend to be inherited together such as those of the major histocompatibility complex. [NIH]

Heartburn: Substernal pain or burning sensation, usually associated with regurgitation of gastric juice into the esophagus. [NIH]

Helicobacter: A genus of gram-negative, spiral-shaped bacteria that is pathogenic and has been isolated from the intestinal tract of mammals, including humans. [NIH]

Hematology: A subspecialty of internal medicine concerned with morphology, physiology, and pathology of the blood and blood-forming

tissues. [NIH]

Hemorrhage: Bleeding or escape of blood from a vessel. [NIH]

Hemorrhoids: Varicosities of the hemorrhoidal venous plexuses. [NIH]

Hepatitis: Inflammation of the liver. [EU]

Hepatobiliary: Pertaining to the liver and the bile or the biliary ducts. [EU]

Hepatocellular: Pertaining to or affecting liver cells. [EU]

Hepatocytes: The main structural component of the liver. They are specialized epithelial cells that are organized into interconnected plates called lobules. [NIH]

Hernia: (he protrusion of a loop or knuckle of an organ or tissue through an abnormal opening. [EU]

Histiocytosis: General term for the abnormal appearance of histiocytes in the blood. Based on the pathological features of the cells involved rather than on clinical findings, the histiocytic diseases are subdivided into three groups: histiocytosis, langerhans cell; histiocytosis, non-langerhans cell; and histiocytic disorders, malignant. [NIH]

Histocompatibility: The degree of antigenic similarity between the tissues of different individuals, which determines the acceptance or rejection of allografts. [NIH]

Humoral: Of, relating to, proceeding from, or involving a bodily humour - now often used of endocrine factors as opposed to neural or somatic. [EU]

Hybridization: The genetic process of crossbreeding to produce a hybrid. Hybrid nucleic acids can be formed by nucleic acid hybridization of DNA and RNA molecules. Protein Hybridization allows for hybrid proteins to be formed from polypeptide chains. [NIH]

Hypertension: Persistently high arterial blood pressure. Various criteria for its threshold have been suggested, ranging from 140 mm. Hg systolic and 90 mm. Hg diastolic to as high as 200 mm. Hg systolic and 110 mm. Hg diastolic. Hypertension may have no known cause (essential or idiopathic h.) or be associated with other primary diseases (secondary h.). [EU]

Idiopathic: Of the nature of an idiopathy; self-originated; of unknown causation. [EU]

Immunity: The condition of being immune; the protection against infectious disease conferred either by the immune response generated by immunization or previous infection or by other nonimmunologic factors (innate i.). [EU]

Immunization: The induction of immunity. [EU]

Immunosuppressant: An agent capable of suppressing immune responses. [EU]

Incidental: 1. small and relatively unimportant, minor; 2. accompanying, but not a major part of something; 3. (to something) liable to occur because of something or in connection with something (said of risks, responsibilities, ...) [EU]

Incontinence: Inability to control excretory functions, as defecation (faecal i.) or urination (urinary i.). [EU]

Infantile: Pertaining to an infant or to infancy. [EU]

Inflammation: A pathological process characterized by injury or destruction of tissues caused by a variety of cytologic and chemical reactions. It is usually manifested by typical signs of pain, heat, redness, swelling, and loss of function. [NIH]

Ingestion: The act of taking food, medicines, etc., into the body, by mouth. [EU]

Invasive: 1. having the quality of invasiveness. 2. involving puncture or incision of the skin or insertion of an instrument or foreign material into the body; said of diagnostic techniques. [EU]

Iodine: A nonmetallic element of the halogen group that is represented by the atomic symbol I, atomic number 53, and atomic weight of 126.90. It is a nutritionally essential element, especially important in thyroid hormone synthesis. In solution, it has anti-infective properties and is used topically. [NIH]

Ischemia: Deficiency of blood in a part, due to functional constriction or actual obstruction of a blood vessel. [EU]

Jaundice: A clinical manifestation of hyperbilirubinemia, consisting of deposition of bile pigments in the skin, resulting in a yellowish staining of the skin and mucous membranes. [NIH]

