Objective: To explore the mechanisms underlying the hypertropia associated with unilateral coronal synostosis.

Methods: In 13 patients with unilateral coronal synostosis, we measured gaze-dependent binocular alignment before and after strabismus surgery, assessed the superior rectus muscle (SRM) pulley using computed tomography, and simulated posterior displacement of the trochlea and superolateral displacement of the SRM pulley.

Results: All the patients had an ipsilateral hypertropia in primary gaze (3-30 diopters) that increased in contralateral gaze and decreased in ipsilateral gaze and that simulated an inferior oblique muscle overaction. Strabismus surgery fully or partially corrected the hypertropia in only 7 of 11 patients. High-resolution computed tomography demonstrated that the pulley of the SRM relative to the inferior rectus muscle was translated 0 to 11.0 mm laterally and up to 2.7 mm vertically. Lateral translation (up to 10 mm) alone or combined with vertical translation (up to 5 mm) of the SRM pulley in the simulated model produced a hypertropia with lateral incomitance. Posterior translation (15 mm) of the trochlea did not induce a significant hypertropia.

Conclusion: Superolateral translation of the SRM pulley creates an imbalance of muscle pulling forces that better accounts for the hypertropia than posterior displacement of the trochlea.

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Hypertropia has a reported prevalence of up to 82% in unilateral coronal synostosis. The hypertropia in primary gaze is ipsilateral to the affected suture, increases in contralateral gaze (adduction of the hypertropic eye), and decreases in ipsilateral gaze (abduction of the hypertropic eye). This pattern of vertical incomitance in lateral gaze is consistent with overaction of the ipsilateral inferior oblique muscle. Apparent overaction of the ipsilateral inferior oblique muscle is attributed to underaction of the antagonist superior oblique muscle. Posterior displacement of the trochlea due to premature closure of the coronal suture and foreshortening of the orbital roof is postulated to weaken the superior oblique muscle relative to the inferior oblique muscle. Therefore, the hypertropia is treated surgically similar to a superior oblique muscle palsy by weakening the ipsilateral inferior oblique muscle alone or combined with weakening of the synergistic inferior rectus muscle (IRM) of the fellow eye.

To further investigate the mechanisms that underlie the hypertropia, we measured gaze-dependent binocular alignment before and after strabismus surgery and delineated the position of the superior rectus muscle (SRM) and IRM pulleys using high-resolution computed tomography (CT). On the basis of these measures, we propose that superolateral translation of the SRM pulley underlies the hypertropia. To test the strength of the trochlear vs the SRM pulley hypothesis, we simulated posterior translation of the trochlea and lateral translation alone or combined with vertical translation of the SRM pulley in a biomechanical model of static binocular eye alignment. We then compared the predicted outcomes with our results.

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Methods

We retrospectively studied 13 children with unilateral coronal synostosis and ipsilateral hypertropia after institutional review board approval was granted from the Children’s Hospital Regional Medical Center and the University of Washington Medical Center. Two of these children also had frontonasal dysplasia. A pediatrician and plastic surgeon specializing in craniofacial disorders evaluated all the patients. Unilateral coronal synostosis was sus-
pected on the basis of the following clinical features: (1) recession and elevation of the ipsilateral superior orbital rim, (2) elevation of the ipsilateral eyebrow and eyelid with contralateral ptosis, (3) displacement of the ipsilateral ear anteriorly, (4) palpable ridging of the ipsilateral coronal suture, and (5) ipsilateral cant of the nose and mouth.\textsuperscript{6,7} In all cases, the craniofacial diagnosis was confirmed by CT documentation of premature closure of 1 coronal suture and upward tilt of the superolateral orbital roof. Children with deformational changes of the face and skull that accompany lying with the head in 1 position (deformational plagiocephaly)\textsuperscript{6,7} or a congenital superior oblique muscle palsy (crouniosclerosis)\textsuperscript{8,9} were excluded.

