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Case Report

Duane Retraction Syndrome - A Case Report

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Abstract

The author presents a clinical picture and therapeutic options for Duane syndrome, which may be a source of diagnostic confusion for its rarity. At the same time, he reports on the possible clinical investigations necessary to diagnose Duane syndrome.

The report presents a disease in a 5-year-old boy who was diagnosed with double-sided Duane syndrome.

Keywords: *Duane Syndrome; Paradoxical Innervation; Surgical Therapy of Duane Syndrome*

Introduction

Duane syndrome (DS) is also called Stilling-Turk-Duane syndrome or congenital retraction syndrome. Disease is known for over 100 years, but etiology and etiopathogenesis, as well as self-diagnosis and treatment is still a topic for discussion.

DS is represented in 2% between ocular disorders. At 80%, it is one-sided and more often, up to 75%, the left eye is affected. Within the sex ratio is 6: 4 in favor of women.

Duane retraction syndrome-1 maps to chromosome 8q13. DS-2 is caused by mutation in the CHN1 gene on chromosome 2q31. DS-3 is caused by mutation in the MAFB gene on chromosome 20q12 [1-6].

The diagnostic feature of DS is torticollis. It is present in up to 2/3 of patients. Although DS is a congenital illness, it is most commonly diagnosed until around the sixth year and light forms even in adulthood. The familiar occurrence is up to 6%. Amblyopia occurs in up to 1/3 of patients and rarely is of a severe grade [7].

Malbran's classification from 1949 divides according to clinical picture DS into three types.

Duane type I syndrome is the most common type, the classic form of the syndrome with up to 70% occurrence and typical symptoms. Abduction is missing or severely restricted. The primary eye position is often parallel, or the eye in light convergence, practically never in divergence. In an abduction effort, there is a slight prominence, the eye slit widens through bulb protruding or the upper eyelid retraction. Adduction is loose or only slightly limited. There is always a retraction of the eye in the orbit and a marked narrowing of the eye (pseudoptosis). Torticollis is very common, when the head is up to 60% inclined to the shoulder in the affected side, while converging occurs or escalates with a straight position of the head.

This is associated with a vertical deviation in the attempt of moving to one or the other direction. The retractable or stationary bulb deviates suddenly upwards or downwards, without otherwise breaking the vertical momentum.

In the primary position, the most common type of deviation is esotropy less than 30 ptd, it will manifest if the abduction limitation is greater than adduction limitation, and if the lateral direct muscle tone in the primary position is less than the medial direct muscle tone. Restriction of adduction may cause clinically manifest lack of convergence.

Duane type II syndrome is rarer (15 - 20%) is often confused with partial paralysis. III. In this type, the abduction is loose or only slightly limited, the bulbs are parallel in the primary position, or a slight divergence is present. The adduction is absent or severely limited, including convergence. It is always associated with significant eye retraction and narrowing of the eye slit as with DS I. Primary position is a slight divergence and when the torticollis is present head inclines to the shoulder of the healthy side (direct holding increases exotropia). Upon adduction, the retraced bulb may suddenly twist upwards or downwards, for mechanical reasons when contracting both horizontal muscles, where the eye slides up or down.

Duane type III syndrome occurs in 10-15%. Typical is a set of atypical images, motion disorders not falling into the previous types, where the bulb may be completely stationary in the horizontal position but retracts and the eye slit narrows in the adduction effort. There are clinical images with vertical momentum disorders - often mixed forms for mechanical causes [10].

Double-sided DS is rare - it occurs in up to 20%. Both sexes are represented equally. In the primary position, bulbs are parallel or alternate esotropy is present [3,5].

Atypical forms of DS are inverse DS and acquired DS.

The inverse type is a typical double-sided congenital paralysis of the abduction associated with retraction of the eye and narrowing of the eye slit in the struggle for abduction. As opposed to all known forms this finding is exceptional in the adducting eye.

The acquired DS is in patients with brain stem tumors in the region of the IV ventricle and has a clinical picture of a double-sided Duane type I syndrome.

The DS is also associated with facial synkinesis.

Rarely, DS may occur in mandibulo-palpebral synkinesis of Marcus Gunn, originating from mixing the nerve fibers of the oculomotorius nerve for the upper eyelid levator and the motor trigeminal fibers for the pterygoideus lateralis muscle.

