3.1 Basics

3.1.1 Anatomy

CHRISTIANTO B. LUMENTA

3.1.1.1 The Skull and Its Solid Contents

The central nervous system has a hard and a soft cover: the skull and the meninges (Figs. 3.1.1–3.1.4).

The skull is divided into the neurocranium and the viscerocranium. The neurocranium encloses the brain, and is formed by the frontal, parietal, temporal, occipital sphenoid and ethmoid bones. The frontal, both parietal, a small part of the temporal and a large part of the occipital bone make up the skullcap, while the orbital roof, the sphenoid, the petrosal, and the suboccipital bones constitute the skull base, which is divided into anterior, middle and posterior fossa. The cranial bones are connected by cranial sutures: the sutura coronalis, the sagittalis and the lambdoidea. In childhood the sutures have not been ossified; therefore, on X-ray films the sutures seem to be gaping. We call the gap the fontanel, which is closed by the end of the second year of life.

The cranial bone is covered by layer of skin called the pericranium.

The brain is covered by the soft meninges, which consists of three layers: the dura mater, the arachnoidea and the pia mater.

The whole nervous system is divided into the central nervous system, which includes the brain and the spinal cord, the peripheral nerves and the vegetative nervous system.

The brain consists of three parts: the cerebrum, the cerebellum and the brain-stem. All body functions are controlled by the cerebrum, while the brain-stem plays the role of an instinctive control system. The cerebellum coordinates all movement impulses coming from the cerebrum, before they reach the muscle via the spinal cord and peripheral nerves.

The cerebrum is in humans the largest part of the brain. It has four lobes: the frontal, parietal, temporal and occipital lobes. The cerebral cortex is structured out of the sulci and gyri. The sulcus centralis differentiates the frontal lobe from the parietal one, and the Sylvian fissure (sulcus lateralis) is the border between the frontal and parietal lobes and the temporal one. The sulcus parieto-occipitalis marks off the parietal lobe from the occipital one. There is, however, no border between the temporal and occipital lobes. If we perform a section through the brain, we recognise the grey and the white matter, in which grey-coloured areas are also localised (brain nuclei). The most important area for the motoric function is the gyrus precentralis. The nerve cells localised to this gyrus nerve cells are the origins of the pyramidal tract, the most important tract of the arbitrary motoric system. Eighty-five percent of the pyramidal tract crosses to the contralateral side at a small mound at the medulla oblongata. In fact, the pyramidal tract goes straight to the anterior horn cells of the spinal cord, but all motoric functions are controlled by connections between the precentral gyrus and other centres of the brain, especially the cerebellum.

The connection tracts between each brain zone are called association and commissural tracts. The most important commissural tracts, which connect the two hemispheres to each other, are localised to the corpus callosum, which can be recognised as white structure in the median section of the brain. While the precentral gyrus represents the motoric control of the body, the postcentral gyrus is responsible for sensoric control, with defined functional areas for different body parts. The two hemispheres are functionally dissimilar; in the normal case of a right-handed person the left hemisphere is the dominant one. The motoric speech centre of Broca – its disturbance causes so-called motoric aphasia – is localised to the lateral part of the frontal lobe, while the sensoric speech centre of Wernicke is situated in the temporal lobe below the Sylvian fissure. A disturbance of the Wernicke speech centre leads to so-called sensoric aphasia, i.e. to be unable to understand.

The cerebellum is localised to the posterior fossa, it has two hemispheres and a middle part (vermis; Fig. 3.1.5). The connection tracts between the cerebrum and the cerebellum pass the cerebellar peduncles. The main functions of the cerebellum are to coordinate and to control all movements of the body.

The following brain areas belong to the brain-stem:

- The diencephalon: consisting of the thalamus, hypothalamus and corpus pineale

- The mesencephalon, including the lamina quadrigemina
- The tegmentum and crura cerebri
- The pons and medulla oblongata

The unarbitrary control of all life functions is performed in the reflex centres of the brain-stem, which consists of the main parts of the tracts to and from the cerebrum.

The pituitary gland is localised in the middle fossa and divided into:

- The adenohypophysis, which produces:
  - Growth hormone (somatotropin, STH, GH)
  - –Adrenocorticotropic hormone (ACTH)
  - Follicle-stimulating hormone (FSH)
  - Luteinising hormone (LH)
  - Interstitial cells-stimulating hormone (ICSH)
  - Thyrotropin hormone (TSH)
  - Prolactin
- The neurohypophysis, which transfers the hormones that are produced in the hypothalamus into the blood circulation:
  - Anti-diuretic hormone (ADH)
  - Oxytocin

The centre of cardiac and blood circulation and the breath function is localised in the medulla oblongata.

3.1.1.2 The Cerebral Blood Vessels

The arterial intracranial blood vessel system (Fig. 3.1.6) consists of:

- The two internal carotid arteries (ICA) with their branches: the ophthalmic artery (OA), the posterior communicating artery (PCommA), the anterior choroidal artery (AChoA), the anterior cerebral artery (ACA) and the middle cerebral artery (MCA)
- The two vertebral arteries (VA) linked to the basilar artery (BA) with its branches: the posterior cerebral artery (PCA), the superior cerebellar artery (SCA), the anterior inferior cerebellar artery (AICA) and the posterior inferior cerebellar artery (PICA)

The two internal carotid arteries are connected by the anterior communicating artery (ACommA), and the basilar artery is connected to the carotid arteries by the posterior communicating arteries. We call this structure at the skull-base the circulus arteriosus or the circle of Willis. The ACA, MCA and PCA come from this circle.

