

those who were not, 91% expressed interest in further education on this topic. Those in a university setting were more likely to be able to identify clinical trials than those in private practice ( $P = 0.0005$ ).

## Discussion

Many early-onset ocular disorders are genetic, offering the possibility for accurate prognosis and family planning if they are diagnosed in a timely manner. Ocular findings may be the presenting clinical manifestation of a syndrome, giving pediatric ophthalmologists the unique opportunity to diagnose multisystem disorders. Pediatric ophthalmologists have a responsibility to recognize a disorder as potentially genetic and either to pursue work-up or refer to the appropriate subspecialist. Once the clinical phenotype is recognized as likely genetic, evaluation by an ophthalmologist with expertise in genetics, a genetic counselor, or medical geneticist should be considered.<sup>1</sup>

Many patients with congenital/juvenile cataracts,<sup>2-4</sup> pediatric glaucoma,<sup>5-7</sup> and congenital/infantile nystagmus<sup>8,9</sup> have a genetic basis for their disorders. For example, in 50 patients diagnosed with bilateral congenital cataracts, putative pathogenic variants were identified in 75% of cases.<sup>3</sup> Nine children from 6 families were diagnosed with metabolic disorders that had been missed by traditional clinical screening algorithms.<sup>3</sup> All children with infantile or developmental bilateral, and in select cases unilateral, cataracts should be referred to either an ophthalmic geneticist or medical geneticist for work-up. Similarly, in a cohort of 202 infantile nystagmus patients, at least 64% had a genetic cause of nystagmus.<sup>8</sup> In a study of 48 infantile nystagmus patients screened using a next-generation gene panel, 58.3% could be genetically diagnosed, with 21% receiving a new diagnosis.<sup>9</sup> An algorithmic approach that includes phenotypic characterization in concert with genetic testing is indicated for congenital nystagmus patients<sup>8</sup> and for all children suspected of having a genetic disorder (Figure 1). Next-generation sequencing has greatly increased the diagnostic yield for congenital/infantile cataracts and nystagmus.<sup>2-4,8,9</sup> Because genetic testing is most accurately interpreted with a pretest clinical diagnosis hypothesis, if referral to a medical geneticist is made, the pediatric ophthalmologist has an important role in providing a detailed clinical diagnosis that will guide genetic testing.

Pediatric ophthalmologists recognize that ocular genetic disorders are common in most practices. The majority of survey respondents identified congenital/infantile cataracts, glaucoma, and congenital nystagmus as potentially genetic. The greatest need expressed in the survey was for continuing education and resources dealing with obtaining genetic testing and identifying eligibility for clinical trials. With the FDA approval of gene therapy for RPE65-related retinal disease, accurate diagnosis of children with congenital nystagmus often becomes actionable. The

Genetic Eye Disease Committee (formerly the Task Force) will offer educational courses and links on the AAPOS website to help address this need. Continuing education about which patients need a genetic work-up will enable AAPOS members to become active participants in the front-line of personalized medicine.

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## Primary IOL implantation in children: the effect of the Infant Aphakia Treatment Study on practice patterns

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**We report the long-term effects on practice patterns at a single institution before (1995-2004) and after (2009-2018) publication**

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of the results of the Infant Aphakia Treatment Study (IATS). The medical records of children <7 months of age who underwent cataract surgery at the Medical University of South Carolina were reviewed. The main outcome measure was the treatment with primary IOL or primary aphakia at the time of surgery. A total of 273 eyes of 183 patients were analyzed. Of the unilaterally affected eyes, primary IOL was the treatment of choice in 28 of the pre-IATS cases (61%) and 3 of the post-IATS cases (6%). In bilaterally affected eyes, primary IOL was the treatment of choice in 18 of the pre-IATS cases (47%) and 11 of the post-IATS cases (23%). According to a year-wise calculation, treatment with primary IOL began to decrease in 2010 and 2011, following publication of the 1-year results of the IATS in 2010. A drastic decrease in primary IOL occurred in 2015, following publication of the 5-year results.

