

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Interventional procedures

Patient Organisation Submission

Insertion of an artificial iris implant for aniridia (IP1738)

Thank you for agreeing to give us your views on this procedure or operation and how it could be used in the NHS.

When we are developing interventional procedures guidance we are looking at how well a procedure or operation works and how safe it is for patients to have.

Patient and carer organisations can provide a unique perspective on conditions and their treatment that is not typically available from other sources. We are interested in hearing about:

- the experience of having the condition or caring for someone with the condition
- the experience of having the procedure or operation
- the outcomes of the procedure or operation that are important to patients or carers (which might differ from those measured in clinical studies, and including health-related quality of life)
- the impact of the procedure or operation on patients and carers. (What are the benefits to patients and their families, how does it affect quality of life, and what are the side effects after the procedure or operation.)
- the expectations about the risks and benefits of the procedure or operation.

To help you give your views, we have provided this template. You do not have to answer every question — they are there as prompts. The text boxes will expand as you type, the length of your response should not normally exceed 10 pages.

Please note, all submissions will be published on the NICE website alongside all evidence the committee reviewed. Identifiable information will be redacted.

About you	
1. Your name	██████████
2. Name of organisation	Aniridia Network
3. Job title or position	Trustee
4. Brief description of the organisation (e.g. who funds the organisation? How many members does the organisation have?)	Aniridia Network is a registered charity funded entirely by charitable donations and run entirely by volunteers. We currently have a total of 839 members with over half of these being people with aniridia themselves and the remainder being mostly parents/carers.
<p>5. How did you gather the information about the experiences of patients and carers to help your submission?</p> <p>(For example, information may have been gathered from one to one discussions with colleagues, patients or carers, telephone helplines, focus groups, online forums, published or unpublished research or user-perspective literature.)</p> <p>This information is based on one to one conversations we have had with patients/carers over a number of years, both face to face and by email/telephone. We also used information gained from discussions within online forums for the aniridia community, both past discussions and those initiated specifically to gather feedback for this submission.</p>	

Living with the condition

6. What is it like to live with the condition or what do carers experience when caring for someone with the condition?

We represent patients with congenital aniridia. Although this condition is named for the characteristic absence of some or all of the iris, usually in both eyes, it is in fact a genetic condition which affects the development of many parts of the eye. Many secondary eye conditions are also associated with congenital aniridia. Conditions such as foveal hypoplasia and nystagmus are very common in people with aniridia leading to low vision from birth and conditions such as cataracts, glaucoma and aniridia keratopathy (clouding of the cornea) can develop at any time during childhood or adulthood leading to further sight loss. Most people with aniridia are registered as “sight impaired” or “severely sight impaired” and we are not aware of anyone with aniridia who meets the eyesight levels required to be able to drive a car in the UK. Clearly any level of sight impairment will impact on many aspects of a patient’s daily life including household tasks (cooking, cleaning etc.), education/employment, getting out and about independently, participating in social, recreational and sporting activities etc. Reduced visual acuity leads to an inability to see detail, including written information without adequate magnification or enlargement which can have a wide ranging impact. A lack of depth perception is also common which can make patients more likely to trip or fall on unexpected slopes or steps, and can make participation in activities such as ball sports difficult, which can particularly impact on children who wish to play with their peers.

Particularly relevant here is the impact of photophobia and glare. Having an enlarged pupil and no iris to control the amount of light entering the eye can make those with aniridia very sensitive to light. This sensitivity can increase with the development of secondary eye conditions such as cataracts and aniridic keratopathy (clouding of the cornea) which can cause the light entering the eye to be incorrectly scattered. The impact of photophobia and glare can be to significantly reduce functional vision. The natural reaction to light that is too bright is to close one’s eyes. Fighting this instinct can be difficult, leading to excess blinking and eye watering. This can then make it difficult for the patient to properly look around them and make full use of the vision they have. Glare and dazzle can act to obscure other objects from view. This can make moving around outdoors more difficult as obstacles may become less visible. Patients with aniridia may find it more or less difficult to move around safely outdoors depending on weather conditions and time of day. For example, bright sunlight, the sun being low in the sky, rain causing reflections off the ground and dazzle from car headlights at night can all be problematic. Even indoors photophobia can be an issue. Sitting facing a window can be very uncomfortable for patients and again glare from a window can obscure objects from view or reduce them to a silhouette. Some patients also report that certain types of electric light, particularly from fluorescent bulbs, cause them a lot of discomfort.

In the short term (a few minutes) photophobia can cause discomfort, over a longer period (tens of minutes or hours) it can cause eye pain or headaches which can be quite painful. This is at least partly caused by the natural reaction to squint in an attempt to reduce the amount of light entering the eye.

It is fairly common for those with aniridia to develop ptosis (drooping eyelid(s)). This is likely due to trying to reduce the amount of light entering the eye. In severe cases the eyelid can then start to impede vision, even in comfortable lighting conditions and surgery may be required.

