Intratrochlear steroid injections in acquired Brown syndrome—a case series

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PURPOSE To present our experience in the treatment of children with acquired Brown syndrome by means of intratrochlear injection of betamethasone.

METHODS The medical records of patients treated with intratrochlear betamethasone in 2016 at the Aravind Eye Hospital, Madurai, were reviewed retrospectively. The following data were collected: pre- and postoperative orthoptic work-up, blood work, and neuroimaging. Betamethasone injection was administered 2-8 weeks following onset of symptoms.

RESULTS Five children (4 girls), 1.5-15 years of age, were included. During the postoperative period, abnormal head posture and elevation in adduction improved in 4 subjects but did not resolve completely. The median vertical deviation was 11.5^{Δ} preoperatively and reduced to 3.5^{Δ} postoperatively. A significant reduction in deviation was demonstrable on diplopia and Hess charting in 2 of the older children. Subject 2, who did not show improvement after injection, was prescribed prism glasses and became diplopia free.

CONCLUSIONS In this case series, children with acquired Brown syndrome of idiopathic or presumed inflammatory etiology showed significant reduction in deviation and symptoms following intratrochlear injection of betamethasone. We recommend that this treatment be considered for children affected by acquired Brown syndrome, especially those in the amblyogenic age group. (J AAPOS 2019;23:23.e1-5)

Brown syndrome refers to limited elevation in adduction that occurs because of changes in the tendotrochlear complex that hinder the movement of the superior oblique muscle. It can be congenital or acquired. Acquired Brown syndrome is uncommon and occurs following a trauma or iatrogenically, especially after a superior oblique tuck. It can also occur secondary to an inflammatory or infectious process in the tendotrochlear complex.[1-3]

The timing and need for surgical treatment of Brown syndrome has often been debated because of the spontaneous resolution seen in some forms of congenital and acquired Brown syndrome.[4] It has been said that the lower incidence of Brown in adults compared to children could be attributed to this spontaneous resolution. A waxing and waning course has also been described in Brown syndrome secondary to an autoimmune inflammation.[1] Also, surgery does not translate to complete cure and often has disappointing results; in some instances a late consecutive superior oblique paresis can occur.

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Infrequently, acquired Brown's syndrome is labeled idiopathic in the absence of a history of trauma or previous surgery and negative laboratory panel work for infectious and autoimmune etiology. Although no inflammation can be demonstrated, we presume that an inflammatory process must have ensued for an acute limitation of elevation to occur. In which case, steroids would help to reduce inflammation and alleviate the symptoms. Injection of steroids in the trochlear area has been described for Brown syndrome secondary to trochleitis wherein there is a palpable and tender swelling in the trochlear region.[5] Steroid injections have also been administered for migraine-associated trochleitis with good results.[6] We describe a series of 5 children with acquired Brown's syndrome of presumed inflammatory etiology treated with intratrochlear betamethasone injection.

Subjects and Methods

This study was approved by the Aravind Eye Hospital Institutional Review Board. The medical records of children treated between January and December 2016 at the Aravind Eye Hospital, Madurai, were reviewed retrospectively. Children with a vertical deviation in the primary position and hence a diagnosis of severe Brown syndrome were included; all patients were treated with intratrochlear betamethasone injection.

All patients underwent blood investigations, including complete blood count, thyroid function test, erythrocyte sedimentation rate, and rheumatoid factor preoperatively to rule out an autoimmune etiology. Neuro-imaging was performed in all to identify any possible inflammation and to rule out trauma,

because clinical history can often be unreliable in children. Oral prednisolone was attempted initially for 1-2 weeks at a dose of 1 mg/kg body weight. Because there was no improvement, the children were then posted for local steroid injection.

All injections were administered under general anesthesia by a single surgeon (SS). The forced exaggerated duction test of Guyton was performed and a tight superior oblique was demonstrated, confirming the diagnosis of Brown syndrome. This was done by grasping the globe at the limbus with two Castroviejo forceps and simultaneously retroplacing, extorting, and rotating it superonasally. This stretches the superior oblique and results in increased restriction. An aggressive forced duction was then repeated several times in an attempt to free the muscle; the muscle remained tight in all included patients. The trochlear area was then painted with betadine, and the trochlea was palpated in the anteromedial part of the roof of the orbit, 4 mm behind the rim. One ml of 4 mg betamethasone was administered at this site through the skin using a 26-G needle under aseptic conditions.