Another synkinesis is the syndrome of crocodile tears - a condition following the facial nerve palsy, when the regenerating fibers for the chewing muscle stray into the lacrimal gland and stimulate them resulting in epiphora.

The ocular anomalies associated with DS are dysplasia of the iris stroma, pupil abnormalities, cataract, heterochromy, colobomes, microphthalmus. Systemic anomalies include Goldenhar syndrome and congenital vestibular disorders.

Diagnosis of DS requires careful anamnesis and ophthalmologic examinations including visual acuity in the distance and near. We investigate refraction in cycloplegia, front segment and ocular background, taking into account the area of macula and fixation.

Special strabological examination includes motility examination on Hess's screen, position and size of deviation using prism tests, Maddox's cross and orthoptic instruments. We need to determine binocularity, domination, attenuation states and retinal correspondence

We mainly rely on DS surgical therapy. For the indication of surgical treatment, we prefer classification according to the compensatory head position.

Only remnant deviations leading to a marked deviation of the bulb and above all torticollis can be corrected surgically. Indicated for surgery are preschool children with unilateral DS and risk of amblyopia, or children with cerebral palsy and deviation of more than 40 degrees, which prevents the creation of a simple binocular vision.

Indication is also a diplopia and a cosmetic factor.

Standard surgical procedures are usually sufficient to achieve a satisfactory state of primary eye position, but do not restore the mobility of the affected muscle.

The aim of surgical treatment is to remove torticollis, to achieve simple binocular vision and to extend the binocular field of vision in the primary eye position.

For deviations in the primary position, on the rigid media muscle, resection is recommended up to 7 mm. The method is also successfully used with the torticollis without deviation.

By resecting the lateral direct muscle of the affected Type I DS eye with esotropy and limited adduction we risk creating severe reductions in adduction and deterioration of bulb retraction in an attempt of adduction [4,6].

Single-sided or double-sided retroposition of the medial direct muscle can significantly reduce adduction (from 30% to complete loss) and cause consecutive exotropia or reduce eye rotation, which the consequences of overdosing that are difficult to manage!

The most well-known technique is the technique of weakening of the internal direct muscle by dosed elongation or partial myotomy according to Gonin-Hollwich.

The advantage of this procedure is the preservation of the original tendon and the basic function of the affected muscle. On this way treated muscle it is possible to successfully reoperated if needed [8,9].

Case Report

10/2008 A five-year-old boy diagnosed with paresis of n. VI on the right side was sent to our department. A sending ophthalmologist prescribed a +1.0 sphere correction on both sides.

From a personal anamnesis:

- Family anamnesis: negative, optht.: father corrected myopia, no strabismus and amblyopia
- Personal history: from the first pregnancy, birth without complications. Born at 42 weeks, birth weight of 3,540g, with physiological postpartum adaptation.
- In a screening pediatric examination, a sinus venosus superior defect and right kidney agenesis with left kidney compensatory hypertrophy
- Allergy: polyvalent - pollen

Ocular Finding

In the primary position, without correction, there is a significant compensatory holding of the head. Typical bow to the left shoulder and twisting of the face with a slight chin lift.

When abduction the right eye is bound from the middle line, the left eye without limitation of motility, the eye slits are symmetrical. Adduction is unlimited, but on the right, with a significant narrowing of the eye slit and a slight retraction of the bulb.

In the passive straightening of the head, the esotropy of the right eye of 15 degrees was present while looking in the distance, 20 degrees in the proximity. The left eye takes over fixation. In the cover test there is alternate fixation. The Bielschowsky test for vertical strabismus in esotropia is negative.

The convergence was directly proportional to the size of the compensatory head holding.

The former correction was without effect on the position.

VOD: 6/6 hooks with corr. Jaeger no. 2 nat

VOL: 6/6 hooks with corr. Jaeger no. 2 nat.

Both eyes: anterior segment and fundus findings are physiological.

Fundus of both eyes: in mydriasis: discs bordered, macula with foveolar reflex, central fixation.

Orthoptic analysis

The convergent compilation present, in the compensating position alternating deviation, in the passive straightening of the head no diplopia, the stereoscope – he does not connect – he alternates.

The cheiroscope – not possible to test for the right eye attenuation, Worth's light attenuation on the right, Bagolini attenuation on the right, and Hess flat.

