
Orbital Decompression in Thyroid Eye Disease

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Abstract

Thyroid-related orbitopathy (TRO) is part of a multisystemic disease mostly related to alterations of thyroid autoimmunity and characterized by a raised intraorbital pressure which causes disfigurement and functional deficits in the affected patient. Reduction of pathologically raised intraorbital pressure can be achieved by means of anti-inflammatory medical treatments or by decompression surgery which consist in enlarging the bony orbit and/or orbital fat removal. In the active phase of TRO, glucocorticoids are the first-line treatment of dysthyroid optic neuropathy; in the case of failed medical treatment, decompression surgery can be a valid alternative. Decompression surgery is the mainstay therapy for the treatment of stable typical disfiguring alterations and/or symptoms of TRO, although it has rarely been linked to reactivation of stable orbitopathy. The multifaceted nature of the disease, the different indications for decompression surgery, surgeon preferences and expertise, variations in orbital osteology, and patients' expectations and attitude toward intervention and possible complications imply the use of many different surgical techniques. Most of the techniques currently used seem to be effective in restoring optic nerve dysfunction, eye position, and/or congestive symptomatology and disfigurement; nevertheless, an unbiased analysis of the current literature in terms of effectiveness versus safety is extremely difficult due to the great heterogeneity of the patients included in the published studies and variations applied to surgical techniques. Fibrosis due to long-lasting orbital disease or possible consequence of retrobulbar irradiation administered in the early phase of TRO has been questioned as being a possible cause of poor distensibility and plasticity of the orbital soft tissues leading to diminished effectiveness of orbital expansion surgery. Recent investigations do not confirm this. Consecutive strabismus may complicate any decompression procedure, but its treatment is not particularly complicated.

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Thyroid-associated orbitopathy (TAO) can be a disfiguring and functionally disabling disease, which in severe cases may result in permanent visual loss. Different degrees of disfigurement and alteration of vision contribute to loss of self-confidence, psychosocial stability and ability to function.

Refraining from smoking, and avoidance of passive smoking, together with prompt restoration of stable euthyroidism and, when necessary, immunosuppression are the

first-line medical treatments. Surgery, which may be necessary to protect visual function in the early active phase of the disease or to correct the stable typical disfigurement and symptoms during the inactive phase which follows, should always be considered rehabilitative as it is aimed at restoring the individual integrity disrupted by the disease and ultimately the lost ability to function. Commonly, however, surgery performed primarily to treat potentially sight-threatening conditions is referred to as 'functional', while procedures primarily aimed at correcting disfigurement and symptoms are referred to as 'rehabilitative'. Besides semantic considerations, a clear-cut distinction between the two does not exist as surgery aimed primarily at restoring function also has positive effects on disfigurement and vice versa [1].

For more than a century, decompression surgery has been used to treat TAO, and through the years it has been subjected to a paradigm shift in respect to indications, approaches and surgical routes. This has been largely due to a better understanding of the multifaceted nature of the disease and a constant attempt to implement the beneficial effects of this type of surgery, while simultaneously decreasing the aesthetic impact of surgical scars, hospitalisation time, convalescence periods and risks for iatrogenic complications in general, and consecutive strabismus in particular. Patients' increased expectancies and a more critical attitude towards surgical interventions are also aspects that have driven the shift in orbital decompression surgery.

This chapter offers an up-to-date overview on orbital decompressions and arguments for clinical choices when dealing with this type of surgery.

Historical Notes and Modern Trends of Decompression Surgery

The earliest surgical approach to bony orbital decompression was published by Dollinger in 1911 [2]. He adapted Kroenlein's technique [3] for removal of an orbital dermoid cyst to increase towards the temporal fossa the volume of the bony orbit. The transfrontal orbital roof decompression advocated by Naffziger in 1931 [4], which was designed to be a possible access to both orbital apices in bilateral dysthyroid optic neuropathy (DON), did not offer an adequate possibility of proptosis reduction. The transfrontal orbital roof approach was soon abandoned as it resulted in a time-consuming and hazardous technique. In addition to this, pulsating exophthalmos, an unwanted side effect induced by extensive removal of the orbital roof, appeared often [5].

Sewall's approach, which followed in 1936 and involved the removal of the medial orbital wall and part of the floor through a transcutaneous route, did not gain more popularity than the two previously described techniques [6].

In 1950, Hirsch [7] adapted the orbital decompression technique, which was described earlier by Lewkowicz [8] to treat orbital complications of paranasal sinusitis. Hirsch's technique entailed removal of the orbital floor.

Soon afterwards, in 1957, Walsh and Ogura [9] proposed a combined approach to the medial orbital wall and floor. Their surgical approach was inspired by the transbuccal access to the maxillary sinus proposed at the end of the 19th century independently by George Caldwell (first) and Henri Luc (later) [10]. Walsh and Ogura's approach was widely used for decades until the early 1980s, when the high incidence of post-decompression diplopia [11] and infraorbital dysesthesia, later calculated to be in the order of 50% [12–14], suggested alternative procedures should be sought.

