Moyamoya Disease
Rare vascular syndrome carries high stroke risk for children

Stroke survivor Malachi Horton
Cover Story

8 A Puff of Stroke
Moyamoya disease is a rare vascular syndrome, usually found in children, for which the first symptom is often a stroke or TIA. We talked with a neurosurgeon who treats moyamoya as well as the mothers of several children with it.

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Stroke survivor and caregiver parents are concerned with how the stroke affects their children. Ashley and Lisa Park were 5 and 3 when their dad had a massive hemorrhagic stroke and brain surgery. Adults now, they share their thoughts on growing up in a stroke family.
Did you know the extreme tightness that can appear in the upper limbs after a stroke can be a treatable condition? This condition—known as spasticity—can develop months after your stroke and is sometimes left untreated. Whether it’s difficulty buttoning your shirt or washing your hands, spasticity can turn the simplest everyday activities into the most complicated tasks. If you’re suffering from spasticity, help is out there. Talk to your doctor about your symptoms today. Relief is possible.

Visit MySpasticityAnswers.com to create a personalized doctor discussion guide.
Non-invasive brain stimulation may help stroke survivors recover speech and language function, according to new research in the American Heart Association journal *Stroke*.

Between 20 and 30 percent of stroke survivors have aphasia, which affects their ability to grasp language, read, write or speak. It’s most often caused by lesions in areas of the brain that control speech and language.

“For decades, skilled speech and language therapy has been the only therapeutic option for stroke survivors with aphasia,” said Alexander Thiel, M.D., study lead author and associate professor of neurology and neurosurgery at McGill University in Montreal. “We are entering exciting times where we might be able in the near future to combine speech and language therapy with non-invasive brain stimulation earlier in the recovery. This could result in earlier and more efficient aphasia recovery and also have an economic impact.”

In the small study in Germany, researchers treated 24 survivors with several types of aphasia. Thirteen received transcranial magnetic stimulation (TMS) and 11 got sham stimulation.

The TMS device is a handheld magnetic coil that delivers low intensity stimulation and elicits muscle contractions when applied over the motor cortex.

During sham stimulation the coil was placed over the top of the head where there is a large venous blood vessel and not a language-related brain region. The intensity for stimulation was lower so that participants still had the same sensation on the skin but no effective electrical currents were induced in the brain tissue.

Patients received 20 minutes of TMS or sham stimulation followed by 45 minutes of speech and language therapy for 10 days.

The TMS groups’ improvements were on average three times greater than the non-TMS group. They used German language aphasia tests, which are similar to those in the United States, to measure language performance of the patients.

“We believe brain stimulation should be most effective early, within about five weeks after stroke...”

“TMS had the biggest impact on improvement in anomia, the inability to name objects, which is one of the most debilitating aphasia symptoms,” Thiel said.

“We believe brain stimulation should be most effective early, within about five weeks after stroke, because genes controlling the recovery process are active during this time window,” he said.
A Secret in the Eyes
Hypertensive retinopathy may hold clues to stroke risk

Your eyes may be a window to your stroke risk.
In a study reported in the American Heart Association journal Hypertension, researchers said retinal imaging may someday help assess if you’re more likely to have a stroke.

“The retina provides information on the status of blood vessels in the brain,” said Mohammad Kamran Ikram, M.D., Ph.D., lead author of the study and assistant professor in the Singapore Eye Research Institute, the Department of Ophthalmology and Memory Aging & Cognition Centre at the National University of Singapore. “Retinal imaging is a non-invasive and cheap way of examining the blood vessels of the retina.”

Worldwide, high blood pressure is the single most important risk factor for stroke. However, it’s still not possible to predict which high blood pressure patients are most likely to develop a stroke.

Researchers tracked stroke occurrence for an average 13 years in 2,907 patients with high blood pressure who had not previously had a stroke. At baseline, each had photographs taken of the retina, the light-sensitive layer of cells at the back of the eyeball. Damage to the retinal blood vessels attributed to hypertension — called hypertensive retinopathy — evident on the photographs was scored as none, mild or moderate/severe.

During the follow-up, 146 participants experienced an ischemic stroke and 15 had a bleeding stroke.

After adjusting for other stroke risk factors, they found the risk of stroke was 35 percent higher in those with mild hypertensive retinopathy and 137 percent higher in those with moderate or severe hypertensive retinopathy.

Even in patients on medication and achieving good blood pressure control, the risk of a blood clot was 96 percent higher in those with mild hypertensive retinopathy and 198 percent higher in those with moderate or severe hypertensive retinopathy.

“It is too early to recommend changes in clinical practice,” Ikram said. “Other studies need to confirm our findings and examine whether retinal imaging can be useful in providing additional information about stroke risk in people with high blood pressure.”

Get to the Hospital!
In mild strokes, early treatment may eliminate risk of disability

In the case of mild or moderate strokes, getting treatment ultra-fast — within 90 minutes of experiencing symptoms — greatly reduces the risk of disability, according to a new study reported in the American Heart Association’s journal Stroke.

The American Heart Association/American Stroke Association recommends getting to a hospital as quickly as possible and within three hours of the onset of symptoms. According to guidelines, clot-busting drugs may be given to treat stroke up to 4.5 hours after the onset of symptoms for some patients.

The study found that survivors with mild to moderate strokes who were given the clot-busting drugs in the first 90 minutes of that 4.5 hour window had little or no disability three months later compared to those who were treated between 90 and 270 minutes.

“Ultra-early treatment increases the likelihood of excellent outcome in patients with moderately severe symptoms, and in secondary analysis also in those with mild symptoms,” said Daniel Strbian, M.D., Ph.D., from the Department of Neurology at Helsinki University Central Hospital in Helsinki, Finland. “All measures must be taken to reduce onset-to-treatment time as much as possible.”

The study included more than 6,800 patients at 10 stroke centers in Europe over 14 years. They were treated intravenously with Alteplase, a clot-busting drug. Patients were separated into three groups based on stroke severity — mild (NIH stroke score 0-6), moderate (NIH score 7-12), or severe (NIH score 13 or higher). Those with mild to moderate stroke benefited most from the ultra-early care.

