Management of Congenital Fourth Cranial Nerve Palsy

William P. Madigan, MD; James D. Reynolds, MD; Mitchell Strominger, MD

Wagner: We’re going to discuss the management of congenital fourth cranial nerve palsy. A 2½-year-old boy presents with a history of persistent head tilt toward the left shoulder. A 14-mm recession of the right inferior oblique muscle was performed 7 or 8 months ago at a different institution. His fixation is central, steady, maintained with either eye and you find a right hypertropia of 8 prism diopters (PD), which increases to 15 PD in left gaze and 18 PD on head tilt toward the right shoulder. There is a significant underaction of the right superior oblique muscle and slight underaction of the previously operated on right inferior oblique muscle. Also noteworthy is that the hypertropia is largest in down gaze to the left. What are the surgical options for a case like this and what goes into your decision-making process?

Reynolds: It is difficult to get measurements on these young children. I like to base my initial surgical judgment on the size of the primary position deviation. A study I reported in the 1980s indicated that 15 PD seems to be the cut-off. If it’s below 15 you can get by with one muscle, but for 15 and above you really need two muscles. If this patient had a deviation of 15 PD, I would have initially weakened the right inferior oblique as the direct antagonist and I would have strengthened the superior oblique. I would have done a myotomy rather than a recession and I would have taken a small tuck in the superior oblique. Given the fact that this patient had inferior oblique surgery previously, I would move to the second muscle. No matter what the pattern is, I really like to tuck the superior oblique as my second surgery.

Madigan: I also use 15 PD as the cut-off point, with one muscle for less than 15 and two muscles for 15 and above. When a really young child presents with a large head tilt and a weak superior oblique muscle, you are going to consider doing the tuck as the first surgery. I try to avoid superior oblique surgery otherwise because a good tuck creates a Brown syndrome with the associated restriction. In these young patients with large head tilts, you must also be prepared, should you find an absent superior oblique tendon, to have an alternate plan. For this particular patient with the hypertropia worse in down and left gaze, I think the second surgery should be a recession of the contralateral (left) inferior rectus muscle.

Strominger: I agree that there seem to be two types of congenital fourth nerve palsy. For a child who presents with a significant head tilt and almost no superior oblique function, I would manage with a tuck primarily. This doesn’t occur often and most cases have some superior oblique function along with significant inferior oblique overaction. In these patients, I do an inferior oblique anterior transposition. I no longer recess the inferior oblique because I have found that some patients who I recessed 10 years ago are showing some inferior oblique overaction. In those cases I try to anteriorize the recessed inferior oblique to see how much more weakening I can get out of it.

In this case, it surprises me that the head tilt is that large with only 8 diopters (D) of right hy-
pertropia in primary position. So there may be associated torticollis. I might try to have the child wear some prisms in the office and place 8 D of base-down prism over the right eye to see if the head tilt improves. If it does, I would consider recessing the contralateral inferior rectus muscle.

**Madigan:** I like that idea. The only thing I would say with regard to the motility examination as it was described is there's actually underaction of the superior oblique. In my experience, most superior oblique palsies have grossly intact function on motility testing, so if you're seeing an underaction, that's a pretty profound palsy and may explain why a second surgery is required.

**Reynolds:** But that again highlights the difficulty of examining very young children. We know it's likely that we will miss some findings in a child that age. I like your point about operating on forgiving muscles and avoiding unforgiving muscles. The inferior oblique is clearly an extremely forgiving muscle. I would never think about going to any other muscle if there is inferior oblique overaction. Superior oblique muscles are not forgiving and you can cause more trouble than you started with.

**Strominger:** I've encountered adolescent patients who had both muscles operated on at the same time. They had an inferior oblique myectomy and a superior oblique tuck. Now the eye doesn't move around much and they have secondary overaction of the contralateral superior rectus muscle and huge upshoots. I've never reoperated by trying to undo someone else's tuck, but if they don't have superior oblique function and now have a primary hypotropia, you have to operate on an weaken the contralateral superior rectus muscle.

**Reynolds:** People get into trouble by making their tucks too big. I like to get a sense of when that muscle begins to resist. When I start to tuck and that superior oblique is pretty tight, I'm prepared to take my suture down if I have positive forced ductions. If I've got a Brown syndrome with a forced duction, then 6 mm is too big a tuck and I need to back off to 4 mm.

**Wagner:** I think that's a good description of a procedure on the superior oblique muscle. Have you experienced intraoperative laxity of the superior oblique in a case like this?

**Reynolds:** I have.

**Strominger:** I've gone in surgically and not found the superior oblique. If you scan cases that have severe superior oblique underaction, you might see the thread of the superior oblique. I'm not sure how you can tuck that.

**Reynolds:** I had a case where the superior oblique inserted on the underbelly of the superior rectus and didn't even insert on the globe. And so the superior oblique is the muscle that's most likely to be anomalous and can be really loose. I respect the superior oblique so much that even on a really loose one, I probably would not follow my own guidelines for doing a tuck because you might take a 15-mm tuck before it started to firm up. I think that's too big a tuck.