The infant aphakia treatment study (IATS) was a randomized clinical trial comparing the use of contact lenses and intraocular lenses (IOL) for the optical correction of monocular aphakia during infancy.<sup>1-3</sup> In a 5-year follow-up, IATS found no significant difference in the median visual acuity of operated eyes in those who underwent primary IOL implantation and those left aphakic; however, there were significantly more adverse events and additional intraoperative procedures in the IOL group.<sup>3</sup> In unilateral cataract cases, the authors of the IATS outcome results report recommended leaving the eye aphakic and correcting vision with contact lenses or aphakic spectacles with secondary IOL implantation planned for later in childhood.<sup>3</sup> A primary IOL in the <7-month-old age group was recommended only if, in the surgeon's judgment, a family would find the contact lens option so burdensome as to result in significant periods of uncorrected aphakia.<sup>3</sup>

The purpose of this study was to evaluate the effect of the results of a prospective randomized clinical trial (IATS) on clinical practice at the Storm Eye Institute at the Medical University of South Carolina (MUSC). We compared the prevalence of primary IOL implantation in the treatment of infantile cataracts before and after the results of the IATS. Although IATS studied outcomes among infants with unilateral cataracts, we included both unilateral and bilateral cases.

## Subjects and Methods

We conducted a retrospective chart review of the medical records of children who underwent cataract surgery between January 1, 1995, to November 30, 2004 (pre-IATS era), and between January 1, 2009, to August 31, 2018 (post-IATS era). The study was approved by the MUSC Institutional Review Board for Human Research and adhered to the tenets of the Declaration of Helsinki. Eyes of children <7-months-old at the time of their original cataract surgery were included in the analysis. All surgeries were performed by one of the authors (MEW). In bilateral cataract cases, both eyes were treated identically with either bilateral aphakia or bilateral pseudophakia. Only one eye of bilaterally operated eyes was chosen for inclusion. The main outcome

Table 1. Primary intraocular lens (IOL) implantation after cataract surgery in children with unilateral and bilateral cataract: before and after results of Infant Aphakia Treatment Study

Laterality of cataract	Primary aphakia, no. (%)	Primary IOL, no. (%)	Total
Unilateral <sup>a</sup>			
Pre-IATS	18 (39.1)	28 (60.9)	46
Post-IATS	48 (94.1)	3 (5.9)	51
Bilateral <sup>b</sup>			
Pre-IATS	20 (52.6)	18 (47.4)	38
Post-IATS	37 (77.1)	11 (22.9)	48
Unilateral + bilateral <sup>c</sup>			
Pre-IATS	38 (45.2)	46 (54.8)	84
Post-IATS	85 (85.9)	14 (14.1)	99
Total: combined groups	123 (67.2)	60 (33.8)	183

Post-IATS, January 1, 2009, to August 31, 2018; Pre-IATS, January 1, 1995, to November 30, 2004.

<sup>a</sup> $P < 0.001$  ( $\chi^2$ ).

<sup>b</sup> $P = 0.02$  ( $\chi^2$ ).

<sup>c</sup> $P < 0.001$  ( $\chi^2$ ).

measured was the treatment of the patient with primary IOL or primary aphakia.

