

CASE STUDIES

Accessory Levator Muscle of the Upper Eyelid: Case Report and Review of the Literature

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The authors describe a supernumerary muscle in each orbit of an elderly male subject. There appear to be no previous reports of this muscle; most reports of anomalies of extraocular muscles describe hypoplasia or aplasia. Thirty-five formalin-fixed cadavers assigned to medical students for dissection were studied. The orbits were dissected by a superior approach which involved removal of the orbital plate of the frontal bone and the superior orbital margin. A supernumerary extraocular muscle was seen in each orbit of one cadaver, located between the superior oblique and levator palpebrae superioris muscles. It originated on the inferior surface of the lesser wing of sphenoid bone and was inserted into the skin of the medial one-third of the upper eyelid. It was innervated by a branch from the superior division of the oculomotor nerve. The insertion of the muscle into the upper eyelid produced a crease running obliquely upwards and medially, from the junction of the medial one-third and lateral two-thirds of the lid margin, towards the medial part of the superior orbital fold. The authors suggest the name *levator palpebrae superioris accessorius* for this muscle in view of its topography and action as tested in the cadaver. The significance of the findings is discussed and the literature on the development of the muscles supplied by the oculomotor nerve is reviewed. Clin. Anat. 11:410–416, 1998. © 1998 Wiley-Liss, Inc.

Key words: supernumerary extraocular muscle; anomalous eyelid muscle

INTRODUCTION

Current anatomical texts (Hollinshead, 1968; Doxanas and Anderson, 1984; Hollinshead and Rosse, 1985; O’Rahilly, 1986; Moore, 1992; Williams et al., 1995) describe six extrinsic ocular muscles, a levator of the upper eyelid (part of which is the superior tarsal muscle of Müller) and the orbitalis muscle (a layer of smooth muscle bridging the inferior orbital fissure).

Abnormalities of the extraocular muscles are rare and most accounts describe unilateral or bilateral absence of individual muscles. Thus, congenital absence of the superior rectus muscle has been reported by Weinstock and Hardesty (1965)—2 cases, Cuttone et al., (1979)—1 case and Diamond et al., (1980)—3 cases. Bilateral absence of inferior oblique muscle was reported to be absent on the right side in one case by Diamond et al. (1980) and on the left side by Ingham et al. (1986)—1 case, and Taylor and Kraft (1997)—1 case. Numerical aberrations of the extraocular muscles are commonly associated with syndromes of premature cranial bone stenosis, notably craniofacial dysostosis (Crouzon’s disease), acrocephalosyndactyly (Apert’s syndrome), and oxycephaly (Diamond et al., 1980).

Other reported abnormalities of extraocular muscles include hypoplasia (mainly congenital absence of the tendon) of superior oblique muscle (Diamond et al., 1980; Botelho and Giangiacomo, 1996) and congenital cysts in the common sheath of the superior rectus and levator palpebrae superioris muscles (Rose and O’Donnell, 1995).

There appears to be no report of numerical variations involving the levator muscle of the upper eyelid. Ballen and Rochkopf (1987) noted in a patient with congenital upper eyelid retraction that the medial and lateral horns of the levator palpebrae superioris were “dense” at the level of the superior transverse (Whitnall’s) ligament. Their report was corroborated by Collin et al. (1990) who examined 22 patients with congenital lid retraction.

Goldberg et al. (1992) reported a high resolution magnetic resonance imaging technique that resolved

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fine details of eyelid structure, such as the orbital septum, levator palpebrae superioris, superior tarsal (Müller's) muscle and tarsal plate. There was no mention of an accessory levator muscle of the upper eyelid or its aponeurosis in their series of patients.

The chance finding of an additional extraocular muscle during a class dissection session prompted a detailed and systematic search of other cadavers assigned to students for the presence of anomalies of extraocular muscles. The aim of this report is to describe the morphology of the supernumerary muscle and to evaluate its possible significance.

