Case Study 8: 3 year old girl with severe developmental delay, CP, seizures and profound visual impairment

Report included:

Lisa is a three-year-old girl with a diagnosis of West Syndrome and cerebral palsy (spastic quadriplegia). She is on medication to control seizure activity (Sabril/Vigabatrin). Pale optic discs were confirmed by Lisa’s ophthalmologist. Lisa is currently undergoing formal assessment of special educational need and her mum is keen to know how well she sees. Mum feels her daughter’s vision is limited and that her visual interest is variable.

Summary of findings:
Lisa has a profound visual impairment probably limited to the perception of light. It is likely that this is primarily due to damage within the brain (cortical visual impairment) but there is also evidence of optic atrophy. Optic atrophy is a condition in which the nerves that carry information from the eyes to the brain are not functioning normally.

Lisa’s response to light is, at best, inconsistent. Sometimes Lisa appears to look at a bright light in a dark room and at other times fails to demonstrate any recognisable response. Whilst improved control of seizure activity may allow her to use what vision she has slightly better, she is likely to retain a severe visual loss. She appears to respond optimally to large light-emitting objects placed centrally and close to her. Lisa should be given plenty of time to ‘see’ such items as it is likely that visual information is processed slowly and it may help to place her hands on visual stimuli to help her locate them and get some meaning from them.

The issue of Lisa’s use of Sabril/Vigabatrin has been raised in the past in relation to its possible impact on visual function. Mum feels that it has been a very effective drug and attempts to wean her from it have not been successful. That being the case, the balance is very much in favour of continuing the treatment, as the additional impact on visual fields is likely to be minimal given the magnitude of her other visual problems.

Lisa has previously enjoyed exposure to a multi-sensory environment through the charity Sense. This will continue to be recommended once she moves on to a formal educational environment with the emphasis on non-visual means of communication and education.
Visual function results:
Refractive error (non-cycloplegic): Low hypermetropia R and L. Spectacles not required
Ocular movements: grossly full
Ocular posture: right/alternating divergent strabismus
Pupil responses: sluggish to light
Vision: variable and inconsistent response to light and changes in room illumination.
Fails to consistently fix or follow light. No blink reflex.
Ophthalmoscopy: pale discs R and L – suggesting optic atrophy

This is a report from the multidisciplinary team at the Special Visual Assessment Clinic. J. Little (optometry), C. Lundy (developmental paediatrics), K. J. Saunders (optometry), M. Shannon (Vision Support teaching) Queries relating to this report? Contact Dr. Little XXXX.

Cc: Parents, General Practitioner, Ophthalmologist, Support teacher for the Visually Impaired, Paediatrician, Educational Psychologist