There is no simultaneous perception or subject superposition troposcope, objective angle+21 with or without correction. Peripheral fusion: no fusion, no fusion of I, II, III degree.

Conclusion

Duane type I syndrome on both sides.

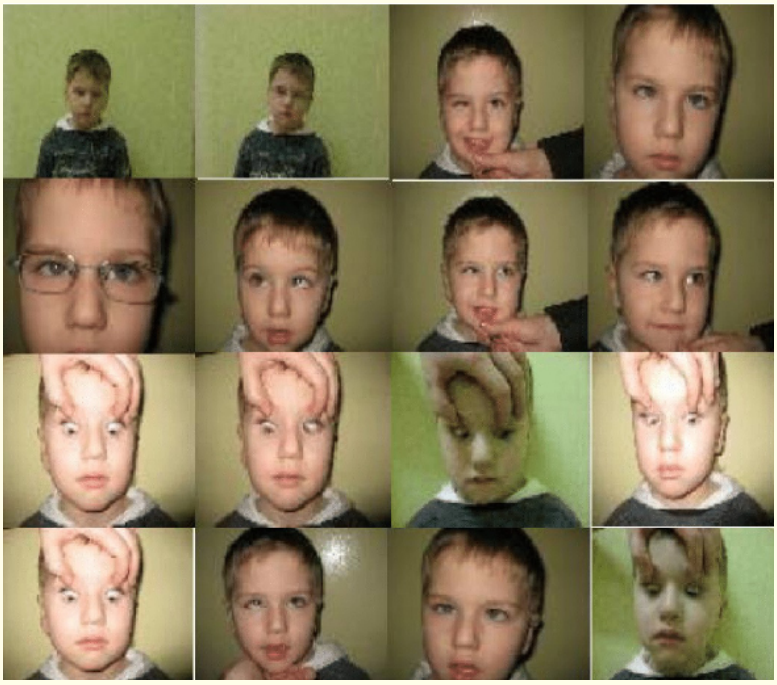


Figure 1.1: *Preoperative clinical finding.*

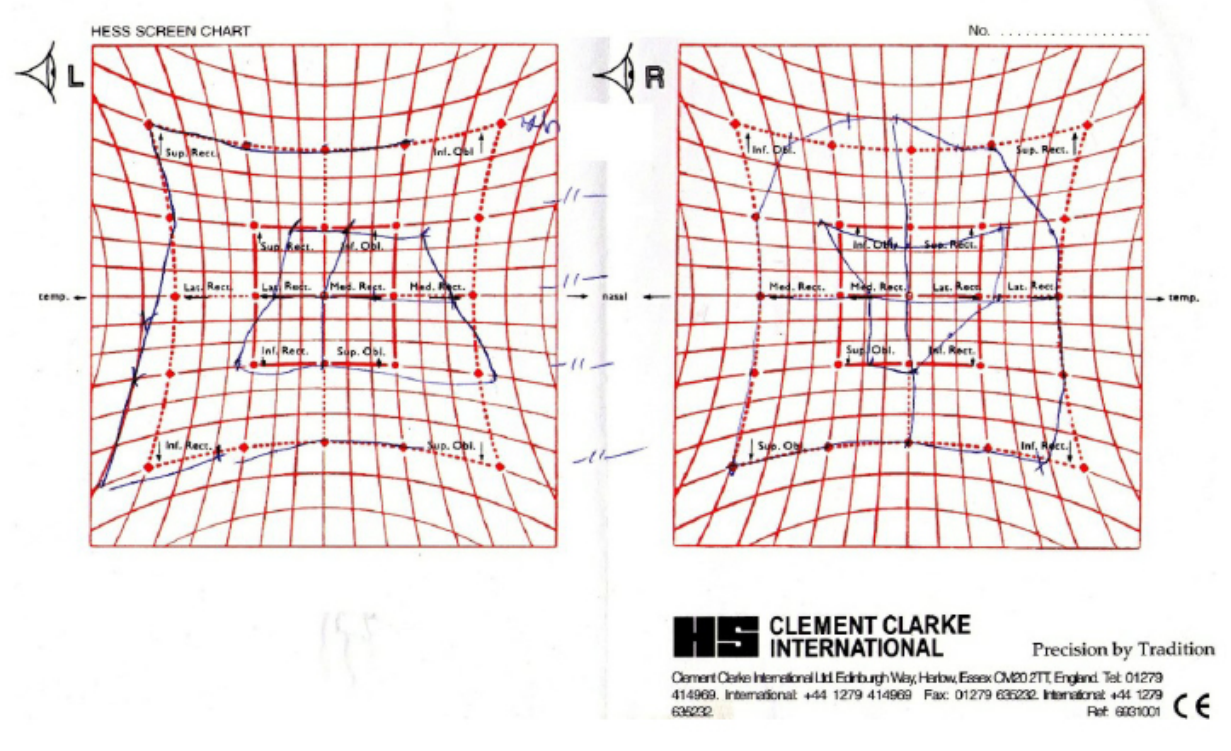


Figure 1.2: *A Preoperative range of motility.*

Operating Solutions

In general anesthesia after cutting the conjunctiva, nasal limbal cut and musculus rectus medialis preparation. The muscle elongated by the technique of Gonin-Hollwich by 4 mm on both sides. The muscles are stiffer and fibrous. Suture of the muscle and conjunctiva by PGA 5-0 individual stitches. Post-operative course without complications and the next day the patient is released into home care.

