12 Skull

12.1 Congenital Osteocutaneous Defect

General considerations

- Regional defect (aplasia) of skin and bones, with an area >2–3 cm^2

Therapy

- Cover the defect with skin flaps

12.2 Encephalocele

General considerations

- Congenital herniation of the brain and the meninges through an opening in the midline of the skull
  - Approximately 60% are occipital
  - Frequently with involvement of the cerebellum
  - Incidence: approximately 10% of neural tube defects
- Encephaloceles are commonly covered with hairy thin skin, which may be pigmented
- Flammeus nevus or angiomatous changes are not rare
- Perforation danger in bulging encephaloceles
- Pulsatile mass that increases in size with crying is a sign of encephalocele
- May be identified antenatally by ultrasonography
- Differential diagnosis
  - Dermoid cyst
  - Hemangioma

- Nasal polyp (diagnosis may be difficult in anterior lesions)
- Teratoma
- Meningioma
- Neurofibroma

Preoperative work-up

- Ultrasonography and MRI (regarding brain deformity) → specification of prolapsed parts of brain
- X-ray → size and location of cranial defect

Operation

- Timing depends on:
  - Condition of the covering skin
  - Pressure
- Aims of the procedure:
  - Restoration of the brain to the cranium
  - Excision of extracranial non-functioning brain tissue
  - Closure of the dura
  - Coverage with periosteal flap
  - Subcutaneous and skin closure
- Simultaneous or subsequent therapy of hydrocephalus, if necessary

Complications

- Meningitis (more common with anterior encephalocele, connection to nose or pharynx)
- Cerebrospinal fluid (CSF) fistula

Prognosis

- Mortality of 20% by 1 year
- Approximately 30% of the survivors expect normal intelligence
- Approximately 30% of the patients with an occipital encephalocele are partially or completely blind
- Convulsions in 25%

12.3 Hydrocephalus

General considerations

- Presence of an excessive amount of cerebrospinal fluid (CSF) with ventricular enlargement, usually under increased pressure, due to an imbalance between production and absorption of CSF
- CSF mainly produced in choroid plexus of the lateral ventricles, a smaller amount originates from the extracellular fluid
- Drainage via the third ventricle, aqueduct of Sylvius, the fourth ventricle, and the spinal central canal
- The fluid escapes into the subarachnoid space via the foramina of Luschka and Magendie
- Absorption through arachnoid granulations and the sagittal sinus
- Normal pressure 5–15 cmH_2O
- Overall incidence 2 per 1000 births, excluding hydrocephalus associated with myelomeningocele, 0.4–1 per 1000 births
- Differential diagnosis
  - Chronic subdural hematoma (hygroma)
  - Macrocephaly/megalocephaly
  - Atrophic encephalopathy
  - Ventricular hemorrhage

Classification

- Communicating hydrocephalus
  - Free flow of CSF between the ventricles and the subarachnoid space
  - Malfunctioning of absorption after infection or hemorrhage
- Non-communicating hydrocephalus
  - Obstruction of CSF drainage in the area of the fourth ventricle caused by:
    - Neoplasm
    - Hemorrhage
    - Infection
    - Dandy–Walker malformation

  - Arnold–Chiari malformation (myelomeningocele)
  - Aqueductal stenosis
- Choroid plexus papilloma with excessive secretion – rare

Signs

- Rising pressure, which can be compensated by the gaping fontanels and the sutures in the neonatal period
- Enlargement of the neurocranium with "sun-set" appearance (superior sclera visible above the iris) due to pressure on the soft supraorbital plates in neonates
- Signs of raised intracranial pressure in the case of closed fontanels
  - Vomiting
  - Seizures
  - Opisthotonus
  - Decreasing level of consciousness
  - Visual disturbance
  - Hypertension
  - Bradypnea
  - Bradycardia
  - Mydriasis
  - Papilledema

Preoperative work-up

- Measure the head circumference and documentation on a centile chart
- Ultrasonography, if fontanel is still open
- Skull X-ray (osteoporotic changes, splaying of suture lines)
- MRI, in special cases CT (tumors, cysts, enlargement of ventricle, hemorrhage)
- Ophthalmoscopy (papilledema)
- Measurement of CSF pressure (Fig. 12.1)
- Scintigraphy scanning for measurement of CSF absorption (rarely needed)

