Aus der Augenklinik und Poliklinik
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DIE HINTERE TENDEKTOMIE ZUR THERAPIE DES KONGENITALEN BROWN-SYNDROMS

THE SUPERIOR OBLIQUE POSTERIOR TENECTOMY AS THERAPY FOR CONGENITAL BROWN'S SYNDROME

Dissertation
zum Erwerb des Doktorgrades der Medizin
an der Medizinischen Fakultät der
Ludwig-Maximilians-Universität München

vorgelegt von
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2011

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Tag der mündlichen Prüfung: 05.05.2011

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1. Introduction

1.1. Definition and clinical features of Brown's Syndrome

The ocular motility disorder defined by Brown has consistent and characteristic features, making it an easily recognizable clinical syndrome (Table 1). The most striking feature is the active and passive limitation of upward gaze in adduction. With rotation of the eye out of the field of vertical action of superior oblique muscle, elevation improves so that less elevation restriction is present in midline and minimal or no elevation deficit is present in abduction.

There are varying degrees of severity of Brown's syndrome and different etiologies. Therefore we can meet varying facultative features, including a widening of the palpebral fissure on adduction, divergence on midline elevation or even in more severe cases primary position hypotropia or a downshoot of the affected eye below the horizontal meridian on adduction.

In mild cases of Brown's syndrome we can observe a normal ocular alignment in primary gaze. In severe cases, a primary position hypotropia prompts an abnormal head posture: most commonly a chin-up position, but sometimes a face turn away from the affected eye or a variable head tilt.

Table 1. Clinical Features of Brown's Syndrome [13, 46, 47]

Typical Features		Variable Features	
1	Limited elevation with restricted forced ductions in adduction	1	Divergence in upgaze producing a V or Y-pattern
2	Less elevation deficiency in midline gaze	2	Minimal or no superior oblique overaction
3	Minimal or no elevation deficit in abduction	3	Down-shoot in adduction
4	Free forced elevation in adduction after superior oblique tenotomy	4	Widened palpebral fissure on adduction
		5	Anomalous head posture with primary position hypotropia
		6	Incyclotorsion accentuated on attemped elevation

1.2. Historical perspective

In 1928 Jaensch described the first case with severe limitation of elevation of the adducted eye after a skiing accident [19]. The clinical picture resembled a paralysis of the inferior oblique muscle, but the forced ductions test showed resistance to elevation of the adducted eye. As a cause, Jaensch suspected a traumatic adhesion between the trochlea and the globe anterior to or at the equator.

In 1950 Brown described an identical anomaly of ocular motility occurred on a congenital basis [3]. However, he understood the disorder as a congenital paralysis of the inferior oblique muscle with consecutive shrinkage of the superior oblique tendon sheath and grouped this entity named "Superior oblique tendon sheath syndrome" together with retraction syndrome, strabismus fixus, fibrosis syndrome - restrictive motility disorders characterized by fibrous changes in the muscles or their tendon sheaths.

Since Brown's original description in the 1950s, after a half a century of collective experience in the diagnosis and treatment of Brown syndrome, it has become clear that there are many anomalies involving the superior oblique muscle, its tendon, surrounding tissue or the trochlea that may contribute to a mechanical restriction of elevation of an adducted eye [4, 7, 17, 31, 35]. For this reason, the older term "Superior oblique tendon sheath syndrome" became ***Brown syndrome***, although the term ***Jaensch-Brown syndrome*** has also been sugested [31].

1.3. Types of Brown Syndrome

In 1973, Brown realized that "Superior oblique tendon sheath syndrome" was actually more complex than he first described, involving different causes and various degrees of severity. He divided the syndrome into two groups: "true" and "simulated" sheath syndrome [4].

The "true" Brown syndrome included congenital, constant and permanent cases and was subdivided into typical and atypical forms. Typical cases had full elevation in abduction, whereas atypical cases had some degree of elevation deficit in abduction, due to a presumed concomitent paresis of the ipsilateral superior rectus muscle.

The "simulated" Brown syndrome included aquired, intermittent or cases showing spontaneous recovery, even if thought to be congenital and constant prior to discovery of regression.

1.4. Anatomical considerations

The superior oblique muscle, the longest extraocular muscle, arises from the body of sphenoid bone above and medial to the optic canal just outside the tendinous ring. The muscle belly (30mm long) runs forward between the roof and medial wall of the orbital cavity and continues with a rounded tendon. The tendon passes through trochlea, a pulley of fibrocartilage that is attached to the trochlear fossa of the frontal bone. After emerging from the trochlea, the tendon bends downward, backward, and laterally, forming an angle of about 54° with the pretrochlear or direct portion of the muscle. After the posttrochlear or reflected part of the tendon passes under the superior rectus muscle, expands in a fan-shaped manner and inserts into the sclera posterior to the equator of the eyeball, forming a convex line of insertion.

According to Fink [10, 11], the anterior end of the insertion lies 3.0 to 4.5 mm behind the lateral end of the insertion of the superior rectus muscle and 13.8 mm behind the corneal limbus. The posterior end of the insertion lies 13.6 mm behind the medial end of the insertion of the superior rectus muscle and 18.8 mm behind the corneal limbus. The width of the insertion of the superior oblique muscle varies greatly from 7 to 18 mm, with an average of 11 mm. The medial end of the insertion lies about 8 mm from the posterior pole of the eye (Fig. 1).

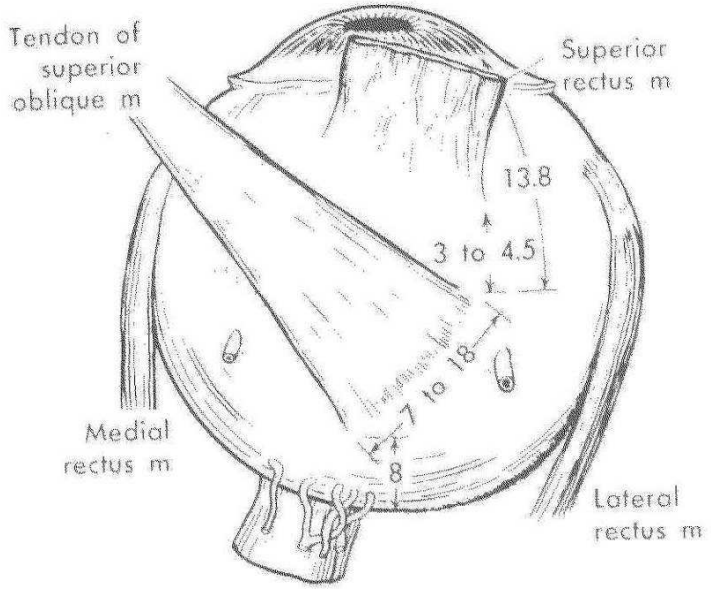


Fig. 1. Relationships of tendons of superior oblique muscle
Right eye, view from above [11]

From all the extraocular muscles, the superior oblique muscle shows the most frecquent anomalies [23]. Many authors [18, 23, 46] communicated cases with the absence of the superior oblique tendon. A redundancy of the tendon, an abnormal posterior insertion of the tendon into Tenon' capsule or an extreme laxity of the tendon were noted in most congenital cranial nerve IV palsy. An aplasia of the trochlea or the Y- splitting of the superior oblique tendon have also been communicated. A tight band at the posterior border of the tendon between the trochlea and the sclera was also observed in some cases with Brown's syndrome [16, 26, 42].

The trochlea is a complex structure attached to the bony orbit at the trochlear fossa located near the junction of the superior and medial orbital rim, with the function to redirect the superior oblique tendon. From a physiologic and kinematic standpoint, the trochlea is the origin of the superior oblique muscle [46].

Because the trochlea is small and firmly affixed to bone and lies deep in the orbital fascia, it was not accessible for surgical exploration from the usual transconjunctival approach by the strabismus surgeons. Penetration of posterior Tenon's capsule and dissection of orbital extraconal fat are required in the approach to the trochlea, creating tissue adhesions in the trochlear region. Thus, its anatomy and physiology were investigated only late, in '80s by Helveston, using fresh human orbital exenterated specimens and autopsy specimens [17].

Four components of the trochlea were described: a cartilage saddle, an intratrochlear portion of superior oblique tendon, a fibrillo-vascular sheath surrounding the tendon and a dense fibrous condensation that secures the trochlear saddle to the bony medial orbital wall [17]. Each fiber of the superior oblique tendon acts nearly independently as a cord from the muscle fibers to the insertion without the presence of interfiber connection (lateral attachments or cross-connecting fibers) found in other extraocular muscles. This finding was verified by Helveston and coworkers and presented as ***a telescoping or slide-by fashion of movement*** : as the tendon passes through the trochlea, each fiber slides farther than the next adjacent and more peripheral fiber (Fig. 2).

Initially, the peripheral fibers move and, in succession, the fibers located more centrally slide relative to the next most peripheral fibers, with the central fibers undergoing maximal excursion and the peripheral fibers the least excursion. The total travel of central fibers appears to be 8 mm in either direction. Also, since in adduction the posterior insertional fibers undergo the maximum excursion, it is proposed that the central tendon fibers insert posteriorly and the peripheral fibers anteriorly.

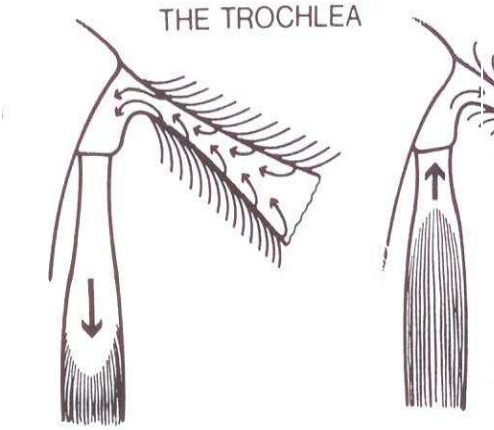


Fig. 2.
The telescoping movement of of the superior oblique tendon through the trochlea [17]. The fibres of the SO tendon move independently with only the central fibres making the full excursion through the trochlea.

Whitnall described a fascial sheath of the reflected tendon of the superior oblique muscle consisting of two layers (2 to 3 mm thick) of strong connective tissue [46]. Parks has rejected the idea that superior oblique tendon could have a separate anterior sheath over it near its insertion as described by Brown [35]. What appears to be a sheath was demonstrated to be reflection of anterior and posterior Tenon's capsule, forming a sleeve that have been mistaken by Brown for a sheath.

Helveston and coworkers described a highly vascular sheath of the intratrochlear portion of the superior oblique tendon [17]. Helveston recognized that movement of the tendon in the trochlear pulley creates a metabolic requirement for repairing "wear and tear" and heat dissipation. This is the only extraocular muscle or tendon that has such a rich vascular supply. The presence of these vessels creates the anatomic basis for possible vascular dilatation or local edema that, if occuring, could lead to restricted passage of the tendon through the trochlea and thus produce a permanent or intermittent Brown syndrome. The resolution of local edema in the trochlear region could also explain the spontaneous resolution seen in acquired and intermittent Brown syndrome cases.

In addition, a bursa-like structure was described by Helveston between the tendon's vascular sheath and the trochlear saddle. Excess fluid accumulation or concretion in this bursa-like space or vascular distension in the sheath could lead to limitation of movement through the trochlear tunnel, causing an acquired Brown syndrome. Wilson and coworkers [47] suggested that if the telescoping movement of the tendon described by Helveston were interfered with an intrinsical anomaly of the trochlea or the tendon, Brown syndrome would result.

1.5. Action of superior oblique muscle

In primary position, the activation of superior oblique muscle causes *incycloduction* and *depression* of the eye and also *abduction*.

When adduction is initiated, the angle between the medial plane of the eye and muscle plane (normal 54°) is reduced progressively and and the superior oblique muscle acts more and more as depressor (Fig. 3). With an adduction of 54°, the superior oblique would be a pure depressor.

When the abduction is initiated, the angle between the medial plane of the globe and oblique muscle plane increases and the superior oblique muscle produces more incycloduction. With 36° of abduction its action is one of pure incycloduction.

So, as a conclusion, the maximum action of the superior oblique muscle as a depressor is in adduction, but in abduction occurs the maximum incycloduction.

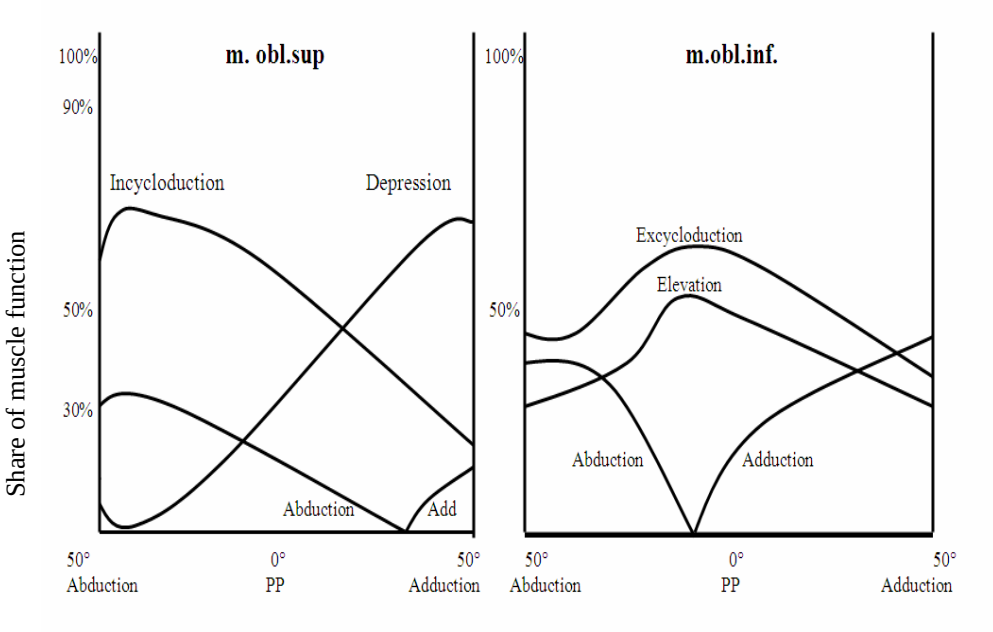


Fig. 3. Representation of superior and inferior oblique muscles function [23]

2. Etiology and pathophysiology of Brown syndrome

The question of etiology is one of the most controversial issues surrounding Brown's syndrome, complicated by the existence both of acquired and congenital cases.