MATERIALS AND METHODS

Thirty-five cadavers assigned to medical students for dissection were studied. They were 65–80 years of age, eight female and twenty-seven male. They had been selected at random and had been embalmed soon after death with a mixture of 10% formaldehyde, glycerol, methylated spirits, and 10% phenol in tap water.

Superficial Dissection of the Eyelids

The eyelids on one side of each cadaver were dissected superficially to display the orbicularis oculi

muscle; the eyelids on the opposite side were initially left intact. Before dissection of the eyelid, features of the canthi, puncta, lid margins, folds, and creases were noted. The orbicularis oculi muscle was then dissected and the arrangement of the fibers of its palpebral and orbital parts studied.

Dissection of the orbit

The orbit was dissected from above. The orbital plate of the frontal bone was fractured by chiseling, and the roof of the orbit removed with a rongeur, care being taken to preserve the periorbita. The skin and tissue overlying the superior rim of the orbit was removed *en bloc*. The superior bony orbital margin was then removed by making two vertical cuts through it (Fig. 1) with an oscillating saw. The bone between the two incisions was freed from the underlying structures. The periorbita was opened, care being taken not to damage the frontal nerve, and the orbital fat was removed piecemeal to expose the superficial structures of the orbit. The levator palpebrae superioris was then transected close to the apex of the orbit and reflected anteriorly. The posterior one-third of the superior rectus muscle was carefully dissected to expose the superior division of the oculomotor nerve. The superior rectus muscle was transected at about

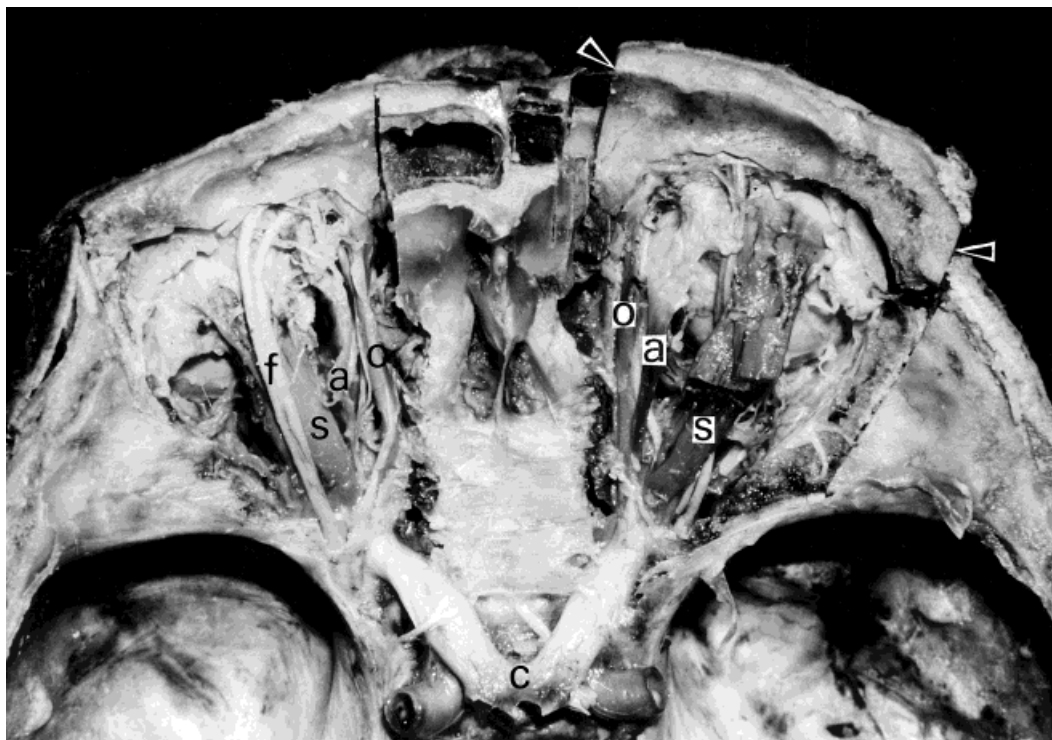


Fig. 1. Superior view of the right and left orbits after removal of their roofs. Arrowheads (►): vertical cuts through the superior margin of the rim; f: frontal nerve; o: superior oblique muscle; s: levator palpebrae superioris; a: levator palpebrae superioris accessorius; c: optic chiasma.

the middle of its belly, and its proximal and distal halves were reflected to expose the trunk of the oculomotor nerve and other deeper placed structures. Measurements of the muscles were made using a pair of dividers and tape measure. Values were recorded to the nearest millimeter.