All the patients underwent detailed eye examinations with emphasis on the assessment of binocular eye alignment in primary gaze and, whenever possible, at eccentricities of 15° up, down, right, and left and with head tilt right and left. Deviations of alignment were quantified using the prism cover test or the Krimsky test depending on patient cooperation. Stereopsis was measured in older patients using the Titmus test. The initial eye examination was performed before craniofacial surgery in 5 children and after craniofacial surgery in the remaining 8 children. At surgery, patients underwent a craniotomy to open the fused suture and frontal advancement to reshape the skull.

Eleven of 13 patients underwent strabismus surgery to correct the hypertropia in primary or contralateral gaze. Surgery consisted of an ipsilateral or bilateral inferior oblique muscle recession alone (4 patients) or combined with a contralateral IRM recession (5 patients) or ipsilateral SRM recession alone (1 patient) or combined with a contralateral IRM recession (1 patient).

To model static eye alignment we used the Orbit 1.8 computer program (Eidactics, San Francisco, Calif).\textsuperscript{10,11} This program takes into account the passive and active forces exercised by each of the extraocular muscles along with the biomechanical properties of the ocular motor plant (orbit, muscle, connective tissues, and muscle pulleys). Eye position in the cardinal directions of gaze is depicted on a Hess plot in Fick coordinates. We compared expected and observed binocular eye alignment predicted by the model after simulating up to 5-mm posterior displacement of the trochlea in 1-mm increments with normal innervation and lateral translation of the SRM pulley up to 10 mm alone or combined with vertical translation up to 5 mm in 1-mm increments. These numerical ranges were selected because they include the full spectrum of translational displacements observed in children with unilateral coronal synostosis or deformational plagiocephaly.

All 13 patients underwent CT of the head and orbits; 8 underwent high-resolution CT using a 2-dimensional scanner (model CBTB-016A; Toshiba Corp, Tokyo, Japan). Patients' heads were stabilized using a foam cushion. Transaxial images of the head and maxillofacial skull were helically acquired using 2:1 pitch. Continuous axial images, 0.5 to 1.0 mm thick, were obtained using a 512×512 matrix covering a 24- by 24-cm square, giving a pixel resolution of 469 pm. Digital CT images of the orbit were exported to a Vitreus 2 workstation (Vital Images Inc, Minnetonka, Minn), where images were normalized in the horizontal, vertical, and frontal planes. Then, serial images, 0.5 to 1.0 mm thick, were batched and recorded onto a compact disc in a DICOM (digital imaging and communications in medicine) file format. The image files were transferred to a Macintosh workstation, where they were analyzed quantitatively using open-source medical imaging software (OsiriX [http://homepage.mac.com/rossetantoine/osiriX/index2.html]).\textsuperscript{12}

Computed tomographic images were initially normalized in the sagittal plane by rotating the parasagittal images until the line connecting the inferior orbital rim with the external auditory canal was aligned with the scanner-defined horizontal meridian. To normalize for rotation in the frontal plane, images were translated/rotated until a line connecting the interhemispheric fissure and crista galli was aligned with the scanner-defined vertical meridian. To normalize for rotation in the horizontal plane, the midpoint of each optic canal was in spatial registration in the same coronal image. This coordinate system allows for relative measurements only because no origin is provided.