Results

Five children (4 girls), 1.5-15 years of age, who presented 1-6 weeks following onset of symptoms, were included (Table 1). Of these the 2 older children presented with a complaint of diplopia; the 3 younger children presented for evaluation of an abnormal head posture noticed by parents. Subject 3, who was 5 years of age, expressed diplopia only on questioning but was then able to comprehend and cooperate for plotting a Hess chart. All 5 underwent a detailed clinical and orthoptic evaluation pre- and postoperatively. Intratrochlear injection was administered between 2 and 8 weeks after onset of symptoms. These patients are described below in chronological order of presentation.

Subject 1

An 8-year-old girl presented with complaint of diplopia of 1 week's duration. The diplopia was binocular, with vertical separation of images, and was more prominent in left gaze. She had had a computed tomography (CT) scan of the brain and orbits elsewhere that showed a pansinusitis, but she was asymptomatic. On examination, she had a small right head tilt with left head turn. Prism cover testing revealed a right hypotropia of 7^{Δ} at distance and of 5^{Δ} at near. She had a limitation of −4 in her right eye on levoelevation. She was given a course of oral azithromycin for 3 days in view of the pansinusitis but experienced no relief of symptoms. Intratrochlear steroid injection was administered 2 weeks after onset of symptoms.

She had a marked improvement of symptoms at 1 week post-injection and vertical deviation for both distance and near had reduced to 2^{Δ}. The limitation on levoelevation also improved. She was reviewed after 3 months and demonstrated similar clinical findings. Hess charting also showed an improved functioning of superior oblique muscle in its field of action. On review 1 year later, the child was orthophoric, but limitation of elevation in adduction persisted at −2.

Table 1. Description of study subjects and their clinical presentation

Subject	Age, years	Sex	Presentation	Eye involved	Time from onset of symptoms to initial presentation, wk
1	8	F	Diplopia	R	1
2	15	F	Diplopia	R	6
3	5	M	AHP	L	2
4	1.5	F	AHP	R	4
5	2	F	AHP	L	1

AHP, anomalous head posture.

Subject 2

Our second patient was a 15-year-old girl, with diplopia in upgaze for the previous 6 weeks following a sudden clicking felt in the trochlear area. She initially ignored her symptoms, but because the diplopia persisted 6 weeks later with no improvement, she presented for evaluation. On examination, there was no pain or tenderness in the trochlear area. She had a very small left head turn and had a right hypotropia of 4^{Δ} at distance and 2^{Δ} at near, with a −4 limitation of elevation in adduction.

Intratrochlear steroids were injected 8 weeks after onset of symptoms, but she was initially lost to follow-up. The deviation remained the same when she was reviewed 4 months later. Because her diplopia was bothersome, she was prescribed prism glasses and has good stereopsis.

Subject 3

A 5-year-old boy presented with a large right head turn and of diplopia of 2 weeks' duration. Oral steroids were tried initially after a negative blood panel and normal neuroimaging. In view of a persistent large left hypotropia of 16^{Δ} at distance and 18^{Δ} at near after 2 weeks, intratrochlear steroids were injected. One month after treatment his head turn had reduced significantly, and he had a left hypotropia of 3^{Δ} at distance and near. Elevation on adduction also improved (Figure 1), and improved field of action was demonstrable on Hess charting (Figure 2).

Subject 4

An 18-month-old girl presented for evaluation of an abnormal head posture noticed for the previous 4 weeks following an episode of fever. She showed a right hypotropia and exotropia in primary position and a limited elevation in adduction. The angle measured by modified Krimsky test was exotropia of 16^{Δ} and hypotropia of 20^{Δ}. Intratrochlear steroid injection was administered 6 weeks following onset of symptoms. At 1 and 3 months after treatment, her head posture had improved, and a small right hypotropia of $<5^{\circ}$ was noted on the Hirshberg light reflex testing.