OBSERVATIONS

The following description relates to one male cadaver in which an additional muscle was noted in the superior part of the orbit, medial to, and in the same plane as, the levator palpebrae superioris. It was present bilaterally, although the right one appeared to be better developed. When the muscle belly was pulled on with a pair of forceps, the upper eyelid was elevated; this movement was more marked on the medial part of the eyelid. Owing to the apparent similarity of its attachments to those of the levator palpebrae superioris, we suggest the name *levator palpebrae superioris accessorius*. There was no evidence of cranial malformation nor of previous orbital or eyelid surgery. The four recti as well as the superior and inferior oblique muscles were normal.

External Features of the Eyelids

The palpebral fissure was of the caucasian type. The inferior orbital, nasojugal, and malar folds of the

lower eyelid (Fig. 2) appeared normal for the age of the subject. The superior orbital fold extended the full width of the upper eyelid. The skin of the lower part of the lateral two-thirds of the upper eyelid was loose and overhung the lid margin, covering the roots of the eyelashes. A well-formed, obliquely placed crease of the upper eyelid began at the junction of the medial one-third and lateral two-thirds of the margin and extended upwards and medially towards the superior orbital fold (Fig. 2). The skin of the medial third of the upper eyelid was more adherent to the subcutaneous tissue and did not fold over the margin of the lid or roots of the eyelashes.

Levator Palpebrae Superioris Muscle

The levator palpebrae superioris muscle was normal on both sides. It took origin from the inferior aspect of the lesser wing of sphenoid and its deep fibers were adherent to the superior surface of the common tendinous ring of the recti muscles. The muscle ran anteriorly superficial to the superior rectus muscle and deep to the frontal nerve. Anteriorly, it passed superficial to the tendon of the superior oblique muscle, and formed a wide aponeurosis distal to the superior transverse (Whitnall's) ligament. The superior transverse ligament is a check ligament passing from the upper surface of the levator palpebrae superioris to the superior margin of the orbit. The aponeurosis split into

