

I Was the Clumsy One:

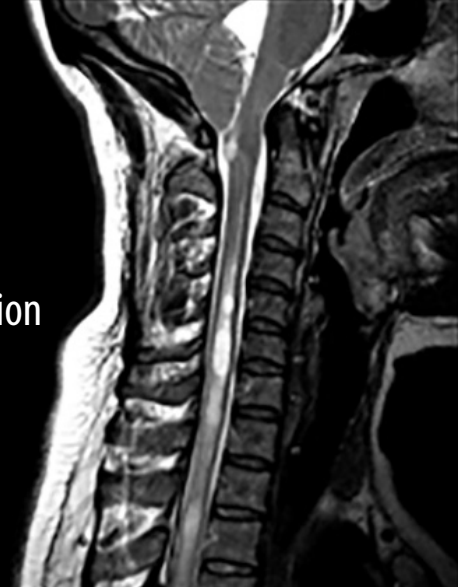
Exploring the Central Nervous System through Type I Chiari Malformation

by

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In this case study, Stacy Ropp, a patient with Chiari malformation, narrates her experience with this condition in four parts. In Part I, she discusses her signs, symptoms, and initial diagnosis. In Part II, she explains her initial treatment; in Part III, she reveals recurring symptoms, and in Part IV, she shares how a second opinion results in the more successful treatment of her condition. Begin this case study by using the following link to watch a video of Stacy explaining her experiences with type I Chiari malformation: <<https://youtu.be/yzP2qkTBiSY>>

Part I – Signs/Symptoms and Initial Diagnosis

As a child, I was the “clumsy” one. The family joke is that I could fall off a chair while sitting on it. I experienced other symptoms over the years, but for me, they were “normal,” such as intense pressure at the base of my skull with sneezing and coughing, the inability to raise my head for an extended period without getting dizzy, and difficulty with memory or concentration. I would gag every time I brushed my teeth and often suffered from severe headaches that started at the base of my skull.

As an adult, my condition finally reached a point that I could no longer disregard my symptoms. In February of 2010, I fell down the steps carrying a carpet cleaner. By the next day, I had severe pain and back spasms, which made standing upright difficult. Since I was seven months pregnant, I opted to see a physiatrist rather than using pharmacological interventions. A physiatrist specializes in the musculoskeletal system and uses exercise and adaptive devices to help patients become functional and pain-free. Since I was pregnant, I could not take most pain-relieving types of drugs because they are contraindicated and could potentially harm the fetus. Contraindications are conditions that prevent a patient from receiving treatment, and pregnancy is a common contraindication for many treatment options.

A physical examination revealed an abnormal Hoffman’s reflex (finger flexor reflex) and hyper-reflexivity (over-responsive reflex activity). These results point to muscular sclerosis (MS) or another central nervous system issue. The plan was to have a complete brain MRI after I delivered my daughter in June 2010.

In July 2010, I underwent brain magnetic resonance imaging (MRI; see Figure 1 on p. 3 below). The initial diagnosis was MS. However, after reviewing the images, the physiatrist questioned a diagnosis of MS because I had a nine-millimeter cerebellar tonsillar herniation. The cerebellar tonsils are on the undersurface of the cerebellum, and cerebellar tonsillar herniation is the elongation of the tonsils through the foramen magnum of the skull and into the spinal cavity. I also had a syrinx of the spine, which is an enlarged fluid-filled cavity in the central canal of the spinal cord.

The physician finally diagnosed me with type I Chiari malformation. The physiatrist immediately referred me to a local neurologist. After researching my new diagnosis, I understood the symptoms I experienced over the years.

In the summer of 2014, I had increased dizziness, which affected my ability to do yoga and personal training. I managed to work through the dizziness with yoga and knew what yoga positions I was unable to do for long periods. The neurologist referred me to physical therapy (PT) as he thought my symptoms were unrelated to Chiari. However, after

doing the course of PT, I did not have any improvement. Indeed, I added dizziness to the symptoms I would have to cope with for the rest of my life.

In February 2015, while doing my routine yoga practice, I had a near syncopal episode (short-term loss of consciousness). Each time I would go into a downward position, I would get lightheaded and feel as though I would pass out. I stopped doing yoga ... one more thing I would not be able to do.

Questions

1. Stacy’s MRI image results indicate abnormalities of the cerebellum of the brain and spinal cord. In what division of the nervous system are the brain and spinal cord?

