

**Case 3**

*M.R.G., male, 15 years old*

February 9, 1978:

This patient turned the head 17 degrees toward the left and lowered the chin to fixate on near and distant objects. The patient exhibited pendular nystagmus in primary position, becoming saccadic in both lateroversions, with fast phase toward the respective version. It was accentuated in infraversion and almost disappeared in supradextroversion. Binocular visual acuity at the blocking position was 20/25; refraction with cycloplegia was +0.5 D OU.

February 13, 1978:

An operation was performed that consisted of 8-mm superior rectus muscle recession OU; 7-mm lateral rectus muscle recession OD; and 5.5-mm medial rectus muscle recession OS.

February 27, 1978:

The horizontal component of the torticollis had disappeared; a slight chin lowering persisted. Binocular visual acuity in primary position was 20/25.

muscles, according to Moacyr Alvaro's technique, may solve the problem.

In cases in which the vertical component is large, vertical transposition of the horizontal rectus muscles is not sufficient for correction. Operating on the vertical rectus muscles is necessary. We prefer to operate only on the vertical rectus muscles, because surgery on the oblique muscles, especially the superior, may cause hypertropia or cyclotropia (or both) in primary position. This strategy coincides with the scheme of von Berger and Dell'Aquila,<sup>42</sup> who described a patient who had horizontal torticollis and a marked chin elevation; they performed an inferior rectus muscle recession and, in a second operation, recessions of the horizontal rectus muscles on the opposite side of the torticollis. We believe such a case can be treated by one surgical procedure, as in Case 3.

In some cases, the head tilt is only vertical or it predominates. Using the same principle posited by Kestenbaum, Pierce<sup>43</sup> presented two cases of nystagmus with the blocking position in infraversion. In one, he performed a 5-mm recession of both inferior rectus muscles and, 14 days later, added a tenectomy of the superior oblique muscles. The patient developed a combined vertical and divergent heterotropia, correction of which required a third

operation. In the second patient, he performed a 4-mm recession of both inferior rectus muscles and a 4-mm recession of the superior oblique muscles. In both cases, he obtained good results.

We prefer to operate on only the vertical rectus muscles. Focosi and Gussinati<sup>44</sup> described a case of nystagmus with the blocking position in supraversion; they performed an 8-mm resection of both inferior rectus muscles and, 8 days later, a 5-mm recession of both superior rectus muscles.

This situation, in which torticollis is only vertical, is fairly rare. We agree with Parks<sup>39</sup> who, for vertical torticollis of less than 25 degrees, performs only a recession of the agonist vertical rectus muscles. If its magnitude exceeds 25 degrees, he performs a recession-resection of the four vertical rectus muscles. He stated that for a horizontal component, he operates on three rectus muscles in each eye. We disagree with this last approach. In such a circumstance, we prefer to perform two operations to avoid the risk of anterior segment ischemia.

Some patients show a torsional component of their torticollis, tilting the head toward one shoulder (see Figure 10-11). Parks<sup>39</sup> believed that this head position was meant to produce a vestibular response, which reduces the nystagmus; for this reason, he does not operate for the torsional component.

In 1964, Harada and Ito<sup>45</sup> proposed a surgical technique to eliminate the torticollis caused by torsional deviation of the eyes. Only recently have such techniques been popularized, thanks mainly to the publications of De Decker.<sup>46</sup> Such techniques can minimize the severity of this problem.

The techniques are based on modern studies of the differential action of the fibers of the oblique muscles. The superior oblique muscle fibers responsible for the torsional action are the anterior fibers; therefore, they must be operated on. They are incycloductors. For intorsion, they must be weakened; for extorsion, they must be shortened or advanced. In the former case, a tenectomy can be performed on the anterior two-thirds of the superior oblique tendon, or they can be recessed. For extorsion, we use Fell's modification<sup>47</sup> of the Harada and Ito technique. We separate one-half of the anterior fibers longitudinally to approximately 1 cm from the scleral insertion. Then we apply two marginal sutures to the fibers at approximately 8 mm from the scleral insertion. Finally, we affix these sutures to the sclera, the anterior suture 4 mm laterally to the lateral end of the superior rectus muscle insertion and the posterior suture 3 or 4 mm behind it. With this procedure, these fibers are advanced and transposed anteriorly, increasing their torsional action significantly.