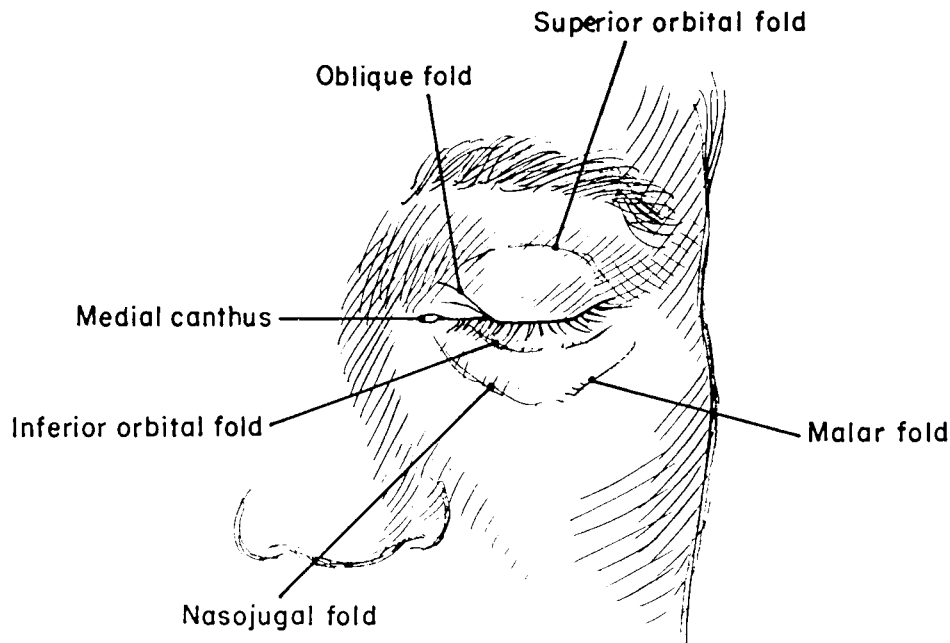


Fig. 2. A drawing of the left side of the face to show the creases and folds of the eyelids. Note that the lateral two-thirds of the upper eyelid has a fold of redundant skin that overhangs the margin of the lid and covers the roots of the eyelashes of the upper eyelid. A deep crease

at the junction of the medial one-third and lateral two-thirds of the upper eyelid gave rise to an oblique fold. At dissection, it was noted that the crease was produced by the insertion of the accessory levator muscle.

superficial (palpebral) and deep (tarsal) lamellae. The palpebral part blended with the orbital septum. Its fibers were traced through the palpebral part of orbicularis oculi to their insertion in the skin of the whole width of the upper eyelid. Fibers from the medial horn of the aponeurosis passed deep to the nasal pad of orbital fat and continued medially to insert on the upper and posterior surfaces of the medial canthal ligament. The lateral horn of the aponeurosis passed between the two parts of the lacrimal gland to be inserted into the lateral orbital (Whitnall's) tubercle and lateral canthal ligament. The deep lamella of the aponeurosis (forming the superior tarsal muscle) was inserted into the upper border of the tarsal plate.

Levator Palpebrae Superioris Accessorius

This new and previously undocumented muscle was found on both right and left sides. It lay between the levator palpebrae superioris and the superior oblique muscles (Figs. 1 and 3a); it took origin at the apex of the orbit medial to, and near, the origin of the levator palpebrae superioris muscle (Fig. 3a), by means of a short, slender tendon mainly from the superior surface of the common tendinous ring of the recti muscles. Part of the tendon was adherent to the fibers of the levator palpebrae superioris muscle, arising from the lesser wing of sphenoid. The proximal part of the muscle was crossed superiorly, immediately distal to the apex of the orbit, by the trochlear nerve running medially towards the superior oblique muscle (Figs. 1 and 3a). The nerve did not give any branches to the accessory muscle. The muscle passed distally, in the superficial part of the orbit, medial to the levator palpebrae superioris. It spread out in the distal half of the orbit to form a wide aponeurosis that continued anteriorly, passing deep to the tendon of superior oblique muscle and the superior transverse ligament (Figs. 3a,b). It descended towards the deep surface of the upper eyelid, superficial to the nasal pad of orbital fat, as a single-layered aponeurotic sheet. Within the eyelid, it was superficial to the medial horn of the aponeurosis of the levator palpebrae superioris. The two aponeuroses were separated from each other,