Those with severe stroke did not benefit as much from the ultra-early treatment. According to Strbian, this does not mean IV thrombolysis is not working in this group, just that it is less effective because there is a higher clot burden.
Reinventing Myself

My life used to be normal, rather uneventful. I had a fulfilling career and was busy, doing things I enjoyed — shopping, reading, going to the bookstore and being with my family. Then I had a hemorrhagic stroke, and things changed drastically.

Initially I did not understand what had happened nor the physical toll of the stroke. The left side of my body was affected. I had to learn to walk, read, use the stairs and perform some daily activities such as writing a check. It was hard work, but after five months in a rehabilitation hospital, I was ready to come home.

I could not do all I did before. Simple tasks were challenging and required more energy. I needed my children to help me do many things. I was not able to return to my job, and I had to surrender my driver’s license.

A lot of my identity came from the different roles I played. I was a nurse, a high school teacher, mother and wife. I attended work-related conferences and drove to wherever I needed to be. I was independent.

After my stroke I experienced a huge loss of independence, and it saddened me. I am still sad about the losses, but I am trying to rejoice in the gains. I survived a stroke, not everyone does. I am able to take care of myself, care for my home and take daily walks. I am still teaching by sharing my story. I am learning that who I am is not about the career I have, the degrees I hold or how many tasks I can manage at one time.

Reinventing myself means looking inside, reevaluating what is important and not letting the stroke define me. Though our family has had to adjust, I am still able to be a mother, wife, sister, aunt and daughter — all roles more precious to me now that I am a stroke survivor. I feel like Dorothy in The Wizard of Oz — the things I was searching for were right in front of me all the time. Who I am does not exist in the external things, but inside of me — the type of person I am, how I use this experience to help others.

It is not easy forging a new path, but I am slowly learning to accept these new challenges. Strokes are devastating because they take so many things from us, but they do not have to rob us of our essence. I am finding new ways to share my talents and passions with others. This is just as challenging as any therapy program. If I do not continue searching for ways to use this experience for something positive, if I allow the stroke to rob me of the essence of who I am, then I will always be a stroke patient, not a stroke survivor.

Denice DeAntonio, Survivor
Fleetwood, Penn.
n April 15, 2006 (yep, tax day), I experienced a hemorrhage of an AVM (arterio-venous malformation) resulting in a stroke. I was airlifted to Washington D.C. Hospital Center where I had surgery. When I woke up in recovery with a tracheotomy, I could no longer walk, talk, read a book or swallow. In addition to the trach, I had a feeding tube and a urinary catheter.

Two months later, I left Washington Rehab Hospital in a wheelchair still unable to walk, read or swallow, and still retaining the feeding tube and catheter. I was able to communicate with a very limited vocabulary and grunting a lot. The next month, my Maryland driver’s license expired.

Prior to my stroke, I had ordered my “dream car,” a Chevrolet Corvette, for my 60th birthday, but had to refuse it when it came in because I was in the hospital. After many years of physical, occupational, speech and vision therapy (the stroke left me with double vision) as well as psychological counseling, I am now walking, talking, reading, running a little, playing basketball, eating and swallowing solid foods and liquids.

Once I had regained my vision and satisfied the Maryland Medical Advisory Board that I was capable of driving a vehicle, I took the driver’s test and regained my driver’s license in August 2007. Then I re-ordered my dream car. I am proud to report that since January 2008, I have been driving a C6 Corvette Coupe. I could not have done it without a fantastic primary caregiver (my wife, Sandi) and oodles of great therapists.

Being one of the fortunate people who survived a stroke, I have found that it takes lots of patience and time for the brain and body to heal. I would further counsel survivors to seek out professional assistance and support during the recovery process. And never, ever, give up on that dream — whatever it may be.

Bill Perrick, Survivor
Woodstock, Md.
Dr. Donna Arnett, immediate past president of the American Heart Association/American Stroke Association, has a couple of firsts to her credit: the first epidemiologist to be president and the first stroke survivor.

A noted researcher and chairperson of the Department of Epidemiology at the University of Alabama at Birmingham School of Public Health, Dr. Arnett had a stroke in 1986 when she was only 27 years old. It was caused by a genetic clotting disorder that she did not know she had.

The Event

Just like every morning, she got up at 5:30 to let out her dog, Nikki. She felt odd, a little fuzzy, but chalked it up to the early hour. When she called Nikki back in, the words came out garbled, which she questioned, but went on to work. She was a clinical nurse at the time and knew the warning signs for stroke; she even thought that what was happening might be a stroke. Nonetheless, she dismissed the possibility because she was so young. There was no pain, and she could not imagine that there would be no pain involved in having a stroke.

At work the fuzzy feeling continued. She recalled, “I remember turning to my boss during the morning conference that happened at 7:30 a.m. every Friday and saying, ‘I think I’m having a stroke.’” He thought she was kidding.

As the morning wore on, symptoms kept piling up. By 9:30, she had started to drool; one side of her face was drooping; her speech was slurred; and she had weakness on her left side. “It’s an eerie feeling to have all these symptoms, knowing that they’re meeting all the criteria of a stroke, but not believing, because of my age, that it was happening to me,” Dr. Arnett said. By the time she got to the emergency room, she could no longer speak.

At the hospital, she had a CT scan, which didn’t show a stroke. Based on her symptoms, however, she was admitted to the hospital for several days. The CT scan was the standard of care in 1986, a decade before the clot-busting drug tPA was approved.
for treating acute ischemic stroke. An MRI three weeks later confirmed stroke signs. She received no treatment.

Her speech came back within two days, but she had lingering cognitive deficits. She had difficulty with math, and could not remember how to weigh patients on the balance-beam scale or work the exercise treadmill testing equipment, which was part of her job. In addition, she could not read a calendar. “I would say to my patient, ‘You need to come back in a week,’ and I didn’t know what a week was. I knew it had seven days, but I would have to put my finger on the calendar and count seven days.”

