Macrocrania

This presenting sign is easy to recognize if the head is too large in relation to the face or to the remaining body with the naked eye. More often, measurement of the head circumference demonstrates a value above the normal range, too high in relation to the body length, or that exceeds one percentile after the other on follow-up.

Because head circumference is measured in the fronto-occipital plane, it may be an indication of a too large skull volume, as the normal values have been obtained from a mean population of different age that show a slightly longitudinal-oval shape.

In children with dolicho- or brachycephaly, the measured values fall often into the upper or lower percentiles for geometric reasons and may therefore simulate a relative macrocrania or microcephalia, for example, in craniostenoses.

In the individual case or in longitudinal measurements, the head circumference does not at all follow a harmonious way as it is recognizable from the common percentile curves. This must be considered in case of small deviations of head circumference values, for example, in constitutional macrocrania.

A wrong measurement is not always excluded, too. Therefore, the mean value of three head circumference measurements should be calculated. After 1 year of age, 87.5%, after 5 years, 93.3% of the final value is attained, and with approximately 13 years (girls) or 15 years (boys), the adult values of 51.5–58 cm are achieved.

In Table 2.1, mainly the surgically relevant pathologies of macrocrania are listed. The significance of some of the pathologies has got smaller in comparison to older similar surveys, in part, due to prenatal ultrasound performed in developed countries for diagnosis of CNS malformations that belong together with the urogenital to the most frequently encountered pathologies.

2.1 Hydrocephalus

Occurrence, Etiology, Forms

Due to prenatal diagnosis and abortion or to a lesser degree due to prevention of spina bifida by preconceptional enrichment of the food with folic acid, congenital hydrocephalus is observed less frequently than peri- or postnatally acquired hydrocephalus, whereas the latter is possibly increased because of increased survival of pre-mature infants with intraventricular hemorrhage (former prevalence 1 isolated hydrocephalus in 1,000 live births).

The percentage of congenital and postnatally acquired hydrocephalus and of the different etiologies depends on the inclusion of children and adults, for example, 41% congenital and 59% acquired types if both ages are considered. Nevertheless, 60–90% of the cases are treated in the first year of life.

There are numerous etiologies and different forms of hydrocephalus. In congenital and con-natal hydrocephalus, malformative or prenatally acquired causes exist such as aqueductal stenosis, spina bifida (due to aqueductal stenosis and Chiari II malformation), Dandy-Walker malformations
(e.g., Dandy-Walker cyst), or hydrocephalus after infections transmitted by the pregnant women (e.g., toxoplasmosis) and after intracerebral hemorrhage (e.g., as expanding porencephalic cyst).

About 2% of the patients with congenital hydrocephalus have an x-linked hydrocephalus that occurs only in boys and may be a part of a MASA syndrome.

Among the peri- or postnatally acquired hydrocephali, previous infections (such as hydrocephalus after pneumococcal meningitis in the western world and after tuberculous meningitis in the western Cape district of South Africa and other parts of the world with high incidence of tuberculosis, or aqueductal stenosis after mumps encephalomeningitis), intraventricular or subarachnoidal hemorrhage (such as in prematurity, trauma to the head, or spontaneously), and brain tumors (such as axial tumors of the posterior fossa) belong to the most common causes.

Hydrocephalus is a disorder of CFS circulation by blockage at different sites of its pathway with dilatation of the prestenotic parts (e.g., triventricular hydrocephalus in aqueductal stenosis or dilatation of all ventricles in obstruction of the foramina of the fourth ventricle), increased intraventricular pressure, and decreased cerebral blood flow. The former are noncommunicating forms, whereas obstructions outside of the ventricles such as those at the level of the basal cisterns belong to the communicating forms of hydrocephalus.

In light of recent research results, the possibility of other CSF absorptive mechanisms must be considered in addition to the classic concept of CSF circulation such as CSF absorption by lymphatics after clearance of CSF along the sheets of the cranial nerves.

Clinical Significance

- The recognition of ventriculomegaly by prenatal screening is important because it may be combined with cerebral and/or extracerebral malformations and/or chromosomal aberrations, and may be the start of progressive hydrocephalus.
- Pathology, pathophysiology, clinical presentation, and prognosis depend not only on the hydrocephalus but also substantially on its etiology.
- The disorder of the cerebrospinal fluid (CSF) circulation can be treated always effectively in contrast to the etiology.
- Non- or delayed treatment leads to death, mental, and neurological deficit.
- Treated hydrocephalus needs lifelong follow-ups.

Clinical Presentation

Prenatal screening and diagnosis: In general, ventriculomegaly is the most frequent issue on ultrasound. In contrast to hydrocephalus that is defined as ventriculomegaly combined with abnormal increase of head circumference, ventriculomegaly means only a lateral ventricle

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**Table 2.1 Differential diagnosis of macrocrania**

<table>
<thead>
<tr>
<th>Category</th>
<th>Conditions</th>
</tr>
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<tbody>
<tr>
<td>Hydrocephalus</td>
<td>Shunt failure, shunt infection, abdominal CSF pseudocyst, slit ventricle syndrome, isolated fourth ventricle hydrocephalus, Chiari II and I malformations</td>
</tr>
<tr>
<td>Constitutional macrocrania (benign familial macrocephaly)</td>
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<tr>
<td>Intraventricular (periventricular/intraventricular) hemorrhage of the newborn</td>
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<tr>
<td>Transient ventriculomegaly, progressive hydrocephalus</td>
<td></td>
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<tr>
<td>Catch-up growth of head circumference</td>
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<tr>
<td>Congenital intracranial cysts</td>
<td></td>
</tr>
<tr>
<td>Arachnoid cyst, Dandy-Walker malformation and variant, porencephalic cyst, septum pellucidum cyst, cavum vergae and velum interpositum cyst</td>
<td></td>
</tr>
<tr>
<td>Subacute and chronic subdural hematoma (hygroma)</td>
<td></td>
</tr>
<tr>
<td>External (extraventricular) hydrocephalus, subdural collection due to meningitis</td>
<td></td>
</tr>
<tr>
<td>Encephalopathies, metabolic disorders with brain involvement</td>
<td></td>
</tr>
</tbody>
</table>

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atrium larger than 10 mm independent of term on ultrasound or MRI (measurements in the coronal and/or axial plane; >15 mm = severe ventriculomegaly). During the second half of gestation, the atriocerebral ratio (ratio between atrial diameter and biparietal brain diameter) decreases normally from 13.6 to 8 %. Ventriculomegaly is caused by CSF accumulation, brain atrophy, or dysgenesis; in one third of the patients, a resolution, and in less than two thirds, stabilization is observed during gestation. The recognition of ventriculomegaly is important because it is often combined with cerebral and/or extracerebral malformations and/or chromosomal aberrations (in up to three fourths and one third, respectively), and can the beginning of progressive hydrocephalus (>10 %). In general, isolated mild ventriculomegaly (10–15 mm) means a postnatal development delay in round 10 %.

A posterior asymmetric enlargement of the ventricles and the parieto-occipital subarachnoid spaces may be a precursor of an external hydrocephalus.

Depending on the age of the child, the type of progression, or stage of hydrocephalus, the clinical presentation is different. The latter include acute, progressive, or chronic, compensated, arrested, or shunted hydrocephalus with overt or insidious blockage or with compensation or arrest.

