1. What’s your opinion about the use of optical aids in aniridia cases?

SK: To my opinion, spectacles work the best. We should work towards getting a good retinoscopy and refine it as much as possible.

PS: Optical aids in terms of refractive correction is very important in the management of aniridia. Correct any refractive error appropriately. Ideally, transitions lenses or tinted glasses for outdoors to reduce glare (see below). Low vision aids including handheld magnifiers.

CP: In my opinion the binocular optical aids can be effective for near vision in patients with aniridia.

AM: Optical aids can be of use and should be tried in any patient with low vision. Their potential benefit can only be discovered when tried on each individual as they may benefit some patients and make no difference to others.

RP: Always issue the full correction.

AL: I think they are important as we want to ensure the patient is optically corrected as best as possible.

2. What wavelength do you consider most effective for filters? Maybe 550 nm?


PS: No great evidence that I could find on this important question. The eye is most sensitive to wavelengths in the region of 555nm (yellow-green). The strength of the filter will reduce glare but also contrast and visual quality. Ideally, the optimal filters should be selected by examining visual acuity, contrast frequency sensitivity, glare sensitivity and subjective selection by the patients. UV filter is important in addition to tilt.

CP: Yes. When the patient is collaborator, I generally indicate subjective testing of different filters for indoor and outdoor use.

AM: I am unaware of any evidence to support a particular wavelength of filters to be used. Again, a “trial and error” approach is advisable for relieving the patient’s photophobia.

RP: There is some literature that advocates the use of 400-500nm filters, but I have no experience of this.

AL: The eye is maximally sensitive to approximately 555nm so 550 nm would be fine.

3. What should be considered in order not to create anti-elevation?
SK: IO insertion should never be put anterior to IR, while operating on the IO. Bunching posterior fibres of IO within 2 mm with no lateral spread is the most important. It is also advisable that when we are disinserting the inferior oblique, we should resect the fibres flush with the sclera to avoid any resection effect.

PS: Savleen had a very good slide with the major points on surgical technique. This webinar is archived on the WSPOS YouTube Channel.

CP: This complication could be avoided by inserting the posterior fibres of the inferior oblique no more than 2 millimetres lateral to the insertion site of the inferior rectus. To minimize the desired antielevator effect, this procedure must be performed bilaterally.

AM: To avoid anti-elevation one should not anteriorise the posterior fibres of the inferior oblique muscle.

BVR: A brief note on the Antielevation Syndrome =>
IOAT procedure can cause limitation of elevation in abduction in the operated eye as well as marked upshoot of the contralateral adducting eye that mimics overaction of the IO muscle of the other eye. This adverse outcome has been named the Antielevation Syndrome.
Steps To Follow To Reduce Antielevation Syndrome =>
(a) Do not place the New insertion of the Anterior fibres of IO muscle more than 2 mm anterior to the inferior rectus muscle insertion.
(b) Posterolateral fibres of the IO muscle must be posterior to the lateral end of IR insertion and do not spread out substantially and rather should be bunched up with the anterior nasal corner making a J-deformity IOAT.
(c) Avoid some of the IO muscle getting resected inadvertently by suture placement proximal to the insertion.
(d) Avoid doing unilateral IOAT

4. Does the traction test have a place in anti-elevation syndrome? (preop-postop)

SK: We should always perform a traction test for elevation in adduction as well as abduction. But the test may be negative in early cases of anti-elevation. Only long-standing cases cause the FDT to be positive (adherence syndrome).

CP: The FDT is useful to make a differential diagnosis of antielevation syndrome with Parks's restrictive syndrome, which presents gradually over time and has a positive duction test, unlike antielevation syndrome, which appears in the immediate postoperative period and has FDT negative.

RP: Yes it can differentiate longstanding bilateral 4th nerve palsy from mechanical restrictions

BVR:
(a) Traction test is typically done to assess for laxity or tightness of Oblique muscles.
1) SO traction test to assess the laxity of SO tendons prior to tucking them in cases of Congenital SO palsy and match it with normal side. Tuck only loose SO tendon & NEVER tuck normal SO muscle.
2) SO traction test is done to feel the tightness in SOM in cases of Brown syndrome and A pattern with SOOA.
3) IO traction test is to assess if complete myectomy done or any posterior 1/8th slip of IO has been left behind which can result in persistent IO overaction.
(b) FDT (forced duction test) is typically done to assess the tightness in Recti Muscles. FDT is helpful in differentiating Adherence syndrome from Antielevation syndrome.
1) Adherence syndrome is seen after IO weakening procedures, in which hypotropia, limitation of elevation in primary gaze, and positive FDT resulting from adhesion between IO and surrounding tissue.
2) Antielevation syndrome typically has resistance in FDT in superotemporal direction due to tethering caused by neurofibrovascular bundle of IO. Hypotropia in primary gaze or severe adhesion around the IO is not found in antielevation syndrome.

AL: Yes.

5. What are the other surgical options for this case?

SK: (a) Explore for posterior fibres of the same eye Inferior Oblique
(b) Operate on the other eye Inferior Oblique
(c) Other eye Superior Rectus weakening-recession or mini-tenotomy

CP: I think that a better option would have been an inferior oblique recession due to mild hypertropia of the left eye in primary position.

AM: Revising the left inferior oblique and “reducing” its anteriorisation, or recessing the right inferior oblique.

MCGN:

6. Apart from everyday lubrication is there anything more we can do for keeping cornea clear?

SK: Early and regular examination to pick up aniridia associated keratopathy and stem cell deficiency is a must. Since it is an inherent problem, we cannot prevent it but only treat it.

PS: Yes, punctal plugs are a good solution in children with dry eyes, especially due to issues with drop compliance and regular drop instillation in children. Important to eliminate preservatives as much as possible.

CP: Use preservative-free lubricants, autologous serum, amniotic membrane, scleral contact lenses.

AM: Daily lubrication is pivotal in prevention of aniridia-associated-keratopathy’s deterioration. Other tools to use are anti-inflammatory drops, autologous serum eyedrops, cyclosporine eyedrops etc. in more advanced stages. In all cases it is advised to use preservative-free products, even for short-term applications. Also, any trauma should be avoided.

RP: Not to my knowledge. In cases of severe keratopathy stem cell transplants are possible. My knowledge on this is limited.

7. Is there any role for autologous serum?

SK: No studies that I am aware of, but authors use it for any ocular surface problem.

PS: Yes, as in all forms of limbal stem cell deficiency and severe dry eye. However, it is challenging from a practical perspective to obtain serum drops based on facilities at different centres. It can also be a huge challenge in children to obtain blood and enough volume of blood for regular treatment.


AM: Autologous serum eyedrops can be used, more so in the second or third decade of life when keratopathy is more advanced and autologous serum is easier to prepare (drawing blood from a young child every month or every few months is not advisable).

RP: Yes