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Impaired functional differentiation for categories of objects in the ventral visual stream: A case of developmental visual impairment

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ABSTRACT

We report the case of a 14-year-old girl suffering from severe developmental visual impairment along with delayed language and cognitive development, and featuring a clear-cut dissociation between spared dorsal and impaired ventral visual pathways. Visual recognition of objects, including faces and printed words, was affected. In contrast, movement perception and visually guided motor control were preserved. Structural MRI was normal on inspection, but Voxel Based Morphometry (VBM) revealed reduced grey matter density in the mesial occipital and ventral occipito-temporal cortex. Functional MRI during the perception of line drawings uncovered impaired differentiation which is normally observed at even younger ages: no local category preferences could be identified within the occipito-temporal cortex for faces, houses, words or tools. In contrast, movement-related activations appeared to be normal. Finally, those abnormalities evolved on the background of chronic bilateral occipital epileptic activity, including continuous spike-wave discharges during sleep, which may be considered as the primary cause of non-specific intellectual disability and visual impairment.

1. Introduction

Although infants already have substantial visual abilities (Gliga and Dehaene-Lambertz, 2007; Frank et al., 2014), full visual development requires extended tuning throughout childhood and into adolescence (Grill-Spector et al., 2008). The various components of the cerebral visual system, and the abilities which they underlie, follow specific developmental trajectories.

The ventral occipito-temporal "what" stream, devoted to the processing of intrinsic objects features (Milner and Goodale, 2008), develops over the first decade, with the progressive differentiation of a mosaic of cortical patches featuring category-selective specialization (for a review see (Grill-Spector et al., 2008)). Thus, while 5–8 year-old children already show well-defined activations in the lateral occipital complex (LOC) to objects, and in the parahippocampal place area (PPA) to places (Scherf et al., 2007), some studies show early preferential activations of the fusiform face area (FFA) to faces (Cantlon et al., 2011) while others do not (Scherf et al., 2007). The PPA then increases in size at least through the age of 11, in parallel with improved visual recognition

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of places (Golarai et al., 2007). Similarly, preferential activation to faces in the FFA positively correlates with age up to ages 12–16, in parallel with improved face recognition (Golarai et al., 2010). Finally, the acquisition of reading is associated with the development of the visual word form area (VWFA) in the left fusiform region (Dehaene and Cohen, 2011). The VWFA is already visible by the age of 7 (Gaillard et al., 2003; Monzalvo and Dehaene-Lambertz, 2013), and its further development parallels the improvement of reading skills (Turkeltaub et al., 2003; Ben-Shachar et al., 2007).

The dorsal occipito-parietal "where" stream, devoted to spatial processing and visual control of actions (Goodale and Westwood, 2004), also develops in an orderly fashion for motion perception and visual-motor control (review in (Braddick and Atkinson, 2011)). In particular motion perception, although present from the first weeks of life, develops steadily up to adolescence (Hadad et al., 2011), with evolving properties of the visual cortex, including area MT/V5 (Wattam-Bell et al., 2010; Rosander and von Hofsten, 2011).

Here we study the case of a young patient who suffered from a severe developmental impairment affecting the perception of visual objects while sparing motion perception and other dorsal stream functions. As compared to the few similar published cases (Ariel and Sadeh, 1996; Joy and Brunsdon, 2002; Eriksson et al., 2003; Gilaie-Dotan et al., 2009, 2011), anatomical and functional imaging allows us to relate this impairment to a probable lack of functional differentiation of the ventral visual stream, which resulted from abnormal electrical activity and cortical development in occipital and temporal visual regions.

2. Material and methods

2.1. Case report

When she first consulted in the neurology department, MJ was a 12-year-old right-handed girl, the first child from genetically unrelated parents. She was born at term (41 wGA) following an eventless pregnancy (birth weight = 2.800 kg (10 p), birth length = 48 cm (−1 SD), birth cranial perimeter = 32.5 cm (−2SD). Her neurological development was considered to be normal (e.g. walking acquired at one year, language before two years). Difficulties with drawing were first noticed at the age of four, associated with some behavioral disorders, especially agitation.