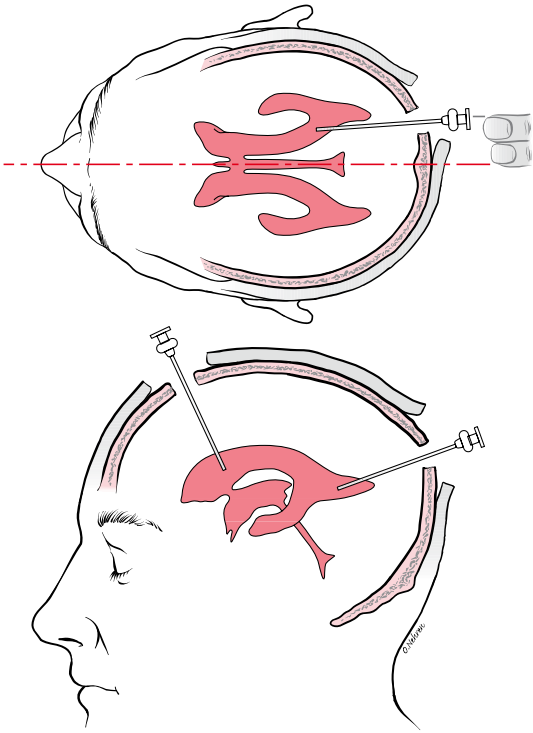

Fig. 12.1 Puncture of the ventricle system

Indications for operation

- Increasing ventricle volume with ballooning of lateral ventricle
- Increasing head circumference >97th centile
- CSF pressure >25 cmH_2O

Operation

- Like other prosthetic insertions, CSF shunts are prone to infections with common microbes from resident skin flora. The initial painting with antiseptics abolishes all vegetative organisms, but during the procedure further baceteria emanate from the pilosebaceous unit. These can infect a shunt system. A system of antisepsis has been shown to reduce infection rates with the offending *Staphylococcus epidermidis* to low levels

- The CSF is drained into the peritoneal cavity (ventriculo-peritoneal shunt = VP shunt) or occasionally the atrium of the heart is used (VA shunt)
- There are many types of shunts, e.g., Silastic slit valves, Holter, Pudenz–Heyer, Hakim-ruby-ball-valve (Codman–Hakim®)
- Ventriculostomy of the third ventricle
- Perioperative antibiotic prophylaxis to prevent wound infection
- Figure 12.2 indicates the surgical steps to treat hydrocephalus

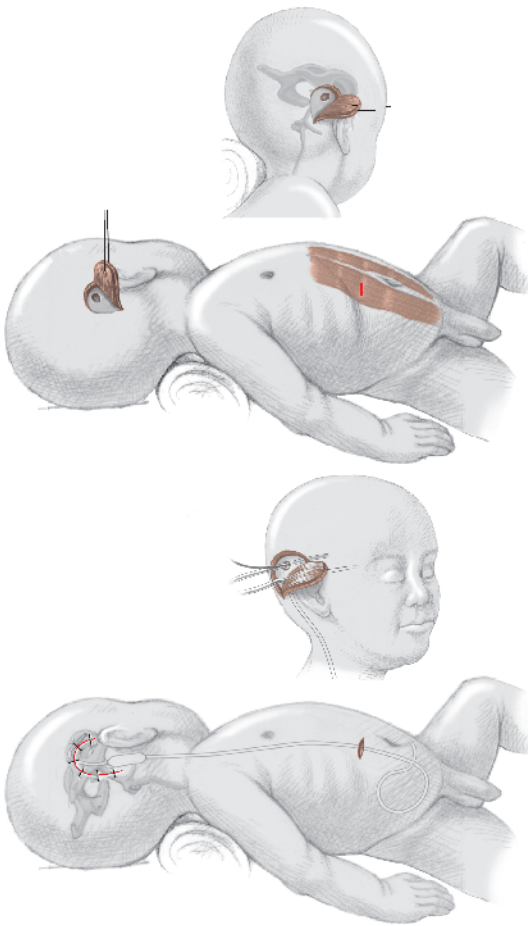

Fig. 12.2 Surgical steps in the treatment of hydrocephalus

Postoperative care

- C-reactive protein (CRP) estimation 6 weeks post surgery to pick up colonized shunts early
- Education of parents about signs of blocked/disconnected shunt, infection and over draining
- Life-time check-ups are required, but especially in the growing period

