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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.[33] 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.

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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.[22,170] 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 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).[3]

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

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[76] 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.[86] 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).[108,126,175]

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).[8]

3. Structural Lesions

a. Post-Enucleation Socket Syndrome (PESS)

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

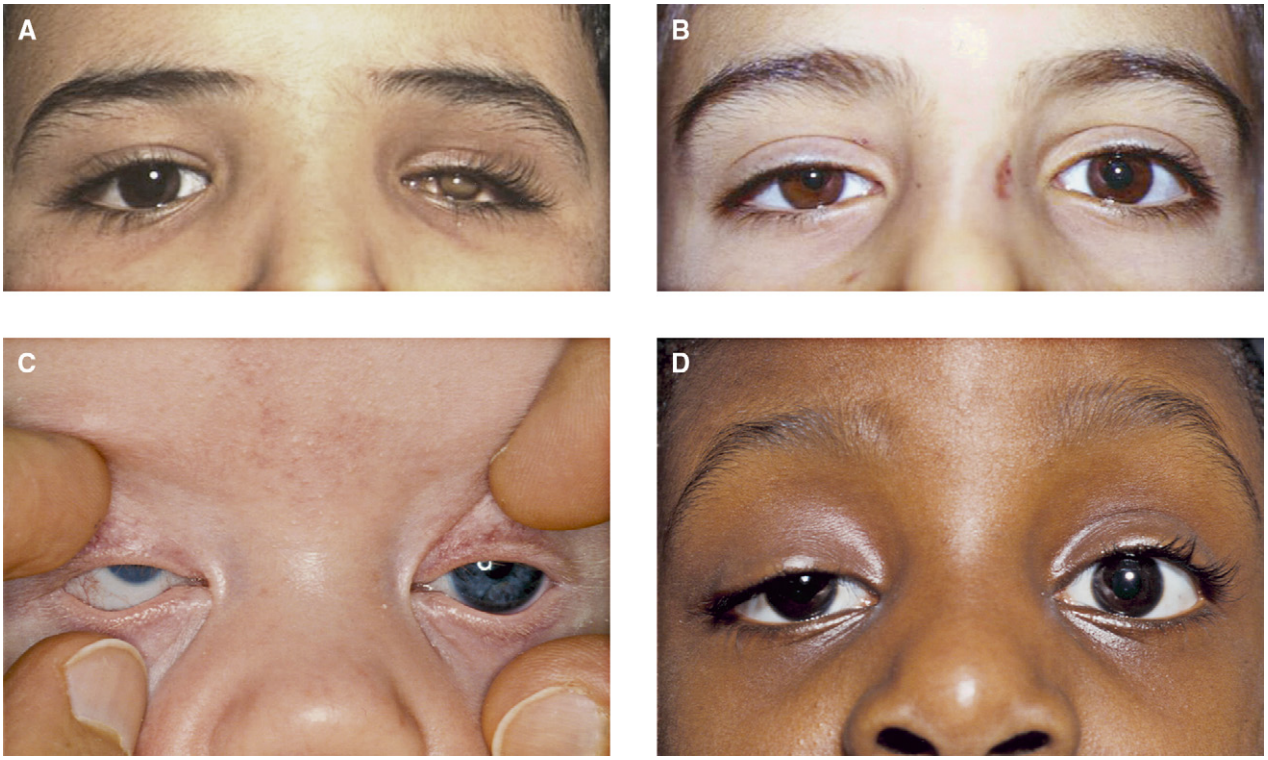


Fig. 2. A: Left pseudoenophthalmos: Phthisis Bulbi. *B:* Right pseudoenophthalmos: Microphthalmos. *C:* Right pseudoenophthalmos: Horner's syndrome. *D:* Right pseudoenophthalmos: Congenital Ptosis.

orbital implants (Fig. 3). It is often associated with a superior sulcus syndrome, which is another common finding in the anophthalmic socket. The causes are reduction of orbital content, and progressive relaxation of the lower eyelid leading to downward passage of the prosthesis, with associated ectropion development and decreased volume of the orbit. The main theory (based on clinical impression) of orbital fat atrophy due to metabolic or circulatory alterations was never proven by clinical or experimental studies.[97,98] To prevent PESS, careful evaluation of an adequate-sized implant and correct placement during surgery are important. Nevertheless, if the syndrome becomes apparent, relaxation of the lower eyelid can be corrected with surgical repair and if necessary support using, for example, autogenous fascia lata.[132] Decreased volume of the orbital content can be corrected also with materials such as autogenous fat[79] or dermis-fat,[69,157,162] autogenous cartilage,[36] autogenous bone, sclera and liquid collagen,[160] silicone,[123,155,167] glass beads,[161] or porous polyethylene,[18,66,147] using several surgical approaches and techniques (intraorbital, subperiostal).

b. Contralateral Proptosis/Exophthalmos

Exophthalmos is defined as a forward displacement of the normal globe in relation to its bony orbit. This may lead to the impression of an enophthalmic condition on the contralateral side. Therefore, any difference in the position of the eyes has to be carefully evaluated for being pseudoexophthalmic or pseudoenophthalmic in respect to its side.

c. Facial Asymmetry

Any bony malformation in the skull resulting in a facial asymmetry can lead to the impression of an enophthalmos due to the asymmetry of the face with anterior or posterior displacement of the whole orbit. The globe, however, might well be in a physiological position in regard to the orbital surroundings.

II. Pathophysiology of Enophthalmos

Three main mechanisms are proposed in the genesis of enophthalmos: enlargement of the orbital container, reduction of the orbital content, and contraction of the orbital content (Table 2).[143]

A. ENLARGEMENT OF THE ORBITAL CONTAINER

Enlargement of the orbital container seems to be the most frequent cause of enophthalmos. Different mechanisms may modify the orbital walls and hence increase the orbital volume. It could be a defect of the orbital wall(s) or an external displacement of these walls. Orbital fractures, chronic maxillary

