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Unique Profile of Visuo-perceptual Skills in a Genetic Syndrome

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Williams syndrome (WS) and Down syndrome (DS) are genetic disorders with characteristic neuropsychological profiles. Subjects with WS show surface similarities to subjects with right hemisphere damage (RHD) in their relative preservation of linguistic skills, their poor visuo-*constructive* skills, and their hierarchical processing biases. Ten adolescents and young adults with WS and nine matched subjects with DS were administered a battery of visuospatial *perceptual* tasks to test whether the profile of performance in WS would resemble that in RHD. It was found instead that the WS subjects showed a distinctive clustering of skills, with particular preservation of facial discrimination, but impairment of other perceptual skills. Subjects with DS showed a more homogeneous profile. The WS profile may map onto the functional dichotomy between ventral and dorsal visual pathways in the cerebral cortex.

INTRODUCTION

Down syndrome (DS) is a leading genetic etiology of mental retardation and is a well-known subject for both medical and psychological research (Lott & McCoy, 1992). Williams syndrome (WS) is a less well-known genetic syndrome affecting multiple organ systems and causing mild to moderate mental retardation (Jones & Smith, 1975; Morris, Demsey, Leonard, Dilts, & Blackburn, 1988). With an estimated incidence of 1 in 25,000 live births, WS is far more rare than DS. Supravalvular aortic stenosis (an otherwise rare congenital cardiac malformation), infantile hypercalcemia, and the unique facial appearance constitute definitive diagnostic evidence of WS.

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The etiology of WS recently has been identified as the deletion of one copy of the elastin gene and possibly of genes contiguous to elastin (Ewart et al., 1993).

Recent neuropsychological interest in WS and DS stems from their distinctive cognitive profiles. A large program of studies comparing matched subjects with DS and WS has shown important domain-specific neuropsychological differences between these groups (Bellugi & Wang, 1992). To wit, studies employing specific linguistic probes have shown better preservation of linguistic skill in WS than in DS, despite comparably poor performance on broad cognitive measures such as verbal IQ (Bellugi, Bihrle, Jernigan, Trauner, & Doherty, 1990; Bellugi, Bihrle, Neville, Jernigan, & Doherty, 1992; Bellugi, Wang, & Jernigan, 1994). The spontaneous speech of adolescents and young adults with WS is fluent and displays a wide variety of grammatical forms, including passives and embedded relative clauses. Furthermore, subjects with WS but not DS have been found to perform well on a number of metalinguistic tasks, such as correcting ungrammatical sentences.

In contrast to their linguistic facility, subjects with WS have been reported to display significant visuo-constructive impairments (Bellugi et al., 1990; Dilts, Morris, & Leonard, 1990; MacDonald & Roy, 1988; Pagon, Bennett, LaVeck, Stewart, & Johnson, 1987). Bellugi et al. (1994) have begun to illuminate a specific pattern to these impairments. As part of the same program of investigation, the WS and DS subjects were tested on the Block Design subtest of the Wechsler Intelligence Scale for Children—Revised. According to standard scoring criteria, the two groups performed equally poorly. However, closer scrutiny of their responses revealed striking differences. While subjects with DS typically maintained the global configuration of the blocks (a 2×2 square) but failed to replicate the internal pattern of the design, subjects with WS failed to maintain the global organization of the blocks. Instead, they placed the four blocks in apparently haphazard, even noncontiguous arrangements (Fig. 1A). The WS and DS errors are reminiscent of those seen in adult subjects with right and left hemisphere damage, respectively (Ben-Yishay, Diller, Mendleberg, Gordon, & Gerstman, 1971).

In order to examine experimentally the processing of hierarchical stimuli, Bihrle et al. (1990) tested the same WS and DS subjects on copying stimuli which had clearly defined global and local featural levels. Both groups again performed poorly, but in very different ways (Fig. 1B). Subjects with WS typically produced only the local forms, sprinkled across the page, and were impaired at reproducing the global forms. Subjects with DS showed the opposite pattern: they tended to produce the global forms without the local forms. The WS pattern once more resembled the pattern seen in adults with right hemisphere damage (RHD) (Delis, Kiefner, & Fridlund, 1988). As Bihrle et al. have suggested (1989) the relative preservation of linguistic function in WS, the poor visuo-constructive ability, and the local bias in hierarchical processing appear similar to the pattern found in RHD. In con-