**Madigan:** I'd like to go back to when we were talking about doing an inferior oblique recession and a superior oblique tuck at the same time. I've never done it that way because I am always afraid of exactly what you're talking about. So as a second stage procedure I would go back to the tuck or do the tuck initially if I felt that was appropriate. But I usually do not do the two procedures together.

**Strominger:** Which procedures do you use then?

**Madigan:** I'd probably do the contralateral inferior rectus as primary. If, despite all my best efforts, I could not find the superior oblique, I would simply taking off the superior rectus and look back there.

**Wagner:** So in a case like that, what would be your option when you set out to correct some of this vertical deviation?

**Madigan:** I do the ipsilateral superior rectus. When I was Marshall Parks' fellow, we had discussions about whether there is such a thing as an absent superior oblique and Edward Wilson made it a project to try to find and document with photographs absent superior obliques. I don't think he ever presented anything conclusive.

**Reynolds:** They're definitely reported.

**Madigan:** New MRIs make it easier to document now, but this was in the 1980s when MRIs were not available or as sophisticated.

**Wagner:** I think most surgeons would agree that they would initially operate on the inferior oblique. We all have experience with Brown's syndrome, which I think you're going to see in cases that you tuck, even if you're extremely careful.

**Reynolds:** I really respect how easy it is to induce Brown's syndrome and how problematic a big tuck can be, so I do small tucks when I do them as final procedures.

**Madigan:** I find interesting this whole idea about the progression of the overaction of the contralateral superior oblique that
you were just talking about. As fellows, we were hoping to find a simple answer that we could publish, but the issue was not that clear-cut and we never published anything because you couldn’t reliably say whether you’re more likely to get a contralateral superior oblique overaction (really a tight ipsilateral superior rectus) when you have a longstanding superior oblique weakness or not.

Wagner: People talk about bilateral superior oblique palsy where you weaken the inferior oblique in one eye and then hypertropia develops in the opposite eye. Have you seen this? What do you think is going on in these cases?

Reynolds: I do believe that there are bilaterals that are so asymmetric that it’s difficult to see that. And again this is an age-related phenomenon. If you’re operating on a 15 year old, it’s easy to unmask them preoperatively. But it’s difficult to get a 2 year old to hold that maximum opposite gaze so you can find that little left hypertropia. And if you do see it for a moment, should you do two muscles on the right eye and one muscle on the left eye? Those are difficult decisions to make and sometimes it’s better to operate on the inferior oblique in the worst eye and see what happens. I always caution parents about unmasking more overaction on the contralateral side. Whenever I do oblique surgery, even if it’s primarily inferior oblique overaction, it’s very asymmetric and unusual but potentially there.

Strominger: I agree. I’ve seen my share of those cases and it’s hard to measure, especially if you have so much inferior oblique overaction, because these eyes become exotropic as they’re looking up and over and you can’t measure the hypertropia. I’ve also seen cases that I thought were bilateral congenital fourth nerve palsies and then 8 years later find an esotropia and realize that it was probably strabismic in nature, especially if they have good superior oblique function to begin with. I worry that there may be something intracranial going on, but so far I’ve never discovered any problem.

Reynolds: That brings up a good question. I image very selectively. How often do you image patients in this situation?

Madigan: I only image if there’s some other related process going on that makes me suspicious that there’s something else beyond the standard congenital superior oblique palsy present.

Strominger: I image more, partly because of my adult patient population. I have discovered at least seven schwannomas and a couple of cysts. These are all adult patients who had the head tilt when they were younger and it became worse as they got older.

Reynolds: So you went back and looked at some old photographs on these adults and they had a tilt 10 years ago and now you’re finding a schwannoma?

Strominger: Yes. So I am trying to figure out why it is getting worse. I’m not imaging children, only adults. Schwannomas are slow growing but they were probably there when the patients were younger, so the question is what findings should have been noted when they were younger.

Madigan: How are you treating the schwannoma after diagnosing it?

Strominger: We’re doing direct radiation treatment to try to prevent them from progressing.

Reynolds: Have you seen the fourth nerve palsy resolve?

Strominger: No. I’ve seen it get better in one case. There’s also a report of two cases in the literature in which one resolved and one got better.

Reynolds: It’s a little disconcerting to think that you could look at photographs that are 10 years old and see that this is an acquired palsy.

Wagner: Thank you all for participating.

This Eye to Eye session was conducted on Thursday, April 3, 2013, during the annual meeting of the American Association for Pediatric Ophthalmology & Strabismus.

Drs. Madigan, Reynolds, and Strominger have no financial or proprietary interest in the materials presented herein. Dr. Wagner is on the speaker’s bureau of Alcon Laboratories.

doi: 10.3928/01913913-20131018-03