

EUGOGO protocol for assessment of Graves' orbitopathy and completion of Case Record Form

Introduction

This protocol has been developed by EUGOGO for the evaluation of patients with Graves' orbitopathy (GO). There are two sets of forms, one for routine clinical use and one for multicentre scientific studies. Both sets assume some background knowledge and experience of GO on behalf of the clinician using them, who can be either an ophthalmologist or endocrinologist with some training in ophthalmology.

The present document sets out the detailed assessment protocol to facilitate both more accurate future comparison, and comparison with others. It should be used in conjunction with the colour atlas which can be downloaded separately for use in clinics, or accessed via the hyperlinks below.

It is hoped that in time this system will be merged with other systems to produce a protocol with worldwide acceptance.

Scoring rules

1. If in doubt, or if features or measurements fall between scores, **always** choose smaller or milder option.

2. Unless otherwise stated, record all dates as mm / yyyy or season / yyyy. In the northern hemisphere spring = 'sp' i.e. March, April, May; summer = 'su' i.e. June. July, August; autumn = 'au' i.e. September, October, November; winter = 'wi' i.e. December, January, February. Hence onset in mid 2010 would score as su/2010

3. The decision as to whether a patient has "active" GO is based on assessment of symptoms, soft tissue signs and change in severity, and influenced by disease duration. However eyelid swelling, eyelid erythema and conjunctival redness should only be scored if thought to be due to active GO. Hence the wider picture should be taken into account when scoring.

SYMPTOMS

1. Pain Pain should only be scored if present for more than a few seconds and more often than just occasionally

2. *Diplopia* Patients who wear **full time** prisms score as "constant" on the subjective diplopia score regardless of whether the diplopia is relieved by prisms.

EXAMINATION

- 1. *Visual acuity* Record corrected distance acuity ideally using Logmar chart. If Snellen is used, record in decimal format.
- 2. **Pupil assessment** Test the relative afferent pupil defect (**RAPD**) with distance fixation, and with care to maintain alignment of your light relative to the patient's visual axis (especially in strabismus).
- 3. **Colour vision** Ideally use HRR plates no.5-10, but otherwise use Ishihara plates. If using either HRR or Ishihara plates, then up to 2 errors still score as 'normal'.
- Soft tissue signs (a e form part of <u>Clinical Activity Score</u>: CAS) Note that conjunctival redness can change rapidly and should be scored prior to touching the patient.

a) <u>eyelid swelling</u> If swelling varies between upper and lower eyelid of an eye then use the more severe lid to score that eye. Only score swelling thought to be due to active GO; i.e. appearance should have changed with disease, and fat prolapse is not scored as 'swelling'. Note only "moderate" or "severe" scores are recorded as CAS positive: "mild" is negative.

Swelling may be very superficial (plates 1a(i),1b(i),1c(i)) or deeper and more like thickened skin (plates 1a(ii), 1b(ii), 1c(ii)). The latter is commoner in younger individuals and harder to distinguish from fatty prolapse, however fat prolapse is less diffuse and individual fat pads are generally distinguishable.

Scoring:

Mild = the patient may be aware of changed appearance, however none of the features defining moderate or severe swelling are present: CAS negative

Moderate = definite swelling but no lower eyelid festoons and in upper eyelid the skin fold becomes angled fold on 45° downgaze: CAS positive

Severe = lower eyelid festoons OR upper lid fold remains rounded on 45° downgaze: CAS positive

- b) <u>eyelid erythema</u> If erythema varies <u>between</u> upper and lower eyelid of an eye then use the more severe lid to score that eye. Only score redness due to active GO, i.e. don't score blepharitis and don't score if whole face is red.
- c) <u>conjunctival redness</u> Assess without slit-lamp 1 meter from patient. Only score redness due to active GO. Score as CAS negative if only "equivocal" or "mild".
- d) <u>chemosis</u> Use a slit-lamp at 60° midway between limbus and lateral canthus to score patient. Distinguish true chemosis (present >1/3 of vertical aperture or prolapsing anterior to grey line of eyelid = CAS positive) from the redundant folds of conjunctiva (conjunctivochalasis) that are common in older subjects (≤ 1/3 of vertical height of aperture and always behind greyline of eyelid = CAS negative).
- e) <u>caruncle and plica inflammation</u> The caruncle is normally yellowish pink and lies medial to plica, which is normally pink. Proptosis makes the caruncle more prominent and may be visible on eyelid closure, unlike the plica.

Score "yes" if caruncle inflamed, but ignore whether visible through closed eyelids or not.

Score "yes" if plica inflamed **or** prolapsed through closed eyelids. A "yes" for either caruncle or plica = CAS positive.

- 5. Eyelid measurements Ask patient to relax, distance fixate in primary gaze, and ensure head in normal position: an imaginary line between the top of the ear and the lateral canthus should be horizontal. If primary distance fixation impossible without head posture, indicate this with * on case record form. If strabismus present then occlude contralateral eye to ensure distance fixation prior to measurement of *palpebral aperture* and *eyelid retraction*. Corneal limbus forms reference point for *eyelid retraction*, so measurements may be + or (click for details). Also measure *levator function* plus
 - a) *lagophthalmos* use pentorch to assess exposure when eyes closed.
 - b) **Bell's phenomenon** (click here for method)

- 6. **Proptosis** (<u>click here</u> for technique). Optimise reproducibility by using same Hertel instrument, same intercanthal distance (ICD)and ideally same observer on each occasion.
- 7. Motility Motility is assessed without spectacle prisms. Head posture is noted first. Compensatory head posture is noted, but corrected prior to prism cover test at distance using a light target. Torsion, monocular ductions and the field of binocular single vision are also assessed. Monocular ductions are assessed using an arc or bowl perimeter, preferably by the same examiner. Careful patient positioning reduces errors induced by head tilt or rotation. Uniocular excursions are plotted for each eye along horizontal and vertical axes using subjective responses where possible while verifying foveal fixation.
- 8. Cornea
 - a) Corneal integrity is assessed using fluorescein
 - b) Risk of corneal breakdown is also assessed (click here for details).
- 9. Intraocular pressure applanation tonometry in primary gaze
- 10. **Optic neuropathy** This is judged on basis of **disc appearance**, plus **acuity**, **afferent pupil defect** and **colour vision**, plus ancillary tests if necessary e.g. visual fields, visual evoked potentials and imaging of orbital apex. Until further data is available, optic neuropathy may be assumed to be present if there is disc swelling, or if two of the other **clinical** features are present. Impaired colour perception carries more weight than other features except disc swelling, and in each instance other causes for the abnormalities should be excluded.
 - a) disc *swelling* or *atrophy* should only be scored if thought due to GO: otherwise score as "*other*" abnormality.