**Definition**

- **Epidemiology**
  Incidence is 0.2–0.5 per million. Girls are affected three times more often than boys. Half of cases occur before age 10 years.

- **Etiology, pathophysiology, pathogenesis**
  Congenital segmental and cystic widening of the common bile duct. In up to 90% of cases, the common bile duct is affected. According to the “common channel” theory, the common bile duct and pancreatic duct drain into a common abnormal orifice. This leads to partial digestion of the wall of the common bile duct by pancreatic enzymes. This in turn leads to a fibrous cystic wall without an epithelial lining. Associated with other biliary anomalies such as biliary atresia, gallbladder anomalies, congenital hepatic fibrosis, or carcinoma of the gallbladder or bile ducts. Kehrer and Todani classification.

**Table 4.4  Todani classification of choledochal cysts**

<table>
<thead>
<tr>
<th>Type</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ia</td>
<td>Cystic enlargement of the common hepatic duct</td>
</tr>
<tr>
<td>Ib</td>
<td>Focal segmental enlargement of the common hepatic duct</td>
</tr>
<tr>
<td>Ic</td>
<td>Fusiform enlargement of the common hepatic duct</td>
</tr>
<tr>
<td>II</td>
<td>Common bile duct diverticulum</td>
</tr>
<tr>
<td>III</td>
<td>Choledochocele affecting only the intraduodenal common hepatic duct</td>
</tr>
<tr>
<td>IVa</td>
<td>Multiple cystic enlargements of the intrahepatic and extrahepatic bile ducts</td>
</tr>
<tr>
<td>IVb</td>
<td>Multiple cystic enlargements of the extrahepatic bile ducts</td>
</tr>
<tr>
<td>V</td>
<td>Caroli disease (multiple cystic enlargements of the intrahepatic bile ducts with cirrhosis of the liver)</td>
</tr>
</tbody>
</table>

**Imaging Signs**

- **Ultrasound findings**
  Findings include a “second gallbladder,” a cystic structure in the porta hepatis. Cystic structure presenting without postprandial contraction. Cyst measures 2–15 cm. May contain stones or sludge. Findings of dilated intrahepatic bile ducts draining into the cyst are diagnostic.

- **CT findings**
  Not necessarily required preoperatively. Unobscured visualization. Distinguishes findings from Caroli disease.

- **MRI findings**
  For precise visualization of ductal anatomy preoperatively recommended. MR cholangiopancreatography: T2-weighted TSE with fat suppression, MIP reconstructions, HASTE or SSFSE sequences.
Cholangiography

ERCP: Risk of pancreatitis • Percutaneous transhepatic technique is used intraoperatively • Intravenous technique is no longer standard.

Nuclear medicine imaging findings

Hepatobiliary imaging • Late filling of the cyst in cholestasis • Dilation of intra-hepatic bile ducts.

Fig. 4.24  Kehr classification of choledochal cysts (from Hofmann V. Ultraschalldiagnostik in Pädiatrie und Chirurgie. Stuttgart: Thieme; 2005).
Fig. 4.25  Choledochal cyst. Doppler ultrasound scan of the upper abdomen. Subhepatic cystic mass (Z), “double gall-bladder” sign. The small arrows are indicating the common bile duct, the long arrow is pointing to the portal vein.

Fig. 4.26a, b  MR cholangiopancreatography. MR image also visualizes the cystic mass (Z) next to the gallbladder (G; a). The cyst communicates with the common hepatic duct. Intrahepatic bile duct dilation (a, b, arrows).
Clinical Aspects

- **Typical presentation**
  Recurrent abdominal pain • Vomiting • Intermittent jaundice • Palpable swelling in the right upper abdomen • Associated pancreatitis.

- **Therapeutic options**
  Conservative treatment • Where complications occur, cyst excision and ductal anastomosis are indicated.

- **Course and prognosis**
  Can resolve spontaneously.

- **Complications**
  Ascending cholangitis • Biliary cirrhosis • Rupture with biliary peritonitis • Malignant degeneration is rare • Postoperative stricture of the anastomosis, cholelithiasis, cholangitis in secondary infection.