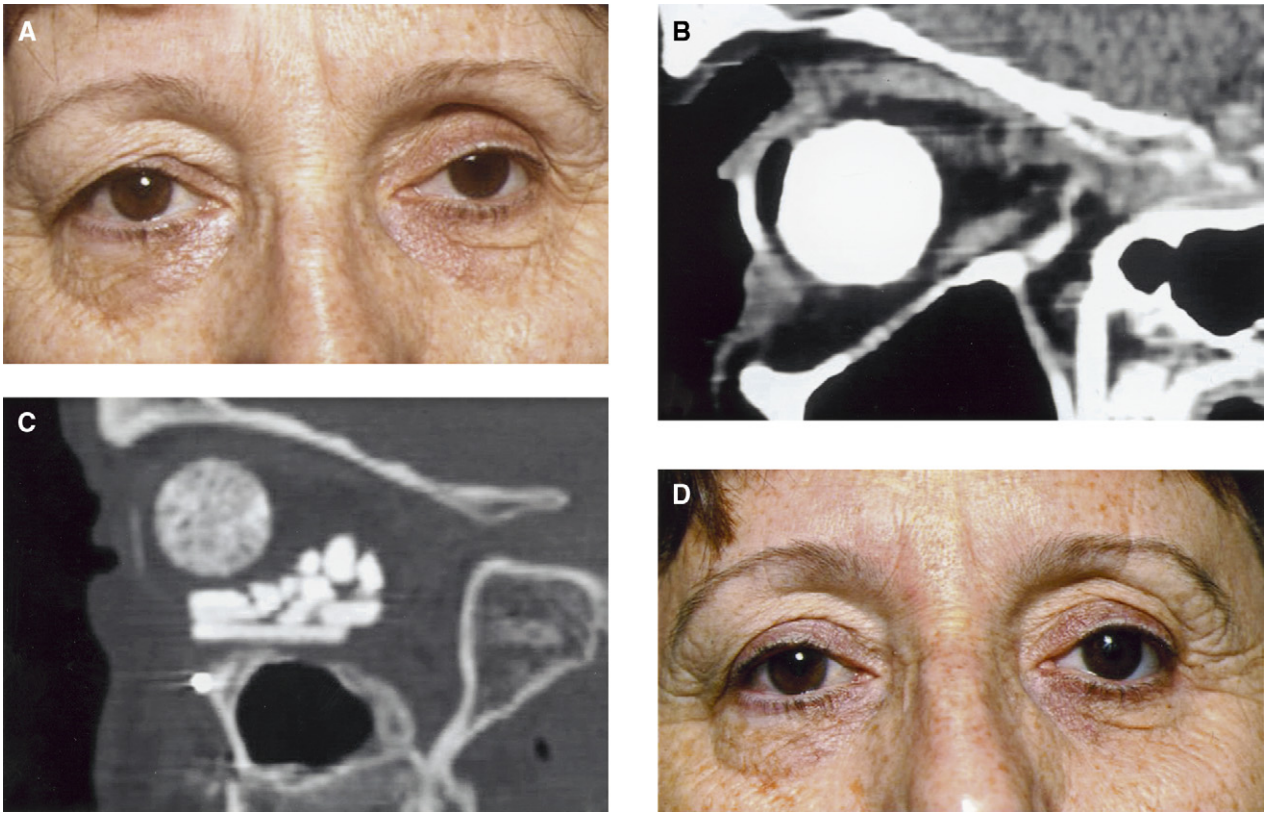


Fig. 3. *A:* Left pseudoenophthalmos: Post Enucleation Socket Syndrome (PESS). *B:* CT-Scan Sagittal view showing the posterior and inferior displacement of the implant. *C:* CT-Scan Sagittal view showing surgical correction by subperiostal orbital grafts. *D:* Postoperative result after six months.

atelectasis, and agenesis of sphenoid wing in Recklinghausen disease are the main diagnoses to consider.

B. REDUCTION OF THE ORBITAL CONTENT

Among the orbital contents, fat and muscle make up most of the volume. Vessels and nerves do not use enough space that their shrinking could be responsible for enophthalmos. Fat atrophy can be age-related, or secondary to orbital varices and radiotherapy. Lipodystrophy may also be part of systemic disease or secondary to medical treatment. Scleroderma and hemifacial atrophy are two rare causes of orbital atrophy.

TABLE 2

Pathophysiology of Enophthalmos

Enlargement of orbital container	Trauma
	Chronic maxillary atelectasis
	Agenesis of sphenoid wing
	Orbital varix
Reduction of orbital content	Age-related fat atrophy
	Orbital varix
	Radiotherapy
	Lipodystrophy
	Linear scleroderma
	Hemifacial atrophy
	Trauma/Surgery
Contraction of orbital content	Breast cancer metastasis
	Trauma/Fibrosis

C. CONTRACTION OF THE ORBITAL CONTENT

Some orbital diseases may induce a posterior displacement of the eyeball by developing an intraconal cellular infiltrate with the potential for contraction. Alternatively, fibrosis and scar formation may lead to contraction with consecutive retraction of the globe. Orbital metastases, particularly breast cancer and post-radiotherapy scarring represent the main etiologies. Congenital fibrosis should be excluded.[64,65,71]

III. Etiologies of Enophthalmos and Management

A. POST-TRAUMATIC ENOPHTHALMOS

The most frequent cause of enophthalmos is the fracture of the orbital floor. The enlargement of the

orbital container may also be secondary to medial orbital wall fracture, which is very often misdiagnosed at the early stage of trauma.[40] Lateral wall and orbital roof fractures are less frequent and seldom associated with enophthalmos. The enlargement of the orbit towards the periorbital sinuses is explained by two main theories. The hydraulic theory postulates that an increased hydraulic force in the orbit caused by posterior eyeball displacement, suddenly increases the intraorbital pressure leading to the rupture of the medial and/or inferior wall. The buckling theory involves a direct trauma to the inferior orbital rim causing mainly the displacement of the floor along the infraorbital channel. The traumatic conditions include sports trauma, vehicular injuries, fighting, and, rarely, orbital or endonasal surgeries.

Enophthalmos could be present immediately after trauma, or appear later after the reduction of orbital hemorrhage and edema. The early detectable enophthalmos is associated with severe orbital contusion. In these cases, periorbital hematoma, edema, subcutaneous emphysema, orbital pain, diplopia, lacrimal system injuries, oculomotor and pupillary dysfunction, epistaxis, and dysesthesia in the V2 distribution are often seen. Larger fractures result in smaller ocular motility dysfunction, due to the fact that the muscle is not strangulated. Eyeball trauma and other facial fractures have to be ruled out by careful examination.