She quickly relearned her job functions, but the calendar was more of a challenge. “I guess reading a calendar and scheduling appointments involves more complex reasoning,” she said. “It literally took me months to work through that and be able to schedule again. And still to this day, I have to look. It’s hard for me to abstract out and remember what two weeks or three weeks is. I have to really look at a calendar.”

Within six months she had recovered completely, and she chalks that up to her age at the time.

The Cause

Three days before her stroke, Dr. Arnett had had a routine procedure. It went fine, but what she didn’t know was that she had a condition that left her prone to clotting after surgery. Clots had formed around the mitral valve in her heart. Over the course of that fateful Friday morning, some of those clots broke off and traveled to her brain’s temporal lobe, creating many small lesions. “I saw them on the MRI — white speckles that lit up my brain like brilliant stars lighting up the sky,” she said in a story for the AHA website.

Of course, considering the current scientific knowledge about stroke, Dr. Arnett was extremely fortunate. The clot-busting drug, tPA, had not yet been FDA-approved at the time of her stroke. tPA is a treatment that demands getting to the hospital as quickly as possible because it cannot be given more than 4.5 hours after stroke symptoms start. Back then, people weren’t as aware of the importance of calling 9-1-1 right away when they saw someone showing warning signs. “Now we know that it’s critical to act if you see someone you know — or even a stranger — showing signs of a stroke,” she said. Dr. Arnett found an expert who identified the cause for her clotting problem and prescribed warfarin, which she has been taking ever since.

An emotional deficit

There was another consequence to her stroke — depression. “It was not a fog because my brain wasn’t foggy, but more of a sadness that was almost chemical or biochemical, like a reaction in my brain to healing,” she said. “But I remember being very sad. I don’t know how much of that was part of the organic process of the brain healing and how much of it was just the sense that you feel of your own mortality when something as major as a stroke happens. It was at least a year before I felt really back to myself.”

She urges other survivors to be kind to themselves when they feel that sadness or depression because it is a natural reaction to stroke. “Don’t be afraid to talk to your doctor or minister or therapist about those feelings because they are real,” she said.

She urges caregivers to be patient with their survivors, especially if there is a speech disorder involved. More than 30 years later, she still recalls how frustrating it was to know what she wanted to say and not be able to say it. “Being unable to speak is an isolating and frightening place to be,” she said. “Patience is a must. Let your loved one heal in their own time.”

Medication Strategy

Because medication adherence is such an important issue for our readers, we asked Dr. Arnett what has helped her take her medication every day for more than 30 years.

“I always take my medication at the same time every day, and I associate it with another activity I do every morning, which is brush my teeth. So linking your regimen of pills to something you know you do every day can really help adherence. I also have the Monday-through-Sunday pillbox so that if I don’t remember if I took it, I can go back and make sure. If you have that thought, ‘Did I take my pills this morning?’ you can go back and check.”
Here are many causes of stroke, but one of the most unusual is moyamoya disease, a blood vessel or vascular disease in which an ischemic stroke or TIA may be the first symptom. Although adults can have it, it is more common in children (age 2-5, although any age is possible). We talked about this disease with Ed Smith, M.D., a pediatric neurosurgeon, co-director of the Neurosurgical Stroke Program at Boston Children’s Hospital and an expert on moyamoya and other vascular diseases of the brain.

A Little Brain Anatomy

Moyamoya causes the two large blood vessels that pass through the front of the neck to the brain — the internal carotid arteries — to narrow over time. This reduces the blood supply to the brain, which raises the risk of a TIA or ischemic stroke.

The disease is different from other artery-clogging diseases like atherosclerosis. “It’s caused by an overgrowth of muscle within the artery that narrows it from the outside in as opposed to plaque clogging it from the inside,” Dr. Smith said.

Another way that moyamoya differs from other types of diseases is that when the brain gets starved for oxygen, it calls for help by either widening tiny existing blood vessels or growing new ones to get additional blood flow in the brain. “It is similar to traffic problems, kind of like when a major six-lane highway is shut down to just one lane, so all the tiny back roads become swollen with traffic and people look for new routes to get where they need to go,” Dr. Smith said. Both the old and new blood vessels get engorged trying to bring extra blood to the brain. Named by a Japanese doctor who described the condition in the 1960s, moyamoya is Japanese for “puff of smoke” because that’s how the engorged vessels appear in X-rays of the head.

The problem is that new and existing blood vessels can’t possibly carry the same amount of blood if the parent arteries (the carotids) shut down, starving the brain of oxygen.

Unlike atherosclerosis, which can be stabilized or reversed, moyamoya is always progressive — the carotid arteries continue to narrow. If left untreated, ultimately moyamoya will lead to strokes and death. Its progression varies, although in children it can progress quickly.

Causation and Epidemiology

According to Dr. Smith there are likely several different conditions that look the same on X-ray, act the same in terms of causing symptoms but are caused by different underlying medical problems. For example, there are number of different genes that seem to be related to Asian populations that get moyamoya but are not present in Caucasian or North American
populations. On the other hand, there are American populations that have a mutation that causes a moyamoya-like condition that is not related to the Asian version. “Among our patients, we have several identical twins, so they have the same genes but one has moyamoya and the other doesn’t,” said Dr. Smith. “So, clearly, number one, there are several different types of causes for moyamoya. And number two, there’s both an environmental influence and a genetic influence.”

In addition, moyamoya is often not recognized as a cause of stroke in some children with sickle-cell disease because their condition mimics moyamoya. “I think there are close to 400,000 people in the U.S. with sickle-cell disease and up to 10 percent of those will have a stroke before they’re age 20,” Dr. Smith said. “Of those, about 40 percent will have some sort of abnormality in the MRI that would look like moyamoya.”

Initial studies suggested that about one in a million people have moyamoya, which extrapolates to roughly 310 cases a year in the U.S. Given the number of cases he sees each year, Dr. Smith thinks that is a gross under-estimation, especially if the percentage of children with sickle-cell is added.

