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Visuospatial Construction

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The ability to see an object or picture as a set of parts and then to construct a replica of the original from these parts is known as visuospatial constructive cognition. Examples of visuospatial construction include drawing, buttoning shirts, constructing models, making a bed, and putting together furniture that arrives unassembled. Visuospatial construction is a central cognitive ability. At the same time, there are enormous individual differences among people in their ability to perform visuospatial constructive tasks. Some individuals draw extremely well; others cannot draw at all. Some people can copy complex patterns accurately and rapidly; others can copy accurately but slowly; still others can copy only simple patterns or none at all. The importance of visuospatial construction for everyday life, coupled with the wide range of ability shown by individuals of the same age, has led to the inclusion of measures of visuospatial construction on virtually every full-scale assessment of intelligence.

In addition to the wide range of ability evidenced in individuals with normal intelligence, the phenotype of at least one neurodevelopmental disorder (Williams syndrome) includes a hallmark weakness in visuospatial construction. In this report, we review what is known about a possible genetic contribution to visuospatial constructive ability. The remainder of the report is divided into four parts. In the first, we consider, in brief, general intelligence (*g*) and spatial intelligence, with a focus on individual differences in visuospatial constructive abilities of people with normal intelligence. With this information as background, the second and third sections focus on the visuospatial constructive abilities of individuals with Williams syndrome or small deletions in the Williams syndrome region; we conclude that there is a specific genetic basis for the extreme difficulties with visuospatial construction evidenced by most individuals with Williams syndrome. In the fourth part, we review

behavioral genetic studies of visuospatial constructive ability, which suggest that a substantial portion of the individual differences found among people of normal intelligence has a genetic basis.

General Intelligence and Visuospatial Constructive Ability of Individuals with Normal Intelligence

The field of psychometrics has a long tradition of characterizing basic dimensions of individual variation in the human population. Much of the focus within this field has been on intelligence. There is wide agreement that human intelligence includes three components: verbal ability, nonverbal reasoning ability, and spatial ability (Carroll 1993; Mackintosh 1998). These components are partially separable but are not completely independent. For example, correlations between tests of the different abilities are typically $\sim .4$ (Plomin 1999 [in this issue]). Thus, people who perform well on tests of one of these components are more likely than would be expected by chance to perform well on tests of the other components; people who perform poorly on one component are relatively likely also to perform poorly on the others. General intelligence is indexed by these correlations. In the first decades of this century, Spearman and Thurstone laid the framework for a controversy about *g* that persists to this day (see, e.g., Mackintosh 1998). Spearman argued for a model of cognition in which *g* was the central driving force behind covariation in specific cognitive skills. Within this framework, *g* may be taken to index a general ability such as executive function (e.g., planning) or speed of processing. Thurstone's opposing point of view suggested that there are multiple cognitive abilities that operate independently; within this framework, *g* may be considered to be derived from overlapping component processes in different domains of intellectual functioning. At the end of this article, we consider behavioral genetic evidence relevant to this controversy.

Attempts to characterize the nature of spatial ability, and the tests that illuminate it, have isolated two groups of tests that assess partially independent abilities. The more powerful group of tests, often called tests of visualization, are closely associated with ability in visuo-

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spatial construction. These tests include block-design tasks, such as those on the Wechsler Intelligence Scale for Children–Revised (WISC-R; Wechsler 1974) or the Differential Ability Scales (DAS; Elliott 1990) (fig. 1) and related tasks that require participants to compose designs from parts or to specify how an object would look after a spatial transformation (e.g., folding, unfolding, slicing, or rotation). Most generally, these are tests that require understanding how objects or configurations can be altered within the spatial domain. It is quite easy to vary the difficulty of these tests by varying the number of spatial elements or the number of transformations required to complete a given task (e.g., Lohman 1988). For example, the difficulty of block-design or pattern-construction tasks varies as a function of the number of blocks required to construct a pattern and the complexity of the design to be constructed (e.g., the number of solid-color surfaces [easy] relative to the number of two-colored diagonally or vertically divided surfaces [hard] needed). Examination of the norms for the Wechsler Adult Intelligence Test–III (WAIS-III; Wechsler 1997) indicates a wide range of ability for individuals considered to be performing within the normal range. Adults at the bottom of the normal range typically are able to construct correctly the two-block patterns and the simplest of the four-block patterns. Individuals performing at the mean of the normal distribution are able to complete all of the four-block patterns and the simplest of the nine-block patterns. Individuals at the top of the normal range are able to construct all of the nine-block patterns quickly and accurately.