There is little that can be done medically to treat photophobia. One option is to use contact lenses with an opaque iris painted on however the corneal clouding (aniridia keratopathy) can be triggered or accelerated by wearing contact lenses. This makes many contact lenses unsuitable altogether for patients with aniridia. Some patients do have success with specialised contact lenses but most are not offered option at all as their ophthalmologist considers it too risky.

The only option for most patients is to try to control the amount of light they are exposed. Indoors this can be done by keeping blinds or curtains closed or tinting windows at home or in the car. It can be more difficult to make these adjustments in shared spaces such as a school or the workplace, and almost impossible in public spaces such as shops and restaurants.

Outdoors patients will often wear a hat with a wide peak or brim (typically a baseball cap), dark sunglasses or both together. Patients typically prefer the wraparound sports style of sunglasses as they sit close to the face so that no light can enter around the side of the lenses. It can be difficult to find suitable sunglasses, particularly for children, and also expensive to have them fitted with prescription lenses (if these are required to improve vision). It can also be difficult to find items such as appropriately tinted swimming goggles to use outdoors or in swimming pools with large windows. Keeping sunglasses on babies and young children can also be challenging. It can be difficult for patients to have to constantly take their sunglasses on and off if they are, for example, going in and out of shops. Patients report that light reactive lenses typically do not react quick enough for their needs.

Patients will often need to wear a hat and/or sunglasses even on cloudy or winter days when other people would not be wearing them, which can make patient's appearance stand out or can make it difficult to dress for more formal occasions. Wearing sunglasses or a hat can make it more difficult to make eye contact which can inhibit social interaction. The effect of ptosis can also be to make the patient appear to be sleepy or inattentive which could also provoke a negative reaction from others. Many people within the congenital aniridia community take pride in their lack of an iris and are not concerned about their eyes' appearance however a minority do want to have eyes which appear more "normal". Sadly this is often as a result of bullying which can have a significant impact on young people.

Advantages of the procedure or operation

7. What do patients (or carers) think the advantages of the procedure or operation are?

Reduced sensitivity to light leading to: less pain or discomfort due to photophobia, better functional vision due to a reduction in glare/dazzle, less reliance on sunglasses and/or hats outdoors.

Some improvement in visual acuity.

A more “normal” appearance to the eye (this would not be regarded as a benefit by all patients)

Disadvantages of the procedure or operation

8. What do patients (or carers) think the disadvantages of the procedure or operation are?

Risk of triggering raised IOP or previously well controlled IOP no longer responding to medication, potentially leading to surgery or sight loss due to glaucoma.

Risk of developing or accelerating the progress of aniridic keratopathy (cornea opacification).

Risk of developing aniridic fibrosis syndrome.

Complications requiring follow up surgery

Concerns about whether the surgery could be riskier for children who are still growing.

Could a pupil of a fixed size cause problems seeing in low light conditions?

Patient population
<p>9. Are there any groups of patients who might benefit either more or less from the procedure or operation than others? If so, please describe them and explain why.</p> <p>Patients with congenital aniridia may have a higher risk of complications from the procedure than those with traumatic aniridia (loss of an iris due to an injury to the eye). This is due to the underlying genetic cause of congenital aniridia and the secondary eye conditions which can often accompany it.</p>
Equality
<p>10. Are there any potential equality issues that should be taken into account when considering this topic?</p> <p>None we are aware of.</p>
Other issues
<p>11. Are there any other issues that you would like the Committee to consider?</p> <p>There are a number of different models of iris implant available on the market, with different models being available for different lengths of time and newer models advertising themselves as having improved technology. We would like to ask the committee to consider if certain models give better outcomes for patients and should therefore be recommended over others.</p> <p>We would also like to ask to the committee to consider if different guidelines are required for using iris implants, depending on whether the patient has congenital aniridia, traumatic aniridia or another iris abnormality.</p> <p>Some patients seem to be unclear about how artificial irises might benefit them and also unaware of potential risks. Iris implants are a relatively common discussion topic in online patient forums, particularly with the recent approval of the use of one particular model by the FDA in the United States. Patients are quite often aware that it is less risky to have iris implant fitted in conjunction with a cataract removal as this eliminates the need for a separate surgery. When the patient then requires cataract surgery, they may be keen to take the opportunity to have iris implants fitted or their ophthalmologist may suggest it without the patient being fully aware of the possible benefits and risks.</p> <p>As many of the congenital aniridia patients who have had iris implants fitted, did so alongside cataract removal, it can be difficult for them to know how much of the benefit they received from the surgery was specifically due to the iris implant.</p>

Key messages

12. In no more than 5 bullet points, please summarise the key messages of your submission.
1. Photophobia can have a significant impact on the functional vision of congenital aniridia patients.
 2. Congenital aniridia patients are already at significantly increased risk of developing secondary eye conditions which can lead to further sight loss. Surgery can increase the risk of these conditions developing or hasten their progression.
 3. There is concern in the patient community about the safety of iris implants, particularly in children.
 4. The patient community would like to know if certain models of iris implant are more suitable for patients with congenital aniridia than others (i.e. reduced risk of complications) and to have access to these via the NHS.
 5. We would like the guidance to include specifics on treating patients with congenital aniridia.

Thank you for your time.

Please return your completed submission to jp@nice.org.uk