Complications

- Infection (peritonitis, ventricular sepsis), which can be reduced by taking great care during the insertion of the shunt (aseptic technique)
- Catheter blockage, disconnection, and breakage leading to signs of raised intracranial pressure
- Over-drainage (slit ventricle syndrome), subdural hematoma, craniosynostosis
- Catheter shortening due to patient growth
- Thrombosis of superior vena cava (VA shunt)
- Abdominal liquor cyst

Prognosis

- This depends on the primary situation, and is influenced by morbidity due to catheter-related complications

12.4 Craniosynostosis

General considerations

- The development of the cranium takes place along the cranial sutures, which close at different ages
- Due to abnormal and early cranial suture closure the skull becomes deformed and the intracranial pressure may rise

Classification

- Craniosynostosis may result in an increase of intracranial pressure when premature union of both coronal and sagittal sutures are involved

- Craniosynostosis should be differentiated from plagiocephaly, which is a condition in which the skull has an abnormal shape not always related to premature synostosis of the sutures but to the position of the head in utero
- Typical congenital craniofacial syndromes are Crouzon syndrome with pansynostosis and Apert syndrome with synostosis of the coronal sutures

Types

- Types of craniosynostosis are listed in Table 12.1

Table 12.1 Types of craniosynostosis

Form	Suture	Characteristics
Scaphocephaly	Sagittal	Elongated head
Brachycephaly	Coronal	Flattened anterior/posterior
Turricephaly	Coronal and sagittal	Tower shaped skull
Trigonocephaly	Frontal	Keel-shaped forehead

Signs

- Striking appearance
- Exophthalmia
- Developmental disorders

Preoperative work-up

- Clinical examination of the skull from the front, back, and always from above
- General neurological status
- X-ray a.p. and lateral
- CT
- Ultrasonography

Indications for operation

- Increase of intracranial pressure
- Cosmetic considerations

Operation

- According to the skull deformation
  - Release craniectomy or cranioplasty

Prognosis

- Very good aesthetic results if early surgery
- Good functional results depending on the severity and the time

12.5 Brain Injuries

General considerations

- Functional impairment of the brain after violent damage
- Anatomical specialties of the child's cranium
  - Open fontanels
  - Wide venous communication between the dura and the skull
  - Possibility of sub- and epidural hematoma without fracture
  - Meningeal arteries are not yet bone covered in a channel
  - Development of brain edema
- Rise of intracranial pressure due to:
  - Intracranial hemorrhage
  - Posttraumatic brain edema
- Skull fractures
  - Linear fissures
  - Non-dislocated linear fractures
  - Depressed fractures
  - Basal skull fractures
- Intracranial hemorrhage
  - Epidural hematoma
  - Subdural hematoma
  - Subarachnoid hematoma
  - Brain hemorrhage
- Always think of a battered child when you see a child with a brain injury

Classification

- Open skull injury: discharge of liquor, meninges or brain tissue
- Closed skull injury: injury of the brain and/or meninges without opening of the skull

Signs of fractures and hematomas

- Table 12.2 lists the signs of fractures and hematomas

Table 12.2 Signs of fractures and hematomas

Linear fissures	Slight signs according to degree of brain involvement
Linear fractures	Slight signs according to degree of brain involvement
Depressed fractures	Focal neurologic signs, signs of increased intracranial pressure
Basal skull fractures	Monocle hematoma, tympanum hematoma, blood and liquor, discharge out of nose, ear or throat
Epidural hematoma	Lucid interval, then increasing unconsciousness till coma, mydriasis on the side of the hematoma, contralateral paresis, positive Babinski, increased reflexes, irregular breathing
Subdural hematoma	Less dramatic than epidural hematoma with longer symptom-free interval (up to 3 days)
Subarachnoid hematoma	Acute headache, unconsciousness, coma

Evaluation

- Advanced Pediatric Life Support (APLS) assessment >11 points (Table 12.3): good prognosis
- APLS <8 points: intubation and respiration

