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DUANE'S RETRACTION SYNDROME

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The retraction syndrome first described by Stilling in 1887 and Türk in 1896 and more exactly defined by Duane in 1905 on the base of a series of 54 cases manifests the following clinical signs:

1. Marked *limitation or absence of the abduction of the affected eye.*
2. *Narrowing of the palpebral fissure on attempting adduction.*
3. Widening of the palpebral fissure on attempting adduction.
4. *Retraction of the globe on attempting adduction.*
5. Deviation of the affected eye upwards or downwards on adduction.

This pattern as been described by Duane is realized in the majority of the cases. Unilateral manifestation (more frequently on the left than on the right side!) is the rule. Bilateral manifestations are a rare occurence. Most of the patients reveal orthophoria with good binocular functions on looking straight ahead. In order to avoid diplopia some patients unconciously move their eyes, respectively their head into the direction of the paretic external rectus, especially in cases where the primary position of the affected eye is slightly esophoric. The presence of a vertical element in the cases of Duane's-Syndrome is a frequent occurence: downshoot or up-shoot on attempting adduction or downdrift on attempating abduction. Not seldom one can find in a typical Duane's-Syndrome also an additional V- or A-pheonomenon.

The *narrowing of the lid fissure on adduction* (varying from some millimeters up to the total closure) is one of the most constant findings and one of the most important symptoms for the diagnosis of the syndrome. The *retraction* of the globe manifests itself also in various degrees; in this connection it has to be mentionned that the narrowing of the palpebral fissure alone may simulate a retraction of the globe. Changes of the palpebral aperture can also occur without retraction. The adduction of the affected eye which according to Duane and other authors should be also markedly limited, is according to our experience not changed at all in the typical syndrome.

From the typical Duane-syndrome, as it has been described above and as it had been named by us as *Duane I*, many cases differ in their symptomatology, although the phenomena of narrowing of the palpebral fissure on adduction and retraction of the globe with eversion of the inferior lid always are to be found. Such an atypical symptomatology of Duane's-syndrome can be according to our newest observations again divided into two groups: Duane's-syndrome II and Duane's-syndrome III.

Duane's-syndrome II manifests instead of the limitation of the abduction a limitation respectively a paresis of the adduction of the affected eye with intact possibility of abduction. On intention to look into the direction of the "paretic" internal rectus a characteristic narrowing of the palpebral fissure occurs, as well a distinct retraction of the globe. Here too anomalies of the vertical movements as described in the typical Duane-syndrome I can be observed. Binocular single vision may be present especially if a compensatory head posture brings both eyes into orthophoric position. The analysis of the electromyographic findings will show that. Duane II is only a special form of the whole group of Duane's-retraction syndromes, where the pathogenetic principle consists in paradoxial innervation of the affected muscles.

In *Duane's-syndrome III* which recently has been observed by us in two patients, there is an apparent paresis of both the external and the internal rectus of the affected eye. On attempting adduction there results a characteristic narrowing of the palpebral fissure and retraction of the globe. In the primary position of the non affected eye, the contralateral affected eye manifests either a parallel or a slightly divergent position. On looking upwards the divergence increases or a divergence become manifest in the sense of a V-syndrome. According to the electromyographic findings also Duane-syndrome III can be classified within the congenital retraction syndromes.

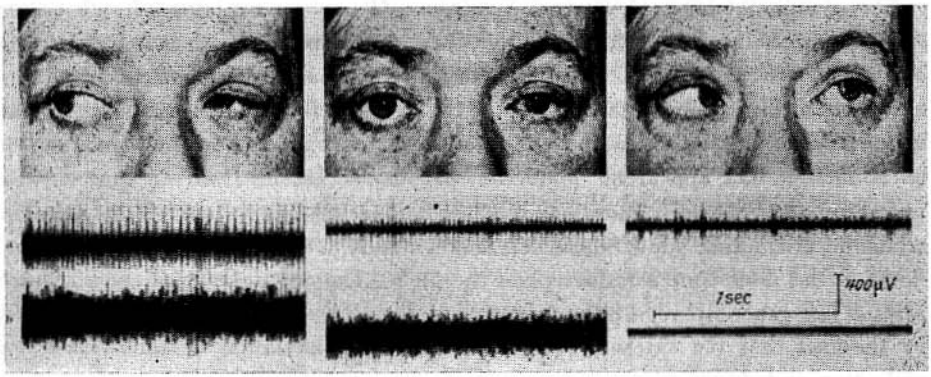

Fig. 1. *Duane Syndrome I*: marked limitation of abduction of the left eye, widening of the palpebral fissure on attempting abduction, narrowing of the palpebral fissure on attempting adduction of the left eye, retraction of the globe on adduction. *EMG* (simultaneous analysis of the external rectus (a) and the internal rectus (b) of the left eye): paradoxic innervation of the external rectus, which manifests maximum electric activity on adduction and minimum on abduction: Normal electric behaviour of the internal rectus: maximum of innervation on adduction, complete inhibition on abduction.