The second group of tests revealing differences in spatial ability is concerned with changes of orientation without changes of shape or configuration. The best known of these tests is mental rotation (e.g., Shepard and Metzler 1971), in which the participant indicates whether two objects are identical except for a change of orientation. Although rotation is often a component of visuospatial construction, ability measured by rotational tests is only partially correlated with ability measured by visuospatial constructive tasks. The rotational tasks can typically be completed more quickly and spontaneously than the constructive tasks, and they may depend more on a well-learned set of unitary spatial intuitions.

Cognitive psychologists have identified three important components of visuospatial construction: spatial working memory, flexibility in the use of spatial reference systems that are necessary for defining spatial properties, and flexibility in the hierarchical organization of objects and configurations. Individual differences in ability for people with normal intelligence have been identified for each of these components (e.g., Just and Carpenter 1985; Pani and Dupree 1994; Shah and Miyake 1996; Mackintosh 1998; Pani et al. 1999). All these

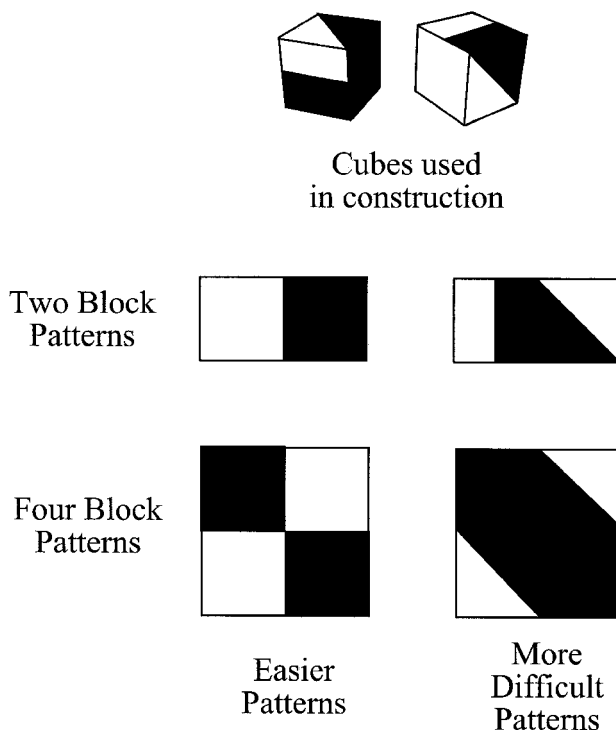


Figure 1 DAS cubes and examples of patterns to be constructed. Each cube has one solid white side, one solid black side, two sides divided diagonally into white and black triangles, and two sides divided vertically into white and black rectangles. Note that the participant is always given the correct number of cubes for the pattern he or she is to construct.

component abilities are likely to be important for success on pattern-construction tasks.

A common finding for spatial tests is that, as a group, males tend to score higher than females (whereas females tend to score higher on verbal tests). Some researchers have suggested that this male-female difference in spatial ability has been diminishing (e.g., Feingold 1988); others disagree (e.g., Masters and Sanders 1993). A number of authors expect male-female differences in spatial ability to remain and have concluded that they are due to the evolution and genetic determination of sexual differentiation (e.g., Silverman and Eals 1992).

Williams Syndrome

Williams syndrome is a contiguous gene disorder (Ewart et al. 1993) involving a hemizygous microdeletion of ~1.5 megabases of chromosome 7q11.23. More than 95% of individuals with Williams syndrome have deletions of this classic length (C. A. Morris, personal communication). Sixteen genes have been mapped to this region (Meng et al. 1998). Williams syndrome is char-

acterized by mental retardation or learning difficulties, a unique cognitive profile, an unusual personality, infantile hypercalcemia, dysmorphic facial features, and supravalvar aortic stenosis (SVAS). The cognitive profile includes relative strength in auditory rote memory, language abilities that are at or somewhat above expectation for overall level of cognitive ability, and extreme difficulty with visuospatial construction. Most individuals with Williams syndrome perform at or below the bottom of the normal range on tests of block design.

