

Alport Syndrome and the Eye

Here is a checklist for you to take to your Optician or Eye Surgeon, summarising the range of changes that can be seen in the eye in Alport syndrome, the symptoms of many of these disorders overlap, so if there is any concern, you should seek the opinion of an eye specialist.

We believe most of these changes develop with age, and are infrequent in teenagers.

But overall, we do not have good data on how common these findings are or how they progress with time, which is what the research group at St Thomas' Hospital are keen to investigate. We can be contacted either through the support group or email us directly. *Mr Moin Mohamed PhD FRCS FRCOphth, Consultant Ophthalmic Surgeon, St Thomas' Hospital, London, SE1 7EH,* (moin.mohamed@gstt.nhs.uk)

Diagnosis	Symptoms	Specialised Tests	Treatment	Comments
Myopia / Short-sightedness	Blurred vision, difficulty seeing in distance, clear when close	Refraction by Optician	Spectacles	If prescription 'irregular', get further testing by eye surgeon you may have lenticonus
Recurrent Corneal Erosions	Sudden sharp pain, watering, sensitivity to bright lights, usually on walking	Slit lamp examination by optician or 'eye casualty' when symptomatic using fluorescein	Lubricating eye drops if frequent, ideally a viscous ointment at night before bed	Settles within 1-2 days, if eye is very red, must exclude infection
Posterior Polymorphous Dystrophy	Blurred vision	Detailed slit lamp examination, endothelial examination by eye surgeon	Nil initially, if severe may benefit from deep lamellar corneal grafting	Very rare finding
Anterior Lenticonus or rarely Posterior Lenticonus	Classic Alport eye finding, a conical deformity of the shape of lens surface. Blurred vision, progressive short-sightedness not correctable with spectacles	Slit lamp examination, Scheimpflug photography, pentacam imaging, wavefront analysis by eye surgeon	Lens replacement surgery	Lenticonus is a specific finding in Alport, >90% of lenticonus seen by eye surgeons is caused by this condition
Cataract	Blurred vision, haze and glare	Slit lamp examination by eye surgeon	Cataract extraction and intra-ocular lens implantation	May relate to steroid use after kidney transplant
Retinal Flecks	A finding typical of Alport's, does not appear to have symptoms	Fundus examination by slit lamp biomicroscopy	None	White spots at the macula and the peripheral retina – not macular degeneration, also seen in carriers
Macular 'Lozenge'	No symptoms in early disease	OCT scan of the macula by eye surgeon (ideally a retinal specialist)	None	The natural history, and late findings in older people is unknown
Giant Macular Hole	Blind spot in central vision	OCT scan by retinal specialist	Vireo-retinal surgery (very specialised eye surgery)	Rare