At the age of six, she presented a first episode of nocturnal clonic convulsions. Clinical examination did not reveal any neurological deficit. However, because of her difficulties at school, a qualitative language assessment was carried out. It revealed global difficulties, with a clear-cut dissociation between better oral abilities and severely impaired graphic abilities, affecting both drawing and writing. Cerebral CT-scan was normal. Repeated EEG recordings showed epileptiform activity, consisting in series of spikes lasting from 5 to 10 s, localized in the occipito-temporal region, with variable left, right, or bilateral lateralization. Those anomalies were sometimes, but not always, associated to clinical signs of absence seizure. A treatment with sodium valproate was introduced, and later associated with ethosuximide. Despite this association, the parents reported the persistence of frequent absence seizures, plus occasional clonic seizures.

At the age of eleven, an MRI-compatible vagal nerve stimulation device was implanted (Cyberonics; VNS therapy Guidelines, 2006), leading to a decrease of seizures. Yet, MJ suffered a clonic seizure a few months later, followed by transient confusion, and by more enduring post-ictal psychosis (paranoid delusions with beliefs relative to black magic, auditory and visual hallucinations) (Devinsky, 2008). She was hospitalized in a psychiatry department. Symptoms resolved entirely within less than two months, but were followed by catatonic symptoms which eventually recovered with lorazepam. No other psychotic or catatonic symptom occurred ever since. At the age of twelve, seizures stopped and ethosuximide was discontinued.

It was during her stay in the psychiatry department at the age of 11 that her major visual impairment attracted medical attention, on the basis of MJ's spontaneous every day behavior. Remarkably, the impairment seemed to affect only some aspects of her visual abilities. She was not able to recognize the most usual objects without touching them, and she identified familiar persons only when they started speaking. Actually, due to those difficulties, she had been admitted two years earlier in a special school for blind children.

In striking contrast with her massively impaired recognition abilities, she was able to make bead necklaces, or to play video games involving moving shapes of different colors. She moved around easily even in unfamiliar places, without bumping into obstacles and was able to skillfully skate in the corridors of the hospital, avoiding chairs, ashtrays or tables. She did not report any subjective deficits in visual motion perception.

Despite some difficulties at maintaining central fixation and at inhibiting saccades to peripheral stimuli, a Goldmann kinetic perimetry was performed, showing no visual field defect. She was submitted to the CADET test of visual acuity (Douche and Badoche, 1987), a method based on matching drawings of objects, and hence possibly underestimating her elementary acuity. She scored 4/10 (i.e. an abnormal score) from a far distance (2.5 m) and 8–10/10 (i.e. a normal score) from "reading" distance, suggesting a dissociation between somewhat impaired far distance and preserved near distance visual acuities.

MJ's navigation abilities and her skill with motion-based video games suggested preserved motion perception. This was confirmed when we had an opportunity to perform the Leuven Perceptual Organization Screening Test (L-POST) when MJ was 17 (Torfs et al., 2014; www.gestaltrevision.be/tests). This battery includes 15 subtests, including 3 subtests designed to assess motion perception: kinetic object segmentation, biological motion, and global motion detection. MJ performed normally on the 3 motion subtests, as compared to age-matched healthy participants. By contrast, she was impaired below the 10th percentile in 10 out of 12 shape-oriented tests. Three years elapsed between the bulk of data reported here and the L-POST test. Naturally, MJ was not subject to any rehabilitation of motion perception, and we may assume that her motion vision had not changed in the interval.

Furthermore, MJ showed no clinical signs of ocular apraxia, optic ataxia, astereognosis, right–left confusion, body schema impairment or autotopoagnosia.

The present study was carried out when MJ was 12 to 14-years-old. MJ had been seizure-free since the age of 12. When she was 12, an anatomical MRI scan was performed on a 1.5 T magnet, including T1, FLAIR, diffusion, and T2* sequences. Images were visually analyzed by a senior neuroradiologist blind to MJ's condition, and were considered to be entirely normal, particularly in the visual cortex. PET-scan did not show any abnormalities of cortical perfusion, notably in occipito-temporal regions. Due to alleged minor dysmorphic signs (large forehead, small ears and big mouth), MJ underwent karyotype screening, chromatography of amino acids and organic acids, study of purine metabolism, assessment of lactic acid, pyruvate and ammoniemia. All results were normal. The *FMR1* mutation, the most frequent microdeletions, and congenital disorders of glycosylation were all absent.

MJ underwent several 24-h-long and standard EEG recordings, using 21 scalp electrodes (10–20 system; MR95 Oxford Médelec FMB system). Despite the absence of seizures, there was still bilateral epileptiform activity in the occipito-temporal regions when