

Is anterior lenticonus the most common ocular finding in Alport syndrome?

Zare et al.¹ report 11 cases of successful phacoemulsification and foldable intraocular lens implantation in patients with anterior lenticonus due to Alport syndrome. They presented all the cases with nice detail. However, I do not think the statement that “anterior lenticonus is one of the most common ocular findings in Alport syndrome and is found in 90% of patients” is correct. The correct statement would be that more than 90% of anterior lenticonus patients are associated with Alport syndrome.²

Anterior lenticonus is a rare bilateral progressive developmental anomaly and manifests as a slowly progressive deterioration of vision, requiring patients to change the prescription of their glasses frequently. It occurs in approximately 25% of patients with X-linked Alport syndrome; when present, it may be a pathognomonic feature of Alport syndrome (Saxena R. Alport Syndrome. EMedicine. Last Updated, January 30, 2007. Available at: <http://www.emedicine.com/med/topic110.htm>. Accessed 6/8/07).

In a study in Finland,³ 34 Alport syndrome patients were examined and ocular abnormalities were found in 32%. Six patients had retinal flecks, and 4 had anterior lenticonus. In 94 cases of Alport syndrome patients studied in 5 separate study groups,⁴ anterior lenticonus was recorded in 22% (Table 1).

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REPLY: Amiraslanzadeh states that our sentence about anterior lenticonus being found in 90% of Alport syndrome patients is not correct. He then says that 90% of anterior lenticonus patients are associated with Alport syndrome, but this does not mean that 90% of Alport syndrome patients have anterior lenticonus.

In a study by Colville and Savige,¹ the prevalence of anterior lenticonus in Alport syndrome is 25%. In a report by Chugh et al.,² the prevalence is 37.8% in 63 patients. In this report, the prevalence of other clinical manifestations is as follows: hematuria, 96.8%; deafness, 82.5%; diminished visual acuity, 66.7%; hypertension, 71.4%; high-frequency sensorineural deafness, 96.8%; bilateral anterior lenticonus, 37.8%; retinal flecks, 22.2%; cataract, 20%; and keratoconus, 6.7%.

The typical ocular associations of Alport syndrome are a dot-and-fleck retinopathy, which occurs in about 85% of patients; anterior lenticonus, which occurs in about 25%; and the rare posterior polymorphous corneal dystrophy.¹ Additional ocular features described in Alport syndrome include other corneal dystrophies, microcornea, arcus, iris atrophy, cataract, spontaneous lens rupture, spherophakia, posterior lenticonus, a poor macular reflex, fluorescein angiogram hyperfluorescence, electrooculogram and electroretinogram abnormalities, retinal pigmentation,¹ macular hole,³ and, rarely, telangiectasia of conjunctiva.⁴

The prevalence of ocular manifestation in Alport syndrome in the literature varies; in some reports the corneal manifestations are high⁵ and in others, lenticular or retinal manifestations are high.^{1,6} Teekhasaene et al.⁵ reported that ocular findings were present in 82.3% of their Alport syndrome patients and that corneal changes such as posterior polymorphous

Table 1. Prevalence of ocular abnormalities in 94 Alport syndrome patients younger than 25 years.

Number of Patients/Men	Number of Patients			
	Any Ocular Abnormality	Anterior Lenticonus	Retinal Flecks	Posterior Polymorphous Dystrophy
51/43	23	10	18	0
9/8	8	4	7	0
6/6	6	1	2	2
8/6	6	4	4	5
20/15	8	2	4	0
Total 94/78	51 (54%)	21 (22%)	35 (37%)	7 (7%)

dystrophy were the most common (64.7%). Colville et al.⁶ reported that dot-and-fleck retinopathy and anterior lenticonus were present in almost 75% of their cases.—*Mohammad Taher Rajabi, MD*

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Scleral fixation technique with scleral cleft

I congratulate Monteiro et al.¹ for their article about scleral fixation via suture burial in a scleral cleft. Scleral suturing is done in cases in which the patient has iris problems, such as a scleral ring, or when the intraocular lens (IOL) requires suturing. In this article, the technique is used to prevent the suture ends from protruding from the conjunctiva by keeping them in the scleral cleft. After looking at other reports on this topic, I would like to make the following points:

1. This method is similar to my scleral fixation method²; however, I do not create a scleral cleft. Surgeons do not need a scleral cleft, flap, or patch to bury the suture and knot.
2. The need to prepare 2 clefts.
3. As mentioned by Monteiro et al., based on the large corneal incision, suturing of the incision could be joined to the scleral cleft. Because of this, the suturing position could be changed to 2 o'clock and 8 o'clock or 4 o'clock and 10 o'clock.
4. In my article on scleral fixation,² the suture end with the knot is buried in the sclera. The edges will not cause problems because the knot and suture edge lie horizontally in the sclera. It is not necessary to cover the knot with a patch, graft, flap, or rotation. Additionally, the technique is easy; once it has been learned, it can be performed quickly.

The suture burial technique has been used successfully in all my cases requiring scleral fixation.

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REPLY: We have been using our technique of scleral fixation for more than 10 years, not only in cases of secondary IOL implantation (fixation of both haptics or fixation of 1 haptic and placement of the other haptic in the sulcus with capsule support), but also in cases in which broken haptics must be resutured.

The scleral cleft is created to bury the polypropylene (Prolene) 10.0 knot in the sclera. Because of the way the suture is fixated to the haptics (sailor loop), the knot does not untie. The scleral suture can be done in all 360 degrees of the globe. The scleral incision is only sutured in cases with thin sclera.

In 10 years of using this technique, we have not had a single case in which the suture ends protruded from the conjunctiva.—*Manuel Monteiro, MD*

Techniques for scleral fixation of IOLs

Monteiro et al.¹ propose a variation to the technique of scleral fixation of intraocular lenses (IOLs). Although details are not provided in the body of the paper, the complications of hyphema, vitreous hemorrhage, and choroidal hemorrhage are mentioned in the abstract. We believe these occurrences may be peculiar to their technique of scleral fixation.

Monteiro et al. make 2 entries into the eye in the region of the ciliary sulcus, the first with a 30-gauge needle, followed by the “blunt” end of the double-armed 10-0 polypropylene (Prolene) suture. Considering the vascularity of the ciliary body, 2 penetrations, the second by a blunt instrument, may tear ocular tissue and thereby predispose these eyes to the aforementioned hemorrhagic complications.²

The authors argue that an advantage of their technique is the lack of scleral flaps. In the absence of a scleral bed, the knot of the anchoring suture is buried in the scleral incision. On careful study of the illustrations, we notice that the anchoring suture is subconjunctival at the conclusion of the procedure. This is not the case when scleral flaps are fashioned, as the anchoring sutures are placed in the scleral bed and covered by the scleral flap as well as the conjunctiva.

This difference is important. As the authors point out, suture exposure might lead to endophthalmitis even after several years. In the case of scleral-fixated IOLs, the anchoring suture communicates directly with the intraocular cavity. We believe the anchoring