Enophthalmos may be missed during the early stages of trauma, and may only be detected weeks or months after the initial trauma. In these cases, motility disorders are often permanent due to the fibrosis. The early CT scan may predict the risk of late enophthalmos according to the extent of the fracture.[183]

The amount of enophthalmos is assessed with Hertel exophthalmometry. If the fracture extends to the lateral orbital rim, the Naugle exophthalmometer may be useful.[151] The Hess-Lancaster test confirms the diagnosis of oculomotor disorders and facilitates the follow-up of the diplopia.

Orbital CT scan (Fig. 4) with axial, coronal, and sagittal sections provides the necessary information about the location and the extent of the fracture(s), the involvement of soft tissues (extraocular muscles,

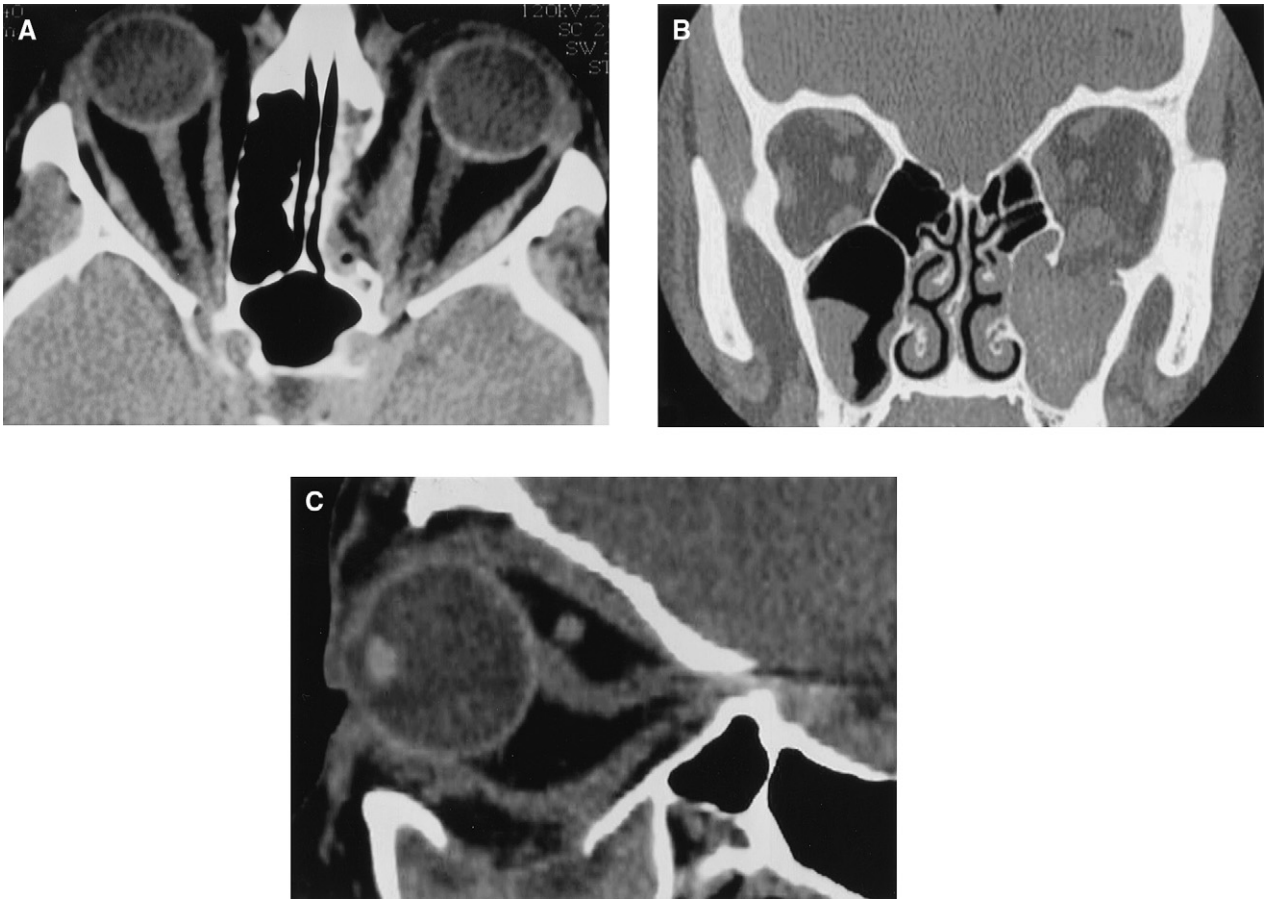


Fig. 4. *A:* CT scan, axial section, left medial wall fracture. *B:* CT scan, coronal section, left floor fracture. *C:* CT scan, sagittal section, orbital floor fracture.

orbital fat),[9,67,134,135] and the risk of late enophthalmos.[46,47,118,153] Orbital volumetric analysis and digital reconstructions may be useful for the improvement of the predictive diagnosis of late enophthalmos and the quality of surgical reconstruction.[24,25,41,48]

Standard radiographies should be avoided and MRI does not provide any further information for the diagnosis. MRI may contribute to the diagnosis of muscular fibrosis, however, in cases of persistent diplopia after fracture reconstruction or muscular restriction in enophthalmic sequelar forms.

The cosmetic demand with or without diplopia is the main indication for surgery in enophthalmic patients. As the enophthalmos appears in cases of larger fractures, there is rarely muscular suffering.The optimal timing for surgery is 1–2 week(s) following the trauma. This period allows for the resorption of orbital edema and hemorrhage.

The correction of enophthalmos is based on two main steps: first, reintegration of the herniated orbital content after a careful dissection by appropriate approach, and, second, reconstruction of the orbital shape and volume.

The transconjunctival approach[14,119] is used in small fractures of the orbital floor, whereas a coronal incision helps for complex orbitozygomatic fractures especially at late stage. In case of orbital floor fracture, the subciliary incision 2 mm below the lid margin, or a palpebral incision in the lid crease, provide a good exposure. The transconjunctival approach with lateral cantholysis permits the same exposure with limited cutaneous scar. The transcaruncular approach[13,53] is often useful for the management of the medial wall fracture with excellent exposure, without any cutaneous scar. In orbitozygomatic fractures, different approaches can be combined: subciliary, palpebral, and transvestibular incisions.[96]

The endoscopic-assisted transconjunctival approach for medial wall fracture provides improved visualization of the fracture site, facilitating bony reduction and the placement of implant.[11,30,121] The endoscopic transnasal approach has been described also for the treatment of medial orbital wall fractures,[105] and the endoscopic-assisted transantral approach has been used to repair orbital floor fractures.[31]

Forced-duction testing at the beginning of surgery provides information about muscular involvement and has to be repeated at the end of surgery to confirm the release of herniated muscle.