**Newborns and infants:** Unspecific symptoms and signs are food refusal, irritability, apathy, and arrest or loss of developmental milestones. On inspection and/or palpation, the following findings are present: macrocrania pre- or postnatally, or progressive increase of head circumference (HC) afterward with crossing of one percentile after the other with disproportion of the large neurocranium in relation to a small face (normal values of HC are at term ≤37 cm in boys and ≤36 cm in girls). Furthermore, distended scalp veins, widening of the cranial sutures, enlarged, tense, and bulging anterior fontanel (the normal anterior fontanel is soft and sunken in the quiet patients held in upright position), and setting sun sign # are encountered.

**Older children:** Depending on the acuteness of development of the disorder of CSF circulation, more or less distinct signs of increased intracranial pressure (ICP) are prominent such as headache (characteristically also at night and in the morning), vomiting, papilledema on fundoscopy, and split cranial sutures on plain skull x-ray. In addition, abducens nerve palsy, paralysis of upward gaze (Parinaud’s syndrome), and other ophthalmological and neurological deficits may be encountered.

In case of compensation, the signs of increased ICP may be discreet except for macrocrania. The preserved language (chatterbox) and memory abilities delude the examiner about the deficits of general and school performance, of behavior, and neuropsychological examination. Episodes of spontaneous exacerbation of hydrocephalus, for example, after head injury may lead to aggravation and specification of the clinical presentation.

**Shunt failure:** Although shunt failure corresponds often to an acute or subacute hydrocephalus with possible clouding of consciousness from apathy to coma, there is often a dissociation of the common symptoms and signs of increased ICP and findings of neuroradiological imaging, and uncommon, isolated complaints are possible such as blurred vision, etc.

In shunted patients, shunt failure must always be considered, and the needed work-ups must be carried out timely because severe deficits may develop, for example, visual loss in approximately 2 % that is completely irreversible in at least one third of the involved children. On checking the valve, the only reliable sign of obstruction is a permanently flat chamber after single decompression (#/#).

The work-up examinations are the same as quoted under slit ventricle syndrome. Ultrasound of the optic nerve sheath yields somewhat earlier and more reliable pathological findings than fundoscopy (lacking papilledema after optic atrophy). Normal are values of <4 and <4.5 mm in infants and older children, respectively. Nevertheless, variation from the asymptomatic baseline value is the most sensitive variable in determining development of hydrocephalus in the individual case.
Natural History
Without treatment, 45% of the patients with hydrocephalus are still alive 7 years after the initial diagnosis, and at least two thirds of them have severe mental and neurological deficits, and all are conspicuous in everyday life. To a lesser degree, the same applies to patients with compensation of hydrocephalus after shunt blockage. As in native hydrocephalus, acute exacerbation of hydrocephalus with fatal outcome is possible even after several years.

After shunting and long-term observation, several types of shunt dependency are observed with the majority remaining shunt-dependent (Fig. 2.1).

Differential Diagnosis, Work-Ups
The differential diagnosis includes pathologies with apparent or real macrocrania, with clouded consciousness or unconsciousness (acute hydrocephalus, spontaneous exacerbation of hydrocephalus, or in shunt failure) and with signs of increased ICP or other neurological presentations. In addition to the pathologies described in this chapter and those quoted in the chapter on clouded consciousness or unconsciousness, the following disorders must be considered: fourth ventricle hydrocephalus, (Arnold) Chiari II and I malformations, and syringobulbia and -myelia; the latter occurs mostly combined with Chiari I and II malformations.

Isolated fourth ventricle hydrocephalus occurs usually after long-term conventional shunting in children with postmeningitic, posthemorrhagic, and hydrocephalus with former shunt infection or due a congenital malformation; communicating hydrocephalus, aqueductal stenosis, or occlusion of the foramina of the fourth ventricle may be present, for example, in Dandy-Walker cyst. The increased volume of the fourth ventricle with or without a clinical symptomatology is due to an imbalance between the supra- and infratentorial ventricular system with increased IVP of the latter and upward shift of the brain stem on condition that there is an aqueductal stenosis or insufficiency on the one hand and an occlusion of the fourth ventricle foramina on the other hand.

Clinically, there is either a slowly progressive symptomatology with ataxia, apathia, and diplopia over years or an acute life-threatening clinical presentation of a posterior fossa mass with cranial nerve deficits and cerebellar tonsil herniation. Treatment option is either a double shunt (ventricular and fourth ventricle catheter with a common valve and distal catheter) or endoscopic stenting in addition to a conventional shunt. Isolated shunt of the fourth ventricle and, to a lesser degree, a double shunt are prone to complications such as primary or secondary injury of the brain stem parenchyma or of the floor of the fourth ventricle or overdrainage with brain stem tethering with the appearance of cranial nerve deficits.

In Chiari II malformation, there is a hindbrain deformity with a small posterior fossa and...
impaction of the posterior cerebellum through the foramen magnum; the elongated fourth ventricle extends as far as the cervical canal and its foramina are obstructed by parts of the cerebellum and arachnoidal adhesions. Precise individual pathoanatomical and flow characteristics can be determined by CT and MRI. By compression and/or distortion of the cranial nerves and the brain stem, life-threatening symptoms may occur such as apneic or cyanotic attacks, respiratory distress syndrome, difficulty in swallowing, and vocal cord paresis. There is no consensus about the most appropriate initial therapy in symptomatic Chiari II malformation. In already shunted children, a thorough evaluation of shunt function is needed because the symptomatology disappears or is lessened by optimal shunt function. In case of life-threatening and persistent severe symptoms, despite a normal shunt function, decompression of the upper cervical canal (occipital decompression is not necessary in large foramen magnum) and dural expansion are indicated. It may lessen the symptoms and those of an associated syringomyelia in at least 75%; the latter needs only a syrinx shunt to the subarachnoid or peritoneal space in case of failure of decompression. Rarely, decompression must be combined with a double ventriculo- and cisterna magna-peritoneal shunt.

In Chiari I malformation, there is a descent of the cerebellar tonsils into the cervical canal. It is diagnosed by MRI (abnormal position of the cerebellar tonsils below the foramen magnum) during work-ups of skeletal abnormalities of the cervical spine, craniocervical junction, or scoliosis in which associated Chiari I malformation is often encountered or due to neurological deficits in the second decade. The symptomatology is caused by compression/distortion of the dura, brain stem, lower cranial nerves, cerebellum, or by an associated syringomyelia. It includes occipital or cervical pain that is paroxysmal (triggered by Valsalva maneuver such as coughing) or persistent (analogous presentation in young children by crying and neck hyperextension), weakness and spasticity of extremities, and in 20 %, sings such as vocal cord paresis, recurrent aspiration, and down beating nystagmus. In addition, weakness of the upper limbs (intrinsic hand muscles) or only absence of superficial abdominal reflexes may be observed. The treatment includes foramen magnum decompression and dural expansion that is not indicated in asymptomatic patients except for those with scoliosis.

**Work-ups:** Ultrasound, CT, and MRI belong to the main imaging procedures #/#/#. They confirm the clinical diagnosis of hydrocephalus, describe its degree (volume of the lateral or all four ventricles, brain mantle thickness, etc.) and the involved parts (e.g., triventricular hydrocephalus), demonstrate possible additional findings, and allow the differential diagnosis from other pathologies. The involved parts of the CSF compartments and additional findings point to the probable site of obstruction and/or possibly to the etiology.

