DJO

Vol. 30, No. 1, July-September 2019

Review Article

# Brown's Syndrome

**Gunjan Saluja, Anju Bhari**

*Squint and Neuro Ophthalmology Services, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi, India*

**Abstract**

*Brown's syndrome, also known as superior oblique tendon sheath syndrome or Janesch Brown syndrome is characterized by limitation of elevation in adduction with divergence in upgaze, and a normal elevation in abduction. The term Brown's plus is used when superior oblique overaction is present along with Brown's syndrome.*

*The syndrome can be congenital or acquired, and a large number of causes have been attributed for the same. Various management options include complete tenectomy of superior oblique, superior oblique recession, tendon lengthening procedures and split tendon lengthening.*

*Delhi J Ophthalmol 2019;30;23-27; Doi http://dx.doi.org/10.7869/djo.473*

***Keywords:*** *Superior oblique tendon sheath syndrome, Brown's syndrome*

## Introduction

The typical presentation of limitation of elevation in adduction, was first described by Janesch in a post traumatic case. Forced duction test was positive in this case, which ruled out inferior oblique palsy, the features were elucidated by a traumatic adhesion between trochlea and globe anterior to the equator, hence did not interfere with the action of superior oblique but caused limitation of elevation.[1]

Later in 1950, Brown described similar features in a congenital instance, and propounded that features could be due to a tight tendon sheath, and he further suggested the term "superior oblique sheath syndrome".[2] It was found that tenotomy was effective in cases in which sheathetomy failed, this description gave a new direction to the pathogenesis and it became clear that a number of anomalies in superior oblique muscle, tendon and trochlea were responsible for these features.

## Relevant anatomy

Superior oblique takes its origin from the superomedial portion of optic foramen, the muscle then runs parallel, close to the upper part of medial wall upto trochlea, it then turns around and runs posterolaterally.

Trochlea acts like a functional origin of superior oblique, and is formed medially by trochlear fossa of frontal bone and laterally by cartilaginous tissue. Superior oblique can be divided into three parts, pretrochlear, which is 10mm distal to the trochlea.

Trochlear portion, 4-6 mm in length and post trochelar portion is tendinous, it passes beneath superior rectus and inserts in a fan shape posterior to the equator.

The posterior fibres in the fan are responsible for depression and anterior fibres are responsible for intorsion.

## Incidence

Incidence of Brown's syndrome is relatively low, being 1 in 450 strabismus cases[3], but the incidence of varies in different studies.[4]

The probability of syndrome being bilateral is 5-10%.[4,5] The syndrome does not follow any particular laterality and has no gender predilection, though Brown in his initial work on 126 patients had described the syndrome in 74 females and 52 males, and suggested it to be more common in females,[6] but further studies did not support this view.[7]

## Etio-Pathogenesis

Brown's syndrome can be congenital, acquired or intermittent.[8]

Congenital cases are characterized by constant limitation, since birth and are not associated with pain or inflammation. Various theories have been proposed for the pathogenesis of Brown's syndrome, each of these theories have been described in brief below.

### Anomalies of tendon sheath

When Brown first described the syndrome in 1950, he attributed it to the delay in development of inferior oblique muscle which resulted in lack of stretch in inferior oblique developing as a check ligament of superior oblique and further led to the shortening of superior oblique tendon sheath. This theory was further supported by his intra-operative findings.

Brown classified the syndrome as, true or simulated. True Brown's was due to anomaly of sheath and was congenital. On the contrary, simulated Brown's was due to any anomaly other than short tendon sheath.

This theory of Brown's was challenged, as the electromyographic studies failed to show paralysis of inferior oblique[9], moreover the surgical techniques also failed to show any sheath on superior oblique.[10]

This gave rise to the new theory of pseudoparalysis of inferior oblique.

