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Neurology Defined

Eponyms in Neurosciences

Kalyan B Bhattacharyya

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SECTION

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Abadie's sign There are two different signs associated with the name of Joseph Abadie of France.

- Loss of pain sensation on squeezing Achilles tendon in tabes dorsalis
- Spasm of levator palpebrae superioris muscle in thyrotoxicosis. This contributes partly to the wide palpebral fissure.

Achilles jerk The other name for ankle jerk.

Adamkiewicz artery It is the largest anterior segmental artery supplying the spinal cord. It typically arises from the left side from the abdominal aorta at the level of 9th to 12th intercostal artery and supplies the lower two-third of the spinal cord through the anterior spinal artery. Sometimes it arises from a lumbar vessel. Occlusion of this artery leads to motor weakness in the lower limbs along with urinary and fecal incontinence with preservation of posterior column functions, since the posterior part of the spinal cord is not supplied by this artery and therefore, spared.



Albert Wojciech Adamkiewicz (1850–1921, Poland)

Adie's pupil Tonic pupil which dilates slowly in darkness and takes a long time to constrict in response to light. The accommodation reflex is normal. It is more commonly seen in women and when associated with absent knee or ankle jerk, it is known as *Holmes-Adie syndrome*, while concomitant impaired sweating in one side of the body is known as *Ross' syndrome*. The lesion lies in the ciliary ganglion, supplying postganglionic fibers

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of parasympathetic nerves to the iris following a viral infection. Abnormal sweating is consequent upon degeneration of the posterior root ganglion. Diagnosis is established by the observation of constriction of the pupil with low dose of pilocarpine, indicating denervation supersensitivity of Walter Cannon.



William John Adie
(1886–1935, United Kingdom)

Adie-Critchley syndrome
Forced grasping and groping reaction in tumor of the contralateral frontal lobe.



Macdonald Critchley
(1900–1999, United Kingdom)

Adrian's all-or-none law
The principle that the strength by which a nerve or muscle fiber responds to a stimulus is independent of the strength of the stimulus. If that stimulus exceeds the threshold potential, the nerve or muscle fiber will offer a complete response; otherwise, there is no response. It was first established by the American physiologist Henry Pickering Bowditch in 1871 for the contraction of heart muscle. However, it was proved experimentally by Edgar Adrian of Cambridge in 1928 which earned him the Nobel Prize in 1932.



Edgar Douglas Adrian
(1889–1977, United Kingdom)

Adson's test A sign of thoracic outlet syndrome. The patient's arm is extended at the elbow and abducted and rotated posteriorly. There is loss of pulsation of the

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radial pulse when the patient rotates his head to the side tested and the examiner rotates and extends the patient's shoulder. The patient takes a deep breath and is asked to hold it and the test is considered positive if there is loss of radial pulse along with the appearance of radicular symptoms like, paresthesia. Sometimes, there is a bruit over the brachial artery. The hand may turn pale and is known as '*white hand sign*'. The test is no longer considered as that reliable, since many normal subjects show diminution of the radial pulse with this maneuver. *Roo's maneuver* is a related test, where the patient elevates and abducts his arms and identical symptoms are reproduced after 3 minutes.

Aicardi syndrome A constellation of agenesis of corpus callosum, infantile spasms and chorioretinal lacunae. Myoclonus, intellectual decline, microcephaly and optic nerve hypoplasia with morning glory appearance in the retina are additional features in some cases. The condition is seen exclusively in girls. It is an X-linked dominant condition at Xp22 locus and male fetuses die in utero. It has been described in boys with Klinefelter's syndrome.



Jean Aicardi
(1926–2015, France)

Alexander's disease A fatal neurodegenerative condition in infants, characterized by developmental delay, enlarged head, seizures, spasticity, dementia and sometimes, idiopathic intracranial hypertension. There is mutation in the GFAP gene and the genetic defect in this autosomal dominant condition is in chromosome 17q21 position, though sporadic cases are commoner. Pathologically, megaencephaly is observed. It is a variety of leukodystrophy characterized by the presence of a variety of protein, *Rosenthal fibers*, in the astrocytes of the brain. Other causes of

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leukodystrophy with enlarged head include Canavan's disease and Van der Knapp's disease or megaencephalic leukodystrophy with subcortical cysts. In India, the latter condition has been described in the Agarwal community by Singhal and in a single Bengali girl by Bhattacharyya.

Alexander's law In spontaneous nystagmus due to unilateral vestibular lesion, the amplitude of the fast component increases when the subject looks towards the quick phase.