The aquired cases involves secondary changes in a previously normal superior oblique (SO) tendon or tendon-trochlear complex, due to trauma to superomedial orbit, inflammation (rheumatoid arthritis, abcess formation or metastasis in the region of the trochlea), iatrogenic changes of the trochlear region-following sinus surgery or strabismus surgery [47].

In congenital forms of Brown syndrome, the etiologic and pathophysiologic mechanism remains enigmatic up to now. For better understanding the etiopathological theories which were under debate since almost 60 years, we have to keep in mind that the SO muscle and tendon must relax as the anterior pole of the adducted eye moves upward. In the same time, the posterior pole of the eye, where the SO tendon inserts, moves away from the trochlea to an abducted and depressed position. This requires a certain degree of relaxation or elasticity of the SO muscle and tendon. To explain the mechanical limitation of elevation in adduction through different mechanisms, Helveston suggested a generic description of the syndrome, according to which the inability to elevate the adducted eye is due to *a failure to increase the distance between the trochlea and the SO tendon insertion*. He described the complex machinery consisting of muscle, tendon and trochlea which is especially vurnerable to developmental defects [17, 18].

2.1. Concept of an anterior tendon sheath

In 1950, at the first American Strabismus Symposium, Harold Whaley Brown debated upon the motility disorders with restrictive character, explained by fibrous changes in the muscle or their tendon sheaths [3]. He grouped in the same entity retraction syndrome, fibrosis syndrome, strabismus fixus, vertical retraction syndrome and a new syndrome which he named "Superior oblique tendon sheath syndrome", a motility disorder defined by restricted elevation of the globe on adduction.

Brown understood the disorder as a congenital paralysis of the inferior oblique muscle with consecutive *shortening of the superior oblique tendon sheath*. According to Whitnall, the tendon sheath of the superior oblique was a structure fixed to the trochlear pulley and fused to the scleral insertion of superior oblique muscle [46]. Brown stated that if this sheath were short it would certainly restrict elevation on adduction.

Fink had described the sheath as a significant membrane having a multitude of minute fibrillar connections to the tendon that prevented the tendon from moving freely through it [11]. Berke described a 2-3 mm thick sheath or areolar tissue along the superior oblique tendon from Tenon's capsule to the trochlea [1]. Under this proposed hypothesis of shortening of the superior oblique tendon sheath, surgical stripping of the sheath or pseudosheath leaving the tendon itself intact has relieved the restriction in some cases of Brown syndrome. Brown achieved full correction of the motility on adduction in 5 of 26 patients operated in this way [4].

Parks observed that the superior oblique tendon does not have a sheath at all. Instead he found that the surrounding tissue of the tendon and Tenon's capsule, through which superior oblique tendon passes, create a sleeve and maybe this sleeve have been mistaken by Brown for a sheath [35]. With modern superior oblique surgery using direct visualization, an anterior "sheath" as described by Brown was not found, the superior oblique tendon having a transparent, avascular capsule, similar to the capsule that envelops the tendons of the other extraocular muscle, giving it a smooth glistening character [7, 36, 47].

In the course of time, Brown himself left his concept of an underlying inferior oblique palsy with reactive tendon sheath shrinkage and rather discussed a *disturbance of the superior oblique tendon itself*. In 1971, Brown stated that multiple etiologies could lead to the clinical pattern he had described [4].

2.2. Anomalies of the superior oblique tendon or trochlea

For at least the majority cases of congenital Brown's syndrome the cause can be found in anomalies of the superior oblique tendon and/or the trochlear apparatus.

Crawford (1980) and later Von Noorden (1982) proposed that the cause of congenital Brown syndrome is an *abnormal tightness of the muscle-tendon complex* [7, 46]. Different degrees of severity of the syndrome are explained by the spectrum of all possible degrees of deficient muscle elasticity.

Arguments for supporting this hypothesis brought Girard in 1956 and later many other authors which recognized that tucking the superior oblique tendon in cases with superior oblique muscle palsy could lead to elevation restriction in adduction (acquired Brown syndrome). That means a tight superior oblique tendon could be the cause of Brown syndrome. This theory was in accord also with good surgical results obtained after tenotomy (cutting the superior oblique tendon just medial to the superior rectus muscle).

Girard postulated that a congenital anomaly of the tendon (retrotrochlear thickening) or anomalies of the trochlea itself could cause an impairment of slippage of the findings of the human trochlea made by Helveston have supported this theory [17, 18].

In 1981 Sevel raised the hypothesis that persistence of fine embryonic trabecular connections between the superior oblique tendon and the intratrochlear sheath may limit free slippage of the tendon through trochlea [42].

In 1996 Mühlendyck found a tight band at the posterior border of the tendon between the trochlea and the sclera in all patients with Brown syndrome [31] and communicated good results after resection of this band (superior oblique posterior tenectomy).

2.3. Anomalies of inferior oblique muscle and adjacent structures

In 1956 Girard found a dense fibrous attachment extending from the insertion of the inferior oblique muscle to the lateral wall of the orbit in a patient with all features of Brown's syndrome [12]. After resection of this band, the resistance to passive elevation of the globe in adduction disappeared.

In 1972 Scott and Knapp reported cases of Brown's syndrome with inferior restrictions and remarked that poor results after superior oblique sheath surgery could be due to these inferior adhesions [40].

In 1975 Parks and Brown found some inferior bands extending from the inferior rectus muscle capsule to orbital floor in a patient with Brown syndrome. The traction test has been improved by cutting this bands, but the active elevation in adduction not [35].

In 1976 Scott described an adaptation of the forced traction test trying to differentiate inferior from superior restriction. Upon testing the passive rotations of the eye, superior restriction is enhanced by depressing the globe into the orbit, inferior restriction is enhanced by proptosing it [40].

2.4. Paradoxical innervation

In 1969 Papst and Stein, after simultaneous electromyography performed on both the superior oblique muscle and the inferior oblique muscle in two patients with congenital Brown's syndrome, found *co-contraction of the inferior and superior oblique muscles* on combined elevation and adduction of the globe and likened Brown's syndrome to Duane's syndrome [34].

In 1971 Feric-Swiwerth and Celic also found evidence of paradoxical innervation of the superior oblique muscle on attempts to elevate the eye in one of three tested patients with Brown's syndrome [9]. They theorized that congenital Brown syndrome is a central innervational disorder and that acquired forms were due to local abnormalities or tenosynovitis.

However, in the same year, Catford and Hart found no paradoxical innervation in patients with acquired Brown's syndrome [5].

In 2004, at the 10th Meeting of the Bielschowsky Society, De Decker reported two cases of superior and inferior oblique co-contraction in electromyography in Brown syndrome [32].

Regarding of the hypothesis of paradoxical innervation, everyone would expect that the forced duction test to become negative in the patient with congenital Brown's syndrome under anesthesia. But this is never the case as Von Noorden observed and this is a counterargument for this hypothesis, although in Duane retraction syndrome intraoperative restriction in adduction/abduction does not disappear under general anesthesia.

2.5. Congenital Brown's syndrome - a CCDD ?

Recently, there has been brought light to the etiologies of some of the other congenital restrictive disorders like Duane's syndrome or congenital fibrosis of extraocular muscles (CFEOM), which Brown has grouped together with "Superior oblique tendon sheath syndrome".

Pathologic, electromyographic and genetic studies [15] have shown that their etiology is rather primarily neurogenic. Current concepts state in these restrictive syndromes a *developmental hypo- or aplasia of the cranial oculomotor nerves and a resulting fibrosis in the target muscles due to non- or paradoxical innervation.*All these different syndromes with congenital, nonprogressive, sporadic or familial developmental abnormalities of the cranial nerves and its nuclei are classified as congenital cranial dysinnervation syndromes (CCDD). In hereditary cases of CFEOM and Duane's syndrome, studies have shown that gene loci that promote the segmental brainstem development are affected [15].

Under the light of the recently defined CCDD, at the 11th Meeting of the Bielschowsky Society, in 2005, Neugebauer [32] discussed the most striking hypothesis that congenital Brown syndrome might be caused by a developmental defect in brainstem differentiation (fourth nerve hypoplasia or aplasia) with consecutive paradoxical innervation of superior oblique muscle by fibres intended to innervate the inferior oblique, the medial rectus, the superior rectus or others.

It is remarkable that this group of CCDD includes syndromes with involvement of the third and sixth nerve (like CFEOM, Duane's syndrome, congenital ptosis) and diseases characterized by a combination of third and fourth nerve involvement (like CFEOM II), whereas a single fourth nerve dysinnervation syndrome was not described, observed Neugebauer.

Most interesting **arguments** brought by Neugebauer supporting the theory that congenital Brown's syndrome could be a misinnervation syndrome are:

1. Duane's syndrome, Crocodile tears, Marcus-Gunn jaw-winking and unilateral ptosis were reported to occur together with Brown's syndrome [46, 47] and also associated with congenital cranial misinnervation disorders [15]. Several reports exist that communicate patients with Brown's syndrome on one side and trochlear palsy on the fellow eye. Clarke reported an incidence of 11% contralateral trochlear palsies in patients with congenital Brown's syndrome [6].
If Brown's syndrome would be a primary developmental defect of the fourth nerve nucleus with paradoxical innervation of the superior oblique muscle, everyone could imagine cases in which the developmental defect occurs, but coinnervation was not established sufficiently, causing a palsy.

2. Superior oblique muscle paradoxical innervation by fibres for the inferior oblique muscle (III nerve) would explain :
- the elevation deficiency in adduction
- discordance between large motility deficiency in adduction and small angle of hypotropia in primary position
Attempting to elevate the globe in adduction, maximal innervation of the inferior oblique muscle occurs. Simultaneous co-contraction of superior oblique muscle together with inferior oblique muscle will hinder the globe from being elevated and elevation deficiency in adduction occurs.
On the other hand, by simultaneous innervation of both antagonist oblique muscles will result an antagonistic movement in the vertical plane, which would explain the small angle of vertical deviation in primary position, in spite of large motility deficiency in adduction.

3. The widening of palpebral fissure could be explained by an anteropulsation of the globe occuring with the co-contraction of both oblique and this phenomenon is comparable to retraction of the globe with co-contraction of the horizontal recti in Duane's syndrome.

4. The tightness of the superior oblique tendon could occur during time, triggered by constant co-contraction.

Future clinical, genetic and MRI findings would be of great interest for clarifing the etiology of congenital Brown's syndrome.

2.6. Radiological findings

Many authors [2, 28, 41] tried to investigate mechanisms of congenital Brown's syndrome by magnetic resonance imaging (MRI).

Most of the abnormalities found on MRI have been reported to be located at the tendon-trochlea complex: an enlargement was noted in some cases, an irregular appearance in shape with intermediate signal intensity in other cases of congenital Brown's syndrome [41].

A new method, high-resolution, multipositional orbital MRI with surface coils, which investigate muscle size and contractility demonstrated a variety of abnormalities in patients presenting with congenital Brown's syndrome, including atrophy or absence of the superior oblique belly [2]. Using this method, Bhola found an inferior displacement of the lateral rectus pulley in adduction, with normal superior oblique tendon-trochlear complex in one congenital and one acquired case of Brown's syndrome. Such cases responded to surgical stabilization of the lateral rectus pulley.

Strong arguments for supporting the hypothesis that congenital Brown's syndrome could be a CCDD brought Kolling in 2008 [25]. He demonstrated in 2 of 4 patients with congenital Brown's syndrome unilateral lacking of fourth cranial nerve on MRI in high resolution technique. Kolling observed also in one case paradoxical contraction of superior oblique muscle on attempting to elevate the globe in adduction.

Up to now, the radiological findings described in congenital Brown's syndrome proved to be various, in connection with the varied hypothesis regarding the etiology of this entity.

3. Treatment of congenital Brown's syndrome

Knowledge and understanding of Brown's syndrome have improved, like the surgical management with refinement of techniques. Precise indication for therapy, however, remains controversial and is clouded by insufficient data on the true incidence of spontaneous regression.

For many authors, the most common management for all forms of Brown syndrome has been observation alone.

3.1. Natural course of congenital Brown's syndrome

It is generally accepted that spontaneous resolution rather occurs in acquired cases than in congenital cases and was reported by several authors [14, 20, 21, 27].

Gregersen [14] reported striking results of a longitudinal long-term study: of 10 patients with congenital Brown's syndrome diagnosed during the first 2 years of life and followed for 13 years, 3 had complete recovery of normal ocular motility and partial improvement was noted in 6 of 10 patients.

Kaban [20] reported a 10% resolution rate in congenital cases over 7 years.