The large venous system of the brain is called the venous sinus, which is a duplication of the dura: the superior sagittal sinus (SSS) and the rectus sinus (RS) → the confluens sinuum (CS) → the transverse sinus (TS) → the sigmoid sinus (SS) → the jugular vein (JV).

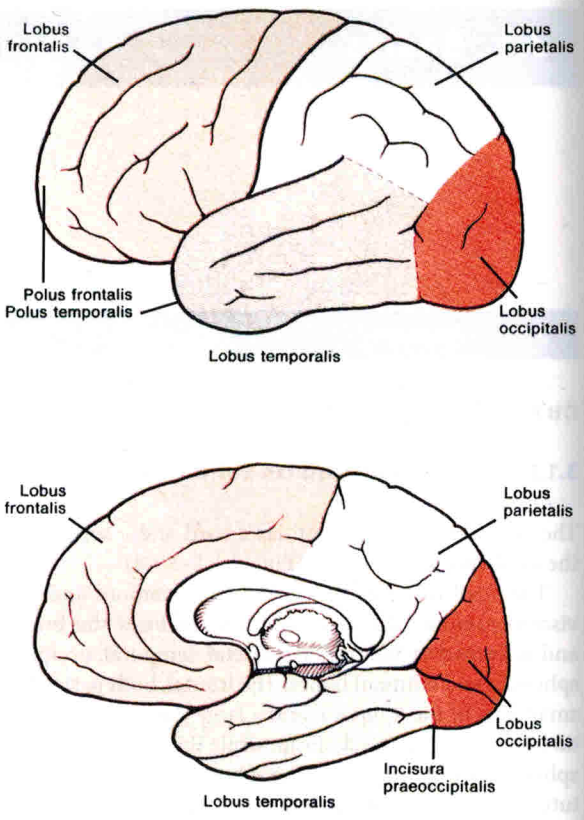


Fig. 3.1.1 The lateral *(top)* and the medial *(bottom)* views of the left hemisphere with each brain region

