

ACQUIRED INTERMITTENT SUPERIOR OBLIQUE TENDON SHEATH SYNDROME (SOTSS) — 3 case reports

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Abstract

Three cases of acquired, intermittent, unilateral Superior Oblique Tendon Sheath Syndrome (SOTSS) or Brown's Syndrome are presented. They presented with differing symptoms and on clinical assessment of ocular movements demonstrated a marked restriction, and at times, a complete absence of elevation in adduction of the affected eye — depicting a marked inferior oblique underaction. This abnormality of movement possibly as a result of prevention of muscle movement through the trochlea was found to be due to a localized inflammation, possibly of the orbit in one case, a systemic disease in another, and of unknown cause in the third. The intermittent nature and 'click' phenomenon will be described and discussed.

Key words: Acquired SOTSS, 'click' phenomenon.

INTRODUCTION

Superior Oblique Tendon Sheath Syndrome (SOTSS) or Brown's Syndrome is a condition characterised by a restriction or absence of elevation in adduction, in an affected eye¹ It is most commonly seen as a congenital condition but as demonstrated by the following cases can be acquired. When acquired the condition can be associated with an intermittent nature and the 'click' phenomenon. It can be acquired at any age^{1,2} but usually seen in childhood^{3,4} and in young adults^{3,6}. The onset of this condition may be sudden or gradual and may eventually disappear completely with or without any treatment^{1,6}.

Inflammation and trauma appear to be the most common aetiological factors responsible for many of the acquired intermittent SOTSS. Some of the inflammatory conditions include — generalised systemic inflammation such as

juvenile or adult rheumatoid arthritis^{2,5,7,8,9,10} and Systemic Lupus Erythematosus (SLE)¹²; localised inflammation of the superior oblique muscle or tendon as a result of a stenosing tenosynovitis (ST)^{2,5,6,8,10,11,12,14}; sinusitis^{7,9}; pregnancy¹⁶; orbital tumors⁸; and inflammation of unknown cause^{11,13}. Trauma to the area of the trochlea^{9,11}; damage to the orbital walls⁸; surgery and/or trauma to the superior oblique muscle, as well as anomalies of the tendon sheath of the superior oblique^{3,7,9}, are other common causes reported for the acquired types of SOTSS.

This interference to normal superior oblique muscle movement through the trochlea is believed to be as a result of some intermittent mechanical restriction or defect of the superior oblique muscles' tendon or its sheath, most commonly involving the area just behind the trochlea, which prevents the free passage of the muscle and therefore no elevation in adduction.

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Figure 1: Case 1 on presentation in primary position and dextro-elevation note marked underaction of left eye in dextro-elevation

This mechanical restriction may be in the form of thickening, swelling, nodules or adhesions of the superior oblique muscle, tendon sheath, or some anomaly of the trochlea or its surrounding region^{8,9,18}.

Stenosing Tenosynovitis (ST) of the hand, a condition usually seen in children or young adults, has been found to show similar mechanisms to that of intermittent acquired SOTSS¹¹. The anomaly appears to lie in the tendon and sheath of the hand and superior oblique muscle, respectively.

Case 1

A thirty-one year old lady presented with an inability to move her left eye up and inwards for two days. She stated that this occurred intermittently over the previous two years beginning with

difficulties focussing then an inability to move the eye, this usually lasting for approximately half a day. She also stated that over the years she felt a 'click' just before full ocular muscle movements of her eye was possible. Pain and diplopia had never been experienced. Her general health was good, she wore glasses for a small hypermetropic astigmatic refractive error, and there was no other relevant ocular family history except that both her children had convergent squints which required surgery.

Ocular examination showed the following:-
Visual Acuity — corrected right and left 6/6 N5
Cover Test — near and distance a small right hypertropia 5▲ increasing on right gaze
Abnormal head posture — slight tilt to left
Ocular Movements — marked left SOTSS, left superior rectus underaction, slight tenderness



Figure 2: Case 1 post steroid treatment, six days later. Full muscle movement of left eye in dextro-elevation

over the left trochlea and injected upper nasal fornix. See figure 1.