The relative horizontal positions of the SRM and IRM were determined from coronal images normalized in the craniooptic system described previously. Measurements of the SRM pulley were taken at a mean±SD of 2±0.5 mm anterior to the back of the globe along the anteroposterior axis of the image sections. We selected this position because it corresponds to the reported normal location of the SRM pulley posterior to which the muscle path is fixed. Demer et al\textsuperscript{13} showed that the SRM pulley inflection is, on average, 7 mm posterior to the globe center in normal eyes (total axial length, 24.5 mm) by magnetic resonance imaging criteria and extends for 14 mm in the anteroposterior dimension by histologic examination. We also selected this coronal section to avoid measurements along the anterior muscle path, where position is gaze dependent\textsuperscript{14} and the muscle is in apposition with the globe and is less clearly delineated by means of CT. To quantify lateral translation of the SRM pulley, we constructed a right triangle formed by a line connecting the centroids of the SRM and IRM and a scanner-defined horizontal reference aligned on the IRM centroid. The horizontal offset is equal to the length of the “opposite” side of this triangle. To quantify vertical translation of the SRM pulley, we constructed a right triangle formed by lines connecting the centroids of the SRM muscles and a scanner-defined horizontal reference aligned on the SRM centroid of the opposite orbit. The vertical offset is equal to the length of the “opposite” side of this triangle. We assumed that the positions of the horizontal and vertical rectus pulleys in the opposite orbit were normal. Rectus muscle centroids were determined using an image analysis program in OsiriX.

Between August 1, 1991, and December 31, 2004, 13 patients with hypertropia due to unilateral coronal synostosis were evaluated in the Pediatric Ophthalmology clinic at Children's Hospital and Regional Medical Center. The Table provides relevant clinical data, including age at strabismus surgery, eye with diagnosis, eye alignment in primary and lateral gaze, and strabismus surgery performed. The defining characteristic was an ipsilateral hypertropia in primary gaze that increased in contralateral gaze and decreased in ipsilateral gaze. The mean age at strabismus surgery was 2.9 years. All but 1 patient underwent craniofacial surgery, including cranioplasty with frontal orbital advancement. Eight (62%) of 13 patients had a compensatory head turn, and 2 (15%) had a head tilt. Seven (88%) of 8 patients who tolerated testing had at most 800 seconds of arc of stereopsis; patient 6, with a 3-diopeter (D) hyperphoria/hypertropia, had 40 seconds of arc of stereopsis.

Figure 1 shows that all but 1 of the patients with unilateral coronal synostosis had an ipsilateral hypertropia in primary gaze ranging from 3 to 30 D. Three patients with a small hypertropia (<4 D) and the 1 patient without a hypertropia in primary gaze had sufficient disparity-driven vergences to maintain eye alignment under bin-
ocular viewing. The mean ± SD amount of hypertropia was 13.1 ± 8.6 D in primary gaze, 22.1 ± 11.0 D in contralateral gaze, and 3.9 ± 4.8 D in ipsilateral gaze.

Figure 2 compares the preoperative and postoperative amounts of hypertropia in primary gaze for 11 patients who underwent strabismus surgery. Surgery fully or partially corrected the hypertropia in 7 patients, of whom only 4 had orthotropia in primary gaze. One patient had no improvement. One patient with normal binocular alignment in primary gaze demonstrated a hypertropia after unilateral inferior oblique muscle recession. Surgery resulted in a hypertropia of the opposite eye in 2 patients.

One explanation for the limited surgical success is that posterior translation of the trochlea does not account for the hypertropia. Computer simulation of binocular eye alignment after a 15-mm unilateral posterior displacement of the left trochlea but without innervational changes is shown in Figure 3. The Orbit 1.8 model predicts vertical deviations of 1.7, 2.8, and 0.4 D in primary, contralateral, and ipsilateral gaze, respectively.

We then proposed that lateral displacement of the SRM pulley could be the underlying pathologic mechanism. Representative coronal CT images from 2 patients with unilateral coronal synostosis are shown in Figure 4A and B. The patient in Figure 4A had a 3-D right hypertropia in primary gaze. Although the ipsilateral orbital roof is translated superolaterally, the CT scan shows no horizontal offset of the vertical rectus muscles. The patient in Figure 4B had a 25-D right hypertropia in primary gaze. Lateral translation of the right SRM pulley is obvious, and there is a

