

THE OFFICIAL
PARENT'S SOURCEBOOK

on

MOYAMOYA
DISEASE

PREPARED FOR DAREN JOHNSON



JAMES N. PARKER, M.D.
AND PHILIP M. PARKER, PH.D., EDITORS

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Dedication

To the healthcare professionals dedicating their time and efforts to the study of Moyamoya disease.

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 Moyamoya disease. All of the *Official Parent'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

In addition to Moyamoya disease, *Official Parent's Sourcebooks* are available for the following related topics:

- The Official Patient's Sourcebook on Adrenoleukodystrophy
- The Official Patient's Sourcebook on Alexander Disease
- The Official Patient's Sourcebook on Alpers Disease
- The Official Patient's Sourcebook on Batten Disease
- The Official Patient's Sourcebook on Canavan Disease
- The Official Patient's Sourcebook on Coffin Lowry Syndrome
- The Official Patient's Sourcebook on Friedreich Ataxia
- The Official Patient's Sourcebook on Incontinentia Pigmenti
- The Official Patient's Sourcebook on Infantile Refsum Disease
- The Official Patient's Sourcebook on Joubert Syndrome
- The Official Patient's Sourcebook on Krabbe Disease
- The Official Patient's Sourcebook on Mobius Syndrome
- The Official Patient's Sourcebook on Neurofibromatoses
- The Official Patient's Sourcebook on Rett Syndrome
- The Official Patient's Sourcebook on Soto's Syndrome
- The Official Patient's Sourcebook on Spinal Muscular Atrophy
- The Official Patient's Sourcebook on Tourette Syndrome
- The Official Patient's Sourcebook on Williams 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 all parents incorporate education into the treatment process. According to the AHRQ:

Finding out more about your [child’s] condition is a good place to start. By contacting groups that support your [child’s] condition, visiting your local library, and searching on the Internet, you can find good information to help guide your decisions for your [child’s] treatment. 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 parents 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

¹ Quotation from <http://www.drkoop.com>.

² The Agency for Healthcare Research and Quality (AHRQ):
<http://www.ahrq.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 parent Internet usage rates. Parents frequently enter their children's 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 children through sound therapies. *The Official Parent's Sourcebook on Moyamoya Disease* has been created for parents 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 Moyamoya disease, 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 Moyamoya disease.

Given parents' 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 Moyamoya disease should affirm that a specific diagnostic procedure or treatment discussed in a research study, patent, or doctoral dissertation is "correct" or your child's best option. This sourcebook is no exception. Each child is unique. Deciding on

appropriate options is always up to parents in consultation with their children's physicians and healthcare providers.

Organization

This sourcebook is organized into three parts. Part I explores basic techniques to researching Moyamoya disease (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 parent networks dedicated to Moyamoya disease. It also gives you sources of information that can help you find a doctor in your local area specializing in treating Moyamoya disease. Collectively, the material presented in Part I is a complete primer on basic research topics for Moyamoya disease.

Part II moves on to advanced research dedicated to Moyamoya disease. 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 Moyamoya disease. 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 covering Moyamoya disease or related disorders. The appendices are dedicated to more pragmatic issues facing parents. Accessing materials via medical libraries may be the only option for some parents, 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 children with Moyamoya disease and their families.

Scope

While this sourcebook covers Moyamoya disease, doctors, research publications, and specialists may refer to your child's condition using a variety of terms. Therefore, you should understand that Moyamoya disease is often considered a synonym or a condition closely related to the following:

- Moya-moya Disease

In addition to synonyms and related conditions, physicians may refer to Moyamoya disease 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 Moyamoya disease:⁴

- 437.5 moyamoya disease

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 Moyamoya disease. 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 parents, 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.

Why "Internet age"? When their child has been diagnosed with Moyamoya disease, parents will often log on to the Internet, type words into a search engine, and receive several Web site listings which are mostly irrelevant or redundant. Parents are left to wonder where the relevant information is, and how to obtain it. Since only the smallest fraction of information dealing with Moyamoya disease is even indexed in search engines, a non-systematic

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."

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 Moyamoya disease, there is, of course, the emotional side to consider. Later in the sourcebook, we provide a chapter dedicated to helping you find parent groups and associations that can provide additional support beyond research produced by medical science. We hope that the choices we have made give you and your child the most options in moving forward. In this way, we wish you the best in your efforts to incorporate this educational approach into your child's 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 Moyamoya disease. The essentials typically include a definition or description of the condition, a discussion of who it affects, the signs or symptoms, tests or diagnostic procedures, and treatments for disease. Your child’s doctor or healthcare provider may have already explained the essentials of Moyamoya disease to you or even given you a pamphlet or brochure describing the condition. 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 the 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 MOYAMOYA DISEASE: GUIDELINES

Overview

Official agencies, as well as federally-funded institutions supported by national grants, frequently publish a variety of guidelines on Moyamoya disease. 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 parent in mind. Since new guidelines on Moyamoya disease 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 guidelines and fact sheets on Moyamoya disease. 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. Among them are 97 scientists who have won the Nobel Prize for achievement in medicine.

⁵ Adapted from the NIH: <http://www.nih.gov/about/NIHoverview.html>.

There is no guarantee that any one Institute will have a guideline on a specific medical condition, though the National Institutes of Health collectively publish over 600 guidelines for both common and rare disorders. 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 Moyamoya disease 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 Neurological Disorders and Stroke (NINDS); http://www.ninds.nih.gov/health_and_medical/disorder_index.htm

Among the above, the National Institute of Neurological Disorders and Stroke (NINDS) is particularly noteworthy. The mission of the NINDS is to reduce the burden of neurological disease—a burden borne by every age group, by every segment of society, by people all over the world.⁶ To support this mission, the NINDS conducts, fosters, coordinates, and guides research on the causes, prevention, diagnosis, and treatment of neurological disorders and stroke, and supports basic research in related scientific areas. The following patient guideline was recently published by the NINDS on Moyamoya disease.

What Is Moyamoya Disease?⁷

Moyamoya disease is a rare, progressive cerebrovascular disorder characterized by the narrowing or occlusion of major blood vessels leading into the brain, and the formation of abnormal blood vessels called *moyamoya vessels*. First described in Japan in the 1960's, the disease has since been found in the United States, Europe, Australia, and Africa. It primarily affects children, adolescents, and young adults, although it has also been seen in people beyond these ages. Females are more frequently affected than males. The cause of the disease is unknown. Researchers suspect a genetic link

⁶ This paragraph has been adapted from the NINDS:

http://www.ninds.nih.gov/about_ninds/mission.htm. “Adapted” signifies that a passage has been reproduced exactly or slightly edited for this book.

⁷ Adapted from The National Institute of Neurological Disorders and Stroke (NINDS): http://www.ninds.nih.gov/health_and_medical/disorders/moyamoya.htm.

because of the 9 percent incidence of the disease found in certain Japanese families. A gene for familial moyamoya disease has been located on chromosome 17q25 and further study of the gene may reveal the cause of the disorder. The name “moyamoya” is Japanese for “cloud of smoke” and was chosen to describe the classic appearance of the abnormal vessels seen in diagnostic tests. Children often present with stroke or recurrent transient ischemic attacks (TIAs), frequently accompanied by hemiparesis (muscular weakness or paralysis affecting one side of the body) or seizures. Adults most often experience stroke (often a hemorrhage). Both children and adults may have disturbed consciousness, speech deficits (aphasia), sensory and cognitive impairments, involuntary movements, and vision problems. Generally, cerebral angiography—a procedure that involves injecting dye into the cerebral arteries before taking an x-ray to make the arteries easier to see—is used to diagnose moyamoya diseases

Is There Any Treatment?

There is no cure for moyamoya disease. Treatment is symptomatic and supportive. Individuals experiencing TIAs and stroke may be given aspirin, vasodilators, or anticoagulants to reduce the risk of future attacks. There are several different types of revascularization (restoration of blood supply) surgery that may be performed in some cases. Children usually respond better to revascularization surgery than adults.

What Is the Prognosis?

Progressive deterioration of cognitive function is seen in the majority of individuals with moyamoya disease. Death usually results from intracerebral hemorrhage.

What Research Is Being Done?

Within the NINDS research programs, moyamoya disease is addressed primarily through studies associated with TIA and stroke. NINDS is the leading supporter of research on stroke and TIA in the United States and sponsors studies ranging from clinical trials to investigations of basic biological mechanisms.

For More Information

For more information, contact:

National Rehabilitation Information Center (NARIC)

1010 Wayne Avenue
Suite 800
Silver Spring, MD 20910-5633
naricinfo@kra.com
<http://www.naric.com>
Tel: 301-562-2400 / 800-346-2742
Fax: 301-562-2401

National Organization for Rare Disorders (NORD)

P.O. Box 8923
(100 Route 37)
New Fairfield, CT 06812-8923
orphan@rarediseases.org
<http://www.rarediseases.org>
Tel: 203-746-6518 / 800-999-NORD (6673)
Fax: 203-746-6481

National Stroke Association

9707 East Easter Lane
Englewood, CO 80112-3747
info@stroke.org
<http://www.stroke.org>
Tel: 303-649-9299 / 800-STROKES (787-6537)
Fax: 303-649-1328

More Guideline Sources

The guideline above on Moyamoya disease 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 Moyamoya disease. Many of the guidelines listed below address topics that may be of particular relevance to your child's specific situation, while certain guidelines will apply to only some children with Moyamoya disease. 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 parents wishing to go beyond guidelines published by specific Institutes of the NIH, the National Library of Medicine has created a vast and parent-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>.

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 <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 parent-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.

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:

- **An Introduction to Genetics and Genetic Testing**

Summary: This online fact sheet answers consumers questions about genetic disorders and metabolism. It discusses genetic counseling, prenatal diagnoses and X-linked disorders.

