

THYROID OPHTHALMOPATHY

A General Overview

INTRODUCTION

Thyroid eye disease, thyroid ophthalmopathy, thyroid orbitopathy, and endocrine orbitopathy are all names, which describe a disorder resulting from inflammation of muscles and fat within the bony box (orbit) surrounding the eyes. Thyroid ophthalmopathy may cause the eyes to bulge forward and the lids to become swollen, red, or retracted. This disease occurs four times more commonly in females, and although any age group may be affected, it is more frequent among middle-aged individuals. Remember, 10% of all women by the age of 55 years have some thyroid abnormality, and 20% of these people will have clinically significant thyroid ophthalmopathy.

ASSOCIATION WITH THYROID ABNORMALITIES

Thyroid ophthalmopathy is typically associated with disorders of the thyroid gland, which is located in front of the lower throat. This gland produces thyroxine, a hormone that affects appetite, metabolism, heart rate, and body temperature among other things. Symptoms associated with abnormally high levels of thyroxine (hyperthyroidism) include hunger, weight loss, nervousness, anxiety, tremors, perspiration, hair loss, premature hair graying, white spots in the skin, menstrual irregularity, and rapid pulse. Low thyroxine levels (hypothyroidism) may cause cold intolerance, fatigue, weight gain, depression, and facial puffiness. Somewhere between six and ten percent of patients with thyroid abnormalities may develop thyroid ophthalmopathy. Sixty to seventy percent of patients develop ophthalmopathy during or after an episode of hyperthyroidism (the remainder may have normal or low thy-

roid levels). **Although thyroid ophthalmopathy is associated with disease of the thyroid gland, the two conditions (thyroid ophthalmopathy and thyroid gland disorder) evolve, progress, and respond to treatment independently.**

CAUSE OF THE DISEASE

The cause of thyroid ophthalmopathy is still unknown; however, it is probably due to a disorder of the body's immune system, resulting in an attack on normal body tissues. Other similar inflammatory diseases include rheumatoid arthritis, lupus erythematosus, and certain types of diabetes. In thyroid ophthalmopathy, the tissues attacked by the immune cells are the muscles, fat, and tear glands surrounding the eyeballs. As a result the following symptoms and signs may occur:

SYMPTOMS AND SIGNS

1. EYEBALL PROPTOSIS

Swelling of the soft tissues around the eye may cause the eyes to protrude (proptosis). There are 6 muscles that move your eye. Four of these, the inferior rectus, superior rectus, lateral rectus and medial rectus, are most frequently involved. These muscles originate behind the eye at the peak of the eye socket and attach to the eye just behind the cornea (the clear portion of the eye overlying the colored part of the eye). The muscles cannot be seen on the surface as they are covered by a thin layer of tissue (the conjunctiva) but may become visible as the blood vessels over their anterior portion become very prominent. The immune system singles out the fibrob-

lasts, support cells within the muscles causing the muscles to enlarge. With muscle enlargement the globe (eyeball) is pushed forward leading to the characteristic "stare." Proptosis frequently occurs asymmetrically, with one eye being more prominent than the other; but generally, both eyes are involved to some degree. As the muscles get larger, 3 things can happen. The eyeball gets pushed forward, the muscles themselves become stiff (the eye may not move normally), and the muscles may press on the optic nerve.

2. EYELID RETRACTION

Inflammation and scarring of the muscles that pull open the eyelids may leave them abnormally open, causing a wide "stare". This may be exaggerated by the proptosis of the eyes. In some cases, the eyelids may be so retracted that they don't completely close, even during sleep.

3. DECREASED TEAR PRODUCTION

The tear glands may become inflamed or scarred, decreasing tear production.

4. DRY EYES OR EXCESSIVE TEARING

Prominence of the eyes, eyelid retraction, and / or decreased tear production may lead to exposure and rapid tear evaporation with foreign body sensation, aching, burning and excessive reflex tearing. Severe cases may be complicated by breakdown of the surface of the eye (corneal ulcer).

5. SOFT TISSUE SIGNS

Poor venous drainage resulting from swollen orbital tissues may cause red-

ness and swelling of the eyelids and conjunctiva (the mucous lining of the eye). In severe cases, the swollen conjunctiva may hang over the edge of the lids looking like a bubble or a blister on the surface of the eye. We strongly advise against the use of over-the-counter ophthalmic decongestants - eyedrops that "take the red out." Such medications can cause very serious problems.