2. Fill in the table below by listing the six major brain regions and describing their functions.

<i>Brain Region</i>	<i>Functions</i>

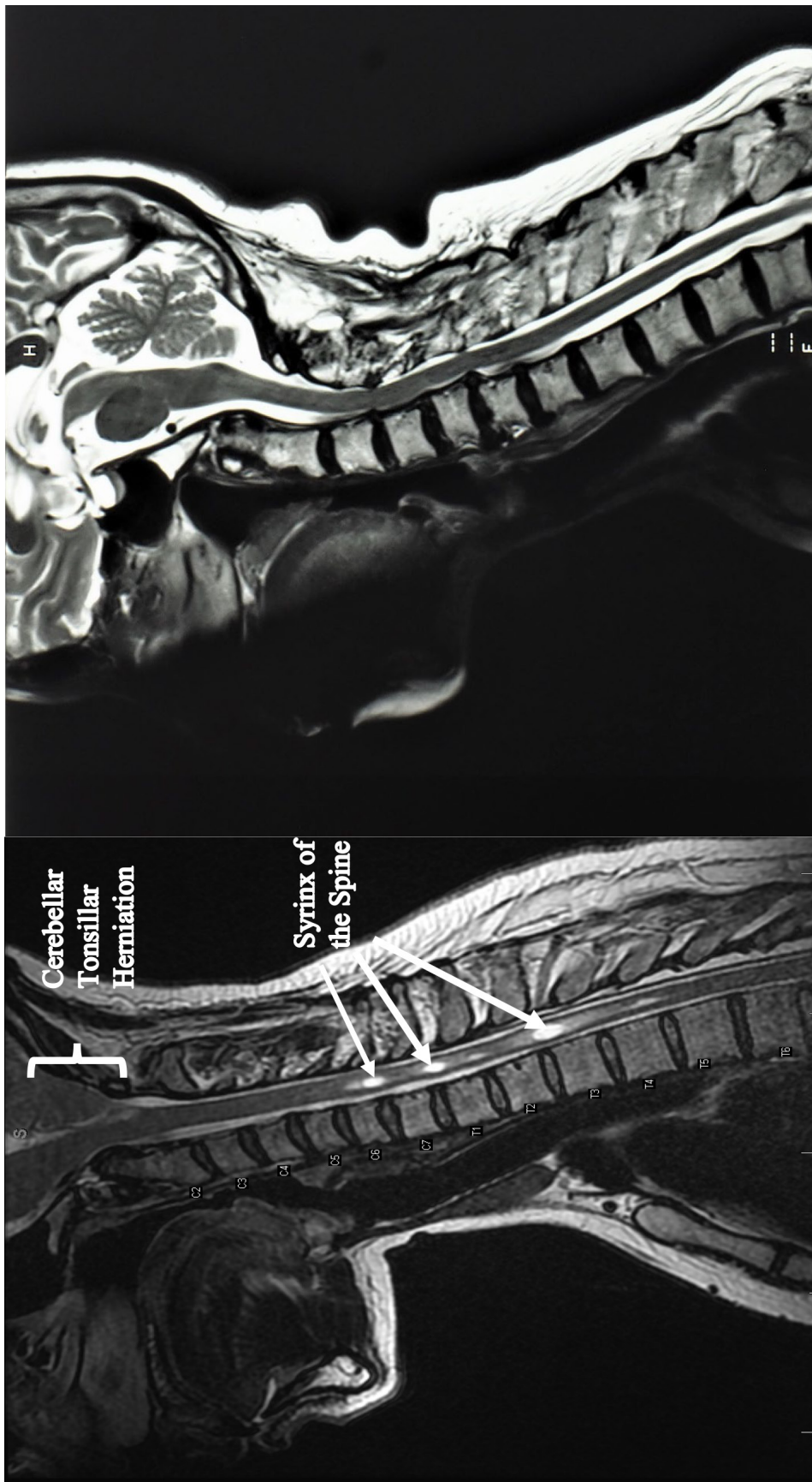
3. Use Figure 1 to answer the following questions:
 - a. What sectional plane did clinicians use to obtain this view of the brain and spinal cord (frontal, transverse or sagittal)?

 - b. What is the normal curvature of the cervical spine, and how does this compare to Stacy’s cervical spine?

 - c. Note the difference in the location of the cerebellum between Stacy and the healthy image. Clinicians refer to this defect as cerebellar tonsillar herniation of the brain. Use Figure 1 and a credible source to explain why patients with Type I Chiari malformation present with cerebellar tonsillar herniation.

 - d. Note the white patches on the spinal cord of Figure 1(A). This is an indication of syrinx of the spine (syringomyelia). Use Figure 1 and a credible source to describe the syrinx of the spine and compare it to a healthy spinal cord.