Lesion: Any pathological or traumatic discontinuity of tissue or loss of function of a part. [EU]

Ligation: Application of a ligature to tie a vessel or strangulate a part. [NIH]

Lipid: Any of a heterogeneous group of flats and fatlike substances characterized by being water-insoluble and being extractable by nonpolar (or fat) solvents such as alcohol, ether, chloroform, benzene, etc. All contain as a major constituent aliphatic hydrocarbons. The lipids, which are easily stored in the body, serve as a source of fuel, are an important constituent of cell structure, and serve other biological functions. Lipids may be considered to include fatty acids, neutral fats, waxes, and steroids. Compound lipids comprise the glycolipids, lipoproteins, and phospholipids. [EU]

Lipodystrophy: 1. any disturbance of fat metabolism. 2. a group of conditions due to defective metabolism of fat, resulting in the absence of subcutaneous fat, which may be congenital or acquired and partial or total.

Called also lipoatrophy and lipodystrophia. [EU]

Lithotripsy: The destruction of a calculus of the kidney, ureter, bladder, or gallbladder by physical forces, including crushing with a lithotriptor through a catheter. Focused percutaneous ultrasound and focused hydraulic shock waves may be used without surgery. Lithotripsy does not include the dissolving of stones by acids or litholysis. Lithotripsy by laser is lithotripsy, laser. [NIH]

Localization: 1. the determination of the site or place of any process or lesion. 2. restriction to a circumscribed or limited area. 3. prelocalization. [EU]

Malabsorption: Impaired intestinal absorption of nutrients. [EU]

Malignant: Tending to become progressively worse and to result in death. Having the properties of anaplasia, invasion, and metastasis; said of tumours. [EU]

Membrane: A thin layer of tissue which covers a surface, lines a cavity or divides a space or organ. [EU]

Methotrexate: An antineoplastic antimetabolite with immunosuppressant properties. It is an inhibitor of dihydrofolate reductase and prevents the formation of tetrahydrofolate, necessary for synthesis of thymidylate, an essential component of DNA. [NIH]

Molecular: Of, pertaining to, or composed of molecules : a very small mass of matter. [EU]

Motility: The ability to move spontaneously. [EU]

Nephrology: A subspecialty of internal medicine concerned with the anatomy, physiology, and pathology of the kidney. [NIH]

Nephropathy: Disease of the kidneys. [EU]

Neural: 1. pertaining to a nerve or to the nerves. 2. situated in the region of the spinal axis, as the neutral arch. [EU]

Neutrophils: Granular leukocytes having a nucleus with three to five lobes connected by slender threads of chromatin, and cytoplasm containing fine inconspicuous granules and stainable by neutral dyes. [NIH]

Niacin: Water-soluble vitamin of the B complex occurring in various animal and plant tissues. Required by the body for the formation of coenzymes NAD and NADP. Has pellagra-curative, vasodilating, and antilipemic properties. [NIH]

Nicotine: Nicotine is highly toxic alkaloid. It is the prototypical agonist at nicotinic cholinergic receptors where it dramatically stimulates neurons and ultimately blocks synaptic transmission. Nicotine is also important medically because of its presence in tobacco smoke. [NIH]

Nosocomial: Pertaining to or originating in the hospital, said of an infection

not present or incubating prior to admittance to the hospital, but generally occurring 72 hours after admittance; the term is usually used to refer to patient disease, but hospital personnel may also acquire nosocomial infection. [EU]

Orthopaedic: Pertaining to the correction of deformities of the musculoskeletal system; pertaining to orthopaedics. [EU]

Overdose: 1. to administer an excessive dose. 2. an excessive dose. [EU]

Paediatric: Of or relating to the care and medical treatment of children; belonging to or concerned with paediatrics. [EU]

Palliative: 1. affording relief, but not cure. 2. an alleviating medicine. [EU]

