Introduction

Examination of a patient with orbital disease begins with a detailed history to discern the chronicity of symptoms, past medical history including systemic medical conditions or neoplasia, and past surgical history and review any corresponding imaging. Orbital examination techniques in the adult and child will help establish differential diagnoses and direct further studies.

History

The history aids in establishing a probable diagnosis and in guiding the initial workup and therapy. Important historical elements will be discussed in the following chapters of this section.

Examination

External Examination

External examination with visual inspection is critical, assessing the position and symmetry of periocular structures, such as the brows, eyelids, canthi, surrounding soft tissues, and bony structures. Visual inspection should include observation for obvious globe deviation, including the direction. Grossly visible changes in the periocular skin and preauricular or submandibular lymph nodes and asymmetries are noted.

Pupils

All patients with suspected orbital disease should undergo a pupillary examination, to help aid in determining optic nerve function. The swinging flashlight test to determine the presence or absence of a relative afferent pupillary defect is helpful to ascertain possible compression of the optic nerve or disruption of the visual system between the optic nerve head and the apex of the orbit. Optic nerve function is further characterized by testing of visual acuity, color plates, and confrontational fields. The efferent pupillary pathway should be tested as well. Anisocoria should be recorded as worse in light (parasympa-

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Examination Techniques

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thetic defect) or in dark (sympathetic defect), and pharmacologic testing can be performed.

Tumors of the lateral orbit may impair ciliary ganglion function to produce a parasympathetic defect, whereas cavernous sinus or superior orbital fissure tumors may result in sympathetic dysfunction.

Extraocular Motility

Extraocular motility (EOM) should be tested in every patient, documenting restriction in motility as well as diplopia. EOM restriction can be documented in either a percentage from 1 to 100, with 100 being normal, or on a scale ranging from -4to +4, with 0 being normal. If a phoria or tropia is found, the cover–uncover test can be useful to help measure deviations with the aid of prisms.

Reduction of EOM can either be a restrictive process or a palsy. To differentiate restriction versus a palsy, patients may undergo forced duction testing. Classically, after a drop of topical anesthetic is placed, a cotton-tipped applicator soaked in 4% lidocaine solution is applied to the muscle away from the direction of gaze limitation for approximately 1 min. The anesthetized muscle is then grasped firmly with toothed forceps and rotated toward the direction of gaze limitation. Resistance indicates a restrictive disorder; free movement is more likely a palsy. If the patient is not amenable to such testing while awake, one can discern restrictive disease from paresis by looking for a "floating" saccade or, basically, the relative speed and comparison of the simultaneous saccades between the two eyes. Standing approximately 3-4 ft directly in front of the patient, the examiner should ask the patient to look at the examiner's nose and then quickly look at his or her finger on an outstretched arm in the four main positions: left, right, up, and down. For example, if the patient has an abduction deficit on the right from 6th nerve paresis, he or she will have a saccade that "floats" to the right, when compared to the fast adducting saccade on the left. If the abduction deficit is due to restriction, the right eye abducting saccade will be limited by a sudden stop.

Fields of single vision and double vision can be mapped using a penlight; Finoff transilluminator, aka muscle light; or a kinetic perimeter.

Eyelid Position and Function

Eyelid position is characterized by marginal reflex distances (MRD). The MRD1 represents the distance from the center of the upper eyelid margin to the corneal light reflex measured in millimeters. The MRD2 represents the distance from the center of the lower eyelid margin to the corneal light reflex. The action of the levator muscle (levator function) is measured as the extent of upper eyelid excursion from downgaze to upgaze with the brows fixated. If present, scleral show is measured from each limbus to the corresponding eyelid margin with the eye in primary position. Upper eyelid ptosis (Fig. 1.1) may imply either mechanical involvement of the levator muscle or palsy, whereas eyelid retraction (Fig. 1.2) suggests proptosis, such as thyroid eye disease or CNS disorder. The upper eyelid may be everted to inspect the palpebral lobe of the lacrimal gland (Fig. 1.3) or by having the patient look down and in and lifting up the upper eyelid. An s-shaped deformity characterized by ptosis and edema laterally is usually associated with