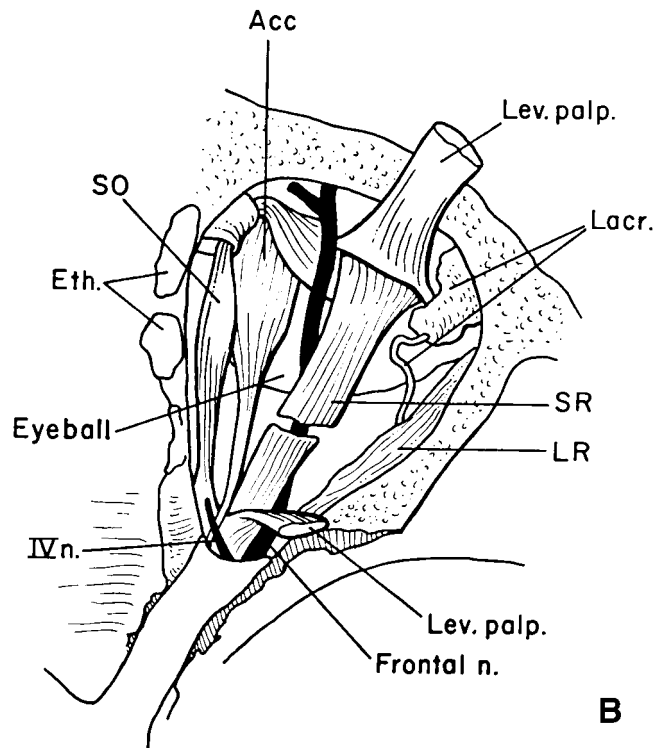
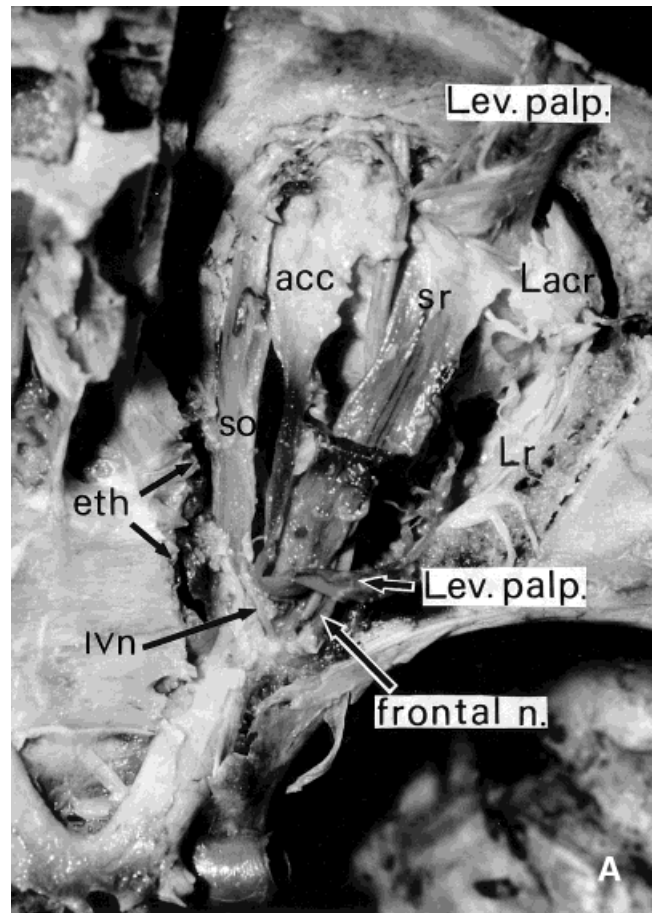


Fig. 3. A: Superior view of the dissected right orbit. **B:** Diagram showing the main features of A. SO: superior oblique muscle; Eth (eth): ethmoidal air sinuses; Lev. palp: levator palpebrae superioris muscle, cut and reflected to show the subjacent superior rectus muscle, SR (sr); Acc (acc): levator palpebrae superioris accessorius; IVn: trochlear nerve crossing the proximal parts of the levator and accessory levator muscles of the upper eyelid to reach the superior oblique muscle. Lacr: lacrimal gland and artery; LR (Lr): lateral rectus muscle. The frontal nerve was displaced to enable the relationships of the superior rectus and levator muscles to be brought to the forefront.

proximally by extensions of the nasal pad of fat, and within the eyelid by loose connective tissue. The aponeurosis of levator palpebrae superioris accessorius fused with the medial part of the orbital septum. Tendinous fibers passing distally from this fusion intermingled with the palpebral part of orbicularis oculi and then inserted into the skin of the medial third of the upper eyelid above the upper margin of the tarsal plate. The insertion of the muscle into the eyelid was associated with a deep, obliquely placed upper eyelid crease (Fig. 2).

On both right and left sides, the accessory muscle was innervated by a branch from the superior division of the oculomotor nerve. A common trunk came off the superior division of oculomotor nerve and ran superiorly, crossing the medial border of the superior rectus muscle. Near the medial edge of levator palpebrae superioris, it bifurcated to give branches which entered the deep surfaces of the proximal parts of levator palpebrae superioris and the accessory muscle respectively.