4. Stacy had an abnormal Hoffman’s reflex (finger flexor reflex), which indicates issues with the central nervous system. Clinicians conduct this reflex test by applying pressure to the middle fingernail bed of a patient’s middle



(A)

(B)

Figure 1. MRI image (A) depicts Stacy's spinal cord, which clinicians used to diagnose her with Chiari malformation. Image (B) is an MRI scan of a healthy spinal cord. The cerebellar location shown in (A) is indicative of a tonsillar herniation. Also, (A) has white patches throughout the spinal cord, which indicate the presence of a spinal syrinx; additionally, the cervical spine is straight. Note that these three abnormalities are absent on the normal (B) image and fit the diagnosis of Chiari malformation.

finger. An abnormal test is the flexion of any phalange and possible adduction of the thumb. Considering this, answer the questions below.

- a. Define *neural reflex*:

- b. Use the following terms to label the different components of a neural reflex illustrated below in Figure 2: *motor neuron*, *sensory neuron*, *interneuron (CNS)*, and *peripheral effector*:

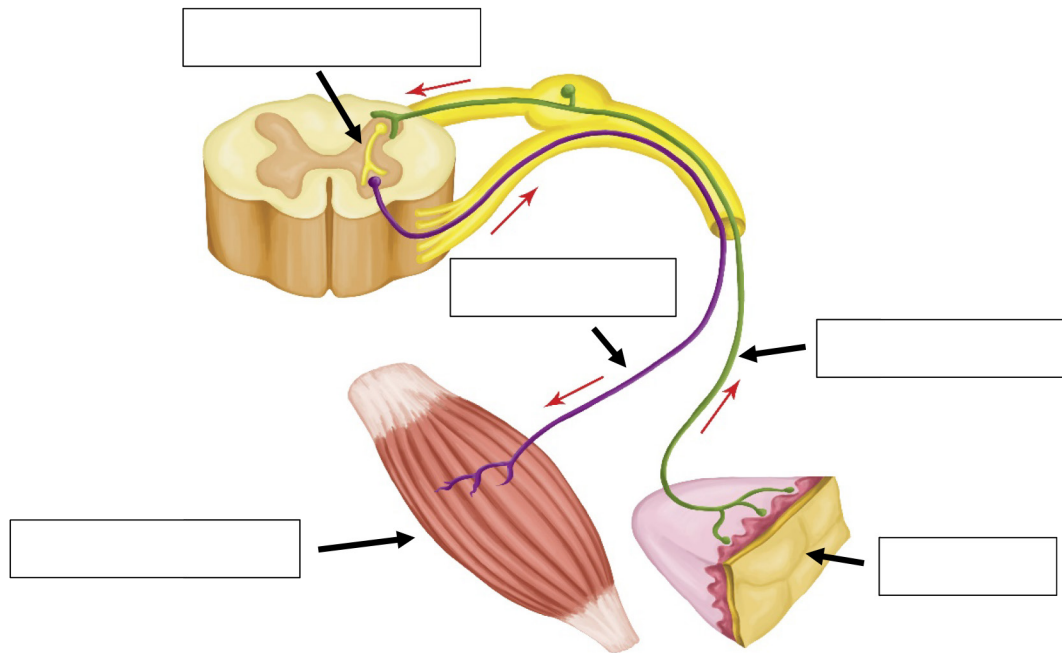


Figure 2. Neural reflex. (Licensed image ©stihii | Shutterstock, ID 214510006.)

- c. Indicate on Figure 2 above where Stacy's abnormal reflex may originate.
 - d. Hoffman's reflex test (finger flexor reflex) can be associated with problems involving the corticospinal tract. What type of information travels the corticospinal tract, and how does this relate to Hoffman's reflex test?
5. Describe syncope and list some common causes.

 6. Use the information you have learned from this part of the case study (Part I) and a credible source to describe and list signs/symptoms of type I Chiari malformation.

 7. List Stacy's signs and symptoms that are consistent with a diagnosis of CM. Briefly explain your answer.

Part II – The Doctor Visit and Initial Treatment

In March 2015, I began to have neck pain while attending a conference in North Carolina. I assumed it was due to the plane and bus ride. By the fifth day, the pain was so severe that I could not sit up after lying prone. It took about 10–15 minutes to roll over to the side of my bed, lower my legs to the floor in a kneeling position, and only then could I stand. By the time I could stand, the pain was so bad I was in tears.

