

title = "Visual and oculomotor disorders", abstract = "Consensus has been reached on the concept that infantile cerebral palsy (CP) is a complex disorder which is limited neither to motor disability nor to the simple association between motor disability and possible disorders of other functions.

Andrea Guzzetta Eugenio Mercuri doi: Maturation of visual function in infants with neonatal brain lesions: Several studies have reported that various aspects of visual function, such as visual acuity, visual fields and optokinetic nystagmus are often impaired in infants with brain lesions of antenatal and perinatal onset. We report our experience with such infants and a more general review of the literature on the maturation of visual function in the first years after birth in normal and brain damaged children. Visual fields Introduction methods for assessing the maturing visual system in early childhood. In the last two decades, however, there have been considerable advances in our Visual impairment, due to damage of central visual understanding of the maturation of visual function pathways, is known as cerebral visual impairment, and in ways to assess it. It is now well accepted that and is frequent in children with brain lesions of the visual system functions mainly at a subcortical antenatal and perinatal onset. Most of the studies level in the newborn and in the first months after reporting the incidence of visual disorders in birth, and becomes progressively integrated with infants and children with such brain lesions have and dominated by cortical processes during the first been published in the last two decades. The first A major breakthrough occurred when it became includes papers describing the incidence of severe possible to assess acuity in the newborn by using visual problems in children with cerebral palsy. Now it is possible to assess aspects of The second group includes papers describing visual function such as, visual fields, attention at visual function in infants with brain lesions of distance, discrimination of colour or orientation onset in the first years of life. Longitudinal Until recently, most studies reporting visual studies have followed the onset and the maturation abnormalities in children with brain damage only of different aspects of visual function in normal used standard ophthalmological evaluation to pro- infants thus providing age-dependent normative vide a gross estimate of the more severe visual data. These tests do not require verbal skills or a abnormalities found in these children. This was great degree of collaboration from the child and mainly due to a lack of appropriate and specific can be used not only in small infants but also in Correspondence: A Guzzetta et al. Visual fields can be assessed using kinetic born and full-term infants with specific patterns of perimetry. The apparatus consists of two per- brain lesions which have been characterized using pendicular black metal strips bent to form two cranial ultrasound and magnetic resonance imaging arcs, each with a radius of 40 cm. The infant is MRI scans. A review of the incidence of visual held in the centre of the arc perimeter. During abnormalities in children with cerebral palsy and a central fixation of a white ball, an identical target more general review on the correlation between is moved from the periphery towards the visual abnormalities and other aspects of develop- fixation point along one of the arcs of the ment will be reported separately. Eye and head movements towards the peripheral ball are used to estimate the outline of the visual fields. Normative data for full-term and preterm infants are available. Optokinetic nystagmus can be elicited by using a niques. Blink reflex, in response to an approaching object, in response to the movement of the pattern in can already be observed in preterm infants,¹ and either direction. Normally, binocular optokinetic is mediated by a tactile sensorial input processed nystagmus is symmetrical from birth onwards, subcortically. The blink reflex is cortically mediated and usually develops 17 weeks after birth. Oculomotor behaviour can be assessed by testing fixation and following reactions. A short period of fixation on a target can be observed by 30 weeks post-menstrual age. At term age, new- borns are generally able to follow a target, such as a red ball, in a full arc. The presence of abnormal eye movements, such as spontaneous nystagmus, can also be noted. Strabismus and eye alignment can be tested by commonly used orthoptic techniques, such as the cover test. Acuity can be tested by using forced choice preferential looking. The infant is presented at eye level on one side of the midline with a target consisting of black and white stripes paired