Checking

When checking after three months, the boy holds his head straight, usually just slightly bending to the left shoulder. The eye position without correction is parallel to the distance. Only a small displacement movement appears in the alternate cover test.

The right eye is without abduction, only a slight twitching and nasal narrowing of the slit, the left eye with a pronounced relaxation from the outside - up to 25 degrees (Figure 2.1).

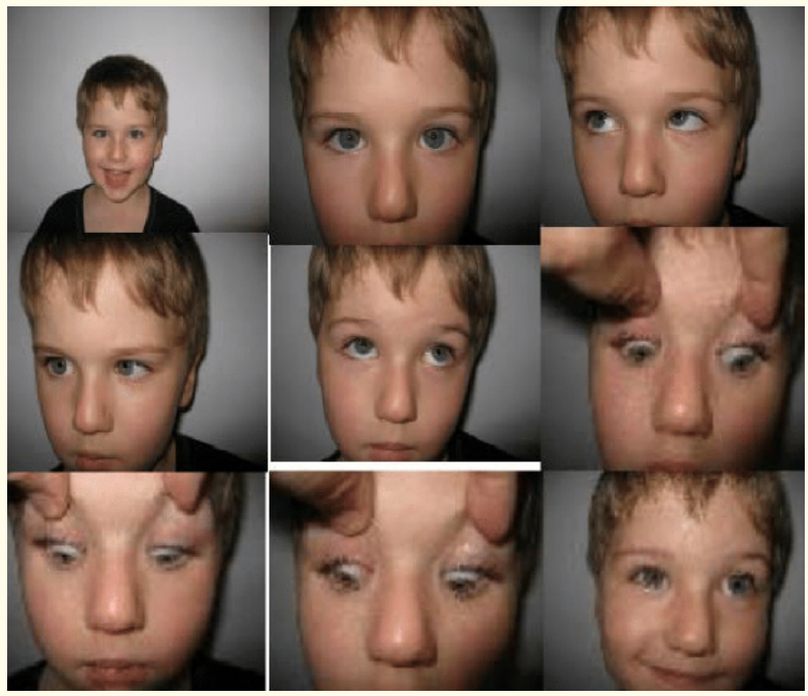


Figure 2.1: *Postoperative clinical finding.*

Hess's canvas: post-operative right eye without abduction with the corresponding motility restriction on Hess's canvas (Figure 2.2).

