Visu-spatial recognition in Williams syndrome: Dissociative performance in non-motor tasks?
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Williams syndrome (WS) is a rare neurodevelopmental disorder (1:20,000), caused by a submicroscopic deletion in the band q11.22-23 of chromosome 7. The WS patients have a unique cognitive phenotype (Bellugi et al, 2001 Clinical Neuroscience Research 1 217–229), classified as mildly to moderately retarded (mean IQ is 55, with a range rarely reaching above 50) associated with generalised difficulties in general problem solving, arithmetic, and typically unable to achieve fully independent living. They also present an unusual socio-emotional and personality attributes, characterised by an excessive sociability. Despite their low IQs, individuals with WS display characteristic patterns of cognitive performance with peaks and valleys of abilities. Specially striking is a well-documented dissociation between relatively spared linguistic abilities and severely impaired visuo-spatial cognition, which is disproportionately impaired (particularly at the level of global organisation). However, there are incongruities within spatial cognition, where WS subjects display preserved areas, with face-processing abilities being a remarkably strong area of performance (Farran et al, 2001 Journal of Child Psychology and Psychiatry 42 719–728). Using two spatial perception recognition tasks (Benton's line orientation test and Benton's test of facial recognition), nonverbal, and both requiring processing pictures (different stimuli—lines and faces), but without involving visuo-constructive abilities, we studied the performance of Williams syndrome subjects. In order to do this, we evaluated a Williams syndrome group (N = 8) in these specific non-motor perceptual tasks to study the referred high performance of individuals with WS on face-processing tasks, despite their severe impairment on the other visually based cognitive tasks.

Substantial loss of chromatic contrast sensitivity in subjects with age-related macular degeneration
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In subjects with normal vision, chromatic contrast sensitivity continues to increase as spatial frequency decreases. When contrast is expressed in terms of combined cone contrast, red—green chromatic sensitivity at a low spatial frequency of, for example, 0.4 cycle deg−1 is higher than luminance contrast sensitivity by a factor between 6 and 10, depending on age (and increasing for still lower frequencies). In the yellow—blue direction, the factor is between 1 and 3. This means that for low spatial frequencies one needs far less modulation of the cones in order to reach a certain chromatic threshold than to detect an equally small achromatic difference. Small differences in colour therefore represent an effective cue for detection and discrimination of objects in normal vision. However, in the case of visually impaired subjects with age-related macular degeneration (AMD) the situation is different. We have addressed this issue as part of a larger study of visual function and AMD. For a selected group of twelve elderly subjects with AMD (mean age 75 years) and no other diseases affecting vision, we have compared achromatic and isoluminant chromatic contrast sensitivities as a function of spatial frequency of sinusoidal gratings. Variability was large between subjects, but, in the AMD group, sensitivity to achromatic contrast was generally less severely affected at low than at high spatial frequencies. Relative to an age-matched control group, the group-average achromatic sensitivity at 0.4 cycle deg−1 was one-third of the normal value. Sensitivity to red—green and yellow—blue contrasts of the same spatial frequency was on average only about one-tenth of the normal age-matched sensitivity. This implies a more dramatic loss of chromatic than of achromatic vision at low spatial frequencies in AMD.

Impairments of colour contrast sensitivity thresholds in cases of damage of chiasma opticum
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The purpose of our study was to estimate how the narrowing of outer visual field sectors affects colour contrast sensitivity. Correlation was made between colour contrast sensitivity thresholds

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