The size and the location of the bony defect will guide the choice and shape of the orbital implant. If surgery is performed late after trauma, especially in orbitozygomatic fractures, osteotomies may be necessary to restore the appropriate orbital shape and volume before the implant placement.

The surgically most challenging patients are those with the sequellar forms of orbital fractures with restrictive fibrosis of the periorbit and the oculomotor muscles. The main risk is worsening of the diplopia.

Controversy persists about the choice of the material used for the fracture repair, which could be autologous (bone or cartilage)[27,28,95,102,104,158,184] or synthetic (resorbable or not).[12,32,48,55,56,66,75,85,87,94,109,122,125,130,147,152,178] The final surgical result depends mainly on the quality of the three-dimensional reconstruction of the orbit (shape and volume) and less on the choice of the orbital implant. However, this material should be easy to model and well tolerated.

Sometimes the correction of enophthalmos is not satisfactory mainly because of undercorrection. Different explanations have to be considered: misdiagnosis of the medial wall fracture, lack of material used for the reconstruction or its resorption, or the absence of reconstruction of the orbital floor convexity behind the equator of the eyeball.

Besides enlargement of the orbital container, two other mechanisms of enophthalmos may play a role in traumatic conditions: fat atrophy and contraction of the orbital content by fibrosis.

B. CHRONIC MAXILLARY ATELECTASIS (SILENT SINUS SYNDROME)

Enophthalmos due to chronic maxillary atelectasis is very often misdiagnosed. The pathophysiology is unknown but it seems that the lateralization of the middle turbinate could be the origin of the sinus disease. In fact, this malposition is almost always present and could be responsible for negative pressure in the maxillary sinus and the subsequent downward displacement of the orbital floor with enlargement of the bony orbit and concurrent enophthalmos.[17,20,21,38,57,72,73,78,81,89,90,99,117,129,142–146,164,165,176]

The patient may present with a history of sinusitis. Enophthalmos is progressive over years without any sign of inflammation (Figs. 5A and 5B).[165] Vertical diplopia may occur during the evolution secondary to the eyeball displacement.[43,180,185]

CT scan or MRI will confirm enophthalmos and enlargement of the orbit with downward displacement of the floor and reduction of the sinus cavity. The maxillary sinus mucosa may be thickened. There is no history or sign of orbital fracture and the orbital content is normal (Figs. 5C–E).[20,44,51,81,92,99,103,129,138,164,182]

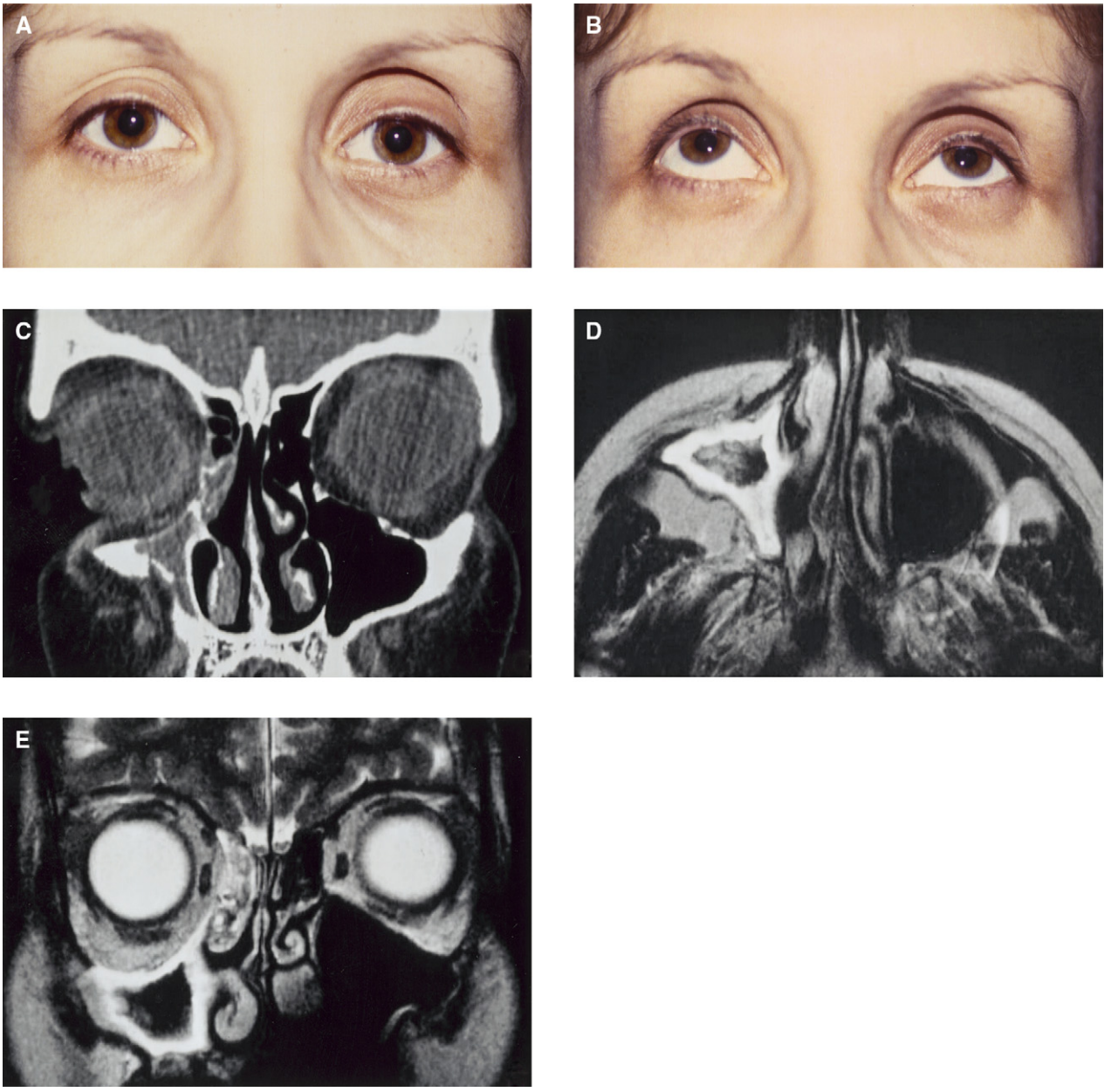


Fig. 5. *A, B:* Left enophthalmos and vertical dystopia. *C:* CT scan, coronal section, "implosion" of the right maxillary sinus and lateral displacement of the middle turbinate. *D:* MRI, axial section, right maxillary sinus atelectasis and thickening of the mucosa. *E:* MRI, Coronal section, Right orbital enlargement and downward displacement of the floor.