For follow-ups, it is important to know that changes of ventricular size in ventriculomegaly are only recognizable from 20 % upward by the naked eye.

With the advent of endoscopic surgery, preoperative evaluation of the type of hydrocephalus (e.g., noncommunicating vs. communicating) and postoperative monitoring (determination of stoma patency, changes in ventricular volume) became indispensable; phase-contrast cine flow MRI and air encephalography are examples of such examinations.

Neurological, ophthalmological, neuropsychological, genetic examinations and laboratory blood and CSF tests (e.g., increased CSF levels of IgM and IgG anti-paramyxovirus in aqueductal stenosis after mumps) allow to demonstrate the etiology of hydrocephalus and possible associated pathologies, malformations, and syndromes for prognostic purposes or to describe the hydrocephalus for further follow-ups.

CSF pressure measurement of 24–48 h by an intracranial route allows the differentiation between shunt-dependent compensated and shunt-independent arrested hydrocephalus in apparently asymptomatic patients who have been shunted or not (##).
Therapy
Most commonly used are the ventriculoperitoneal (VP) shunt and endoscopic procedures such as third ventriculostomy (ETV). In contrast to the generally applicable shunting, endoscopic procedures need specific indications because the success rate depends on the age, on the site of CSF blockage, and on the etiology, respectively. Both methods have different advantages and drawbacks.

Fetal surgery by placement of a ventriculo-amniotic shunt has been disappointing so far. Actually, the prerequisites that concern meaningful indications (e.g., type of hydrocephalus in which there will be irreversible damage if left untreated) and suitable surgical methods are not yet established for a renewed trial. The outcome depends specifically on the time of onset of hydrocephalus in general and/or on the stage, type, and clinical category of congenital hydrocephalus (perspective classification of congenital hydrocephalus).

Shunt implants are expensive especially in case of repeated shunt failure. Shunt revisions performed in time are nearly impossible in not accessible parts of the third world.

In contrast to the differential pressure valves that produce an unphysiologically negative ICP pressure in upright position, the newer generation of valves (hydrostatic valves, programmable valves, antisiphon devices, and so-called variable-resistance or flow-regulated valves) overcome or lessen overdrainage but may be combined with the drawbacks of significant risks of shunt insufficiency. Radiological signs of overdrainage are postponed, and occurrence of SVS is possibly decreased.

Shunt surgery (Fig. 2.2): If one lateral ventricle is larger than the other, this side is chosen for shunting. The child is in supine position with the head turned to the contralateral side, and the planned incisions are marked: transverse incision lateral to the umbilicus and hockey stick incision in the parieto-occipital angle or lateral to the anterior fontanel. The skin is covered by a film, and the parts of the shunt never touch the naked skin. After the cranial and abdominal incision, the shunt passer is introduced from one to the other incision and replaced by a thread that is used to pull through the peritoneal catheter. After a burr hole in the parieto-occipital angle or 1 cm in front of the coronal suture and laterally to the midline, the ventricular is introduced with its tip in the frontal horn, the ICP measured, and CSF for culture, protein, and cells collected. Afterward, the ventricular catheter is connected with an intervening Rickham ventriculostomy reservoir and
the chosen valve with the peritoneal catheter. After a transverse incision of the fascia of the rectus muscle and longitudinal spreading of its fibers, the peritoneal catheter is introduced in the abdominal cavity for 30–40 cm that allows free motion and sufficient length for the first decade of life. In contrast to sutures to fix the reservoir and the sites of connection safely, the peritoneal catheter is not restraint by sutures. The incisions are carefully closed in two and four layers, respectively.

Shunts are prone to the following complications:

1. **Lifelong shunt dependence** in most of the patients; only about 10% of the patients become shunt-independent with shunts used in former times.

2. **Shunt failure (dysfunction).** A prognosis for the individual patient is difficult; the history of a straightforward shunt implantation without time delay is one of several prognostically favorable factors. Possible results are as follows: 0.15 shunt revision per patient per year observation time; after 7 and 12 years has 60 and 53%, respectively, of a shunt population, zero or one shunt revision experienced; and the remainder patients had 2 or more shunt revisions.

   The **main causes of shunt dysfunction** are ventricular or central catheter obstruction by debris or in small or slit ventricles and growth of the body length (with final localization of the end of the peritoneal catheter outside the peritoneal cavity) or disconnection of parts of the shunt system. Less frequent causes are abdominal CSF pseudocyst, failure of the valve due to technical properties or unintentional changes of adjustable valve systems by MRI, distal or proximal catheter migration, localized trauma to the shunt, and complications in alternative methods such as in ventriculopleural or gallbladder shunt.

   The **abdominal CSF pseudocyst** is mostly caused by shunt infection (occurrence 0.7–4.5%) and only rarely due to silicone allergy (due to loose, unbounded silicone oil) that leads to adhesions between the intestinal loops by connective tissue and compartmentalization of the drained CSF as a pseudocyst. Clinically, abdominal pain and distension is combined with signs of shunt dysfunction. The same proceeding is indicated as in shunt infection. After cure, reimplantation of a new ventriculoperitoneal shunt is often possible, or another distal route must be chosen. In case of allergy, systems with extracted silicone must be used.

3. **Shunt infection.** Shunt infection is mostly introduced by surgery with *Staphylococcus epidermidis* as the most frequently observed germ. Less frequently is colonization of shunts by puncture of the reservoir, after ulceration of the overlying skin in very young infants, or during septicemia. It may be combined with shunt dysfunction.

   By appropriate surgery, it is possible to keep the infection rate as low as ≤1% of interventions. Shunt infection is treated at best by removal of the shunt, external ventricular drainage, and systemic and, if needed, by topic antibiotics. Reinsertion of a new shunt after clearance of the CSF infection is possible within 10 days to 2–3 weeks. The results of implantation of shunts impregnated by antibiotics to avoid infection are only partially convincing.

4. **Overdrainage** is one of the main problems in shunt surgery and is caused by siphon effect in upright position.

In **slit ventricle syndrome (SVS),** overdrainage of the ventricles by the commonly used shunts in the past becomes symptomatic at different times after shunting in infancy. Overdrainage is not only recognizable by an SVS but also primarily by asymptomatic radiological findings such as small or slit ventricles (ventricular volume below the normal value for age and sex or ventricles not or only recognizable as minute structure) and diminished skull growth (low modulus). Clinically, the head circumferences fall gradually below the 50%.

SVS is characterized by intermittent, more or less dramatic and threatening episodes of severe headache, heliophobia, varying degrees of lethargy, and/or nausea and vomiting in an otherwise healthy child. In a shunt population, up to two thirds have small or slit ventricles on neuroimaging, but only a part of them had an SVS (1–37%).
The differential diagnosis includes low CSF pressure syndrome combined with a sensitivity to low CSF pressure, intermittent shunt dysfunction, and/or cerebral vasomotor instability similar to migraine. The same disorders occur probably one behind the other or combined in the same patient and episode.

The aim of diagnostic work-up is mainly to exclude complete or partial obstruction of the ventricular catheter by shuntogram (no entry of contrast into the ventricles or only a trace), neuroimaging (enlargement of slit ventricles occurs only delayed if at all), and ICP recording by the Rickham reservoir (no or false-negative recording due to encapsulation of the catheter tip). In addition, careful clinical examination including fundoscopy and optic nerve ultrasound is irreplaceable tools.

