Any types of primary and secondary tumor can arise in the orbit (table 1). Orbital processes are rare; accurate figures on their frequency are hard to obtain because of differences between the groups of patients presenting with orbital processes to different types of physicians. Orbital surgery lies in a border area between the various surgical specialties concerned with the anatomically adjacent structures.

This article concerns orbital tumors and their clinical presentation and treatment. Particular emphasis is laid on the choice of the best surgical approach. Minimally invasive approaches are now increasingly used. One of the more recently developed approaches, such as the transconjunctival (figure 1), pterional (frontotemporal) contralateral, or pterional extradural approach, can be used as alternative to a classic extracranial approach (lateral orbitotomy) or a more extensive transcranial approach (1, 2, 3). Some specialists may choose to operate by way of the neighboring paranasal sinuses (transethmoidal or transmaxillary approach).

Part of this article is devoted to the special topic of orbital meningiomas, which are classified by location (4). The treatment options in this particular area have been dramatically transformed since 2002; these lesions are increasingly being treated with stereotactic radiosurgery (5, 6, 7). Unfortunately, the overall incidence of non-infectious, non-inflammatory lesions in the orbit is so small that there are as yet no major randomized studies concerning their treatment (i.e., studies of evidence level A1 or A2). The available data are derived from comparative studies with small numbers of patients (evidence level B) and from non-comparative studies (evidence level C). The authors discuss selected papers from the literature and their own clinical experience. Infectious and inflammatory disorders, such as endocrine orbitopathy, will not be discussed.

**Symptom-oriented differential diagnosis**

Orbital tumors frequently impair vision. Visual acuity is often reduced by malignant and inflammatory/infectious processes (79%), as well as by optic nerve gliomas (67%); it is less commonly reduced by meningiomas (45%) and other benign processes (39%). The patient’s quality of life can also be substantially impaired by pain, which is a prominent feature of inflammatory/infectious disorders (96%) and meningiomas (70%).

Proptosis can be caused by nearly any type of orbital process and impairs cosmesis (table 2).
The mean duration of symptoms until diagnosis is 4.2 months for malignant tumors, 15 months for gliomas, and 28 months for meningiomas. The mean age at the initial manifestation of disease is 19 years for optic gliomas, which is considerably lower than that for neurinomas (42 years) or meningiomas (54 years) (table 3) (8).

### Topographical distribution

The intraconal space is delimited by the conus, which connects the four rectus muscles to each other. The most common intraconal processes affecting the optic nerve are optic nerve glioma, optic sheath meningioma, and lymphoma. Other intraconal processes include cavernoma, neurinoma, and metastases. The extracranial compartment takes up only a small amount of space within the orbit, surrounding the muscular conus like a tube. The most common extracranial processes are dermoid tumor (dermoid cyst) and pleomorphic adenoma of the lacrimal gland (8).

The subperiosteal compartment is defined as the potential space between the periosteum and the bony orbit. Mucocele is the most common process affecting this compartment. The bony orbit and the sphenoid wing can be the site of an osteoma, a malignant tumor, or
fibrous dysplasia, but the most common process affecting these structures is a sphenoid wing meningioma. Meningiomas are often found at the orbital apex, the superior orbital fissure, and the cavernous sinus. The intracranial periorbital dura mater heading toward the optic nerve can be infiltrated by an optic sheath meningioma or by other types of meningioma of variable size and extent. Pleomorphic adenoma and carcinoma arise in the lacrimal gland and duct system. Lymphoma and infections can affect the preseptal segment or the lids (8).

**Histology**

The most common orbital tumor is a benign, slowly-growing meningioma arising from the medial portion of the sphenoid wing, the anterior clinoid process, or the optic nerve sheath. The peak incidence of orbital meningioma is between the ages of 30 and 50. Women are more commonly affected than men. Clinically, meningiomas manifest themselves with gradual loss of visual acuity, accompanied by optic nerve atrophy or proptosis. Spheno-orbital meningiomas, i.e., tumors arising from the bone of the sphenoid wing and also involving the orbit, involve bone and possibly also muscle over a wide area and may require a correspondingly extensive resection (9, 10, 11). Tumors of this type account for only about 4% of all intracranial meningiomas. Most meningiomas grow along the medial portion of the sphenoid wing and enter the orbit from there. On the other hand, optic sheath meningiomas often grow along the optic canal into the cranial cavity (figure 2).