In the acquired and intermittent forms of Brown's syndrome with inflammatory origin, the impaired slippage of the tendon is caused by the hypertrophy and constriction of the trochlea and tendon sheath associated with localized swelling of the superior oblique tendon. It is easy to understand that in such cases sudden release of the restriction and full rotation of the globe nasally and upward occurs after the inflammation subside.

In constant, congenital Brown's syndrome, considered a stable disorder, spontaneous improvement is hard to explain. Some authors supposed that spontaneous recovery is due to the enlargement of the trochlear ring with growth. Taking into account the hypothesis of misinervation, Neugebauer supposed that during the time, triggered by constant co-contraction, consequent lesions and elongations in the superior oblique tendon could occur, facilitating the slippage of the tendon trough the trochlea [32].

Another argument for high percent of spontaneous resolution in constant, congenital Brown's syndrome is the fact that Brown syndrome is encountered less frequently in adults. Of the 126 patients described by Brown, 89% were identified in children under the age of 12 years [4, 47]. This suppots the theory that *spontaneous resolution in cases of congenital Brown's syndrome is probably more common than previously recognized*.

3.2. Surgical treatment

3.2.1. **Indication for surgery**

In patients without spontaneous resolution of Brown's syndrome, surgery may be a treatment option.

Indication for surgery in congenital Brown's syndrome include the presence of a primary position hypotropia and/or an anomalous head posture. A large downshoot in adduction causing psychosocial stress to the patient is considered a relative indication for the surgery.

The goal of the treatment is to correct hypotropia in primary position, to reduce objectionable downshoot and head posture, to increase upgaze and expand the binocular diplopia-free fields. When binocular vision is normal in primary position and without an extreme anomalous head posture, the surgery is not advisable. These patients could experience diplopia when they attempt to elevate the involved eye in adduction, but they will learn to avoid this position of gaze.

3.2.2. **Surgical procedures**

Based on the incorrect supposition that congenital Brown syndrome is due to a contracted "anterior sheath" surrounding the superior oblique tendon, *dissecting and stripping the sheath*, as originally advocated by Brown, became the first recommended surgical procedure. The few reported sheathectomy results were uniformly unsuccesful [4, 12, 35].

After almost 20 years, the surgical results obtained by Brown in 36 cases of sheathectomies were dissapointing: only in 5 patients were obtained full rotations, 13 improved mildy, in 15 patients he reported no changes and overcorrection in 3 patients [4].

Although isolated reports of *superior oblique tenectomy/tenotomy* appeared since 1955 (Nutt), in 1970 Crawford recommended first tendon weakening in congenital Brown's syndrome, based on the theory that a tight superior oblique tendon caused the motility deficit. After tenotomy of the superior oblique, restriction of elevation the eye in adduction was usually eliminated, but a high percent of patients (ranges from 40% to 85%) developed iatrogenic superior oblique palsy with longer followup and 11%-42% of them loss of binocularity [7, 36, 46].

To reduce the incidence of postoperative superior oblique palsies, Parks studied several different superior oblique tendon weakening procedures like *tenectomy of posterior tendon, Z-tenotomy, split tendon lengthening or tendon recession* [35, 36].

Technically, all these procedures proved to be more difficult, requiring considerably dissection and, consequently, more scarring and finally have shown discouraging results.

Later studies of Parks regarding the intermuscular septum existing along the entire sub-Tenon's course of the superior oblique tendon are valuable [36]. Parks has shown that dissecting the anterior and posterior borders of the tendon from the intermuscular septum followed by tenotomy along the medial border of the superior rectus muscle produces a high incidence of superior oblique muscle palsy (5 patients of 6 operated). Of 15 patients in whom the intermuscular septum was preserved, only 3 (20%) developed a superior oblique palsy. Parks concluded that the intact intermuscular septum may serve as insertion for the proximal end of the cut tendon, transmitting the superior oblique force to the distal severed tendon. Furthermore, the intact intermuscular septum prevents forward migration of the tendon segments which may alter normal vector forces of the superior oblique muscle.

In an effort to lower the number of overcorrections, in 1987 Parks and Eustis used *a combination of the superior oblique tenotomy* and 14 mm *inferior oblique recession* approaches [37]. Reoperation for overcorrection was not necessary, but inferior oblique underaction was seen postoperatively in 44% of their patients, so that inferior oblique recession was reduced from 14 mm to 10 mm.

Von Noorden reported that 50% of patients operated for congenital Brown syndrome developed the classic features of a superior oblique palsy one year after complete superior oblique tenectomy [46], but this consecutive superior oblique palsy responded well to a subsequently recession of the contralateral inferior rectus muscle or recession of the ipsilateral inferior oblique muscle.

Other authors described irreversible strabismus problems (incomitant vertical deviation with significant torsion, cyclovertical diplopia especially in downgaze, anomalous head postures) following superior oblique tenotomy/ tenectomy even it was combined with recession of inferior oblique muscle [39]. This highlights the importance of selecting patients in whom surgery is truly indicated and of tailoring the surgical procedure to minimize overcorrections.

In 1989 Wright introduced a technique in which a segment of silicone retinal band is sewn between the cut ends of a tenotomized superior oblique tendon to control the amount of weakening [48]. Three of four patients with Brown syndrome showed excellent ocular motility postoperatively and none developed superior oblique palsy. Later results in 2000 [50] of the *silicone superior oblique tendon expander* have shown that 14 of 15 patients improved motility, with normal version in 10 patients, 3 were undercorrected and 2 overcorrected (only one requiring an inferior oblique weakening procedure). The silicon expander provided good results in resolution of the downshoot in adduction. The potential development of downgaze restriction after placement of the expander and in some cases postoperative inflammatory reaction or extrusion of silicone band are a potential disadvantage.

In 1999 Stager reported a relatively high success rate and a low rate of mild undercorrection with expanders [43]. All of the operated eyes had resolution of the downshoot in adduction, but 5 (20%) required reoperation for overcorrection. Some of the patients in Stager's study had 9 or 10 mm spacers, which he discontinued to use because of overcorrection. The overcorrection rate in eyes with 5 to 8 mm expanders was 12.5% (2 of 16 eyes). Comparable with the results of silicone superior oblique tendon expander were the the results of Stolovitch and Leibovitch after the *superior oblique split tendon lengthening*, a technique consisting in a Z–cut across the tendon with end-to-end suturing [44].

In order to avoid the complications of silicone superior oblique tendon expander, like limitation in downgaze or postoperative inflammatory reaction Keskinbora [24], Suh [45] and Yazdian [51] proposed the *superior oblique tendon spacer with application of nonabsorbable adjustable suture* as a lengthening method of superior oblique tendon, a procedure which seems to be technically easier than a silicone expander.

In 1996 Mühlendyck described a tight band at the posterior border of the tendon between the trochlea and sclera, which he found in all 31 patients he has operated for congenital Brown's syndrome [31]. He reported a normalisation of the passive elevation and an improvement of active elevation in adduction after the *superior oblique posterior tenectomy*, altough in 9 cases a second operation was necessary. A consecutive superior oblique palsy was seen only in one case.

This communication of Mühlendyck is not in contradiction with the studies of Crawford and Parks. The latter described a tight superior oblique tendon in only 2 cases of 24 with congenital Brown's syndrome [35]. In Park's cases, the direct visualization of the more posterior part of superior oblique tendon was not possible, because he made always the surgical investigation along the nasal border of the superior rectus muscle and not along the temporal border, as Mühlendyck did [31].

Later on, in 2005 Gräf performed *superior oblique tendon recession* in 22 cases with congenital Brown's syndrome. He reported only a slightly improvement of elevation in adduction (median 5 deg) inspite of free passive motility at the end of the operation [13]. At a late control, in 2-10 years after surgery, the hypotropia (median 0 deg) and the elevation in adduction (median 15 deg) were significantly improved.

Taking into account the hypothesis of co-contraction of both superior and inferior oblique muscle, some authors (Papst and Stein, Neugebauer) proposed a combined surgery with recession of the superior oblique or partial tenotomy of the tendon and tuck in the inferior oblique for diminished innervation of the inferior oblique for elevation and thus less coinnervation of the superior oblique muscle. Neugebauer obtained a reduction of deorsoadduction by 8° in 5 operated cases with this combined technique [32].

4. Questions

As we have seen, various surgical procedures described for the treatment of congenital Brown's syndrome often show disappointing results, some of them followed by severe complications as superior oblique palsy, overcorrection, scarring with limitation of ocular rotations.

Through this study, we try to reevaluate Mühlendyck's results, which have not been confirmed by others authors since their first publication in 1996.

Since 2001, *the superior oblique posterior tenectomy* (Mühlendyck technique) was performed in cases with congenital Brown syndrome in the University Eye Hospital, Ludwig-Maximilians-University Munich, Germany.

Evaluating retrospectively all operated patients with congenital Brown's syndrome, we were especially interested in:

1. Is there a uniform etiological factor in cases of congenital Brown syndrome, like a tight band at the posterior border of the superior oblique tendon, as Mühlendyck described it?

2. Could the *superior oblique posterior tenectomy* significantly improved the passive and the active elevation of the affected eye in adduction and the head posture?

3. Are the results of this procedure comparable to the results of superior oblique tenotomy/recession or silicon superior oblique tendon expander?

5. Patients and methods

5.1. Inclusion criteria

Between 2001 and 2006, 21 patients with congenital Brown's syndrome (23 eyes) were operated using *superior oblique posterior tenectomy* as primary procedure in the Department of Ophthalmology, Ludwig-Maximilians-University Munich.

Before the surgery, all the patients underwent complete ophthalmological and orthoptic examination. Diagnosis was based on active and passive limitation of upward gaze in adduction (Table 1.).

Special care was taken to include only truly congenital cases of Brown syndrome and to avoid mixing with acquired cases. Cases with inferior oblique palsy, blow-out fracture, congenital fibrosis syndrome, monocular elevation deficiency and adherence syndromes were excluded.

Inclusion criteria consisted on:

1. congenital Brown syndrome cases presented with:
    - primary position hypotropia
    - significant anomalous head position
    - or large downshoot in adduction
2. a complete orthoptic examination pre- and postoperatively including:
    - measurements of vertical deviation (VD) and horizontal deviation (HD) in primary position, lateral gaze, up- and down gaze, elevation in adduction
    - testing binocular vision, stereo acuity
    - assesing of anomalous head posture at distance fixation
3. a superior oblique posterior tenectomy as primary procedure performed
4. follow up of at least 3 months postoperatively.

All relevant data of all cases are summarized in table 2 and table 3.

Table 2. Summary of the preoperative findings and postoperative results

Subj	AGE yrs	Sex	Eye	Preop Binocular Vision	Postop Binocular Vision	Preop Head Posture (deg)	Postop Head Posture (deg)	Preop VD in PP (deg)	Postop VD in PP 1 month	Postop VD in PP 3 months
1	2	M	L	-	-	Chin up 15°	Chin up 10°	+20°	+8°	+8°
2	3	M	R	4 in AHP	4 in AHP	Chin up 10°	Chin up 0°	0°	0°	0°
3	4	F	L	3,1 in AHP	in AHP	Chin up 15°	Chin up 2°	+12°	+6°	+8°
4	4	F	L	4	4	Face turn 2°	0°	+1°	0°	0°
5	4	M	L	0	0	0°	0°	+2°	+5°	+3°
6	4	M	R	0in PP, 4 in AHP	4 in PP and AHP	Chin up 15°	Chin up 10°	-9°	-4°	-2°
7	5	F	L	0	0	Head tilt 5°	0°	+13°	+6°	+4°
8	5	F	L	0	1	Chin up 10° Face turn 15° Head tilt 10°	Chin up 0° Face turn 5° Head tilt 2°	+8°	+5°	+7°
9	5	M	R	0	1	Face turn 5°	Face turn 2°	-5°	0°	0°
10	6	F	R	0	0	Head tilt 10°	0°	-12°	-7°	-7°
11	6	F	L	0	1	0°	0°	+3°	0°	0°
12	6	M	R/L	4 in AHP	4 in AHP	Chin up 15°	Chin up 2°	-1°	0°	0°
13	7	M	L	0	0	Head tilt 2°	0°	+6°	+5°	+4°
14	7	M	L	0	0	0°	0°	+10°	+5°	+5°

15	8	F	L	4	4	Chin up 10° Face turn 15°	0°	0°	0°	0°
16	11	M	R	4 in AHP	4 in AHP	Chin up 15°	Chin up 5°	-13°	-2°	-2°
17	11	M	L	1 in PP, 4 in AHP	4 in PP	Chin up 10° Head tilt 10°	0°	+5°	+1°	+1°
18	12	M	R / L	3,1 in AHP	2 in AHP	Chin up 10° Face turn 2°	0°	-1°	0°	0°
19	17	M	L	4 in AHP	4 in AHP	Face turn 10°	0°	+1°	+3°	+5°
20	24	M	R	4 in AHP	2 in AHP	Chin up 10°	Chin up 5°	-15°	-7°	-8°
21	29	F	L	3,4	3,5	0°	0°	+13°	+4°	+4°

Abbreviations: R - right eye, L - left eye, AHP - abnormal head position, PP - primary position, VD - vertical deviation, deg - degrees
Binocular vision: 0 - suppression, 1 - Bagolini positive, 2 - stereo Fly positive, 3 - stereo circles positive (number of circles +), 4 - Lang II positive
Vertical deviation: –VD deg when the right eye is involved and +VD deg when the left eye is involved

Legend - table 3:
Monocular elevation (ME) in adduction: mm below(–) or over horizontal (+)
Passive motility in adduction: 0 – free, 1 - almost free, 2 - mild restriction, 3 – severe restriction
Tightness of the posterior part of the SO tendon: 0 - normal posterior tendon, 1 - slightly tight, 2 - tight, 3 - very tight
Subjective results: 2 - excellent results, 1 - satisfactory results, 0 - no changes, -1 - worse results

Table 3. Summary of the preoperative findings and postoperative results

Subj	Preop ME (mm) R/L	Postop ME 1 month	Postop ME 3 months	Preop passive motility R/L	Postop passive motility R/L	Tight posterior part of the SO tendon R/L	Subjective results
1	-2	-1	-1	3	3	3	0
2	-1	1	1	3	0	2	0
3	1	2	1.5	3	1	2	-1
4	-1	0	-1	3	1	3	0
5	-1	1.5	1.5	3	0	1	0
6	0	1.5	1.5	3	0	3	0
7	3	5	5	3	1	2	2
8	0	4	3	3	2	3	2
9	2.5	5	5	3	0	2	1
10	1	4	5	3	0	3	0
11	0	3	3	3	0	3	2
12	0/0	3/3	4/2	3/2	1/1	2/2	2
13	0	3	1	3	0	3	1
14	1	2	2	3	0	3	1
15	0	2.5	2.5	3	0	3	2
16	1	2	2	3	0	3	1
17	0	2	2	3	1	0	1
18	1.5/0	4/2.5	4/2.5	3/3	0/0	2/3	2
19	1.5	2	2	3	0	1	2
20	0	5	5	3	0	3	2
21	1.5	2.5	2.5	3	1	3	2

5.2. Methods

Before the operation, one month and three months after the operation, prism alternate cover test in five positions of gaze was used to determine the horizontal (HD) and vertical deviation (VD). In 2 children of 2 and 3 years old who could not cooperate sufficiently to allow cover testing, the horizontal and vertical deviation were estimated using Hirschberg's method, based on the premise that 1 mm of decentration of the corneal light reflection corresponds to about 7 deg.
We noted –VD(deg) when Brown's syndrome involved the right eye and +VD when the left eye was affected.