### Pseudoparalysis of inferior oblique

Pseudoparalysis of inferior oblique can be explained by factors intruding the normal contractile function of inferior oblique, which can be either due to short tendon of superior oblique or due to anomalous check ligaments on the insertion of inferior oblique. On this basis Giard,[11] classified Brown's syndrome into four types, type1 is caused due to retraction of tendon of superior oblique, type 2 due to postoperative retraction of superior oblique tendon sheath, type 3 due to anomalous check ligaments on the insertion of inferior oblique and type 4 was explained by the swelling on superior oblique tendon passing through trochlea.

It was suggested that the check ligaments extending from inferior oblique to the lateral wall of orbit, limit the elevation in adduction.

**Anomalies of superior oblique tendon**

Crawford, later observed that doing sheathotomy alone did not bring favorable results, he reported experimental and surgical results in 28 patients of Brown's syndrome for 4 surgical procedures, which included Z tenotomy (n=9), split tendon lengthening (n=2), complete tenotomy (n=16) and tenectomy (n=2). He found tenotomy of superior oblique to have the best results and concluded that the syndrome did not result from a tight sheath alone, instead was a consequence of a tight tendon[12], cutting the tendon of superior oblique medial to superior rectus was an effevctive method of management.

**Anomalies in development of tendon**

Sevel, for a better understanding of the pathogenesis of Brown's, studied the embryology of superior oblique tendon and trochlea. He found that in early phases of development trochlea and tendon are indistinguishable, as the development progresses, the structures can be differentiated but a trabeculae continues to persist between the tendon and trochlea. Fine trabecular remnants continue to persist on both sides of trochlea throughout the life. The persistence of thick trabecular remnants can lead to Brown's.[13]

**Anomalies of trochlea**

Trochlea acts as a functional origin of superior oblique, Helveston et al studied the anatomy and physiology of trochlea. They described trochlea to be composed of four components, a cartilage "saddle," an intratrochlear portion of superior oblique tendon, fibrillo- vascular sheath surrounding the tendon, a dense fibrous condensation that secures the trochlear saddle and contents of bony medial orbital wall. A bursa like structure was found to be present between tendon sheath and trochlea, helping in the smooth movement of muscle. Hence, accumulation of fluid in bursa can result in the limitation of elevation.[14]

This mechanism can contribute to both congenital and acquired causes of Brown's syndrome.

Impaired slippage of superior oblique tendon through trochlea has been observed in several patients with intermittent and congenital Brown's syndrome.[15] The limitation may improve spontaneously with an audible click, hence the term, superior oblique click syndrome was proposed.[16]

Impaired slippage of tendon through trochlea can be attributed to the retrotrochlea thickening of superior oblique tendon.

**Brown's a Congenital Cranial Dysinnervation Syndrome**

Papst and Stein did electromyographic studies and found co-contraction of superior oblique and inferior oblique in Brown's syndrome, similar to Duane's retraction syndrome.[21] Later Ellis J et al proposed that Brown's syndrome can be due to congenital cranial dysinnervation. Anomalies in the development of trochlear nerve, can lead to poor differentiation of tendon-trochlea complex resulting in restricted movement of tendon through trochlea.[22] This theory has been further supported by MRI findings which show hypoplasia of trochlear nerve.[23]

Brown's syndrome has also been found to be associated with crocodile tears, cardiac abnormalities and congenital ptosis.[24]

**Acquired Brown's**

The causes of acquired Brown's syndrome can be attributed to the etiologies causing the shortening of superior oblique tendon or due to damage to trochlea.

The various etiologies of acquired Brown's syndrome, along with the proposed pathogenesis have been summarized in Table1.