Source: Nemours Foundation

<http://www.healthfinder.gov/scripts/recordpass.asp?RecordType=0&RecordID=2269>

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 Moyamoya disease. 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 parents. 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:

Angiography: Radiography of blood vessels after injection of a contrast

medium. [NIH]

Anticoagulants: Agents that prevent blood clotting. Naturally occurring agents in the blood are included only when they are used as drugs. [NIH]

Aphasia: Defect or loss of the power of expression by speech, writing, or signs, or of comprehending spoken or written language, due to injury or disease of the brain centres. [EU]

Arteries: The vessels carrying blood away from the heart. [NIH]

Cerebral: Of or pertaining of the cerebrum or the brain. [EU]

Cerebrovascular: Pertaining to the blood vessels of the cerebrum, or brain. [EU]

Consciousness: Sense of awareness of self and of the environment. [NIH]

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

Paralysis: Loss or impairment of motor function in a part due to lesion of the neural or muscular mechanism; also by analogy, impairment of sensory function (sensory paralysis). In addition to the types named below, paralysis is further distinguished as traumatic, syphilitic, toxic, etc., according to its cause; or as obturator, ulnar, etc., according to the nerve part, or muscle specially affected. [EU]

Prenatal: Existing or occurring before birth, with reference to the fetus. [EU]

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

Seizures: Clinical or subclinical disturbances of cortical function due to a sudden, abnormal, excessive, and disorganized discharge of brain cells. Clinical manifestations include abnormal motor, sensory and psychic phenomena. Recurrent seizures are usually referred to as epilepsy or "seizure disorder." [NIH]

Symptomatic: 1. pertaining to or of the nature of a symptom. 2. indicative (of a particular disease or disorder). 3. exhibiting the symptoms of a particular disease but having a different cause. 4. directed at the allaying of symptoms, as symptomatic treatment. [EU]

Venereal: Pertaining or related to or transmitted by sexual contact. [EU]

CHAPTER 2. SEEKING GUIDANCE

Overview

Some parents are comforted by the knowledge that a number of organizations dedicate their resources to helping people with Moyamoya disease. These associations can become invaluable sources of information and advice. Many associations offer parent 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 child's physician can be a valuable source of guidance and support.

In this chapter, we direct you to resources that can help you find parent organizations and medical specialists. We begin by describing how to find associations and parent groups that can help you better understand and cope with your child's condition. The chapter ends with a discussion on how to find a doctor that is right for your child.

Associations and Moyamoya Disease

In addition to associations or groups that your child's 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 child's insurance provider to find out if the cost will be covered):

⁸ Churches, synagogues, and other houses of worship might also have groups that can offer you the social support you need.

- **Families with Moyamoya Support Network**

Address: Families with Moyamoya Support Network 4900 McGowan Street SE, Cedar Rapids, IA 52403

Telephone: (817) 261-6003 Toll-free: (800) 433-5255

Background: Families with Moyamoya Support Network is a not-for-profit voluntary health organization composed of families who are affected by Moyamoya Disease. Moyamoya Disease is a progressive disease that affects the blood vessels in the brain (cerebrovascular). The disorder is characterized by narrowing and/or closing of the main artery to the brain (carotid). This lack of blood may cause paralysis of the feet, legs, or upper extremities. Headaches, various vision problems, mental retardation, and psychiatric problems may also occur. The Network seeks to share information and support to help affected individuals and families cope with the impact of this rare disease. Established in 1992, the organization provides a forum through which members can receive emotional support and learn about Moyamoya Disease, treatment options, and sources of treatment. Educational materials produced by the organization include a bibliographic list of medical journal articles about Moyamoya Disease and brochures.

Relevant area(s) of interest: Moyamoya Disease

- **March of Dimes Birth Defects Foundation**

Address: March of Dimes Birth Defects Foundation 1275 Mamaroneck Avenue, White Plains, NY 10605

Telephone: (914) 428-7100 Toll-free: (888) 663-4637

Fax: (914) 997-4763

Email: resourcecenter@modimes.org

Web Site: <http://www.modimes.org>

Background: The March of Dimes Birth Defects Foundation is a national not-for-profit organization that was established in 1938. The mission of the Foundation is to improve the health of babies by preventing birth defects and infant mortality. Through the Campaign for Healthier Babies, the March of Dimes funds programs of research, community services, education, and advocacy. Educational programs that seek to prevent birth defects are important to the Foundation and to that end it produces a wide variety of printed informational materials and videos. The March of Dimes public health educational materials provide information encouraging health-enhancing behaviors that lead to a healthy pregnancy and a healthy baby.

Relevant area(s) of interest: Adrenoleukodystrophy, Coffin Lowry Syndrome, Incontinentia Pigmenti, Joubert Syndrome, Moyamoya Disease, Rett Syndrome, Tourette Syndrome, Williams Syndrome

- **The Arc (a national organization on mental retardation)**

Address: The Arc (a national organization on mental retardation) 500 East Border Street, Suite 300, Arlington, TX 76010

Telephone: (817) 261-6003 Toll-free: (800) 433-5255

Fax: (817) 277-3491

Email: thearc@metronet.com

Web Site: <http://thearc.org/>

Background: The Arc is the largest organization in the United States that is solely devoted to improving the lives of all children and adults with mental retardation. The organization offers support to families affected by mental retardation and fosters research and educational programs on the prevention of mental retardation. The Arc is committed to securing opportunities for all people with mental retardation. To this end, the organization emphasizes personal opportunities for choice in education, housing, employment, and entertainment. The Arc is further committed to reducing the incidence and limiting the consequences of mental retardation through research, advocacy, and mutual support. The Arc provides leadership in the field of mental retardation and develops necessary human and financial resources to attain its goals. In addition, the Arc provides a wide variety of educational materials for parents, teachers, health care professionals, and others, including a regular newsletter, handbooks, instruction packets, reports, booklets, audio-visual aids, posters, and brochures. Many materials are available in Spanish.

Relevant area(s) of interest: Adrenoleukodystrophy, Coffin Lowry Syndrome, Joubert Syndrome, Moyamoya Disease, Rett Syndrome, Williams Syndrome

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 parent 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 Moyamoya disease. 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 "Moyamoya disease" (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 "Moyamoya disease". 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 "Moyamoya disease" (or synonyms) into the "For these words:" box, you will only receive results on organizations dealing with Moyamoya disease. 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 medical conditions. 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 “Moyamoya disease” (or a synonym) in the search box.

Online Support Groups

In addition to support groups, commercial Internet service providers offer forums and chat rooms to discuss different illnesses and conditions. WebMD[®], for example, offers such a service at their Web site: <http://boards.webmd.com/roundtable>. These online 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 child’s doctor or other healthcare providers, as the treatments or discoveries you hear about may not be scientifically proven to be safe and effective.

Finding Doctors

All parents must go through the process of selecting a physician for their children with Moyamoya disease. While this process will vary, the Agency for Healthcare Research and Quality makes a number of suggestions, including the following:⁹

- If your child is 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.

⁹ This section is adapted from the AHRQ: www.ahrq.gov/consumer/qntascii/qntdr.htm.

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 child’s 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>.¹⁰ 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 expertise in various rare medical conditions. The Metabolic Information Network (MIN), 800-945-2188, also maintains a database of physicians with expertise in various metabolic diseases.

Finding a Neurologist

The American Academy of Neurology allows you to search for member neurologists by name or location. To use this service, go to <http://www.aan.com/>, select “Find a Neurologist” from the toolbar. Enter your search criteria, and click “Search.” To find out more information on a particular neurologist, click on the physician’s name.

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 expertise in various rare diseases. The Metabolic Information Network

¹⁰ 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.

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

Selecting Your Doctor¹¹

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

Working with Your Child's Doctor¹²

Research has shown that parents who have good relationships with their children's doctors tend to be more satisfied with their children's care. Here are some tips to help you and your child's doctor become partners:

- You know important things about your child's symptoms and health history. Tell the doctor what you think he or she needs to know.
- Always bring any medications your child is currently taking with you to the appointment, or you can bring a list of your child's medications including dosage and frequency information. Talk about any allergies or reactions your child has had to medications.
- Tell your doctor about any natural or alternative medicines your child is taking.
- Bring other medical information, such as x-ray films, test results, and medical records.
- Ask questions. If you don't, the doctor will assume that you understood everything that was said.
- Write down your questions before the doctor's visit. List the most important ones first to make sure that they are addressed.
- Ask the doctor to draw pictures if you think that this will help you and your child understand.

¹¹ This section has been adapted from the AHRQ: www.ahrq.gov/consumer/qntascii/qntdr.htm.

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

- Take notes. Some doctors do not mind if you bring a tape recorder to help you remember things, but always ask first.
- Take information home. Ask for written instructions. Your child's doctor may also have brochures and audio and videotapes on Moyamoya disease.

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

Broader Health-Related Resources

In addition to the references above, the NIH has set up guidance Web sites that can help parents find healthcare professionals. These include:¹³

- 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:

Neurology: A medical specialty concerned with the study of the structures, functions, and diseases of the nervous system. [NIH]

Psychiatric: Pertaining to or within the purview of psychiatry. [EU]

¹³ You can access this information at:
<http://www.nlm.nih.gov/medlineplus/healthsystem.html>.

PART II: ADDITIONAL RESOURCES AND ADVANCED MATERIAL

ABOUT PART II

In Part II, we introduce you to additional resources and advanced research on Moyamoya disease. All too often, parents 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 Moyamoya disease. In Part II, as in Part I, our objective is not to interpret the latest advances on Moyamoya disease 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 Moyamoya disease is suggested.

CHAPTER 3. STUDIES ON MOYAMOYA DISEASE

Overview

Every year, academic studies are published on Moyamoya disease 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 Moyamoya disease. 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 Moyamoya disease 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 Moyamoya disease, 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 "Moyamoya disease" (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:

- **Pathologic Basis of the Symptomatic Epilepsies in Childhood**

Source: *Journal of Child Neurology*. 13(8):361-371, August 1998.

