Arachnoid Cysts

989

Copyright (C) 1994 National Organization for Rare Disorders, Inc.

**IMPORTANT**
It is possible that the main title of this report (Arachnoid Cysts) is not the name you expected. Please check the SYNONYMS listing to find the alternate name(s) and disorder subdivision(s) covered by this report.

**Synonyms**
Cysts, Arachnoid

*Information on the following diseases can be found in the Related Disorders section of this report:*
Acoustic Neuroma
Arachnoiditis
Brain Tumors, General
Dandy-Walker Syndrome
Empty Sella Syndrome
Epidermoids
Midline Caves of the Brain
Porencephaly
Syringomyelia
Bobble-Head Doll Syndrome
Hyperprolactinemia
Panhypopituitarism

**General Discussion**

**REMINDER**
The information contained in the Rare Disease Database is provided for educational purposes only. It should not be used for diagnostic or treatment purposes. If you wish to obtain more detailed information about this disorder, please contact your personal physician and/or the agencies listed in the "Resources" section of this report.

Arachnoid Cysts are fluid-filled sacs that occur on the arachnoid membrane that covers the brain and the spinal cord. There are three membranes covering these components of the central nervous system: dura mater, arachnoid, and pia mater. Arachnoid cysts appear on the arachnoid membrane, and they may also expand into the space between the pia mater and arachnoid membranes (subarachnoid space).
Arachnoid Cysts

While Arachnoid Cysts in general are not rare, they are extremely rare in some locations. Arachnoid Cysts do not usually appear within the two hemispheres of the brain (cerebrum) or within the cavities (ventricles) of the brain.

Symptoms

Arachnoid Cysts may be present at birth (congenital) and they may cause no symptoms throughout an individual's life. They are often discovered when a person seeks medical attention for headaches or seizures. When Arachnoid Cysts increase in size over time they can cause symptoms to appear, especially if they press against a cranial nerve, the brain, or the spinal cord. The symptoms are determined by the location of the cyst on the arachnoid membrane.

Major symptoms of Arachnoid Cysts may include an enlarged head (macrocephaly), disorders of hearing or vision, headaches, dizziness and instability, delayed motor development (psychomotor retardation), epileptic seizures, and/or premature sexual development. Symptoms of Arachnoid Cysts may also include nausea, loss of the sense of smell (olfactory loss), loss of coordinated movement (ataxia) in the arms and legs, and/or paralysis of lower limbs (paraplegia). In children, an enlarged head, continuous bobbing of the head, skin that is very sensitive to the touch (hypsersensitivity), and/or mental retardation may also be symptoms.

Arachnoid Cysts of the posterior fossa may cause vague symptoms such as dizziness and/or headache, which are often associated with inner ear problems. Individuals with long-standing complaints related to the ear (otologic problems), and who have normal hearing tests, should be referred for specialized diagnostic tests such as computerized tomography (CT scan) or magnetic resonance imaging (MRI).

Arachnoid Cysts frequently cause symptoms that may also occur in other disorders such as brain tumors or malformations of the brain. Differential diagnosis is usually possible by imaging techniques such as CT scan or MRI.

Complications of Arachnoid Cysts can occur when a cyst is damaged because of trauma. Trauma can cause the fluid within a cyst to leak into other areas (e.g., subarachnoid space). Blood vessels on the surface of a cyst may tear and bleed into the cyst (intracystic hemorrhage), increasing its size. If a blood vessel bleeds on the outside of a cyst, a collection of blood (hematoma) may result. In the cases of intracystic hemorrhage and hematoma, the individual may have symptoms of increased pressure within the cranium and signs of compression of nearby nerve (neural) tissue.

Premature sexual development may occur secondary to a Suprasellar Arachnoid Cyst. Symptoms of this type of cyst may include head nodding, abnormal gait, and/or abnormalities of vision.

Causes

The cause of Arachnoid Cysts is unknown. In some people they may be present at birth (congenital). It is believed that they may occur as a result of developmental disturbances of the membranes that cover the brain and the spinal cord (the meninges), or they may develop in response to a prenatal (before birth) inflammation of the meninges. It has been reported that in some families Arachnoid Cysts appear to affect more family members than would be expected by chance (familial).