A CT scan and full blood examination were ordered and were normal. Oral steroids (prednisolone 5mg) were prescribed to relieve the discomfort. At review in six days, she was much happier and ocular movements were full. See figure 2. Medication was ceased and when reviewed four months later she was symptom-free and her ocular examination was normal.

Case 2

A forty year old lady presented complaining of intermittent vertical diplopia and an inability to look up and to the right. She stated that at times she would forcefully move the eye into this position whereby it was accompanied by a 'click' followed by a discomforting 'tight' sensation of the left eye. She had noticed this intermittently for approximately three months. She was on medication for hypertension and suffered from sinus problems. She had presented to the clinic six months earlier after suffering food poisoning with the complaint of difficulty when focussing. At this time, all tests including ocular movements were unremarkable.

Ocular examination on this visit showed the following:-

Visual Acuity — right and left 6/6, N5 uncorrected

Cover Test — near and distance — orthophoric in primary position.

On right gaze — right hypertropia

Convergence — to 6 cms

Stereopsis (TNO) — 60 sec'

Ocular Movements — marked left SOTSS with no other over or under actions and some discomfort around the left eye.

A CT scan revealed no abnormality of the orbit or sinuses. She was given a course of oral steroids (prednisolone). One month later she still had a marked left SOTSS which was evidently worse in the morning and again could be overcome by forceably moving the eye into the affected gaze whereby she felt a 'click' and was able to demonstrate a full range of movement. This could be demonstrated several times during the consultation. At this stage the diagnosis of left superior

oblique tendonitis was made and she was given an injection of Decadron to the trochlea.

One month later she stated that the injection had helped for several days, but the condition had since reoccurred with the muscle abnormality now being constant, vertical diplopia worse, and the eye was no longer 'clicking'. She complained of muscle aches and joint pain and was sent for a full blood examination. Rheumatoid factor was negative. Antinuclear antibodies were present in a titre of 1:40 which is indicative of an underlying systemic disorder, particularly that of Systemic Lupus Erythematosus (SLE).

At the next visit, ocular examination showed a constant marked left SOTSS which could eventually be forced into full movement by extreme effort and slight pressure to the area of the left trochlea. See figure 3 & 4.

This patient is still being investigated by her GP and on last examination was given an injection of Depomedrol (longer lasting steroid) to the trochlea.

Case 3

A five year old boy presented because his parents were concerned that their son's eyes "didn't seem to be co-ordinated" and that his vision seemed to have deteriorated since hitting his head during a fall two weeks previously. There was no bruising or direct injury to either eye. He was a healthy boy with no relevant ocular family history and whose kindergarten eye test had been normal the year before.

Ocular examination revealed the following:-
Visual Acuity — right and left 6/6 (Sheridan Gardner)

Cover Test — near and distance — orthophoric in primary position.

On left gaze — left hypertropia

Convergence — to 6 cms

Stereopsis (TNO) — 60 sec'

Ocular Movements — marked right SOTSS with no other over or under actions. On attempting to move the eyes into laevoelevation the boy complained of pain and was very reluctant to move the eye into this position. Five minutes later, on repeated testing of ocular movements, a full range of movement was elicited.

DIAGNOSIS:

No.