6. GLAUCOMA

Some patients will develop glaucoma, which is increased pressure inside the eye. This condition is generally not painful, but if unrecognized and untreated it can cause slowly progressive vision loss.

7. DOUBLE VISION

Inflammation and scarring of the muscles that move the eyes may lead to impaired eye movement. In mild cases, one might feel a pulling sensation when moving the eyes. With more advanced disease, double vision may occur when looking in certain directions. The inferior rectus muscle (located beneath the eye) tends to be more often affected than others. When it becomes stiff, the globe cannot move up normally. This often results in double vision with one image seen on top of the other. In some cases, eye movement can be very restricted and the eyes may become obviously misaligned with constant double vision.

8. OPTIC NERVE COMPRESSION

Severe swelling of the tissues near the

back of the eye may press on the optic nerve, the cable transmitting visual signals from the eye to the brain. Early symptoms include fading of colors and graying of vision. Permanent visual loss may occur if this complication is not recognized and promptly treated. This severe loss of vision fortunately, occurs only in about 5% of the patients with thyroid orbitopathy and may be reversible if the pressure on the optic nerve is relieved.

COURSE OF THE DISEASE – ACTIVE & INACTIVE PHASES

Thyroid ophthalmopathy typically has an active, inflammatory phase lasting 6-24 months (rarely as long as 3-5 years). After the inflammation has died down, individuals may be left with any of a number of structural changes, which might require treatment. Recurrences of the active phase are very uncommon (less than 1% of people). Unfortunately, we have no test to tell when a person has passed from the "active" phase to the "inactive" phase, and we rely on your and our powers of observation. We assume that if the eyes have not changed in appearance or function for 6 months, then you have entered the "inactive phase".

Every person's thyroid eye disease follows a unique course. Some people may have minimal symptoms or signs and others may have a sudden onset of severe complications such as vision loss or major eyelid or eye swelling and redness.

We are still unable to predict accurately which complications a particular individual will develop. Patients must therefore be followed on a regular basis during the

active phase of their disease. Any additional signs and symptoms should be reported **immediately**, in case specific treatment is needed. **A concerned telephone call to your physician is never "a bother," but a smart and responsible action.**

SMOKING AND STRESS

An association between smoking and increased severity of thyroid ophthalmopathy has clearly been demonstrated, especially in women. **All patients who smoke and have thyroid disease, should make a particularly strong effort to stop smoking.** Ask your physician for help. Stress is also associated with exacerbation of thyroid eye disease. **Stress reduction techniques may be very beneficial.**

DIAGNOSIS AND INVESTIGATIONS

Thyroid eye disease is diagnosed by the clinical features described above. It is confirmed by ultrasound and CT scan showing the enlarged muscles around the eyes. Other tests may be ordered to document visual function and eye movements. Photographs, for example, document the appearance of the eyes to judge progression over time.

Also, blood and urine tests are used to monitor the activity of the thyroid gland and the inflammatory process. In addition, nuclear medicine scans of the thyroid gland may be ordered by your endocrinologist or primary care physician.

TREATMENT OF ACTIVE PHASE

I. THYROID GLAND MODULATION

The Endocrinologist may prescribe various medications to suppress or augment the thyroid hormone level. A radioactive iodine drink or thyroid removal surgery may be offered to destroy portions of an overactive thyroid gland. Although these treatments are very important for the patient's general well being, they do not appear to directly influence the course of eye disease in most patients.

2. EYE MEDICATIONS

Mild exposure symptoms, dry eyes, and even excessive tearing can often be relieved with lubricating teardrops and ointments.

3. SALT RESTRICTION AND HEAD ELEVATION

Swelling symptoms may be decreased by cutting down on salt in your diet and elevating the head of your bed by placing bricks under the feet at the head of the bed.

4. ANTI-INFLAMMATORY MEDICATIONS

Moderate to severe inflammation and redness of the eyelids and conjunctiva may be treated with corticosteroids or other immune-modulating drugs. Some people respond to these medications, but others do not. If a person is going to respond to these medications, they do so rapidly (within 3 to 14 days). In cases of optic nerve compression, these medicines may be used

in addition to radiation therapy or surgery. Because some of these medications have potentially serious side effects, they are not used in mild cases of thyroid ophthalmopathy.