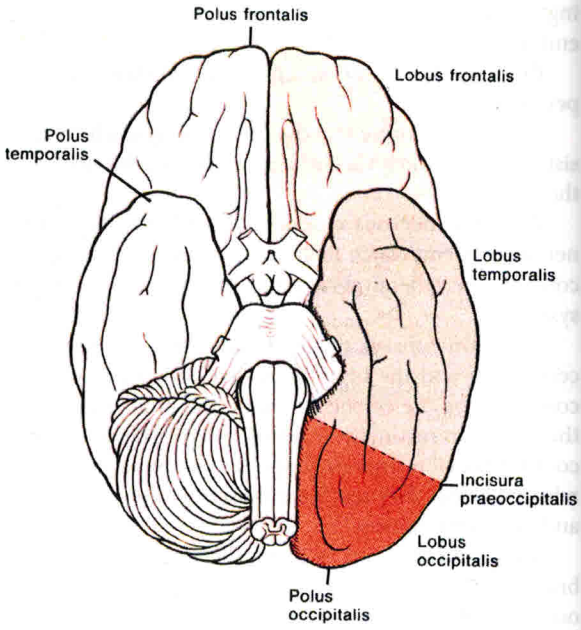


Fig. 3.1.2 The basal view of the brain without the cerebellum

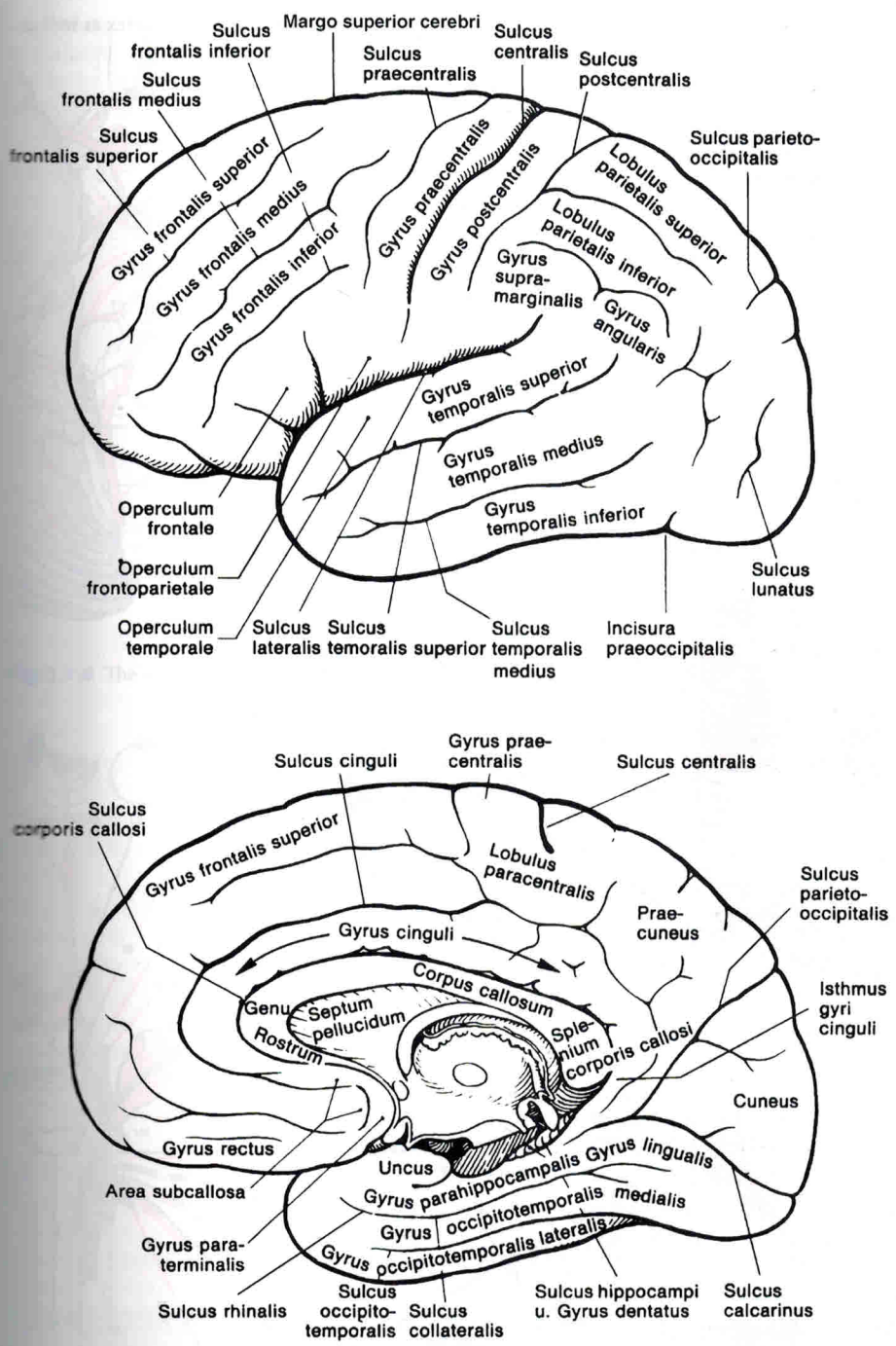


Fig. 3.1.3 The brain gyri *(top)* and sulci *(bottom)* in lateral and medial view

3.1.1.3 The Cerebrospinal Fluid and Ventricle System

Cerebrospinal fluid (CSF) is produced in the ventricles by the plexus choroideus, and washes around the central nervous system within the subarachnoid space.

The CSF circulates in the ventricle system of the brain:

- From both lateral ventricles via the foramen of Monro to
- The third ventricle via the aqueduct to

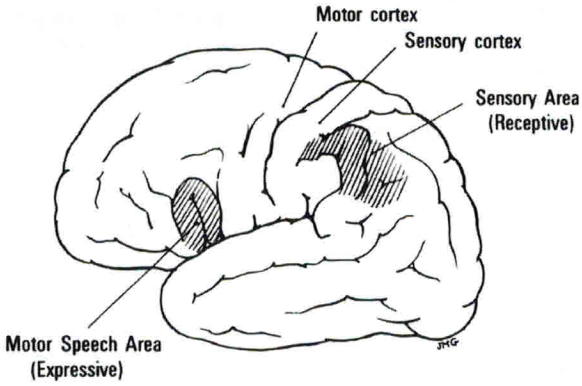
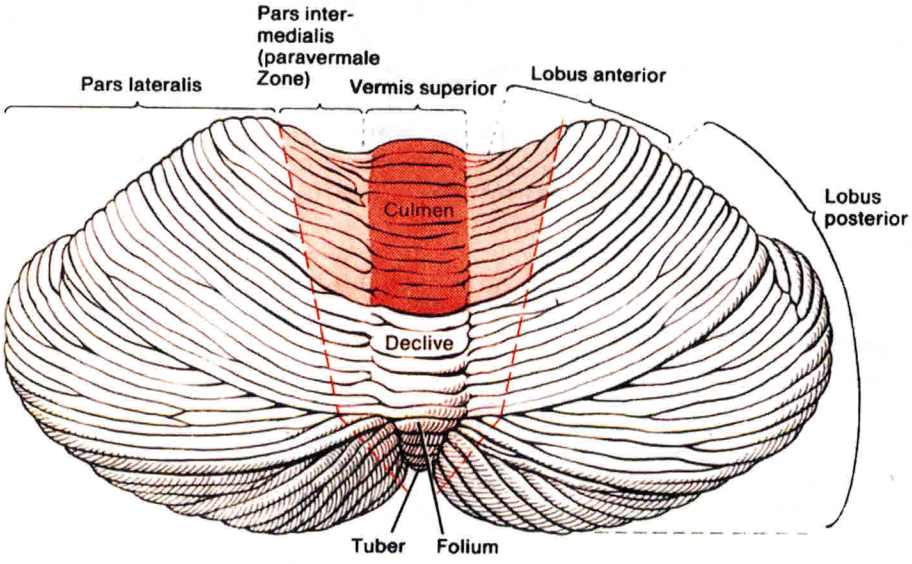


