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Thyroid Eye Disease Double Vision and Loss of Vision

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Sir Robert Graves, MD., first described the association of hyperthyroidism, goiter and exophthalmos in 1835. Since then, other eye-related findings have been described in patients with Graves' disease, including swelling and edema of the conjunctival surface of the eye (chemosis), prominence of the tiny blood vessels overlying the white (sclera) part of the eye, as well as where the muscles attach to the eye, swelling in the tissues around the eyes (periorbital swelling), lid lag, lid retraction, visual loss, and limitation of eye movement (ophthalmoparesis). When the full array of ocular symptoms and signs are clearly present, the diagnosis of Graves' disease is readily established. This occurs in only three to five percent of cases. Hence, there are many patients with less overt manifestations, and who are more difficult to diagnose. The purpose of this bulletin is to review the abnormalities of

lid function and ocular mobility that occur in patients with Graves' disease.

Lid retraction gives the hyperthyroid patient their "bug-eyed" appearance. The eyes seem prominent either because they pushed outward, a term referred to as proptosis or exophthalmos, or because the upper lids are abnormally pulled back and the white portion of the eye (sclera) shows more than usual. Normally, the upper lid just touches the upper border of the iris and the lower lid just covers the lower iris margin, so there is no "scleral show" between the top and bottom of the iris and the surrounding sclera. When scleral show is present with the eyes looking straight ahead, there is lid retraction, and when it occurs as the patient slowly moves their eyes downward, it is called lid lag. Lid retraction and lid lag may occur in one or both eyes; it can be asymmetric, intermittent, and sometimes quite subtle.

Although lid lag and lid retraction occur most commonly in patients with thyroid eye disease, it can also be seen in a number of other neuro-ophthalmic disorders. From the lower animals up to and including man, a standard set of six ocularotary muscles in each eye move the eyes in the horizontal, vertical and oblique planes. They may be grouped into three pairs: horizontal (the medial and lateral recti), vertical (the superior and inferior recti), and oblique (the superior and inferior oblique).

The medial rectus moves the eye along a horizontal plane toward the nose. Its antagonist, the lateral rectus muscle, moves the eye in the opposite direction or horizontally away from the nose. The superior rectus muscle primarily raises the eye; the inferior rectus muscle lowers it. And the superior and inferior oblique muscles allow the eye to rotate diagonally.

Under-action of any one or more of these eye muscles results in the symptom of double vision (diplopia).

Inefficient eye muscle function arises from two processes: either weakness of the eye muscle itself or mechanical restriction. Weakness may result from interruption of the nerve supply to the eye muscle (denervation). Diseases that affect the cranial nerves which supply the eye muscles result in denervation palsy. Consequently, the eye muscle or muscles respond slower and less extensively than normal. In contrast to denervation, an eye muscle may also not function efficiently if it is infiltrated, engorged, entrapped or swollen. Being bulkier than normal, the eye muscle stretches less extensively because of mechanical inefficiency.

The eye muscles of patients with Graves' disease are enlarged, bulky, firm, and rubbery, owing to swelling (inside the cells), scarring (inside the muscle fibers), and a heavy infiltrate of protein cellular substances (collagen, mucopolysaccharides, lymphocytes and plasma cells). These cellular and semi-solid substances which deposit in the muscles hamper eye movement by

mechanical restriction. For instance, if the medial rectus muscle is involved, the eye becomes tethered toward the nose. Hence, the eye does not move well looking to the side because the lateral movement of the eye is hampered by the bulkiness of the medial rectus. This same infiltrate of protein and cells affects the fat tissue behind the eye, further accounting for the exophthalmos, feeling of pressure, and excessive tearing that is so characteristic of Graves' disease. While the changes just mentioned are seen most commonly in typical Graves' disease with thyrotoxicosis, they can also occur in euthyroid Graves' disease, Hashimoto's thyroiditis, and on rare occasions, in hypothyroidism.

These eye muscle changes usually do not affect the right and left eyes evenly or equally. For this reason, one eye may move faster and farther than the other, resulting in double vision (diplopia). Such patients complain of seeing two images with both eyes open. To lessen or totally avoid the double vision, they often compensate by turning their head, or changing the position of their face or neck in order to fuse the images into one. They may have to close or patch one eye in order to regain single vision.

The diplopia of thyroid eye disease ranges from mild to severe, temporary to permanent. When mild, double vision may only occur when the patient looks far upward, downward or to the right or left. When severe, double vision intrudes on straight ahead gaze. In the early stages, eye problems related to an improperly operating thyroid (dysthyroid ophthalmopathy) may mimic other neurological and ophthalmological causes of double vision. Examples are myasthenia gravis, multiple sclerosis, brain tumors and strokes. It is not uncommon for the correct diagnosis to take weeks and sometimes months to establish. The critical clinical test in a patient who complains of diplopia, which helps distinguish between eye muscle weakness versus mechanical restriction, is the forced duction test. This consists of the doctor applying a forceps to the front surface of the eye and attempting to passively move the eye in the direction of apparent limitation, while the patient sits still. When an eye muscle is weak from denervation, the patient may not be able to move the eye voluntarily, but the examiner can passively move the globe in the desired direction. If the eye cannot be moved passively, it obviously is being tethered by restrictive forces.