Pancreas: An organ behind the lower part of the stomach that is about the size of a hand. It makes insulin so that the body can use glucose (sugar) for energy. It also makes enzymes that help the body digest food. Spread all over the pancreas are areas called the islets of Langerhans. The cells in these areas each have a special purpose. The alpha cells make glucagon, which raises the level of glucose in the blood; the beta cells make insulin; the delta cells make somatostatin. There are also the PP cells and the D1 cells, about which little is known. [NIH]

Pancreatitis: Inflammation (pain, tenderness) of the pancreas; it can make the pancreas stop working. It is caused by drinking too much alcohol, by disease in the gallbladder, or by a virus. [NIH]

Parasitic: Pertaining to, of the nature of, or caused by a parasite. [EU]

Parenteral: Not through the alimentary canal but rather by injection through some other route, as subcutaneous, intramuscular, intraorbital, intracapsular, intraspinal, intrasternal, intravenous, etc. [EU]

Pathogen: Any disease-producing microorganism. [EU]

Penicillamine: 3-Mercapto-D-valine. The most characteristic degradation product of the penicillin antibiotics. It is used as an antirheumatic and as a chelating agent in Wilson's disease. [NIH]

Pentoxifylline: A methylxanthine derivative that inhibits phosphodiesterase and affects blood rheology. It improves blood flow by increasing erythrocyte and leukocyte flexibility. It also inhibits platelet aggregation. Pentoxifylline modulates immunologic activity by stimulating cytokine production. [NIH]

Peptic: Pertaining to pepsin or to digestion; related to the action of gastric juices. [EU]

Percutaneous: Performed through the skin, as injection of radiopacque material in radiological examination, or the removal of tissue for biopsy accomplished by a needle. [EU]

Perinatal: Pertaining to or occurring in the period shortly before and after

birth; variously defined as beginning with completion of the twentieth to twenty-eighth week of gestation and ending 7 to 28 days after birth. [EU]

Peritonitis: Inflammation of the peritoneum; a condition marked by exudations in the peritoneum of serum, fibrin, cells, and pus. It is attended by abdominal pain and tenderness, constipation, vomiting, and moderate fever. [EU]

Pharmacist: A person trained to prepare and distribute medicines and to give information about them. [NIH]

Pharmacologic: Pertaining to pharmacology or to the properties and reactions of drugs. [EU]

Porphyria: A pathological state in man and some lower animals that is often due to genetic factors, is characterized by abnormalities of porphyrin metabolism, and results in the excretion of large quantities of porphyrins in the urine and in extreme sensitivity to light. [EU]

Postoperative: Occurring after a surgical operation. [EU]

Potassium: An element that is in the alkali group of metals. It has an atomic symbol K, atomic number 19, and atomic weight 39.10. It is the chief cation in the intracellular fluid of muscle and other cells. Potassium ion is a strong electrolyte and it plays a significant role in the regulation of fluid volume and maintenance of the water-electrolyte balance. [NIH]

Predisposition: A latent susceptibility to disease which may be activated under certain conditions, as by stress. [EU]

Prednisone: A synthetic anti-inflammatory glucocorticoid derived from cortisone. It is biologically inert and converted to prednisolone in the liver. [NIH]

Prevalence: The number of people in a given group or population who are reported to have a disease. [NIH]

Progressive: Advancing; going forward; going from bad to worse; increasing in scope or severity. [EU]

Prosthesis: A man-made substitute for a missing body part such as an arm or a leg; also an implant such as for the hip. [NIH]

Proteins: Polymers of amino acids linked by peptide bonds. The specific sequence of amino acids determines the shape and function of the protein. [NIH]

Pruritus: Itching skin; may be a symptom of diabetes. [NIH]

Receptor: 1. a molecular structure within a cell or on the surface characterized by (1) selective binding of a specific substance and (2) a specific physiologic effect that accompanies the binding, e.g., cell-surface receptors for peptide hormones, neurotransmitters, antigens, complement fragments, and immunoglobulins and cytoplasmic receptors for steroid

hormones. 2. a sensory nerve terminal that responds to stimuli of various kinds. [EU]

