

Case Study 4: 4 year old girl with aniridia and significant VI

Report included:

Clare was referred for a visual assessment by her paediatrician who requested information regarding Clare's visual function. Clare has aniridia and WAGR syndrome. She is under the ongoing care of the hospital ophthalmology and orthoptic/optometric team and has been prescribed glasses from the hospital clinic (both clear and tinted lenses). Her father reports good compliance.

Clare has a significant visual impairment. Her early visual development was delayed and while her visual responsiveness has improved she has many visual difficulties in addition to the underlying aniridia of the WAGR syndrome. In addition to poor vision Clare has a significant refractive error (need for glasses) nystagmus (eye wobble), photophobia (discomfort in bright conditions), cataract, strabismus, absent stereopsis (lack of 3D vision) and reduced contrast sensitivity (difficulties with visual material that is at low contrast). All these factors will contribute to her visual impairment.

Significant visual impairment and multiple ocular problems

Although Clare's level of vision is difficult to assess, she worked very hard today and **enclosed are images of an appropriate size and complexity** for someone with Clare's level of vision. Visual information, educational and recreational should be at least this size. A rule of thumb is that objects will need to be at least 12x larger or 12x closer for Clare to be able to see them as well as someone with typical vision.

Clare's contrast sensitivity was significantly reduced compared to age norms. Reduced contrast sensitivity is known to cause difficulties with face recognition, identifying edges and contours (including steps/kerbs) and orientation in new environments. This must be considered, in conjunction with size, when providing play and educational material for Clare. When drawing or writing Clare may find using a soft pencil or marker (one that makes a thick, dark mark) more visible than an ordinary pencil or coloured crayons.

When working with Clare today I noticed that she was less visually responsive to visual stimuli at a distance of more than about one metre. She is likely to do better with play and educational material placed within approximately one metre and presented in as simple a format as possible. Too much information on a page/surface/plate is likely to reduce Clare's ability to access what is being presented.

Proximity best

Clare has a strabismus (squint/turn) in her left eye. She mainly uses her right eye to view the world. Because she is only using one eye she will not have 3D stereoscopic vision and this may make mobility more challenging for her, particularly when coupled with reduced vision and contrast sensitivity.

To summarise, in addition to needing glasses with a dark tint, Clare demonstrates;

- **Significantly reduced vision** requiring play/education material to be approximately 12x larger or closer than for an average person.
- **Significantly reduced contrast sensitivity** requiring play/education material to be bold, distinct, highly contrasting.
- **Lack of stereopsis** meaning that she does not have pure 3D vision and may find judging distances and some other tasks involving hand/eye coordination challenging.
- Evidence of **difficulty with crowded visual material**. Clare is likely to have more chance of engaging visually with material that is presented in isolation with visual distractors minimised (e.g. play/education material placed on a plain, uncluttered table or an unpatterned carpet).
- More **visual interest in objects within approximately 1m**.

I will review Clare at her father's request, prior to her starting school next year. If you have any comments or queries relating to this report, please contact me.

Technical details

Current spectacle correction (six months old, hospital Rx, one pair dark tint, one pair clear, both good condition):

R +2.50/-2.50x180 L +2.00/-2.00x165

Visual acuity with current Rx (Kay Crowded Pictures) under habitual viewing conditions (BE open, fixing with RE):

@ 1m 6/72

@ 33cm 6/76

Contrast sensitivity with current Rx (Cardiff contrast test): 12.5 (8% contrast)

Accommodative function (dynamic retinoscopy): grossly normal through own Rx

Ocular posture: L exotropia, nystagmus, occasional tonic updrift to right.

Seen by: AN Other, _____ Senior Optometrist

Cc: Parents, Paediatrician, General Practitioner, Ophthalmology, Paediatric low vision clinic, support teacher for the visually impaired, therapists