Electromyographic findings:

When recording simultaneously the action potentials from the internal rectus and the external rectus of the affected eye one can prove that the Duane-syndromes are caused by a disturbance of the distribution of the innervation between these two muscles, a phenomenon which has been called by Esslen and Papst *paradoxic innervation.*

In *Duane's-syndrome I* the external rectus shows a pathologic innervational pattern. On adduction the electric activity within the external rectus increases which represents just the contrary of the normal innervation. On attempting abduction the external rectus manifests a distinct decrease of innervation up to complete disappearance of electric activity. The internal rectus however shows normal electric behaviour with a maximum of innervation on adduction and inhibition on abduction. The external rectus of the affected eye in Duane's-Syndrome I has its maximum of innervation on adduction and its minimum of innervation on attempting abduction. The clinical finding of "abducent paralysis" is explained by the fact of the weak or completely failing innervation of the external rectus on attempting abduction. Because of the simultaneous innervation of internal and external rectus on adduction there results a retraction of the globe backward into the orbit.

The symptimatology of *Duane's-Syndrome II* can be explained by the electromyographic findings in a surprisingly easy way. In spite of the clinical paresis of the internal rectus there exists a practically normal electric behaviour of this muscle in all direction of gaze. However the external rectus manifests a maximum of innervation on adduction and a second equally strong maximum of innervation on abduction. The appearent "abduction paresis" in Duane II is not caused by a paresis of the internal rectus, but by a paradoxic co-innervation of the external rectus on attempting adduction which makes impossible a real adduction and produces at the same time a retraction of the globe. The absence of the abducent paralysis (as generelly regarded typical of a Duane-Syndrome) can be explained by the fact that the external rectus has a second maximum of innervation on abduction and therefore can move easily in this direction.

Also the symptomatology of *Duane's-syndrome III* can be explained by the aid of electromyography performed simultaneously on the internal and external rectus of the affected eye. Here the phenomenon of the paradoxic innervation of the external rectus manifests itself in a degree nowhere else to be observed. The normal agonist and antagonist-relationship between these two muscles is completely abolished. It seems as the two muscles of the affected eye would represent parts of one and the same muscle innervated by the same nerve. In the primary position both muscles, the external and the internal rectus, manifest a rather strong and equal innervation. The pattern of electric discharge in both muscles reveals to be practically the same. On attempting abduction there results a complete inhibition of the firing in both muscles, replaced by synchroneus discharges of 30-60 m/sec. duration. These synchroneus discharges appear in both muscles in such a similar way that one would assume moth muscles to be innervated by the same nerve. On attempting adduction there is a distinct increase of the electric activity both in the external and internal rectus of about the same intensity. The result is that the globe cannot be moved towards the nose. Instead of an adduction movement there results a distinct retraction of the globe. The impossibility of abduction is caused by the practically complete inhibition of the external rectus on attempting abduction. The described synchroneous discharges in both muscles on attempting abduction are not enough to enable an abduction movement but only a sort of nystagmus ratractorius .

Apart from Duane's-retraction syndromes of the type I, II and III there exist *other forms of paradoxical innervation*, namely simultaneous innervation of the external rectus with the inferior rectus, with the superior rectus or with several other eye muscles. All these syndromes manifest an apparent more or less pronounced limitation of the abduction and sometimes also narrowing of the

lidfissure and retracition of the globe on attempting adduction. Thus the Duane-syndromes can be placed within a still larger group of congenital disorders of innervation in which a synergistic innervation of muscles which are innervated by different nerves, occurs. Even with the aid of electromyography it is not yet possible to determinate exactly where these paradoxical innervations are centrally located. It might be possible that in future associated neurological symptoms will be able to give further information about the level of these congenital neur-motor lesions which seem to be in connexion with an embryonic maldevelopment. Here it might be of interest to point to the fact that Duane-syndromes may be associated with other developmental anomalies like Kippel-Feil-syndrome, syndactylia, shortened legs, maldevelopment of chest muscles, marked asymmetry scapulae, coloboma outer canthus, heterochromia, hypoplasia maxilla, lens opacities, krokdile tears, nystagmus, deafness etc., abnormalities which may be of significance in pin-pointing the ethiology.

With regard to the ethiology of the Duane-retraction syndrome one could imagine a lesion or anomaly within the posterior longitudinal

