

Impact of habilitational strategies on quality of life and visual performance in children with cortical visual impairment

Cerebral visual impairment (CVI), accounts for half of all blind children in the UK. Extensive clinical experience suggests that structured clinical history taking has been helpful in both diagnosis and habilitation for children with CVI. Software is available that generates an individualised habilitational strategy from a pre-defined database that are selected based on questionnaire responses. Tailored habilitational strategies can have a profound impact on quality of life but this has not been proved formally. Children with CVI have a wide variety of symptoms and further understanding of which groups of patients respond to which particular strategies is urgently required. The strategies used will be developed on an individual basis and based on the findings of a battery of clinical assessment tools. Comparing quality of life scores before and 6 months after implementation will investigate the effectiveness of the strategies while MRI and electrophysiological measures (visual evoked potentials) will determine any changes in visual pathway structure and function.

Participants in the study will receive a full medical and neurological history work up. The medical assessment will include a full ophthalmological and orthoptic assessment and will receive refractive correction if needed. GMFCS, cognitive level, medical history and presence of ocular conditions obtained by applicants from medical notes. The participants will also undergo clinical visual evoked potential (VEP) testing to determine the presence of any visual pathway pathology. The student will be responsible for the behavioural assessment of the participants. Testing will include the questionnaire that specifically investigates problems known to arise in children with CVI. Quality of life will be assessed employing the PedsQL (child and parent versions). The student will also be responsible for carrying out (initially under supervision) multi channel cognitive VEPs in those patients who are old enough to co-operate adequately. The cognitive VEPs will recorded to stimuli in an odd-ball (mismatch negativity and P300) will provide valuable information regarding higher visual processing. The student will co-ordinate and analyse brain MRI examinations. The imaging study is designed to assess optic radiation structure using tractography and to map cortical thicknesses using T1-weighted MRI.

Employing the 'Insight database tool the student will also plan the habilitation intervention. This strategy will include advice for the home and school. A structured interview will be used to explore each intervention strategy and implementation will be quantified for each strategy, allowing for report of non-implementation. Participants will be reassessed at 3 and 6 months after the start of the habilitation intervention.

Data on the process and conduct of, and compliance with, the intervention will be analysed to evaluate the effectiveness of the habilitation strategies on quality of life and visual function assessed by visual evoked potentials. Baseline analysis will involve description of baseline questionnaire data and comparison with demographic, ophthalmological and medical findings. The questionnaire data will be divided into the subgroups (e.g. items related to dorsal versus ventral stream processing problems) based on previous analyses of reliability of subgroups in the questionnaire data⁵ within a diagnosis of CVI. The subgroup scores will be compared with quality of life data to see which symptoms have most impact at baseline.

At 3 months, patients/parents will be asked to complete a questionnaire to assess their concordance with and experience of the intervention. A parent-reported compliance score will be recorded with an opportunity given for report of reasons for non-compliance if necessary. The questionnaire scores and quality of life scores will be compared to baseline both in total and by subgroup and baseline predictors of an improved quality of life after the intervention will be sought. Linear and multivariate regression analysis will be used, as appropriate.

Little experimental work exists to guide choice of outcome parameters and effect size. The primary outcome will be quality of life and the relationship to the habilitation strategies. We hypothesize there may be (beneficial) changes in important non-visual areas of the children's with CVI functioning, that will improve after tailored strategies.

Other outcomes will include objective data on visually evoked potentials (VEPs), in particular those that are responsive to stimuli eliciting "visual attention" - a key part of visual perceptual function.