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Graves' Disease – Predicting Outcomes

The most important aspect of predicting outcomes in ophthalmic Graves' disease is to recognize the two distinct patterns of presentation associated with it.

The first pattern (which we refer to as Type I Graves' disease) appears to be an orbital proliferative process leading to increased fat compartments in the orbit without significant disruption of extraocular muscle (EOM) function. These patients may present with proptosis, eyelid retraction, corneal exposure, and dramatic alteration of midfacial appearance. Type I disease notably does not present as an inflammatory process, and does not lead to cicatrization of EOM's with diplopia. Proptosis and eyelid retraction are highly variable from mild to severe, and Hertel measurements up to the mid 30's. In the more severe Type I patients, spontaneous prolapse of the globe may be seen. Symptoms in Type I disease arise from proptosis and eyelid retraction, with corneal exposure. Compressive neuropathy has not been observed in Type I disease, regardless of severity.

The second pattern of presentation in ophthalmic Graves' disease is predominately an inflammatory and cicatricial process which we refer to as Type II Graves' disease. This is characterized by external inflammatory signs such as chemosis and hyperemia accompanied by asymmetric proptosis and eyelid retraction, cicatricial myopathy with diplopia and pressure pain phase lasting up to six to nine months, inflammation subsides, but restrictive diplopia, proptosis and eyelid retraction remain. Compressive optic neuropathy is common in Type II disease, perhaps as high as 36 percent.[1]

Type II disease is even more strongly associated with cigarette smoke than is Type I disease (89% are active smokers at time of presentation).[2]

Since compressive optic neuropathy has only been associated with Type II disease, the diagnostic recognition of Type I versus Type II disease becomes critical. Patients with inflammatory orbitopathy and cicatricial myopathy with large EOM's at the apices should be followed very closely and treated aggressively, anticipating the likelihood of optic neuropathy. Patients with Type I disease also often require intervention but for the indications of corneal risk from exposure.

Treatment of Ophthalmic Graves' Disease

Type I patients may be treated supportantly with ocular lubricants and nocturnal moisture chambers. When exposure keratopathy becomes clinically significant, surgical alternatives, such as orbital decompression to lessen proptosis and eyelid retraction repair to prevent lagophthalmos and exposure, are usually quite successful. Notably, diplopia following decompression surgery in Type I patients is extremely rare, and should not be a consideration in this population.

We treat the acute inflammatory symptoms of Type II disease with pulse high dose prednisone, preferring oral to intravenous administration. We recommend orbital decompression relatively early in the course of Type II disease in view of the high risk of optic neuropathy, as well as our and others' observations that orbital decompression combined with pretreatment with steroids is highly effective in significantly reducing the acute orbital inflammation and providing protection from optic neuropathy.[1] We have also observed that while strabismic angles may change following decompression in Type II patients, overall ductions significantly improve and may improve final outcomes of strabismus surgery in Type II patients. We may rarely use radiation therapy as an adjunct to surgical intervention in unusually severe cases.

Rituximab

Rituximab is a new addition to treatment options in ophthalmic Graves' Disease. Rituximab has been shown in few and small series to decrease orbital inflammation in Graves' disease, for up to twelve months following administration. Rituximab has only been noted to decrease proptosis by 1.5 mm in one study, so it is not known whether Rituximab will be as effective in decreasing optic neuropathy as is orbital decompression. That observation, in addition to the very high cost of the drug (up to $20,000), may limit the eventual role of Rituximab.

References

[1] Kikkawa, D et al: *Reduction of orbital inflammation following decompression for thyroid-related orbitopathy.* BioMed Research International, Vol 2013, ArticleID 794984

[2] Nunery, W.R et al: *The association of cigarette smoking with clinical subtypes of ophthalmic Graves' disease.* Ophthalmic Plastic and Reconstructive Surgery. 9(2):77-82, 1993.

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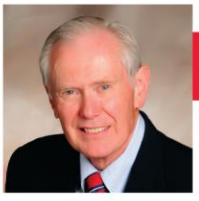

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