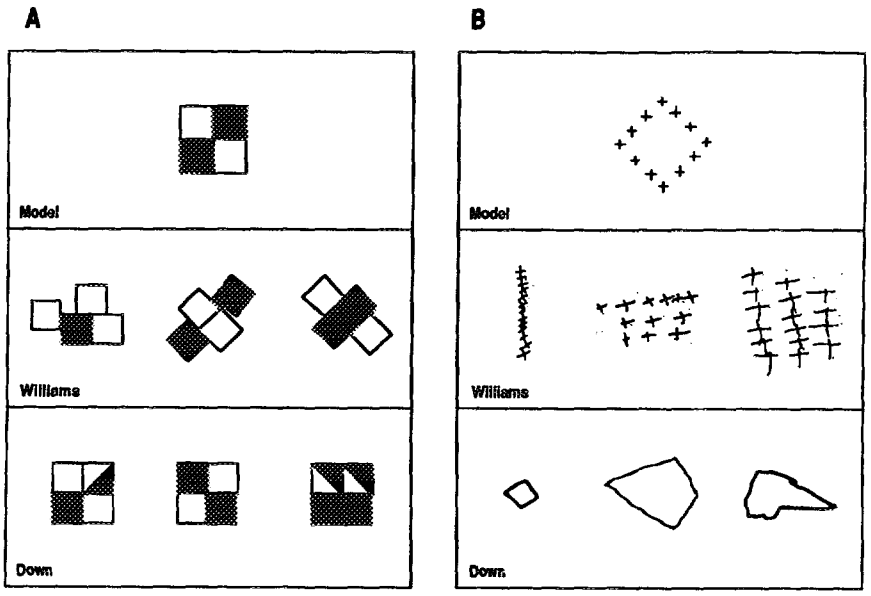

FIG. 1. Contrast between Williams and Down syndrome subjects on visuo-constructive tasks. Characteristic error patterns are evident on the Block Design subtest of the Wechsler Intelligence Scale for Children—Revised (A), and on the Delis Hierarchical Processing Task (B).

trast, the impairment of language abilities and the global hierarchical bias found in DS are consistent with the pattern seen in left hemisphere damage (LHD).

The current study was designed to further explore visuospatial abilities in WS and DS, focusing on visuo-perceptual rather than visuo-constructive task performance. The same WS and DS populations that were tested above, were tested now on a battery of visuospatial perceptual tasks. While both cerebral hemispheres contribute to visuospatial processing, the tasks administered here were chosen for their greater susceptibility to damage of the right hemisphere than of the left. This allowed us to compare the neuropsychological profiles resulting from genetic perturbations of the neurodevelopmental process with the profiles resulting from lateralized brain lesions in the mature adult.

METHODS

Subjects

Ten subjects with WS (5 female, 5 male) and 9 subjects with DS (6 female, 3 male) had been recruited for a multidisciplinary study of the neurobiological bases of language and cognitive

development (Bellugi et al., 1990, 1992). The diagnoses of DS had been confirmed by karyotype examination. The WS diagnoses had been established by pediatric geneticists, based on the characteristic constellation of major stigmata. WS and DS subjects were in comparable special educational placements for the "educable mentally retarded," and the groups were matched on age (WS: 15.7 ± 3.1 (mean ± *SD*) (range 11–18 years); DS: 15.0 ± 2.6 (range 11–20)) and full-scale IQ (WS: 48.9 ± 7.1, DS: 50.0 ± 7.9). As part of the larger study, magnetic resonance images (MRIs) of the brain had been obtained for 7 of the subjects with DS and 8 of the subjects with WS (Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993). Routine clinical interpretation of the MRIs revealed no focal lesions or focal atrophy in any of the subjects. One female subject with DS showed mild generalized atrophy. (The results of morphometric MRI analyses are discussed below.)

Tasks

In order to confirm previous findings of visuo-constructive impairment, the Developmental Test of Visuo-Motor Integration (VMI) was administered to both WS and DS groups. It is a standardized, normed measure which requires subjects to copy 24 drawings of increasing difficulty. Drawings were marked correct or incorrect according to standard criteria (Beery, 1982). One point is awarded for each correct item until three consecutive items are failed.

The battery of visuo-perceptual measures included tests of visual neglect, judgment of line orientation, visual closure, face discrimination, orientation-invariant object identification, and a broad-based measure of overall visuo-perceptual abilities (see Table 1). All are known to be more sensitive to right hemisphere lesions than to left (Benton, Hannay, & Varney, 1975; Bruyer, 1986; Warrington & Taylor, 1978; Wasserstein, Zappulla, Rosen, Gerstman, & Rock, 1987). Visual neglect was tested with Albert's Test of Visual Neglect (Albert, 1973). On this test, subjects are asked to cross each of 40 lines which are widely distributed across a sheet of paper. Performance was assessed qualitatively, to determine whether there was consistent failure to cross lines located on only one side of the sheet.