Fig. 1.1 Right upper eyelid with ptosis. Note the right brow is also elevated due to the patient's use of the frontalis muscle in an attempt to lift the ptotic right upper eyelid. The left upper eyelid is also pseudo-retracted and would likely descend to a more normal position with ptosis correction on the right



Fig. 1.2 Bilateral upper and lower eyelid retraction, left greater than right from thyroid eye disease



Fig. 1.3 Prominent palpebral lobe of lacrimal gland, visible beneath the upper eyelid

lagophthalmos should also be evaluated as part of the cranial nerve exam detailed below.

Globe Position

Proptosis

By evaluating the patient in the submental view (chin-up position), the examiner can qualitatively look for globe protrusion or retrusion relative to the canthal angle and the nasion (Fig. 1.5). To quantify the degree, three common exophthal-mometry tools exist: the Hertel, which is most commonly used (Fig. 1.6); the Naugle, which is useful for patients with abnormal lateral orbital rims (Fig. 1.7); and the Luedde, which is more feasible to use in children (Fig. 1.8).

The Hertel exophthalmometer quantifies the anterior protrusion of the eye by measuring the distance in millimeters from the anterior lateral orbital rim to the front surface of the cornea. The reading is taken with a base measurement of the separation of the positioning arms of the tool to help reference subsequent measurements on the same device. The Naugle exophthalmometer measures anterior globe position relative to the superior and inferior orbital rims. This method provides a more accurate assessment in those with lateral rim fractures, iatrogenic repositioning of the lateral rim, or orbital rim defects. The Luedde exophthalmometer measures globe protrusion unilaterally from the lateral orbital rim.



Fig. 1.4 Salmon-colored lymphoma in the inferior fornix

lacrimal gland enlargement. Lymphoma can result in a salmon-colored conjunctival mass that is visible upon inspection of the fornix (Fig. 1.4). Orbicularis strength, Bell's phenomenon, and



Fig. 1.5 Submental view of proptotic globes from Graves' disease (**a**). Child with left proptosis from orbital dermoid (**b**)

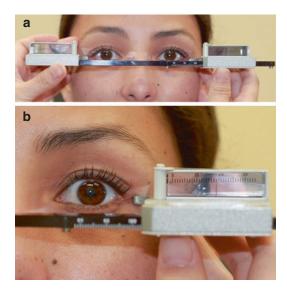


Fig. 1.6 Hertel exophthalmometer. While resting the Hertel instrument on both lateral rims, the base number is recorded on the ruler for consistency (**a**), and the amount of exophthalmos is measured by aligning the red bars then recording the number at which one sees the anterior surface of the cornea (**b**). The examiner and patient should be at eye level

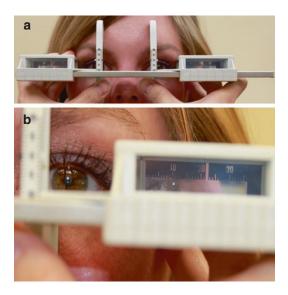


Fig. 1.7 Naugle exophthalmometer. In patients with lateral orbital rim defects, the Naugle can be used by resting the posts on the forehead and the maxillary prominence at the pupillary axis (a), aligning the red mark with the clear bar, and then recording the number at the anterior surface of the cornea (b)



Fig. 1.8 In children, the clear Luedde ruler is placed at the lateral orbital rim, and the distance to the anterior corneal surface is measured

It consists of a clear bar with millimeter markers. The anterior corneal surface can be visualized through the bar to determine the millimeters of protrusion. This can be positioned on the lateral orbital rim without a device in front of the eyes and is easier to use in children who reflexively more away and close their eyes with the other tools.

Hyperglobus or Hypoglobus

Orbital or periorbital neoplasms often displace the globe. Nonneoplastic conditions such as thyroid eye disease, trauma, and silent sinus syndrome may cause similar examination findings, and further imaging studies, such as computed tomography (CT) or magnetic resonance imaging (MRI), may be indicated.

