

PEDIATRICS

New Research Sheds Light on Intermittent Exotropia

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INTERVIEWING MICHAEL P. CLARKE, FRCOPHTH, SEAN P. DONAHUE, MD, PHD,
AND K. DAVID EPLEY, MD

Surgical correction of intermittent exotropia is a bread-and-butter procedure for many pediatric ophthalmologists. But a recent study in the *British Journal of Ophthalmology* (BJO)¹ and an accompanying editorial² call into question the notion that intermittent exotropia is straightforward or fully understood.

These articles challenge some of the dogma that has guided the management of children with this condition in the past. In particular, researchers are reconsidering the indications for treatment; and, when surgery is needed, what procedure is most appropriate.

Bewildered at the Squint Club

The BJO editorial, titled “The Many Enigmas of Intermittent Exotropia,”² recounts a meeting of the Squint Club (an annual gathering of strabismus experts) several years earlier, at which Arthur Rosenbaum (now deceased) presented a paper contending that intermittent exotropia was the most perplexing and difficult form of strabismus. According to the editorial, Dr. Rosenbaum’s choice of topic and conclusions surprised many Squint Club members because intermittent exotropia had traditionally been viewed as an uncomplicated and easily treatable form of strabismus.

But Dr. Rosenbaum argued that a number of issues still needed to be explored. For example, he noted that the natural history of the disorder had not been adequately defined and that it was

not clear whether the condition does in fact deteriorate over time, as has been widely believed. Further, the clinical indications for surgery remained unclear; and the long-term outcomes of surgical treatment were sometimes disappointing, with high rates of persistent or recurrent exodeviations and consecutive esodeviations.

What the New Research Shows

The BJO study, conducted by Buck et al. in the United Kingdom,¹ supports Dr. Rosenbaum’s premise that intermittent exotropia is not fully understood. This study followed an observational cohort of 460 children with intermittent exotropia who were younger than 11 years old. Each child was examined at one of 26 hospital ophthalmology clinics between May 2005 and December 2006. During a two-year period, the study investigators compiled data on each child’s angle of strabismus, near stereoacuity, visual acuity, control of intermittent distance exotropia (measured with the Newcastle control score [NCS]), and type of treatment performed. The main outcome measures were changes in clinical status two years after enrollment or six months after surgery.

Surprising results. Outcome data were available for 371 of the 460 children: 195 (53 percent) did not undergo treatment; 63 (17 percent) were treated only for reduced visual acuity (pure refractive error and amblyopia); 50 (13 percent) were treated nonsurgically to improve control (spectacles, occlu-

Intermittent Exotropia



(1A) Although the eyes of this boy are initially aligned, his right eye starts to drift outward (1B) as he looks at distance.

sion, prisms, or exercises), and 63 (17 percent) underwent surgery. Two children (0.5 percent) developed constant exotropia. Only the surgically treated patients showed clinically significant improvements in angle or NCS. However, five (8 percent) of the surgically treated children required a second operation within six months because of initial overcorrection. At the six-month follow-up, an additional 13 (21 percent) of the surgically treated chil-

dren had manifested overcorrection.

Study investigator Michael P. Clarke, FRCOphth, a consultant pediatric ophthalmologist and head of ophthalmology at Newcastle upon Tyne Hospitals in the United Kingdom, said he was surprised by the relatively low rate of cure found in the study as well as the high rate of overcorrection. “I am now probably less upbeat with parents about the results of surgery than I was previously. And there is an argument to be made for waiting until children are older on the basis that some of the variability in outcome may relate to difficulties in getting precise measurements in small children. That leaves you waiting until they are older before you do the surgery.” Dr. Clarke believes that, in general, pediatric ophthalmologists were more optimistic about being able to successfully correct intermittent exotropia 10 or 20 years ago than they are today.

When Is Surgery Appropriate?