**Table 1: Etiology of acquired Brown's syndrome**

| Etiology | Proposed Pathogensis |
|---|---|
| Rheumatoid Arthiritis[17]<br>Sjogren's syndrome[18]<br>Systemic lupus erythematosus[19] | Causes inflammation and fibrosis, leading to thickening of muscle entering trochlea |
| Trauma<br>Periocular surgery<br>Peribulbar block<br>Blepharoplasty<br>Double plate Molteno implant[20]<br>Sinus surgery | Leads to scarring and shortening of the tendon |
| Cyst in superior oblique tendon | Impaired slippage of muscle through trochlea |
| Superior oblique tendon tucking procedure<br>Thyroid myopathy | Tightens the superior oblique tendon |
| Metastasis to superior oblique tendon | |
| Blow out fracture<br>Fat adherence<br>Secondary to inferior oblique surgery | Limits the passive limitation of adduction |

**Clinical features**

1. Gender No particular gender predilection.
2. Laterality: Although by his earlier work, Brown suspected right side to be involved more commonly, however later reports did not support the evidence.
3. Presenting features: Patients frequently complaints of diplopia, and inability to elevate the involved eye.
4. Examination
   a. Extra ocular movements: There is limitation of elevation in adduction, with normal elevation in abduction; down shoot in adduction may be a variable feature. There is divergence in upgaze, which occurs due to slippage of globe against a tight tendon of superior oblique. Hence, causing a characteristic Y pattern.

      Depression in adduction and elevation in abduction are normal. Brown's syndrome when associated with superior oblique over action, has been termed Brown's plus.
   b. Head posture: Chin elevation with head tilt to the opposite side is the usual head posture adapted to prevent diplopia in primary.

c. Forced duction test: Forced duction test is positive which helps to differentiate Brown's syndrome from inferior oblique palsy. Exaggerated forced duction test for obliques has been described by Guyton,[25] the test is performed by holding the globe at 3 and 9 O' clock position, followed by retro placement of globe so as to create stretch on the tendon; the globe is then forcibly extorted and moved supero nasally. A tight superior oblique tendon causes the globe to slip.

d. Sensory adaptation: Suppression is rarely seen.

Classification of Brown's syndrome

Brown's syndrome can be classified as congenital or acquired Brown's has also been classified by Stager et al[26] as:

- Mild- No hypotropia in primary position
- Moderate- Hypotropia in adducted position
- Severe- Hypotropia in primary position

**Differential diagnosis**

Differential diagnosis include, primary paralysis of inferior oblique, double elevator palsy, congenital fibrosis of extra ocular muscle, thyroid ophthalmopathy and adherence syndrome.

1. Inferior oblique palsy: Inferior oblique palsy is associated with A pattern, a positive head tilt test and a negative exaggerated forced duction test.
2. Double elevator palsy is characterized by limitation of elevation in both abduction and adduction and can result from, a tight inferior rectus, or from superior rectus palsy.
3. Congenital fibrosis of extra ocular muscle (CFEOM), is characterized by limitation of all extra ocular movements and convergence on attempted up gaze.

## Management

**Conservative management**

Observation is the most common conservative management option, as spontaneous resolution is likely to occur in intermittent and acquired causes, and in less likely in long standing congenital cases. An underlying etiology for the acquired cases must always be investigated. Auto-immune causes like rheumatoid arthritis, Sjogren's syndrome, systemic lupus erythematosus must be ruled out. Imaging can be helpful to exclude the presence of cyst or secondary metastasis.

Depot steroid injections can help in early resolution of Brown's syndrome associated with auto-immune causes like rheumatoid arthritis.[27]

## Surgical management

Indications of surgery

- Diplopia in primary position
- Hypotropia in primary
- Anomalous head posture
- Loss of binocularity

Various surgical procedures have been described for the management of Brown's syndrome:

**1. Sheathectomy**

Brown, first had described dissection and stripping of the sheath of superior oblique, leaving the tendon intact.

**2. Complete superior oblique tenectomy**

Complete resection of superior oblique tendon, first advocated by Jacobie yields favorable results[28], but can result in superior oblique palsy due to complete dissection of superior oblique.

Hence was usually done along with inferior oblique weakening procedure.[29,30]

**3. Wright's silicone expander**

Wright[31] introduced the use of silicone band expander to lengthen the superior oblique tendon.