Beginning in the early 1980s, several other approaches, including the swinging eyelid [15], coronal [16, 17] and transcaruncular [18, 19] approaches, were proposed or popularised, and in 1989 the concept of balanced decompression was introduced [20]. This technique, which theoretically should have minimised the risk of iatrogenic diplopia, later appeared to be associated with a higher risk for such a complication as compared with removal of the lateral orbital wall alone or with inferomedial and 3-wall techniques [21, 22].

In 1990 the transnasal endoscopic approach was introduced for treating exophthalmos and DON [23]; once again, the lateral orbital wall, and in particular its deep portion, was promoted as being the region of first choice for orbital decompression. Its removal, associated with a low risk of consecutive diplopia or severe complications such as cerebrospinal fluid leak, perfectly fit the needs of this increasingly demanding patient population [24–26].

It was reported that in moderate-to-severe TAO, the removal of the deep lateral wall as a part of a rehabilitative coronal approach 3-wall decompression enhances exophthalmos reduction by 32%, without increasing the risk of consecutive diplopia as compared with traditional more conservative 3-wall decompression. The same study, however, confirmed the known high interindividual variability of the volume of the deep lateral wall. In light of this, the deep lateral wall, more so than the region of first choice for orbital decompression, is to be considered an effective although not always available zone of possible orbital volume expansion when dealing with rehabilitative decompression surgery [25]. The effect of pure lateral wall decompression on exophthalmos reduction may be modest if not associated with medial wall removal, but in these cases the risk of consecutive diplopia rises. However, the effect of lateral wall decompression can be implemented by intraconal fat removal without substantially increasing the risk for iatrogenic strabismus. The removal of the lateral orbital wall and intraconal fat has even been beneficial in reducing pre-operative primary-gaze diplopia [27].

Pure orbital fat decompression was first described by Moore in 1920 [28]. A mean exophthalmos reduction of 6 mm and an improvement of extraocular eye motility with new-onset strabismus in only 4% of the cases was reported by Olivari in the late 1980s [29], but the same results were not confirmed by other authors or by Olivari himself. With fat removal orbital decompression, Trokel et al. [30] could obtain only a modest reduction of exophthalmos, which was on the order of 1.8 mm, rising to 3.3 mm only for those patients with preoperative Hertel measurements greater than 25

mm. Better results were reported by Adenis et al. [31], who were able to obtain an average reduction of exophthalmos of 4.7 mm with complications limited to motility disturbances only; however, the incidence of new-onset strabismus was much higher than previously reported by Olivari. According to a more recent and larger series published by Olivari and co-workers [32], although exophthalmos reduction remained in the same order of his earliest study, new-onset strabismus rose to 20.2%. In the last years, the combination of bone decompression associated with fat removal has gained popularity in view of its claimed safety and increased effectiveness as compared with bone or fat decompression alone [33–35].

At present, orbital decompression is mostly achieved by means of osteotomies which involve the lateral and medial orbital walls as well as the orbital floor. When necessary, fat is removed from the inferolateral orbital quadrant as this zone, which does not host important orbital structures, appears to be the safest orbital site for lipectomy [36]. The most effective and safest sequence of osteotomies and lipectomies to be used to gradually implement the effects of decompression surgery continues to be a subject of debate [33, 35, 37]. Beveled osteotomies and onlay alloplastic periorbital implants, although sporadically used to camouflage more than to reduce exophthalmos, remain of uncertain effectiveness [38–40], and their edges, which are sometimes visible, may devaluate the final rehabilitative cosmetic result.

Indications for Orbital Decompression

Originally, decompression surgery was limited to the cure of sight-threatening conditions such as optic neuropathy refractory to medical therapy or exposure keratopathy unresponsive to lubricants, bandages and eyelid surgeries. More recently, the indications of orbital decompression were extended to the treatment of disfiguring exophthalmos and symptoms. Eyeball subluxation, which may be a possible cause of acute DON and exposure keratopathy, postural visual obscuration in patients with congestive inactive TAO, and choroidal folds represent other more recently recognised indications for decompression surgery [41].

DON occurs in up to 5% of patients with TAO, and if left without treatment it is a potentially blinding condition in 30% of the cases [42, 43]. Older age, male gender and smoking are factors associated with an increased risk for DON [44, 45]. Compression and stretching, an abnormally short optic nerve, vasculopathy and inflammation have been quoted as possible causes of optic nerve dysfunction in TAO [35, 46, 47]. Nevertheless, the latter remains purely a theoretical speculation which has never been confirmed by necroscopic histopathological studies [48].

The most widely accepted mechanism underlying DON is compression of the nerve or its vasculature by the enlarged extraocular muscles [49]. The randomised clinical trial of Wakelkamp et al. [50] comparing surgical decompression to medical decompression as a first-line treatment for DON led to the conclusion that immediate

decompression surgery did not result in a better outcome in terms of increased visual acuity; instead, intravenous followed by oral glucocorticoids appeared to be the therapy of first choice. Orbital decompression possibly followed by different combinations of immunosuppressive treatments can be used in the case of failure or partial success of systemic glucocorticoids alone.

