

Focus

The singing syndrome They have pixie-like faces, a cheerful nature and a low IQ. But they sure can sing. ANNE McILROY delves into the mysterious world of people with a rare genetic disorder

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Lisa Walsh is preparing for a concert in Montreal where she will sing a few jazz standards with two of Duke Ellington's children. At 24, she has already performed on stage in Los Angeles and Las Vegas and is featured on a compact disc of hit tunes from musicals.

"I love to sing for people and I love jazz," she says.

Ms. Walsh, a born performer, was also born with a rare neurogenetic disorder called Williams syndrome. It has given her a pixie-like face, starry eyes and an affinity and talent for music. She has perfect pitch, can hear a melody once and pick it out on the piano and can sing pieces of classical music, such as Brahms's Requiem, by heart.

But she can't add two plus two.

People with Williams syndrome have low IQs and trouble reasoning. They also have poor spatial perception, which makes it hard for them to tie their shoes or cross a street safely. Most have to live with their parents or in a group home.

They fascinate scientists because they are so talented in some ways, so disabled in others. Many are musical, and some, like Ms. Walsh, are gifted. New research, in which she has participated, suggests that their brains are actually wired differently when it comes to music. When she sings Cry Me A River to a crowded concert hall in June, a part of her brain that has nothing to do with music in most people will be rippling with electrical and chemical activity.

"Imagine opening up the hood of the car and finding all the parts are doing things they weren't supposed to be able to do," says **Daniel Levitin**, a professor at McGill University in Montreal who does experiments on music and the human mind.

Like many researchers, he is fascinated by Williams syndrome, because it challenges our notions of intelligence and because it offers a rare glimpse of the role genes play in brain development, brain function and human behaviour.

The syndrome is estimated to occur in one in 20,000 births, and has probably been around for centuries. Afflicted children often have elfin features, including small upturned noses, and some have a unique star patterns in their irises. Their looks, cheerful and outgoing nature and ability to make haunting and lyrical songs have led some researchers to speculate that they were the original pixies and "little people" in the folklore of different cultures.

The syndrome was first identified 40 years ago, when J. C. P. Williams, a heart specialist in New Zealand, noted that a small group of his patients had distinct facial features and similar cardiovascular problems and seemed to be mentally retarded.

In the years that followed, doctors identified other traits. Babies often have trouble digesting food and sleep poorly, possibly because they are extremely sensitive to noise. Children are slow to develop, are often short and can wrinkle prematurely.

Doctors always suspected that it was genetic, and in 1993, the cause of the disorder was discovered. In 95 per cent of cases, children born with Williams are missing a tiny section of genetic material on one copy of chromosome No. 7.

Every cell in our bodies, except for sperm and eggs, has two copies of 23 chromosomes. In people with Williams syndrome, one copy of No. 7 is perfect, but the other has what is known as a micro-deletion. It means that they are missing one copy of about 20 genes, including the gene for elastin, the protein that gives skin and other organs their elasticity and keeps blood vessels and arteries flexible.

Recent research has shown that in the remaining 5 per cent of people with Williams, that same section of chromosome No. 7 is inverted, or flipped over, knocking those genes out of operation.

Genes carry the directions for making proteins that perform various crucial functions in the body. Because they have only one copy of some genes, people with Williams syndrome produce smaller amounts of some proteins, which can have serious consequences in their bodies. Their lack of elastin, for example, explains why they suffer from heart problems and wrinkle prematurely.

Three of the genes they are missing have been generally linked to the brain, although researchers don't know exactly what they do. It seems likely that they don't make enough of a protein crucial for the brain to develop or function normally. That deficit has led to what researchers call asymmetrical intelligence or fractionated brain functioning. The people are disabled in some ways, but normal or gifted in others.

People with Williams syndrome tend to have strong verbal and social skills, and are good at recognizing faces -- perfect cocktail party personalities. In some cases, the people they meet don't realize at first that there is anything different about them.

Their musicality has been documented only in recent years. Howard Lenhoff, a professor emeritus of biological sciences at the University of California, has a daughter with Williams syndrome. He didn't notice Gloria's musical talent until her bat mitzvah at the age of 13. He had scheduled a private ceremony because he didn't think she would be able to stand up and sing from the Bible in front of a crowd. She performed flawlessly.

Dr. Lenhoff started wondering if other children like Gloria might be musical, and conducted experiments that revealed many were. Subsequent work by other researchers showed that many people with Williams syndrome have perfect pitch, love music more than the average person and respond to it with great emotion.