with a uniform grey background on the other side. Example of testing visual acuity by means of Teller The level of acuity is measured as the finest acuity cards. Visual defects and neonatal brain lesions degrees. Figure 5 shows details of the maturation of various aspects of visual function in the first year of life. Incidence and type of visual impairment in preterm and full- term infants with specific patterns of brain lesions Fig. Example of testing visual fields by means of the kinetic perimeter. Periventricular leukomalacia whereas monocular optokinetic nystagmus shows Several studies have assessed visual acuity in a better response to stimulation in a temporonasal infants with periventricular leukomalacia. Acuity is generally normal in the infants with. A central target is used as a affected in infants with periventricular leukomalacia fixation stimulus before the appearance of the grades 3 and 4. While in some trials the ment in children with periventricular leukomalacia central target disappears simultaneously with is significantly associated with lesions in the the appearance of the peripheral target non- peritrigonal white matter, with the involvement of competition in others the central target remains optic radiation Fig. Normal chil- Visual impairment is common and generally dren can reliably shift their attention in a severe in infants with subcortical cystic leukoma- situation of non-competition during the first lacia,17,19,20,22 while it is less common in infants with weeks after birth, but brisk refixations in a cystic periventricular leukomalacia. Absent or delayed a latency of more than 1. Visual evoked potential can be recorded by using flash or orientation-reversal and phase-reversal stimuli. Using flash stimuli it is possible to follow the normal or abnormal maturation of the visual pathway. For orientation reversal visual evoked potentials, stimuli peri- odically change orientation between 45 and Fig. Stimuli used for testing fixation shift in infants. Stimuli used for testing phase shift and orientation reversal visual evoked potentials in infants. Graph illustrating the maturation of some visual functions in the first year of life. The difference may be explained by the mus or inferior colliculi, or to the bleeding of the distribution of the lesion, the mature infants having germinal matrix at the origin of the optic radiations cystic lesions that extend further into the subcor- and the posterior thalami. Therefore these effects tical white matter with subsequent higher risk of may be transient, as there is no major tissue abnormal visual outcome. Permanent effects are not so common Other studies have also demonstrated that, in as the parenchymal involvement with grade IV addition to visual acuity, other aspects of visual lesions is more often in mid-anterior parietal lobe function, such as visual fields and eye movements and not so often in sites affecting the primary are also frequently affected in these children. Visual defects and neonatal brain lesions Fig. Male infant aged 16 months with cerebral visual Fig. Male infant aged 2 weeks with delayed visual impairment. While infants with stage 1 abnormal. It is important to recognize that not all lesions involving the occipital lobes are associated with impaired visual function but in our experience infants who have lesions involving both the hemispheres and the basal ganglia putamen and caudate invariably have persistent and severe abnormalities of various aspects of visual function. Visual abnormalities may also be found in the first months after birth in infants with isolated basal ganglia lesions but these tend to recover by the end of the first year. Female infant age 15 months with cerebral visual Although visual acuity is generally normal in impairment. Unlike adults with similar lesions who show reciprocal connections between visual cortical a reliable association between the involvement of areas and basal ganglia. In other words damage to the subcortical structures may, by inhibiting a general exchange Visual impairment in infants with of information within the brain, preclude the possibility of functional reorganization of the brain lesions: Visual function can be abnormal in children with From our experience and from the review of the a normal ophthalmological examination and literature investigating visual function in infants normal optic radiations and visual cortex. This with brain lesions of antenatal and perinatal onset may be explained by the involvement of parts of it is clear that visual abnormalities are frequent in the brain other than geniculostriate pathways, infants with brain lesions but that the association such as the frontal or the temporal lobes, which between lesions in the visual pathway and visual are known to be associated with visual attention impairment does not always follow the rules or with other aspects of visual function. In some observed in adults with lesions involving the cases visual attention, and more generally visual visual pathway. The early use of neonatal brain function, may also be disturbed by other clinical MRI has given us considerable insight into