In nystagmus, generally torsion is very marked, so operating on only the superior oblique muscle is not sufficient. Increasing or reducing the torsional action of the inferior oblique muscle is necessary. The functional difference between the different fibers of this muscle is not as marked as in the superior oblique muscles because of the inferior oblique muscle's very long arc of contact (that of the superior oblique muscle being very short) and its very short tendon. The operations must be more radical in such cases. To weaken its extorsional action, we perform a myectomy of the anterior two-thirds of its fibers at 1 cm from the scleral insertion and an approximately 5-mm recession of the posterior fibers. We take care not to transpose them anteriorly. We must be careful because the reinsertion area is very near the inferolateral vortex vein. To increase the extorsional action of the inferior oblique muscles, we perform an approximately 5- or 6-mm myectomy of all its fibers and reinsert it more anteriorly. The anterior end of the muscle is inserted approximately 3 mm posterior to the inferior end of the lateral rectus muscle insertion, and the posterior end is inserted approximately 6 mm behind, near the inferior margin of the lateral rectus muscle.

In nystagmus with torsional torticollis, one of the eyes obviously is intorted (the one on the same side as the head tilt), and the other is extorted. Therefore, all four oblique muscles are operated on. The anterior fibers of the superior oblique muscle are weakened, the inferior oblique muscle in the eye on the side of the head tilt is moved anteriorly and resected, and the superior oblique anterior muscle fibers are advanced and moved anteriorly. This procedure is coupled with a myectomy of the anterior fibers and recession of the posterior fibers of the inferior oblique muscle. This therapeutic method follows Kestenbaum's principle and has provided satisfactory results, although sometimes it is insufficient to correct the torticollis completely.

The foregoing procedure may produce in primary position a hypertropia that must be corrected by recession of the superior rectus muscle of the hypertropic eye (the one in which we did the extorsional operation). However, if the operation is performed carefully, such a negative outcome is fairly rare.

Some colleagues have reported good results with the Anderson operation (i.e., only recessing the agonist muscles).

Gradstein et al.<sup>48</sup> pointed out the danger of failing to diagnose periodic alternating nystagmus. If a Kestenbaum operation is performed on such patients, they will turn the head markedly to the opposite side after the operation. If only recessions are performed (Anderson's pro-

cedure), the unoperated muscles can be recessed, and the null zone can be recentered.

Zubcov et al.<sup>49</sup> proposed that the operation to correct torticollis should produce an exophoria and that the amount of postoperative exophoria is more important than is the type of operation. These notions are based on the theory that the exophoria will induce convergence. We do not have any experience with this procedure, but we believe that preoperative investigation is required to determine whether affected patients truly block the nystagmus in convergence (see Figure 10-10).

### Nystagmus and Strabismus

Frequently, patients are found to have both strabismus and nystagmus. The typical case is Ciancia's syndrome (early esotropia with bilateral abduction deficiency) described by Ciancia in 1962<sup>50</sup> and more recently called *blocking nystagmus syndrome* (*das nystagmus blockierung syndrom*) by Adelstein and Cüppers<sup>51</sup> and Cüppers.<sup>52</sup>

Correcting this type of esotropia is difficult. One of the proposed hypotheses in such cases is that the nystagmus is blocked in extreme convergence (see Chapter 3), but we do not accept this theory. For practical purposes, these patients actually block each eye in adduction.

The aforementioned characteristics do not necessarily fit every case. Some patients present with strabismus, with or without amblyopia, in which the fixating eye has a null zone that does not coincide with the primary position. This condition demands that such patients adopt a compensatory torticollis, which may be in adduction or abduction, and the deviation may be convergent or divergent (Figure 10-14). Surgery must aim at correcting both the strabismus and the torticollis.

The latter can be corrected only by operating on the fixating eye; sometimes, the procedure corrects both defects (e.g., a hypothetical case of esotropia in which the dominant eye blocks the nystagmus in adduction). A combined recession of the medial rectus muscle and resection of the lateral rectus muscle of this eye tends to solve both problems. The nystagmus correction may require a more extensive operation than that required to correct the esotropia. The nystagmus surgery results in a secondary exotropia, which should be corrected with a second procedure, operating on the deviated eye according to the size of the deviation; this procedure will not influence the nystagmus.

In summary, the fixating eye should be operated on with the goal of correcting the torticollis. A second surgical procedure can be performed to correct any eventual remaining strabismus. This surgery should be performed on the nonfixating eye.