Summary: The authors summarize the etiologies of the symptomatic epilepsies of childhood. The etiologies are (1) malformative, (2) metabolic, (3) neoplastic, (4) hypoxic-ischemic, (5) infectious, and (6) unknown. Malformative etiologies include (1) lissencephaly, types I through IV; (2) regional cortical dysplasia, including focal cortical dysplasia and perisylvian dysplasia; (3) heterotopia; (4) hemimegalencephaly; (5) microdysgenesis; and (6) vascular malformations, including arteriovenous malformation, cavernous angioma, and arteriovenous aneurysm of vein of Galen. Metabolic disorders associated with childhood epilepsy include (1) acute intermittent porphyria, (2) Alpers syndrome, and (3) biotinidase deficiency. Tumors and phakomatoses in childhood epilepsy include (1) gangliogliomas, (2) dysembryoplastic neuroepithelial tumor, (3) desmoplastic cerebral astrocytoma of infancy, (4) desmoplastic infantile ganglioglioma, (5) pleomorphic xanthoastrocytoma, (6) subependymal giant cell astrocytoma of tuberous sclerosis complex, (7) Sturge-Weber syndrome, and (8) tuberous sclerosis complex. Vascular lesions in childhood epilepsy include (1) hypoxic-ischemic lesions in infants, (2) focal cortical infarction in infants and children, (3) moyamoya disease, and (4) intracranial hemorrhages. Infectious etiologies include (1) bacterial infections, (2) viral infections, (3) toxoplasmosis, and (4) cysticercosis. Ammon's horn sclerosis, a lesion of unknown etiology, is the most common lesion seen in temporal lobe resections for intractable complex partial epilepsy. The authors review the pathologic processes involved in each type of symptomatic epilepsy. 1 figure, 1 table, 113 references.

Federally-Funded Research on Moyamoya Disease

The U.S. Government supports a variety of research studies relating to Moyamoya disease and associated conditions. These studies are tracked by the Office of Extramural Research at the National Institutes of Health.¹⁴ 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 well as topics related to Moyamoya disease 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 Moyamoya disease 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 Moyamoya disease:

- **Project Title: Neurobiology of Disease in Children Conferences**

Principal Investigator & Institution: Maria, Bernard L.; Pediatrics; University of Florida Gainesville, Fl 32611

Timing: Fiscal Year 2001; Project Start 5-APR-2001; Project End 1-AUG-2001

Summary: This grant application describes for symposia titled "Neurobiology of Disease in Children", to be held in conjunction with the 2001 to 2005 annual Child Neurology Society (CNS) meetings. Each of the conditions selected as symposia topics- leukodystrophy, tuberous sclerosis complex, neurofibromatosis, Rett syndrome, and Tourette syndrome- are significant to the child neurology community. The symposia directors are Dr. Hugo Moser (leukodystrophy) Dr. Steven Roach (tuberous sclerosis complex), Dr. Bruce Korf (neurofibromatosis), Dr. Alan Percy (Rett syndrome), and Dr. Harvey Singer (Tourette syndrome). CNS members will constitute the largest segment of the projected 100 to 150 attendees. The symposia will bring together

¹⁴ 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).

clinicians, scientists, caregivers, and National Institutes of Health (NIH) program officers to determine how research findings can be translated to enhance clinical understanding and affect clinical practice. Moreover, symposia attendees will identify the clinical and basic research needed to advance diagnostic accuracy and develop safe and effective therapies for these disorders. In 1998, the NIH funded a symposium on Joubert syndrome held as a satellite meeting of the annual CNS conference that will serve as a model for the proposed scientific sessions. The overall aims are (1) to review scientific advances and determine their relevance to current and future clinical practice in child neurology; (2) to coordinate efforts among various clinical and basic science disciplines, lay organizations, and NIH to review current research initiatives and define future directions; and (3) to disseminate symposia proceedings to ensure that clinical and basic science disciplines are informed about scientific advances, current research initiatives, and future directions. We believe the "Neurobiology of Disease in Children" symposia will contribute to advancing the field of child neurology by facilitating the exchange of information and coordination of research efforts across disciplines and organizations. The project will help further develop a symposium model that may be applied to a broader range of conditions that affect the child's nervous system.

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

- **Project Title: Regulation by Retinoic Acid Synthesized in the Choroid Plexus**

Principal Investigator & Institution: Mccaffrey, Peter; ; Eunice Kennedy Shriver Ctr Mtl Retardatn Waltham, Ma 02254

Timing: Fiscal Year 2000

Summary: The transcriptional activator retinoic acid is one of the most teratogenic substances known, and through accidental exposure of human embryos to the acne drug Acutane(R), cerebellar development was found to be particularly vulnerable. A high susceptibility to systemically applied retinoic acid could indicate that the developing cerebellum is naturally disposed to respond to endogenously synthesized retinoic acid and that a precisely regulated spatial and temporal distribution of the endogenous compound is essential for normal development. Our preliminary observations are consistent with this notion: with a novel technique for the detection of retinoic-acid synthesizing enzymes we find very little synthesis in the cerebellum proper, but high levels in the closely apposed choroid plexus. Moreover, enzyme levels in the choroid plexus shown two maxima that seem to parallel two processes in cerebellar morphogenesis: the first during

Purkinje cell development at late fetal stages and the second during the postnatal stage of granule cell development; cultures of the developing cerebellum reveal a potent growth-promoting effect secreted by the choroid plexus. Here we propose to test the hypothesis that the metencephalic choroid plexus represents a paracrine organ for the developing cerebellum, influencing neuronal differentiation through spatially and temporally regulated retinoic acid secretion. In detail, we plan to address the following topics: (1) We will characterize the distribution of retinoic acid synthesis in the developing metencephalon by functional assay, and by *in situ* hybridization with a probe to the choroid plexus enzyme that we recently isolated for the mouse. (2) We will assay the extent to which retinoic acid accounts for the choroid-plexus neurite inducing activity, and what isomer is synthesized *in vivo*. (3) We will test for effects of thyroid hormone, a factor known to influence cerebellar development and which interacts with retinoic acid through the direct cooperation between the thyroid hormone receptor and the cerebellar retinoic acid receptor RXR. (4) We will attempt to perturb the distribution of retinoic acid in the developing cerebellum *in vivo* by unilateral lesioning of the choroid plexus and by application of retinoic-acid soaked beads. (5) We will try to isolate the human homolog of the murine retinoic-acid generating enzyme, in order to facilitate future work on cerebellar abnormalities in humans. This is exemplified by the Acutane(R)-induced cerebellar malformation which resembles the sporadically occurring Dandy-Walker syndrome and Joubert syndrome, in which ataxia and mental retardation are observed.

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

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.¹⁵ If the publisher has a Web site that offers full text of its journals, PubMed will provide links to that site, as well

¹⁵ 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.

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 Moyamoya disease, simply go to the PubMed Web site at www.ncbi.nlm.nih.gov/pubmed. Type "Moyamoya disease" (or synonyms) into the search box, and click "Go." The following is the type of output you can expect from PubMed for "Moyamoya disease" (hyperlinks lead to article summaries):

- **Neovascularization (angiogenesis) after revascularization in moyamoya disease. Which technique is most useful for moyamoya disease?**
Author(s): Houkin K, Kuroda S, Ishikawa T, Abe H.
Source: *Acta Neurochirurgica*. 2000; 142(3): 269-76.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10819257&dopt=Abstract
- **Neuronal impairment of adult moyamoya disease detected by quantified proton MRS and comparison with cerebral perfusion by SPECT with tc-99m HM-PAO: a trial of clinical quantification of metabolites.**
Author(s): Harada M, Miyoshi H, Uno M, Okada T, Hisaoka S, Hori A, Nishitani H.
Source: *Journal of Magnetic Resonance Imaging : Jmri*. 1999 August; 10(2): 124-9.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10441014&dopt=Abstract
- **Neuropsychological recovery from childhood moyamoya disease.**
Author(s): Bowen M, Marks MP, Steinberg GK.
Source: *Brain & Development*. 1998 March; 20(2): 119-23.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9545184&dopt=Abstract
- **Neurorehabilitation outcome in moyamoya disease.**
Author(s): Moore DP, Lee MY, Macciocchi SN.
Source: *Archives of Physical Medicine and Rehabilitation*. 1997 June; 78(6): 672-5.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9196479&dopt=Abstract

- **Non-anastomotic bypass surgery for childhood moyamoya disease using dural pedicle insertion over the brain surface combined with encephalogleomyosynangiosis.**
 Author(s): Yoshida YK, Shirane R, Yoshimoto T.
 Source: Surgical Neurology. 1999 April; 51(4): 404-11.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10199294&dopt=Abstract
- **Noonan syndrome associated with moyamoya disease: report of one case.**
 Author(s): Tang KT, Yang W, Wong J, Lee KY.
 Source: Acta Paediatr Taiwan. 1999 July-August; 40(4): 274-6.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10910629&dopt=Abstract
- **Ocular ischemic syndrome in a child with moyamoya disease and neurofibromatosis.**
 Author(s): Barrall JL, Summers CG.
 Source: Survey of Ophthalmology. 1996 May-June; 40(6): 500-4.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8724642&dopt=Abstract
- **Ocular malformations, moyamoya disease, and midline cranial defects: a distinct syndrome.**
 Author(s): Bakri SJ, Siker D, Masaryk T, Luciano MG, Traboulsi EI.
 Source: American Journal of Ophthalmology. 1999 March; 127(3): 356-7.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10088755&dopt=Abstract
- **Perioperative factors related to the development of ischemic complications in patients with moyamoya disease.**
 Author(s): Sato K, Shirane R, Yoshimoto T.
 Source: Child's Nervous System : Chns : Official Journal of the International Society for Pediatric Neurosurgery. 1997 February; 13(2): 68-72.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9105739&dopt=Abstract
- **Perioperative management protocols for children with moyamoya disease.**
 Author(s): Nomura S, Kashiwagi S, Uetsuka S, Uchida T, Kubota H, Ito H.

Source: Child's Nervous System : Chns : Official Journal of the International Society for Pediatric Neurosurgery. 2001 April; 17(4-5): 270-4.

http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11398948&dopt=Abstract

- **Phakomatosis pigmentovascularis type IIIb associated with moyamoya disease.**

Author(s): Tsuruta D, Fukai K, Seto M, Fujitani K, Shindo K, Hamada T, Ishii M.

