Strokes and Down Syndrome
Brian Chicoine, MD
Medical Director

Strokes occur when the blood supply to the brain is compromised, which deprives the brain of needed oxygen and glucose. Strokes can be caused by atherosclerotic disease (a disease in which the inside of the arteries becomes narrower due to the build-up of plaque). Did you know that strokes due to atherosclerotic disease have been found to be less common in people with Down syndrome? Similarly, myocardial infarctions (heart attacks), which can also be caused by atherosclerotic disease, are less common in people with Down syndrome. Overall, atherosclerotic disease is less common in people with Down syndrome. The reason for this is not known. Some have called Down syndrome “atheroma-free,” meaning people with Down syndrome develop little narrowing of the arteries due to plaque.

However, some people with Down syndrome still get strokes. Why is that the case if there is less development of atherosclerosis?

There are 4 conditions that may predispose a person with Down syndrome to a stroke:

1. **Heart disease:**
   In some individuals who had congenital heart disease (heart disease we are born with) or other disease of the heart valves, structural changes in the heart may cause a situation in which a clot may form within the heart (not in the arteries to the heart). If some of the clot “breaks free,” it can travel to the brain and cause a stroke.

   In addition, some individuals with congenital heart disease have a hole between the left side of the heart and the right side of the heart. In that situation, if the person develops a blood clot in their veins (as opposed to in their arteries) the blood clot can break loose, travel through the heart, and go into the brain. In this situation, it typically presents as a person having a longstanding “hole in the heart” who then develops a blood clot in their leg veins that breaks loose and travels through the heart to the brain and causes a stroke. However, it should be noted that this scenario is not common.

   Also, some individuals are predisposed to abnormal heart rhythms (sometimes as a complication of other heart problems) and some of these rhythms are associated with increased clots within the heart that, again, can cause a stroke.

2. **Moyamoya disease:**
   This is more common in people with Down syndrome. It is associated with blockage of arteries in the back of the brain and the development of arteries for blood to flow around the blockage. When Moyamoya causes a stroke, most commonly it is due to...
reduced blood flow due to the blockage. However, it can also cause strokes due to bleeding into the brain.

3. **Hypercoagulation disorders:**

   People with Down syndrome seem more predisposed to developing conditions in which the blood clots more readily than normal. If the location that it clots is in an artery in the brain, it can cause a stroke. Some of these conditions are caused by auto-immunity (a condition in which the body’s immune system attacks cells in other parts of the body as if they were foreign or infectious cells). Some auto-immune diseases such as Systemic Lupus Erythematosus can cause abnormal clotting. Primary coagulation disorders (e.g. Factor V Leiden mutation, Protein S deficiency, and others) can also cause abnormal clotting and lead to strokes.

4. **Alzheimer’s disease:**

   In Alzheimer’s disease, the build-up of plaque associated with amyloid (different than the build-up of plaque in atherosclerotic disease) can cause blockage or bleeding from small blood vessels, usually in the deeper structures of the brain. This is called microangiopathic disease or amyloid angiopathy. Due to a greater tendency for the build-up of amyloid in their brains, people with Down syndrome seem more susceptible to strokes from this cause. Strokes from this would occur more commonly later in life.

In addition, while not actually a stroke, the effect of a seizure may, at times, mimic the symptoms of a stroke. Usually those symptoms revert to normal after a period of time following the seizure.

If a person with Down syndrome presents with a stroke, particularly if they are younger, a careful evaluation should be done to assess for the cause. We recommend discussion with your health care provider to review assessment and treatment plans. Due to the variety of conditions contributing to strokes in people with Down syndrome, consultation with a neurologist, cardiologist, hematologist, and/or other specialists may be indicated after discussion with your provider.