

# Growing Up Different

## Activity 3: Grades 9-12 Williams Syndrome



As you have learned, Williams Syndrome is a rare disorder (1 in 25,000 births) characterized by physical and developmental problems. Scientists have tracked this disorder to a mutation on chromosome 7. People with Williams Syndrome are missing a small segment of this chromosome.

Most people with Williams Syndrome also lack the gene responsible for the manufacture of elastin. This gene maps to the same missing area of chromosome 7.

Elastin is a protein that allows the tissues within our blood vessels, skin and heart to stretch. Without this elastic property, the function of the tissues is compromised.

In addition to having a compromised elastin level, most people with Williams Syndrome experience the narrowing of blood vessels. Both the lack of tissue resiliency and the narrow diameter of the vessels produce hypertension (high blood pressure). Narrowed vessels within the heart and lung further exacerbate the condition and may require surgical correction.

### **OBJECTIVE**

This activity page will offer:

- An insight into elastin and its association with Williams Syndrome
- An experience in taking blood pressures
- An observation of how vessel diameter affects pressure

### **Blood Pressure Connection**

People with Williams Syndrome must have their blood pressure monitored periodically. In a doctor's office, this procedure is most often performed with a

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### Part 1 - Measuring Blood Pressure

#### QUESTIONS

1. Why is it important for people with Williams Syndrome to have their blood pressure monitored periodically?  
**(Most people with Williams Syndrome will have some sort of vessel or heart problem. A blood pressure measurement offers a "window" on the functioning of the circulatory system.)**
2. What do the two numbers of a blood pressure mean?  
**(The first number (systolic) identifies the maximum surge pressure of the blood. The second number (diastolic) identifies the resting pressure within the vessel.)**

### Part 2 - Compromised Diameter

#### QUESTIONS

1. Through which of the two tubes did you blow the hardest?  
**(The straw.)**
2. Why was it easier to blow air through the larger diameter tube?  
**(The larger diameter offered more space for moving air. Since there was less resistance, the air flowed with less effort.)**
3. If we were to apply this experience to Williams Syndrome, what would the straw and paper tube represent?  
**(The straw represents the smaller diameter blood vessels that are often present in people with this syndrome. The wider-diameter paper tube represents the larger diameter of healthy blood vessels.)**

cuff, stethoscope and mercury-filled manometer. At home, it can be done using an automatic blood pressure monitor.

The automatic device will measure blood pressure and display it as two numbers--e.g., 110/80. The first number is the systolic pressure (110). The systolic pressure is the force of the maximum surge. The second number (80) is the diastolic pressure. This is the "resting pressure" or baseline from which the systolic arises.

## **PROCEDURE**

### **Part 1-Measuring Blood Pressure**

#### **Materials**

- Blood pressure monitor (automatic cuff)

#### **Steps**

1. Work with a partner. Before obtaining a pressure, deflate the cuff. You can release air from the cuff by pressing the release valve on the bladder.
2. Position the cuff around the upper arm. Secure it with the Velcro strap.
3. Inflate the cuff with a series of quick pumps. The pressure should go up to about 150. Stop pumping.
4. The air will automatically seep from the cuff. As the air escapes, the device will begin monitoring the blood vessel, recording both the systolic and diastolic pressures.
5. As the cuff deflates, the systolic pressure will be the first reading recorded and displayed.
6. The next reading is the diastolic pressure.
7. Obtain three measurements for the right arm.
8. Record your blood pressure. Do you have high blood pressure?\* In adults, hypertension is diagnosed as a blood pressure of 140/90. In kids (who have more resilient and less obstructed vessels), hypertension is diagnosed at lower blood pressures. Hypertension is also influenced by other factors such as exercise, diet and lifestyle.

\*If you have high blood pressure, see the school nurse to find out what you can do about it.

## **QUESTIONS**

1. Why is it important for people with Williams Syndrome to have their blood pressure monitored periodically?
2. What do the two numbers of a blood pressure mean?

## **PROCEDURE**

### **Part 2 - Compromised Diameter**

#### **Materials**

- Drinking straw (new)
- Paper (clean sheet)
- Tape

#### **Steps**

1. Roll a sheet of clean paper into a tube. The tube should have a diameter of about 1 inch (about 2.5 cm).
2. Blow through the tube as hard as you can. Note any resistance or difficulty in blowing air through the tube.
3. Remove the straw from its wrapping. Blow through the straw as hard as you can. Note any resistance or difficulty forcing air through the straw.

#### **QUESTIONS**

1. Through which of the two tubes did you blow the hardest?
2. Why was it easier to blow air through the larger diameter tube?
3. If we were to apply this experience to Williams Syndrome, what would the straw and paper tube represent?

#### **Applying What You Have Observed**

How might a decreased diameter affect the health of a blood vessel? To compensate for the restriction, the heart must pump harder to move blood at the same rate through the vessels. This harder pumping action raises blood pressure. At a high enough pressure, the blood vessels may rupture and cause a stroke.

#### **EXTENSIONS**

##### **Resilient Pump**

The heart and other circulatory structures contain elastin. Elastin allows a structure to stretch, then return to its "resting" length. If muscle cells can only actively contract, what is the relationship between elastin and the pumping action of the heart tissue?

With your instructor's permission, examine the bladder of a blood pressure monitor. If possible, detach the bladder from its connecting tube. Observe the function of the one-way valve. How does this valve control the flow of air? How is the bladder similar to a heart? How is it different? Although the bladder does not contain elastin, it behaves as if it does. Explain.

## **Aging and Elastin**

Elastin is manufactured primarily when we are young. The older we get, the less elastin is produced. Think about it. What is the connection between this protein and the appearance of wrinkles? How might research breakthroughs affect the appearance of skin as we age?

## **The Old Fashioned Way**

Learn how to take blood pressures using a manual cuff and stethoscope [here](http://www.madsci.org/experiments/archive/859422898.Bi.html).  
([www.madsci.org/experiments/archive/859422898.Bi.html](http://www.madsci.org/experiments/archive/859422898.Bi.html))

## **Web Connection**

### **[Overview of Williams Syndrome and its symptoms](http://www.williams-syndrome.org/facts.htm)**

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### **[Informational site for parents, family members and others posted by the Williams Syndrome Foundation](http://www.wsf.org/)**

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### **[A site that describes elastin and its role in the body](http://www.wsf.org/MEDICAL/layreader/elastin.htm)**

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The activities in this guide were contributed by Michael DiSpezio, a Massachusetts-based science writer and author of "Critical Thinking Puzzles" and "Awesome Experiments in Light & Sound" (Sterling Publishing Co., NY).

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