Horizontal and vertical globe displacements are measured in millimeters from the central pupil to vertical midline and horizontal canthal line, respectively. For vertical displacement, one can draw an imaginary line horizontally across a patient's pupillary axis and determine if the pupil of the other eye is higher or lower, which could suggest hyperglobus (Fig. 1.9) or hypoglobus (Fig. 1.10), respectively. Care must be taken to ensure the patient's head is in primary position, without any tilt, and that the line is parallel to the ground.

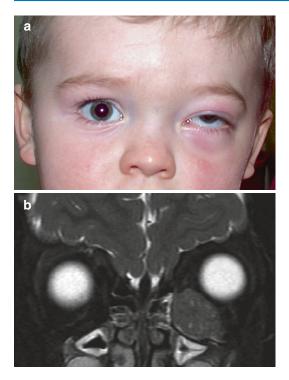


Fig. 1.9 A 23-month-old boy with left hyperglobus from desmoplastic small round cell tumor/round cell sarcoma, grade 3/3. Clinical appearance (**a**) and coronal MRI (**b**)

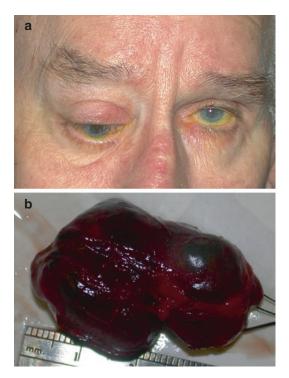


Fig. 1.10 Right hypoglobus from large cavernous hemangioma. Clinical appearance (**a**) and gross resected specimen (**b**)

Palpation

The examiner should palpate any abnormal areas for tenderness or a mass, assess the degree of resistance to retropulsion of each globe, and check for local adenopathy. The lacrimal gland area should be palpated for fullness and tenderness. Sensation to evaluate sensory nerve function is evaluated with tactile stimulation by touch. Areas of reduced sensation or hypesthesia are noted (see below CN V).

Resistance to Globe Retropulsion

The examiner places both forefingers over the anterior portion of the globe with the eyelids closed and gently pushes posteriorly on the globe. The degree of resistance is recorded on a relative scale. Orbital mass lesions often produce increased resistance to manual globe retrodisplacement.

Slit Lamp Examination

The slit lamp examination typically focuses on the corneal surface and the posterior pole in patients with a suspected orbital neoplasm. The corneal surface is evaluated for signs of exposure, and the posterior pole is evaluated for signs of ocular or optic nerve compression or congestion.

Fundus Examination

Orbital mass lesions may result in choroidal folds, optic disc edema, pallor, or shunt vessels (Fig. 1.11).

Cranial Nerves V and VII

Sensation to light touch in each dermatome of the trigeminal nerve, V1–V3, V2 may be tested using a tissue or wisp of cotton, including testing of the corneal blink reflex. Each motor branch of the facial nerve is also evaluated. Loss of muscle function may be graded on a relative scale comparing the weak side to the normal side. Bell's

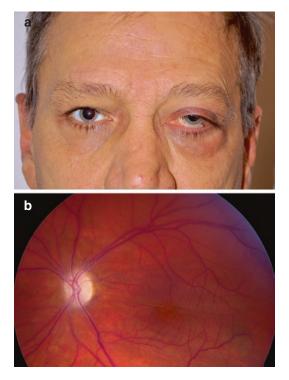


Fig. 1.11 Orbital mantle cell lymphoma. Clinical appearance with hyperglobus on the left (**a**). Note optic atrophy and choroidal folds (**b**)

phenomenon testing is performed in all patients with weak facial nerve function or lagophthalmos by asking the patient to squeeze his or her eyes shut, while the examiner tries to open them to evaluate if the eye supraducts sufficiently for corneal protection.