Except for complete ventricular catheter obstruction and/or life-threatening increased ICP, delayed surgery should be performed whenever possible with preliminary application of i.v. infusion, steroids, analgetics (or antimigrainous drugs), and Trendelenburg’s position. Shunt revision includes replacement of the valve and, if needed, of the ventricular catheter. Prophylaxis by implantation of a new generation of valves at the time of the primary shunt is superior to the former. The new generation of valves (hydrostatic and programmable valves, antisiphon devices, and variable-resistance and so-called flow-regulated valves) overcome or lessen the overdrainage of the former differential pressure valves but are combined with the drawbacks of significant risks of shunt insufficiency.

Epilepsy is observed in up to one fifth of a shunt population. Because it is mostly a sequel of the cause of hydrocephalus and not of its treatment, it is not a typical shunt complication. On the other hand, exacerbation of epilepsy or its deterioration may be a sign of shunt dysfunction.

Neuroendoscopic treatment of hydrocephalus may include aqueductal stenosis and fourth ventricle outlet obstruction, isolated fourth ventricle hydrocephalus, complex compartmentalized hydrocephalus, and hydrocephalus caused in congenital intracranial cysts. It is contraindicated in communicating hydrocephalus. The proof of open subarachnoid spaces may be difficult even by modern neuroimaging (cine flow MRI, if not available, lumbar air encephalography instead of flow studies with isotopes or contrast), for instance, in secondary aqueductal stenosis after hemorrhage, meningitis, long-term shunting, or due to the hydrocephalus itself.

The aim of treatment, the applicable procedures, and the results are different for the quoted pathologies. In isolated aqueductal stenosis or fourth ventricle outlet obstruction, the endoscopic third ventriculostomy (ETV) creates a communication to the basal subarachnoid spaces and cisterns. The success rate depends on the age and probably on the etiology, for example, worst outcome in patients younger than 3 months and with another than a simple idiopathic aqueductal stenosis, and amounts to ≥70 % with ≥1 year of age. In distal membraneous aqueductal stenosis and isolated fourth ventricle hydrocephalus, aqueductoplasty (or interventriculostomy) with stenting in the latter may be considered which allows an unrestricted flow and equalizes the pressure difference between the ventricles.

For complex compartmentalized hydrocephalus, the option is elimination of multiple shunt systems that is achieved by fenestration between isolated intraventricular compartments and the ventricles. The success rate is 60 % and more with the best results in unilateral (monoventricular) hydrocephalus. In hydrocephalus caused by congenital intracranial cysts, their expanding and obstructive effect is eliminated by ventriculocystomy or cystocisternostomy with fair results.

The drawbacks of neuroendoscopic techniques are a significant learning curve, inadvertent bleedings, and neurological deficits, for instance, temporary or permanent oculomotor paresis or disconjugated gaze, reclosure of the created communications, for example, with the need of a secondary ETV, and pitfalls due to an insufficient information from the work-ups.

After neuroendoscopic treatment, long-term follow-ups are necessary by neuroimaging and clinically because the presence of a functional stoma is not always equal to a significant clinical recovery. On the other hand, the symptoms
may improve and do not always completely resolve. After surgery, the ventricular volume falls to a lower value than preoperatively, and stabilization of the volume of the ventricles is achieved within 3–6 months, although it does not return to normal values for age and sex. In case of persistent or recurrent symptomatology and/or persistently increased (as preoperatively) or increasing ventricular volume, reclosure or insufficiency of a functional stoma must be considered. Unfortunately, the long-term effect of moderately enlarged ventricles on the outcome has not yet been studied thoroughly, and exacerbation of hydrocephalus with acute neurological deterioration has been observed after minor head injury in the same way as in compensated hydrocephalus.

Prognosis
It depends on the cause and stage of hydrocephalus as well as on the care and experience of the involved surgeons for the primary surgery and the long-term follow-ups.

In up to two thirds of the patients with isolated hydrocephalus, a normal intelligence (school performance, neuropsychological tests) can be observed with independent lifestyle. Thirty percent of the patients can be educated for practical tools of daily life, and less than 10% are dependent on somebody else’s help (personal hygiene, food intake, locomotion, and body position).

The prognosis is possible at school age in 90%. Neuropsychological tests are necessary prior to the choice of the career or the job due to a possibly reduced performance in special fields.

In connatal hydrocephalus (head circumference at term above the normal values), up to 53% will have no mental deficit and the remainder different degrees of mental retardation.

### 2.2 Constitutional Macrocrania (Benign Familial Macrocephaly)

#### Occurrence, Clinical Significance
Frequent constitutional macrocrania is observed in some sibs and specific regions. It is diagnosed in up to two thirds of children allocated to a tertiary center for work-up of a too large head.

- Severe pathology may be considered due to the large head by parents and general practitioner.
- On the other hand, macrocrania may be observed sometimes in syndromes, hamartomas, encephalopathies, metabolic disorders with brain involvement, and skeletal dysplasias such as achondroplasia.
- Genetic or familial constitutional microcrania exists by analogy with constitutional macrocrania.

In constitutional microcrania, the neurocranium is well-proportionate to the face. The head circumference is below or close to the second percentile and below the percentile of the body length, and there is a normal intelligence. Constitutional microcrania is an important differential diagnosis of microencephalia. Occasionally, it is associated with benign external hydrocephalus.

#### Clinical Presentation
The history yields often members of the same family with large heads. The large head is adjusted to the face but not necessarily to the body length. The head circumference lies above or close to the 98th percentile and runs parallel with it, although short-term increases above the individual growth curve may be observed for several reasons. The head circumference of one of the parents is often above the normal values, and photographs of family members may demonstrate macrocrania.

#### Differential Diagnosis, Work-Up Examinations
The differential diagnosis may be clinically difficult if the head circumference does not increase harmoniously. In addition to the differentiation from the pathologies quoted in this chapter, the disorders listed under clinical significance must be considered in case of abnormal findings in history, general and neurological examinations, or in neuropsychological tests and their course.

In infants with open fontanel, ultrasound allows to exclude some of the major causes of
Later and on follow-up, MRI (or CT) excludes most of the differential diagnoses and reassures parents and other involved people. Nevertheless, clinical follow-up including the course of neuropsychological development is indicated together with the common medical checkups.

### Prognosis
The prognosis is excellent. During pregnancy, descendants of members with familial macrocephalia need special consideration of the head size of the unborn patient and if large of their parturition.

### 2.3 Intraventricular (Periventricular/Intraventricular) Hemorrhage of the Newborn (in Prematurity)

#### Occurrence
In contrast to the intracranial hemorrhage (IVH) of the newborn at term that occurs less frequently and mostly after birth injury (for instance, after instrumental delivery and less often in platelets and coagulation disorders, its general prevalence is 5–6 per 10,000 live births), the so-called intraventricular hemorrhage is observed in the immediate postpartal period in 40 % and more of premature infants (<35 gestational week and <1,500 g) and in the newborn small for date.

The originally germinal matrix, periventricular hemorrhage can be divided into four grades of which mainly grade III and IV with intraventricular hemorrhage and ventricular dilatation and parenchymatous hemorrhage, respectively, are of clinical significance in the context of macrocephaly.