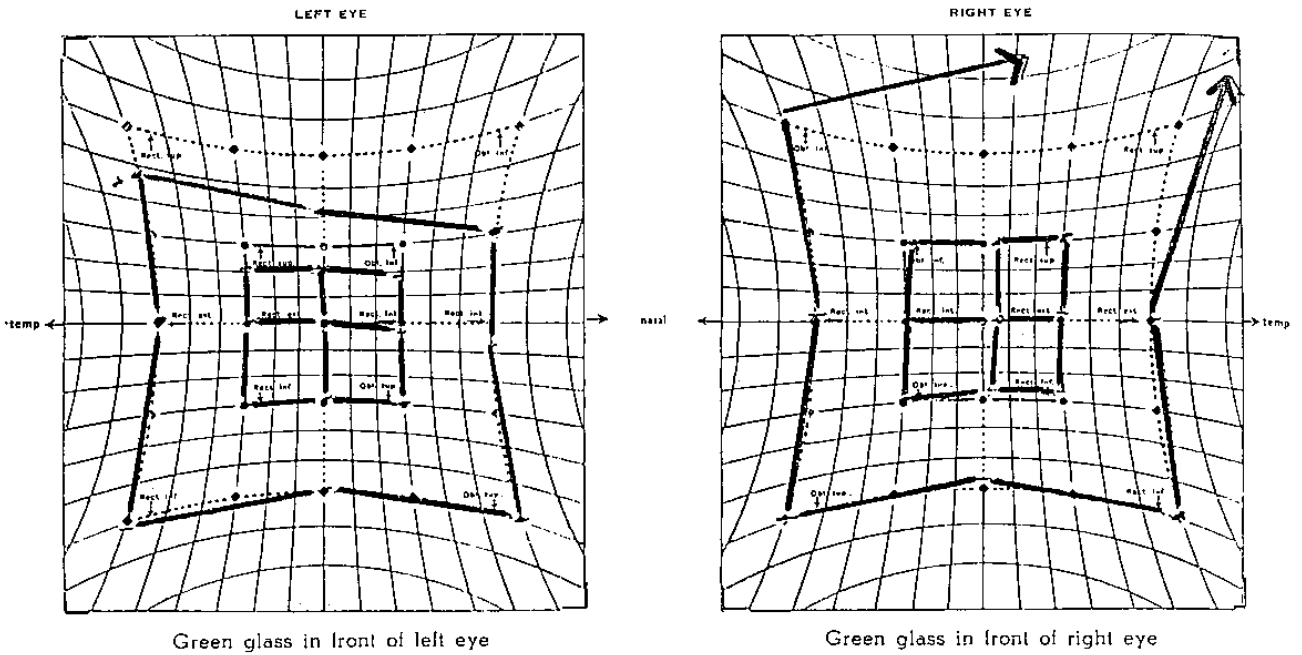


Figure 3: Case 2 Hess chart showing muscle under and overactions before 'click'

DIAGNOSIS:

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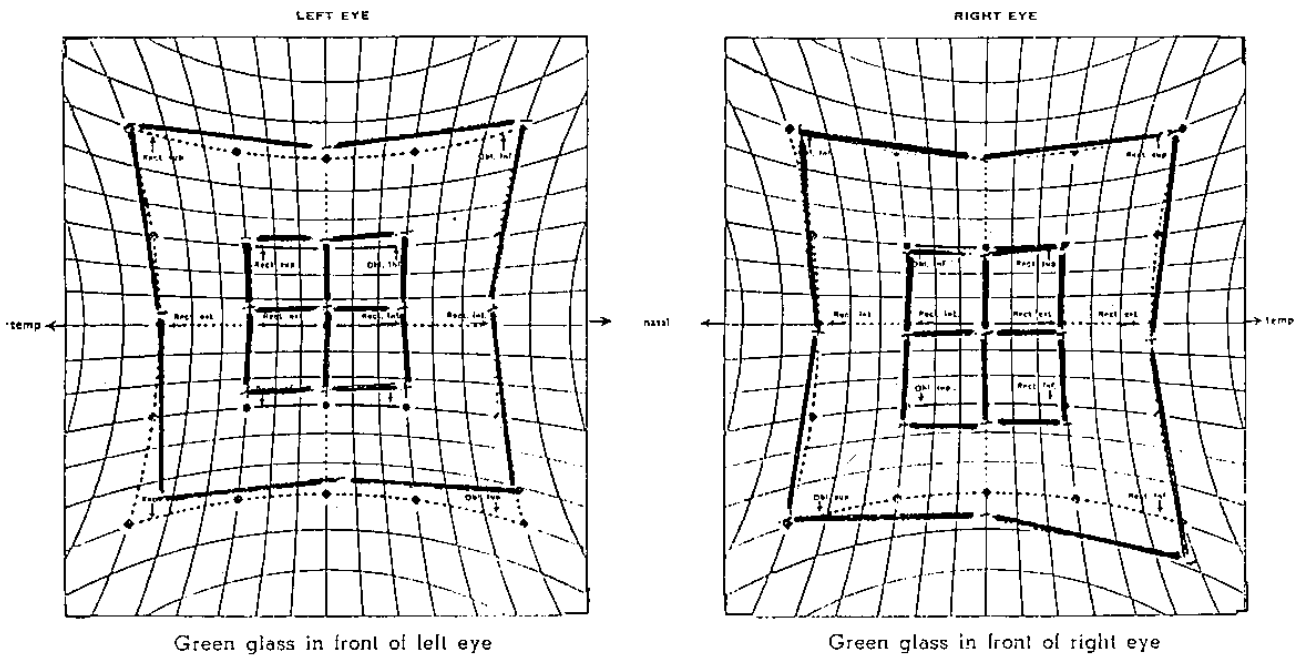