Source: Pediatric Dermatology. 1999 January-February; 16(1): 35-8.

Review.

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Author(s): Komiyama M, Yasui T, Sakamoto H, Fujita K, Sato T, Ota M, Sugita M.
Source: Pediatric Neurosurgery. 2000 August; 33(2): 100-4.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11070437&dopt=Abstract

- **Cardiopulmonary bypass in a patient with moyamoya disease.**
Author(s): Wang N, Kuluz J, Barron M, Perryman R.
Source: Anesthesia and Analgesia. 1997 May; 84(5): 1160-3. No Abstract Available.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9141954&dopt=Abstract
- **CBF change with aging in moyamoya disease.**
Author(s): Oyama H, Niwa M, Kida Y.
Source: J Neurosurg Sci. 1998 March; 42(1): 33-6.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9766270&dopt=Abstract
- **Central deafness in a young child with Moyamoya disease: paternal linkage in a Caucasian family: two case reports and a review of the literature.**
Author(s): Setzen G, Cacace AT, Eames F, Riback P, Lava N, McFarland DJ, Artino LM, Kerwood JA.
Source: International Journal of Pediatric Otorhinolaryngology. 1999 April 25; 48(1): 53-76. Review.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10365973&dopt=Abstract
- **Cerebellar hemorrhage associated with moyamoya disease.**
Author(s): Ise H, Tokunaga H, Aizawa M, Sawaura H.
Source: Neurol Med Chir (Tokyo). 1998 April; 38(4): 225-8.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9631637&dopt=Abstract
- **Cerebral blood flow measurement as an indicator for an indirect revascularization procedure for adult patients with moyamoya disease.**
Author(s): Kohno K, Oka Y, Kohno S, Ohta S, Kumon Y, Sakaki S.
Source: Neurosurgery. 1998 April; 42(4): 752-7; Discussion 757-8.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9574639&dopt=Abstract
- **Cerebral blood flow velocities in an infant with moyamoya disease.**
Author(s): Ipsiroglu OS, Eichler F, Stockler-Ipsiroglu S, Trattnig S.
Source: Pediatric Neurology. 1999 October; 21(4): 739-41. Review.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10580888&dopt=Abstract

- **Cerebral hemodynamics and metabolism in moyamoya disease--a positron emission tomography study.**
Author(s): Kuwabara Y, Ichiya Y, Sasaki M, Yoshida T, Masuda K, Ikezaki K, Matsushima T, Fukui M.
Source: Clinical Neurology and Neurosurgery. 1997 October; 99 Suppl 2: S74-8.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9409411&dopt=Abstract
- **Cerebral hemodynamics in patients with moyamoya disease and in patients with atherosclerotic occlusion of the major cerebral arterial trunks.**
Author(s): Obara K, Fukuuchi Y, Kobari M, Watanabe S, Dembo T.
Source: Clinical Neurology and Neurosurgery. 1997 October; 99 Suppl 2: S86-9.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9409413&dopt=Abstract
- **Cerebral hypoxia after hyperventilation causes "re-build-up" phenomenon and TIA in childhood moyamoya disease. A near-infrared spectroscopy study.**
Author(s): Kuroda S, Houkin K, Hoshi Y, Tamura M, Kazumata K, Abe H.
Source: Child's Nervous System : Chns : Official Journal of the International Society for Pediatric Neurosurgery. 1996 August; 12(8): 448-52; Discussion 453.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8891362&dopt=Abstract
- **Cerebral ischemic complications following intracranial bleeding in patients with moyamoya disease--three case reports.**
Author(s): Iwama T, Kotani Y, Yamakawa H, Nagata I, Hashimoto N, Sakai N.
Source: Neurol Med Chir (Tokyo). 2001 September; 41(9): 450-3.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11593973&dopt=Abstract
- **Cerebral oxygenation state in childhood moyamoya disease: a near-infrared spectroscopy study.**
Author(s): Lin Y, Yoshiko K, Negoro T, Watanabe K, Negoro M.

Source: Pediatric Neurology. 2000 May; 22(5): 365-9.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10913728&dopt=Abstract

- **Cerebral perfusion imaging evaluates pharmacologic treatment of unilateral moyamoya disease.**
 Author(s): Kuroki M, Nagamachi S, Hoshi H, Flores LG 2nd, Ohnishi T, Jinnouchi S, Futami S, Watanabe K.
 Source: Journal of Nuclear Medicine : Official Publication, Society of Nuclear Medicine. 1996 January; 37(1): 84-6.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8544009&dopt=Abstract

- **Cerebral revascularization for moyamoya disease in adults.**
 Author(s): Srinivasan J, Britz GW, Newell DW.
 Source: Neurosurg Clin N Am. 2001 July; 12(3): 585-94, Ix. Review.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11390316&dopt=Abstract

- **Cerebral revascularization for moyamoya disease in children.**
 Author(s): Houkin K, Kuroda S, Nakayama N.
 Source: Neurosurg Clin N Am. 2001 July; 12(3): 575-84, Ix.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=11390315&dopt=Abstract

- **Cerebral revascularization using omentum and serratus anterior muscle free flap transfer for adult moyamoya disease: case report.**
 Author(s): Yoshioka N, Tominaga S, Inui T.
 Source: Surgical Neurology. 1996 November; 46(5): 430-5; Discussion 435-6.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8874540&dopt=Abstract

- **Changes in cortical CBF and vascular response after vascular reconstruction in patients with adult onset moyamoya disease.**
 Author(s): Watanabe H, Ohta S, Oka Y, Kumon Y, Sakaki S, Sugawara Y, Tanada S.
 Source: Acta Neurochirurgica. 1996; 138(10): 1211-7.
http://www.ncbi.nlm.nih.gov/80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8955441&dopt=Abstract

- **Characteristics of intracranial aneurysms associated with moyamoya disease. A review of 111 cases.**
Author(s): Kawaguchi S, Sakaki T, Morimoto T, Kakizaki T, Kamada K.
Source: Acta Neurochirurgica. 1996; 138(11): 1287-94. Review.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8980731&dopt=Abstract
- **Childhood moyamoya disease: hemodynamic MRI.**
Author(s): Tzika AA, Robertson RL, Barnes PD, Vajapeyam S, Burrows PE, Treves ST, Scott RM.
Source: Pediatric Radiology. 1997 September; 27(9): 727-35.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9285733&dopt=Abstract
- **Chorea: an unusual presenting feature in pediatric Moyamoya disease.**
Author(s): Parmar RC, Bavdekar SB, Muranjan MN, Limaye U.
Source: Indian Pediatrics. 2000 September; 37(9): 1005-9. No Abstract Available.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=10992339&dopt=Abstract
- **Clinical and angiographic follow-up of childhood-onset moyamoya disease.**
Author(s): Ezura M, Yoshimoto T, Fujiwara S, Takahashi A, Shirane R, Mizoi K.
Source: Child's Nervous System : Chns : Official Journal of the International Society for Pediatric Neurosurgery. 1995 October; 11(10): 591-4.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=8556726&dopt=Abstract
- **Clinical and neuroradiological findings of moyamoya disease in Italy.**
Author(s): Battistella PA, Carollo C.
Source: Clinical Neurology and Neurosurgery. 1997 October; 99 Suppl 2: S54-7.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9409406&dopt=Abstract
- **Clinical features of moyamoya disease in sibling relations under 15 years of age.**
Author(s): Hamada JI, Yoshioka S, Nakahara T, Marubayashi T, Ushio Y.

Source: Acta Neurochirurgica. 1998; 140(5): 455-8.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9728245&dopt=Abstract

Vocabulary Builder

Aneurysm: A sac formed by the dilatation of the wall of an artery, a vein, or the heart. The chief signs of arterial aneurysm are the formation of a pulsating tumour, and often a bruit (aneurysmal bruit) heard over the swelling. Sometimes there are symptoms from pressure on contiguous parts. [EU]

Arteriovenous: Both arterial and venous; pertaining to or affecting an artery and a vein. [EU]

Assay: Determination of the amount of a particular constituent of a mixture, or of the biological or pharmacological potency of a drug. [EU]

Ataxia: Failure of muscular coordination; irregularity of muscular action. [EU]

Cerebellar: Pertaining to the cerebellum. [EU]

Cerebellum: Part of the metencephalon that lies in the posterior cranial fossa behind the brain stem. It is concerned with the coordination of movement. [NIH]

Choroid: The thin, highly vascular membrane covering most of the posterior of the eye between the retina and sclera. [NIH]

Cortical: Pertaining to or of the nature of a cortex or bark. [EU]

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

Ganglioglioma: Rare indolent tumors comprised of neoplastic glial and neuronal cells which occur primarily in children and young adults. Benign lesions tend to be associated with long survival unless the tumor degenerates into a histologically malignant form. They tend to occur in the optic nerve and white matter of the brain and spinal cord. [NIH]

Granule: A small pill made from sucrose. [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]

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

Infarction: 1. the formation of an infarct. 2. an infarct. [EU]

Intermittent: Occurring at separated intervals; having periods of cessation of activity. [EU]

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

Lobe: A more or less well-defined portion of any organ, especially of the brain, lungs, and glands. Lobes are demarcated by fissures, sulci, connective tissue, and by their shape. [EU]

Malformation: A morphologic defect resulting from an intrinsically abnormal developmental process. [EU]

Morphogenesis: The development of the form of an organ, part of the body, or organism. [NIH]

Neoplastic: Pertaining to or like a neoplasm (= any new and abnormal growth); pertaining to neoplasia (= the formation of a neoplasm). [EU]

Neuronal: Pertaining to a neuron or neurons (= conducting cells of the nervous system). [EU]

Pathologic: 1. indicative of or caused by a morbid condition. 2. pertaining to pathology (= branch of medicine that treats the essential nature of the disease, especially the structural and functional changes in tissues and organs of the body caused by the disease). [EU]

Pediatrics: A medical specialty concerned with maintaining health and providing medical care to children from birth to adolescence. [NIH]

