

## Research Article

# Is Amblyopia Really Rare in Brown Syndrome?

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## Abstract

**Purpose:** Evaluating the sensory status and accompanying horizontal and vertical deviations in Brown syndrome cases.

**Method:** Visual acuity, affected eye, refractive error, presence of amblyopia, horizontal and vertical deviations were evaluated retrospectively in twenty one Brown syndrome cases observed in Ankara Ataturk Training and Research Hospital Strabismus Unit between 2006 and 2017.

**Result:** The mean follow-up period was 4.31±3.88 years for twenty-one patients in this study. The left eye was affected in 47.6% (n=10) of the cases and 9.5% (n=2) were bilateral. Amblyopia was observed in 28.6% (n=6) of the patients. In four patients, amblyopia was on the same side as Brown syndrome. Thirteen patients (61.9) were orthophoric. Anisometropia was detected in five (23.8%) patients Anisometropia and strabismus were observed together in four of these five patients.

**Conclusion:** Brown syndrome patients are mostly orthophoric while accompanying horizontal and vertical deviations were present in 38.1% of the cases and amblyopia in six patients (28.6%). Detection of deviations and possible amblyopia secondary to deviation is critical in Brown syndrome because of their impact on binocular sensory development.

**Keywords:** Amblyopia; Anisometropia; Brown syndrome

## Introduction

Brown syndrome is a rare ocular motility disorder, characterized by restriction of elevation in adduction due to abnormalities in the trochlea or superior oblique tendon sheath [1]. This syndrome may be congenital, acquired, intermittent or constant. Although its etiology remains uncertain, various causes have been associated with Brown syndrome [2].

The reason of congenital Brown syndrome is an abnormality of the superior oblique tendon or trochlea, while acquired Brown syndrome is caused by various reasons including infection, scleritis, systemic inflammatory disease such as rheumatoid arthritis and systemic lupus erythematosus, trauma, orbital metastatic deposits in the extraocular muscles, mucopolysaccharidosis, glaucoma drainage device implantation, scleral buckling, frontal sinusitis, orbital wall fracture or sinus surgery [2-10].

Variable clinical features can be seen in Brown Syndrome such as a positive forced duction test, down-shoot in adduction, V pattern exotropia in the up gaze, abnormal head position including chin up and contralateral face turn, and hypotropia in primary position [3].

Amblyopia, a neurological developmental disorder that affects 3-5% of the population [13,14], is characterized by monocular visual acuity loss in an anatomic healthy eye, despite optimal refractive correction. There are several causes for amblyopia including uncorrected refractive error, anisometropia, strabismus and visual deprivation (such as cataract, opaque cornea, complete ptosis, prolonged uncontrolled patching) [15-19].

Several studies have reported that amblyopia is not common in

Brown syndrome because of rare incidence of suppression [11,12]. Although some case reports are available, there is limited literature on clinical finding with accompanying anisometropia and deviations in Brown syndrome cases. We aimed to report evaluation of sensory status and accompanying horizontal-vertical deviations in Brown syndrome cases.

## Materials and Methods

The medical records of twenty-one patients that visited Ataturk Training and Research Hospital Strabismus Unit from 2006 to 2017 with the diagnosis of Brown syndrome were retrospectively reviewed. Congenital and trauma induced acquired Brown syndrome patients having no ocular and sinus surgery, with a minimum of six-month follow-up was included in this study. Patients with amblyopia due to ocular media opacity, retinal disorders and systemic and autoimmune diseases, tumors, orbital and sinus surgery history were excluded from the study. The medical records of the patients including demographic features, affected eye, best-corrected visual acuity, presence of amblyopia, the amount of horizontal-vertical deviations and refractive errors were reviewed. Visual acuity was evaluated by Snellen chart. For statistical analysis the visual acuity was converted to LogMAR. Amblyopia was defined as a difference of two or more lines between the best-corrected visual acuity of two eyes. Anisometropia was accepted as the difference of 1.50D or more (sphere or cylinder) between the two eyes. The amount of manifest deviation was assessed by using prism cover test. Statistical analysis of the data obtained in this study was performed using the Statistical package for Social Sciences (SPSS) 20 program. Categorical data were analyzed using frequency and percentages. The comparisons between eyes with and without Brown syndrome were made by Mann Whitney U test. The p