#### Clinical Significance
- Intraventricular hemorrhage may lead to death and to neuropsychological deficits of the surviving patients. In the extremely low-birth-weight infants (<1,000 g), about 35 % will die, and only 5–30 % of the surviving will have a normal neuropsychological outcome.
- Up to 50 % and more premature newborns with intraventricular hemorrhage may develop ventriculomegaly of different degree; the most severe ventricular dilatations occur significantly in grade III and IV intraventricular hemorrhages.
- Most of the ventriculomegalies are transitory, but their clinical significance is not well-established, and less than 10–15 % (and more in the extremely low-birth-weight infants) with intraventricular hemorrhage develop posthemorrhagic hydrocephalus within the neonatal period or delayed after 1/2–2½ years after temporary stabilization or regression of the initial ventriculomegaly.
- Posthemorrhagic hydrocephalus is mostly communicating. Nevertheless, combined or isolated aqueductal stenosis or closure of the fourth ventricle outlets may be observed as well.

#### Clinical Presentation
The clinical presentation of the intraventricular hemorrhage is unspecific and depends on the magnitude of hemorrhage. Apathy or stupor, convulsions or tremulousness, in- or decreased muscle tone, apneic spells or need for artificial ventilation, bulging anterior fontanel, paleness and anemia are possible symptoms and signs.

In transitory ventriculomegaly, the observed ventricular dilatation is stable or regressive, and the increase of head circumference is less than 2 cm per week, and clinical signs are not demonstrable.

In progressive hydrocephalus, the observed ventricular dilatation is increasingly combined with some of the unspecific signs quoted above or with signs of increased intracranial pressure such as tense and prominent anterior fontanel. The abnormal increase of head circumference (≥ 2 cm or more per week) lags typically behind in relation to the ultrasound findings of the developing posthemorrhagic hydrocephalus in the premature infant.
These criteria are somewhat arbitrary like a working hypothesis, and the transition from transitory ventriculomegaly to progressive hydrocephalus may occur insidiously and only be confirmed by follow-up.

Work-Ups
To recognize a possible peri- and intraventricular hemorrhage, all neonates at risk (premature and low-birth-weight infants at term) must be followed by ultrasound in regular intervals. In case of a probable posthemorrhagic hydrocephalus, MR or CT is indicated. In case of equivocal clinical and sonographic findings or if the former neuroimaging is not available, a ventricular tap by the anterior fontanel with measurement of the intraventricular pressure (and examinations of the CSF leukocytes and cultures) is a useful adjunct. Amplitude-integrated EEG activity is an example of continuous functional cerebral monitoring (increased discontinuity of background activity and onset of nearly isoelectric pattern) that precedes abnormal sonographic and clinical signs and can be used for the indication of surgical intervention.

Therapy
In the early stage of progressive ventriculomegaly with hematohydrocephalus, an external ventricular drainage (EVD) is indicated to avoid secondary brain damage by the increased intraventricular pressure and to evacuate the hemorrhagic CSF. By proper handling of the EVD and collection of the CSF, there is no time limit. Less useful alternative methods are subgaleal shunt, serial punctures of a subgaleal reservoir, or lumbar punctures.

The possible negative role of transitory ventriculomegaly for the functional outcome, the development of posthemorrhagic hydrocephalus in intraventricular hemorrhages, and their avoidance by appropriate measures is still a matter of debate. Nevertheless, the normal intraventricular pressure in premature infants is lower than in neonates at term and estimated to be zero cm water or less.

In early hydrocephalus, a ventriculoperitoneal shunt should be performed, whereas in delayed hydrocephalus with proven isolated aqueductal stenosis, an ETV is a reasonable alternative.

Prognosis
The prognosis of intraventricular hemorrhage depends on the stage and the gestational age, and birth weight of the patient. In posthemorrhagic hydrocephalus, the time of continuous normalization of the ICP by different measures and the number of shunt complications are additional prognostic factors. On the other hand, spontaneous arrest of shunted hydrocephalus may occur after several years occasionally.

2.4 Catch-Up Growth of Head Circumference

Occurrence
An abnormal growth of head circumference may be observed in the period of maximum brain growth (first trimenon) after severe illnesses, malnutrition, artificial respiration, and in preterm infants or with low birth weight for date. Sometimes, it may be combined with a prominent and tense anterior fontanel and other signs of increased intracranial pressure typical for this age group.

Clinical Significance
- If the quoted accompanying disorders and/or their treatment are not considered, the abnormal increase of head circumference is interpreted as a presenting symptom of a severe underlying pathology, and work-ups are performed; ultrasound is sufficient under these circumstances.

Prognosis
The abnormal head growth returns to normal values for the individual patient within months provided the development is normal.
2.5 Congenital Intracranial Cysts

Occurrence, Types, Cyst Volumes
Due to prenatal investigations with ultrasound and MRI, an increased number of congenital intracranial cysts are recognized (>1:17,000 fetuses). Significant new informations can be gained by regular pre- and postpartal long-term follow-ups of such patients that differ from those of previous retrospective studies with variable conditions of admission. They concern the occurrence, distribution of types and locations, and natural history of congenital intracranial cysts.

In a cohort of postnatally symptomatic congenital cysts, about 50% are arachnoid cysts (malformative intracranial cysts), 35% are Dandy-Walker malformations, and 15% are porencephalic cysts. In contrast to the former entities of congenital intracranial cysts that have a developmental origin, the latter are mostly prenatally acquired pathologies. The greatest volumes may be encountered in arachnoid cysts (mean volume for the convexity type 100 ml), followed by porencephalic cysts (mean volume 90 ml) and Dandy-Walker cysts (mean volume 50 ml). The remainder cysts reveal a mean volume of 10 ml or more. If measured, there is often a CSF pressure difference between cyst and ventricular system (by a mean value of 60 mm H₂O).

Clinical Significance
• Two thirds of the symptomatic congenital intracranial cysts are diagnosed in the first year of life and 50% of these in the neonatal period, mostly due to macrocrania or hemimacrocaphaly combined with signs of increased ICP.
• Congenital intracranial cyst is a potpourri of different disease entities (namely, the three main groups and additional pathologies) and of different types of the same disease entity. Therefore, their clinical presentation and natural history, treatment, and prognosis may differ considerably.
• An increasing number of intracranial cysts are recognized already in the second half of pregnancy that puts enormous pressure on the obstetrician and the attending pediatrician

and/or surgeon (fetal board) from the involved parents.
• On the other hand, the increasing knowledge gained by careful prenatal investigations and postnatal follow-ups or autopsies allows more differentiated recommendations about the unborn patient and for its parents.

Disease Entities
Arachnoid cysts (malformative intracranial cysts): They arise in the second half of pregnancy as a hypoechogenic mass of variable size. The adjacent normal brain and/or ventricles are often shifted or lifted up. Two thirds of the cysts are supratentorial #, and one third are infratentorial including the incisural (dumbbell-shaped) cysts #. The most frequent types are interhemispheric and retrovermian cysts, respectively. Three fourths of the cysts do not increase in size during pregnancy, whereas 20% increase. Although >10% are combined with ventriculomegaly, only a small portion develops hydrocephalus during pregnancy.

On the other hand, nearly 30% of the prenatally recognized arachnoid cysts become symptomatic after birth, among other things due to an increase of the cyst or due to hydrocephalus in early infancy. Hydrocephalus may be due to obstruction (posterior fossa cysts), due to displacement or trapping of parts of the ventricles, and due to additional involvement of the arachnoid space (communicating hydrocephalus).