Table 12.3 Glasgow Coma Scale (adapted as Children's Coma Scale <4 years)

Points	Check/condition
Best motor response (6 grades)	
6	Carrying out request (obeying command): the child moves spontaneously or to your request
5	Localizing response to pain: put gentle pressure on the patient's fingernail bed with a pencil then try supraorbital and sternal pressure: purposeful movements towards changing painful stimuli is a localizing response
4	Withdrawal to pain: pulls limb away from painful stimulus
3	Flexor response to pain: pressure on the nail bed causes abnormal flexion of limbs, decorticate posture
2	Extensor posturing to pain: the stimulus causes limb extension (adduction, internal rotation of shoulder, pronation of forearm), decerebrate posture
1	No response to pain
Best verbal response (5 grades) adapted for children <4 years	
5	Fixes and follows objects, is orientated to sounds, smiles, may cry but interacts
4	Crying, inappropriate fixation, irritable
3	Cries only to pain
2	Inconsolable, irritable, moans to pain
1	No response
Eye opening (4 grades)	
4	Spontaneous eye opening
3	Eye opening in response to speech: any speech, or shout, not necessarily request to open eyes
2	Eye opening in response to pain: pain to limbs as above
1	No eye opening to pain

Investigations

- Exclude other injuries
- Exclude skull fractures
- Neurological status
- Intracranial pressure signs – check-up

- Ultrasonography, if fontanel is still open
- X-ray
- CT according to clinical history and condition
- Consider repeat CT after a few hours or next day

Therapy for fractures and hematomas

- Treatment for fractures and hematomas is outlined in Table 12.4

Table 12.4 Treatment for fractures and hematomas

Cerebral concussion	Close observation, give parents head injury leaflet informing them on what they need to pay attention to or observe
Cerebral contusion	Admission and close observation
Cerebral compression	Admission and intracranial pressure prophylaxis
Linear fissures	Conservative management
Linear fractures	Usually conservative
Depressed fractures	If depression > thickness of calvarium and signs: operative
Basal skull fractures	Usually conservative
Epidural hematoma	Trepanation and operative removal
Subdural hematoma	Trepanation and early operative removal

General therapeutic concepts

Acute phase

- Chest up 15°–20°
- Normothermy
- Control of respiration; intubation and ventilation with slight hyperventilation
- Sufficient infusions to stabilize circulation and correct electrolyte imbalance
- Monitoring (ECG, pulse oximetry, blood gas analysis, blood tests)
- Treat shock with urine output monitoring
- Continuous re-check of Glasgow Coma Scale score
- Regular check of pupil status

- Temperature check (consider central hyperthermia)
- Nasogastric tube for aspiration prophylaxis and early high caloric nutrition – not with basal fracture
- Measurement of intracranial pressure if necessary
- Rehabilitation

Prognosis

- Very good for mild cerebral injuries
- Morbidity (e.g., epilepsy)

12.6 Skull Fractures

General considerations

- Of the skull fractures in childhood, 75% are linear temporoparietal fractures
- These fractures heal within 1–2 months without any residual complications
- The following must be excluded:
  - An epidural or other intracranial bleeding
  - A depression fracture
  - A leptomeningeal cyst (developing fracture)
    - When the dura ruptures and the meninges become interpositioned, and the fracture develops rather than heals

Signs

The following signs are not always present and may be signs of brain injury:

- Decreasing consciousness
- Headache
- Vomiting (may be a sign of high intracranial pressure)
- Bleeding or CFS from the ear is a sign of skull base fracture
- Liquorrhea from the nose

Investigations

- Careful clinical evaluation including Glasgow Coma Score
- Examination of liquid coming from the nose (liquor from a fronto-basilary fracture)
- Otoscopy
- Radiology and CT as indicated

Therapy

- Observation (pulse, blood pressure, pupillary reflex, etc.)
- Specific therapy needed depending on situation
- Operative therapy of depressed fracture and often of intracranial haemorrhage

Prognosis

- Simple fractures: very good
- According to severity

Prevention

- It is recommended that all females be commenced on folic acid before pregnancy which drops the risk of hydrocephalus significantly
- In some countries pregnant women are screened for α-fetoprotein in the serum and amniotic fluid. The affected fetus is then aborted with parental agreement