

Presence of Fusion in Albinism after Strabismus Surgery Augmented with Botulinum Toxin (Type A) Injection

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It is commonly accepted that albino patients with strabismus rarely achieve binocularity and depth perception after strabismus surgery. The presence of retino-geniculo-cortical misrouting, a hallmark of the visual system in albinism, does not necessarily cause total loss of binocular vision, however, not even in albino patients with strabismus. Recently some degrees of stereopsis were reported in albinism patients with minimal clinical nystagmus, if any, in the absence of strabismus. It is possible that patients with albinism and strabismus have binocular visual potential which appears after strabismus correction and provides appropriate postoperative alignment in the long term. Here we present two cases of clinically diagnosed oculocutaneous albinism, an 18-year-old girl and a 16-year-old boy, both with exotropia ≥ 40 prism diopter, who gained acceptable alignment and fusion after surgical correction of their strabismus as demonstrated on Bagolini testing. In cases of albinism accompanied by visual pathway abnormalities and strabismus, binocular visual potential is not impossible, and some levels can be expected. Thus, these patients, like other cases of strabismus, may benefit from treatment of strabismus at an earlier age to achieve appropriate alignment, cosmetic satisfaction, and a possibly increased chance of fusion.

Key Words: Albinism, Botulinum toxins, Strabismus

In albinism, excessive decussation of retinostriated projections in the optic chiasm leads to changes in the lateral geniculate laminae, abnormalities in binocular vision and retinal correspondence. Despite these abnormalities, some patients with oculocutaneous albinism (OCA) and no significant strabismus have normal binocular vision [1]. Considering potential fusion in these patients, binocular vision can be expected to develop or improve after strabismus correction. There is no preferred procedure for strabismus correction in albino patients, and there is scarce literature on binocular vision after strabismus correction [2,3]. Here we report two cases of OCA with exotropia who gained fusion after bilateral lateral rectus recession augmented with bilateral botulinum toxin A (BTA) injection. To our

knowledge, this is the first report of such results in strabismic patients with albinism.

Case Reports

Two patients presented with clinically diagnosed OCA (diffuse skin and hair hypopigmentation, diffuse iris transillumination, foveal and optic nerve dysplasia and fundus hypopigmentation [Figs. 1 and 2]) and exotropia $\geq 40\Delta$. They both had mild horizontal jerk nystagmus without abnormal head posture, and neither had a history of strabismus surgery. Patients underwent a full ophthalmic examination, including ocular motility assessment, cycloplegic refraction, and anterior and posterior segment examination. To avoid the effect of angle kappa, deviation was measured using the alternate prism cover test. Particular attention was paid to the presence of fusion. Binocular function tests were performed preoperatively and postoperatively. Fusion was tested with Worth 4-dot test, Bagolini lenses and stereoacuity with near and distance Randot test.

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The distance Randot test was performed at 40 cm instead of 3 m to assess higher stereopsis thresholds. These tests were conducted 4 months after surgery when the effect of BTA would have reached a minimum and surgical results would be stable.

Case 1

Case 1 was an 18-year-old girl with refraction in her right and left eye of $+4.0 / -1.5 \times 180^\circ$ and $+4.0 / -2.0 \times 180^\circ$, respectively, and a best-corrected visual acuity (BCVA) of 2 / 10 in both eyes. Examination showed 45 Δ exotropia and 8 Δ hypotropia in the left eye due to an overactive superior oblique muscle and no fusion. She underwent 8 mm bilateral lateral rectus recession, left superior oblique posterior tenotomy, and intraoperative bilateral 12.5 U BTA (Dysport; Ipsen Laboratories, Porton, UK) injection to these muscles with no complications. Follow-up examinations revealed 8 Δ left esotropia at 2 weeks and 1 month, and 6 Δ left exotropia at month 4, which remained constant up to 12 months. The patient demonstrated fusion on Bagolini test (Fig. 3). Before surgery, the patient had no perception



Fig. 1. Fundus photograph of the right eye of the patient, case 1.



Fig. 2. Fundus photograph of the right eye of the patient, case 2.

in the Bagolini test; however, at 4 months after surgery, the patient was able to perceive the major parts of both arms of the X generated by Bagolini lenses, despite small areas of suppression around the fixation light.

Case 2

Case 2 was a 16-year-old boy with refraction in his right and left eye of $+3.5 / -1.75 \times 20^\circ$ and $+3.25 / -2.0 \times 155^\circ$ and a BCVA 2 / 10 and 3 / 10, respectively. He had 50 Δ left exotropia and no fusion. He underwent 9 mm bilateral lateral rectus recession, augmented with intraoperative bilateral 12.5 U BTA (Dysport) injection, with no complications. On week 2 and month 1 follow-ups, he had 10 Δ left esotropia which decreased to 6 Δ left exotropia from the 4th month postoperative follow-up onwards; fusion in the Bagolini test was also observed at 4 months after surgery (Fig. 4).

Discussion

In albinism, the hallmark of the visual pathway is the misrouting of retino-geniculo-cortical fibers. Abnormal crossing of the projections from the central regions of the temporal retina and paucity of ipsilateral projections lead to changes in lateral geniculate laminae, abnormal function of binocular visual cells in the cortex, and abnormal retinal correspondence. This defective development in the visual pathway is thought to be responsible for the absence of stereopsis and fusion in many albino patients [4], but binocular vision can still be normal in some patients.

Apkarian and Reits [5] studied global stereopsis in 18 albino patients using a TV generated Randot stereotest and reported stereopsis in 9 patients (6 of the remaining 9 patients had tropia); they attributed their findings to the compensating role of corpus callosum. Laboratory assessments of Cobo-Lewis in 5 patients without clinical stereopsis demonstrated the existence of detectable stereopsis of several thousand arc seconds or better. They concluded that



Fig. 3. Preoperative (A) and 4 months postoperative (B), case 1.

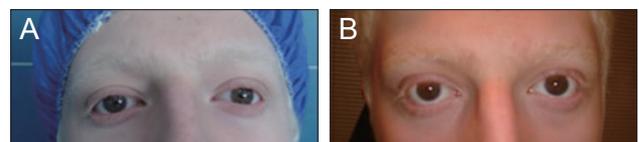


Fig. 4. After preoperative (A) and 4 months postoperative (B), case 2.

the stereopsis threshold is probably higher and undetectable through routine tests [6]. Boyle et al. [2] examined 45 albinos with 20 / 100 or better visual acuity and less than 10 Δ deviation, and found stereopsis in 19 cases; 5 patients without nystagmus had fine stereopsis. This presentation of fine stereopsis could be due to a normal path of peripheral projection of the temporal retina, and the peripheral visual fields having both ipsilateral and contralateral projections and normal retinal correspondence [6].

The surgical approach in exotropia greater than 40 Δ is the 3-muscle procedure, or the 2-muscle procedure with intraoperative BTA injection into one of the recessed muscles [7]. The objective is to create balance between agonistic and antagonistic muscles, and achieve acceptable alignment even after the effect of BTA subsides. An essential factor in this process is peripheral fusion of any degree [8]. Considering that retino-geniculo-cortical fibers are not misrouted in the peripheral visual field, the patient may experience different degrees of alignment at the beginning, peak, and end of the BTA effect, and may have a greater chance to achieve fusion.

Neither of the two cases presented herein had fusion pre-operatively, even when their strabismus was corrected with a prism. However, both achieved acceptable alignment after surgery in the presence of mild nystagmus, and, since Bagolini lenses create less dissociation, they demonstrated fusion in this test.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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