**Table. Clinical Features of 13 Patients With Unilateral Coronal Synostosis**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age at Strabismus Surgery, y</th>
<th>Eye With Diagnosis</th>
<th>Eye Alignment</th>
<th>Strabismus Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2.9</td>
<td>Left</td>
<td>14 D LHT, 10 D ET</td>
<td>18 D LHT, 12 D ET</td>
</tr>
<tr>
<td>2*</td>
<td>4.3</td>
<td>Left</td>
<td>20 D LHT</td>
<td>&gt;30 D LHT</td>
</tr>
<tr>
<td>3</td>
<td>4.3</td>
<td>Right</td>
<td>15 D RHT</td>
<td>ND</td>
</tr>
<tr>
<td>4</td>
<td>2.8</td>
<td>Right</td>
<td>20 D RHT</td>
<td>&gt;25 D RHT</td>
</tr>
<tr>
<td>5</td>
<td>3.3</td>
<td>Right</td>
<td>12 D RHT</td>
<td>14 D RHT</td>
</tr>
<tr>
<td>6</td>
<td>11.0</td>
<td>Right</td>
<td>3 D RHT</td>
<td>10 D RHT</td>
</tr>
<tr>
<td>7</td>
<td>3.7</td>
<td>Right</td>
<td>25 D RHT</td>
<td>&gt;30 D RHT</td>
</tr>
<tr>
<td>8</td>
<td>4.7</td>
<td>Right</td>
<td>0-30 D RHT</td>
<td>40 D RHT</td>
</tr>
<tr>
<td>9</td>
<td>0.8</td>
<td>Right</td>
<td>20 D ET</td>
<td>20 D ET</td>
</tr>
<tr>
<td>10*</td>
<td>2.1</td>
<td>Right</td>
<td>0-3 D LHT</td>
<td>12 D LHT</td>
</tr>
<tr>
<td>11</td>
<td>2.3</td>
<td>Left</td>
<td>20 D ET</td>
<td>16 D LHT</td>
</tr>
<tr>
<td>12</td>
<td>1.9</td>
<td>Left</td>
<td>8 D LHT</td>
<td>16 D LHT</td>
</tr>
<tr>
<td>13</td>
<td>2.9</td>
<td>Left</td>
<td>0-3 D LHT</td>
<td>12 D LHT</td>
</tr>
</tbody>
</table>

Abbreviations: D, diopters; ET, esotropia; IOM, inferior oblique muscle; LHT, left hypertropia; LIO, left inferior oblique muscle; LIRM, left inferior rectus muscle; LMRM, left medial rectus muscle; LRM, lateral rectus muscle; MRM, medial rectus muscles; ND, no data; Ortho, orthotropia; RIO, right inferior oblique muscle; RHT, right hypertropia; RIRM, right inferior rectus muscle; RSRM, right superior rectus muscle.

*These patients had the concurrent presence of frontonasal dysplasia.
6.7-mm horizontal offset of the vertical rectus muscles. The superior oblique muscle bellies in both patients are similar in cross-sectional size, and the horizontal rectus pulleys are normally aligned. By comparison, Figure 4C is a representative CT scan from 1 of 2 patients with unilateral coronal synostosis combined with frontonasal dysplasia and a 30-D right hypertropia. The scan reveals that the SRM pulley is translated 11.0 mm laterally and 2.7 mm vertically. The ipsilateral superior oblique muscle is underdeveloped, and the ipsilateral lateral rectus muscle is translated inferiorly.

Computer simulation of the binocular alignment effects of SRM pulley translation reproduces some of our clinical observations. Figure 5 shows the horizontal and vertical eye position after a simulated 10-mm lateral translation of the left SRM pulley predicted by Orbit 1.8. The model predicted the presence of a 6° (12 D) hypertropia in primary gaze that increased to 10° (20 D) in contralateral gaze and decreased in ipsilateral gaze. However, the Orbit 1.8 model also predicted that lateral displacement of the SRM pulley would produce a Y-pattern exotropia. We did not observe this strabismus pattern in any patients.

