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Clinical Presentation of Hydrocephalus

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1. Introduction

The two key determinants of clinical presentation of hydrocephalus are the age of onset and the acuity of the rise in intracranial pressure.

In fetus, while minor degree of hydrocephalus often goes undetected, severe cases of obstructive variety present with the following features: 1 .the head is felt larger, globular and softer than the normal head 2.the head is high up and difficult to push down into pelvis 3.fetal heart sound is situated high up above the umbilicus 4.internal examination during labor may reveal widening of sutures and tense fontanelle. In breech presentation however, the diagnosis is not made until the after-coming head is arrested at the brim. In the communicating variety, often the head is of normal size at birth and its enlargement starts only at 6-12 weeks of age.

In Infancy and early childhood (prior to 2 years of age), progressive enlargement of the head is the commonest manifestation of hydrocephalus as sutures have not united firmly. Occipitofrontal circumference should be measured after 24 hours of birth when moulding and overriding of sutures have disappeared. The head continues to enlarge and appears to be disproportionately larger than the rest of the body. The head is quite heavy and the child is not able to hold it without support. The child starts becoming less playful and does not feed properly. Milestones tend to get delayed. On examination, the child's head is found to be large in proportion to the body with bossing of frontal bones giving an inverted triangular appearance to the head. Serial head circumference should be taken to identify whether it is progressive (active) or arrested hydrocephalus. The fontanelle are widely open, bulging, tense and non pulsatile and the scalp veins are engorged, strikingly so when the infant cries. Sutures are widely separated. Cracked pot sound may be heard on percussion of the head due to separation of sutures and is called "Macewen's sign".Bruits over fontanels on auscultation of skull with the bell of the stethoscope indicates vascular origin of hydrocephalus (vein of Galen aneurysm or other vascular malformation).Transillumination of skull should be done in all cases. Normally, the halo of light around the rim of the illuminator extends upto 1 cm in the occipital region and upto 2 cm in frontal region in term babies. Excessive transillumination indicates abnormal collection of fluids as in hydranencephaly where the whole skull may glow with light or Dandy Walker syndrome where posterior part of skull transilluminates owing to the fluid accumulation in the posterior fossa.

The limbs may show increased tone and brisk reflexes “spastic paraparesis”. This results from stretching and distortion of paraventricular corticospinal tracts arising from leg area of motor cortex. These fibres have a longer distance to travel around the ventricles than those supplying the face and the upper limbs. However, mild spasticity and weakness of upper limbs is not uncommon particularly in advanced cases. Spine examination should be performed in all cases to look for presence of spina bifida (commonly associated with Chiari Malformation II).

Downturning of eyeballs with visibility of sclera above the iris is called ‘Sunsetting Sign’ and is a frequent finding. It is due to pressure on the superior quadrigeminal plate against the free edge of the tentorium causing a supranuclear paresis. It may be intermittent to begin with but later becomes continuous. Other ocular disturbances include unilateral or bilateral abducens nerve paresis, nystagmus, ptosis, strabismus and diminished papillary light responses. Optic atrophy can occur due to compression of the chiasm and optic nerves by a dilated anterior portion of the third ventricle. Papilledema is rare because rising tension is easily buffered by sutural diastasis.

Fig. 1. Congenital Hydrocephalus: Downturning of eyeballs with visibility of sclera above the iris (Sunsetting Sign).

In earlier stage, child may be quite playful, pick up the objects, put them into mouth, recognize the parents and follow light and objects. As hydrocephalus progresses, further destruction of cerebral cortex occurs, child tends to become listless, stops taking interest in the surroundings and regression of earlier achieved milestones occurs. There is unusual somnolence, persistent vomiting, failure to thrive and visual loss. Finally a decerebrate state ensues.

In early to late childhood (2 years and above), neurological symptoms caused by increased intracranial pressure or by focal deficits referable to the primary lesion is the predominant finding and appear before any significant change in head size. The clinical picture of various space occupying lesions depend on their site of origin. A unique but rare hydrocephalic syndrome “the bobble head doll syndrome” is related to obstructive lesions in or around

third ventricle and is characterized by bobbing of head forward and back involuntarily mimicking a bobble head doll. This can be inhibited voluntarily and disappears during sleep. A symptom very closely associated with head bobbing is the presence of ataxia. Several patients were reported to have difficulty in walking, running, and climbing steps because of the bobbing. One likely explanation for such symptom is interruption of the patient’s ability to balance which rely on input from various sources namely, the vestibular, ocular and somatosensory due to constant bobbing. The head bobbing is a neurological phenomenon and stems from dilated third ventricle impinging on the medial aspects of the dorsomedial nucleus of the thalamus.

1.	Hydrocephalus
2.	Hydranencephaly
3.	Subdural Effusion
4.	Thickened skull bones (achondroplasia, osteopetrosis, pyknodysostosis, craniometaphyseal dysplasia, orodigitofacial dysostosis , rickets, leontiasis ossea, etc.)
5.	Cerebral Gigantism
6.	Mucopolysaccharidosis
7.	Cerebral lipidosis (gangliosidosis)
8.	Metachromatic leukodystrophy
9.	Fragile X Syndrome
10.	Porencephaly
11.	Subdural Hematoma
12.	Intracranial Tumor
13.	Glutaryl-1-Coenzyme A dehydrogenase deficiency

Table 1. D/d of Enlarged Head in Newborn, Infancy and Children

Fig. 2. CT scan of the brain at the level of basal ganglia showing enlarged lateral ventricles suggestive of hydrocephalus.

