Brown Syndrome - A Review

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Introduction

Brown syndrome was first described by Harold W Brown in 1949 as the superior oblique tendon sheath syndrome.1 Brown believed that the syndrome was due to the congenital paralysis of the inferior oblique muscle leading to a short anterior tendon sheath of the superior oblique. The incidence of this condition is 1 in 450 cases of strabismus; approximately one in 20,000 live births.[2] The actual incidence may be higher as many cases are asymptomatic. There is equal predilection for both sexes in congenital Brown syndrome. Wright noted that 5% cases are bilateral and idiopathic Brown syndrome has a higher preponderance in females (63%) and traumatic acquired Brown in males (82%).[3]

Etiology

Congenital Brown syndrome was initially believed by Brown to be due to a short anterior tendon sheath. Another possible cause was a congenital or acquired anomaly of the superior oblique tendon limiting passage of the tendon through the trochlea. The concept of a short or inelastic tendon is a well accepted theory based on the fact that a tenotomy can relieve the limitation to elevation in adduction. Wright demonstrated by his computer model in 1999, that a tight or inelastic muscle-tendon complex was the best fit for Brown syndrome pattern of deviation.[3] Spontaneous resolution of Brown syndrome can be explained by the possible cause of the problem being in the trochlea or tendon trochlea complex. Persistent embryonic trabeculae between the tendon and the trochlea can inhibit the passage of the tendon through the trochlea. Helveston proposed the theory of abnormal telescoping where he showed that the tendon-slackening distal to the trochlea comes from a telescopic elongation of the central tendon.[4] Some patients with idiopathic click can be explained by the presence of a trochlear problem. Chronic movement of the superior oblique tendon through the trochlea can result in a traumatic tenosynovitis with tendon swelling and stenosis of the surrounding sheath - this has been named as the trigger-thumb analogy theory.

Acquired Brown syndrome can be a result of surgeries on the superior oblique like a tuck, scleral buckling procedures and external valves like Molteno or Ahmed valve. These can create adhesions and prevent full relaxation of the tendon. Sinus operations near the trochlea can also cause Brown syndrome. Dog bites in this region can also cause Brown syndrome with superior oblique palsy the "canine tooth syndrome". The click syndrome is caused by inflammation leading to dilatation of the tendon limiting movement through the trochlea. This can occur commonly following rheumatoid arthritis, systemic lupus erythematosus or Sjogren syndrome.

Genetic transmission has been reported though in most cases it is sporadic. Occurrence in monozygotic twins has been reported. Autosomal dominant inheritance with incomplete penetrance and variable expression has been proposed.

Clinical features Classification

"The most common classification is congenital and acquired. Congenital cases are less likely to improve spontaneously and more likely to need surgery. Acquired cases especially inflammatory can improve spontaneously.

"Brown syndrome has also been classified as

a) mild - no hypotropia in primary or adducted position
b) moderate - hypotropia in adducted position

c) severe - hypotropia in primary position

"A third classification was proposed by Jampolsky5

a) True Brown syndrome - no hypotropia in primary position or down gaze

b) Brown syndrome plus - vertical deviation in primary position or adduction ± head posture.

Clinical features

The diagnostic features in Brown syndrome include deficient elevation in adduction, less deficiency in the midline, minimal or no elevation deficiency in the abducted position, minimal or no superior oblique overaction, V pattern with divergence in upgaze and restricted forced ductions. Anomalous head posture with hypotropia in the primary position or a downshoot in adduction may be present. Audible click and tenderness in the trochlear region may be present in inflammatory cases. V pattern exotropia may be present in bilateral involvement.

Spontaneous resolution of congenital Brown syndrome over years has been noted. The click syndrome represents a stage towards resolution. Inflammatory conditions may show waxing and waning especially cases of rheumatoid arthritis. Improvement is unlikely after scleral buckling or glaucoma surgery.

Differential diagnosis

1. Isolated inferior oblique palsy: This is characterized by overaction of the superior oblique muscle and positive Parks' three step test. Forced duction test is free in inferior oblique palsy.
2. Double elevator palsy: Limitation of elevation is present in both adduction and abduction. In addition the patients have ptosis or pseudoptosis.
3. Congenital fibrosis syndrome: The differences include restricted elevation in abduction and esotropia on attempted upgaze.
4. Blow out fracture of the inferior orbital wall: The elevation deficiency is more marked in abduction. Imaging reveals a fracture and there may be associated enophthalmos.
5. Thyroid ophthalmopathy: The elevation deficiency is worse in abduction than adduction.
6. Adherence syndrome: During inferior oblique surgery adhesions may form due to fat prolapse and limit elevation in abduction.

Management Evaluation

The vision needs to be checked to rule out amblyopia. Abnormal head posture if present is indicative of the presence of fusion. Forced duction testing needs to be performed to confirm the diagnosis.