Lacrimal System

Attention should be directed to the superotemporal orbit to evaluate for fullness or tenderness of the lacrimal glands. The lacrimal secretory function can be measured using Schirmer's testing. This can be performed typically by placing a small strip of filter paper in the lateral conjunctival sac of bilateral lower eyelids for 5 min with the eyes closed. Basal tear secretion can be tested after placing topical anesthetic to prevent tearing from irritation. Normal wetting is 15 mm or more, mild dryness 9–14 mm, moderate 4–8, and severe less than 4 mm.

A tumor or malignancy may also involve the lacrimal drainage system and present as tearing. The excretory drainage patency is determined by irrigation of the lacrimal system with a cannula. Even in the absence of a tumor, lacrimal outflow obstruction alone can cause enlargement of the lacrimal sac and fullness in the medial canthal region. This more common benign lacrimal pathology usually begins below the medial canthus. If there is a lesion above the medial canthal tendon or presence of bloody tears, an imaging study evaluating for neoplasm should be performed. If there is a dacryocystitis, dacryocystorhinostomy (DCR) is usually indicated. Abnormal mucosa noted at the time of dacryocystorhinostomy is biopsied. If lymphoma is suspected, fresh tissue is sent to pathology for flow cytometry and lymphoma evaluation is performed.

Nasal Endoscopy

Intranasal examination using an endoscope can detect intranasal disease causing secondary orbital or lacrimal signs.

Special Issues in Examination of Children

The examination of the child with orbital pathology requires more creativity and adjustments depending on age of the patient and cooperation. Asking the parent to hold or feed an infant often facilitates the physical examination. Usage of small toys to attract the attention of the child is often critical in evaluating ductions and versions. Through observation alone, the evaluator may gather important information regarding eyelid and globe position, external periocular soft tissue changes, ocular motility, and vision. The examination should also include observation of any changes of globe position with crying, which may indicate a vascular malformation.

Patient cooperation, however, may limit the ability to perform a complete physical examination in the office. Thus, some children require sedation or general anesthesia to complete the physical examination. Communication with the pediatrician regarding suspected etiology helps to determine the need for additional systemic evaluation. Systemic workup may include serologic testing, genetic studies, or imaging studies.

Complete Eye Examination

Orbital tumors can affect sensory visual function by producing compressive or glaucomatous optic neuropathy, refractive errors, or keratopathy. Any cause of visual dysfunction in the pediatric group may produce amblyopia. Detailed visual assessment can help localize an orbital tumor and determine whether amblyopia needs to be acutely addressed. In children, assessment requires a cycloplegic retinoscopy and refraction. Eyelid position and pupillary testing should be evaluated prior to placing drops for dilation. Versions, ductions, and strabismus measurements should be noted.

In older children, color plates and visual fields may help to better characterize optic nerve function, especially if the examiner is considering an underlying glioma. In younger children, measurement of visual evoked potential (VEP or VER) may be helpful in assessing optic nerve function. This test is one of many tools used to monitor optic nerve compression in fibrous dysplasia. Evaluation of stereopsis may help distinguish a long-standing tropia from strabismus due to a new orbital process. Comparison with old photos and history from the parents can be utilized. A standard or portable slit lamp allows for the most detailed anterior segment evaluation. However, a penlight with or without a 20D lens for magnification may be used. Conditions such as lymphangioma, neurofibromatosis, or capillary hemangioma may present with anterior segment findings. Posterior pole examination follows and may reveal findings such as choroidal folds due to an orbital mass effect, optic disc pallor due to a glioma or other tumor compression, or orbital invasion from a primary intraocular tumor.

Orbital Examination

Globe Displacement

The examiner assesses globe position qualitatively with the child in the chin-up position. Although an exophthalmometer may provide an objective measure, patient cooperation may limit its accuracy. The Luedde device is particularly valuable for evaluation of globe position in children, who often find it less intimidating because it is smaller and placed on the side (Fig. 1.8). The Luedde instrument offers accurate measurements with the patient in the supine position and can be used during an examination under anesthesia.

Summary

Each step of the examination aids in disease localization and characterization to ultimately help formulate a treatment plan.