Recurrence: The return of a sign, symptom, or disease after a remission. [NIH]

Reflux: A backward or return flow. [EU]

Remission: A diminution or abatement of the symptoms of a disease; also the period during which such diminution occurs. [EU]

Respiratory: Pertaining to respiration. [EU]

Retrograde: 1. moving backward or against the usual direction of flow. 2. degenerating, deteriorating, or catabolic. [EU]

Riboflavin: Nutritional factor found in milk, eggs, malted barley, liver, kidney, heart, and leafy vegetables. The richest natural source is yeast. It occurs in the free form only in the retina of the eye, in whey, and in urine; its principal forms in tissues and cells are as FMN and FAD. [NIH]

Sarcoidosis: An idiopathic systemic inflammatory granulomatous disorder comprised of epithelioid and multinucleated giant cells with little necrosis. It usually invades the lungs with fibrosis and may also involve lymph nodes, skin, liver, spleen, eyes, phalangeal bones, and parotid glands. [NIH]

Sclerotherapy: Treatment of varicose veins, hemorrhoids, gastric and esophageal varices, and peptic ulcer hemorrhage by injection or infusion of chemical agents which cause localized thrombosis and eventual fibrosis and obliteration of the vessels. [NIH]

Secretion: 1. the process of elaborating a specific product as a result of the activity of a gland; this activity may range from separating a specific substance of the blood to the elaboration of a new chemical substance. 2. any substance produced by secretion. [EU]

Sedimentation: The act of causing the deposit of sediment, especially by the use of a centrifugal machine. [EU]

Selenium: An element with the atomic symbol Se, atomic number 34, and atomic weight 78.96. It is an essential micronutrient for mammals and other animals but is toxic in large amounts. Selenium protects intracellular structures against oxidative damage. It is an essential component of glutathione peroxidase. [NIH]

Serum: The clear portion of any body fluid; the clear fluid moistening serous membranes. 2. blood serum; the clear liquid that separates from blood on clotting. 3. immune serum; blood serum from an immunized animal used for passive immunization; an antiserum; antitoxin, or antivenin. [EU]

Species: A taxonomic category subordinate to a genus (or subgenus) and superior to a subspecies or variety, composed of individuals possessing common characters distinguishing them from other categories of individuals

of the same taxonomic level. In taxonomic nomenclature, species are designated by the genus name followed by a Latin or Latinized adjective or noun. [EU]

Spectrum: A charted band of wavelengths of electromagnetic vibrations obtained by refraction and diffraction. By extension, a measurable range of activity, such as the range of bacteria affected by an antibiotic (antibacterial s.) or the complete range of manifestations of a disease. [EU]

Subclinical: Without clinical manifestations; said of the early stage(s) of an infection or other disease or abnormality before symptoms and signs become apparent or detectable by clinical examination or laboratory tests, or of a very mild form of an infection or other disease or abnormality. [EU]

Systemic: Pertaining to or affecting the body as a whole. [EU]

Thermoregulation: Heat regulation. [EU]

Thyroxine: An amino acid of the thyroid gland which exerts a stimulating effect on thyroid metabolism. [NIH]

Topical: Pertaining to a particular surface area, as a topical anti-infective applied to a certain area of the skin and affecting only the area to which it is applied. [EU]

Toxicity: The quality of being poisonous, especially the degree of virulence of a toxic microbe or of a poison. [EU]

Toxicology: The science concerned with the detection, chemical composition, and pharmacologic action of toxic substances or poisons and the treatment and prevention of toxic manifestations. [NIH]

Toxin: A poison; frequently used to refer specifically to a protein produced by some higher plants, certain animals, and pathogenic bacteria, which is highly toxic for other living organisms. Such substances are differentiated from the simple chemical poisons and the vegetable alkaloids by their high molecular weight and antigenicity. [EU]

Transdermal: Entering through the dermis, or skin, as in administration of a drug applied to the skin in ointment or patch form. [EU]

Transplantation: The grafting of tissues taken from the patient's own body or from another. [EU]