Fig. 3.1.4 Localisation of motor and sensory cortex as well as of motor speech and sensory area



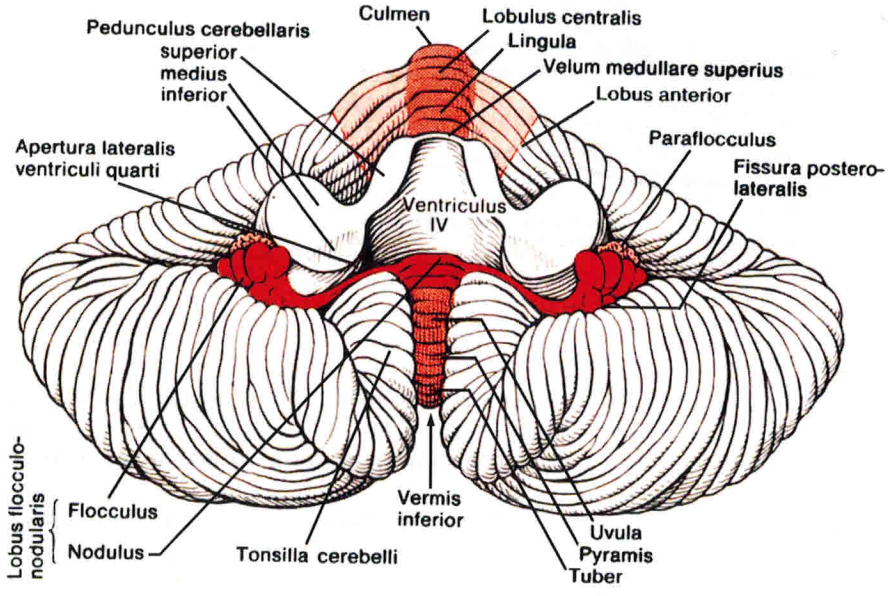


Fig. 3.1.5 The cerebellum: view from the top and from below

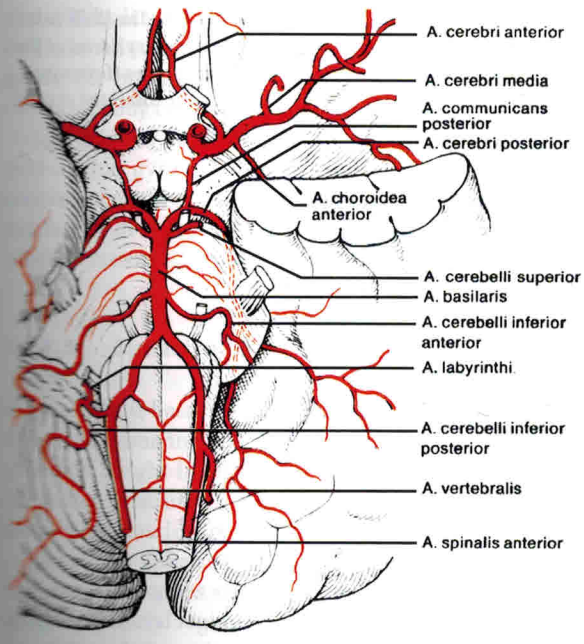


Fig. 3.1.6 The arteries at the base of the brain

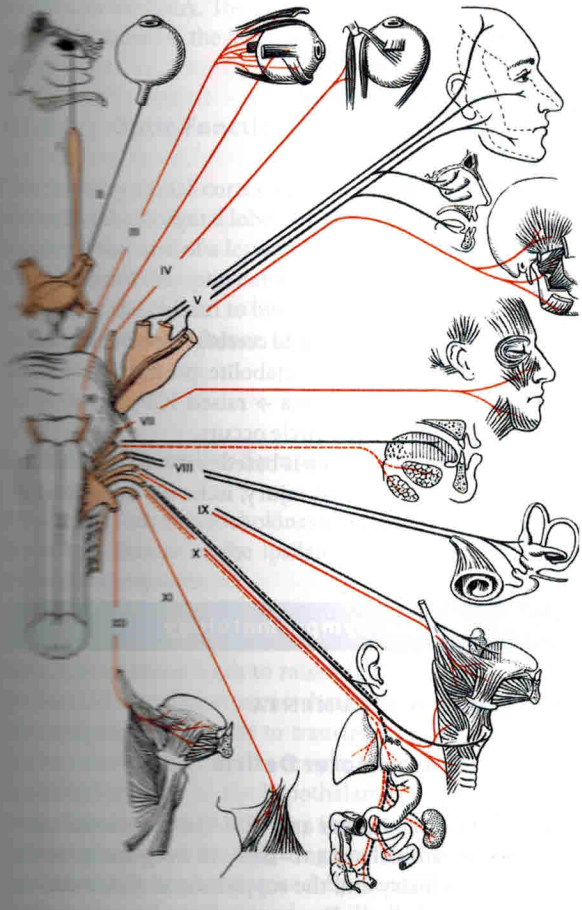


Fig. 3.1.7 The cranial nerves

- The fourth ventricle via the foramen of Magendie (medially) and the foramina of Luschka (laterally) to
- The spinal canal

Resorption of the CSF takes place in the arachnoid, in Pachioni's granulations, and in the root sheath of the spinal nerves.