Differential Diagnosis

- **Hematoma**
  - History of trauma
  - Coagulation disorders
  - Usually exhibit intermediate signal characteristics, not purely cystic

- **Cystic duodenal duplication**
  - Directly adjacent to duodenum
  - Biliary system appears normal
  - Often incidental finding

- **Pancreatic pseudocyst**
  - History of pancreatitis
  - Localized, walled off collections of pancreatic secretions
  - Circumscribed pancreatic necrosis

- **Mesenteric cyst**
  - Usually lies in the mid and lower abdomen
  - Directly adjacent to small bowel structures
  - Normal biliary system

- **Hepatic cyst**
  - Intrahepatic location
  - Dysontogenetic lesion
  - Normal biliary system
  - No growth tendency

- **Biloma**
  - Intrahepatic or subcapsular location
  - Iatrogenic lesion, often postoperative

- **Gallbladder hydrops**
  - Findings may include stone in the infundibulum of the gallbladder

- **Duodenal ectasia in annular pancreas**
  - Passage of food through the ectatic duodenum
**Tips and Pitfalls**

Avoid direct aspiration of the cyst due to the risk of biliary peritonitis. ● Postoperative biliary air on ultrasound must not be confused with stones.

**Selected References**


Definition

- **Epidemiology**
  - Brain tumors are the second most common tumor disorder in children and adolescents after the leukemias. Incidence is 4:100,000. 50% of these tumors are located in the posterior cranial fossa.
    - **Medulloblastoma**: Most common tumor of the posterior cranial fossa (40% of cases). Usually occurs before age 10 years. More common in boys than girls (1.5:1).
    - **Pilocytic astrocytoma**: Most common brain tumor and second most common tumor of the posterior cranial fossa in children. Usually occurs before age 20 years. Peak age is 5–9 years. No sex predilection.
    - **Ependymoma**: Third most common tumor of the posterior cranial fossa in children. Accounts for 10% of all brain tumors. Peak age is 5–6 years. A third of the affected children are under age 3 years. No sex predilection.
    - **Epidermoid cyst**: Third most common mass of the cerebellopontine angle and internal auditory canal. Rare intracranial mass.

- **Etiology, pathophysiology, pathogenesis**
  - **Medulloblastoma**: Belongs to the primitive neuroectodermal tumors (PNET). Arises from the vermis cerebelli. Tumor growth is usually round and displaces adjacent structures. Progressive growth gradually obliterates the fourth ventricle. This leads to hydrocephalus. Tumor spreads by direct extension (into the cerebellar peduncles and/or to the floor of the fourth ventricle, brainstem, spinal cord, and supratentorial region) or by metastasis via the CSF (to the supratentorial region, into the leptomeninx, and into the spinal canal). Extracranial metastases can also occur in rare cases. WHO grade IV.
  - **Pilocytic astrocytoma**: Arises from precursor cells of the astrocytes in the cerebellar hemispheres. Slow-growing, circumscribed, often cystic tumor. Metastasizes and degenerates only very rarely. Spontaneous regression can occur. Often occurs in the cerebellum. Less often involves the optic nerve, optic chiasm, hypothalamus, thalamus, basal ganglia, and cerebral hemispheres. Rarely involves the brainstem. Progressively compresses the fourth ventricle, leading to hydrocephalus.
  - **Ependymoma**: Arises from the ependyma. Presumably results from genetic defects. There are four subtypes: Cellular, papillary, clear cell, and tanycytic. Two-thirds of all lesions are infratentorial (on the floor of the fourth ventricle), one-third are supratentorial. Tumor is usually lobulated and circumscribed. It can contain cysts. Occasionally necrosis and hemorrhage are present. Calcifications occur in 50% of lesions. The tumor can expand through the lateral apertures of the fourth ventricle as far as the cerebellopontine angle and into the basal cisterns; it can expand posteriorly through the median aperture into the cisterna magna. Spinal ependymomas are very rare in children. In up to 20% of cases, the tumor metastasizes via the CSF.
  - **Epidermoid cyst**: Arises during embryogenesis from ectoderm enclosed within the neural tube. Usually outside the midline. Most often at the cerebellopontine angle. Less often in the fourth ventricle. Cyst wall consists of squamous epithelium, the contents of crystalline cholesterol, and cellular debris. Grows very slowly. Encases neurovascular structures.
Imaging Signs