The monocular elevation in adduction was measured in mm below(–) or over horizontal (+).

Because most of the patients were children - some with poor cooperation or exclusion of the dark red light, it was not possible to measure the cyclodeviation with Harms' tangent screen.

The binocular vision was assesed using Bagolini glases, Titmus stereotest and Lang II test. A numeric rating was used to describe the degree of binocular vision:

- 0 - suppression
- 1 - Bagolini positive
- 2 - stereo Fly positive
- 3 - stereo circles positive, number of circles +
- 4 - Lang II positive

The presence of binocular vision detected only in abnormal head position (AHP) was noted.

Intraoperative forced ductions test showed restriction to elevation in adduction in all operated subjects. The preoperatively and postoperatively passive motility in adduction was evaluated as follows:

- 0 - free
- 1 - almost free
- 2 - mild restriction on attempted elevation in adduction
- 3 - severe restriction on attempted elevation in adduction

The tightness of the posterior part of the tendon was assesed using the following numeric rating :

- 0 - normal posterior tendon
- 1 - slightly tight posterior tendon
- 2 - tight posterior tendon
- 3 - very tight posterior tendon

On the patients with associated eso-/exotropia, the correction for horizontal deviations was performed together with superior oblique posterior tenectomy.
In 2 cases, superior oblique tendon recession was performed as an additional procedure after superior oblique posterior tenectomy and in 1 case a superior rectus resection as a secondary procedure was necessary.

At three months after operation, the postoperative results were estimated subjectively by the parents as follows:

- 2 - excellent results
- 1 - satisfactory results
- 0 - no changes
- -1 - worse results

All 21 patients with congenital Brown's syndrome were followed for at least 3 months postoperatively. A long-term follow up for 8 cases ranged from 6 months to 24 months.

5.3. Surgical approach

5.3.1. **Forced duction test**

Before the operation, under general anesthesia, *a forced duction test* was performed with the eyelids separated by an eyelid speculum. Using two forceps, the globe was grasped at the 4 and 10 o'clock limbal position for right eye and at the 2 and 8 o'clock limbal position for left eye and an attempt was made to elevate it in adduction.

In every operated case, passive elevation in adduction was restricted, whereas passive elevation and depression in both the abducted and midline positions were checked and confirmed to be normal. Forced retroplacement of the globe with forceps during attempted passive elevation in adduction places the superior oblique muscle on strech and accentuates the restriction in Brown syndrome.

5.3.2. **Surgical technique**

The surgical approach for superior oblique posterior tenectomy was that described by Mühlendyck [31].

An incision is made through the conjunctiva and Tenon's capsule in the upper temporal quadrant, close to the temporal border of the superior rectus muscle.

One muscle hook engages the superior rectus muscle and the eye is turned downward and inward by applying traction on the muscle hook, held by the assistant. The upper edge of the wound is retracted upward using a small Desmarres lid retractor. The tendon of the superior oblique can be seen as a glistening white band. A small Graefe hook is used to engage the anterior part of the tendon and a traction silk suture can be passed through it and then pull forward. The posterior part of the tendon is engaged on a muscle hook. In each operated case this 1/3 anterior part of the tendon proved to be slack comparing with the 2/3 posterior part of the tendon, which was found tight.

After the excision of a 8 mm band of the posterior superior oblique tendon from the insertion, the muscle hook under the superior rectus muscle is removed and the conjunctival wound is closed with 3 stitches of 7-0 Vicryl.

At the end of operation, the forced duction test is perfomed to determine the degree of restriction postoperatively when elevating the adducted eye.

6. Results

6.1. Epidemiologic features

6.1.1. **Age**

For 21 cases with congenital Brown's syndrome operated between 2001 and 2006, in the Department of Ophthalmology, Ludwig-Maximilians-University Munich, the median age at the time of surgery was 8.5 years.

The youngest patient was 2 years old when the surgery was performed - the operation was indicated so early because it was a severe form of congenital Brown's syndrome with +20°VD in primary position and the monocular elevation in adduction was 1.5 mm below the horizontal. The oldest pacient operated was 29 years old.

71% of operated patients with congenital Brown syndrome were over 5 years old.

6.1.2. **Laterality**

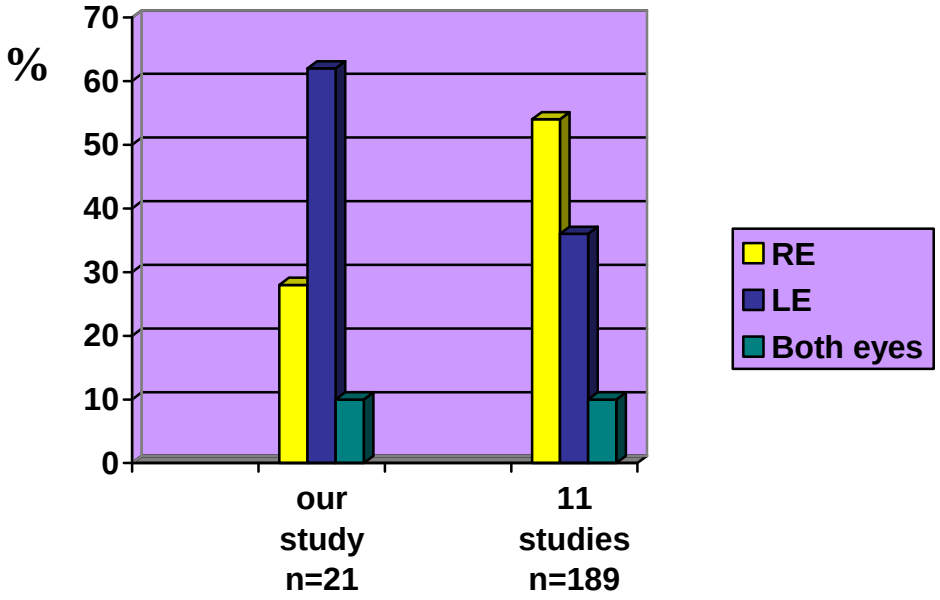
From the analyze of our series of 21 patients with congenital Brown's syndrome resulted that the left eye (LE) was affected in 62% of cases, the right eye (RE) in 28%, both eyes in 10%.

At the 11th Meeting of Bielschowsky Society in 2005, Neugebauer presented a review of 11 studies pooled from the literature - especially the highly elaborated reviews of Brown, Wright, Wilson and herself (totally 189 cases with congenital Brown's syndrome). In a total of these 11 studies pooled from the literature by Neugebauer [32], the right side was affected in 54%, the left side in 36% and both sides in 10%.

Comparing our series with the data of Neugebauer (Fig. 4), we remark in our study a tendency to affect rather the left eye, probably due to the lower number of patients included in our study.

Many studies [46, 47], like our results showed that Brown syndrome may be bilateral in approximately 10% of cases.

Fig. 4. Comparison between our study and 11 studies (Neugebauer)



6.1.3. **Sex distribution**

Regarding the sex distribution, our study showed the affection of 62% males and 38% females, while the data of 11 studies showed a affection of 55% females and 45% males.

6.1.4. **Heredity**

Most cases of congenital Brown's syndrome seem to occur spontaneously.

The few familial cases have led authors to postulate autosomal dominant inheritance with incomplete penetrance and variable expression, although recessive transmission has been proposed in some cases [29, 30].

In our retrospective study, none of 21 operated patients with congenital Brown's syndrome doesn't show inheritance.

6.1.5. **Comorbidity**

Brown syndrome occurs, as a rule, as an isolated anomaly. However, exceptions have been reported, including an association with Duane's syndrome, crocodile tears, unilateral congenital ptosis, Marcus-Gunn jaw-winking, thus entities that are in context with congenital cranial misinnervation disorders [32, 46, 47].

Several reports exist that describe patients with congenital Brown's syndrome on one side and trochlear palsy or at least strabismus sursoadductorius on the fellow eye [6, 32].

In our series of 21 patients, no association with any ocular syndrome enumerated above has been noted.

6.2. Coexisting horizontal strabismus and amblyopia

Regarding coexisting horizontal strabismus, of 21 cases with congenital Brown's syndrome operated between 2001 and 2006, 6 presented associated esotropia and 1 exotropia (Table 3). In all 7 cases, the correction for horizontal deviations was performed together with superior oblique posterior tenectomy.

Regarding amblyopia, we noticed at the time of surgery a visual acuity difference more than 1 line in only 3 patients (a incidence of amblyopia of 14% at the time of surgery), although 11 patients (52%) have patched one eye to improved the visual acuity in a mean period of 2.8 years (range 0.5 years to 5 years). Probably, the true incidence of amblyopia is higher than reported in some studies, because many cases have already improved the visual acuity when the surgery was performed.

6.3. Fusion

Brown reported fusion in some field of gaze in 98 of his series of 126 patients[4]. Gräf noted the highest degree of stereopsis (Lang I Stereotest, Titmus-Test) preoperatively and postoperatively in 11 of 22 patients with congenital Brown's syndrome and lower degree of stereopsis in 2 cases before and 3 cases after superior oblique tendon recession [13].

Preoperatively, in our study, 12 of 21 patients presented some degrees of binocular vision:

- 4 patients in primary position (3 the highest degree of stereopsis and 1 only Bagolini positive)
- 8 patients only in abnormal head position (and all of them fine stereopsis)
- the rest of the patients presented suppression before the surgery.

Postoperatively, after the superior oblique posterior tenectomy, we noticed that:

- 3 patients gained Bagolini positive and 1 the highest degree of stereopsis in primary position.
- 2 patients improved the binocular vision in primary position and 1 in compensatory head position.

In 2 patients of this series of 21, we remarked that the fusion was worse through the operation.

6.4. Abnormal head position

Head postures (chin up, face turn, head tilt) were evaluated in all patients with congenital Brown's syndrome pre- and postoperatively (Fig. 5a, 5b, 5c).

A compensatory head position was remarked before superior oblique posterior tenectomy on 17 patients (81%):

- most of them - 7 (34%) presented a chin up position of 10° to 15°
- 3 (14%) presented a face turn away from the affected eye of 2° to 10°
- 3 patients (14%) a head tilt to the shoulder on the side of the affected eye of 2° to 10°
- 4 (19%) a combined compensatory head posture (chin up ± head tilt ±face turn)

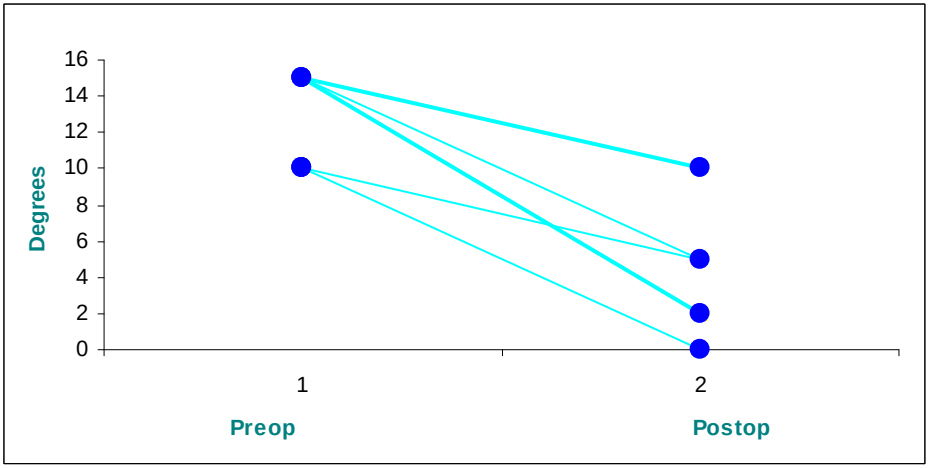
Postoperatively, of 17 patients with compensatory head position:

- 12 had a complete resolution of anomalous head posturing
- 5 patients improved their head posture.

