Definition

- **Epidemiology**
  Prevalence of 3:100,000 ● First presenting symptom of demyelinating disease in 12–30% of cases ● Approximately 50% incidence of bilateral visual impairment.

- **Etiology, pathophysiology, pathogenesis**
  Acute inflammation of the optic nerve (CN II) ● Autoimmune diseases (e.g., systemic lupus erythematosus, disseminated encephalomyelitis) ● Parainfectious or viral etiology (e.g., cytomegalovirus, rubella, mumps, herpes, toxoplasmosis) ● Radiation-induced (exposure of approximately 10 Gy or more).

Imaging Signs

- **Modality of choice**
  Gadolinium-enhanced MRI.

- **CT findings**
  CT often shows no abnormalities ● Possible thickening of the optic nerve ● Nerve may enhance after contrast administration.

- **MRI findings**
  Intraorbital and intracanalicular thickening of the optic nerve ● Mixed punctate and streaky enhancement after gadolinium administration (especially of the intracanalicular nerve) ● Increased T2-weighted signal intensity ● Sequences with combined fat and water suppression (SPIR FLAIR) are more sensitive for detecting optic nerve lesions.

- **Pathognomonic findings**
  Thickened optic nerve showing enhancement after gadolinium administration on T1-weighted fat-suppressed imaging.

Clinical Aspects

- **Typical presentation**
  Viral: Visual deterioration 10–14 days after underlying disease ● Central scotoma ● Afferent pupillary defect.

- **Treatment options**
  Steroid therapy ● Interferon is given for disseminated encephalomyelitis.

- **Course and prognosis**
  Unilateral optic neuritis has a good prognosis with cortisone therapy ● Visual impairment persists in up to 15% of cases, depending on the underlying disease ● Recurrence rate approximately 20%.

- **What does the clinician want to know?**
  Diagnosis ● Intracerebral foci ● Exclusion of a mass.
Thornwaldt Cyst

Definition

- **Epidemiology**
  - Most common congenital mass of the nasopharynx
  - Incidence: 4%
  - Peak occurrence at 15–60 years of age
  - Usually an incidental finding, detected in 1–5% of cranial MR examinations.

- **Etiology, pathophysiology, pathogenesis**
  - Synonym: Pharyngeal bursa
  - Benign cyst of the posterosuperior nasopharynx based on an embryogenic variant
  - Midline cyst located in the submucous plane of the posterosuperior nasopharynx
  - Usually results from inflammatory obstruction of a persistent embryonic communication between the primitive pharynx and notochord
  - Usually asymptomatic
  - Rarely undergoes abscess formation.

Imaging Signs

- **Modality of choice**
  - MRI.

- **CT findings**
  - Incidental finding
  - Cyst located in the posterior midline of the nasopharynx
  - Iso- or hyperdense to muscle
  - Small cysts are difficult to diagnose
  - As in MRI, lesion delineation is usually improved by contrast administration.

- **MRI findings**
  - Cyst shows intermediate to high T1-weighted signal intensity, depending on its protein content
  - Cyst wall occasionally shows enhancement after gadolinium administration
  - T2-weighted and STIR sequences usually show a uniformly hyperintense, benign cyst with well-defined margins.

- **Selected values**
  - Cyst from several millimeters to 3 cm in diameter
  - Chronic inflammatory changes in cysts > 2 cm.

- **Pathognomonic findings**
  - Midline cyst with high T1-weighted and T2-weighted signal intensity located in the submucous plane of the posterior pharyngeal space.

Clinical Aspects

- **Typical presentation**
  - Over 99% are clinically silent
  - Rarely causes eustachian tube compression, nasal speech, or abscess formation
  - Rare cases show chronic infection with “Thornwaldt syndrome”: Pharyngitis, halitosis, stiff neck, and occipital headache.

- **Treatment options**
  - Treatment is unnecessary in asymptomatic cases
  - Antibiotic therapy
  - Transoral excision or marsupialization for chronically infected or painful cysts.

- **Course and prognosis**
  - Cases detected incidentally do not require follow-up
  - Operative treatment is curative.
Fig. 5.1  Unenhanced T2-weighted MR image shows a Thornwaldt cyst located in the anterior midline between the bellies of the longus colli muscle in the roof of the nasopharynx. The cyst content is usually hyperintense on T2-weighted images.