The features of raised intracranial pressure are evident in almost all instances and includes frontal headache aggravated in the morning, improving with upright posture and associated with nausea and vomiting. The cracked pot sound is prominent on skull percussion. Fundoscopy may show papilledema. Additional features seen in late group are endocrine changes including small stature, gigantism, obesity, precocious or delayed puberty, menstrual irregularities, absent secondary sexual characteristics and central diabetes insipidus. They are probably caused by compression of hypothalamic pituitary axis by an enlarged third ventricle (a particular risk in aqueductal stenosis) resulting in abnormal hypothalamic pituitary function. Spastic diplegia is common. Thought and behavior may be affected adversely. Learning disabilities are fairly common and these children are credited with better verbal IQ than performance IQ. While the severity of hydrocephalus can differ considerably between individuals, some are of average or even above-average intelligence. Patients may develop motion and visual problems, coordination problems, or may be clumsy. Perceptual motor deficits and visual spatial disorganization follow as a result of stretched corticospinal fibres of parietal and occipital cortex due to dilated posterior horns of lateral ventricles. About one in four develops epilepsy.

2. Normal Pressure Hydrocephalus

Normal Pressure Hydrocephalus (NPH) is a clinical symptom complex, characterized by the classic triad of gait abnormality, dementia and urinary incontinence (commonly referred to as "wet, wobbly and wacky"). NPH occurs either as idiopathic or secondary condition, roughly in equal proportions. While NPH secondary to an identifiable cause can occur in all age groups, idiopathic NPH is most common in adults over 60 years of age without any sex predilection. These manifestations are believed to arise from dysfunction of periventricular white matter tracts, particularly those sub serving frontal lobe connections. Gait difficulty is often the first clinical manifestation. It is the effect of expansion of the ventricular system (particularly the lateral ventricles) and subsequent traction on the lumbosacral corticospinal fibers arising from the motor cortex. It is also believed to be the most responsive feature to shunting. It is classically described as "magnetic gait"; the patient's feet appear to stuck to the floor, steps are characteristically short with decreased stride length and height and a broad base. This may resemble parkinsonian gait at a glance but is distinguished by a narrow base and absence of tremor or rigidity. Postural stability is impaired, and a history of falls may be reported.

The cognitive disturbance of NPH is likely to be frontal in nature with psychomotor slowing, decreased attention and concentration, and apathy. The patient is slower in timed tasks, performs poorly on tests of divided attention and executive function, has difficulty with fluency tests, and has poor learning and better preserved recognition memory. The dementia is believed to be the consequence of stretching of frontal and limbic fibers that travel in the periventricular region. The Mini Mental State Examination may be an insensitive measure of cognitive impairment in NPH since it exhibits a frontal subcortical pattern rather than a cortical pattern and neuropsychological tests may prove to be a better tool in its characterization as well as diagnosis of coexisting dementia conditions (including Alzheimer's dementia and vascular dementia which are also highly likely in advancing age).

The third component, urinary incontinence, often begins as urgency and frequency rather than incontinence per se. However, overtime, true urinary incontinence ensues and is

accompanied by a lack of concern to urinary symptoms, reflecting its probable origin in the frontal lobe.

Fig. 3. Enlarged lateral ventricles with thinning out of cerebral cortex in a patient with normal pressure hydrocephalus.

Other features of NPH may include long tract signs with spasticity of lower limbs, hyperreflexia and extensor plantar responses (upper motor neuron signs). In very late stages, frontal release signs, akinetic mutism, and quadriparesis may occur.

3. References

- [1] Fishman, MA. Hydrocephalus. In: Neurological Pathophysiology, Eliasson, SG, Prensky, AL, Hardin, WB (Eds), Oxford, New York 1978.
- [2] Carey, CM, Tullous, MW, Walker, ML. Hydrocephalus: Etiology, Pathologic Effects, Diagnosis, and Natural History. In: Pediatric Neurosurgery, 3rd ed, Cheek, WR (Ed), WB Saunders Company, Philadelphia 1994.
- [3] Chumas P, Tyagi A, Livingston J. Hydrocephalus--what's new? Arch Dis Child Fetal Neonatal Ed 2001; 85:F149.
- [4] Blackburn BL, Fineman RM. Epidemiology of congenital hydrocephalus in Utah, 1940-1979: report of an iatrogenically related "epidemic". Am J Med Genet 1994; 52:123.