35% of all optic nerve tumors are optic sheath meningiomas. These lesions account for

<table>
<thead>
<tr>
<th>Common symptoms of orbital tumors (8)</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proptosis</td>
<td>92</td>
</tr>
<tr>
<td>Visual field defect, loss of visual acuity</td>
<td>74</td>
</tr>
<tr>
<td>Diplopia, strabismus</td>
<td>66</td>
</tr>
<tr>
<td>Pain</td>
<td>34</td>
</tr>
<tr>
<td>Lacrimation</td>
<td>23</td>
</tr>
<tr>
<td>Conjunctival edema</td>
<td>22</td>
</tr>
<tr>
<td>Inflammation</td>
<td>13</td>
</tr>
</tbody>
</table>

1% to 2% of all meningiomas and 1.7% of all orbital tumors. The authors' classification scheme distinguishes intraorbital (type 1) from intracanalicular (type 2) optic sheath meningiomas and from those that are both intraorbital and intracanalicular (type 3). When an intraorbital tumor (type 1) begins to affect visual acuity, it should be treated with radiation at first (5, 6, 7), and any large exophytic tumor mass should be resected. For type 2 tumors, surgical decompression of the optic canal can stave off the impending loss of vision. Type 3 tumors growing toward the intracranial compartment are best treated with extradural
decompression of the optic canal, intracranial tumor resection, and postoperative irradiation of
the intraorbital portion (4). The long-term prognosis is poor for vision, but good for patient
survival. Vascular processes are the second most common type of mass affecting the optic
nerve; capillary hemangioma accounts for most cases in children (12, 13, 14). The eyelids
are often involved as well. As long as visual acuity is unimpaired, these lesions can be
treated expectantly, as they sometimes regress spontaneously in 1 to 2 years. Cavernoma,
the most common vascular malformation in the orbit, is the second most common type of
orbital process in patients aged 31 to 40 and the third most common type in patients aged
41 to 50 (15). Orbital cavernomas account for 4% of all orbital processes and 9% to 13% of
all cranial cavernomas of the head. These intraconally situated lesions cause slowly
progressive, painless proptosis and, later, impairment of visual acuity. They rarely present
with an acute, clinically significant hemorrhage. At surgery, a cavernoma can be collapsed
by electrocoagulation but must then be completely removed because of the danger of
regrowth if it is left in place. If surgery is performed early, the patient’s vision may recover.

“Developmental venous anomalies” (DVAs) are a heterogeneous group of venous
malformations; different subtypes of DVA were previously called varices, varicoceles or
venous angiomas. Unlike intracranial DVAs, orbital DVAs can bleed, exert mass effect, and
produce proptosis that worsens with the Valsalva maneuver. DVAs should be treated if they
cause severe pain or significant cosmetic disturbance. The treatment can be endovascular,
surgical, or a combination of both (8).

Hemangiopericytoma is a rare (< 1%) vascular wall tumor of adulthood. It behaves
rather aggressively, is locally invasive, and undergoes malignant degeneration in 30% of
cases, possibly giving off distant metastases. The prognosis, however, is good as long as the
tumor is diagnosed early and completely resected.

Lymphangioma is a benign, hamartomatous, cystic tumor of childhood, sometimes
associated with facial or palatal cysts. It accounts for up to 4% of orbital processes, depending
on the case series. Unlike a capillary hemangioma, a lymphangioma does not enlarge if the
patient performs a Valsalva maneuver. These tumors grow diffusely, crossing anatomical
boundaries, and they form extensive cysts that are often the site of hemorrhage (chocolate
cysts). A lymphangioma may enlarge during an episode of respiratory infection because of
lymphoid proliferation of connective tissue. Either of these two processes can manifest
itself as acute proptosis. Acute worsening of vision or progressive proptosis are the only
indications for surgical resection or for the puncture and aspiration of large cysts.

Optic nerve glioma (pilocytic astrocytoma) usually arises in childhood, particularly in
children with neurofibromatosis (12). It accounts for 4% of all orbital tumors, 66% of all
primary tumors of the optic nerve, and 1% to 5% of all gliomas in childhood. The initial
manifestation is usually visual impairment. Small children may present with amblyopic
strabismus or nystagmus.