We noticed in those 5 cases with improvement of head posture :

- 2 had after operation a slight chin up position of 5°
- 2 presented postoperatively a chin up position of 10°
- 1 patients who had before operation a combined compensatory head posture (chin up and head tilt of 10° and face turn of 15°) remained with a face turn away from the affected eye of 5° and a head tilt of 2°

Fig. 5a. Comparison between preoperative and postoperative chin up position



Legend: Thick line – more patients with same data

Fig. 5b. Comparison between preoperative and postoperative face turn

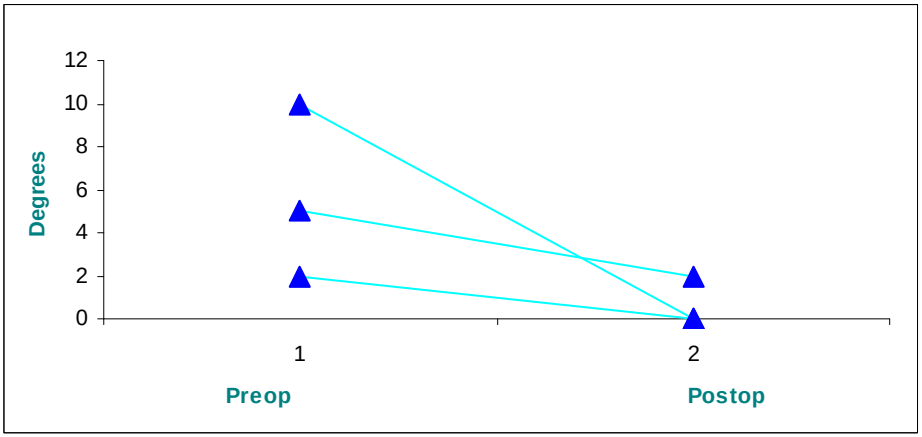
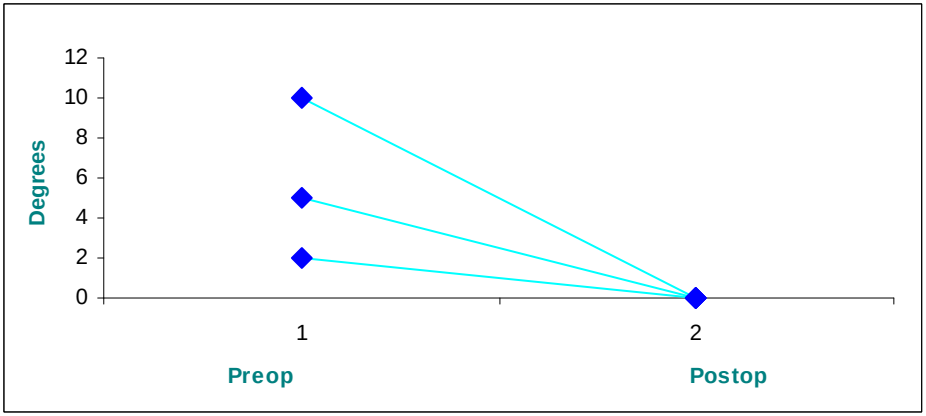


Fig. 5c. Comparison between preoperative and postoperative head tilt



Using Parametric Paired T-test, we compared pre- and postoperative abnormal head position. Between the 3 types of compensatory head posture (chin up, face turn, head tilt), we found statistically significant difference before and after operation. The chin up position decreased statistically significant after superior oblique posterior tenectomy (p=0.001).
For the other two types of posture (face turn, head tilt), even p-values are very close to the significance threshold 0.05 (p=0.043 for "face turn" and p=0.037 for "head tilt"), the affirmation of the existence of a statistically significant difference pre- and postoperatively must be made with prudence.

6.5. Intraoperative findings

6.5.1. **Passive motility**

In our series of 21 patients (23 eyes) with congenital Brown's syndrome, preoperative forced ductions test proved:

- on 22 eyes severe restriction to elevation in adduction
- on 1 eye mild restriction

After superior oblique posterior tenectomy, passive motility became free or almost free on the majority of operated patients (21 eyes). In 1 eye, on attempted elevation in adduction we noted postoperatively unchanged severe restriction inspite of superior oblique posteror tenectomy and in another eye mild restriction.

6.5.2. **The tightness of the posterior part of the superior oblique tendon**

In a series of 31 cases with congenital Brown's syndrome operated between 1980 and 1995, Mühlendyck found a tight band at the posterior border of the superior oblique tendon between the trochlea and the sclera in all patients [31].

In our study on 23 eyes (Fig. 6), the data found in clinical records showed that posterior part of the superior oblique tendon was found:

- very tight in 13 eyes
- tight in 7 eyes
- slightly tight in 2 eyes
- normal in 1 eye

In summary, in our study, a tight or very tight posterior part of the superior oblique tendon was found in 87 % of operated eyes with congenital Brown's syndrome.

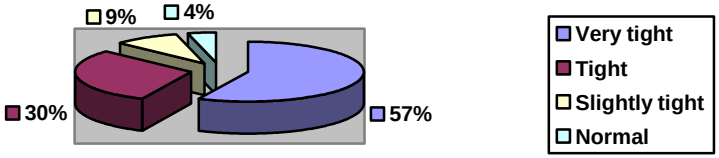


Fig. 6. The tightness of the posterior part of the tendon

By using nonparametric Spearman correlation test, we tried to correlate the tightness of the posterior part of the superior oblique tendon with preoperative, early postoperative or late postoperative VD in primary position. We found a moderate correlation, statistically significant for a risk level of 0.05 between the superior oblique tendon tightness and VD in primary position ($p=0.023$ for preoperative VD, $p=0.03$ for early postoperative VD in primary position, $p=0.018$ for late postoperative VD in primary position). Late postoperatively, the statistical significance of p-value is improved ($p=0.018$) and we remarked a good correlation between the SO tendon tightness and the VD in primary position.

By using nonparametric Spearman test, we noted a moderate correlation between SO tendon tightness found intraoperatively and binocular vision improvement ($p=0.033$) or head position improvement after superior oblique posterior tenectomy ($p=0.035$).

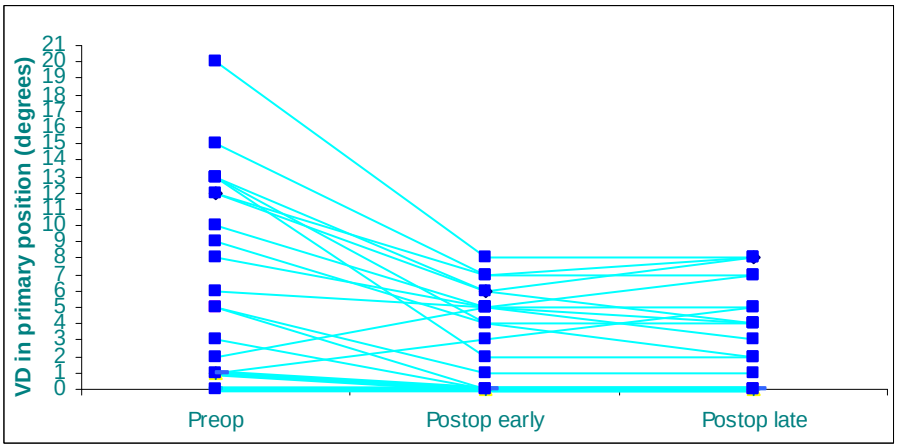
6.6. Alignment of the eyes and ocular rotations

6.6.1. **Vertical deviation in primary position**

Before the operation, the vertical deviation (VD) in primary position varied from 0 to 20 deg hypotropia (mean 7 deg) - Table 2.
Early postoperatively, one month after superior oblique posterior tenectomy, the vertical deviation in primary position was 0 to 8 deg hypotropia (mean 3 deg) - Fig. 7a, 7b.
Late postoperatively, three months after superior oblique posterior tenectomy, the vertical deviation in primary position was 0 to 8 deg hypotropia (mean 3 deg).

Postoperatively, we saw no overcorrections in our cohort.

Fig. 7a, 7b. Comparison between preoperative and early/late postoperative vertical deviation in primary position (1 month/3 months)



Legend: Thick line – more patients with same data

Fig. 7a

The graph called box plots (Fig. 7b) contains the middle 50% of the data. The line in the box indicates the median value of the data.

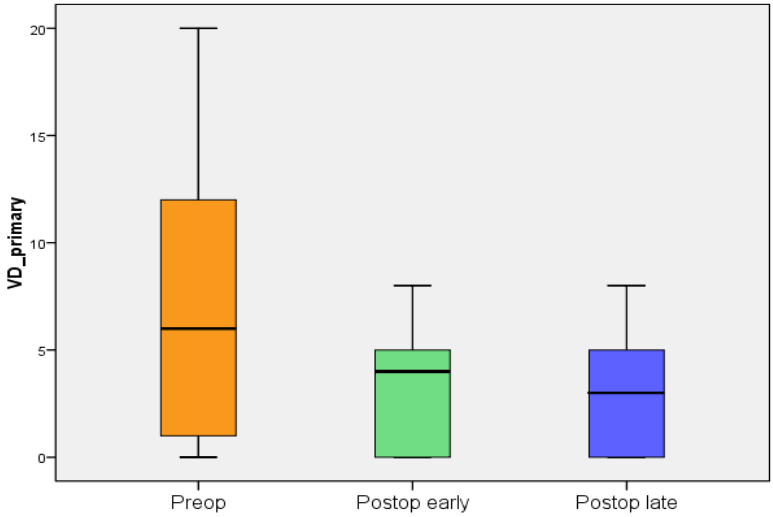


Fig. 7b

Analyzing the distribution of vertical deviation in primary position provided by box plots (Fig. 7b), we remarked a decreased value of hypotropia obtained after superior oblique posterior tenectomy. No significant differences between the early postoperative VD and the late postoperative VD can be identified.

By applying the One-way ANOVA multivariate test, we found that vertical deviation (VD) in primary position differs statistically significant between the preoperative group and early postoperative group (P=0.011) and between the preoperative group and late postoperative group (P=0.011), the VD in primary position having a constant trend in decreasing the value through the operation.

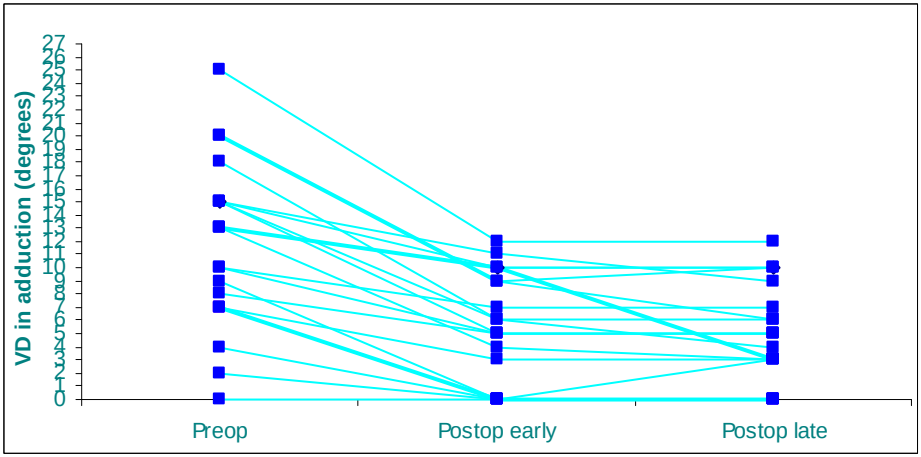
The early postoperative and late postoperative groups do not differ statistically significant.

6.6.2. **Vertical deviation in adduction**

Preoperatively, the vertical deviation (VD) in adduction measured with prism alternate cover test varied from 0 to 25 deg hypotropia (mean 11.5 deg).
Early postoperatively, vertival deviation in adduction was 0 to 12 deg hypotropia (mean 5 deg) - Fig. 8a, 8b.
Three months after superior oblique posterior tenectomy, the vertical deviation in adduction was 0 to 12 deg hypotropia (mean 5 deg).

Postoperatively, we have not noticed any consecutive superior oblique palsy (i.e. depression deficit in adduction) in our cohort.

Fig. 8a, 8b. Comparison between preoperative and early/late postoperative vertical deviation in adduction (1 month/3 months)



Legend: Thick line – more patients with same data

Fig. 8a

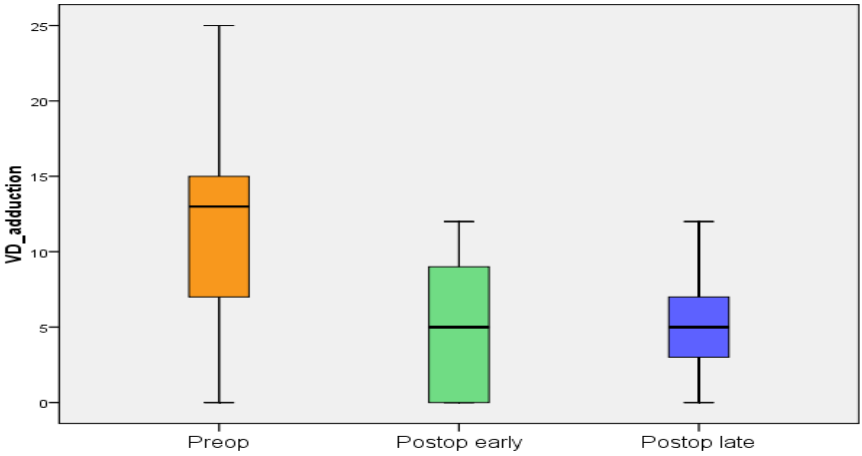


Fig. 8b

Comparing the distribution of VD in adduction between preoperative-early postoperative-late postoperative group (Fig. 8b), we noted a decreased value of hypotropia in adduction obtained after superior oblique posterior tenectomy. No significant differences between the early postoperative VD in adduction and the late postoperative VD in adduction can be identified.