The most common causes of misdiagnosis of thyroid eye disease include the following: (1) the conspicuous absence of other systemic or laboratory signs of thyrotoxicosis; (2) subtle, elusive or atypical ophthalmosparesis (apparent limitation of eye movement) that was either minimal or variable; and (3) failure of the examiner to recognize mild lid lag and/or to perform the forced duction test.

Enlargement and infiltration of the extraocular muscles results in vascular engorgement of the small vessels which supply these eye muscles. Two of the eye muscles, the medial and lateral recti, attach to the eye in a relatively visible portion of the globe. By asking a patient to look all the way to their right, the pink muscular insertion of the right medial rectus can be seen, and looking into an extreme left gaze exposes the right lateral rectus muscular insertion. In thyroid eye disease, the vascular perfusion of these muscular insertions increase, so the superficial vessels become engorged and twisted, resulting the bloodshot eyes of the active phase patient.

Treatment of Double Vision

The treatment of double vision in patients with thyroid eye disease depends upon the degree of extraocular muscle

involvement. For instance, double vision which is present only in extreme gazes – up, down, right or left – is managed more conservatively than double vision in straight ahead gaze. With the former, there is very little need for medical or surgical intervention. Since double vision looking straight ahead can be an occupational handicap, there are an array of medical and surgical therapy options. Before any treatment is prescribed, the examiner must be certain of the following: (1) that vision in each eye, related to optic nerve function, is normal; (2) that visual loss, if present, is not related to drying of the front surface of the eye due to protrusion or ineffective blinking (exposure keratopathy), in which case the use of lubricating agents might be helpful; and (3) that the deviation between eyes cannot be neutralized simply by the application of Fresnel prisms.

The majority of patients with all forms of thyroid disease can be managed conservatively. Many whose double vision is present in straight ahead gaze can be managed by using prisms, either pressed on or ground into their prescription glasses. The experienced examiner measures the patient's eyes for vertical, horizontal or oblique misalignment. The

nine cardinal positions of gaze are: straight ahead, eyes to the right and left, and eyes up and to the left and down and to the left. After quantifying these deviations, a plastic prism can be affixed to prescription lenses which neutralizes the misalignment in straight ahead gaze and gives the patient single vision with both eyes open. Because this prism consists of a soft plastic material, it often distorts clarity. But it is only used temporarily. If the patient finds the prism acceptable because it corrects their double vision, and if the misalignment of the globes remains stable, this amount of prism can be ground into prescription lenses.

Treatment of Visual Loss

If the eye muscles become sufficiently enlarged, they may compress the optic nerve, they can interfere with the visual information from the eye back to the brain. Slowly developing compressive optic neuropathy is characterized by blurred vision, a change in the perceived sharpness of color vision, and greater difficulty seeing in darkness or reduced light. The patient with optic nerve involvement on one side will often say that the eye does not see colors as brightly or as vividly as the other eye. Further, they also find themselves putting on more

lights to see as well with the affected eye. Since these symptoms may go unnoticed in the presence of one good eye, patients with thyroid ophthalmopathy should be examined at regular intervals depending on the severity of the symptoms and signs. Severe double vision, proptosis, or evidence that the optic nerve is being compressed from the overhanging eye muscles, warrants immediate and sometimes aggressive therapeutic intervention. This may consist of a short course of oral corticosteroids, irradiation of the orbits, or decompressive surgery.

Oral corticosteroids may provide some temporary relief for the patient and sometimes enormous benefits. However, they should not be continued for long periods due to the uncomfortable and sometimes serious side effects which result from chronic administration of corticosteroids. Careful follow-up is necessary in these patients since cortisone and its derivatives may precipitate diabetes, hypertension, glaucoma, peptic ulcer disease, softening of the bones and fluctuations in mood. These mood changes can range from mild depression to frank psychosis.

Orbital irradiation or surgical decompression of the orbital structures may be necessary in patients with severe ophthalmopathy, particularly if there are signs that the optic nerve is being compressed (compressive optic neuropathy). Orbital irradiation consists of usually giving 1,600 to 2,000 rads in eight to ten divided doses delivered by way of ports on either side of the head that are aimed at the apex of the orbit. The patient is lying down, and kept very still, in the same position, for the 8 to 10 treatments, which are delivered about a 10-day to two week time period. The low dose of radiation plus the care that is taken to avoid irradiation of the eyeball itself helps avoid such complications as cataract formation and damage to the retina and optic nerve.

In summary, the double vision and loss of vision which occurs in patients with thyroid eye disease can be managed effectively. It is important to have regular evaluations by a specialist who sees a lot of patients with thyroid ophthalmopathy. Precise measurements of eye muscle and visual function should be recorded each visit and compared from one visit to the next. A subtle change may indicate improvement or worsening. In most cases, the double vision of thyroid eye

disease can be managed conservatively with patching or prisms. If the symptoms persist unmitigated by these measures, eye surgery can be done to relieve the double vision by realigning the muscles.

Involvement of the optic nerve is a more frightening complication. Immediate intervention is indicated. If visual improvement is noted with steroids and persists after the drug is tapered, no further therapy may be necessary. If, however, visual loss relapses with lowered dosages of prednisone, then more steroids (for a short amount of time), irradiation and/or decompressive surgery should be considered.

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