Figure 4: Case 2 Hess chart showing full muscle movements after 'click'

There was no sign of proptosis or obvious pain over the right trochlea and pressure to the area of the trochlea did not produce a 'click' or full ocular movements. However, some twenty minutes later a marked right SOTSS was again observed which recovered to full movements within moments.

Six weeks later, the ocular examination showed a constant right SOTSS. Repeated attempts to move the eye into laevoelevation or applying pressure to area of right trochlea did not achieve full movement. Since this last visit the parents have noticed the restriction of movement only intermittently. On review three months later similar intermittent ocular movements were seen.

DISCUSSION

On ocular examination, all three cases showed a marked intermittent SOTSS. The hypothesis that this condition mainly affects young adults and children is consistent with the three cases presented in this paper.

All three cases experienced different presenting symptoms. This is important because it shows that a variable range of symptoms exist for this condition. Cases 1 and 2, experienced difficulties with focussing prior to the onset of the muscle movement abnormalities. It was not possible to accurately determine whether focussing was a problem on questioning the young boy, however, the fact that the parents felt he was not seeing well could indicate a focussing problem. Blurring or 'haziness' of vision have been noted by others^{9,17}.

The onset of the presenting symptoms could not be attributed to any previous injury or illness in cases 1 and 2. However, a fall experienced by the young boy, two weeks prior to presentation may have been a predisposing factor in producing the right SOTSS.

Pain and discomfort was experienced by all three cases, this being in the region of the upper, nasal canthal area and experienced when attempting to move the affected eye into the affected gaze. This was overcome by avoiding the affected gaze in all cases. It was overcome completely in case 1, by a short course of oral

steroids, and temporarily overcome by injections of decadron and depomedrol into the trochlea in case 2.

The muscle restriction observed in each case had an intermittent nature, in that, the restriction changed from moment to moment and visit to visit. With the change in muscle restriction, from marked restriction to full muscle movement it was possible to observe a sudden release of the muscle along with an audible 'click' which was also felt by the patients. The muscle restriction in each case could be overcome either by extreme effort, repeated attempts, or by applying pressure to the area of the trochlea with an index finger.

Interestingly, cases 1 and 2 complained that the muscle restrictions and 'clicking' were worse in the morning and becoming more frequent. This could indicate that the restriction is as a result of some unknown causal entity occurring when the eye is not being used, but improves as the day progresses and with repeated eye movement.

It appears that the 'click' is always experienced and felt in the area of the trochlea. The 'click' most likely indicates a sudden release of the muscle as it passes through the trochlea overriding any obstruction and therefore resulting in full muscle movement. The 'click' phenomenon is regarded to be a stage in the resolution of the condition⁹. The 'wearing down' of swelling or enlargement of trochlea with growth is believed to be a cause of this intermittent condition^{3,9}.

CONCLUSION

The muscle abnormalities, intermittent nature, 'click' phenomenon and symptoms presented in these three cases show some of the common signs associated with acquired, intermittent SOTSS as reported by others.

As seen the symptoms and possible causes are numerous, therefore it is most important to attempt to determine the cause of the condition before any form of treatment is considered. A thorough systemic examination should be carried out.

Ocular movements should be tested routinely on all patients, particularly adults, as this condi-

tion may be a common phenomenon which is being missed and may be an important sign leading to the diagnosis of some underlying systemic disease.

ACKNOWLEDGEMENT

I would like to thank the three patients, Drs Barry Lansdell and Patrick Lockie for use of their patients and advice. The orthoptic staff at The Royal Victorian Eye and Ear Hospital and Lincoln for all their encouragement, help and advice in writing this paper. Many thanks also to the medical illustration staff at The Royal Victorian Eye and Ear Hospital for the photography.

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