In symptomatic arachnoid cysts, there is a significant peak in the two first years of life, although they can be observed throughout the whole childhood. They have no or only an insufficient communication with the arachnoid space. In some of the cysts, an intermittent symptomatology is possible due to spontaneous regression and regrowth by cyst rupture or changing communication.

Signs of increased ICP often combined with macrocrania are the leading symptoms in more than 50% and are recognized mainly in infancy, about 30% have focal signs corresponding to the cyst, and more than 15% have seizures. In up to 15%, arachnoid cysts are discovered by chance by CT for different reasons or in same proportions
after head injury. In the latter group, several previously silent arachnoid cysts, for example, middle cerebral fossa (temporobasal) cysts, become symptomatic due to an impact on or a rupture of the cyst (<10% of the patients) with intracranial, subarachnoidal, or subdural hematoma or due to simple cyst emptying.

Some clinical features need special attention: hemimegalencephaly (convexity and middle fossa cysts) with temporal bulging and possible exophthalmos in the latter; large posterior fossa, gait disturbances, and truncal ataxia (posterior fossa cysts including cysts of the cerebellopontine angle) with eighth (hearing loss, tinnitus, or Meniere’s disease), seventh nerve palsy, or hemifacial spasms in the latter. Optic atrophy, visual field defects, and hypopituitarism occur in sellar cysts. In bobble-head doll syndrome, there is a rhythmic head bobbing in toddlers during walking that disappears in recumbent position. The causative cystic lesion in the region of the dilated third ventricle is often a suprasellar arachnoid cyst that leads to an intermittent obstruction of the foramina of Monro.

The frequency, pattern of symptoms, and their great variation are explained in part by the preference of Sylvian fissure (middle cerebral fossa) and cerebellomedullary types (posterior or posterolateral cerebellar surface) of arachnoid cysts (35 and 15%), cerebral convexity #, and sellar cysts (each >10%). Interhemispheric and quadrigeminal plate cysts are the less frequently observed arachnoid cysts. Asymptomatic arachnoid cysts are encountered in 1 of 1,000 adult autopsies.

Dandy-Walker malformations (Dandy-Walker cyst and variant or complex): This group of, at first glance, similar malformations occurs in 1 of 25,000–35,000 live births. With the advent of new neuroimaging techniques, efforts have been made to define true Dandy-Walker malformation (Dandy-Walker cyst #) with regard to a safer prognostication by the following features: A large median posterior fossa cyst which communicates largely with the fourth ventricle, an upward displaced tentorium, an anterolateral displacement of seemingly normal cerebellar hemispheres, and a vermis which is either rotated, raised, small, and comes in contact with the tentorium (partial agenesis of the cerebellar vermis), or a malformed and dysplastic vermis.

The latter feature allows differentiation of two prognostic groups. Dandy-Walker complex includes a wide variety of similar malformations that fit not to the described features and have not uniform prognoses.

Hydrocephalus occurs in 70–100% of the two prognostic groups of true Dandy-Walker malformation. A major challenge of Dandy-Walker malformation combined with hydrocephalus is a possible compartmentalization of the fourth ventricle and its cyst from the supratentorial ventricular system by a primary or secondary aqueductal stenosis or insufficiency with the development of isolated fourth ventricle hydrocephalus. In contrast to the arachnoid cysts, numerous associated central nervous system and systemic malformations (cardiovascular, urogenital, intestinal, facial, and extremities) or genetic disorders are observed in Dandy-Walker malformations. All may have an impact on survival and the necessary treatment options whereas the cerebral anomalies (excluding hydrocephalus) and genetic disorders on the neuropsychological prognostication.

Clinically, most of the true Dandy-Walker malformations are recognized in the neonatal period or in the first year of life due to the bossing of the posterior fossa (if looked for precisely on clinical examination #) and/or macrocrania combined with signs of increased intracranial pressure. Later or with the development of an isolated fourth ventricle hydrocephalus, subtle or distinct (delayed motor development and ataxia), and sometimes life-threatening posterior fossa signs can occur which may simulate a malignant posterior fossa tumor.

In porencephalic cysts, a focal deficiency within the cerebral parenchyma is filled with fluid, and most of these congenital cysts communicate with the lateral ventricle. True congenital forms are restricted to one vascular territory (often absence of the middle cerebral artery). In the more frequently, later in pregnancy acquired forms, anoxemia, intracerebral hemorrhage #, and theoretically ventricular puncture, closed
Macrocrania
head injury, and penetrating wounds of the brain are possible causes. With the advent of ultrasound and MRI applied in pregnancy, their development can be observed prior birth, for instance, after a large intracerebral hemorrhage of the fetus which ends up later with a porencephalic cyst. In general, such intrauterine events need postnatal follow-ups and the symptomatic ones treatment.

Some of them exert a mass effect on follow-up by an enormous increase of their size and/or an insufficient communication with the ventricle with shifting of the midline and compression of the brain and the adjacent ventricles.

Clinically, macrocrania or hemimacrocephaly combined with signs of increased intracranial pressure (possibly associated with bulging and thinning of the skull overlying the cyst), lateralizing signs corresponding to the involved brain area (hemiparesis or focal seizures which can be lessened or disappear after decompression), and unspecific signs such as apathy, irritability, and failure to thrive can be observed. In addition, often delay of mental and motor development is observed.

By the distribution and the shape of the defect, and by the finding of an avascular area, a differentiation between the two forms of porencephalic cyst is sometimes possible.

Differential Diagnosis, Work-Ups
The differential diagnosis includes other, less frequently symptomatic congenital intracranial cysts such as subcallosal cysts (septum pellucidum cyst, cavum vergae, and cavum velum interpositum cyst), neuroepithelial cysts (colloid cyst of the third ventricle), Rathke’s cleft cyst, and acquired cystic lesions such as chronic subdural hematoma and hygroma, leptomeningeal cyst, hydatid cysts, and cystic tumors.

The septum pellucidum cyst is mostly communicating with the ventricles. It may be a cause of concern because it is found on neuroimaging in about 80 % of full-term neonates. Because septum pellucidum cyst is found by chance only in 10 % of adults, most of them disappear with time and are asymptomatic. Rarely recurrent headache and vomiting in children can be caused by a not or only insufficiently communicating septum pellucidum cyst that leads to intermittent obstruction of the CSF flow from the lateral ventricles by a ball-valve mechanism with biventricular hydrocephalus. The cavum vergae cyst is either a large septum pellucidum cyst with posterior expansion or a solitary cavum of the latter site. Cavum velum interpositum is a rostral extension of the quadrigeminal plate cistern and may point to an obstruction of CSF flow at the site of the interpeduncular and chiasmatic cistern.

The work-up examinations include CT and MRI; the former is replaced increasingly by MRI because it allows a more precise delineation and definition of the individual intracranial cyst, for instance, in Dandy-Walker malformation, with sagittal planes and T2-weighted images, and may replace the former dynamic studies of CSF flow with contrast or isotopes in arachnoid cysts by visualization of flow phenomena, for example, in the differentiation of a posterior fossa cyst from a large cisterna magna.

Depending on the type of recognized congenital intracranial cyst, further work-up examinations become necessary to exclude possible CNS or systemic malformations and genetic disorders.