The amount of CSF in adults is around 120–180 ml.

3.1.1.4 The Cranial Nerves

All 12 cranial nerves (Figs. 3.1.7, 3.1.8) come directly from the brain:

- CN 1: the olfactory nerve → the smell function
- CN 2: the optic nerve → vision
- CN 3: the oculomotor nerve → movement of the eye ball and pupil reaction
- CN 4: the trochlear nerve → rotation of the eye ball laterally and downwards
- CN 5: the trigeminal nerve → sensory function of the face and the mastication function
- CN 6: abducent nerve → rotation of the eye ball laterally
- CN 7: facial nerve → the mimic function
- CN 8: vestibulocochlear nerve → the balance and hearing functions
- CN 9: glossopharyngeal nerve → taste and the swallowing function
- CN 10: vagal nerve → sensory and motoric function of the tongue and mouth floor, as well as the parasympathetic function
- CN 11: accessory nerve → head flexion and shoulder elevation
- CN 12: hypoglossal nerve → tongue movement

3.1.2 Pathophysiology

CHRISTIANTO B. LUMENTA

Although the weight of the brain is around just 2% of that of the whole body, it needs around 20% of the cardiac minute volume for its blood flow. The cerebral blood flow (CBF) remains constant, as long as the systolic blood pressure is not lower than 60 mmHg and not higher than 150 mmHg, the so-called autoregulation of the brain. It controls the width of the blood vessels in the brain depending on the pH, pO_2 and pCO_2 values of the blood, and thus the CBF. A shock occurs if the systolic blood pressure falls below 60 mmHg. This situation leads to a disturbance of the brain's autoregulation. The CBF and the brain blood volume decrease. The result is unconsciousness or irreversible brain function damage, if it persists.

3.6 Developmental and Acquired Anomalies

3.6.1 Hydrocephalus in Adults

AXEL TROST, CHRISTIANTO B. LUMENTA

3.6.1.1 Basics

Hydrocephalus is a syndrome that is not yet fully understood. For almost a century Dandy's observations in 1919 paved the way to hydrocephalus being referred to as obstructive or communicating. Our knowledge of hydrocephalus has developed stepwise, based on fundamental observations in diagnostic procedures such as pneumoencephalography, cisternography, computed tomography (CT) and magnetic resonance imaging (MRI).

While childhood hydrocephalus acts on a developing brain with soft and growing borders, in adults space within the skull is fixed, and the relationship among brain tissue, cerebrospinal fluid (CSF), and blood changes physiologically as a reaction to changing brain volume caused by ischemic or traumatic damage, toxic or degenerative changes. The adult brain may be more vulnerable due to comorbidities such as hypertonic small vessel disease, small-infarction syndrome, white matter lesions, and gliotic fibrosis. Many degenerative states and diseases of the brain can result in hydrocephalus. Thus, adult hydrocephalus may not be a disease per se, but a multi-etiological clinical entity as a result of chronic changes in an aging or damaged brain.

Hydrocephalus is one of the few almost completely reversible causes of dementia and gait disturbance in the elderly. Adult hydrocephalus markedly impairs mobility in affected persons. Falls and other problems associated with gait disorders, and the enforced immobility that ensues, are a major source of preventable morbidity and mortality.

Adult hydrocephalus is usually characterized by enlarged ventricles, compared with normal or compressed extracerebral subarachnoid spaces. However, normal ventricular size does not guarantee that hydrocephalus will not occur. CSF pressure in adult hydrocephalus may be elevated, but can fluctuate within normal ranges or even low. Ventricular enlargement in the presence of "normal" CSF pressure seems to be related to periodic wave-like changes in CSF pressure.

In occlusive or obstructive hydrocephalus CSF flow is impaired, thus producing an increase in ventricular pressure and ventricular size. In communicating hydrocephalus the relationship between CSF production and CSF resorption is impaired. While CSF production is almost constant, CSF resorption seems to be subject to change, owing to many states, resulting in elevated resistance to CSF outflow.

Normal pressure hydrocephalus (NPH) is usually referred to as being first described by Hakim and Adams in 1965. NPH can be idiopathic (iNPH), typically beginning in the sixth decade of life or secondary (sNPH), which may be a result of subarachnoid hemorrhage, brain injury, meningitis, posterior fossa surgery or tumors, including carcinomatous meningitis at any age.

Synonyms are Hakim–Adams syndrome, low-pressure hydrocephalus, malresorptive hydrocephalus, aresorptive hydrocephalus, and adult-onset hydrocephalus.

3.6.1.2 Diagnostic Procedures

Proper diagnosis is both fundamental and difficult, for treatment is not without risk. Hydrocephalus due to pure atrophy (hydrocephalus ex vacuo) will not respond to a shunt. After the intervention of shunting procedures in NPH patients, many patients with dementia of other origin were shunted, resulting in severe complications. Therefore, the aim of diagnostic studies is to find the optimal surgical procedures.

There is no commonly accepted standard for diagnosis at the moment. Usually, NPH is diagnosed by a combination of clinical signs and radiological findings.