**TREATMENT = SURGERY**

There are no effective drug treatments for this condition, so essentially, the solution is to build a new plumbing system. “The brain doesn’t care where the blood comes from, whether it’s from the carotid or some other place, as long as it’s getting blood with oxygen,” said Dr. Smith.

With moyamoya, it is impossible to reconstruct the internal carotids coming through the neck. Attempts to do angioplasty to stretch them open have failed because unlike atherosclerosis, moyamoya is a disease that covers the length of the artery, not just one spot.

There are two types of surgery — direct and indirect. Both use the same source of blood to build a new supply — branches of the external carotid artery in the scalp, which are typically not affected by moyamoya.

In the direct technique, a blood vessel in the scalp is cut in half and one half is sewn directly into an artery in the brain that supplies blood to the brain’s cortex. This supplies blood immediately, but it is harder to do on children because their blood vessels are so small.

The indirect technique uses the same scalp blood vessels, along with other blood vessels in the lining of the brain (the dura or meninges). These vessels are surgically implanted under the skull, and allow an entirely new system of blood vessels to grow — a process called angiogenesis. These new vessels take several weeks to months to grow. Indirect surgery is more commonly used in children; the direct technique is more common in adults.

The really good news with this procedure is that once done, there is almost never a need to do it again. Though their stroke deficits are not reversed, many children are able to lead normal lives.

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**Haleigh’s Story**

At the end of June 2012, Michelle Martin of New Orleans noticed her six-year-old daughter Haleigh experiencing involuntary body movements while attending a friend’s birthday party. Ten days later, Haleigh was diagnosed with an involuntary movement disorder called chorea and sent to Children’s Hospital of New Orleans to see what was causing it. After testing, she was diagnosed with moyamoya. “We were also told that she had already had four strokes, and the chorea was a result of those,” Michelle said.

Haleigh was given an “Arnold Schwarzenegger-dose of steroids for the chorea” and daily aspirin to help prevent future strokes. The Martins were told the only treatment for moyamoya was surgery, and her records were sent to Boston Children’s where she had indirect surgery on September 4, 2012. During the time between diagnosis and surgery, Haleigh had a fifth stroke.

As a result of her strokes, when Haleigh gets excited or experiences anxiety, the chorea movements return. She also has subtle learning and speech issues, which are noticeable to her family. She cannot take any medicine that can cause the blood vessels to constrict, like decongestants or antihistamines. “It can be hard watching her not getting any relief and having to let some things run their course,” her mother said. “Also, because she is restricted from contact sports and activities, as well as anything that can cause her to become overexerted, our biggest challenge is finding the balance between protecting her and allowing her to be a normal seven year old.”

After Haleigh was diagnosed, Michelle replayed earlier events and realized she was there when Haleigh had two strokes. “I was devastated and beat myself up that I didn’t recognize the signs,” she said. “Immediately after the surgery, it was like having a newborn again because my husband and I found ourselves checking to make sure she was breathing. Now, we try not to focus on what she can’t do, but on finding those things that she can do.”
Malachi’s Story

When Malachi Horton was 18 months old, he lost movement in his right arm and leg and was unable to sit, stand or walk. “We took him to three different emergency rooms before he was diagnosed as having a stroke,” his mother Erin Horton said.

At first doctors thought he had a clotting disorder, but a cerebral angiogram detected moyamoya disease. It also showed a number of possible TIAs. The Horton’s family doctor explained that Malachi would need brain surgery as soon as possible. “Without it, the strokes would continue until they took his life,” Erin said. To reduce the risk of another stroke, he was kept cool and well hydrated and given low-dose aspirin.

He had the indirect-type of surgery at Boston Children’s Hospital on June 29, 2010, and his prognosis is good. However, his stroke was not without consequences: his right side is weaker than his left, and he has limited fine motor skills in his right hand. “We also see him get angry or agitated quickly,” Erin said. “I have been told this is common in people with a history of strokes.”

Because of moyamoya, the Hortons are much more proactive about Malachi’s medical care. “We take time to ask questions and do our own research,” Erin said. “We are more vocal about our concerns and voice them when we feel they are not being addressed. Malachi’s ultimate diagnosis of moyamoya would not have changed, but perhaps the severity of his stroke would have changed if our concerns had been taken seriously at the first sign.”

Claire’s Story

Between August and November 2012, nine-year-old Claire Sargent had a series of excruciating headaches. Her doctor ordered an MRI, which showed abnormalities that proved to be three hemorrhagic strokes. She was referred to a neurologist near her hometown of Grand Rapids, Mich. A follow-up MRI showed she had moyamoya disease, and Claire was referred to a neurosurgeon.

She had indirect-type surgery in December, four months after her first headache.

“Claire is not progressing at the average rate for this kind of indirect bypass,” said her mother, Andrea Sargent. “Her prognosis is still unknown.” Her blood flow at her six month checkup was mostly unchanged. “She is still at extremely high risk of additional strokes and TIAs, but our hope is that each day her risk lessens.”

Claire was an honor roll student before her strokes, diagnosis and surgery, plus a TIA in April 2013. Today, “she has some cognitive deficits in the areas of working memory, sustained auditory attention and visual organization, with a cognitive impairment in the area of executive functioning,” her mother said. Her education going forward will be guided by a lengthy Individual Educational Plan. She started 5th grade in September.

Andrea doesn’t know if Claire’s deficits are due to the strokes or surgery. She tires more quickly and her thought patterns are different. “She is forever altered by what she’s been through,” Andrea said, “but remains a mostly happy-go-lucky pre-teen, although she struggles mightily now that things take longer and are harder to do. It causes tremendous frustration and anxiety, which is why she was recently diagnosed as having an adjustment disorder with anxiety.”

Claire can’t do a lot of activities that she used to because she’s still at an extremely high risk of stroke. Dehydration, over-stimulation and fatigue can increase her chances of having another stroke. Her doctor says her body is expending a lot of energy simply healing from the trauma of surgery.