Given the pathophysiological hypothesis, an early middle meatal antrostomy could improve the aeration of the sinus and possibly reverse the course of the disease.[6,17,19,26,70,171] Later, diplopia and cosmetic demand are the main indications for surgery.[37,82] Subperiosteal orbital floor grafting is a reconstructive option as a secondary procedure. Many surgeons recommend simultaneous sinus drainage and orbital reconstruction, unless marked sinus infection is present.

The main complications are persistence or deterioration of diplopia, undercorrection and overcorrection of enophthalmos, and reaction against synthetic orbital implants.

C. RECKLINGHAUSEN DISEASE

The absence of the sphenoid wing in some cases of Recklinghausen disease explains the pulsating enophthalmos,[127] or proptosis (Fig. 6). There is a communication between the brain and the orbital content.[50,61,107,150] Treatment is based on the interposition of grafts by a neurosurgical approach. Autologous bone grafts may be resorbed and biomaterials are preferable.[163]

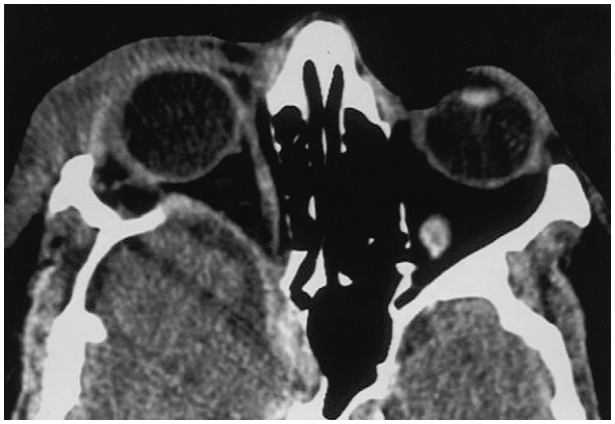


Fig. 6. CT scan, axial section, absence of the right sphenoid wing, Recklinghausen disease.

D. ORBITAL VARIX

Vascular lesions (lymphangioma, cavernous angioma, capillary angioma) in the orbit are more often responsible for proptosis rather than enophthalmos.[68]

Varix represents a venous anomaly, mainly occurring in the superior ophthalmic vein (SOV), and rarely in the inferior vein or both veins. When the dilated vein is empty, enophthalmos appears, and when the venous pressure is high and the varix is filled, the globe is pushed forward and is proptotic (Valsalva maneuver; Figs. 7A and 7B). Sometimes there are also venous malformations of the eyelids and the episclera guiding the physician towards this etiology and the diagnostic Valsalva maneuver. The enophthalmos is explained by orbital fat atrophy and sometimes enlargement of orbital space.[63]

Some activities may participate in the development of these venous abnormalities: sports, yoga, and playing music instruments.

Patients suffer from intermittent orbital pain or positional proptosis. The Valsalva maneuver confirms the venous participation and guides further investigations. In fact, MRI and CT scan could be normal on decubitus positioning, and the orbital varix visible only on procubitus positioning (Figs. 7C–F). Furthermore, spiral CT during Valsalva maneuver using a single breath hold technique could lead to the diagnosis of this venous anomaly, even in patients who are asymptomatic.[148]

Ultrasound-Doppler examination shows a low flow, compatible with venous lesions. During ultrasound, the Valsalva maneuver is often useful to show the variability of the lesion.

Orbital varix could be complicated by hemorrhage or thrombosis. Hemorrhage is responsible for sudden pain and proptosis, oculomotor limitation, and sometimes visual loss. CT scan or MRI shows a diffuse intraconal extravasation of blood. Emergency drainage could be indicated according to the functional signs. The prognosis is very often benign.

A particular clinical presentation may be pointed out and called the *blocking syndrome*. The patient presents with a sudden painful unilateral proptosis. CT scan or MRI shows an oblong limited intraconal process (Fig. 7G). There is a spontaneous clinical and radiological regression. This syndrome could be explained by the filling of the varix followed by outflow limitation. There is no change in the varix, and the Vasalva maneuver persists after the resolution of proptosis.

The treatment of orbital varix is very often conservative and surgical resection is indicated mainly in cases of anterior thrombosed lesions.

In some cases, intraoperative direct venography and embolization by cyanoacrylate glue may greatly facilitate the excision of the venous malformation.[100]

E. BREAST CANCER METASTASIS

Different cancers (breast, stomach, lung, prostate) may be responsible for enophthalmos by contraction of orbital tissues.[4,5,29,39,45,59,74,101,110,140,149,156,166,172,179] Breast cancer metastasis should be considered in the initial differential diagnosis for appropriate-aged females with non-traumatic enophthalmos (Figs. 8A and 8B). Enophthalmos due to orbital metastasis could be the first manifestation of the cancer.[74,115] Early diagnosis may improve the final outcome and prognosis.

The metastatic retrobulbar infiltrate seems to have a potential for contraction explaining the eyeball displacement and oculomotor disorders.[101]

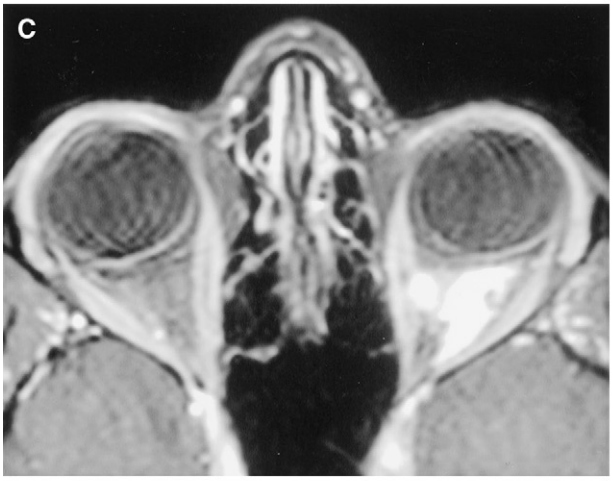
The course of the disease is very often insidious and can be subacute or more often chronic. The enophthalmos appears progressively. The inflammatory signs may be mild or absent. Eye movements are also progressively restricted secondary to the evolution of intraconal infiltration, and diplopia is a frequent motivation for consultation at this stage. The vision is unaltered. The disease may be unilateral or bilateral.