Morphological and Topographical Differences Between Levator Palpebrae Superioris and Levator Palpebrae Superioris Accessorius

The size of the muscles. The two muscles differed in size. The depth of the orbit (distance from apex to the superior orbital margin along the axis of the orbit) in this subject was 35 mm. Eighteen millimeters from the apex, the widths of the levator palpebrae superioris accessorius and levator palpebrae superioris muscles were approximately 3 mm and 7 mm, respectively; at the level of the superior transverse ligament (33 mm from the apex), the widths of their aponeuroses were approximately 10 mm and 15 mm, respectively. The thickest part of the fleshy belly of the accessory muscle was 0.5 mm while the fleshy part of the levator palpebrae superioris was about 1.0 mm thick until it became aponeurotic.

Relationship to the tendon of superior oblique muscle. The aponeurosis of the levator palpebrae superioris accessorius muscle passed deep to the superior oblique tendon as the latter emerged from its trochlea, whereas the levator palpebrae superioris (lying superior to superior rectus muscle) passed superficial to the distal part of the superior oblique tendon as it approached its insertion into the eyeball.

Aponeuroses and insertions. The aponeurosis of the accessory muscle differed from that of levator palpebrae superioris in having only one lamella which was inserted into the skin of the eyelid above the upper border of the tarsal plate. It had no attachment to the tarsal plate.

DISCUSSION

This is, to our knowledge, the first report of an additional extrinsic ocular muscle acting on the upper eyelid. The muscle was bilateral. Although its occurrence is intriguing, being an isolated case, no firm conclusions can be drawn on its significance. Judging from its relationship to levator palpebrae superioris and superior oblique muscles, and its mode of insertion into the upper eyelid, the muscle seems to be a separate structure. Its contraction would contribute to the elevation of the medial part of the upper eyelid hence the suggestion to call it *levator palpebrae superioris accessorius*. Three questions arise: Is it a vestigial muscle or does it signify an adaptation to some special need? Could it be merely a developmental aberration?

The apparent lack of previous reports of this muscle in lower animals makes it unlikely that it is vestigial. Yapp (1965) showed that lower vertebrates had six extra-ocular muscles, homologous to human extrinsic eye muscles, as well as mobile eyelids, but did not possess levator palpebrae superioris muscle. He described the eyelids as consisting of muscular folds of skin. Comparative morphologic evidence adduced by Holmes (1975) confirmed the accounts of Yapp (1965) in fish, amphibia, reptiles, birds, and quadruped mammals. Williams et al. (1995) suggested that levator palpebrae superioris was a phylogenetically new muscle, formed as a later delamination from the superior rectus muscle to serve the upper eyelids in the higher tetrapods. The weight of comparative anatomical evidence, therefore, appears to rule out the possibility that the levator palpebrae superioris accessorius is a vestigial muscle.

The extrinsic ocular muscles develop from three pro-otic somites (Neal, 1918), while their connective tissue components are derived from neural crest cells. Gilbert (1952) showed that those extraocular muscles innervated by the oculomotor nerve developed from the most rostral of the three, the premandibular somite which formed from mesoderm originating in the prechordal plate. Superior rectus muscle appeared on the 24th day of gestation whereas medial and inferior recti, and inferior oblique muscles were formed between the 28th and 30th days of gestation. Levator palpebrae superioris was the last to form, appearing early in the eighth week by delamination from the medial aspect of the superior rectus muscle (Gilbert, 1957). The oculomotor nerve reached the vicinity of the developing eye early in the fifth week and quickly innervated these muscles.

Cuajunco (1942) suggested that in man, all skeletal muscle formation is completed by mid-fetal stage and their innervation established by the 15th week of

gestation. No new muscles are formed after this period. This implies that the accessory muscle probably differentiated before the eyelids became functional, and makes it very unlikely that it was formed to subserve an intercurrent functional need.