Figure 6A depicts the relationship between 1-mm incremental displacements of the SRM pulley and the amount of hypertropia predicted by Orbit 1.8. Overall, the model predicts a nonlinear increase in the amount of hypertropia with increasing lateral displacement of the SRM pulley up to 10 mm. Adding vertical translations of 1 to 5 mm to lateral translations of the SRM pulley linearly increases the amount of hypertropia predicted by the model. Vertical translations of the SRM pulley of 1 to 5 mm induce more than 2-fold–higher amounts of hypertropia in primary gaze than lateral translations of the same magnitude. Figure 6B demonstrates that vertical translations of the SRM pulley, unlike horizontal translations, are not associated with differential amounts of hypertropia in lateral gaze.

Translations of the SRM pulley for 8 patients with high-resolution axial CT data are compared with the amounts
of observed hypertropia in Figure 7. The distributions for displacement of the SRM pulley varied from 0 to 11.0 mm for lateral translations and from 0 to 2.7 mm for vertical translations. Lateral displacements of the SRM pulley of less than 4 mm were associated with little or no hypertropia in primary gaze. In comparison, lateral displacements of 4 mm or more were associated with an ipsilateral hypertropia of 20 to 30 D in primary gaze. The amounts of hypertropia predicted by the model for lateral translations of the SRM pulley up to 10 mm alone or combined with vertical translations of 5 mm are represented by the curvilinear tracings. The amount of hypertropia observed clinically was consistently more than the amount predicted by the model.

The present study investigated the mechanisms that underlie the hypertropia associated with unilateral coronal synostosis using quantitative measures of gaze-dependent binocular alignment and extraocular rectus muscle pulley locations. The hypertropia associated with unilateral coronal synostosis has been attributed to posterior displacement and weakening of the superior oblique muscle relative to the inferior oblique muscle.1-3 In support of this finding, this study and previous studies have shown that the hypertropia in primary gaze increases in contralateral gaze and decreases in ipsilateral
gaze and is associated with apparent overaction of the ipsilateral inferior oblique muscle. However, this theory is not supported by the following observations. First, hypertropia does not occur with deformational plagiocephaly in which the orbital rim can be displaced posteriorly by similar amounts. Second, simulated posterior displacement of the superior oblique muscle up to 15 mm without changes in innervational inputs in a computer model of static eye alignment did not produce more than 2 D of hypertropia in primary gaze. Substantial increases in innervational inputs were required to induce even modest amounts of hypertropia. Third, unlike superior oblique muscle palsies, for which surgery is highly successful, the same surgical strategy did not consistently produce binocular alignment.

An alternative possibility is that the hypertropia is a consequence of lateral translation of the orbital roof rather than posterior displacement of the orbital rim. Previous imaging studies have demonstrated that there is oblique elongation of the superolateral segment of the orbital roof due to superolateral shift in the angulation of the sphenoid bone, the so-called harlequin-shaped deformity. The configuration of the remaining orbital walls, particularly the orbital floor, remains relatively normal. Such alterations in orbital anatomy could be associated with shifts in the paths of the extraocular muscles. High-resolution magnetic resonance imaging of healthy individuals demonstrates that near the globe equator there are inflections in the muscle path where the muscle is mechanically coupled by connective tissue sleeves to the orbital periosteum. Therefore, we postulated that the SRM pulley, like the orbital roof, was translated superolaterally.

High-resolution orbital CT images (Figure 4) showed that unilateral coronal synostosis is associated with lateral or superolateral translations of the SRM pulley. The amount of SRM pulley translation was consistently higher in patients with increased amounts of hypertropia. The Orbit 1.8 model predicted a nonlinear increase in the amount of hypertropia as the SRM pulley was translated laterally up to 10 mm. The Orbit 1.8 model also predicted that this lateral translational effect was amplified by vertical translation of the SRM pulley. Computer simulation of lateral translation, but not vertical translation, of the SRM pulley also predicted different amounts of hypertropia in lateral gaze. Collectively, the orbital CT image data and model predictions of eye alignment suggest that lateral translation of the SRM pulley provides a better account for the hypertropia associated with unilateral coronal synostosis than posterior displacement of the trochlea.