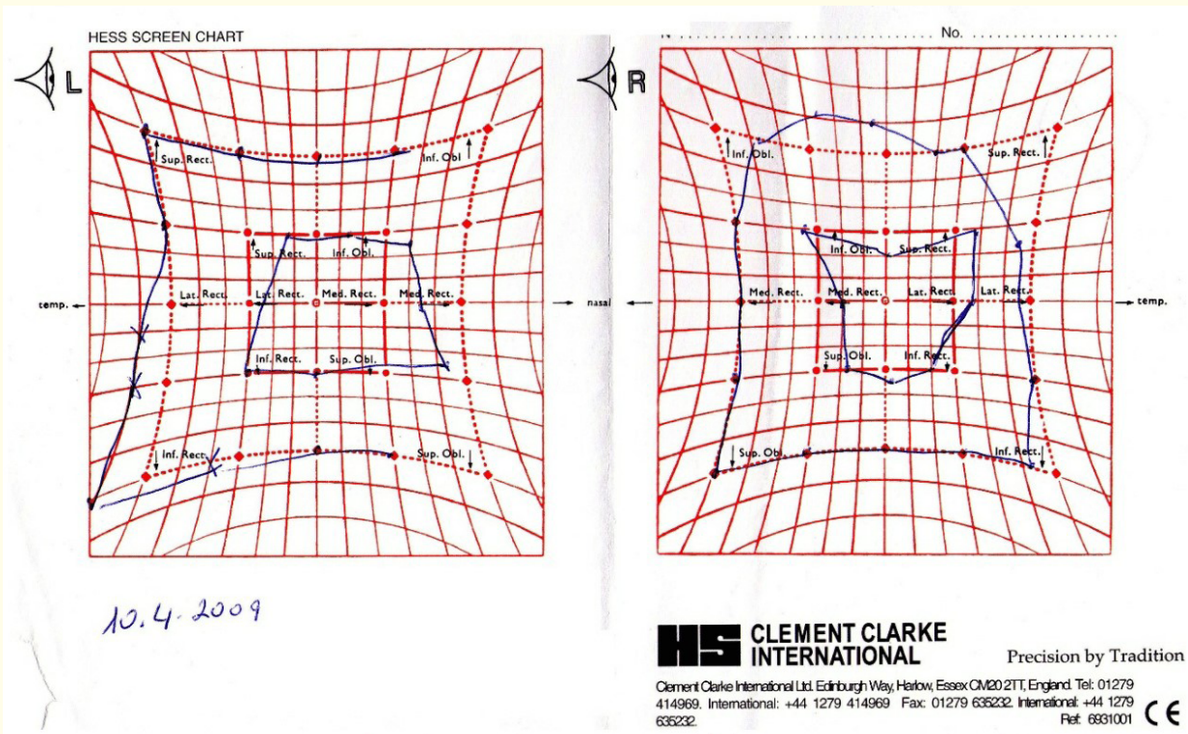
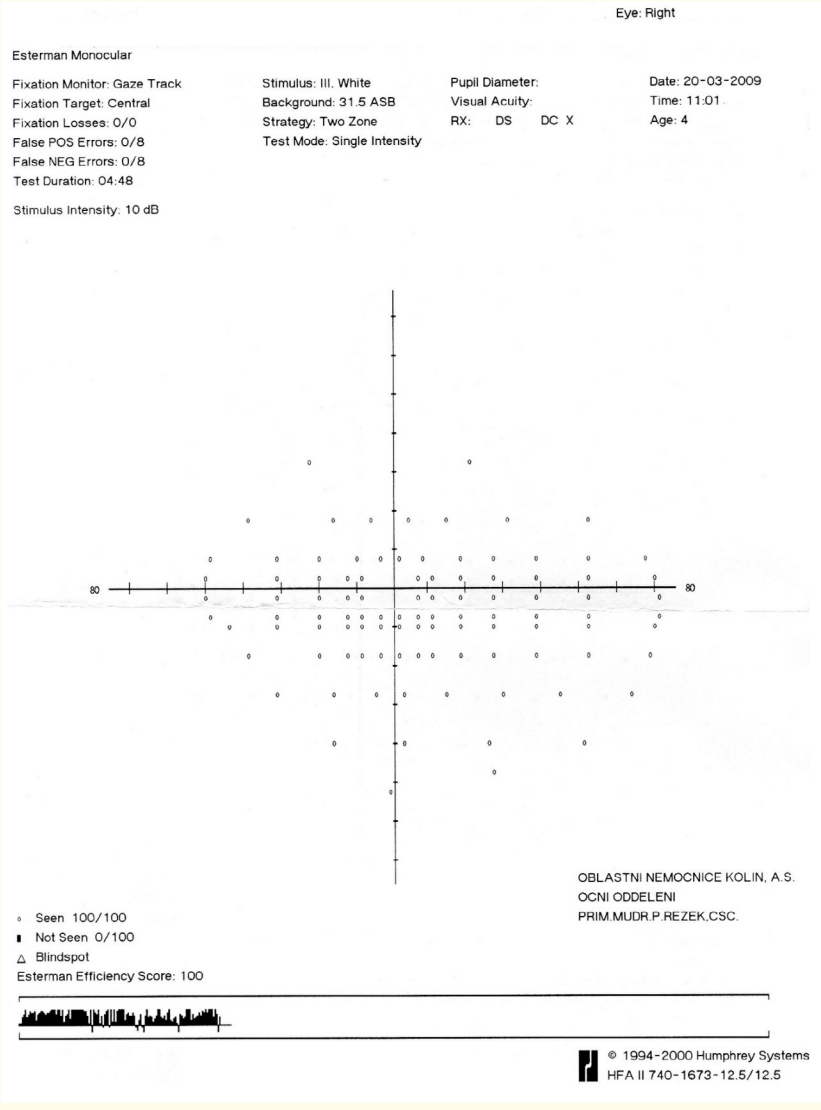