5. RADIATION THERAPY

X-ray therapy has been shown to reduce inflammation and is offered in cases of thyroid orbitopathy complicated by moderate to severe soft tissue signs, optic nerve compression, and in some cases, a progressive muscle scarring with ocular misalignment (**RADIATION THERAPY IS NOT PERFORMED ON DIABETICS, AND IT IS USED ONLY WITH EXTREME CAUTION IN PREGNANT WOMEN**). Mrs. Bush (prior First lady) had this treatment.

6. ORBITAL DECOMPRESSION

Despite the effectiveness of medications, radiotherapy, or a combination of both, there are some people who continue to have threatened compression of the optic nerve with visual loss. In these people, the last resort to salvage vision is orbital decompression, which consists of enlarging the eye socket by opening up some of the sinuses behind the eye, thus relieving the pressure on the optic nerve. This procedure is more commonly used to allow the eye to settle back in cases of extreme protrusion, but a number of special variations on this procedure may be used to prevent blindness.

TREATMENT IN THE INACTIVE PHASE

After you have passed into the inactive phase of your thyroid eye disease, you may wish to consider a number of procedures to recapture some of your normal eye function and normal appearance.

I. ORBITAL DECOMPRESSION SURGERY

The orbital space may be enlarged by surgical removal of one or more of the bone walls surrounding the eyes and orbital soft tissues. This surgery is performed for certain cases of severe, cosmetically troubling proptosis and eye exposure or vision-threatening nerve compression. Orbital decompression may be complicated by misalignment of the eyes.

2. MUSCLE ALIGNMENT SURGERY

Covering one eye immediately relieves double vision. It does not matter which eye is covered. It may be possible to optically realign eyes with the use of prisms either applied to glasses or ground into lenses, although this may not be effective until things stabilize. When double vision cannot be corrected with prisms, eye muscle surgery may be necessary. In most cases, physicians choose to wait until the double vision is stable. If we operate on a patient who is undergoing progressive change, we may correct them now but have things change within the next few months. Often multiple muscle operations are necessary. It is sometimes not possible to completely remove double vision, but the goal is to remove double vision looking straight ahead and in reading position,

as these are the most important directions of sight. Surgery can straighten eyes to allow single vision in straight gaze and down gaze. This procedure is often delayed for six or more months after the active stage has died out to be sure that no further progression in muscle scarring is occurring. Muscle alignment is usually performed after completion of anti-inflammatory medications, X-ray therapy, or orbital decompression. Nationally, this complication occurs on average 33% of the time. Using advanced techniques, we see this complication roughly 5% of the time and are often able to predict which patients will have this problem. In selected individuals, we are able to perform a surgery called "Small Bone Decompression" and have not had a single case of eye misalignment with this technique.

3. COSMETIC EYELID SURGERY

The upper eyelid may be lowered or the lower eyelid may be raised in cases of eyelid retraction with "stare". Eyelid surgery can often improve the appearance of a proptotic (sticking out) eye and avoid the need for orbital decompressive surgery. Blepharoplasty (surgical removal of redundant thickened skin and fat bulges from the eyelids) may also be recommended. These operations are performed at least six months after the active inflammation has subsided and after any necessary muscle alignment has been completed.

FREQUENTLY ASKED QUESTIONS

The doctors tell me they fixed my thyroid and that it is now normal. Why are my eyes acting up?

In Graves' disease the thyroid gland is stimulated by the immune system to secrete too much hormone. This excess hormone results in nervousness, palpitations, weight loss, diarrhea, tremors, and a feeling of being hot all the time. Treatment is aimed at limiting the thyroid gland's ability to make thyroid hormone. This may be done with medications, surgery, or radioactive iodine; usually resulting in normalization of thyroid production (occasionally requiring thyroid replacement). This does not, however, affect the primary auto-immune process and the immune system may continue to target other tissues; in particular the extraocular muscles. Orbital symptoms may even worsen following treatment with radioactive iodine. The eye and orbit changes must be treated separately as outlined.

The steroids made my eyes much more comfortable. Can't I just continue taking them?

Steroid therapy may be effective in halting the inflammatory phase of thyroid orbitopathy and partially shrinking the muscle swelling. Steroid side effects are very common with continued treatment. If there are still problems with eye movements (double vision), exposure problems (irritation and foreign body sensation), or decreased vision then surgery should be considered.

Why can't you fix my eyelids now?

Eye muscle surgery on the vertically acting muscles may change the eyelid position. Thus we do not want to perform eyelid surgery until we have done any possible muscle surgery.

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