Fig. 5.2  T1-weighted image after gadolinium injection (same patient as in Fig. 5.1). Here the cyst contents appear iso- to hypointense to muscle. The hyperintense ring surrounding the cyst is composed of the cyst wall and enhancing pharyngeal mucosa after gadolinium administration (arrow).
### Differential Diagnosis

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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<tbody>
<tr>
<td>Adenoid hyperplasia</td>
<td>Diffuse, usually paramedian hyperplasia of lymphatic tissue, permeated by enhancing bands</td>
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<tr>
<td>Adenoid retention cyst</td>
<td>Low T1-weighted signal intensity, often located in the lateral recess, multiple, characteristic heart- or pear-shaped configuration</td>
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<tr>
<td>(mucosal cyst)</td>
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<tr>
<td>Choanal polyp</td>
<td>Low T1-weighted signal intensity, obstructs the nasopharyngeal space from the front</td>
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<tr>
<td>Rathke cleft cyst</td>
<td>Incomplete embryonic occlusion of Rathke pouch, caudal cyst, usually located in the sphenoid bone</td>
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<tr>
<td>Cephalocele</td>
<td>May be found in the nasopharynx, but shows a definite communication with cerebral structures</td>
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### Tips and Pitfalls

Cysts with a characteristic appearance on imaging are rarely misdiagnosed.

### Selected References

- Ikushima I et al. MR Imaging of Tornwaldt’s Cysts. AJR 1999; 172: 1663–1665
Differential Diagnosis

- **Mass (e.g., optic glioma, meningioma)**
  - Circumscribed optic nerve expansion or mass, enhancing after contrast injection
- **Orbital pseudotumor**
  - Pain
  - May involve all orbital structures
- **Radiation neuropathy**
  - Rare
  - Prior history of radiotherapy

Tips and Pitfalls

Cerebral imaging should be done to exclude a demyelinating disease.

Selected References

Definition

- **Epidemiology**
  Most common (90–95%) of all branchiogenic malformations (cysts, fistulas) ● Usually clinically silent in newborns ● Often first recognized in adolescents and adults ● Initial diagnosis usually made at 20–40 years of age.

- **Etiology, pathophysiology, pathogenesis**
  Cyst in the lateral cervical triangle ● Arises from the second (or occasionally the third) branchial arch ● In the sixth week of embryonic development, the second branchial arch overgrows the third and fourth arches and the second through fourth branchial clefts ● Persistent communication results in cysts and fistulas.

Imaging Signs

- **Modality of choice**
  MRI, CT.

- **CT findings**
  Cystic mass (10–25 HU) lateral to the neurovascular sheath (up to 10 cm in diameter) ● Displaces the submandibular gland anteromedially, displaces the sternocleidomastoid muscle posterolaterally ● Often located near the mandibular angle; occasionally parapharyngeal or anterior to neurovascular sheath ● Septation and intracystic hemorrhage (density) are rare ● Only infected cysts show enhancement of the thickened wall after contrast administration.

- **MRI findings**
  T1-weighted signal intensity depends on protein and blood content (low = hypointense, high = hyperintense) ● High T2-weighted signal intensity ● Well-circumscribed, noninfiltrating mass ● Intense enhancement of the wall after gadolinium enhancement is seen only in infected cysts.

- **Pathognomonic findings**
  Nonenhancing smooth-bordered cyst located medial to the neurovascular sheath, anterior to the sternocleidomastoid muscle, and posterior to the submandibular gland.

Clinical Aspects

- **Typical presentation**
  Soft, usually asymptomatic mass in the region of the mandibular angle or lateral neck ● May become infected ● Infection characterized by pain and lymph node swelling ● Openings of sinus tracts on the skin surface are visible at birth ● These may drain mucus.

- **Treatment options**
  Complete cystectomy with adequate margins to remove any sinus tracts.

- **Course and prognosis**
  Excellent prognosis after complete resection ● Infection hampers surgical removal.
**Fig. 9.1** Infected branchial cleft cyst. Postcontrast CT. A cyst at the level of the right mandibular angle shows central low density with a thickened, enhancing wall. The sternocleidomastoid muscle has been displaced posterolaterally and the neurovascular sheath medially.

**Fig. 9.2** Unenhanced T2-weighted MR image of a branchial cleft cyst in the left submandibular region. The center of the cyst is markedly hyperintense, and the cyst wall shows intermediate signal intensity. The sternocleidomastoid muscle has been displaced posterolaterally, the neurovascular sheath medially.
**Differential Diagnosis**

*Inflammatory or malignant lymph node enlargement*
- Central enhancement after contrast administration (in the absence of central necrosis)
- Usually multiple, distributed along vessels

*Cystic hygroma*
- Usually multilocular
- Often larger and septated
- Most common in children younger than 2 years of age

*Abscess*
- Usually incites inflammatory reaction in surrounding tissue

*Hematoma*
- No enhancing wall
- Signal changes

*Thymic cyst*
- Located at a more caudal level and within the neurovascular sheath
- Cystic mass, sometimes with a spongelike appearance

*Cystic neurinoma*
- Lateral to the neurovascular sheath

**Tips and Pitfalls**

May be confused with abscess or hematoma. Differentiating feature: Relationship to neurovascular sheath.

**Selected References**

