

# **MAJOR REVIEW**

# Diagnosis and Management of Enophthalmos

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**Abstract.** Enophthalmos is a relatively frequent and misdiagnosed clinical sign in orbital diseases. The knowledge of the different etiologies of enophthalmos and its adequate management are important, because in some cases, it could be the first sign revealing a life-threatening disease. This article provides a comprehensive review of the pathophysiology, evaluation, and management of enophthalmos. The main etiologies, such as trauma, chronic maxillary atelectasis (silent sinus syndrome), breast cancer metastasis, and orbital varix, will be discussed. Its objective is to enable the reader to recognize, assess, and treat the spectrum of disorders causing enophthalmos. (**Surv Ophthalmol 52**:457–473, 2007. © 2007 Elsevier Inc. All rights reserved.)

**Key words.** breast cancer metastasis • enophthalmos • fat atrophy • orbital fracture • pseudoenophthalmos • scleroderma • silent sinus syndrome • trauma • varix

# I. Diagnosis of Enophthalmos

#### A. DEFINITION

Enophthalmos is a posterior displacement of the eyeball within the orbit in an antero-posterior plane due to several etiologies.<sup>33</sup> The volume of the globe is normal. In case of unilaterality, a difference of more than 2 mm between the two eyes can be considered diagnostic. It is the opposite of exophthalmos (proptosis) where the globe is pushed forward.

# **B. CLINICAL PRESENTATION**

# 1. Symptoms

Subjective complaints depend strongly on the etiology and severity of enophthalmos. The most common disturbances are facial asymmetry and double vision. Sometimes, the patient may consider the disorder as a ptosis or contralateral proptosis.

#### 2. Clinical Examinaton

Enophthalmos is often obvious during the inspection of a patient's face. The diagnosis is simplified in cases of unilaterality or major asymmetry. Indirect clinical signs contribute to the diagnosis of enophthalmos and include deep superior sulcus, narrowing of the palpebral fissure (pseudoptosis), and lagophthalmos.

The position of the globe in the orbit has a high variability due to age, sex, and ethnic background. The best position for the clinical recognition of enophthalmos is asking the patient to look up with the head tilted back, and the observer being in front of the patient (Fig. 1). Objective and quantitative measurement can be achieved by Hertel exophthalmometry. In case of orbital fractures with displacement of the lateral orbital rim, other devices using a frontal support are necessary (e.g., Naugle exophthalmometer). Concomitant vertical misalignement (hypoglobus) is often present.



Fig. 1. Left enophthalmos.

#### C. RADIOLOGICAL IMAGING

Radiological investigations, computed tomography (CT) scan and magnetic resonance imaging (MRI), confirm and also quantify enophthalmos. Axial sections in the neuro-ocular plane provide reproducible measurements and can be used for follow-up comparison. <sup>22,170</sup> Coronal and sagittal sections are equally important for the analysis of the surrounding tissues and sinuses. The CT scan serves as the reference for the analysis of the bony structures (orbital container), whereas the MRI is more relevant for the observation of the globe surrounding soft tissues (orbital content).

# D. PSEUDOENOPHTHALMOS

The definition of true enophthalmos has been described in previous sections; therefore, it is important to distinguish disorders that may initially appear as enophthalmos, due to lid malpositions, globe size anomalies, or structural deviations (Table 1), but are not associated with an actual axial displacement of the globe.

### 1. Globe

## a. Phthisis Bulbi

Phthisis bulbi is defined as a shrinking of the globe often following injury, surgery, infection, or

TABLE 1
Etiologies of Pseudoenophthalmos

Globe	Phthisis bulbi
	Microphthalmos, microcornea
	Refractive-Anisometropia
Altered lid position	Horner's syndrome
_	Ptosis
	Contralateral lid retraction
Structural lesions	Post Enucleation Socket Syndrome
	(PESS)/Anophthalmic socket
	Contralateral exophthalmos
	Facial/Bony asymmetry

disease. Due to the reduced volume, the eye will appear sunken in to the orbit and the lids will seem ptotic without actual axial displacement of the globe in relation to its surrounding structures (Fig. 2A).<sup>3</sup>

#### b. Microphthalmos, Microcornea

Microphthalmos is defined as a congenitally small eye with reduction of the volume of the globe in the absence of other ocular anomalies. 42,49 On the basis of the small corneal diameters the diagnosis is obvious and seldom missed, even in young infants. As described for phthisis bulbi, the volume reduction of the globe or anterior segment will make the eye appear enophthalmic. Microphthalmos could be part of hemifacial microsomia (Fig. 2B).

# c. Refractive-Anisometropia

In case of significant anisometropia the shorter eye may lead to the wrong impression of being enophthalmic. It should be noted that the general rule that 3 diopters translated into 1 mm of biometric axial length may sometimes be misleading, given the widespread use of refractive surgery.

#### 2. Altered Lid Position

### a. Horner Syndrome

The syndrome named after Johann Friedrich Horner<sup>76</sup> has generally been described with miosis, ptosis, and enophthalmos, as well as anhidrosis. A lesion at any point along the oculosympathetic pathway will result in this syndrome with symptoms on the same side and anisocoria.<sup>86</sup> Anisocoria is more apparent in dim illumination, and the affected pupil shows dilation lag. Light and near pupillary reactions are intact. The eyelid is ptotic because of paresis of the sympathetically innervated Müller's muscle. There seems to be apparent enophthalmos (pseudoenophthalmos) due to the ptosis and because the lower eyelid may be elevated; exophthalmometry readings, however, are generally equal (Fig. 2C). <sup>108,126,175</sup>

### b. Ptosis

Ptosis is defined as a drooping of one or both eyelids. It may be complete or incomplete, varying in degree of severity. As described in the Horner syndrome section, a blepharoptosis can lead to the impression of an enophthalmos (Fig. 2D).<sup>8</sup>

#### 3. Structural Lesions

#### a. Post-Enucleation Socket Syndrome (PESS)

Marked pseudoenophthalmos frequently occurs after enucleation with or without the use of intra-

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