

Beyond nature vs. nurture: Williams syndrome across cultures

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Nobody questions that the color of our eyes is encoded in our genes. When it comes to behavior the concept of "DNA as fate" quickly breaks down – it's been long accepted that both genes and the environment shape human behavior. But just how much sway the environment holds over our genetic destiny has been difficult to untangle.

Scientists at the Salk Institute for Biological Studies have found a clever way to sort one from the other: They compared the social behavior of children with Williams syndrome — known for their innate drive to interact with people — across cultures with differing social mores. Their study, published in a forthcoming issue of *Developmental Science*, demonstrates the extent of culture's stamp on social behavior.

"Overall, a consistent result has emerged from our research," summarizes lead author Ursula Bellugi, director of the Laboratory for Cognitive Neuroscience at the Salk. "Regardless of age, language or cultural background, Williams syndrome social phenotype is shaped both by genes and interactions with the environment."

The current research is just one piece in a puzzle that a large collaboration of scientists under the umbrella of a long-running Program Project from the National Institutes of Child Health and Human Development has been trying to piece together over the last decade. Led by Bellugi, the researchers are looking to Williams syndrome to provide clues to some of the mysteries of the genetic basis of behavior. Much of the research revolves around the work of molecular geneticist Julie R.

Korenberg, a professor in the Department of Pediatrics at UCLA and an adjunct professor at the Salk Institute, who has been studying the genetic basis of Williams syndrome for the last decade.

Virtually everyone with Williams syndrome has exactly the same set of genes with one strand missing, a small set of genes on chromosome 7, but some rare cases with different size deletions sparked the interest of researchers. One unusually shy and introverted little girl retained at least one gene from the GTF2i family that most people with the disorder have lost. This finding convinced Korenberg and her collaborators that this short stretch of DNA may contain the gene (or genes) responsible for the hypersociability among children with Williams syndrome.

"Although a certain amount of variability exists with the Williams syndrome population, the clear genetic basis presents an unusual opportunity to search for the genetic underpinnings of human social behavior and social characteristics, such as trust and over-friendliness," explains Bellugi.

Identified more than 40 years ago, Williams syndrome occurs in an estimated one in 20,000 births worldwide. It arises from a faulty recombination event during the development of sperm or egg cells. As a result, almost invariably the same set of about 20 genes surrounding the gene for elastin is deleted from one copy of chromosome seven, catapulting the carrier of the deletion into a world where people make much more sense than objects do. Despite a myriad health problems and a generally low IQ, children with Williams syndrome are loquacious, sociable, and irresistibly drawn to strangers.

To determine the extent to which this behavioral profile is universal across culture, the researchers settled on two vastly differing environments: the United States and Japan, whose cultural differences are said to be aptly summarized in two proverbs: In America, "The

squeaky wheel gets the grease," while in Japan, "The nail that stands out gets pounded down."

Using a questionnaire developed by Salk researchers, Bellugi and first author Carol Zitzer-Comfort, a professor at California State University in Long Beach, asked parents in the U.S. and Japan to rate the tendency of their child to approach others, their general behavior on social situations, their ability to remember names and faces, their eagerness to please other people, their tendency to empathize with others' emotional states, and the tendency for other people to approach their child.

Despite the differences in upbringing, in both countries children with Williams syndrome were rated significantly higher in global sociability and their tendency to approach strangers than were their typically developing counterparts. But cultural expectations clearly influenced social behavior, since the sociability of normal American kids was on par with Japanese Williams syndrome kids, whose social behavior is considered out of bounds in their native country.

Says Zitzer-Comfort: "It really is an intriguing illustration of the interaction between nature and nurture," but notes that there might be alternative explanations. Japanese parents, for one, rated their children generally lower on the 7-point scale of the questionnaire. "Perhaps the stigma of having a 'different' child in Japan affected the ways in which parents ranked their child's degree of sociability," speculates the scientist.

In an earlier study, published last year, Bellugi and her colleagues collected oral narratives from children and adolescents with Williams syndrome in the U.S., France, and Italy and came to a similar conclusion. Not only are Williams syndrome kids natural born storytellers, who hook their audiences with expressive and affective narratives, but — no matter where they grew up — they did their countrymen significantly

better.

Source: Salk Institute

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