"Music is my favourite way of thinking," one child once told researchers.

Not all are musical. Some are deeply attracted to loud noises, and love leaf blowers.

Others are like Gloria Lenhoff, now in her 40s, who can sing 2,000 songs in 25 different languages and once brought a Bosnian porter at the airport to tears by singing him a song in his native tongue.

How does she do it?

There is some evidence that the actual physical structure of the brain is different in people with Williams syndrome. Autopsies have shown they have smaller brains, and the upper back parts of the brain that deal with spatial abilities are underdeveloped, says Albert Galaburda, a neurology professor at Harvard University.

But parts of the brain known to be important to both music and language have been found to be unusually large. Dr. Galaburda's microscopic examinations have also found larger cells in the part of the brain that processes sound.

Dr. Levitin was building on Dr. Galaburda's work when he decided to study how music is processed in the brains of people with Williams syndrome. He used functional Magnetic Resonance Imaging (fMRI), a new tool that allows scientists to take pictures of living brains at work.

He compared people with Williams to a control group without the disorder. The results were striking. Normal people process music through intense activity in well-defined network contained within a relatively small region of the brain, but people with Williams recruit many widely dispersed areas of the brain, calling on primitive parts, such as the brain stem, and the amygdala, which is tied to emotional responses.

"Their brains are wired differently," says Dr. Levitin, who published his findings this week in the scientific journal *NeuroImage*.

It is a rare glimpse of the role genes play in the development of the brain, Dr. Levitin says. The picture is still murky. It's not clear if the missing Williams syndrome genes actually cause structural damage, and whether the rewiring Dr. Levitin found was a response to that damage.

There is some evidence that the human brain may be more flexible, more adaptable than previously believed. Research shows that people who have been profoundly deaf from birth use the part of their brain that processes sound for another purpose, Dr. Levitin says.

Ms. Walsh was one of the young adults who took part in Dr. Levitin's brain-imaging experiment. Nancy Walsh, Lisa's mother, was intrigued by the snapshot the results have given her into the workings of her daughter's mind.

Her daughter was diagnosed with Williams syndrome when she was a year old. By the time she was 3, she was entranced by music, and could more easily pick up a song off the television than talk. "The only toys she played with were musical toys," her mother says. "It had to have a musical element in it or she wasn't interested."

In school, she loved choir, and was never nervous about performing. "In Grade 1, she was the little red hen in a musical. And we were floored when she stepped up to the stage and belted out that little red hen song."

When she was 14, she started going to a music camp in Massachusetts that had been established by Dr. Lenhoff and other parents of children with Williams syndrome. "She found herself with that group of people, who had an affinity for each other and for creating music," her mother says.

Mrs. Walsh was inspired by Dr. Lenhoff and his daughter Gloria, and decided that she and her husband would help make music more important in Lisa's life. They found a voice teacher for her, and she began performing in local adult choirs.

After high school, Ms. Walsh attended Concordia University's Centre for the Arts in Human Development, which trains graduate students to use the arts as therapy for the disabled. She starred in a musical it put on, and her voice was so strong, so rich and vibrant, the centre asked her to sing on a CD it produced.

The centre is organizing the June fundraising concert at Montreal's Oscar Peterson Concert Hall where Ms. Walsh will perform.

She now attends a special music school in Massachusetts that was established to help children with Williams syndrome take their musical skills to the next level, and also to teach them how to live independently, how to tidy their rooms and manage money.

There are two streams. One group of students is working toward being an assistant to a music therapist, either at a daycare centre or a seniors home. That wasn't appropriate for Ms. Walsh. "If you asked to her take a little old lady in a wheelchair somewhere, she might hit the wall three times on her way," her mother says.

Instead, Ms. Walsh is perfecting her performing skills. She and the other students in her group may start singing in seniors homes. She sings with a Montreal classical choir, and is in demand as a performer at fundraisers. She brought down the house at the Beverly Hills Hilton and has also sung at the Mirage Hotel in Las Vegas.

"It is amazing when you think about it. It is not like a professional person doing this as her business, but it is still amazing," Mrs. Walsh says.

Ms. Walsh says she sometimes gets nervous before she goes on stage, but loves performing and seeing the audience's reaction. "They listen to my music and they think I do a good job."

Anne McIlroy writes on science for The Globe and Mail.

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