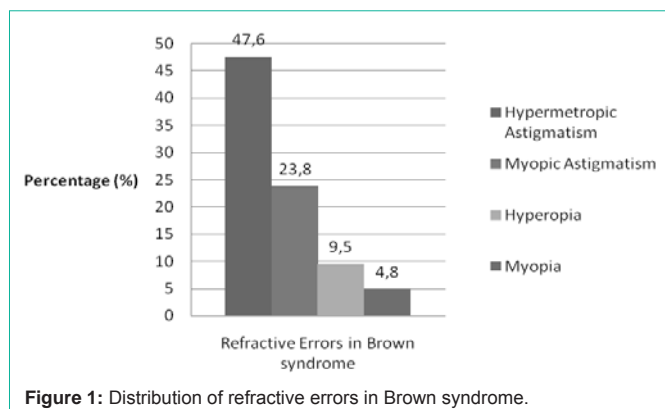


Figure 1: Distribution of refractive errors in Brown syndrome.

values less than 0.05 were accepted as statistically significant.

## Results

The mean follow-up period was  $4.31 \pm 3.88$  years for twenty-one patients. Seventeen cases (81%) were female and the mean age was  $10.62 \pm 5.07$  years. The left eye was affected in 47.6% ( $n=10$ ) of the cases and 9.5% ( $n=2$ ) of the cases were bilateral.

The mean best corrected visual acuity was  $0.91 \pm 0.16$  and  $0.91 \pm 0.19$  for the eye with Brown syndrome and the other eye respectively ( $p=0.23$ ).

The mean corrected spherical refraction error was  $+0.80 \pm 2.02$  in the eye with Brown syndrome;  $+0.06 \pm 1.41$  for the other eye without Brown syndrome ( $p=0.14$ ) while corrected astigmatic refractive error was  $-0.08 \pm 1.28$  for the eye with Brown syndrome and  $-0.36 \pm 1.24$  for the other eye ( $p=0.35$ ). Ten patients (47.6%) had hypermetropic astigmatism, five patients (23.8%) had myopic astigmatism 2 patients (9.5%) had hyperopia and 1 patient (4.8%) had myopia. Three of all patients (14.3%) had no refractive error (Figure 1).

Amblyopia was observed in 28.6% ( $n=6$ ) of the patients. The mean best corrected visual acuity of amblyopic eyes was  $0.56 \pm 0.26$ . In four patients, amblyopia was on the same side as Brown syndrome (Figure 2). Amblyopia and deviations (two of the patients have esotropia) were seen in three patients on the same side as Brown syndrome.

Anisometropia was detected in five (23.8%) patients and four patients had high refractive error in the eye with Brown syndrome. Amblyopia due to anisometropia and deviations was observed in four of these five patients (Figure 2).

61.9% ( $n=13$ ) of the patients were orthophoric while deviations were seen in 38.1% ( $n=8$ ) of them. Isolated hypotropia, hypotropia with exotropia, esotropia and exotropia was observed in 14.3% ( $n=3$ ), 4.8% ( $n=1$ ), 9.5% ( $n=2$ ), 9.5% ( $n=2$ ) of the cases respectively (Figure 3). In addition, V pattern deviation in upward gaze was observed in 23.8% ( $n=5$ ) of the patients.

The mean horizontal near and distance deviations were  $15\Delta$  ( $12-18\Delta$ ) and  $11\Delta$  ( $4-20\Delta$ ) respectively. The mean vertical deviation was  $15\Delta$  ( $12-20\Delta$ ) at near and  $13\Delta$  ( $10-16\Delta$ ) at distance.