MRI is highly superior to CT scan in this pathology. It shows the intraconal infiltrate

Fig. 7. A: Left enophthalmos with vascular anomalies of the left lids. *B:* Positive Valsalva maneuver. *C:* MRI, axial section, left intraconal vascular lesion, patient on decubitus. *D:* MRI, axial section, left intraconal vascular lesion, patient on procubitus. *E:* MRI, coronal section, left intraconal vascular lesion, patient on decubitus. *F:* MRI, coronal section, left intraconal vascular lesion, patient on procubitus. *G:* CT scan, axial section, "Blocking syndrome."


A


B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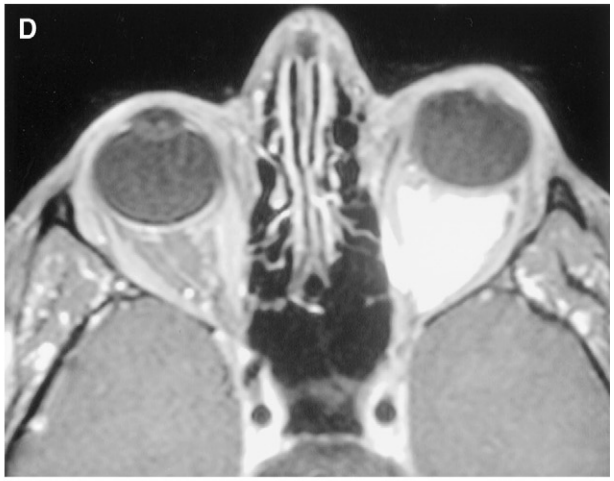
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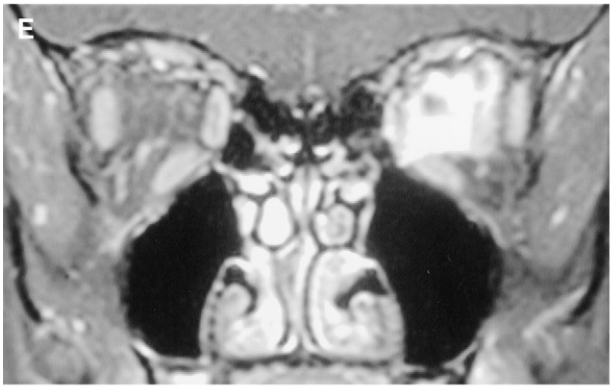
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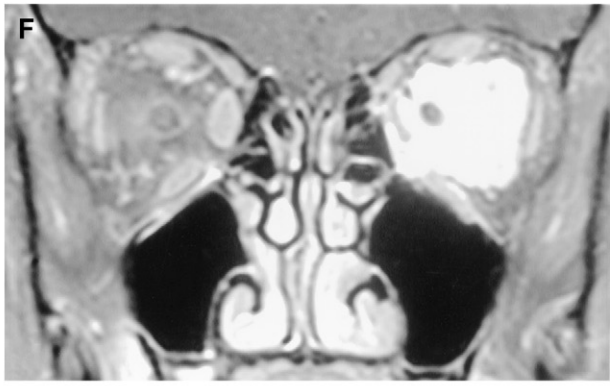
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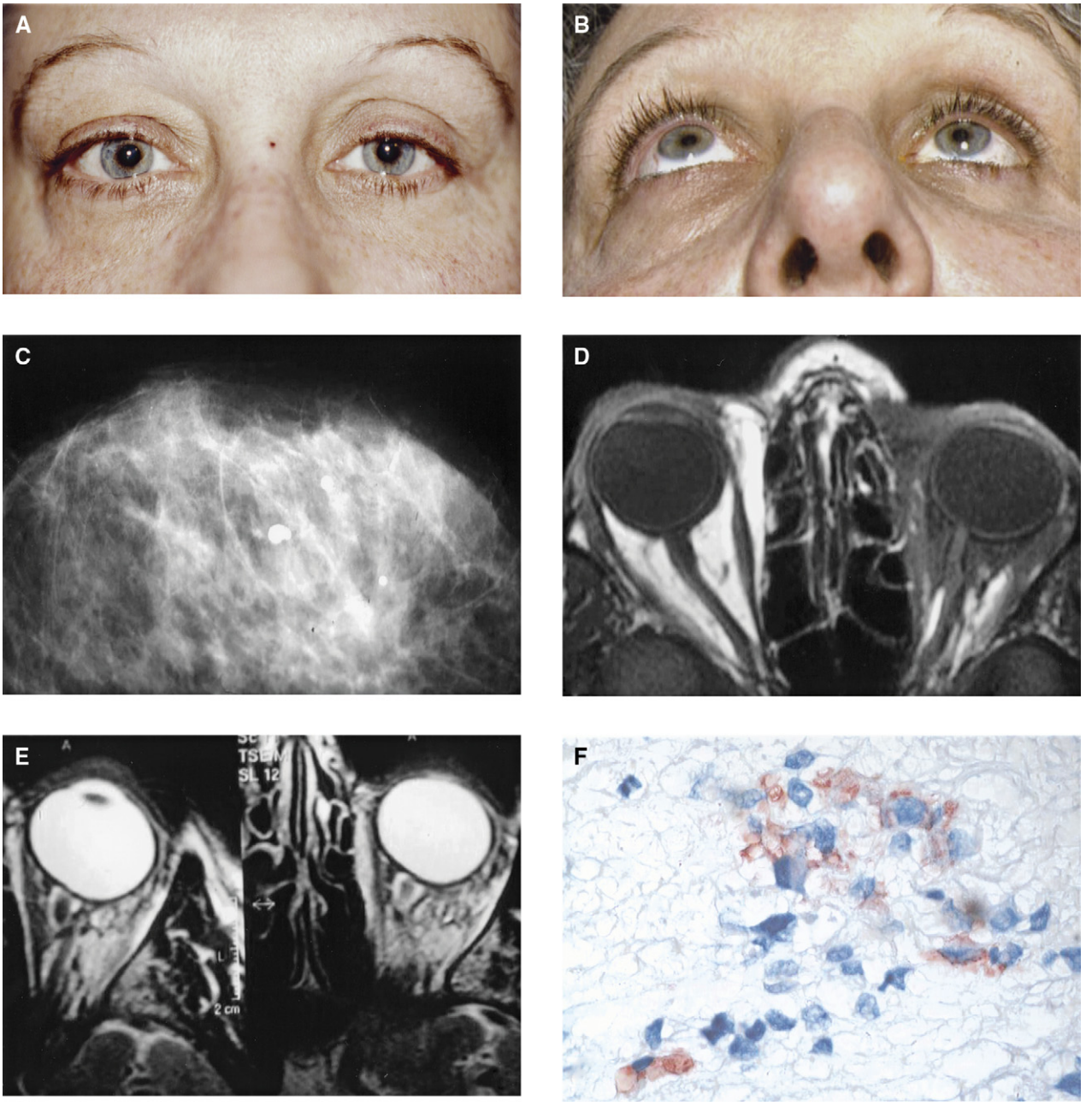