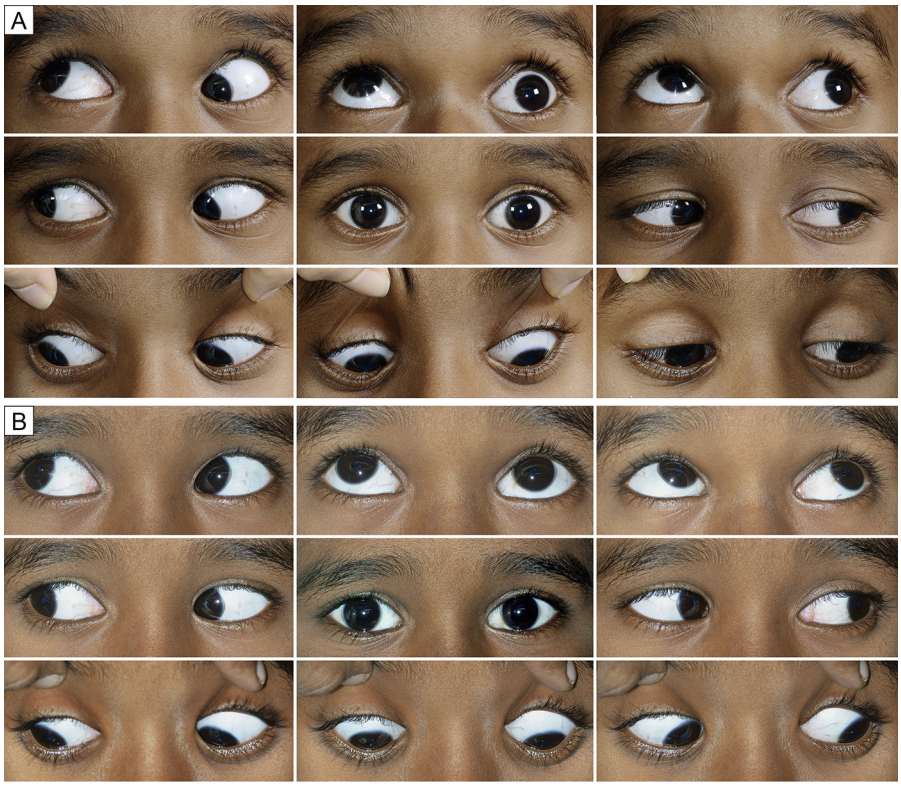

FIG 1. Clinical nine-gaze photographs of subject 3 before (A) and 1 month after (B) intratrochlear betamethasone injection.

Subject 5

A 2-year-old girl presented for evaluation of an abnormal head posture noticed for the previous 1 week. The parents noted that she had had a cold 10 days prior to onset of head tilt. On examination, the patient had a left head tilt with a small chin elevation. Elevation on adduction was −3 in her left eye. She was not cooperative for measurement of vertical deviation. A hypotropia of 16^{Δ} was noted in the left eye on the modified Krimsky testing. Intratrochlear steroid injection was administered 2 weeks after onset of symptoms, in view of our clinical diagnosis of acquired Brown syndrome and promising results seen with our earlier patients. At 1 month after the procedure, the head posture had improved dramatically and the child appeared orthotropic (Figure 3).

Discussion

Of the 5 patients, 4 showed improvement in head posture and reduction in vertical deviation in primary gaze on the first postoperative visit. In all patients, the limitation of elevation in adduction improved but persisted to some degree. As the measure of limitation may vary among examiners, Hess charting was performed in the 3 older children and demonstrated a marked improvement in the field of action in the superior oblique muscle.

Although variations exist in the timing of injection and follow-up varied, our results are promising (Table 2). The median vertical deviation was 11.5^{Δ} before treatment and reduced to 3.5^{Δ} after ($P = 0.094$). In the one older child that did not show improvement in vertical deviation, the angle was small preoperatively, and the steroid injection was administered 8 weeks following onset of diplopia.

Brown originally described this syndrome and its subtypes as the true sheath syndrome and the simulated sheath syndrome.[7,8] The latter was further classified into three types: Spontaneous recovery, intermittent, and acquired cases. The terms *congenital* or *acquired* and *intermittent* or *constant* are most frequently used to describe this condition. Based on the severity of vertical deviation, Brown syndrome is classified as mild, moderate, and severe.