The treatment of optic nerve glioma depends on its location. Tumors involving the optic
chiasm and hypothalamus are usually treated with external beam irradiation or seed
implantation. The goal of treatment in such cases is the reduction, or at least stabilization,
of tumor size. Children below the age of 5 years are treated at first with chemotherapy only.
The authors favor surgical debulking of large suprasellar tumors. Intraorbital optic nerve
tumors that have grown into the cranial cavity should be resected only when the patient’s
vision is severely impaired or proptosis is severe. The optic nerve is transected behind the

| TABLE 3 |
|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|
| **Frequency of orbital processes by age (8)** | 0–10 years | 11–20 years | 21–30 years | 31–40 years | 41–50 years | 51–60 years | > 60 years |
| Capillary hemangioma | Rhabdo- | Mucocele | Meningioma | Meningioma | Lymphoma | Lymphoma |
| Rhabdo- | myosarcoma | myosarcoma | inflammation | cavernoma | Mucocele | Meningioma | Metastasis |
| Optic nerve glioma | Dermoid cyst | Inflammation | cavernoma | Mucocele | Meningioma | Lymphoma | Lymphoma |
| Rhabdo- | neurofibroma | Adenoid | cystic | carcinoma | Meningioma | Meningioma | Metastasis |
| myosarcoma | | carcinoma | | | | | |
globe, at the orbital apex, and intracranially just anterior to the optic chiasm through a combined extra- and intradural pterional approach.

The clinical course and prognosis of optic nerve glioma also depend on its location. Involvement of the optic tract or other postchiasmatic structures increases the likelihood of visual impairment and also increases mortality. If the hypothalamus is involved, the mortality rises from 5% to 50%.

Neurinoma usually arises in adulthood, neurofibroma in childhood. Neurinoma accounts for 1% to 4% of all orbital tumors. Patients under 30 years old with multiple neurofibromas are suffering from neurofibromatosis. They cause progressive exophthalmos, while the visual acuity is preserved. Because these tumors tend to develop on the sensory branches of peripheral nerves, oculomotor function also remains intact for a long time. The motility of the globe is restricted only when the tumor becomes large enough to impede its movement. These well-demarcated tumors can often be completely neurosurgically removed in their early stage, with a good prognosis. Neurofibromas rarely undergo secondary degeneration.

Fibrous dysplasia most commonly involves the bone of the orbital roof (< 1% of all orbital tumors). These lesions grow slowly, often compromising the optic canal. In symptomatic cases, the optic nerve is decompressed in its canal. Osteomas most commonly arise from bone of the paranasal sinuses (< 1% of all orbital tumors). They only rarely primarily involve the orbital bone itself. They are removed only if symptomatic.

Pleomorphic adenoma, a histologically mixed, benign tumor, is the most common benign epithelial tumor of the lacrimal gland (1% to 4%). It usually arises in middle age. It may involve bone. These lesions can be cured by total resection with preservation of the capsule. Residual tumor fragments can undergo secondary degeneration.

Rhabdomyosarcoma is the most common malignant orbital tumor of the first decade of life (1% to 3% of orbital processes). It is usually found extraconally in the superior nasal quadrant and may additionally involve the conjunctiva, uvea, and lid. Orbital rhabdomyosarcoma has the best prognosis among all locations, provided that the paranasal sinuses are not involved. The 3-year recurrence-free survival rate is 91%. For most patients, the initial manifestation is massive proptosis with inferotemporal displacement of the globe, accompanied by lid edema and conjunctival injection. Anteriorly situated rhabdomyosarcomas can be totally resected. Posteriorly situated ones are biopsied for diagnostic purposes and then treated with a combination of radiation and chemotherapy (17).

Adenoid cystic carcinoma of the lacrimal gland arises in middle age (< 1%). Infiltration of sensory nerves by the tumor causes pain. These tumors have a poor prognosis, even when they are radically resected; they tend to recur locally and to give off distant metastases (the presence of metastases worsens the prognosis). Brachytherapy can be used for local tumor control as an alternative to enucleation of the globe.

Metastases are more common with advancing age (18). In various studies, orbital metastases account for 1% to 13% of all orbital tumors. The primary tumor may be in the breast, lung, prostate, gastrointestinal tract, or kidney. Metastasis occurs by way of the bloodstream; thus, by the time an orbital metastasis becomes apparent, the cancer is often widespread, and there are not uncommonly intracranial tumor deposits as well. Diplopia and pain accompany orbital metastases more commonly than they accompany primary orbital tumors. A multiplicity of suddenly arising symptoms is typical of orbital metastases. Their treatment is palliative; above all, the creation of new deficits should be avoided.

The diagnosis of an orbital metastasis must nonetheless be confirmed by biopsy, because these lesions have an extensive differential diagnosis. Total excision should be attempted only if the lesion is small, well circumscribed, and easily accessible. Symptomatic improvement for six to twelve months can be achieved with radiotherapy. Even if the orbital tumor can be locally controlled, however, the overall prognosis with respect to systemic disease progression and survival is generally poor.