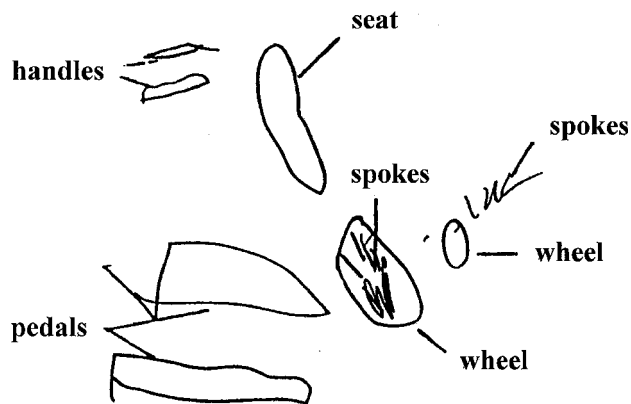
To address genotype/phenotype correlations involving cognition, a quantitative specification of the Williams syndrome cognitive profile (WSCP) is important. To provide such a specification, we operationalized the WSCP as a particular pattern of subtest scores on the DAS. The specific criteria are listed in Frangiskakis et al. (1996) and Mervis et al. (1999). Of 84 individuals with Williams syndrome tested, 74 fit the WSCP, yielding a sensitivity of .88. In contrast, of 56 individuals in the same IQ range but with other disabilities or low-normal intelligence, only 4 fit the WSCP, yielding a specificity of .93. The group of 56 individuals included 20 who had been clinically diagnosed with Williams syndrome but were later found not to have a deletion of 7q11.23. Only 2 of the 20 fit the WSCP. Thus Williams syndrome, as defined molecularly, is associated with an identifiable and very distinctive cognitive profile (Morris and Mervis 1999).

The hallmark difficulty individuals with Williams syndrome have with visuospatial construction activities is most clearly illustrated by their performance on tasks of block design (also referred to as pattern construction). Accordingly, the DAS Pattern Construction subtest is a critical component of the assessment of the WSCP. In pattern-construction tasks, the participant is shown a target design and is asked to reproduce this design from cubes. The cubes and examples of two- and four-block patterns are illustrated in figure 1. To complete this task successfully, the participant must analyze the target pattern into the parts available on the cube, locate the correct parts on the cubes, and arrange the parts into the correct configuration to match the target pattern. This process requires flexibility in moving back and forth between the local organization of the parts of the pattern and the global organization of the pattern as a whole (Pani et al. 1999).

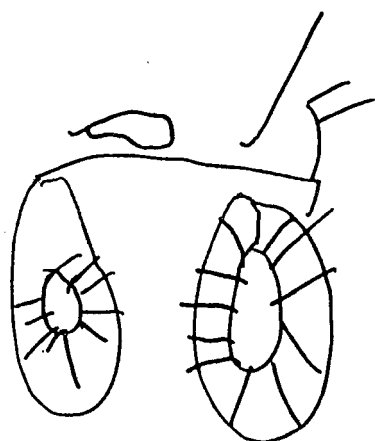
Bellugi and colleagues (e.g., 1988, 1994) provided the first extensive discussion of the difficulty evidenced by individuals with Williams syndrome on block-design tasks and other measures of visuospatial construction, and they concluded that the visuospatial constructive abilities of individuals with Williams syndrome were deviant rather than simply delayed. Bellugi et al. (1988, 1994) reported that adolescents and young adults with Williams syndrome had extreme difficulty copying even

a simple four-block checkerboard pattern—the easiest pattern on the block-design subtest of the WISC-R. (Most normally developing 6-year-olds complete this item correctly.) Although many of the participants used the correct block surfaces, most did not maintain the overall (global) 2×2 arrangement of the blocks, leading to broken configurations. Bellugi et al. (1988, 1994) argued that the visuospatial constructive problems of individuals with Williams syndrome were due to attention to the local elements (parts), at the expense of the global element (the object as a whole). Bellugi et al. (1988, 1994) also noted that, when individuals with Williams syndrome drew pictures of objects, they focused on the parts of the object (local characteristics), rather than the object as a whole (global organization). This led to drawings in which object parts were scattered over the page (fig. 2a).