Alice in Wonderland syndrome It is a syndrome complex of micro- or macropsia, pelopsia (a feeling that subjects are nearer than they actually are), teleopsia (reverse of pelopsia), etc., and altered perception of body image. This is associated with migrainous aura or intake of psychotropic or psychedelic agents, like LSD. The name comes from the description of Lewis Carroll in Alice in Wonderland and it is believed that Carroll himself suffered from migraine.

Alpers' disease Also known as Alpers-Huttenlocher syndrome, it is a progressive neurodegenerative disorder of mitochondrial DNA depletion, characterized by delayed milestones, intractable seizures, particularly epilepsia partialis continua, myoclonus, areflexia, spasticity, dementia, optic atrophy and deafness. It is an autosomal recessive disorder occurring typically in childhood and is fatal. The disease can manifest as myopathic, encephalomyopathic, or hepatopathic patterns. The concerned genes are TK2 in myopathic, SUCL2 and RRM2B in encephalomyopathic, and DGUOK, POLG and MPV17 in hepatopathic variety.

Alport syndrome An oculorenal syndrome characterized by a triad of clinical findings consisting of hemorrhagic nephritis, sensorineural hearing loss and lenticonus, keratoconus, and cataracts. Diffuse leiomyomatosis and dissection of the aorta are occasionally observed. Most of the cases are of X-linked recessive inheritance with mutation in the COL4A3, COL4A4 and COL4A5 gene.

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Alzheimer's baskets Condensed clumps of filaments between the nerve cells seen in advanced cases of Alzheimer's disease.

Alzheimer's cell type 1 Large multinucleated astrocytes found in glial tumor and progressive multifocal leukoencephalopathy.

Alzheimer's cell type 2 Swollen astrocytes with large nucleus and a significant nucleolus with little cytoplasm. They are metabolically active and found in cortex, brainstem, cerebellum and thalamus. They are classically found in hyperammonemic conditions like, hepatic encephalopathy, hepatocerebral degeneration and Wilson's disease.

Alzheimer's disease (The reader is referred to any standard textbook of neurology)



Alois Alzheimer (1864–1915, Germany)

Alzheimer's sclerosis Degeneration of the middle and smaller cerebral blood vessels at a cellular level.

Alzheimer's stain A staining method for the detection of Negri bodies in rabies.

Anderson's disease The other name for glycogen storage disease type IV characterized by enzyme deficiencies affecting either glycogen synthesis, glycogen breakdown, or glycolysis, classically within the muscles. Of the various manifestations, the neuromuscular ones consist of late childhood development of myopathy and mild muscular weakness.

Angelman syndrome It is characterized by intellectual and motor developmental delay, microcephaly, speech and balance problem and seizures. The affected children are usually of an affable nature, hyperexcitable and they often flap their hands. They exhibit the tendency to handle water. The condition is caused by lack of function of the gene UBE3A, in chromosome 15, which is inherited from the mother. Sometimes, it is due to

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inheritance of two copies of the chromosome from the father and none from the mother. *Prader–Willi syndrome* is a related condition where the disease is inherited from the father.

Anton syndrome Also known as *Anton–Babinski syndrome*, it is a condition where a blind person denies blindness and in order to prove their point, they tend to confabulate as if they can see. This happens with bilateral occipital infarction following posterior cerebral infarction, or as a complication of JC virus infection following the use of natalizumab in multiple sclerosis. It has also been described in rare cases of adrenoleukodystrophy, where both the occipital lobes undergo dysmyelination. Thus, it is a variety of visual anosognosia.



Gabriel Anton
(1858–1933, Austria)

Antoni A and B patterns In schwannoma, two varieties of cells are found in histological study. The fibrillary, polar and elongated tissue is known as *Antoni A pattern*, whereas the loose, less dense microcystic tissues in close proximity is known as *Antoni B pattern*. Antoni A is also known as *Verocay bodies*.

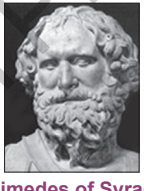
Apert syndrome A variety of acrocephalosyndactylism, where there is pronounced malformations in the bones in the skull, face, hands and feet, affecting the first branchial arch. Various malformations of the skull like, brachycephaly (the commonest variety, where the coronal and lambdoid sutures undergo fusion, leading to a transversely broad face and the parietal bones form the sides and the top of the head), trigonocephaly, dolicocephaly, plagiocephaly, turricephaly and oxycephaly have been described depending on the nature of fusion of cranial sutures in uterus. Syndactylism is a common accompaniment and the eyes protrude due to small orbit following premature fusion of the sutures between the

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frontal, sphenoid, zygomatic and maxillae. There is mutation in the FGFR2 gene which produces fibroblast growth factor and immature cells turn into osseous tissue. The condition is inherited as an autosomal dominant trait.