3.6.1.3 Clinical Signs

The most important clinical signs are the so-called Adams–Hakim Triad: gait disturbance (shuffling), which usually precedes the other symptoms; cognitive deficit; and urinary incontinence. As these are common symptoms of many age- and degeneration-related diseases of the brain, it is often not possible to differentiate hydrocephalus from senility, Alzheimer's disease, and other

forms of dementia. The combination of NPH and other diseases is common. Co-morbidity may complicate diagnosis, but it is not a negative prognostic factor per se.

Gait disturbance is the most important sign and should preferably be the first one to appear. Gait disturbance is referred to as the stretching of the fibers of the pyramidal tract caused by ventricular enlargement. Reduced gait velocity, owing to a diminished and highly variable stride length, may be found in both Parkinson's disease and NPH. It may be distinguished by a broad-based gait pattern with outward rotated feed and diminished height of the steps, which is typical of NPH. External cues will only mildly improve gait in NPH as opposed to good effects being obtained in Parkinson's disease. Unsteadiness and multiple steps on turning is another typical pattern in NPH.

Differential diagnosis in cognitive deficits and urinary incontinence may be even more difficult. Many patients with NPH are at an age at which they suffer from diseases that may result in the same symptoms.

Dementia in NPH usually starts with memory impairment, followed by impaired wakefulness, bradyphrenia (mental slowness), and bradykinesia.

Urinary incontinence takes place often unwittingly and may be exacerbated by gait disturbance, preventing the patient from reaching a toilet in time. Fecal incontinence is rare.

Headache, dizziness, vertigo, and psychiatric disturbances may also be symptoms of NPH.

3.6.1.4 Radiological Findings

Computed tomography and MRI allow for the evaluation of ventricular enlargement and brain atrophy, but diagnosis should not be based on these procedures alone.

Typical signs of hydrocephalus are said to be symmetrical communicating quadri-ventricular enlargement and flow void in the aqueduct of Sylvius. Ventricular enlargement may be detected as rounding of the frontal horns, enlargement of temporal horns ≥2 mm on both sides and upward bulging of the corpus callosum in MRI. A discrepancy between an enlarged third and a normal fourth ventricle is often seen. Cortical sulci are usually compressed apically and in the interhemispheric fissure, whereas the Sylvian fissure may be enlarged. Ventricular enlargement is calculated by various indices, e.g., Evan's ratio >0.3. Evan's ratio means the ratio of distances between the frontal horns of the lateral ventricles to the maximum biparietal diameter measured on axial slices in CT or MRI at the level of the foramen of Monro. Periventricular lucency (low density on CT, high intensity on T2-weighted MRI) adjacent to the frontal and temporal horns is often said to be a sign of hydrocephalus as transependymal CSF absorption or bulk flow, but has no significant value. Neither cortical or subcortical atrophy signs of cerebrovascular disease per se does not predict NPH. Multiple white matter lesions as result of cerebrovascular disease, on the other hand, may be a negative prognostic sign.

Magnetic resonance imaging cine flow studies show unspecific CSF flow phenomena in the aqueduct of Sylvius, which some authors believe to be of prognostic value for treatment. Most important in CT and MRI findings is the exclusion of severe brain atrophy. When clinical signs and radiological findings alone do not allow for diagnosis, CSF pressure and dynamics should be evaluated.

3.6.1.5 Spinal Tap

Withdrawal of CSF is most often used to further evaluate the patient.

Normal CSF pressure in NPH is 5–15 cm H_2O in lying position. Higher values are usually found in hydrocephalus of other origins. Withdrawal of CSF (spinal tap) of 40–50 ml of CSF is performed via lumbar puncture. The patient is examined before and several hours after spinal tap. Improvement in walking ability and consciousness is thought to give proof of CSF absorption disturbance related hydrocephalus. If the result of this test is not clear either it can be repeated after some time (days), or a temporary spinal catheter may be inserted and CSF drained 500–1,000 ml/72 h, thus simulating the effect of shunting. The effect of CSF drainage has to be evaluated clinically for at least 3 days in order to achieve reliable results.

3.6.1.6 Continuous CSF Pressure Monitoring

Invasive methods to give objective criteria for CSF pressure are continuous intracranial pressure (ICP) monitoring via ventricular drainage or intraparenchymal sensor. Epidural pressure recording is less invasive, but gives less reliable values, which is especially true of absolute pressure values. The pressure monitoring has to be recorded for at least 72 h to be able to compare ICP values and pressure curves for at least two nights, as the first night after anesthesia is often not representative. During daytime there may be too many artifacts due to activities of daily life and nursing procedures. The records are evaluated for absolute values of ICP and for specific breath- and pulse-dependent changes in ICP. Sometimes, pressure peaks at >20 cm H_2O and typical wave forms can be recorded. The time span of B-waves is recorded and set in relation to the recorded time. As B-waves seem to be associated with REM phases during sleep, up to 20% of the time is regarded as normal; if they are present more than 50% of the time, hydrocephalus is presumed.

CSF Dynamics

...e of these procedures does not result in a certain ..., CSF dynamics may be examined. While con... recording of CSF pressure is installed via either ... or lumbar drainage, artificial CSF is added or ... by bolus injection or via continuous, prefer... computer-assisted, intrathecal infusion.