The finding that individuals with Williams syndrome have great difficulty with a range of visuospatial construction tasks has been replicated repeatedly (see reviews by Mervis et al. 1999; Morris and Mervis 1999). However, recent research suggests that the difficulties evidenced by individuals with Williams syndrome are developmental—part of the normal developmental sequence of acquisition of these abilities—rather than deviant. Bertrand et al. (1997) found that normally developing 4- and 5-year-olds often produced drawings like the bicycle in figure 2a; production of this type of drawing is part of the normal developmental sequence of learning to draw. Longitudinal investigations of drawing ability of individuals with Williams syndrome also fit a developmental model: clear improvement is shown over time, often resulting in drawings in which the parts of objects are clearly integrated into a coherent whole, as illustrated by the bicycle in figure 2b (Bertrand and Mervis 1996). Performance on pattern-construction tasks also improves significantly with age (Fonaryova Key et al. 1998; Mervis et al. 1999). Detailed analysis of the errors made by individuals with Williams syndrome on the DAS pattern-construction subtest (Fonaryova Key et al. 1998) indicates that children produced almost twice as many broken configurations as adults for four-block patterns. The majority of adults did not produce any broken configurations. Furthermore, the proportion of incorrect patterns produced by adults for which the correct block surfaces were used but placed in the wrong position or orientation—the type of error closest to the correct pattern—was more than double that for the children. Finally, Pani et al. (1999) have shown that global perceptual processing by individuals with Williams syndrome is largely normal. All these findings are consistent with a developmental rather than a deviance account of the acquisition of visuospatial construction abilities by individuals with Williams syndrome.



Age: 9 years 7 months



Age: 12 years 11 months

Figure 2 Two drawings of a bicycle by a girl with Williams syndrome, aged 9 years 7 months (*top*), and 3 years later, aged 12 years 11 months (*bottom*). In both cases, the child was given a blank piece of paper and asked to draw the best bicycle that she could. The labels on the top drawing were provided spontaneously by the child.

There are also clear individual differences among persons with Williams syndrome in performance on measures of visuospatial construction. For our sample of participants, standard scores on the DAS pattern-construction subtest, at all ages, range from 20 (the lowest possible score) to 38 (which is at the 12th percentile, definitely within the normal range). On the Developmental Test of Visuo-Motor Integration (VMI; Beery 1989), standard scores range from <55 (the lowest possible score) to 80 (9th percentile). From a qualitative perspective, some adolescents with Williams syndrome produce integrated drawings that are recognizable as the

intended object; at the other extreme, some adolescents produce unrecognizable drawings composed of isolated object parts strewn across the page (Bertrand and Mervis 1996). On the DAS pattern-construction subtest, ~10% of adults with Williams syndrome are able to correctly construct even the most difficult four-block patterns, albeit very slowly (Fonaryova Key et al. 1998). Of the remaining adults, some are able to construct simple four-block patterns and some complex two-block patterns; others can construct only the simplest two-block patterns.

Performance of individuals with Williams syndrome on measures of visuospatial construction was highly correlated with performance on measures of other components of intelligence, as would be expected, given our earlier discussion of *g*. For example, even after we controlled for the effect of chronological age, the performance of a sample of 50 individuals with Williams syndrome on pattern construction correlated .46-.59 with three measures of verbal ability, .48 with a measure of verbal working memory, and .57 with a measure of nonverbal reasoning ability (Mervis 1999). The individuals who performed best on pattern construction were those who had relatively good vocabularies, verbal working memory, and “nonverbal” reasoning abilities. Observation of participants during testing indicated that most tried to work their way through both the pattern-construction and the nonverbal reasoning (matrices) problems verbally, rather than trying to use a spatial strategy. Thus, participants tended to use a compensatory strategy to solve pattern-construction problems, attempting to turn spatial problems into verbal ones. The participants with the highest overall intelligence were the ones most successful with this strategy, as evidenced by their relatively good performance on pattern-construction problems. For these and the remaining Williams syndrome participants, overall verbal ability and nonverbal reasoning were consistently superior to level of performance on visuospatial construction tasks.