Plexus: A network or tangle; a general term for a network of lymphatic vessels, nerves, or veins. [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]

Postnatal: Occurring after birth, with reference to the newborn. [EU]

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]

Resection: Excision of a portion or all of an organ or other structure. [EU]

Sclerosis: A induration, or hardening; especially hardening of a part from inflammation and in diseases of the interstitial substance. The term is used chiefly for such a hardening of the nervous system due to hyperplasia of the connective tissue or to designate hardening of the blood vessels. [EU]

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]

Technetium: The first artificially produced element and a radioactive fission product of uranium. The stablest isotope has a mass number 99 and is used diagnostically as a radioactive imaging agent. Technetium has the atomic symbol Tc, atomic number 43, and atomic weight 98.91. [NIH]

Teratogenic: Tending to produce anomalies of formation, or teratism (= anomaly of formation or development : condition of a monster). [EU]

Toxoplasmosis: An acute or chronic, widespread disease of animals and humans caused by the obligate intracellular protozoon *Toxoplasma gondii*, transmitted by oocysts containing the pathogen in the feces of cats (the definitive host), usually by contaminated soil, direct exposure to infected feces, tissue cysts in infected meat, or tachyzoites (proliferating forms) in blood. [EU]

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

CHAPTER 4. BOOKS ON MOYAMOYA DISEASE

Overview

This chapter provides bibliographic book references relating to Moyamoya disease. You have many options to locate books on Moyamoya disease. 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 parents, however, prefer online sources (e.g. www.amazon.com and www.bn.com). In addition to online booksellers, excellent sources for book titles on Moyamoya disease 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.

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 "Moyamoya disease" (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:¹⁶

¹⁶ 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

- **Genetic polymorphisms and susceptibility to disease.** Author: edited by M.S. Miller, M.T. Cronin; Year: 2000; London; New York: Taylor & Francis, 2000; ISBN: 0748408223
<http://www.amazon.com/exec/obidos/ASIN/0748408223/icongroupinterna>
- **Genetics and public health in the 21st century: using genetic information to improve health and prevent disease.** Author: edited by Muin J. Khoury, Wylie Burke, Elizabeth J. Thomson; Year: 2000; Oxford; New York: Oxford University Press, 2000; ISBN: 0195128303 (alk. paper)
<http://www.amazon.com/exec/obidos/ASIN/0195128303/icongroupinterna>
- **Moyamoya Disease edited by Kiyonobu Ikezaki, Christopher Loftus.** ; Year: 2001; Rolling Meadows: American Association of Neurological Surgeons, c2001; ISBN: 1879284804
<http://www.amazon.com/exec/obidos/ASIN/1879284804/icongroupinterna>
- **Moyamoya disease.** Author: Jiro Suzuki; Year: 1986; Berlin; New York: Springer-Verlag, c1986; ISBN: 0387157786 (U.S.: pbk.)
<http://www.amazon.com/exec/obidos/ASIN/0387157786/icongroupinterna>
- **Operative techniques in pediatric neurosurgery.** Author: edited by A. Leland Albright, Ian F. Pollack, P. David Adelson; Year: 2001; New York: Thieme, 2001; ISBN: 0865778469 (TNY)
<http://www.amazon.com/exec/obidos/ASIN/0865778469/icongroupinterna>

Chapters on Moyamoya Disease

Frequently, Moyamoya disease will be discussed within a book, perhaps within a specific chapter. In order to find chapters that are specifically dealing with Moyamoya disease, an excellent source of abstracts is the Combined Health Information Database. You will need to limit your search to book chapters and Moyamoya disease using the "Detailed Search" option. Go 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

which the phrase is found. Currently, the majority of the links are between the books and 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>.

typing in “Moyamoya disease” (or synonyms) into the “For these words:” box, you will only receive results on chapters in books.

General Home References

In addition to references for Moyamoya disease, 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):

- **Adams & Victor’s Principles Of Neurology** by Maurice Victor, et al; Hardcover - 1692 pages; 7th edition (December 19, 2000), McGraw-Hill Professional Publishing; ISBN: 0070674973;
<http://www.amazon.com/exec/obidos/ASIN/0070674973/icongroupinterna>
- **American Academy of Pediatrics Guide to Your Child’s Symptoms : The Official, Complete Home Reference, Birth Through Adolescence** by Donald Schiff (Editor), et al; Paperback - 256 pages (January 1997), Villard Books; ISBN: 0375752579;
<http://www.amazon.com/exec/obidos/ASIN/0375752579/icongroupinterna>
- **The Children’s Hospital Guide to Your Child’s Health and Development** by Alan D. Woolf (Editor), et al; Hardcover - 796 pages, 1st edition (January 15, 2001), Perseus Books; ISBN: 073820241X;
<http://www.amazon.com/exec/obidos/ASIN/073820241X/icongroupinterna>
- **Clinical Neuroanatomy Made Ridiculously Simple (MedMaster Series, 2000 Edition)** by Stephen Goldberg; Paperback: 97 pages; 2nd edition (February 15, 2000), Medmaster; ISBN: 0940780461;
<http://www.amazon.com/exec/obidos/ASIN/0940780461/icongroupinterna>
- **Helping Your Child in the Hospital: A Practical Guide for Parents** by Nancy Keene, Rachel Prentice; Paperback - 176 pages, 3rd edition (April 15, 2002), O’Reilly & Associates; ISBN: 0596500114;
<http://www.amazon.com/exec/obidos/ASIN/0596500114/icongroupinterna>
- **It’s Not a Tumor!: The Patient’s Guide to Common Neurological Problems** by Robert Wiedemeyer; Paperback: (January 1996), Boxweed Pub; ISBN: 0964740796;
<http://www.amazon.com/exec/obidos/ASIN/0964740796/icongroupinterna>
- **Medical Emergencies & Childhood Illnesses: Includes Your Child’s Personal Health Journal (Parent Smart)** by Penny A. Shore, William Sears (Contributor); Paperback - 115 pages (February 2002), Parent Kit

Corporation; ISBN: 1896833187;

<http://www.amazon.com/exec/obidos/ASIN/1896833187/iconegroupinterna>

- **Neurology for the Non-Neurologist** by William J. Weiner (Editor), Christopher G. Goetz (Editor); Paperback (May 1999), Lippincott, Williams & Wilkins Publishers; ISBN: 0781717078;
<http://www.amazon.com/exec/obidos/ASIN/0781717078/iconegroupinterna>
- **Taking Care of Your Child: A Parent's Guide to Complete Medical Care** by Robert H. Pantell, M.D., et al; Paperback - 524 pages, 6th edition (March 5, 2002), Perseus Press; ISBN: 0738206016;
<http://www.amazon.com/exec/obidos/ASIN/0738206016/iconegroupinterna>

Vocabulary Builder

Acuity: Clarity or clearness, especially of the vision. [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]

Extrapyramidal: Outside of the pyramidal tracts. [EU]

Neuroanatomy: Study of the anatomy of the nervous system as a specialty or discipline. [NIH]

Neurosurgery: A surgical specialty concerned with the treatment of diseases and disorders of the brain, spinal cord, and peripheral and sympathetic nervous system. [NIH]

Pulmonary: Pertaining to the lungs. [EU]

Tomography: The recording of internal body images at a predetermined plane by means of the tomograph; called also body section roentgenography. [EU]

CHAPTER 5. PHYSICIAN GUIDELINES AND DATABASES

Overview

Doctors and medical researchers rely on a number of information sources to help children with Moyamoya disease. 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 medical conditions, 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 Neurological Disorders and Stroke (NINDS); neurological disorder information pages available at http://www.ninds.nih.gov/health_and_medical/disorder_index.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.¹⁷ 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:¹⁸

- **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 cancer-oriented databases:
http://www.nlm.nih.gov/databases/databases_cancer.html

¹⁷ Remember, for the general public, the National Library of Medicine recommends the databases referenced in MEDLINEplus (<http://medlineplus.gov/> or <http://www.nlm.nih.gov/medlineplus/databases.html>).

¹⁸ 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 Moyamoya disease, the following are particularly noteworthy.

The NLM Gateway¹⁹

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.²⁰ 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,

¹⁹ Adapted from NLM: <http://gateway.nlm.nih.gov/gw/Cmd?Overview.x>.

²⁰ 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).

researchers, librarians, students, and, increasingly, parents and the public.²¹ To use the NLM Gateway, simply go to the search site at <http://gateway.nlm.nih.gov/gw/Cmd>. Type “Moyamoya disease” (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	349060
Books / Periodicals / Audio Visual	2577
Consumer Health	294
Meeting Abstracts	2575
Other Collections	100
Total	354606

HSTAT²²

HSTAT is a free, Web-based resource that provides access to full-text documents used in healthcare decision-making.²³ 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.²⁴ Simply search by “Moyamoya disease” (or synonyms) at the following Web site: <http://text.nlm.nih.gov>.

²¹ 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 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.

²² Adapted from HSTAT: <http://www.nlm.nih.gov/pubs/factsheets/hstat.html>.

²³ The HSTAT URL is <http://hstat.nlm.nih.gov/>.

²⁴ Other important documents in HSTAT include: the National Institutes of Health (NIH) Consensus Conference Reports and Technology Assessment Reports; the HIV/AIDS

Coffee Break: Tutorials for Biologists²⁵

Some parents 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.²⁶ 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.²⁷ This site has new articles every few weeks, so it can be considered an online magazine of sorts, and intended for general background information. Access the Coffee Break Web site at <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/clinweb/>.
- **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

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.

²⁵ Adapted from <http://www.ncbi.nlm.nih.gov/Coffeebreak/Archive/FAQ.html>.

²⁶ 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.

²⁷ 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 record system; see the following Web site:
<http://www.cml.upmc.edu/cml/imageengine/imageEngine.html>.

- **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 <http://www.lexical.com/Metaphrase.html>.

The Genome Project and Moyamoya Disease

With all the discussion in the press about the Human Genome Project, it is only natural that physicians, researchers, and parents want to know about how human genes relate to Moyamoya disease. 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).²⁸ 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.