K. David Epley, MD, a pediatric ophthalmologist in private practice in Kirkland, Wash., believes that the *BJO* study provides some important new information. “Only 0.5 percent of patients in the study decompensated to the point that their eyes were constantly drifting,” he said. “Most of the kids who underwent surgery were losing vision or losing depth perception, but their eyes were not drifting out all the time.”

Dr. Epley added that the proportion of children in the study who had surgery was surprisingly low. “The surgery rate of 17 percent was lower than what most of us think of as the number of kids who need surgery. When it comes to taking kids to surgery, there is a huge variation in how we all practice. Some of us advocate early surgery, whereas others advocate waiting as long as possible. But we all believe that about a third of these kids will end up having surgery at some point, and some people quote a higher proportion than that.”

Dr. Epley’s approach is to try conservative measures first, provided that the child is not losing vision, can still

see normally, and has not lost stereopsis. “If these are normal, we will recommend exercises or do some patching or even prescribe glasses to exercise the convergence system,” he said.

“A lot of kids go through childhood having intermittent sessions of these conservative measures and don’t require further treatment. But some kids will decompensate and lose vision or depth perception, and that is my cue to move on to something more interventional. If a child is losing ground and can’t maintain or recover that ground through a conservative approach, then we will move on to surgery.”

Seeking consensus.

Sean P. Donahue, MD, PhD, professor and vice chairman of ophthalmology at Vanderbilt University School of Medicine, addressed the many unknowns inherent in treating young patients with intermittent exotropia. “Consensus is lacking with regard to when to intervene surgically and regarding the risks of performing surgery versus letting the natural history play out. This is not just with respect to a child’s age at surgery but also with the degree of deterioration or lack of control required before you initiate surgery.” He added that even when surgical intervention is warranted, there is no consensus about the best procedure to treat the condition.

Ongoing Studies

Dr. Donahue, who is also the protocol chair and a member of the Pediatric Eye Disease Investigator Group (PEDIG) executive committee, described two studies on intermittent exotropia that are underway. The first study will explore the natural history of intermittent exotropia by comparing patching against no treatment. Patients in this study, which will have a three-year follow-up period, are randomized to receive no treatment or to undergo six months of patching of one eye for two hours per day. The main outcome measure will be the degree of deterioration in each study group. More than 200 children will be enrolled in these trials, which will be conducted at more than 50 designated study centers.

Parents of children who experience deterioration of control will have the option of enrolling their child in a surgical study designed to evaluate the recess-resect procedure versus bilateral recession. “Some people believe that the best approach is to do symmetric surgery because each eye sees well and each eye drifts,” said Dr. Donahue. “In this case, you would operate on the lateral rectus muscle of each eye—the bilateral recession procedure. Others believe that you should weaken the lateral rectus and tighten the medial rectus of the same eye—the recess-resect procedure.”

“We know that each procedure works relatively well, in the 70 percent range for intermediate-term outcome,” Dr. Donahue continued. “But we don’t know whether one procedure has a higher risk of initial overcorrection or if that overcorrection can cause deterioration in binocular vision or might even be helpful in ensuring long-term stability. Those are some of the questions we are trying to answer.”

More than meets the eye.

Dr. Donahue expressed some concern that three years of follow-up may not be sufficient to evaluate recurrence. “These kids may do well for three years, but the problem with intermittent exotropia is that it is not an eye problem or an eye muscle problem. It’s a problem with the brain. That’s why all of the putative treatments—whether they are some type of eye exercises or vision training or eye muscle surgery—have a high risk of recurrence; they don’t fix the brain. That is the big black box that we eventually have to get to the bottom of. What we are currently doing with surgery is an orthopedic solution to a neurologic problem.”

1 Buck D et al. *Br J Ophthalmol*. 2012;96(10):1291-1295.

2 Hoyt CS, Pesic A. *Br J Ophthalmol*. 2012;96(10):1280-1282.

Dr. Clarke reports no related financial interests. Dr. Donahue is a consultant for Diopsys, iScreen, PlusOptix, and REBIScan. Dr. Epley is a lecturer for Alcon.