The different varieties of lymphoma are diseases of old age. Lymphoma can be either intra- or extraconal, and may arise in the conjunctiva (< 8%). Non-Hodgkin lymphoma is extranodal in 40% of cases; 5% to 14% of these affect the orbit. The symptoms include proptosis, restrictive limitation of oculomotor function, and, later, visual impairment. Non-Hodgkin lymphoma is difficult to distinguish radiologically from inflammatory syndromes or lymphatic hyperplasia. Among the MALT (mucosa-associated lymphatic tissue) syndromes, those more commonly found in the orbit (e.g., in the conjunctiva or lacrimal gland) are...
classified as extranodal marginal zone lymphomas (EMZL). These diseases have a good prognosis; once the diagnosis is confirmed by biopsy, they can be effectively treated with moderately intense radiotherapy. The most common type of orbital lymphoma, which is also the most aggressive and prognostically least favorable type, is diffuse large-cell B-cell lymphoma (DLCL). Aggressive lymphomas must be treated primarily with chemotherapy (19, 20, 21, 22). 15% to 20% of EMZL and 50% of DLCL become systemic at some point in their course; these types of lymphoma must therefore be followed up at close intervals.

Dermoid cysts are the most common type of orbital cyst (2% of all orbital processes). They are of congenital origin and become clinically evident in childhood or young adulthood. They should not be opened during surgical excision so as not to produce an inflammatory reaction. They should be completely removed, as they otherwise tend to recur. Aneurysmal bone cysts are benign destructive lesions of bone that enlarge by local extension; they are rarely found in the orbit (0.25% of orbital lesions). Their optimal treatment is currently a matter of debate. The options include radiotherapy, surgery, and embolization.

Operative approaches
The choice of approach depends on the location, size, demarcation, and histological type of the lesion (please see additional table at the end of this article). The least traumatic approach should be chosen. The procedures that can be performed range from open biopsies for confirmation of a diagnosis to the subtotal resection of diffusely infiltrating processes with preservation of function and the complete excision of well-circumscribed lesions. Fine-needle biopsies are suitable for confirmation of the diagnosis when the primary lesion is known (e.g., in metastatic solid tumors or lymphoma) but do not provide enough tissue for more thorough immunohistochemical studies. Very extensive surgery, with wide excision of bone followed by complicated reconstructive techniques, is rarely necessary. In general, open operations in the orbit are performed by one of two types of approach: transcranial or direct extracranial.

A transcranial approach is preferable for processes extending behind the orbit into the cranial cavity or located in the optic canal or the superior orbital fissure. A classic pterional approach is standard and is used by the authors for orbital apex tumors as well. If the tumor involves the posteromedial portion of the orbit, inferomedial to the optic nerve, a pterional approach from the opposite side will be needed. An exclusively extradural pterional approach can be used to decompress the optic nerve in the optic canal and superior orbital fissure, as well as for tumor biopsy or resection (2).

The supraorbital approach occupies an intermediate position between the extra- and transcranial approaches. Well-circumscribed processes superior to the optic nerve can be reached either intra- or extraconally through an eyebrow incision. The supraorbital approach is particularly useful for the removal of cavernomas and neurinomas (15, 16).

Processes located exclusively within the orbit can be reached by a wide variety of relatively noninvasive extracranial approaches, including lateral orbitotomy, the transthyroidal and transconjunctival approaches (figure 1), and the frontal trans-sinusoidal approach (1). Lateral orbitotomy is useful for well-circumscribed periorbital and intraconal tumors and for tumors located dorsally, basally, and laterally to the optic nerve, as well as for tumors of...
the lacrimal gland. The authors favor a transconjunctival approach for basal, medial intra- and extraconal tumors, and particularly for biopsies of intraconal processes. Even if the tumor is larger than the globe itself, it can still be removed by this approach without any sacrifice of bone (1).

**Recommendations**

- Any patient with visual loss, proptosis, or impaired ocular motility should undergo MRI to detect or exclude an intraorbital or intracranial process.
- Patients found to have one of these rare tumors should be referred to a specialized center for further treatment.
- Orbital surgery is a difficult and delicate undertaking that requires microsurgical technique.

**Summary**

The surgical treatment of orbital processes occupies a border area between different surgical specialties, including otorhinolaryngology, oral/maxillofacial surgery, ophthalmology, and neurosurgery. The spectrum of pathology is broad, and the potential operative approaches are numerous. A good understanding of the variety of disease processes in the orbit, and of the available interdisciplinary approaches, is needed so that the optimal mode of treatment can be determined. Collaboration across specialties improves the chances of successful treatment.

**Conflict of Interest Statement**

The authors declare that they have no conflict of interest as defined by the guidelines of the International Committee of Medical Journal Editors.

Manuscript received on 11 April 2006; final version accepted on 14 July 2006.