Individuals with Small Deletions of 7q11.23

Because the Williams syndrome deletion involves ≥ 16 genes, genotype/phenotype correlations would be extremely difficult to identify on the basis of participants who had classic Williams syndrome. Individuals with more-subtle mutations, involving only one or a few genes, are therefore crucial to genotype/phenotype correlation studies. This strategy has implicated one of the deleted genes, *elastin*, in the vascular pathology associated with Williams syndrome (including SVAS), certain facial characteristics of Williams syndrome, and connective tissue problems associated with Williams syndrome, such as inguinal hernias and hoarse voice (see

summary in Morris and Mervis 1999). Individuals with mutations of only *elastin* did not share other Williams syndrome characteristics.

The strategy of studying individuals with small deletions has also been used to implicate *LIM-kinase1* in the visuospatial construction difficulties associated with Williams syndrome (Frangiskakis et al. 1996). Two kindreds with SVAS and small deletions of 7q11.23 (a total of 13 affected members) were identified. Phenotypic characterization indicated that, as expected, most kindred members with deletions had SVAS, a few facial characteristics of Williams syndrome, and some of the connective tissue problems associated with Williams syndrome. As in previous studies (Ewart et al. 1993; Morris et al. 1993), variability of expression and incomplete penetrance for autosomal dominant SVAS was found. Continued phenotypic characterization indicated that family members with deletions had not had infantile hypercalcemia, did not fit the Williams syndrome personality, and did not have mental retardation. Performance of affected members on measures of auditory rote memory and language was similar to that of unaffected relatives. However, most affected members performed less well on measures of visuospatial construction than would have been expected for their overall level of ability. These individuals fit the WSCP. None of the unaffected members shared any Williams syndrome characteristics, including the WSCP. All family members had IQs in the low-average range.

Molecular genetic characterization indicated that deletion size was ~84 kb in one kindred and ~300 kb in the other. Continued characterization indicated that the smaller deletion included only *elastin* and one other gene, *LIM-kinase1*. As indicated above, kindreds with mutations of only the *elastin* gene do not evidence any cognitive or personality aspects of the Williams syndrome phenotype. In fact, *elastin* is only negligibly expressed in the brain. In contrast, *LIM-kinase1* is strongly expressed in different regions of the brain, with highest expression in the cerebral cortex. This pattern of findings indicates that hemizygous deletion of *LIM-kinase1* contributes to the visuospatial constructive difficulties of individuals with Williams syndrome (Frangiskakis et al. 1996). We have since characterized three additional kindreds with small deletions and low-average intelligence. Affected members of these kindreds fit the WSCP, but none of their unaffected relatives did (authors' unpublished data). Members of all five kindreds tended to "talk" their way through pattern-construction problems.

Tassabehji et al. (1999) have reported findings from three additional individuals (from two families) with small deletions of 7q11.23, including both *elastin* and *LIM-kinase1*. On the basis of cognitive features of these participants, Tassabehji et al. (1999) concluded that hap-

loinsufficiency for *LIM-kinase1* may be necessary for impaired visuospatial construction but is not sufficient. However, all three individuals were of higher intelligence than members of the kindreds we studied; and two of the three had above-average intelligence and excellent reasoning abilities. Given these strengths, these individuals would be expected to successfully use verbal compensatory strategies to solve pattern-construction problems. Thus, not surprisingly, these individuals did not fit the WSCP, which includes limits on the absolute level of performance on DAS pattern construction, consistent with mental retardation or borderline-normal or low-average intelligence.

In summary, the hallmark cognitive weakness of individuals with Williams syndrome is visuospatial construction. Studies of kindreds with small deletions of 7q11.23 indicate that hemizygous deletion of *LIM-kinase1* contributes to these difficulties. Although visuospatial construction is the area of greatest difficulty for most individuals with Williams syndrome, there are still individual differences among people with Williams syndrome in level of visuospatial constructive ability, which correlate strongly with individual differences in verbal ability and in nonverbal reasoning ability. Any genetic basis for these individual differences is likely due (at least primarily) to regions of the human genome other than 7q11.23.

Behavioral Genetic Studies of Visuospatial Construction

Our research demonstrated that hemizygous deletion of *LIM-kinase1* on chromosome 7 forms a foundation for the deficit in visuospatial constructive cognition evidenced in Williams syndrome. The genetic basis of normal variation in visuospatial construction, however, is less clear. Barring other evidence, defining DNA variation in *LIM-kinase1* and associating genetic variation with phenotypic variation is a reasonable place to begin the search for genes influencing visuospatial construction. It is possible, however, that *LIM-kinase1* is necessary for the development of normal visuospatial construction but that it does not account for a significant amount of the variation in the normal range. Regardless of the role of *LIM-kinase1*, the genetic underpinnings of visuospatial construction as a complex behavioral trait are likely to follow a quantitative trait loci (QTL) model described by a multigene system, with each gene contributing to the distribution of individual differences in visuospatial construction.