There is evidence to suggest that some cases of eyelid and extraocular muscle disorders or anomalies are hereditary. Gillies et al. (1995) reported that some patients with congenital lid retraction expressed "an unusual dominantly inherited variant of congenital extraocular muscle fibrosis syndrome." Botelho and Giangiacomo (1996) also showed that congenital superior oblique palsy (characterized by hypoplasia or absence of the tendon of superior oblique muscle) was inherited as an autosomal dominant characteristic.

Although there is no direct evidence on the developmental origin of the levator palpebrae superioris accessorius, its innervation from the oculomotor nerve suggests that it probably develops, in common with the levator palpebrae superioris, from the premandibular somite. Diamond et al. (1980) re-opened the debate on the developmental origin of the levator palpebrae superioris muscle. In two of their subjects, who had congenital absence of the superior rectus muscle bilaterally, the levator palpebrae superioris muscles were present, well-developed, and did not exhibit any impairment of function. Diamond et al. (1980) surmised that the superior rectus muscle had undergone secondary atrophy after the differentiation of the levator palpebrae superioris. The apparent inconsistency could also be explained by the finding of Sevel (1981) that the different parts (origin, belly, and insertion) of individual extraocular muscles, including the levator palpebrae superioris muscle, developed simultaneously from multiple foci of mesodermal condensations and fused later on during development. This theory could also clarify the occurrence of hypoplasia of extraocular muscles, e.g., congenital absence of the tendon of superior oblique muscle. More work is needed to clarify the developmental origin of the extraocular muscles, especially muscles of the eyelid, and it might also shed some light on the origin of the levator palpebrae superioris accessorius muscle.

The possibility of the accessory muscle being a developmental aberration cannot be ruled out easily, although its bilateral occurrence, its clear-cut topographical differences from the levator palpebrae superioris and the presence of a well-established innervation appear to make it unlikely. The present evidence is insufficient to enable any firm deductions to be made concerning the significance of this accessory muscle. The question arises, is there a need for an additional levator muscle of the eyelid? The levator complex (consisting of the levator palpebrae superi-

oris, its aponeurotic extensions to the eyelid and the superior tarsal muscle of Müller) supports the normal eyelid, and is active continuously for long periods during waking hours (except during blinking) to keep the eyelids elevated. There are no reports to suggest that the muscle achieves full relaxation during blinking. Mustardé (1989) showed that the involuntary contraction of Müller's muscle was largely responsible for keeping the eyelid raised during waking hours, helped by the reciprocal relaxation of the orbicularis oculi muscle. Information is incomplete on the factors involved in the control of the continuous activity of the levator in keeping the eyelid raised (Conn, 1995). By varying the degree of elevation of the eyelid, the levator palpebrae superioris influences the amount of light entering the eye, and an accessory levator could help in the fine control of this function. Movements and posture of the eyes and eyelids are also important in human communication by body language. The respective roles played by the striated and non-striated parts of the levator palpebrae superioris in communication are well known. Through its attachment to the medial part of the upper eyelid, the levator palpebrae superioris accessorius could contribute an additional dimension to eye communication.

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REFERENCES

- Ballen, P.H. and L. Rochkopf 1987 Congenital retraction of the upper lid. *Ophthalmic Surg.* 18:689-690.
- Botelho, P.J. and J.G. Giangiacomo 1996 Autosomal-dominant inheritance of congenital superior oblique palsy. *Ophthalmology* 103:1508-1511.
- Collin, J.R., L. Allen and S. Castronuovo 1990 Congenital eyelid retraction. *Br. J. Ophthalmol.* 74:542-544.
- Conn, P.M. 1995 The Oculomotor System. In *Neuroscience in Medicine*. Philadelphia: Lippincott, pp. 249-260.
- Cuajunco, F. 1942 Development of the human motor end-plate. *Contrib. Embryol. Carnegie Inst. Washington* 30:127-152.
- Cuttone, J., P. Brazis, M. Miller and S. Folk 1979 Absence of the superior rectus muscle in Apert's syndrome. *J. Pediatr. Ophthalmol. Strab.* 16:349-354.
- Diamond, G.R., J.A. Katowicz, L.A. Whitaker, G.E. Quinn and D.B. Schaffer 1980 Variations in extraocular muscle number and structure in craniofacial dysostosis. *Am. J. Ophthalmol.* 90:416-418.
- Doxanas, M.T. and R.L. Anderson 1984 *Clinical Orbital Anatomy*. 1st Ed. Baltimore: Williams and Wilkins, pp. 62-88.