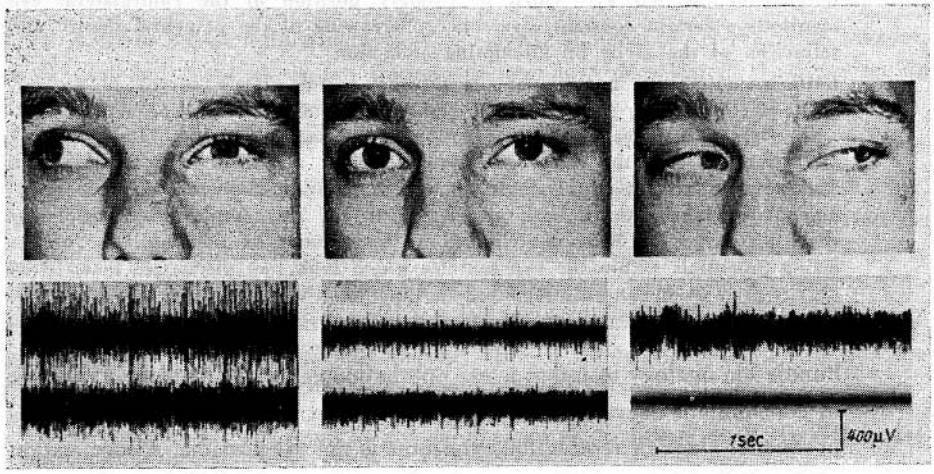

Fig. 2. ***Duane Syndrome II*: marked limitation resp. failure of adduction of the left eye, normal abduction, narrowing of the palpebral fissure and retraction of the left globe on attempting adduction.**
EMG **(simultaneous analysis of the external rectus (a) and the internal rectus (b) of the left eye):**
paradoxic innervation of the external rectus manifesting one maximum of innervation on attempting adduction and another maximum on abduction. Normal electric behaviour of the internal rectus: maximum of innervation on attempting adduction, complete inhibition on abduction of the left eye.

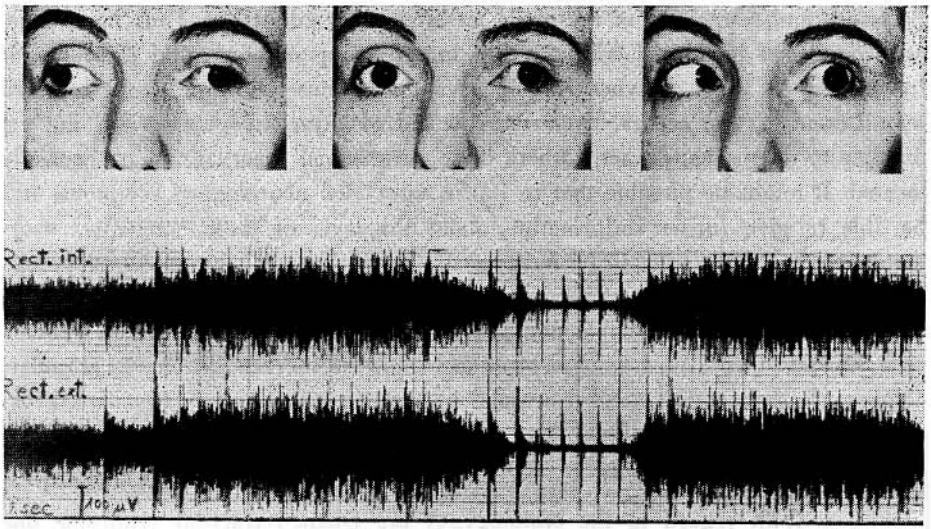

Fig. 3. *Duane Syndrome III*: practically complete limitation of abduction and adduction of the left eye which in primary position of the contralateral eye manifests a certain constant divergence. Narrowing of the palpebral fissure and retraction of the globe on attempting adduction.

EMG (simultaneous analysis of the internal rectus (lower curve) of the left eye): in the primary position of the right eye intensive innervation of both recti with slight prevalence of the innervation of the external rectus. On attempting adduction (gaze to the right) intensive electrictic action in both muscles with strikingly equal pattern of discharge. On attempting abduction distinct phenomenon of inhibition in both muscles, disturbed by completely synchroneous short discharges of 30-60 msec duration (take notice of the similarity of the action potentials within the two muscles which look like discharges taken from nearby parts of one and the same muscle innervated by one and the same nerve!).

bundle (in analogy to internuclear ophthalmoplegia). Considering the lack of concomitant neurological symptoms an exact localization of the process producing Duane's reotraction syndrome is not yet possible. A localization of the disturbance on still higher levels of the cortico-bulbar gaze cerntes seems very improbable, last but not least because in the Duane-syndrome both the command movements and the follow movements and the optically eliciated movements are intact. An other possibility of explanation would be an aberrant innervation on the level of the peripheral eye muscle nerves. In the case of the Duane-syndromes one should assume an aberrant innervation of muscles which are innervated by different nerves

(abducens and oculomotorius). For purely anatomical reasons this seems to be very unlikely; it would be possible only on the base of a hypothesis that in very early embryonic periods the nerves get into contact with wrong muscles.

SUMMARY

There are three forms of the Duane-retraction syndrome which from the clinical standpoint of view can be differentiated:

1. *Duane I* with palsy of abduction, narrowing of the lid fissure and retraction of the eye in adduction;

2. *Duane II* with palsy of adduction, normal abduction and narrowing of the lid fissure and retraction on adduction;

3. *Duane III* with palsy of adduction and abduction and narrowing of the lid fissure and retraction on attempting adduction. By means of electromyography it can be demonstrated that these "palsies" are due to a paradoxical innervation between external and internal rectus. The Duane-rectraction syndrome is to be regarded as a special form of congenital central innervation disorder where a pathological synergistic innervation occurs between muscles which work normally mainly as antagonists.

The electromyographic studies have been performed in collaboration with Dr. Esslen, Director of the Research Departament of the Neurological Clinic, University of Zurich.

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