- [5] Fernell E, Hagberg G, Hagberg B. Infantile hydrocephalus epidemiology: an indicator of enhanced survival. *Arch Dis Child Fetal Neonatal Ed* 1994; 70:F123.
- [6] Bondurant CP, Jimenez DF. Epidemiology of cerebrospinal fluid shunting. *Pediatr Neurosurg* 1995; 23:254.
- [7] Yasuda T, Tomita T, McLone DG, Donovan M. Measurement of cerebrospinal fluid output through external ventricular drainage in one hundred infants and children: correlation with cerebrospinal fluid production. *Pediatr Neurosurg* 2002; 36:22.
- [8] Frawley GP, Dargaville PA, Mitchell PJ, et al. Clinical course and medical management of neonates with severe cardiac failure related to vein of Galen malformation. *Arch Dis Child Fetal Neonatal Ed* 2002; 87:F144.
- [9] Graf WD, Born DE, Sarnat HB. The pachygyria-polymicrogyria spectrum of cortical dysplasia in X-linked hydrocephalus. *Eur J Pediatr Surg* 1998; 8 Suppl 1:10.
- [10] Schrander-Stumpel C, Fryns JP. Congenital hydrocephalus: nosology and guidelines for clinical approach and genetic counselling. *Eur J Pediatr* 1998; 157:355.
- [11] Fransen E, Van Camp G, Vits L, Willems PJ. L1-associated diseases: clinical geneticists divide, molecular geneticists unite. *Hum Mol Genet* 1997; 6:1625.
- [12] Sasaki-Adams D, Elbabaa SK, Jewells V, et al. The Dandy-Walker variant: a case series of 24 pediatric patients and evaluation of associated anomalies, incidence of hydrocephalus, and developmental outcomes. *J Neurosurg Pediatr* 2008; 2:194.
- [13] Kirkpatrick M, Engleman H, Minns RA. Symptoms and signs of progressive hydrocephalus. *Arch Dis Child* 1989; 64:124.
- [14] Löppönen T, Saukkonen AL, Serlo W, et al. Accelerated pubertal development in patients with shunted hydrocephalus. *Arch Dis Child* 1996; 74:490.
- [15] Rekate, HL. Treatment of Hydrocephalus. In: *Pediatric Neurosurgery*, 3rd ed, Cheek, WR (Ed), WB Saunders Company, Philadelphia 1994.
- [16] Drake JM, Kestle JR, Milner R, et al. Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus. *Neurosurgery* 1998; 43:294.
- [17] Langley JM, LeBlanc JC, Drake J, Milner R. Efficacy of antimicrobial prophylaxis in placement of cerebrospinal fluid shunts: meta-analysis. *Clin Infect Dis* 1993; 17:98.
- [18] Casey AT, Kimmings EJ, Kleinlugtebeld AD, et al. The long-term outlook for hydrocephalus in childhood. A ten-year cohort study of 155 patients. *Pediatr Neurosurg* 1997; 27:63.
- [19] Forward KR, Fewer HD, Stiver HG. Cerebrospinal fluid shunt infections. A review of 35 infections in 32 patients. *J Neurosurg* 1983; 59:389.
- [20] Shapiro S, Boaz J, Kleiman M, et al. Origin of organisms infecting ventricular shunts. *Neurosurgery* 1988; 22:868.
- [21] Siomin V, Cinalli G, Grotenhuis A, et al. Endoscopic third ventriculostomy in patients with cerebrospinal fluid infection and/or hemorrhage. *J Neurosurg* 2002; 97:519.
- [22] Scarrow AM, Levy EI, Pascucci L, Albright AL. Outcome analysis of endoscopic III ventriculostomy. *Childs Nerv Syst* 2000; 16:442.
- [23] Javadpour M, Mallucci C, Brodbelt A, et al. The impact of endoscopic third ventriculostomy on the management of newly diagnosed hydrocephalus in infants. *Pediatr Neurosurg* 2001; 35:131.
- [24] Kulkarni AV, Drake JM, Mallucci CL, et al. Endoscopic third ventriculostomy in the treatment of childhood hydrocephalus. *J Pediatr* 2009; 155:254.
- [25] Sacko O, Boetto S, Lauwers-Cances V, et al. Endoscopic third ventriculostomy: outcome analysis in 368 procedures. *J Neurosurg Pediatr* 2010; 5:68.

- [26] Libenson MH, Kaye EM, Rosman NP, Gilmore HE. Acetazolamide and furosemide for posthemorrhagic hydrocephalus of the newborn. *Pediatr Neurol* 1999; 20:185.
- [27] Whitelaw A, Kennedy CR, Brion LP. Diuretic therapy for newborn infants with posthemorrhagic ventricular dilatation. *Cochrane Database Syst Rev* 2001; :CD002270.
- [28] International randomised controlled trial of acetazolamide and furosemide in posthaemorrhagic ventricular dilatation in infancy. International PHVD Drug Trial Group. *Lancet* 1998; 352:433.
- [29] Haines SJ, Lapointe M. Fibrinolytic agents in the management of posthemorrhagic hydrocephalus in preterm infants: the evidence. *Childs Nerv Syst* 1999; 15:226.
- [30] Whitelaw A. Repeated lumbar or ventricular punctures in newborns with intraventricular hemorrhage. *Cochrane Database Syst Rev* 2001; :CD000216.
- [31] Berger A, Weninger M, Reinprecht A, et al. Long-term experience with subcutaneously tunneled external ventricular drainage in preterm infants. *Childs Nerv Syst* 2000; 16:103.
- [32] Heep A, Engelskirchen R, Holschneider A, Groneck P. Primary intervention for posthemorrhagic hydrocephalus in very low birthweight infants by ventriculostomy. *Childs Nerv Syst* 2001; 17:47.
- [33] Hoppe-Hirsch E, Laroussinie F, Brunet L, et al. Late outcome of the surgical treatment of hydrocephalus. *Childs Nerv Syst* 1998; 14:97.
- [34] Klepper J, Büsse M, Strassburg HM, Sörensen N. Epilepsy in shunt-treated hydrocephalus. *Dev Med Child Neurol* 1998; 40:731.
- [35] Bourgeois M, Sainte-Rose C, Cinalli G, et al. Epilepsy in children with shunted hydrocephalus. *J Neurosurg* 1999; 90:274.
- [36] Caraballo RH, Bongiorno L, Cersósimo R, et al. Epileptic encephalopathy with continuous spikes and waves during sleep in children with shunted hydrocephalus: a study of nine cases. *Epilepsia* 2008; 49:1520.
- [37] Lindquist B, Carlsson G, Persson EK, Uvebrant P. Learning disabilities in a population-based group of children with hydrocephalus. *Acta Paediatr* 2005; 94:878.
- [38] Brookshire BL, Fletcher JM, Bohan TP, et al. Verbal and nonverbal skill discrepancies in children with hydrocephalus: a five-year longitudinal follow-up. *J Pediatr Psychol* 1995; 20:785.
- [39] Stoll C. Problems in the diagnosis of fragile X syndrome in young children are still present. *Am J Med Genet* 2001; 100:110.
- [40] Zvulunov A, Weitz R, Metzker A. Neurofibromatosis type 1 in childhood: evaluation of clinical and epidemiologic features as predictive factors for severity. *Clin Pediatr (Phila)* 1998; 37:295.
- [41] Furuta T, Tabuchi A, Adachi Y, et al. Primary brain tumors in children under age 3 years. *Brain Tumor Pathol* 1998; 15:7.
- [42] Tomita T, McLone DG. Brain tumors during the first twenty-four months of life. *Neurosurgery* 1985; 17:913.
- [43] Nard, JA. Abnormal head size and shape. In: Gartner, JC, Zitelli, BJ. *Common & Chronic Symptoms in Pediatrics*, Mosby, St. Louis, 1997.
- [44] Rios A. Microcephaly. *Pediatr Rev* 1996; 17:386.
- [45] Fenichel, GM. Disorders of cranial volume and shape. In: *Clinical Pediatric Neurology: A Signs and Symptoms Approach*, 5th ed. Elsevier Saunders, Philadelphia 2005. p. 353.