By applying the One-way ANOVA multivariate test, we found that vertical deviation (VD) in adduction differs statistically significant between preoperative group and early postoperative group (P=0.001) and between preoperative group and late postoperative group (P=0.001).

There is no statistically significant difference between early postoperative results (at 1 month) and late postoperative results (at 3 months) regarding hypotropia in adduction.

6.6.3. **Monocular elevation in adduction**

The monocular elevation in adduction varied from -2 mm below horizontal to 2.5 mm over horizontal (mean 0.5 mm) before the operation (Table 3).

Postoperatively, the mean value of the monocular elevation in adduction was 2.5 mm (range -1 mm to 5 mm).

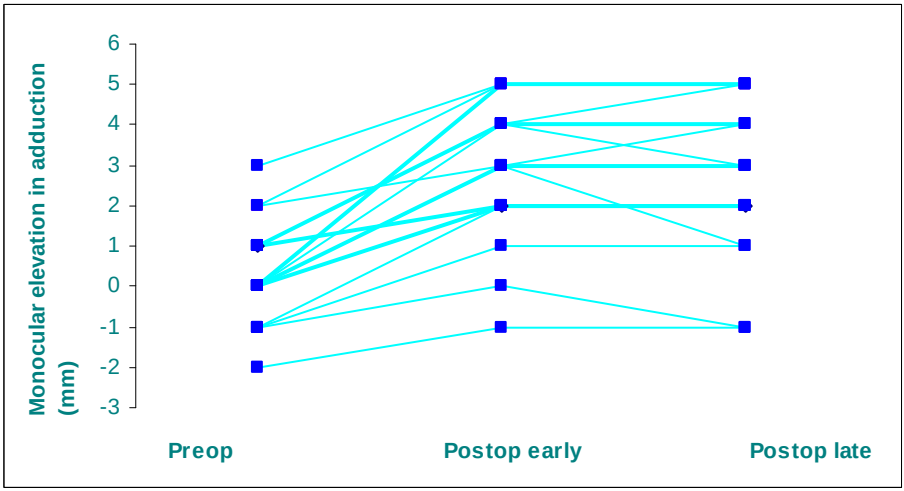
Inspite of free or almost free passive motility at the end of the operation in the majority of patients, we remarked only a slightly improvement of the active monocular elevation in adduction of 0.5 mm to 5 mm (mean 2.25 mm).

At three months postoperatively, 11 cases (52%) had an improvement of active monocular elevation in adduction of 1 mm to 3 mm. 6 patients (29%) had an improvement of more than 3 mm and 4 patients (19%) showed no improvement.

No patient achieved free active elevation in adduction postoperatively.

Regarding the distribution of monocular elevation in adduction in our lot of patients with congenital Brown's syndrome (Fig. 9b), we remarked an increased value of active monocular elevation in adduction obtained after superior oblique posterior tenectomy. The median value of monocular elevation in adduction had slightly decreased in the late postoperative period comparing with early postoperative period. But applying the One-way ANOVA multivariate test, we found that the early postoperative and late postoperative groups do not differ statistically significant (p=0.001).

Fig. 9a, 9b. Comparison between preoperative and early/late postoperative monocular elevation in adduction (1 month/3 months)



Legend: Thick line – more patients with same data

Fig. 9a

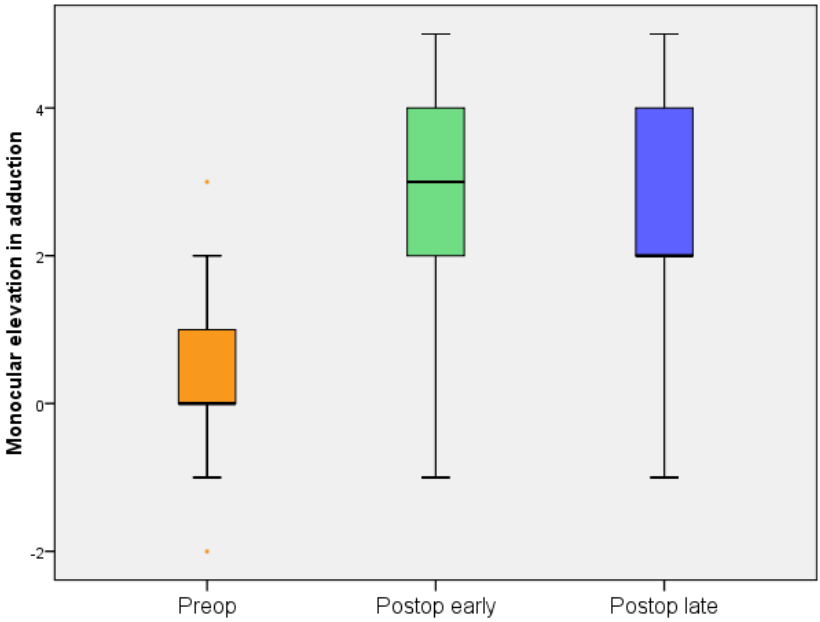


Fig. 9b

Monocular elevation in adduction differs statistically significant between the preoperative group and early postoperative group (P=0.001) and between the preoperative group and late postoperative group (P=0.001).

As a conclusion, hypotropia in primary position and in adduction decreased significantly and monocular elevation in adduction increased significantly. There was no significant difference between early and late postoperative results.

6.6.4. **Monocular elevation in abduction**

The monocular elevation in abduction was measured in mm below/over horizontal on only 12 patients of 21. The results showed:

- unchanged measurements before and after operation in 9 cases (75%)
- an improvement of monocular elevation in abduction in 3 cases (25%) with 1.5 mm to 3.5 mm (mean 2.16 mm).

6.6.5. **V or A pattern**

Using prism alternate cover test, we evaluated the changes in magnitude of horizontal deviation (HD) in gaze up 25°, primary position and gaze down 25°.

A V-pattern in which the difference in horizontal deviation between upward and downward gaze is 15Δ (7.5°) or more is considered clinically significant.

Any A-pattern is considered clinically significant if it measure 10Δ (5°) or more difference between downgaze and upgaze.

In Y-pattern, we notice a difference between primary position and upgaze, with increasing divergence and no difference between primary position and downgaze.

In our study, we noted preoperatively the presence of a clinically significant V-pattern on 6 patients of 21 operated and a clinically significant A-pattern on 3 cases of 21. 3 patients presented Y-pattern.

Postoperatively, a V-pattern was remarked on 1 patient and an A-pattern in 1.

The 2 cases in which the A-pattern resolved through the operation gained binocular vision in primary position (Bagolini positive) and the parents have seen excellent results after surgery.

On 5 cases in which V-pattern resolved through the operation, we noted a correction of abnormal head position with an improvement of monocular elevation in adduction of 2 mm to 5 mm, while the parents reported excellent results in 3 cases and no changes after surgery in 2 cases.

6.6.6. **Consecutive superior oblique palsy**

In a series of 31 patients operated for congenital Brown's syndrome, after superior oblique posterior tenectomy, Mühlendyck reported only one case with consecutive superior oblique palsy [31].

In our series, none of the 21 operated patients developed consecutive superior oblique palsy (i.e. depression deficit in adduction) at the follow-up of 3 months postoperatively.

6.6.7. **Long-term follow-up**

Eight patients were examined 6 - 24 months after primary procedure.

On 5 patients with congenital Brown's syndrome, the measurements of the vertical deviation in primary position, the monocular elevation in adduction and head position remained unchanged at the late postoperative control.

Three patients had further surgery (2 cases - superior oblique recession and 1 case - superior rectus resection). We noticed in these 3 cases an improvement of:

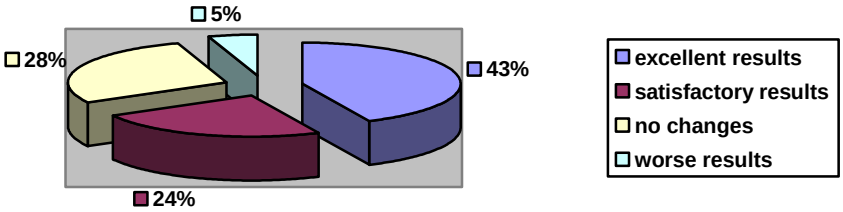
- the vertical deviation in primary position of 4 deg to 7 deg
- the monocular elevation in adduction of 1mm to 3mm.

6.7. Subjective results

At three months after superior oblique posterior tenectomy performed on 21 patients with congenital Brown's syndrome, the postoperative results were judged subjectively by the parents as follows:

- excellent results in 9 cases (43%)
- satisfactory results in 5 cases (24%)
- no changes in 6 cases (28%)
- worse results in 1 patient (5%)

Fig. 10. Subjective results after superior oblique posterior tenectomy



Of those 6 patients with no remarkable changes after surgery, the surgeon noticed intraoperatively a very tight tendon on 4 patients and a slightly tight one on 2 patients. After the operation, the passive motility proved to be free in 4 cases, almost free in 1 case and in the last case remained severe restriction.

The most important problem that we had to face was linked to the differentiation in quantification of the subjective perception of the postoperative results and those of head position. To establish a uniform appreciation we have allocated some scores of the variation of the head position (0 for insufficient improvement, 1 for substantial improvement, 2 for very good improvement) and we correlate them with the scores of the subjective results (0 for no chages, 1 for satisfactory results, 2 for excellent results). By applying a nonparametric Spearman correlation between the two scores, we noted a moderate correlation between the subjective results and the improvement of head position through operation (p=0.039).

6.8. Distribution of cases depending on vertical deviation in primary position

Of 21 patients who underwent surgery for congenital Brown's syndrome:

- 13 had a VD of <10° in primary position preoperatively (group I)
- 8 had a VD ≥10° in primary position before the operation (group II).

In these 2 groups, we evaluate postoperatively mean VD in straight gaze, the improvement of monocular elevation in adduction (Table 4), the abnormal head position, the passive motility in adduction and the tightness of the posterior part of the superior oblique tendon.

Table 4. Mean preoperative and postoperative results in group I and II

	Mean preop VD in primary position (°)	Mean postop VD in primary position (°)	Mean improvement of monocular elevation in adduction (mm)
Group I, n=13 (preop VD <10°)	3.2°	1.6°	2.1 mm
Group II, n=8 (preop VD ≥10°)	13.5°	5.7°	1.9 mm

Although the mean improvement of monocular elevation in adduction was not significantly different in the two groups, we remark a most pronounced effect of superior oblique posterior tenectomy in reducing VD in primary position in the group II (mean improvement of VD in straight gaze by 7.8°) comparing to group I (mean improvement of VD in primary position by 1.6°).

Intraoperatively, both groups showed a tight or a very tight posterior tendon:

- 10 of 13 cases in group I
- all 8 cases in group II.

In 2 cases from group I, in which the posterior part of the tendon was found slightly tight, we noticed that VD in primary position increased slightly after superior oblique posterior tenectomy. In these 2 cases, a further surgery (recession of the superior oblique tendon) was performed and we noted an improvement of the vertical deviation in primary position of 4 deg to 7 deg and an improvement of the monocular elevation in adduction of 1 mm to 3 mm.

Regarding postoperative passive motility in adduction, we found similar results in both groups:

- in group I, 11 cases (85%) proved free/almost free passive motility after operation and a mild restriction in 2 cases (15%)
- in group II, 7 cases (88%) showed free/almost free passive motility postoperatively and in one case (12%) severe restriction to elevation in adduction, maybe due to some inferior restrictive bands.

Also the postoperative findings regarding compensatory head position were very similar in both groups :

- 9 patients of group I corrected totally abnormal head position and 2 improved it postoperatively
- 4 cases of group II corrected the abnormal head posture, 1 improved it after operation and in 1 case in which we remarked severe restriction to elevation in adduction also postoperatively, the abnormal head position remained unchanged.

Regarding fusion after operation:

- in group I, 5 patients of 13 improved binocular vision after superior oblique posterior tenectomy and all of them in primary position
- in group II, only 2 patients of 8 improved the binocularity, 1 in primary position and 1 in compensatory head position

These last results showed a better chance to improve binocularity after superior oblique posterior tenectomy when the initial hypotropia in primary position is smaller.

7. Discussion

7.1. Indication for surgery

Although the etiopathology of congenital Brown's syndrome proved to be heterogenous [3, 4, 9, 16, 18, 25, 31] and several surgical techniques were developed by many authors, the indications for surgery remained unanimous valid for all surgeons [13, 23, 31, 32, 46, 47]:

When binocular vision is normal and comfortable with the eye aligned in primary position without an extreme head posture, surgery will not be recommended. On the other hand, when there is a significant anomalous head posture, when the involved eye is hypotropic in primary position, binocular vision is impaired and this entity does not resolve spontaneously until the age of 6-10 years, surgery should be considered. If a patient needs a correction of eso-/exotropia, the associated limitation of elevation in adduction could be corrected in the same surgery with the horizontal deviation.