In biomechanical terms, the hypertropia represents an imbalance in the static pulling force of the SRM relative to the IRM. Total pulling force of an extraocular muscle can be represented by the sum of passive and active forces exerted on its rotation vector. The observed hypertropia could be due to an imbalance of the passive forces between opposing vertical recti resulting from asymmetrical increases in muscle tension. Superolateral translation of the pulley stretches the SRM relative to the IRM. Because there is a hyperbolic relationship between muscle length and tension, muscle stretch could lead to nonlinear increases in passive tension of the SRM relative to the IRM. Assuming that innervational inputs to both SRMs are equal across the horizontal plane, the resulting imbalance of static forces could lead to a hypertropia in primary gaze that increases nonlinearly with lengthening of the muscle path.

Lateral translation of the SRM can also produce a vertical imbalance of static muscle forces due to the loss of coplanarity in the muscle pulling directions. The pulling direction of each rectus extraocular muscle is defined by its anterior path, which functionally originates at the muscle pulley and terminates at the tendinous muscle insertion. When orbital anatomy is normal, the anterior muscle paths of the SRM and IRM course 23° oblique to the sagittal plane for globe rotation, and their rotation planes are coplanar. In this configuration, the muscle forces generated by paired vertical rectus muscles are balanced. Lateral offset of the SRM pulley creates an angular misalignment in the pulling direction of the SRM and IRM relative to the vertical rotation plane of the globe. In this configuration, the pulling force of the SRM is in closer alignment with the vertical rotation plane of the globe and is increased by the tangent of the angular misalignment. For example, a lateral offset of the SRM pulley of 23° would create a force imbalance of approximately 42% (tangent of 23°) in the vertical direction. The coplanar rotation mechanism can also contribute to vertical incomitance in lateral gaze owing to the shifting relationship between the relative pulling forces of opposing vertical rectus muscles and the vertical rotation axis of the globe.

Although translation of the SRM pulley correctly identified the laterality of the hypertropia, the magnitude of the observed hypertropia was consistently larger than the predicted hypertropia. Therefore, lateral translation alone does not fully account for the hypertropia. We propose...
that lateral translation induces an upward directional bias in the affected eye but that the magnitude of that effect is amplified by vertical translation of the SRM pulley, a breakdown of disparity-driven vergences due to poor binocularity, or changes in the length of vertically acting extraocular muscles. Furthermore, in the subset of patients with concurrent frontonasal dysplasia, a superimposed weakness of the superior oblique muscle likely contributes to the hypertropia. We agree that in patients with such complex craniofacial disorders or generalized craniosynostoses, the underlying pathologic mechanism likely involves complex displacements of multiple pulleys and additional pathologic abnormalities. In addition, the Orbit 1.8 model also predicted a Y-pattern exotropia that was not observed in these patients. However, in young children, assessment of eye alignment is difficult at vertical eccentricities of 30°, where the model predicts significant exotropia.

The 2 patients with concurrent frontonasal dysplasia deserve special mention because the pathologic process extended beyond translation of the SRM pulley. Both patients demonstrated translation of at least 1 additional pulley and superior oblique muscle deficiency. We conclude that in patients with such complex craniofacial disorders or more generalized craniosynostoses, the mechanisms underlying the strabismus likely involve complex displacements of multiple pulleys and additional pathologic abnormalities.

Poor surgical outcomes are likely due to failure to address the biomechanical consequences of superolateral translation of the SRM pulley. Before surgery, patients with unilateral coronal synostosis should have orbital imaging studies to assess the lateral displacement of the SRM pulley and to document laterality. On the basis of these data, we recommend weakening of the ipsilateral SRM alone or combined with a contralateral IRM recession. One alternative approach would be to selectively alter the pulling direction of the SRM or the opposing IRM by sutureal fixation of the muscle to the globe posterior to the equator (Faden procedure), thereby repositioning the pulley.19 The subset of patients with an attenuated superior oblique tendon may benefit from weakening of the ipsilateral inferior oblique muscle.

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