Independently, if the pathophysiology of optic nerve dysfunction is sustained by enlargement of the extraocular muscles or increased volume of the orbital fat compartment, or both, it has been hypothesised that when the volume of the soft tissues is reduced by fat removal or the volume of the bony orbit is expanded by any type of osteotomy, critical relief of pressure at the apex may be achieved. Data are available to support this idea [2, 51]. The actual trend for surgically addressing DON is, however, to remove the medial wall which is the only surface which reaches the orbital apex [35].

In TAO, increased palpebral fissure width, exophthalmos, blink rate, lid lag, lagophthalmos, deficit of elevation and poor Bell's phenomenon can all be potentially connected with alterations of the ocular surface. The literature concerning the alteration of the ocular surface occurring in TAO is not extensive [52], but according to the findings of Gilbard and Farris [53] the widened palpebral fissure has a prominent role in determining the alterations of the ocular surface. Although the effect of decompression surgery on severe corneal alteration has never been specifically addressed, most of the studies dealing with orbital decompressions report reduction in symptoms associated with exposure keratopathy [54]. This may be linked to a reduction in eyelid aperture induced by decompression surgery. One of our studies showed a clear reduction of eyelid fissure based equally on decreased upper and lower lid displacement in about 50% of patients presenting with pre-operative increased eyelid aperture, and decompressed with the 3-wall coronal approach, which leaves upper and lower lid retractors undisturbed [55].

Eyeball subluxation is rare, but deserves urgent referral to a specialist centre [56, 57]. It is a recurrent complication which represents a potential cause of visual loss, which may benefit from orbital decompression [1, 56, 57].

Sub-optimal optic nerve perfusion, which can occur in elderly or diabetic patients affected by TAO, may be responsible for visual obscuration whenever there is a drop in systemic blood pressure due to postural changes. The vascular embarrassment of the optic nerve, dependent on elevated intraorbital pressure, can be relieved very effectively by orbital decompression, and leads to an immediate cessation of postural visual obscuration [58].

Choroidal fold due to eyeball indentation subsequent to enlarged extraocular muscles had been demonstrated to respond to orbital decompression [59], and it is conceivable to expect a better response to treatment in the case of short-lasting folds.

Visible deformity, particularly involving the face, has always induced aversion, prejudice and isolation. Patients with facial disfigurement suffer from intrusions such as staring or comments. At the root of patient's distress lies societal pressure to con-

form to an idealised appearance. The obsession with appearance devalues those who do not match the perceived ideal and stigmatises those with visible disfigurement [60].

Although disfiguring exophthalmos affects as many as 62% of patients with TAO [61], most of patients with TAO do not require surgical intervention. However, a large case series demonstrated the cumulative probability of having surgery for patients with TAO increases in the course of their orbitopathy from 5% one year after diagnosis to 9.3, 15.9 and 21.8% at year 2, 5 and 10, respectively. The need for surgery was significantly related to age, with an overall risk for intervention 2.6 times greater in patients older than 50 years [44].

Most of the studies dealing with orbital decompressions have indicated that this type of surgery is associated with lessening of subjective perception of retro-ocular tension. In the early 1990s, Khan et al. [62], using the McGill Pain Questionnaire and visual analogue scales, specifically addressed this issue, and although their study was not free of biases, it seemed to confirm that orbital discomfort significantly responded to orbital decompression. When rehabilitative orbital decompression is aimed at addressing the tiresome retro-ocular tension that may characterise the post-inflammatory stage of TAO, surgery can be performed with minimally invasive techniques leading to minimal, if any, impact on extraocular motility or complications in general [63].

Timing of Rehabilitative Orbital Decompression

Depending on the severity of TAO, surgical rehabilitation can be more or less extensive, the full treatment consisting of decompression surgery, squint surgery, eyelid lengthening, and eyebrow- and blepharoplasty. Surgery is performed in that order since the preceding step may influence the necessity and extension of the step that follows. In particular cases the rehabilitation can be favourably sped up by carrying out more than one procedure at the same time [1].

When rehabilitative decompression surgery is necessary, it can be undertaken after 4–6 months of stable euthyroidism and orbitopathy, with the stability of TAO assessed based on the absence of signs of inflammation and no changes in disease severity. Decompression surgery has been advocated soon after achievement of stability, as it has been questioned whether fibrosis due to long-lasting orbital disease is a possible cause of poor distensibility and plasticity of the soft orbital tissues resulting in scarce effectiveness of orbital expansion surgery [64–66]. A study by our group, however, demonstrated that long-lasting TAO or pre-operative radiotherapy, which may represent another possible cause of fibrosis of the soft orbital tissues, does not adversely interfere with the results of orbital bony decompression. Therefore, when the stabilisation of Graves' disease and of the orbitopathy have occurred for some months, rehabilitative surgery can be started at any time and no adverse effects from common preceding treatment such as retrobulbar irradiation are to be expected [55, 67].