The tendon is secured with two 5-0 mersilene sutures, following which transection is done, and a piece of silicon band 240, 2 mm wide and 5 to 8 mm in length is inserted between the cut ends, care must be taken to minimally disturb the adjoining tissues to prevent adhesions in post-operative period, the procedure must be preferably done through temporal approach.

This procedure maintains the fan shaped insertion of superior oblique, which is one of its major advantage, but the procedure also has several drawbacks like persistent inflammation, formation of adhesions and extrusion of band.

**4. Chicken suture lengthening**[32]

Knapp initially described the chicken suture technique for lengthening the superior oblique tendon, this was achieved by using a loosely tied non absorbable suture between the cut ends of the superior oblique tendon, the procedure is reversible and also helps in maintaining the anatomical fan shaped insertion

**5. Suture tendon lengthening**[33]

In this technique as described by Suh et al, two non absorbable sutures are placed 4 mm apart, the tendon is then cut and further with the help of a slip knot the distance between the two ends of tendon is adjusted.

**6. Loop tenotomy**

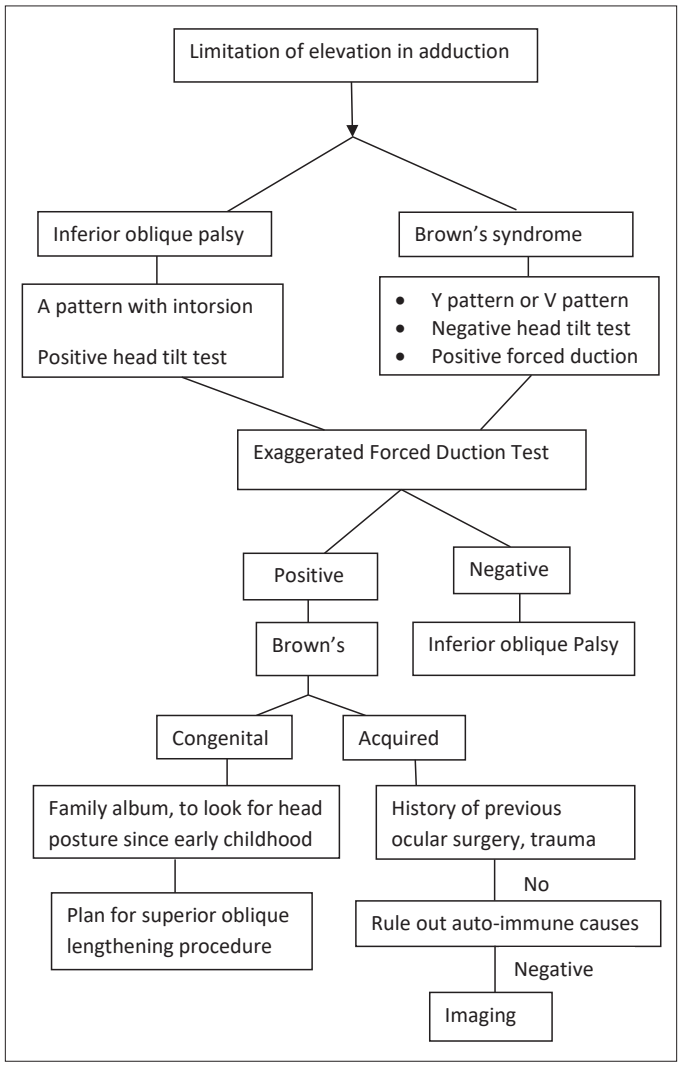
Loop tenotomy can be performed with the help of a non absorbable suture, superior oblique is hooked and dis inserted, and is further secured with a non-absorbable 5-0 ethibond suture. Sutures are then passed through the insertion site and a loop of 10 mm or more is made depending on the forced duction test.

**7. Split tendon lengthening procedure**

Split tendon lengthening procedure introduced by Ciancia is not very popular. In this procedure superior oblique tendon is divided into two and the cut half widths are further joined. The procedure has been found to improve hypotropia of 12 pd.[34]

Table 2 shows a simplified approach for the management of Brown's syndrome.