how problems often occurring in children with brain visual function is related to the site and extent of lesions, such as severe oculomotor impairment brain lesions that occur in the perinatal period. The or severe epilepsy. A proportion of infants show transient visual impairment with a gradual recovery⁴⁰ which, in. Visual abnormalities tend to be more frequent some cases, occurs in the first months after birth. This term is used to describe probably due to the low incidence of visual infants with reduced vision at birth, which abnormalities in the preterm infants with mild subsequently improves by the end of the first leukomalacia or with haemorrhages. Whilst in year of life. Delayed visual maturation can be preterm infants lesions of the occipital visual an isolated finding, or associated with ocular cortex are generally associated with impaired and neurodevelopmental abnormalities. The different extrageniculo-striate pathways,^{42,43} the restoration visual behaviour might be explained by the of normal excitability to the surviving neurons, difference in type and site of lesions, as in severe or the recruitment of neurons adjacent to the periventricular leukomalacia the lesions are lesion. One possible explanation is that the brain after the insult. In full-term infants the severity of visual tures causes delayed visual maturation in the impairment seems to be more related to the first months when visual function is mainly concomitant involvement of basal ganglia and subcortical, and that vision improves in parallel thalami. The role played by basal ganglia and with the onset of more mature aspects of cortical thalami in visual maturation is still not fully visual functioning. Further imaging studies in understood. The neuroanatomical basis for infants without obvious perinatal problems are these findings may result from the extensive needed to exclude the possibility that minor Review article: Visual defects and neonatal brain lesions lesions might also occur in that group and be 6 Atkinson J, Braddick OJ. Development of optokinetic responsible for the delayed visual maturation. Cognition and Visual Perception. Lawrence Erlbaum Associates, Visual evoked potentials disturbance of visual development in children with in infants and children. J Clin Neurophysiol ; 9: Early predictors of cerebral visual impairment in occurrence and extent of visual impairment, the infants with cystic leukomalacia. Neuropediatrics correlation between imaging findings and function ; Stepwise decrease in VEP born at term. This is probably due to the involve- latencies and the process of myelination in the ment of the complex extrastriate visual pathways human visual pathway. Brain Dev ; Effects of preterm extrauter- Future studies with larger cohorts of infants with a ine visual experience on the development of the human visual system: Dev Med wider variety of lesions and longer follow-up will Child Neurol ; Orientation- mechanisms underlying visual development. Orientation- reversal and phase-reversal visual evoked potentials Acknowledgements in full-term infants with brain lesions: Cerebral visual from the colleagues of the Visual Development Unit impairment in preterm infants with periventricular University College, London and the Paediatric leukomalacia. Pediatr Neurol ; Cerebral visual impairment in periventricular leukomalacia: Am J Neuroradiol ; Dev Med Child Neurol ; The spectrum of leukomalacia using cranial ultrasound. Behav References Brain Res ;

Chapter 2 : Andrea Guzzetta

Request PDF on ResearchGate | Visual and Oculomotor Disorders | Consensus has been reached on the concept that infantile cerebral palsy (CP) is a complex disorder which is limited neither to motor.