Figure 2.2: *Postoperative range of motility Hess's canvas – visual field range is improved in abduction.*

Perimetric examination of both eyes postoperatively: complete field of view without defects and limiting. (Figure 2.3).



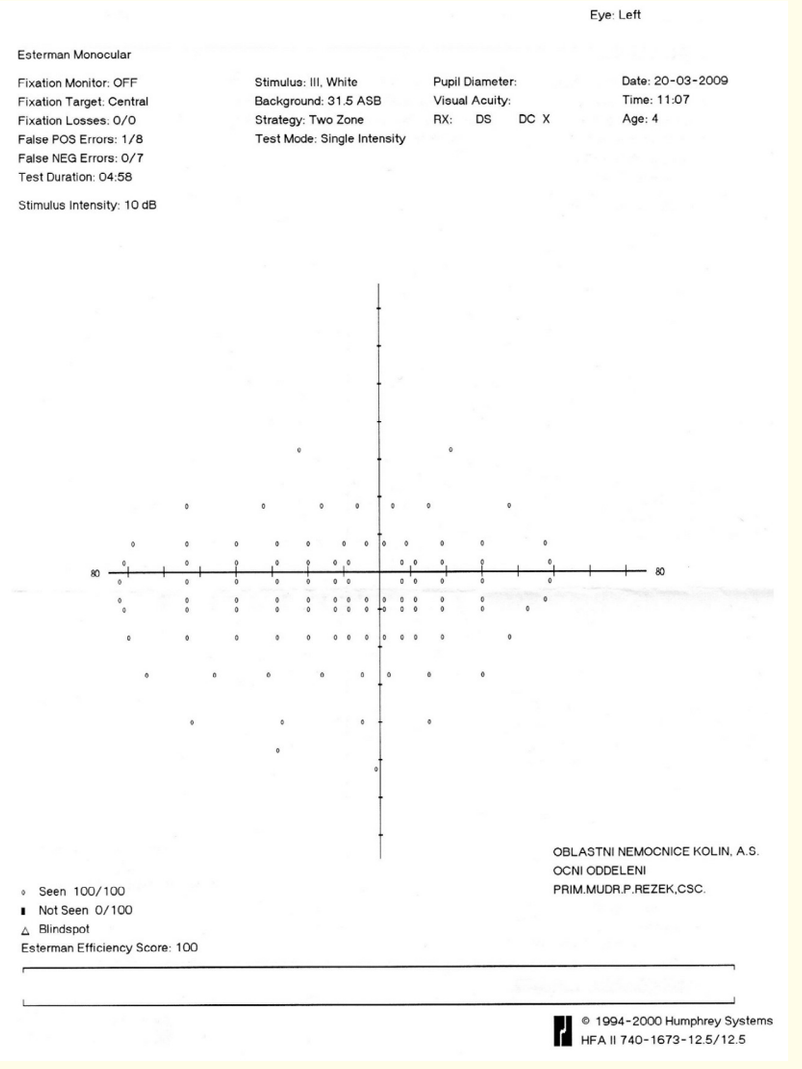


Figure 2.3: *Postoperative screening neurological perimeter C135 - both eyes: normal range of visual field.*

Post-operative care has been done with orthoptic exercises.

Discussion

DS should be distinguished from paresis of nerve VI, Moebius syndrome, congenital ocular apraxia and congenital or familial esotropy, orbital injuries of musculus rectus medialis, convergent and accommodative excesses, orbital pseudotumors, Graves disease, and atrophy of musculus rectus lateralis after surgery.