**Table 2: Management of Brown's syndrome**



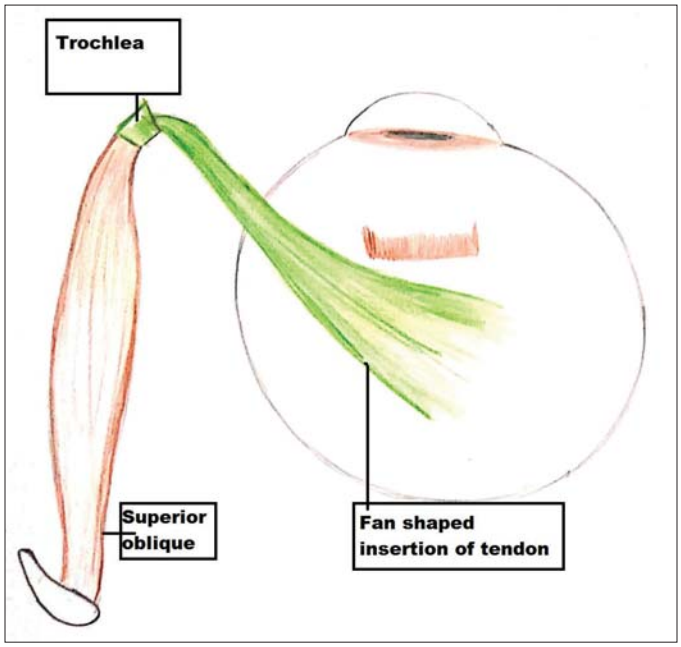




***Figure 1:*** *Anatomy of superior oblique*

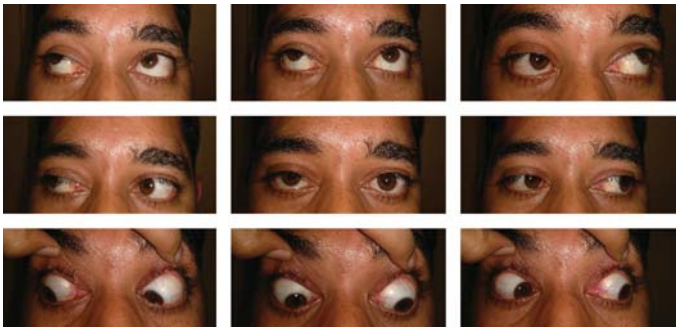


***Figure 2:*** *9 gaze picture of a patient of Brown's syndrome, showing limitation of elevation in adduction*

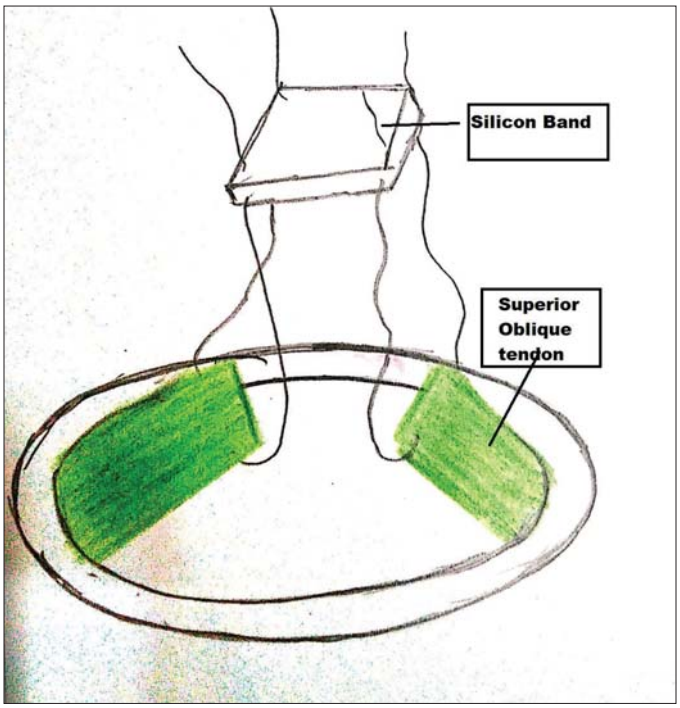



***Figure 3a:*** *Schematic diagram of silicon band expander procedure*

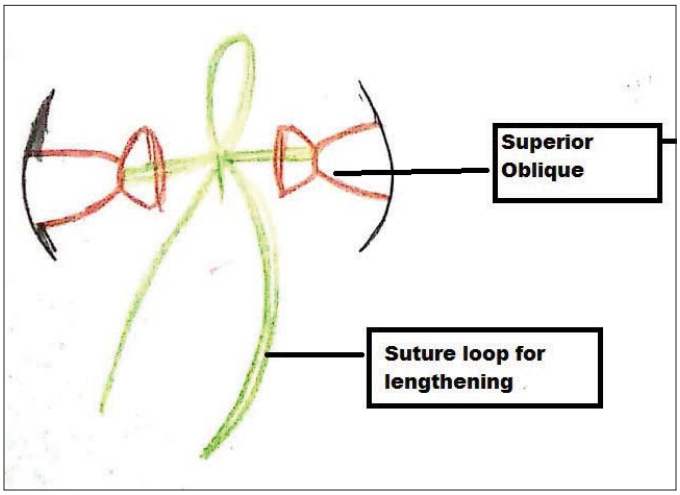



***Figure 3b:*** *Schematic diagram of suture lengthening procedure*

## Reference


1. Janesch PA. Paresen der schragen Heber, Graefes Arch. Ophthalmol, 1928; 121:113.
2. Brown HW. Congenital structural muscle anomalies. In Allen JH ed: Strabismus Ophthalmic Symposium I. *St Louis Mosby-Year Book,* 1950; p 205.
3. Raynor R, Hiatt RL: Bilateral Brown's superior oblique tendon sheath syndrome. *Ann Ophthalmol,* 1970; 2:506-508.
4. Crosswell HH, Haldi B A, The superior oblique tendon sheath syndrome. A report of two bilateral cases. *J Pediatric Ophthalmol.* 1967; 4:8.
5. Wright KW. Brown's syndrome: Diagnosis and management. *Trans Am Ophthalmol Soc,* 1999; 97:1023-109.
6. Brown HW: True and simulated superior oblique tendon sheath syndrome. *Dot Ophthalmol,* 1973; 34:123-136.
7. Eustis HS, O'Reilly C, Crawford JS: Management of superior oblique palsy after surgery for true Brown's syndrome. *J Pediatr Ophthalmol Strabismus,* 1987; 24:10-7.
8. Costenbader FD, Albert DG. Spontaneous regression of pseudoparalysis of inferior oblique muscle. *Arch Ophthalmol,* 1958; 59:607.
9. Catford, GV, Dean Hart JC. Superior oblique tendon sheath syndrome: An electromyographical study. *Br J Ophthalmol,* 1971; 55:155.
10. Parks MM, Brown M. Superior oblique tendon sheath syndrome of Brown. *Am J Ophthalmol,* 1975; 79:82-86.
11. Girard LJ. Pseudoparalysis of the inferior oblique muscle. *South Med J,* 1956; 49(4):342-6.