When Claire gets tired, her balance also suffers. She has taken more than a few tumbles down the stairs after an overactive day. “For now, Claire can’t be her normal, overactive, bouncy, busy little self,” her mother said. “She needs to take things a little easier, something she’s not fond of doing.”
Arianna’s Story

In the summer of 2003, five-year-old Arianna Carter of Herrin, Ill. began having symptoms that no one could explain. “She was playing with her sister and her left side seemed to go numb on her,” said mother Tammy Carter. Fortunately she was okay within a few minutes, and Tammy chalked it up to rough-housing. That fall Arianna began school, and her teachers noticed she was having similar episodes and needed a nap afterwards.

Her parents took Arianna to a specialist who thought she had DeJorges Syndrome because of learning disabilities that seemed consistent with it. Tammy began researching to see what could be done to help Arianna.

The numbness episodes persisted. During one, Arianna’s face drooped. Alarmed, Tammy took her to her pediatrician, who recommended a neurologist. “When I explained her symptoms as stroke-like, the neurologist laughed and said ‘Kids don’t have strokes, Mrs. Carter. You must be over-exaggerating a seizure.’”

However, Arianna’s neurologist said if she continued to have episodes, to take her to the nearest ER and tell them her doctor wanted an MRI. “Soon we were in the local ER,” her mother recalled. “When I told them what the doctor wanted, I was told they weren’t going to do it — too expensive. They said children do things for attention. Or it could be an ear infection. And again, we were told that children don’t have strokes, but seizures are common.”

The family was dealing the best they could. Tammy watched her daughter’s every move. After another incident of strange behavior, Tammy took Arianna to the ER in another town. She was in the middle of a numbness episode when they arrived. They did a CT scan, and the physician said it looked okay to him, although there was a blur on it, probably because she moved. “He wasn’t sure what was going on with her but did tell us to get to her neurologist as soon as we could and to come back if she had more episodes,” Tammy said.

The neurologist called the next morning, after receiving Arianna’s CT scan. “I will never forget what she said,” Tammy recalled. “Arianna is a very sick little girl. Take her to your local ER. There an ambulance will pick her up and take her to St. Louis Children’s Hospital. You have to go NOW!” When Tammy asked why she couldn’t just drive her to St. Louis, two hours away, the doctor said that she might not survive the drive. “My heart dropped. I quickly made arrangements for my other daughter and called my husband. We were at the ER within 30 minutes.”

The doctor in St. Louis explained that the blur on the CT scan was a brain anomaly. Tammy was asked to explain the events of the previous months. “I was relieved to see someone taking me seriously,” Tammy said. Arianna was admitted for testing.

The test results showed Arianna had moyamoya disease. In January 2004 the Carters went to Boston for surgery, the direct technique. Further tests showed she’d had a major frontal lobe stroke and that the right side of her brain was 98 percent blocked and the left side 16 percent blocked. After surgery, Arianna had another massive stroke, which led to brain death. Her parents took her off life support on January 15, 2004.

After Arianna’s death Tammy was told she couldn’t hold the doctors in their small town responsible because “they just can’t be expected to know about these rare diseases. I find that unacceptable,” Tammy said. “So I try to educate as many as I can so that they do know. I don’t want another parent to be told, ‘Children don’t have strokes’ or ‘She’s just trying to get attention.’ I want to share her story to open eyes, not to dampen the hopes of anyone who has moyamoya. I want to help them prevent her story from being repeated.”

Moyamoya in Adults

The adult version of moyamoya disease typically starts affecting people in their mid-40s and progresses more slowly than in children.

In children and adults, twice as many females have moyamoya compared to males.

Ischemic stroke is the most common outcome for adults and children with moyamoya, but adults sometimes have bleeding strokes because of higher blood pressure.
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Health Insurance Marketplaces opened in every state plus the District of Columbia in October. These Marketplaces provide a single location in each state where individuals and small businesses can shop for private health insurance that fits their budgets. For the first time in our nation’s history, people will not be turned down or charged higher premiums for pre-existing medical conditions. Financial assistance will be available to help make premiums affordable for people who don’t get health insurance through their workplace.

THE MARKETPLACES — THE PLACE TO START

The Marketplaces — sometimes referred to as exchanges — provide a new, easier way to shop for and buy health insurance. Through the Marketplaces, you can choose a private health plan that best meets your family’s needs and budget. You can see all of your choices in one place, determine your premium, deductibles and out-of-pocket costs up front and compare health insurance plans before you decide to enroll. Depending on where you live, your Marketplace could be run by your state or by the U.S. Department of Health and Human Services. You’re eligible to buy coverage through the Marketplaces if you’re a U.S. citizen or lawfully residing in this country and are not incarcerated. Small employers with 50 or fewer full-time workers can use the Marketplaces to shop for coverage for their employees.

If you already have health insurance through your employer, as most Americans do, you’ll still be eligible for coverage under your employer’s plan and won’t need to use the Marketplace. Your coverage also will continue if you’re covered through a public program like Medicare, Medicaid or another government plan.

PENALTY FOR NOT BEING COVERED

As of Jan. 1, everyone will be required to have some type of health insurance coverage, unless no affordable insurance is available. Individuals have until March 31, 2014, to obtain coverage and still be in compliance with the requirement.

Those who can afford coverage but don’t obtain it will be subject to a tax penalty. In 2014, this penalty will be $95 per adult or 1 percent of your taxable income, whichever is greater.
The new Health Insurance Premium Tax Credit will help make the premiums affordable to qualified individuals and families buying health insurance coverage through the Marketplaces. The amount of the tax credit will depend on income and family size. Those with the lowest incomes may be eligible for a health plan with no premium.

Qualified individuals can choose how they receive their tax credit. They can take it right away and pay a lower monthly premium, take it at the end of the year by subtracting it from the taxes they owe or use it to get a bigger refund.

Every health insurance plan sold through the Marketplaces is required to offer ten categories of Essential Health Benefits, covering services important to heart disease and stroke patients. They include hospital care, doctor’s office visits, emergency services, prescription drugs, lab tests, rehabilitative care, and preventive screenings and services. The services covered are intended to be similar to the benefits provided by a typical employer plan.

