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SCOPING REVIEW OF HOMONYMOUS HEMIANOPIA IN CHILDHOOD

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Cerebral visual impairment (CVI) encompasses a heterogeneous group of disorders and a spectrum of types of visual impairments. Research is needed to characterise the different forms of CVI and identify the specific needs of these groups to inform individualised patient care. Homonymous hemianopia (HH) is a definable visual field defect that affect some children with CVI. As part of a new research programme, we conducted a scoping review of the literature on HH in children and young people to map current knowledge and identify evidence gaps.

We used the PRISMA extension for Scoping Reviews methodology. Multiple online databases were searched using terms associated with 'homonymous hemianopia' and 'children'. This yielded 1588 papers which were screened by two reviewers. Of these 1001 were excluded at abstract screen and a further 415 excluded after full text review, with full text unavailable for 15. Data were extracted and charted from 157 studies and additional grey literature.

Interim analysis shows reported studies are predominantly from high income countries with a paucity of higher-level evidence, and a preponderance of case reports. Most papers reported causative pathology and diagnosis of HH. There was minimal attention to or evidence relating to intervention. Child-specific grey literature on HH was limited.

This review collates the current evidence-base for HH in children. It demonstrates the important evidence-gap relating to intervention in these cases that would help inform more individualised care. Similar scoping reviews may be prove useful in assessing the evidence relating to other definable groups within the CVI umbrella.



UNEXPECTED RETINOPATHY IN A PATIENT PRESENTING WITH BILATERAL OPTIC DISC SWELLING

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A 12-year-old boy presented with 5 day history of blurry vision, 'wobbly eyes', tinnitus and difficulty seeing at night. Local ophthalmology noted bilateral optic disc swelling and referred him urgently for neurological investigations.

Clinical Findings: At presentation VA was RE 0.00 and LE 0.2 with normal Ishihara colour vision. His extraocular movements were full without manifest strabismus. Fundoscopy showed bilateral optic disc swelling. Electrophysiology unexpectedly revealed a functionally cone isolated retina with markedly abnormal rod function. Pattern VEPs indicated bilateral macular pathway dysfunction affecting left eye more than right eye. Wide field imaging showed bilateral diffusely scattered yellow-white flecks in the midperiphery of each eye. His

kinetic visual fields were moderately restricted bilaterally. MRI showed a Chiari 1 malformation with cerebellar tonsil herniation, but LP opening pressure was normal.

Differential diagnosis included RDH5 retinopathy or vitamin A deficiency. On questioning he reported a diet restricted to only meat and biscuits. His vitamin A levels were subnormal at 0.14 umol/L (reference range 0.9-2.5umol/l) and he was started on high-dose Vitamin A supplements.

Four months after supplementation retinal appearances had normalised, the rod ERGs recovered, nyctalopia and visual field restriction resolved. PVEPs had improved but an element of LE macular pathway dysfunction remained. Optic disc swelling settled leaving mild temporal pallor, particularly of the LE with some RNFL loss.

It is important to recognise nutritional Vitamin A deficiency in children as prompt recognition and treatment can improve symptoms, reverse retinal pathology which we have demonstrated with electrophysiological findings



MOG ASSOCIATED ENCEPHALITIS PRESENTING AS IDIOPATHIC INTRACRANIAL HYPERTENSION

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A young Caucasian male (7y) with normal BMI was atypical for his provisional diagnosis of Idiopathic Intracranial Hypertension (IIH), that resolved following a Lumbar Puncture (LP). At 8y he presented with a 2-week history of headaches and vomiting that started some weeks after flu vaccination and an upper respiratory infection.

Visual Acuity (VA) and colour vision were normal. Ocular motility was full. Fundoscopy and OCT showed recurrence of papilloedema, with enlarged blind spots on Kinetic perimetry.

LP opening pressure was 30cm H2O and CSF white cells were elevated (23). Repeat brain and spine imaging showed new white matter signal changes in keeping with neuroinflammation, as well as enhancement of the left optic nerve extending to the chiasm and optic tract. VA, colour vision and pupillary reactions remained normal.

Pattern VEP peak times were prolonged from the left eye compared to right eye to small check widths, consistent with relative macular-cortex pathway dysfunction. Hemifield PVEPs were slightly prolonged and reduced from the bitemporal fields indicating chiasmal dysfunction. Normal PERGs excluded PVEP delay associated with primary RGC disease.

Further investigations showed oligoclonal band and serum-MOG antibody positivity.

Management: Initial treatment with Acetazolamide 125mg bd for a week, following LP, was changed to IV methylprednisolone followed by oral prednisolone.

Symptoms improved significantly following LP and steroid treatment. He will be followed in a Demyelination Clinic.

MOG-associated disease has been reported with raised intracranial pressure and should be considered especially in children with atypical clinical phenotype for IIH.