- **Ultrasound findings**
  Most intracranial tumors occur after age 2 years • By then the fontanellae are no longer patent and only the temporal bone is available as an acoustic window.
  *Medulloblastoma*: Increased echogenicity • Occasionally cysts and calcifications • Obstructive hydrocephalus.
  *Pilocytic astrocytoma*: Hyperechoic solid component • Usually large anechoic cystic component • Hydrocephalus.

- **Contrast CT findings**
  *Medulloblastoma*: Solid, isodense to hyperdense mass in the roof of the fourth ventricle • Small cysts or necroses are present in 40–50% of all lesions • Calcifications are rare • Hemorrhages are very rare • Over 90% of cases involve hydrocephalus • Tumor tissue enhances homogeneously.
  *Pilocytic astrocytoma*: Mass with cystic component isodense to CSF and solid component hypodense or isodense to brain parenchyma • Often there is a halo of decreased density in the surrounding brain parenchyma (edema) • Calcifications and hemorrhages are rare • Hydrocephalus is usually present • The solid component enhances homogeneously, necrosis inhomogeneously • The cystic component enhances in only half of all tumors • Contrast agent occasionally fills the cysts.

*Fig. 7.16a, b* Medulloblastoma in a 9-year-old boy. MR images. On the T2-weighted image (a), tumor is isointense to gray matter with isolated hyperintense cysts. The post-contrast T1-weighted image (b) shows inhomogeneous enhancement of the tumor tissue.
Ependymoma: Mass on the floor of the fourth ventricle, usually isodense to brain tissue • May spread into the cerebellopontine angle and cisterna magna • Calcifications are common • Hemorrhages and cysts occasionally occur • Contrast enhancement is variable and inhomogeneous.

Epidermoid cyst: Hypodense mass (isodense to CSF) • Calcifications are present in up to 25% of cases • Rare variant: Dense epidermoid • Usually does not enhance after contrast administration.

**MRI findings**

**Medulloblastoma:** Preoperative staging • Postoperative follow-up • Hypointense to gray matter on T1-weighted images • Isointense on T2-weighted images • Hyperintense on proton density and FLAIR images • Reduced diffusion on DWI • Inhomogeneous enhancement on T1-weighted images • Leptomeningeal enhancement occurs where the tumor spreads via the meninges.

**Pilocytic astrocytoma:** T1-weighted images: Solid component is hypointense or isointense to gray matter • Cyst contents are isointense or slightly hyperintense to CSF.

T2-weighted images: Solid component is hypointense to gray matter • Cyst contents are isointense or slightly hyperintense to CSF.

FLAIR: Solid component is hyperintense • Cyst contents are hyperintense to CSF. Contrast-enhanced T1-weighted images: Markedly inhomogeneous enhancement of the solid component • Cyst walls only occasionally enhance.

*Fig. 7.17a, b* Pilocytic astrocytoma in a 2-year-old boy. MR axial FLAIR (a) and sagittal T1-weighted post-contrast (b) images. Large inhomogeneous tumor with solid and cystic components and secondary internal hydrocephalus (used with the kind permission of Dr. G. Hahn, Department of Pediatric Radiology, Institute of Diagnostic Radiology and Polyclinic, Carl Gustav Carus University Medical Center, Dresden).
**Ependymoma:** T1-weighted images: Hypointense to isointense • Calcifications and hemorrhages appear as slightly hyperintense areas • Cyst contents are hyperintense to CSF.

T2-weighted images: Isointense to hyperintense • Cystic areas appear hyperintense • Calcifications and hemorrhages appear as hypointense areas.