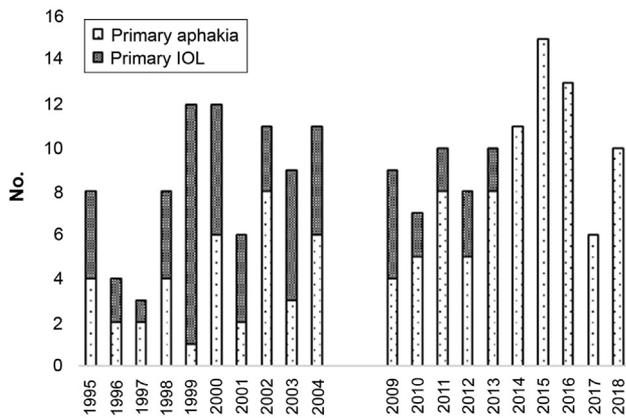
## Results

A total of 273 eyes of 183 patients were reviewed for surgical outcomes of a primary IOL compared to primary aphakia. There was no significant difference in the average age of our patients pre-IATS ( $2.61 \pm 1.99$  months of age [ $n = 84$ ]) and post-IATS ( $2.56 \pm 1.66$  months of age [ $n = 99$ ]) ( $P = 0.9$ ). Before IATS, there were 84 cases: 46 of unilateral cataracts and 38 of bilateral cataracts. After IATS, there 99 cases: 51 of unilateral cataracts and 48 of bilateral cataracts.

Of the unilaterally affected eyes, primary IOL was the treatment of choice in 28 (61%) of the pre-IATS cases and 3 (6%) of the post-IATS cases (Table 1). In bilaterally affected eyes, primary IOL was the treatment of choice in 18 (47%) of the pre-IATS cases and 11 (23%) of the post-IATS cases (Table 1). Between 2014-2018, there have been 43 cases of cataract surgery in infants <7 months of age, all of which were performed with primary aphakia (Figure 1).

## Discussion

In this study, we examined 273 eyes of 183 patients preceding and following the IATS enrollment years. We report a reduction of treatment of infantile cataract with primary IOL from 61% to 6% in unilaterally affected patients and 47% to 23% in bilaterally affected patients, with a total reduction of primary IOL from 55% to 14%. It can be concluded that the results of the IATS have affected our surgical decision making in infantile cataracts, not only in unilaterally affected cases but also in bilaterally affected cases. For bilateral cataract during infancy, aphakic glasses or contact lens use may be a reasonable option. Before



**FIG 1.** Year-wise distribution of number of eyes with primary IOL implantation and primary aphakia. Note, 2018 = eyes operated before August 31, 2018.

IATS, in unilateral cases, primary IOL implantation was more often chosen when compared to bilateral cases (61% vs 47%). Primary IOL implantation after IATS is less common in both the unilateral and the bilateral cataract cases compared to the pre-IATS era.

Our results show that in the post-IATS era, more bilateral-ataract cases were implanted than unilateral cases (23% vs 6%). From our data we cannot be sure why the reduction of primary IOL implantation has been more common in the unilateral group. Perhaps it is because the IATS was performed for infants with unilateral cataract, and thus this group more closely matches the randomized trial data. Parents were partners in the treatment decision, and our recommendations for primary aphakia may have been stronger when the infant fit all of the criteria used in the IATS, including unilateral involvement. A recently published survey on global practice patterns in the treatment of infantile cataracts reported primary IOL implantation by 7% and 4% of pediatric cataract surgeons for patients undergoing unilateral and bilateral cataract surgery, respectively.<sup>4</sup>

A year-wise calculation shows that the decrease in primary IOL implantation began in 2010 and 2011 slightly, following the publication of the 1-year results of the IATS in 2010. A greater decrease occurred in 2015, following the publication of the 5-year results of the IATS.<sup>3</sup> The IATS design publication in 2010 stated it was yet to be determined whether a primary IOL implantation would lead to an improved visual acuity outcome and whether any improvement would be offset by a higher incidence of postoperative complications.<sup>1</sup> The IATS 1- and 5-year results showed no improvement in visual acuity outcome when an IOL was implanted in infants with unilateral cataract but a substantial increase in unexpected returns to the operating room compared to those left aphakic.

The results of this study are limited by the fact that it reports the effects of IATS on treatment decisions of a

single surgeon; nevertheless, we believe these data are generalizable. The IATS had specific exclusion criteria (eg, corneal diameter <9 mm, eyes with stretched ciliary process). No eyes were excluded in this retrospective study. We would have been less likely to implant in the IATS-excluded eyes before IATS as well as after IATS.

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## Driver licensing and motor vehicle crash rates among young adults with amblyopia and unilateral vision impairment

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**This retrospective cohort study investigated whether unilateral vision impairment (UVI) or amblyopia are associated with driver licensing and crash risk among young adults. Electronic health**

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