Fig. 8. A, B: Left enophthalmos. *C:* Mammogram: calcifications, breast cancer. *D:* MRI, axial section, T1, left intraconal infiltration. *E:* MRI, axial section, T2, left intraconal infiltration. *F:* Microscopic view of the infiltration of orbital tissues by malignant cells showing keratin expression.

behind the posterior pole of the eye with enophthalmos (Figs. 8D and 8E). There is no orbital wall fracture.

Further investigations concern mainly gynecological status: breast examination, mammogram (Fig. 8C), breast ultrasound, and particularly breast cancer circulating markers (i.e., CA 15-3, CEA), which may be the only sign of generalized cancer at this stage. A breast biopsy should be considered in case of clinical or radiological mass. The diagnosis of breast cancer metastasis to the orbit is confirmed by orbital intraconal biopsy (Fig. 8F). The presence of estrogen and progesterone receptors, identified by fluorescent histochemical techniques, may guide the medical treatment.[139] Orbital biopsy should be discussed with oncologists in case of negative systemic investigations.

Surgery may be difficult because of the topography of the lesions and the visual risk. Different approaches are possible, such as a conjunctival approach with disinsertion of the medial or lateral rectus muscle, or lateral osteotomy.

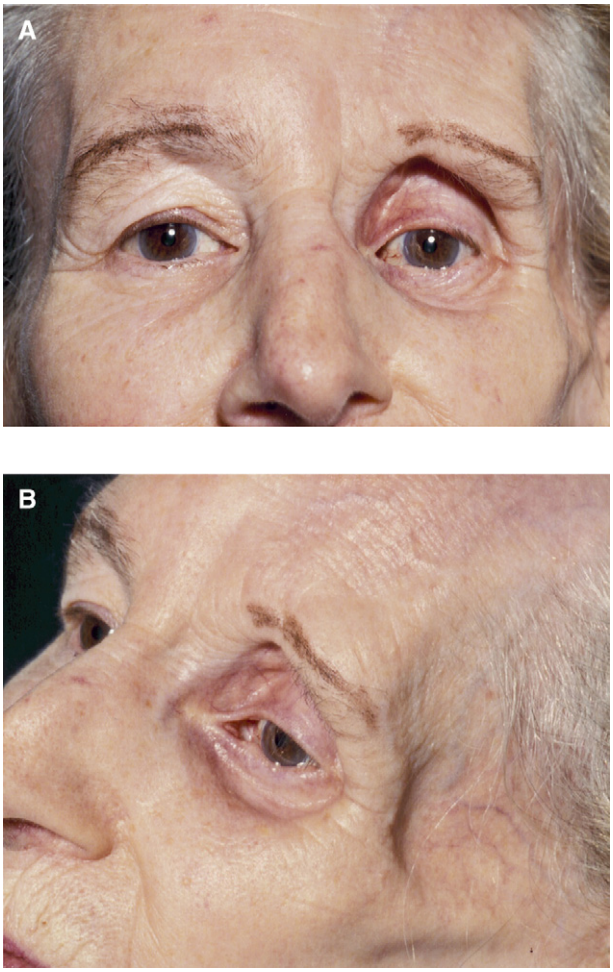

Fig. 9. A, B: Left enophthalmos: scleroderma.

The treatment of the generalized cancer is multidisciplinary, including oncologists and gynecologists, and is always based on systemic drugs (chemotherapy, anti-hormones). The indication for orbital biopsy should be discussed, and orbital radiotherapy may be considered as an adjuvant treatment.

F. LINEAR SCLERODERMA/PARRY-ROMBERG SYNDROME (HEMIFACIAL ATROPHY)

Linear scleroderma and Parry-Romberg syndrome can both be associated with enophthalmos due to orbital atrophy. The two diseases also share other common features. However, the relationship between linear scleroderma "en coup de sabre", and Parry-Romberg syndrome remains controversial and unclear.[83,106,181]

Linear scleroderma is a relatively rare disorder characterized by localized, progressive fibrosis of skin, subcutaneous fat, blood vessels, and muscles usually in the V1 dermatome ("coup de sabre") (Figs. 9a and 9b). The skin is involved first and appears indurated. Ophthalmic manifestations may include the mentioned atrophy in the orbit and periocular region, sclerosis or inflammation of the eyelids, orbit, or globe.[23,136,154,168,169,177] Serologic abnormalities may include anti-nuclear antibodies, anti-single-stranded DNA antibodies, and rheumatoid factor. Eosinophilia may be present and may correlate with disease activity. A polyclonal IgG and IgM hypergammaglobulinemia may also be present and is found more often with severe cases and with clinical progression.[173] According to histopathology, there are two phases: an early inflammatory phase with coarsened collagen bundles in the reticular dermis and perivascular lymphocytic infiltrates, and a second late sclerotic phase in which the collagen bundles become hyalinized, replacing muscle and subcutaneous fat. Therapeutic management options have included topical, intralesional, or systemic steroids; vitamin E; vitamin D3; phenytoin; retinoids; penicillin; griseofulvin; interferon-(x);[128] D-penicillamine; antimalarials; colchicines; antiplatelet therapy; ultraviolet A phototherapy with or without psoralens; and surgery.[80] One case of enophthalmos due to systemic scleroderma has also been described in the literature.[93]