## Discussion

We investigated clinical findings 21 cases with Brown syndrome. We observed that amblyopia was present in 28.6% ( $n=6$ ) of patients

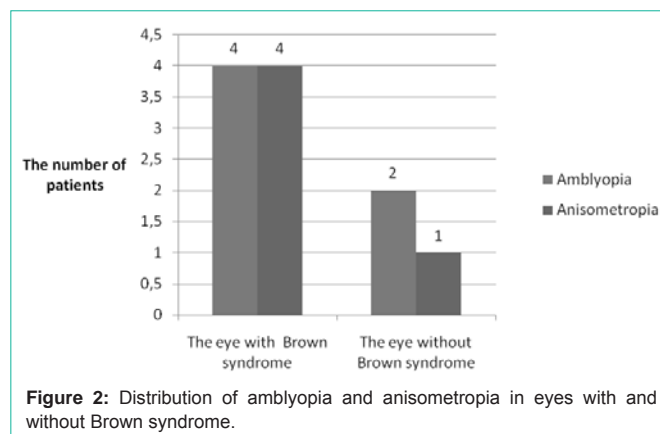


Figure 2: Distribution of amblyopia and anisometropia in eyes with and without Brown syndrome.

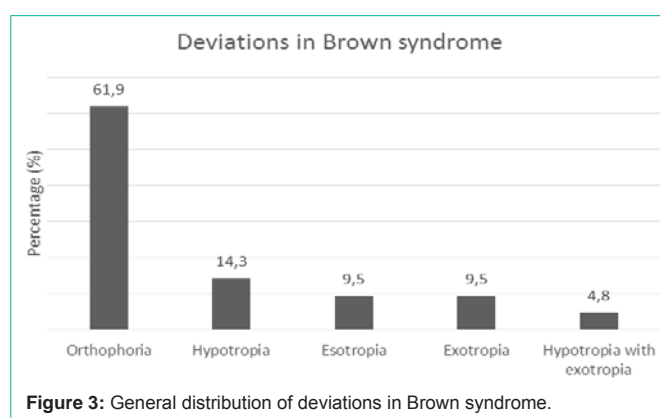


Figure 3: General distribution of deviations in Brown syndrome.

and horizontal or vertical deviations in 38.1% of the patients with Brown syndrome.

The most common refractive error that can induce amblyopia is uncorrected hyperopia which is more common due to the natural distribution of refractive error at the young age group. Similarly, hyperopic astigmatism (47.6) was most common refractive error in this study.

Nonetheless there are studies that offer different views on the effect of anisometropia on amblyopia. According to several studies, the depth of amblyopia increases as the amount of anisometropia increases [20,21]. But there are also studies advocating that there is no association between anisometropia and amblyopia in Brown syndrome because of rare incidence of suppression [11,12] whereas severe amblyopia was presented in one case report [25]. Similarly, Brown et al. have reported that amblyopia and refractive errors have no significant importance in Brown syndrome. Although in the study of Sekeroglu et al 7 patients (15.9%) had amblyopia and 14 patients (31.8%) had anisometropia [26], anisometropia and strabismus were not reported as significant amblyogenic factors in Brown syndrome.

Strabismus causes inhibition in the retinocortical pathway and in vivo studies have shown decreased metabolic activation of the visual cortex [27]. Previous studies have reported that strabismic amblyopes have denser suppression than anisometropic amblyopes [14-28].

According to the study of Sekeroglu et al exotropia was found to be the most common strabismus in cases with Brown syndrome. However, in our study we found that anisometropia and strabismus

were seen in Brown syndrome in association with amblyopia.

Similar to previous studies there were more female patients (81%) in this study [12]. We observed that the left eye was involved in 47.6% (10/21 patients) of our cases and 9.5% (n=2) of the patients were bilateral while the right eye was involved (70/126 patients) in the study of Wright et al [12]. This difference may depend on the number of patients as well as the possible differences in the populations studied.

We have some limitations in this study. Since it is a retrospective study some information related to sensorial status is missing. Patients without symptoms are usually lost to follow up resulting in variable follow up information. Limited size of the study group should also be considered while interpreting the results. The number of patients should be increased to confirm the high prevalence of amblyopia and anisometropia in Brown syndrome.

## Conclusion

In conclusion we have observed that Brown syndrome patients are mostly orthophoric, although horizontal and vertical deviations and amblyopia can be seen in contrast to previous reports presenting lower incidence [11,12]. We think that the detection of symptoms such as deviations, anisometropia and amblyopia is important for early treatment and normal binocular sensory development in Brown syndrome patients.

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