Go to <http://www.ncbi.nlm.nih.gov/Omim/searchomim.html> to search the database. Type "Moyamoya disease" (or synonyms) in the search box, and click "Submit Search." If too many results appear, you can narrow the search

²⁸ 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.

by adding 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 Moyamoya disease:

- **Moyamoya Disease**
Web site: <http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispim?252350>
- **Moyamoya Disease 2**
Web site: <http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispim?607151>

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:

- **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>
- **Nervous System:** Mind and body.
Examples: Alzheimer disease, Amyotrophic lateral sclerosis, Angelman syndrome, Charcot-Marie-Tooth disease, epilepsy, essential tremor, Fragile X syndrome, Friedreich’s ataxia, Huntington disease, Niemann-Pick disease, Parkinson disease, Prader-Willi syndrome, Rett syndrome, Spinocerebellar atrophy, Williams syndrome.
Web site: <http://www.ncbi.nlm.nih.gov/disease/Brain.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>

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/>, 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 "Moyamoya disease" (or synonyms) and click "Go."

Jablonski's Multiple Congenital Anomaly/Mental Retardation (MCA/MR) Syndromes Database²⁹

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 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.

²⁹ Adapted from the National Library of Medicine:
http://www.nlm.nih.gov/mesh/jablonski/about_syndrome.html.

The Genome Database³⁰

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 "Moyamoya disease" (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 Moyamoya disease (sorted alphabetically by title; hyperlinks provide rankings, information, and reviews at Amazon.com):

- **Atlas of Pediatric Physical Diagnosis** by Basil J. Zitelli, Holly W. Davis (Editor); Hardcover, 3rd edition (March 1997), Mosby-Year Book; ISBN: 0815199309;
<http://www.amazon.com/exec/obidos/ASIN/0815199309/icongroupinterna>
- **The 5-Minute Pediatric Consult** by M. William Schwartz (Editor); Hardcover - 1050 pages, 2nd edition (January 15, 2000), Lippincott,

³⁰ Adapted from the Genome Database:
<http://gdbwww.gdb.org/gdb/aboutGDB.html#mission>.

Williams & Wilkins; ISBN: 0683307444;

<http://www.amazon.com/exec/obidos/ASIN/0683307444/icongroupinterna>

- **The Behavioral Neurology of White Matter** by Christopher M. Filley; Paperback - 279 pages; 1st edition (September 15, 2001), Oxford University Press; ISBN: 019513561X;
<http://www.amazon.com/exec/obidos/ASIN/019513561X/icongroupinterna>
- **The Cerebellum and Its Disorders** by Mario-Ubaldo Manto, Massimo Pandolfo; Hardcover - 1st edition (January 2002), Cambridge University Press; ISBN: 0521771560;
<http://www.amazon.com/exec/obidos/ASIN/0521771560/icongroupinterna>
- **Clinical Neurology** by David A. Greenberg, et al; Paperback - 390 pages; 5th edition (February 9, 2002), Appleton & Lange; ISBN: 0071375430;
<http://www.amazon.com/exec/obidos/ASIN/0071375430/icongroupinterna>
- **Clinical Neurology for Psychiatrists** by David M. Kaufman; Hardcover - 670 pages, 5th edition (January 15, 2001), W. B. Saunders Co.; ISBN: 0721689957;
<http://www.amazon.com/exec/obidos/ASIN/0721689957/icongroupinterna>
- **Comprehensive Neurology** by Roger N. Rosenberg (Editor), David E. Pleasure (Editor); 1280 pages, 2nd edition (April 1998), Wiley-Liss; ISBN: 0471169587;
<http://www.amazon.com/exec/obidos/ASIN/0471169587/icongroupinterna>
- **Emergent and Urgent Neurology** by William J. Weiner (Editor), Lisa M. Shulman (Editor); Hardcover - 571 pages; 2nd edition (January 15, 1999), Lippincott, Williams & Wilkins Publishers; ISBN: 0397518579;
<http://www.amazon.com/exec/obidos/ASIN/0397518579/icongroupinterna>
- **Nelson Textbook of Pediatrics** by Richard E. Behrman (Editor), et al; Hardcover - 2414 pages, 16th edition (January 15, 2000), W B Saunders Co; ISBN: 0721677673;
<http://www.amazon.com/exec/obidos/ASIN/0721677673/icongroupinterna>
- **Neurology in Clinical Practice: Volume I: Principles of Diagnosis and Management, Volume II: The Neurological Disorders (2-Volume Set, Includes a 12-Month Subscription to the Online Edition)** by W. G. Bradley, et al; Hardcover - 2413 pages, 3rd edition, Vol 1-2 (January 15, 2000), Butterworth-Heinemann; ISBN: 0750699736;
<http://www.amazon.com/exec/obidos/ASIN/0750699736/icongroupinterna>
- **Neuroscience: Exploring the Brain** by Mark F. Bear, et al; Hardcover - 855 pages, 2nd edition (January 15, 2001), Lippincott, Williams & Wilkins Publishers; ISBN: 0683305964;
<http://www.amazon.com/exec/obidos/ASIN/0683305964/icongroupinterna>

- **Office Practice of Neurology** by Martain A. Samuels, Steven F. Feske; Hardcover, Churchill Livingstone; ISBN: 0443065578;
<http://www.amazon.com/exec/obidos/ASIN/0443065578/icongroupinterna>
- **Patient-Based Approaches to Cognitive Neuroscience** by Martha J. Farah (Editor), Todd E. Feinberg (Editor); Paperback - 425 pages (April 3, 2000), MIT Press; ISBN: 0262561239;
<http://www.amazon.com/exec/obidos/ASIN/0262561239/icongroupinterna>
- **Principles of Neural Science** by Eric R. Kandel (Editor), et al; Hardcover - 1414 pages, 4th edition (January 5, 2000), McGraw-Hill Professional Publishing; ISBN: 0838577016;
<http://www.amazon.com/exec/obidos/ASIN/0838577016/icongroupinterna>
- **Review Manual for Neurology in Clinical Practice** by Karl E. Misulis, et al; Paperback, Butterworth-Heinemann Medical; ISBN: 0750671920;
<http://www.amazon.com/exec/obidos/ASIN/0750671920/icongroupinterna>

Vocabulary Builder

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

PART III. APPENDICES

ABOUT PART III

Part III is a collection of appendices on general medical topics relating to Moyamoya disease and related conditions.

APPENDIX A. RESEARCHING YOUR CHILD'S MEDICATIONS

Overview

There are a number of sources available on new or existing medications which could be prescribed to treat Moyamoya disease. While a number of hard copy or CD-Rom resources are available to parents 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 child's medications. You may also want to research medications that your child is currently taking for other conditions as they may interact with medications for Moyamoya disease. Research can give you information on the side effects, interactions, and limitations of prescription drugs used in the treatment of Moyamoya disease. 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 Child's Medications: The Basics³¹

The Agency for Health Care Research and Quality has published extremely useful guidelines on the medication aspects of Moyamoya disease. Giving your child medication can involve many steps and decisions each day. The AHCQRQ recommends that parents take part in treatment decisions. Do not be afraid to ask questions and talk about your concerns. By taking a moment to ask questions, your child may be spared from possible problems. Here are some points to cover each time a new medicine is prescribed:

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

Do not hesitate to tell the doctor about preferences you have for your child's medicines. You may want your child to have a medicine with the fewest side effects, or the fewest doses to take each day. You may care most about cost. Or, you may want the medicine the doctor believes will work the best. Sharing your concerns will help the doctor select the best treatment for your child.

Do not be afraid to "bother" the doctor with your questions about medications for Moyamoya disease. You can also talk to a nurse or a pharmacist. They can help you better understand your child's treatment plan. Talking over your child's options with someone you trust can help you make better choices. Specifically, ask the doctor the following:

- The name of the medicine and what it is supposed to do.
- How and when to give your child the medicine, how much, and for how long.
- What food, drinks, other medicines, or activities your child should avoid while taking the medicine.
- What side effects your child may experience, and what to do if they occur.
- If there are any refills, and how often.
- About any terms or directions you do not understand.
- What to do if your child misses a dose.

³¹ This section is adapted from AHCQRQ: <http://www.ahcpr.gov/consumer/ncpiebro.htm>.

- If there is written information you can take home (most pharmacies have information sheets on prescription medicines; some even offer large-print or Spanish versions).

Do not forget to tell the doctor about all the medicines your child is currently taking (not just those for Moyamoya disease). This includes prescription medicines and the medicines that you buy over the counter. When talking to the doctor, you may wish to prepare a list of medicines your child is currently taking including why and in what forms. 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 Child's Medications

Because of historical investments by various organizations and the emergence of the Internet, it has become rather simple to learn about the medications the doctor has recommended for Moyamoya disease. 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.³²

While the FDA database is rather large and difficult to navigate, the Pharmacopeia 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 child's doctor's office.

Reuters Health Drug Database

The Reuters Health Drug Database can be searched by keyword at the hyperlink: <http://www.reutershealth.com/frame2/drug.html>.³³

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 information on prescribing and drug interactions. Information can

³² Though cumbersome, the FDA database can be freely browsed at the following site: www.fda.gov/cder/da/da.htm.

³³ Adapted from *A to Z Drug Facts* by Facts and Comparisons.

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 children with Moyamoya disease--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 Moyamoya disease or potentially create deleterious side effects in patients with Moyamoya disease. You should ask the physician about any contraindications, especially as these might apply to other medications that your child 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 your child to experience an unexpected side effect. Drug interactions may make medications less effective, cause unexpected side effects, or increase the action of a particular drug. Some drug interactions can even be harmful to your child.

Be sure to read the label every time you give your child a nonprescription or prescription drug, and take the time to learn about drug interactions. These precautions may be critical to your child's 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 give your child a medication. When the doctor prescribes a new drug, discuss all over-the-counter and prescription medications, dietary supplements, vitamins, botanicals, minerals and herbals your child takes. Ask your pharmacist for the package insert for each drug prescribed. 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 Moyamoya disease. Exercise caution--some of these drugs may have fraudulent claims, and others may actually hurt your child. 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 Moyamoya disease. The FDA warns to watch out for³⁴:

- Secret formulas (real scientists share what they know)
- 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**.