- [46] Nellhaus G. Head circumference from birth to eighteen years. Practical composite international and interracial graphs. *Pediatrics* 1968; 41:106.
- [47] Roche AF, Mukherjee D, Guo SM, Moore WM. Head circumference reference data: birth to 18 years. *Pediatrics* 1987; 79:706.
- [48] Rollins JD, Collins JS, Holden KR. United States head circumference growth reference charts: birth to 21 years. *J Pediatr* 2010; 156:907.
- [49] Grummer-Strawn LM, Reinold C, Krebs NF, Centers for Disease Control and Prevention (CDC). Use of World Health Organization and CDC growth charts for children aged 0-59 months in the United States. *MMWR Recomm Rep* 2010; 59:1.
- [50] Committee on Nutrition American Academy of Pediatrics. Failure to thrive. In: *Pediatric Nutrition Handbook*, 6th ed, Kleinman, RE (Ed), American Academy of Pediatrics, Elk Grove Village, IL 2009. p.601.
- [51] Bushby KM, Cole T, Matthews JN, Goodship JA. Centiles for adult head circumference. *Arch Dis Child* 1992; 67:1286.
- [52] Daymont C, Hwang WT, Feudtner C, Rubin D. Head-circumference distribution in a large primary care network differs from CDC and WHO curves. *Pediatrics* 2010; 126:e836.
- [53] Varma, R, Williams, SD, Wessel, HB. Neurology In: *Atlas of Pediatric Physical Diagnosis*, 5th ed. Zitelli, BJ, Davis, HW (Eds). Mosby Elsevier, Philadelphia 2007. p. 563.
- [54] Opitz JM, Holt MC. Microcephaly: general considerations and aids to nosology. *J Craniofac Genet Dev Biol* 1990; 10:175.
- [55] Williams CA, Dagli A, Battaglia A. Genetic disorders associated with macrocephaly. *Am J Med Genet A* 2008; 146A:2023.
- [56] Olney AH. Macrocephaly syndromes. *Semin Pediatr Neurol* 2007; 14:128.
- [57] DeMyer W. Megalencephaly: types, clinical syndromes, and management. *Pediatr Neurol* 1986; 2:321.
- [58] Day RE, Schutt WH. Normal children with large heads--benign familial megalencephaly. *Arch Dis Child* 1979; 54:512.
- [59] Lorber J, Priestley BL. Children with large heads: a practical approach to diagnosis in 557 children, with special reference to 109 children with megalencephaly. *Dev Med Child Neurol* 1981; 23:494.
- [60] Weaver DD, Christian JC. Familial variation of head size and adjustment for parental head circumference. *J Pediatr* 1980; 96:990.
- [61] Aoki N, Oikawa A, Sakai T. Serial neuroimaging studies in Sotos syndrome (cerebral gigantism syndrome). *Neurol Res* 1998; 20:149.
- [62] Gleeson, JG, Dobyns, WB, Plawner, L, Ashwal, S. Congenital structural defects. In: *Pediatric Neurology Principles and Practice*, 4th ed. Swaiman, KF, Ashwal, S, Ferriero, DM (Eds). Mosby Elsevier, Philadelphia 2006. p. 399.
- [63] Zahl SM, Wester K. Routine measurement of head circumference as a tool for detecting intracranial expansion in infants: what is the gain? A nationwide survey. *Pediatrics* 2008; 121:e416.
- [64] Alvarez LA, Maytal J, Shinnar S. Idiopathic external hydrocephalus: natural history and relationship to benign familial macrocephaly. *Pediatrics* 1986; 77:901.
- [65] Ment LR, Duncan CC, Geehr R. Benign enlargement of the subarachnoid spaces in the infant. *J Neurosurg* 1981; 54:504.
- [66] Kumar R. External hydrocephalus in small children. *Childs Nerv Syst* 2006; 22:1237.