The motility defect of Brown syndrome has consistent and characteristic features, making it easily recognizable clinically.[9] The most striking feature is an inability to actively or passively elevate the affected eye in full adduction, with identical results on version or duction testing[10]

Acquired Brown syndrome has been described[1-3] in connection with inflammatory diseases such as rheumatoid arthritis, systemic lupus erythematosus, idiopathic tenosynovitis, sinusitis, and scleritis, and primary and secondary tumors of the orbit. Other causes include blunt trauma to the orbit, sinus surgery, excessive tucking of the superior oblique tendon, and scleral buckling surgery. In some cases, acquired Brown syndrome is idiopathic.[8] Although it is difficult to discern trochlear abnormalities on neuroimaging, all our patients were imaged prior to injection to look for signs of trauma.

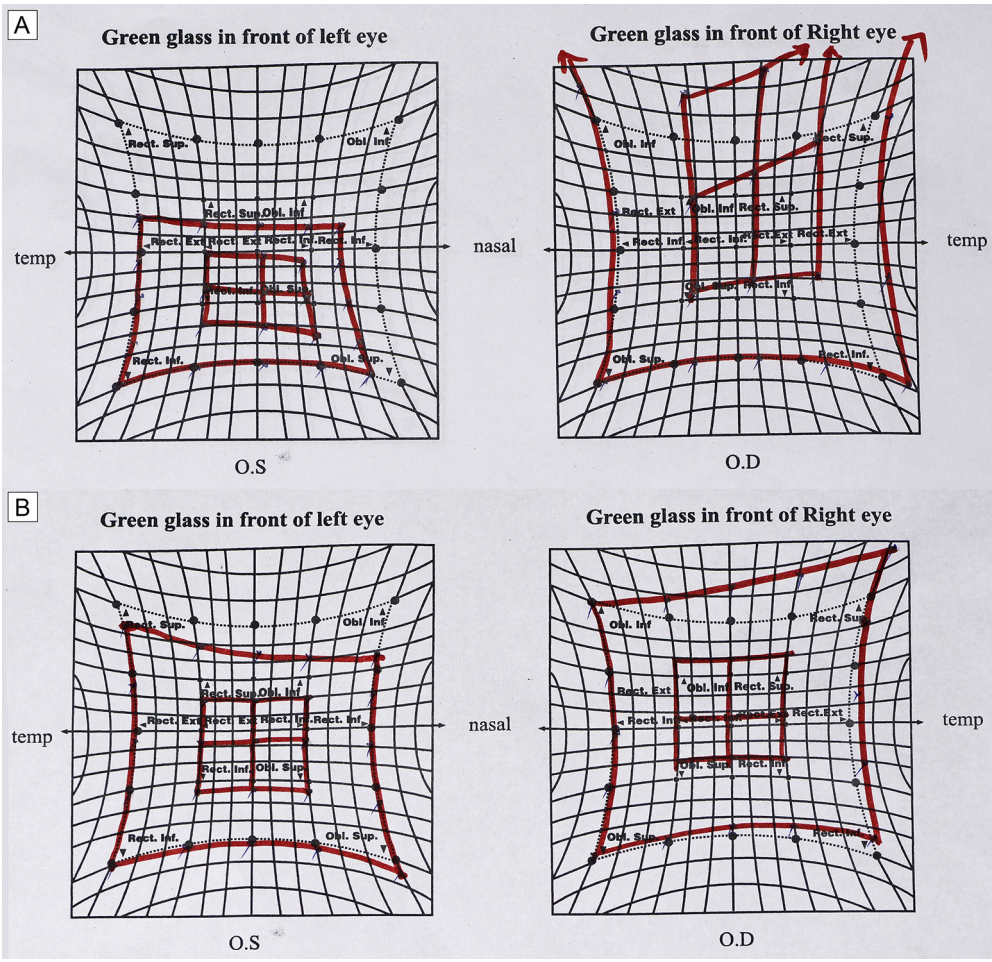

FIG 2. Hess charting of subject 3 before (A) and 4 months after (B) injection.