There are a variety of ways to shop, apply for and enroll in health coverage: online through the Marketplace website (healthcare.gov), in person, by mail, or by phone (1-800-318-2596).

Every Marketplace is required to have available trained, certified counselors called Navigators. Navigators are not insurance agents, but people who can help you understand and complete the application and explain the different insurance options. In addition, licensed insurance agents and brokers can help you apply and enroll.

A three-page application will allow individuals to find out whether they qualify for the premium tax credit or for free or low-cost coverage through Medicaid or the Children’s Health Insurance Program.

The Marketplace website and application are available in Spanish. The toll-free number can also help people in Spanish and up to 150 other languages.

Some consumers using the website to shop and apply for coverage have experienced difficulties. Although changes are being made to improve the website, remember that you can also apply by phone or in-person through Navigators and insurance agents.

Once the initial open enrollment period ends, people who are uninsured have to wait until the next open enrollment period to sign up for coverage. However, you may be eligible to enroll outside the open enrollment period in special circumstances, including losing your health insurance coverage, getting married or moving. If you have or adopt a child, that child is immediately eligible to be signed up for coverage.

Finally, individuals who qualify for Medicaid or the Children’s Health Insurance Program can apply and be enrolled at any time throughout the year.

Starting Jan. 1, individuals with pre-existing medical conditions can no longer be turned down for coverage or charged a higher premium because of their health status. Also, for most health plans, not just plans sold through the Marketplaces, annual dollar limits on coverage are no longer permitted. Most health plans will now be required to cover the routine costs associated with participating in clinical trials for patients with life-threatening conditions.

STOP    Ask for identification and a phone number where the person may be reached later. If the person refuses to give this information for any reason, or tries to pressure you into signing any document, you should immediately hang up, close the door or walk away.

Do NOT volunteer your Social Security number or a credit/debit card number to anyone offering insurance unless you personally know them. Likewise, do NOT sign any paperwork or write a check.

CALL    Contact your state department of insurance or your state Marketplace. The insurance company or agent or broker, as well as the Navigator, must be registered or licensed in your state before they can sell coverage or counsel consumers through your state’s Marketplace.

CONFIRM    Before giving personal information or signing any documents, confirm the authorization status of any insurance company, agent or broker, or Navigator trying to provide assistance.

From National Association of Insurance Commissioners website
Studies show younger people are having strokes more often than even a generation ago. As a result, more children are being raised in stroke families. And we’ve talked to enough survivor and caregiver parents to know that they are concerned with how the stroke affects their children. With that in mind, we talked with two sisters about their experience growing up in a stroke family.

Ashley Park Pryor was five years old and her sister, Lisa, was three when their father, Steve Park, had a hemorrhagic stroke and brain surgery as the result of a ruptured AVM. Overnight, the Park family’s lives changed dramatically. Steve could no longer work in refrigeration and air conditioning repair, which had provided a comfortable life for his family. The girls’ mother, Charlene, who had been a stay-at-home mom, had to go to work to support the family. They went from enjoying a secure middle-class life to struggling to keep their house and car and even to put food on the table. “It was a lot of beans and potatoes and hamburger for the first couple of years,” Charlene said. It took nine months before they started receiving Social Security Disability benefits.

Consequences

“I think Dad’s stroke changed my childhood completely,” said Ashley, who is now 27 and lives in North Texas with her husband. “When he went in for surgery to remove the AVM, I had to stay with friends of the family. It was the first time I had ever spent any significant time away from home, and I did not adjust well to it.” After Steve came home, the carpets were removed and toys had to be put away so he wouldn’t trip. Because of weakness on one side, he could no longer pick the girls up or chase them or do other “dad-things.”

“I feel that Lisa and I had to grow up faster than other kids our age,” Ashley said. “For instance, we immediately started doing our own laundry and other chores around the house. I’m pretty sure no other girls at my school were regularly mowing the lawn and helping with plumbing, air conditioning and car maintenance.”

A positive effect of their father’s stroke was that the girls got to see their father more. “Before the stroke, Dad worked pretty long hours,” said Lisa, now 25 and living in North Texas. “After the stroke, he was the stay-at-home parent. He was always there to help me with homework or anything else I needed.” The stroke happened so early in both girls’ childhood, it was just something that was part of him. Lisa has some fuzzy memories of the pre-stroke life, “but they’re really indistinct. In terms of actual distinct memories, this has always been the norm,” she said.

A different kind of education

Despite his stroke, Steve still knew how to fix things, and Ashley became his “extra hand.” In the process, she became adept at auto and air conditioning repair as well as plumbing. “He still taught me things that dads teach,” she said. “He taught me to drive, change a tire, change the oil. Because of his lessons, I know how to parallel park in very tight conditions. He taught me how to fish, including gutting and cleaning what we caught.” Even though she’s married and no longer lives with her parents, she and Steve recently replaced the brakes of Charlene’s car.
Early on, living with a stroke survivor showed Ashley how much discrimination there is toward people who are different. Charlene recalled being at the grocery store with the girls and seeing a disabled person, and Lisa asked if he’d had a stroke. “I told her I didn’t know,” Charlene said. “And then she said, ‘We don’t want to stare.’ In school both girls gave up PE time to work with the developmentally disabled children.”

The family’s situation taught the daughters the value of determination. “I know my parents are really strong to have something so life-changing happen to both of them, and yet they worked with it. Neither of them ever gave up,” Ashley said.

When it became clear he could no longer work as a repairman, Steve went back to school. “Seeing Dad go back to school had a huge impact on how I view education,” Lisa said. “He was incredibly diligent and dedicated to his studies, so that was an inspiration for me to try as hard as I could, too.” Lisa recently graduated from Texas Women’s University.

The stroke affected the girls’ view on gender roles because their mother and father essentially switched places after the stroke: “Dad was home taking care of my sister and me and Mom was working. Before, it was the opposite.” Ashley said. “I think helping Dad with the cars, plumbing and air conditioning also impacted my views on what tasks are gender-segregated.”

Recently Ashley was diagnosed with asthma. “I like to think I follow Dad’s example of not focusing on or mourning all the things I can’t do now. Instead I try to focus on figuring out what all I can still do, despite the asthma.”