Non surgical management

Spontaneous improvement has been known to occur. Hence it may be prudent to observe cases where there is no threat to binocularity. Elevation in adduction exercises can improve the condition in congenital cases or in cases where there is intermittent Brown syndrome. Injection of corticosteroids has been reported to improve the Brown syndrome in patients with inflammatory disease. Systemic treatment of the underlying disease may improve cases of acquired Brown syndrome.

Surgical management Indications

When there is a loss of binocularity, with the chance of development of amblyopia and the child does not develop an abnormal head posture, surgery is indicated. Mild Brown syndrome needs to be observed. If there is presence of primary position hypotropia and unacceptable downshoot on adduction, surgery can be considered. Acquired cases due to the presence of a scleral explant or a glaucoma filtering valve need to be operated.

Surgical techniques

Sheathectomy was proposed by Brown initially, but the surgery did not yield satisfactory results.1 Superior oblique and trochlear luxation consists of removing the tendon from the trochlea by luxating the trochlea, this procedure is no longer practiced. Technically easier

procedures include superior oblique tenectomy or tenotomy. Though elevation can improve with this surgery, a significant proportion (50-80%) of patients develop superior oblique palsy as Brown syndrome is generally not associated with superior oblique overaction.6 A Z-tenotomy of the superior oblique was also tried, though this was also unsatisfactory and associated with superior oblique palsy.7 Simultaneous inferior oblique recession was advised by Parks and Eustis when performing a tenotomy or tenectomy.8 While this technique reduced the risk of postoperative superior oblique palsy, about 20% of the patients still had superior oblique underaction. Superior oblique tendon expander surgery was proposed by Wright in 1991.9 In this technique a retinal silicone band about 5 to 7 mm is cut and sewn to cut ends of the superior oblique tendon with non absorbable sutures. High success rate was reported with a low incidence of superior oblique palsy. A chicken suture can be passed in lieu of an expander to retain the cut ends of the superior oblique together. Superior oblique recession produces a graded slackening of the tendon.10 Undercorrections are common and the problem with recessing the tendon is that it changes the characteristics of the superior oblique tendon insertions and results in postoperative complication of limited depression.

Complications of surgery include superior oblique palsy which is the most common complication. Palsy usually develops slowly and may be difficult to correct. Undercorrection can occur following surgery, but the effect may improve gradually over time. Silicone expander surgery if performed incorrectly can lead to adhesions and rarely may extrude.9 Care must be taken not to damage the sheath of the superior oblique. Inadequate care during surgery can lead to damage to the superior rectus muscle.

It is important to remember that Brown syndrome cannot be cured but one can only attempt to improve the field of binocular single vision, improve the elevation in adduction and eliminate head posture. Hence prudence is advised before attempting any surgery for Brown syndrome.

Table 1. Differentiating features of Brown syndrome, primary superior oblique overaction and inferior oblique paresis.

	Brown syndrome (inelastic superior oblique muscle-tendon complex)	Primary superior oblique overaction	Inferior oblique paresis
Limitation of elevation in adduction	Usually severe (-3to-4); common	Usually mild; not common	Usually severe (-3to -4); not common
Bilateral involvement	Rare (5-10%)	Common	Unusual
Vertical deviation	None or small (<10 PD)	Bilateral small(<10 PD)	Unusually large (>10 PD)
Superior oblique overaction	None or minimal	Yes, marked	Yes, marked
Pattern	None or V patter Y-subtype with divergence in upgaze	A-pattern Lambda subtype with divergence in downward gaze	A-pattern often convergence in upgaze
Fundus torsion	None in primary or downgaze, intorsion in upgaze	Intorsion in primary, increasing in downgaze	Intorsion in primary, increasing in upgaze
Head tilt test	Negative	Negative	Positive
Forced duction	Positive	Positive	Positive

Figure 1: A 3 year old girl presented with Right Brown syndrome. Note the absence of elevation in levoelevation of the right eye.

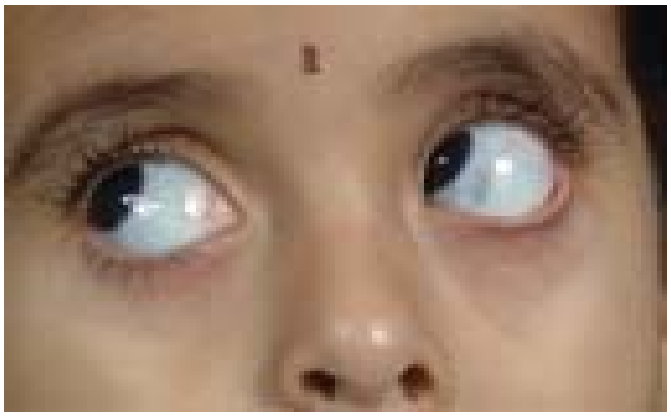

Figure 1a: Dextroelevation shows the eyes are elevating fully;