FLAIR: Tumor is more clearly demarcated • Cyst contents are markedly hyperintense.

T1-weighted images with contrast: Slight to moderate inhomogeneous enhancement.

**Epidermoid cyst:** T1-weighted images: Slightly hypointense to CSF • Can resemble a complex arachnoid cyst • Occasionally septated • A dense epidermoid is hyperintense.

T2-weighted images: Isointense or hyperintense to CSF.

FLAIR: Hyperintense.

Contrast-enhanced T1-weighted images: Slight or absent marginal enhancement.

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**Clinical Aspects**

**Typical presentation**

**Medulloblastoma:** Symptoms of cerebellar involvement (ataxia of the trunk and extremities, intention tremor, nystagmus) • Signs of increased intracranial pressure (vomiting, headache, sixth cranial nerve palsy) • Symptoms of local tumor spread (cranial nerve palsy, dysregulation in vital centers, deficits in long pathways).

**Pilocytic astrocytoma:** Symptoms of increased intracranial pressure and cerebellar involvement.

**Ependymoma:** Symptoms of increased intracranial pressure • Symptoms of cerebellar involvement • Occasional neck pain • Torticollis • Vision loss.

**Epidermoid cyst:** Remains clinically asymptomatic for many years • First symptoms usually appear around age 40 • Symptoms depend on the location • Headache • Cranial neuropathies (fifth, seventh, and eighth cranial nerves).

**Therapeutic options**

**Medulloblastoma:** Radical surgery is best wherever possible • Chemotherapy • Irradiation of the entire CNS (in children over age 3).

**Pilocytic astrocytoma:** Resection • Adjuvant combined radiation and chemotherapy is indicated to treat residual tumor.

**Ependymoma:** Complete tumor resection • Postoperative radiation therapy • Efficacy of chemotherapy has not been established.

**Epidermoid cyst:** Resection.

**Course and prognosis**

**Medulloblastoma:** Prognosis depends on the age of the child, the size of the residual tumor postoperatively, and evidence of distant metastases (M classification).

**Pilocytic astrocytoma:** Where total resection of the tumor is feasible, the 10-year survival rate is nearly 100%.

**Ependymoma:** In up to 20% of cases, metastases are present at the time of the diagnosis • Resectability is a decisive factor in the prognosis • Where total resection...
is possible, the survival rate is 51–80% • Where only subtotal resection is possible, it decreases to 0–26% • Prognosis in infants younger than 1 year is very poor. *Epidermoid cyst:* Prognosis is good where the cyst is completely resected.

**Complications**

*Medulloblastoma:* Hydrocephalus • Neurologic deficits • Pain • Treatment-related complications such as endocrinopathy, retarded growth, leukomalacia and encephalomalacia, microangiopathy, hearing loss including deafness, and secondary CNS malignancies.

*Pilocytic astrocytoma:* Identical to medulloblastoma.

*Ependymoma:* Identical to medulloblastoma.

*Epidermoid cyst:* Residual cyst wall left in situ after resection often leads to recurrence.

**Differential Diagnosis**

*Choroid plexus papilloma* – More common in the lateral ventricles (70% of cases)
  - Lobulated appearance
  - Highly homogeneous enhancement
  - Lesser mass effect
  - Choroidal artery dilated

*Hemangioblastoma* – Older patients
  - Nodule adjacent to the pia mater
  - Marked homogeneous enhancement of the nodule
  - Tumor lacks soft tissue component

*Brainstem gliomas* – See section on “Brainstem Gliomas”

*Atypical teratoid or rhabdoid tumor* – Younger children
  - Usually indistinguishable from medulloblastoma
  - Very heterogeneous appearance
  - Tumor appears as a cystic and solid hemorrhagic mass
  - Variable enhancement

**Tips and Pitfalls**

Examination of the spinal axis is indicated in medulloblastoma to exclude drop metastases • Pilocytic astrocytomas compress the fourth ventricle, medulloblastomas fill it • Ependymomas are far less common than medulloblastomas and pilocytic astrocytomas.

**Selected References**