As a measure of overall visuo-perceptual abilities, the Motor-Free Visual Perceptual Test (MVPT) was administered (Colarusso & Hammill, 1972). It is a standardized, normed measure used frequently for educational placement. Its 36 items are divided into six subtests. These include Baseline (matching target to an identical choice stimulus), Figure-Ground Discrimination (choice stimulus embedded in another figure), Spatial Relationships (choice stimulus transformed in color, size, or rotation), Visual Memory (target and choice stimuli presented sequentially rather than simultaneously), and Visual Closure (choice stimulus is an incomplete version of target). The final subtest, Visual Discrimination, requires the subject to discern which of four stimuli is nonidentical to the other three. The Benton Judgment of Line Orientation (Benton, Hamsher, Varney, & Spreen, 1983) also was administered. It requires subjects to choose which of eleven lines have the same orientation as two target lines. Two consecutive correct answers on the 5-item pretest are required before the subject is administered the 30-item test.

Three tasks were administered to test visual closure. The Gestalt Closure subtest of the Kaufman Assessment Battery for Children (Kaufman & Kaufman, 1983) requires the subject to name or describe an incomplete inkblot drawing. It is a descendant of Street's Gestalt completion test (Street, 1931), although it uses entirely new stimuli. The 25 stimuli are ordered by difficulty level, and administration is continued until all items at a level are failed. The Mooney Closure Faces test (Mooney, 1957) uses extremely high contrast pictures of faces, containing only black (shadow) and white (highlight). Subjects must classify each picture as "girl," "boy," "grown-up woman," "grown-up man," "old woman," or "old man." All 51 items were administered to each subject. Some items had two correct responses, according to Lansdell's revised criteria (1968). Finally, the Anomalous Contours test (Hamsher, 1978) consists of 15 figures whose boundaries are illusory (after Kanizsa, 1976). These figures are

formed by the mental interpolation of imaginary boundaries which depend on the conformation of other figures. One point is awarded for each figure that the subject can either trace or name.

Three tests which draw on object identification abilities were administered: the Benton Test of Facial Recognition, the Inverted Faces task, and the Canonical-Noncanonical Views test. On Benton Faces (Benton et al., 1983), subjects must decide which one or three of six choice faces, shown in different lateral rotations or with different shadowing, match an upright target face. To create the Inverted Faces task (Doherty & Bellugi, 1990), the Benton Faces test was modified by turning the six-choice faces upside-down. This manipulation is known to interfere with the specialized processing of faces (see Diamond and Carey, 1986, for a discussion). Because pilot testing showed fatigue effects, the "short form" (13 items, maximum score 27) of each test was employed. Short-form scores are highly correlated with full-test scores ($r \geqq 0.88$) in normal and brain-damaged populations (Benton et al., 1983). The Canonical–Noncanonical Views test (Carey & Diamond, 1990) is an object naming test, developed as a child-version of a similar test by Warrington and Taylor (1973). It consists of two subtests of 25 pictures each. In the first subtest, objects are shown in noncanonical orientations, e.g. a teapot viewed from above. The same 25 objects are depicted in the second subtest, from a canonical perspective, e.g. a teapot viewed from the side. The Noncanonical Views score was the percentage of objects correctly named out of those which were named correctly in the Canonical subtest.

Procedures

Each subject was tested individually over a several-month period. Due to subjects' scheduling exigencies, the testing period was longer for some than others. However, the WS and DS groups did not differ in age for any given measure. The tasks were interspersed with other cognitive and linguistic tasks. Subjects were carefully instructed to ensure that the task demands were understood. On the Mooney test, for example, subjects were drilled by asking them to classify their friends, relatives, and members of the testing laboratory as "girl," "boy," "grown-up man," etc., as required in the test itself.

Analysis of variance (ANOVA) procedures were used for statistical comparisons. A one-way ANOVA was used to compare WS and DS performance on the single visuo-constructive test (VMI). Two multivariate ANOVAs (MANOVA) were performed to compare visuo-perceptual test scores. A 2 group × 6 subtest MANOVA was used to compare the groups on the MVPT. A second 2 group × 6 task MANOVA was used to compare performance across the other visuo-perceptual tasks. For both MANOVAs, analysis of the raw scores failed to satisfy the requirement for multivariate homogeneity of the dispersion matrices. Therefore, arcsine transformations of the percent correct scores were used for analysis. Test scores also were examined for sex effects by ANOVA, and for age effects by Kendall rank correlation.