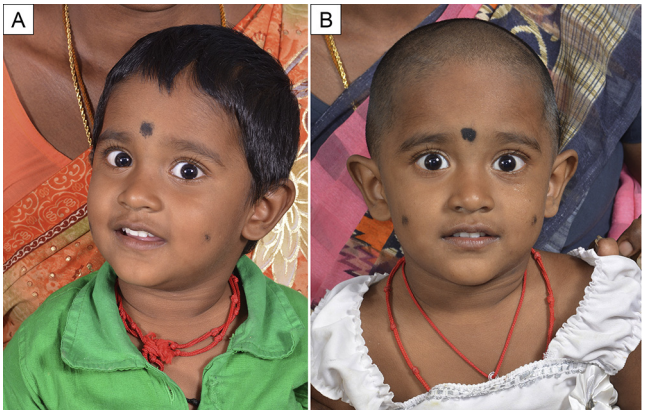

FIG 3. Subject 5 before (A) and 1 month after (B) injection.

Steroid injection in the trochlear region for acquired Brown syndrome of inflamatory origin was first reported by Hermann[5] in 1978. The 2 patients he described exhibited local inflammatory signs in the trochlear region and were diagnosed with stenosing tenosynovitis. A noticable improvement was seen after injection of 40 mg methylprednisolone acetate in the trochlear area. Beck and Hickling[11] also describe using an orbital injection of 1 mL of 40 mg methylprednisolone acetatenate over the superomedial aspect of both eyes 1 week apart in a patient with bilateral superior oblique tendon sheath syndrome complicating rheumatoid arthritis. They reported improvement both subjectively and objectively after the steroid injection. Peritrochlear steroid injection containing 1 mL of 3 mg of dexamethasone and 3 mg of methylprednisolone has also been described to produce quick relief in patients with trochleitis associated with migraine headache.[6]

The patients in our series were diagnosed with idiopathic acquired Brown syndrome because no etiological factor could be identified. We ruled out a congenital etiology, because there was no abnormal head posture seen in photographs of all 5 patients prior to the onset of symptoms. Although no inflammation can be demonstrated, we presume that some inflammatory process must have caused the acute limitation of elevation. Because there was a demonstrable improvement with

Table 2. Details of orthoptic evaluation before and after injection

	Before injection					After injection		
	Deviation, PD					Deviation, PD		
Case	Distance	Near	Elevation in adduction	Time to injection	Follow-up, mo	Distance	Near	Elevation in adduction
1	L-HT 7	L-HT 5	−4	2	2	Ortho	L-HT 2	−3
2	L-HT 4	L-HT 2 XT 3	−4	8	4	L-HT 3; XT 3	L-HT 3; XT 3	−3
3	R-HT 16 XT 2	R-HT 18 XT 2	−4	4	1	R-HT 3	R-HT 3	−2
4	—	L-HT 20; XT 16	−3	6	1	—	Small L-HT	−2
5	—	R-HT 16	−3	2	1	—	Ortho	−3

HT, hypertropia; *Ortho*, orthotropia; *PD*, prism diopter; *XT*, exotropia.

steroid injection, we thus classified these cases as resulting from a presumed inflammatory process. Kushner[12] has found that steroid injection is more effective in the cryptogenic cases of inflammatory Brown syndrome than in cases associated with identifiable autoimmune disease. We speculate that the absence of any improvement in subject 2 could be attributed to the long delay in treatment.

It might be argued that improvement in these 5 patients would have occurred spontaneously and was not due to the effect of the steroid injection. However, none of the patients showed any improvement when observed for the first 2 weeks or with a trial of oral steroids, and considerable improvement was seen 1-2 weeks following injection in the children that presented for early follow-up examination.

Although our results are limited by the small sample size, a randomized clinical trial would be impossible to undertake with such a rare condition. Also, because the study was retrospective, there was no uniformity in the duration of oral steroids, timing of injection and follow-up. There could also have been subjective variations in interpreting the limitation of muscle action. Nevertheless, Hess charts and clinical photographs are convincing evidence of improvement, and, based on our experience, we recommend intratrochlear steroid injection in cases of acquired severe Brown syndrome of presumed inflammatory etiology following necessary blood work and neuroimaging.

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