12. Crawford JS. Surgical treatment of true Brown's syndrome. *Am J Ophthalmol,* 1976; 81:289-295.
13. Sevel D. Brown's syndrome - a possible etiology explained embryologically. *J Pediatr Ophthalmol Strabismus,* 1981; 18(5):26-31.
14. Helveston EM. The influence of superior oblique anatomy on function and treatment. The Bielschowsky Lecture. *Binocul Vis Strabismus Q,* 1999; 14(1):16-26.
15. Leone CR, Leone RT. Spontaneous cure of congenital Brown's syndrome. *Am J Ophthalmol,* 1986; 102:542.
16. Roper- Hall MJ, Roper- Hall G: The superior oblique "click" syndrome. In: Proceedings of the 2nd International Orthoptic Congress. *Amsterdam: Excerpta Medica,* 1972; pp.360-366.
17. Hickling P, Beck M. Brown's syndrome: an unusual ocular complication of rheumatoid arthritis. *Ann Rheum Dis,* 1991; 50(1):66.
18. Brahma AK, Hay E, Sturgess DA, Morgan LH. Acquired Brown's syndrome and primary Sjogren's syndrome. *Br J Ophthalmol,* 1995; 79(1):89-90.
19. Whitefield L, Isenberg DA, Brazier DJ, Forbes J. Acquired Brown's syndrome in systemic lupus erythematosus. *Br J Rheumatol,* 1995; 34(11):1092-4.
20. Dobler AA, Sondhi N, Cantor LB, Ku S. Acquired Brown's syndrome after a double-plate Molteno implant. *Am J Ophthalmol,* 1993; 116(5):641-2.
21. Stein JH, Papst W. Electromyographic studies on the pathogenesis and therapy of superior oblique tendon sheath syndrome. *Ber Dtsch Ophthalmol Ges,* 1969; 69:618-624.
22. Ellis FJ, Jeffery AR, Seidman DJ, Sprague JB, Coussens T, Schuller J. Possible association of congenital Brown syndrome with congenital cranial dysinnervation disorders. *JJ AAPOS,* 2012; 16(6):558–564.
23. Kaeser PF, Kress B, Rohde S, Kolling G. Absence of the fourth cranial nerve in congenital Brown syndrome. *Acta Ophthalmol,* 2012; 90:310-13.