³⁴ This section has been adapted from
<http://www.fda.gov/opacom/lowlit/medfraud.html>.

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):

- **Current Therapy in Neurologic Disease** by Richard T. Johnson, et al; Hardcover - 457 pages, 6th edition (January 15, 2002), Mosby-Year Book; ISBN: 0323014720;
<http://www.amazon.com/exec/obidos/ASIN/0323014720/icongroupinterna>
- **Emerging Pharmacological Tools in Clinical Neurology** by MedPanel Inc. (Author); Digital - 66 pages, MarketResearch.com; ISBN: B00005RBN8;
<http://www.amazon.com/exec/obidos/ASIN/B00005RBN8/icongroupinterna>
- **Goodman & Gilman's The Pharmacological Basis of Therapeutics** by Joel G. Hardman (Editor), Lee E. Limbird; Hardcover - 1825 pages, 10th edition (August 13, 2001), McGraw-Hill Professional Publishing; ISBN: 0071354697;
<http://www.amazon.com/exec/obidos/ASIN/0071354697/icongroupinterna>
- **Neurology and General Medicine** by Michael J. Aminoff (Editor), Hardcover - 992 pages, 3rd edition (March 15, 2001), Churchill Livingstone; ISBN: 0443065713;
<http://www.amazon.com/exec/obidos/ASIN/0443065713/icongroupinterna>
- **Neurology and Medicine** by Hughes Perkins; Hardcover - 415 pages, 1st edition (December 15, 1999), B. M. J. Books; ISBN: 0727912240;
<http://www.amazon.com/exec/obidos/ASIN/0727912240/icongroupinterna>
- **Pharmacological Management of Neurological and Psychiatric Disorders** by S. J. Enna (Editor), et al; Hardcover - 736 pages, 1st edition, McGraw-Hill Professional Publishing; ISBN: 0070217645;
<http://www.amazon.com/exec/obidos/ASIN/0070217645/icongroupinterna>

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 child's doctor or your friends 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 Moyamoya disease. Finally, at the conclusion of this chapter, we will provide a list of readings on Moyamoya disease 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?³⁵

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 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 acupuncture and massage, homeopathy, vitamins or herbal products, and acupuncture.

What Are the Domains of Alternative Medicine?³⁶

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

³⁵ Adapted from the NCCAM: <http://nccam.nih.gov/nccam/fcp/faq/index.html#what-is>.

³⁶ Adapted from the NCCAM: <http://nccam.nih.gov/nccam/fcp/classify/index.html>.

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 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 illness, 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 a medical condition is the manifestation of alterations in the processes by which the body naturally heals itself and emphasizes health restoration rather than treatment for the condition itself. 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 medical conditions 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 conditions.

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 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 Child's Treatment?

A critical issue in pursuing complementary alternatives mentioned thus far is the risk that these might have undesirable interactions with your child's medical treatment. It becomes all the more important to speak with the 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 everyone.³⁷

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 child's healthcare provider, whether a physician or practitioner of complementary and alternative medicine. Competent healthcare management requires that the practitioner know of all conventional and alternative therapies that your child is taking.

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 Moyamoya Disease

Having read the previous discussion, you may be wondering which complementary or alternative treatments might be appropriate for Moyamoya disease. 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.

³⁷ Adapted from <http://www.4woman.gov/faq/alternative.htm>.

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 parents to search for articles that specifically relate to Moyamoya disease 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 "Moyamoya disease" (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 Moyamoya disease:

- **A case report of Moyamoya disease: successfully treated with Chinese medicine.**
 Author(s): Hiyama Y, Itoh T, Shimada Y, Shimada T, Terasawa K.
 Source: Am J Chin Med. 1992; 20(3-4): 319-24.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=1471616&dopt=Abstract
- **Hyperventilation technetium-99m-HMPAO brain SPECT in moyamoya disease to assess risk of natural childbirth.**
 Author(s): Kume N, Hayashida K, Shimotsu Y, Matsunaga N.
 Source: Journal of Nuclear Medicine : Official Publication, Society of Nuclear Medicine. 1997 December; 38(12): 1894-7.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=9430464&dopt=Abstract
- **Stroke in children.**
 Author(s): Pavlakis SG, Gould RJ, Zito JL.
 Source: Adv Pediatr. 1991; 38: 151-79. Review.
http://www.ncbi.nlm.nih.gov:80/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=1927699&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):

- **Alternative and Complementary Treatment in Neurologic Illness** by Michael I. Weintraub (Editor); Paperback - 288 pages (March 23, 2001), Churchill Livingstone; ISBN: 0443065586;
<http://www.amazon.com/exec/obidos/ASIN/0443065586/icongroupinterna>
- **Healthy Child, Whole Child: Integrating the Best of Conventional and Alternative Medicine to Keep Your Kids Healthy** by Stuart H. Ditchek, M.D. and Russell H. Greenfield; Paperback - 464 pages (June 2002), Harper Resource; ISBN: 0062737465;
<http://www.amazon.com/exec/obidos/ASIN/0062737465/icongroupinterna>
- **Radical Healing: Integrating the World's Great Therapeutic Traditions to Create a New Transformative Medicine** by Rudolph Ballentine, M.D., Linda Funk (Illustrator); Paperback - 612 pages; Reprint edition (March 14,

2000), Three Rivers Press; ISBN: 0609804847;

<http://www.amazon.com/exec/obidos/ASIN/0609804847/icongroupinterna>

- **The Review of Natural Products** by Facts and Comparisons (Editor); Cd-Rom edition (January 2002), Facts & Comparisons; ISBN: 1574391453;
<http://www.amazon.com/exec/obidos/ASIN/1574391453/icongroupinterna>

For additional information on complementary and alternative medicine, ask your child's 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

APPENDIX C. RESEARCHING NUTRITION

Overview

Since the time of Hippocrates, doctors have understood the importance of diet and nutrition to 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 for Moyamoya disease. Any dietary recommendation is based on age, body mass, gender, lifestyle, eating habits, food preferences, and health condition. It is therefore likely that different patients with Moyamoya disease may be given different recommendations. Some recommendations may be directly related to Moyamoya disease, while others may be more related to general health.

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

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 child's 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 eyes, hair, bones, and skin; sources of vitamin A include foods such as eggs, carrots, and cantaloupe.
- **Vitamin B¹**, also known as thiamine, is important for the nervous system and energy production; food sources for thiamine include meat, peas, fortified cereals, bread, and whole grains.
- **Vitamin B²**, also known as riboflavin, is important for the 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 B³**, 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 B⁶**, 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 B¹²** 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 medical conditions, 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; 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, the doctor may encourage deviations from the average official recommendation based on your child's 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:³⁸

- **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.

³⁸ Adapted from the FDA: <http://www.fda.gov/fdac/special/foodlabel/dvs.html>.

- **RDIs (Reference Daily Intakes):** A set of dietary references based on the Recommended Dietary Allowances for essential vitamins and minerals 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?³⁹

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.”⁴⁰ According to the ODS, dietary supplements can have an important impact on the prevention and management of medical conditions and on the maintenance of health.⁴¹ The ODS notes that considerable research on the effects of dietary supplements has been conducted in Asia and Europe where

³⁹ This discussion has been adapted from the NIH:
<http://ods.od.nih.gov/whatare/whatare.html>.

⁴⁰ 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.

⁴¹ 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.”

the use of plant products, in particular, has a long tradition. However, the overwhelming majority of supplements have not been studied scientifically. 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 Moyamoya Disease

The NIH maintains an office dedicated to 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.⁴² 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

⁴² 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.

periodically as this database is frequently updated. More studies can be found by searching the Full IBIDS Database. Healthcare professionals and 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 “Moyamoya disease” (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 “Moyamoya disease” (or a synonym):

- **A case report of Moyamoya disease: successfully treated with Chinese medicine.**
 Author(s): Department of Japanese-Oriental Medicine, Toyama Medical and Pharmaceutical University Hospital, Japan.
 Source: Hiyama, Y Itoh, T Shimada, Y Shimada, T Terasawa, K Am-J-Chin-Med. 1992; 20(3-4): 319-24 0192-415X
- **Moyamoya disease of childhood as a cause of recurrent cerebral ischemic attacks--a case report.**
 Author(s): Department of Paediatrics, Aga Khan University, Karachi.
 Source: Ibrahim, S Hyder, S S J-Pak-Med-Assoc. 1996 January; 46(1): 17-8 0030-9982

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:

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]

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

Intestinal: Pertaining to the intestine. [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]

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]

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]

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]

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.⁴³

⁴³ 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):⁴⁴

- **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/>

⁴⁴ 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.lsuhscc.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 Resource 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://nmlm.gov/>
- **National:** NN/LM List of Libraries Serving the Public (National Network of Libraries of Medicine), <http://nmlm.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.html#conshealth.html#
- **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/koopp1.shtml>
- **Pennsylvania:** Learning Resources Center - Medical Library (Susquehanna Health System), <http://www.shscare.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://hwh.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 CHILD'S RIGHTS AND INSURANCE

Overview

Parents face a series of issues related more to the healthcare industry than to their children's medical conditions. This appendix covers two important topics in this regard: your responsibilities and your child's rights as a patient, and how to get the most out of your child's medical insurance plan.

Your Child's 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 child's rights as a patient.⁴⁵

⁴⁵Adapted from Consumer Bill of Rights and Responsibilities:
<http://www.hcqualitycommission.gov/press/cbor.html#head1>.