RESULTS

A summary of group mean scores by task is provided in Table 1. Results from the VMI confirmed the previously reported visuo-constructive impairments in both WS and DS. While these groups have not shown quantitative differences on other tests of visuo-constructive ability, and the VMI performance of both groups was depressed relative to chronological age, the DS group did score significantly better than the WS group ($F(1, 17) = 18.11$, $p < .01$). No significant effects of age or sex were found, either across groups or within groups, on the VMI or on any other test administered. Qualitatively, the WS responses on the VMI again showed an impairment in global coher-

TABLE 1
Visuospatial Performance in Williams and Down Syndromes

Tasks	Scores (mean ± *SD*)		
	Williams	Down	(Possible)
Visuo-constructive ability			
Visual motor integration*	7.5 ± 1.3	10.8 ± 2.0	24
Neglect			
Albert's visual neglect	(No evidence of neglect)		(No omissions)
General perceptual ability			
Motor-free visual perception	25.5 ± 4.9	24.4 ± 5.4	36
Line orientation			
Benton line orientation	(Most failed pretest)		30
Visual closure			
Gestalt closure	17.1 ± 4.7	14.9 ± 3.1	25
Mooney closure faces	36.2 ± 5.2	33.8 ± 5.4	51
Anomalous contours	12.3 ± 2.4	13.8 ± 1.5	15
Face and object recognition			
Benton faces*	21.8 ± 2.1	14.7 ± 2.5	27
Inverted faces	15.6 ± 3.1	13.2 ± 2.5	27
Canonical views	23.3 ± 1.1	22.7 ± 1.3	25
Noncanonical views**	75.9 ± 10.2%	66.4 ± 8.9%	100%

* $p < .01$, Williams syndrome vs. Down syndrome.
** $p < .05$, Williams syndrome vs. Down syndrome (see text for scoring method).

ence. Although these subjects may correctly depict the requisite components of each figure, these "local" features were not oriented correctly with respect to each other (Fig. 2). Impairments in hierarchical organization were not found in the DS group.

On Albert's Test of Visual Neglect no subject showed consistent, lateralized failure to cross lines. On the Benton Judgment of Line Orientation, only two subjects with WS and one with DS passed the pretest. On the test proper, these three subjects all scored in the range considered "severely defective" for adults (Benton et al., 1983). No further analysis was performed on these data.

Overall visuo-perceptual ability, as assayed by the MVPT, was impaired but closely matched in WS and DS. (One subject with DS refused to complete the last two subtests of the MVPT. For statistical purposes, he was assigned a score equal to the mean for all other subjects on those subtests.) The 2 group × 6 subtest MANOVA showed no difference between WS and DS groups (Hotelling's $T^2 = 0.29$, exact $F(6, 12) = 0.58$, $p > .05$).

Despite the close matching of DS and WS performance on the MVPT and on full-scale IQ, analysis of the other visuo-perceptual results was enlightening. A 2 group × 6 task (Benton Faces, Inverted Faces, Noncanonical Views, Gestalt Closure, Mooney Closure, and Anomalous Contours) MANOVA showed a significant difference between WS and DS groups (Hotelling's $T^2 = 3.99$, exact $F(6, 12) = 7.98$, $p < .01$). Univariate statistics

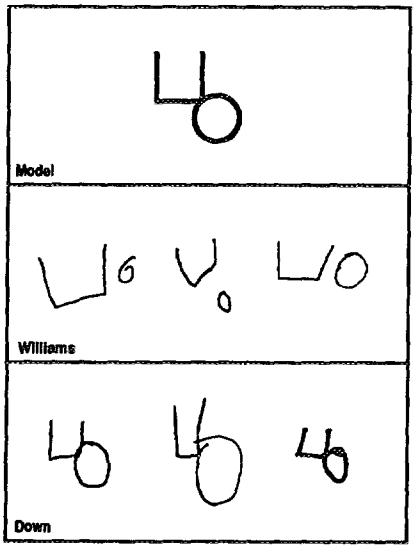

FIG. 2. Contrast between Williams and Down syndrome subjects on the test of visuo-motor integration. On a standardized copying task, Williams and Down syndrome subjects showed the same pattern of hierarchical processing biases that they showed on other visuo-constructive tasks.