Francesca Tinelli Elisa Fazzi Epilepsia, 45 7: Several studies have reported behavioral Results: Twenty-two children had at least one abnormal re- and electrophysiological evidence of visual impairment dur- sult on one or more of the tests assessing visual function at T0. The underlying mech- Visual impairment at the spasm onset was related to the sleep dis- anisms are, however, poorly understood, and little has been re- organization rather than to the hypsarrhythmic pattern in awake ported about the correlation between visual impairment, EEG EEG. After 2 months, both EEG features become significantly patterns, and brain lesions. The aim of the study was to as- linked to visual function. Visual function improved in several sess visual function at the onset of spasm and 2 months cases after 2 months, in parallel with the seizure regression. Twenty-five infants with West syndrome were en- Conclusions: The study supplies new evidence of the involve- rolled and studied with a a full clinical assessment including ment of visual function in West syndrome. The presence of ab- a battery of tests specifically designed to assess visual function, normal visual findings in infants without lesions on brain MRI b a video-polygraphic study, and c brain magnetic resonance suggests that visual abnormalities are due not only to brain injury imaging MRI. Besides brain neuroimaging and EEG compar- but also to epileptic disorder per se. New insight is also provided ison with visual function, an intra-EEG analysis was performed into the possible mechanisms underlying clinical and EEG ab- to investigate the possible relation of EEG patterns to fluctuating normalities. West syndromeâ€”Visual functionâ€” visual behavior fixation and following. It has been previously reported that various aspects of The aim of this study was twofold: Visual abnormalities that can already be de- 2 months thereafter, and second, to correlate these find- tected by using behavioral and electrophysiological tests ings with EEG and magnetic resonance imaging MRI during the first weeks of the disease and even before 1â€”4 patterns. More specifically we wished to establish a the include poor visual responsiveness 5â€”8 , abnormal visual range and severity of visual abnormalities in West syn- evoked potentials 9,10 , and deficits in other aspects of drome; b the possible relation between visual abilities, visual function such as fixation shift 4. EEG patterns, and brain lesions; and c the role played The mechanisms underlying visual abnormalities, how- by the epileptic disorder in modulating the fluctuation of ever, are not fully understood, and little has been reported visual skills. The main clin- on an inborn preference for a pattern black and white ical data are reported in Table 1. As to the etiology of cases, gratings of different stripe widths over a uniform field, four were cryptogenic, four were associated with tuberous depicted on cards with decreasing stripe widths. Acuity sclerosis, and the remaining symptomatic patients had dif- values were expressed in minutes of arc or cycles per de- ferent causes: Visual fields were assessed by using for two patients, infectious disease for one, and a suspected kinetic perimetry, according to the technique described in metabolic disease for the last one. Oculomotor behavior was as- assessment at the spasm onset T0 and again, with the sessed by testing fixation, pursuit, and visual attention. This assessment was scored according to the criteria re- ported in Table 2. The global score, from 0 to 24, was the Clinical assessment result of the sum of the scores for each item. Clinical assessment included an ophthalmic examina- tion and a battery of tests specifically designed to assess Video-EEG visual function 11 in the first years of life. This method is based electrodes for the youngest infant, and deltoid surface Epilepsia, Vol. Hypsarrhythmic features were 1. Intra-EEG analysis of visual behavior fixation and scored according to criteria previously described. The visual performance was scored from 0 to 3 ac- cording to the performance level: In case of fluctuating visual performances, state hypsarrhythmia severe, moderate, mild, and ab- each score was considered in relation to the EEG sent and sleep organization normal and abnormal versus features during the period concurrent to the visual global neurovisual scores. Moreover, a multivariate anal- test. A hypsarrhythmia scoring system based on ysis was conducted by using the generalized linear model three features slightly modified from Kramer et