7.1.1. Spontaneous resolution of congenital Brown's syndrome

Most of the studies showed that spontaneous improvement occurred in almost half of patients with constant congenital Brown syndrome [21, 46, 47]. Others authors (Lee) reported an even higher percentage of spontaneous improvement, up to 75 % [8] and for this reason he recommended to adopt "wait and see" approach with these children.

Only few older patients with congenital Brown's syndrome require operation. This may be another argument for the high rate of spontaneous resolution of this ocular motility disorder. An easy explanation for the few cases of congenital Brown syndrome accidentally discovered at adult age could be the fact that the adults rarely elevate the eyes in normal life and for this reason, the moderate forms of the syndrome could be easily overlooked.

In our study on 21 patients with congenital Brown's syndrome, most patients were under 8 years of age. Only 6 cases were aged 11, 12, 17, 24, 29 years. The youngest patient was 2 years old when the surgery was performed – the operation was indicated so early because it was a severe form of congenital Brown's syndrome with 20 deg hypotropia in primary position and the monocular elevation in adduction was 1.5 mm below the horizontal. Of 17 operated children under 8 years of age, 7 had a coexisting eso-/exotropia and the associated limitation of elevation in adduction has been resolved in the same surgery with the correction of horizontal deviation.

7.2. Laterality and sex predilection

Despite Brown's original impression [4] that the condition occurs more often in females (58%) and in the right eye (62%), *subsequent reports have failed to substantiate a sex or laterality predilection [8, 13, 46, 47]*.

In our series of 21 patients with congenital Brown's syndrome, the left eye was affected in 62% of cases, the right eye in 28%, both eyes in 10% (Fig. 4).

At the 11th Meeting of Bielschowsky Society in 2005, Neugebauer presented a review of 11 studies pooled from the literature - especially the highly elaborated reviews of Brown, Wright, Wilson and herself (totally 189 cases with congenital Brown's syndrome). In a total of these 11 studies pooled from the literature by Neugebauer [32], the right side was affected in 54%, the left side in 36% and both sides in 10% - Fig. 4.

Many studies [46, 47], like our results showed that Brown syndrome may be bilateral in approximately 10% of cases.

Under the same aspects, sex distribution was analyzed: our study showed the affection of 62% males and 38% females, while the data of 11 studies showed a affection of 55% females and 45% males. This tendency to affect rather the males was reported also by Gräf [13], who noted that of 22 operated patients with congenital Brown's syndrome, 13 were males (59%). But in the same study, he reported that the right eye was prevalent involved (68%).

In summary, there is no strong evidence to support a statement on either laterality or sex predilection.

Using Two-Sample Kolmogorov-Smirnov test, we checked up the similarity between the distribution of the eyes (LE-RE-both eyes) between our lot (n=21) and Neugebauer lot (n=189).

The Two-Sample Kolmogorov-Smirnov test presumes as null hypothesis the identity between the two distributions of frequencies. The result suggests that we can accept this presumption, so the distributions of the affected eye (LE-RE-both eyes) between the two cohorts are statistically similar (P=0.996). There are no statistically significant differences between the proportions of the affected eye between our lot of patients and the Neugebauer lot of patients with congenital Brown's syndrome.

Using the same Two-Sample Kolmogorov-Smirnov test, we verified the similarity between the distribution of women-men in our lot (n=21) and Neugebauer lot (n=189). The distributions of the frequencies of women and men respectively, in these 2 lots of patients with congenital Brown's syndrome are statistically similar (P=0.964).

7.3. Heredity

Of a series of 126 cases, Brown reported only 2 familial cases [4]. Wright reported in his own study a incidence of inheritance of 3% in Brown's syndrome [46]. Many others authors (Parks and Eustis, Magli, Lowe, Katz) described familial occurrence and mirror reversal which was observed in monozygotic twins [22, 27, 29, 37].

The few familial cases have led authors to postulate autosomal dominant inheritance with incomplete penetrance and variable expression, although recessive transmission has been proposed in some cases [30].

However, most cases of congenital Brown's syndrome seems to be sporadic. Also in our retrospective study, none of 21 operated patients with congenital Brown's syndrome did show a positive family history. Further pedigree and family population studies are needed to prove a mode of inheritance.

7.4. Comorbidity

Brown's syndrome occurs, as a rule, as an isolated anomaly. However, exceptions have been reported, including an association with Duane's syndrome, crocodile tears, unilateral congenital ptosis, Marcus-Gunn jaw-winking, thus entities that are in context with congenital cranial dysinnervation disorders (CCDD) [32, 46, 47].

Several reports exist that describe patients with congenital Brown's syndrome on one side and trochlear palsy or at least strabismus sursoadductorius on the fellow eye [6, 32].

The coincidence with congenital fourth nerve palsy is a strong argument supporting the theory that congenital Brown syndrome could be a misinnervation syndrome (Neugebauer). If congenital Brown's syndrome would be a primary developmental defect of the fourth nerve nucleus with paradoxical innervation of the superior oblique, one can imagine cases in which developmental defect occurs, but coinnervation is not established sufficiently [32].

In our series of 21 patients, no association with any ocular syndrome enumerated above has been noted.

Brown and later Mühlendyck [4, 31] described the coexistence of congenital Brown syndrome with an ipsilateral superior rectus palsy**,** this association being found also in one patient of our study. After performing a 3.5 mm superior rectus resection as a secondary procedure, we remarked an improvement of active monocular elevation in adduction from 4 mm to 7 mm. Gräf however postuled that following a long lasting limitation of elevation, structural changes of superior rectus muscle and even a secondary contraction of inferior rectus could appear, explaining the further limitation of elevation after superior oblique muscle sugery [13].

7.5. Coexisting horizontal strabismus and amblyopia

In congenital Brown's syndrome, Eustis noted a 15% incidence of coexisting horizontal strabismus [47]. Sanford-Smith warned that a threat to binocular vision exists in these patients and he reported that 6 of 19 patients developed secondary esotropia [38].

We found a similar percent of horizontal strabismus in our study: of 21 cases, 6 presented with associated esotropia and 1 with exotropia. In all 7 cases, the correction for horizontal deviations was performed together with superior oblique posterior tenectomy. Postoperatively, only 2 cases gained Bagolini positive, the rest of the patients presented suppression before and after the surgery (Table 2).

Regarding amblyopia, Brown found it to be insignificant in his series of 126 patients. Clark and Noel reported 7 cases with amblyopia in their series of 28 Brown's syndrome patients [6]. Gräf found amblyopia in only 2 patients of 22 with congenital Brown's syndrome he operated [13].

In our study, we noticed at the time of surgery a visual acuity difference more than 1 line in only 3 patients (a incidence of amblyopia of 14% at the time of surgery). However 11 patients (52%) had patched one eye to improved the visual acuity in a mean period of 2.8 years (range 0.5 years to 5 years). Probably, the true incidence of amblyopia is higher than reported in some studies, because many cases have already improved the visual acuity when the surgery was performed.

7.6. Abnormal head position and binocular vision

Preoperatively, in our study an abnormal head position was noted in 17 patients (81%) of 21 with congenital Brown's syndrome.

Regarding head posture after superior oblique posterior tenectomy, we noted either a complete resolution (12 cases) or an improvement of the compensatory head position (5 cases) in all operated patients ($p=0.001$). It is remarkable that all cases with face turn or head tilt totally resolved the compensatory posture, while most cases with chin up position improved only partially after superior oblique surgery (Fig. 5a, 5b, 5c).

In spite of the minimal-to-no anomalous head posture postoperatively, only 4 patients gained some degrees of binocular vision through operation and another 3 improved the binocular vision in primary position or in compensatory head position.

We noticed also that the fusion was obtained prevailing in patients with Brown's syndrome with preoperative small hypotropia in primary position.

Of 7 cases with congenital Brown's syndrome operated also for coexisting eso-/exotropia, only 2 gained Bagolini positive, the rest of the patients presented suppression before and after the surgery.

7.7. The tightness of posterior part of the superior oblique tendon

Comparing with the results of Mühlendyck, who found a tight band at the posterior border of the superior oblique tendon between the trochlea and the sclera in all 31 operated patients with congenital Brown syndrome [31], we found a tight posterior part of the tendon in only 87 % of operated eyes with congenital Brown's syndrome (in 20 of 23 operated eyes).

In a recent study done by Hartmann [16], exact exploration of the superior oblique tendon reveals variable pathological findings. Of 18 operated patients with congenital Brown's syndrome, 5 (27%) showed spindle-shaped thickening of the superior oblique tendon in the trochlea region, in 4 cases (23%) the insertion of the posterior part of the tendon was found as extended nasally and the remaining 9 patients (50%) demontrated intraoperativelly tight bands reaching from the trochlea region and inserting at the globe behind the posterior border of the tendon insertion.

In a study done by Gräf in 2005, none of the 22 operated cases with congenital Brown syndrome showed a pathologic fibrotic component in the posterior part of the tendon between the trochlea and the sclera. Gräf postulated that this accessory tight band described by Mühlendyck could be an artefact, the posterior border being normally "tight" in comparison to the anterior one due to the course of the superior oblique tendon in relation with the globe's rotation center [13]. However he may have used a different way of looking at the tendon. As he always did a full muscle recession he may have missed partial tightness of the posterior part of the muscle.

Maybe this apparent "tightness" of the posterior part of the superior oblique tendon is due to the very pointed angle between the posttrochlear part of the superior oblique tendon and pretrochlear one [13], as Fink found this angle very variable, between 20° to 71°. In a case with an angle approaching of 20°, the posterior end of the insertion is extended too nasally and the posterior part of superior oblique tendon would seem too tight on attempted elevation in adduction on passive ductions test.

As a conclusion, when we found no intraoperative changes of the posterior border of the superior oblique tendon - when the tendon was slack, the cause of congenital Brown's syndrome may come from changes of the superior oblique tendon in the trochlear or pretrochlear region.

7.8. Alignment of the eyes and ocular rotations

Although postoperative passive motility was free or almost free on the majority of operated patients with congenital Brown's syndrome, we remarked only a slightly improvement of:

- hypotropia in primary position of 1 to 12 deg (mean 4 deg)
- monocular elevation in adduction of 0.5 mm to 5 mm (mean 2.25 mm).

At three months postoperatively or at a long-term follow-up, the measurements of the vertical deviation in primary position, the monocular elevation in adduction and head posture remained unchanged, except for 3 cases in which a second operation was performed and a further improvement of vertical deviation in straight gaze and elevation in adduction was seen.

In congenital Brown's syndrome, after superior oblique posterior tenectomy, hypotropia in primary position/in adduction decreased statistically significant ($p=0.011/p=0.001$), while monocular elevation in adduction increased statistically significant ($p=0.001$). There was no statistically significant difference between early and late postoperative results, regarding hypotropia or monocular elevation in adduction.

7.9. Subjective results

At three months follow-up, the results after superior oblique posterior tenectomy in congenital Brown's syndrome were estimated by the parents as beeing excellent and satisfactory in 67% cases. No changes were noted in 28% cases and worse results only in 5% cases (1 patient).

8. Comparison of the surgical results of superior oblique posterior tenectomy to other methods used for the management of congenital Brown's syndrome

For comparison of superior oblique posterior tenectomy to other procedures like superior oblique tenotomy / recession or silicon superior oblique expander, it is mandatory that the same methods for pre- and postoperative measurements were used for a quantitative comparison of the different results in these studies.

A detailed comparison between the newest methods- *silicone superior oblique tendon expander* or *superior oblique split tendon lengthening* was difficult because quantitative data on the elevation in adduction are lacking, most of the american studies using a semiquantitative grading scheme for the effect of oblique muscle surgery, for example noting with -1 a minimal underaction and - 4 a maximal underaction [48-51].

From this point of view, we could compare the results of superior oblique posterior tenectomy with those of Gräf [13], who performed a recession of the superior oblique tendon in 22 cases with congenital Brown's syndrome – Table 5.

Table 5. Comparison of superior oblique posterior tenectomy to recession of superior oblique tendon (Gräf)

	SO posterior tenectomy n=21	SO recession n=22
Age (years)	2 to 29 (mean 8.5)	4 to 17 (mean 7)
Preoperative VD in Primary position	0 to 20 deg (mean 7 deg)	0 to 12 deg (mean 7 deg)
Postoperative VD in Primary position	0 to 8 deg (mean 3 deg)	0 to 6 deg (mean 1 deg)
Late postoperative improvement of monocular elevation in adduction	mean 2.25 mm	mean 15 deg
Complete resolution of AHP (abnormal head posture)	12 of 17 cases	12 of 16 cases
Simultaneous surgery for eso-/exotropia	7	9
Overcorrections	-	2

There is no significant difference between the results of this two methods (superior oblique posterior tenectomy versus recession of superior oblique tendon) regarding pre- and postoperative vertical deviation in primary position. With superior oblique posterior tenectomy, we noted a mean preoperative hypotropia of 7 deg (range from 0 to 20 deg) and a mean postoperative VD of 3 deg (varied from 0 to 8 deg) in primary position. Gräf found similar values in his study: a mean preoperative VD in primary position of 7 deg (range from 0 to 12 deg) and a mean postoperative VD of 1 deg (varied from 0 to 6 deg).

In spite of free or almost free passive motility on elevation the globe in adduction, we remarked after superior oblique posterior tenectomy only a slightly improvement of the monocular elevation in adduction with a mean value of 2.25 mm, corresponding with the postoperative elevation in adduction obtained by Gräf of mean 15 deg.

The abnormal head posture was reduced immediately after surgery: 12 patients of 17 had a complete resolution of anomalous head posturing in our cohort. These results are comparable with those of Gräf, who reported postoperatively in 12 cases of 16 any abnormal head posture.