did not show differences on any of the tests of visual closure (Gestalt Closure, $F(1, 17) = 1.88, p > .05$; Mooney Closure Faces, $F(1, 17) = 1.00, p > .05$; Anomalous Contours, $F(1, 17) = 3.20, p > .05$). However, significant contrasts were found on the face and object identification tasks. First, subjects with WS performed much better than those with DS on Benton Faces ($F(1, 17) = 41.23, p < .001$). Benton Faces raw scores showed almost no overlap between WS and DS groups. Subjects with WS also performed better on Noncanonical Views ($F(1, 17) = 4.97, p < .05$). Both groups averaged about 23 items correct on Canonical Views ($F(1, 17) = 1.34, p > .05$), suggesting that the Noncanonical results are not attributable to group differences in naming ability for the items pictured. Finally, the WS group showed a trend toward better performance on Inverted Faces ($F(1, 17) = 3.47, p < .10$). Qualitatively, subjects with WS seemed to answer more quickly and to perform with greater facility than subjects with DS on all three object identification tasks. The WS and DS group profiles are contrasted in Fig. 3.

DISCUSSION

The contrasts found between WS and DS show that profiles of visuospatial task performance may differ significantly despite comparable impairments of general cognitive ability and comparable performance on broad perceptual

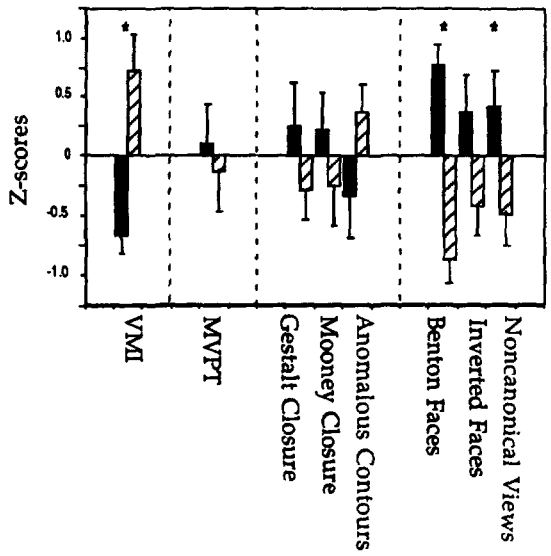

FIG. 3. Profile of visuospatial performance in Williams and Down syndromes. Average group performance is shown for a visuo-constructive task (VMI), a general measure of visuospatial perception (MVPT), three tests of visual closure (Gestalt Closure, Mooney Closure Faces, and Anomalous Contours), and three tests of object identification (Benton Faces, Inverted Faces, and Noncanonical Views). (For the purposes of illustration only, average Z scores were calculated for each group, based on the pooled results across groups. Graph shows averages ± standard errors of the mean.) (■) Williams and (❐) Down syndromes. *Significant group differences (see text for details).

measures such as the MVPT. The WS performance profile was distinguished most by a peak on Benton Faces, where the WS group mean fell squarely in the middle of the "average" range for normal adults (Benton et al., 1983). By comparison, both groups performed in the range of pre-school and young school-aged children on the VMI, MVPT, and Gestalt Closure tasks, and in the "severely defective" range for adults on Benton Line Orientation, according to published norms (Beery, 1982; Benton et al., 1983; Colarusso & Hammill, 1972; Kaufman & Kaufman, 1983). The WS profile also showed relative strength on Noncanonical Views, while the DS profile showed relative strength on the visuo-constructive task, the VMI. Each of these genetic syndromes thus is associated with a performance profile that is distinct from the other and, in the case of WS, is distinct from the profile found at any normal developmental age.

The strong performance of WS subjects on Benton Faces distinguishes them from the RHD population as well. Although both cerebral hemispheres may contribute to the processing of face stimuli, studies of brain-damaged subjects and of normal controls suggest that the role of the right hemisphere

is critical (Bruyer, 1986). This WS strength also stands in accord with the extreme interest in and attentiveness to faces that have been reported for children with WS (Bertrand, Mervis, Rice, & Adamson, 1993).