- Gilbert, P.W. 1952 The origin and development of the head cavities in the human embryo. *J. Morphol.* 90:149–187.
- Gilbert, P.W. 1957 The origin and development of the human extrinsic ocular muscles. *Contrib. Embryol. Carnegie Inst. Washington* 36:59–78.
- Gillies, W.E., A.J. Harris, A.M. Brooks, M.R. Rivers and R.J. Wolfe 1995 Congenital fibrosis of the vertically acting extraocular muscles: A new group of dominantly inherited ocular fibrosis with radiologic findings. *Ophthalmology* 102: 607–612.
- Goldberg, R.A., J.C. Wu, A. Jesmanowicz and J.S. Hyde 1992 Eyelid anatomy revisited. Dynamic high-resolution magnetic resonance images of Whitnall's ligament and upper eyelid structures with the use of a surface coil. *Arch. Ophthalmol.* 110:1598–1600.
- Hollinshead, W.H. 1968 The Eyelids, Orbit and Eyeballs. In *Anatomy for Surgeons: Vol. I: The Head and Neck*. New York: Hoeber Medical Division, Harper and Row, pp. 107–181.
- Hollinshead, W.H. and C. Rosse 1985 Orbit and Eye. In *Textbook of Anatomy* 4th Ed. Philadelphia: Harper and Row, pp. 958–986.
- Holmes, E.B.M. 1975 Muscular system. In *Manual of Comparative Anatomy: A Laboratory Guide and Brief Text*. Dubuque: Kendall/Hunt, pp. 141–174.
- Ingham, P.N., S.T. McGovern and J.L. Crompton 1986 Congenital absence of the inferior rectus muscle. *Aus. N.Z. J. Ophthalmol.* 14:355–358.
- Moore, K.L. 1992 The Orbit. In *Clinically Oriented Anatomy*. 3rd Ed. Baltimore: Williams and Wilkins, pp. 702–719.
- Mustardé, J.C. 1989 New horizons in eyelid reconstruction. *International Ophthalmology Clinics* 29:237–246.
- Neal, H.V. 1918 The history of the eye muscles. *J. Morphol.* 30:433–453.
- O'Rahilly, R. (ed.) 1986 The Orbit. In *Gardner-Gray-O'Rahilly Anatomy* 5th Ed. Philadelphia: Igaku-Shoin/Saunders, pp. 646–647.
- Rose, G.E. and B.A. O'Donnell 1995 Congenital orbital cysts associated with the common sheath of the superior rectus and levator palpebrae superioris muscles. *Ophthalmology* 102:135–138.
- Sevel D. 1981 A reappraisal of the origin of human extraocular muscles. *Ophthalmology* 88:1330–1338.
- Taylor, R.H. and S.P. Kraft 1997 Aplasia of the inferior rectus muscle. A case report and review of the literature. *Ophthalmology* 104:415–418.
- Weinstock, F.J. and H.H. Hardesty 1965 Absence of superior recti in craniofacial dysostosis. *Arch. Ophthalmol.* 74:152–153.
- Williams, P.L., L.H. Bannister, M.M. Berry, P. Collins, M. Dyson, J.E. Dussek and M.W.J. Ferguson (eds.) 1995 The extraocular muscles. In *Gray's Anatomy: The Anatomical Basis of Medicine & Surgery*. 38th Ed. New York: Churchill Livingstone, pp. 1353–1359.
- Yapp, W.B. 1965 Sense Organs. In *Vertebrates: Their Structure and Life*. New York: Oxford University Press, pp. 279–321.