It is necessary to distinguish between DS and acquired neurogenic paresis. In the case of DS, there is orthophoria in the primary position, while in the acquired paresis there is a large deviation caused by hyperfunction or contracture of the antagonist.

So far, most published studies show that:

- Retroposition surgery - resection in a patient with DS II may improve abduction and eliminate compensatory head holding. This procedure should be performed in patients with esotropy, mild bulb retraction and good pre-operative adduction of the affected eye.
- Preoperative esotropic size of at least 15 - 20 pdt in the primary position reduces the risk of over-correction [11,12].
- The limitation of lateral direct muscle resection to 3.5 - 4.0 mm minimizes the risk of deterioration of adduction and retraction of the bulbus (Figure 3) [1,2].

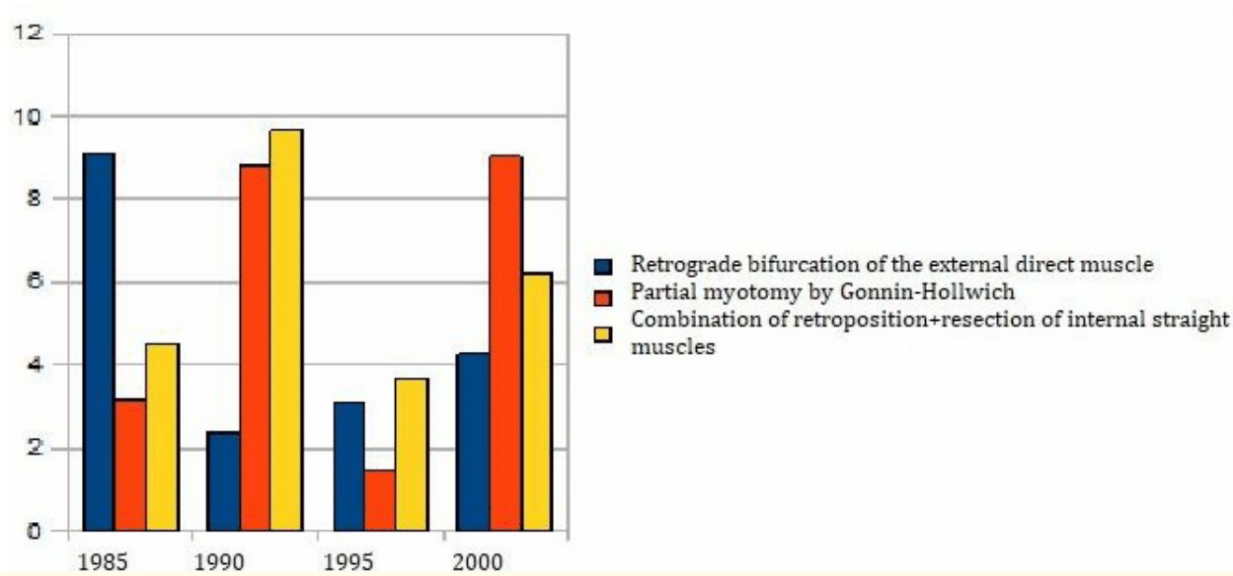


Figure 3: *In this illustration, it is obvious lately is the retreat from traditional techniques, such as retroposition - resection and the popularity of partial myotomy increases for most accurate final postoperative result.*

In the postoperative period, it is necessary to check regularly from the beginning of the healing process itself and consequently also the need for orthoptic - pleoptic exercise. It is unconditionally necessary to use full refractive correction, in the treatment of amblyopia occlusive methods and, last but not least, the enhancement of motility and binocularity itself in the orthoptic examination. Only then can we tell that the patient is treated in a comprehensive way. Regular check by the ophthalmologist should take place until adulthood.

Conclusion

For our patient, we chose a surgical method to loosen and weaken the internal rectus muscle - a curable elongation according to Gonin-Hollwich. Postoperatively with very good cosmetic effect minimizing torticollis and with virtually parallel primary position of the eyes. The patient is dispensaries and in the aftercare we rely on orthoptic exercises in an attempt to create a simple binocular vision.

When examining patients with strabismus, it is necessary to focus on careful complex examinations, not only to address the primary diagnosis of "first sight", but also to think about other options within differential diagnosis. This is necessary for the right diagnosis and successful treatment.

An essential part of treatment in patients with such complicated type of strabismus is the precise strabological analysis and subsequent postoperative orthoptic exercises.

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