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 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.
- **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 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 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 parents with sufficient information and opportunity to decide among treatment options consistent with the informed consent process.
- Discuss all treatment options with a parent 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 parents the opportunity to refuse treatment for their children 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 parents.
- Abide by the decisions made by parents 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 parents about medically necessary treatment options for their children.
- 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.⁴⁶

Parent Responsibilities

To underscore the importance of finance in modern healthcare as well as your responsibility for the financial aspects of your child's care, the President's Advisory Commission on Consumer Protection and Quality in the Healthcare Industry has proposed that parents understand the following "Consumer Responsibilities."⁴⁷ In a healthcare system that protects consumers' rights, it is reasonable to expect and encourage consumers to assume certain responsibilities. Greater involvement by parents in their children's 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 your child's healthy habits.
- Work collaboratively with healthcare providers in developing and carrying out your child's agreed-upon treatment plans.
- Disclose relevant information and clearly communicate wants and needs.

⁴⁶ 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.

⁴⁷ Adapted from <http://www.hcqualitycommission.gov/press/cbor.html#head1>.

- Use the insurance company's internal complaint and appeal processes to address your concerns.
- 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 the community.
- Become knowledgeable about health plan 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.
- 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.⁴⁸ The U.S. Department of Labor, in particular, recommends ten ways to make your health benefits choices work best for your family.⁴⁹

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 family's 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 face. Cheapest may not always be best. Your goal is high quality health benefits.

⁴⁸ More information about quality across programs is provided at the following AHRQ Web site:

<http://www.ahrq.gov/consumer/qntascii/qnthplan.htm>.

⁴⁹ Adapted from the Department of Labor:

<http://www.dol.gov/dol/pwba/public/pubs/health/top10-text.html>.

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 your family. 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 family's benefits and your legal rights under the Employee Retirement Income Security Act (ERISA), the federal law that protects your family's 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 family's 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 your family 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 a 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 your family receives. 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.

Medicaid

Illness strikes both rich and poor families. For low-income families, Medicaid is available to defer the costs of treatment. In the following pages, you will learn the basics about Medicaid as well as useful contact information on how to find more in-depth information.

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. You can find more information about Medicaid on the HCFA.gov Web site at <http://www.hcfa.gov/medicaid/medicaid.htm>.

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.⁵⁰ NORD programs ensure that certain vital drugs are available "to those families 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 medical conditions. 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:⁵¹

- 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>

⁵⁰ Adapted from NORD: http://www.rarediseases.org/cgi-bin/nord/proserv#patient?id=rPIzL9oD&mv_pc=30.

⁵¹ You can access this information at: <http://www.nlm.nih.gov/medlineplus/healthsystem.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>
- 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 Moyamoya disease 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>

MOYAMOYA DISEASE 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.

Acne: An inflammatory disease of the pilosebaceous unit, the specific type usually being indicated by a modifying term; frequently used alone to designate common acne, or acne vulgaris. [EU]

Acuity: Clarity or clearness, especially of the vision. [EU]

Aneurysm: A sac formed by the dilatation of the wall of an artery, a vein, or the heart. The chief signs of arterial aneurysm are the formation of a pulsating tumour, and often a bruit (aneurysmal bruit) heard over the swelling. Sometimes there are symptoms from pressure on contiguous parts. [EU]

Angiography: Radiography of blood vessels after injection of a contrast medium. [NIH]

Anticoagulants: Agents that prevent blood clotting. Naturally occurring agents in the blood are included only when they are used as drugs. [NIH]

Aphasia: Defect or loss of the power of expression by speech, writing, or signs, or of comprehending spoken or written language, due to injury or disease of the brain centres. [EU]

Arteries: The vessels carrying blood away from the heart. [NIH]

Arteriovenous: Both arterial and venous; pertaining to or affecting an artery and a vein. [EU]

Assay: Determination of the amount of a particular constituent of a mixture, or of the biological or pharmacological potency of a drug. [EU]

Ataxia: Failure of muscular coordination; irregularity of muscular action. [EU]

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]

Cardiology: The study of the heart, its physiology, and its functions. [NIH]

Cerebellar: Pertaining to the cerebellum. [EU]

Cerebellum: Part of the metencephalon that lies in the posterior cranial fossa behind the brain stem. It is concerned with the coordination of movement. [NIH]

Cerebral: Of or pertaining of the cerebrum or the brain. [EU]

Cerebrovascular: Pertaining to the blood vessels of the cerebrum, or brain. [EU]

Choroid: The thin, highly vascular membrane covering most of the posterior of the eye between the retina and sclera. [NIH]

Consciousness: Sense of awareness of self and of the environment. [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]

Cortical: Pertaining to or of the nature of a cortex or bark. [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]

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

Extrapyramidal: Outside of the pyramidal tracts. [EU]

Ganglioglioma: Rare indolent tumors comprised of neoplastic glial and neuronal cells which occur primarily in children and young adults. Benign lesions tend to be associated with long survival unless the tumor degenerates into a histologically malignant form. They tend to occur in the optic nerve and white matter of the brain and spinal cord. [NIH]

Granule: A small pill made from sucrose. [EU]

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

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]

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

Infarction: 1. the formation of an infarct. 2. an infarct. [EU]

Intermittent: Occurring at separated intervals; having periods of cessation of activity. [EU]

Intestinal: Pertaining to the intestine. [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]

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

Lipid: Any of a heterogeneous group of fats 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]

Lipoprotein: Any of the lipid-protein complexes in which lipids are transported in the blood; lipoprotein particles consist of a spherical hydrophobic core of triglycerides or cholesterol esters surrounded by an amphipathic monolayer of phospholipids, cholesterol, and apolipoproteins; the four principal classes are high-density, low-density, and very-low-density lipoproteins and chylomicrons. [EU]

Lobe: A more or less well-defined portion of any organ, especially of the brain, lungs, and glands. Lobes are demarcated by fissures, sulci, connective tissue, and by their shape. [EU]

Malformation: A morphologic defect resulting from an intrinsically abnormal developmental process. [EU]

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

Morphogenesis: The development of the form of an organ, part of the body, or organism. [NIH]

Neoplastic: Pertaining to or like a neoplasm (= any new and abnormal

growth); pertaining to neoplasia (= the formation of a neoplasm). [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]

Neuroanatomy: Study of the anatomy of the nervous system as a specialty or discipline. [NIH]

Neurology: A medical specialty concerned with the study of the structures, functions, and diseases of the nervous system. [NIH]

Neuronal: Pertaining to a neuron or neurons (= conducting cells of the nervous system). [EU]

Neurosurgery: A surgical specialty concerned with the treatment of diseases and disorders of the brain, spinal cord, and peripheral and sympathetic nervous system. [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]

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]

Paralysis: Loss or impairment of motor function in a part due to lesion of the neural or muscular mechanism; also by analogy, impairment of sensory function (sensory paralysis). In addition to the types named below, paralysis is further distinguished as traumatic, syphilitic, toxic, etc., according to its cause; or as obturator, ulnar, etc., according to the nerve part, or muscle specially affected. [EU]

Pathologic: 1. indicative of or caused by a morbid condition. 2. pertaining to pathology (= branch of medicine that treats the essential nature of the disease, especially the structural and functional changes in tissues and organs of the body caused by the disease). [EU]

Pediatrics: A medical specialty concerned with maintaining health and providing medical care to children from birth to adolescence. [NIH]

Plexus: A network or tangle; a general term for a network of lymphatic vessels, nerves, or veins. [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]

Postnatal: Occurring after birth, with reference to the newborn. [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]

Prenatal: Existing or occurring before birth, with reference to the fetus. [EU]

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

Psychiatric: Pertaining to or within the purview of psychiatry. [EU]

Pulmonary: Pertaining to the lungs. [EU]

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]

Resection: Excision of a portion or all of an organ or other structure. [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]

Sclerosis: A induration, or hardening; especially hardening of a part from inflammation and in diseases of the interstitial substance. The term is used chiefly for such a hardening of the nervous system due to hyperplasia of the connective tissue or to designate hardening of the blood vessels. [EU]

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]

Seizures: Clinical or subclinical disturbances of cortical function due to a sudden, abnormal, excessive, and disorganized discharge of brain cells. Clinical manifestations include abnormal motor, sensory and psychic phenomena. Recurrent seizures are usually referred to as epilepsy or "seizure disorder." [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]

Symptomatic: 1. pertaining to or of the nature of a symptom. 2. indicative

(of a particular disease or disorder). 3. exhibiting the symptoms of a particular disease but having a different cause. 4. directed at the allaying of symptoms, as symptomatic treatment. [EU]

Technetium: The first artificially produced element and a radioactive fission product of uranium. The stablest isotope has a mass number 99 and is used diagnostically as a radioactive imaging agent. Technetium has the atomic symbol Tc, atomic number 43, and atomic weight 98.91. [NIH]

Teratogenic: Tending to produce anomalies of formation, or teratism (= anomaly of formation or development : condition of a monster). [EU]

Thermoregulation: Heat regulation. [EU]

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

Tomography: The recording of internal body images at a predetermined plane by means of the tomograph; called also body section roentgenography. [EU]

Tone: 1. the normal degree of vigour and tension; in muscle, the resistance to passive elongation or stretch; tonus. 2. a particular quality of sound or of voice. 3. to make permanent, or to change, the colour of silver stain by chemical treatment, usually with a heavy metal. [EU]

Toxoplasmosis: An acute or chronic, widespread disease of animals and humans caused by the obligate intracellular protozoon *Toxoplasma gondii*, transmitted by oocysts containing the pathogen in the feces of cats (the definitive host), usually by contaminated soil, direct exposure to infected feces, tissue cysts in infected meat, or tachyzoites (proliferating forms) in blood. [EU]

Transfusion: The introduction of whole blood or blood component directly into the blood stream. [EU]

Venereal: Pertaining or related to or transmitted by sexual contact. [EU]

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

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 Acronyms & Abbreviations** by Stanley Jablonski (Editor), Paperback, 4th edition (2001), Lippincott Williams & Wilkins

Publishers, ISBN: 1560534605,

<http://www.amazon.com/exec/obidos/ASIN/1560534605/iconegroupinterna>

- **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/iconegroupinterna>
- **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/iconegroupinterna>
- **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/iconegroupinterna>
- **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/iconegroupinterna>
- **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/iconegroupinterna/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/iconegroupinterna>
- **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/iconegroupinterna>
- **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/iconegroupinterna>
- **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/iconegroupinterna>

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