Another dad-thing Steve did for Ashley — walking her down the aisle when she got married a few years ago.

Advice for others

Charlene remembers how scared the girls were that they were going to lose their father, so she took them to the hospital after surgery, even though they could not go in the room. “My advice to parents is don’t hide anything from your children,” she said. “That is the worst thing you can do. I couldn’t believe the relief on their faces when they saw that Daddy was still alive.”

“When you’ve been through that kind of trauma,” Steve said, “everything else is secondary — bad grades, car wreck, losing a job. I think they learned that young and grew up with a strong set of values. I think we all learned that relationship matters more than things and money.”

95% Say It’s Important To Receive SC

For years the regular surveys of our readers have let us know that almost everyone thinks it’s important to extremely important that they continue to receive the great information in Stroke Connection. Please help us continue to serve as many stroke families as we possibly can by making your gift of support today. We appreciate you.

strokeassociation.org/scgive
SmallTalk™ from Lingraphica

The SmallTalk Family of 13 free apps offer users extreme portability for practicing their speech or communicating more effectively on the go.

The SmallTalk Aphasia App turns the iPod Touch, iPhone, or iPad into a mobile accessory to the AllTalk device. It allows users to take their favorite icons, phrases and videos from the AllTalk with them. This app provides a vocabulary of pictures and videos that talk in a natural human voice (male or female). While these apps are designed to work with the AllTalk speech generating device, they can be used on their own.

aphasia.com/products/apps/smalltalk

In addition, there are apps for dysphagia, oral motor exercise, conversational phrases, daily activities and pain scale, among others.

MyTalkTools (free lite version)

MyTalkTools is an app that displays a picture of words, and the survivor simply taps the picture to speak the idea. The screen has a large green checkmark for yes and a large red x for no. It’s designed for survivors with aphasia who can’t recognize words. MyTalkTools also has categories for items such as food, drinks and greetings. If the survivor’s aphasia does not allow him or her to say or recognize words, he or she may be able to communicate through the pictures in this app.

mytalktools.com/dnn/Purchase.aspx

Rehab Equipment Exchange

This national network is a FREE service to facilitate the exchange and transfer of used rehabilitation equipment. Individuals can use the website to buy, sell or donate used rehab items of many types, including wheelchairs, mobility aids, ADL aids, communication devices, assistive technology and exercise equipment.

Developed in 2004, this website is managed by a physical therapist. Hundreds of rehab items have been exchanged in the United States and a few other countries. This is a valuable resource for stroke families who only need equipment for a short time, need items that insurance doesn’t cover, cannot pay for pricey new items, or have used equipment to sell or donate.

rehabequipmentexchange.com

These apps, tips and information are provided as a resource to our readers. They have not been reviewed or endorsed by the American Stroke Association.
Fingernail Tips from Survivor David Layton

Tip #1: One thing I have found particularly helpful is a fingernail brush attached with stainless steel screws to a board cut to fit in the bathroom sink. I use it to clean my hand and fingernails after working in my garden. The board helps secure the brush while I scrub my fingernails and hand across the brush.

Tip #2: Another idea for fingernail maintenance is to use a spring clamp to hold a fingernail file to a desktop or countertop, so I can move the nails of my working hand across the file. We all got the idea that nail clippers were necessary for nail maintenance back when we had two working hands, but filing without using nail clippers will do the job.

Tips from Survivor Adrienne Statfeld

I have weakness in the fingers of my left hand. I’ve found the following items to be helpful during my post-stroke life.

• **Elastic shoelaces** to replace laces that need to be tied. Now I only buy slip-on shoes and sneakers that come with elastic bungee laces.
• **Button-hook device** — Using this device makes getting dressed and undressed easier. It only requires moderate dexterity from the fingers on one hand while buttoning a shirt without the device requires the cooperation of the fingers on both hands. Of course, clothing with fewer fasteners or hook-and-loop fasteners instead of buttons or zippers would be easier, but would require that I buy a new wardrobe.

• **Extra-long shoehorn** — Saves unsteady survivors from bending over to get their shoes on.
• **Dental floss** that comes attached to a plastic pick so only one hand is necessary. Just because you’ve had a stroke doesn’t mean you can neglect dental hygiene.
• **“Prep n Pop”**: This device, advertised on infomercials, is a winner. It allows you to stab a fruit or vegetable and hold it in place while you peel or slice it.

I suggest you keep items, even if you feel you no longer need them. Shower seats, grab bars and over-the-toilet commodes might seem less important following recovery, but they can decrease the likelihood of a fall.

New Survivor Tips Library on the American Stroke Association Website

The American Stroke Association has recently launched a new area in our *Life After Stroke* section of the ASA website specifically for survivors to share their tips and tricks for all kinds of activities of daily living. This site is powered by volunteers who submit their own videos, photos or written tips for everything from cooking techniques, to grooming and bathing tips, to tips for communicating with someone with aphasia.

Many of these tips are also being featured on the ASA’s Facebook page. Each week we share tips on #WhatWorksWednesday. Follow us at [facebook.com/strokeconnectionmag](http://facebook.com/strokeconnectionmag) or [facebook.com/americanstroke](http://facebook.com/americanstroke) to have these weekly tips show up in your Facebook feed every Wednesday. You can also be a part of #MotivatedMonday and #TalkItOutTuesday as people share the little things that keep them motivated and discuss various stroke-related topics.

Do you have some great tips or adaptations that could help other survivors? Or looking to find new and better ways to do some things yourself? Visit [StrokeAssociation.org/tips](http://StrokeAssociation.org/tips) to check out the latest tips or to submit one of your own.
Books About Recovery

Brain Injury Rewiring for Survivors
By Carolyn E. Dolen
Idyll Arbor, Inc. | ISBN 978-1-82883592
Learn how to rewire spiritually, emotionally, cognitively, physically, socially and vocationally. This book has simple solutions that will help you climb the recovery mountain. Every chapter has ideas to help you get back into life. The author is a brain injury survivor. She also wrote a companion book for caregivers: Brain Injury Rewiring for Loved Ones; ISBN 978-1-82883714.