Behavioral genetic research and traditional psychological research can inform molecular work concerning the genetic basis of visuospatial construction. Twin and adoption studies indicate that visuospatial construction,

as measured by block-design tests, is heritable, with the proportion of variance accounted for by genes in the range of .44–.68 in older children and adults (Rose et al. 1979; Tambs et al. 1984). There is also evidence that the h^2 estimates of visuospatial construction increase from toddlerhood to childhood (Cardon 1994) and may include both additive and nonadditive effects (Pedersen et al. 1992b). Beyond estimating the heritability of visuospatial construction, behavioral genetic work draws on the rich psychological tradition that attempts to disentangle the complex phenotype concerning the relation between specific cognitive skills, such as visuospatial construction, and general cognitive ability, as described at the beginning of this article.

Current research has begun to focus on whether models such as those of Thurstone or those of Spearman better account for the genetic underpinnings of visuospatial construction. Is it the case that there are multiple independent skills with a fairly simple mapping between single genes and individual skills, or are we more likely to find a complex system of genes that influence visuospatial construction, other specific cognitive skills, and g ? Recent work with sophisticated multivariate behavioral genetic models indicates that the answer may lie somewhere between the views of Spearman and Thurstone. Analyses of genetically informative data by means of a hierarchical model provide evidence that specific cognitive skills can be accounted for, in part, by genetic influences that are specific to individual skills as well as common to many of them. Pedersen et al. (1992a), for instance, used multiple regression methods coupled with a powerful twin-adoption design to demonstrate that a significant amount of variation in block-design scores can be attributed to genetic influence that affects block-design but not other WISC-R subtests. At the same time, however, a large portion of the variance in block-design scores was accounted for by genetic influences that are shared with many other cognitive skills. In fact, consistent with purely phenotypic models, block design was found to be one of the most heavily g -loaded abilities relative to other commonly measured cognitive skills. Therefore, it appears that there are genetic influences on visuospatial construction that are unique to this cognitive skill, as well as genetic influences that are mediated by g .

There is some evidence that the hierarchical model implying both unique and general genetic influences on cognitive abilities may be reflected at the molecular level. Petrill et al. (1996) examined the relations between eight DNA markers known to be associated with cognitive ability and the subtests of the WISC-R. They found significant associations between two markers (EST083 and HLA) and the block-design subtest. These two markers also significantly predicted scores on many other cognitive subtests included in this study, indicating that they

most likely influence a basic process central to g . In a second analysis, the effects of g (full-scale WISC-R IQ) were removed prior to examination of associations between individual markers and block design. One marker, ADH5, significantly predicted block-design scores even after Petrill et al. accounted for the influence of g . Moreover, block design was the only WISC-R subtest to be significantly associated with this marker. Thus, normal variation in visuospatial construction is likely associated both with genes that influence cognitive ability in general and with genes whose influence is relatively specific to visuospatial constructive ability.

In summary, large individual differences in visuospatial constructive ability are found among people with normal intelligence. Individual differences in visuospatial construction are also found for individuals with Williams syndrome. However, the range of these individual differences is restricted to the lower part of the normal range or, in most cases, below the bottom of the normal range. The visuospatial constructive abilities of individuals with Williams syndrome are also substantially lower than expected on the basis of overall level of cognitive ability, suggesting specific genetic involvement in the determination of visuospatial construction. At the same time, visuospatial constructive ability is strongly correlated with both verbal ability and nonverbal reasoning ability. Studies of kindreds with small deletions have implicated hemizygous deletion of *LIM-kinase1* as contributing to the difficulty that individuals with Williams syndrome have with visuospatial construction. Behavioral genetic studies have indicated that visuospatial constructive abilities have a substantial genetic component, most likely fitting a QTL model, with some genes involved in determining visuospatial constructive ability affecting cognitive ability in general, and others that are likely to be relatively specific to visuospatial construction. This QTL model likely accounts for individual differences within both the normal population and the Williams syndrome population.

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