Desperate, I decided to make an appointment with a chiropractor. Unfortunately, the visit resulted in little relief of my pain. I contacted my primary care physician (PCP), who ordered a cine MRI, a special MRI used to observe cerebrospinal fluid (CSF) flow. The MRI showed that I had minimal CSF flow, my tonsillar herniation was now at 18 mm, and the syrinx was much larger, encompassing much of my spinal cord from spinal cord sections C3–T4 (see Figure 3, next page). I saw a neurosurgeon that Friday. He recommended surgery within 72 hours due to the size of the syrinx pressing on my spinal cord and the lack of CSF. The operation would include partial decompression, which consisted of shaving off some of my C1 vertebra (atlas) and removing a piece of my occipital bone to allow more room for the CSF to flow naturally and thus minimize the syrinx. This surgery is less invasive compared to a complete decompression surgery process and, therefore, a shorter recovery time. Total decompression surgery involves cutting into the dura mater of the brain, which increases the chance of infection. I opted for the less invasive partial decompression, which went well, and I was able to return to work full time within one month. I completed a 5 K obstacle course two months after surgery. Although my progress was exciting, the surgeon did not cover the space once protected by bone with a metal plate, and the soft tissue of my brain was vulnerable to injury.

Questions

1. List Stacy's signs and symptoms before her surgery. What do her signs/symptoms and MRI scans indicate regarding the progression of her condition?
2. What does C3–T4 mean in the above text?
3. What are the functions of cerebral spinal fluid (CSF), and what is the typical sequence of CSF flow in the CNS?
4. How is the CSF interrupted in Stacy's central nervous system?
5. How does the interruption of Stacy's CSF impact the neural tissue of the spinal cord?
6. List the structures that protect the brain from superficial to deep and explain how a full decompression surgery without any protective patches or plates covering the brain could have compromised Stacy's brain.

