Eurylepharon

JAMES A. KEIPERT
Royal Children's Hospital, Melbourne, Australia

Eurylepharon is defined by Duke-Elder (1964) as a symmetrical enlargement of the palpebral aperture occurring as a primary abnormality associated with large eyelids, and by Waardenburg, Franceschetti, and Klein (1961) as bilaterally symmetrical large eyelids opening much wider than the normal, especially at the lateral but also at the medial end. Waardenburg and others (1961) also stated that the presence of the condition at birth and the absence of any primary ocular abnormality distinguished eurylepharon from the secondary enlargement of the palpebral aperture which may accompany a variety of ocular abnormalities.

The condition was first described by Desmarres (1854), and subsequent reports in the German and Dutch literature by Schreiber (1924), Lindberg (1928), Seefelder (1930), and Weve (1936) are quoted by Duke-Elder (1964). Only one further case report had been found; the patient described by Gupta and Kumar (1968) did not show abnormal width of the palpebral apertures, and might be considered by some as a case of normal long lid fissures rather than eurylepharon.

The condition may be inherited. Waardenburg and others (1961) often found long lid fissures inherited through two generations with no difference between sexes, and had also seen eurylepharon in a father and daughter.

The average length of the lid fissures at different ages is given by Duke-Elder and Cook (1963) and Waardenburg and others (1961); it increases from 18.35 mm. at birth to 29.68 mm. at 24 to 26 years, one half of this increase occurring in the first 4 years of life. The length is slightly less for the female.

Case report
The typical clinical features of eurylepharon were present at birth, and are shown at the age of 4 months (Fig. 1a, b). Mild ectropion of the lower lids was present, especially laterally. At the age of 2 months he was not noted to blink, but he could close his eyes in sunlight or strong light, and he could keep them closed while he remained in such a situation. His eyes remained open during sleep. His eyes were therefore rarely closed, despite which corneal or conjunctival irritation or infection did not occur.

The corneae appeared to be normal in size. Ocular tension was normal. At the age of 6 months the palpebra apertures measured 3 x 1.7 cm. (R) and 2.9 x 1.6 cm. (L). At the age of 7½ months, exophthalmometer readings were 16.5 mm. (R) and 14 mm. (L).

At the age of 8½ months bilateral tarsorrhaphy was performed with a good result (Miss Lena McEwan). The cosmetic appearance was improved (Fig. 2a, b).

He still sleeps with the lids open almost to the width of the iris, and the eyeball rotated upwards. With ordinary blinking the eyelids do not meet. When he screws the lids up, they meet on the left side but remain slightly apart on the right.

At the age of 3½ years, refraction showed the right eye to be normal and a minor degree of hypermetropic astigmatism in the left. He has made normal physical and intellectual progress, and is contented.

Discussion
The palpebral portion of the orbicularis oculi muscle can act under voluntary control, or may act reflexly, closing the eyelids gently when blinking or during sleep. It also holds the lids in contact with the globe. The orbital portion more frequently acts under voluntary control, and closes the lids more forcibly. The lacrimal portion draws the eyelids and lacrimal papillae medially, and exerts traction on the lacrimal fossa. The palpebral portion is opposed by the levator palpebrae superioris, and the orbital portion by the occipitofrontalis (Warwick and Williams, 1973; Last, 1968).

A specific cause for eurylepharon is not suggested in any of the previous reports in the English literature. Gupta and Kumar (1968) say that tension of the skin, pull of the platsmas, defective separation of the lids, and localized displacement of the lateral canthi have been mentioned in the literature as possibly explaining the abnormal enlargement of the palpebral aperture, but they state that the cause is still unknown.

The most obvious features in eurylepharon are the increased length and width of the palpebral apertures, the slackness of the eyelids, and the decreased width and rather wasted or hollowed-out appearance of the eyelids. These features, together with the absence or deficiency of blinking, the failure to close the eyes during sleep, the tendency to lateral displacement of the palpebral aperture, and the mild ectropion of the lower eyelids, would all appear to be consistent with a congenital hypoplasia or absence.
FIG. 1(a) Patient aged 4 months, full face
(b) Patient aged 4 months, looking down

FIG. 2(a) Patient aged 8½ months, after tarsorrhaphy, full face
(b) Patient aged 8½ months, looking down

of the palpebral and lacrimal portions of the orbicularis oculi muscle as a cause of this interesting and rare congenital anomaly. This could be proved only by biopsy of the eyelid, but unfortunately this would not appear to offer any possibility of direct surgical correction.

Summary
The rare congenital anomaly of euryblepharon is reported in a male infant. It is suggested that euryblepharon is due to congenital hypoplasia or absence of the palpebral and lacrimal portions of the orbicularis oculi muscle.

I am grateful to Dr. David Gale for making the diagnosis of euryblepharon, and for the results of tonometry and refraction, to Miss Judith Quilter, Librarian, The Royal Victorian Eye and Ear Hospital, and the Australian Medlars Service for perusal of the literature, and to Mr. H. G. Berkshire for the photographs.

References


Additional references
WOLTER, J. R. (1972) ibid., 9, 179