Surgical Dose-Response Relationship in Patients with Down Syndrome with Esotropia: A Comparative Study

Osman Bulut Ocak, Asli Inal, Ebru Demet Aygit, Serap Yurttaser Ocak, Can Ozturker, Ahmet Demirok, Birsen Gokyigit

1University of Health Sciences Beyoglu Eye Research and Training Hospital, Istanbul, Turkey
2University of Health Sciences Okmeydani Research and Training Hospital, Istanbul, Turkey

Abstract

Objectives: This study is a comparison of the effectiveness of bimedial rectus recession surgery in patients with Down syndrome and those with normal neurological development.

Methods: Records of patients with Down syndrome (age range: 2-17 years) who underwent bimedial rectus recession surgery for esotropia (ET) between April 2005 and April 2016 were reviewed retrospectively. A control group was also selected from age-matched patients with normal neurological development who underwent the same surgical procedure during the period. Ocular alignment was measured with the Krimsky test. Surgical success was defined as within 10 prism diopters (PD) of orthotropia 1 year after surgery. The case and control groups were compared in terms of preoperative and postoperative esodeviation angle at 1-year follow-up, the size of the bimedial rectus recession, and postoperative surgical success.

Results: A total of 21 patients with Down syndrome and 42 control subjects were included. The groups did not differ in either preoperative (Down syndrome group: 39.73±8.47 PD; control group: 37.91±7.65 PD) or postoperative near deviation angle (Down syndrome group: 5.45±11.45 PD; control group: 2.36±7.13 PD) or size of bimedial rectus recession (Down syndrome group: 4.68±0.40 mm; control group: 4.78±0.38 mm). Surgical success had been achieved in 15 patients with Down syndrome (80.90%), and in 35 control patients (83.33%) at 1-year follow-up. There was a significant difference between the preoperative and postoperative deviation angles in both groups (p<0.05).

Conclusion: Surgical success rate was similar in ET patients with or without Down syndrome who underwent bimedial rectus recession surgery.

Keywords: Bimedial rectus, Down syndrome, esotropia, recession surgery.

Introduction

Down syndrome, which causes neurological developmental delay, is the most common genetic disease (1). Epidemiological studies report a strabismus frequency in this syndrome of between 20% and 65% (2-6). It has been demonstrated in studies that standard bimedial rectus recession operations performed in cases with esotropia (ET) with genetic disease causing neurological developmental delay may result in over-correction (7-10). Studies conducted with only Down syndrome patients have demonstrated that the results of standard surgeries are similar to those of ET cases with normal neurological development (11-13).

The present study is a comparison of the efficacy of standard bimedial rectus surgery applied in ET patients with Down syndrome and in ET patients with normal neurological development.
Table 1. Standard bimedial rectus recession surgery doses

<table>
<thead>
<tr>
<th>Esotropia amount (PD)</th>
<th>Surgical doses (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>3.0</td>
</tr>
<tr>
<td>20</td>
<td>3.5</td>
</tr>
<tr>
<td>25</td>
<td>4.0</td>
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<tr>
<td>30</td>
<td>4.5</td>
</tr>
<tr>
<td>35</td>
<td>5.0</td>
</tr>
<tr>
<td>40</td>
<td>5.5</td>
</tr>
<tr>
<td>50</td>
<td>6.0</td>
</tr>
</tbody>
</table>

PD: Prism diopters.

Table 2. Comparison of patients with Down syndrome and control group

<table>
<thead>
<tr>
<th></th>
<th>Down syndrome</th>
<th>Control group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(n=21)</td>
<td>(n=42)</td>
</tr>
<tr>
<td>Sex, n (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>10 (47.61)</td>
<td>21 (50)</td>
</tr>
<tr>
<td>Female</td>
<td>11 (52.39)</td>
<td>21 (50)</td>
</tr>
<tr>
<td>Age (years)</td>
<td>7.37±4.81</td>
<td>7.65±4.53</td>
</tr>
<tr>
<td>Type of ET, n (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infantile</td>
<td>14 (66.66)</td>
<td>19 (45.24)</td>
</tr>
<tr>
<td>Acquired</td>
<td>7 (33.34)</td>
<td>23 (54.76)</td>
</tr>
</tbody>
</table>

ET: Esotropia.

Methods

The patient records of 110 patients with Down syndrome under 18 years of age from the hospital's pediatric ophthalmology and strabismus unit between April 2005 and April 2016 were reviewed retrospectively. All cases of bimedial rectus recession surgery were examined. The surgical dose advised in the literature for standard bimedial recession surgery was applied in all cases (Table 1). For each Down syndrome case included, 2 ET cases with normal neurological development of a similar age were included in the control group. In both groups, patients who were operated on for ET with a surgery other than bimedial rectus recession, who had been followed up for less than a year postoperatively, who had a history of previous strabismus surgery, or who were diagnosed with secondary strabismus (paralytic strabismus, etc.) were excluded. Cases with retardation in neurological development other than Down syndrome were also excluded.

The age, preoperative deviation measures, the extent of recession in the surgery, and the postoperative deviation measures in the first year were examined in the Down syndrome patients and in control group patients. Deviation measurements were performed using the Krimsky test in Down syndrome patients, and the prism cover test or the Krimsky test in the control group. Deviation measurements of 10 prism diopters (PD) and under in the first year postoperatively were accepted as surgical success. Undercorrection or overcorrection above 10 PD was considered unsuccessful. Statistics were analyzed using SPSS for Windows, Version 16.0 (SPSS Inc., Chicago, IL, USA). A p value less than 0.05 (p<0.05) was accepted as significant. The Mann-Whitney U test was used to compare the groups.