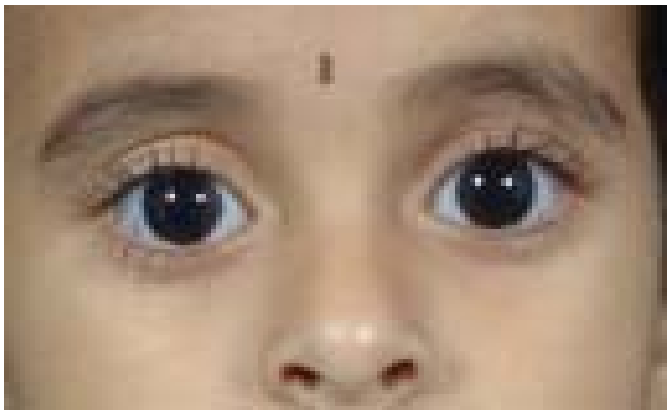

Figure 1b: Primary position does not show any deviation

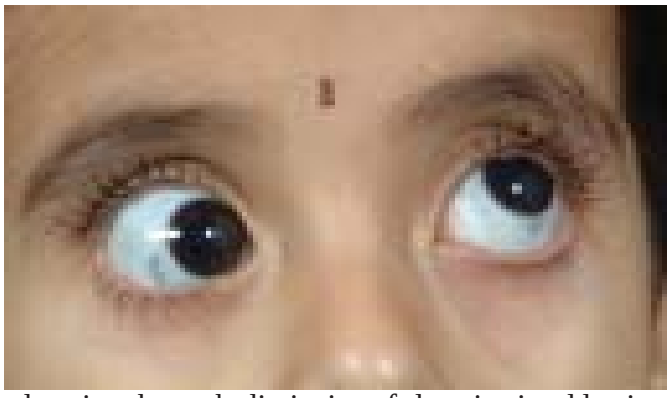

Figure 1c: Levoelevation shows the limitation of elevation in adduction of the right eye.

Figure 2: Superior oblique expander surgery:

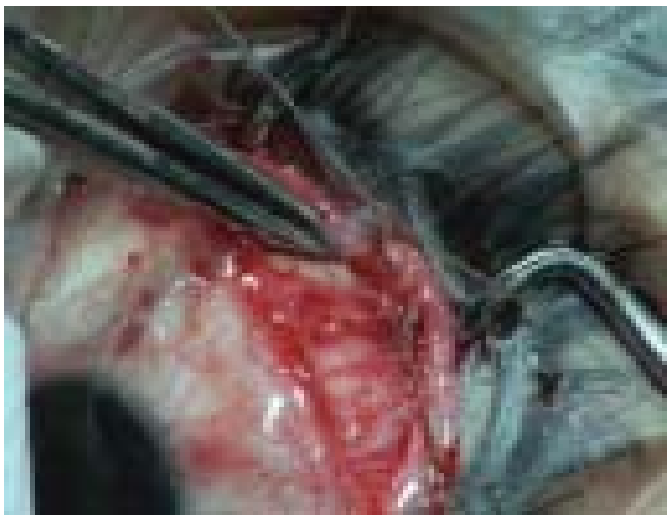

Figure 2a: The superior oblique tendon is located and 2 double armed non absorbable sutures (5-0 ethibond) are passed a few millimeters apart. The tendon is cut between the sutures.

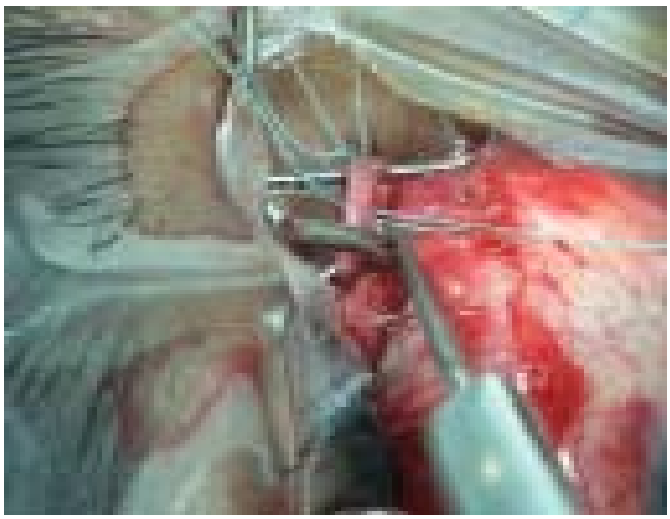

Figure 2b: The silicon band (5- 7 mm in size) is cut and secured between the cut ends of the superior oblique tendon, thus lengthening the tendon.