Live or Die — A Stroke of Good Luck
By Richard L. Burns
This is a memoir about the author’s recovery from a stroke he had more than 40 years ago when he was 38. He went on to a successful career in advertising as the creator of the Fruit of the Loom characters. This is the story of his recovery, what he learned from the ordeal and how his life and values changed with each step he took to heal himself.

The Stroke Recovery Book: A Guide for Patients and Families
By Kip Burkman, MD
The author is a rehab physician who has worked with thousands of stroke patients and their families. He understands the fear and confusion they experience. He provides answers to many commonly asked questions about personality changes, emotional and cognitive impairments, speech and language problems, hemiparesis, and bladder and bowel problems. He also answers questions about medical equipment and the type and level of care needed at home.

Managing Stroke: A Guide to Living Well after Stroke
Editors Paul Rao, Ph.D.; Brendan Conroy, M.D.; Christine Brown, M.A., C.C.C.
National Rehabilitation Hospital Press | ISBN 978-09661676-8-9
This is an easy-to-use guide from a nationally recognized rehabilitation hospital. It covers stroke from every angle — medical, emotional and social — and puts the power of recovery in the patient’s hands. It covers everything from recurrent stroke to sex after stroke, to navigating the complexities of insurance coverage.

Between Hope and Despair: Living after a Stroke
By Rick and Nancy Davis
Booksurge | ISBN 978-1-4392-3040-4
Survivor Rick Davis teamed up with his wife Nancy to tell the story of how he recovered in less than two years and ultimately walked a half-marathon after an ischemic stroke initially left him completely paralyzed.

On the Dark Side of the Moon: A Journey Toward Recovery
By Mike Medberry
In the spring of 2000, Mike Medberry, a longtime conservation advocate, had a stroke in the remote wilderness called the Craters of the Moon in Idaho. He was rescued after lying for hours alone in one of the harshest, yet most beautiful landscapes in the country. This story interweaves Medberry’s own struggle to speak, walk, write and think with the struggle to protect this brutal, lava-bound landscape. Medberry’s recovery and his struggle to protect the Craters of the Moon is a story of renewal, restoration, accommodation and ultimately, of finding workable compromises to some of life’s most difficult problems.

These summaries are provided as a resource to our readers. The books have not been reviewed or endorsed by the American Stroke Association.
It was New Year’s Day in NYC, week-old snow molded over the curbs, and the low January sky looked like a gigantic sheet of brushed aluminum. The doors of St. Vincent’s Hospital opened magically, allowing Marilyn to wheel me into the subzero arctic air, all but scattering rose petals in our path. After two months of medical incarceration, the powers-that-be decided I was ready to face the outside world…or my insurance ran out. Either way I was a free man.

If ever a wheelchair needed a heater it was that morning, but I was so pumped up my metabolism was burning hotter than a nuclear meltdown. I leaned back as if I were in a Barcalounger, closed my eyes and let the idea of never being stabbed again with a syringe at 4 in the morning sink in. Then I turned to Marilyn and said, “We’re outta here.”

On that note we coasted to the corner, hung a left and headed home. As we cruised along I felt like George Bailey running down Main Street in “It’s a Wonderful Life,” savoring all those things he once took for granted. Of course Greenwich Village is slightly different than Bedford Falls.

In my case I was looking forward to seeing Tammy, the transgender manicurist at “Think Pink” who has so many piercings she resembles a walking “connect the dots” game. Or Julio, “Planet Donut’s” barista, with a dozen glazed crullers labeled “Honey-Dip” tattooed on his forearms. And JB, the skeletal owner of “Partners & Crime” mystery bookstore who dresses like Gene Simmons, kabuki makeup and all. Oddballs? Yup. But they were my oddballs. And I knew I’d get a nice homecoming wave as we passed by.

Instead, I did a series of theatrical double takes, because my good old West Village ‘hood had turned into a suburban mall littered with “Capital One,” “Starbucks” and “Super Cuts.” All we needed was a “Sunglass Hut” to complete the picture. I looked up at Marilyn and asked, “How long was I in that bed-pan hotel again?” It was as if I had been in a long, Rip Van Winkle-esque sleep and woke up to an unrecognizable world, not to mention an unfamiliar body.

Then I spotted a red illuminated sign that hadn’t morphed into something else. Who would have thought I’d have warm fuzzy feelings about a chain like “CVS.” Still, I knew the cashier with the gold tooth who calls me “doll face” in her husky Lauren Bacall voice would give me that quirky Village flavor I longed for. Silly? Yes. But these were the kind of day-to-day connections I missed in the hospital. I said to Marilyn, “You know what I’m in the mood for? Making change.” Finally, those 60 days of excruciating cognitive math exercises could be put to practical use. So in we went.

But alas, “CVS” had gone DIY. We immediately found ourselves surrounded by a barrage of female robo-voices prissily demanding, “CHOOSE YOUR LANGUAGE…SCAN YOUR REWARDS CARD…PLACE YOUR ITEMS IN THE BAG…PUT THE COUPON IN THE SLOT…TAKE YOUR RECEIPT!” Not a “doll-face” in the bunch. Sure, they prefaced everything with the word “please,” but you know they didn’t mean it. And when did we go from being customers to employees having to do everything ourselves — minus the benefits and 401(k)?

Nevertheless, putting up with this grief is way better than a 4 a.m. syringe-stabbing. Hey, it was January 1 and I was back among the living. A new year, a new world, a new me. I’ll adapt.
Could you have symptoms of spasticity?

Did you know the extreme tightness that can appear in the upper limbs after a stroke can be a treatable condition? This condition—known as spasticity—can develop months after your stroke and is sometimes left untreated. Whether it's difficulty buttoning your shirt or washing your hands, spasticity can turn the simplest everyday activities into the most complicated tasks. If you’re suffering from spasticity, help is out there. Talk to your doctor about your symptoms today. Relief is possible.

to create a personalized doctor discussion guide.