24. Karsenti G, Karsenti D, Zaluski S, Mercadier B. Association of Stilling-Duane syndrome and Brown's syndrome with crocodile tear syndrome and other congenital anomalies. *Bull Sot Ophtalmol Fr,* 1984; 84:661-2.
25. Guyton DL. Exaggerated Traction Test for the Oblique Muscles. *Ophthalmology,* 1981; 88(10):1035–1040.
26. Stager DR, Parks MM, Stager DR, Pesheva M. Long-term results of silicone expander for moderate and severe Brown syndrome (Brown syndrome 'plus'). *J AAPOS,* 1999; 3:328–332.
28. Jacobi KW. Tenectomy of the superior oblique in the tendon sheath syndrome (Brown). *Klin Monbl Augenheilkd,* 1972; 160(6):669-74.
29. McNeer KW. Untoward effects of superior oblique tenotomy. *Ann Ophthalmol,* 1972; 4:747-55.
30. Parks MM, Eustis HS. Simultaneous superior oblique tenotomy and inferior oblique recession in Brown's syndrome. *Ophthalmology,* 1987; 94(8):1043-8.
31. Wright KW. Results of the superior oblique tendon elongation procedure for severe Brown's syndrome. *Trans Am Ophthalmol Soc,* 2000; 98:41-8.
32. Scott AB, Knapp P. Surgical treatment of the superior oblique tendon sheath syndrome. *Arch Ophthalmol,* 1972; 88:282-6.
33. Suh DW, Guyton DL, Hunter DG. An adjustable superior oblique tendon spacer with the use of nonabsorbable suture. *J AAPOS,* 2001; 5(3), 164–171.
34. Moghadam AA, Sharifi M, Heydari S. The results of Brown syndrome surgery with superior oblique split tendon lengthening. *Strabismus,* 2014; 22(1):7-12.



**Cite This Article as:** *Saluja G, Bhari A. Brown's Syndrome.*

**Acknowledgments: Nil**

**Conflict of interest: None declared**

**Source of Funding: None**

**Date of Submission: 1 August 2019**
**Date of Acceptance: 2 October 2019**


## Address for correspondence


**Gunjan Saluja** MD, DNB
Squint and Neuro Ophthalmology Services, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India
Email id: gunjansaluja2015@gmail.com


**Quick Response Code**