...esponses of CSF pressure owing to multiple bo... allow for estimation of correlation between ... and volume (P/V). For better visualization this ... is usually shown in semi-logarithmic form; ... of the curve is called the pressure volume in... (PVI). The PVI allows the elastance of the brain to ... .

...es in CSF pressure caused by continuous added ... fluid allow for calculation of resistance to ... (CSF R_{out}). For correct interpretation of CSF dynamics via ... route, spinal stenosis and aqueductal stenosis ... be precluded.

...ed outflow resistance (R_{out} >10 mmHg/ml/min, ... mmHg/ml/min is said to be almost predictive), ... with improvement in symptoms following drain... (> 2 h, is considered useful in identifying those pa... most likely to benefit from implantation of a per... shunt.

Additional/Useful Diagnostic Procedures

...chological testing of cognitive and motor func... most helpful in differentiating between hydro... and dementia of other origin. They allow for an ... and reliable assessment of functional outcome after ... ment. This is especially worthwhile during the ... process before and after CSF withdrawal either ... tap or by lumbar drainage.

...gical and gynecological examination is essential ... urinary incontinence owing to dysfunction of ..., prostate or pelvic floor.

...therapy and occupational therapy examina... be of help in the evaluation of gait disturbance ... .

...clide cisternography may show evidence of ... block, prolonged activity in the ventricles or ... resorption over convexity, but nowadays does ... enough diagnostic power to warrant this inva... consuming method.

...rospinal fluid studies have been thoroughly ... There is no evidence of relevant diagnostic ..., but comparisons of CSF before and after shunting ... to predict response to shunting. Further follow... may indicate shunt failure.

Electroencephalography and evoked potentials are abandoned as diagnostic procedures in NPH.

3.6.1.9 Therapy

The management of hydrocephalus is fraught with complications. Approximately 53% of shunts fail and require further surgery within the first 2 years of surgery. Infection rates at most centers vary between 5 and 15% per procedure. Of all the medical devices that are implanted in the body, shunts for hydrocephalus probably have the highest failure rate.

3.6.1.9.1 Conservative Therapy

There is no useful conservative therapy at the moment. If spinal tap improves the patient's state for some time, it may be repeated, if necessary, to delay surgery.

Drugs to decrease CSF production (furosemide, acetazolamide) will not work for long enough without severe side effects.

3.6.1.9.2 Operative Therapy

Reducing CSF production by destroying the choroid plexus by surgical removal, radiation therapy, local chemical destruction or by immunotoxins associated with choroid-specific antibodies is rarely practiced. The resulting reduction is unsafe and temporary, because the choroid plexus is only one of several sites of CSF production.

Surgical procedures are aimed either at restoring CSF flow within the cranium or at lowering CSF outflow resistance by creating alternative routes for CSF drainage within the cranium or to other sites of the body where CSF can easily be absorbed.

Complications include overdrainage- and underdrainage-related problems, dislocation or disruption of the implanted devices, shunt infection and seizures. Infection is a major problem in shunt surgery. Perioperative single-shot antibiosis is commonly advocated. In at-risk patients antibacterial impregnation of shunt hardware may be warranted.

Mostly, a ventriculo-peritoneal shunt is used. Alternatives are ventriculo-atrial, ventriculo-pleural or lumbo-peritoneal shunting. Ventriculo-renal and ventriculo-gallbladder shunts are rarely used. Ventriculo-sinusoidal shunting seems to provide for the most physiological procedure, although the results are still preliminary. Some surgeons use endoscopic inner shunting procedures (e.g., third ventriculo-cisternostomy, aqueductoplasty) to avoid shunts or as a first procedure, although results are equivocal.

Typical problems of peritoneal shunts are infections within the peritoneal space, formation of pseudocysts because of omental encasement, mostly in cases of chronic infection, laceration of abdominal organs, and unintentional injury to the distal catheter during abdominal surgery. Atrial shunts are prone to more serious infections and injury to the endocardium, leading to sepsis, endocarditis, glomerulonephritis and renal failure. Formation of thrombi at the tip of the distal catheter may cause repeated, often unapparent pulmonary thrombosis, leading to pulmonary hypertonus and cardiac failure. Thrombosis of the jugular or subclavian veins, or even the superior cava vein, has been reported.

Pleural shunts require adequate absorption within the pleural space. This may be insufficient in patients with pneumonia, pleuritis, during artificial ventilation, and in diseases or other circumstances leading to pleural effusions. Because of the physiologically negative interpleural pressure, pleural shunts are especially prone to overdrainage.

If the shunt is implanted correctly, a remarkable improvement in symptoms will appear within a few days. The sooner NPH is diagnosed and treated, the better the results of shunting.

In many patients, improvement will last for several months to many years, until secondary worsening occurs. This often gives rise to repeated testing of shunt patency and further diagnostic procedures.