Aran-Duchenne disease The other name for amyotrophic lateral sclerosis.

Archimedes spiral This refers to drawing a spiral smoothly and elegantly from the center of the page to the periphery. In normal subjects it is well organized. However, in tremulous conditions, like in essential tremor or Parkinson's disease, it is clumsy and the gaps between the curves are irregular. In Parkinson's disease, the space between the lines is short, while the reverse is true for essential tremor.



Archimedes of Syracuse (287–212 BC, Greece)

Argyll Robertson pupil A classic sign of neurosyphilis, particularly in tabes dorsalis. The pupils are small, eccentric, and irregular in the margin, reacts to accommodation but not to light reflex. The iris is brownish in color. The lesion is thought to be in the pretectal area of the midbrain and the Edinger–Westphal nucleus of the Warwick oculomotor nucleus complex. Syphilitic iritis producing local scarring is the possible etiology of the iris pathology. Other mesencephalic lesions like, diabetes mellitus with autonomic neuropathy, multiple sclerosis, Wernicke's encephalopathy, sarcoidosis, optic nerve disease, among other conditions, can also present with such a clinical condition and they have been assigned the name, *pseudo Argyll Robertson pupil*. A colorful name, *prostitute's pupil*, has also been assigned to it. *Reverse Argyll Robertson pupil*, where light reflex is normal but accommodation reflex is defective it is sometimes found in postencephalitic parkinsonism and Burkitt's lymphoma.

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Argyll Robertson
(1837–1909, United Kingdom)

Arnold–Chiari malformations Structural defects in the cerebellum consisting of downward displacement of the cerebellar tonsils through the foramen magnum, sometimes causing non-communicating hydrocephalus. Classically, it is divided into four types, Type I, where there is tonsillar ectopia below the foramen magnum, sometimes with syringomyelia. The medulla and the brain stem may be elongated and kinked, while craniovertebral anomalies may be associated. Type II consists of displacement of the cerebellar vermis. It is sometimes associated with spina bifida and lumbar myelomeningocoele. Type III is associated with occipital encephalocoele, while Type IV is pure cerebellar hypoplasia, associated with Dandy–Walker syndrome. Cerebellar and pyramidal tract signs are the presenting features (Fig. A.1).

Figure A.1: Arnold–Chiari malformations. Note the descent of cerebellar tonsil below the foramen magnum





Hans Chiari
(1851–1916, Austria)

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Julius Arnold
(1835–1915, Germany)

Ashworth scale A scale to assess the degree of spasticity. The grading runs from 0 or no spasticity to 4, when there is extreme spasticity in flexion or extension in ascending order of severity.

Asperger syndrome A disorder of development, it is characterized by difficulties in social interaction and nonverbal communication. Language is unaffected and intelligence is within normal limits. Some of them display stereotyped behavioral patterns like, preoccupation with one activity like stacking cans in a rack or arranging books for a long period of time, and the condition is often considered as a mild autism spectrum disorder. Flapping or twisting body movements or bending the whole body is a characteristic feature in some subjects. The speech is marked by limited prosody, tangentiality, circumstantiality and verbosity. Sometimes they can make socially inappropriate statements when strangers approach them, often known as verbal dysdecorum, or may yawn loudly during conversation. Some may show unusual hypersensitivity to sound, light and touch. Some have difficulty in falling asleep or maintaining it. No specific inheritance pattern is yet recognized.

Axenfeld-Schürenberg syndrome This is a congenital unilateral third cranial nerve palsy with mydriasis and alternate phases of pupillary constriction and contraction of muscles supplied by the oculomotor nerve. This leads to elevation of the upper lid and nasal deviation of the eye. This is autosomal dominant in inheritance and is also known as cyclic oculomotor palsy.

Avellis syndrome A variety of alternating paralysis involving the soft palate and the vocal cord

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of one side and hemianesthesia on the other side. It is usually the complication of occlusion of vertebral artery in lesions in the nucleus ambiguous and pyramidal tract in brain stem infarction. Sometimes, it is associated with Horner's syndrome with involvement of vagus and glosso-pharyngeal nerves.

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