Informed consent was obtained from the parents of all participants. The study was conducted in accordance with the Declaration of Helsinki, and received approval from the Beyoglu Eye Research and Training Center Ethical Committee.

Results

Of 110 cases of Down syndrome, 35 underwent strabismus surgery for ET (31.81%). In all, 21 patients who underwent bimedial rectus recession surgery were included in the study; 10 were male (47.61%) and 11 were female (52.39%). The mean age of the patients was 7.37±4.81 years (range: 2-17 years). Fourteen cases were diagnosed with infantile ET (66.66%) and 7 cases were diagnosed with acquired ET (33.34%). Of the 42 cases in the control group, 21 patients were male (50%) and 21 were female (50%). The mean age of the control group patients was 7.65±4.53 years (range: 2-17 years). In this group, 19 had infantile ET (45.24%) and 23 had acquired ET (54.76%) (Table 2).

When the Down syndrome group and the control group were compared, the mean age (7.37 years, 7.65 years, respectively), the mean preoperative near and distance deviation measures (near: 39.73 PD, 37.91 PD; distance: 32.27 PD, 29.09 PD, respectively), the mean surgical dose (4.68 mm, 4.78 mm, respectively), mean postoperative near and distance deviation measures (near: 5.45 PD, 2.36 PD; distance: 3.90 PD, 0.98 PD, respectively) were similar. Comparison of the 2 groups revealed no significant differences (p>0.05) (Table 3).

Surgical success was achieved in 15 (80.90%) of the Down syndrome cases and in 35 cases (93.33%) in the control group. There was no statistically significant difference between the infantile and acquired ET cases in either group in terms of surgical success (p>0.05). Undercorrection was detected in 2 of the Down syndrome cases and in 1 case in the control group, while overcorrection was observed in 1 case in the control group. No surgical complications were detected in any of the cases.
Discussion

Down syndrome (Trisomy 21) is the most common genetic disease causing neurological developmental delay (1). In cases with Down syndrome, disorders of the eyelids, refractive disorders, blepharitis, nasolacrimal duct obstruction, nystagmus, cataract, iris pathologies, optical disc and retinal disorders, amblyopia, and strabismus are encountered in varying frequencies (2-5). The presence of strabismus in these studies was found to be 20% to 65%; however, the presence of strabismus was 31.81% in our study (2-6).

The presence of ET in Down syndrome cases with strabismus is between 18% and 70% (2-6). Yurdakul et al. (2) reported in a study that among 45 Down syndrome cases, 8 of 9 cases with strabismus had ET. Similarly, Jaeger et al., (4) in their study of 75 cases, detected 28 (37.3%) ET cases in 31 cases of strabismus. In our study, the presence of ET among strabismus cases in Down syndrome patients was determined to be 31.81%.

The standard bimedial rectus recession operation performed in ET cases has been demonstrated to lead to overcorrection when there is genetic neurological developmental delay (8-11). Pickering et al. (8) reported an improvement of more than 5.28 PD correction after bimedial rectus recession surgery performed in accordance with the standard table in patients with neurological development retardation. Habot-Wilner et al. (11) demonstrated a success rate of 37.5% in 11 patients who underwent standard bimedial rectus recession surgery.

Studies of only patients with Down syndrome have demonstrated a high success rate with standard bimedial rectus surgery (7, 12, 13). A surgical success rate of 85% was reported by Yahalom et al., (12) and it was 76% in the study of Perez et al. (13). In our study, the success rate was 80.90%. Yahalom et al. (12) reported undercorrection in 2 patients of 14 who underwent standard surgery, and Perez et al. reported 2 undercorrections and 2 overcorrections in 17 patients (13). In our study, undercorrection was detected in 2 Down syndrome cases out of 21, and no overcorrection was observed in that group.

In their controlled studies, Motley et al. (7) and Perez et al. (13) did not find any differences between Down syndrome cases and cases with normal neurological development in terms of postoperative deviation measures after standard bimedial rectus recession surgery. Motley et al. (7) demonstrated a difference of less than 1 PD between the 2 groups in terms of postoperative deviation measures. Perez et al. (13) did not detect any significant differences in terms of preoperative deviation measurements, the applied surgical recession doses, or surgical success between the 2 groups. In this study, preoperative and postoperative deviation measurements have not been assessed separately as near and distance deviation. However, both near and distance prism measures of all cases were assessed preoperatively and postoperatively. In our study, there was no significant difference between the 2 groups preoperatively and postoperatively in terms of near and distance measures. Furthermore, no significant differences were found in terms of surgical recession dose.

We determined that in Down syndrome cases with ET, bimedial rectus recession surgery in accordance with the standard table was successful and that undercorrection and overcorrection rates were similar to those of cases without a neurological developmental disorder.

Disclosures

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship Contributions: Involved in design and conduct of the study (OBO, AI, BG, AD); preparation and review of the study (EDA, SYO, CO); data collection (OBO, AI, EDA, BG); and statistical analysis (OBO, SYO, CO).

References