- [67] Menkes, JH, Sarnat, HB, Flores-Sarnat, L. Malformations of the central nervous system. In: *Child Neurology*, 7th ed. Menkes, JH, Sarnat, HB, Maria, BL (Eds) Lippincott, Williams & Wilkins, Philadelphia, 2006. p.284.
- [68] Hellbusch LC. Benign extracerebral fluid collections in infancy: clinical presentation and long-term follow-up. *J Neurosurg* 2007; 107:119.
- [69] Andersson H, Elfverson J, Svendsen P. External hydrocephalus in infants. *Childs Brain* 1984; 11:398.
- [70] Gherpelli JL, Scaramuzzi V, Manreza ML, Diamant AJ. Follow-up study of macrocephalic children with enlargement of the subarachnoid space. *Arq Neuropsiquiatr* 1992; 50:156.
- [71] Pettit RE, Kilroy AW, Allen JH. Macrocephaly with head growth parallel to normal growth pattern: neurological, developmental, and computerized tomography findings in full-term infants. *Arch Neurol* 1980; 37:518.
- [72] Hamza M, Bodensteiner JB, Noorani PA, Barnes PD. Benign extracerebral fluid collections: a cause of macrocrania in infancy. *Pediatr Neurol* 1987; 3:218.
- [73] Nickel RE, Gallenstein JS. Developmental prognosis for infants with benign enlargement of the subarachnoid spaces. *Dev Med Child Neurol* 1987; 29:181.
- [74] Bosnjak V, Besenski N, Marusić-Della Marina B, Kogler A. Cranial ultrasonography in the evaluation of macrocrania in infancy. *Dev Med Child Neurol* 1989; 31:66.
- [75] Ravid S, Maytal J. External hydrocephalus: a probable cause for subdural hematoma in infancy. *Pediatr Neurol* 2003; 28:139.
- [76] Lago P, Rebsamen S, Clancy RR, et al. MRI, MRA, and neurodevelopmental outcome following neonatal ECMO. *Pediatr Neurol* 1995; 12:294.
- [77] Canady AI, Fessler RD, Klein MD. Ultrasound abnormalities in term infants on ECMO. *Pediatr Neurosurg* 1993; 19:202.
- [78] Lorch SA, D'Agostino JA, Zimmerman R, Bernbaum J. "Benign" extra-axial fluid in survivors of neonatal intensive care. *Arch Pediatr Adolesc Med* 2004; 158:178.
- [79] The Head. In: Green, MG (Ed), *Pediatric Diagnosis Interpretation of Symptoms and Signs in Children and Adolescents*, 6th ed, WB Saunders, Philadelphia, 1998. p.4.
- [80] Firth, HV, Hurst, JA, Hall, JG. Macrocephaly. In: *Oxford Desk Reference: Clinical Genetics*, 1st ed. Oxford University Press, Oxford 2005. p. 162.
- [81] Smith R, Leonidas JC, Maytal J. The value of head ultrasound in infants with macrocephaly. *Pediatr Radiol* 1998; 28:143.
- [82] Krauss JK, Halve B. Normal pressure hydrocephalus: survey on contemporary diagnostic algorithms and therapeutic decision-making in clinical practice. *Acta Neurochir (Wien)* 2004; 146:379.
- [83] Tisell M, Höglund M, Wikkelsø C. National and regional incidence of surgery for adult hydrocephalus in Sweden. *Acta Neurol Scand* 2005; 112:72.
- [84] Vanneste JA. Diagnosis and management of normal-pressure hydrocephalus. *J Neurol* 2000; 247:5.
- [85] Petersen RC, Mokri B, Laws ER Jr. Surgical treatment of idiopathic hydrocephalus in elderly patients. *Neurology* 1985; 35:307.
- [86] Black PM, Ojemann RG, Tzouras A. CSF shunts for dementia, incontinence, and gait disturbance. *Clin Neurosurg* 1985; 32:632.
- [87] Marmarou A, Young HF, Aygok GA, et al. Diagnosis and management of idiopathic normal-pressure hydrocephalus: a prospective study in 151 patients. *J Neurosurg* 2005; 102:987.

- [88] Hebb AO, Cusimano MD. Idiopathic normal pressure hydrocephalus: a systematic review of diagnosis and outcome. *Neurosurgery* 2001; 49:1166.
- [89] Bech RA, Juhler M, Waldemar G, et al. Frontal brain and leptomeningeal biopsy specimens correlated with cerebrospinal fluid outflow resistance and B-wave activity in patients suspected of normal-pressure hydrocephalus. *Neurosurgery* 1997; 40:497.
- [90] Bech RA, Waldemar G, Gjerris F, et al. Shunting effects in patients with idiopathic normal pressure hydrocephalus; correlation with cerebral and leptomeningeal biopsy findings. *Acta Neurochir (Wien)* 1999; 141:633.
- [91] Graff-Radford NR, Godersky JC. Symptomatic congenital hydrocephalus in the elderly simulating normal pressure hydrocephalus. *Neurology* 1989; 39:1596.
- [92] Krefft TA, Graff-Radford NR, Lucas JA, Mortimer JA. Normal pressure hydrocephalus and large head size. *Alzheimer Dis Assoc Disord* 2004; 18:35.
- [93] McComb JG, Bradley WG Jr, Safar FG, et al. Is a large hat size hazardous to your health? *AJNR Am J Neuroradiol* 2004; 25:1454.
- [94] Krauss JK, Regel JP, Vach W, et al. Vascular risk factors and arteriosclerotic disease in idiopathic normal-pressure hydrocephalus of the elderly. *Stroke* 1996; 27:24.
- [95] Graff-Radford NR, Godersky JC. Idiopathic normal pressure hydrocephalus and systemic hypertension. *Neurology* 1987; 37:868.
- [96] Ritter S, Dinh TT. Progressive postnatal dilation of brain ventricles in spontaneously hypertensive rats. *Brain Res* 1986; 370:327.
- [97] Bradley WG Jr, Whittemore AR, Watanabe AS, et al. Association of deep white matter infarction with chronic communicating hydrocephalus: implications regarding the possible origin of normal-pressure hydrocephalus. *AJNR Am J Neuroradiol* 1991; 12:31.
- [98] Krauss JK, Regel JP, Vach W, et al. White matter lesions in patients with idiopathic normal pressure hydrocephalus and in an age-matched control group: a comparative study. *Neurosurgery* 1997; 40:491.
- [99] Bradley WG. Normal pressure hydrocephalus: new concepts on etiology and diagnosis. *AJNR Am J Neuroradiol* 2000; 21:1586.
- [100] Kuriyama N, Tokuda T, Miyamoto J, et al. Retrograde jugular flow associated with idiopathic normal pressure hydrocephalus. *Ann Neurol* 2008; 64:217.
- [101] ADAMS RD, FISHER CM, HAKIM S, et al. SYMPTOMATIC OCCULT HYDROCEPHALUS WITH "NORMAL" CEREBROSPINAL-FLUID PRESSURE. A TREATABLE SYNDROME. *N Engl J Med* 1965; 273:117.
- [102] Lenfeldt N, Larsson A, Nyberg L, et al. Idiopathic normal pressure hydrocephalus: increased supplementary motor activity accounts for improvement after CSF drainage. *Brain* 2008; 131:2904.
- [103] Sudarsky L, Simon S. Gait disorder in late-life hydrocephalus. *Arch Neurol* 1987; 44:263.
- [104] Stolze H, Kuhtz-Buschbeck JP, Drücke H, et al. Comparative analysis of the gait disorder of normal pressure hydrocephalus and Parkinson's disease. *J Neurol Neurosurg Psychiatry* 2001; 70:289.
- [105] Iddon JL, Pickard JD, Cross JJ, et al. Specific patterns of cognitive impairment in patients with idiopathic normal pressure hydrocephalus and Alzheimer's disease: a pilot study. *J Neurol Neurosurg Psychiatry* 1999; 67:723.