In a few rare cases, Intracranial Arachnoid Cysts may be inherited as an autosomal recessive trait. In
Arachnoid Cysts

recessive disorders, the condition does not appear unless a person inherits the same defective gene for the same trait from each parent. If an individual receives one normal gene and one gene for the disease, the person will be a carrier for the disease, but usually will not show symptoms. The risk of transmitting the disease to the children of a couple, both of whom are carriers for a recessive disorder, is 25 percent. Fifty percent of their children risk being carriers of the disease, but generally will not show symptoms of the disorder. Twenty-five percent of their children may receive both normal genes, one from each parent, and will be genetically normal (for that particular trait). The risk is the same for each pregnancy.

There are several reports in the medical literature of Arachnoid Cysts and intramedullary spinal cord cysts occurring together in people who have had spinal surgery and/or spinal cord trauma.

Affected Population

Arachnoid Cysts affect males and females equally. Some scientists feel that Intracranial Arachnoid Cysts occur more frequently in males. These cysts may occur at any age and have been found in all races and geographic locations. Arachnoid Cysts are not rare in the general population, but they rarely occur in certain areas of the central nervous system.

Related Disorders

Symptoms of the following disorders can be similar to those of Arachnoid Cysts. Comparisons may be useful for a differential diagnosis:

Acoustic Neuroma (Cerebellopontine Angle Syndrome) is a benign (noncancerous) tumor of the 8th cranial nerve. The early symptoms of an Acoustic Neuroma include a ringing sound in the ear (tinnitus) and/or a hearing loss. These symptoms occur due to pressure from the tumor on the 8th cranial nerve. (For more information on this disorder, choose "Acoustic Neuroma" as your search term in the Rare Disease Database.)

Arachnoiditis is a progressive inflammatory disorder of the arachnoid membrane, with possible involvement of the brain and spinal cord. Cerebral involvement leads to symptoms such as severe headaches, visual disturbances, dizziness, nausea, and vomiting. Spinal involvement may cause pain, weakness, and paralysis. (For more information on this disorder, choose "Arachnoiditis" as your search term in the Rare Disease Database.)

Brain Tumors are growths in the brain that can be either cancerous (malignant) or noncancerous (benign). The symptoms of malignant and benign brain tumors are similar, and depend on the type of tumor and its location. Symptoms may include recurrent headache, seizures, emotional instability, nausea, and/or facial pain or numbness. There are many different types of Brain Tumor. (For more information on these disorders, choose "Brain Tumor" as your search term in the Rare Disease Database.)

Dandy-Walker Syndrome is characterized by congenital accumulation of fluid in the brain (hydrocephalus) resulting from obstruction of passages between the fourth ventricle of the brain and the subarachnoid space (foramina of Magendie and Luschka). Hydrocephalus may be accompanied by headache, episodes of visual disturbances, and/or abnormally large amounts of fluid in the tissue of the optic disk (papilledema) in affected infants. (For more information on this disorder, choose "Dandy-Walker Syndrome" as your search term in the Rare Disease Database.)
Arachnoid Cysts

Empty Sella Syndrome is a rare brain disorder in which the depression across the bone that contains the pituitary gland (sella turcica) appears as an extension of the subarachnoid space and is filled with cerebrospinal fluid. The syndrome may be primary or may be secondary to a pituitary tumor, irradiation, or surgery on the pituitary gland. Symptoms may include headaches, impaired vision, and obesity. Sex-specific differences may be seen, such as excessive hair growth (hirsutism) in women and reduced sex drive (libido) in men. (For more information on this disorder, choose "Empty Sella" as your search term in the Rare Disease Database.)

Epidermoids are tumors that are thought to arise from fragments of skin that have been displaced (ectopic epithelium). This can be a result of traumatic injury, inflammation, or a disturbance of embryonic development. Symptoms typically depend on the location of the tumor, and may include headache, confusion, numbness and partial paralysis of the legs (paraparesis), sensory disturbances, and/or back and leg pain.

Midline Cysts of the Brain are cavities that are normally present within the brain. A change in shape and/or size of these cavities may indicate that a lesion is present (e.g., a tumor or cyst). Midline caves may form sacs that put pressure on different parts of the brain and cause symptoms similar to those of a tumor, such as headache and/or nausea. CT scan or MRI examination can identify midline caves.

Porencephaly is a major congenital malformation of the brain that is characterized by cysts or the formation of cavities (cavitations) within the brain. The symptoms of porencephaly usually include mental retardation, spastic paralysis, mild paralysis of cranial nerves (ophthalmoplegia), and/or epilepsy. Porencephaly can be diagnosed by CT scan examination.