Translated from the original German by Ethan Taub, M.D.

**REFERENCES**


Corresponding author
PD Dr. med. Uta Schick
Neurochirurgische Klinik, Wiedau Kliniken
Universitätsklinikum Giessen und Marburg
Zu den Rehwiesen 9
D-47055 Duisburg, Germany
Uta_Schick@web.de

ADDITIONAL MATERIAL SEE NEXT PAGE
# ADDITIONAL TABLE

## Approaches to the orbit

<table>
<thead>
<tr>
<th>Approach</th>
<th>Indications</th>
<th>Contraindications</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lateral orbitotomy</td>
<td>Well-circumscribed periorbital and intraconal tumors, dorsal, basal, and lateral to the optic nerve; lacrimal gland tumors</td>
<td>Processes lying medial or mediobasal to the optic nerve; limited access to the apex</td>
<td>Good exposure, well-tolerated procedure</td>
<td>Visible surgical scar</td>
</tr>
<tr>
<td>Transethmoidal</td>
<td>Extracranial tumors medial to the optic nerve, traumatic injury of the optic canal</td>
<td>Superior, basal, and lateral tumors, intraconal processes</td>
<td>Well tolerated, usually used by ENT surgeons, no brain retraction</td>
<td>Limited exposure, approach through unsterile sinuses, more bleeding</td>
</tr>
<tr>
<td>Frontal trans sinusoidal</td>
<td>Tumors or trauma of the frontal sinus, retention cysts involving the orbit, extracranial processes extending into the frontal sinus</td>
<td>Extensive frontobasal injury, intraconal tumors</td>
<td>Minimally invasive, particularly for retention cysts</td>
<td>Visible surgical scar, risk of infection, limited indications</td>
</tr>
<tr>
<td>Transmaxillary</td>
<td>Basal lesions, whether intra- or (preferably) extraconal, contacting the maxillary sinus</td>
<td>Lesions above the optic nerve</td>
<td>Well tolerated, performed in collaboration with ENT surgeons</td>
<td>Limited exposure, hemorrhage, approach through unsterile sinus</td>
</tr>
<tr>
<td>Transconjunctival</td>
<td>Basal, medial intra- and extraconal tumors, biopsy of intraconal processes</td>
<td>Very small lesions, orbital apex tumors, lateral, superior, and extraconal tumors</td>
<td>Minimally invasive, ideal for cavernomas, excellent cosmetic result</td>
<td>Only for highly experienced surgeons (collaboration with ophthalmologists is recommended)</td>
</tr>
<tr>
<td>Combined extra- and intracranial approach</td>
<td>Well-circumscribed intra- and extraconal processes above the optic nerve</td>
<td>Processes below the optic nerve</td>
<td>Minimally invasive extradural approach with minimal manipulation of the orbital structures and brain, no limitation by the size of the lesion, excellent cosmetic result</td>
<td>Permanent hypesthesia of the forehead</td>
</tr>
<tr>
<td>Supraorbital approach, via eyebrow incision</td>
<td>Intraorbital optic nerve glioma growing into the cranial cavity, and lateral tumors of the optic nerve</td>
<td>Orbital apex tumors, tumors below the optic nerve, complex tumors infiltrating bone</td>
<td>Good general exposure</td>
<td>Traumatizing approach requiring brain medial retraction</td>
</tr>
<tr>
<td>Frontolateral, pterional</td>
<td>Ideal approach for tumors of the superior orbital fissure, optic canal, orbital apex, and intraorbital optic nerve; tumors located dorsal to the optic nerve; and lateral extra- and intraconal tumors</td>
<td>Tumors medial and basal to the optic nerve, lacrimal gland tumors</td>
<td>Broad exposure, minimal brain retraction, good access to the extra- and intradural and intraorbital compartments</td>
<td>No disadvantages</td>
</tr>
<tr>
<td>Pterional extradural</td>
<td>Ideal for decompression of the optic nerve in the optic canal, suitable for periorbital tumors and for tumors near the superior and inferior orbital fissures and the cavernous sinus</td>
<td>Processes in the anterior portion of the orbit</td>
<td>Well tolerated, no brain retraction, clear exposure</td>
<td>Detailed anatomical knowledge is essential</td>
</tr>
<tr>
<td>Pterional contralateral</td>
<td>Tumors of the medial orbital apex, aneurysms of the ophthalmic artery</td>
<td>Not suitable for other processes</td>
<td>Direct approach to medial processes</td>
<td>Difficult without navigation, injury to the olfactory n. is possible, may involve extensive brain retraction</td>
</tr>
</tbody>
</table>