Therapy, Prognosis
Surgery is indicated in symptomatic congenital intracranial cysts in which the symptoms and signs are related to the cyst and can disappear after surgery. Preventive surgery in large or enlarging asymptomatic cysts is a matter of discussion (elimination of expansive and/or obstructive effect with the aim to provide normal development of the involved adjacent brain structures). It should be considered at least in young infants in whom normalization is possible as shown in hemimacrocephaly and local bossing with thinning of the skull and in older children, in whom electrophysiological examinations and psychological testing demonstrate abnormalities.

The available treatment options are shunting (cyst or ventriculoperitoneal shunt, or double shunt), open surgery with microscopic resection of the cyst, fenestration, and establishment of a communication to the ventricles, cisterns, or subarachnoid space, and endoscopic surgery with
fenestration to the ventricles and/or cisterns. For the last procedure using stereotactic guidance or a neuronavigation system, success rates up to 70–80 % have been reported that may be possible also in long-term shunting.

Each treatment option has advantages and disadvantages. Shunting is associated with possible revisions, shunt dependency, and the insertion of a cyst tube may be tricky due to the tight cyst membrane. Open surgery may be difficult and combined with major complications, and reclosure of fenestration occurs commonly in childhood.

Probably, each case needs an indication for a specific treatment option, and each of them should be available and, if necessary, be combined in succession until precise indications are available based on large numbers and the principle of evidence-based medicine.

Prognosis depends on the group and type of congenital intracranial cyst. Except for some cases with long-lasting symptomatology, additional congenital anomalies, or acquired perinatal pathology, there is a normal development in arachnoid cysts (90 %). In Dandy-Walker malformations, the more frequently observed group with only partially agenetic vermis has a normal development in a similar percentage as in arachnoid cysts, whereas the group with dysplastic vermis or the variant forms reveals retardation in all or in a changing number, respectively. Almost all patients with porencephalic cyst have some degree of psychomotor retardation.

In a cohort of arachnoid cysts with either shunts (1/3) or open surgery, the symptomatology disappeared or improved, and the cyst was not any more visible or smaller in about three fourth of the cases, and there was a positive trend between reduction of the cyst size and outcome. In a cohort of congenital intracranial cysts with 80 % shunts, a normal psychomotor development and/or normal school placement were achieved in about 60 %, and delayed psychomotor development and/or special school requirement, or marked psychomotor development and/or inability to attend school in about 40 %. Although there was a reduction of the cyst volume in at least two thirds (most strikingly for the convexity arachnoid cysts and porencephalic cyst by 55 and 41 %, respectively), no uniform pattern of correlation with outcome could be ascertained.

### 2.6 Subacute and Chronic Subdural Hematoma (Hygroma)

#### Occurrence, Etiopathogenesis
This type of subdural hematoma # occurs mostly in the first 2 years with a peak incidence between 1 and 6 months of life and preferred in boys because the acute or repeated trauma receives no attention and/or may be accompanied by less dramatic symptoms than in the other age groups. It is less frequent than the former figures of hydrocephalus and spina bifida or as constitutional macrocrania. In the older literature, more than 50 % of the small children with chronic subdural hematoma had no history of birth or another accidental trauma. Neglected or only conservatively treated acute subdural hematoma is an important cause of chronic subdural hematoma according the newer literature.

A population-based study tells that still 53 % of serious or fatal traumatic brain injuries in the first 2 years of life are because of child abuse. Two inflicted brain injuries in 10,000 children ≤2 years of age in North Carolina and 0.13 shaken infants in 1,000 live births in Switzerland with a mortality of 16 % are the available data about head injury in battered child syndrome. These data also permit an estimation of the prevalence of chronic subdural hematoma independent of the possible causes.

#### Clinical Significance
- The cause of chronic subdural hematoma may be an inflicted traumatic brain injury, especially if a history of a reasonable injury is missing.
- Chronic subdural hematoma may lead to death if it is not recognized or too late and in up to one to two thirds to severe neuropsychological deficits depending on the cause.
- Early diagnosis and treatment of chronic subdural hematoma interrupts additional
spontaneous, inflicted, or accidental hemorrhages and improves therefore the prognosis.

Clinical Presentation
Specific symptoms and signs for chronic subdural hematoma do not exist. It is rather the history of an infant who is not doing well: anorexia, failure to thrive, intermittent low-grade fever, and resistance to be cuddled in the arms of the attending nurse. Or the combination of the following symptoms and signs that are ambiguous per se and arranged according to their frequency in a large cohort of children: convulsions, fever of different types, vomiting and hyperactive reflexes ($\geq 50\%$), restlessness, irritability, or apathy, tense, prominent anterior fontanel, anemia, and large head (about $40\%$–$30\%$). In at least one fifth of the cases, abnormal fundoscopic findings are observed; the mainly asymmetric retinal hemorrhages are characteristic of subdural hematoma as is the change of the skull shape into a squared or “box-like” form with biparietal bossing.

Periods of loss or clouding of consciousness or stupor in the history in up to two thirds of the cases point to an initial or repeated blow(s) to the head. Beyond infancy, signs of increased intracranial pressure become prominent; nevertheless, lateralizing neurological signs are often missing.

Differential Diagnosis
It includes subdural fluid collections such as those due to bacterial meningitis, and if the findings of neuroimaging are considered as well, subarachnoid fluid collections of different etiopathogenesis.

In the former situation, the large fluid collections ($>10$–$15$ ml) are yellow, clear or cloudy, bloody or purulent (subdural empyema), follow with time the same pathoanatomical course as in subdural hematoma with membrane formation, and reveal their identity by persistent fever in spite of adequate antibiotic treatment of meningitis, by irritability, and failure to thrive. External drainage or rarely open surgery is necessary to cure large and symptomatic postinfectious subdural fluid collection.

To the latter belong to the benign extracerebral fluid collections (extraventricular hydrocephalus) that may be observed in infancy beyond the neonatal period by macrocrania (head circumference $>90$th percentile with increased growth velocity) and possibly by a squared forehead. In $10$–$15\%$ of the cases, there is a motor delay of varying degree. On neuroimaging, enlargement of the anterior subarachnoid spaces that includes the interhemispheric and Sylvian fissures, possibly a mild ventricular dilatation specifically of the frontal horns, and no increased signal intensity on T1-weighted MRI is seen. Rarely, clinical signs of increased ICP (such as tense anterior fontanel, irritability, setting sun phenomenon), acute or subacute abnormal head growth, and arrest of motor development may be observed. Medical treatment (acetazolamide and furosemide) or in case of failure even temporary EVD is indicated in such patients. In general, improvement of motor delay and decreased head growth occur after 1–2 years of age and mild developmental delay in $<$10\%. Benign external hydrocephalus may be complicated by acute subdural hematoma.

Work-Ups
They include ultrasound, MRI, or CT, and subdural puncture with examination of the subdural fluid.

The latter may be performed for diagnostic and/or therapeutic reasons. Today, it is replaced by MRI and surgery, respectively. If a subdural puncture must be performed, some precautions must be taken: meticulous asepsis, puncture lateral to the edge of the anterior fontanel through the coronal suture, and fluid should be allowed to drip from the needle (no use of a syringe).