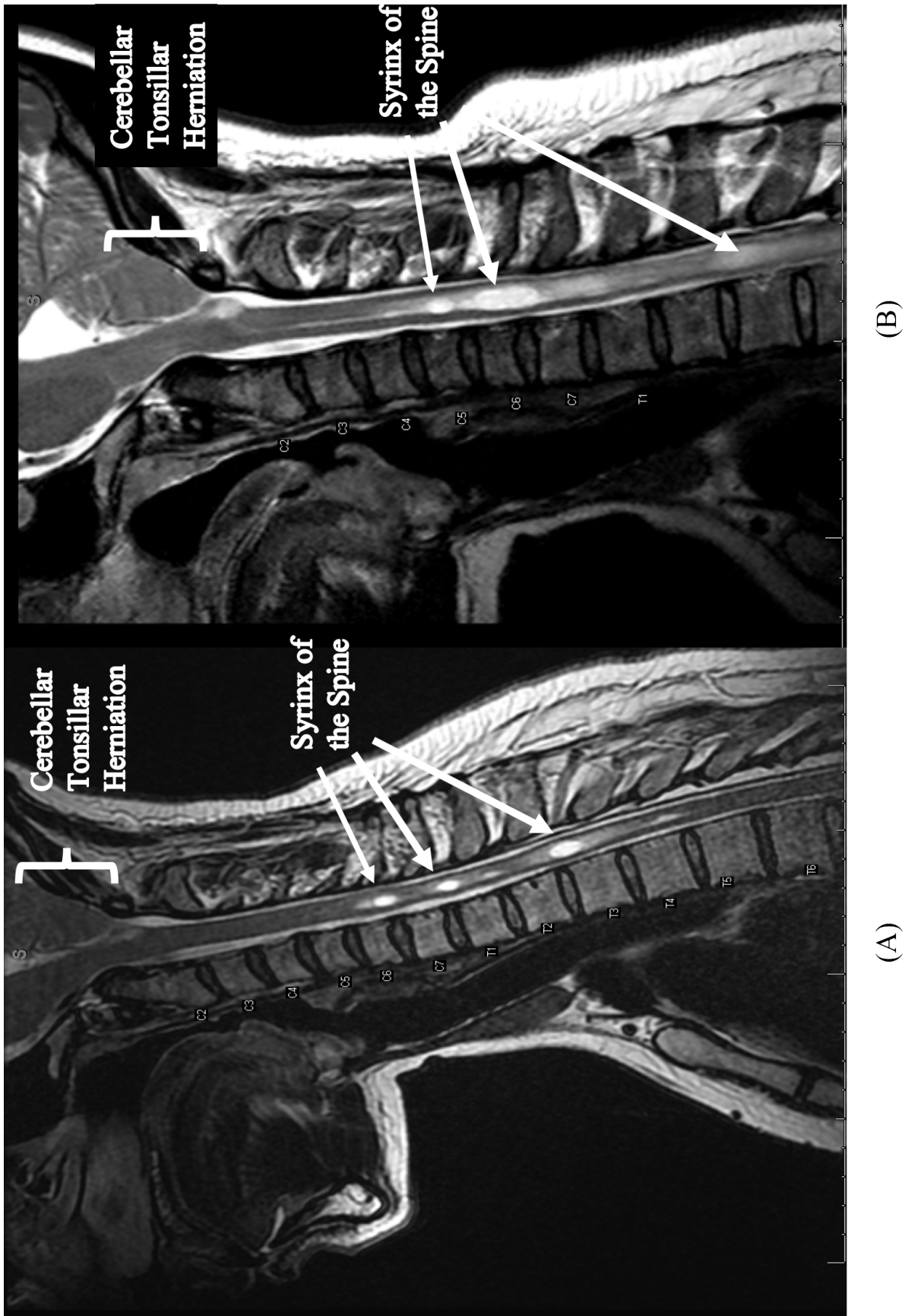


Figure 3. MRI image (A) is of Stacy's posterior brain and inferior spinal cord (July 2010). Image (B) is an MRI scan of the same region (March 2015). Notice the increased size of the tonsillar herniation to 18 mm, and the much larger syrinx in most of her spinal cord from C3–T4.

Part III – Recurring Symptoms

In February 2016, I began having increased dizziness while raising or lowering my head or while walking. Then one day, I had a sudden bout of severe head pain. Up until this point, I had been headache free since my surgery, so I decided to head to the emergency room. The MRI of the brain noted some improvement in the tonsillar herniation; it decreased from 18 mm to 16 mm. However, the emergency room physician did not order a cervical MRI to check the syrinx. I returned to my neurosurgeon, who thought the headaches and dizziness were unrelated to the Chiari. He could not explain the reason for the reoccurrence of the headaches or dizziness but stated my surgery was a success.

I contacted my PCP, and together we decided to contact John Hopkins Hospital (JHH) to see a Chiari specialist and assess the syrinx. At this time, I started experiencing tingling and numbness in both arms and legs.

Questions

1. What is your response to the neurologist’s opinion that the headaches, dizziness as well as tingling and numbness in Stacy’s limbs were unrelated to her Chiari condition?
2. Medical professionals often recommend a second medical opinion. List possible obstacles patients may encounter trying to obtain a second medical opinion and explain why it is vital for Stacy to overcome these obstacles.

Part IV – Second Opinion

In mid-April 2016, the neurosurgeon at JHH recommended having a full decompression surgery because the MRI illustrated poor improvement of the syrinx from the prior surgery. At the beginning of May, I had a complete decompression surgery, which included removing more of the occipital bone and stretching the arachnoid meninges. Surgeons placed a patch and metal plate over my brain and my occipital bone, respectively. The recovery period for this surgery took much longer, and I was unable to return to work until August 2016. And, I was unable to participate in strenuous activity for the next 12 months.

Since the second surgery, my headaches have greatly improved. I do get headaches when the barometric pressure drops or with any significant change in the weather. The numbness and tingling have improved as well. Many of my other symptoms, dizziness when raising my head, lack of balance, gagging when brushing teeth, I will always have because of the Chiari. The newest symptom, my head feeling like a 1,000-pound weight at the end of the day, will improve overtime as I re-strengthen my neck muscles. The muscles that attach to my occipital bone were cut and are no longer naturally attached to my skull. I will need to have regular MRI scans to monitor the syrinx, but the hope is that the second surgery was enough that the herniation and the syrinx will not worsen or cause further nerve damage.

Questions

1. Describe the location of the occipital bone and list the brain regions protected by this bone.
2. Explain why Stacy’s headaches, numbness, and tingling improved as a result of the decompression surgery.
3. As a group, fill in the table below with information about the cranial nerves responsible for the gag reflex.

<i>Nerve Name and Number</i>	<i>Location</i>	<i>Function</i>	<i>How does the nerve location and function relate to Stacy’s signs and symptoms?</i>

4. List the layers of the brain meninges from superficial to deep. Include in your answer the function of the arachnoid meninges and speculate why the surgeon stretched her arachnoid meninges during the decompression surgery.
5. Many muscles originate in the occipital bone. Collectively, what are their muscle actions? How does this contribute to her head “... feeling like 1000 pounds ...”?
6. Compare and contrast the procedures of the two surgeries.