There is an ongoing debate as to which valve should be used. Conventional differential pressure valves provide ample drainage of CSF. However, they produce an unphysiological state of CSF pressure. In normal individuals, CSF pressure in the lying position is somewhere between 5 and 15 cm H_2O, and in the upright position between −5 and −10 cm H_2O. CSF is absorbed into the blood, usually during the night in the horizontal position. In shunted individuals with differential pressure valves with no gravitational accessories, on the other hand, CSF will be drained in the upright position according to hydrostatic forces into the peritoneal space. Thus, CSF pressure in the upright position is lowered to somewhere between −20 and −40 cm H_2O. In the lying position CSF has to be produced to regain normal CSF pressure. Therefore, the diurnal rhythm of CSF absorption and CSF pressure is just opposite to that in individuals without shunts. Gravitational valves provide for almost physiological relations concerning CSF pressure and diurnal CSF flow. They almost prevent overdrainage-related complications, but seem to more often produce underdrainage (because they do not have a daily hydrodynamic flush during erection). Often, the ventricular size is not or only to a small degree reduced by gravitational valves, despite good clinical results. This makes radiological control of the shunt function more difficult as in differential pressure valves where usually a significant reduction in ventricular size will be the result of normal shunt function. The results of the Dutch Normal-Pressure Hydrocephalus Study showed better clinical results with low-pressure valves but an increase in overdrainage-related complications.

Externally adjustable valves are more expensive but offer the chance to react to complications like overdrainage-related subdural collections by choosing a higher opening pressure and by dialing down in minimally responsive patients or in the case of secondary failure because of an ongoing disease. Some of these externally adjustable valves may be checked by external magnetic instruments, others require radiological examination by X-rays. Artifacts, unintended changes in opening pressure or even destruction of mechanical parts by MRI procedures represent further problems associated with these valves. Some externally adjustable valves now have integrated brakes to prevent unintended changes in opening pressure by MRI or other magnetic influences.

Ventriculo-sinusoidal shunts avoid differential pressure depending on the position. They seem to provide in most physiological procedures, draining ventricular CSF into the venous sinuses of the cranium. Their very short tubes allow for a minimized risk of damage to the shunting hardware. On the other hand the long-term patency of shunts and the sinusoidal drainage site is uncertain and results are still preliminary.

Endoscopic third ventriculostomy (ETV) is an alternative to shunting that may allow patients with hydrocephalus to avoid shunt implantation or allow a patient with a non-functioning shunt to have it removed. It is usually not intended for patients with iNPH, but for adults with sNPH or aqueductal stenosis. An opening is made in the floor of the third ventricle just in front of the corpora mamillaria. The opening allows CSF to flow out of the ventricle to the subarachnoid space via a bypass. Care must be taken to avoid damage to the basilar artery and the oculomotor nerves. Primary complications are fever, bleeding and disequilibrium owing to damage to hypothalamic structures. If successful, ETV avoids implantation of a foreign body and the increased risk of shunt infection and malfunction. Overdrainage-related problems are avoided, as there is no major intracranial pressure difference. Often the ventricular size is not or only to a lesser degree reduced after ETV, although good clinical recovery is observed. Secondary closure of the ventriculostomy may occur. Diagnosis of the resulting recurrence of hydrocephalus is difficult and based on clinical signs and missing flow void at the former site of the ventriculostomy.

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Useful Links

- Hydrocephalus Association www.hydroassoc.org
- Hydrocephalus Database Project www.hydrocephalusdatabase.org
- Arbeitsgemeinschaft Spina bifida und Hydrocephalus www.ASBH.de

3.6.2 Congenital Arachnoid Cysts

GIANPIERO TAMBURRINI, CONCEZIO DI ROCCO

3.6.2.1 Synonym

Congenital arachnoid cysts are also called leptomeningeal cysts. This term includes neither secondary "arachnoid" cysts (i.e., post-traumatic, post-infectious, etc.), lined with diseased arachnoidal membranes, nor glio-ependymal cysts, lined with glial tissue and epithelial cells.

3.6.2.2 Definition and Etiology

Congenital arachnoid cysts are developmental lesions that arise from the splitting or duplication of the arachnoid membrane (thus they are in fact intra-arachnoid cysts).

The etiology of these lesions has long been the subject of debate. The most accepted theory is that they develop from a minor aberration in the development of the arachnoid mater from around week 15 of gestation onward, when the cerebrospinal fluid (CSF) is generated to gradually replace the extracellular ground substance between the external and the internal arachnoid membrane (endomeninx). The malformative hypothesis is supported by the common location of arachnoid cysts at the level of normal arachnoid cisterns, their occasional occurrence in siblings, the presence of accompanying anomalies of the venous architecture (i.e., the absence of the Sylvian vein), and the association with other congenital anomalies (agenesis of the corpus callosum and Marfan syndrome).

A further unclarified subject is why arachnoid cysts tend to expand. Electron microscopy and ultracytochemical analysis have demonstrated increased Na+ and K+ pump activity within the cyst walls compared with normal arachnoid, supporting the theory of active CSF production by the cyst-lining membranes; the latter are morphologically similar to the subdural neuroepithelium and to the neuroepithelial lining of arachnoid granulations. On the other hand, cine-MRI and direct endoscopic views have shown evidence that some arachnoid cysts may enlarge by trapping CSF within them by a ball-valve mechanism. The pressure gradient for the movement of CSF into the arachnoid cyst would be ensured by transient increases in cerebrospinal fluid pressure, especially those increases brought about by cerebral artery systolic oscillations or by pulsations transmitted through the veins.

Specific problems in the definition of the pathogenesis concern intraventricular arachnoid cysts. For some authors, they represent a kind of "internal" meningocele;