Ultrasound or more precisely MRI allows to recognize the subdural hematoma, its laterality, and site (in infancy, $80$–$85\%$ are bilateral and mostly frontoparietal and interhemispheric), its composition and stage, the width of the subdural space, a possible ventriculomegaly, and some differentiation from other fluid collections. The resolution of the subdural hematoma, the establishment of cerebral atrophy, or progressive communicating hydrocephalus (in about $10\%$) can
2.7 Encephalopathies, Metabolic Disorders with Brain Involvement

be demonstrated by postoperative radiological follow-up,

Additional examinations include the current status of development, ophthalmological and neurological findings. If a battered child syndrome is suspected, a throughout clinical and radiological search for corresponding findings and confrontation with the caregivers are necessary.

Therapy

Infants with head injury and only small acute subdural hematomas on neuroimaging need clinical and radiological follow-up because it may lead after an early stage of apparent resorption of the hematoma with widening of the subdural space to gross chronic subdural hematoma within 2–4 months.

In the past, treatment was started with repeated daily subdural taps over 2–3 weeks and more up to dryness with reported success in three fourth of the cases with chronic subdural hematoma.

When the subdural fluid gets clear and the protein level is less than 250 mg%, a bi- or unilateral subduroperitoneal shunt with a common distal part, a purpose-designed subdural catheter, and a low pressure valve is performed or alternatively beyond early infancy a subduropleural shunt with or without valve. The whole system should be removed within 6 months because resolution of the hygroma occurs in a few months and the subdural catheter becomes adherent afterward, must be left behind, and may be a source of complications.

Alternatively, burr hole, minicraniotomy, endoscopic washout, or continuous external subdural drainage is proposed. Temporary EVD has been proposed either as intermediate measure until the subdural fluid gets clear or instead of shunting (as definitive treatment in >90%). Because the time of resolution is variable in chronic subdural hygroma and the risks of infection of the EVD increase with time, a closed system and aseptic handling are necessary.

Today, craniotomy with membrane stripping is less frequently performed because there is questionable evidence that the mechanical constriction leads to brain damage #.

According to the current opinions, prognosis depends more on the cause(s) of the chronic subdural hematoma of infancy than on the treatment with good outcome in two thirds and moderate to severe disability in one third.

2.7 Encephalopathies, Metabolic Disorders with Brain Involvement

Occurrence, Etiopathogenesis

Encephalopathies of different causes such as toxic, metabolic, anoxemic, traumatic, and inflammatory may lead to increased growth of head circumference of minor degree in infants and small children.

Examples of metabolic disorders are the mucopolysaccharidoses, GM2-gangliosidoses, and glutaraciduria type I. Macrocrania develops by two mechanisms: storage of metabolites in brain and skull with increase of volume and/or brain damage with secondary atrophy and dilatation of the subdural space that becomes prone to hematoma and hygroma in minor head injury.

Clinical Significance, Clinical Presentation

- The quoted pathologies with macrocrania are infrequent disorders. Nevertheless, they may have a differential diagnostic and/or prognostic significance in the individual case. The prognostic significance increases if they are combined with hydrocephalus.

In glutaraciduria type I, an increasing head size belongs to the first signs of the disorder, the corresponding clinical (microencephalic macrocrania, possible retinal hemorrhages) and CT findings (dilatation of subdural space with hematoma and hygroma) must be differentiated from battered child syndrome, and the chronic subdural hematoma needs possibly surgical treatment. In Hurler’s disease (as one of the seven types of mucopolysaccharidoses) and Tay-Sachs’ disease (GM2-gangliosidosis), macrocrania is just one of several other signs, for example, typical features (gargoylism) and hepatomegaly in
the former and neurological signs including loss of developmental milestones in the latter. Macrocrania is caused by storage of the corresponding metabolites. In Hur"ler’s disease, the thickened leptomeninx and small subarachnoid cysts lead additionally to progressive hydrocephalus that needs shunting.

Red flags that refer to the possibility of such causes of macrocrania are the history of such encephalopathies or familiality of such metabolic disorders, arrest or loss of further neuropsychological development, facial dysmorphia, and other organ involvements.

2.8 Achondroplasia (Chondrodysplasia)

Occurrence
Achondroplasia is the most frequent bone dysplasia, is mostly due to a de novo mutation, and has an autosomal dominant inheritance.

Clinical Significance
- Disproportionate or absolute macrocrania is common.
- Macrocrania may be caused by megalencephaly, excessive growth of the calvaria, and/or hydrocephalus.
- Hydrocephalus is present in up to 50 %. It is mostly a communicating hydrocephalus with stable ventricular size and without clinical signs of increased ICP. It is caused by venous congestion (due to jugular foramen and thoracic inlet obstruction) and/or by obstruction of the basal cisterns and the fourth ventricle outlets (due to distortion of the brain stem).
- The communicating hydrocephalus may get superimposed by an intermittent and/or progressive hydrocephalus due to aqueductal stenosis (following dynamic changes of brain morphology such as tectal beaking) in up to 20 %.
- Up to 75–80 % have or develop chronic pneumopathy for different reasons.
- Up to 50 % have or develop neurological complications, among other things due to compressive cervicomedullary or multisegmented syndrome (mostly in the third and fourth decade) or cervical myelopathies (mostly in young children).

Clinical Presentation
There is a disproportionate dwarfism combined with rhizomelic, short limbs (mean body length in adulthood 125 cm). The large brachycephalic head displays a narrow cranial base and a sunken root of the nose #. In addition, a small thoracic cavity is combined with a thoracolumbar kyphoscoliosis and with crura vara.

The head circumference lies above or close to the 98th percentile or at a higher percentile than the body length. On follow-up, the head circumference parallels the normal values at a higher level. In case of progressive hydrocephalus of infancy, the slope takes an acute upward turn. In general, all symptoms are progressively worsening with age.

Differential Diagnosis, Work-Ups
The differential diagnosis includes other disorders which lead to macrocrania, particularly in infancy, to dwarfism, and to similar symptoms and signs as in achondroplasia.

Work-ups are necessary in every case right from the start and or if the child becomes symptomatic during the clinical long-term follow-up: cranial CT and MRI including the craniocervical junction, for instance, in case of suspected progressive hydrocephalus or compressive cervicomedullary syndrome. Because the ICP may be increased in moderately dilated ventricles in spite of absence of clinical signs, continuous ICP measurement allows recognition of such cases. Somatosensory evoked responses allow identification of early ongoing compressive syndromes.

Therapy, Prognosis
For the indication of treatment of hydrocephalus, there are two options: either shunting only in case of symptomatic hydrocephalus or by checking for increased ICP. Surgery is indicated in ICP >15 mmHg and/or pressure waves in symptomless cases with moderately increased ventricular size. In craniocervical and other compressive
syndromes, decompressive surgery is indicated; it must consider in the former situation the venous congestion in the region of the cranial base.

The prognosis of the individual case depends not only on the time and effectiveness of treatment of hydrocephalus but also on the general intellectual outcome of achondroplasia. Although most of the patients have at worst only a slight delay of psychomotor development and a normal IQ with mild cognitive defects without evidence of progression, up to 10–20 % display a significant delay in psychomotor development and low IQ values. In general, life expectancy is reduced in achondroplasia.

Webcodes

The following webcodes can be used on www.psurg.net for further images and data.

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Bibliography

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Walker M (2005) Looking at hydrocephalus: where are we now, where are we going? Childs Nerv Syst 21:524–527

Arnold Chiari Malformation


Shunt Surgery


Endoscopic Surgery


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Section 2.3


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Section 2.7


Section 2.8


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