Syringomyelia is a slowly progressive neurological disorder in which the formation of cavities (cavitation) occurs in the spinal cord. The cavity (syrinx) is usually situated near the middle of the spinal cord (paramedian) and may extend up into the brain stem area (medulla oblongata) or down as far as the base of the spine (lumbar region). Symptoms may include loss of pain and temperature perception in the arms and chest, spreading to the shoulders and back. (For more information on this disorder, choose "Syringomyelia" as your search term in the Rare Disease Database.)

The following disorders may be associated with Arachnoid Cysts as secondary characteristics. They are not necessary for a differential diagnosis:

Bobble-Head Doll Syndrome is a rare disorder that appears in childhood and is characterized by accumulated fluid in the brain (hydrocephalus) and/or obesity. Symptoms may include a continuous bobbing of the head, rhythmical bending and straightening of the head and arms, a generalized fine tremor, skin that is very sensitive to the touch, mental retardation, and/or impaired vision. Bobble-head Doll Syndrome can be caused by any obstruction that results in the enlargement of the third ventricle of the brain. Draining the fluid from the cyst, removal of the cyst, and/or insertion of a shunt are standard therapies.

Hyperprolactinemia occurs when there is an abnormally high amount of prolactin in the blood. Hyperprolactinemia has been associated with the extension of Arachnoid Cysts into the suprasellar cistern. Symptoms in women may include absence or abnormal cessation of menstruation (amenorrhea).
Arachnoid Cysts

and/or spontaneous milk flow from the breasts (galactorrhea). Symptoms in men have been reported to include impotence and/or abnormally decreased function of the testes.

Panhypopituitarism is a rare disorder characterized by a generalized reduction or cessation of pituitary gland function. This can result when an expanding Intracellar Arachnoid Cyst causes the pituitary gland to become flattened. Such a cyst can be mistaken for a pituitary tumor. A CT scan or MRI may be necessary for proper diagnosis.

Standard Therapies

If it is decided that treatment of an Arachnoid Cyst is necessary, therapy usually consists of surgical removal and/or drainage of the cyst. After the cyst is drained, it may be necessary to surgically insert a device to provide a passageway for fluid from the cyst to one of the reservoirs for bodily fluids (a shunt). This may prevent a recurrence of fluid build-up.

If the subarachnoid space has become blocked by an Arachnoid Cyst, it may also be necessary to place a shunt between a ventricle and the space between the layers of the membrane that lines the cavities of the body (the peritoneal cavity). This will bypass the blockage and provide an adequate passageway for cerebrospinal fluid to circulate.

Investigational Therapies

In one study, three individuals who had bilateral hearing loss as a result of Arachnoid Cysts were treated with diuretics. This therapy resulted in improvement in symptoms, with no evidence of enlargement of the cysts during several years of follow-up. More research is needed to determine whether diuretic drugs may help other people with this type of Arachnoid Cyst.

This disease entry is based upon medical information available through February 1994. Since NORD's resources are limited, it is not possible to keep every entry in the Rare Disease Database completely current and accurate. Please check with the agencies listed in the Resources section for the most current information about this disorder.

Resources

For more information on Arachnoid Cysts, contact the referrals listed below, which are in alphabetical order.

National Organization for Rare Disorders, Inc. (NORD)
P.O. Box 8923
New Fairfield, CT 06812-8923
(203) 746-6518
(203) 746-6481 (Fax)
(800) 999-6673
(203) 746-6927 (TDD for the hearing impaired)
e-mail: orphan@nord-rdb.com
Home Page: http://www.nord-rdb.com/orphan

Brain and Pituitary Foundation of America

Date: 1/14/97 11:57:54 AM  Last Edit Date: 6/20/96
Arachnoid Cysts
281 East Moody Avenue
Fresno CA 93720-1524
Telephone: (209) 434-0610

NIH/National Institute of Neurological Disorders and Stroke
31 Center Dr MSC 2540
Building 31 Rm 8A16
Bethesda MD 20892
Telephone: (301) 496-5751
Toll Free: (800) 352-9424

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
TDD: (817) 277-0553
e-mail: thearc@metronet.com

Worldwide Education and Awareness for Movement Disorders
Mt. Sinai Center
One Gustave L. Levy Place Box 1052
New York NY 10029
Telephone: (212) 241-8567
Toll Free: (800) 437-6682
e-mail: wemove@smtplink.mssm.edu.

References


TEXTBOOK OF UNCOMMON CANCER, 1st Ed.: C.J. Williams, Editor; John Wiley & Sons, Ltd.
Arachnoid Cysts