- [106] Tullberg M, Hultin L, Ekholm S, et al. White matter changes in normal pressure hydrocephalus and Binswanger disease: specificity, predictive value and correlations to axonal degeneration and demyelination. *Acta Neurol Scand* 2002; 105:417.
- [107] Sudarsky L, Ronthal M. Gait disorders among elderly patients. A survey study of 50 patients. *Arch Neurol* 1983; 40:740.
- [108] Temml C, Haidinger G, Schmidbauer J, et al. Urinary incontinence in both sexes: prevalence rates and impact on quality of life and sexual life. *Neurourol Urodyn* 2000; 19:259.
- [109] Savolainen S, Paljärvi L, Vapalahti M. Prevalence of Alzheimer's disease in patients investigated for presumed normal pressure hydrocephalus: a clinical and neuropathological study. *Acta Neurochir (Wien)* 1999; 141:849.
- [110] Golomb J, Wisoff J, Miller DC, et al. Alzheimer's disease comorbidity in normal pressure hydrocephalus: prevalence and shunt response. *J Neurol Neurosurg Psychiatry* 2000; 68:778.
- [111] Savolainen S, Hurskainen H, Paljärvi L, et al. Five-year outcome of normal pressure hydrocephalus with or without a shunt: predictive value of the clinical signs, neuropsychological evaluation and infusion test. *Acta Neurochir (Wien)* 2002; 144:515.
- [112] Hamilton R, Patel S, Lee EB, et al. Lack of shunt response in suspected idiopathic normal pressure hydrocephalus with Alzheimer disease pathology. *Ann Neurol* 2010; 68:535.
- [113] Malm, J, Eklund, A. Idiopathic normal pressure hydrocephalus. *Practical Neurology* 2006; 6:14.
- [114] Barron SA, Jacobs L, Kinkel WR. Changes in size of normal lateral ventricles during aging determined by computerized tomography. *Neurology* 1976; 26:1011.
- [115] Gyldensted C. Measurements of the normal ventricular system and hemispheric sulci of 100 adults with computed tomography. *Neuroradiology* 1977; 14:183.
- [116] Black PM. Idiopathic normal-pressure hydrocephalus. Results of shunting in 62 patients. *J Neurosurg* 1980; 52:371.
- [117] Thomsen AM, Børgesen SE, Bruhn P, Gjerris F. Prognosis of dementia in normal-pressure hydrocephalus after a shunt operation. *Ann Neurol* 1986; 20:304.
- [118] Børgesen SE, Gjerris F. The predictive value of conductance to outflow of CSF in normal pressure hydrocephalus. *Brain* 1982; 105:65.
- [119] Palm WM, Saczynski JS, van der Grond J, et al. Ventricular dilation: association with gait and cognition. *Ann Neurol* 2009; 66:485.
- [120] Krauss JK, Droste DW, Vach W, et al. Cerebrospinal fluid shunting in idiopathic normal-pressure hydrocephalus of the elderly: effect of periventricular and deep white matter lesions. *Neurosurgery* 1996; 39:292.
- [121] Boon AJ, Tans JT, Delwel EJ, et al. Dutch Normal-Pressure Hydrocephalus Study: the role of cerebrovascular disease. *J Neurosurg* 1999; 90:221.
- [122] Tullberg M, Jensen C, Ekholm S, Wikkelsø C. Normal pressure hydrocephalus: vascular white matter changes on MR images must not exclude patients from shunt surgery. *AJNR Am J Neuroradiol* 2001; 22:1665.
- [123] Tullberg M, Ziegelitz D, Ribbelin S, Ekholm S. White matter diffusion is higher in Binswanger disease than in idiopathic normal pressure hydrocephalus. *Acta Neurol Scand* 2009; 120:226.