There were some differences between our results and Gräf results regarding a delayed improvement of active elevation in adduction after superior oblique tendon recession and overcorrections. Gräf noted that the elevation in adduction was significantly improved at a late postoperative control from 5 deg median to 15 deg median, while our results remained unchanged postoperatively. We noticed no overcorrections in our cohort, while Gräf reported 2 cases with overcorrections, e.g. trochlear palsies causing diplopia to the patient in downgaze. The posterior tenectomy however was free of overcorrections. We therefore recommend this safer approach.

On 2 cases of our series a superior oblique recession was performed as a second operation after superior oblique posterior tenectomy. We noted a further improvement of VD in primary position and of monocular deviation in adduction.
We need more surgical cases to conclude that the recession of superior oblique tendon can be used as an additional method to improve alignment of the eye and the ocular rotations, after superior oblique posterior tenectomy.

Regarding the newest surgical method for congenital Brown's syndrome (*silicone superior oblique tendon expander)*, Wright [50] reported good results in resolution of the downshoot in adduction. 14 of 15 patients improved motility, with normal version in 10 patients, 3 were undercorrected and 2 overcorrected (only one requiring an inferior oblique weakening procedure). The potential development of downgaze restriction after placement of the expander and in some cases postoperative inflammatory reaction or extrusion of silicone band are a potential disadvantage and severe side effects which cannot be seen after superior oblique posterior tenectomy.

9. Proposed therapeutic algorithm in congenital Brown's syndrome

The surgical management of congenital Brown's syndrome remains a complicated task. The efficiency of all surgical procedures described until now proved to be variable. Presumably, this variability was caused more by the heterogenous etiology of Brown syndrome rather than by surgical technique.

For the future, it will be better if the surgeon will *adapt the surgical technique to the intraoperative or even to the radiological findings*. I purpose the following algorithm (Fig. 11).

In a case of congenital Brown's syndrome, if intraoperative forced ductions test shows severe restriction to elevation in adduction and we find a tight or very tight posterior part of the superior oblique tendon, the first step is to perform a *superior oblique posterior tenectomy*. In case we found intraoperatively a normal or slightly tight posterior tendon, we should perform superior oblique tendon recession.

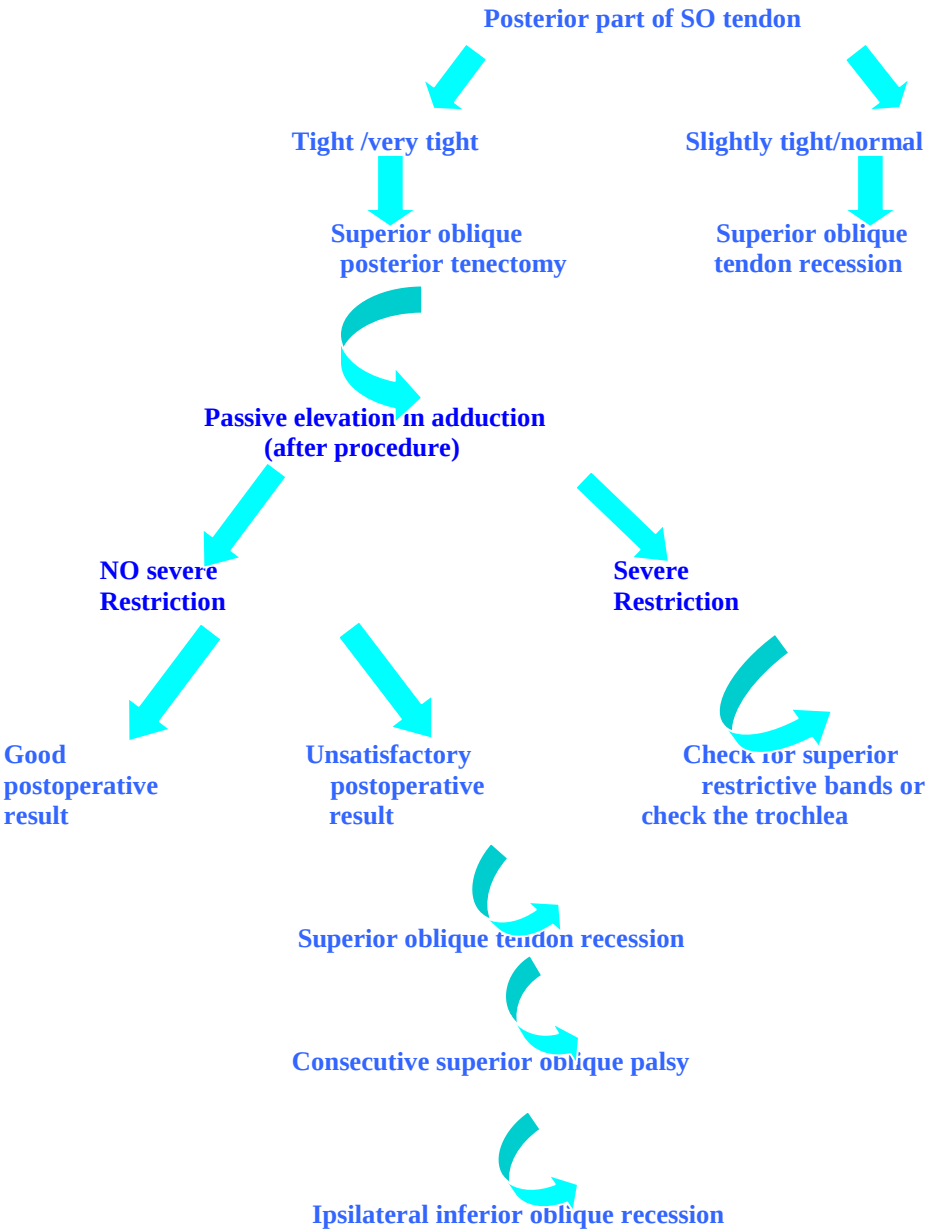
In the second step, after superior oblique posterior tenectomy, if postoperative forced ductions test shows still severe restriction to passive elevation in adduction, the surgeon should check the tendon – trochlea complex or to check if superior restrictive bands hinder the elevation in adduction. These superior tight bands reaching from the trochlea region and inserting at the globe behind the posterior border of the SO tendon insertion could only be found when the upper nasal posterior quadrant of the globe is explored [16]. If any kind of these obstacles exists, we should eliminate them.

If passive motility proved to be free or almost free postoperatively, it will be better to see the results at three months follow-up.

If the results of superior oblique posterior tenectomy are unsatisfactory, I recommend as a second procedure the *superior oblique tendon recession*, an effective, safe and even reversible surgical procedure.

If the operated patient developed in months the classic features of a superior oblique palsy, the surgeon will have to perform recession of the ipsilateral inferior oblique muscle.

Fig. 11. Proposed therapeutic algorithm in congenital Brown's syndrome



10. Abstract

Introduction: Since more than 50 years, various surgical procedures have been described for congenital Brown's syndrome. However most showed low success rates and some even severe side effects.
The aim of this retrospective study was to evaluate the results of superior oblique posterior tenectomy. This technique was introduced in 1996 by Mühlendyck. Since this first description no other results have been published by others.
Patients and methods: 21 patients with congenital Brown's syndrome (aged 2 to 29 years) were operated between 2001 and 2006, in the Department of Ophthalmology, Ludwig-Maximilians-University Munich. In all patients, intraoperative forced ductions showed severe passive restriction of elevation in adduction and superior oblique posterior tenectomy was performed as a primary procedure. The squint angle (vertical and horizontal deviation in primary position, lateral gaze, up/down gaze), active elevation in adduction, abnormal head posture at distance fixation, binocular vision (in primary position and anomalous head posture) were assessed in each case.
All the measurements were performed 1 day before, 1 month and 3 months after surgery. Eight patients were examined 6-24 months after primary procedure.
Results: Intraoperatively, a tight or very tight posterior part of the superior oblique tendon was found in 87 % of operated eyes.
At the end of the operation, passive motility in adduction became free (14 eyes) /almost free (7 eyes) on the majority of operated patients (totally 23 eyes).
Inspite of free passive motility, the active monocular elevation in adduction was only slightly improved by 0.5 mm to 5 mm (mean 2.25 mm), like hypotropia in primary position, which was improved by 1 to 12 deg (mean 4 deg).
Better results regarding hypotropia in primary position were noted when the preoperative vertical deviation in primary position was more than 10 deg. However in cases with preoperative hypotropia less than 10 deg, a better fusion was obtained.
Preoperatively, 17 patients showed an abnormal head posture. Postoperatively, 12 of them totally gave up their posture and 5 improved partially.
Of 8 cases with a long-term follow-up, 5 showed unchanged measurements of vertical deviation in primary position, monocular elevation in adduction and head posture. 3 patients with a long-term follow-up had further surgery and an improvement of vertical deviation in straight gaze and active elevation in adduction.

Conclusion: The use of superior oblique posterior tenectomy significantly improves abnormal head posture and also improves alignment and ocular rotations in patients with congenital Brown's syndrome. We did not see any serious side effect like consecutive superior oblique muscle underaction (as in superior oblique tenotomy or recession) and no foreign body extrusion (as in silicone superior oblique tendon expander). So the superior oblique posterior tenectomy is a safe and effective procedure with regard to the head posture.

The fact that the passive motility had dramatic improved postoperatively, but the active elevation in adduction improved only slightly, suggests a paretic/ dysinnervational component to the superior oblique in some patients.

From this point of view, a therapeutic algorithm depending on intraoperative/ radiological findings in congenital Brown's syndrome is proposed.

11. Zusammenfassung

Hintergrund: Seit mehr als 50 Jahren, wird die schwierige, operative Behandlung des kongenitalen Brown-Syndroms kontrovers diskutiert.
Ziel dieser retrospektiven Studie war die Bestimmung des Effektes der hintere Obliquus-superior-Tendektomie, die 1996 erstmals von Mühlendyck vorgestellt wurde und zu der bisher noch keine weiteren Daten publiziert wurden.
Patienten und Methoden: Insgesamt wurden 21 Patienten (im Alter von 2 – 29 Jahren) in der Studie eingeschlossen, die wegen eines kongenitalen Brown-Syndroms in der Augenklinik der Ludwig-Maximilians-Universität München zwischen 2001 und 2006 operiert wurden. Bei allen Patienten war präoperativ die passive Hebung in Adduktion sehr deutlich eingeschränkt und es erfolgte eine hintere Obliquus-superior-Tendektomie.
Die Schielwinkel wurden im Prismen-Abdecktest, die monokulare Exkursion nach Folgebewegungen, die Kopfzwangshaltung (KZH) bei Fernfixation, das Binokularsehen in Primärposition und in Kopfzwangshaltung bestimmt. Die Messungen erfolgten 1 Tag präoperativ, 1 Monat und 3 Monaten postoperativ. Acht Patienten erschienen zu einer Spätkontrolle nach 6-24 Monaten.
Ergebnisse: Am Ende der Operation war die passive Hebung in Adduktion in meisten Fällen frei (bei 14 der 23 Augen) oder fast frei (bei 7 der 23 Augen).
Trotz freier passiver Motilität, war die aktive Hebung in Adduktion nur wenig gebessert (0.5mm - 5 mm, Median 2.25 mm), wie auch die vertikale Abweichung in Primärposition (1° - 12°, Median 4°).
Eine stärkere Reduktion des Höhenschielen in Primärposition wurden beobachtet, wenn die vertikale Abweichung in Primärposition präoperativ über 10° war. Besser Binokularfunktion ergaben sich bei VD, die unter 10° war.
Präoperativ, nahmen 17 Patienten eine Kopfzwangshaltung (KZH) ein. Postoperativ, haben alle 17 Patienten die KZH vollständig aufgegeben (12) oder nur noch eine geringe KZH eingenommen (5).
Bei 5 der 8 Patienten mit einer späten Kontrolle hatten sich die vertikale Abweichung in Primärposition, die aktive Hebung in Adduktion und die KZH nicht geändert. Bei 3 Patienten ergab sich nach einer weiteren Operation eine deutliche Besserung.

Schlussfolgerung: Die hintere Obliquus-superior-Tendektomie verbessert deutlich die Kopfzwangshaltung und reduziert das Höhenschielen in Primärposition und die Hebungsfähigkeit in Adduktion bei kongenitalem Brown-Syndroms. Im Gegensatz zu anderen Operationsverfahren ist sie eine sichere Methode ohne das Risiko der konsekutiven Obliquus-superior-Parese (wie bei der Tenotomie oder der Rücklagerung des gesamten Muskels) oder der Extrusion von Fremdmaterial (wie bei der Obliquus-superior-Sehnenverlängerung).
Die Variabilität der Effekte dieser Therapie ist vermutlich auf die heterogene Ätiologie des Brown-Syndroms zurückzuführen. Die auch postoperativ stark eingeschränkte aktive Hebung bei passiv freier Motilität deutet auch eine paretische / dysinnervationelle Komponente bei einigen Fällen des kongenitalen Brown-Syndroms hin.

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Danksagung

Besonders bedanken möchte ich mich bei Herrn Prof. Dr. Oliver Ehrt für die Überlassung des Themas und die hervorragende Betreuung während der Bearbeitung. Ein ganz besonderer Dank geht an Herrn Prof. Dr. Klaus- Peter Boergen und an Herrn Prof. Dr. Günther Rudolf, die mich immer bezüglich der komplizierten Fälle und Operationen gut beraten haben.

Ebenso gilt mein Dank den Orthoptistinnen der Sehschule für deren Vorarbeit und Unterstützung.

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