al. Each feature was scored on a scale performed during the EEG versus the three different EEG from 0 to 3 according to growing severity: Analysis of EEG in awake and sleep states. Visual function at T0 Relation of hypsarrhythmic awake EEG and sleep All patients had a normal ophthalmologic examination. Statistical analysis showed no significant and 22 had at least one abnormal result. Besides fixation and pursuit, In contrast, sleep patterns, in particular, spindle organization the most discriminating parameters in our series seemed adequate for age, were related to visual performances to be ocular motility and visual field: Seventeen patients at onset of the syndrome and 14 after 2 months had a fluctuation in the visual performances Visual function and brain MRI findings of fixation and pursuit concurrent with EEG monitoring. Both cryptogenic and tuberous sclerosis patients had Comparing the results of the visual test with three different generally good visual abilities, whereas the other symptomatic EEG parameters delta maximum voltage, delta frequency, multivariate mal visual findings. Only three 14, 18, and 24 of the 17 analysis generalized linear model analysis did not find cases in the latter group had normal results on visual tests, any statistical significance. EEG patterns and visual function Visual function after 2 months T1: Some improvement, in contrast, had a better association with visual improvement of visual function was observed Table 3. This findings, as infants with a recognizable sleep organization more frequently concerned those patients more often had good visual function. It is therefore difficult to make a fair comparison. Results showing that a disorder DISCUSSION of sleep EEG organization, rather than hypsarrhythmia This study confirms previous observations that visual during awake EEG, is associated at the onset of West syndrome with poor visual performance are not surprising, first appear 5,6,16, Conversely, acuity, visual field, and visual attention. However, the most severe and persistently related to low scores in visual function only after the first 2 months. This could be because transitory with lesions involving the basal ganglia, central and posterior cortical areas, as well as in those with optic radiation, drug-responsive infants with epilepsy is not generally associated with poor visual function. This can weaken the was not always related to the presence of brain lesions, statistical correlation between EEG pattern and visual function at T0. In such infants, the presence of abnormal visual findings also were common West syndrome. In contrast, a persistent hypsarrhythmia normal visual function suggests the possible effect of the at T1, like sleep disorganization, becomes another index epileptic disorder per se, as previous studies have reported of the severity of West syndrome. The possible variations of visual attention were related to EEG changes. Previous studies reported that visual impairment at the 6. Cognitive assessment of onset of West syndrome is often fluctuating, but although infants with West syndrome: Dev Med Child Neurol ; Cortical visual impairment in activity 24, more recent studies reported that it can also children with infantile spasms. Infantile spasms as a cause of acquired perinatal visual loss. We did not find any significant correlation between in- 9. Pattern reversal evoked ictal EEG features and performances of fixation and potentials in infantile spasms: The poor association might reflect the severity. Evoked potentials in infantile spasms. Even though one cannot exclude some cortical;9: Visual disorders in children with brain lesions, 1: Eur J found to be normal even in infants with severe holoprosencephaly;5: The acuity card procedure: Invest Ophthalmol Vis Sci function, depending on the integrity of ascending brain- ; It is not surprising that fixation and pursuit are abnormal in infants with years of life in healthy term newborns: West syndrome, as the disruption of sleep organization and Mohn G, van Hof-van Duin J. Development of the binocular and the dysfunction of ascending pathways with imbalance of monocular visual fields of human infants during the first year of sensitive inputs probably due to a brainstem disorder have life. Clin Vis Sci ;1: The poor correlation patterns and correlation with etiology and outcome. Sudden mental deterioration with convulsions in infancy. Arch Dis Child ; Early prognostic indicators with West syndrome and provide further insight into their of outcome in

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infants with neonatal cerebral infarction: Predicting favorable outcome in idiopathic West syndrome. This study was supported by a grant from Neonatal behavioral organization and 3-month visual discrimination. Visual abnormalities and oculomotor liability and short-term prediction of neurobehavioral status. Child cerebral EEG discharges: Colombo J, Horowitz FD. Behavioral state as a lead variable in 2. Preceding symptoms to neonatal research. J Child Health ; Okumura A, Watanabe K. Clinico-electrical evolution in pre- and quantification of infantile spasms. The visual evoked potential in neonates with occipital lesions and holoprosencephaly. Behavioral inattention in West syndrome. Visual inattention in West syndrome: Chaos, balance and development; thoughts on selected drome: Epilepsia ;31 suppl 3:

Chapter 3 : Visual and oculomotor disorders – Italian Ministry of Health

The issue of visual-perceptive development (and of its disorders) in CP must be viewed within this framework, also considering the central role it plays in the child's neuromotor, cognitive, and affective development, becoming the first tool for the interaction with the surrounding world.

Chapter 4 : Andrea Guzzetta – Research Output – Italian Ministry of Health

Cerebral palsy (CP) is known to be associated with disorders of the visual system including refractive errors, poor visual acuity, accommodative dysfunction, oculo-motor disorders, and cerebral visual impairment.

Chapter 5 : Visual disorders in children with brain lesions | Giovanni Cioni - www.nxgvision.com

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Chapter 6 : Visual and oculomotor disorders - CORE

Visual abnormalities that can already be detected by using behavioral and electrophysiological tests during the first weeks of the disease and even before include poor visual responsiveness, abnormal visual evoked potentials, and deficits in other aspects of visual function such as fixation shift.

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Very early upper limb interventions for infants with asymmetric brain lesions Boyd, R., Perez, M. & Guzzetta, A. Sep Cerebral Palsy in Infancy: Targeted Activity to Optimize Early Growth and Development.