In contrast to linear scleroderma, Parry-Romberg syndrome (facial hemiatrophy) is characterized by a disappearance of fat in the dermal and subcutaneous tissues on one side of the face. It occurs mainly in females, usually within the first two decades of life, and is slowly progressive.[124] The affected side of the face is bony, and the skin is thin, wrinkled, and darkened or brown in its advanced form. The facial hair may turn white and fall out, and the sebaceous glands become atrophic. Muscles and bones are usually not involved. Sometimes the atrophy becomes bilateral and involvement of the ipsilateral upper extremity and half of the body has been described. The disorder may be associated with neurological features.[113] Other ocular involvement besides enophthalmos are refractive changes,[88] ptosis, restrictive and paralytic strabismus, coloboma, heterochromia, retinitis, and uveitis.[2,7,10,16,23,52,54,62,114,159,174] The condition is a form of lipodystrophy. Histologically, Parry-Romberg syndrome resembles the sclerosis and perivascular leucocytic infiltration seen in linear scleroderma, but in Parry-Romberg syndrome there is preservation of elastic fibers.[133] Treatment approaches include cosmetic tissue augmentation of affected areas, consisting of transplantation of skin and subcutaneous fat,[58,120,131,141] as well as symptomatic treatment of neurologic signs, anti-inflammatory regimens, and stellate ganglion blocks.[35,111,114]

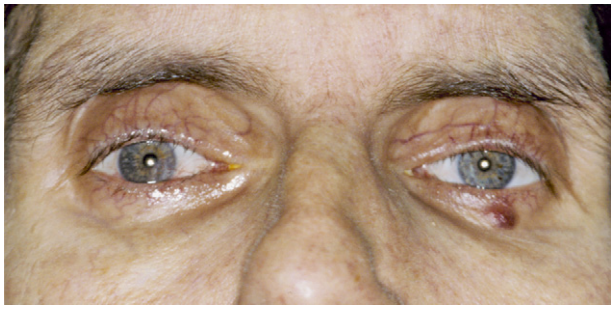

Fig. 10. HIV infection-related lipodystrophy inducing bilateral enophthalmos.

G. AGE-RELATED FAT ATROPHY

There is a natural involution of orbital fat with age. This change also involves the other parts of the face and temporal region. The age-related enophthalmos is bilaterally symmetric without any symptoms or pathologic findings. This fact is the basis for lipofilling or liposculpting for facial rejuvenation. Although the position of the globe will not change, the increased fullness of the lids will improve the cosmetic aspect.[34]

H. HIV INFECTION–RELATED LIPODYSTROPHY

HIV-infected patients often present with bilateral enophthalmos some years after the initiation of treatments (Fig. 10). Enophthalmos results from the loss of subcutaneous fat in the face, mainly at the temporal areas and at the cheeks. These findings are induced mainly by nucleoside treatment and rapidly lead to an enophthalmos due to the loss of orbital fat.[112]

I. RADIOTHERAPY

The lateral effects of radiotherapy on the orbit depend mainly on the age of the patient. Therapeutic radiation in childhood (mostly for retinoblastoma or rhabdomyosarcoma) can efficiently treat facial or orbital malignancy. Nevertheless, it might also lead to visible disfigurement including enophthalmos due to irradiation of developing structures that are in close proximity but not part of the intended target.[116] These alterations explain the effort to develop techniques for more accurate treatment of the tumoral region sparing the surrounding tissues, that is, with proton-radiation therapy.[77] The incidence for significant late enophthalmos was found to be 28% by the Intergroup Rhabdomyosarcoma Study.[137] Osseous hypoplasia and atrophy of orbital content are at the origin of these deformities. Therefore, younger children are more susceptible with the possibility of more severe secondary skull deformations and enophthalmos after radiotherapy. Consequently, visual fields can be affected by direct compression of the optic nerve due to the primary orbital disease, radiotherapy, and enophthalmos.[1] A characteristic facial appearance (hourglass facial deformity) can be induced following bilateral orbital irradiation. The radiologic findings of hypotelorism, enophthalmos, depressed temporal bones, atrophy of the temporalis muscles, narrow and deep orbits, and a depressed nasion can be seen early.[60,186]

Treatment of post-irradiation-related enophthalmos is quite challenging. Surgery in these tissues is unpredictable because the vascularity of the region is uncertain. Acceptable results can be achieved using osteotomies, tissue expansion, repositioning, and bone grafting.[15,84,91]

Radiotherapy for malignant tumors in adults (e.g., lymphomas or metastasis) could be responsible for tissue scarring, fat atrophy, and resultant enophthalmos.

IV. Summary and Conclusion

The knowledge of the different etiologies of enophthalmos is important for further diagnostic steps and appropriate treatment. Although trauma is the main cause of acquired enophthalmos, different other etiologies should be kept in mind: 1) breast cancer metastasis because of the prognostic importance of early diagnosis, 2) orbital varix because of the need to consider a diagnostic Valsalva maneuver, and 3) chronic maxillary atelectasis because of the possible reversibility in cases of early diagnosis.

V. Method of Literature Search

MEDLINE was used to search the literature from 1980 to 2006. Supplemental sources including Index Medicus and references contained in identified articles were used. The English abstracts of foreign language articles were also included as well as our personal reference libraries citing articles in French and German. Keywords searched were: *enophthalmos, enophthalmia, enophthalmos and fractures, enophthalmos and implants, enophthalmos AND Horner, enophthalmos AND ptosis, pseudoenophthalmos, enophthalmos AND anophthalmos enophthalmos AND facial hemiatrophy, enophthalmos AND scleroderma enophthalmos AND lipodystrophy, enophthalmos AND fibrosis, enophthalmos AND orbital asymmetry, enophthalmos AND fat atrophy, enophthalmos AND metastasis enophthalmos AND breast cancer enophthalmos AND orbital varix, enophthalmos AND neurofibromatosis, enophthalmos AND chronic maxillary atelectasis, enophthalmos*

AND silent sinus syndrome, enophthalmos AND radiotherapy. We excluded animal reports and animal studies. A few selected articles published before 1980 are included for historical purposes or where no other, more recently published articles could be found.

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