Spontaneous comments by some WS subjects suggest that they were using a feature-matching strategy on Benton Faces, rather than attending to the global configuration of the faces. If that were the case, the WS subjects might have been expected to show an advantage on Inverted Faces as well. Though such a trend was seen, the results did not achieve statistical significance. This may have resulted from inadequate statistical power or floor performance in the DS group or, alternatively, from the mental rotation requirement of the Inverted Faces task. (Recall that the six choice faces were upside-down, but the target face was upright.) Some of the Benton Faces stimuli also are shown in slight rotation, but on a different axis and to a much lesser extent than for Inverted Faces. Although some contend that mental rotation is required also for Noncanonical Views, this argument is disputed (Farah & Hammond, 1988) and there is no explicit demand for rotation. Mental rotation abilities have not been studied in WS and DS.

Wasserstein et al. (1987) have argued that the right hemispheric dependency of visual closure tasks may be most specifically tested by the Anomalous Contours test. No group differences emerged on this or the other tests of visual closure, despite their varying stimulus characteristics and task demands. Previous reports from the focal lesion literature suggest that performance on the Mooney tends to group with performance on other closure tests rather than with other tests of facial perception (Wasserstein et al., 1987), and the results here show a similar pattern. On the single test for which published norms are available (Gestalt Closure), both syndromic groups performed in the range of young school-aged children. The results therefore suggest that the WS and DS impairments on visual closure are roughly commensurate with their general cognitive impairment.

The inadequacy of an RHD model of cognitive abilities in WS is reflected by the remarkable WS performance on Benton Faces and on Noncanonical Views, and the absence of evidence for a neglect syndrome. Recent studies of semantic and discourse skills in WS also provide evidence that the surface similarities between the WS and RHD profiles are misleading. On tests of semantic fluency, subjects with WS perform very well, and much better than those with DS (Bellugi et al., 1994). In RHD, semantic fluency is typically impaired, despite the preservation of fluency in spontaneous speech and on tasks of phonemic fluency (Laine & Niemi, 1988). WS subjects also show well-preserved discourse skills and use a rich variety of linguistic affective devices, sometimes in exaggerated abundance. For instance, Reilly, Klima, and Bellugi (1991) found on a picture book storytelling task that WS subjects used a wealth of affective prosody (in phrases such as "*Froggie,* come *back* again") and semantic devices ("Well, *what do you know*? A frog family!" "*Gadzooks!* The boy and the dog start flipping over.") for the purpose of

conveying affective meaning. The WS stories also were much more cohesive and well structured. Studies of adults with focal brain lesions have shown that these skills also depend on right hemispheric integrity (Wapner, Hamby, & Gardner, 1981).

Morphometric studies using MRI brain scans from the same WS and DS subjects as those studied here, have shown that despite overall cerebral volume reductions in both syndromes, there is no lateralized pattern of cerebral malformation in either. Instead, each shows a different but characteristic pattern of regional preservation of brain volume (Jernigan & Bellugi, 1990; Jernigan et al., 1993; Wang, Doherty, Hesselink, & Bellugi, 1992a; Wang, Hesselink, Jernigan, Doherty, & Bellugi, 1992b). These *in vivo* neuroimaging studies, as well as neuropathological studies of a recently-obtained postmortem WS brain (Galaburda, Wang, Bellugi, & Rossen, 1994), are beginning to suggest that the performance profiles described here may be related to parietal lobe abnormalities.

One alternative to the right–left division of higher cortical function may be drawn from the distinction between ventral and dorsal visual pathways in the cerebral cortex (Jernigan et al., 1993). Animal studies have elucidated the distinct anatomic and psychophysical properties of these pathways, and researchers have begun to relate these pathways to human neuropsychological competence. (See Livingstone and Hubel (1988) for a review.) While the two pathways do interact, it is hypothesized that the ventral pathway mediates orientation-invariant object identification, while the dorsal pathway is responsible for motion and depth perception and visual closure. The WS group's superior performance on Noncanonical Views and Benton Faces on the one hand, versus their performance on tests of visual closure and line orientation on the other, maps onto the ventral/dorsal dichotomy. Their bias for local processing of hierarchical stimuli similarly may be consistent with that model (Badcock, Whitworth, Badcock, & Lovegrove, 1990; Hughes, Fendrich, & Reuter-Lorenz, 1990).

Further neuropsychological studies are required to specify the fundamental cognitive processes underlying facial and object perception in WS and other facets of the WS and DS profiles of visuospatial performance. Analyses of the covariance of performance on different cognitive probes will provide insight into the relationship of various visuospatial abilities to each other and their dissociability from general cognitive skill. Their covariance with cerebral morphometrics will provide one strategy to define the neurological substrates of visuospatial cognition.

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