- [124] Bradley WG Jr, Whittemore AR, Kortman KE, et al. Marked cerebrospinal fluid void: indicator of successful shunt in patients with suspected normal-pressure hydrocephalus. *Radiology* 1991; 178:459.
- [125] Krauss JK, Regel JP, Vach W, et al. Flow void of cerebrospinal fluid in idiopathic normal pressure hydrocephalus of the elderly: can it predict outcome after shunting? *Neurosurgery* 1997; 40:67.
- [126] Hakim R, Black PM. Correlation between lumbo-ventricular perfusion and MRI-CSF flow studies in idiopathic normal pressure hydrocephalus. *Surg Neurol* 1998; 49:14.
- [127] Jack CR Jr, Petersen RC, O'Brien PC, Tangalos EG. MR-based hippocampal volumetry in the diagnosis of Alzheimer's disease. *Neurology* 1992; 42:183.
- [128] Holodny AI, Waxman R, George AE, et al. MR differential diagnosis of normal-pressure hydrocephalus and Alzheimer disease: significance of perihippocampal fissures. *AJNR Am J Neuroradiol* 1998; 19:813.
- [129] Savolainen S, Laakso MP, Paljärvi L, et al. MR imaging of the hippocampus in normal pressure hydrocephalus: correlations with cortical Alzheimer's disease confirmed by pathologic analysis. *AJNR Am J Neuroradiol* 2000; 21:409.
- [130] Miyoshi N, Kazui H, Ogino A, et al. Association between cognitive impairment and gait disturbance in patients with idiopathic normal pressure hydrocephalus. *Dement Geriatr Cogn Disord* 2005; 20:71.
- [131] Graff-Radford NR, Godersky JC, Jones MP. Variables predicting surgical outcome in symptomatic hydrocephalus in the elderly. *Neurology* 1989; 39:1601.
- [132] De Mol J. [Prognostic factors for therapeutic outcome in normal-pressure hydrocephalus. Review of the literature and personal study]. *Acta Neurol Belg* 1985; 85:13.
- [133] Stolze H, Kuhtz-Buschbeck JP, Drücke H, et al. Gait analysis in idiopathic normal pressure hydrocephalus--which parameters respond to the CSF tap test? *Clin Neurophysiol* 2000; 111:1678.
- [134] Wikkelso C, Andersson H, Blomstrand C, et al. Normal pressure hydrocephalus. Predictive value of the cerebrospinal fluid tap-test. *Acta Neurol Scand* 1986; 73:566.
- [135] Walchenbach R, Geiger E, Thomeer RT, Vanneste JA. The value of temporary external lumbar CSF drainage in predicting the outcome of shunting on normal pressure hydrocephalus. *J Neurol Neurosurg Psychiatry* 2002; 72:503.
- [136] Kahlon B, Sundbärg G, Rehncrona S. Comparison between the lumbar infusion and CSF tap tests to predict outcome after shunt surgery in suspected normal pressure hydrocephalus. *J Neurol Neurosurg Psychiatry* 2002; 73:721.
- [137] Malm J, Kristensen B, Karlsson T, et al. The predictive value of cerebrospinal fluid dynamic tests in patients with th idiopathic adult hydrocephalus syndrome. *Arch Neurol* 1995; 52:783.
- [138] Chen IH, Huang CI, Liu HC, Chen KK. Effectiveness of shunting in patients with normal pressure hydrocephalus predicted by temporary, controlled-resistance, continuous lumbar drainage: a pilot study. *J Neurol Neurosurg Psychiatry* 1994; 57:1430.
- [139] Haan J, Thomeer RT. Predictive value of temporary external lumbar drainage in normal pressure hydrocephalus. *Neurosurgery* 1988; 22:388.
- [140] Duinkerke A, Williams MA, Rigamonti D, Hillis AE. Cognitive recovery in idiopathic normal pressure hydrocephalus after shunt. *Cogn Behav Neurol* 2004; 17:179.

- [141] Krauss JK, Regel JP. The predictive value of ventricular CSF removal in normal pressure hydrocephalus. *Neurol Res* 1997; 19:357.
- [142] Symon L, Dorsch NW. Use of long-term intracranial pressure measurement to assess hydrocephalic patients prior to shunt surgery. *J Neurosurg* 1975; 42:258.
- [143] Crockard HA, Hanlon K, Duda EE, Mullan JF. Hydrocephalus as a cause of dementia: evaluation by computerised tomography and intracranial pressure monitoring. *J Neurol Neurosurg Psychiatry* 1977; 40:736.
- [144] Stephensen H, Andersson N, Eklund A, et al. Objective B wave analysis in 55 patients with non-communicating and communicating hydrocephalus. *J Neurol Neurosurg Psychiatry* 2005; 76:965.
- [145] Krauss JK, Droste DW, Bohus M, et al. The relation of intracranial pressure B-waves to different sleep stages in patients with suspected normal pressure hydrocephalus. *Acta Neurochir (Wien)* 1995; 136:195.
- [146] Katzman R, Hussey F. A simple constant-infusion manometric test for measurement of CSF absorption. I. Rationale and method. *Neurology* 1970; 20:534.
- [147] Boon AJ, Tans JT, Delwel EJ, et al. Dutch normal-pressure hydrocephalus study: prediction of outcome after shunting by resistance to outflow of cerebrospinal fluid. *J Neurosurg* 1997; 87:687.
- [148] Meier U, Bartels P. The importance of the intrathecal infusion test in the diagnostic of normal-pressure hydrocephalus. *Eur Neurol* 2001; 46:178.
- [149] Malm J, Lundkvist B, Eklund A, et al. CSF outflow resistance as predictor of shunt function. A long-term study. *Acta Neurol Scand* 2004; 110:154.
- [150] Vanneste J, Augustijn P, Dirven C, et al. Shunting normal-pressure hydrocephalus: do the benefits outweigh the risks? A multicenter study and literature review. *Neurology* 1992; 42:54.
- [151] Lins H, Wichart I, Bancher C, et al. Immunoreactivities of amyloid beta peptide((1-42)) and total tau protein in lumbar cerebrospinal fluid of patients with normal pressure hydrocephalus. *J Neural Transm* 2004; 111:273.
- [152] Kudo T, Mima T, Hashimoto R, et al. Tau protein is a potential biological marker for normal pressure hydrocephalus. *Psychiatry Clin Neurosci* 2000; 54:199.
- [153] Tullberg M, Rosengren L, Blomsterwall E, et al. CSF neurofilament and glial fibrillary acidic protein in normal pressure hydrocephalus. *Neurology* 1998; 50:1122.
- [154] Tullberg M, Månsson JE, Fredman P, et al. CSF sulfatide distinguishes between normal pressure hydrocephalus and subcortical arteriosclerotic encephalopathy. *J Neurol Neurosurg Psychiatry* 2000; 69:74.
- [155] Dixon GR, Friedman JA, Luetmer PH, et al. Use of cerebrospinal fluid flow rates measured by phase-contrast MR to predict outcome of ventriculoperitoneal shunting for idiopathic normal-pressure hydrocephalus. *Mayo Clin Proc* 2002; 77:509.
- [156] Egeler-Peerdeman SM, Barkhof F, Walchenbach R, Valk J. Cine phase-contrast MR imaging in normal pressure hydrocephalus patients: relation to surgical outcome. *Acta Neurochir Suppl* 1998; 71:340.
- [157] del Mar Matarín M, Pueyo R, Poca MA, et al. Post-surgical changes in brain metabolism detected by magnetic resonance spectroscopy in normal pressure hydrocephalus: results of a pilot study. *J Neurol Neurosurg Psychiatry* 2007; 78:760.