Urology: A surgical specialty concerned with the study, diagnosis, and treatment of diseases of the urinary tract in both sexes and the genital tract in the male. It includes the specialty of andrology which addresses both male genital diseases and male infertility. [NIH]

Vascular: Pertaining to blood vessels or indicative of a copious blood supply. [EU]

Veins: The vessels carrying blood toward the heart. [NIH]

Viral: Pertaining to, caused by, or of the nature of virus. [EU]

Viruses: Minute infectious agents whose genomes are composed of DNA or RNA, but not both. They are characterized by a lack of independent metabolism and the inability to replicate outside living host cells. [NIH]

General Dictionaries and Glossaries

While the above glossary is essentially complete, the dictionaries listed here cover virtually all aspects of medicine, from basic words and phrases to more advanced terms (sorted alphabetically by title; hyperlinks provide rankings, information and reviews at Amazon.com):

- **Dictionary of Medical Acronymns & Abbreviations** by Stanley Jablonski (Editor), Paperback, 4th edition (2001), Lippincott Williams & Wilkins Publishers, ISBN: 1560534605, **http://www.amazon.com/exec/obidos/ASIN/1560534605/icongroupinterna**
- **Dictionary of Medical Terms : For the Nonmedical Person (Dictionary of Medical Terms for the Nonmedical Person, Ed 4)** by Mikel A. Rothenberg, M.D, et al, Paperback - 544 pages, 4th edition (2000), Barrons Educational Series, ISBN: 0764112015, **http://www.amazon.com/exec/obidos/ASIN/0764112015/icongroupinterna**
- **A Dictionary of the History of Medicine** by A. Sebastian, CD-Rom edition (2001), CRC Press-Parthenon Publishers, ISBN: 185070368X, **http://www.amazon.com/exec/obidos/ASIN/185070368X/icongroupinterna**
- **Dorland's Illustrated Medical Dictionary (Standard Version)** by Dorland, et al, Hardcover - 2088 pages, 29th edition (2000), W B Saunders Co, ISBN: 0721662544, **http://www.amazon.com/exec/obidos/ASIN/0721662544/icongroupinterna**
- **Dorland's Electronic Medical Dictionary** by Dorland, et al, Software, 29th Book & CD-Rom edition (2000), Harcourt Health Sciences, ISBN: 0721694934, **http://www.amazon.com/exec/obidos/ASIN/0721694934/icongroupinterna**
- **Dorland's Pocket Medical Dictionary (Dorland's Pocket Medical Dictionary, 26th Ed)** Hardcover - 912 pages, 26th edition (2001), W B Saunders Co, ISBN: 0721682812, **http://www.amazon.com/exec/obidos/ASIN/0721682812/icongroupinterna/103-4193558-7304618**
- **Melloni's Illustrated Medical Dictionary (Melloni's Illustrated Medical Dictionary, 4th Ed)** by Melloni, Hardcover, 4th edition (2001), CRC Press-

Parthenon Publishers, ISBN: 85070094X, **http://www.amazon.com/exec/obidos/ASIN/85070094X/icongroupinterna**

- **Stedman's Electronic Medical Dictionary Version 5.0 (CD-ROM for Windows and Macintosh, Individual)** by Stedmans, CD-ROM edition (2000), Lippincott Williams & Wilkins Publishers, ISBN: 0781726328, **http://www.amazon.com/exec/obidos/ASIN/0781726328/icongroupinterna**
- **Stedman's Medical Dictionary** by Thomas Lathrop Stedman, Hardcover - 2098 pages, 27th edition (2000), Lippincott, Williams & Wilkins, ISBN: 068340007X, **http://www.amazon.com/exec/obidos/ASIN/068340007X/icongroupinterna**
- **Tabers Cyclopedic Medical Dictionary (Thumb Index)** by Donald Venes (Editor), et al, Hardcover - 2439 pages, 19th edition (2001), F A Davis Co, ISBN: 0803606540, **http://www.amazon.com/exec/obidos/ASIN/0803606540/icongroupinterna**

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