- [158] Kristensen B, Malm J, Fagerland M, et al. Regional cerebral blood flow, white matter abnormalities, and cerebrospinal fluid hydrodynamics in patients with idiopathic adult hydrocephalus syndrome. *J Neurol Neurosurg Psychiatry* 1996; 60:282.
- [159] Owler BK, Pickard JD. Normal pressure hydrocephalus and cerebral blood flow: a review. *Acta Neurol Scand* 2001; 104:325.
- [160] Tedeschi E, Hasselbalch SG, Waldemar G, et al. Heterogeneous cerebral glucose metabolism in normal pressure hydrocephalus. *J Neurol Neurosurg Psychiatry* 1995; 59:608.
- [161] Pujari S, Kharkar S, Metellus P, et al. Normal pressure hydrocephalus: long-term outcome after shunt surgery. *J Neurol Neurosurg Psychiatry* 2008; 79:1282.
- [162] Sand T, Bovim G, Grimse R, et al. Idiopathic normal pressure hydrocephalus: the CSF tap-test may predict the clinical response to shunting. *Acta Neurol Scand* 1994; 89:311.
- [163] Gustafson L, Hagberg B. Recovery in hydrocephalic dementia after shunt operation. *J Neurol Neurosurg Psychiatry* 1978; 41:940.
- [164] Raftopoulos C, Deleval J, Chaskis C, et al. Cognitive recovery in idiopathic normal pressure hydrocephalus: a prospective study. *Neurosurgery* 1994; 35:397.
- [165] Fisher CM. The clinical picture in occult hydrocephalus. *Clin Neurosurg* 1977; 24:270.
- [166] Hughes CP, Siegel BA, Coxe WS, et al. Adult idiopathic communicating hydrocephalus with and without shunting. *J Neurol Neurosurg Psychiatry* 1978; 41:961.
- [167] Czosnyka Z, Czosnyka M, Richards HK, Pickard JD. Laboratory testing of hydrocephalus shunts -- conclusion of the U.K. Shunt evaluation programme. *Acta Neurochir (Wien)* 2002; 144:525.
- [168] Ringel F, Schramm J, Meyer B. Comparison of programmable shunt valves vs standard valves for communicating hydrocephalus of adults: a retrospective analysis of 407 patients. *Surg Neurol* 2005; 63:36.
- [169] Boon AJ, Tans JT, Delwel EJ, et al. Dutch Normal-Pressure Hydrocephalus Study: randomized comparison of low- and medium-pressure shunts. *J Neurosurg* 1998; 88:490.
- [170] Weiner HL, Constantini S, Cohen H, Wisoff JH. Current treatment of normal-pressure hydrocephalus: comparison of flow-regulated and differential-pressure shunt valves. *Neurosurgery* 1995; 37:877.
- [171] Raftopoulos C, Massager N, Balériaux D, et al. Prospective analysis by computed tomography and long-term outcome of 23 adult patients with chronic idiopathic hydrocephalus. *Neurosurgery* 1996; 38:51.
- [172] Malm J, Kristensen B, Stegmayr B, et al. Three-year survival and functional outcome of patients with idiopathic adult hydrocephalus syndrome. *Neurology* 2000; 55:576.
- [173] Lundkvist B, Eklund A, Kristensen B, et al. Cerebrospinal fluid hydrodynamics after placement of a shunt with an antisiphon device: a long-term study. *J Neurosurg* 2001; 94:750.

Hydrocephalus

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Description of hydrocephalus can be found in ancient medical literature from Egypt as old as 500 AD. Hydrocephalus is characterized by abnormal accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain. This results in the rise of intracranial pressure inside the skull causing progressive increase in the size of the head, seizure, tunneling of vision, and mental disability. The clinical presentation of hydrocephalus varies with age of onset and chronicity of the underlying disease process. Acute dilatation of the ventricular system manifests with features of raised intracranial pressure while chronic dilatation has a more insidious onset presenting as Adams triad. Treatment is generally surgical by creating various types of cerebral shunts. Role of endoscopic has emerged lately in the management of hydrocephalus.

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